Psychological Wellbeing of Siblings of Children and Young People with Chronic Health Conditions

Mhairi McKenzie Smith

Prepared under the supervision of Professor Roz Shafran and Dr Snehal Pinto Pereira

Population, Policy & Practice Research and Teaching Department, University College London Great Ormond Street Institute of Child Health

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Declaration

I, Mhairi McKenzie Smith, confirm that the work presented in this thesis is my own. Where information has been derived from other sources. I confirm that his has been indicated in the thesis.

N.B. Some content in chapters is taken from, or based on, work published by the candidate, or submitted for publication in peer-reviewed journals. Where text has been taken from material published by the candidate, this has been indicated in the thesis, including a citation for the relevant publication. The inclusion of the content of the papers in chapter form adheres to University regulations (https://www.ucl.ac.uk/academic-manual/sites/academic-manual/files/chapter_5_part_a_research_degree_regulations_2019-20.pdf, p.18. Section 5.1.2.)

Abstract

The psychological wellbeing of individuals with a brother or sister with a chronic illness or condition is at a unique risk. There is a need to further consider the impact on sibling psychological wellbeing from their own perspective within the family unit in order to build a holistic understanding of sibling's need for support.

This thesis includes 10 chapters. The 1st chapter provides a review of the existing literature on sibling psychological wellbeing along with an introduction and overview of the thesis. The 2nd chapter is a systematic review and meta-analysis that synthesises 17 studies and 1,264 participants to consider the effectiveness of support offered to siblings. Chapter 3 introduces a large qualitative research project involving siblings with a brother or sister with cystic fibrosis and their immediate families. The results of the large qualitative study are presented in Chapters 4 to 7, and a family level multiple casestudy analysis in Chapter 8. Chapter 9 increases the depth of understanding of these findings by presenting a mixed methods analysis using the qualitative results and quality of life measures from the siblings and their immediate families. Chapter 10 describes how the results from Chapters 2 to 9 fit within the scope of existing research and theory, and identifies how best to support siblings.

Results of this thesis suggest that the evidence of effectiveness of sibling support is inconsistent and measures used may lack the necessary sensitivity. Although siblings generally did not identify a large impact on themselves, it was clear from both the qualitative analysis and the mixed methods study that many siblings used avoidance coping mechanisms. When it became more difficult to avoid their brother's or sister's health condition siblings felt an impact to a greater extent. Interventions may be best targeted at siblings during stressful times.

Impact Statement

This thesis adds to the existing understanding of the experiences of siblings with a brother or sister with a chronic illness or condition. The findings from this thesis highlight a continual lack of clarity on the best approach and methodology to use when evaluating the impact on siblings' psychological wellbeing. The results highlight the possible benefits of using a mixed methods approach to enhance the understanding of the experiences of siblings and provide possible explanations for the apparent lack of findings when only quantitative measures were used. The clear use of avoidance coping mechanisms by siblings found in this thesis could go some way to explaining the lack of substantial findings in previous research that used quantitative measures only.

This thesis also emphasises the need to consider the immediate family members when conducting sibling research. There is a need to move away from relying solely on parental proxy, instead using multiple respondents from the family unit allows for a greater holistic understanding of the sibling experience. This thesis builds on previous research by emphasising the importance of positive family relationships and the interconnectedness of family outcomes.

High treatment burden and mortality rate has previously been shown to have a negative effect on sibling wellbeing. This thesis focuses on siblings with a brother or sister with cystic fibrosis, allowing for in-depth exploration of the potential influence of a high treatment burden and mortality rate on sibling psychological wellbeing. However, the results of this thesis do not highlight high treatment burden and mortality as a pinnacle factor. In keeping with theories of sibling wellbeing there appear to be a multitude of factors that influence sibling outcomes. Further developments of this thesis are needed to understand the factors that should be targeted for support.

Finally, beyond further research, the findings from this thesis could be used to build new and enhance existing support available for siblings. Despite a level of resistance displayed by siblings to engage with support services, the meta-analysis included in this thesis highlights a potential improvement in outcomes for siblings when they received support of any kind. However, there is an inconsistency in the methods and evaluations of existing sibling support, and when developing new sibling support the use of research evidence and consistency need to be prioritised. Further, given the findings from the qualitative and mixed-methods work included in this thesis, support may not need to be continuous for siblings as it may be more beneficial during periods of high stress in their families. There is a need for clinicians and the immediate family unit to recognise the potential impact and need for support in siblings, even when the sibling chooses to ignore it.

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Glossary of Terms

Term	Definition
Adolescence	The definition of adolescence has changed over time in order to be more developmentally appropriate. Rather than the previously accepted 10–19 years, a definition of 10–24 years has been proposed as corresponding more closely to adolescent growth and popular understandings of this life phase (Sawyer, Azzopardi, Wickremarathne, & Patton, 2018).
Avoidant Coping Style/Mechanisms	Avoidant coping is a form of coping mechanism. Please see the definition of coping mechanisms given below. Avoidance coping mechanisms involve an individual directing their attention away from threatening information in an attempt to 'escape' from the stressful event they are faced with (Greenaway et al., 2015).
Chronic Illness or Condition	A chronic illness or condition is defined as one that is medically diagnosed and reproducible using valid methods or instruments, has been present for longer than three months or has occurred three or more times in the past year and is likely to reoccur, and is not (yet) curable or is highly resistant to treatment (including mental health conditions) (Mokkink, Van Der Lee, Grootenhuis, Offringa, & Heymans, 2008).
Coping Strategies/Mechanisms/Methods	Coping has been defined as a response to perceived stress involving "constantly changing cognitive and behavioural efforts to manage specific external and/or internal demands that one appraised as taxing or exceeding the resources of the person" (Lazarus & Folkman, 1984).
Cystic Fibrosis (CF)	"Cystic fibrosis (CF) is an inherited condition that causes sticky mucus to build up in the lungs and digestive system". Those with CF generally have an intensive treatment regime, which can include multiple medications and physiotherapy sessions.
Emotional and Behavioural Difficulties	A problem identified in emotions (anxiety/worry, low mood/depression) or behaviour (disruptive behaviour).

¹ https://www.nhs.uk/conditions/cystic-fibrosis/

Family Dynamics/System	Family dynamics are the ways that family members interact with each other. It often refers to the interconnectedness of family outcomes. This emphasises the concept that "any individual member of the family is inextricably embedded in the larger family system and can never be fully understood independent of that system" (Cox, 2010).
Holistic Approach	Relating to the whole of something or to the total system instead of just to its parts.
Internalising and Externalising Disorders	A dichotomous classification of child psychopathology. Internalising disorders incorporate difficulties experienced internally by the child, for instance anxiety and depression. Whereas, externalising disorders are more overtly displayed by the child i.e. behaviour problems (Achenbach & Edelbrock, 1978).
Mental Health Disorder/Condition	"Generally characterised by a combination of abnormal thoughts, perceptions, emotions, behaviour and relationships with others." Mental health disorders include: depression, bipolar affective disorder, schizophrenia, dementia, intellectual disabilities and developmental disorders including autism" (WHO, 2017).
Peer Support	A rudimentary explanation of peer support could be "the giving of assistance and encouragement by an individual considered equal". For further discussion of the complexities of peer support see Dennis (2003).
Psychological Functioning and Psychological Wellbeing	Psychological functioning is defined as "an individual's ability to achieve their goals, both within themselves and in the external environment. This includes their emotions, behaviour (both internalising and externalising behaviours), social skills and their overall mental health" (Preedy & Watson, 2010). Psychological wellbeing is closely related to the definition of psychological functioning as further discussed on page 15.
Psychosocial Wellbeing	A combination of psychological and social wellbeing which closely relates to the concepts of psychological wellbeing and quality of life (Dean, Orford, Staines, McGee, & Smith, 2017).

Quality of Life	Typically defined as a measure of wellbeing that encompasses five dimensions: physical wellbeing, material wellbeing, social wellbeing, emotional wellbeing, and development and activity (Felce & Perry, 1995).
Reference/ Reference Density/Proportional Reference Density	A reference in NVivo is a portion of text associated with a specific theme or subtheme. The reference density describes the number of references that occur within a theme or subtheme. The proportional reference density represents the percentage of a participants interview that includes references for a specific theme or subtheme.
Sibling	For the purpose of this work a sibling is an individual with a brother or sister diagnosed with a chronic illness or condition as previously defined.

A Sibling's Experience (1)

Presented below is my personal motivation for completing this thesis. My first-hand account of being a sibling describes the impact I have felt due to having an older sister with myotonic dystrophy and how this directly relates to the approach taken throughout this thesis.

The vast majority of researchers I have met that work on projects considering the lives of siblings of individuals with a chronic illness or condition are they themselves siblings, and I am no exception to the rule. I am the younger sister of Claire. Both Claire and my father have a chronic illness called myotonic dystrophy. My knowledge and understanding of the condition remain somewhat limited in a biological sense, which was interestingly echoed by several of the siblings I spoke to throughout my PhD work. What I do believe I have developed, and continue to develop, is a sound understanding of the day-to-day social and psychological impact their condition has had on both myself and my mum. Growing up my family had several "typical" aspects; like most "normal" families we most definitely had our share of arguments and frustrations with each other, but we also shared some great moments and a joy of music. In several ways, I believe that my life has been enhanced by the presence of myotonic dystrophy.

There is one story I have grown up hearing that I believe portrays the relationship I had with my sister throughout our childhood, and to an extent our relationship now in our adult lives. Claire is five years my elder, and one day following school when I was just 3 years old I was taken to collect her from school, but some boys in the playground were picking on Claire and the way she spoke. I am informed that I stormed over to these eight-year old boys and told them, I'm sure in a very friendly manner, that they were to leave my sister alone and they scampered off. This instinct to protect and defend Claire remains despite the years that have passed.

Myotonic dystrophy comes with a cacophony of complications and side effects, many of which are not regularly recognised or are poorly understood. The condition itself affects Claire in many ways which I, and certainly the general public, do not well understand. At this point in time the developmental and learning difficulties that affect Claire, I can say with confidence, have caused the largest disturbance on our lives, despite these not being regularly recognised as related to myotonic dystrophy. This lack of understanding meant that, despite my mother's protests, Claire was not tested for any learning difficulties until she was 18, at which point she was diagnosed with dyslexia and provided with support from her college. Claire received no additional support throughout primary or secondary school.

Having to be the "older sister" from a very young age did mean that I needed to mature, in some ways, at a quicker rate than my peers with "healthy" siblings. This is a fact I only came to recognise in my adult years. For instance, I quickly learnt that Claire found it difficult and upsetting

when I was able to do something that she was struggling with, therefore, I became quite adept at faking a lack of ability at video games. Claire has always wanted to remain the "big sister", offering me great support and guidance where I need it. The degenerative nature of myotonic dystrophy and the difficulties, both social and physical, that come with this have definitely put a strain on my relationship with Claire. However, we remain close and some of my best memories growing up are making Claire laugh to the point of tears, although in hindsight this is not great for someone with a weak oesophagus.

My mum and I have always shared a close bond, and I know that she has gone above and beyond to ensure that I never felt I had to go without or that there was any disparity in the way she treated Claire and I. While I may have largely been oblivious to the extent of effort my mum put in throughout my childhood, the determination and stubbornness of my mother to make my life as positive as possible has definitely become clearer to me as I have become older. My mother has always been the fiercest protector of both Claire and I. The unfortunate reality is that my mother will not always be around, and as she gets older and the demands of myotonic dystrophy continue to increase in our lives, she will not be able to care for Claire alone. This is typically an unspoken truth in our family. I have begun to take steps towards accounting for this now in my life, which has also highlighted to me several ways in which I am different to my peers with healthy siblings as even now I am having to make choices and take on responsibilities perhaps beyond my years.

My experiences were the largest driver in my decision to undertake a project considering the lives of sibling of individuals with a chronic illness or condition. It has also driven the way in which I approached designing my research questions, for instance the recognition that physical and mental health are not distinct categories presented in the systematic literature review presented in Chapter 2, and the importance of considering the experiences of the full family as in Chapters 3 to 9.

My hope is that this work will be able to provide a useful addition to the existing literature considering siblings, and will be used to enhance recognition and support of this ill-considered group. As a sibling myself, I know that I took great solace in talking to friends with a brother or sister with a chronic illness or condition; this and the support offered by my immediate family whom were already stretched thinly, was the only support I received.

Chapter 1: Thesis Introduction and Background

1.1. Thesis Introduction

The subject of this thesis is the psychological wellbeing of siblings with a brother or sister with a chronic illness or condition. The psychological wellbeing of individuals with a brother or sister with a chronic illness or condition, referred to as "sibling" throughout this thesis, is at unique risk. This Chapter begins by presenting the empirical basis for studying the psychological wellbeing of siblings, followed by an overview of the content and the aims of the thesis.

1.1.1. Overarching Thesis Aim

The overarching aim of this thesis is to enhance the understanding of the psychological wellbeing of siblings in families where one child or young person has a chronic illness or condition, and to consider the implications this has for future research and support provided for siblings.

1.2. Central Definitions and Terminology

Throughout this thesis there are several pieces of terminology that are used regularly and must be defined. These terminologies were briefly summarised in the glossary on pages 9-11. The central subject of this thesis is siblings. Within this thesis the term **sibling** will consistently refer to an individual that has a brother or sister with a diagnosed chronic illness or condition (definition of which is given below). In previous research the term "healthy sibling" has frequently been used to describe this individual, however this suggests that the sibling is not affected in anyway and that their brother or sister is "unhealthy" which may not always be true from the perspective of the family, as is shown in the qualitative project presented in Chapters 3 to 8.

A chronic condition or illness has previously been defined in multiple different ways. Differences in definition can lead to problems in research (Perrin et al., 1993). Although there are generally a few common aspects amongst definitions there are key differences that can lead to the inclusion or exclusion of certain conditions. The definition appropriated in this work is as follows: A chronic illness or condition can be defined as one that is medically diagnosed and reproducible using valid methods or instruments, has been present for longer than three months or has occurred three or more times in the past year and is likely to reoccur, and is not (yet) curable or is highly resistant to treatment (including mental health conditions) (Mokkink et al., 2008). As this definition includes mental health conditions the wording of "illness or condition" has been chosen carefully to reflect the view that not all chronic conditions are illnesses (Boorse, 1975). The use of language to describe chronic mental health conditions is contentious and subject to debate (Kenny et al., 2016).

The World Health Organisation defines a **mental disorder** as "generally characterised by a combination of abnormal thoughts, perceptions, emotions, behaviour and relationships with others". Mental disorders included in this definition include: depression, bipolar affective disorder,

schizophrenia, dementia, intellectual disabilities and developmental disorders including autism (WHO, 2017). This definition of a mental disorder needs to be distinguished from that of **psychological functioning**, which is defined as "an individual's ability to achieve their goals, both within themselves and in the external environment. This includes their emotions, behaviour (both internalising and externalising behaviours), social skills and their overall mental health" (Preedy & Watson, 2010).

The central concept of impact considered in this thesis is **psychological wellbeing**. This term relates closely to the definition of psychological functioning. Further consideration needs to be given to the term wellbeing, particularly as the definition of wellbeing is continually developing. It is suggested that high wellbeing is positively related to good mental health and can be made up of the following ten components: competence, emotional stability, engagement, meaning, optimism, positive emotion, positive relationships, resilience, self-esteem, and vitality (Huppert & So, 2013). As such **psychological wellbeing** is defined for this work as an individual's ability to achieve their goals, both within themselves and in the external environment. This includes behaviour (both internalising and externalising disorders), social skills, emotions, emotional stability, engagement, relationships, resilience, vitality, and their overall mental health (Huppert & So, 2013; Preedy & Watson, 2010).

The definition of psychological wellbeing used for this work also encompasses elements of frequently used definitions for quality of life (QoL). **Quality of life** is typically defined as a measure of wellbeing that encompasses five dimensions: physical wellbeing, material wellbeing, social wellbeing, emotional wellbeing, and development and activity (Felce & Perry, 1995). This definition relates closely to the definition of psychological wellbeing given above.

1.3. Empirical Basis

While there has been a vast amount of time and effort given to research on the psychological wellbeing of those with a chronic illness or disability (Merikangas et al., 2015; Parry-Langdon, Clements, Fletcher, & Goodman, 2008), the impact on their immediate family has typically been given less attention (Barlow & Ellard, 2006). Research on the immediate family has largely focused on parents and has suggested that parents of children and young people with a chronic illness or condition have poorer psychological wellbeing relative to other parents with unaffected children (Besier et al., 2011; Grootenhuis & Last, 1997; Ingerski, Shaw, Gray, & Janicke, 2010). Further research has also highlighted the long-term psychological risk in parents (Mailick Seltzer, Greenberg, Floyd, Pettee, & Hong, 2001). Given the increasing recognition that siblings are highly probable to take on a caregiving role for their brother or sister in later life, the importance of considering their mental health and wellbeing is also increasing (Burke, Fish, & Lawton, 2015; Chen & Lukens, 2011; Dimitropoulos, Klopfer, Lazar, & Schacter, 2009; Hatfield & Lefley, 2005). More research is being published about this population on a regular basis; despite this increase in the evidence base, there remains little consensus on how to support siblings of children with a chronic illness or condition (Hartling et al., 2014). The

following literature review draws on the evidence available that considers the psychological wellbeing, quality of life, family relationships and dynamics, support services, and theoretical approaches to sibling wellbeing, in order to identify the gaps in research and support for siblings that need to be addressed.

1.3.1. Sibling Prevalence in the UK

It is estimated that anywhere between 13-32% of children and young people (0-19 years) suffer from a chronic condition or illness (Fraser et al., 2012; Van Cleave, Gortmaker, & Perrin, 2010; Wijlaars et al., 2016), and around 18%² of families have two or more children (OECD 2016), which implies that approximately 2-6% of children have a brother or sister with a chronic illness. Given the population of the UK (Office for National Statistics, 2016), we can estimate that there could be up to 700,000 siblings of children and young people with a chronic condition or illness currently living in the UK. The only UK wide charity dedicated to siblings of individuals with disabilities suggests that further to this estimate of young siblings, there is also a minimum of 1.7 million adult siblings with a disabled brother or sister in the UK³

1.3.2. Psychological Wellbeing in Siblings

Literature regarding sibling psychological wellbeing dates back to the 1970s and yet to this day the results of such work remain inconsistent and indeterminate. Some literature suggests an elevation in psychological distress and mental health disorders in siblings (Cadman, Boyle, & Offord, 1988), other studies suggest that siblings have comparable psychological and psychosocial outcomes to their peers with healthy brothers and sisters (Bischoff & Tingstrom, 1991), and finally a small number of studies suggest a potential for positive effects and experiences in siblings (Houtzager, Grootenhuis, Hoekstra-Weebers, Caron, & Last, 2003; O'Brien, Duffy, & Nicholl, 2009; Petalas, Hastings, Nash, Reilly, & Dowey, 2012; Sharpe & Rossiter, 2002).

A meta-analysis of 51 studies looking at the psychological impact of having a sibling with a chronic illness or condition found a significant overall negative impact (mean effect size (M_d) = -0.20, 95% Confidence Interval (CI) (-0.23,-0.16)) and specifically a significant negative impact on psychological functioning (M_d = -0.22, 95% CI (-0.26, -0.17)), peer activities (M_d = -0.29, 95% CI (-0.36,-0.22)), and cognitive development (M_d = -0.24, 95% CI (-0.44,-0.04)) (Sharpe & Rossiter, 2002). This is consistent with the findings in a more recent meta-analysis, which included 13 additional studies to

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² OECD-32 average proportion of households with two or more children is reported as 54.6%, which is multiplied by our estimated prevalence estimates (13–32%) to achieve our estimate of children that have a sibling with a chronic condition. "Children" in this instance are generally defined here as dependent resident children under 25 and include both biological children and step- or adopted

³ https://www.sibs.org.uk/about-sibs/aims-history-strategy/ - accessed 13/5/2019

Sharpe and Rossiter's meta-analysis (Vermaes, van Susante, & van Bakel, 2012) and found a significant, although small, negative effect on psychological functioning (mean effect size (d₊) = -0.14, 95% CI (-0.17, -0.10)). It has been noted that there is a particularly high potential for internalising rather than externalising disorders in siblings. An increase in internalising disorders, i.e. emotional symptoms including anxiety and depression, has been observed in several studies (Cadman et al., 1988; Fisman et al., 1996; Hastings, 2003b; Verté, Roeyers, & Buysse, 2003), and is supported by both meta-analyses (Sharpe & Rossiter, 2002; Vermaes et al., 2012). Suggestions as to why internalising behaviours are more prevalent in siblings included children not wishing to burden parents further (Sidhu, Passmore, & Baker, 2006), the quality of the family environment (Verté et al., 2003), and factors relating to the child themselves e.g. age, sex (Hastings, 2003b). Alderfer et al. (2010) found that siblings of children with cancer did not present with a greater number of psychiatric disorders. However, there were a significant number of siblings that presented with symptoms of post-traumatic stress disorder, including avoidance, re-experiencing, and arousal. Post-traumatic symptoms have also been noted in siblings of children and young people with Prader-Willi syndrome, a complex genetic disorder characterised by a range of developmental abnormalities, both physical and psychological (Mazaheri et al., 2013).

Psychological wellbeing comparable to a normative population has been found in sibling of those with cancer (Alderfer et al., 2010), renal failure (Batte, Watson, & Amess, 2006) and epilepsy (Wood, Sherman, Hamiwka, Blackman, & Wirrell, 2008). In their study on the wellbeing of siblings of children with disabilities Emerson and Giallo (2014) initially found lower psychological wellbeing in siblings relative to their peers with 'typically developing' brother or sisters. However, they noted that the effect was small and was eliminated when environmental adversities such as maternal mental health and education were taken into consideration. Despite environmental adversities, such as maternal mental health, potentially being attributed to the presence of a disability in the family, Emerson and Giallo (2014) concluded that the psychological wellbeing of siblings of children and young people with a disability was comparable to that of their peers.

Social and emotional outcomes are the most consistently noted psychosocial effects of having a brother or sister with a chronic illness or condition. It is known that having a brother or sister with a chronic illness or condition can pose social challenges for siblings which can reduce the sibling's social functioning (Bluebond-Langner, 2000; Clemente et al., 2003; Houtzager, Grootenhuis, Hoekstra-Weebers, & Last, 2005). There are multiple difficulties associated with having a brother or sister with a chronic condition or illness that could have a detrimental effect on sibling's socialising abilities. The requirement for parents to focus on their ill child reduces the physically and mentally capacity of parents, disrupting family routines on a daily basis and sometimes resulting in the sibling taking on responsibilities that interfere with their ability to socialise with peers (Alderfer et al., 2010; Fullerton, Totsika, Hain, & Hastings, 2017; Lobato et al., 2011).

The effects of being a sibling of a child with a chronic illness or condition on schooling have been considered in a recent systematic review (Gan, Lum, Wakefield, Nandakumar, & Fardell, 2017). Negative psychological wellbeing in siblings can affect their school life. Difficulties siblings experience include school absences (Barlow & Ellard, 2006), troubles with interacting with classmates and teachers (Prchal & Landolt, 2012; Sidhu, Passmore, & Baker, 2005), and problems with school work (Prchal & Landolt, 2012; Williams et al., 2009). There is the possibility that siblings may have poorer educational attainment compared to their peers (Breining, 2014), which may have repercussions in the long-term (Wolfe, Song, Greenberg, & Mailick, 2014). Research considering difficulties at school for siblings has largely focused on siblings of children and young people with cancer (Alderfer et al., 2010), but there is also evidence to suggest similar difficulties in siblings of children with Type 1 diabetes (Sleeman, Northam, Crouch, & Cameron, 2010), and autism (Lobato et al., 2011).

There is research that suggests potential positive effects of having a sibling with a chronic illness or condition. The majority of the suggested positives for siblings are psychosocial in nature. An increase in maturity has been found in several studies (Alderfer et al., 2010; Grossman, 1972), greater warmth and understanding towards their sibling (Fisman et al., 1996), and prosocial behaviour (Ferrari, 1984; Lobato, Faust, & Spirito, 1988) have also been suggested. Currently, there is limited acknowledgement and understanding of these positive findings but it has been suggested that they could potentially act as protective factors for mental health outcomes in siblings (Fisman et al., 1996). Much of the research on sibling psychological wellbeing does not facilitate the option of considering positive outcomes due to the measures and methods adopted that are generally selected due to a 'negative narrative' approach taken by the researcher (Hastings, 2016). Hastings (2016) suggested that the typically found 'negative narrative' should be dropped and the potential for positive outcomes should be given greater consideration in future research.

Quality of Life in Siblings: Measuring quality of life (QoL) in siblings allows several aspects of life such as health, social-life, and schooling to be incorporated into one measure. It has been suggested that measuring QoL in siblings may be the most appropriate way in which to assess the impact of having a brother or sister with a chronic illness or condition (Hartling et al., 2014). For instance, the Paediatric Quality of Life Inventory (PedsQL) (Varni, Seid, & Rode, 1999) and the EQ-5D (Brooks, 1996) consider several aspects of life and have been used in studies considering sibling QoL (Limbers & Skipper, 2014). QoL relates closely to the concept of psychological wellbeing as defined for this work on page 15. The limited research available on QoL in siblings of children with chronic physical health conditions appears to also produce somewhat inconsistent findings as those on the psychological and psychosocial wellbeing of siblings. Of the nine studies included in a systematic review considering sibling QoL when they have a brother or sister with a chronic physical health condition, conducted by Limbers and Skipper (2014), only four considered the quality of life of the sibling relative to a normative sample. Three studies reported higher or similar health related quality

of life (HRQoL) (Havermans, Croock, Vercruysse, Goethals, & Diest, 2015; Havermans et al., 2011; Wood et al., 2008) and one study reported lower HRQoL (Packman et al., 2005) in siblings compared to peers with an unaffected brother or sister. Since the review by Limbers and Skipper (2014) it has also been found that siblings of children with chronic fatigue have a comparable QoL to that of a normative sample (Velleman, Collin, Beasant, & Crawley, 2016). Although measuring QoL the previously mentioned studies did not use a consistent measure, which may explain some of the variance in findings. It is still possible to conclude that, as with the psychological wellbeing of siblings, it is challenging to determine exactly how QoL in siblings is affected by the presence of a chronic physical condition.

Few studies are available that have specifically evaluated the QoL in siblings of children and young people with a chronic mental condition. In general the studies on the QoL of siblings with brother or sister with a mental health condition appear to find a consistent negative effect. Barnett and Hunter (2012) found a significantly lower QoL in their sample of siblings with a brother or sister with mental health problems and 80% of siblings of children with eating disorders had a negatively affected QoL in the study by Areemit, Katzman, Pinhas, and Kaufman (2010). Studies that considered neurological chronic conditions such as Autism Spectrum Disorders (ASD), epilepsy, and cerebral palsy, typically also find a lower QoL in siblings (Marciano & Scheuer, 2005; Rana & Mishra, 2015). In the study by Rana and Mishra (2015) the majority (64%) of the siblings included suggested that they did not have sufficient knowledge of their brother's or sister's condition. The influence of knowledge on psychological functioning is further considered in section 1.3.6.

1.3.3. Family Relationships and Dynamics

Family relationships can be disrupted by the presence of a chronic illness or condition (Ma, Roberts, Winefield, & Furber, 2017). As with the majority of sibling research the evidence on sibling relationships is inconsistent and greatly varies in the methods used (Ma et al., 2017). The presence of a chronic illness or condition can result in both negative and positive effects on the relationship between a sibling and their brother or sister (Ma et al., 2017). Within Sharpe and Rossiter (2002) meta-analysis, six papers considered the sibling relationship and reported a positive effect size of 0.12 [95% CI (-.06,0.3)], however this was not statistically significant. Siblings can experience feelings of guilt for being "healthy", anxiety for their brother or sister, or embarrassment at their brother or sister behaviours, which may affect the relationship between them (Petalas, Hastings, Nash, Dowey, & Reilly, 2009). Siblings of children with ASD have been noted to have typically poorer relationships with their brother or sister, relative to a normative sample of siblings with neuro-typically brothers and sister (Kaminsky & Dewey, 2001). Siblings with a brother or sister with a physical chronic condition such as cancer, muscular dystrophy, or a congenital heart defect, report a similar sibling relationship quality to siblings with a brother or sister with ASD (Fullerton et al., 2017). There is also the potential that a close sibling relationship (i.e. greater warmth) could result in greater externalising behaviours in

siblings in conditions such as cancer (Labay & Walco, 2004). The sibling relationship is not just important for development, it is also potentially one of the longest-lasting relationships individuals can have (Ma et al., 2017). The knowledge of potentially needing to become a carer for their brother or sister in the future can put a strain on the sibling relationship and the sibling's psychological wellbeing (Arnold & Heller, 2018). The relationship between siblings and their brother or sister is considered from the perspective of all the family in the qualitative project results presented in Chapters 4 to 8.

There is also a high potential for the sibling-parent relationship to be affected by the presence of a chronic illness or condition in the family, and this is explored in Chapters 4, 7, and 8. Siblings may feel as though they are second to their brother or sister. Parents are required to focus their attention on the child with the chronic illness or condition which can lead to feelings of jealousy or neglect in the sibling (Williams et al., 2010). Parents may also, either consciously or subconsciously, parent the sibling and their brother or sister differently (Deal & MacLean, 1995). Parents being stricter on, but also expecting more from, the sibling can foster feelings of resentment and anger in the sibling towards both their parents and their brother or sister (Ma et al., 2017). Overall, relationships between parents and siblings in families with a child with a mental health problem appear to be generally similar to the norm in terms of positive aspects. Differences in negative aspects, such as intrusive involvement appear to exist in mother-sibling relationships (Ma et al., 2017). Family dynamics are highly interconnected with family relationships (Cox, 2010). Family dynamics have also been noted to be affected by the presence of a chronic illness or condition. There may be poorer family cohesion and increased tension between parents resulting in a higher rate of divorce in parents of children with chronic illnesses or conditions (Slowik, Willson, Loh, & Noronha, 2004). Having a child with a chronic illness or condition limits parents abilities to socialise outside of the family, and could affect their ability to work (Kvist, Nielsen, & Simonsen, 2013; Mailick Seltzer et al., 2001).

1.3.4. Long-Term Outcomes for Siblings

The need to investigate and understand the impact on siblings is becoming steadily more important given the increasing need for siblings to take over as caregivers for their brother or sister in later life (Burke et al., 2015; Chen & Lukens, 2011; Dimitropoulos et al., 2009; Hatfield & Lefley, 2005). In 1997 it was found that 10% of siblings ended up living with their brother or sister with intellectual disability in adulthood, and this number was expected to grow (Freedman, Krauss, & Seltzer, 1997). Studies suggest that female siblings tend to take over a larger amount of caring responsibilities for their brother or sister relative to male siblings (Burke, Taylor, Urbano, & Hodapp, 2012). Siblings with their own children and those with poorer relationship with their brother or sister when younger appear to be less willing to take on caring responsibility (Bigby, 1998; Greenberg, Seltzer, Orsmond, & Krauss, 1999; Griffiths & Unger, 1994; Krauss, Seltzer, Gordon, & Friedman, 1996).

Adult sibling caregivers tend to exhibit a greater number of negative outcomes relative to their peers with unaffected brothers and sisters (Greenberg et al., 1999). Poorer educational outcomes (Wolfe et al., 2014) and greater participation in risky health behaviours, such as excessive drinking and smoking, have been noted in adulthood for siblings of individuals with cancer (Buchbinder et al., 2016; Lown et al., 2013). Several of the previously reported outcomes, including poorer psychological wellbeing, could potentially last into adulthood if left undetected and unsupported (Orsmond, Kuo, & Seltzer, 2009). Siblings of adults with ASD have been noted to have an increased risk of depression compared to a normative sample (Rai et al., 2018). In a large longitudinal study on adult siblings of individuals with mental illness adult siblings were found to have a higher probability of a previous depressive episode compared to a normative sample of individuals with "healthy" brothers or sisters (Taylor, Greenberg, Seltzer, & Floyd, 2008). Sibling taking over caring responsibility for their brother or sister in adulthood places them under greater strain (Dimitropoulos et al., 2009). However, Tomeny, Ellis, Rankin, and Barry (2017) suggest that there is the possibility of a caring having a positive effect on the sibling relationship. Although this thesis only includes siblings up to the age of 24, there were several adult siblings above 24years of age that expressed an interest in the work and one provided a poignant account of their experience (given on page 219).

As the understanding of chronic illness and conditions continues to improve there are broader aspects that may affect siblings that need to be considered. Adult siblings with a brother or sister with autism that had previously presented with a level of autistic behaviours were found to have poorer social relationships and greater mental health difficulties compared to siblings that presented with no autistic behaviours in childhood (Howlin, Moss, Savage, Bolton, & Rutter, 2015). Furthermore, the potential for siblings to be carriers of genes associated with a chronic genetic condition, such as cystic fibrosis (CF), and the possibility of passing the condition on to their own children has been shown to affect family decision making and psychological wellbeing in siblings (Fanos & Johnson, 1995; James, Hadley, Holtzman, & Winkelstein, 2006; Lerman, Croyle, Tercyak, & Hamann, 2002; Sobel & Cowan, 2000).

1.3.5. Understanding the Experiences from the Sibling's Perspective

Despite the potential benefits of qualitative research, which are further considered in Chapter 3, a limited amount has been completed on the experiences of siblings (Knecht, Hellmers, & Metzing, 2015). Previously parents' perspective on sibling experiences have been given precedent (Hames & Appleton, 2009; Patterson, Holm, & Gurney, 2004; Wennick, Lundqvist, & Hallström, 2009; Williams et al., 2009). Although there is a scarcity of qualitative evidence considering sibling psychological wellbeing it is still possible to extract a number of key themes and implications for support from the existing evidence.

As found in quantitative research on sibling wellbeing, qualitative findings appear diverse and inconsistent. Diverse findings have been noted in sibling health, family relationships, and sibling development (Knecht et al., 2015). A notable finding from qualitative research on sibling wellbeing, which is consistent with the quantitative findings on psychological wellbeing, is that siblings regularly report a higher level of internalising behaviours e.g. emotional distress, rather than externalising behaviours e.g. behavioural problems (Knecht et al., 2015). Acute periods in their brother's or sister's illness, e.g. time of diagnosis, and their brother or sister presenting behavioural difficulties have been related to higher reported levels of distress and anxiety in siblings (Hames & Appleton, 2009). Siblings report an adjustment to the illness over time which allows them to cope with their emotions better (Hames & Appleton, 2009; Petalas et al., 2009; Read, Kinali, Muntoni, Weaver, & Garralda, 2011).

Psychosocial behaviours are more commonly addressed in the qualitative literature and the findings are generally consistent, although still limited by the quantity and detail in which they have been considered. The evidence suggests that several siblings appear to show social isolation and withdrawal (Malcolm, Gibson, Adams, Anderson, & Forbat, 2014; Petalas et al., 2009) but also a greater potential for empathy (Alderfer et al., 2010), compassion (Wilkins & Woodgate, 2005), and patience (Bellin & Kovacs, 2006). Siblings regularly feel a lack of understanding of their experiences and their brother's or sister's condition from others (Opperman & Alant, 2003; Petalas et al., 2009; Read et al., 2011). Support from friends and other family members is viewed positively (Barr & McLeod, 2010; Waite-Jones & Madill, 2008). It is echoed by many siblings with brothers or sister with a range of chronic illnesses and conditions that they felt a strain in the family due to a lack of time for them (Areemit et al., 2010; Barr & McLeod, 2010; Patterson et al., 2004). Contradictorily a potential for an increase in family cohesion has also been suggested (Read et al., 2011; Wennick et al., 2009).

Siblings can often feel a level of responsibility toward their brother or sister (Areemit et al., 2010; Patterson et al., 2004). Both parents and siblings have suggested that there is a need for siblings to mature quicker than their peers (Molinaro, Rollo, Fletcher, & Schneider, 2018). Qualitative research into the experiences of adult siblings of individuals with autism suggested that although siblings recognised the difficulty of growing up with someone with autism they had a positive commitment to providing care for their brother or sister in later life (Tozer & Atkin, 2015). This acceptance of caring responsibilities was also noted by Read et al. (2011) in siblings of children with Duchenne muscular dystrophy. Where their brothers and sister can independently manage their condition, such as in the case of Type 1 diabetes, siblings may not be needed to provide support with their brother or sisters health condition, rather they may be a source of emotional support (Wennick et al., 2009).

Coping methods in siblings have been investigated qualitatively. An active involvement in their brother or sisters care was believed to be a positive coping technique for younger siblings as it helped improve their understanding of the chronic illness or condition (Van Riper, 2003), and a lack of

information has been connected to distress in the sibling (Areemit et al., 2010; Hames & Appleton, 2009; Opperman & Alant, 2003; Waite-Jones & Madill, 2008). Further coping mechanisms utilised by siblings were typically passive (Haukeland, Fjermestad, Mossige, & Vatne, 2015) and included normalising the presence of the chronic illness or condition in their lives, which can be aided by the presence of the condition from early childhood (Malcolm et al., 2014; Read et al., 2011) and choosing to live in the present and have a positive outlook (Petalas et al., 2009; Wennick et al., 2009).

UK based studies have previously reported that siblings and their parents felt there was currently insufficient acknowledgement and support for siblings (Petalas et al., 2009; Tozer & Atkin, 2015). None of the siblings or parents included in a study considering sibling experiences when their brother or sister has epilepsy had heard of or been offered any support, but thought they would benefit from support (Hames & Appleton, 2009). Current support available to sibling is reviewed and evaluated in Chapter 2 of this thesis. Areas of need and suggestions for future support are captured in the qualitative project first presented in Chapter 3 and the subsequent mixed methods analysis.

1.3.6. Potential Influencing Factors

Identifying siblings at the greatest risk of poor QoL or psychological wellbeing could help the development and targeting of support services. Targeting support at those in the greatest need is imperative to resource-limited services, such as in the UK National Health Service (NHS). The wellbeing of siblings is a highly complex and dynamic subject, affected by many external factors. Literature has attempted to identified potential risk and protective factors for sibling wellbeing, but is limited by the great expanse of the potential factors. For simplicity and ease of understanding the existing evidence has been split broadly into seven categories that are relevant to this thesis: family dynamics, parental factors, knowledge of the chronic illness or condition, condition severity and treatment burden, sibling support, coping strategies, and demographic factors, each of which are discussed in turn below. These factors are considered in the thesis in two ways; they are highlighted in the systematic review and meta-analysis and incorporated into the semi-structured interview schedules used in the interviews with all family members included in the qualitative project presented in Chapter 3.

Family Dynamics: Family related factors have been suggested that may help identify those at risk. Daniels, Moos, Billings, and Miller (1987) found that less family cohesion and expressiveness were related to an increased psychological risk in siblings. Positive family functioning has been noted as a potential protective factor in siblings of children with Down's syndrome, yet not in those with pervasive developmental disorder (Fisman et al., 1996). Barnett and Hunter (2012) found that siblings of children receiving treatment for mental health problems were more likely to live in poorly functioning families. In their relatively large longitudinal study that considered sibling of children with cancer up to 24months after diagnosis, Houtzager et al. (2004) found that chaotic family circumstances negatively affect sibling psychological wellbeing. Stability in adaptation to changes, however, promoted better

outcomes in siblings as has also been noted in further studies since this work (Giallo & Gavidia-Payne, 2006). Giallo and Gavidia-Payne (2006) found that effective communication and problem-solving within families were associated with better adjustment in siblings whose brothers and sisters had a range of disabilities.

Parental Factors: Parental factors, such as poor maternal mental health (Lindström, Åman, & Norberg, 2010) and higher levels of parental stress (Giallo & Gavidia-Payne, 2006) have been associated with poorer psychological wellbeing in siblings. Behavioural difficulties in the child with the chronic illness or condition apply a greater stress to parents which in turn affects the sibling and their internalising and externalising behaviours(Walton, 2016; Wood et al., 2008). A greater maternal awareness of the sibling's attitude to their brother or sister illness has also been linked to better psychological outcomes in siblings (Taylor, Charman, & Fuggle, 2001). The amount of time a parent spends with the sibling has been shown to affect sibling outcomes in some cases (Murray, 2001). Parents spending more nights in hospital with their ill child was associated with a greater amount of adjustment problems in siblings with a brother or sister with CF (O'Haver et al., 2010).

Knowledge of Chronic Illness or Condition: Several studies have proposed a link between poor psychological wellbeing and siblings lacking understanding of their brother's or sister's condition (Carpenter, Sahler, & Davis, 1990; Houtzager, Grootenhuis, & Last, 2001; Sidhu et al., 2006). A limited understanding of their brother's or sister's condition has also been linked to poorer adaption in siblings (Evans, Jones, & Mansell, 2001). A negative impact on the sibling relationship is also associated with poor understanding in siblings (Roeyers & Mycke, 1995). The QoL of siblings may potentially be influenced by the attitude, in relation to their understanding, with which they approach their brother's or sister's condition, as has been noted in siblings of children with cancer (Canter et al., 2015). Improving the sibling's understanding of their brother's or sister's condition has been linked to reduced anxiety levels (Houtzager et al., 2001; O'Haver et al., 2010), and is viewed positively by both siblings and their parents (Loos & Kelly, 2006; Sidhu et al., 2005). Interventions that involve informational support have been found to improve the psychological wellbeing of siblings (Williams et al., 2003). There is also some evidence that suggests siblings who try to understand the meaning behind their brother's or sister's condition (often referred to as interpretive control) have more negative outcomes (Houtzager et al., 2005). There may be a balance to be struck in the level of information siblings should seek and be given.

Condition Severity and Treatment Burden: When considering potential predictive factors in their meta-analysis, Vermaes et al. (2012) reported that gender, birth-order, and diagnosis were not significantly associated with behavioural problems. They did however find that when the child has a chronic condition that is associated with a higher mortality rate and more intrusive treatment the

siblings were significantly more likely to have greater internalising and externalising problems, along with less positive self-attributes. This is corroborated by the findings from Rodrigues and Patterson (2006) who found condition severity to be significantly associated with family functioning in families where one child has a chronic illness or condition, and Incledon et al. (2015) who highlighted two studies that suggested disease severity to be negatively associated with QoL in siblings. However, Giallo and Gavidia-Payne (2006) did not find a significant relationship between the severity of the child's disability and the psychological outcomes of the sibling.

Sibling Support: Social support for siblings from those close to them can help to improve their psychological wellbeing (Incledon et al., 2015), in particular friends (Alderfer & Hodges, 2010), and other siblings in a similar position (Sidhu et al., 2005). Positive and consistent social support has been shown to reduce both internalising and externalising behaviours in siblings of children with cancer (Barrera, Fleming, & Khan, 2004). Emotional support from, and time with, parents along with greater parental awareness, have been noted to act as protective factors for sibling psychological wellbeing (Incledon et al., 2015). Conversely, Giallo and Gavidia-Payne (2006) noted that the siblings within their sample who had previously attended a sibling support group were more likely to have adjustment difficulties, which the authors argue could either suggest that sibling groups are detrimental to the psychological wellbeing of certain siblings or that the siblings that attend such group are those with the poorest psychological wellbeing.

Coping Strategies: There has been little research specifically on coping strategies in siblings. Higgins, Bailey, and Pearce (2005) study suggests that the coping strategy adopted by caregivers and families of child with ASD was not related to family cohesion or adaptability. However, Incledon et al. (2015) found that siblings choosing to positively reframe the way in which they viewed their brother's or sister's illness was associated with a reduction in anxiety, loneliness, and insecurity scores, and an improvement in emotional QoL (Houtzager et al., 2005; Houtzager et al., 2004).

Demographic Factors: Walton (2016) investigated correlates of sibling adjustment in families with a child with ASD. They found multiple demographic factors that acted as either a protective or risk factor for sibling internalising and externalising behaviours. Lower family income, being a male sibling, and a lower number of children in the family were associated with a greater number of externalising behaviours in siblings. Whereas, having a brother with ASD and being a younger sibling were associated with a greater number of internalising behaviours. Further studies have reported effects of birth-order, for instance, Limbers and Skipper (2014) report on two studies that found being the older sibling of a child with a chronic illness or condition could lead to a higher level of psychological distress, contrary to the findings in Walton (2016). There are several explanations given for the findings in Limbers and Skipper (2014), including the concept that younger siblings are more likely to be protected from sensitive information about their brother's or sister's condition by parents. The meta-

analysis by Vermaes et al. (2012) concurs with the findings from Walton (2016) in that they found no significant effect on sibling wellbeing from birth-order overall.

The socioeconomic status (SES) of the family has been connected to siblings' behavioural outcomes (Giallo & Gavidia-Payne, 2006). In the study by Giallo & Gavidia-Payne (2006) the relationship found between SES and adjustment in siblings was affected by parental stress and family resilience. Given less parental stress and positive family functioning, siblings from lower SES families may not necessarily be at an increased risk of poor psychological adjustment. This was reinforced by the findings from a previous study which noted that although SES and behavioural outcomes in siblings were significantly related, the relationship was mediated by maternal mood and family cohesion (Williams et al., 2002).

It is known that cultural differences can have a large effect on the way in which families deal with a chronic illness or condition (MCCubbin, Thompson, Thompson, MCCubbin, & Kaston, 1993). Culture has been found to alter sibling outcomes (Lauderdale-Littin & Blacher, 2017; Long et al., 2013; Smith & Elder, 2010; Vermaes et al., 2012). In their study Lauderdale-Littin and Blacher (2017), found that Anglo mothers reported lower stress levels relative to Latino mothers. Furthermore, they suggest that the caregiving expectation of siblings is different between Anglo and Latino mothers, with Anglo mothers being less likely to expect the sibling to provide care for their brother or sister in the future.

The vast majority of studies considering the psychological wellbeing of siblings concludes by emphasising the need to incorporate screening of siblings in the care for their brother or sister (Dia & Harrington, 2006; Rana & Mishra, 2015). Should regular screening of siblings and their families be undertaken it may be possible to assess factors such family dynamics, social support and sibling knowledge, and together with information on condition severity, treatment burden, and sibling birth order, it may be possible to identify siblings that are at the greatest risk of poorer psychological wellbeing and adjustment in order to provide support. Walton (2016) conclude their work by emphasising that individually each risk factor identified had only a small connection to sibling outcomes and the siblings most at risk of clinically significant psychological distress were those exposed to multiple risk factors, as was found in other studies also (Macks & Reeve, 2007; Walton & Ingersoll, 2015).

1.3.7. Methods and Approaches to Understanding Sibling Psychological Wellbeing

Multiple issues have been proposed about the methods with which sibling psychological wellbeing is researched and measured. Following their review of the literature on sibling psychological wellbeing, Hartling et al. (2014) suggested that the measures typically used in sibling research, such as the Strengths and Difficulties Questionnaire (SDQ) may not appropriate or sensitive enough to accurately capture the impact on siblings and typically provide results of no-effect. Issues with the measures regularly used in psychological research has encouraged researchers to move towards either

using a measure specifically designed for sibling research (Kao, Plante, & Lobato, 2009), or using a type of measure that incorporates aspects of life that may be more relevant to the sibling's situation, for instance QoL (Limbers & Skipper, 2014). However, measurement of QoL in siblings is also problematic. Moyson and Roeyers (2012) emphasise the need to consider the QoL of siblings from their own perspective and not through parental proxy. They found that when siblings of children with intellectual disability are asked about their QoL, the nine themes identified by siblings as important to their QoL (joint activities, mutual understanding, private time, acceptance, forbearance, trust in well-being, exchanging experiences, social support and dealing with the outside world) were different from the aspects traditional included in a family level QoL measure. Family QoL has been evaluated in several studies (Davis & Gavidia-Payne, 2009; Mazaheri et al., 2013), although these may not be representative of the impact on siblings QoL.

Parental proxy is regularly used in research involving children, including sibling research. (Limbers & Skipper, 2014). The accuracy and relevance of parental proxy has been questioned in sibling research. Parents tend to over report psychological symptoms in siblings relative to sibling self-report (Mazaheri et al., 2013), but parents appear to underreport social difficulties (Barlow & Ellard, 2006; Sleeman et al., 2010). There are positives to continuing to incorporate multiple respondents' perspectives in sibling research. A parent may notice an aspect a sibling doesn't and vice versa (Eiser & Morse, 2001b; Kraemer et al., 2003; Rankin, Tomeny, & Barry, 2017). It may be beneficial to also incorporate the perspective of the young person with the chronic illness or condition, although they are frequently not included in sibling research (Kovshoff, Cebula, Tsai, & Hastings, 2017). Perspectives of the whole family are incorporated into the qualitative research project presented in Chapters 3-8.

Much of the research on sibling psychological wellbeing has adopted either quantitative or qualitative methods, yet there are advantages to research that incorporates both. Mixed methods research, which brings together both quantitative and qualitative methods, is known to be beneficial beyond the use of each of these methods individually as each can corroborate and support the other (Packman et al., 2005). Velleman et al. (2016) implemented mixed-methods research in an attempt to enhance understanding of siblings of children with chronic fatigue syndrome, using the qualitative evidence they gathered through interviews to enhance their interpretation of the quantitative measures taken on QoL and psychosocial wellbeing. Further research that uses this approach is needed in the area of sibling psychological wellbeing in order to better understand how best to support siblings in the family context. Chapter 9 presents a mixed methods analysis that draws together the views of multiple respondents with the QoL of both the sibling and their immediate family.

1.3.8. Supporting Siblings' Psychological Wellbeing

How best to support siblings of children and young people with a chronic illness or condition is still uncertain (Hartling et al., 2014), as could be expected given the ambiguity of the existing

literature considering the impact and experiences of siblings. The need and potential benefits of supporting siblings of children with a chronic illness or condition has been recognised by not just parents and siblings but also clinical staff that may provide the support (Nolbris & Nilsson, 2017).

Despite limited clarity in understanding of the impact on siblings and the underlying mechanisms psychological wellbeing support services have been developed for siblings. There is a large variability in the support offered to siblings (Tudor & Lerner, 2015). In their review of support for siblings of children with ASD, Thomas, Reddy, and Sagar (2016) highlighted a high level of heterogeneity in several aspects of the included studies, such as methodology, age of participants, and measures used. They pinpoint this as particularly limiting on the conclusions they are able to draw on the effectiveness of support for siblings of children with ASD. Literature reviews considering interventions for sibling psychological wellbeing, such as the one completed by Thomas et al. (2016), have typically focused on one diagnosis or have segregated the sibling population in a way that reduced the number of studies they were able to include (Hartling et al., 2014; Tudor & Lerner, 2015).

Several different forms of support have been proposed for siblings. Camps that provide support for siblings have been considered in several pieces of literatures (Sidhu et al., 2006), and are generally viewed positively. Sibling support groups have also been developed and evaluated (Roberts, Ejova, Giallo, Strohm, & Lillie, 2016), along with family-centred support (Besier, Holling, Schlack, West, & Goldbeck, 2010; Giallo & Gavidia-Payne, 2008). As with the development, the evaluations of sibling support services vary greatly in their methods. There are a few outcomes that have been consistently considered in several of the evaluations, for instance the siblings understanding of their brother's or sister's condition or illness has been measured in multiple evaluations (Thomas et al., 2016).

Segregating siblings by the type of illness or condition their brother or sister has, e.g. physical or mental, is not currently supported by the literature, yet much of the existing support has done this. This thesis does not consider physical and mental health conditions separately. Where an attempt has been made to support one diagnosis in isolation there tends to be limited consideration given to the severity of the child's condition (Thomas et al., 2016), which has been highlighted as a potentially crucial factor for sibling adjustment (Roberts et al., 2016). The Medical Research Council (MRC) emphasise the importance of underpinning interventions using the appropriate theories and developing them systematically based on existing empirical evidence (Craig et al., 2008). The use of empirical evidence needs to be given more consideration when developing sibling support services.

1.3.9. Theoretical Approaches to Understanding Sibling Wellbeing

Theories that have previously been used in an attempt to clarify the impact on sibling include the diathesis-stress model (Orsmond et al., 2009), Family Systems Theory (Cridland, Jones, Magee, & Caputi, 2014), the Double ABCX model (Pozo, Sarriá, & Brioso, 2014), and the biological systems model (Bronfenbrenner, 2005). Each model approaches the subject in a slightly different manner; Some have

been used to consider one chronic illness or condition in particular, for instance, a diathesis-stress model of sibling functioning was proposed for those with a brother or sister with an ASD by Bauminger and Yirmiya (2001). Whereas other models, such as Family Systems Theory (Bowen, 1974), have been applied more broadly.

Bowen's family system theory treats the family as a single interconnected emotional unit. It proposes that an individual cannot, and should not, be considered in isolation. Rather it is important to consider the interdependent nature of emotions between family members. Dr Bowen used systems thinking to develop the theory through the integration of human behaviour and family research. Family relationships are likely to be some of the longest emotional connections an individual has (Reynolds et al., 2015) and it is presumed that the complex nature of emotional interdependence in a family unit presumably evolved to promote cohesiveness and cooperation in families (Kerr, 2000). Family system theory highlights the difficulties that are likely to be experienced by family members when another member of the family has a chronic illness or condition. Taylor, Burke, Smith, and Hartley (2016) used family systems theory as a basis for their research. They highlighted the fact that although family systems theory is widely accepted, research in families with a child with an intellectual developmental disability have typically only considered one view point in a family and are thus limited in their ability to understand the experiences of a family. This closely relates to the following work which considers multiple view points within families with cystic fibrosis.

Family systems theory has a clear relevance to the work in this thesis, however, this work is centred around The Sibling Embedded Systems Framework (Kovshoff, Cebula, Tsai, & Hastings, 2017). This framework attempted to condense multiple theories, including family systems theory, into a new framework that enhanced the contextualisation of siblings' experiences. Kovshoff et al. (2017) formed the Sibling Embedded Systems Framework to consider the numerous dynamic factors affecting siblings with a brother or sister with autism. The framework attempts to provide a map of the interactions between factors influencing outcomes of siblings. These factors are split into nested levels or "systems". The micro-, meso-, exo-systems, and macro-system, as shown in Figure 1.1. The micro- and meso-system include factors that the sibling experiences directly either within themselves or from external sources. The micro- and meso-systems are nested within the exo-system, which includes factors that the sibling may be indirectly involved with. Finally, the exo-system is nested in the macrosystem, which includes broader societal level factors. The factors included within the nested systems interact to form sibling outcomes. Kovshoff et al. (2017) suggest various outcomes that could be considered using the framework model, including psychological outcomes, relationship quality, and academic achievement, and highlight how these outcomes in turn feedback into the system.

Given the uncertainty about the direction and magnitude of the impact on siblings and the multitude of potentially influencing factors, to be viable theoretical models need to include a vast

number of factors, which can make them very challenging to test and validate. Difficulty in testing theories can leave them underutilised but experimental manipulations of key variables can illuminate key maintenance mechanisms (Clark, 2004). Qualitative projects, such as that included in this work can also be used to enhance the understanding of factors that siblings themselves identify as being integral to the impact they experience.

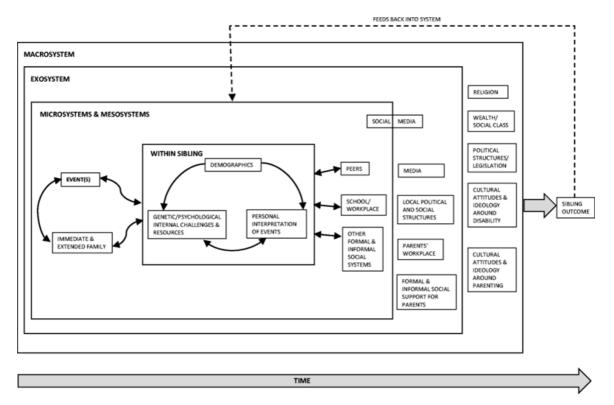


Figure 1.1. The Sibling Embedded Framework from Kovshoff et al. (2017)

1.3.10. Summary and Identified Gaps in the Literature

From the above literature review it is clear that there is little consensus on the impact on sibling psychological wellbeing and how siblings should be supported. Evidence is mixed in the direction and magnitude of the impact on sibling psychological wellbeing and QoL. This is also reflected in the qualitative evidence that suggests a potential for both negative and positive aspects of being a sibling. Further, there are multiple factors, including family related factors, demographics, and factors relating to their brother's or sister's condition that could have an effect on the extent of the impact sibling experience. Consideration for such factors when developing and targeting interventions may be important but the evidence on these again remains mixed and unclear. Given this uncertainty in the impact and underlying factors it is no wonder that there is a large variety in existing support for siblings. Finally, there is a lack of clarity in how support services have been developed for siblings and if they are derived from theories that have been proposed to explain the impact on siblings.

There are several gaps in the sibling literature that are very apparent, and need to be addressed. There is a need to systematically review all available evaluations of support services that have been offered to siblings. Currently there are two reviews available on the effectiveness of support for siblings (Hartling et al., 2014; Tudor & Lerner, 2015) however, as previously argued, these studies are limited due to arbitrarily segregating the sibling population into physical and mental health conditions, which is not clearly supported by the literature. This thesis considers mental and physical health conditions together for the first time. It has been well established that a relationship exists between physical and mental health (Prince et al., 2007; Scott, Lim, Al-Hamzawi, & et al., 2016). Those with chronic conditions regularly report poorer mental health symptoms especially anxiety and depression (Lavigne & Faier-Routman, 1992), and children with neurological conditions frequently report a greater number of externalising problems (Freilinger et al., 2006). It is important to consider and aggregate all available evidence on the effectiveness of existing sibling support services in order to allow a greater depth of understanding to be developed and common aspects of support to be identified. This will provide helpful information for the development of existing and new sibling support services.

Inconsistency in quantitative findings and the use of measures not sensitive enough to detect psychological impact in siblings is a key issue within existing sibling research. The clearest way to address this issue is to take a population that has been regularly assessed as one at risk of poor psychological wellbeing, such as siblings of individuals with a highly intrusive condition, and consider their experiences. Using a robust qualitative methodology and gathering a sufficiently large sample will also help address the uncertainty. This qualitative work will help contribute stronger more reliable evidence on siblings and the support they believe is needed. Evidence was also provided in support of using a mixed-methods approach. Where possible combining both qualitative and quantitative evidence will enhance research and form more robust and trustworthy estimates of the impact on siblings, something which is much needed given the current level of uncertainty.

Methodology should be at the forefront of all sibling research going forward. There is a large remit of evidence against using solely parental report in research and the use of self-report is encouraged. This could be further enhanced by the use of multiple respondents, for instance including all the family. Considering reports from the parent and the sibling's brother or sister would enhance the picture of the sibling experience, as is clear from the previously presented literature the sibling experience is a highly complex and dynamic subject that is influenced by many factors, some of which relate to the parent or the individual with the chronic illness or condition.

1.4. Chapter Summary

This chapter has provided an empirical grounding to the subject of sibling psychological wellbeing. Initially presented was the current understanding of the direction and magnitude of the impact on sibling psychological wellbeing. As there are high levels of heterogeneity in the quantitative

evidence on psychological wellbeing in siblings, to provide further clarity on the subject the qualitative evidence base on sibling's experiences was also considered. Factors that were highlighted, from both the quantitative and qualitative evidence, as potentially mediating or moderating the impact on siblings were considered. Given an increasing concern about the level of uncertainty in the literature and the large amount of variation in the methods used, reflection was then given to methodology in sibling research and how this may be improved. Following the evidence on impact, potential influencing factors, and methodology a brief overview of the support services that are currently or have previously been provided to siblings was subsequently given. Finally, evidence on the limited theoretical work surround siblings and their psychological wellbeing was presented. Given this empirical evidence gaps in the current literature were identified. To conclude a thesis overview including the aims and objectives will be given, which identifies how this thesis will address the identified gaps.

1.5. Thesis Overview

Chapter two of this thesis presents a systematic review of 17 studies and a meta-analysis containing eight studies that synthesises the available evidence on the effectiveness of interventions targeting the psychological wellbeing of siblings that have a brother or sister with a chronic illness or condition either physical or mental. Chapter three introduces a large national qualitative project investigating the experiences of families living with a chronic condition. CF is used as a case study for this work as it is a chronic condition known to have a high treatment burden and mortality rate, which was identified as a potentially influencing factor on sibling wellbeing. Subsequently Chapter four presents the results of a thematic analysis of 19 sibling interviews, Chapter five and six presents the results from interviews with 25 young people and 25 adults with CF, and Chapter seven presents the results of 25 parent interviews. Chapter eight further synthesis the results given in Chapters four to seven, considering the views and experiences expressed across the 94 interviews. Chapter nine continues to use CF as a case study, although the focus now shifts to the quality of life of families living with CF and how this relates to their experiences and sibling psychological wellbeing through a mixed methods analysis. Finally, Chapter ten draws together the evidence presented through all previous chapters to discuss the implications of the findings for understanding how to best support sibling psychological wellbeing.

1.5.1. Thesis aims and objectives

This thesis will aim to address the identified gaps in the literature and form a robust and well-rounded picture of the role support could play in the lives of siblings of children and young people with a chronic illness or condition. Specifically, the aims and objectives of the thesis are to:

- Systematically review and synthesise the evidence the available in existing evaluations of sibling support services available up to the end of 2016 (Systematic Review and Meta-Analysis: Chapter 2)
- 2. Develop and conduct a methodically robust, large sampled, national, qualitatively research project involving families living with a chronic condition that is known to have a high treatment burden and mortality rate (namely CF) in collaboration with a third-sector charity looking to enhance their support for siblings (Qualitative Research Project, Introduction & Methods: Chapter 3)
- 3. Consider the experiences of siblings from their own perspective (Siblings Qualitative Research Project Result: Chapter 4)
- 4. Use multiple viewpoints to consider the experiences of families and synthesise their perspectives to form a holistic understanding of the experiences of siblings and their immediate families living with CF (Parents, Young People with CF, and Adults with CF Qualitative Research Project Results: Chapters 5-7, Family Synthesis Qualitative Research Project Results: Chapter 8)
- 5. Enhance the findings from the qualitative research project with the use of mixed methods analysis. Connecting the QoL of families with their qualitative experiences (Mixed Methods Approach to Understand the Experiences of Families living with CF: Chapter 9)
- 6. Synthesise the findings from the systematic review and meta-analysis, qualitative research project, and mixed methods project to explore the findings within the context of existing research and theories of sibling impact. Concluding with suggestions for future research and support for siblings of children and young people with a chronic illness or condition (*Discussion and Conclusion: Chapter 10*)

Chapter 2: Support for Siblings and Their Families

This chapter and therefore, some of the included text, was taken from the published paper (Appendix 2.1): Smith, M. M., Pinto Pereira, S., Chan, L., Rose, C., & Shafran, R. (2018). Impact of Wellbeing Interventions for Siblings of Children and Young People with a Chronic Physical or Mental Health Condition: A Systematic Review and Meta-Analysis. Clinical Child and Family Psychology Review, 21(2), 246-265.

2.1. Introduction

Following on from the empirical evidence presented in Chapter 1, this chapter presents a systematic review and meta-analysis of the effectiveness of support provided to sibling of children or young people with a chronic illness or condition. By collating and synthesising the available literature on sibling support services and their effectiveness, this work will enhance the understanding of how siblings are currently supported, what services look like and who provides them and their effectiveness. As this work is the first to include all siblings, potentially influencing factors will be considered fully allowing the use of divisions in future research and the targeting of support to be more relevant and, hopefully, effective.

2.2. Background

2.2.1. Interventions and Evaluations

Wellbeing interventions have been suggested to help improve psychological outcomes (including anxiety, depression, stress, self-esteem and coping) of siblings of children and young people with a chronic illness or condition. These interventions have taken various forms, including group interventions (Heiney, Goon-Johnson, Ettinger, & Ettinger, 1990; Houtzager et al., 2001; Lobato & Tlaker, 1985; Smith & Perry, 2005), sibling training (Ferraioli, Hansford, & Harris, 2012), camps (Kiernan, Gormley, & MacLachlan, 2004; Sidhu et al., 2006), and family-based support (Besier et al., 2010; Giallo & Gavidia-Payne, 2008). A range of populations have been targeted, some have been disease specific (Dolgin, Somer, Zaidel, & Zaizov, 1997) while others have taken a broad approach (Cadman et al., 1988). The content and duration of the interventions are highly varied. Camp interventions are typically formulated from the concept of therapeutic recreation (Fine & Fine, 1996), which focuses on enjoyment and freedom in recreation. While other studies, particularly group interventions, have focused on psychoeducational components (Giallo & Gavidia-Payne, 2008; Granat, Nordgren, Rein, & Sonnander, 2012; Lobato & Kao, 2002).

Evaluations of interventions are limited and typically associated with methodological issues including small sample sizes (Marszalek, Barber, Kohlhart, & Cooper, 2011), a lack of intervention integrity tracking (Kryzak, Cengher, Feeley, Fienup, & Jones, 2015), and large heterogeneity (Ali et al., 2014). A previous systematic review that considered interventions for siblings of children with a chronic illness or condition and included articles published between 1985 and 2008, adopted a broad

approach and included 14 papers (Hartling et al., 2014). The definition of chronic illness or condition used in this review was unclear. Hartling et al.'s review found a large inconsistency in treatment effects on behavioural and emotional outcomes and highlighted the importance of the sensitivity of the measures used as several of the included studies reported the child to be within the 'normal' range of mental health prior to the intervention. It is suggested that this may cause a ceiling effect on results as their scores are unlikely to continue to improve beyond their current point. The most recent review by Tudor and Lerner (2015) included 16 papers, both qualitative and quantitative, that considered interventions for psychological functioning targeted specifically at siblings of children with developmental disabilities (DD). Tudor and Lerner initially argued that the experience of typically developing siblings of children with DD was distinguishable from siblings of children or young people with physical disabilities, yet within their conclusion they acknowledged that the best services for siblings may not make that distinction.

2.2.2. Aims of This Review

In summary, previous systematic reviews have suggested that there is a need for interventions to improve psychological wellbeing in siblings of children and young people with a chronic illness or condition. When considering this subject it is important to remember that children with a physical health condition have an increased likelihood of having a mental health condition (Lavigne & Faier-Routman, 1992) and that there is a close relationship between physical and mental health. There have been calls for physical and mental health to be more closely integrated (Prince et al., 2007; Scott et al., 2016). It is therefore of value to investigate the impact of the interventions regarding the psychological wellbeing of siblings of children and young people with either a chronic physical or mental condition or both. Additionally, no meta-analysis has been conducted on the effectiveness of psychological interventions for siblings of children and young people with chronic illness or condition. The aims of this review are to contribute to the literature by:

- 1. Conducting a systematic review to synthesise the literature that evaluates wellbeing interventions offered to siblings of children and young people with a chronic illness or condition
- 2. Conducting a meta-analysis to quantitatively evaluate the impact of the interventions included in the systematic review

2.3. Methods

2.3.1. Sources & Search Strategy

A systematic search was conducted, following the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines (Moher, Liberati, Tetzlaff, Altman, & Group, 2009) and Cochrane recommendations (Higgins & Green, 2011). Electronic database searches were completed along with reference list and citation hand searches, and grey literature searches. The

following databases were used: PsycINFO, EMBASE, CINAHL, PubMed, Scopus and Web of Science, and PsycExtra was used to search for grey literature. The search strategy was piloted in November 2016 and following review was re-run in January of 2017, by three independent researchers (MMS, CR & LC), to include all literature up to the end of 2016. The search strategy was built using the Participant, Intervention, Comparator, Outcome (PICO) framework, as suggested in PRISMA guidelines (Shamseer et al., 2015). The broad themes included in the search strategy were sibling, chronic condition, intervention, and mental health. The search strategy was adapted to each database. The full search strategy can be found in Appendix 2.2.

2.3.2. Study Selection

Studies were included if they evaluated an intervention offered to siblings of children and young people with a chronic health condition, as defined previously. It was required that the two children live together (or were of an age where it is assumed they would still live together i.e. below 18 years of age). The sibling must be considered "healthy" themselves and not a donor for the ill child or young person.

The intervention could take any form, provided it aimed to improve the psychological wellbeing of the sibling, and reported an outcome that is related to the mental health of the sibling, including direct psychological outcomes e.g. anxiety, depression, and stress, as well as related factors e.g. knowledge, social support, self-esteem, relationships, coping and adjustment. Family level interventions were not included unless there were sufficient (at least one) sibling specific outcome, as described above, reported.

Included studies could be mixed methods if they report the result of at least one quantitative measure. Any form of trial was accepted if it evaluated the effectiveness of the intervention, this included pre-post design trials. Studies were excluded if unavailable in English or French. Studies that involved bereaved siblings, and studies that looked specifically at sibling donors were also excluded.

2.3.3. Risk of Bias Assessment

The Effective Public Health Practice Project Quality Assessment Tool (EPHPP) (Thomas, Ciliska, Dobbins, & Micucci, 2004) was used to evaluate the quality of all papers included in this review. This tool was chosen as it has been shown to have a higher inter-rater reliability relative to the Chronic Collaboration Risk of Bias Tool (Armijo-Olivo, Stiles, Hagen, Biondo, & Cummings, 2012); and is appropriate for use across different study designs compared to other tools such as the ROBINS-I, which is only appropriate for non-randomised trials (Sterne et al., 2016). This allowed confidence in the consistency and reliability of the assessments.

The EPHPP evaluates studies on eight components: selection bias, study design, confounders, blinding, data collection methods, withdrawals and drop-outs, intervention integrity, and analyses. The

ratings for all but intervention integrity and analyses are combined to give the study an overall rating of Strong, Moderate or Weak.

Searches, study selection, quality assessment, and data extraction were completed by three independent researchers: MMS, CR, and LC. Any discrepancies were dealt with through discussion and if a consensus could not be reached the opinion of an additional independent researcher was sought.

2.3.4. Data Extraction and Analysis

A data extraction form was created, using the Effective Practice and Organisation of Care (EPOC) data collection form (EPOC 2013) as a base, to ensure sufficient data was collected from each study included in the systematic review and meta-analysis.

It was expected that there would be large heterogeneity in outcome measure used across studies, therefore a random-effects meta-analysis was conducted. A Standardised Mean Difference (SMD) and Restricted Maximum Likelihood (REML) technique was used to estimate effect sizes and weights in STATA 14 (StataCorp 2017). The SMD was estimated using Hedge's g technique; allowing for a smaller sample size relative to Cohen's d method, which is typically used in meta-analyses in this subject area (Cuijpers, 2016).

The SMD technique allows the combination of different scales that are measuring the same outcome. For instance, the Strengths and Difficulties Questionnaire (SDQ) (Goodman, 1997) and the Child Behaviour Checklist (CBCL) (Achenbach, 1991), both measure behavioural outcomes and have been noted to have highly correlated scores (Goodman & Scott, 1999).

This review was registered on PROSPERO (International Prospective Register of Systematic Reviews), registration number CRD42017056740 (Appendix 2.3).

2.4. Results

2.4.1. Search results

1,536 papers were identified from the initial searches. After removal of duplicates 980 records were screened. 913 were excluded based on title and abstract (n=904), format (n=7), and language (n=2). 67 full text articles were assessed for eligibility and 17 were included in the qualitative synthesis. Eight studies were included in the meta-analysis. Five of the papers included reported an outcome measure of behaviour, and five reported on knowledge change following intervention. The nine papers not included in the meta-analysis either did not report sufficient data, used a different study design, or did not use either a behaviour or knowledge outcome measure. The flow of papers through the process of eligibility can be seen in Figure 2.1 In the initial search ten reviews were identified including three systematic reviews (Hartling et al., 2014; Prchal & Landolt, 2009; Tudor & Lerner, 2015). Rather than including the reviews, as there were discrepancies with the inclusion criteria, it was decided that the individual papers from each should be reviewed against the eligibility criteria.

2.4.2. Baseline Characteristics

Across the 17 included studies there were 1,264 participants. Age of participants ranged from 6-15 years, with an average of 10.47 years⁴. There was a relatively even gender balance in the overall sample, with 53% of participants being female. Further demographic information can be found in Table 2.1.

2.4.3. Quality Assessment

Of the included studies eight (47%) were considered of weak quality (Besier et al., 2010; D'Arcy, Flynn, McCarthy, O'Connor, & Tierney, 2005; Evans et al., 2001; Giallo & Gavidia-Payne, 2008; Heiney et al., 1990; Houtzager et al., 2001; Kiernan et al., 2004; McLinden, Miller, & Deprey, 1991), seven (41%) were rated as moderate (Cebula, 2012; Dolgin et al., 1997; Granat et al., 2012; Kryzak et al., 2015; Lobato & Kao, 2002; Phillips, 1999; Williams et al., 2003), and only two (12%) were rated as strong (Sidhu et al., 2006; Smith & Perry, 2005). The two strong studies were both rated strong in the confounders, data collection methods, and withdrawals and drop-outs components of the EPHPP, and moderate in the remaining three components. The one RCT study included in the review was rated weak overall (Giallo & Gavidia-Payne, 2008). One paper was scored N/A for Withdrawals and Drop-Outs as it had one time point only and therefore quality assessment in this area was irrelevant for this paper (Cebula, 2012). A table of quality assessment results can be found in Appendix 2.4.

2.4.4. Interventions

Nine of the 17 studies included in this review were group-based interventions (53%); the next most frequent form of intervention was camp based interventions (18%). The studies were conducted in mainly high income, predominately Caucasian countries including Germany, UK, The Republic of Ireland, Australia, Canada, the Netherlands, and Sweden. The largest number of studies coming from one location was four, which were all based in the USA. The duration of the interventions ran from 4 days (Sidhu et al., 2006) to 96 months (Cebula, 2012), with a median and mode duration of 6 days. There was little consensus in the approach taken in the interventions, even between studies that used similar designs. Further details about the included interventions can be found in Table 2.2.

Of the 17 papers included six focused on physical illnesses, four focused on mental conditions, and the remaining seven focused on a combination of physical and mental conditions. Several studies focused on specific conditions, for instance four of the 17 studies offered interventions to siblings of children with cancer. Of the six papers that focused on physical illnesses much of their samples were made up of siblings of children with cancer (minimum 47.9%).

38

⁴From the twelve studies which reported average age

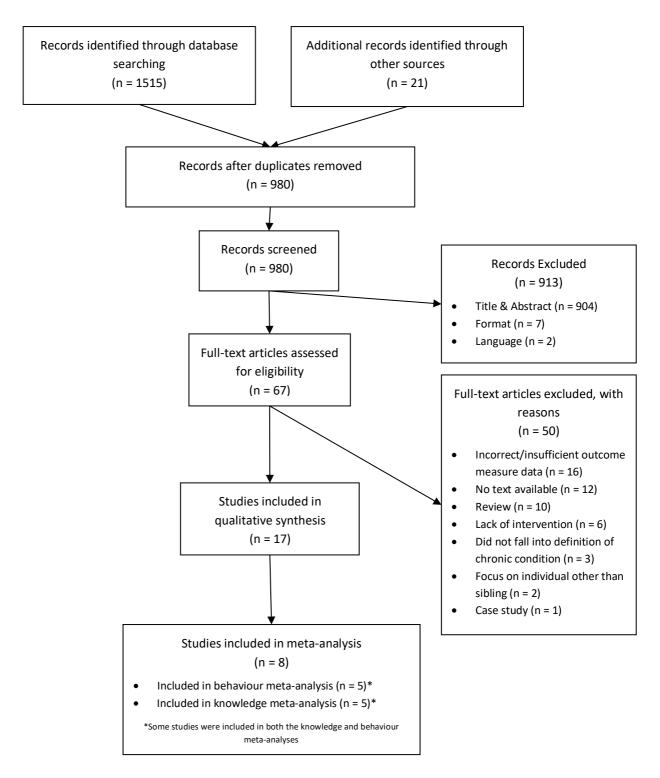


Figure 2.1. PRISMA Flow Diagram

Author(s)	Year	Total n	Experimental n	Control n	Other n (specified)	Age, yrs. Mean (SD) or Range	Gender (% Female)	Siblings Condition
				Chronic	Physical Illnesses o	or Conditions		
								Cystic fibrosis (20.5%)
Besier et al.	2010	259	259	N/A	N/A	8.58yrs (3.3)	45.60%	Congenital heart disease (31.7%)
								Cancer (47.9%)
Dolgin et al.	1997	23	23	N/A	N/A	11.71yrs (3)	48%	Cancer (100%)
Sidhu, Passmore, & Baker	2006	26	26	N/A	N/A	8-13yrs	52%	Acute lymphoblastic leukaemia (65%) Acute myeloid leukaemia (7.6%) Brain tumours (7.6%) Neuroblastomas (7.6%) Osteogenic sarcoma (3.8%) Hepatoblastoma (3.8%)
Heiney et al.	1990	14	7	7	N/A	9-15yrs	57%	Cancer (100%)
Houtzager, Frootenhuis, & Last	2001	24	24	N/A	N/A	11.3yrs (3.1)	62.50%	Cancer (100%)
Kiernan et al.	2004	119	119	N/A	N/A	11.5yrs (2.4)	45%	Cancer (52.2%) Haematological-related illness (21.7%) Not reported (26.1%)
				Chronic	Mental Illnesses o	r Conditions		
Kryzak et al.	2015	15	15	N/A	N/A	6-14yrs	40%	Autism spectrum disorder (100%)

								Mild mental retardation				
Phillips	1999	180	90	90	N/A	9-12yrs	60%	Moderate mental				
								retardation				
Smith & Perry	2005	26	26	N/A	N/A	10.63yrs (2.1)	54%	Autism				
Jimin & Ferry	2003	20	20	14//	14/1	10.03 y 13 (2.1)	3470	Addisin				
								Learning disabilities and				
Evans et al.	2001	28	28	N/A	N/A	6-12yrs	68%	associated challenging				
					cal & Mental Illnes			behaviour				
						Intervention: 11.1yrs (2.2)		Cystic fibrosis (4.4%)				
						, , , ,	50%	Diabetes (34.9%)				
Williams	2003	252	79	102	Partial Treatment: 71	Partial: 11yrs (2.5)		Spina bifida (9.5%)				
								Cancer (8.7%)				
						Control: 11.2yrs (2.5)		Developmental disabilities				
								(42.5%)				
		132								Applied Debergiousel Applysis		Autism: ABA (80%); ABA
					26 (5)	Applied Behavioural Analysis (ABA): 9.08yrs (2.4)	ABA: 53%	Control (73%); Post-ABA				
								(88%); Post-ABA Control: (85%)				
					26 (Post- Applied			Other (Asperger, ASD, HFA):				
					Behavioural		ABA-Control:	ABA (20%); ABA-Control				
Cebula	2012		45	45	Analysis (ABA)	ABA-Control: 9.25yrs (3.3)	60%	(27%); Post-ABA (12%);				
					& Post-ABA		3373	Post-ABA control (15%)				
					Control)	Post-ABA: 9.92yrs (2.3)	Post-ABA: 42%	Additional Diagnoses: ABA				
						, , ,		(16%); ABA control (13%);				
						Post-ABA Control: 9.67yrs (3.1)	Post-ABA	Post-ABA (15%); Post-ABA				
						. , ,	Control: 46%	control (15%)				
								Disability (a physical or				
D'Arcy et al.	2005	5 16	16	N/A	N/A	8-10yrs	45.45%	intellectual; disability, or a				
								combination of both)				

								Down's syndrome 4				
								Autism 5				
						Intervention: 11.75yrs (2.9)		ADHD 1				
Giallo & Gavidia-								Polymicrogyria 1				
Payne	2008	21	12	9	N/A		57.14%	Multiple disabilities 3				
layiic								Cystic fibrosis 1				
						Control: 11.00yrs (2.3)		Congenital heart disorder 2				
						Control. 11.00y13 (2.3)		Multiple illnesses 1				
								Williams syndrome 2				
	2012	2 54	54	N/A	N/A	8-12yrs	61%	ADHD 9				
Granat et al.								Asperger syndrome 7				
Granat et al.								Physical disability 8				
								Intellectual disability 17				
		2 54										Physical disabilities (26%)
							56%	Autism spectrum disorder				
								(23%)				
Lobato & Kao	2002		54	N/A	N/A	8-13yrs		Mental retardation (21%)				
								Medical disorders (17%)				
								Combined psychiatric and				
								learning disorders (13%)				
McLinden, Miller, &		91 11	1 6	5		9.82yrs (2.2)		Mentally retarded (45%)				
Deprey	1991				N/A		64%	Physically handicapped (9%)				
2-5651								Multiply handicapped (45%)				

Table 2.1. Summary of Demographics

Author(s)	Year	Name of Intervention	Who Delivered Intervention	How often/how many sessions were involved	Sessions	Protocol Available	Protocol Adherence Recorded	Intervention offered to other members of family	
Chronic Physical Illnesses or Conditions									
Besier et al.	2010	Family-Oriented Rehabilitation Programme	Psychosocial Team	4-Week Programme. Session offered 1-3 times per week	Psychoeducational group, exercise, relaxation, supportive/psychotherapy, parent-child sessions	Individually arranged treatment protocols	N/A	ill child admitted for rehabilitation. Parents also treated according to individually arranged protocols	
Dolgin et al.	1997	Structured Group Intervention	Clinical social worker, a child life specialist and a supervising psychologist.	Six group sessions were held on consecutive weeks	In addition to group discussions concerning their experience of the illness and its impact, subjects took part in arts and crafts and other creative activities in order to encourage interaction among participants and to promote nonverbal expression of relevant feelings and themes.	Detailed structure available	Unclear	No	
Sidhu, Passmore, & Baker	2006	Camp Onwards	Group facilitators (undertook pre- camp training workshop)	4-day	The program aimed to provide an opportunity to develop peer support networks and social competencies; provide age appropriate information on cancer, treatment and its impact on all the family; facilitate activities, that encourage the expression of feelings; and impart strategies to enhance adjustment to the family stressors in a safe environment.	Manual (soon to be published at point of paper publication)	No	No	

Heiney et al.	1990	Sibling Support Group	Co-therapists: a fellow in child psychiatry, and a paediatric oncology nurse	Seven 1-hour Sessions	The group was organised so that each session focused on specific topic: introduction and orientation, diagnosis, treatment, school, coping, family relationships, and the future.	No	No	Concurrent Parent Group
Houtzager, Frootenhui s, & Last	2001	Support Group for Siblings	Led by two well- trained psychologists	Five weekly sessions	First session: getting to know each other, second session: changes, third session: emotions related to illness, fourth session: Paediatric Oncologist invited to talk, final session: siblings visit the oncology ward.	No	No	No
Kiernan et al.	2004	The Barretstown Gang Camp	Unclear/Camp Staff	10-day sessions	Core Activities: Music, theatre, photography, arts and crafts, wordsmith, woodwork, canoeing, fishing, horse-riding, adventure, archery and camping. Periphery Activities: hangout, and evening activities. Social Activities: Cottage chat, rest hour and the opportunity to meet other from different countries (1).	No (more info: https://ww w.barretsto wn.org/)	No	Camp for children with life threatening illnesses and their siblings
				Chronic Mental	Illnesses or Conditions			
Kryzak et al.	2015	The Support and Skills Program (SSP)	Special Education Teacher, School Councillor, Volunteers, a Psychology Doctoral Student	Seven 2hr sessions	Focused on developing a network of peers who face similar family challenges, learning about Autism Spectrum Disorders (ASD) and coping strategies.	No	No	Support for Child with ASD
Phillips	1999	After-School Program	Six team leaders (community centre staff), and seven volunteers	15-week, after- school (3- 5:30pm) every weekday	Group Discussions, Recreation and Homework assistance	No	No	No
Smith & Perry	2005	Sibling Support Groups	Treatment, Research, and		Exercises, games, and activities that were fun and promoted group	No	No	No

			Education for Autism and Developmental Disorders (TRE- ADD) Staff	Weekly for 8 consecutive weeks	cohesion, providing information sessions on autism and related disorders, and facilitating discussion relating to feelings and attitudes associated with living with a brother or sister who has a developmental disability			
Evans, Jones, & Mansell	2001	Sibling Support Groups	Facing the Challenge' multi- disciplinary team. Comprised of nurses, a psychologist and outreach workers.	Three consecutive full days, and then on a weekly basis for six evenings. A final day at a local theme park.	Had a problem-solving focus. The majority of activities were of an educational and informative nature. Leisure activities were also used.	No	No	No
	T	T	Cl	hronic Physical & M	ental Illnesses or Conditions			
Williams	2003	Intervention for Siblings: Experience Enhancement (ISEE)	Paediatric nurse clinicians	5-days	Structured teaching about the brother/sister's illness, psychosocial session, a 5-day residential summer camp, and two booster sibling session and parent sessions	Brief Protocol Available	No	Parent sessions
Cebula	2012	Applied Behavior Analysis (ABA)	Mother/partner, outside agency, or parents & outside agency	2-96months/5- 40hrs per week	N/A	No	No	Intensive home- based intervention for child with autism. ⁵

⁵ Arguably ABA, although focused on the child with autism, teaches parents key behavioural strategies founded on the behavioural principles of learning and motivation, including reinforcement, extinction, stimulus control, and generalization (Granpeesheh, Tarbox, & Dixon, 2009). ABA strategies include the promoting of wanted behaviours, the fading of behaviour prompts, shaping approximations of wanted behaviours and chaining together the steps of a wanted behaviour. These strategies are likely to be relevant to parenting siblings also. As such ABA interventions are considered as 'family level interventions in this study.

D'Arcy et al.	2005	SibShops	Unclear. Clinical Psychologist conducted interviews	Once a month for four consecutive months	Consists of high and low energy activities, interspersed with discussion about disability and each siblings' experiences	No - based on the model developed by Meyer and Vadasy (1994)	Unclear	No
Giallo & Gavidia- Payne	2008	Sibstars	A clinician with postgraduate psychology training	Six weeks. Weekly telephone support offered to sibling and parent	After the first face-to-face session, each week families were required to read an information booklet and complete the practice activities provided.	Yes	Programme adherence checklist was used	Involved Parents
Granat et al.	2012	Sibling Group Intervention	Clinical staff from an outpatient habitation centre	weekly two-hour sessions	Content intended to increase knowledge and problem-solving skills	A manual (in Swedish) for clinical practice was compiled.	No	Separate education groups being provided for parents
Lobato & Kao	2002	SibLink	Doctoral level trainees in psychology or psychiatry	Six 90-minute sessions	Activities alternated between explicitly focused "main events" and other more social-recreational activities	Manuals available on request	No	Parent Group
McLinden, Miller, & Deprey	1991	Sibling Support Group	School Psychologists	Six week, 1-hour per week	Focused on developing participants' acceptance of both negative and positive feelings about their siblings. Information was provided and numerous activities were utilized.	No - based on Lobato (1985)	No	No

Table 2.2. Intervention Details

2.4.5. Systematic Review

Physical Conditions: Of the six papers that focused entirely on chronic physical illnesses or conditions, four of them focused exclusively on cancer diagnoses, while the remaining two incorporated Congenital Heart Disease, CF and other Haematological-related illnesses. Across these six studies there were nine different outcomes considered and 13 different measures used.

There was significant improvement in self-esteem (Kiernan et al., 2004; Sidhu et al., 2006), behaviour (Besier et al., 2010), knowledge, attitude and feeling, mood (Dolgin et al., 1997), and anxiety (Houtzager et al., 2001; Sidhu et al., 2006). Three studies examined Quality of Life (QoL), and all found significant improvements (Besier et al., 2010; Kiernan et al., 2004; Sidhu et al., 2006). There was no significant change in "coping" (Heiney et al., 1990), and affect (Kiernan et al., 2004).

Mental Conditions: Four of the 17 papers focused solely on children with a sibling with a chronic mental illness or condition. Two focused on autism spectrum disorders (Kryzak et al., 2015; Smith & Perry, 2005), one on learning disabilities (Evans et al., 2001) and one on mental retardation (Phillips, 1999). Across these four papers, ten outcomes were considered, and 12 different measures used.

There were significant improvements in self-esteem (Evans et al., 2001), sibling involvement (Evans et al., 2001), social support (Phillips, 1999), anxiety and depression (Kryzak et al., 2015; Phillips, 1999). There was no significant improvement in sibling interaction (Kryzak et al., 2015)⁶, sibling relationship, family functioning (Phillips, 1999), or coping and adjustment (Smith & Perry, 2005). Mixed results were noted from knowledge tests: Kryzak et al. (2015) found no significant improvement in Autism Sibling Knowledge, while a significant improvement in Autism knowledge was noted by Smith and Perry (2005). Evans et al. (2001) reported an improvement in knowledge about their siblings learning disorder but provided no statistical evidence.

Both Physical and Mental Conditions: Seven of the 17 papers did not specifically look at either physical or mental chronic illnesses or conditions. The study by Cebula (2012) noted that between 13-16% of their participants had additional diagnoses of physical health conditions, therefore their study is included in this section, rather than in the mental condition focused section. Prevalence studies suggest that children with a chronic physical illness or condition are more likely to have emotional/behavioural problems and psychiatric diagnoses (Hysing, Elgen, Gillberg, Lie, & Lundervold, 2007), and the same is true of young people with autism where comorbidity is regularly found, including psychological difficulties and physical conditions (Matson & Goldin, 2013). None of the included studies that examined only a mental or physical chronic illness or condition reported comorbidity and they were therefore unable to consider how comorbidities may influence siblings psychological functioning.

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⁶ As determined through behavioural observations of sibling dyads

Across these seven studies there were 15 outcomes considered, and 26 different measures used. Positive significant findings were found in the three studies that used a measure of "intervention impact" (Cebula, 2012; McLinden et al., 1991; Williams et al., 2003), in the two that evaluated coping and adjustment (Giallo & Gavidia-Payne, 2008; Lobato & Kao, 2002), and in the studies that looked at stress, family functioning (Giallo & Gavidia-Payne, 2008), mood (Williams et al., 2003), and connectedness (Lobato & Kao, 2002). Non-significant findings were found in the two studies that considered parent related variables (Cebula, 2012; Giallo & Gavidia-Payne, 2008). Of the seven papers, five considered behaviour (Cebula, 2012; Giallo & Gavidia-Payne, 2008; Lobato & Kao, 2002; McLinden et al., 1991; Williams et al., 2003) and knowledge (Granat et al., 2012; Lobato & Kao, 2002; McLinden et al., 1991; Williams et al., 2003) and knowledge (Granat et al., 2012; Lobato & Kao, 2002; McLinden et al., 1991; Williams et al., 2003), while two looked at attitude and feelings (McLinden et al., 1991; Williams et al., 2003) and sibling relationship (Cebula, 2012; McLinden et al., 1991), and one considered social support (Cebula, 2012), all of which produced mixed evidence.

Comparison: Coping and adjustment, knowledge and self-esteem were the only outcomes considered in all three categories (mental, physical, combined). Both (Giallo & Gavidia-Payne, 2008) and Lobato and Kao (2002) looked at a combination of health conditions and found a significant improvement in coping and adjustment, whereas Smith and Perry (2005) and (Heiney et al., 1990), who considered mental and physical conditions respectively, found no significant improvement. Both of the studies that found significant improvements in coping and adjustment involved the parents of the sibling, which may indicate that, although both studies were combined studies, this finding may be explained by factors other than the consideration of combined physical and mental conditions. The results for knowledge were spread across the types of study and there appeared to be no clear divide across physical, mental or combined studies. Self-esteem was considered in nine papers, of which six found significant improvements following intervention (Evans et al., 2001; Kiernan et al., 2004; Phillips, 1999; Sidhu et al., 2006; Smith & Perry, 2005; Williams et al., 2003), and three did not (Cebula, 2012; D'Arcy et al., 2005; McLinden et al., 1991). The three that found no significant associations were all combined studies, and only one of the six significant results was a combined study (Williams et al., 2003). It is unclear if this is due to study design. In the six papers that found a significant association, three were camps (Kiernan et al., 2004; Sidhu et al., 2006; Williams et al., 2003), and three were group support (Evans et al., 2001; Phillips, 1999; Smith & Perry, 2005). Two of the papers that did not find significant associations, were also a group support evaluation (D'Arcy et al., 2005; McLinden et al., 1991), and one was a service for the ill child or young person (Cebula, 2012).

2.4.6. Efficacy and Effectiveness

Due to the large heterogeneity in outcomes, small sample sizes and in some instances, poor study design it is challenging to compare the efficacy and effectiveness of the interventions. The only RCT included in this review, (Giallo & Gavidia-Payne, 2008) had a sample size of only 21 across both

O.treatment (12) and control (9). They offered a family-based psycho-educational based intervention called SibStars. Using seven outcome measures (including an evaluation of the intervention) they found a significant improvement in stress, coping and adjustment, the emotional symptoms subscale of the SDQ (behaviour), and family functioning.

As 11 of the 17 studies (65%) adopted a within-subjects pre-post design, without the use of a control, the results of these papers should be carefully interpreted and no assumptions of causality can be made. Within the discussion of two of the studies, consideration was given to the value of time spent together between the child and parent as a by-product of the intervention, but this was not accounted for this in their analysis (Houtzager et al., 2001; Williams et al., 2003).

2.4.7. Meta-Analysis

There were eight papers that could be included in the meta-analysis since they reported on the same outcomes using the same study groups and time points (Sutton et al., 2000). Of these eight studies, three were group interventions, four were family-based interventions, and one was an intervention for the child with the health condition. Two studies looked at mental conditions, two looked at physical conditions and four looked at a combination of the two. These numbers were too small to conduct a sub-group analysis in this meta-analysis so the studies were only considered for their results relating to behaviour (internalising, externalising and total score) and knowledge.

Behaviour: The results of the SMD meta-analysis on behaviour was split into three categories: internalising, externalising, and total score. Further, these papers were separated by whether they looked at pre-post measures in the treatment group, or compared the intervention group post treatment (Tx) to a control (Cntrl) group. Five papers were included in the analysis. The forest plots for each of the analyses can be found in Figures 2.2 and 2.3. Cebula (2012) report the SDQ from three different subjects (child, parent, and teacher), to be consistent with other studies, only the parent report was included in the meta-analysis.

The pooled SMD estimates in the pre-post analysis (Figure 2.2) all indicated improvement in behavioural outcomes on both SDQ and CBCL (as reflected through a reduction in score, relative to the baseline score). The pooled SMDs were as follows: internalising (SMD = -0.34 [95% CI (-0.50, -0.18); p<0.001]), externalising (SMD = -0.29 [95% CI (-0.45, -0.13); p<0.001]) and total score (SMD = -0.44 [95% CI (-0.6, -0.28); p<0.001]). In contrast, the meta-analysis of studies comparing a treatment group to a control group resulted in no significant difference in any of the scales of behavioural difficulties considered (Figure 2.3).

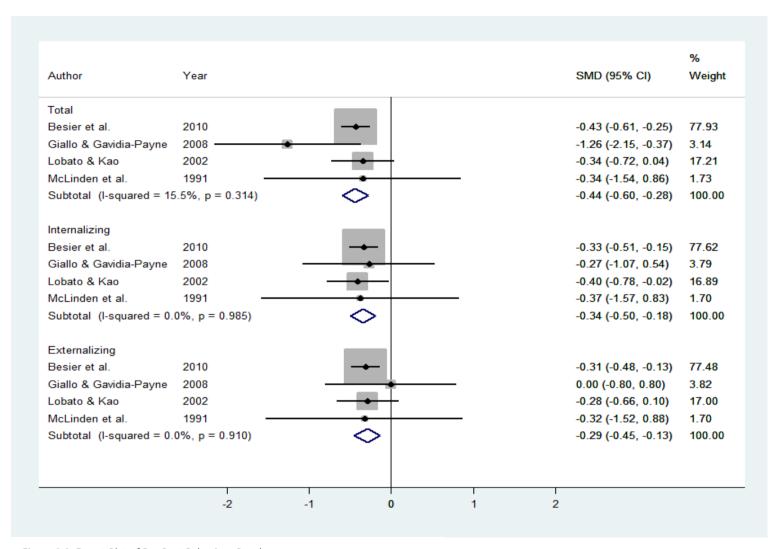


Figure 2.2. Forest Plot of Pre-Post Behaviour Results

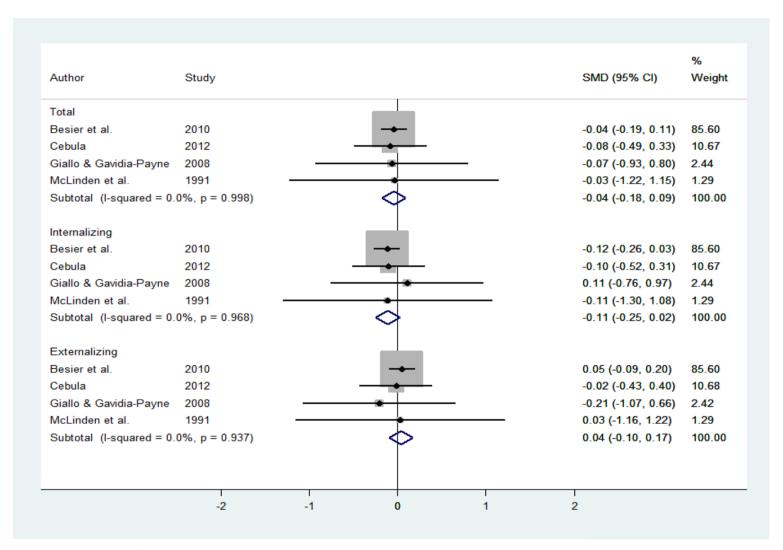


Figure 2.3. Forest Plot of Treatment (Tx) - Control (Cntrl) Behaviour Results

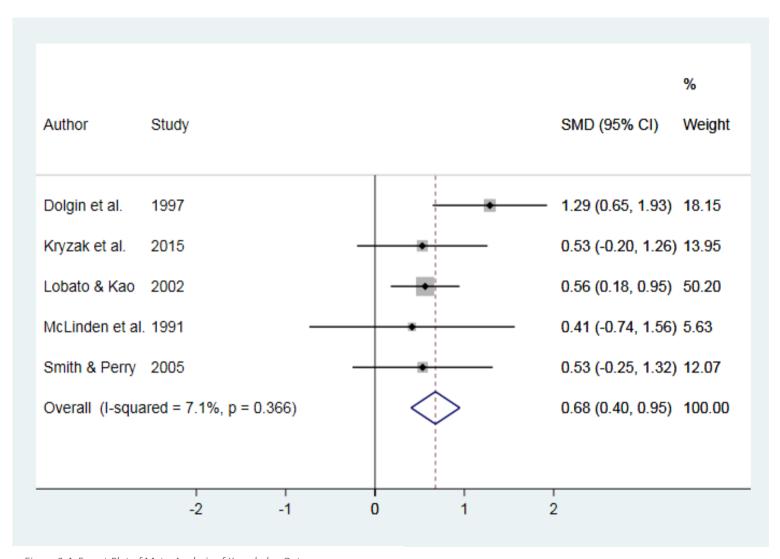


Figure 2.4. Forest Plot of Meta-Analysis of Knowledge Outcomes

Knowledge: No two papers used the same measure for knowledge. The results of the meta-analysis that included five studies demonstrated that overall there was a small significant improvement in knowledge following the intervention. Only one paper (McLinden et al., 1991) reported control group results, therefore this study design was not considered in this meta-analysis, rather all studies included used a pre-post treatment study design. The results of this analysis can be seen in the forest plot shown in Figure 2.4. The pooled SMD estimate for knowledge improvement following intervention is 0.68 [95% CI (0.40, 0.95); p<0.001].

Bias: Bias in a meta-analysis may be attributed to a range of sources including reporting biases, poor methodological design or chance. Typically, a funnel plot would be used to test for the presence of bias. A funnel plot plots the treatment effect (SMD) against study precision (standard error (se)). If the funnel plot is not symmetrical within the 95% confidence interval it is taken as a sign that there is bias present. To ensure that the asymmetry is not attributable to chance it is recommended to conduct a test for funnel asymmetry, such as Egger's Test (Egger, Smith, Schneider, & Minder, 1997). Egger's Test is not recommended if there are fewer than ten studies included in the analysis, as this is unlikely to distinguish true bias from chance due to low test power (Sterne et al., 2011). Therefore, due to the small number of studies included in this meta-analysis, no formal test for bias was completed.

2.5. Discussion

The literature evaluating interventions for siblings of children and young people with a chronic illness or condition is diverse and produces varied results. Studies included in this review included 1,294 siblings of children with a range of chronic illnesses or conditions, used various techniques to help improve the sibling's psychological wellbeing and a range of measures to evaluate several outcomes. Each of these sources of heterogeneity provides significant challenges for conducting a systematic review and meta-analysis. The heterogeneity of interventions and samples makes it particularly challenging to make firm conclusions on the effectiveness of interventions using a pre-post study design. Despite the challenges, the current meta-analysis provided some evidence of effectiveness of interventions for siblings of children and young people with a chronic illness or condition. The analysis of knowledge scores using pre-post measures indicated a small significant positive effect on knowledge following the intervention. Offering a knowledge component in an intervention could help facilitate the sibling's understanding of their brother's or sister's condition. By increasing understanding, it is possible the sibling will feel they have more control and this may help increase their coping skills (Heiney et al., 1990) although there has been relatively little work in this area.

When the treatment group were considered pre-and-post treatment there was a significant improvement in their internalising and externalising behaviours, as well as total score (reflected by a reduction in scores). Yet when we consider treatment group post intervention relative to a control group, there was no difference in behaviour scores. One suggestion as to why this may be is that the research process increased the salience of the needs of the sibling to parents in the control group which means their

outcomes also improved. Data collected in the included studies did not allow for this factor to be considered. Future research should attempt to account for such contamination effects. Further explanations include a lack of difference from control at pre-treatment, low sensitivity of measures, or potentially a bias from parent reported measures, consideration for each potential explanation will follow.

The sensitivity of the measures used in evaluations of this type should be further considered. For instance, a few of the included studies noted that the siblings were within the 'normal' range before receiving the intervention, and this had the potential to cause a ceiling effect on gains from the intervention (Giallo & Gavidia-Payne, 2008; Kiernan et al., 2004; McLinden et al., 1991). As previous meta-analyses (Sharpe & Rossiter, 2002; Vermaes et al., 2012) have found a significant negative effect on sibling's psychological functioning there appears to be a discrepancy between these samples and some of the samples considered in this review. This may be attributed to the samples themselves being unrepresentative, or may be related to the measures used in these evaluations.

Another explanation as to why results differed by study design may be that the range in parent or sibling report measures resulted in either a downward (under-reporting) or upward (over-reporting) bias on the results. This single-rater bias has been noted in the literature (Rivers & Stoneman, 2003). Within the papers included in this review three consider the effect of parental reporting bias on results. Cebula (2012) compared the results given to the SDQ by parents, teachers and siblings and found that siblings view themselves significantly more negatively compared to their parents. Sidhu et al. (2006), recognised that parents are only able to report on the externalising behaviours of the sibling, whereas the sibling could report on their internalised problems, and their perceptions of these distresses were generally greater than parents. Finally, Lobato and Kao (2002) found a difference in ratings on the Sibling Perception Questionnaire (SPQ), and bring into question the sensitivity of parent reported results using SPQ.

The positive impact of the interventions on behaviour may operate via a change in the parent-sibling dyad. For instance, parents may spend more time with siblings as a direct result of being involved in the intervention, or it may be that parents gain a higher awareness of the sibling's needs due to the intervention. The increased time spent together between the sibling and parent could have an influence upon the result, but it is challenging to formally record this for it to be considered in the analysis (Houtzager et al., 2001; Williams et al., 2003).

Several of the studies that considered both physical and mental chronic illnesses or conditions gave justification as to why they chose to do so (Giallo & Gavidia-Payne, 2008; Williams et al., 2003), but there is a lack of evidence as to which approach, disease-specific or broad, produces the optimum results. Evidence put forward by Vermaes et al. (2012) suggests that illnesses with a high morbidity and mortality may act as the largest moderating factors. Therefore, it may be beneficial to focus on siblings of children and young people that have a high impact, high mortality rate condition, regardless of whether the conditions was categorised as physical or mental.

2.5.1. Strengths and Limitations

This review is the first to synthesise the current literature evaluating interventions offered to siblings of children with chronic physical or mental illnesses or conditions or both. Previous reviews have attempted to separate out the two groups, by physical or mental conditions, and no previous meta-analysis has been completed in this area. The use of the broad approach allows a more complete picture of interventions currently available to siblings, by considering studies that have evaluated interventions focused on siblings of children and young people with a chronic mental or physical illness or condition together. It has also highlighted how it may not be the most advantageous approach to consider these groups separately. Consideration has been given to various forms of interventions and has highlighted the importance of more robust and replicable research in this area.

The heterogeneity of the studies included in the review make it difficult to draw firm conclusions regarding the effectiveness of psychological interventions aimed at siblings of children with chronic illnesses or conditions. The studies were typically subject to low sample sizes, poor methodology, and short follow-up periods; less than a quarter (24%) of the included studies report on a follow-up beyond 1-month post-intervention. Although it is advantageous that this analysis has included studies that consider both physical and mental conditions, this may also have increased the level of heterogeneity and reduced the clinical relevance, relative to reviews that have focused on solely mental or physical conditions (Tudor & Lerner, 2015). It should also be noted that the included studies were all from developed countries and therefore the results from this analysis cannot be generalised to those in low and middle-income countries.

Only eight studies were included in the meta-analysis due to a lack of consistent and compatible data. Of the included eight, five used an uncontrolled pre-post study design (Besier et al., 2010; Dolgin et al., 1997; Kryzak et al., 2015; Lobato & Kao, 2002; Smith & Perry, 2005), which makes causality difficult to establish. The four studies included in the meta-analysis that used a treatment-control design used various control groups, one used a waitlist control (Giallo & Gavidia-Payne, 2008), one used the participants that refused to participate in the intervention (McLinden et al., 1991), another used a data sample from the general population (Besier et al., 2010) and finally, Cebula (2012) used a retrospective design and thus their control were willing subjects who have not previously/were not currently using the intervention which limits the generalisability of the findings.

Further limitations of the included studies include a lack of acknowledgement for potential positive impacts of having a sibling with a chronic illness or condition. There is also a lack of consideration for the influence of parent-report relative to child-reported outcomes. Cebula (2012) report measures from parents, siblings and teachers to attempt to deal with this issue. In her analysis she found that the child or young person reported themselves more negatively on two of the five domains of the SDQ, but they also appeared to have a slightly more positive perception of the sibling relationship, particularly empathy which

may be due to the parents' greater attention to negative interactions (Cebula, 2012; Rivers & Stoneman, 2003).

Across all studies there were 23 outcomes considered, sufficient data were reported to combine behaviour and knowledge scores in a meta-analysis. The results of this analysis were limited by the small sample sizes of previous studies, along with methodological problems due to the lack of consistency in measures being used, type and protocol of interventions, and time-points being considered. It would be more beneficial if intervention studies that evaluated the same form of intervention, with the same type of population and using the same outcome measures could be statistically combined; unfortunately, with the current literature this is not possible.

2.6. Directions for future research

The primary recommendation from this review and meta-analysis is the need for stronger evidence, such as RCTs, which also capture a larger more representative sample of the population. Identification of the tools that should be used in evaluations also requires deliberation to help encourage consistency across studies. Future studies should be conducted that include siblings of children with both mental and physical conditions. It may also be important to consider potential moderating factors, including protective factors, which could help target and tailor support services to those most in need. For instance, considering different populations based on moderating factors i.e. high/low burden, using the same intervention protocol would provide more informative evaluations in this area of research.

2.7. Conclusion

This review and meta-analysis improved upon the current literature by combining the existing findings in a systematic and robust manner, providing transparent results and reducing potential sources of bias. It is concluded that psychological wellbeing interventions for siblings of children and young people with chronic physical and mental illnesses or conditions lead to an improvement in illness knowledge and an improvement in externalising and internalising behaviour scores, when using a pre-post one group study design. The findings from the systematic review are mixed and inconsistent, which emphasises the need for additional work that better establishes the needs, appropriate methodologies and evaluation techniques for interventions offered to siblings of children with chronic illnesses or conditions⁷.

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⁷ In the Viva, (Hastings 2003) was brought to my attention as a paper that was not detected within the search strategies. This indicates that the search terms may not have been optimal. However, Hastings (2003) does meet the inclusion criteria as it considers the behavioural adjustment of sibling of children with Autism following ABA intervention. Many of Hastings's findings agreed with the papers already included, and Hastings (2003) rated weak quality overall using the EPHPP. Thus, inclusion of Hastings within the systematic review did not substantively change the conclusions. In the updated meta-analysis, the SMD estimate for the total difficulties score changed from -0.04(-0.18, 0.09) to -0.02(-0.14,0.09), the internalising subscale changed from -0.11(-0.25, 0.02) to -0.04(-0.19,0.1), and the externalising score changed from 0.04(-0.10,0.17) to 0.06(-0.05, 0.18). Therefore, the meta-analysis/systematic review results are robust to the inclusion/exclusion of the Hastings paper.

A Sibling's Experience (2)

Presented below is a first-hand account of being a sibling given by Liam* (14) whose brother Noah* has cystic fibrosis.

"I was just two and a half when my brother Noah was born, I couldn't understand at first why I could not see him and I was being cared for by my grandparents. This meant that I was not able to see my parents for a week or more as Noah was taken by ambulance to hospital and I was far away so I wasn't able to see him. Whilst Noah was at the hospital my mum and dad were staying with Noah without me, however a week later I was able to join them and see him daily. And that was the start of living life with a brother with CF.

Now Noah is almost 12 I know a lot about CF and how it is to live with a brother with it and know his life is always going to be much different to mine but he is always outside playing rugby and scooting and is making use of his life.

Even now I found it very hard to talk to anyone about it even Noah and my parents. I don't ever like going to the hospital with him, it upsets me seeing him in that environment. Whenever Noah goes into hospital I always feel bad because I feel like I'm a left out because my parents are always with him and not ever spending time with me. But thankfully Noah has not been in hospital for a long spell from over a year now.

Although Noah's start to life was not great, we now both play rugby and are always outside enjoying our local environment. With Noah's medications and the care from the hospital we can now share an active life together, but I do secretly worry about how his life will be as an adult."

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 $[^]st$ Names and locations have been changed in order to protect the identity of the sibling and their brother

Chapter 3: A Qualitative Examination of the Daily Impact of Cystic Fibrosis – Background & Methods

This project was funded by the Cystic Fibrosis Trust and a full report of the findings was sent to the Trust in order to help them to enhance and develop support services for families living with cystic fibrosis. The candidate was included as a co-investigator in the funding confirmation (Appendix 3.1).

3.1. Introduction

Qualitative research allows a full and detailed picture of the experiences of siblings to be built. Given the disparity in quantitative evidence on the impact on siblings' psychological wellbeing, using qualitative evidence to develop a greater understanding of siblings' own perspectives on their experiences could help to enhance the appropriateness of future research and support. The following chapter provides the background and methodology for a large qualitative project that considered the experiences of siblings with a brother or sister with cystic fibrosis (CF) and their immediate family members. By considering multiple perspectives in family research it is possible to build a holistic understanding of the impact on siblings. The project is focused on siblings that may be at a higher risk. Given the previous findings from both Chapter 1 and 2 it could be thought that a condition with a high treatment burden and mortality rate, such as CF may be associated with a greater impact on siblings (Vermaes et al., 2012). This project utilises CF as an exemplar condition from which more general conclusions can be drawn.

3.2. Background

3.2.1. Cystic Fibrosis (CF)

Cystic fibrosis (CF) is the most common inherited genetic condition in Caucasian individuals (Hamosh et al., 1998), affecting approximately 10,500 people in the UK (CF Trust Registry Report 2018). The symptoms of the condition can be severe, for example persistent coughing, frequent lung infections and in some cases infertility. However, the way CF affects individuals varies greatly, largely due to the 1,400 possible mutations of the faulty gene (Alibakhshi, Kianishirazi, Cassiman, Zamani, & Cuppens, 2008). CF causes a thick mucus to build up in parts of the body including the lungs and digestive system (Donaldson et al., 2006). This leads to difficulties, particularly with infection, and can cause complications such as Cystic Fibrosis Related Diabetes (CFRD). The treatment and management of CF can be extensive and typically includes medication, physiotherapy, and diet adjustments. This treatment regime is time consuming, and can take over an hour each day (Ziaian et al., 2006). Currently, CF remains a progressive, incurable, and, for the majority, a fatal disease (Lask, Bluebond-Langner, & Angst, 2001). CF was previously seen as a childhood illness, however, those born with CF in 2017 had a median predicted survival age of 47 years. This is a notable improvement from the median predicted survival age for those born ten years earlier of 35.2 years (CF Trust Registry Report 2007),

and a drastic improvement on the six months life expectancy given to those born in 1938 (Andersen, 1938). Individuals with CF and their families are continually facing new challenges that have yet to be very well understood, which will be discussed in the following background section. The need for further research on topics affecting families with CF has been recognised (Hollin et al., 2019).

3.2.2. Siblings and Parents of Those with CF

As the conditions surrounding individuals with CF continue to develop and change, the needs and experiences of the family need to be reconsidered. The family has been highlighted in several pieces of literature about CF, mental health, QoL and development (Berge & Patterson, 2004). How families function when they include a child with CF has been considered widely in the literature. Research has typically emphasised parents, especially mothers as the primary caregivers, although some have also highlighted the importance of balanced family coping (i.e. both mothers and fathers) (Patterson & Garwick, 1994). Generally, the findings suggest that families adjust well to a life with CF and have outcomes similar to that of the general population. There is also evidence to suggests that families with CF could have better outcomes than families with a child with Autism or Diabetes (Berge & Patterson, 2004). Berge and Patterson (2004)'s review of the evidence pertaining to families with a child with CF established five key areas where CF impacted a family. The first was treatment compliance and physical health status, where the evidence suggested that greater parental involvement negatively related to treatment adherence (Foster et al., 2001) and good family coping and greater parental time at home were positively related to the child with CF's health (Patterson, McCubbin & Warwick, 1990). Secondly, they highlighted the importance of family functioning both positive and negative. A lower level of family stress was found if the child with CF has older siblings (Cappelli et al., 1988; Johnson, Muyskens, Bryce, Palmer, & Rodnan, 1985). Positive family functioning was more influential than disease severity on outcomes for both the child and family (Olmsted, Lewis, & Khaw, 1982). The final three topics highlighted by Berge and Patterson (2004) concerned the psychosocial problems of the child with CF, their siblings, and parents respectively. They presented findings that suggested the potential for psychological distress in each group. However, the literature they cite is limited to 2002 or earlier, at which point the median life expectancy for an individual with CF was around 20 years less than it was in 2017 (Jackson & Goss, 2018).

The intensive treatment regimen, life-limiting nature of the CF and the genetic transmission can cause negative psychological outcomes for parents including feelings of anxiety, stress and guilt (James et al., 2006). In a sample of 650 caregivers over a third were found to present symptoms of anxiety, nearly double that of the community sample (Besier et al., 2011). Besier et al. (2011)'s sample of caregivers also reported greater symptoms of depression relative to a community sample, and both symptoms of anxiety and depression were associated with a lower life satisfaction. The mental health of the parent has been linked to outcomes for their child (Brucefors, Hochwälder, Sjövall, & Hjelte, 2015; Cappelli et al., 1988; Czyzewski, Mariotto, Bartholomew, LeCompte, & Sockrider, 1994),

therefore it continues to be important to support parents and carers both for themselves and their child.

Research on the impact on siblings of those with CF is limited, as was acknowledged by Berge and Patterson (2004) in their review of the evidence. In the study by Havermans et al. (2011) it was suggested that siblings of children with CF reported a higher QoL than those with a healthy brother or sister. Although this improvement in QoL was reduced if the sibling was older than their brother or sister with CF, or if their brother or sister had had a recent pulmonary exacerbation. This thesis continues to develop the understanding of the impact on siblings with a brother or sister with CF and helps identify further aspects of the sibling experience that need consideration.

3.2.3. Psychological Wellbeing of Those with CF

Families living with a chronic illness or condition have complex and highly interconnected outcomes (McCubbin, 1988). As discussed in Chapter 1 the use of multiple respondents in family research allows a holistic understanding of the sibling experience to be created. Therefore, consideration needs to be given to the existing evidence on the experiences of those with CF also.

Quality of Life: One study that considered QoL in children and young people with chronic respiratory illnesses found that those with CF had poorer psychological health, emotional function, school functioning and overall QoL scores relative to those with asthma (Bodnár et al., 2015). Bodnár et al. (2015) also found that the QoL of those with CF was poorer than the general population. The authors suggested these findings may be due to the additional treatment burden and the progressive nature of CF relative to asthma. In several other studies it was suggested that in those with CF QoL is comparable to their contemporaries without a chronic illness or condition (Congleton, Hodson, & Duncan-Skingle, 1996; Havermans, Colpaert, & Dupont, 2008).

Various factors have been found to directly relate to QoL in those with CF. It is regularly noted that women with CF have worse QoL relative to men (Arrington-Sanders et al., 2006), which may be related to the lower life expectancy of females relative to males (FitzSimmons, 1993). Disease severity, culture, and mental health have also been noted to have a close relationship with QoL (Abbott, 2009; Arrington-Sanders et al., 2006; Havermans et al., 2008). The presence of anxiety or depression in adults with CF has been highlighted to lower QoL across several subscales (Havermans et al., 2008). How QoL relates to health markers, such as lung function, in CF has been investigated in several studies in both children and adults (Gee et al., 2005; Riekert, Bartlett, Boyle, Krishnan, & Rand, 2007).

Mental Health: The mental health of those with CF is frequently found to be comparable to those without a chronic illness or condition (Havermans et al., 2008; Szyndler, Towns, van Asperen, & McKay, 2005). Psychological need has, however, been found in both young people and adults with CF. Young people with CF who are subject to key risk factors such as alienation from family members or a later diagnosis may have an increased rate of psychological distress (Szyndler et al., 2005). White, Miller, Smith, and McMahon (2009) identified a higher incidence of several DSM disorders in their

sample of 53 young people with CF compared to the general population, including anxiety and elimination disorders. The key interest of the study by White et al. (2009) related to the implications of such diagnoses on adherence to medication and treatment regimens. They found that young people who have an anxiety disorder are more likely to adhere to their treatment regimen, although they questioned whether those with anxiety inflated their self-reported level of treatment adherence.

Adults with both CF and anxiety symptoms have been found to report poorer vitality and emotional functioning. Adults with depression symptoms also reported lower emotional functioning along with greater eating disturbances and poorer body image (Havermans et al., 2008). Despite only 30% of their participants screening positive for depressive symptoms, Riekert et al. (2007), highlighted a negative relationship between lung function, Health-Related QOL (HRQoL), and depressive symptoms. Given the increasing median survival age of individuals with CF the mental health and quality of life of adults is of growing interest. Challenges which were previously not considered for those with CF, such as employment and retirement, provide additional complexities to a life with CF and have the potential to add additional emotional distress (Habib et al., 2015).

Coping Mechanisms: Coping has received a large amount of attention in the literature on those with CF (Abbott, 2003; Abbott, 2001a). It is reported that those with CF and their families have developed ways in which they can cope with the additional stresses put upon them by CF (Venters, 1981). How individuals with CF develop throughout childhood and adolescence may affect their ability to cope with the stresses of CF (Ernst, Johnson, & Stark, 2010). In peer interactions, it has been found that adolescents with CF tend towards less social engagement and more assertive behaviours, which may allow the development of both negative and positive behaviours further down the line (Meijer, Sinnema, Bijstra, Mellenbergh, & Wolters, 2000).

3.2.4. Previous Qualitative Research in CF

The vast majority of previous research in CF is quantitative in nature. High quality qualitative research in this area would help to develop a more in depth and well-rounded understanding of a life with CF (Braun & Clarke, 2006). This is particularly important given the lack of explanation for the inconsistent findings on psychological wellbeing and QoL in those with CF. Qualitative research may help to shine a light upon the complex reality of living with CF. Previous qualitative research on CF has typically focused on young people's perspectives and experiences. A systematic review of qualitative studies involving children and young people was conducted in 2014 (Jamieson et al., 2014). From their synthesis Jamieson et al. (2014) concluded that young people with CF reported greater resilience, lifestyle restriction, resentment of chronic treatment, temporal limitations, emotional vulnerability, and transplant expectation and uncertainty. The authors take these outcomes to reinforce the importance of patient-centred care and its role in improving patient outcomes.

Parents and siblings have again received a limited amount of attention in previous qualitative research and are quite regularly only involved in order to give their perspective on the impact CF has

had on their child or brother or sister, rather than focusing on their own experiences (Savage & Callery, 2005). The work that has looked at parents experiences has highlighted how large an impact parents feel and the importance of coping methods (Wong & Heriot, 2008). How a parent copes has been linked to outcomes in both young people with CF and siblings (Ernst et al., 2010).

3.2.5. Cystic Fibrosis Research in the UK

Qualitative research into the experiences and opinions of families living with CF is lacking in general, although this is particularly the case in the UK. How individuals experience a life with CF is likely to differ by country given the difference in healthcare systems and policies (Abbott, 2001b). Within the UK itself there are differences in the provision of care and costs encountered by those with chronic illnesses or conditions (Taylor-Robinson & Schechter, 2011). How this changes the experiences of siblings is not currently known but is important to consider when conducting research within the UK.

A multicentred approach has previously been used in CF research (Bradley, Blume, Balp, Honeybourne, & Elborn, 2013) but it has not yet been done qualitatively within the UK. The benefits and potential challenges of multicentred research have been considered previously (Appel, 2006). Some advantages of multicentre research include greater recruitment and generalisability, as, for example, it is hard to say the experiences of a participant in England are likely to be representative of those of people in Scotland. Disadvantages include the added costs and logistical requirements, however in the case of this research this was viewed as a cost worth incurring to improve the quality and representativeness of the findings.

3.2.6. Support Services and the Third Sector

The third sector plays a vital role in reaching out to those in need and offering crucial support, in whichever form this may be. The qualitative and mixed-method projects as presented in Chapters 3 to 9 of this thesis were funded by the Cystic Fibrosis Trust and the results of these projects will be used to aid the Cystic Fibrosis Trust in the design and development of support services. The Cystic Fibrosis Trust is a UK based charity that focuses on "living a life unlimited by cystic fibrosis". Currently they provide a range of support services, including a helpline for anyone affected by CF, financial support, a range of information resources, an opportunity to connect with others online through social media and online forums, along with support for travelling with CF8. Through funding this work the charity wanted to learn how best they may be able to support the whole family through a life with CF, along with improving their understanding of the impact CF has on an individual's life as they move into and through adulthood.

⁸ https://www.cysticfibrosis.org.uk/the-work-we-do/support-available

3.3. Qualitative Research

Qualitative research methodology is not a singular method but rather a paradigm that guides researchers through a series of assumptions and choices to form an appropriate research project. Qualitative research projects aim to enhance the contextual understanding of the meaning and experiences of participants through the analysis of non-numerical data. Whereas, quantitative research uses statistical methodology to control for measurable numerical elements of a phenomena. When choosing between qualitative and quantitative paradigms the researcher has to make certain assumptions concerning knowledge (epistemology) and reality (ontology). These assumptions shape the research project, from the methodology employed to the type of questions asked (Hathaway, 1995). Assumptions underlying quantitative methodologies tend to assume that all phenomena can be observed, measured and tested systematically. Approaches such as positivism underlie much of quantitative research (Hathaway, 1995). The results from qualitative research are often regarded as being "grounded in the human experience" (Sandelowski, 2004). Although approaches to qualitative research are diverse they tend to have a commonality in their belief of the subjective nature of experiences and individual social construction of reality and meaning. Common approaches to qualitative research include constructivism or interpretivism (Searle & Willis, 1995).

Given the decision to use a qualitative paradigm, researchers can use a variety of data collection techniques and sources to consider their research question. Common sources of qualitative data include interviews, focus groups, observations and imagery. One of the most common data collection method used is an interview. Qualitative research tends to use either a semi-structured or unstructured interview technique (Edwards & Holland, 2013). Mason (2017) explains the three components involved in a qualitative interview. The first component involves an interviewer and at least one interviewee interacting through dialogue, typically either face-to-face or over the phone. Secondly, the interviewer has certain topics they want to discuss via a flexible and fluid structure. Finally, they suggest that an interview involves the creation of meaning and understanding through the interaction between the interviewer and the interviewee, this is also commonly known as co-production. Furthermore, Edwards and Holland (2013) emphasise that the interview is a learning process for both the interviewer and the interviewee. Through interacting with each other they are able to learn about one another and, in particular the interviewer, can use this knowledge going forward.

Researchers must acknowledge their own positionality and reflect upon this in order to provide sufficient transparency and rigor in their work (Austin & Sutton, 2014; Dodgson, 2019). However, reflexivity can often be challenging for researchers despite being recognised as highly important in qualitative research as researcher objectivity is generally not possible (Mitchell, Boettcher-Sheard, Duque, & Lashewicz, 2018). The concept of bracketing has been suggested to allow a researcher to isolate and disregard their own past experiences limiting the influence this has over

data collection or analysis (Dowling & Cooney, 2012). However, Smith and Shinebourne (2012) argue that it is not feasible for a researcher to separate their prior experiences and data interpretation and Ashworth (1995) suggested that the researchers' past experiences and feelings are "intricately interwoven" into the research study. This thesis does not accept the theory of bracketing and instead presented the student's past experiences as a sibling and their personal interpretation of this phenomenon at the start of the thesis in A Sibling's Experience 1 (pages 12-13). The intention of this was to inform the reader of the context within which the researcher approached the project.

Qualitative research provides a contextual understanding of participants' experiences of a phenomenon of interest. Hathaway states that "The meaning of a particular utterance or interaction can be understood and has meaning only within the specific context in which it occurred". The Increasing critique faced by qualitative methodologies has promoted researcher to consider how best to assure the research community of the trustworthiness of qualitative research. Conducting robust and reliable analyses helps to ensure findings are as widely accepted and considered as possible (Attride-Stirling, 2001). Reaching the trustworthiness criteria set forward by Lincoln and Guba (1985), which mirrors that accepted for quantitative research, may be one way in which to do this. Trustworthiness incorporates five components: credibility, transferability, dependability, confirmability, and a clear audit trail that includes reflexivity. The trustworthiness criteria will be further discussed in section 3.3.4 in relation to the qualitative methodology used for this work.

3.3.1. Qualitative methods

There are several forms of analysis or methodologies that can be used in a qualitative research project. Some methodologies commonly used in qualitative research include: case studies, phenomenology, grounded theory, content analysis, and thematic analysis.

Case studies are one of the most common qualitative analysis techniques methodologies. Two key approaches to case study methodologies have been proposed by Stake (1995) and Yin (2003). Both approaches use a constructivist paradigm. Yin (2003) proposed that a case study method should be used when considering questions of "how" and "why". They also suggest that case study methods should be used when it is not possible or desirable to directly manipulate study participants. Further case studies allow for consideration of the context of phenomenon and, therefore, can be beneficial in studies where context is important. Prior to conducting a case study analysis, the case, boundaries, type of case study, and any propositions need to be determined. Propositions are comparable to hypotheses in quantitative research, and both Yin (2003) and Stake (1995) argue that they are necessary elements of case study research and can help with the development of a conceptual framework that guides the research (Austin & Sutton, 2014). Yin (2003) categorises case studies into three types: single, holistic, and multiple-case studies. Chapter 8 utilises a multiple-case study design to consider the family level experience and this methodology is further considered in section 8.3.2 (pages 191-192).

Grounded theory (Glaser, Strauss, & Strutzel, 1968) uses an inductive, also commonly referred to as "data driven", approach that believes data should inform the development of theory (Charmaz, 1996). In grounded theory, the interpretation of data, as much as possible, should not be guided by preconceived ideas the researcher holds for this to be achieved it is recommended that the researcher complete the data collection and analysis concurrently, and delay any literature review until the analysis is complete (Charmaz, 1996). The fundamental drive of grounded theory is theory construction and development, which should directions the use of the theory from sampling to the presenting of findings.

Interpretative phenomenological analysis (IPA) is also commonly used in qualitative research. IPA originated from two theoretical concepts, phenomenology and symbolic-interactionism. Phenomenology was formed through an attempt to build a theory of consciousness and symbolic-interactionism suggests it is only possible to access meaning through an interpretative process (Biggerstaff & Thompson, 2008). Conducting IPA requires the researcher to interpret the data in order to establish the meaning that participants express for their experiences, rather than taking the data at face value (Smith, Jarman, & Osborn, 1999).

Thematic Analysis is often regarded as a foundational method for qualitative research. Braun and Clarke (2006) were the first to propose this methodology. Thematic analysis allows complex qualitative data to be reduced into easily understandable themes and conclusions. These themes and conclusions can be used to inform further understanding, research, and potentially policy decisions. Thematic analysis is the methodology used in Chapters 4-7 and, therefore, is considered in greater detail below.

3.3.2. Thematic Analysis

Thematic analysis requires researchers to make a series of decisions prior to conducting the analysis and, occasionally, prior to data collection. First researchers must choose between an inductive, also known as "data-driven" or "bottom-up", or a deductive, also known as "theoretical" or "top-down", approach to the analysis. As an inductive approach is driven by the data, it may result in findings that bear little resemblance to the questions asked of participants (Braun & Clarke, 2006). An inductive approach tends to produce a richer fuller account of the entire data set, relative to a deductive approach. Secondly, researchers must also choose between a latent or semantic approach to theme development. A latent approach requires the analyst to consider underlying motivations to their data, whereas a semantic approach does not go beyond what is written in the data and does not attempt to infer meaning. Thirdly and finally, the epistemological approach taken to the analysis must be chosen. Researchers can choose to approach the data with a realist or constructionist manner. A realist approach assumes that there is one 'true' experience and that what participants say articulates exactly what they mean and reflects the 'true' experience of an event. Constructionists approach the data with the belief that experiences and meaning are a socially developed phenomenon and therefore that

there is not one 'true' experience. In constructionism what participants say may not necessarily reflect their 'true' experience.

Despite thematic analysis being a widely used qualitative analysis technique, previous work that used the analysis technique provide only vague explanations of their methods, regularly glazing over how they went about conducting their analysis. Braun and Clarke (2006) attempted to explain how they believed thematic analysis should be conducted to help researchers improve their future analyses and outputs. They put forward six steps for conducting thematic analysis. The six steps are (1) familiarisation with the data, (2) coding, (3) searching for themes, (4) reviewing themes, (5) defining and naming themes, (6) creating a narrative analytical account. The methods put forward by Braun and Clarke (2006) have been readily accepted and utilised by the field.

Further approaches to thematic analysis have been suggested, following the foundational work of Braun and Clarke (2006). Castleberry and Nolen (2018) proposed a five-stage process for thematic analysis, which follows the format of compiling, disassembling, reassembling, interpreting, and concluding. Despite attempts to demarcate thematic analysis by Braun and Clarke (2006) and Castleberry and Nolen (2018), there is a continued lack of clarity as to how best to conduct thematic analysis (Braun & Clarke, 2014). Each proposed process of thematic analysis shares similarities and all recognise that thematic analysis is not a linear process despite step-wise recommendations. Rather thematic analysis is an iterative reflective process that is likely to require researchers to revisit several of the steps at various stages in the research process. A brief overview of the stages of thematic analysis according to Braun and Clarke (2006) and Castleberry and Nolen (2018) will now be given.

Familiarisation with the data/compiling: Familiarisation with your data is a key component in both Braun and Clarke (2006) and Castleberry and Nolen (2018)'s approaches. "Immersing" yourself in the data when conducting any qualitative analysis is vital to ensure the researcher has a good understanding of their data prior to completing the analysis. To become familiar with the data it is recommended researchers transcribe the data themselves, read the data repeatedly, and actively engage with the data.

Coding/disassembling: Once the researcher is familiar with the data they can begin the process of coding. Coding data involves selecting pieces of text from within the data and placing them in meaningful groups (Tuckett, 2005). Codes should be as basic as possible, and leave room to be complied into more complex groupings, known as themes, further into the analysis process. There is no clear point at which researchers should stop coding. One recommendation is when no further codes can be developed when investigating a new piece of data, the researcher can be confident in their analysis (Braun & Clarke, 2006). How coding is conducted is largely related to the approach the researcher has chosen towards their data as previously discussed.

Searching for themes/reassembling: Once all the data has been organised into codes, the researcher can begin to develop themes. Themes are broader grouping of data, made up of previously

assembled codes. It is possible for codes to appear in several themes, if relevant. Themes are more likely to be guided by the research questions the researcher is attempting to answer. How to organise your data into themes is subject to personal preferences although there are multiple possible methods. Tools such as thematic maps can be used to establish how themes relate to each other and in the development of sub-themes (groups of codes within themes).

Reviewing themes: Once potential themes and sub-themes have been developed they need to be reviewed and considered. There is no definitive guide as to how much data constitutes a theme or sub-theme, this is a judgement call for researchers. Some themes may be highly related and judged to be more appropriate as one larger theme. Themes need to be considered at two levels, one in relation to the data within themselves, and the other as a component of the whole data set. Thematic maps are recommended by Braun & Clarke (2006) at this point to ensure that all data is captured and that it makes sense.

Defining and naming themes: Following the reviewing themes, the researcher now has a clearly organised set of themes that are appropriate for their data. They now need to provide a clear title and description as to what they are, what they include, and why they are interesting for their data. This may be achieved through defining what the data is and what it is not (inclusion and exclusion criteria).

Creating a narrative analytical account/Interpreting & concluding: When a researcher writes the report of their thematic analysis they provide a story to the reader which connects all the data they have gathered and interprets it in a way that is easy to understand, relevant, and provides evidence of the merit and validity of their work (Braun & Clarke, 2006). Providing examples from the data allows readers to further appreciate how presented themes and sub-themes were developed and relate to the research question given. An analytic narrative is required that goes above a description of the data.

3.3.3. Selection of Qualitative Methodology

Thematic analysis was selected for this qualitative research project as it is a flexible and accessible method that does not require assumptions to be made about the underlying philosophical or theoretical foundations of the data. The simplicity of the method allows those who are new to qualitative research to be able to quickly acquire the level of understanding needed to conduct the analysis, and glean useful insights from large qualitative data sets.

Using thematic analysis allows complex data sets to be summarised in a useful way, and requires the researchers to follow a well-structured and organised approach to analysis. Given the use of well-structured and organised methods it is possible to produce a clear and organised final report of results (King, 2004). This was beneficial to both the student and the third-sector funders of the research project. Other techniques, such as IPA could have been used to reach the objectives of both the research project and the third-sector funder. However, the complexity and scale of the sample included in the research project, and the in-depth nature of IPA would have made the research project

unmanageable. IPA is typically completed on smaller sample sizes to allow a rich interpretation of the individual experiences to be presented (Smith & Shinebourne, 2012).

The research project was completed primarily by the student; however, they were supported by a research team at several points during data collection and the initial analysis. Given the clear structure and outputs from thematic analysis it also supports ease in working with a team of researchers (Braun & Clarke, 2014). However, the flexibility that is stressed as an advantage of thematic analysis can also cause potential inconsistencies between team members (Braun & Clarke, 2006; Holloway & Todres, 2003). Techniques such as peer debriefing (Nowell et al. 2017) were used to negate this disadvantage. Alternatively approaches such as framework approach (Bennett et al., 2018) could have been used, as they allow even greater structure in the analysis, with little space for variability and individual interpretations. However, limiting the level of qualitative interpretation of the data was undesirable, particularly given the novelty of the subject matter and participant experiences being considered.

3.3.4. Robustness of Thematic Analysis

The reliability and validity of the findings presented from thematic analysis has been put into question (Guest, MacQueen, & Namey, 2012). However, work has been conducted in how to address this potential pitfall, which is incorporated into the methods used in this thesis (Nowell, Norris, White, & Moules, 2017). Nowell et al. (2017) apply the four criteria of trustworthiness, (credibility, transferability, dependability, and confirmability) presented by Lincoln and Guba (1985) to thematic analysis and propose a step-by-step approach to reach trustworthiness. They propose a six-phase method to thematic analysis. Within each phase there are multiple means of establishing trustworthiness; for instance, within the first phase "familiarising yourself with the data" they suggest researchers have prolonged engagement with the data, store the raw data in well-organised archives, and maintain records of their thoughts on potential codes or themes along with field notes and reflexive journals. Clear and consistent documentation is suggested throughout each phase as a means to increase trustworthiness of the analysis. Another means of establishing trustworthiness that is utilised in this work is researcher triangulation. Nowell, Norris et al. (2017) recommend the use of multiple researchers in Phase 2: Generating Initial codes, Phase 3: Searching for themes, Phase 4: Reviewing themes, and Phase 5: Defining and naming themes. Particularly in phase 2 it is known that credibility is enhanced if the data are analysed by multiple researcher (Côté & Turgeon, 2005).

3.3.5. The Research Project

The current study aims to build on the lack of understanding of the lives of people with CF and their families and use this information to build a holistic understanding of the impact on siblings with a brother or sister with CF. By conducting a qualitative investigation of young people with CF (11-24yrs), adults with CF (25yrs+), siblings (11-24yrs) and parents/carers, from across the UK this work aims to address areas of limited knowledge such as the new challenges faced by the aging CF

population, the impact of having a brother or sister with CF, and the experiences of existing support. Once these populations are better understood, more useful and effective support services can be subsequently developed.

3.4. Methods

3.4.1. Research Team Responsibilities and Distribution of Tasks

The candidate was a named co-investigator on the primary grant for this project. Their role within the project was to lead in the development and execution of the project with the support from the research team that included two primary investigators, research assistants, and placement student while acting as the primary point of contact for the funders and supporting clinicians across the UK.

3.4.2. Sample

There are four distinct but highly related groups included in this study. The groups are: young people with CF (aged between 11-24years), adults with CF (aged 25years and above), siblings (aged between 11-24years) of individuals with CF, and parents or guardians of individuals with CF. Where possible full families were recruited, however this was not a necessity. The age range applied to young people with CF and siblings was chosen in accordance with the scope for potential support services from The Cystic Fibrosis Trust, along with current literature regarding the period of adolescence lasting from 10 to 25years of age (Sawyer et al., 2018).

A target sample size of 25 participants in each group was initially set following discussions with an experienced qualitative researcher. Sample size decisions in qualitative research are not informed through a data driven methodology as is usually found in quantitative research. Decisions about qualitative sample sizes should focus on the context and subject they are based within (Boddy, 2016). The theory of data saturation is often used in determining sample sizes in qualitative research (Mason, 2010). Given the subject matter, the analysis approach to be taken, and the breath of potential participants from across the UK it was decided that data saturation was likely to occur around 25 participants in each group. However, it was necessary to continually review data as it was being collected to gauge if saturation was reached prior to recruitment of 25 participants. Saturation is typically stated to have been reached when it is not possible to generate any further themes from the new data being collected.

3.4.3. Multi-Centre and Multi-Method Recruitment

Participants were recruited through multiple centres across the UK, as well as through the Cystic Fibrosis Trust's social media and website. Great Ormond Street Hospital (GOSH) acted as the only site for this study, while multiple other CF clinics across the UK were Participant Identification Centres (PICs). At PICs potential participants were informed of the study and provided with the relevant information sheets (Appendix 3.2). Interested patients were then welcome to either contact the team themselves or give their clinician permission to provide the research team with their contact

information. The research team attended clinic at GOSH themselves, but first approach was still completed by a member of the patient's clinical team. Complications from the potential of cross-infection limited the amount of time patients were able to spend talking to the research team in a communal waiting room, this resulted in a very similar process at GOSH to that done at PICs. The Cystic Fibrosis Trust used two news posts on their website⁹, along with their social media to promote the research project. This proved particularly beneficial in recruiting siblings to the research project. Those who found out about the project through these channels were sent the appropriate information sheets by the research team.

It was required for participation that the individual in the family with CF provided a completed QoL measure, in order for this to be considered in the mixed methods analysis presented in Chapter 9. The option was available for the individual with CF to consent to an interview and QoL measure or just a QoL measure. If the individual chose not to provide a QoL measure their parent or sibling was ineligible to participate in the study.

3.4.4. Interview Schedules and Process

Following a review of the relevant literature, semi-structured interview schedules were formed through a collaborative process. The research team worked with CF clinicians, staff members at the Cystic Fibrosis Trust, and members of the public to develop interview schedules that represented the objectives of the research project while also being sensitive to certain topics that had the potential to cause distress. Engaging with the relevant members of the public, for instance feedback on the parents interview schedule was sought from a parent in attendance with their child at clinic at Great Ormond Street Hospital, was particularly beneficial in ensuring the questions included in the schedules were relevant, appropriate, and sensitive.

While the schedules varied slightly based upon the study group and age of the participant, each included several key topics, allowing a degree of comparability (Knox & Burkard, 2009). Each schedule involved questions on day-to-day impact, mental health, coping strategies, support service and awareness. A question on their experiences of transitioning between child and adult health care services was asked of those aged 16 years and upwards. Siblings were asked about how they viewed the time their parents spent with them and their brother or sister, as well as the genetic aspects of CF (where age appropriate). Parents were asked about coping skills and parenting approaches. As interview schedules were semi-structured they were there to guide the interviewer through the key areas, however the interviewer was open to asking questions not included on the schedule in response to the information provided by the participant (Hill et al., 2005). Each interviewer was required to have training in Good Clinical Practice (GCP), Information Governance and one of several interview training options available from MindEd¹⁰, along with supervision from an experienced researcher prior to

⁹ https://www.cysticfibrosis.org.uk/news/siblings-and-cf

¹⁰ https://www.minded.org.uk/

conducting any interview. Clinical Psychologists were available to the team should there be any concern raised for a participant. All interview schedules can be found in Appendix 3.3.

Following either recruitment from GOSH or identification at one of the PICs located across the UK, the research team contacted the participant to organise a time to conduct the interview. Due to location constraints interviews were typically completed over the phone. While there is concern about the validity of phone interviews relative to face-to-face (Rubin & Rubin, 2011) this has since been disputed (Sturges & Hanrahan, 2004). Phone interviews were thought to be appropriate in this instance particularly as it helped to facilitate recruitment from across the UK.

3.4.5. Demographics and QoL measures

Each participant was asked to provide some basic demographic data followed by a QoL measure. The demographics collected varied based on the group and age of participants. The demographics collected included: age, gender, racial background, marital status, education level, work status and family income. The QoL measure completed depended on the age of the individual and whether or not they had Cystic Fibrosis. All participants with CF completed an age specific Cystic Fibrosis Questionnaire-UK (CFQ-UK). Children and young people without CF under the age of 18 completed the KIDSCREEN-10, and anyone aged 18 or above without CF completed the World Health Organisation Quality of Life — BREF measure (WHOQOL-BREF). The outcomes of the quality of life measures are considered in the mixed-methods analysis presented in Chapter 9. Further details about each measure used are given within the methods section of Chapter 9.

3.4.6. The Analysis Process

Each interview was transcribed verbatim by either a member of the research team or an external service. Each verbatim transcript that was used in this study, whether transcribed externally or internally, was reviewed by a member of the research team against the original audio in order to ensure the highest accuracy as possible. This also helped to further familiarise the researchers with the text prior to analysis.

The qualitative analysis technique chosen for this research was thematic analysis. The approach to thematic analysis taken in this work is guided by both the work of Braun & Clarke (2006) and Castleberry & Nolan (2018). In accordance with Braun and Clarke (2006)'s discussion of the decisions required for qualitative thematic this work uses an inductive approach; this means that the data guided the analysis, rather than the research question guiding the process. This approach was chosen as it is known that an inductive approach provides a richer description of the data, which is particularly beneficial in areas where research is currently lacking. It was also decided that a semantic approach would be taken to the data, in that no attempt was made to look beyond what the participants' responded to the interviews. Finally, a realist (or essentialist) approach was taken to the

data, meaning the accounts put forward by the participants in interviews were taken to be true of their experiences.

Several methods as suggested by Nowell et al. (2017) are used within this work to help increase the reliability and credibility of the results, including peer debriefing of both codes and themes, reflexive writing (note taking throughout process by researchers) and member checking (checking results with the participants themselves). An example of a summary sent to participants for

Summary of the main themes from your interview

You spoke about how having a sister with CF has not affected your school life, although sometimes when she has to take her tablets very early it wakes you up. You also said that your sister's CF hasn't affected your friendships. As you have moved into high school recently, aside from friends from Primary school most of your new friends do not know about your sister's CF.

You said it can be quite hard sometimes talking to friends about your sister's CF, because of the things she has to do at the hospital when she's unwell. A couple of years ago when she was unwell most of the time, it was hard for you to go through that because seeing her unhappy all the time made you feel unhappy. You said you tried to manage by going through the daily basis of your life. You feel you are quite close to your sister, and can sometimes be a bit protective over her.

You mentioned that you find it particularly difficult when she goes to the hospital sometimes, for a check-up or a blood test. You said you sometimes feel like you hear can hear her screams, and one time when she came back she was screaming a lot. This made you feel a little bit worried.

You know that CF affects your sister's lungs, eating and digestive system, and that she has to take tablets to help her puke. You said you learnt these things mainly through asking your mum. You spoke about how you think some of your friends know about CF and take time to learn about it, but most do not really do so. You said you like to talk about your sister's CF with your best friend, and you think it helps a bit to talk to someone about it.

You do not think many people in the UK know about CF as not many people have it, and also would not know about it in their daily life, unlike cancer. You agreed that it is good for

Figure 3.1. Example Summary sent to Participants for Member Checking

the purpose of member checking can be seen in Figure 3.1. All analysis for this work was conducted in QSR International's NVivo 11 qualitative data analysis software.

3.4.7. Ethics

Ethical Approval; This study was reviewed and approved by London Bromley Research Ethics Committee (registration number 17/LO/0348; Appendices 3.4 & 3.5). All participants received verbal and written information about the study, and all patients gave written informed consent before inclusion. Further capacity and capability approvals were received from all sites and participant identification centres (PICs) (Appendix 3.7). An ethical amendment was made to the project following an increase in the funding to allow a sample of parents to also be included in the work (Appendix 3.6).

Informed Consent: Once potential participants had had an appropriate amount of time to familiarise themselves with the information sheets provided to them, they were given the opportunity to ask any questions. Only once the research team was assured that all queries had been answered, written consent was taken. if the participant was below 16years of age parent/guardian consent was required. In the circumstance where the parent/guardian consented on the child's behalf, the child was asked to provide written informed assent. Participants were informed that their participation was voluntary and that they could withdraw at any time, without providing a reason and without their medical care or legal rights being affected. Blank copies of the consent and assent forms can be found in Appendix 3.8.

Ethical and Safety Considerations: There was a possibility that the interviews or questionnaires could cause participants distress. In order to minimise the risk to participants each interview schedule was reviewed by an appropriate member of the public, e.g. a parent of a child with CF reviewed the parent interview schedule prior to it being used. Patient and public involvement like this has been used to increase the appropriateness of interview schedules previously (Brett, Staniszewska, Mockford, Herron-Marx, et al., 2014). The research team involved in this project was made up of several clinically trained researchers, who were well placed to provide support or sign-posting to relevant information if distress occurred.

Patient Confidentiality and Data Security: Identifiable data for each participant was collected during the registration process, including name and date of birth. The research team ensured that all data was handled correctly in accordance with General Data Protection Regulations (GDPR). All electronic files that contained any identifiable data were stored either on a secure university server or in paper study documents stored in a locked cabinet within a locked room, each only accessible to the study team.

Chapter 4: Results - Siblings with a Brother or Sister with Cystic Fibrosis

4.1. Introduction

Chapter 1 acknowledged a need to further develop the understanding of how siblings are impacted and, due to high levels of heterogeneity in quantitative analysis, recommended this be done through methodically rigorous and robust qualitative research. Chapter 2's systematic review of the literature on sibling support reiterated the potential influence the severity and burden of the illness or condition could have on the level of impact the sibling experiences, as first noted in Chapter 1. Given the evidence from Chapters 1 and 2, Chapter 3 introduced a large qualitative research project involving families of individuals with cystic fibrosis (CF), a condition known to have a high treatment burden and mortality rate. This chapter presents the first findings from that qualitative project. While it is important to understand the family context and multiple view points on the sibling experience, emphasis should be given to self-reported experiences (as discussed in Chapter 1) and as such the siblings' first-hand experiences are presented first. Subsequently the experiences of their brothers and sisters will be presented, and finally those of their parents.

4.2. Background

Existing literature on siblings with a brother or sister with CF is limited and the need for further research in this area has been acknowledged (Berge & Patterson, 2004). Much of the literature that has looked at sibling experiences and wellbeing has used either a parent report or a parent-completed proxy measure, rather than directly considering the siblings themselves (Moyson & Roeyers, 2012). Recent research has suggested that younger siblings of a child with CF appear to have psychological wellbeing outcomes comparable to their peers (Havermans et al., 2008) and potentially a higher QoL relative to the norm (Havermans et al., 2011). Given that CF is known to have a high treatment burden and mortality rate it would be expected that there to be a notable impact on these siblings yet it does not appear to be the case. Why outcomes for siblings with a brother or sister with CF are comparable, or potentially better, to the norm has not been clearly investigated.

Given the demands and fluctuations that exist in a life with CF there are many ways in which the sibling's psychological wellbeing may be affected. During periods where their brother or sister is experiencing particularly poor health the sibling is likely to experience a greater level of psychological distress (Bluebond-Langner, 2000). It may also be that during these periods the sibling is able to spend less time with their parent, which may be detrimental to familial relationships (Ma et al., 2017). Parents tend to need to focus more attention on the ill child and the sibling can often feel left out (Foster et al., 2001; Williams et al., 2010). Relationships with their brother or sister play an important role in siblings adjusting to the presence of a chronic illness or condition. A more positive sibling relationship is associated with better adjustment and a greater willingness to support their brother or sister (Bigby, 1998; Greenberg et al., 1999).

Siblings have a further complexity in a life with CF as they have a risk of being a genetic carrier and could potentially pass CF onto their own children (Fanos & Johnson, 1995). The diagnosis of carrier status and the decision to be tested has been considered in a small number of studies, which highlight this as an area that could impact sibling psychological wellbeing (Crozier, Robertson, & Dale, 2015). Another complexity of CF, and a number of other chronic illnesses or conditions, is its invisibility. CF being invisible means that 'others' unaware of their brother's or sister's diagnosis are not able to tell they have CF from simply looking at or interacting with their brother or sister. Invisible illnesses have been associated with negative misconceptions and assumptions being made, which can have a negative effect on mental health of siblings (Chudleigh, Browne, & Radbourne, 2019).

How siblings cope with having a brother or sister with CF has not received a lot of attention in existing literature. There is literature that suggests siblings of children and young people with a chronic illness or condition use cognitive restructuring and normalisation as methods of coping (Malcolm et al., 2014; Ross & Cuskelly, 2006). A good level of knowledge and understanding of their brother's or sister's condition could also positively influence psychological wellbeing, as has previously been found in siblings of children and young people with an eating disorder (Areemit et al., 2010), as was also highlighted in the findings from the systematic review and meta-analysis presented in Chapter 2.

Given the continual improvements in CF diagnosis and treatments the way the family is impacted is changing rapidly. Research is challenged to keep up with the continually changing experiences of families with CF. Notable literature, such as that published by Bluebond-Langner (1991), is rapidly becoming out of date.

4.3. Aims and Objectives

The overall aims and research questions of this project, as stated in section 3.2.10 of Chapter 3 are considered throughout this chapter. However, this chapter focuses on siblings and thus required its own specific aims and research questions. The aim of this chapter is to build a picture of the experiences of siblings with a brother or sister with CF and how they are impacted by the presence of CF in their lives. Consideration is also given to sibling support services, including any previous experiences and opinions on the need for sibling support. To address these aims the following research questions were proposed:

- 1. How do siblings with a brother or sister with CF view the impact of CF on their lives? Specifically, do they identify a direct effect on their own psychological wellbeing?
- 2. Do siblings believe that they are currently being well supported, both informally and formally? If not, are there areas of their lives or a specific type of support which they believe could be beneficial?

4.4. Recruitment of Siblings

In total 19 siblings were recruited. The majority of the siblings were recruited through online advertisements (n=9), while the remaining were recruited from hospitals in London (n=5) and Cardiff (n=6). Around 75% of siblings that provided consent to be contacted were reached. Of those reached 23 consented to participate in the study. It was not possible to complete the study with four siblings who agreed to participate. This results in a participation rate of roughly 61%. Of the 19 siblings included in the study, nine had a brother or sister included in the young person interviews, none had a brother or sister included in the adult interviews, and nine had a parent included in the parent interviews; 10 had a brother or sister with CF only complete a QoL measure. Siblings proved to be particularly challenging to recruit for this research project, and unfortunately is was not possible to reach to the initial sample size goal of 25. Multiple potential barriers to recruitment exist that could explain this issue. Much of the contact information received initially for siblings was their parents, particularly while recruitment was being completed only in clinics. It quickly became clear that siblings did not regularly attend clinics with their brother or sister, whether they were a young person or an adult with CF. It was also the case that the clinical team was not always aware if the individual with CF had a sibling, and if they were aware they may not have been certain about the age of the sibling, which reduced the likelihood of them approaching about the sibling aspect of the study. As recruitment was slow in an attempt to increase numbers advertisements were placed on the Cystic Fibrosis Trust website and social media platforms.

Due to the preventative measures that have to be put in place at CF clinics due to cross-infection risks, it was not possible for the research team to speak to families while waiting for appointments. If the family wished to speak to the research team after receiving information about the study it had to be in their clinical room following their appointment, which could last for several hours.

4.5. Data Collection and Analysis

Data collection and inputting was coordinated by the candidate (MMS), with the support of other researchers (CL, NK, LA, JL and RS). Informed consent and, where appropriate, assent were collected through confidential email or post, unless the interview was completed in person in which case consent was collected at time of interview. Demographics and QoL measures were completed over the phone or in person prior to the interview, where this was not possible due to factors such as understanding or age, they were sent in the post with a return envelope. Seventeen of the interviews with siblings were conducted over the phone, with the remaining two completed in person. The sibling interviews were conducted in majority by one researcher (MMS), who conducted 18 of the interviews, the one remaining interview was conducted by NK. Ensuring interviewer consistency where possible was important in order to minimise interviewer variability, which has been widely discussed as having

the potential to alter participant responses (Biemer & Lyberg, 2003). Reducing interviewer variability does not alleviate other interviewer effects including interviewer experience and attitudes, which have been found to change over the course of a study (Olson & Peytchev, 2007).

Interviews with siblings lasted on average 30 minutes. The longest interview lasted 61 minutes, and the shortest interview was 12 minutes. Four sibling interviews were transcribed verbatim by MMS but due to time constraints the remaining 15 sibling interviews were sent to an external transcription service. MMS further familiarised herself with the transcripts by reading through each at least once while listening to the audio, prior to conducting the preliminary analysis of the sibling interviews. Codes developed by MMS were cross-checked by two other researchers using a code-book method, as also used by Ulph, Leong, Glazebrook, and Townsend (2010). Themes were established through an iterative process. MMS maintained a theme development document that detailed the thought process behind the development of themes from the codes. These methods helped to ensure that the analysis conducted was as robust as possible. Robustness was also enhanced through respondent validation. Following the initial code and theme development a short summary of the results was compiled for each participant to review and sent to them via confidential email or in the post as requested. Participants were asked to reply with comments should they feel that the themes were not representative of their experiences. Only one sibling returned the summary with comments, which were incorporated into the analysis. Therefore, it is assumed that the themes presented below met with the sibling's expectations and true experiences.

4.6. Participant Demographics

Sibling ages ranged from 11 to 24 years (median = 16). 10 had a sister with CF, while nine had a brother with CF. Twelve (63%) were the younger sibling. All included siblings were white and the majority were from high income families (57% of those with available information). Six siblings had a parent included in the study while three had two parents included in the study. Demographics were collated across families where possible and can be seen in Table 4.1 below. As it was not a requirement for full families to participate there are multiple points of missing data, which are shown by blanks in Table 4.2.

Sibling	Age	Sex	Marital Status	Employment Status	Household Income	Brother/Sister	Parent 1	Parent 2
S1	11	М		Student	35,000-50,000	Y11	P14	
S2	11	F		Student		Y12	P18	
S3	12	F		Student	15,000-25,000	Y14	P1	
S4	13	F		Student	50,000+	Y21	P13	P11
S5	13	F		Student	35,000-50,000	Y3	P22	P21
S6	23	F	Single	Employed Full/Part Time		A9		
S7	13	М		Student		Y4		
S8	14	F	Single	Student		Y7	P8	
S9	16	М	Single	Student	50,000+	Y9	P23	P24
S10	15	М	Single	Student	50,000+	Y2	P10	
S11	14	М	Single	Student	50,000+	Y27	P25	
S12	18	F	Single	Student		Y28		
S13	20	F	Single	Student		Y22		
S14	21	F	Single	Student		Y30		
S15	22	М	Single	Employed Full/Part Time		Y31		
S16	22	F	Single	Student		Y23		
S17	22	F	Single	Employed Full/Part Time		A1		
S18	22	F	Single	Employed Full/Part Time		A10		
S19	24	F	Living with Partner	Employed Full/Part Time		A7		

Table 4.1. Sibling Demographics

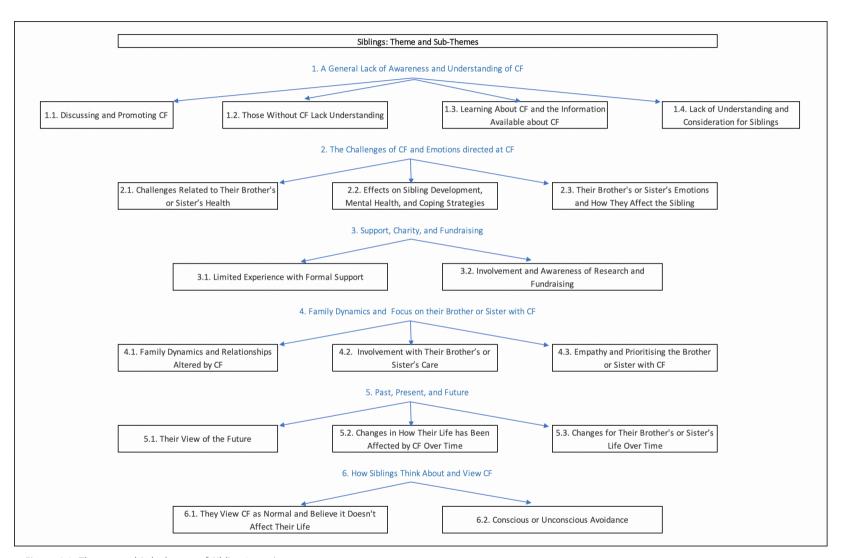


Figure 4.1. Themes and Sub-themes of Sibling Interviews

4.7. Themes

From the qualitative thematic analysis conducted on the 19 sibling interview transcripts six themes were formed:

- 1. A General Lack of Awareness and Understanding of CF
- 2. The Challenges of CF and Emotions Directed at CF
- 3. Support, Charity, and Fundraising
- 4. Family Dynamics and Focus on their Brother or Sister with CF
- 5. Past, Present, and Future
- 6. How Siblings Think About and View CF

Each theme contains multiple sub-themes. The themes and sub-themes are given in rank order of reference density and is displayed in Figure 4.1. Each theme is discussed below with verbatim examples taken from transcripts.

4.7.1. Theme 1: A General Lack of Awareness and Understanding of CF

In all sibling interviews a distinct lack of awareness and understanding for CF was identified. Four sub-themes were identified within this theme. The sub-themes identified were:

- 1. Discussing and Promoting CF
- 2. Those Without CF Lack Understanding
- 3. Learning about CF and the Information Available about CF
- 4. Lack of Understanding and Consideration for Siblings

4.7.1.1. Discussing and Promoting CF

Most of the siblings included in this sample were comfortable with CF and open to discussing it with friends, family, and school or work colleagues. There were some siblings who did not wish to discuss CF at all, even when being offered support.

"I sort of did try to like talk to like a psychologist about it. But then I just, I just don't really like talking about it so it would be like work" (S7)

Siblings that were open to talking about CF each had a different approach to explaining CF, with the majority focusing on a simple explanation of the symptoms associated with CF. Some siblings felt that although they could give a basic description of CF there were intricacies that they were unable explain well. Many siblings that were open to discussing CF were also aware that it is a very personal subject for their brother or sister, which meant they were hesitant to discuss it in too much detail without their brother's or sister's permission first.

"It's often easier for [Brother] to talk about than for me to talk about... because you know it's his own body, his own condition, his experiences... and so he can kind of control the conversation a bit

more in that way whereas for me it's kind of a combination of okay well this is my brother's medical information that I'm sharing, so kind of trying to be wary of how much I share because it's, you know it's not mine" (S16)

When asked if they felt there was a good awareness of CF siblings generally believed that awareness was poor, unless an individual was directly affected by CF. Many of the siblings saw a benefit to increasing awareness of CF in the general public. Suggestions sibling gave for how to raise awareness usually involved a reference to campaigns from other charities that the sibling thought were effective and successful.

"I guess sort of things like social media campaigns or TV campaigns and things like that where, you know, you see or you hear adverts for Cancer Research every five minutes on the radio but you don't hear things like that for CF" (S6)

4.7.1.2. Those Without CF Lack Understanding

Siblings recognised their brother or sister with CF as the expert in their condition. Siblings typically suggested that those without CF, including themselves, lacked a good understanding of all aspects of a life with CF.

"I don't really know about the mechanisms, but I know like, I do like know the basics and stuff but like I don't know into depth about a lot of it" (S16)

Siblings identified friends, partners, 'others', and schools as also potentially lacking a good understanding of CF. The siblings' experiences with understanding from their school varied and some had experienced difficulties due to a lack of understanding. Their own level of understanding varied from a shallower understanding, such as that shown in the quote from S16 above, to those with a detailed knowledge of their sibling's medical care, as demonstrated by S3 in the quote below.

"she was like changing medicines from like Colomycin[©] or Tobi[©], or like when she got the I-neb, she went from Colomycin[©], then Tobi[©] to Promixin[©]...I know what the medicines do and like how they... how it helps her" (S3)

Some siblings believed that their understanding of CF had improved over time, and that when they were younger their lack of understanding could have potentially been a source of tension and conflict with their brother or sister. For instance, being unable to understand the differences in the way they were treated caused tension for some siblings.

"when I was very young, then I wasn't massively aware of what it actually meant and so obviously like it, it kind of, all it really meant to me was that she got to eat extra chocolate and sweets cause she needed calories I didn't" (S6)

Several of the siblings highlighted barriers they saw to increasing understanding. For instance, a number of the siblings believed the invisibility of CF was an issue for understanding. May of the siblings thought of CF as a highly individual condition and believed that each person with CF had a different experience. This was also highlighted as a difficult for understanding. Barriers to understanding allowed "others" to make assumptions about CF and how it affected individuals, including some common misconceptions about CF.

"people must look at it and think they... not a lot of people know that it is not contagious. So, if you say you've got cystic fibrosis, people may be like oh stay away, I don't want to get it but when really, they can't get it, because it's a part of your genes, it's unless they like... they've got the cystic fibrosis carrier gene, then they have nothing to do with it. They just make assumptions to like say 'oh it's a like a harmful disease'" (S3)

4.7.1.3. Learning about CF and the Information Available about CF

Siblings increased their understanding of CF in various ways. Frequently siblings were not able to directly identify how they learnt about CF, rather they felt they had gained their understanding through living with their brother or sister.

"it is mostly just from living around somebody with CF that I know about it. Because you learn about all the different things used to treat it and all of that" (S9)

One method that was mentioned by several siblings was learning from their families. If the siblings had questions about CF many would approach either their parents or their brother or sister to get a better understanding. Several siblings in this study mentioned approaching their brother or sister with CF directly with their questions.

"I think as I've got older I've got a lot better at actually speaking to my brother about it... like we will chat about it now and again, but he makes light of it when sometimes I want a serious conversation which is annoying" (S14)

Siblings learnt about CF at school in some capacity. For several of the siblings CF was included on their school curriculum. While some had fond experiences of learning about CF at school, several did not enjoy the experience.

"it's the only time I've ever walked out of a class actually... I did go and apologise to her afterwards actually and she felt very guilty too, but it was a new science teacher so she didn't know my brother and she didn't know me [...] but she was basically just kind of talking about how awful it was and how you, you know, you kind of wouldn't wish it on anyone sort of thing, and I just remember thinking 'no, I can't deal with this right now'" (\$16)

If at this point siblings felt they still did not have a sufficient understanding, they may search for online resources. Opinions about the information available online differed between siblings. Some siblings disliked the information available online as they believed it only highlighted negative points about CF, which they didn't wish to focus on.

"as I was a teenager I would like google things, you know when I got access to the computer and stuff, I would just google it. But then it scared me so I didn't like looking at it" (S19)

Other siblings believed the information available online could be a helpful resource, not only for themselves but for anyone looking to learn about CF. Although many siblings further stipulated that the information needed to be from a reputable source such as the Cystic Fibrosis Trust website.

"there's a lot of very good information out there. I think, you know always [be] wary of things like Wikipedia, but you know the CF Trust and other websites have very good information, that kind of gives enough that you can get an understanding, but without being kind of too bogged down in too much medical jargon" (S16)

Siblings did not appear to gain much knowledge about CF from their brother's or sister's care team. Only be a small number of siblings were addressed directly by their brother's or sister's care team and offered information about CF. Some siblings sat in on their brother's or sister's appointments, although many made the conscious decision to not want to sit in on appointments.

"The hospital they came to my house... We had like a little like thing where they would like tell me what it is about and stuff and they would answer all my questions" (S8)

4.7.1.4. Lack of Understanding and Consideration for Siblings

Relatively few siblings highlighted an absence of information and knowledge of the impact on siblings themselves. The siblings that did were clear that they felt there was a lack of consideration for siblings.

"I think I did always feel left out, as a sibling, that's sort of something that was kind of on my mind... you know when CF's being written about, like CF affects parents and children, and I'd be like "what about the sibling?"" (S17)

Several of the siblings suggested that it may be the case that they themselves did not truly understand the impact that CF had had on their own lives. Although as they got older it appeared that siblings were better able to acknowledge and process the impact CF had on their life as was the case for S18.

"I think when I was younger I didn't kind of understand like the true extent of it, like I never really thought about it and I guess..." (\$18)

4.7.2. Theme 2: The Challenges of CF and Emotions directed at CF

CF comes with several challenges including both everyday difficulties, such as medications and treatments, and larger less frequent complications such as pulmonary exacerbation (an increase in pulmonary symptoms (Goss & Burns, 2007)). These challenges affect siblings as well as those with CF. Three sub-themes were identified within this theme. They were:

- 1. Challenges Related to Their Brother's or Sister's Health
- 2. Effects on Sibling Development, Mental Health, and Coping Strategies
- 3. Their Brother's or Sister's Emotions and How They Affect the Sibling

4.7.2.1. Challenges Related to Their Brother's or Sister's Health

One subject that almost all siblings brought up as a challenge for themselves was when their brother or sister was going through a period of poor health, particularly when they were in hospital. During their brother's or sister's hospital stay siblings talked about how they missed and worried about them.

"I think it could very easily kind of consume you, particularly when you know he's especially unwell or when he's in hospital, obviously it, you know, it's easy to think about it a lot because it's kind of more acute and I think that, you know that then impacts all kinds of other areas of your life"

(S16)

Another challenge faced by siblings, particularly when their brother or sister was in hospital, is the distribution of their parent's attention and time. It was frequently the case that the sibling had to stay with other family members while their parents were having to attend to their brother or sister. However, while some siblings did have negative emotions and memories about their time staying with other family, most did not view it as an issue.

"when I was younger he'd go into hospital for long periods, I would then stay at my auntie's and I'd say impacted me ... quite a bit then, but I'd still enjoy myself cause I'm with the family, it was a bit more fun" (S12)

Siblings highlighted their brother's or sister's worsening health as a particular challenge to them, their daily lives, and their emotions. Their brother's or sister's deteriorating health meant that the sibling's brother or sister was faced with further restrictions on their lives. Their brother or sister may also have to face more serious medical situations and decisions, such as those regarding lung transplants. One sibling in particular highlighted how this had been a very challenging period for them.

"you sometimes don't want to speak about, too much about ... particularly with the transplant, how like big an operation it is. You kind of like struggle sometimes to kind of ... discuss the kind of ... the possible outcomes of it" (S18)

Their brother or sister being unwell affected the emotions of many of the siblings and in turn this had an effect on their day-to-day lives. For instance, the inability to avoid thoughts of CF made it difficult for some siblings to concentrate on school or work.

"He was hospitalised a few months ago and he, when he was quite poorly and, I was finding it quite tricky to you know concentrate on my work at uni because I was worried about him and thinking about him" (S16)

CF is regularly regarded as having a high treatment burden, which has been related back to the level of impact siblings experience. When asked about the impact of daily treatments siblings appeared to not see the daily treatments as a large inconvenience in their lives. This was particularly true for siblings close in age or younger than the brother or sister with CF, as is shown in the below quote.

"She's very independent with CF... if she has any issues [they] will be solved between her and my dad. So, he helps her out a lot, but like in terms of like getting her medicine and things, but when she, actually like her taking everything and doing all her like nebuliser and everything is basically completely her. She's able to be sort of very independent and stuff" (S15)

4.7.2.2. Effects on Their Development, Mental Health, and Coping Strategies

When asked about how they viewed themselves relative to their peers, the majority of siblings did not view themselves as any different to their contemporaries. However, some siblings mentioned a few slight differences they perceived between themselves and their peers. A few siblings thought they had a higher level of maturity, potentially due to an increase in the amount of responsibility and independence they had from a younger age.

"I know I'm more mature than a lot of people my age. And that could just be down to personality and things, but that, aspect of my life probably does play a role in that" (S11)

Others suggested that they have less sympathy for others, for instance when a friend or someone outside of their family was suffering from a mild cold they were less likely to be inclined to provide comfort.

"If someone's telling you something really minor and if [brother] were to be in hospital then I think I would be a bit, I wouldn't ever be rude to someone, but in my head, I'd be a bit like, oh, you, like that's nothing, sort of thing, even though it's clearly something to them..." (S12)

Siblings appeared to believe that their mental health was not affected by their brother's or sister's CF. However, when asked further prompting questions some siblings were able to begin to identify negative feelings and emotions they had in relation to certain aspect of their brother's or

sister's CF. Many readily exhibited feelings of worry, concern and fear for their brother or sister. When the sibling mentioned feelings of low mood or depression this was typically directly related to their brother's or sister's health. For many of the siblings focussing their thoughts on their brother's or sister's diagnosis and the life-shortening implications of it cause a large amount of distress. This was particularly apparent in siblings older in age, as was the case for 24 years old S19.

"a bit of confusion mainly, and a lot of fear. That is something I remember hugely, and it still fills me with fear now. Mainly fear, yeah. Yeah, mainly fear is a word I associate with it..." (S19)

One coping technique that was presented by many of the siblings was avoidance. In particular when thinking about life expectancy and complications their brother or sister were likely to face, several siblings suggested that the best method to deal with these emotions was by avoiding them. However, a smaller number of siblings recognised that ignoring their emotions may not be the best technique in the long-term.

"I think burying and kind of sticking your head in the sand about it isn't going to help anybody either because I think that means you know when the time does come and when he, you know, when he is particularly poorly or when he does die, I think you know then I'll be, I'd be, I'm prepared for it and it wouldn't be too much of a shock. I think kind of finding that middle ground of acknowledging it and discussing it and being aware of it, but not letting it rule our lives..." (S16)

4.7.2.3. Their Brother's or Sister's Emotions and How They Affect Them

The mental health of their brother or sister with CF appeared to greatly influence the sibling. For some of the siblings it was especially difficult when their brother or sister had to do something for their CF that they did not find enjoyable, such as injections. Low mood or upset in their brother or sister was challenging for the sibling.

"It is quite difficult in that respect just because I know how upset he is. But he hides it well. He just breaks down every so often obviously. But otherwise, he is so strong you wouldn't know anything was wrong." (S19)

It was also difficult for siblings when their brother or sister had to spend time in hospital. Concern for their brother or sister during this time led several of the sibling to become upset and many tried to ease the burden on both their brother or sister and their parents when they could.

"...being on your own in hospital is quite lonely and I think seeing him a bit down was a bit like, oh, a bit gloomy, but I think trying to perk everyone up and make it a bit of a better situation" (S12)

Some siblings felt a continual need to take on the responsibility to keep their brother's or sister's moods high. Especially as several of the siblings believed that it was important for their brother

or sister to maintain a positive outlook as the sibling believed this was connected to their brother's or sister's health.

4.7.3. Theme 3: Support, Charity, and Fundraising

Within a family with CF siblings can frequently be left out when it comes to support, as the focus is typically on those with CF and their parents. Two sub-themes were identified within this theme. They were:

- 1. Limited Experiences with Formal Support
- 2. Involvement and Awareness of Research and Fundraising

4.7.3.1. Limited Experiences with Formal Support

The majority of siblings had received very little formal support. Siblings were generally unaware of where they could reach out to in order to be supported. However, several siblings had received some informal support from friends and family. Although some families were not overly open about CF, the majority of siblings knew that should they want to talk to someone they could always approach their parents.

"They've [parents] always you know said you can come and talk to us, about anything if you want to and we're always here for you" (S16)

A small number of the siblings were fortunate enough to receive some support from their brother's or sister's care team, although this was rare and typically only happened if they went to the hospital with their brother or sister. One sibling was offered support by way of a member of the clinical team coming to their home to provide information about CF and increase their understanding of what their brother or sister was going through.

"The hospital they came to my house and they... like... We had like a little like thing where they would like tell me what it is about and stuff and they would answer all my questions" (S8)

In terms of more formal support, this tended to have to be sought out by the sibling or their parents and the support that was found did not always meet the expectations of the sibling and siblings, therefore, chose to not engage with the support for extended periods of time.

"I used to go to like a young carers group for a bit so you know people, most people who are from similar, have similar things going on ... we'd all meet up and go out and do stuff. I did that for a bit. I just, I don't know, I just found it uncomfortable, I just didn't really want to do it, so I didn't last very long there" (\$14)

Another issue that siblings found with formal support was that it did not directly relate to their brother's or sister's CF and this limited the amount those offering the support were able to understand, which the siblings felt reduced the amount that they could get out of the service.

"it was probably because of not being specifically for siblings with CF, that you know, counselling at school, where the person, the counsellor probably might not know what CF is, or something like that you, you know, it's not related to it, then you don't feel that kind of connection where you think oh they probably don't understand or something like that, yeah." (S17)

Given the limited knowledge, experiences, engagement and perceived benefits from currently available support it could be beneficial to promote and develop relevant and appropriate services for siblings with a brother or sister with CF. However, when siblings were asked if they felt they needed support several believed they did not need any additional support. A number of siblings suggested a general need for support in families with CF.

"support for families. There are some, you know other illnesses and other, you know trauma and stuff there are support services available. I'm not really aware of any specific to CF, so I really think that would be something really awesome if that could happen" (S6)

Despite a lack of previous engagement siblings were willing to consider using support services if they were offered them. One service many of the siblings thought could be beneficial was the opportunity to talk to other siblings of individuals with CF.

"as a sibling I think, yeah, it would be quite nice to actually speak to other people. Or even being the help. You know if someone was really struggling and needed to talk to someone else. I think it would be really beneficial and helpful." (\$13)

Opinions on how helpful talking to other siblings would be were generally positive, with the majority of siblings stating it would be good to talk to someone that 'understands'. When asked how they would like to go about talking to others, several of the siblings presented a preference for face-to-face contact rather than through any other method.

"I think maybe kind of for a face to face thing, so like if there's anyone kind of you know, locally, sort of thing. I think that's quite a good way of doing it. Social media can be good as well, but like I quess it's just personal preference. Yeah, I prefer kind of meeting people face to face really." (S17)

Further support that a few siblings suggested could be beneficial for them was help in knowing how to explain their circumstances to others, particularly if their brother or sister was in poor health. One sibling spoke about a "template" their parents gave their brother that he was able to use in order to explain CF to others, and how they felt this may also be helpful for siblings.

"a template of something that you could say to people, something that you could say to teachers if you were having a particularly difficult day, if your sibling was in the hospital or something you could say to friends, if there was some kind of template to help you word that when you were younger because I think that's quite difficult when you don't really know, err, how to explain things and how to explain how you're feeling, err and you know something that you could download that gives you a few templates for those kind of things would probably have been really helpful." (S16)

As the siblings got older there was an increasing concern for certain CF related topics, such as the genetic inheritance of CF. Although several of the siblings were screened for carrier status when they were young, there was a small number that still had to decide when and if to be tested. This issue was particularly focused around considering to have children of their own.

"when it comes to the stage in my life where I think I want to have children I'll definitely need some support and guidance there if whoever I am with has, is carrier or not, I think I'd need to know all the implications of being a carrier and things like that cause to me I don't think it's worth risking to have a child with CF" (S12)

Contrary to what was expected from the current literature, (Burke et al., 2015), no sibling mentioned a concern about having to care for their brother or sister at a later time point. This may relate to the young age of the included sample, the increasing independence of those with CF, or the increasing life expectancy of those with CF. It is now more likely that those with CF will get married and caring responsibilities could fall to their partner rather than siblings.

4.7.3.2. Involvement and Awareness of Research and Fundraising

Numerous siblings spoke about participating in fundraising activities, typically for the Cystic Fibrosis Trust. Parents were usually the driving force behind fundraising, although there were a couple of siblings who pushed the subject matter themselves. Participating as a family appeared to make fundraising an enjoyable and positive activity for some of the siblings.

"we've done quite a lot of like sponsored like, walks and cycles and then we done like, cake sales and stuff like that...Well, my whole family do it...we did do it quite a lot at school" (S7)

Several of the siblings identified an interest in research, with a small subset actively trying to be involved in studies themselves or encouraging their brother or sister to take part.

"I don't know what everyone else is like but I know we're always interested in taking part in stuff like that [research]...I mean if you can do it, like, why not?" (\$13)

Older siblings were generally more aware of current research. Younger siblings tended to have a simpler understanding and hold the view that research should be focused on, and able to produce,

a cure for CF. Although, hope for a cure was not exclusive to the younger siblings as demonstrated in the quote from 23year old S6 below.

"I think the most important thing for them to do with their money is to research and find a cure, hopefully, yeah, without a doubt I would say that's the most important thing" (S6)

At the other extreme, there were siblings that chose to avoid everything to do with fundraising and research and namely the Cystic Fibrosis Trust.

"I actively avoided it [the Cystic Fibrosis Trust] for a long time because, err, I just didn't want to think about it. I didn't, you know I didn't want to engage with CF any more than I had to..." (\$16)

4.7.4. Theme 4: Family Dynamics and Focus on their Brother or Sister with CF

The relationships between siblings, their parents and their brother or sister with CF may be affected due to the presence of CF. Four sub-themes were identified within this theme. They were:

- 1. Family Dynamics and Relationships Altered by CF
- 2. Involvement with Their Brother's or Sister's Care
- 3. Empathy and Prioritising the Brother or Sister with CF
- 4.7.4.1. Family Dynamics and Relationships Altered by CF

Several of the siblings mentioned they felt as though their relationship with their brother or sister was like any other typical sibling relationship. This included both positives and frustrations that are common between brothers and sisters as they grow up.

"well obviously we probably do like just general brother and sister, obviously argue a bit and stuff, but I think we've maybe grown a bit closer as we've grown up just cause we're probably both maturing a bit, so there's you know I would say, I wouldn't say, well we're not, we're close enough as siblings" (S15)

Siblings who thought they were closer to their brother or sister than other siblings may be suggested that this could be due to the amount of time they spent with their brother or sister. Many siblings spent time with their brother or sister while they completed their physiotherapy.

"I get to spend like, quite a lot of time with her when she does like, her exercising and stuff like that, so that, that brings us a bit closer" (S5)

Parents regularly have to spend time away from the sibling, particularly when their child with CF has to spend periods of time in hospital. Time away from the sibling has the potential to alter their relationship (Ma et al., 2017). Despite this, when asked about this the siblings included in this study generally reported that their parents adjusted their schedules and routines to limit the disruption on them.

"I think they always tried to do it so my mum would go during the day and then my dad would go in the evening so I'd always kind of get... and, oh, at the weekend they'd alter, like alternate. So, I'd always get to see like at least one of them..." (S18)

Siblings mentioned that they felt a high level of concern for their parents and found it difficult when they saw their parents upset. Some siblings went out of their way to try and not make things more difficult for their parents.

"you know obviously of course we're all going to be worried, so I don't feel the need to kind of add to that concern by letting them know that I'm having a particularly rough time. I'd rather deal with that with you know a few friends that I trust or you know something like that rather than necessarily going to them all the time" (\$16)

4.7.4.2. Involvement with Brother's or Sister's care

There continues to be improvements in the treatments, medicines, and earlier diagnoses for CF. This has helped increase the average life expectancy for those with CF. As siblings and their brother or sister with CF get older it could be expected that the sibling may take on a greater role in their care. This did not seem to be the case for the siblings included in this sample. While the sibling is young it appeared that their involvement in their brother's or sister's care was limited. Siblings typically did not attend hospital appointments unless their parents were unable to organise alternative care for them.

"not really, no, we went for a few if mum couldn't find a babysitter or whatever, but usually it was just mum and [Sister] that went" (S6)

Two ways that siblings were involved with their brother's or sister's care were participating in exercise with their brother or sister and visiting them when they were in hospital. Siblings felt that by being there for their brother or sister while they spent time as an inpatient they could help their brother or sister through an isolating and difficult period.

"Cause she was unwell and she had to stay in hospital for like two weeks. So I came to see her, like every day after school. And normally she would just be watching, movies on a DVD player... Yeah [watched movie with sister], like one or two" (S1)

Some siblings, however, viewed visiting their brother or sister in hospital as a requirement rather than a voluntary support they were offering.

"...I don't think it was ever I wanted to go see her, I think it was more I was taken along with..."

(S11)

A few of the older siblings chose to support their brother or sister to a greater extent, including going with them to appointments. When siblings were involved with their brother's or sister's care they tended to try to keep it light hearted and fun

"... I think it was last year I, he had to go and have some sort of scan or lung function general check-up thing and mum wasn't available to go with him and I had the day off college anyway, so I went up to [location] with him and we took the train up and all of that and then took the train home and everything and that was, we kind of made a bit of a day out of it and made it a bit more different to like a normal trip up to the hospital" (\$12)

The siblings also understood the importance of exercise for their brother's or sister's health and viewed this as an area they were able to support. Siblings also noted that their being active with their brother or sister may potentially be of benefit for themselves also.

"Cause like it sort of keeps me in to shape at the same time. Cause like with my sister and stuff she has to have a lot of exercise. So, like I will do it all with her and stuff. Like we will stay behind school and go to the fitness suite and stuff" (S8)

4.7.4.3. Trying to Empathise and put their Brother or Sister with CF First

Many of the siblings spoke about trying to always put their brother or sister with CF first, putting aside not only their own wants but also expecting other people within the family to prioritise their brother or sister. It was clear that siblings thought about their brother or sister with CF a lot, whether they felt they had a close relationship with them or not. Several of the siblings expressed a large amount of sympathy for their brother or sister. For some this altered how they allowed themselves to feel about their own wellbeing.

"If she's going through all of that, what reason do I have to complain with what I'm going through? I feel like that's kind of how I feel. But I still do complain a lot of the time! [But] It is a reminder of I do not have the worst scenario" (S11)

Putting their brother or sister with CF first appeared to come automatically to several of the siblings. This compassion translated into a number of the siblings being very protective of their brother or sister, as is shown in the quote below.

"Definitely, like, I'm like quite protective over him and obviously I want him to make sure that he's taking his medicine and he's alright, and you know he's like good doing his breathing exercises and using that big machine that he's got and everything and, you know, if he does have any issues or he's like scared about something he can like, come and talk to me about it." (\$13)

4.7.5. Theme 5: Past, Present, and Future

Several of the siblings involved in the study have seen their brother or sister grow up with CF and how they have changed over time. The question of future with CF was previously associated with negative connotations but this is becoming less of a problem. The increasing life expectancy of those with CF could have long-term implications for the sibling. Three sub-themes were identified in this area. They were:

- 1. Their View of the Future
- 2. Changes in How Their Life has been Affected by CF Over Time
- 3. Changes for Their Brother's or Sister's Life Over Time

4.7.5.1. Their View of the Future

When asked about the future the majority of siblings focused on the mortality of their brother or sister and the role that CF plays within this. Each sibling had a slightly different approach in dealing with the knowledge of the shorter life expectancy of those with CF. Most siblings tried to limit the amount this changed the way they treated their brother or sister.

"I found it quite difficult trying to navigate knowing that we, he wasn't going to be around forever, so wanting to kind of make the most of time with him but also not wanting our relationship to be based on kind of me wrapping him in cotton wool or me choosing to, you know if he was being annoying choosing to let it go because he's going to die" (\$16)

While others chose to remain optimistic and focused on the increasing life expectancy and how this may allow their brother or sister to live a "normal" life.

"...there's all these statistics ...if you look up life span for someone with cystic fibrosis, like people say it's 28, like that's the average ...like... how long they live to. But there's like people who have lived like past 28 and had children, like living normal life even though they have cystic fibrosis" (S3)

What was true for all siblings, however they chose to approach it, was that the discussion of life expectancy caused them distress and upset. This made it particularly challenging for siblings who had the subject presented to them in a school class, such as was the case for S14 quoted below.

"then they'd talk about life expectancy and things like that and that would probably be the bit that would upset me about it obviously" (S14)

While life expectancy dominated any conversation on the future, a few older siblings' thoughts began to move towards having children of their own. Siblings talked about their understanding of how CF may affect themselves having children in the future, although not many saw this as an issue. Thoughts of their own future were often tied to feelings of guilt for focusing on the impact on their future, rather than that on their brother or sister.

"I feel like when I was younger and I used to be like thinking about future things, I'd think, oh, my God what if I can't have children because whoever I'm with is a carrier, but as you get older I think you realise that there's so many different things and it's really probably not going to affect me that much if I think about it compared to how it's affected [brother]" (S12)

Siblings continued to identify positives that may not have been immediately apparent to others. For instance, on the subject of having their own children, the below sibling highlighted how their good understanding of CF prepared them better for having a child with CF relative to their parents.

"When my parents, when [sister]...when my parents found out that [sister][had CF]...they didn't understand much, they didn't know, really, much about it. But, for me, if my child happens to have CF, I'd know all about it." (S11)

4.7.5.2. Changes in How Their Life Has Been Affected by CF Over Time

As the siblings got older the way that their brother's or sister's CF impacted upon their life changed. How their interactions with their brother or sister changed varied from sibling to sibling. There were a few key points which were raised throughout several of the sibling interviews. How the siblings interacted with their brother's or sister's treatments changed for several of the siblings. Some siblings found their brother's or sister's increasing independence more challenging and preferred to be involved with their brother's or sister's care.

"when I was younger, I quite liked the idea that I was also part of like looking after her, or I was you know, involved in it somehow. Whereas as I got older, I felt like I guess I probably felt like I was less, I don't know, less involved or something, or yeah ..." (S17)

A reduction in involvement with their brother's or sister's care may also mean that the impact on the sibling's day to day life reduces.

"I'd say that when I was younger it impacted me more because sometimes I'd have to do physio with him in the mornings and thing when he didn't want to and things like that but as I've got older he does it a lot more independently so he doesn't need someone to actually like physically tap his chest every morning and do that physio" (S12)

Other aspects that lessened as their brother or sister got older and, as a result, lessened the impact on siblings, varied from person to person. For some hospitalisations reduced following the initial diagnosis. When the number of hospitalisations their brother or sister experienced reduced the level of impact on the siblings also greatly reduced, as they no longer needed to see their brother or sister 'unwell' and were less likely to be displaced with other family members.

"nowadays it doesn't really impact me as much, but obviously when I was younger he'd go into hospital for long periods, I would then stay at my auntie's and I'd say impacted me quite a bit then, but I'd still enjoy myself cause I'm with the family, it was a bit more fun, but I think it did impact a bit more when I was younger than it does now" (S12)

As their brother or sister got older their independence from the family increased. The sibling's brother or sister may have chosen to move away from home for university or work. This separation from their brother or sister reduced the amount of interaction the sibling had with their brother or sister. By reducing the amount of contact the sibling had with their brother or sister things like hospitalisations were less obvious to siblings and the level of impact the sibling experience reduced accordingly.

"[sister] goes to uni and stuff...I'm more and more used to her not being at home. So, even when she does go into hospital, it doesn't feel much different" (S11)

Not all siblings felt a benefit from moving away from their brother or sister, and some found it challenging to separate from them for longer periods of time. One sibling in particular highlighted how they have felt an increasing need to spend time with their sister given her increasing age and deteriorating health.

"I guess what I've kind of discovered more in the past year cause she kind of ... I did always spend a lot of time with [sister] but like I kind of want to make the most of like when I do see her now..."

(S18)

Further to the changes in their brother's or sister's life, the siblings went through changes over time that altered the level of impact CF had on their lives. Primarily the siblings highlighted how their understanding of CF changed and the role it played in the impact of CF on their life. As siblings gained a greater understanding of CF they were able to understand the implications of CF for their brother or sister. Although this appeared to have a negative effect on some siblings, others mentioned the positives they associated with an increase in understanding.

"I think it was just the general like growing up, as I grew an understanding of why she needed or like a bit more attention. I think as I grew up with that it all sort of clicked into place a bit." (S15)

The most obvious negative taken away by siblings from their brother or sister getting older is the negative developments in their health. Having a greater understanding and, therefore, more knowledge about the implications of these changes for their brother or sister provided an additional challenge that siblings needed to deal with.

"I think as you've gotten older you know there's been ... more sort of developments with his condition that maybe weren't there when he was younger but because I'm older as well, and because we can both sort of understand what's happening, it's maybe had a bit more of an impact. You know it's maybe a bit more real than it was when I was younger." (\$13)

4.7.5.3. Changes for Their Brother's or Sister's Life Over Time

Siblings were very aware of how their brother's or sister's life changed and how this affected them. A small number of the siblings brought up key monuments in their brother's or sister's life that they noted as having a large impact. Several of the siblings' brother or sister had already transitioned from paediatric to adult CF care services. The few siblings that spoke about transitioning didn't appear to feel a notable effect of the move themselves but some saw it as associated with increasing independence for their brother or sister.

"then obviously when he turns 18, um, go to the adult clinic. I think that was a bit of a, a bit more of a change than anything else because we all see him as, as wee [name], you know? Wee boy. So, I think it was quite difficult watching him having to go in ... do that themselves." (S13)

Another element that several siblings noted as associated with increasing independence for their brother or sister with CF was moving away from home for opportunities such as university. Most siblings were able to deal with the increasing independence of their brother or sister well, but one sibling in particular found this a very challenging time.

"I think when she went to university, it was like such a big thing, but I got really, really down, really depressed and really like didn't know how to sort of deal with it, and when we used to go and visit her in [Location], and ... because she wasn't even that far away, because it was from [Location] to [Location], but I found it like really hard to deal with, because she wasn't at home anymore" (S17)

A positive development identified that was viewed as improving the lives of the sibling's brother and sister was the continued improvement in medications and treatments for CF. One sibling spoke about technology and how this reduced the amount of time it took their brother to complete his physiotherapy.

"I've seen like the development of technology with it...So that the physio used to take ages and now it can take probably less than half an hour if [brother/sister]'s quick" (S10)

4.7.6. Theme 6: How Siblings Think About and View CF

Siblings have different perspectives and approaches as to how they view CF as part of their lives. Two sub-themes were identified within this theme:

1. They View CF as Normal and Believe it Doesn't Affect Their Life

2. Conscious or Unconscious Avoidance

4.7.6.1. They View CF as Normal and Believe it Doesn't Affect Their Life

Several siblings suggested that to them, as they have grown up with CF, it is normal. For these siblings there is no notable difference in their lives due to their brother or sister having CF.

"I don't know what it's like to not have a sibling with CF. So, I've just grown up with it, so everything is, to me it's just ... I don't even notice it really, in a way." (\$17)

That is not to say that they assume themselves to be the same as their peers with healthy brothers and sisters but rather it usually suggests that they have normalised the presence of CF in their life.

"that's just been a part of my life that, for me, is [as] normal as me just talking about my sister's personality or something" (S11)

This normalisation can lead the sibling to assume that CF does not impact upon their life. The majority of siblings in the sample reported this normalisation of CF in their life, yet continuing with their interview and interpreting the findings above we can gather that CF does in fact affect their life in some way. This impact may be not be obvious to the sibling initially and require deeper thought and consideration to be illuminated.

4.7.6.2. Conscious or Unconscious Avoidance

Siblings approached thinking about CF in their daily lives in two distinct ways. There were those that chose not to think about CF. For instance, in the case of S19 they chose not to think about CF due to the distress that it caused them.

"I just wanted to ignore it all really. So, I didn't ever talk about it until I was a bit older" (S19)

For S19 this was a very conscious decision to not think about CF, whereas for other siblings the decision to not think about CF appeared unconscious. A small number of siblings also mentioned forgetting about their brother or sisters CF. When their brother or sister with CF was well siblings would forget about CF and see "just" their brother or sister, as is the case in quote below.

"she's like, "Oh I don't feel up to it today." I'm always like, "Oh just do it," kind of thing sometimes. Do you know what I mean? Cause like I don't really ... Cause she's always had it I don't really see it as somebody with a condition. I just see it as my sister" (\$18)

This can often result in the sibling feeling guilty because they feel they have forgotten the amount that their brother or sister has to go through every day while living with CF. There were also

instances where siblings felt like they were treating their brother or sister unfairly as they were forgetting to consider their illness and treated them as if they did not have a chronic illness.

"I mean he still does wind me up, like yesterday he didn't come back from hospital because he felt ill but I was fuming at him for it but then when you take a step back, I mean you've got to think, oh, well you know he is actually ill" (S14)

Summary of Results

Theme 1: A General Lack of Awareness and Understanding of CF

- Siblings believed there is a general lack of awareness of CF, which should be improved
- Siblings were largely comfortable and open to talking about CF, but they did not like to give too much detail as they saw that as their brother's or sister's personal information
- Siblings believed anyone without CF, including themselves, lacked a good understanding of CF, and that their brother or sister was the expert on the subject
- Siblings thought the individuality of CF was a large barrier to others understanding
- Siblings mainly learnt about CF from their parents, brother or sister, school, and online
- The information they received from school or saw online was subject to criticism, but the Cystic Fibrosis Trust website was generally viewed as trustworthy and helpful
- The siblings believed that the impact on siblings is under acknowledged and not well understood

Theme 2: The Challenges and Emotions Directed at CF

- Discussion of life expectancy caused all siblings distress, but otherwise siblings didn't believe they were greatly affected
- Several siblings tried to use avoidance as a way of coping while some tried to focus on the positives. Both coping mechanisms were harder to maintain when their brother or sister was ill.

Theme 3: Support, Charity, and Fundraising

- Little formal support was used by siblings. Where it was used it was not typically targeted enough or well received
- Informal support was regularly received from friends and parents
- Siblings did not think they needed more support, but would consider taking support if offered to them
- Peer support was highlighted as a key area that siblings would be interested in

Theme 4: Family Dynamics and Focus on their Brother or Sister with CF

- Relationships between siblings, their brother or sister, and parents were largely thought of as "typical"
- Older siblings typically believed they were closer to their brother or sister, and their family in general
- Siblings were aware of all their parents did for them and tried to reduce the impact on they had on their parents
- Siblings were sympathetic and protective of their brother or sister and regularly put them first
- Siblings were not highly involved in their brother's or sister's care, but many took on a more emotionally supportive role, particularly when their brother or sister was in hospital

Theme 5: Past, Present and Future

- Their brother or sister becoming more independent reduced the day-to-day impact on siblings
- Being apart from their brother or sister caused a few siblings distress, particularly as they saw their brother's or sister's health deteriorating

Theme 6: How Siblings Think About and View CF

- CF was viewed as "normal" by the majority of siblings
- Siblings regularly forgot about their brother's or sister's CF when they were 'healthy'

4.8. Discussion

The thematic analysis results presented in this chapter begin to demarcate the experience of siblings with a brother or sister with CF. For these results to be used effectively to enhance future research and support for siblings they need to be interpreted within the context of the existing literature.

CF affects roughly 10,500 people in the UK (CF Trust Registry Report 2018), yet overall siblings believed that there was a poor level of awareness and understanding in the general public about CF. How poor understanding and awareness in the general population affects siblings has not previously been considered. However, poor understanding has been associated with an increase in psychological problems in young people with epilepsy (Benson, Lambert, Gallagher, Shahwan, & Austin, 2015). Poor understanding by key people, such as teachers and friends, in the siblings' lives could be particularly detrimental to their wellbeing (Gan et al., 2017). As CF is regularly regarded as an invisible illness, siblings felt that others made several incorrect assumptions about CF and that this could change the way they treated their brother or sister, which has also previously been associated with mental health difficulties (Haukeland et al., 2015). Several of the siblings also spoke about how they thought there was a lack of recognition for siblings. Not recognising and supporting potential psychological need in childhood can lead to further difficulties in later life, and the potential for negative health behaviours in adult sibling has been noted (Buchbinder et al., 2016).

A key highlight from the presented analysis was a high level of difficulties for siblings around fluctuations and deteriorations in their brother's or sister's health. Fluctuations such as pulmonary exacerbations and hospital stays have previously been acknowledged as a concern for families living with CF (Havermans et al., 2011). Worrying about their brother or sister appeared to be a key issue for many of the siblings during these times. Previously it has been suggested that a disparity in parental attention between the sibling and their brother or sister was connected to poorer wellbeing in siblings and this was particularly an issue around periods of fluctuation in the chid with CF's health (Murray, 2001). However, the majority of the siblings included in this study felt as though they had sufficient time and attention from their parents, even when they had to stay away from home with other family members for periods of time. Siblings recognised the effort their parents put in to adjusting their lives to compensate for the time they spent away from the sibling. This concurs with the study by O'Haver et al. (2010) that found no significant association between parent caretaking while their chid with CF was hospitalised and sibling internalising and externalising behaviours.

The progression of their brother's or sister's illness was a concern for all siblings. Although younger siblings tended to be more optimistic about the future than adult siblings. Several of the adult siblings were very emotional when discussing the future, and a few felt the need to be both physically and emotionally closer to their brother or sister as their health worsened. This need to be closer has

been found in other siblings with a terminally ill brother or sister (Bluebond-Langner, 1989). There were no siblings in this sample that solely took on a caring role for their brother or sister. Many siblings wanted to provide support for their brother or sister and parents when they could. Some older siblings began to offer lifts or company at hospital appointments. Providing company for their brother or sister when they were staying in hospital was suggested by both young and old siblings as a way they were able to help. Siblings that reported a closer relationship with their brother or sister appeared to be more willing to offer up and provide support than those with either a 'normal' or conflicting relationship. Therefore, the importance of a positive sibling relationship on the probability of providing future care and support, as was previously found in siblings (Bigby, 1998), was reemphasised by the siblings in this study.

Given much of the previous family research it would be reasonable to expect a strain on family relationships to be found (Barnett & Hunter, 2012). However, in this study several siblings described their family relationships as typical and unaffected by CF and there were a couple of siblings that believed CF brought their family closer together. A stronger family relationship has been found to improve psychological wellbeing in siblings (Giallo & Gavidia-Payne, 2006). The siblings' perspective on their family relationships differed a little dependent on their age. Moving away from home and having a level of separation was viewed positively by some, but many siblings found it more challenging to have a distance between them and their family. Increasing independence in adolescent and adult siblings has not been greatly considered in the literature but how this affects family dynamics and relationships could be beneficial to investigate further.

When siblings were asked about their thoughts and experiences of being tested for carrier status for the majority of the siblings there was very little concern. Many of the siblings had been tested already, particularly the siblings that had an older brother or sister with CF. Older siblings had a better understanding of the implications of their carrier status but the idea of being a carrier of an affected gene did not appear to cause them a large amount of distress, unlike what has been suggested in the literature (Fanos & Johnson, 1997). Siblings believed that they were actually advantaged due to their knowledge of genetics and inheritance. Siblings thought that they would be better able to care for a child with CF thanks to their enhanced understanding of the condition. Being a carrier did not appear to affect family planning decisions in siblings. Siblings did acknowledge that if they wanted to have children they would want their partner to be tested for carrier status. Advancements in the diagnosis and treatment of CF help siblings to feel more comfortable should they be a carrier.

Controlling and normalising the presence of CF in their life were prominent coping strategies mentioned when siblings were asked how they dealt with difficult situations relating to their brother's or sister's CF. These coping techniques have previous been suggested as positive influences on outcomes in siblings of children with a chronic health condition (Incledon et al., 2015). Changes to

routines and long periods of separation from their brother or sister where the sibling has limited control caused the greatest amount of distress. In keeping with the findings from previous research, and connected to the problems around fluctuations and progression of their brother's or sister's health, it appears that coping is more difficult for siblings during periods of change in their brother's or sister's health (Houtzager et al., 2005). During these periods it is more challenging for siblings to avoid CF, which was another coping technique commonly used by the siblings.

Intuitively from the above it would be anticipated providing siblings with support may be beneficial, particularly around periods of poor health in their brother or sister, as has also been discussed in previous research (Hartling et al., 2014) and the results of Chapter 2. However, when asked very few of the siblings were engaged with any service that may be able to offer them support. Siblings did not perceive themselves as in need of support and when asked tended to suggest a need for support in the rest of their family. A number of siblings suggested that if they were proactively offered support they would be willing to take it up. Most siblings agreed that they would be interested in talking to other siblings about their experiences although there were hesitations about talking to other siblings who may have a brother or sister in worse health. The concept of talking to others that were able to "understand" was viewed positively by all.

4.9. Conclusion

The experiences of the siblings involved in this research project both concurred and deviated from what would be expected given the existing literature. Although they recognised it, the day-to-day burden of CF did not appear to impact on siblings a great amount, rather notable changes from the day-to-day such as hospitalisations had a greater impact. This is not in keeping with the general concept that the high treatment burden of chronic illnesses or conditions is associated with a lower psychological wellbeing in siblings (Vermaes et al., 2012). The mortality of their brother's or sister's condition does appear to play a role in their psychological wellbeing and distress, in keeping with the literature (Vermaes et al., 2012). Adult siblings in this sample did not appear to take on a focal caregiving role, but rather supported the care provided by parents and their brother's or sister's partner, as has also been noted in adult siblings of individuals with Type 1 Diabetes (Wennick et al., 2009). Finally, it is unclear how best to support the siblings of those with CF, there is no clear point of engagement as they do not regularly attend clinic and often choose to avoid all things to do with CF. Hearing the perspective of other members of their families may help to either identify how best to engage siblings or indirectly supporting siblings through their family.

Chapter 5: Results - Young People with Cystic Fibrosis

A sub-sample (n=13) of the interviews with young people with CF included in this chapter were analysed for a dissertation submitted to The University of Manchester for the degree of BSc Psychology with Professional Placement in the Faculty of Biology, Medicine and Health. The analysis included in the dissertation are separate from that shown below.

5.1. Introduction

It has been suggested that by using multiple respondent in family research, it is possible to build a holistic understanding of individual experiences (Reczek, 2014), which is particularly important given the highly interconnected nature of families (Williams et al., 2002). Many of the siblings included in the previous chapter reported that their experiences with CF were directly related to the wellbeing of their brother or sister. Eliciting the impact of CF on their brother or sister with CF is necessary to provide a complete picture of the impact on siblings. This chapter facilitates a greater depth of understanding of the sibling experiences by presenting the findings from 25 interviews with young people with CF included in the large qualitative research project described in Chapter 3.

5.2. Background

CF is a chronic illness that, despite an increasing life expectancy, is as of yet incurable. Improvements in both the treatment and diagnosis of CF have allowed the expected survival age to increase greatly over the last 40 years (Stephenson, Stanojevic, Sykes, & Burgel, 2017). New-born screening programmes (NBS) have drastically improved the speed of diagnosis allowing appropriate care to be put in place earlier (Castellani et al., 2009). The presence of a chronic condition, such as CF, in childhood has been associated with adjustment difficulties in young people (Barlow & Ellard, 2006). Ernst et al. (2010) considered developmental and psychosocial issues related to CF and suggested that there is evidence to suggest both challenges and positive correlates of psychosocial development in young people with CF.

Relationships between young people with CF and their immediate family play a crucial role in their social development (Ernst et al., 2010). Attachments between the young person and their parents are often formed at a similar time point to the diagnosis of CF (Ernst et al., 2010). Diagnosing CF in a young person is a complex and emotional process (Havermans, Tack, Vertommen, Proesmans, & de Boeck, 2015). During this period it is likely that the level of psychological distress a parent experiences is high (Tluczek, Koscik, Farrell, & Rock, 2005) and yet the relationship between parents and their child with CF does not appear to be greatly affected. This suggests a resilience that allows positive attachments to be formed despite high parental stress and changes in behaviour (Solomon & Breton, 1999).

The relationship between siblings is typically one of the longest standing relationships an individual has and the importance of the relationship for social development has been highlighted (Ma et al., 2017). Several areas of disruption exist when the sibling's brother or sister has a chronic illness or condition. Parents of children with CF have to spend a higher proportion of their time with their child with CF. Parents attend clinical appointments and hospitalisations for their child with CF which can cause feelings of resentment and jealousy in the sibling towards their brother or sister (Berge & Patterson, 2004; Williams et al., 2009).

Peer relationships play an important role in children and young people's social development (Hartup, 1993). The way young people interact with their peers can be affected by the presence of a chronic illness or condition (Meijer et al., 2000). Young people with CF face further complications in meeting and interacting with their peers with CF due to a risk of cross-infection (Schaffer, 2015). Interactions with peers with CF can increase young people's awareness of the permanency of CF and cause them emotional difficulties (D'Auria, Christian, Henderson, & Haynes, 2000). Despite CF affecting roughly 10,500 people in the UK public awareness and understanding remains low (Braido et al., 2015). Awareness and acceptance help young people to be more open with their condition, which can improve adjustment and peer relationships (Casier et al., 2011).

Evidence regarding the psychological wellbeing of young people with CF is mixed. There is evidence that suggests poorer mental health outcomes in young people with CF. In particular higher rates of internalising behaviours, such as anxiety and depression, have been found (Szyndler et al., 2005). There is also evidence that suggests the mental health and QoL in young people with CF is comparable to their peers without a chronic illness or condition (Havermans et al., 2008). Positive adjustment is also suggested in the young person's immediate families (Berge & Patterson, 2004). Positive adjustment may be due to the use of effective coping mechanisms (Wong & Heriot, 2008). Psychological distress in young people with CF may centre around particular time points, such as the transitioning between care services, rather than occurring at all times. The process of transitioning from paediatric care to adult care services has frequently been investigated (Heath, Farre, & Shaw, 2017). Transitioning has improved over time and there now appears to be limited disturbance to the young person (Chaudhry, Keaton, & Nasr, 2013). Another time point that can have negative psychological effects on young people is their first inpatient stay (Hanline, 1991; Hegarty, Macdonald, Watter, & Wilson, 2009). Inpatient stays impact on both the young person and their immediate family as the reality of the permanency and progressive nature of CF is highlighted during this time (Pfeffer, Pfeffer, & Hodson, 2003).

The way in which young people with CF cope has been investigated (Abbott, 2003) and also placed in the context of their immediate family (Wong & Heriot, 2008). Proposed methods of coping in young people include avoidance and positive reframing, similar to that found in siblings. Positive

reframing involves techniques such as acceptance, normalisation, and adaptation (Wong & Heriot, 2008). The use of routines, for instance, has commonly been reported as a practical way young people can adapt their lives to cope with the intrusion of CF.

Young people with CF were supported through various channels, including their clinical team, charities such as The Cystic Fibrosis Trust, and their immediate family. The support available to young people has improved over time and is now generally viewed positively (Coyne, Sheehan, Heery, & While, 2017). Given continual advancements in technology and understanding, young people with CF are facing challenges that were previously less of a concern for those with CF, such as entering the job market. There is a continual need to consider the support services available for young people with CF.

Given the highly interconnected nature of families, particularly those living with a chronic illness or condition (Cox, 2010; Williams et al., 2002) each of the above concerns for young people with CF has either a direct or indirect effect on their siblings. Family relationships in a life with CF are highly centred on the health and treatment of the young person with CF as acknowledged in both Chapters 3 and 4, family dynamics play a crucial role in sibling adjustment (Ma et al., 2017). By considering the young person with CF's experiences it is also possible to enhance the understanding of the sibling experience.

5.3. Aims and Objectives

As with the sibling results presented in Chapter 4, consideration was given throughout this chapter to the overall aims of the qualitative project as presented in section 3.2.10 of Chapter 3. The aim of this chapter was to reconsider the experiences of young people with CF and the implications for their immediate family. To address this, the following research questions were proposed:

- 1. How do young people with CF view the impact of CF on their lives? Specifically, do they identify an effect on their and their immediate family's psychological wellbeing?
- 2. Do young people believe they and their family are being well supported? If not, are there areas where they believe support could be beneficial?

5.4. Young Person Recruitment

Following discussion between the research team and an experienced qualitative researcher an initial aim of 25 participants was selected. The reasons for this target sample size were addressed in Chapter 3. In total 31 young people with CF were recruited, although only 25 of these young people completed an interview, in keeping with the initial target. The further six completed only a QoL measure. The majority of the young people were recruited from clinics across the UK, London (n=11), Cardiff (n=9) and Glasgow (n=1). A further four young people were recruited through online advertisements. As it was not a requirement for full families to participate there were a range of immediate families included for the young people. Sixteen of the young people also had a parent included in the project and four had two parents included. Fifteen had a sibling included in the

previously presented analysis, while one had a sibling with CF included also involved in the young person analysis.

Recruitment of young people with CF was more straight forward than the recruitment of siblings. The target recruitment figure was reached quicker for young people than for the other groupings and as such some interested parties were unable to complete an interview with the team. Due to the breath of the age range for inclusion in the young person category (age 11 to 24), it was possible to recruit young people from both paediatric and adult clinics, therefore young people were recruited at all 7 locations across the UK and were able to self-refer through the online advertisements. This may also have added to the ease with which young people were recruited.

5.5. Data Collection & Analysis

Data for each group were collected concurrently. Data collection and analysis were coordinated by MMS with the support of other researchers. The consent and data collection procedures followed for the young people mirrored that completed with the siblings. Twenty-two of the interviews with the young people were completed over the phone, and the remaining three were completed face-to-face either in the participants home (n=1) or in GOSH (n=2). To maintain interviewer consistency (Biemer & Lyberg, 2003) the majority of young person interviews were conducted by CL (n=19), with the remaining conducted by MMS (n=5) and NK (n=1). Interviews with young people lasted on average 32 minutes. All interviews were transcribed verbatim by a member of the research team (CL (n = 18); LA (n=5); MMS (n=2)). All interview transcripts were cross-checked against the original audio by another member of the research team. The analysis process was completed by MMS and CL. Codes were checked using a code-book method by NK and MMS (Ulph et al., 2010). No young people responded to the participant validation check with any concerns about the coding and theme structure developed from their interview.

5.6. Participant Demographics

The age of the young people ranged from 11 to 24 years (median = 14). Twenty of the young people were currently students, one was unable to attend school due to their health, and the remainder were either working or seeking work. All of the young people were of white ethnicity, and half of those with available information were from high income families (£50,000+ per annum). Fifteen of the young people also had a sibling without CF included in the study, 14 had one parent in the study, and four had two parents included in the study. Further details about the young people can be seen in Table 5.1., missing data is again shown in blank cells within the table.

Young	Age	Sex	Marital	Employm	Young	Age	Sex	Marital	Employm	Househol	Sibling	Parent 1	Parent 2
Person 1			Status	ent Status	Person 2			Status	ent Status	d Income			
Y1	11	F		Student						15,000-25,0	000	P6	
Y2	11	М		Student						50,000+	S10	P10	
Y3	11	F		Student						35,000- 50,000	S5	P22	P21
Y4	11	М		Student							S7		
Y5	11	F		Student						50,000+		P2	Р3
Y6	12	М		Student						25,000-35,0	000	P4	
Y7	12	F		Student							S8	P8	
Y8	12	М		Student						50,000+		P9	
Y9	12	М		Student						50,000+	S9	P23	P24
Y10	13	М		Student						50,000+		P12	
Y11	13	М		Student						35,000- 50,000	S1	P14	
Y12	13	M		Student							S2	P18	
Y13	13	F		Student	Y16 ¹¹	14	F	Single	Student	15,000-25,0	000	P20	
Y14	13	F		Student						15,000- 25,000	S3	P1	
Y15	13	М		Student								P5	
Y17	16	М	With a Partner	Student									
Y18	15		Single	Unable to A Health	ttend School	Due to							
Y19	16	F	Single	Student									
Y20	16	М	Single	Out of Work								P7	

Y21	17	М	Single	Employed For	ull/Part			50,000+	S4	P13	P11
Y22	18	М	Single	Out of Work					S13		
Y23	24	М	Single	Employed Fo	ull/Part				S16		
Y24	20	F	Single	Student							
Y25	20	М	Single	Student				35,000-50,	000	P19	
Y26	20	F	Single	Student							
Y27	21	F	Single	Student				50,000+	S11	P25	
Y28	22	М	Single	Employed For	ull/Part				S12		
Y29	22	F	Single	Employed For	ull/Part						
Y30	24	М	With a Partner	Student					S14		
Y31	24	F	Single	Out of Work					S15		

Table 5.1. Young Person with CF Demographics

¹¹ Y13 and Y16 were sisters both with CF

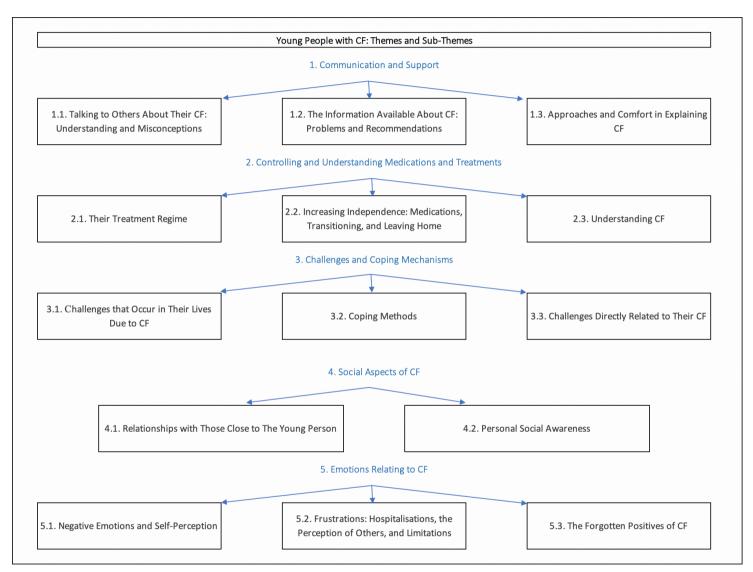


Figure 5.1. Themes and Sub-themes of Young People with CF Interviews

5.7. Themes

From the qualitative thematic analysis conducted on the 25 young person interview transcripts, five themes were identified:

- 1. Communication and Support
- 2. Controlling and Understanding Medications and Treatments
- 3. Challenges and Coping Mechanisms
- 4. Social Aspects of CF
- 5. Emotions Relating to CF

Each theme contained multiple sub-themes. The themes and sub-themes are given in rank order of reference density and displayed in Figure 5.1. Each theme is discussed below with verbatim examples taken from transcripts.

5.7.1. Theme 1: Communication and Support

Communicating about CF can be challenging for young people with CF. How open they are about their CF and how comfortable they feel discussing it with others can have a large effect on their mental health and wellbeing (Casier et al., 2011). Three sub-themes were identified in this theme:

- 1. Talking to Others About CF: Understanding and Misconceptions
- 2. The Information Available About CF: Problems and Recommendations
- 3. Approaches and Comfort in Explaining CF

5.7.1.1. Talking to others about CF: Understanding and Misconceptions

All of the young people mentioned that the level of understanding others had of CF altered how they felt about speaking to them about CF. Many of the young people mentioned that it would be better if others were aware of CF.

"if they knew about it already or, know it exists, it'd be easier to explain, you know." (Y20)

This was particularly the case for the young people who were less comfortable discussing their CF. Even with an increase in awareness some of the young people believed that others would still not understand what it was like to have CF. Misconceptions about CF led the young people to worry about how they were viewed by others. Others having misconceptions about CF changed how the young person felt about disclosing and discussing their CF with them.

"people have the educational knowledge about it which always seems a lot worse than how I feel it is? So I kind of worry they're going to go oh I know what that is, I know how rubbish that is and then take that view rather than actually... everyone's different and I'm not as bad as maybe the majority of people are" (Y27)

When someone in a position of authority, such as a teacher, lacked awareness or understanding young people were more resistant to discuss their CF with them, which could lead to negative complications for the young person.

"... I think it would help me more because I don't normally say if I'm feeling unwell to a teacher. I think it might help my confidence to say that maybe... because I have seen a lot of people when they go up to say they're not feeling very well just get sent straight back down so... I just don't see the point" (Y15)

One person the young people felt that would be able to understand what they were going through and that they would benefit from talking to about their CF is other young people with CF.

"Someone that you could like go to and they wouldn't like... there'd be no judgy or kind of, they'd be good at understanding... They'd know about it and it's fine... you do feel this fear of people not getting it." (Y29)

Many young people felt that others with CF were best positioned to empathise and understand their experiences, allowing them to offer appropriate advice and support. However, a few of the young people mentioned that speaking to another young person with CF in worse health than them may be discouraging and scary.

Several of the young people mentioned that they were happy to speak to their family about their CF. A smaller number of the young people didn't wish to discuss their CF with their family preferring instead to talk to their friends.

"Not really [talk to family]. Because I think everyone just kind of like deals with it. And like knows everything about it. So, I think talking to my friends is kind of better" (Y5)

5.7.1.2. The Information Available About CF: Problems and Recommendations

Sources of information about CF the young people mentioned during their interviews included the Cystic Fibrosis Trust, other online resources, their care team, parents, and school. Their care team provided the young people with the majority of the information they needed. The young people felt that this information was helpful as it was reliable and readily available. However, there were a few circumstances when the young people did not feel they gained sufficient detail from their clinical team.

"The hospital did give me information, but it wasn't like kind of enough... it wasn't fully explained" (Y5)

When young people wanted more details they typically sought it out online, as was the case for Y5 quote above. However, when young people used online resources, they were cautious of the

information they read and trusted. Most young people saw the Cystic Fibrosis Trust website as a resource they could use for reliable information about CF.

", sometimes I would go look on their [the Cystic Fibrosis Trust] website first cause they do have some like useful, helpful things and videos on there...You can trust the information they give you as such like you don't have to second guess it and think oh is this right? Like, it is going to be right because it is from them." (Y19)

A source of information that several of the young people highlighted as being particularly untrustworthy and regularly out-of-date was that offered in school textbooks. Several of the young people had been faced with information that was outdated while being taught about CF in school. This information could be quite upsetting for the young people. Providing the young person's friends and school mates with an inaccurate picture of CF also upset the young person.

"textbooks don't explain it great at all... That was something that kind of bothered me at school I think but like the biology textbooks would be like 'they can't do exercise' and like 'the life expectancy is like 30s' and that's all changed now" (Y29)

To better inform teachers and schoolmates about CF several of the young people chose to either give a presentation themselves about their CF or brought in someone from their care team or the Cystic Fibrosis Trust to present. A few young people were more reserved and cautious of this level of openness about CF. However, the majority of young people thought informing teachers about CF would help them, as in the quote below.

"I think before you actually start school they should send like someone out to meet with one of them [teacher], just to like explain what happens in our lives compared to other people's...Because if they would understand it would make your life much more easier [sic]" (Y8)

While reliable information about CF is easily accessible from sources such as the Cystic Fibrosis Trust, several young people doubted that others were aware that this information was available. Many young people doubted that others had even a basic awareness about CF and, therefore, did not believe they would go looking for information.

"like cancer research and stuff, you know that kind of stuff is everywhere in a way, so when someone mentions it, it is just common knowledge that this person has it and you know what it is. But with CF I haven't seen much about it anywhere besides if I was specifically looking for that." (Y20)

5.7.1.3. Approaches and Comfort in Explaining CF

Many of the young people stated that they were open to talking about CF with others. Although many young people amended this to being open when asked direct questions about CF.

"I am open with my CF and like I do tell people if they want to know but I wouldn't just openly... If they didn't ask about why I was coughing or like why I had Creon or something, I wouldn't necessarily just go up and say 'I have CF'. But that's just me personally" (Y19)

Several of the young people believed that being open to questions from others about CF allowed them to help reduce the likelihood and number of misunderstandings others had about CF.

"if they're genuinely curious about it I'm more than happy to talk about it, more than happy to give them as much knowledge as they are looking for. I much prefer it when someone asks me a question rather than just assume something" (Y27)

Besides increasing others understanding of CF and reducing misunderstandings, some young people also saw being open about CF as potentially beneficial for their own wellbeing.

"I think like being more open about things and actually talking about it more has helped, which I wasn't doing at University, particularly [...] I kind of just didn't tell people that I was in hospital I just sort of... but now looking back, it's like, why didn't I?." (Y29)

A few young people were previously very open about their CF but chose to become more selective over who they told about their CF and the information they gave as they got older. The young people didn't want to be known and defined purely by their diagnosis.

"I used to be very happy telling people and talking to people and then I kind of realized that I don't want to tell everybody just straight away and to kind of, make sure they know who I am first, and realise that it's not what defines me." (Y27)

How young people brought up the subject of CF and the level of detail they provided was frequently mentioned as an uncertainty. How others perceived the young person was very important to them and was a key source of concern for many.

"I don't actually know which friends know I have it...It sounds weird but I'm never sure... Like if I, if I haven't remembered consciously telling them in like secondary school let's say, I don't know if they're aware, like other friends have told them or not or if they are not aware. Like generally I am not really sure. Because I have never come out and actually been like 'Do you know...' because I feel like by this point it's kind of a bit weird. It's like, I have known you like 11 years or something, it's weird to suddenly go 'By the way...'." (Y31)

A few of the young people had negative experiences when they spoke to others about their CF. Their diagnosis was a sensitive and private topic for young people and it can be disarming to trust others with this information.

"I literally only told two people until I was 18 so, so they were the only ones that I trusted...Really good friends of mine yeah. I trust them for years and they didn't even realise that I had it." (Y24)

When young people explained their CF to others, they typically give a very brief explanation that touched upon the key biological factors of CF. For instance, many young people mentioned their lungs and the fact that CF is a genetic condition. Sometimes the young person may choose to also mention other aspects such as their digestive problems or complications such as diabetes. The descriptions provided were clearly tailored to the aspects of CF that affected the young person giving the description. Some young people also chose to alter the way they described CF dependent on who they were talking to or the circumstances in which the topic came up.

"a genetic condition, causing a build-up of mucus, affecting multiple organs. It's really... it depends on how it's coming up like, I just say it affects a lot of my day, like everyday life. But then sometimes I won't say that, depending on... because I don't want it to be like a massive big deal but sometimes it is, it is a massive big deal, so it's sort of... it does vary." (Y29)

5.7.2. Theme 2: Controlling and Understanding Medications and Treatments

All of the young people had some level of understanding and control over their own medications. Both knowledge and independence with medications and treatments changed over time, which altered the way the young people felt about their CF. Three sub-themes were identified within this theme. They were:

- 1. Their Treatment Regime
- 2. Increasing Independence: Medications, Transitioning, and Leaving Home
- 3. Understanding CF

5.7.2.1. Their Treatment Regime

The young people understood why they were doing their treatments and what they needed to do in order to stay well. However, several of the young people found interrupting their daily activities in order to complete their treatments frustrating.

"it's quite annoying when I do my treatments cause I know that I could be doing like better things and that and it can get quite annoying. Yeah because I don't really like doing my nebulisers and stuff so it's a bit annoying because I don't I'd rather be doing better things" (Y3)

Further to the interruption the young peoples' medications and treatments had outside of school time, the young peoples' schooling was frequently disrupted. Issues faced because of medications and treatments at school typically related to the young person having to miss-out on something or a concern about others perception.

"with my friends, we're like eating and they instantly pick up their stuff and just go walk for like a walk, and I'm just still sitting there behind sorting out all my medication. And I end up like losing them. And, and then I have to do more medication afterwards if my lungs are bad. And I have to like leave a lesson, which makes you feel a bit odd and like different" (Y11)

Numerous young people mentioned that they recognised and understood the benefit to them of being active and participating in sports and exercise. Several of the young people associated their positive health with the fact that they had maintained an active life style.

"Yeah no but, dance is probably...it's a really active sport however you get a lot of little breaks during it [...] I've been doing it for as long as I remember so I can imagine that I'd be worse without it"

(Y27)

5.7.2.2. Increasing Independence: Medications, Transitioning, and Leaving Home

All of the young people had some level of independence with their medications and treatments. For instance, the majority were able to complete their physio and nebulisers independently. As the young people got older they steadily took on an increasing amount of responsibility for their treatment and medications. Y19, articulated a step by step process of becoming responsible for their own treatments.

"at the beginning I was just managing the physio by myself and then it come to like managing nebs by myself and then it was like setting up my neb, putting my neb away things like that and it gradually got more... one step at a time kind of thing" (Y19)

Several of the young people still maintained some level of involvement from parents, whether that was help with ordering their medications and ensuring they have an adequate supply or a quick check in from their parents every so often to ensure they are doing it correct, as was the case for Y19.

"I can manage all of my treatments myself now which is quite good but sometimes like every so often or every few days or couple of weeks or something they [parents] might come and check, like check my weekly medicine or watch me do my neb and physio or something.... it is quite nice to have that little bit of help as well" (Y19)

A landmark for several of the young people was the moment they moved out of their parent's house. This increase in their independence, which quite often came when they began university, typically came with a large amount of responsibility for their CF treatments. Without any regular supervision or prompting from parents, several of the young people dropped off in their adherence to their treatments and medications.

"at the very beginning, when I very first started uni, I felt, I did it quite strongly, it was my first time being independent, doing it all by myself. And that quickly teetered off into very rarely doing it"

(Y27)

Despite the potential to slip away from taking their medications, the majority of the young people enjoyed their increasing independence. Despite their independence parents regularly continued to want to check-in with the young person and this was not always viewed very positively. However, the young person did typically empathise and understand that their parents were used being in control and may find it difficult to let go.

"My parents still struggle not to sort of nag me and such to like "have you done your medicines" they'll say and they'll comment...they've had like 18 years of responsibility of me and now I have to do it on my own. So, it was a jump. I did feel it" (Y24)

Parents typically attended paediatric clinic appointments and were fully informed on all things to do with their child's health. When the young person was young parents would lead during clinic appointments. However, as young people get older the clinicians are often more likely to address them directly and their parents begin to take on a supporting role.

"when I was younger they [parents] used to answer most of my questions that the doctors would ask whereas the past 2 years they've let us answer on our own and then if we ever needed help or were struggling with words to think of they'd chip in and help us" (Y13)

Going into hospital was a daunting task for a few of the young people, particularly those who were younger or were uncomfortable with their care team. As the young person got older and developed a greater confidence in them self and their CF it became easier for them to speak to their care team and tell them how they were feeling.

"it's still really scary going to the hospital but it just maybe seems because I'm a bit more grown up I don't feel as isolated from things and I can maybe ask whereas as a child I couldn't vocalise necessarily how I was feeling as much" (Y26)

Transitioning from paediatric services to adult services was a key move towards independence several of the young people had made. The young people who were either in the middle of the process or were just beginning to talk to their clinical teams about transitioning offered very little opinion on the matter. Young people that had already gone through the process had a range of experiences and emotions on the matter. Nerves were common in those about to transition.

"I feel a little bit nervous but a little bit excited just to see what it is like, just a little bit of a difference really" (Y19)

Introducing the young person to the adult care team and the new environment provided the young person with comfort that helped them to see the move as exciting rather than the daunting prospect it could have been.

"They gave me a mini tour of like the CF unit and things like that. So, I feel a little bit more confident going in there now knowing where I am going to go on my first clinic appointment and I feel a little bit more confident because I met quite a few of them which makes me feel a little bit more relaxed about it and a little bit more excited than nervous. Because if I was going in not knowing any of the doctors I would feel really nervous about it" (Y19)

While the majority of the young people expressed a similar positive experience to Y19 there were also a few that found the transition a lot more emotionally challenging

"that was very similar to like leaving home from going to university for my parents. Because I, I missed paediatric as soon as I went to adult. I really missed it..." (Y24)

Close bonds with their paediatric care team along with the change in physical environment the young person had come to know caused some of the young people to miss paediatric services.

"It's very different. Paediatric is a lot more homely... I feel comfortable in that building because I feel like it's got more attention and like the staff are more needy and helpful whereas when I went to adult it was completely different. I kept on having different doctors all the time, I still do. I still don't know who's the ones I have" (Y24)

5.7.2.3. Understanding CF

Having a good understanding of CF, including the treatments and medications, reduced the impact of CF on the lives of many of the young people. A poor understanding of CF was associated with difficulties the young person had experienced.

"as a child you don't necessarily have a proper grasp of what's going on so for me it wasn't major things, it was being held back from lunchtimes and things like that, but I think that for someone else with CF if they're held back from other things and there's not that total understanding I think that was the more difficult thing" (Y26)

The majority of the young people believed their understanding of CF had improved as they got older. Many of the young people mentioned gaining an understanding by studying CF at school. However, studying CF at school was challenging for several of the young people, particularly due to inaccuracies in the information they were provided with and teachers lacking understanding. A few young people did not have entirely negative experiences when studying CF at school.

"I found it fine and I remember being in the exam and writing about it being like I hope I've actually got this right...I know this I definitely do, I've had it my whole life" (Y27)

Some of the young people, particularly those a little bit older, expressed some interest in research related to CF. For instance, several young people took interest in new medications that were either mentioned by their care team or featured in research they had seen. Young people were very positive about research and expressed excitement at the fact that others were trying to improve the lives of those with CF.

"...I sometimes look up on the website or the doctors have explained to us about what they've been up to lately. I feel really excited to know that they're actually working on doing something and improving" (Y13)

5.7.3. Theme 3: Challenges and Coping Mechanisms

Growing up with a chronic illness such as CF provided additional challenges for young people. How they chose to cope with the challenges varied. Three sub-themes were identified within this theme. They were:

- 1. Challenges that Occur in Their Lives Due to CF
- 2. Coping Methods
- 3. Challenges Directly Related to Their CF

5.7.3.1. Challenges that Occur in Their Lives Due to CF

Young people highlighted four main challenges they experienced - school, work, travel and meeting others with CF. Many young people mentioned particular challenges they experienced relating to their school or work life. Challenges faced at school largely focused on missing out.

"like I've got like really important things going on in school and I have to sit there doing 3 nebs, 3 or 4 nebs, it can take forever and I can miss it. I've done that a few times...missed like important talks.

About, like I missed the first set of choosing options at GCSE's? I completely missed that" (Y11)

When the young person had to miss classes due to their CF, for instance for an appointment or because their symptoms were flaring up, such as is the case for Y11 quoted below, the information they received in order to catch-up was not always sufficient or easy to understand. This meant it was hard for them to catch-up without seeking additional support.

"I've been off school with my chest...It can be a bit annoying, and I, I don't always understand what I'm doing because the teachers that have given me my work aren't my actual teachers, like my subject teachers...So I don't always understand it, but some of my friends would text me a picture of what's on the board and that helps make it a bit better" (Y11)

Some young people had far more positive experiences with their school and mentioned the dedicated support they were given from the school to ensure they were able to catch-up with the materials they missed. As the young people get older and considered their move into the job market they knew that they had to make additional considerations compared to their peers. CF limited the aspirations of some young people.

"I'm still thinking about it because CF restricts most jobs that I'd like to be. Some jobs like the police was one but I can't run for long distances. And fireman I can't be near, near the smoke" (Y2)

If the young person was not sure which job they wished to pursue they were aware that their CF was likely to interfere and continue to cause issues in the long-term.

"I have no idea what I want to be or I just hope that my CF doesn't get in the way... I feel like if I do get worse over time then I'll have to start doing more treatments and it'll affect my time if I get a job and my working hours" (Y16)

The majority of the young people were open about their CF with employers. Viewing themselves as disabled was difficult for a few of the young people when applying for jobs and choosing whether to tell potential employers about their CF.

"it's difficult because when you do applications it says do you consider yourself to have a disability and that is such a broad kind of, and I don't and I think just me personally I wouldn't see someone with diabetes and think oh you're disabled it's just something that you have to live with but I think when you're kind of going for a proper grown up job it's something that you really should be you should be saying just for your future benefit I suppose" (Y26)

The young people were aware that work would be challenging to balance with their CF treatment regime. However, the young people did not feel they had sufficient or appropriate advice about how to balance work and treatments. The young people who were about to begin working were interested to hear how others with CF managed their treatments while at work.

"because they're always telling me at hospital and stuff 'oh yeah you can do your nebulisers in your car at lunch' and things and I am just like 'is that practical?'. Yeah, you know, I am always like 'really?'" (Y31)

A challenge which was brought up by several of the young people was that of travelling. The young people highlighted the need to be organised to ensure they took all the required medications with them and going through airport security as concerns. For the younger participants that still relied on their parents for support with various aspects of their treatments going away for even one night was a challenge.

"If you wanted to go away on like a school trip or something one of my parents has to come and do it on the overnight thing and do my physio...my dad had to come and he stayed out somewhere else and then he came in in the morning and in the evening to do my physio" (Y8)

It is known that individuals with CF should not be in close proximity to each other as there is a high risk of cross infection. This in itself is a challenge for the young people. Not solely due to not having the opportunity to meet someone else with their condition but the limitation it can put on the places they can go.

"being around other people with CF other than my sister, I can't do it so if I want to go to a place where another person with CF is I can't so that kind of disappoints me a bit whereas everyone else can go anywhere whereas I'm limited to some places" (Y13)

Several of the young people commented on a level of irony in some of the fundraising activities they had heard about being done for CF as they were activities that took place in one central venue where it was not possible for anyone with CF to attend.

"I think it was someone was putting on a concert or something the other day or there was a comedy thing and it was literally like no people with CF can go to it. It makes it awkward, you know, you know the only theatre where it's like 'yeah come and support one of these' but can no one with the disease actually go" (Y31)

5.7.3.2. Coping Methods

There were several different coping methods mentioned by the 88% of young people who spoke about them during their interviews. Some of the coping methods were conscious efforts made by the young person while other coping methods were unconscious but came through in the young person's responses to the interview questions. One conscious and common method used by young people to cope with their CF was forming a clear routine and planning ahead. This was particularly applied to their medications and treatments.

"I think I like to be prepared for things. Like I am very I think organised I think. Is probably the way
I deal with it is if I am like, I always want to know things in advance" (Y31)

When the young person's routine was disrupted or they felt unable to maintain a structured routine the young person felt a lack of control and were more likely to struggle, as was the case for Y25.

"it was quite difficult initially, trying to find a, a routine, the time to do all my medication, keep up with work and still have time to like do the obvious things, you know, cook and everything else. So

initially it was quite difficult. As time went on it got easier and now I'm in a steady routine and I get everything done" (Y25)

Another common coping technique mentioned by young people was to put CF out of their mind and "forget" about it. This avoidance technique, as was also seen previously in siblings, has been found in young people with a chronic illness or condition previously (Abbott, 2003). Other people frequently supported this form of coping as they treated the young people the same as others, which was generally appreciated by the young person as it allowed them to effectively avoid CF and on occasion forget it all together.

"Sometimes I feel like I don't have CF at all because I just feel like normal to them" (Y19)

"it's possibly not the best way to deal with it but I generally just try and ignore it I think" (Y26)

Forgetting about their CF is only possible when the young person has accepted and normalised the presence of CF in their life. The young people viewed CF as something they have had throughout their life and as such "normal" for them, allowing them to forget.

"I've got to be honest I don't really think about CF if I'm thinking about myself. In a way I forget that I have it at times. Not because it doesn't affect me, because... I mean I've got a habit down and I just do everything automatically, I don't see anything like out of the blue like or anything strange..."

(Y25)

Despite the young people being experts in their condition several found it difficult to articulate the impact CF had on them. Normalisation, adaption, and avoidance coping may explain why this was found.

"It's probably a bit hard for me to say what it's like to have CF in some regard because there might be a lot of things that just seem normal to me that other people would notice as different"

(Y21)

When the young person wanted to speak to someone about a difficulty they were having with their CF it was predominately their parents they would speak to first.

"if I'm worried about something I talk to my mum and things and about blood tests and tablets and stuff and changes in my medication" (Y12)

Other young people chose to speak to friends, their clinical team, or their teachers. Most of the young people understood that it was beneficial for them to speak to someone when they were struggling.

"I usually either talk to Mum or the teachers and just talk it out a bit instead of holding it in" (Y2)

Some young people mentioned specific techniques that they used to get through difficult times. However, techniques that the young people were able to identify tended to be associated with negative coping, although viewed as a necessity by the young people.

"I sort of, sort of let it out I guess. I sort of, you know, might shout a bit...I don't turn to, to anything, I don't turn to food or whatever. I'm not like that. But I sort of learn that I, that I have to live with it and that while, you know, it does feel good to, to shout it out, you know, that's not really going to help" (Y10)

5.7.3.3. Challenges Directly Related to Their CF

It was harder for younger participants to recognise and articulate the challenges they had faced that were directly related to their CF. One challenge that was identified was remembering to take their medications, particularly their Creon, when at school or with friends and the side effects they suffered if they forgot.

"sometimes I would forget because I'm with my friends and then I'd get a bad belly and then I'll
have to go to the nurse" (Y12)

The treatment requirements of CF and the strain that this put on their life was also highlighted by several of the older young people. Issues they experience as a repercussion of their treatment regime included being more tired due to having to get up early and concerns about their increasing number of treatments and medications.

"life is harder in general when you are older anyway. So, I think having that [CF] on top of it is probably, you know, it doesn't make it easier I don't think, that's the problem. Like it's, you always end up just having more treatments, you never seem to get less" (Y31)

Side effects from new medications were challenging and, occasionally, upsetting for the young people.

"I started on a drug... in the first few days it was making my chest tight which is when I was doing sport, I found it sometimes hard to breathe but that was over in like 3 days and now I've got a problem with like throwing up because of it and I guess I can't just... It just gets boring having, like throwing up, it's not a good feeling" (Y8)

The effects and complications of CF were challenging for many of the young people. Having to take additional breaks relative to their peers when they got out of breath was an issue for some of the young people. A smaller number of the young people suffered from more severe complications such as CF related diabetes (CFRD). Adjusting to the extra requirements following a diagnosis with CFRD was

challenging for some participants, such as Y29 below, while others saw it as something they simply had to fit into their existing routine and then normalise.

"the bigger transition for me was being diagnosed with diabetes when I was 14. So, it was still while I was in paediatric care but that was another dimension that I had to adjust to and that, the adjustment to that was a lot harder" (Y29)

"it's not a pleasant prospect having to be taking medicine every single day for the rest of your life" (Y26)

The greatest challenge the young people identified in relation to their CF was the permanency of CF and the deterioration of their health as they got older. Concerns about their worsening health were mentioned by several young people.

"You're not likely to get better in any way so it [their health] is worse or at the best it will just sort of stay the same" (Y21)

5.7.4. Theme 4: Social Aspects of CF

Having CF is a daily reality for the young people included in this study, and various aspects of CF have the potential to interrupt their relationships on a daily basis. Their CF may also change the way the young person views themselves. Two sub-themes were identified within this theme. They were:

- 1. Relationships with Those Close to the Young Person
- 2. Personal Social Awareness

5.7.4.1. Relationships with Those Close to the Young Person

Throughout their life young people with CF spend a large amount of time with their care team. Most of the young people believed it was beneficial to have a good relationship with their care team and, on a whole, this appeared to be the case. Having a positive relationship with their care team allowed the young people such as Y25, to feel comfortable in approaching them with questions about CF.

"I'm quite open and the CF team I have got are very, they are very good... you feel like you can just speak to them and tell them what's going on... if I have anything going on, I know I can give them a ring or go down and they'll sort it for me, so it's good." (Y25)

As the young people get older they develop new relationships. Some of the young people included in this study were thinking about the potential complications their CF may bring to their relationships, for instance having children. Most young people were aware that they may have difficulties with fertility but that options were available to them should they wish to have children. Their children potentially inheriting CF appeared to be more of a concern for the young people.

"when I'm older if I ever did want to have children or anything it's definitely something... it's a conversation you would need to have, it would be reckless not to and it would just it would be difficult to know if the person you were with was a carrier or something like that because then that would obviously mean your child probably would have CF and that's kind of more of a worrying thing and it doesn't worry me all the time but it is something that I'm kind of aware of the older I'm getting" (Y26)

CF placed a strain on relationships between the young person and their immediate family, particularly parents. The concern parents had for their child's health could on occasion cause parents to get irritated or frustrated with the young person.

"every now and then if we forget to do our physio my parents can get quite defensive whereas we don't attack back at them because we understand why they're being defensive" (Y13)

The young people did not believe they had a strained relationship with their siblings, either believing them to be closer or 'typical'. Having another person in their immediate family with CF that understood what they were going through allowed young people to become closer to their brother or sister with CF. Young people generally believed their relationship with siblings without CF were as would typically expected between siblings.

"my older sister the one that has it I can really relate to her a lot her situation and I back her up cause I know how she feels and so on whereas my younger sister it doesn't really change anything she never really asks she's gotten used to it...she's quite normal with it really" (Y13)

Relationships with peers were also affected by the young person's CF. A small number of young people had experienced bullying related to their CF at school.

"I was quite bullied at school... People are not nice to people that are different at school. Like people with invisible like illnesses" (Y24)

A few young people were hesitant to tell their peers about their CF as they were concerned that their peers would treat them differently if they knew about it. However, once the young person told their friends about their CF they were generally understanding and supportive.

"most of them were really supportive and like understood and they didn't change the way they thought about me, they like treated me the same which was what I wanted, that was what I was scared of really...It was really nice to like know they had... they were supporting about it and didn't treat me any differently" (Y19)

5.7.4.2. Personal Social Awareness

The majority of young people included in this study viewed themselves as relatively 'healthy' compared to others with CF. The young people frequently referred to how they believe their circumstances could be worse and that they were fortunate compared to others with CF.

"You could be sitting, hooked up to something 24/7 really. There are some people with cystic fibrosis which are, which is a lot worse than what I've got and the way I've got it now" (Y10)

"But as far as CF goes I'm, I think I'm generally considered kind of healthy. I've got sort of a more common variation it's... Doesn't affect me as much as some people" (Y21)

The young people saw themselves and their CF as highly individual. Seeing themselves as different from others with CF and also different to those without. Being different was not necessarily a negative for all the young people.

"It kind of makes you feel like, like you're different in a good way and so it has something that equals you out from other people. And it's like having diabetes for example, it kind of shows that, that everyone has their way of being different and I think CF is ours and people with diabetes, diabetes is theirs, and other people have theirs" (Y8)

A few of the young people acknowledged that other people had complications in their lives such as diabetes, that did not make them either better or worse off than the young person, just different.

"I mean everyone has issues but I think they probably just carry on like sort of deal with issues, just carry on whereas I feel mine is like an ongoing one" (Y31)

How others viewed the young person was raised as a concern for over half of the young people. This was particularly mentioned in relation to others seeing the young person taking medications. In more than one case the young person believed that others would think they were a drug addict if they saw them taking their tablets. Several young people were hesitant to take their tablets in front of other people which could have implications for their health if they choose not to take their medications.

"I find a lot of people like to judge as well. Especially if you are taking like pots of... like if I am taking 12 tablets at once everyone thinks I am like a drug addict or something sometimes which is sad" (Y24)

"when you are sitting there taking pills you don't want them to think you are a drug addict or something...you know, in front of all these people...you know you don't want them to think less" (Y20)

5.7.5. Theme 5: Emotions Relating to CF

The young people included in this study expressed mixed emotions about their CF. Many of these emotions were negative, however the vast majority of the young people attempted to remain positive when possible. Three sub-themes were identified in this theme. They were:

- 1. Negative Emotions and Self-Perception
- 2. Frustrations: Hospitalisations, The Perceptions of Others, and Limitations
- 3. The Forgotten Positives of CF

5.7.5.1. Negative Emotions and Self-Perception

Some of the young people had a much more difficult time with their CF and these young people typically experienced a greater number of more emotional challenges. Most of the young people had difficulties with their medications and how they made them feel, this was particularly true for the younger participants in the sample who had to take medication while at school. Young people also spoke negatively about the time they had to take away from activities they wanted to be doing to complete their treatments and take their medications.

"it does get you a bit depressed at times cause you know you want to do more stuff that, you know, anyone else would do but you can't do it, you know? ...in a way it kind of restricts some of the things you can do like if I wanted to be out most of the day you can't, you have to come back and you know do all your antibiotics and stuff like that, you know" (Y20)

Some of the medications prescribed for CF have side effects that can affect the young person's appearance. This can be detrimental to the young person's self-esteem, which is already a known vulnerability in those with CF (Bregnballe, Thastum, & Schiøtz, 2007).

"I'm on these antibiotics at the moment I have to wear 50-degree, 50 SPF sun cream because it's making my skin and my face really red. So, it's making me really self-conscious...I feel self-conscious of my looks because of it" (Y24)

How others treated the young person was often challenging for the young person. Some young people were anxious because they believed that others were hesitant to be friends with them when they found out they had CF.

"if I became friends with someone if they did have CF and it would maybe it would maybe be scary for me if I got to know someone like that in case they got ill or in case I had to do any additional things so I think for me I maybe feel like why would someone want to continue being pals with me or whatever if they knew which is ridiculous rationally speaking I know people wouldn't but there's that kind of that anxiety at the back of your mind I suppose" (Y26)

5.7.5.2. Frustrations: Hospitalisations, the Perception of Others, and Limitations

The young people experienced several frustrations as a result of their CF. Many of these frustrations and tensions were related to having to go into hospital. Negative emotions about hospital visits were not necessarily isolated to solely the time when the young person was in hospital.

"when I'm about to come into hospital I just, I know I am not right because I'm coming in, obviously I am going to have to have a PICC line and what not. It's just, I just get moody" (Y17)

The challenges and frustrations of being in hospital mentioned by young people included treatments, boredom, isolation, some negative experiences with staff, and having to missing school, university or work. Generally, the young people did not enjoy missing school, however it was the young people that were either going through exams or studying more intensively who particularly felt frustration at missing school or university. Pushing to continue attending school or work even when feeling unwell could put the young person's health at risk.

"I've got to be honest like, you know, when you're younger and you miss school and you don't think it's a big deal like but I think if I have to miss like a lecture at university now it would be a lot different [...] if you miss a lecture you, you do fall quite far behind. So, it would be one of those things where rather than miss a lecture I'll actually maybe try and force myself to go in even if I was ill" (Y25)

Due to the isolation young people experienced when staying in hospital the young people had lots of time to get lost in their thoughts. While in the hospital it is hard for the young person to move their thoughts away from CF. This forced focus on CF removes the option for young people to use avoidant coping techniques, which, as discussed previously, were commonly found and suggested to improve outcomes in young people and siblings.

"It's obviously hard, being in hospital. It's a funny place and you don't want to be there, but you have to be there. And it's a place where you have to concentrate on CF, and you have to concentrate on what your life is like in that sense, whereas normally your, my concentration is diverted away from CF" (Y27)

Only a few of the young people appeared to have an issue with their treatments but several expressed frustrations at their treatments interfering with activities and the need to wake up earlier than their peers. This early waking and additional burden on the young people often left them feeling tired and sluggish.

"I think overall having CF can be a bit tiring, just like cramming everything in. And the things that people talk to you and asking questions, it can just get a bit like kind of boring in a way, having to do it over and over again." (Y8)

Having to plan ahead and incorporate their medications and treatments into their daily regime was frustrating at times for the young person. This was particularly true when the young person wanted to travel or stay away from home for a few nights.

"you can't just be like 'oh I want to go to France' I have to then got to think about the medical stuff I have got to take with me and it just frustrates me and then in a way it just makes you think 'well, you know, what's the point, I'm going to pack all this stuff and then I've got to organise for it all to go through and stuff' and it's just annoying" (Y20)

A common frustration expressed by several of the young people was directed toward their school, and certain teachers with whom the young person had had a particularly negative experience. A lack of understanding and the invisibility of CF cause issues for the young person. Having someone the young person could trust at school or work helped the young person to feel less frustrated.

"I say to them [Teachers] "I need to use the bathroom, my lungs don't work properly" and yeah, and they're like "you're fine, I've seen you in school, you're fine, sit down" ... I get really annoyed and frustrated. And I just keep going on about it and then... I was telling one of the teachers about it and he said "if that happens just walk out of the classroom, if they shout at you I'll shout at them" (Y11)

One young person felt a further irritation from the invisibility of CF and also the issue of cross-infection in CF. These issues combined meant the young person was wary of other people as they could potentially also have CF, which could in turn make them unwell.

"You can't hardly tell if the child's got CF... I could actually walk up to somebody now they probably could have CF. They might probably have CF. But like, I have to be really careful though because like anybody in my street they could actually have CF right now so that kind of is actually is really annoying" (Y7)

5.7.5.3. The Forgotten Positives of CF

There were a small number of the young people who described positive aspects they found from having CF. The most frequently referenced positive of CF by young people was an increase in their confidence relative to their peers.

"I think it helps confidence...Because it's like, you have been through things that other people won't have been through so that boosts your confidence a little bit in my opinion" (Y15)

"I think it boosts my confidence quite a lot to be fair because it tells me that I'm not alone it shows me stuff like other people and how much they're suffering worse than me and it boosts my confidence knowing that I'm lucky to be born with such a smaller case than ..." (Y13)

Summary of Results

Theme 1: Communication and Support

- Young people with CF believed people generally have misconceptions about CF, which made them hesitant to talk about their CF
- Many young people were open to talking about their CF when they were asked direct questions about it
- How they brought up the subject of CF and the level of detail they provided others with was highlighted as an area of uncertainty
- Peer support was welcome, however young people were worried it could be discouraging and scary depending on the other person's health and experiences
- Young people were cautious over the information they received about CF. They highlighted some sources as trustworthy such as their clinical team and The Cystic Fibrosis Trust
- School textbooks and Google were highlighted as being poor information sources that were frequently out-ofdate

Theme 2: Controlling and Understanding Medications and Treatments

- Gaining independence by transitioning or moving away from home was generally viewed positively by the young people
- Some young people were less adherent to their medications and treatments when they left home
- Knowing who they were going to see and where following transitioning provided comfort to the young people
- The majority of the young people recognised the benefits of being active

Theme 3: Challenges and Coping Mechanisms

- Having to plan far ahead and organise their days around their treatments was annoying for many of the young people
- Having children in the future was not seen as too large a concern and something only to be considered when needed
- Treatments and medications were challenging for young people, particularly due to the permanency of their CF and the belief that the amount they had to do was only likely to increase
- Thinking about how CF would affect their future working ability was difficult for the young people
- Managing their treatments while in work, and the support available about this was also a concern for the young people
- Travelling with CF was mentioned by several young people as arduous due to their treatments and medications
- The young people coped with these challenges through either normalisation or avoidance

Theme 4: Social Aspects of CF

- Young people were concerned about how other people viewed them
- Relationships with siblings were viewed as "typical" brother sister relationships
- Young people didn't want their friends to treat them any differently, so were sometimes hesitant to tell them about their CF
- The young people saw themselves and their experiences with CF as highly individual

Theme 5: Emotions Relating to CF

- Young people were frustrated by missing school particularly when studying difficult subjects or going through exams
- Side-effects from medications were detrimental to young peoples' self-esteem
- Comparison to others could often make the young person feel down

5.8. Discussion

These results affirm and add to the current understanding of the experiences and psychological wellbeing of young people with CF. Interpreting the results within the existing research will help to illuminate the implications of this chapter for both young people with CF and their siblings.

Relationships between the young person and their immediate family were generally viewed as 'typical', particularly the relationship between the young person and the sibling. As many of the young people appeared to be in good health and had a parent figure caring for their medications and treatments, the young people were able to focus their attention elsewhere and avoid negative aspects of their CF. Avoidance coping in young people with CF has previously been suggested as a passive way for them to cope with the intrusion of CF in their everyday lives (Abbott, 2003). In keeping with the work by Abbott (2003) several of the young people spoke about their use of adaptive coping, where they felt they were able to incorporate CF into their lives through routine and acceptance. This was particularly apparent in relation to things the young people were unable to avoid and ignore, such as treatments, periods of illness, and limitations due to symptoms and side-effects.

The lengthy daily treatment routines were challenging for young people. Young people found it particularly difficult when their treatments disrupted their social or school/work life. The high treatment requirements of CF can often result in the young people missing social occasions (Jamieson et al., 2014), which was highlighted by many of the young people included in this sample. Furthermore, many of the young people were self-conscious of having to take medication in front of friends and colleagues. This could lead to non-adherence, which could be problematic for the young person's health (Bregnballe, Schiøtz, Boisen, Pressler, & Thastum, 2011).

The young people regularly noticed how others viewed them and this affected them in several ways, including a reluctance to take medications and treatments in front of others, which could have long term ramifications for their health. Young people with a chronic illness or condition viewing themselves as different from their peers without chronic illnesses or conditions has been found before and can cause issues with the social functioning in young people (Meijer et al., 2000). Meijer et al. (2000) found that the diagnosis of the young person did not relate to their level of social functioning but rather all young people with a chronic illness or condition were more withdrawn and shier compared to their peers. Many of the young people in this sample appeared to be shy and withdrawn when they believed that others lacked understanding or held misconceptions about CF. More generally, in agreement with siblings, the young people believed that others had a poor understanding of CF. How others misunderstand CF and the associated myths relates to the availability and accuracy of information (Rubin, 2014). Concerns about the information available about CF were raised by young people.

As their understanding of CF improved and the potential for CF related complications increases (Cohen-Cymberknoh, Shoseyov, & Kerem, 2011) the young person's concerns for the future intensified. In particular the young people showed concerns about the permanency and progression of CF. The majority of young people were concerned about the increasing amount of medication they were having to take and the potential side-effects the medication may have. When asked about the future the young people acknowledged that it was likely to come with challenges for them that they could require additional support with. For instance, those moving to university or into work were concerned about maintaining their treatment routines in a sustainable way. Little is known about how young people feel about these challenges as much of the existing literature on aging in CF focuses solely on the transitioning period from paediatric to adult care services (Brumfield & Lansbury, 2004; Dupuis, Duhamel, & Gendron, 2011). Potentially due to this focus the process of transitioning did not appear to be a large issue for the young people who were going through or had gone through the process of transitioning. Many of the young people viewed the move to adult services as positive and looked forward to their increasing independence. There was a small number of the young people who were a bit older and had transitioned at a time when the services may have been lacking compared to current provision.

5.9. Implications for the Sibling Experience

Although there was no direct question on siblings included in the young person interview schedule, interviewers would often ask about the young person's relationship with their sibling or the young person would choose to talk about their sibling. When asked about their siblings the young people often mentioned that they thought their relationship was typical. Many referenced having the "typical brother/sister" problems. Some young people recognised their sibling was likely to have experienced some impact due to the amount of time their parents had to spend with them, but it didn't seem to be a topic they had openly discussed.

The young people sometimes acted out at home when frustrated with their medications or treatments, this could be distressing for the sibling in both terms of family tension and their brother or sister being distressed. Further, many of the young people spoke about being self-conscious when taking their medications in school, and this could potentially lead to non-adherence, which may worry the sibling. Siblings with a brother or sister with a chronic illness or condition have been noted to find it challenging when their brother or sister has social difficulties (Floyd, Purcell, Richardson, & Kupersmidt, 2009). Poor understanding and misassumptions about CF were mentioned by both the young people and the siblings. The idea of others holding misassumptions made young people more hesitant to talk about their condition. Siblings were also hesitant to discuss CF but because they saw it as a personal subject for their brother or sister. As such, the openness of the young person with CF may relate to how comfortable the sibling is talking about CF and the impact it has on their own life.

5.10. Conclusion

The young people with CF appeared to adapt and cope well to a life with CF. This is generally in agreement with the existing literature (Ernst et al., 2010; Havermans et al., 2008). Despite the progressive and permanent nature of CF, the young people appeared relatively unaffected, which may relate to their generally positive experiences with both formal and informal support. The young people were happy with the support they had received from their clinical team and their immediate family. The young people had limited interactions with third party support services such as The Cystic Fibrosis Trust, although perceptions of the information and fundraising they provide were positive. As with siblings, one of the most positively viewed ideas for additional support was peer support. Reservations about peer support included difficulties with speaking to others online and potentially speaking to others in the worse or better health than themselves.

Chapter 6: Results - Adults with Cystic Fibrosis

6.1. Introduction

Cystic fibrosis (CF) is a progressive condition, however, thanks to improvements in medications, treatments and diagnosis the life expectancy of those with CF continues to rise (Stephenson et al., 2017). As the epidemiology of CF continues to develop and change the understanding of the experiences and impact on adults is limited relative to the understanding of young people with CF (Habib et al., 2015). Family relationships can be some of the longest experienced by an individual and it is likely that the impact of a chronic illness or condition continues to be felt within the family into adulthood (Shepherd et al., 1990). Several of the siblings included in Chapter 4 had an adult brother or sister with CF and therefore, it is important to consider the perspective of adults with CF also.

6.2. Background

As the expected survival age for those with CF continues to increase it has been recognised that there is a need to further delineate the experiences of adults with CF and their support needs. As they move into adulthood (25 year + (Sawyer et al., 2018)) those with CF are at an increased risk of complications such as CFRD, liver problems, and the need for transplant (Cohen-Cymberknoh et al., 2011). Adults with CF also have increasing treatment demands as new medications and treatments tend to be additional rather than replace existing treatments for those with CF (Sawicki, Sellers, & Robinson, 2009). A review considering the Health-Related Quality of Life (HRQoL) of both adolescents and adults with CF highlighted a significant association between age and perceived treatment burden following adjustment for several potential external factors including BMI, sex, and frequency of pulmonary exacerbations (Habib et al., 2015).

The stage of life when individuals move from adolescence into adulthood is associated with several developmental and social challenges that can cause psychological distress even in those without a chronic illness or condition (Sawyer et al., 2018; Zarrett & Eccles, 2006). This is further complicated by the presence of CF (Hamlett, Murphy, Hayes, & Doershuk, 1996). As with the research on sibling and young people with CF, the evidence on the psychological wellbeing of adults with CF appears to be mixed. One study in particular reported high rates of depression in adults (Riekert et al., 2007), whereas another suggested that adults experienced similar levels of mental health symptoms compared to the norm (Cronly et al., 2018). In their review on the HRQoL of adolescents and adults with CF, Habib et al. (2015) collated the evidence and found a negative correlation between age and emotional functioning (correlation coefficients of -0.23 to -0.28), and age and health perceptions (correlation coefficient -0.22), although these relationships were no longer statistically significance following adjustment for several externalities. As stated previously, the health of those with CF tends

to deteriorate with age and as such they are at risk of more frequent periods of ill health, which has been linked to poorer mental health outcomes (Riekert et al., 2007).

Relationships both within and outside the adult's family are complicated by the presence of CF in adulthood (Pfeffer et al., 2003). Family dynamics typically change greatly during the period of adolescence rather than adulthood. Transitioning from paediatric to adult care services is regularly completed by the time the individual is 18 years old and during this period family dynamics go through a large shift (Dupuis et al., 2011). As the individual progresses into adulthood they become highly independent with their medications and treatments and parents can feel as though they are losing control (Akre & Suris, 2014). As the focus shifts to the future for the adult and their health there are new points of stress and anxiety that can cause difficulties in family relationships. Although there is currently a limited understanding of this it has been found that the time when awaiting a transplant is associated with psychological difficulties in families of those with CF (Kurland & Orenstein, 2001).

Relationships with peers are also challenged by CF in adulthood. Adults with CF have been reported to experience difficulties with romantic relationships (Pfeffer et al., 2003). Relationships may suffer as adults with CF are resistant to let others in, as the idea of putting others through the difficulties they experience is linked to high levels of anxiety in adults (Pfeffer et al., 2003). There is a focus in the literature on understanding how issues with fertility and the potential for genetic inheritance alter adults' relationships and views of the future (Fair, Griffiths, & Osman, 2000). The use of carrier testing, for instance, has been considered in both parents and adults with CF, and a large proportion of those at risk of passing on the affected gene show a high willingness to use prenatal diagnosis, although the decision of aborting for CF is much less accepted (Henneman et al., 2001).

In the literature considering adults and their future there is also a recognised need to better understand how CF affects an adult's working life. Adults with CF want to be in work and are more likely to be actively employed than their peers, even when awaiting transplant (Burker, Carels, Thompson, Rodgers, & Egan, 2000). However, it is also recognised that CF interrupts adult working life in several ways including the potential for stigma in colleagues (Burker, Sedway, Carone, Trombley, & Yeatts, 2005). What appears to affect adults to the greatest extent is the treatments and medications that they are required to administer during working hours, for instance individuals with access devices were more concerned about their careers (Gee et al., 2005).

Adults can also have regularly difficulties with adhering to their treatment and medications (Williams, Mukhopadhyay, Dowell, & Coyle, 2007). In the study by Abbott, Dodd, Bilton, and Webb (1994) not completing treatments (not adhering) was connected to the adults perception of the benefits of adhering; adults who saw an immediate benefit following exercise were more likely to be compliant to their exercise routine. It is important for adults to adhere to their treatments and medications to maintain good health, as such promoting adherence has been suggested to be a vital

component of the adult's care. The development of support to address adherence has been recommended (Kettler, Sawyer, Winefield, & Greville, 2002).

Given the above literature and the increasing proportion of those with CF that are now living into adulthood, there is a need to better support adults with CF. To do this effectively the experiences and psychological wellbeing of adults with CF needs further consideration (Cronly et al., 2018). Given the limited understanding of the experiences of adults themselves there is also a limited understanding of the siblings of adults with CF (Wennström, Isberg, Wirtberg, & Rydén, 2011). Gathering evidence from the adults will enhance the understanding of the sibling experience.

6.3. Aims and Objectives

As in both Chapter 4 and 5, consideration was given through this chapter to the overall aims of the qualitative project as presented in section 3.2.10 in Chapter 3. The purpose of this chapter was to consider the experiences of adults with CF and the implications for their immediate family. To address these aims the following research questions were proposed:

- 1. How do adults with CF view the impact of CF on their lives? Specifically, do they identify an effect on their and their immediate family's psychological wellbeing?
- 2. Do adults with CF believe they and their family are being well supported? If not, are there areas where they believe support could be beneficial?

6.4. Recruiting Adults with CF

The majority of adults were sourced from a hospital in Glasgow (n=16), with the remaining recruited at clinics in London (n=3), Cardiff (n=1), Manchester (n=1) and through online advertisements (n=4). A further four adults completed a QoL measure only. The contact information was received for a total of 54 adults with CF, of which 55% were reached. The remaining were not contacted as data saturation was reached. The majority of the adults participated without their families. Of the 25 adults interviewed, only one had a single parent included in the study and one had two parents included in the study. As with young people, the recruitment of adults was far easier than the recruitment of siblings and the target was reached quickly.

6.5. Data Collection and Analysis

All data collection and analysis were coordinated by MMS with the support from other researchers, CL, NK, LA, JL and RS. All participants provided informed written consent prior to participating. All interviews with adults were completed over the phone. A QoL measure was completed by all adults, this was done either over the phone or where this was not possible through the post. Adult interviews were largely conducted by NK, with the remaining completed by MMS (n=10). Interviews with adults lasted on average 39 minutes. All adult interviews were transcribed verbatim by NK, and due to a high familiarity with the data initial code production was also completed

by NK. All transcripts were reviewed by MMS and code checking was also completed by MMS. The production of themes was an iterative process conducted by MMS and NK.

Following the initial code development, a short summary of the results were built for each participant to review and sent to them via confidential email or in the post. Participants were asked to respond with comments should they feel that the themes were not representative of their experiences. One adult responded with comments, which were incorporated into the analysis, therefore it is assumed that the themes presented below met with the adult's expectation and true experiences.

6.6. Participant Demographics

Adults ages ranged from 26 to 52 years (median = 34). Fifteen were male, while the remaining 10 were female. Seventeen adults worked either full or part time, one was a student, and six were unable to work due to their health. All adults were also of white ethnicity. Further demographics are shown in Table 6.1. below. Only a few of the family members of adults participated (four siblings and three parents), therefore there is a large amount of missing data for adults. For instance, household income was only given for two adults and is, therefore, not shown in the table.

Adult	Age	Sex	Marital Status	Employment Status	Household Income	Sibling	Parent 1	Parent 2
A1	25	F	With a Partner	Employed Full/Part Time		S17		
А3	26	M	Single	Employed Full/Part Time				
A4	27	F	Single	Employed Full/Part Time				
A5	27	F	Single	Student				
A6	28	F	Single	Employed Full/Part Time				
Α7	27	M	Single	Out of Work		S19		
A8	28	M	Single	Out of Work				
A9	29	F	Married	Unable to Work due to Health		S6		
A10	29	F	With a Partner	Employed Full/Part Time		S18		
A11	30	M	Single	Out of Work				
A12	31	M	With a Partner	Employed Full/Part Time				
A13	31	M	With a Partner	Employed Full/Part Time				
A14	31	F	Married	Employed Full/Part Time	50,000+		P15	
A15	32	M	With a Partner	Employed Full/Part Time				
A16	31	M	With a Partner	Unable to Work due to Health				
A17	34	F	Married	Employed Full/Part Time				
A18	35	F	Married	Employed Full/Part Time				
A19	35	M	With a Partner	Unable to Work due to Health				
A20	36	M	With a Partner	Employed Full/Part Time				
A21	36	M	Married	Unable to Work due to Health	15,000-25,000		P17	P16
A22	37	F	Married	Employed Full/Part Time				
A23	37	M	Married	Employed Full/Part Time				
A24	45	M	Married	Employed Full/Part Time				
A25	48	M	Single	Employed Full/Part Time				
A26	48	F	Married	Employed Full/Part Time				
A27	49	М	With a Partner	Unable to Work due to Health				
A28	50	M	With a Partner	Employed Full/Part Time				
A29	50	F	Married	Employed Full/Part Time				
A30	52	F	Divorced/Separated	Unable to Work due to Health				

Table 6.1. Adults with CF Demographics

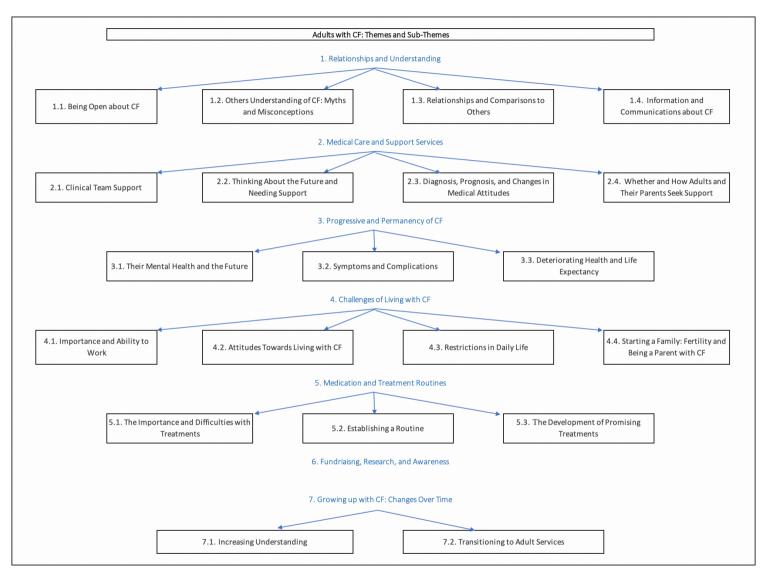


Figure 6.1. Themes and Sub-themes of Adults with CF Interviews

6.7. Themes

From the qualitative thematic analysis conducted on the 25 adult interview transcripts, seven themes were identified:

- 1. Relationships and Understanding
- 2. Medical Care and Support Services
- 3. Progressive and Permanency of CF
- 4. Challenges of Living with CF
- 5. Medication and Treatment Routines
- 6. Fundraising, Research, and Awareness
- 7. Growing up with CF: Changes Over Time

Each theme contains multiple sub-themes. The themes and sub-themes are given in rank order of reference density and displayed in Figure 6.1. Each theme is discussed below with verbatim examples taken from transcripts.

6.7.1. Theme 1: Relationships and Understanding

Adults suggested that awareness of CF had improved. Although they believed that knowledge, understanding, and attitudes to those with CF still varied significantly. Within this theme four subthemes were identified:

- 1. Being Open about CF
- 2. Others Understanding of CF: Myths and Misconceptions
- 3. Relationships and Comparisons to Others with CF
- 4. Information and Communication about CF

6.7.1.1. Being Open about CF

Many of the adults were comfortable discussing and explaining their condition to others but there were a few adults that chose to share information only if asked a direct question about CF. Several adults felt that the information was personal and didn't want to provide more details than necessary.

"I don't talk ... volunteer information, you know if they ask then I haven't got a problem saying anything... I wouldn't say embarrassed about it or anything it's just, you know it's just one of them things" (A27)

There was a small number of adults that felt very uncomfortable talking about CF and rarely shared information on their condition, particularly with people they were not familiar with.

"I just I kind of find it difficult to tell people, like you know I don't know when to bring it up, like you know do you do it in the first date do you do it in, do you just kind of randomly drop it in like "oh by the way" (A5)

As with young people many of the adults were unsure of how best to explain CF to others. The way that they approached explaining CF to others differed although typically they offered a simple and brief explanation with very little personal detail.

"it's not something that I go out of my way to tell people. If someone asks me about it, or they ask me about my cough, or whatever, I just say I've got cystic fibrosis. And if they don't know what it is, I kind of give them, you know, a very brief you know, idea of what it is" (A11)

Many adults struggled to share the true extent of their difficulties, even with those close to them. Adults generally believed their relationships with loved ones were unaffected by CF. However, the adults tried to protect those they were close to by not telling them the full extent of their problems. Many of the adults felt guilty sharing the impact CF had on them because they understood how it distressed their loved ones.

"it makes them [parents] so upset when I've told them that I am feeling bad so I have just to hide that, well I choose to just hide that" (A14)

6.7.1.2. Others Understanding of CF: Myths and Misconceptions

Contrary to what was found in the previous chapters considering siblings and young people, many of the adults felt that the general public were typically aware of CF. Even if they were not familiar with all the details adults thought that others would have at least an idea of CF.

"Most of them I don't actually have to explain it. I'll just say I've got cystic fibrosis, and they'll at

least have heard of it" (A11)

Despite adults believing there was good awareness of CF they did not believe this translated into a good understanding. The exception adults believed was others that were close to someone else with CF.

"people are probably familiar with the logo and the CF Trust but actually unless you've been got some sort of personal connection to it people wouldn't know what it is" (A12)

Some adults spoke about the reactions of other to finding out about their CF. Several adults also shared their feelings and experiences of explaining CF to others. Most adults believed that because CF is an invisible condition, many people could be judgemental or hold misconceptions about the disease.

"My recent experience I think is my neighbours don't easily understand why on earth I would be thinking about retiring, you know. I think there's a lot of people don't know what CF and what it does [...] And it's probably a pretty good example of one your invisible illnesses." (A21)

Even once others are aware of what CF is, many adults thought that the invisibility of the condition can prevent them from truly comprehending what it is like to have the condition.

"I think it's probably very difficult to understand actually what it is like, you know in the sense that you know some people can completely over react to it and some people you know, probably don't really understand the impact it has on you or it can have on your life you know." (A3)

6.7.1.3. Relationships and Comparison to Others with CF

Many of the included adults felt "lucky" compared to others with CF. The adults believed each person with CF had different experiences.

"I'm aware everybody's totally different, different mutations, different treatments, different"
(A21)

The possibility of talking to others with CF about their experiences interested many of the adults. Several of the adults remembered being able to meet others with CF face-to-face prior to the guidance on cross-infection being released. Adults were not overly enthusiastic about the use of online forums, such as those offered by the Cystic Fibrosis Trust. However, they were unable to identify a more suitable way to talk to other with CF.

"people tend to go there [forum] when they have a problem or when they are really sick and bored at hospital and not when they are happy and I think it is a difficult space" (A4)

The varying presentations and severities of CF meant that several of the adults had had friends with CF who had passed away. The adults typically found this very difficult and as a result many tried to avoid talking with other people with CF.

"Especially if I start seeing other people that are like ill and that, needing transplants or are dying or whatever, it just makes very, very depressed, often for days [...] and these forums used to do that to me, so I just said ... and I wasn't even actually engaged in them, this was just reading them. It's sad to say that I don't read anything" (A11)

6.7.1.4. The Information Available Online

CF being a highly individual condition was seen as a barrier to understanding by many adults. Adults believed that much of the information available online did not explain the variations that can be found in those with CF well.

"You can Google cancer, yeah but there's so many different types [of CF] and it affects people differently" (A27)

There were several adults that believed the information available online about CF was unreliable. There were also other adults that thought people would worry when they read information on the internet and felt that they had to deter people from researching it online.

"I also find that most of the time people sit down and look it up they completely panic in fact like every time I go out with someone or like all of my previous boyfriends I always had to say like right so

I have CF, don't google it" (A4)

6.7.2. Theme 2: Medical Care and Support Services

Adults were found to be generally happy with the support and medical help they had received. Within this theme four sub-themes were identified:

- 1. Clinical Team Support
- 2. Thinking About the Future and Needing Support
- 3. Diagnosis, Prognosis, and Changes in Medical Attitudes
- 4. Whether and How Patients and Their Parents Seek Support

6.7.2.1. Clinical Team Support

The majority of adults were very pleased with the support and information they had received from their clinical team. The clinical team appeared to be, as with young people, the key resource for support and information for adults. Clinical teams often filled a space where adults did not know of an alternative support option.

"if [son] wants, if he's got questions and if he wants to come in they [consultants] will sit with him and chat to him if he wants to, you know they've been really great, but there's only so much, you know there's just not, there's just nothing out there really" (A22)

Adults with CF have a multidisciplinary care team as standard, however, a number of the adults still described how they believed a specialist CF team and in particular a source of psychological support was important to them.

"I've got a very good psychologist team in my, in the unit I go to... If you want to get upset or talk to somebody in-depth about things it's a nice support network to have. I think that's helped me tremendously really" (A24)

6.7.2.2. Thinking About the Future

Many of the adults discussed their thoughts about the future and support they believed they may need. In particular, adults spoke about improving support for parents with CF, travel insurance

costs, and access to sports. This is particularly important given the increasing survival age of those with CF and lack of understanding of the experiences of this aging population.

"...it's so new, yeah absolutely and I think there is a lot of research lacking in the older adult areas because you know, yeah, it's, it is new. You know what do you do when you have to retire, basically almost retire? You know I stopped my work at thirty-five. Nobody else I know stopped work at thirty-five and I joined the daytime book club and everyone was over sixty-five and they were lovely and they were welcoming and I really enjoyed it but you know how do I stay thirty-five when I've actually got the body of somebody much older than me" (A22)

Adults recognised that there were services, such as those available from the Cystic Fibrosis Trust, that were currently underutilised and undervalued. Some adults suggested ways in which this could be improved.

"when you go into your CF clinic there is zero Cystic Fibrosis Trust material, you know like, you know pamphlets, leaflets. You know advocacy information or just like contact information. None of that is visible at all... When that could probably be a very useful tool. Maybe the CF Trust could think about doing something like that, making those resources available in the clinics" (A13)

6.7.2.3. Diagnosis, Prognosis, and Changes in Medical Attitudes

The adults acknowledged that changes in medications and treatments over the years had helped improve their health. The majority of participants talked about their initial prognosis being quite negative and described how medical attitudes and care had changed over the years. For instance, several had been allowed to meet others with CF before the issue of cross-infection was identified.

"I am so thankful that we had that opportunity because that isn't the case now. I completely understand why we can't mix, but I do think it's a huge missed opportunity" (A22)

A few of the adults included in this sample received their diagnosis of CF late. Early diagnoses are known to improve outcomes (Castellani et al., 2009), however, multiple adults included in this study received their diagnosis in adulthood. A late diagnosis can cause emotional difficulties in those with CF and their families (Mérelle et al., 2003; Szyndler et al., 2005). Receiving a late diagnosis can come as a surprise to those who had not realised they had the condition for many years.

"They said well because it's a genetic thing they said "well maybe we should, test you as well".

so, they did test me and then it was yes, you've got cystic fibrosis. But otherwise I wouldn't have

known, so, until probably later on" (A48)

For many of the adults receiving a diagnosis was a relief as they could finally understand the symptoms they experienced and they were then able to access the medical care they needed.

"It wasn't so much a struggle to be told it was cystic fibrosis. In a way, I was relieved that they'd found something, you know, that I did get a diagnosis, and it was over, I felt once I was transferred to the team I was actually getting the medical help that I needed" (A30)

6.7.2.4. Seeking Support

Amongst adults there seemed to be a general lack of awareness regarding the available support services. Many adults mentioned that they were not sure where to look for support or what there was available.

"...my problem... I couldn't identify exactly what services were available" (A13)

There also appeared to be a resistance in many adults to seek support, even when offered guidance and the options for referral.

"she can maybe refer me to the GP, to maybe see if we could get someone more local to speak to me, but I haven't really pursued it... I've just been kind of procrastinating and I'm not really sure if it's something I want to start doing again" (A11)

As with young people and sibling engagement with support services and third-sector charities largely seemed to be through the adult's parents.

"oh yeah, yeah, my mum's in to like all that group stuff and she pops into all that" (P46)

6.7.3. Theme 3: Progressive and Permanency of CF

CF is characterised by various medical symptoms and a deteriorating health. This poses many restrictions for participants, such as feeling constantly tired and some needing transplants. The progressive nature of CF is known to negatively affect the mental health of adults with CF (Riekert et al., 2007). Within this theme three sub-themes were identified:

- 1. Their Mental Health and the Future
- 2. Symptoms and Complications
- 3. Deteriorating Health and Life Expectancy

6.7.3.1. Their Mental Health and the Future

Most adults talked about the negative impact CF had on their psychological wellbeing including feelings of anxiety, low mood, hopelessness, self-harm, and a distrust of their medication. The most common area that was raised as an issue for adults' mental health was the future and an inability to feel excited about it.

"it is having quite a big psychological impact on me now [...] just probably inability to look to the future any more... I don't feel that I have a long future anymore so I don't, it doesn't feel exciting to look ahead it feels frightening" (A14)

A smaller number of adults talked about positive impacts CF had on their wellbeing and their attitude towards life. For instance, adults believed they were able to perceive things differently and appreciating life more compared to others without CF.

"CF has helped me to be happy I think I appreciate things and I am very clear about what I want...
because I guess I know that my life is mostly likely to be a little bit shorter than most other people's
and because I know that I have to struggle to get where I want to be, that gives you a different
appreciation of time and life ... I suppose" (A4)

Although it appeared that a negative mental health in adults was due to thoughts of the future it is also possible that a negative mental health in the present made it more difficult for adults to think of the future.

"I think I am excited for the future, I think the challenge for me is my mental health perspective is something that I can struggle with quite significantly and that's a broader issue in terms of you know trying to find the joy in things and you know suffering from bouts of depression essentially and anxiety" (A12)

6.7.3.2. Symptoms and Complications

Concerns about the side effects relating to CF were far more apparent in adults than young people. Adults appeared to particularly struggle with feelings of exhaustion.

"just worn out and tired and feeling physically um that I just it feels like a massive uphill like the steepest hill in front of you... and you have no choice but to just keep trying to claw up just yeah very very hard" (A14)

As with young people, adults frequently compared themselves to their healthy peers. Many adults spoke about how their CF symptoms prevented them from being able to do things others without CF were able to do.

"I can't do as much as my peers, friends, especially with my son, like taking him to the park and play football, I can't do much when I'm there, I just kind of stand about, I can't run around, you know, and play" (A23)

Transplants were recognised as a stressful experience by adults. However, previous literature suggests that adults with CF are well adept and able cope with the stress (Burker et al., 2000). The adults in this study who were awaiting a transplant did appear to struggle. Adults that had already had a transplant expressed how grateful they were for their transplant and how they viewed it as a second chance.

"Being honest I think about the transplant every single day, you think about the phone call...

Sometimes it makes you happy, it was a big occasion to be honest. You think about the bad times before it and you say, thank God... I had CF lungs for 43 years and so that's, I've got a chance at something else. That's the way I look at this" (A25)

6.7.3.3. Deteriorating Health and Life Expectancy

The negative prognosis of CF caused several of the adults' distress. The fact that this was out of their control and the knowledge that they have a permanent incurable condition is known to cause mental health difficulties in adults (Riekert et al., 2007). Many adults struggled with their deteriorating health and accepting the progressive nature of their condition meaning that their health is unlikely to improve drastically.

"The biggest struggle I think personally is the mental aspects of knowing that it is a progressive condition, knowing I could not be very well today and I could be better tomorrow if I did some airway clearance or some exercise or do my treatments really well so I can get a bit better compared to how I am, but I can never be as good now as I was when I was you know 16 for example so knowing that it is a progressive condition" (A15)

"I feel like, in a way like having a terminal cancer diagnosis but you are doing the chemotherapy to see how long you'll get so because I am getting more and more ill and I know that I feel it all the time in my everyday stuff I can't do [...] so I don't feel like the treatment is controlling it is just like dampening a fire" (A14)

6.7.4. Theme 4: Challenges of Living with CF

Several of the daily challenges adults with CF experienced have been touched upon in the previous themes. All of the adults included in this study agreed that they faced challenges relating to their CF on a daily basis. Within this theme four sub-themes were identified:

- 1. Importance and Ability to Work
- 2. Attitudes Towards Living with CF
- 3. Restrictions in Daily Life
- 4. Starting a Family: Fertility and Being a Parent with CF

6.7.4.1. Importance and Ability to Work

The majority of adults talked about the importance of being able to work. Many adults mentioned that their CF affected their ability to work, the type of work they could do, the quality of their work, and their potential for career progression. Having an understanding and supportive work environment was important for several of the adults. One of the most common concerns and difficulties faced when adults looked for employment was that people would not understand. Finding

a boss that was understanding and accommodating made it much easier for adults with CF and reduced the impact CF had on their work life.

"I think initially when I was looking for work there was challenges I think people weren't as understanding as what this job that I have got now they are very understanding, I think that maybe the biggest challenge for me" (A6)

"he [manager] was understanding he also didn't penalise me for not being able to do all the hours...any hours I was already scheduled to do, I was still getting paid for...I didn't suffer financially just because, I ended up in hospital" (A13)

6.7.4.2. Attitudes Towards Living with CF

Many of the adults viewed CF as part of who they were. Adults tried to accept CF in their lives to try and reduce how much it interrupted their lives. Only a small number of the adults allowed themselves to have an exclusively negative view of CF. The majority of adults tried to maintain a positive outlook.

"it takes a little while to get used to but of course at the end of the day what can you do? It's there, it's ... so you just sort of get on with it and accept it at some point, you know" (A27)

"I think the best thing I can say about having CF is not to let it get you down. I spent years thinking that my condition would hold me back. People with CF are capable of anything. It's important for younger people with CF to realise that it's not a hindrance, as much as it's a pain, and it may seem a hindrance at time. Having CF can make you stronger. You fight more to survive" (A16)

6.7.4.3. Restrictions in Daily Life

Several daily activities completed by the adults such as driving, doing house chores, travelling, and working were disrupted by their CF. When the adults gained independence with their treatments, such as when they go to university, the daily impact of CF increased. At this stage adults with CF were better able to distinguish between themselves and their peers without CF. Adults compared themselves to others in terms of their work life, social life, family life, and general health.

"I still go out from time to time, but you know, it's like it is a pain to kind of like ... whenever I'm going out with friends, I have to kind of know at the time where we're going and carry it all... you know, I'd have to bring all my treatments forwards, in order to go out for a night out, and things like that" (A11)

"it was just when I was with my friends and stuff and doing the course I had to leave... for IV for two weeks at a time, so I was missing two weeks of the courses, I just couldn't catch up" (A8)

6.7.4.4. Starting a Family: Fertility and Being a Parent with CF

Many adults talked about fertility and having children. The adults highlighted a lack of support and information on the subject and how that affected their decision to start a family.

"well I guess there is a few things that are of interest to me specifically at the moment thinking about you know starting a family not too long away" (A12)

Those with children talked about being a parent with CF, the challenges they faced and worries they had regarding their children. In particular, the adults were worried about how their children would be affected by the adult's CF and about the lack of support for parents with CF.

"[...] and I can't meet up with other families that have CF because I can't be near them so I can't say to him you know look at these people or why don't you ask them how they're dealing with or let's read this book together about mummy has you know there's nothing there. So, it's been up to me to sort of manage it really" (A22)

6.7.5. Theme 5: Medication and Treatment Routines

All the adults spoke about the need to have a treatment routine. Several of the adults struggled with their medications and maintaining a routine. Within this theme three sub-themes were identified:

- 1. The Importance and Difficulties of Treatments
- 2. Establishing a Routine
- 3. The Development of Promising Treatments

6.7.5.1. The Importance and Difficulties of Treatments

The vast majority of the adults recognised the importance of completing their treatments and taking their medication. There were several difficulties that made maintaining treatment routines challenging. One such issue, which was similarly expressed by young people, was a concern for the side-effects from medications, in particular effects on their appearance. Negative side effects discouraged the adults from continuing with their treatments.

"when I was on steroids I felt absolutely horrendous I had real bloating and weight... I felt like I didn't want to leave the house I didn't want anyone to even look at me. I wouldn't go back on them even if it was the only medication that would save my life I wouldn't go back on them because I didn't want to exist when I was on them" (A14)

Beyond the more immediate side effects adults were also worried about the long-term effects of taking medication for such a long period of time.

"something that occasionally nags in my head is you know I've been on antibiotics every bloody day of my life for such a long time. Now we, you know research or whatever has shown that that's okay up to a point, but what are going to be, what are the impacts of all of these different medications going to be long term?" (A22)

As all of the adults had full responsibility for their care they also had difficulties with practical issues. For instance, some had difficulties with maintaining a supply of their medications

"I do have problems with the pharmacy sometimes they don't order enough of the stuff or they ...
get it wrong somehow and then just being like shit I am not going to have enough pulmozyme if I go
away and that definitely means I am going to spend some time in hospital" (A4)

All these difficulties can lead to disengagement and a lack of enthusiasm in adults who have tried to maintain their treatment routines. This is likely to have a negative effect on their health.

"I used to do all the medication, I guess I was thinking yes it will help keep me well, it didn't seem to make any difference, so I ended up in hospital, come out of hospital didn't do my treatments still ended up in hospital so I was like yeah no matter what I do with it don't work" (A19)

6.7.5.2. Establishing a Routine

Integrating medications and treatments into everyday life by establishing a clear routine allowed adults to normalise the presence of CF in their lives. Establishing a routine was challenging for adults with CF for several reasons including ever increasing treatment demands. As individuals with CF get older the amount of medications and treatments they require is only likely to increase (Sawicki et al., 2009). Adapting to additional treatments was difficult for adults, although given time and support they generally managed to adapt.

"I started off with like one or two treatments and it wasn't taking up a lot of time, it was fine. But then gradually introduced one here and one there over the years and you sort of, you get to a point when you just integrate them into your routine and it just becomes a thing that you do" (A20)

6.7.5.3. Thoughts on the Development of Promising Treatments

Many of the adults included in the study were optimistic about the future of medications and treatments for those with CF.

"I think with all the research and all the new drugs are coming out now as well I think the future's a lot brighter for people with CF than it was, there's more hope now than ever before" (A29)

There were still several adults that remained sceptical of the development of new treatments and the chance of a cure. For many of the adults a cure had been discussed as a possibility for a very long time.

"it feels like it's all just around the corner. It feels like it's really exciting. It feels like there's a lot that might be happening but it always, it, I remember so clearly sitting...I might have been thirteen and my consultant saying to me and my they've identified the gene, I think there'll be a cure by the time you're fifteen, so that's fifteen and I'm now 37" (A22)

6.7.6. Theme 6: Fundraising, Research, and Awareness

Many of the adults saw the importance of being involved with research and fundraising. Despite issues with cross-infection, which prevent those with CF from being involved in group activities, many of the adults tried to stay involved with fundraising.

"next year they've got like a golfing charity day so I was thinking about maybe a few of my friends, they play golf, I might get them to sort of a golf day and raise some money that way so, yeah so you know I mean every little helps I suppose" (A27)

Research was also very interesting for adults and many chose to be involved in as much research as possible.

"I am interested in probably research, is the best thing concentrating on research to help look for better drugs and things like that" (A11)

The majority of adults talked about the importance of continuing to raise awareness. A few of the adults believed that those with CF should be actively involved in helping to improve awareness of CF. Still, there were a couple of adults who thought that it was not necessary to further improve awareness as they thought those interested in learning about CF were able to easily research it themselves.

"I think it's probably also a role that we, as people with CF, I think we have an not an obligation but rather if we want things to improve we should take a little bit of responsibility to inform people [...] that can only happen if you know we talk to people around us and it's when you know you see some advertising campaigns out there we can do as much as we can in terms of advertising and raising awareness that way" (A13)

6.7.7. Theme 7: Growing up with CF: Changes Over Time

Several aspects of an individual's life change as they grow up, which is still true of those with CF but with additional complexities and challenges. Two sub-themes were identified in this theme:

- 1. Increasing Understanding
- 2. Transitioning to Adult Services

6.7.7.1. Increasing Understanding

As the adults grew up with CF their understanding of the condition increased and this had both positive and negative effects. Gaining more information about the prognosis of their condition could have negative effects on their mental health.

"nobody really said anything and so when I did realise that this wasn't as straightforward as life
might be, that was a massive, massive shock" (A22)

Moreover, for many adults one of the biggest changes that came with a greater understanding of CF was the way they perceived their future.

"Obviously I understood that I had this thing wrong with me, but I just kind of accepted it, and you know, I never really gave it too much thought, I never really thought about the future too much, whereas now I tend to dwell on it" (A11)

6.7.7.2. Transitioning to Adult Services

Most of the adults included in this sample that had experienced the process of transitioning from paediatric to adult CF services appeared to have had quite difficult experiences. This is discordant to the findings presented in the previous chapter considering the experiences of young people with CF who all reported largely positive experiences.

"...it was really daunting, really scary... to be going to a hospital that is filled with adults, when actually you just want to spend time on the computer on the children's ward, playing with your game, you don't want to be going in and having to be surrounded by adults" (A17)

There were some adults who experienced a smooth transition and had a very positive experience. As mentioned previously there has been a large focus in research on the process of transitioning and this may have helped to improve the service. The improvements in experiences could also relate to the increasing number of individuals going through the process of transitioning and the service learning to adapt over time.

"the changeover was great from what I can remember it's been that long ago but yes, the two teams working together were great and the hand over [...] it was a gradual sort of process which was great" (A6)

Summary of Results

Theme 1: Relationships and Understanding

- Adults with CF were generally open about their CF and happy to answer questions others ask of them
- Some adults were selective in how much they told their families as they did not wish to over burden them
- The majority of adults were positive about peer support through online forums, with a minority concerned that the forums largely focused on the negatives of CF
- Many adults had become friends with others with CF that had since passed away, which made them more hesitant to speak to others with CF
- Several of the adults believed that the invisibility of CF lead others to be judgemental
- Information available online about CF was largely criticised for being inaccurate

Theme 2: Medical Care and Support Services

- The physical effects of CF were a large concern amongst adults, particularly fatigue
- Adults received most of their support from their clinical team and were generally very pleased with it
- Most adults were unsure of what support was available to them and lacked motivation to engage
- Adults noted that their parents were typically actively engaging with some form of support

Theme 3: Progressive and Permanency of CF

- The adults believed that CF prevented them from enjoying the simple things relative to their peers without CF
- The vast majority of adults felt a negative impact of CF on their mental health
- A few adults noted some positives they believe to have come from their CF
- Adults struggled with the poor prognosis, permanency, and degenerative nature of their condition

Theme 4: Challenges of Living with CF

- Adults generally saw transplants as a challenge. However, those who had received a transplant saw it as a second chance
- It was important to most of the adults that they were able to continue working, however the majority found that their CF greatly affected the type of work they could do, what hours they could do, and the potential for career progression
- The need to plan everything in advance restricted the adults' day-to-day lives
- Fertility issues worried several of the adults and affected their decision to have a family
- The few adults who were already parents in the sample believed there was lack of support for parents with CF

Theme 5: Medication and Treatment Routines

- _Adults used routines in order to cope with the demands of their CF, although some struggled to maintain them and found it challenging when new elements were added
- Adults had concerns about their medications affecting them in the long-term and some found the short-term side-effects detrimental on their self-esteem

Theme 6: Fundraising, Research and Awareness

- The majority of adults believed awareness of CF should be raised and thought those with CF should be involved
- Many of the adults were optimistic for drug research, although sceptical about a potential cure

Theme 7: Growing up with CF: Changes Over Time

- Many of the adults had difficult experiences while transitioning into adult services, but believed this had since improved
- Several of the adults believed their understanding of the impact of CF on their own lives had greatly improved as they got older

6.8. Discussion

The seven themes and 20 sub-themes presented above demonstrated multiple key points about the daily experiences and impact on adults with CF. A large proportion of the results from the interviews with adults with CF corresponds to the findings previously discussed in either the sibling or young person results chapters. Despite adults rarely discussing their siblings, understanding the adults' experiences within the context of the existing research could support understanding of family life and how siblings could be supported.

Adults with CF expressed a larger effect on their mental health relative to the young people with CF (Chapter 5). All of the adults believed there was an impact on their mental health, however previous literature has suggested that on average their psychological wellbeing is comparable to norms (Cronly et al., 2018). Negative effects on the adults' mental health related largely to the progressive nature of the condition. For instance, adults found it increasingly difficult to think of the future and not feel an effect on their mental health. Some of the adults held a particular concern about their medications affecting them in the long-term and some found the short-term side-effects detrimental on their self-esteem. This affected the adherence to medications for some adults (Kettler et al., 2002). Nevertheless, many of the adults remained optimistic for drug research. A few adults did also note some positives they believed came from their CF, e.g. a greater appreciation for life, which may warrant further consideration in future research.

Active adaption coping techniques, such as the use of routines to integrate treatments into their daily life, were mentioned by many of the adults. Despite this there were also several adults that spoke about being disengaged with their treatments and a small number acknowledged poor adherence. Poor adherence tended to be apparent in adults that didn't view a benefit from maintaining their treatments, which concurs with the evidence presented by Kettler et al. (2002). The addition of new medications or treatments to existing routines also proved challenging and was mentioned by a number of adults. Evidence suggests that the additive nature of CF treatment and medications is challenging for adults with CF (Sawicki et al., 2009).

Of particular concern to the majority of adults with CF was the physical effects of CF, particularly fatigue. As their health worsened adults increasingly struggled predominantly with the poor prognosis, permanency, and degenerative nature of their condition. The evidence on the association between clinical outcomes such as respiratory impairment and the psychological wellbeing of adults with CF is mixed (Pfeffer et al., 2003). Thoughts of the future have been negatively associated with psychological outcomes (Habib et al., 2015), however research on the perception of the future in adults with CF is not as extensive. Complications such as transplant or CFRD were highlighted by many of the adults as a challenge to their day-to-day lives. Previous evidence suggests that adults with CF awaiting a lung transplant adapt well and are able to continue working (Burker et al., 2000). However, adults in this study considering a transplant were quite anxious about it. Adults were particularly

worried about having to stop working and further medications or complications that may come from having a transplant. In agreement with the work by Burker et al. (2000) the adults that had received a transplant were very positive about their life and the chance they had been given from the transplant.

Despite it being important to most of the adults that they were able to continue working, the majority found that their CF greatly affected the type of work they could do, what hours they could do, and the potential for career progression. Concerns about their working life, including taking medications at work, have previously been linked to negative HRQoL in adults with CF (Burker et al., 2005; Gee et al., 2005). Adults also felt a negative effect of misunderstandings of CF in their employers. Poor understanding in work colleagues has previously been related to negative psychological outcomes in adults (Burker et al., 2005). Some adults had begun to think about a need to stop work or retire and they were finding that there was a lack of understanding from both employers and friends.

Several of the adults were worried about issues relating to fertility or parenting. Many were happy with the support and guidance they had received about potential fertility issues from their clinical team. Most adults only wanted to consider the issue of fertility when it was relevant to them during their stage of life, in disagreement with the suggestion from Fair et al. (2000) that discussions about fertility should start in early adolescence. What was particularly apparent from the discussion on this topic was that the few adults who were already parents believed there was a severe lack of support for parents with CF.

Adults received the majority of their support from their clinical team and were generally very pleased with it, as was also the case for the young people with CF included in Chapter 5. Given their time spent in CF clinics adults were more aware of the changes they had seen in services over time. For instance, many of the adults had had difficult experiences with transitioning from paediatric to adult services but believed this had since improved. Given that transitioning has received a large amount of the attention in the literature (Coyne et al., 2017), it is highly likely that improvements to services have been implemented since the adults in this sample transitioned between care services, particularly given the changing population distribution of CF (Stephenson et al., 2017).

The adults lacked awareness of what support was available to them outside of their clinical team. If the adults were aware they typically lacked motivation to engage with support. Adults demonstrated a unique concern about participating in peer support. Several adults had previously been able to meet and become friends with others with CF. Many of the adults' friends with CF had since passed away, which made them more hesitant to speak to others with CF. Several adults suggested that their parents continued to be actively engaged with some form of support for themselves, although no mention was made of siblings' need for support. The use of support by the families of adults with CF is not currently well understood. There appears to be a separation between those with CF and their families in adulthood. Adults chose to be selective in how much they told their

families to try and alleviate the burden their family felt as a result of their CF. Despite being asked about family relationships very little was said by adults on the subject, and in particular on their relationship with their siblings. This is again an area which could benefit from further consideration.

6.9. Implications for the Sibling Experience

Throughout the 25 interviews with adults with CF very few mentions were made to siblings and their experiences, which may be due to the fact that no direct question on siblings was included in the Adult interview schedule. However, it is possible to bring together the findings from the sibling interviews (three of which were with siblings of adults with CF) and the results of this chapter to form potential implications for the sibling experience. There often appeared to be a separation between those with CF and their families in adulthood. Adults chose to be selective in how much they told their families to try and alleviate the burden their family felt as a result of their CF.

Adults with CF were anxious about the increasing health demands and progressive nature of CF, and the implications this had on various aspects of their life. While this was likely to affect siblings while they lived with their brother or sister, very few adults live with their sibling (McGuffie, Sellers, Sawicki, & Robinson, 2008). However, the siblings that mentioned they had a close relationship with their adult brothers or sisters also tended to be very emotional about the long-term health implications and found it challenging when their brother or sister was upset. Some siblings, particularly those whose adult brother or sister was growing increasingly unwell expressed some separation anxiety and often changed their lives in order to be near their brother or sister. Given that many of the adults had seen other people with CF pass away, it was likely that their siblings may also have been aware of individuals with CF passing away, which could cause a great amount of emotional distress. Adults themselves lacked motivation and awareness to engage with support from outside of their clinical team. This lack of engagement may also make it challenging for siblings to engage, particularly as siblings mentioned that they often felt as though CF was a personal subject for their brother or sister.

6.10. Conclusion

The psychological wellbeing of the adults included in this study appeared to be affected by the presence of CF. The adults were aware of this impact and could identify multiple areas that they believed this affect came from. There was limited discussion and consideration given by the adults to how their CF affected their relationships with the rest of their family. Despite this, it is still important that the impact felt by adults is considered when building an understanding of the impact on the rest of the family, including siblings.

Chapter 7: Results - Parents with a Child with Cystic Fibrosis

7.1. Introduction

The literature on parental psychological wellbeing suggests that they experience feelings of uncertainty and concern when their child has a chronic illness or condition (Barlow & Ellard, 2006). Parental mental health can influence outcomes in their children (Brucefors et al., 2015). An emphasis on self-report and multiple-perspectives in sibling research, rather than solely parental proxy is needed (Kraemer et al., 2003; Mazaheri et al., 2013). By considering the parents' perspectives of both their own and siblings' experiences an enhanced and more well-rounded picture can be established of the impact on siblings.

7.2. Background

When a child has CF their parents are at a particular risk of negative psychological wellbeing. Besier et al. (2011) found that parents of a child with CF presented high levels of anxiety and depression symptoms. The intensive treatment regimen, the life-limiting nature, and the genetic transmission of CF can cause negative outcomes for parents including feelings of anxiety, stress and guilt (James et al., 2006). There is a known bi-directional relationship between the mental health of parents and their children (Pardini, 2008). Positive mental health in parents has also been linked to better health outcomes for their child (Brucefors et al., 2015; Cappelli et al., 1988; Czyzewski et al., 1994). Negative psychological wellbeing in parents is likely to negatively affect both their child with CF and siblings in the family.

The treatment burden of CF affects not only the young person but also their parents who typically supports their child in completing the demanding routine of medications and treatments (Wong & Heriot, 2008). Parents take on a large amount of responsibility for not only their child's treatments but also for keeping them 'healthy' more generally (Foster et al., 2001). This daily intrusion takes a large amount of time and effort from parents. Parents of children with CF regularly lack time for themselves, which can result in negative health outcomes (Lee et al., 2009) and social problems for the parent (Besier et al., 2011). Maintaining a social life can be difficult for parents with a child with a chronic illness or condition. Wong and Heriot (2008) found that receiving social support improved parents' ability to cope. Parents of children with CF are limited in their ability to interact with other parents with children with CF due to the risk of cross-infection. Peer support for parents of children with a chronic illness or condition is particularly beneficial as it helps parents speak to others who better understand their experiences (Nicholas & Keilty, 2007). Parents feel an effect of the level of understanding of others. Misunderstandings and negative views of CF have been connected to negative outcomes in parents (Pakhale et al., 2014).

When their child is going through a period of ill health parents show particularly high levels of stress and anxiety (Besier et al., 2011). The time of diagnosis has also been found to cause parents a

very high level of emotional distress (Mérelle et al., 2003). Parents find the process of transitioning from paediatric to adult CF services challenging (Heath et al., 2017). The increasing independence of their child can leave parents feeling a loss of control and requires the parent to trust their child to maintain their own health (Akre & Suris, 2014). It has been suggested that these are time points when it is much harder for parents to avoid thinking about the progressive nature of CF and the future implications of the condition for their child, which could be the greatest source of their emotional distress (Berge & Patterson, 2004).

Positive family relationships and cohesiveness have been linked to improved psychological wellbeing in parents (Berge & Patterson, 2004). The work by Olmsted et al. (1982) suggests that family functioning is more influential than disease severity on outcomes for both the child and family. Parents play a crucial role in family functioning and can often place the needs of their family above their own (Wong & Heriot, 2008). In families with both a child with CF and a sibling parents can struggle to effectively balance their time between their children and this has been connected to distress in both the sibling and the parent (Foster et al., 2001). Martial relationships are frequently reported to be stressed if a child has a chronic illness or condition (Rivers & Stoneman, 2003). Although, this is not what has been found in parents with a child with CF (Eddy et al., 1998).

Despite the evidence suggesting potential negative psychological wellbeing in parents there is also evidence that suggests good adjustment and coping in parents with a child with CF (Besier et al., 2011). Parents may be able to use coping mechanisms effectively to process the presence of CF in their life (Wong & Heriot, 2008). Families are generally found to adapt well to CF (Patterson, McCubbin & Warwick, 1990). Parents are able to access support from a range of sources, including their child's clinical team and third-sector charities. There are still gaps in the existing support and literature on parents. A greater need for supporting parents during the period of transitioning between paediatric and adult care teams has been highlighted (Dupuis et al., 2011).

The parent plays an important role in the psychological wellbeing and adjustment of families with a child with CF (Ma et al., 2017). Parents have a unique view point on the impact that CF has on not only their life but also that of their child with CF and siblings. It is beneficial to incorporate the parent's perspective into research, as has previously been done (Kraemer et al., 2003). However, the use of parental proxy only is not recommended as self-report provides the most relevant and reliable results (Eiser & Morse, 2001a).

7.3. Aims and Objectives

As with the previous result chapters consideration was given throughout this chapter to the overarching aims of the qualitative project; to enhance the understanding of the lives, needs, challenges and aspirations of those with CF and their immediate families as well as inform the development and provision of appropriate and relevant support. The purpose of this chapter was to

consider the experiences of parents of children with CF and the implications for the rest of their family. To address these aims the following research questions were proposed:

- 1. How do parents of a child with CF view the impact of CF on their lives? Specifically, do they identify an effect on their and their immediate family's psychological wellbeing?
- 2. Do parents believe they and their family are well supported? If not, are there areas where they believe support could be beneficial?

7.4. Parent Recruitment

As with young people and adults with CF recruitment of parents was completed up to the target of 25 with relative ease. Parents were recruited from across the UK, from hospitals in London (n=11), Cardiff (n=9), and Glasgow (n=3). The remaining were recruited through online advertisements (n=2). Fewer parents were recruited from adult clinics (n=4), as it was far rarer for parents to attend routine clinic appointments. Contact information was received for a total of 67 parents. Of these parents 30 agreed to participate, however it was not possible to complete the study with five of the parents as three did not answer their phone and two no longer wished to take part. Of the 25 parents, 19 had a child in the young person group, two had a child in the adult group, five also had a partner included in the parent category, and nine had a child in the sibling group.

7.5. Data Collection and Analysis

Data collection and analysis was run in accordance with the details previously given in Chapters 3 – 6. All data collection was organised and managed by MMS. Consent was taken for all parents prior to interview along with a QoL measure. MMS was assisted primarily by LA in the collection of data from parents. The majority of the interviews were conducted by MMS (n=8) and NK (n=7) with the remaining completed by other researchers (LA (n=4); RS (n=3); HAW (n=2); CR (n=1)). Interviewer consistency was more challenging to ensure with the parents due to limitations in the research team. Twenty-two of the interviews with parents were completed over the phone and the remaining three were completed in person either at GOSH or at the participant's home. Interviews with parents lasted an average of 45 minutes. All parent interviews were transcribed verbatim by LA. MM along with NK cross checked all transcripts against the original audio to ensure as high an accuracy as possible. The analysis process was completed by MMS and supported by LA. Cross-validation of the coding of the transcripts was completed by MMS and NK.

As with the previous analyses, respondent validation was used. Parents were asked to review the initial codes and themes established from their interviews and respond with any concerns they had. No parents responded, therefore no changes to the analysis was made. Given the approach of this analysis, it is assumed that the results below are representative of the true experiences of this sample.

7.6. Participant Demographics

Parents ages ranged from 32 to 68 years (median = 49). Twenty-two were parents of young people with CF while three were parents of adults with CF. Seventeen of the participants included in this group were female and eight were male. The majority of participants were parents, one grandparent with guardian responsibilities was also included. As with each of the previous groups, all parents were of white ethnicity. Only one parent reported being a full-time carer and the majority of parents were working either full-time (24%) or part-time (28%). Four parents did not disclose their yearly household income. A majority (52%) of those that provided household income information earned above £50,000 per year. Further demographics are shown in Table 7.1 below.

Parent 1	Age	Sex	Marital Status	Employment Status	Parent 2	Age	Sex	Marital Status	Employment Status	Household Income	Sibling	Child w. CF
P1	57	F	Living with Partner	Full Time Carer						15,000-25,000	S3	Y14
P2	36	M	Living with Partner	Employed (Full-Time)	Р3	36	F	Living with Partner	Employed (Part-Time)	50,000+		Y5
P4	50	F	Married	Employed (Full-Time)						25,000-35,000		Y6
P5	43	F	Divorced/Separated	Employed (Part-Time)								Y15
P6	39	F	Married	Out of Work						15,000-25,000		Y1
P7	51	F	Widowed	Unable to Work								Y20
P8	36	М	Divorced/Separated	Out of Work							S8	Y7
P9	48	F	Married	Employed (Part-Time)						50,000+		Y8
P10	46	F	Married	Employed (Part-Time)						50,000+	S10	Y2
P12	52	М	Married	Employed (Full-Time)						50,000+		Y10
P13	52	М	Married	Unable to Work	P11	56	F	Married	Employed (Full-Time)	50,000+	S4	Y21
P14	44	F	Married	Employed (Part-Time)						35,000-50,000	S1	Y11
P15	56	М	Married	Employed (Full-Time)						50,000+		A14
P17	62	F	Married	Retired	P16	69	M	Married	Retired	15,000-25,000		A21
P18	32	F	Living with Partner	Unable to Work							S2	Y12
P19	48	F	Married	Self-Employed						35,000-50,000		Y25
P20	36	F	Divorced/Separated	Employed (Part-Time)						15,000-25,000		Y13, Y16
P22	41	М	Married	Employed (Full-Time)	P21	41	F	Married	Out of Work	35,000-50,000	S5	Y3
P23	53	F	Married	Self-Employed	P24	53	М	Married	Self-Employed	50,000+	S9	Y9
P25	44	F	Married	Employed (Part-Time)						50,000+	S11	Y27

Table 7.1. Parent Demographics

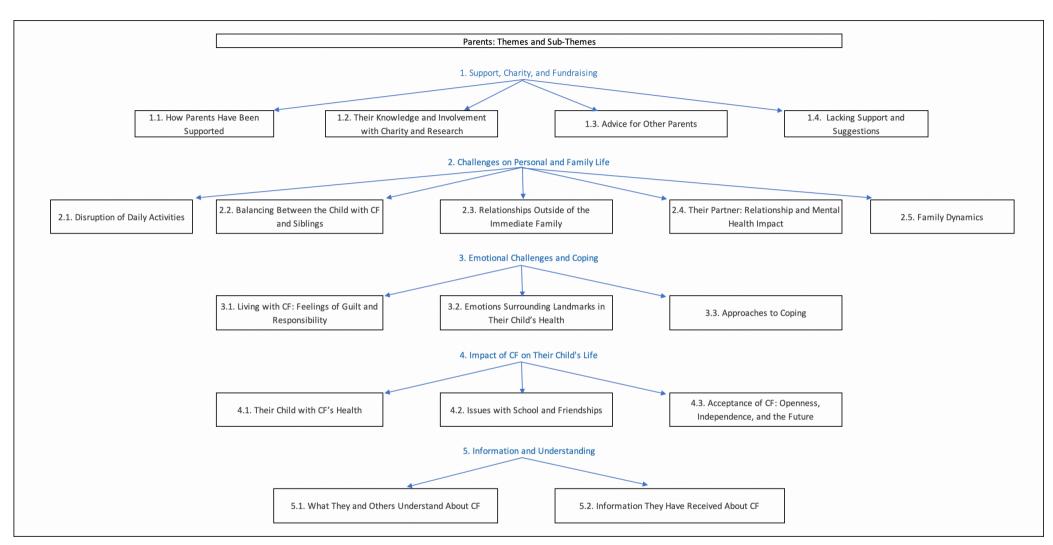


Figure 7.1. Themes and Sub-themes of Parent Interviews

7.7. Themes

From the qualitative thematic analysis conducted on the 25 parent interview transcripts five themes were identified:

- 1. Support, Charity, and Fundraising
- 2. Challenges on Personal and Family Life
- 3. Emotional Challenges and Coping
- 4. Impact of CF on Their Child's Life
- 5. Information and Understanding

Each theme contains multiple sub-themes. The themes and sub-themes are given in rank order of reference density and displayed in Figure 7.1. Each theme is discussed below with verbatim examples taken from transcripts.

7.7.1. Theme 1: Support, Charity, and Fundraising

Parents of a child with CF are able to access a range of support services from various sources including charities such as the Cystic Fibrosis Trust and their child's care team. Whether parents chose to use it and what services they use is unclear. Parents regularly choose to engage through other activities such as fundraising and research. Five sub-themes were identified within this theme:

- 1. How Parents Have Been Supported
- 2. Their Knowledge and Involvement with Charity and Research
- 3. Advice for Other Parents
- 4. Lacking Support and Suggestions

7.7.1.1. How Parents Have Been Supported

Nearly all parents mentioned that they had used some form of support, either formal or informal. One of the most predominately mentioned and positively viewed types of support was peer support. Peer support was highlighted as particularly beneficial around difficult time points e.g. their child's diagnosis.

"What I found very helpful when [child with CF] was first diagnosed is the doctor immediately put us in contact with another family that happened to be in the hospital at the time. And we could sit and talk to them and find out things" (P25)

By consulting their peers, it was possible for parents to gather what they viewed as reliable information from a source other than their child's care team or the internet.

"And I think If you google things it can give you crazy diagnoses and stuff whereas if you just ask someone who has actually been through it, it, can totally take the weight off your mind of actually what it is" (P6)

Talking to other parents not only had the benefit of being a source of information that the parents felt they could trust but it also allowed them to speak to someone whom they believed understood what it was like to be in their shoes.

"I just feel that they [other parents] really do understand what it's like to have to nag your children, to have to be worried about your children, to have such huge issues and huge conversations on such serious matters with young children, you know no-one gets it like them" (P10)

Most of the support from other parents was received informally, although a small number of parents were involved in more formal peer support through organisations such as the Cystic Fibrosis Trust.

"I'm one of their [the Cystic Fibrosis Trust] befrienders so if families come forward on the helpline wanting advice then they'll put out an email to the volunteers to say can we match you up to this family" (P17)

Despite enthusiasm for peer support many parents were concerned about speaking to other parents. Parents typically viewed their child's experiences with CF as highly individual and worried about speaking to other parents whose child was in better or worse health than their own.

"I almost feel guilty because [child with CF] is quite healthy and then you speak to other people that are having more issues. And I almost feel guilty to say oh actually [child with CF] is fine" (P25)

Other informal support received by parents included support from friends and family. Quite frequently this support was as simple as having someone to talk to about their child and the difficulties they were having. Most parents believed their friends and family were supportive of them and their children and available to help and comfort them should they need it. Several parents chose to lean on their partner when they needed support. If the parent was not the primary caregiver it was harder for the parent to do this as they did not wish to over burden their partner. The clinical team at the hospital provided support to the parents in several ways, including being available to give parents reassurance and information. Several of the parents suggested that the hospital was their main source of support, and that they were well supported by them.

"the CF nurses are fantastic, absolutely fantastic. And they talk to you like you're a human being.

the doctors are really really good, but sometimes, you come across doctors that haven't really got

much people skills, whereas the nurses have definitely got the people skills" (P3)

7.7.1.2. Their Knowledge and Involvement with Charity and Research

Several parents were aware of the Cystic Fibrosis Trust or other CF charities. Many of the parents chose to not engage with support services offered by them. While this echoes the engagement

of those with CF and siblings, it does not agree with the way parents were perceived by their children in these previous chapters.

"the CF Trust I mean we've never really been we're aware of it but we've never really tapped into it" (P16)

Despite a resistance to engage with the services from the Cystic Fibrosis Trust several of the parents had completed some form of fundraising activity for them.

"we've done a lot, my husband did the London marathon a few times for CF, we raised about

1500 pounds for the CF Trust" (P19)

Opinions about research on CF were varied. Some parents had a negative outlook on research and, in particular, the funding of drugs they had seen were available in other countries. Other parents were excited by research and the prospect of participating in it. Parents recognised the benefit of research and the potential to improve outcomes for their child, but many continued to mention that there needed to be a balance between research and supporting those currently in need.

"I know that's important but you can't just do all research and then leave the people in hospital...

basically juggling that money to make the best for everybody" (P19)

7.7.1.3. Advice for Other Parents

When asked about what advice the parents would have for other parents of children with CF they gave a range of responses but there were several key messages which appeared frequently. Parents recommended other parents try to focus on the positive and particularly on the increasing life-expectancy of those with CF.

"well anyone who finds out [their child has CF] now has got the best chance ever of the child having a long life which is great and they must take heart from that and not to be too scared of it"

(P23)

Other, more practical, recommendations parents had for those who had just found out their child had CF included setting a clear routine quickly, and becoming informed about CF. Becoming informed about CF was mitigated by some parents who mentioned a benefit from not over informing yourself, particularly on the things that could go wrong. Parents also agreed that the source of the information used needed to be reliable.

"just read up about it you know don't read don't google it on the internet or anything silly like that" (P22) A few parents emphasised the need to treat the child with CF as a 'normal' child, while also making sure the child is kept well and completed their treatments.

"treat him like a normal child even though he has got health issues you care about the health issues but [treat] them like any other child" (P7)

7.7.1.4. Lacking Support and Suggestions

As with young people, parents appeared happy with the support they had received. There were areas that parents identified a need for further support. Some parents mentioned that they lacked awareness of the support available to them.

"I don't know what there is beyond information I don't know what there is for actual caring support" (P12)

Many of the parents offered suggestions of practical support that they believed could be beneficial to them or others. Multiple of the support suggestions were for their children with CF and many related to the dietary requirements and restrictions for those with CF.

"it would be a good idea if you could have a recipe book that also had the fat contents of all the ingredients" (P11)

Further to this the several parents highlighted how they believed siblings were the ones in need of support. Several parents felt that their child without CF was not opening up to them about their difficulties or was "suppressing" their emotions as they do not feel like the priority for support in their family.

"I think my biggest thing out of all of it is the lack of support I think for siblings" (P5)

"Well I'm sure that my daughter who doesn't have CF maybe has repressed talking about things that she might feel because maybe she has felt that in the context of the CF in the family that it wasn't a legitimate" (P14)

One area that was mentioned by several parents where they believed they needed additional support was their child transitioning from paediatric to adult CF services and in the process their child becoming more independent. Parents were the only group to identify the transition period as one that they saw a need for better support during.

"going from us supervising everything to [CF Child] starting to take care of it himself I think that is an area that you could maybe get some support" (P24)

Parents also felt a need for support with responding to questions from their child about more sensitive topics relating to CF.

"I think going forward as [CF Child] gets older maybe how we approach talking to her in depth about it maybe cause we haven't had the conversation where everything's on the table... I'm not sure how I would cope having that conversation so maybe yeah some tips and stuff on how to and that when that talk does come along how to approach it" (P22)

The parents were generally unwilling to regularly lean on others within their family. Parents thought that having the opportunity to talk to someone outside of their immediate family would be positive.

"I think there's a natural reluctance to burden the others in your family with your own personal (issues) around CF" (P14)

Parents believed there was an issue with the information currently available about CF. They emphasised a need for clearer information for parents about CF, as illustrated by P24.

"information which I think could benefit people in general would be along the lines of when do you need to worry about a persistent cough you know for how long do they have it and then you would say you need to contact them" (P24)

As previously discussed, parents believed that encouraging more peer support amongst parents would allow them to share information in a way that would support them through difficult times.

"I think if we could encourage the parents to get together and share their stories so that they didn't feel so alone and that they could talk about their fears and that they could see actually they might be going through some difficult years but you know seasons pass" (P10)

Peer support for their child with CF was also highlighted as a potentially beneficial support service. However, parents generally viewed their child talking via social media or online negatively as they were concerned about supervision of conversations online. Some parents suggested that phone conversations or emails would be more appropriate.

"I think that it would be good if there was some way that young people could communicate with each other I know the Trust are doing more to do with that but I've got such issues with social media I just think it's hideous cause there's been talk of a Facebook group and all that stuff and I don't want her at 13 years old being on Facebook. I would want to know more about how that's going to be monitored and supervised and ran before I would let her do anything like that but I think sometimes she would like to talk to somebody else" (P17)

Further areas that a small number of the parents highlighted for potentially needing support with included leaving their child with others, single parenting, governmental benefits, and geographical limitations to support.

7.7.2. Theme 2: Challenges on Personal and Family Life

The parents and their families were faced with many challenges in their lives that were either directly or indirectly related to CF. Five sub-themes were identified within this theme:

- 1. Disruption of Daily Activities
- 2. Balancing Between the Child with CF and their Siblings
- 3. Relationships Outside of the Immediate Family
- 4. Their Partner: Relationship and Mental Health Impact
- 5. Family Dynamics

7.7.2.1. Disruption of Daily Activities

Many of the parents felt that CF had a large impact on their daily lives. The impact experienced by parents appeared to lessen as their children became older and were able to take greater responsibility for their treatments. However, parents tended to feel a level of responsibility for keeping their child well even when their child was fully independent.

"it just affects everything about your life. Absolutely everything on a daily basis. I mean personally for me, I can't really make any plans because you don't know whether they are going to be well from one day to the next" (P1)

"they're a little older now so it has got a little bit easier because when they were younger it was hard it was really hard" (P20)

Other parents believed there was very little impact on their day-to-day life, however they generally followed this up by mentioning mechanisms they have put in place in order to reduce the daily impact of CF. The use of routines was discussed by several of the parents as a useful technique for reducing the daily impact of CF. Parents spoke about using treatment routines to help reduce the impact of CF on day-to-day life. When a treatment routine was interrupted by the addition of new medications parents were faced with a challenge both in adjusting to a new routine and experiencing a period of time where the reality of their child's health condition was highlighted.

"what used to throw us was when any added, either medication or nebulisers or if she became unwell that's when it almost brings it back to reality of what we're dealing with... you then get used to that new change and it then again becomes normal until something else is thrown in and then you go through a period of it being not stressful but kind of the knowing what it entails again" (P21)

All parents felt an effect of CF on their work life. Adjustments frequently had to be made by either the parent or their partner to ensure that someone was able to care for the child or deal with any health issues that may come up.

"my husband gave up his career I kept my career...so he gave up work to look after [CF Child] and then to look after his sister as well when she was born" (P11)

Even when parents were able to keep their job they still felt a strain because of their child's CF. Parents regularly had to either reduce their working hours or rely on their employer being very understanding.

"there are times well when [CF Child] has been in hospital I think if I'd of had a different job I
would have possibly been vulnerable to losing my job" (P11)

The disruption to parents' work life was largely focused on either when the child was ill or when they were younger. As their child gained more independence the impact reduced, although some parents found it hard to adjust back to working life.

"I went part time and obviously after having the children and I think the only effect it probably had on my work life and still now I feel reluctant to go back full time just in case [CF Child] has any needs" (P21)

Practical limitations affect both the parent and their child on a regular, if not daily, basis. For instance, while the parent controls their child's treatments and medications it can be very difficult for the two to spend time apart.

"I mean it adds a complication if we're all going out or if [CF Child] is going to go and stay somewhere else then you know things have to be packed and things have to be explained to people that he's going to and so on so yes there is an impact" (P13)

A small number of the parents also suggested that some of their child's behaviour related to their CF and placed a strain on their daily life. For instance, those with CF often have complicated relationships with food and this can be an issue for parents when trying to go somewhere that their child is expected to eat.

"it's just difficult when you're out and about because he's so picky about what he likes to eat"
(P5)

Some parents believed that their own health was affected by their child's CF. Health effects in parents have been found previously (Lee et al., 2009). These effects could be either positive or negative on the parent's health and appear to relate to their level of emotional adjustment to CF.

"Yes, for sure [CF affects their general health] because I am overweight and eat and drink more than is sensible for good health probably do that as a coping mechanism I recognise it has an adverse effect say on sleep" (P14)

"If anything, I thank [the CF child] because I got into running because of her, cause when she was three, I did a 10K for the CF Trust... since I have been running, I'm even happier than I was before, so I feel like I kind of owe her to that, because I don't think I'd have ever got into it if it wasn't, wasn't for the fact that she had CF" (P3)

7.7.2.2. Balancing Between Their Children with and without CF

Parents spoke about how they balanced their time and resources between their children with CF and their siblings. Parents have to spend more time with their child with CF for various reasons, which both they and the sibling found challenging. Many of the parents spoke about the need to consciously compensate for the lack of attention the sibling received.

"I think I would say I have physically not been able to spend as much time with my other son [CF Child] needs come first" (P23)

"sometimes [you] find yourself overcompensating a little bit like because we spend so much time and effort on [CF Child] you find you have to make that effort and say to [Sibling] what do you want to do let's do something for you because you don't want them to feel like they are not important"

(P19)

Despite parents attempting to compensate both parents and siblings expressed emotional difficulties from the disparity. Parents mentioned that siblings, on occasion, expressed feelings of jealousy against their brother or sister with CF.

"I do remember him years ago saying it's not fair I wish I had CF obviously about the attention and I turned around sharply and said no you don't you really don't but [child] who has the CF will often say oh I'm so glad I've got this because of all the attention he was getting" (P10)

Parents also mentioned that they felt guilt for the difference in the way they parented their child with CF and their sibling, although they believed that this was inevitable and unavoidable.

"[sibling] always says you let him get away with everything and I know I do sometimes because he's got hard time with the other things he has to do I cut him more slack I know I do but he obviously picks up on that and criticises me for it so you can't win" (P23)

Despite these clear differences in treatment and attention, several of the parents believed that if the sibling was asked they would likely say that CF did not affected their life.

"he will tell you that it doesn't affect his life in any way whatsoever. There's little things that you wonder if that's because...for example he never complains if he's poorly [...] And I wonder if that's just his personality or if that's because when you've got a common cold, you feel grotty, but you look at someone like who's that's with common cold all her life, and you think it's not so bad. So, then I do wonder if that's why is like that is it because he compares himself to?" (P25)

As CF played less of a role in their relationship several parents believed their relationship with sibling was a "very easy relationship" (P4). A close bond between siblings and their brother or sister with CF, while a positive thing, had the potential to cause parents more stress when their child with CF was unwell due to the siblings emotional and behavioural responses.

"you can see very clearly that when he was in hospital they [siblings] almost become sort of like enlarged versions of their worst character...so one of his sisters you know got a lot crosser a lot more frequently and the other one became more worried" (P9)

Some parents used siblings as a measure of 'normality' and tried to balance the way they treated their children with CF against how they treated a 'normal' child.

"he had to be treated normally as well because he was normal we wanted him to be everything that his brother and sister were so you had to have that in your fore thought and your planning always that he's not wrapped in cotton wool that he's got to be sustainable in this world" (P16)

7.7.2.3. Relationships Outside of the Immediate Family

Relationships outside the immediate family were challenged by the presence of CF. A number of the parents had negative experiences with their extended family due to a lack of sympathy and understanding. The lack of understanding in their extended family reduced the confidence of both the parent and the family member to care for the child with CF independently.

"my Mum wouldn't have him, my sister wouldn't have him, for ages because they were scared of him" (P4)

Children with CF are at a heightened risk of infection and small illnesses such as colds could be much riskier for them. Having friends and family that tried to understand and be considerate was generally a very positive thing, however it may also reduce the parent's opportunities to socialise.

"quite often because I've got good friends of mine who are very considerate they would ring up at the last minute when we were about to go and see them for the weekend and she's got a cold do you think you better not come or do you still want to come? And you automatically feel well I can't come now because if I go and then [CF Child] gets a cold I'll never forgive myself" (P23)

Friends' level of understanding greatly influenced a parent's willingness to maintain friendships. Some parents chose not to remain friends with others who they believed were inconsiderate of their child's CF.

"I think your tolerance of people changes and I think you only then literally have people in your life that are going to stay there and be there and tolerate and understand what you're going through" (P5)

Their child's treatment routine put a strain on several parents' social lives. Having to be with their child when completing treatments prevented many parents from being spontaneous. However, their child's health also meant that parents were less able to stick to plans and had to regularly cancel when their child was unwell.

"we've cancelled plans last minute and it's never a question I don't ever feel like I offend anybody and I think the truth is family first, kids come first it's never been a question of being worried if we have to cancel something it's just if you can't go you can't go there will always be another time" (P19)

There was also a couple of parents that viewed the impact on their social life as comparable to what they believed having any child would be like.

"having a child like massively impacts your social life. So, I don't know if, perhaps CF did any more than just having not, like a child..." (P3)

Almost all the parents had a positive relationship with their child's paediatric team. Parents whose children were going through or about to go through the process of transitioning to adult service, regularly mentioned that they thought leaving the paediatric team was going to be difficult for them.

"yeah [CF Child] will be moving up to adults in a few years and I'm going to miss them [paediatric CF team] so much because I call them part of my family because they've been there since day 1 and they they're such an amazing team as well" (P18)

7.7.2.4. Their Partner: Relationship and Mental Health Impact

A number of the parents believed that their relationship with their partner had been affected by their child's CF. Parents identified both positive and negative effects on their relationship with their partner. The expectation is for a chronic illness or condition to place a strain on marriages (Hartley et al., 2010), however there were parents included in this sample that believed their relationship was strengthened by having a child with CF.

"the doctor had said to her oh you guys have done really well to stay together because a child this sick often it breaks the couple up you know they said that and she told me that you know so I thought we did quite well that we were still there still together I think what doesn't break you makes

you stronger" (P10)

This is not to say that parents did not recognise the challenges their child's CF placed on their marriage. An effect was particularly apparent during stressful periods such as when the child was in poorer health.

"if things go wrong you both get stressed and tense and then you start blaming each other for it...

and you just punch holes in each other because you can so that's a thing that's bad about it you use
each other as a bit of a punch bag when you're feeling bad about him or yourself or whatever so I
think we're both quilty of that to be fair" (P23)

In the majority of families there was one parent that took on the role of primary caregiver and the level of involvement by the other parent varied. Where the primary caregiver did not receive much support from their partners they felt a strain on the relationship.

"I'd have liked him [partner] to take more of an interest in it and would've liked him to be more hands on then, obviously he done physio sometimes in the afternoons and stuff but even then it was hard it was really hard because he didn't really know what he was doing half the time" (P20)

Many of the parents chose to be selective about their communications in order to limit the impact they had on others in their immediate family, including their partners

"she's [partner] the person I'd most likely to share such a feeling with by the same token she's also the person I'd least likely share it with because I know how much it's affected her too" (P14)

Several of the parents spoke about how challenging they found it to spend time alone with their partner following their child's diagnosis with CF. The mental health of the parent and their partner were affected by the limited amount of time they were able to spend alone together.

"I think that did have a different effect on our mental health that we felt we could never get a babysitter and go out" (P11)

A few parents believed that their partner's mental health had particularly suffered. While other parents thought their partner was the stronger one of the two of them and managed to maintain a better mental health.

"in contrast, I would say my husband is naturally quite an anxious person and I definitely think it has at times been a struggle" (P9)

"I think I would say my husband definitely was a stronger person and really got us through it all.

He was very positive" (P25)

7.7.2.5. Family Dynamics

Relationships between siblings and their brother or sister with CF were viewed by many of the parents as affected by CF. Although tension between their children could not necessarily be entirely attributed to the presence of CF.

"they could be like that anyway but is there a resentment there, I don't know and hard to know because like a lot of things you just tend to blame everything that's bad on CF life" (P23)

Many of the parents identified a tendency in siblings to be protective over their brother or sister with CF. A small number of parents hoped that the sibling relationship was strong enough that the sibling would be able and willing to give support should their brother or sister need it.

"I'd like to think that, if me and [partner] were, I don't know, away or something like that, when they're older, if [CF child] needed a hand or needed something like that, then she could call up one of her sisters" (P3)

The majority of parents strived to be open about CF with their children. How they spoke about CF varied from family to family. Some parents chose to limit the amount of time they spoke about CF within the family. The majority of the parents tried to speak about CF in a way that didn't focus on the negatives.

"I don't bring it up all the time. We do discuss it, we don't go into massive discussions about it because I just want it to be normal I don't want it to be a daily thing with them" (P20)

"we've always been open and I know it sounds a bit shit but we've always been quite jokey about things as well because to sort of lighten the mood about things we've never sort of been a family to sit there and be oh my god you know [CF Child]'s got it's like you can do what you want to do just get on with it and that's the way we've always been" (P19)

7.7.3. Theme 3: Emotional Challenges and Coping

Having a child with CF put an emotional strain on all parents in one way or another. How parents coped with these emotions and challenges greatly varied. There were three sub-themes identified within this theme:

- 1. Living with CF: Feelings of Guilt and Responsibility
- 2. Emotions Surrounding Landmarks in Their Child's Health
- 3. Approaches to Coping

7.7.3.1. Living with CF: Feelings of Guilt and Responsibility

The time of diagnosis was mentioned as a particularly emotionally challenging period for parents. Several parents mentioned feelings of guilt surrounding this time. The guilt the parents felt related to various aspects of CF. At the time of diagnosis genetic inheritance caused parents a particularly large amount of distress and guilt.

"if you have a child that you know you've given them something then you have that element of guilt don't you, that you've done that" (P5)

The guilt of giving CF to their child lead P3, and a few other parents to choose to not have any more children.

"I just think for me and [partner] to then risk having another baby with CF, it'll be a bit selfish to knowingly... possibly bringing another child into the world with CF [...] so, we just made the decision like, ethically I don't think it's right to bring another child into the world with CF" (P3)

Many of parents expressed feelings of guilt around times when their child was ill from an infection, such as pseudomonas. Parents tended to take full responsibility for their child getting ill and blamed themselves.

"he was positive for pseudomonas you know and I still have guilt about that now because sometimes I used to wash him in the bath and I think I wonder if I did it because I wasn't aware of the dangers then you know" (P10)

Parents also experienced guilt when leaving their child. While a small amount of anxiety and concern is common in parents leaving young children, this was further complicated by the medications, treatments, and increased risk of illness in children with CF.

"I remember going away for one weekend, and coming home and she was so ill... I felt it was my fault that I should never have left her, but my parents thought it was their fault because they didn't look after her properly. It wasn't anybody's fault, it just happened" (P1)

Further guilt was felt by several of the parents when they had to enforce a treatment routine on a resistant child. Despite knowing it is beneficial for the child's health, several parents felt guilt for making them do treatments.

"it's something he's got to do and it's for his own good and get on with it so and then that kind of gets you... you can find yourself feeling a bit guilty. Getting a young child to do those sorts of things but it is for their own good and it reassures you when somebody says you have to be strong you've got to do it" (P13)

7.7.3.2. Emotions Surrounding Landmarks in Their Child's Health

Throughout a life with CF there are various key time points and complications that tend to be associated with heightened emotions. Many parents spoke about how these moments affected their mental health. The first landmark the parents identified as coming with a high level of emotion was the period surrounding the diagnosis of their child. Several parents expressed how this period was particularly challenging.

"I think the fact that when I was told my child did have it I was like Oh my God my world was ripped up and thrown into little pieces and thrown on the floor cause this wonderful baby that I'd waited for so long has all this other stuff to deal with" (P4)

"when he was first diagnosed he was quite ill you go through that awful terrified and grieving phase learning about the whole disease and what it might mean" (P23)

Parents recognised that the period when their child was transitioning from paediatric to adult CF services was an emotionally challenging period. Transitioning was potentially more challenging for the parent than their child, "it was lovely for [CF Child] it was a bit harder for me letting go" (P19). While parents highlighted leaving the team they had known for a long time as difficult, the aspect that appeared to be most challenging during this period was the child's increasing responsibility for their own medications. Trusting their child and letting go of control over their health was challenging for many of parents.

"that is a bit of a scary time, to have to leave these people that we've had his whole life and to move on to somewhere new and to think of him as an adult going to adult clinic and being more responsible for everything rather than me and his father" (P11)

"we had a whole thing of going to the adult clinic introduction meeting and they asked you to express your hopes and fears and worries and you know worries was we won't understand what's going on with [CF Child]'s treatment, they said that shouldn't be a worry that's just the way it is so" (P12)

There were a few parents that were concerned about the issues with fertility their child may face. Talking to their child about the fertility issues of CF appeared to be an emotional and challenging topic for parents.

"If I had to pick one of the difficult times that because [CF Child]'s always been oh I'm going to have 4 kids and you think sooner or later I'm going to have to tell him" (P19)

The most emotionally charged subject concerning their child's CF related to the shorter life expectancy. Negativity in their child about their expected life expectancy was very difficult for parents

to hear. Many parents choose to avoid the conversation about life expectancy with their child as they hope their child will have the same life expectancy as those without CF.

"and I spoke to him about it and he said what's the point; I'll be dead in 5 years" (P7)

"we haven't gone down the route really of speaking to him about life expectancy and things like that because you don't want for him, cause you know he might have a normal touch wood fingers crossed a near normal life expectancy if these treatments come on board and they help him because he's already healthy at the moment" (P24)

7.7.3.3. Approaches to Coping

There were many common forms of coping mechanisms used amongst parents. Parents regularly reported methods of positive reframing coping. Positive reframing coping methods could be used in conjunction with other techniques, such as comparisons to others or avoidance.

"I suppose we always looked for the positives A rugby player who had CF and somebody else and these people can do everything and he's realised he can do so I think it is about staying positive and also saying there are people a lot worse off rather than I never wanted him to be one of those children who sits there and feels sorry for himself" (P19)

The vast majority of the parents used routines in order to incorporate and manage the high treatment burden of CF and reduce the impact this had on their lives "Yeah, I think be organised is a major thing and having a routine" (P17).

A few of the parents highlighted how they felt that being open and talking to others was a key way they were able to deal with difficulties. This may also relate back to the enthusiasm for peer support identified previously.

"I'm a talker so my way of coping I think is to talk to friends in work and to problem share as opposed to storing it up" (P10)

Suppressing emotions when needed to provide strength to others was reported by parents. This also re-emphasises the tendency for parents to put the rest of their family before themselves.

"you sort of you have to switch off...you had to be hard because he needed you there and there is no way that you could be there crying your eyes out because that wasn't going to help him out" (P7)

There were also those parents who chose to avoid the negative emotions they may have when times are difficult by keeping themselves distracted by other things "I try to keep myself like busier to take my mind off it" (P8).

7.7.4. Theme 4: Impact of CF on Their Child's Life

Each parent had their own perspective on how CF had impacted on their child's life. Difficulties experienced by their children often pass on to the parent and impact parents' psychological wellbeing. Three sub-themes were identified within this theme:

- 1. Their Child with CF's Health
- 2. Issues with School and Friendships
- 3. Acceptance of CF: Openness, Independence, and the Future

7.7.4.1. Their Child with CF's Health

Several parents spoke about how they viewed their child as a 'healthy' CF patient, "she's generally a fairly healthy CF patient in the grand scheme of things" (P25). Even if their child had had previous inpatient stays, a long period without being hospitalised was acknowledge as a very positive sign of their child's good health.

"she went back into the hospital a few times... since then she's doing pretty well and that's been going on for like 8 years" (P8)

The increasing amount of treatment their child was having to do, their deteriorating health, and further complications of CF, such as CFRD, concerned parents. As their child's treatment requirements increase the daily impact of CF increased and it becomes harder for parents to avoid CF.

"we had a few years when everything was normal and so we had his sister because it was all just like we didn't it didn't really have that much of an impact he wasn't on many nebulisers you know but obviously as time has gone on his nebulisers have increased, treatments have increased and IVs, he's now got a port he's had a port fitted" (P5)

Parents agreed that their child with CF's mental health was greatly impacted. Particularly of concern for parents was their child having a negative outlook on their life expectancy. This negativity from their child could impact on both the parent and the child's daily lives.

"recently I asked for [CF child] to see a child psychologist because she was getting a bit, at school she's like I'll be dead I don't have to do it. And she really got that attitude and it's really affecting her school life so I have, for the first time, asked for help" (P1)

Anger was also raised as a potential issue for a few young people. Anger from their child tended to centre around the child's treatments. Parents often had this anger directed at them when trying to ensure their child completed their treatments which caused tension between them and their child.

"I think frustration for me and for him because he loses his rag he has a real issue with anger"
(P10)

7.7.4.2. Issues with School and Friendships

Several parents highlighted issues with school and friendships they believed were particularly challenging for themselves and their child with CF. The child with CF regularly had to take medication at school. Parents need to be confident in either the young person taking their own medications or their school to support them.

"At school he self-medicates with his Creon sometimes if he feels like it, other times he won't.

Sometimes I have to ring the medical lady and say look I know he's not taking his tablets can you, and the school are very supportive and they will check up on him for me" (P4)

All the young people were required to take Creon at meal times to help their bodies correctly digest their food. Having to take their Creon caused a disturbance to the young person's lunch time and socialising with friends. The young person with CF being concerned about how they were viewed by others when taking their medications was highlighted by multiple parents as a difficulty. Some parents thought this increased the chance of their child not taking their medications, which was a key concern.

"she's seen an impact from school you know where people will obviously stare and stuff like that when you're taking tablets and because she's hypoglycaemic so she's testing her bloods a lot and that sort of thing so there's certainly reactions from that" (P6).

Parents tend to be anxious and worry about their child's move from primary to high school. Several parents mentioned their nervousness surrounding their child's greater independence with their medications when at high school. Other parents were more concerned about a new environment with children that may not have previously aware of their child's CF.

"then you go to secondary school and you've got all these strange kids asking why you take and he's actually sort of hidden away to take his and things like this so that's a bit upsetting" (P23)

The level of understanding and support the school showed both the young person and their parents greatly altered their experiences. Ignorance in teachers was associated with very negative experiences.

"When [CF child] was at her primary school, I felt very isolated because the headmistress just wouldn't allow [CF child] to do anything. She couldn't go on school trips and I did feel, I felt isolated for her cause she couldn't go. But not now because she goes to secondary school and they just let her go on everything" (P1)

A small number of parents had difficulties making schools understand CF and their child's needs while at school. The parents often sought the help of their child's clinical team when they were unable to handle the issue themselves.

"in class he might have to get up 2 or 3 times ... and go to the bathroom and each time they'd say why in front of the classroom of people so I did go down and ask them if they could print a little card so [CF Child] would just show the teachers to make them aware he's not just trying to fob off school if he needs the toilet he really needs the toilet and they did it but they weren't the most helpful and I think what helped the CF nurses they wrote letters to the school for me suggesting that they give him a card and explaining and the school were much better after they'd received those letters some school some teachers think that as a parent you're just trying to make an excuse so it was a big help when the hospital wrote so that was good" (P19)

Even when there was one member of staff that had a key responsibility for their child while at school, some parents were surprised to find them lacking in a good understanding of CF.

"I think it was a bit shocking that the school secretary who deals with the illnesses and whatever she didn't know they shouldn't even mix she wasn't aware" (P23)

On the other hand, there were parents that spoke very highly of their school and how supportive they believed them to be.

"[CF Child]'s just started the school she started in September and they seem to be really good really supportive" (P21)

Several of the young people included in the study had learnt about CF within school lessons. Parents were particularly sceptical of the information that was provided to the young people while learning about CF.

"And I think we sort of, you know, the kids both my kids have obviously done GCSEs Biology and they cover cystic fibrosis in that so, again, I said to the kids, know that they're giving you the worst case scenario, know that you probably know more than the teacher does about it, you know, deal with it, don't be embarrassed to go out to point out what's wrong because it's no longer like that.

Don't think either of my kids did that, but, I sort of said you can do" (P25)

Another key area that caused an impact on both the parent and young person was school absences. Missing school was challenging as the young person missed materials and had to try to catchup. It was also difficult for the young person as friendships changed quickly while at school. Each of these also indirectly impacted on the parent.

"it's always difficult to go back after a period of time being off cause the friendship groups change and you know she does struggle a little bit trying to find her way back in and get established again in that group" (P17)

The child with CF was limited in the amount they could do with their friends and often missed social occasions. If the child's friends were understanding and sympathetic this may not greatly affect the young person. However, for some young people their friends still appeared to struggle to understand, particularly at the beginning of friendships.

"I think it affected [CF Child]'s a bit when he was younger because a lot of his friends wanted to do things and some days he'd go and other days he couldn't and I think kids are strange in they sort of go oh he's being funny and that's just him but now they now I don't think it does now because a lot of people understand it the older you get they understand it's not a problem" (P19)

7.7.4.3. Acceptance of CF: Openness, Independence, and the Future

How accepting the young person was of their CF affected their and their parent's day-to-day lives. Many parents believed that their child became less open with their CF as they get older.

"he seems to have become more I suppose private about it since he's started at senior school"

(P24)

Other parents thought that they had an open relationship with their child and their child spoke to them about any concerns they have.

"I don't know but she, she was quite happy to, to sort of talk about it and... with us anyway. so yeah, I think it's just been open all along and talking about things" (P25)

The young person becoming selective with their communication about CF appeared to apply to both people they already knew and new people.

"she doesn't want everybody to know the first thing to know about her is that she's got CF and I think it was quite interesting for her in the sense that she realized it actually makes no difference"

(P25)

Loss of control of their child's health care as they got older and they became more independent was difficult for many parents. Some parents were concerned that their child did not necessarily see the role that medication played in their life and therefore may choose to stop taking it. There were a smaller number of parents that believed their child was going to be responsible with their medications as the child appeared to have accepted the need for them in their life.

"she's been really good with it, she's really taken on board you know how she's doing things for herself and just accepting it that it is part of her life" (P25)

Their child's increasing independence was challenging for some parents but most understood that it was a positive thing and wanted to encourage it. For a few of the parents this meant making a conscious effort to change their behaviours. Some parents had more difficulty than others with changing their own behaviours.

"I've said to him I will stay out of secondary school as much as I can because that's what I should do" (P4)

"I know he's more than responsible and he looks after himself well when he is away but I like to sort of make sure he's fed healthy stuff when he's home" (P19)

Their child's future was a tricky subject for parents. Parents were generally pragmatic about the limitations CF put on their child's ability to do certain jobs. However, as with most parents, the parents in this study wanted their children to achieve what they wanted in life, including getting the job they wanted.

"I encourage him to do whatever he wants to do to be fair as long as he's happy I don't really care as long as he's not sitting on his butt doing nothing I don't really care. yeah as long as he's happy" (P5)

7.7.5. Theme 5: Information and Understanding

All the parents largely held common opinions on the information they have received about CF and the understanding others had of CF. Two sub-themes were identified within this theme:

- 1. What They and Others Understand About CF
- 2. Information Received About CF

7.7.5.1. What They and Others Understand About CF

Generally, parents believed there was poor awareness and understanding of CF. Many parents mentioned how they themselves were unaware of CF prior to their child's diagnosis. Those who were aware typically had a medical background, and even in such cases they still did not believe they had a strong understanding, such as for P23.

"I think that myself included we were relatively very very ignorant about this before [CF Child] was diagnosed with it" (P23)

If an individual had a connection to someone with CF parents believed they would have some level of understanding about CF "when I mentioned cystic fibrosis, they say "what's that all about?", no-one knows what it is" (P8). Despite having someone in their family with CF a few parents

commented on how they found that their extended family had a poor understanding of CF. This often led to the extended family making incorrect assumptions about the child's CF, "they'll just be like oh he'll get better and he isn't going to get better you know" (P5).

Parents believed that their friends were also relatively ignorant of CF. For most of the parents their close friends learnt about CF as a consequence of their friendship.

"I would say my closer friends definitely do understand now, but that's through experiencing it with us" (P25)

Other people that the parents expected to have some level of understanding of CF but in actuality appeared to lack both awareness and understanding was their child's school, as was previously discussed in 7.7.4.2.

Parents believed that it was important to raise awareness of CF, and made some suggestions as to how this could be achieved. Parents' suggestions for improving awareness often related to how they had seen other charities raise awareness and included the use of social media and other media.

"people understand the word cancer because there is so much about cancer which is a good thing but there is other illnesses out there that people need to be more aware of" (P7)

7.7.5.2. Information Received About CF

Parents often discussed the information they have received or gathered about CF since their child's diagnosis. The information available to parents through the internet was generally subject to a high level of criticism, "I went to google and the first thing I read is the life expectancy is early- is midteens" (P25). Several of the parents were recommended against using any online information sources, particularly when they first received the news of their child's diagnosis.

"the advice our nurse gave us when he was first diagnosed she said don't go on the website and I do think she was right for the same reason because as soon as you go onto the forums it's people who are not having a good time or have got too much time on their hands" (P9)

Although P9 believed it was beneficial to avoid online forums, other parents saw the benefit in being able to seek advice from other parents easily.

"I am part of several Facebook groups that are just people asking for advice, you know that sort of thing [...] And I think If you google things it can give you crazy diagnoses and stuff whereas if you just ask someone who has actually been through it...it can totally take the weight off your mind of actually what it is" (P6)

A vast amount of the information parents received about CF was from their child's clinical team. This information was generally viewed very positively and regarded as reliable by parents. Parents often showed a preference for this information over the information available online.

"I always speak to them about any concerns or confusions that you've got because they are always happy to answer questions and to give advice and obviously it's the best kind of advice there are. I've seen quite a lot of online stuff going on in social media and so on group set up for sort of people with CF and so on and there are people on there asking questions and it drives me nuts I think why are you asking these questions to unqualified people. You need to speak to your CF nurse or the consultant when you're next down there you know don't be throwing it out to the public to get advice from the people that know best you know' (P13)

There was a small number of parents who had issues with the information they had received from their child's clinical team. The parent's negative perceptions typically came from the feeling that the information being received was not the full picture and the parent was not being treated as an expert in their child's condition.

"I used to sit there and say well tell me what you really think" (P10)

Several parents thought one of the best sources of information was other parents with a child with CF. Speaking to other parents was on occasion preferable to support from their child's clinical team.

"I think when you're first told your child got CF, rather than like five nurses, a doctor, a physio, a dietician come and talk to you, you should talk to the parents" (P1)

The Cystic Fibrosis Trust website was highlighted by many of the parents as being a helpful source of information. Parents believed the website was a great source of information about CF. Some parents had also reached out the Cystic Fibrosis Trust in other ways, although this did not always end positively.

"obviously, the Cystic Fibrosis Trust site is a massive help I've got so much information from there" (P20)

"I didn't really get an awful lot from the CF Trust when I did ring all that was done... I was just registered in and that was it... there was actually a local lady from the CF Trust who was supposed to contact me but that never happened" (P6)

Another key point mentioned by several of the parents was that the amount of information could be quite overwhelming and upsetting. This was particularly in reference to the information they

received when their child was first diagnosed, when too much information at one time was an issue for a few parents.

"when she was first diagnosed obviously I had information overload and if you look too far ahead it can be quite upsetting" (P21)

"sometimes finding out more information just frightens you in a way" (P11)

Summary of Results

Theme 1: Support, Charity and Fundraising

- For parents the hospital was largely their main source of support and they were generally happy with that
- Peer support was viewed very positively by most parents as it allowed them to speak to someone who understood
- A small number of parents were concerned about talking to other parents as they thought they may feel guilt for their child being 'healthy' or were worried about hearing negative stories
- Parents did not like talking to others online and did not think this was a good idea for their child to do so either
- Talking to their child about difficult subjects related to CF was an area that parents highlighted as needing support
- Most parents were aware of the Cystic Fibrosis Trust and involved in fundraising but few engaged with support offered

Theme 2: Challenges on Personal and Family Life

- Finding a balance between children with and without CF is difficult for parents
- Several of the parents believed that if asked their child without CF would say that CF had not affected their life but this was not true and siblings actually needed greater support
- Parents worried about CF straining their relationships with their children
- Parents believed the relationships between siblings and their brother or sister with CF was affected by CF
- Parents tended to prioritise the needs of others in their family
- Having to complete treatments with their child limited the amount of spontaneity parents were able to have, however parents also found it difficult to make plans in case their child with CF became ill
- Some parents were able to continue in the job they currently had, but either they had to reduce their hours or their partner had to adjust their work in order to account for this

Theme 3: Emotional Challenges and Coping

- Having a routine reducing the amount of impact treatments had on parents' lives
- Parents expressed feelings of guilt for the genetic inheritance of CF, their child getting sick, and enforcing treatment regimens; they also felt a great responsibility to keep their child healthy
- Parents suggested coping methods that involved positive reframing and avoidance
- Transitioning from paediatric services was potentially more challenging for parents than their children, particularly due to their child's increasing independence
- A few parents found it difficult to change their own behaviours as their child became more independent
- Some parents had an issue with their child's school understanding CF and some got the hospital involved to help

Theme 4: Impact of CF on Their Child's Life

- A few parents believed that their child became less open and more selective with their communication as they get older
- Several of the parents spoke about how they viewed their child as 'healthy'
- Parents were concerned over the increasing amount of treatments and complications their child faced
- Parents agreed that their child's mental health was impacted by their CF and found their child having a negative outlook on life expectancy particularly difficult

Theme 5: Information and Understanding

- Parents were particularly sceptical of the information provided to their child whilst learning about CF at school and online
- Feelings about online discussion forums were mixed
- Information about CF could sometimes be overwhelming for parents

7.8. Discussion

Parents with a child with a chronic illness or condition are well documented to be at an increased risk of poor psychological wellbeing (Besier et al., 2011). The thematic analysis of the 25 interviews collected from parents with a child with CF presented in this chapter appear to be consistent with what is expected from the previous literature. All parents identified challenges to their psychological wellbeing relating to their child's CF.

Many of the parents included in this chapter had children both with and without CF and the vast majority of these parents spoke about how they found balancing their time between their children difficult, as was previously found by Williams et al. (2010). Balancing between their children went beyond the parent's time, as many recognised disparities in both the amount of attention they gave their children and the way they parented. However, several parents believed this disparity was inevitable. There was an effort made by the parents to even out the way they treated each of their children to try and maintain 'normality'. Previous research that has touched upon this adjustment in parent's behaviour has highlighted how having to put effort into this adjustment of their behaviours can cause stress and anxiety in parents (Nabors et al., 2013).

Parents believed there was an impact on sibling psychological wellbeing. The majority of the parents believed that the sibling was likely to underreport the impact they experienced. This corresponds with the existing literature, which suggests a lack of significant problems in siblings and a disparity in parental proxy and sibling self-reported measures (Alderfer et al., 2010; Moyson & Roeyers, 2012). This also relates to the findings from the sibling analysis presented in Chapter 4 that suggested a high propensity for siblings to cope through the use of avoidant coping mechanisms. All parents with children without CF (siblings) believed there was a need for further support for siblings discordant with what was found in siblings, this reemphasises the benefits of not using parental proxy and suggests a need for further consideration.

Past research has suggested that family relationships and dynamics are disrupted by the presence of a chronic illness or condition (Ma et al., 2017). Berge and Patterson (2004) reviewed the research on families living with CF and found inconsistent effects on family relationships. They did suggest a potential increased stress in families with a child with CF compared to families with healthy children. The parents in this study identified strain in the relationship between themselves and their child, and between their child with CF and siblings. The parents largely felt a strain due to the requirement they placed on themselves to ensure that their child with CF maintained their treatment routine, which was on occasion met with resistance from the child. Tension between parents and the child during treatments reduced as the child becomes more independent with their treatments (Heath et al., 2017). The strain between siblings appears to be more complex and parents believed that in childhood it largely related to the disparity in parental attention. The potential for positive family

relationship effects have been found in those with Down's syndrome (Fisman et al., 1996) and needs further consideration in families with CF.

Poor family functioning has been associated with adjustment problems in children with CF and their siblings (Fisman et al., 1996; Olmsted et al., 1982). However, parents suggested that a strong bond between the child with CF and the sibling increased the distress siblings experience when the child with CF was going through a period of ill health. This concurred with the findings presented in Chapter 4 and previous literature on siblings of children with cancer (Labay & Walco, 2004).

All the parents agreed that their child's mental health was impacted by their CF, which does not necessarily agree with the findings from either the existing literature or the results in Chapter 5, both of which suggest psychological wellbeing comparable to the norm except at particular times of stress (Havermans et al., 2008; Szyndler et al., 2005). Parents' mental health appeared to be strongly affected by feelings of guilt both for the genetic inheritance of CF and for the maintenance of their child's health. Guilt for genetic inheritance and the negative effect this has on parent's mental health has been widely recognised in previous research (James et al., 2006) and could affect how parents perceive the impact on their child with CF.

There were two time periods that appeared to affect the parent the most. One was the time of diagnosis, and the distress experienced by parents during this time has previously been highlighted (Mérelle et al., 2003). The other time that cause parents a great deal of stress and anxiety was the period where their child transitioned from paediatric to adult CF services. Dupuis et al. (2011) found distress and uncertainty in parents whose children were about to go through the process of transitioning, echoing the findings in this chapter. The source of the distress in this sample did not appear to be due to the service delivered by either the paediatric or adult care team but rather due to the increasing independence of the child with CF. Parents feeling a loss of control over their child's medications and treatments caused them a high level of distress. It is also a possibility that the distress experienced by parents during this time had an effect on sibling wellbeing.

CF intruded on parents daily lives, affecting their ability to socialise, work and have time alone; these findings were anticipated given the previous literature (Besier et al., 2011). The parents appeared to be well adjusted to dealing with these limitations on their daily lives. The work by Wong and Heriot (2008) investigated methods of coping seen in parents of children with CF. They split the identified coping mechanisms into helpful and unhelpful techniques. Several unhelpful techniques such as self-blame and retreating from social relations were seen in the parents included in this chapter. There was also positive coping in the form of compliance (i.e. development of routines to ensure the child did all treatments), cognitive restructuring and seeking social support. Although avoidance coping was still present in parents, it was less present than in siblings and young people.

This may be due to the need for them to be consciously aware of CF at many times throughout the day.

As with those with CF, the key source of support and information for parents was their child's care team. Parents were generally happy with the support they received from the care team. Parents identified a general lack of awareness of CF along with a potential for stigma against CF, in agreement with young people and siblings. Misconceptions about a health condition can cause emotional distress in both those with the condition and their immediate family members (Pakhale et al., 2014). The support and information parents identified as needed was largely focused on other members of their family. Parents tended to put the rest of their immediate family first.

Parents spoke at length about the benefit they envisaged from increasing the amount of peer support amongst families with CF. Despite some reservations about the potential for their child either being healthier or in worse health than others, parents thought speaking to someone they identified as able to understand as beneficial. Peer support in parents of chronically ill children has been shown to reduce feelings of isolation, improve knowledge, and increase feelings of being understood (Nicholas & Keilty, 2007). Parents also thought peer support could benefit their children with CF and siblings. However, parents were very resistant to this being done through any online media, which may prevent the younger participants from engaging with forums such as those offered by the Cystic Fibrosis Trust.

As with adults with CF parents are now facing challenges that previously would not have been a concern but due to the improvement in survival CF is no longer only a childhood illness. Parents indicated they could use support in talking to their child with CF about more sensitive and complicated issues to do with their CF, such as the fertility. There is very little support offered to parents on this subject currently as there remains a limited understanding of how best to go about it.

7.9. Implications for the Sibling Experience

The implications for sibling experiences are clearer from the 25 parent interviews than they were from the young people or adults with CF, despite there again being no direct question about siblings included in the parent interview schedule. Parents acknowledged difficulties in balancing their time between their children with CF and siblings. Although most siblings felt this wasn't a big issue for them, it seemed to be quite a large source of guilt for the parent. This could in turn affect the sibling, as sibling and parent mental health have previously been linked (Williams et al., 2002). Parents identified an impact on sibling psychological wellbeing and thought that siblings underreported the impact they experienced. This may relate to potential avoidance coping in siblings, as well as siblings wanting to limit the amount of distress they caused their parents. The suggestion from parents that a strong bond between the child with CF and the sibling may increase the distress siblings experience, was more apparent in the sibling of adults with CF. Importantly it was clear that parents' psychological

wellbeing was affected by CF and this is known to have an impact on the rest of the family (Williams et al., 2002). Siblings are highly likely to be influenced by how their parents approach CF and how CF alters their family dynamics (Olmsted, Lewis, & Khaw, 1982).

7.10. Conclusion

Parents showed the greatest amount of emotional distress of all the family and yet focused their attention on improving the lives and outcomes of the rest of the family. The effect on siblings was not forgotten by parents but they thought a disparity in the way they interacted with their children was inevitable; many continued to put the child with CF first but tried to compensate for this with the sibling. Both discordances and agreements were seen between this analysis and the analyses of other family members described in Chapters 4 to 6.

Chapter 8: The Family Perspective: Family Level Case Studies

8.1. Introduction & Background

The work presented so far in this thesis has emphasised the need to consider multiple respondents' viewpoints in sibling research to build a holistic understanding of the sibling experience. Research has established the importance of considering the parents' experiences of living with a child with a chronic illness or condition (Bally et al., 2018) but little attention has been given to integrating sibling and parent perspectives. Issues with the appropriateness of parental proxy measures (Moyson & Roeyers, 2012) and the complexity of the family interactions (Jessup et al., 2018) highlight the need to consider multiple perspectives, including siblings, in order to fully understand the experiences of siblings. The following chapter presents a multiple-case study consideration of families with at least one sibling, parent, and individual with CF that completed an interview. This chapter also supports the aim of developing appropriate support services for families living with CF. As it has been suggested that supporting parents may help improve sibling outcomes (Gunlicks & Weissman, 2008). Furthermore, this chapter aims to delineate the experiences of families living with CF with the hope that this will encourage better support and understanding for families living with CF.

8.2. Aims & Objectives

The primary objective of this chapter was to consider the holistic experiences of families of children with CF in the UK. Given the findings from the thematic analysis of the separate study groups, the initial aim of this chapter was to identify families that participated in interviews and qualitatively describe their experiences as a family unit. Subsequently, given the overall objective of this thesis, it was important to draw out the difference and similarities across these case-studies and identify the implications for siblings and CF family support.

8.3. Methods

8.3.1. Data Collection and Participant Recruitment

An overview of the recruitment methods and data collection methods for this study are given in Chapter 3. Further details for each group considered were given within their respective results chapter.

8.3.2. Multiple Case Study Methodology

The qualitative research methodology selected for this work was a multiple case study methodology. As previously mentioned in section 3.3.1. case studies are a commonly used methodology in qualitative research. An alternative approach to qualitative data analysis that may have been appropriate for this work could have been IPA. IPA involves an interpretative approach that aims to establish meaning behind the data (Smith et al., 1999). As the primary objective of this work was to provide a holistic overview of the experiences of families living with CF in the UK, and this is a

relatively novel population, an overview of the experiences as can be achieved through multiple casestudy methodology was viewed as more appropriate.

Yin (2003) coined the phrase multiple case-study methodology, whereas Stake (1995) termed this form of analysis collective case studies. Both authors described the methodology as a series of case studies that are not considered in isolation. Furthermore, both used a constructivist epistemological paradigm. Multiple case-studies allow researchers to discover differences both between and within cases. In order to compare between cases, it is important that cases be selected and presented carefully, to ensure a holistic understanding can be developed (Baxter & Jack, 2008). Ambiguity in the unit of analysis, and the use of the term more generally, in case-studies has previously been highlighted as a concern (Grünbaum, 2007). Previously work has suggested that multiple units of analysis can be included within one project, dependent on the phenomenon of interest (De Massis & Kotlar, 2014). Moreover, the unit of analysis has often been suggested to be interchangeable with a description of the case (Carolan, Forbat, & Smith, 2016). The unit of analysis for this chapter, therefore, is a family living with CF. By carefully considering the case studies included in a multiple case study methodology it may be possible to either establish similar results (a literal replication) or predicts contrasting results but for predictable reasons across the units of analysis (a theoretical replication) (Yin, 2003).

Conducting a multiple case study research project can be time consuming and expensive. However, the evidence produced can often be considered robust and reliable (Baxter & Jack, 2008). As this work follows on from that done in chapters 4,5,6 and 7 much of the draw backs of the time and resource consumption can be mitigated and as such using this methodology here is another way to use the already collected data to build more robust and reliable evidence about family experiences with CF.

8.3.3. Propositions

Propositions in qualitative multiple case-study analyses are comparable to hypotheses in quantitative research. Propositions can be informed by past research, the researchers' personal or professional experiences, or existing theories (Baxter & Jack, 2008). Propositions are not a requirement and are not always present in multiple case-studies. Where propositions are present, they can often be helpful in guiding the project and form the basis of a conceptual framework.

Given the researcher's high level of involvement with the data collection and analysis, along with presentation of the findings in chapters 4 to 7, it is improbable to suppose they will hold no propositions of the experiences of families living with CF. As such, the findings from the thematic analysis may be considered as propositions for the following multiple case-study analysis.

8.4. Results

The multiple case studies presented in this analysis are at a family unit level, and facilitate comparisons between family experiences but also within a family, as a story is formed through the

multiple perspectives of a life with CF within the family. Families where at least one parent, one sibling, and one individual with CF in it were included in this analysis and their experiences compiled into one case study. Following the methods suggested by Mccarthy, Holland, and Gillies (2003) incorporating multiple perspectives from a family in a case study allows the researcher to have an overview, or "bird's eye view", of the agreements and disagreements within a family unit.

8.4.1. Family Demographics

The sample for this study were taken from that collected for the previously presented thematic analyses. Eight families were recruited that contained at least one parent, one sibling and one individual with CF. A summary of the demographics collected for these families is shown in Table 8.1. All families were of white ethnicity and contained a young person with CF; while two families had two parents involved.

Family	Sibling	Sex	Age	YP	Sex	Age	Parent	Sex	Age	Marital Status	Employment Status	Parent	Sex	Age	Marital Status	Employment Status	Household Income
1	S3	F	12	Y14	F	13	P1	F	57	Living with Partner	Full Time Carer						15,000-25,000
2	S8	F	14	Y7	F	12	P8	M	36	Divorced/Se parated	Out of Work						
3	S10	М	15	Y2	M	11	P10	F	46	Married	Employed (Part-Time)						50,000+
4	S4	F	13	Y21	M	17	P13	M	52	Married	Unable to Work	P11	F	56	Married	Employed (Full-Time)	50,000+
5	S1	М	11	Y11	M	13	P14	F	44	Married	Employed (Part-Time)						35,000-50,000
6	S2	F	11	Y12	M	13	P18	F	32	Living with Partner	Unable to Work						
7	S5	F	13	Y3	F	11	P22	М	41	Married	Employed (Full-Time)	P21	F	41	Married	Out of Work	35,000-50,000
8	S11	M	14	Y27	F	21	P25	F	44	Married	Employed (Part-Time)						50,000+

Table 8.1. Family Demographics

8.4.2. Family 1

Within family 1 there was a 12-year-old female sibling, her 13-year-old sister with CF, and their 57-year-old female guardian (referred to as parent throughout the case study). All the interviews were conducted in person. MM interviewed the sibling, CL interviewed the young person, and NK interviewed the parent.

Although both the sibling and the parent seemed comfortable with the researchers they mentioned that they were hesitant to talk about CF with others in too much detail as it wasn't their condition. The young person did not express much emotion throughout her interview, and yet one of the first things the parent said in her interview was that they had sought psychological support for the child with CF because they had recently said "I'll be dead, I don't have to do it [school work]". The increasing treatments the child had to complete put them in a poor emotional state which was also hard for the parent and sibling.

All members of this family spoke about the strains on daily life and relationships they experienced due to CF. However, the parent and the sibling largely focused on how CF had a daily impact on the child with CF rather than themselves. The parent was very clear about her inability to work and the social restrictions they experienced due to having a child with CF. The parent felt a high level of guilt and responsibility for the child with CF, which meant that they were reluctant to leave the child in someone else's care. The parent spoke about several changes that had to be made in her life in an attempt to prevent the child getting ill, this included the sibling having to sleep on the sofa if they or their sister ever had a cold. This parent was also particularly concerned about the child with CFs relationship with food. It was expected that members of the family make compromises and sacrifices to support the child with CF with her eating.

The young person spoke about challenges they had with catching up with school work due to absences. The sibling also recognised this as a difficulty for her sister, but didn't believe, despite noting they were often late and distracted at school due to CF, that it had a large impact on them. The sibling continually expressed empathy and consideration for her sister but did not recognise an impact on her own life, often stating that it was "normal". However, the parent did recognise an impact on the sibling and mentioned that when younger the sibling had expressed jealousy and "wished they had CF" due to the attention the young person with CF was getting.

As the child with CF was entering her teenage years there were several issues relating to her increasing independence that came up in all three interviews. For instance, the young person was unable to spend long periods of time away from home due to her medications and treatments. The sibling felt guilty for being able to be away from home and was often resistant to doing so without her sister. Both the parent and the sibling were concerned about the young person not doing her medications and becoming ill. The child with CF would occasionally be reluctant to complete their treatments and this led to tension between the parent and the child, placing a strain on their

relationship. The sibling had a very thorough understanding of the medications and treatments her sister did, and tried to continually encourage her sister to complete her treatments.

The child with CF, and to a certain extent the sibling and the parent, mentioned that they believed that if you didn't have CF or know someone directly affected by it you could not fully understand. The parent took it upon themselves to inform the young person's friends that came over to the house about the medications and treatments the child was completing. The parent thought that the best source of information and understanding was other parents with children with CF. The parent was also particularly unhappy with what they thought was a lack of sufficient information from the hospital when her child was going in for procedures, such as a gastronomy. However, no one in this family had sought to find out what further support was available for them although when support had been suggested to them the parent had chosen to use it.

8.4.3. Family 2

Family two consisted of a 14-year-old female sibling, her younger sister (12-years-old) with CF, and their 36-year-old father. The parent and sibling interview were conducted by MM and the young person interview completed by CL. All of the interviews were completed over the phone. The parent for this family did not report their income and was the only parent in the case series to report a marital status of divorced/separated.

The parent and the young person in this family spoke a large amount about the relationship and social impact they felt from the presence of CF. The parents' work and social life had been greatly affected as, unfortunately, he felt that he had been denied jobs due to his daughter's CF. The parent also mentioned that the mother to the two girls had left soon after their daughter had received the diagnosis of CF.

The young person reported experiencing bullying at school multiple times due to their CF. The bullying was focused around her medications and others quizzing her about CF. It made the young person resistant to talking about her CF and taking medications in front of others. The parent was aware that since their daughter had started high school she was more likely to miss taking her medications. The sibling and their friends would keep an eye out for the young person during the school day and the school offered the young person a sanctuary where she could go to when upset to talk to someone.

The sibling tried to support her sister in a number of other ways. The sibling would often help her sister when trying to explain what CF was to others. However, the sibling also mentioned limiting the amount she spoke about CF to others as she knew it would upset her sister. The young person felt that if understanding of CF was better she would be far more comfortable in discussing her CF. Although the young person and her father thought you were unlikely to truly understand unless you also had CF. Being able to speak to someone who understood, but did not pity them, was suggested as helpful by both the young person and the parent.

The relationship amongst the family was generally reported very positively and both the young person and the sibling felt comfortable speaking to their parent about CF. The parent thought that his daughters may have a closer relationship than other siblings. However, the parent also recognised that the sibling was affected by her sister's CF and thought that there were times when the sibling was jealous of the treatment her sister was getting. The sibling mentioned that they felt guilt for being able to do things that her sister could not. The parent also mentioned that the sibling was more mature in his eyes compared to her peers and often helped out with her sister.

The family all spoke about the benefit they saw from remaining active while living with CF. The parent thought that the young person did so much exercise it was not necessary for her to always complete her physio treatments. The sibling often spent her spare time going with her sibling to complete some form of exercise, and enjoyed the opportunity to keep her sister company.

The parent believed that they had received good support from his child's clinical team, particularly around the time of diagnosis. The family had a particularly difficult period around the time of diagnosis which did cause the father to have periods of low mood but he felt that wasn't an issue anymore, and it was "all good now". Furthermore, the young person had not considered asking for any further support, despite suggesting she could use some help with school work. This sibling was one of the few siblings that had received direct support from her sister's clinical team. A member of the clinical team came to their house to talk to the sibling about CF and help them understand. Despite this support the sibling was still scared about reading inaccurate information about CF which could cause her to get upset.

Each member of this family tried to maintain a positive mindset, though some was potentially through avoidance. For instance, consistently throughout her interview the young person repeated the phrase "but other than that it is fine" and, similarly, the sibling said "everything is fine" several times in her interview.

8.4.4. Family 3

Family 3 was made up of a 15-year-old male sibling, his 11-year-old brother with CF, and their 46-year-old mother. They were one of the higher income families involved in the study. Again, all interviews were conducted over the phone. MM was the interviewer for both the sibling and the parent, and CL interviewed the young person.

The parent in this family had a detailed knowledge of the medical aspects of CF and gave a thorough account of her traumatic birth experience. For a while following the birth of the young person the parent had to spend long periods of time going in and out of hospital. The parent recognised this as a hard time for the sibling, especially as the sibling was used to being an only child that received all the attention. The parent mentioned that the sibling had been jealous of his brother and had once said "I wish I had CF". The sibling did briefly mention this being a difficult period but appeared somewhat

resistant to say it had a big impact on them. The sibling said that he believed there was no impact on him from his brother's CF.

The parent identified a significant impact of CF on her day-to-day life. The social and physical impacts of CF were the most challenging for both the young person and the parent. The parent thought her social life was affected because she was uncomfortable with leaving the young person at home with anyone else. The parent experienced a strong feeling of guilt each time her child got ill and this made it harder to trust someone else to care for the young person. Furthermore, the parent also mentioned how challenging it was to balance her work and the child with CF, especially in the first year or so following diagnosis.

The young person in this family reported that he had suffered from bullying at school due to physical effects of their CF. Despite the bullying this young person was happy to explain what CF was to others if they asked and had even presented about CF to his class. However, the young person mentioned that he was only comfortable talking about his CF in more detail with his close friends. The young person mentioned that he was still happy to take his medications and complete his treatments in front of the friends.

The young person reported that having a treatment routine helped them to cope. The parent also thought that having a routine was beneficial, but thought her main methods of coping were talking with friends, faith and religion. The parent also stated that she coped because she had to.

The reported relationships within this family were complex, particularly relating to the sibling. The sibling and their brother mentioned that they did have a very close relationship and didn't often talk about CF. The parent thought that CF had brought her closer to her husband and their son with CF, despite having to nag the young person to do his medications. However, she thought CF had put a "huge ridge" between her and the sibling.

The sibling mentioned that he was briefly concerned about the genetic inheritance of CF, until he gained the understanding that allowed him to not worry about it. Both the parent and the young person recognised a much larger impact on their mental health. The parent mentioned a strong feeling of grief when she thought of the life-limiting nature of CF. The parent also suggested that the young person showed signs of social, anger, and developmental difficulties, which had led to problems at school.

The parent had several issues with the support she had received. She stated that she often felt like she was being spoken down to and not given full information about her child's clinical care. The parent has also had difficulty in accessing consistent psychological support for the young person either through school or the clinical team. Neither the sibling or the young person had particularly good awareness of available support, and the sibling believed he had never received any formal support. Support the parent was particularly keen to participate in and encourage was peer support. Like the pervious families, she believed that it was unlikely that someone could understand what they went

through without going through a similar experience. While the parent mentioned that she took great solace in talking to other mothers she also believed that her child's clinical team actively discouraged her from doing so. The parent did not believe that there was currently sufficient support available, however, they had also not sought any information about the services available outside of the hospital and school.

This young person, despite only being 11, already believed that CF would put limitations on his life and the jobs he would be able to do. For instance, he had considered joining the police or the fire service but ruled both out due to his CF. The young person ended his interview by saying that "CF is hard and it limits your expectations of life". The parent mentioned that she was very open and honest with the child about the negatives of CF. She believed the young person would find out the negatives at some point and it was better that it came from her. However, the parent also thought that she tried to maintain a positive outlook overall. She believed her child was "healthy" for having CF, and that the "inevitable wouldn't happen".

8.4.5. Family 4

Within family four there was a 13-year-old female sibling, her 17-year-old brother with CF, their 52-year-old father, and 56-year-old mother. Both parent interviews were completed by LA, MM led the sibling interview, and CL led the young person interview. In this family the mother had continued to work full-time while the father had stopped work to care for their children.

The father believed there was a huge impact of CF on the family's day-to-day lives. However, he also believed that as the young person with CF was his first child there was an element of having any child having a big effect on your life and that he didn't know any other way than with CF.

Both parents felt an impact on their work from having a child with CF. As despite the father taking on full time caring responsibilities for the children, there was still a strain on the mother's work and she felt fortunate to have an understanding work place. Furthermore, the father has since struggled to get back into full-time employment. The young person was also in full time work and mentioned that so far he had not had any issues with his employers or fitting his medications and treatments around a working day. Although the young person did suggest that it was simpler to manage his routine when at school.

The mother recognising that there was an impact on her mental health from CF, although she thought the daily impact had reduced over time. In particular she mentioned being scared when her son was ill, to the extent that she would wake up throughout the night whenever he was coughing. When the young person with CF was younger the parent's mental health was also affected by an inability to go out and socialise as they were not comfortable leaving the child in anyone else's care, and didn't have a family support network they could rely on. The young person with CF also acknowledged that sometimes he could have periods of low mood, although he was not sure why, and he wasn't very comfortable in talking to family or friends about it.

The young person was currently going through the process of transitioning between paediatric and adult care services. While the parents were very anxious and concerned the young person with CF didn't report having any concerns, rather he mentioned being quite excited. The parents were worried about leaving behind the care team they had known for so long, as well as the increasing independence of their son and "being distanced a bit" from his care. However, the father stated that he had full confidence in his son to continue his treatments independently and the parents felt as though they had been prepared as much as possible for the change. The parents also saw the benefit of having more time for themselves now that their children were older and more independent. Beyond transitioning to adult services, the young person was also considering moving away from home, and as such was in the process of taking on full responsibility for his medications and treatments. The process of filling and maintaining prescriptions seemed to be the one causing most concern, particularly for the father.

Both parents felt as though they could speak to and lean on each other whenever they needed support. However, the mother also thought that being able to participate in peer support and offer advice to other parents would be positive, although the individuality of children with CF meant it may not always be helpful to compare experiences. The young person had recently joined an online chat forum for people with CF, which he thought was helpful for them as although his friends without CF tried to understand, the young person thought it wasn't easy for him to do so.

The mother recognised that there was support available for herself and her family should they ask, however she had never felt the need to ask. Both parents appreciated the way they had been supported, and thought being given the information they need when they needed, particularly around the time of diagnosis, was a good way to ensure they did not become overwhelmed.

The parents considered having a second child very carefully before having the sibling. The four-year age gap between the sibling and her brother was suggested to potentially reduce the impact on the sibling. It was also mentioned that the relationship between the sibling and her brother was not very close. The sibling knew some information about CF which she thought she had gained from living with her brother, but she had no real interest in learning anything further about CF. The sibling was open to talking about CF if anyone had a question and thought it could be interesting to meet another sibling but reported that she wasn't in a rush to do so. She didn't know of any support services available to her and wasn't sure she would engage with any support services even if she was offered them.

This family put a lot of emphasis and thought into the diet needed for someone with CF. The parents thought this could have been one source of tension felt by the sibling, as her brother was on a high fat diet that she was not allowed. The parents built lots of recipes with a high fat content, to the point that the mother mentioned creating a recipe book for families with CF. Along with the focus on diet the parents also emphasised the benefits of raising a child that loves sports and wants to stay active, although the father worried he had failed to bring his children up this way.

Overall the family considered themselves "lucky" as the young person with CF was a relatively healthy person with CF. The young person thought it was hard for him to explain what a life with CF was truly like as it was normality for them, "a lot of things that just seem normal to me other people would notice as different".

8.4.6. Family 5

Family five consisted of an 11-year-old male sibling, his 13-year-old brother, and their 44-year-old mother. MM led the interview with the sibling, the parent interview was led by LA and the young person was led by CL. This family's income was in the mid-range for the sample.

The parent in this family believed that as it was her oldest child that had CF, to a certain extent it was all they knew. However, she did note a few ways their life had been impacted by CF. For instance, she mentioned that she had found it hard to balance her part-time work and caring for her children, but believed she was fortunate that her employers had generally been very understanding and flexible.

In terms of her mental health the parent thought that one of the hardest periods for her was the time leading up to diagnosis when she wasn't sure what was wrong with her child. The time following diagnosis was also difficult as she did not have a family support network to rely on, and felt somewhat socially isolated. The parent mentioned that the young person found it particularly difficult when the topic of life expectancy was brought up.

The parent believed she may have kept the sibling from maturing as quickly as may have been expected. When she was trying to juggle two children, one with CF, it was sometime easier for her to restrict the independence of the younger sibling. This sibling believed they may have a closer relationship with his brother compared to other siblings. The sibling initially said in his interview that there was no effect of CF on their school life, that they had little involvement with their brother's treatments and that he was comfortable with talking to either his friends or parents about CF. However, the sibling proceeded to talk about how he found it very difficult when his brother had to spend time in hospital. Their brother being upset either due to the procedures he was having to undergo or because he was lonely in hospital caused the sibling to also become upset.

The young person felt many impacts on his school life from his CF. For instance, the young person was absent and missed GCSE class selection, and the GCSEs he was allowed to take were limited by his CF e.g. he was unable to take geography because he couldn't complete all the field trips. The young person also talked about being made to sit out of PE or having restrictions placed on what he was allowed to do by teachers. This young person also mentioned that he had had issues at school due to poor communications between teachers, meaning he had not been allowed to leave the classroom when he need to go to the bathroom. This was very frustrating for the young person and his mother. When the school did support the young person, it was often be flawed. For instance, although they gave the young person the option to sit in another room to complete their class when he was having

difficulty breathing, this made it more challenging for the young person to follow some of the work and he needed his friends to text them pictures of the teacher's board.

School friendships were also challenging for the young person. The young person mentioned that they had experienced bullying at school due to the side effects of medication he took for his CF. The parent also noticed that the young person found it hard when he returned to school after an absence due to a potential shift in friendship groups. Occasionally the young person also felt he was left behind by friends during a break when he needed to organise his medications.

Although the young person did most of his treatments independently they would on occasion forget to take their medication and need reminding by their parents. The young person was not entirely comfortable taking his medications in front of others, which prevented him from going out with friends and sometimes resulting in him hiding when taking his medications.

The support the young person with CF had received from their care team was great according to the parent. However, the parent thought support services lacked family level consideration. Although the parent thought it would be helpful to continue participating in peer support with other mothers of children with CF, it was important to remember that no two children are the same. The parent also thought that a form of peer support could be beneficial for the children with CF, but she was unsure of how this should be handled and concerned about the use of social media. The young person also wanted to speak to someone else with CF as he thought that they were more likely to understand. However, he also thought it was easier to discuss certain aspects of his CF than others, he found certain things hard to explain, and other parts uncomfortable to discuss. The sibling agreed that it was unlikely that someone without CF would be able to actually understand.

The parent suggested a few techniques she used to cope on daily basis. These included being organised, setting a routine, taking each day as it came and not looking too far ahead. She also thought she had benefited from participating in fundraising activities with the Cystic Fibrosis Trust so that she felt as though she was "doing something". The young person was aware of the Cystic Fibrosis Trust but only through fundraising activities, which he considered to be very important in order to support research into medications for CF.

8.4.7. Family 6

Family six consisted of an 11-year-old female sibling, their 13-year-old brother with CF and their 32-year-old mother. MM led the interview with each member of the family, and all interviews were completed over the phone. The parent indicated that she was unable to work, however later explained that this was not due to her child's CF. The mother sat in on the young person's interview and chose to prompt the young person and contribute at several points during the interview.

Early on in her interview the parent identified them self as a worrier and this came across through many things she said during the interview. For instance, the parent liked to keep up to date with information about CF, although believed she sometimes tried to take in too much information.

Furthermore, despite the young person with CF being only 13 the parent was already very concerned about the transition from paediatric to adult care services and leaving behind their current care team who she saw as "like family". The parent was trying to gradually let go and give her child greater independence but she continued to sit with the young person while he did his treatments just to ensure that he did everything correctly.

In terms of the family relationships, the parent thought that there may be a tension between the young person and the sibling. The parent thought that disparities in the way she had to treat her children could cause tension. For instance, the child with CF was allowed a high fat diet while the sibling wasn't. More generally the parent found it hard to balance between the two children and tried to compensate for the time she needed to spend away from the sibling. However, the sibling thought they were quite close with their brother.

The sibling stated that CF didn't affect her life. However, she subsequently went on to discuss how she worried about her brother and the number of treatments he had to complete. The sibling found times when her brother was in hospital quite upsetting. Although she mentioned being well supported by her mother and extended family during these difficult times.

This sibling also took an interest in her brother's treatments and medication and enjoyed learning about CF when she sat in on her brother's clinical appointments. The sibling also mentioned that she enjoyed supporting her brother through his treatments but mainly because she liked being able to boss her brother around.

The young person's school life was particularly affected on a daily basis. The young person mentioned that he found it hard to fit all of his treatments and medications into a school day, although he believed he had now got used to it. The young person noticed that he was different from his peers in a few ways. For instance, he was unable to follow the healthy eating diet promoted by their school, however he was quite happy about being able to eat fattier foods than his friends. Interestingly this young person also believed that he was more confident than his peers, particularly thanks to his interactions with their clinical team.

The parent tried to be open and honest about CF with her children and mentioned that she was quite comfortable talking about CF with others if it came up in conversation. The parent thought that the more she spoke about it the more she would find other people with connections to CF. The sibling was also willing to talk about CF should someone bring it up, but otherwise she chose to limit the amount of information she shared with others. Both the parent and the sibling believed there was limited awareness and understanding of CF. By increasing awareness, the parent thought other people would be more comfortable talking about CF. Speaking to other parents and siblings was viewed positively as they thought they were likely to understand their experiences better. This was also the case for the young person who thought speaking to other people with CF could be helpful for him as his friends were not truly able to understand what it was like to have CF.

As with the sibling, when they young person became worried or upset he would speak to his mother. The parent believed the support he had received from his clinical team and extended family was great. The young person's school also appeared to offer them regular support, including an additional evening class to help them catch up and a toilet pass with access to a private toilet. The young person did believe that he could benefit from additional support with staying engaged with his treatments as he often found them boring and got distracted while completing them. The parent was particularly appreciative of the support that the child's clinical team had provided them with, especially in helping to find alternative medications that reduced the length of the child's treatment routine.

Neither the young person or the sibling was particularly aware of the Cystic Fibrosis Trust or any support services available to them. However, the parent had previously used support from the Cystic Fibrosis Trust and was quite comfortable in reaching out when she was in need.

8.4.8. Family 7

In family 7 there was a 13-year-old female sibling, their 11-year-old sister with CF, their 41-year-old father and their 41-year-old mother. All interviews were conducted over the phone. The sibling and mother interviews were led by MM, the young person and father interviews were led by NK. They were a middle-income family, and the father had continued to work full time while the mum had recently left work and was currently seeking part-time work.

At only 11 the child with CF was quite independent with her medications and treatments. The mother thought that the young person could sometime rush through her treatments, so chose to keep an eye on her. The young person also mentioned that she sometimes got emotional when doing her daily medications and treatments as she was aware that it took time away from other activities she would prefer to be doing. The increasing independence of her child was already a concern for the mother, and both parents thought it was going to be difficult for them to let go but didn't think it would be an issue for the child with CF.

Despite the child with CF being younger than the sibling the mother didn't believe she knew what life was like without CF, and found it hard to see the impact it had on her life. The mum also believed that the family had adapted well to a life with CF through the use of a clear daily routine. However, it was challenging for the routine when new medications or treatments needed to be added. The mother also believed that she was able to cope because she tried to live in the "here and now" and not think too far into the future.

The mother felt a social and work impact of CF on her life. She was concerned about placing burden on other family and friends and chose to limit the amount she spoke about CF with them. The mother thought it was easier to talk to other parents with children with CF as they were better able to understand. This led the mother to seek peer support through online sources.

The mother was reluctant to go back to full time work for concerns of the demands of CF and how this would affect her while at work. However, both parents believed their employers, especially around the time of diagnosis, had been very understanding.

Both parents felt an impact on their mental health. The father believed his mental health related closely to the health of his child; when his child with CF was unwell he thought more about the future which made him angry and upset. The mother had been diagnosis with postnatal depression following the birth of the young person. The mother was concerned that the sibling internalised the impact they felt, while the father was concerned that the young person was growing increasingly affected. The mum hoped that should the sibling be going through a hard time she would be able to reach out and talk to someone. Both the sibling and her mum thought it was particularly difficult for the sibling when her sister went to stay in hospital. During this time the sibling said she missed and worried about her sister.

The family relationships and dynamics were affected by the presence of CF. The parents thought that all the family were closer and their relationships stronger due to CF. The mum thought that her husbands level-headed nature allowed him to support her when she was struggling. The mum found it hard to discuss CF with the young person and was selective about which topics she discussed with the child. In particular, the issue of life expectancy was not a discussion she liked to have with her children. The father also thought it was best to keep conversations about CF light hearted.

None of the family were particularly comfortable with discussing CF with others. The father thought that discussing CF with other people could make them awkward. All of the family agreed awareness of CF was poor and that it would be easier to discuss CF with people that understood. Both the young person and the sibling were more comfortable in talking to their family when they were upset. However, the father didn't want to talk to his partner as he worried about burdening them. The support suggested by the family centred around communications about CF, particularly within the family unit.

The sibling had little active involvement with her sister's care. However, she thought that sitting with her sister while she completed her treatments had helped them build a closer relationship. The mum also thought that the sibling and her sister had a closer relationship. The sibling mentioned that she liked to exercise with her sister, and tied this in to how she viewed fundraising as very important as they planned to run a marathon together. The father also emphasised how important he thought it was to raise his children to live an active lifestyle.

The family believed the support they had received was generally good. The parents had used psychological support from their clinical team previously but were not sure how to approach the idea with the sibling. The information received at the time of diagnosis from the clinical team was viewed as potentially too much and overwhelming for the parents. This led the parents to seek support from the Cystic Fibrosis Trust around the time of diagnosis. The young person was aware of the Cystic

Fibrosis Trust but had never felt a need to investigate the support services available to them. The sibling hadn't used any support services and did not believe she needed support.

Overall, the family tried to maintain a positive outlook. The father mentioned a belief that certain things would not affect his family. The mum hoped that an upcoming hospitalisation would be a "reset" that would allow the young person to have another long period of good health.

8.4.9. Family 8

Family 8 included a 14-year-old male sibling, their 21-year-old sister with CF and their 44-year-old mother. All interviews were led by MM and conducted over the phone. This family had lived in a different country for the first part of the child's treatment before moving to the UK, and thus offered several observations on difference in care.

This family was distinct from the others involved in the study as the young person with CF was at university and did not currently live at home. This allowed the parent to be less involved in the young person's care. Although the parent had made a conscious effort to "back-off" and allow her child with CF to become more independent she was still concerned. The young person gaining independence was a gradual process, including a period where the parent would regularly text her daughter to check she was doing her treatments. The young person believed she had gained full independence with her treatments only that year. However, even though the young person attended adult care services her mother still attended many of her appointments.

The hospital the young person selected to attended for her adult care was closer to her university and further from the family home. When her daughter was recently in for an inpatient stay the parent tried to make the trip every day, which was quite physically tiring for the parent. The young person appreciated her family making the trip to visit and acknowledge it was difficult. The young person mentioned that she was particularly grateful that her mother was there with her. The young person mentioned seeing her mother as "a comfort blanket".

Her daughter having to have a hospital stay made the parent feel as though she should start to nag her about her treatments again. The parent believed that the adult care services seemed to be much more patient guided, and thought her daughter was much better prepared for the change in services thanks to a paediatric consultant that addressed the young person directly. The young person also spoke about this and how they found it beneficial. Generally, the young person thought her experience of transitioning between care services had been easy.

The young person had found adhering to her medications more challenging since she moved away from home. She mentioned finding it difficult building a new routine and being comfortable taking her medications in front of new people. Maintain her motivation and treatment consistency was particularly challenging without the support of her family. However, once she entered a relationship she found the support of her partner encouraged her to consistently complete her medications and treatments

The parent thought that there were periods the family had gone through that were more difficult for her mental health than others. For instance, she mentioned issues her daughter had with eating when she was younger being a particularly difficult issue. More generally the parent believed that she did on occasion have sad moments, particularly when she was thinking of the future. However, the parent wanted to focus on the here and now, "she's quite healthy now, just be quite positive on that". The young person also spoke about a period of time during high school where she struggled with her mental health and often had thoughts of "why me?". Although the young person thought she had adapted and were now able to cope better.

The sibling identified them self as a naturally anxious person and thought his anxiety increased when his sister was in hospital. Despite this, the sibling didn't want to go visit his sister in hospital, rather he mentioned going only because his parents were going.

In terms of her work life the parent was thankful that she had been able to continue working thanks to her family support system. Although generally positive the young person had experienced mixed responses from her employer when she had missed work when she needed to go into hospital. Inpatient stays had also interrupted the young person's education at times, but she had been able to apply for special circumstances to limit the interruption.

The parent didn't believe they felt any effect from CF on her relationships either within the family or with friends. Both the young person and the sibling thought they had a potentially closer relationship due to CF. However, the sibling also mentioned that he didn't get along very well when they were younger, and thought this could relate to the time their sister spent away from home while in hospital.

The young person and the sibling were taught about CF at school. The parent encouraged them to recognise themselves as an expert in the topic and to correct the teachers if they believed the teacher was incorrect. Neither the young person or the sibling had an issue with studying CF, however, the parent was not very happy about some of the information presented to her children, and shared this fact with the school. The young person wanted to learn about CF but she didn't want to over-inform herself unnecessarily, "I like being informed when I need to be informed".

The parent recognise that it was hard for her friends to fully understand what a life with CF was like unless they had experienced it. When the parent was first informed of her child's CF she used the internet to investigate CF, at which point she was faced with the worst-case scenario. The parent now saw themselves as on a mission to educate people that CF is not all doom and gloom.

General the family was very open with their communication about CF and tried to keep discussions light hearted when possible. The parent would openly discuss aspects such as genetic inheritance with the sibling and lung transplant with the young person. However, as the young person had gotten older she had chosen to change the way she communicated about CF outside of the family. When she moved to university she found it hard telling new people about her CF, as she was previously

used to it being common knowledge. Given this she chose to limit the who she told, what she told them and when, as she "did not want to be defined by their CF".

Overall the family felt that the understanding of CF in the general public was limited. However, they were willing to speak to others should they have questions. The young person would rather people spoke to her about CF than continued to hold misconceptions about CF. The young person thought that due to the personal nature of CF it was good for her to get to explain her experiences with CF should someone she knows be unsure.

The parent thought that the sibling restricted the impact he expressed. The parent believed the sibling would say there was no impact on him, and that he was less willing to tell the parent when he was ill due to his sister's CF. The sibling, separately, also agreed with this and said he didn't like to complain too much as "if she is going through all that, what reason do I have to complain with what I'm going through". Further, the sibling also suggested he could sometimes find it hard when other people complained.

The family had made friends with other families with children with CF before the infection risk was known. They still stayed in touch with many of the families and found it interesting to hear about their experiences. Although the parent had positive experiences with peer support she also felt an amount of guilt talking to other parents whose children were not as well as her own. Rather the parent preferred to rely on her family when looking for support. Both the young person and their sibling also thought that the support they received from their parents was best. The parent had tried to offer the sibling counselling but he had refused to take it.

The family was aware of the Cystic Fibrosis Trust however they had never investigated or used their services. Rather they tended to focus and prioritise fundraising. The young person thought it could be beneficial to have greater transparency about where the money was going from fundraising. Research was highlighted by the sibling as being an area he considered important, and the young person was also very interested in being involved in future research.

Overall the sibling thought it was hard to say there was no impact on his life, but he personally didn't notice a big impact. The sibling mentioned that he hadn't noticed a difference with his sister now living away from home, and thought this could be because of the time his sister spent in hospital. The parent thought the biggest problem for the sibling, particularly when he was younger was the split of the parent's time. However, the sibling thought his parents had always had enough time for him.

8.5. Discussion

It has previously been suggested that families living with CF are able to adapt well (Berge & Patterson, 2004). From the eight case-studies presented above it is clear that families have varied experiences and some are far more affected than others. Both similarities and disparities in experiences, emotions, and opinions were evident within families included in this study. Multiple case-studies allow for comparisons both within and between cases (Baxter & Jack, 2008; Mccarthy et al.,

2003), and therefore, to fully appreciate the holistic understanding of the sibling experience within the family unit, it is important to further discuss agreements and disagreements between the cases.

Parents must frequently focus their attention on their child with CF (Foster et al., 2001). Several parents believed that this disparity in treatment between the sibling and the young person negatively affected both the sibling-young person and the parent-sibling relationship. One parent (family 3) described the "large ridge" they saw between them and the sibling due to time spent away from the sibling when their child was first diagnosed with CF. However, across all families the siblings did not believe this disparity caused them any difficulties. Although compensatory behaviours were suggested in the thematic analysis, it was only the sibling in family 6 that mentioned their parents trying to compensate.

The siblings included in these case-studies agreed with the findings from the thematic analysis that overall, they viewed very little impact on themselves. However, there were certain events that were highlighted in multiple cases by siblings and their parents as potentially affecting them. Several siblings mentioned that when their brother or sister spent a period of time in hospital they worried about them (families 3, 5, 6, 7 and 8). Furthermore, when the sibling was older than their brother or sister the diagnosis period could be challenging given the amount of time parents need to spend away from home, this was particularly apparent in the parent's contributions to case-study 3.

Siblings that mentioned they felt a closer relationship with their brother or sister tended to view a greater impact on them, particularly in terms of their mental health. A close relationship between siblings has been associated with greater impact in previous studies (Ma, Roberts, Winefield, & Furber, 2017). Being close to their brother or sister led siblings, such as those in family 1 and 2 to express feelings of worry in daily life (such as at school). Parents form families 1, 2 and 3 spoke about siblings being jealous of their brother or sister with CF. This jealousy generally linked to the attention the young person with CF received, and lessened when the sibling gained a greater understating of CF and the impact it had on their brother or sister.

Across many of the families the siblings did not believe there was an impact on their social life. However, the siblings from families 1 and 2 mentioned that they could often felt guilty because they were able to participate in activities and attend social occasions that their brother or sister could not. Feelings of guilt have also been noted in siblings of young people with autism (Petalas, Hastings, Nash, & Duff, 2015). A couple of parents also spoke about feelings of guilt. The parents from families 1 and 3 felt guilt for when their child became unwell. The risk of their child becoming unwell along with the treatment requirements reduced the ability for parents to leave the child in someone else's care, as mentioned by the families 1, 3, 4 and 5. Despite this strain, the parents' relationships did not appear to be significantly strained by the presence of CF, as may have been expected given previous literature (Rivers & Stoneman, 2003). Several parents mentioned they felt closer to their partner due to CF

(families 3, 4 and 7). However, two parents also mentioned they were divorced, although in both cases this was from the point of diagnosis rather than a strain from living with CF.

Only one sibling in the case studies had received direct support from their brother's or sister's clinical team (family 2). The sibling from family 6 received indirect support as they believed sitting in on their brother's clinical appointments helped them to learn about CF. The majority of siblings mentioned that they were well supported by their families and that they did not believe they required any additional support. Several parents believed the sibling was impacted to a greater extent than the sibling expressed. Parents from families 1 and 3 were concerned about the sibling internalising their feelings and believed greater support for the sibling would be helpful.

In agreement with findings from all the thematic analysis each family appeared to believe that there was poor awareness of CF in the general public. The vast majority of participants also believed that it was hard for others to truly understand a life with CF unless they were living it. Following from this belief participants limited the amount they spoke about CF with others, as they felt it was highly probable that those they spoke to would hold incorrect assumptions and beliefs about CF. Participants with CF were concerned about how they were viewed by others, as has been noted in other illnesses (Murray Jr, Daniels, & Murray, 2006). Siblings and parents also regularly limited their communications about CF as they viewed it as personal information for the individual with CF.

Peer support has been described as an opportunity to speak with someone who understands (Dennis, 2003). The families included in this analysis agreed with this view and the majority believed peer support could be beneficial. Parents were particularly enthusiastic about peer support for themselves, although due to their view of their child as different from others with CF, they thought comparing notes about children with CF could be difficult. The mum from family 8 believed that she could feel guilty when speaking to other parents whose children were in worse health.

Overall, the families tried to maintain a positive outlook, although this was regularly done through avoidance coping mechanisms, as mentioned in the thematic analysis results. For instance, in family 2, the sibling repeated the phrase "everything is fine" several times, the parent said "it's all good new" multiple times and the young person said "but other than that it is fine" regularly in their interviews. This may support the family in not thinking too far into the future with a life-limiting condition such as CF, although could have long term implications. Based on the above we can suggest that the need for support in families with CF appears to centre around recognition of need in siblings, access and promotion of available support services, and increasing independence in those with CF and how this affects the whole family.

8.6. Strengths and Limitations

Limitations of this work include those that apply to the initial thematic analysis presented in chapters three to seven. Further limitations of this study relate to the case study methodology. Limitations of a multiple case study methodology include the time consuming and resource demanding

nature of the methodology, however, this disadvantage was absorbed by the thematic analysis project already completed (Baxter & Jack, 2008). This work is also strengthened by the rigorous and trustworthy thematic analysis that underlies it. Further, the use of multiple respondents and the inclusion of families from across the UK, allowed a holistic understanding of the family experiences to be presented.

8.7. Conclusion

These findings can be used to enhance the understanding of families living with CF. Particular emphasis is needed for a greater understanding and recognition of the experiences of all the family by clinicians and others involved in the care of those with CF. The family unit is highly interconnected in a life with CF and key aspects such as the strain on siblings has not yet been fully addressed. Providing consistent support throughout life with CF is a necessity, although this needs to be dynamic, as well as patient and family centred in order to provide the most relevant support in this ever-changing environment.

Chapter 9: The Quality of Life of Sibling with a Brother or Sister with CF

9.1. Introduction

When a member of the family has CF the experiences of each family member appear to be highly interconnected (Williams et al., 2002). Despite the interconnectedness of the QoL of siblings and their immediate family members it is not suggested that sibling QoL should be evaluated at the family level (Moyson & Roeyers, 2012). Within this chapter the impact on siblings with a brother or sister with CF is further developed using a mixed methods approach. Utilising quantitative measures of QoL alongside the previously presented qualitative results increases the breadth and depth of understanding of the experiences of the siblings with the context of the family (Johnson, Onwuegbuzie, & Turner, 2007).

9.2. Background

Psychological measures may be insufficiently sensitive and clinical thresholds not readily met (Hartling et al., 2014). The impacts on siblings and their families are broad and as QoL measurement incorporates several different aspects of life including psychological wellbeing it has been suggest to be a more appropriate measure (Hartling et al., 2014).

9.2.1. QoL in Siblings with a Brother or Sister with CF

The evidence on QoL in siblings with a brother or sister with CF is limited. Literature tends to have focused on other chronic illnesses or conditions such as cancer and epilepsy (Bansal, Sharma, Bakhshi, & Vatsa, 2014; Limbers & Skipper, 2014; Wood et al., 2008). Studies have suggested sibling QoL is comparable to that of a normative sample (Bansal et al., 2014; Havermans, Croock, et al., 2015). Other studies have noted a slightly lower QoL in siblings (Packman et al., 2005; Wood et al., 2008). Finally, it has also been suggested that siblings may have a more positive QoL compared to their peers in certain domains (Havermans et al., 2011). Fundamentally large heterogeneity in the study methods, including the QoL measure, convenience sampling issues, control group use, and the use of parental proxy measures, may explain some of the difference in QoL found (Limbers & Skipper, 2014). Parental proxy-reports have been found to result in both reports of higher and lower QoL scores relative to sibling-self report (Mazaheri et al., 2013).

Characteristics of the sibling, their brother or sister, and their wider family have also been connected to variation in QoL scores. Siblings older than their brother or sister with CF may have poorer outcomes, particularly in terms of self-esteem (Limbers & Skipper, 2014). Although it may not be possible to isolate a direct relationship between sibling QoL and CF (Havermans et al., 2011). There is an overall scarcity in evidence considering adult sibling QoL, although there is some evidence that suggests poorer mental health outcomes (Hallion, Taylor, & Roberts, 2018), and a greater number of risky health behaviours (Buchbinder et al., 2016; Lown et al., 2013), both of which can negatively affect QoL. There is also a small pocket of research on the effect of gender but this needs further

consideration (Barlow & Ellard, 2006). Furthermore, a greater CF severity has been connected to poorer QoL in siblings of those with CF (Havermans et al., 2011).

9.2.2. QoL in the Sibling's Immediate Family

The relationship between QoL in siblings and their immediate family has been discussed in the literature. The QoL reported by siblings is typically higher than that of their brother or sister, as has been found in studies considering children with cancer (Bansal et al., 2014; Norris, Moules, Pelletier, & Culos-Reed, 2010), and epilepsy (Baca, Vickrey, Hays, Vassar, & Berg, 2010).

The QoL of young people with CF compares to that of normative samples (Havermans et al., 2008). The lung function of adults with CF correlates closely with both their depressive symptoms and quality of life (Riekert et al., 2007). Cronly et al. (2018), however, found that mental health variables explained a greater amount of the variance in QoL scores than physical health indicators. Mental health outcomes in parents have also been linked to the health of their child with CF (Brucefors et al., 2015). Yet in the study by Staab et al. (1998) QoL scores in parents were not significantly associated with the severity of their child's condition, rather the QoL of parents was strongly associated with the use of coping mechanisms.

9.2.3. Multi-Respondent Mixed Methods Research

The potential for variation in sibling experiences and QoL is highlighted by the previous research on this subject, which has a high level of heterogeneity both in the methods used and the results found. Considering the experiences of siblings with a brother or sister with CF alongside their QoL through mixed methods research will help to illuminate how they relate and generate a greater breadth and depth of understanding (Johnson et al., 2007). Mixed Methods research allows the production of knowledge that may not have been available through the use of one method alone and a greater confidence in study findings (White, Judd, & Poliandri, 2012).

Given the interconnectedness of the family and disparity in the results from parental proxy-reported and self-reported measures, the use of multi-respondent methods has been recommended for family-centred research (Kraemer et al., 2003). This is important in sibling research, and in particular, research on sibling QoL, given the disagreement found between parental proxy-reported and self-reported measures in siblings (Mazaheri et al., 2013). Using a multi-respondent mixed-methods approach to establish relationships between sibling self-reported QoL and experiences, and the self-reported QoL of their immediate family in a life with CF will allow a more well-rounded picture of the sibling's life to be established within the context of their family.

9.3. Aims and Objectives

This chapter's primary objective is to provide an initial tentative consideration of any potential trends between the qualitative research findings presented in chapter 4 and the QoL data provided by

siblings and their families through the use of a multi-respondent mixed-methods analysis. Through this analysis this chapter aims to consider the following points:

- 1. How does the quality of life of the siblings, parents, and those with CF compare to each other?
- 2. Are any visible trends present between the amount siblings spoke about the pre-established themes and sub-themes and the QoL scores of the siblings, their parents, their brother or sister, and family level demographic factors?

9.4. Methods

9.4.1. Demographics and QoL Outcomes

Each participant was asked to provide some basic demographic data followed by a QoL measure. The demographics collected were tailored by participant group. Across families the demographics collected included: age, gender, racial background, marital status, education level, work status and family income. The QoL measure completed depended on the age of the individual and whether or not they had Cystic Fibrosis. Anyone with CF completed an age specific CFQ-UK. Children and young people without CF under the age of 18 completed the KIDSCREEN-10. Anyone aged 18 or above without CF completed the World Health Organisation Quality of Life BREF measure (WHOQOL-BREF). A copy of each QoL measure can be found in Appendices 9.1 – 9.5.

Cystic Fibrosis Questionnaire-UK (CFQ-UK): The Cystic Fibrosis Questionnaire (Quittner, Buu, Watrous, & Davis, 2000) was developed in order to assess disease specific health related quality of life in a range of populations with CF. Scores on the CFQ-UK can range from 0 to 100, with a higher indicating better QoL. The measure is available in multiple languages including French, English, German, and Dutch. There are three versions of the CFQ, which facilitates completion by anyone with CF aged 6 years and above. The CFQ-UK incorporates nine quality of life domains, three symptoms scales and one health perception scale for anyone aged 14years and above. The CFQ-UK for those aged between 11-13 years produces scores for seven QoL domains and two symptom scales. The measure has been well validated and shown strong reliability (Quittner et al., 2012). High correlation has been established between the CFQ and physiological measures that are regularly used as markers of health in CF, such as Forced Expiratory Volume 1 (FEV1) (Gee, Abbott, Conway, Etherington, & Webb, 2003). An age appropriate version of this measure was completed by anyone with CF.

KIDSCREEN-10: The KIDSCREEN-10 is one of a range of KIDSCREEN instruments available and was developed from the longer KIDSCREEN-27. It is a 10-item generic measure of quality of life, which can be used to assess health related quality of life in children and adolescents between 8-18 years of age. Scores on the KIDSCREEN-10 range from 0 to 100, with higher score indicating better QoL. The KIDSCREEN-10 maintains a good internal consistency reliability (Cronbach's Alpha =.82), and a good test retest reliability/stability (r=.73; ICC=.72). Further to this there is evidence that the KIDSCREEN-10 can distinguish between different groups of children that may be in need of further support or

intervention, for instance children with a low score on the family affluence scale (FAS, effect size d=.47), with behavioural problems or with a high number of psychosomatic complaints as rated on the Strengths and Difficulties Questionnaire (SDQ, effect size d=1.30, d=1.69 respectively) display a significantly lower health related quality of life (HRQoL) as rated on the KIDSCREEN-10 relative to their contemporaries (Ravens-Sieberer et al., 2010). This measure was completed by siblings between 11 and 17 years of age.

World Health Organisation Quality of Life Measure - BREF (WHOQOL-BREF): The WHOQOL assessment is a 100-item measure of quality of life which is regularly administered in research (Skevington, 1999). The WHOQOL-BREF is a shortened version of the 100-item WHOQOL and contains just 26-items (Group, 1998). The WHOQOL-BREF contains one item from each of the 24 facets of QOL included in the WHOQOL-100 along with two additional 'benchmark' questions on general QOL and health. Results from the WHOQOL-BREF are calculated in four separate domains and while it is common to consider each sub-scale individually (Ohaeri, Awadalla, & Farah, 2009), there has also been previous work which has considered an average of the four domains to create an overall QoL estimate (Bolghan-Abadi, Kimiaee, & Amir, 2011). Each sub-scale of the WHOQOL-BREF is measured on a scale of 0 to 100, with a higher score indicating better QoL. The WHOQOL-BREF has demonstrated good to excellent psychometric properties of reliability, and promising results in initial tests of validity in both healthy and unwell samples (Skevington, Lotfy, & O'Connell, 2004). This measure was completed by parents and siblings aged 18 years or above.

9.4.2. Data Analysis

There are various approaches that can be taken to mixed methods research. This work takes a convergent approach (Creswell, 2014), which brings together a qualitative and quantitative data set that have previously been analysed separately to compare the evidence in both.

Qualitative Analysis: As discussed in Chapter 3 thematic analysis was used to develop themes and sub-themes from 94 interview transcripts. Each verbatim transcript was analysed according to the work Braun & Clarke (2006) and Castleberry & Nolan (2018). An inductive semantic and realist approach was taken to the analysis in order to provide a rich account of the sibling experience. All analysis for this work was conducted in QSR International's NVivo 11 qualitative data analysis software.

Quantitative Analysis: Demographic and QoL data were initially considered through basic descriptive analyses using SPSS software (IBM SPSS Statistics for Macintosh, Version 25.0). Mann-Whitney U Tests were used to compare the QoL scores from each group. The Mann-Whitney U Test compares two groups on a single, ordinal variable with no specific distribution (Mann & Whitney, 1947). The Mann-Whitney U test is non-parametric as it does not require a specific distribution of the outcome variables, alternatively the parametric t-test assumes a normal distribution. Given the small sample sizes included in this work it is not possible to establish a normal distribution. Further, a skewed

distribution is typically expected in QoL scores, therefore, a non-parametric test was appropriate (McKnight & Najab, 2010).

Given the breadth of ages of siblings included in this study (11 to 24yrs), and that there is no single QoL measure validated across the age range, it was necessary to use two different, validated QoL measures. As discussed above the KIDSCREEN-10 was completed by 10 siblings between the ages of 11 and 17yrs and WHOQOL-BREF was completed by nine siblings aged 18yrs and above. Both the KIDSCREEN-10 and WHOQOL-BREF can be transformed onto a scale of 0 to 100 and had a similar distribution of scores and hence the transformed scores were used.

Mixed Methods Analysis: The mixed methods analysis was completed using NVivo 12.4.0 Software. Demographic and QoL data were imported from SPSS into NVivo. Within NVivo it is possible to assign attributes to each participant or 'case' based upon demographic and QoL data. Attributes gathered through other family members, such as household income, which was only collected from parents and adult siblings, were assigned to each individual in that household. Finally, each case was associated with the relevant coded transcript and as such their responses within themes and subthemes. Where an interview was not completed and only QoL was available a transcript was not associated with the case.

A series of Crosstab Queries were used in NVivo 12 in order to establish if there were trends between the themes established in the sibling thematic analysis, QoL scores, and specific attributes associated with the sibling and members of their immediate family. Running a Crosstab query in NVivo produces a Crosstab matrix, an example of which is shown in Figure 9.1. A Crosstab matrix places participant attributes against previously coded qualitative data. The cells in the matrix represent the number of coded references for a theme given an attribute.

A "proportional reference density" was formed by estimating the average contribution of a sibling to the total number of references included in each theme and the proportion (%) of references they provided within their own interview. This proportional reference density was plotted against the attribute of interest e.g. sibling QoL. Trend lines were then applied to each plot and visually inspected by the student. If any potential trends were identified during the visual inspection the trend lines were kept and are shown on the relevant plots included in the results section.

This approach to mixed methods analysis is a first attempt at exploring potential trends between sibling experiences, QoL, and demographics in families with CF. The results of this analysis should be carefully considered. This is particularly important given the following caveat; The number of references does not necessarily equate to the significance of a theme for the sibling. The nature of the references provided by the sibling are not included in this analysis. For instance, siblings that have an emotional experience may be hesitant to discuss it and therefore provide a smaller number of references, but this is not to say that they are less affected. The number of references provided by a

sibling may also reflect the joint construction of meaning between the sibling and the interviewer. Even though there was as much consistency as possible in interviewer, there is still the potential for variation in the interview as interviews were semi-structured. Interviewers were free to probe to a greater extent about different topics in each interview which could have resulted in a varying number of references from siblings. Further, as the selection of trend lines to display was not statistically based but rather subjective the results need to be considered tentatively.

Query Example						
Nodes	Sex = Male (n=6)	Sex = Female (n=13)	Total (n=19)			
A General Lackerstanding of CF	73	311	384			
Changes Overiew of the Future	12	113	125			
Family Dynamicr Sister with CF	44	133	177			
How They Thinout and View CF	17	64	81			
The Challengess Directed at CF	41	172	213			
Total	187	793	980			

Figure 9.1. Example of a Crosstab Matrix

9.5. Results

9.5.1. Qualitative Analysis

19 siblings completed an interview, and were considered in the qualitative analysis. Demographic information for these siblings and their families can be found in Table 4.1 on page 79. The thematic analysis of the 19 sibling interviews resulted in six themes and 17 sub-themes. A flow chart of the sibling theme and subthemes can be found in Figure 4.1. on page 80. The six themes were: "A General Lack of Awareness and Understanding of CF", "The Challenges and Emotions Directed at CF", "Support, Charity, and Fundraising", "Family Dynamics and Focus on their Brother or Sister with CF", "Past, Present, and Future", and "How Siblings Think About and View CF". See the results section of chapter 4 (p81) for an in-depth discussion of the themes and sub-themes.

9.5.2. Quantitative Analysis

9.5.2.1. Demographics

In total 104 participants completed a QoL measure. The 104 participants were made up of 19 siblings, 25 parents, 31 young people with CF, and 29 adults with CF, and came from a total of 59 families. The median age of included siblings was 16 (range 11 - 24 years). Ten (53%) siblings were below 18 years of age and as such completed the KIDSCREEN-10, with the remaining nine (47%) completing the WHOQOL-BREF. Six (32%) of the siblings were older than their brother or sister with

CF. Thirteen (68%) of the siblings were female. Nine (47%) had a sister with CF, and 9 (47%) were the same sex as their brother or sister with CF.

Siblings & Their Families	Siblings (n=9)	Brother or Sister with CF (n=19)	Parents (n=9)			
WHOQOL-BREF Subscales (median (IQR))						
Physical	69 (56-69)		63 (53-69)			
Psychological	69 (69-75)		63 (56-69)			
Social	75 (69-94)		75 (75-80)			
Environmental	88 (81-88)		78 (69-88)			
	CFQ-UK Subscales (median (IQR))					
CFQR Body Image		89 (67-89)				
CFQR Digestion		67 (67-89)				
CFQR Eating		100 (67-100)				
CFQR Emotion		71 (69-83)				
CFQR Health Perception		56 (56-56)				
CFQR Physical		78 (65-93)				
CFQR Respiratory		72 (64-75)				
CFQR Role		75 (50-83)				
CFQR Social		72 (64-75)				
CFQR Treatment Burden		56 (39-61)				
CFQR Vitality		50 (33-58)				
CFQR Weight		67 (67-100)				

Table 9.1. QoL outcomes for Siblings and their Families

9.5.2.2. How does the quality of life of the siblings, parents, and those with CF compare to each other?

The median global QoL in siblings that completed the KIDSCREEN-10 was 75 (Interquartile range (IQR): 70-83.75) and 75 (IQR: 69-80) in siblings that completed the WHOQOL-BREF. There was no statistical difference found between the global QoL scores in individuals with CF or parents dependent on if they had a sibling/child without CF also included in the study. Furthermore, there was no statistical difference between siblings and their brother or sister with CF's global QoL scores. However, siblings did have statistically better QoL than their parents (*U=34*, *p=.011*); parent median global QoL was 71.8 (64-75.2) and siblings median global QoL across both the KIDSCREEN-10 and WHOQOL-BREF was 75 (70-80.75). Table 9.1. presents further details about subscale QoL scores for the WHOQOL-BREF and the CFQ-UK. The siblings that completed the KIDSCREEN-10 are not included in Table 9.1. as no sub-scales are available for this measure.

9.5.3. Mixed Methods Analysis

9.5.3.1. Is there a relationship between the level of reporting on the previously established sibling themes and the QoL of siblings, their parents, or their brother or sister?

Sibling Quality of Life and its Relationship to Their Self-Reported Experiences:

Two slight trends were visible between sibling QoL scores and their self-reported experiences. Figure 9.2. displays the percentage of their interview each sibling spent discussing each of the five previously identified themes given their global QoL score. Trend lines were applied in order to visually identify trends between the amount siblings spoke about each theme and their QoL scores. A slight upward trend was identified between the theme: "A General Lack of Awareness and Understanding of CF" and QoL scores. When considering the subthemes, it was possible to suggest that the previously identified slight upward trend was potentially driven by siblings with high QoL speaking more about "Discussing and Promoting CF". A slight downward trend was visible between the theme: "Changes Over Time and Their View of the Future" and sibling QoL scores. This trend appeared to relate to a negative trend between the amount siblings spoke about the subtheme of "Changes in How Their Life Has Been Effected by CF Over Time" and their QoL score. These findings, although strongly caveated, potentially agree with the findings reported in Chapter 4. Siblings talking less about the future having a higher QoL may reflect the negative effect on siblings mental wellbeing when thinking about their brother's or sister's future. Further, siblings with a higher QoL speaking more about others awareness could suggest they were more comfortable and open about CF.

The Sibling's Brother's or Sister's Quality of Life and its Relationship to Sibling Self-Reported Experiences: The QoL of the sibling's brother or sister with CF is likely to contribute to the sibling experiences. QoL scores provided by their brother or sister with CF (either a young person (n=14) or an adult (n=5) with CF), were considered against the amount each sibling spoke about each of the sibling themes as shown in Figure 9.3. Siblings whose brother or sister had a lower QoL spoke more than siblings whose brother or sister had a high QoL. Siblings may have had more to discuss if their brother or sister was struggling and had a low QoL. Whereas, siblings with a brother or sister with a high QoL could feel relatively little impact and have less to discuss.

The previously noted positive trend between sibling QoL and the theme of "A General Lack of Awareness and Understanding of CF" is reversed when the QoL of the sibling's brother or sister is considered. When this is investigated further, by considering the sub-themes, the driving force behind this slight negative trend appeared to be two sub-themes. Both "Discussing and Promoting CF" and "Those Without CF Lack Understanding" were discussed less by sibling with a brother or sister with a higher QoL. Further, a positive trend was also visible between the amount siblings spoke about the subtheme "Learning about CF and the Information Available about CF" and the sibling's brother's or sister's QoL.

A greater treatment burden in those with CF has been linked to poorer outcomes in siblings (Vermaes et al., 2012). As such it was of interest to consider the subscale included in the CFQ-UK that considered the level of treatment burden and any trends this had with the sibling experience. There was a visible negative trend between the amount siblings spoke overall and their brother's or sister's score on the treatment burden subscale. A higher treatment burden (higher score) in their brother or sister appeared to be associated with siblings talking more overall. Figure 9.4. suggests that treatment burden was negatively related to the amount siblings spoke about the theme of "Changes Over Time and Their View of the Future". Further, a positive trend was visible between treatment burden scores and the theme "The Challenges and Emotions Relating to CF". As a low score on this subscale suggests a lower treatment burden in those with CF, these trends could suggest that siblings with a brother or sister experiencing a high treatment burden discussed the future more and the challenges of CF less than siblings with a brother or sister with a low treatment burden.

The Siblings' Parents Quality of Life and its Relationship to Sibling Self-Reported Experiences:

A positive trend was observed between parental QoL and the amount siblings spoke about the themes "The Challenges of CF and the Emotions Directed at CF" and "Family Dynamics and Focus on Their Brother or Sister with CF". A negative trend was visible between parent QoL and the theme "A General Lack of Awareness and Understanding of CF", as was also found when considering the sibling's brother's or sister's QoL. These relationships are shown in Figure 9.5. No clear trends were seen between the subthemes in "A General Lack of Awareness and Understanding of CF" or "The Challenges of CF and the Emotions Directed at CF" and parental QoL. However, several trends were visible between the sub-themes within "Family Dynamics and Focus on Their Brother or Sister with CF" and parent QoL. Two positive trends were observed, when their parents QoL was higher siblings spoke more about "Attention on Brother or Sister" and "Family Dynamics and Relationships Altered by CF". A negative trend was seen between the amount siblings spoke and parent QoL for the subtheme of "Trying to Empathise and Put Their Brother or Sister First".

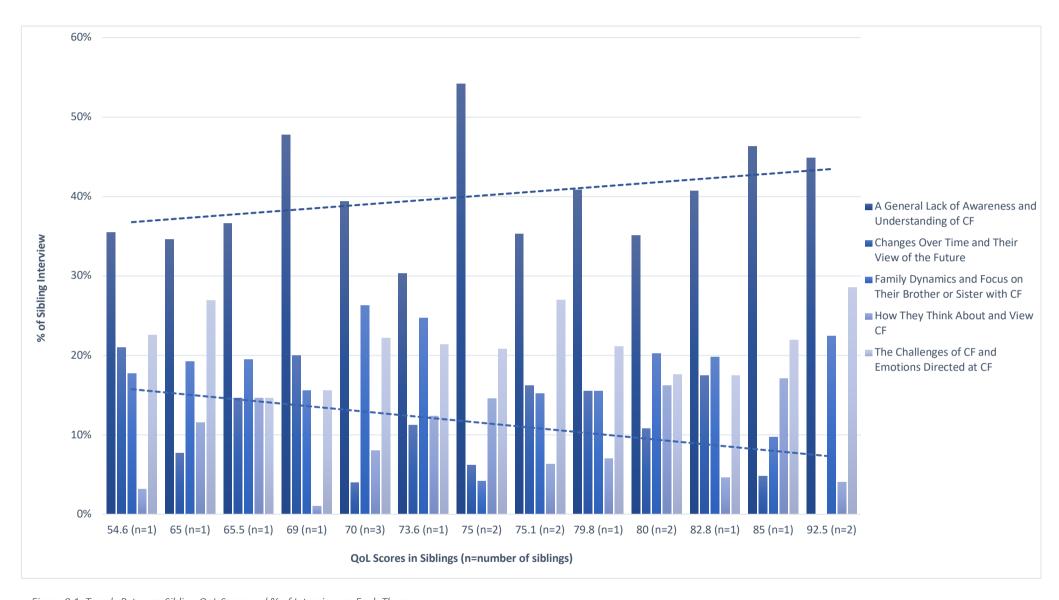


Figure 9.1. Trends Between Sibling QoL Score and % of Interview on Each Theme

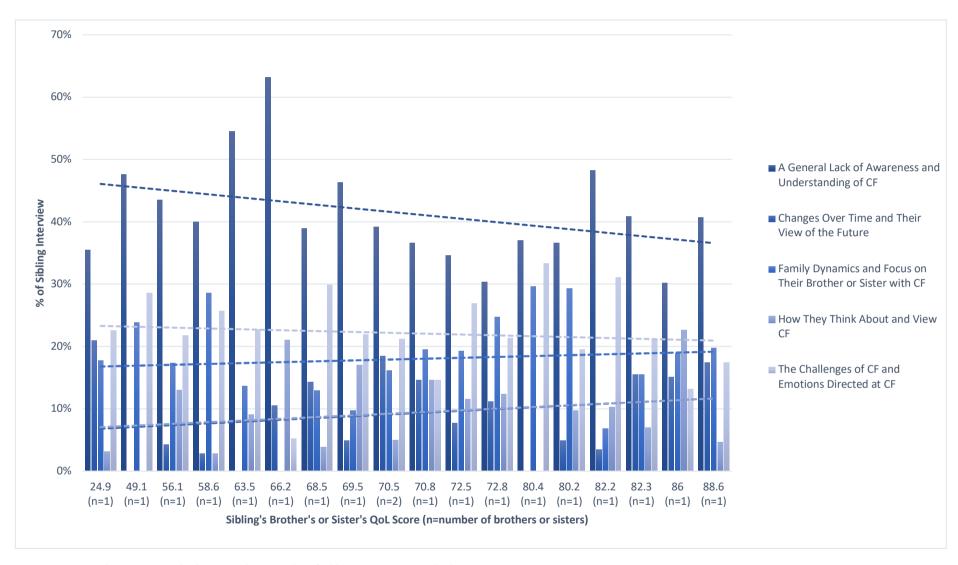


Figure 9.2. Trends Between Brother's or Sister's QoL and % of Sibling Interview on Each Theme

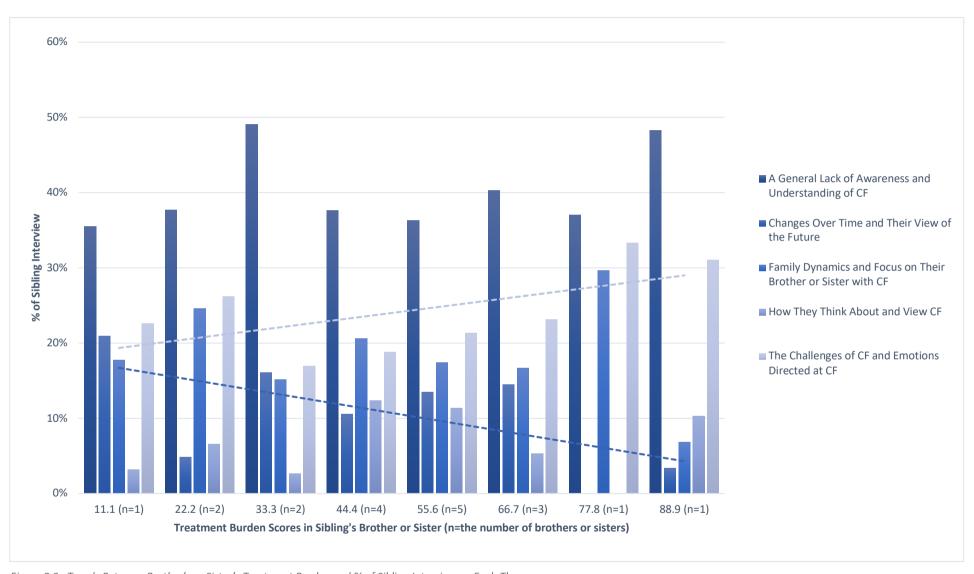


Figure 9.3 . Trends Between Brother's or Sister's Treatment Burden and % of Sibling Interview on Each Theme

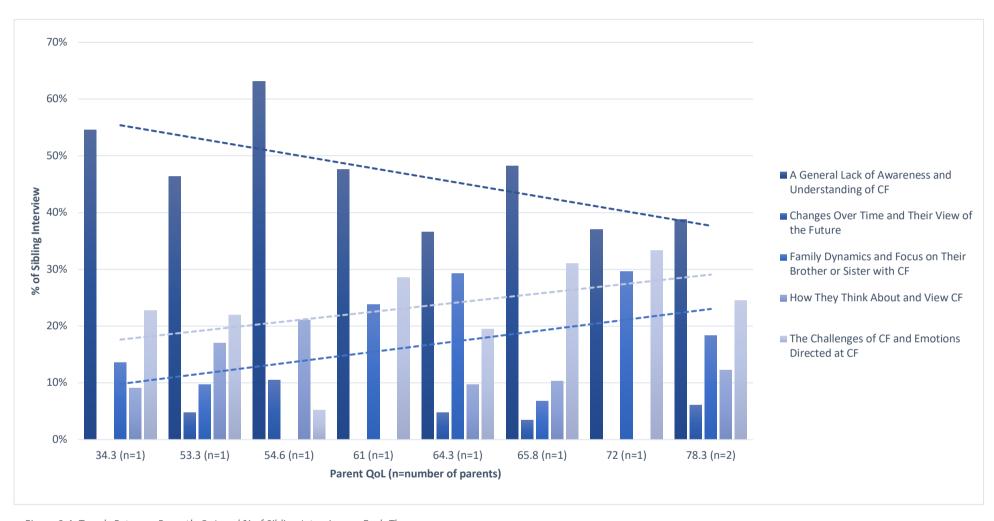


Figure 9.4. Trends Between Parent's QoL and % of Sibling Interview on Each Theme

9.5.3.2. Is it possible to identify demographic factors, such as the birth-order of the sibling, that relate to the number of references provided by siblings?

Previous research in the area of sibling QoL has suggested various demographic factors that may influence the impact on siblings (Vermaes et al., 2012). Factors such as the birth-order of the sibling, i.e. if the sibling is younger than their brother or sister, and the siblings gender relative to their brother or sister have been suggested to potentially influence QoL outcomes in siblings (Limbers & Skipper, 2014; Walton, 2016). Several demographic factors were collected for this research project. Demographics were considered in relation to the sibling experience.

The figures in Table 9.2. represent the difference in the number of references provided by two groups of siblings based on a demographic factor. Sibling experiences did not appear to relate greatly to any particular demographic. Younger siblings spoke slightly more about the theme "Changes Over Time and Their View of the Future" compared to older siblings. Siblings of young people with CF talked slightly more about a lack in awareness of CF. However, neither birth-order or age of their brother or sister appeared to have a notable effect on the sibling experiences. The second two columns in Table 9.2. are focused on variables relating to the gender of the sibling and their brother or sister with CF. The way sibling spoke was nearly identical in both comparisons (brother or sister with CF, and same sex or different sex to brother or sister with CF).

	Younger vs. Older	Young Person vs. Adult with CF (as Brother or	Brother vs. Sister with	Same Sex vs. Different Sex
	Siblings	Sister)	CF	as B/S
A General Lack of Awareness and Understanding of CF	-2%	7%	1%	-1%
Changes Over Time and Their View of the Future	8%	-6%	-1%	-1%
Family Dynamics and Focus on Their Brother or Sister with CF	-5%	-1%	0%	3%
How They Think About and View CF	2%	3%	1%	-2%
The Challenges of CF and Emotions Directed at CF	-2%	-3%	-2%	2%

Table 9.2. Comparison of Proportional Referencing for Sibling Themes Based on Demographics

Two variables relating to the parents of the sibling were considered but did not provide a large insight into the way sibling spoke across the five themes. There were two data points that deserved a brief acknowledgment. When considering the marital status of the parent the siblings who had married parents spoke more about the theme of "Family Dynamics and a Focus on Their Brother or Sister".

Siblings with parents that acted as full-time carers for their brother or sister spoke less about the theme of "Family Dynamics and a Focus on Their Brother or Sister".

Overall, demographic factors appeared to have a minimal influence on the way siblings spoke about their experiences. Factors related to the sibling's parents did not appear exert a large influence on the experiences of siblings, but what influence it did have was isolated to themes that directly related to the family environment.

9.6. Discussion

The QoL of siblings is comparable to the QoL of their brother or sister with CF. This is not necessarily unexpected given previous literature that has found QoL scores comparable to norms in both siblings (Bansal et al., 2014; Havermans, Croock, et al., 2015) and those with CF (Havermans et al., 2008). It appeared that the burden, in terms of QoL, was felt by parents whom had significantly lower QoL scores compared to siblings.

Despite a lack of evidence of a negative effect on average QoL scores in siblings, that is not to say that they are not impacted by having a brother or sister with CF. Two of the sub-themes included within the theme "A General Lack of Awareness and Understanding of CF" showed a trend with sibling QOL scores. A higher QoL in siblings was also noted to relate to the amount that they spoke about their past experiences and the future prospects associated with CF. This could link back to coping in siblings as discussed in chapter 4 as it suggested that siblings often chose to "stay in the here and now" and avoid thinking of CF in order to cope.

It has previously been suggested that when siblings have a brother or sister with a chronic illness or condition that is associated with a high treatment burden there may be a greater impact on sibling wellbeing (Vermaes et al., 2012). Accordingly, a trend was visible that suggested siblings were less likely to talk about emotions and challenges, and more likely to talk about the future when their brother or sister had a high treatment burden. Trends were also visible between the QoL of their brother or sister with CF and the previously established themes in the sibling thematic analysis.

The themes and sub-themes relating to family dynamics displayed trends with parent QoL. Further, a positive trend was visible between parent QoL scores and the amount siblings spoke about the theme "The Challenges of CF and the Emotions Directed at CF". This is particularly important in the family context where it was previously suggested that siblings will often limit the amount of distress they express to try and alleviate strain on other family members. The positive trend between "The Challenges of CF and the Emotions Directed at CF" and parent QoL emphasises that siblings may be more willing to talk about CF given that their parents are coping well and have a high QoL. This continues to highlight the interconnected nature of the family in a life with a chronic illness or condition.

Finally, the results above suggest that, in this sample, demographic factors such as gender and parental marital status have little effect on the sibling experience. The one demographic that presented a possible trend was age, as older siblings spoke more generally. Overall, however, due to the shallow nature with which these trends should be interpreted it is not possible to say for certain as to why any of the above trend exists.

9.6.1. Strengths & Limitations

The use of mixed-methods analysis is both an advantage and limitation of this study. Using mixed methods techniques allows a greater insight into the experiences of siblings than would have been achieved through only quantitative or qualitative analysis (Creswell, 2014). The increasing popularity of mixed methods emphasises that when used correctly and appropriately the consideration of the two types of data captures a more well-rounded and translatable picture of the sibling experience (Johnson et al., 2007). However, the use of mixed methods in this work was limited by the tentative nature of the analysis, as initially considered in the methodology section 9.4.2. Furthermore, the underlying assumption equating the frequency siblings spoke about themes and the significance they applied to these themes, is flawed and limits the possible interpretations of the findings.

Previously acknowledged strengths of the qualitative research project incorporate into this study such the use of robust and transparent methodology and incorporating multiple respondents within each family strengthen this tentative initial exploration of trends between sibling experiences and QoL in the family.. For instance, if the QoL of the sibling's brother or sister with CF had not been collected it would not have been possible to connect the sibling experiences with the level of treatment burden their brother or sister experienced.

Unfortunately, it is not possible to conduct a research project without limitations. Despite the overall sample size for the qualitative project, given the lack of requirement to recruit whole families throughout the study the sample size available for the quantitative and mixed methods analysis was restricted. The small sample size limited the type of quantitative analysis possible with the QoL results. Furthermore, the QoL measures completed by participants were selected to minimise the burden on participants, however the combination of the WHOQOL-BREF and KIDSCREEN-10 scores for siblings limits the clinical applicability of the findings.

9.7. Conclusion

In conclusion, the above results highlight some of the intricacies of the sibling experience when living with a brother or sister with CF that would not have been available had the qualitative or quantitative analysis be considered in isolation. While there appears to be a difficulty with measuring the impact on siblings using traditional measures there are certain time points, such as when their

brother or sister is ill, and coping behaviours, such as avoidance, identified throughout the work which may help the development and targeting of research and support in the future.

A Sibling's Experience (3)

Shown below is a first-hand account of being a sibling given by Emma* (28), whose older sister

Charlotte* has cystic fibrosis

"Cystic fibrosis has always been a part of my life. I've grown up with it, dealt with its consequences and cried over it. It is however, not me that has CF, it's my older sister.

She is an incredible human being, who will take on anything you could possibly have to throw at her; whatever it is, she might find it hard, but she will find a way to deal with it. She is three years older than me, so I was the annoying sibling that came along and ruined her sole access to Mum and Dad.

We didn't get along when we were younger, we would fight and slam our doors to the point where Dad took them off the hinges because he'd had enough, but she has always without a doubt been my hero.

Before I knew there was anything out of the ordinary about Charlotte and even afterwards, I wanted to be like my big sister. I wanted to play with her when she had friends over, I joined her in ballet and gymnastics classes and even followed her to the same university to do the same course. I used to ask when I would be old enough to start doing my puffer (nebulisers) each morning and when I would need to start taking Creon before all my meals and when I was told I would never need to, I thought I was being very hard done by and that it was outrageously unfair. Why was Charlotte so special? She had her own doctors, her own daily medication, she even got to frequently lie across Mum or Dad's lap and have her back massaged. Why couldn't I have all that?

I remember looking through photos from our childhood with my Mum and asking indignantly why there were so many more photos of Charlotte than me as a new-born. I was quickly put in my place with this response: 'We didn't think Charlotte would survive.'

It wasn't until I was about 12 years old and Charlotte was 15, that I truly realised that Charlotte was living with something that I was lucky not have. During a bad winter Charlotte had her first hospital stay and CF took its toll and brought Charlotte down emotionally for the first time that I'd ever seen. She was out of school away from her family and friends and the majority of the time isolated to a hospital room because of the risk of cross contamination with other CF patients. I wanted to visit as much as possible, but I still had to go to school and try and get on with life as best I could without my big sister around. I couldn't cycle to school with her, I couldn't ask her for help with my homework (not that I did very often, but the option wasn't even there anymore) and there was no one to argue with over the last bit of cheese. I got shipped between family and friends some evenings, while Mum and Dad went to visit Charlotte without me and although it was a great novelty to have fried eggs and

bacon for dinner, it wasn't so much fun to be having this novel dinner without my sister. She was only in hospital for a week or two, but it felt like an age and after a while it started to take its toll on me; I started to feel like the child that everyone had forgotten about and nobody really cared about, but that also left me feeling guilty because I wasn't the daughter stuck in hospital constantly attached to an intravenous drip pumping antibiotics into my body to fight a lung infection I couldn't fight by myself.

Quite selfishly at times, Charlotte's need for IV antibiotics worked in my favour. During Charlotte's second year at university she had a lung infection and had to come home during the term. By this point she was able to complete her courses of IVs at home, which meant she was able to come and support me at my last international school hockey tournament. It was great to have her there and it wouldn't have happened had she not had CF.

As Charlotte and I got older, the fighting from our childhood lessened (I can't say it stopped, as we still have our rather large disagreements on occasions) and I would happily call her my best friend, she's even agreed to be my Maid of Honour at my wedding next summer. I've been with her to checks up to play heads up and keep her entertained while we wait for the dieticians, physiotherapists and doctors to file through to poke and prod her. I've struggled to watch my big sister be punctured both physically and emotionally deflated over and over again while a nurse tries to find a vein to put yet another IV-line in. I've even fainted after being asked to help hold something in place in her vein. There have been times when I've tried to protect her on the tube at rush hour after having an IV line put in as I've escorted her on to a train where her husband would meet her at the other end. There are many things I've hated doing for Charlotte and her CF, but I wouldn't have done any of them any different. I wish I could say that I would swap places with her in a heartbeat, but I know that I am nowhere near as strong as she is.

I've been left feeling furious, when my brother-in-law has told me about the sly and ill-informed comments Charlotte has received on the netball court: 'maybe you should give up the smoking habit.' Charlotte has never touched a cigarette in her life, as a small child I used to snap cigarettes in half so that they couldn't be smoked near her. People's ignorance and their ability to jump to conclusions, that couldn't be more wrong are one of the most infuriating things about this often-silent condition (I've always refused to call it a disease).

I've tried to do my bit and raise money for the CF Trust, and I know that my small contribution will help in some way to finding that so far elusive cure for Cystic Fibrosis. I'm also well aware that the slight discomfort I feel during my fundraising Tough Mudders, were nothing compared to what CF patients deal with every single day of their lives.

Charlotte has been one of the lucky ones. there are CF patients out there who have had much harder lives and far more severe problems and complications than her, but I am selfishly and eternally grateful that it's them and not my big sister."

Chapter 10: Discussion & Conclusion

10.1. Introduction & Objectives

The evidence presented within this thesis builds on and enhances existing sibling research. The primary aim of this chapter is to interpret the findings from this thesis in relation to the overarching aim of increasing the understanding of the psychological wellbeing of siblings in families where a child or young person has a chronic illness or condition. Furthermore, consideration is given to the implications the findings have for theories of sibling adjustment and the support provided for siblings. The thesis objectives initially presented in section 1.5.1 of Chapter 1 are reiterated and considered in relevant points of this chapter. This chapter places the key findings from each of the included chapters within the existing literature and highlights both the strengths and limitations of the work. Finally, a picture of how research and support for siblings could be developed and improved is presented.

10.2. Overview of Key Findings

To fully understand how the findings from this thesis fit within the overarching aim and existing empirical evidence, it is first important to establish the key findings from each section and how they relate.

10.2.1. Chapter 2: Literature Review and Meta-Analysis of Support for Siblings

The evidence on the effectiveness of support services for siblings of children and young people with a chronic illness or condition is greatly limited due to high levels of heterogeneity. High variability in support design and the evaluation measures used reduces the ability to generalise the results beyond the populations they involve. Thus, the first conclusion to be drawn from this work is a need for consistent and robust methodology. While there is some evidence that suggests an improvement in sibling knowledge and behavioural outcomes from support, this needs to be considered carefully given the limitations of this work. What can be taken from the result presented in Chapter 2 is that there appear to be factors that may be influential on the level of adjustment in siblings. For instance, several studies proposed that poorer outcomes in siblings were associated with having a brother or sister with a condition with a greater treatment burden and mortality rate. Finally, the evidence presented suggested a lack of support for a condition specific approach to sibling support. Therefore, it may be useful to consider a support service for siblings that focuses on certain aspects of their bother's or sister's condition rather than dividing conditions by physical or mental conditions.

10.2.2. Chapter 4: The First-Hand Sibling Experience

As conditions with a high treatment burden and mortality rate were previously linked to poorer adjustment in siblings, it was anticipated that siblings with a brother or sister with CF would experience a high level of difficulties, however this did not appear to be the case. Fluctuations in the sibling's brother's or sister's health, along with the prognosis of their health condition did appear to

cause siblings of individuals with CF emotional distress. However, siblings reported being able cope with these stressors through the use of various coping strategies, in particular the use of avoidant coping. Despite a potential scope for providing siblings with a brother or sister with CF with support being identified, engaging siblings with support appeared to be a likely challenge. Reports of positive relationships between siblings and their immediate family members appeared to mitigate some of the impact experienced by siblings. Sibling recognised that their parents did their best to compensate for the time and attention they had to give to their brother or sister. Family dynamics and relationships appeared to play a critical role in the use of coping mechanisms and the ability to adapt to change in the siblings.

10.2.3. Chapter 5: Children's and Young People's Experiences of Living with CF

Relative to siblings, children and young people with CF were affected to a greater extent by CF on a daily basis. The young people also believed their family relationships, including that with any siblings, to be typical. However, the young people also appeared to be able to adapt well and used a number of coping skills in order to accommodate CF in their life. Many of the children and young people used an avoidance technique similar to that found in the siblings. Other young people adopted techniques of cognitive reframing and adaptation, in which they integrated and accepted the presence of CF in their lives. As with siblings the young people seem most affected during fluctuations in their health, such as during hospitalisations. Young people received adequate support during times of poor health and change, and spoke highly of the support they received from their clinical care team. An area of concern for children and young people was the ignorance of others, particularly given the invisibility of their illness, and the availability of accurate information. Relationships also appeared to have a key role in how the young people were able to adapt to the presence of CF.

10.2.4. Chapter 6: Experiences of Adults with CF

Adults with CF appeared to have similar experiences to those of young people with CF, however as they were typically in poorer health than the young people the intrusion of CF in their lives was greater. The adults did not speak about their siblings and the impact CF had on them, rather they spoke about their parents being affected. Adults echoed the concern about the ignorance of others and the availability of poor information mentioned by both young people and siblings. Further concerns for the adults that were not found in young people stemmed largely from complications with their health, such as increasing limits to their physical activity from symptoms associated with CF and family planning decisions. Adults' coping was similar to both that of siblings and young people with CF, with several of them using avoidance techniques where possible. It becomes much more challenging to avoid CF as their health worsened and acceptance and adaption coping techniques became more prominent. Adults had positive opinions of the current support provided to them by their clinical team

but expressed more mixed experiences with the support they had previously received and suggested they had seen improvements in services over time.

10.2.5. Chapter 7: Parents Experiences of Life with CF

Parents were notably the most affected and forthright about the impact of CF on their lives. Parents were far more emotional about CF and how it affected not only them but also the rest of their family. Parents frequently mentioned feelings of guilt and responsibility for their child with CF. They also spoke about how they noticed a negative impact on siblings and were concerned for their wellbeing. Parents coped through several methods, but primarily they used control and cognitive reframing to balance the intrusion of CF in their life. At times where parents felt a loss of control, such as during the period of transitioning between paediatric and adult CF care, parents were particularly affected. Parents believed that the support they had received was generally good. Support for parents primarily came from the care team. As with siblings and those with CF, parents reported coping through avoidant coping strategies. This was particularly apparent in the denial of the long-term implications of CF. Family dynamics were also important to the parent's wellbeing and adjustment. Parents felt a strain on their relationships with both siblings and their child with CF. However, parents consistently chose to put the rest of their family first.

10.2.6. Chapter 8: A Multi-Respondent Depiction of Family Experiences with CF

Families that included a child with CF appear to adapt and cope well and view themselves and their relationships as largely normal. By considering multiple respondents in a life with CF it was possible to enhance the understanding of the circumstances of siblings and how the family experiences interrelate. All family members echoed several of the same thoughts and experiences in their interviews. For instance, all groups typically believed that there was a high level of ignorance about CF in the general public and that many people tend to make false assumptions about CF. While overall, the support received from clinical teams was viewed positively there remained scope for support beyond this, particularly for siblings. A frequently suggested support was peer support, despite reservations about talking to others in a better or worse situation and the use of online forms. Although the daily impact of CF was felt much greater by parents and those with CF, all participants were impacted during periods of fluctuation. Several participants in each group mentioned the use of avoidance coping mechanisms and adaptation (the use of routines). In general, families tried to maintain a positive outlook.

10.2.7. Chapter 9: Mixed Methods Analysis of the Quality of Life in Siblings

Clear relationships between the QoL of siblings and the level of detail they provided for themes and sub-themes established in the qualitative analysis were found. The QoL of siblings was comparable the QoL of their brother or sister, but significantly better than their parents QoL. The QoL evidence re-emphasised the tendency for siblings to use avoidance coping mechanisms. A lower QoL

in their brother or sister reduced the amount that siblings wanted to talk about CF and the emotional burden they expressed. The QoL of the parents also related to the amount siblings spoke about the emotional impact they felt from CF. The results highlighted a potential relationship between parent and the brother or sisters QoL on sibling experience and reemphasised the need to consider the whole family in sibling research to build a full picture of the impact on sibling experiences.

10.3. Discussion Points

The key conclusions given above add to the existing sibling literature in several ways. They highlight multiple discussion points that need to be further considered and contextualised within the scope of the existing literature, and subsequently incorporated into future research and support.

10.3.1. Measuring the Impact on Siblings (Chapters 2 & 9)

The results from the review and meta-analysis in Chapter 2 highlighted concerns about how the impact on siblings was measured. Primarily there appeared to be two key concerns when measuring sibling outcomes. One concern was the use of measures that were not be sufficiently sensitive to capture the impact on siblings (Hartling et al., 2014) and the other was the use of parental proxy, which doesn't often correspond to that given by siblings (Barlow & Ellard, 2006; Mazaheri et al., 2013). Measure sensitivity was highlighted as a concern as, despite small effects on internalising and externalising behaviours of siblings being found following intervention, authors suggested there may have been a ceiling effect on the measure of effectiveness for their intervention given the number of siblings that were classified in the normal range prior to intervention (Giallo & Gavidia-Payne, 2008; Kiernan et al., 2004; McLinden et al., 1991). This could be due to siblings not reaching diagnosis criteria on psychological measures (Hartling et al., 2014). QoL may be a more appropriate holistic way to measure sibling outcomes. The results presented in Chapter 9 suggested that sibling QoL scores were comparable to their brother or sisters. Instruments tailored to measure sibling wellbeing, which consider specific factors that have been identified as crucial to sibling adjustment, such as the QoL of their parents, may be more appropriate. The findings from Chapter 9 corroborate the expected benefits of mixed methods research and should be taken as encouragement for future research to also employ this methodology.

Parental proxy was frequently used within the studies included in Chapter 2, however there were concerns raised about the potential bias these may cause (Cebula, 2012; Lobato & Kao, 2002; Sidhu et al., 2006). it is known that siblings describe QoL differently to parents (Moyson & Roeyers, 2012). When parental proxy is used there is a tendency for over-reporting of psychological symptoms and underreporting of social difficulties (Barlow & Ellard, 2006; Mazaheri et al., 2013). Collecting both the perspective of the parent and the brother or sister with CF, as has previously been recommended in families of autistic children (Kovshoff et al., 2017), was important for developing a well-rounded understanding of the sibling experience. Collecting evidence from multiple respondents has been

shown to enhance understanding (Kraemer et al., 2003) and within this work it aided the understanding of how interrelated the experiences of siblings and parents are, aspects which are common across the whole family e.g. selective communications, and topics that were only viewed as important by the sibling themselves e.g. the personal nature of CF.; each of which can be used to help enhance future sibling research and support.

Heterogeneity characterises much of the research and findings in sibling research. Heterogeneity was raised as a measurement issue that was particularly crucial and prominent in the systematic review and meta-analysis presented in Chapter 2. Inconsistency in reported treatment effects from sibling support services was previously acknowledged (Hartling et al., 2014) but there is a limited consideration of the source and implications of the heterogeneity. The systematic review and meta-analysis included in this thesis is challenged by three key sources of heterogeneity: the diagnosis of the sibling's brother or sister, the protocol (or lack thereof) of the support service, and the measure used to evaluate the psychological wellbeing of the sibling. Despite these sources of heterogeneity, it was possible for the meta-analysis to establish a small treatment effect on internalising- and externalising-behaviours, and an improvement in knowledge. These results need to be considered with caution, but should be used to inform future research in the area of sibling support and specifically the evaluation of interventions in the future.

10.3.2. Coping Styles in Siblings (Chapter 4, 8, & 9)

A central finding from this thesis focuses on the coping styles used by siblings. The qualitative analysis of sibling interviews presented in Chapter 4 drew out the tendency for siblings to adopt avoidance coping techniques. This finding was also reiterated and reinforced by the mixed-methods analysis presented in Chapter 9, which found that siblings spoke less about their emotions when the sibling's brother or sister had a poorer QoL. Avoidance coping techniques are known to reduce stress and help to increase hope and courage. Yet, avoidance coping is also connected with a greater intrusion from threatening material and emotional numbness (Roth & Cohen, 1986). This corresponds with the findings from this thesis as siblings felt a high intrusion from threatening material, such as when their brother or sister was in poor health. Siblings could also potentially exhibit 'emotional numbness' as when first asked the siblings did not believe had an impact on their lives, however, following further questioning there were clear areas of impact. Avoidance coping has previously been noted in siblings of individuals with autism (Kovshoff et al., 2017) and also bereaved siblings (Cohen & Katz, 2015). Cohen and Katz (2015) linked the use of avoidant behaviours to poor adjustment post the death of their sibling. Affective communication skills were viewed as highly important by those with avoidant attachment styles in a study by Carr and Wilder (2016). This may contribute to the desire siblings expressed in Chapter 4 for others to be informed about CF in an effective and reliable way. Carr and Wilder (2016) also note that avoidant individuals were less inclined to seek support from friends. Although this was not an obvious finding within this thesis, as several of the siblings included in the

qualitative analysis were open to talking to their friends about CF. Hesitation was noted in talking to individuals the siblings thought 'wouldn't understand', which could sometimes include friends. Finally, the tendency for older siblings to recognise and report a greater amount of emotional distress supports the findings of a previous meta-analysis considering avoidant and non-avoidant coping strategies (Suls & Fletcher, 1985), which suggested that avoidant coping was associated with greater positive-adaptation but only in the short-term. It is also worth highlighting that there is work that does not corroborate the findings of the use of avoidance coping styles, such as in siblings of individuals with psychiatric disorders as found by Pos et al. (2015).

Another slightly less frequently adopted coping mechanism found in the sibling was positive cognitive restructuring. Previously this form of coping has been positively associated with QoL, anxiety, loneliness and insecurity in siblings with a brother or sister with a cancer diagnosis (Houtzager et al., 2005; Houtzager et al., 2004). This form of coping is generally viewed as more positive relative to avoidance coping (Greenaway et al., 2015) and positive coping was a target for several of the interventions included in Chapter 2. Although this is not directly measured there is a suggested connection between knowledge of their brother's or sister's condition and positive coping in siblings (Heiney et al., 1990). The slight increase in knowledge found in the meta-analysis in Chapter 2 could imply the potential for more positive coping in siblings. Another factor that has been connected to better coping in siblings is positive family functioning (Fisman et al., 1996). This can go some way to explain why siblings appeared to have more significant improvements in coping and adjustment when their parents are involved in an intervention for the sibling, as identified in the results of Chapter 2.

10.3.3. Factors that Shape the Impact on Siblings (Chapter 1, 2-9)

Throughout this thesis there have been several factors suggested that could help explain the level of impact experienced by siblings and differences in adjustment. Chapter 1 presented the factors that had previously been proposed, these included several aspects that were investigated in subsequent work. For instance, the condition severity and treatment burden of their brother's or sister's chronic illness or condition was previously connected with poorer outcomes and experiences in siblings (Incledon et al., 2015; Rodrigues & Patterson, 2006; Vermaes et al., 2012); as was supported by the results of the systematic review in Chapter 2. This motivated the choice of CF as a case study for subsequent in-depth qualitative analysis as CF is known to be associated with high levels of morbidity and mortality. It was, therefore, expected that the experiences reported in interviews with siblings of those with CF would be negative, however for most this was not the case. Siblings included in the analysis presented in Chapter 4 were on the whole well-adjusted and several were able to highlight positive aspects from having a brother or sister with CF. This was reinforced by the findings from the QoL data. A factor that related to their brother's or sister's condition and did appeared to influence sibling wellbeing and adjustment, was periods of fluctuation and change. For instance, in the

case of CF these fluctuations could include hospitalisations, CF related complications, pulmonary exacerbations, or transplant operations.

Demographic factors that were previously associated with sibling wellbeing and were further considered within this thesis included age, birth-order and gender. Within the mixed methods analysis presented in Chapter 9 it was possible to isolate each of these factors and consider if they had any influence over the references provided by siblings for each theme established in the thematic analysis. Contrary to the findings from Walton (2016) there was no effect of gender on the reported experiences of siblings. Evidence suggested that the age of the sibling, independent of their brother's or sister's age, did influence the sibling's contribution to the themes established in Chapter 4. The findings suggested that older siblings spoke about the emotional distress they experienced a greater amount than young siblings. This may connect back to the discussion on coping mechanisms and the use of avoidance coping being both positive in the short-term (i.e. for younger siblings) and encouraging emotional numbness (Roth & Cohen, 1986). Unfortunately, given both a lack of variety in certain factors (e.g. ethnicity) and an inability to accurately measure other factors (e.g. socioeconomic status) it was not possible to account for several potentially influencing factors in the systematic review and meta-analysis, the qualitative analysis, and the mixed methods study.

A lack of understanding of their brother's or sister's condition has previously been associated with poorer outcomes in siblings, including negative effects on the sibling relationship and greater anxiety (Carpenter et al., 1990; Houtzager et al., 2001; Sidhu et al., 2006). A lack of knowledge has been suggested to reduce the adaptation and coping ability in siblings (Evans et al., 2001). The capacity for support targeting the psychological wellbeing of sibling to improve knowledge was a clear outcome of the meta-analysis presented in Chapter 2. The siblings and their families included within the qualitative analysis also spoke at length about both their understanding and the understanding and awareness of 'others'. Concerns about understanding focused around the tendency for 'others' to focus on negatives and make false assumptions about CF. The level of understanding of siblings themselves varied greatly. Siblings included in the analysis presented in Chapter 4 generally believed they had sufficient information and access to the information they needed. When seeking further information about their brother's or sister's condition siblings tended to approach either their parent or their brother or sister. Siblings viewed their brother or sister as the expert in their condition. Therefore, despite the evidence presented in Chapter 2, siblings were typically not overtly concerned with learning about their brother's or sister's condition. Again, this may relate back to the use of avoidant coping as learning would require the sibling to focus attention on their brother's or sister's chronic illness. There is evidence that suggests sibling's use of interpretive control, which is when they need to understand the meaning behind their brother's or sister's condition, can result in negative outcomes (Houtzager et al., 2005). These results suggest that knowledge improvement may not necessarily be a solution in the long-term and as in the case of the siblings included in Chapter 4 it may be positive to allow siblings to seek information from reliable sources as and when they feel they need it.

10.3.4. Family Dynamics and Relationships (Chapter 2, 8 & 9)

Positive family functioning has previously been connected with better outcomes in siblings (Fisman et al., 1996). Although no direct measure of family functioning was taken during this work there were other family related factors, which closely relate to family functioning, that can be further considered. Houtzager et al. (2004) found that stability within families in adaptation to change promoted positive outcomes in siblings. This relates to both the qualitative and mixed methods findings that periods of fluctuation or change were found to be the most stressful for siblings. Families being able to adapt well to change could limit the impact felt by siblings during periods of fluctuation (Giallo & Gavidia-Payne, 2006).

The family is highly interconnect and it is known that sibling outcomes are influenced by their parents and brother or sister (Giallo & Gavidia-Payne, 2006; Lindström et al., 2010; Walton, 2016; Wood et al., 2008). Chapter 2 suggested that siblings could benefit from the inclusion of their parent in the intervention they received. This can be associated with several possible explanations. If the intervention improved the mental health of their parent it may also improve sibling outcomes, as poor maternal mental health is associated with poorer sibling adjustment (Lindström et al., 2010). Including the parent in the intervention will increases the parent's awareness of sibling's needs. Including the parent in the interview will both require and encourage the parent to spend a greater amount of time with the sibling, which has been connected with better sibling outcomes (Murray, 2001). Within this thesis it was possible to identify that the QoL of the parent was significantly worse than that in the siblings, and that a poorer parental QoL reduced the amount that siblings wished to discuss their family dynamics. The parent having a higher QoL appeared through some mechanism (e.g. better treatment of brother or sister condition, more time for sibling) to give the sibling more space to focus on themselves as they felt less of a need to prioritise their brother or sister. Nevertheless, what was fundamentally clear from the work presented in this thesis, and in particular in Chapters 8 and 9, is that the experiences of the family are highly interconnected, as has previously been suggested (Ma et al., 2017). Furthermore, there is a benefit to understanding and completeness from the inclusion of multiple respondents from the family in sibling research (Kraemer et al., 2003).

The relationships siblings have both within their families and with their peers are crucial for development and have been found to be impacted by the presence of a chronic illness or condition (Ma et al., 2017). Despite a lack of robust evidence, it is worth highlighting that relationships were evaluated as an outcome for multiple of the interventions included in the systematic review. Evidently relationships were viewed as outcome worth considering, however, there were limited findings and a lack of consistency in measurement. Within the qualitative analysis many of the siblings viewed their

family relationships as 'normal. A few older siblings mentioned that they felt closer to both their brother or sister and parents. Previously it has been suggested that a close relationship with their brother or sister could increase the probability of siblings taking on a caring role for their brother or sister in later life (Bigby, 1998). Being close to their brother or sister did appear to make siblings more open to supporting them both practically and emotionally, as shown in Chapter 4. However, A close relationship with their brother or sister also has the potential to increase emotional distress in siblings (Sharpe & Rossiter, 2002).

Siblings that receive social support from those close to them such as friends and family can benefit from improved psychological wellbeing (Incledon et al., 2015). Emotional support from and time with parents along with greater parental awareness, have been noted to act as a protective factor for sibling psychological wellbeing (Incledon et al., 2015). It has been suggested that support from other siblings is viewed positively by siblings (Giallo & Gavidia-Payne, 2006; Sidhu et al., 2005). The siblings included in the qualitative analysis, however, had concerns about speaking to others that were either having a better or worse experience compared to them. Those with CF and parents echoed this concern about peer support.

10.3.5. Supporting Siblings (Chapter 2, 4, & 7 – 9)

Chapter 2 provides in-depth and noteworthy findings on the support currently received by siblings of children and young people with a chronic illness or condition. Points can also be raised from the qualitative research project that contribute to the understanding of sibling support. Evidently there is an issue with heterogeneity in the support currently provided to siblings and, despite this previously being acknowledged (Tudor & Lerner, 2015), little has been done to encourage consistency. Furthermore, there are issues with the way in which several of the existing interventions have been evaluated. Very few of the studies included in the reviewed used a robust methodology and almost half (47%) of the studies were considered of weak quality. There was one RCT contained within the review but due to other issues in the study design it was rated weak overall. The recommendation made at the end of Chapter 2 to encourage further robust RCTs in this area should not be forgotten.

Intervention Development Suggestions: In keeping with the objective of this work to support the design and development of support services the findings from this thesis will now be considered within an existing framework for intervention development. When developing a complex intervention, the Medical Research Council (MRC) (2006) recommended using questions to guide the development and suggest that if any response to the questions is unclear an evaluation should not yet be undertaken. The suggested questions were:

 Are you clear about what you are trying to do: what outcome you are aiming for, and how you will bring about change?

- Does your intervention have a coherent theoretical basis? Have you used this theory systematically to develop the intervention?
- Does the existing evidence ideally collated in a systematic review suggest that it is likely to be effective or cost effective?
- Can you describe the intervention fully, so that it can be implemented properly for the purposes of your evaluation, and replicated by others?
- Can it be implemented in a research setting, and is it likely to be widely implementable if the results are favourable?
- Have you done enough piloting and feasibility work to be confident that the intervention can be delivered as intended?
- Can you make safe assumptions about effect sizes and variability, and rates of recruitment and retention in the main evaluation study?

The results presented in Chapter 2 highlighted multiple existing studies on sibling support that do not address these seven questions. There was a lack of clear use of theory to help in the development and structuring of the interventions, along with a limited use of previous literature, potential due to the novelty of the subject matter. Several of the studies did provide or were working to establish a protocol for the intervention, but this was not available for all, and furthermore there was very limited consideration of adherence both from siblings and those delivering the programme to the structures provided. Recruitment of siblings provides an additional challenge and many of the studies used small convenience samples which limit the applicability of the study.

The challenge of sibling recruitment was briefly discussed in Chapter 4. Particularly given the regular use of avoidance coping techniques it can be difficult to find siblings not only engaging with sources of support but also willing to participate. Quite frequently for younger siblings it could be under the pressure of a parent that they partake in support. Beyond a resistance to engage with sources of support many siblings and also parents included in the qualitative analysis suggested a more fundamental lack of awareness of need for siblings and as such a lack of service provision. Many of these siblings and parents were unaware of services that were available for siblings, and as such there is a complex situation emerging where there is a want for greater recognition but a lack of engagement. How best to deal with this complex situation needs to be further considered.

Even though there are concerns about the support currently available for siblings along with a lack of engagement, the results in Chapter 2 also provide a promising outlook for future support as despite challenges improvements in the internalising and externalising behaviours of siblings were found along with an improvement in knowledge. Furthermore, although the majority of the siblings included in Chapter 4 were not actively engaged with support, there were many that would still be open to support should they be approached about it and the points which continue to be drawn out

as potentially important to support siblings at is during fluctuations and changes. Therefore, there is the potential for support to fill a gap which is currently not being addressed.

10.3.6. Engaging Siblings in Research on the Development of Support

Patient and Public Involvement (PPI) in research is receiving increasing attention as a potential mechanism for ensuring that research is implemented in clinical practice and makes a significant difference to the relevant population (Crocker, Boylan, Bostock, & Locock, 2017). PPI was used at several points through this thesis, including in the development of the interview schedules used for the qualitative research project. Using PPI in the development of interview schedules can help to ensure sensitive topics are approached in the best way (Brett, Staniszewska, Mockford, Herron-Marx, et al., 2014).

A useful way to receive feedback from the relevant public on several aspects of a study, in particular the design and content of research protocols and documents, is through the use of patient and public research advisory groups (Brett, Staniszewska, Mockford, Herron-Marx, et al., 2014). At the commencement of this thesis there was a limited presence of siblings within existing research advisory groups and despite a push to increase PPI no group had been developed specifically for siblings. Due to this identified gap in the PPI resources a research group made up of siblings of young people with developmental disabilities and/or chronic illness were recruited using social media, national charity links, and posters. In the designing of this group another PPI group based in London was approach to help ensure the group was appropriate for the purpose. 10 siblings were recruited to the group. Siblings were all aged 11-18. They were siblings of individuals with autism (n=7), Down's Syndrome (n=2) and epilepsy (n=1). During the first meeting of the Sibling Research Advisory Group (SRAG) the siblings were asked to contribute their thoughts and opinions towards three research projects focused on siblings. For this thesis feedback was sought on which aspects of being a sibling the group viewed as important and influencing on their wellbeing. Furthermore, the siblings were each asked to evaluate the group following their attendance at the group, and all agreed that there was a need for the group. The evaluation also indicated that the siblings found the day enjoyable and supportive, some of the feedback received can be seen in Appendix 10.1.

A subsequent meeting of the SRAG was held and the feedback on the planning and organising of a public engagement event, Sibling Research Networking (SIREN), was sought from the siblings. Along the same lines as patient and public involvement there has also been an emphasis on the need for more and better public engagement in research (Shippee et al., 2015). The National Institute for Health Research (NIHR) distinguish between involvement, participation and engagement in research. They define public engagement as an activity where information and knowledge about research is provided and disseminated¹². Sibling Research Networking (SIREN) was an open day that aimed to do

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¹² https://www.invo.org.uk/find-out-more/what-is-public-involvement-in-research-2/

three things: increase public awareness of the importance and positive impact of research into siblings' physical and mental health, give members of the public the opportunity to influence future sibling research, and bring together researchers and charities working in sibling health to encourage further collaborative work. In order to achieve these aims anyone interested in sibling research was invited to attend including families that include siblings, researchers and health care professionals. The event featured a series of discussion tables followed by suggestion development for the future of sibling research. Attendees composed posters of what they thought was the most important consideration to make for future sibling research. A couple of examples of the posters can be seen in Figures 10.1 and 10.2. Following the event attendees were asked to complete an evaluation form, which can be found in Appendix 10.2. Overall, all attendees believed the day was worthwhile and provided them with a lot of thoughts to take forward in their future sibling research.

Given the issues with engaging and recruiting siblings previously discussed, as could have been anticipated recruitment of siblings to both SRAG and SIREN was challenging. Many of the siblings in attendance at SRAG were encouraged to attend by their parents, which may potentially reduce their level of engagement. Furthermore, it became clear to the researchers in SRAG, that despite efforts to ensure clarity in the purpose of the group, many siblings and parents viewed the group as an opportunity for peer support. Although the use of SRAG as support is unintentional, the use of it as such reinforces the desire for support for siblings and a gap in the provision. The poor attendance at SIREN by families may relate to several of the factors previously discussed, however one parent at SIREN believed that a reduced capacity in parents due to having a child with a chronic illness or condition could result in parents making the choice to not acknowledge the need of the sibling. Despite these concerns involvement and engagement were seen as positive and worthwhile activities that enhance the appropriateness and reliability of the findings included in this thesis. Fundamentally, it is also important to remember that engagement activities such as SIREN continue to help increase the salience of sibling need. Increasing salience can help to encourage both parents and siblings to acknowledge the impact of having a brother or sister with a chronic illness or condition.

10.3.7. Theoretical and Clinical Implications of This Thesis

To further enhance the relevance and applicability of the findings from this thesis it is important to consider how they contribute to the existing theoretical models of sibling impact. A Diathesis-Stress model (Orsmond et al., 2009) may have been appropriate in the case of siblings with a brother or sister with cystic fibrosis given the genetic basis of the condition. Siblings did not appear to be greatly concerned about the genetic aspects related to CF and critically it did not appear to have any influence over the experiences of the siblings included in this study; in fact, many of the siblings were positive about being able to control how being a carrier could affect their lives. As such it is

proposed that this model may be more appropriate in conditions where the genetic impact on siblings is more acute and regularly felt.

The Sibling Embedded Systems Framework (Kovshoff et al., 2017) was developed for siblings with a brother or sister with an Autism Spectrum Disorder (ASD) but appears to also be a helpful theory for understanding the experiences of the siblings included in this thesis. The theory also allows the integration of the findings from the systematic review, meta-analysis and mixed methods study. The inclusion of numerous dynamic and interacting factors within each system of the framework, as shown in Figure 1.1 (page 30), allows several factors discussed in this thesis to be applied within the scope of an established framework.

Of most relevance to the thesis is the key external factor included in the micro- and meso-system called 'event'. Kovshoff et al. (2017) describe the 'event' as an external factor that impacts on the sibling and relates to the research being conducted which can be either a positive or negative element and does not necessarily need to be directly related to the child with the condition. For instance, it may be landmarks such as transitioning or diagnosis. From this work it is proposed that what may be best considered as said 'event' is periods of fluctuation or change, particularly periods of ill health for their brother or sister. Further the sibling's personal internal interpretation of the 'event' is included within the model, which could be considered from this thesis as the coping mechanism they use to deal with the 'event' and as such for many of the siblings that use an avoidant approach the presence of the 'event' reduces their ability to avoid their brother's or sister's health condition and increases the intrusion they feel.

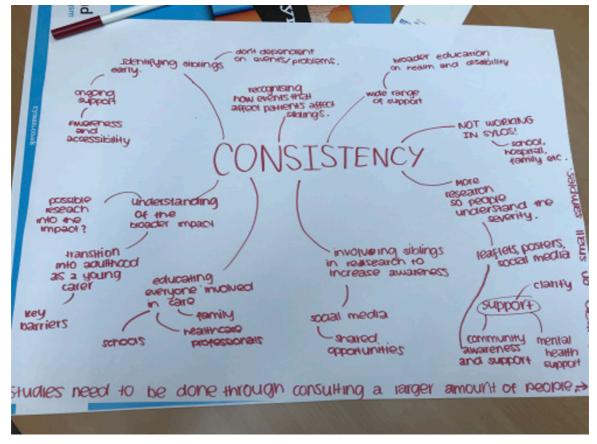


Figure 10.1. Future Sibling Research Suggestion Poster 1

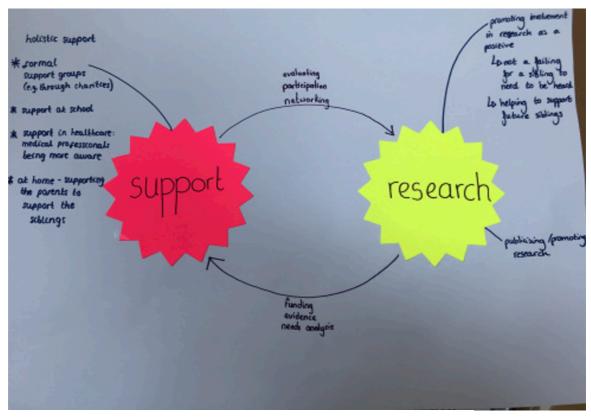


Figure 10.2. Future Sibling Research Suggestion Poster 2

The other internal factors proposed in the model were not clearly drawn out in findings of this thesis. While the systematic review and meta-analysis suggested some demographic factors that may be influential there is limited consensus and robust evidence in this area. The one demographic which was related to outcomes in the mixed methods analysis was the age of the sibling independent of the age of their brother or sister, in that older siblings appear to be report more emotional difficulties. This could work closely with the 'time' aspect included in the Sibling Embedded Systems Framework, and could be an advancement of this work.

The framework also includes factors that directly affect the sibling but that do not occur internally for instance their immediate family and peers. Each of these factors was considered in length previously. Sibling's immediate family relationships and dynamics were highlighted as particularly important though as sibling's experiences appeared to relate to the QoL of their parents. These social connections and relationships are vital for siblings as they appear to be the key source of support and information when needed and although peer support (from other siblings) was generally viewed positively it may not be as simple as this as there were hesitations and a potential for a negative effect.

Many of the factors included in the both the exo-system and the macro-system were either not measured or there were issues with the measurement taken. There were several participants included in the qualitative analysis that discussed the topics, particularly those included in the exosystem. Parents discussed their experiences with work and also support services. Generally, most parents had positive experiences with the support they had received overall, despite a few having some poorer experiences at certain points in time. Further, parents' experiences at work varied somewhat, there were many who felt they could not continue in full-time employment as they would not find an employer that understood. Those that had an employer that was understanding typically viewed themselves as very fortunate. Finally, the remaining factor included in the exo-system which was considered by all participants in the qualitative analysis was social media and the media in general. All participants were sceptical about the interpretation of CF commonly seen in the media and the tendency for it to focus on the negatives. There were sources which were viewed and more reliable that participants encouraged other to use, such as The Cystic Fibrosis Trust website.

Unfortunately, it was not possible to consider the factors included in the macrosystem of the model within this thesis. Factors such as culture, religion, and legislation have been noted to impact on siblings. Further work exploring the appropriateness of other factors and theories from which the Sibling Embedded Systems Framework was drawn, in a range of diagnoses could help to reinforce the framework. Given the breadth of the framework, however, it is challenging to form a single study that includes all factors and as such it may be necessary to conduct multiple trials or consider the use of population level data to test aspects of the model.

Beyond the application of the findings to theories of sibling impact, it is also important to consider the potential clinical implications of the results. Siblings evidently have a limited amount of exposure to clinical teams, however there are a few ways that siblings may be reached and certain points in time when it may be more beneficial to do so. Although siblings themselves are not always in attendance at clinics their parents are. The systematic review and meta-analysis presented in Chapter 2 suggested that even an increase in the salience of the potential impact on siblings could help improve sibling outcomes, and from the mixed methods analysis results we can see that the emotional experiences of siblings are linked to the QoL of their parents. Therefore, supporting parental wellbeing and giving them information about sibling wellbeing could allow them to provide the siblings with the level of support they need.

Furthermore, the suggestion that clinical measures may be insufficiently sensitive has drawn some away from the recommendation of regular screening of siblings (Hartling et al., 2014), although there are also others that believe screening the whole family including the siblings regularly would be a positive step forward (Dia & Harrington, 2006; Rana & Mishra, 2015). What may be most appropriate to further consider given the results of this thesis is screening siblings when their brother or sister is going through a period of ill health or change, for instance at the time of diagnosis, which has been given a great amount of attention for both the individual with the chronic condition (Stein et al., 2019) and their parents (Mérelle et al., 2003; Modi, 2009) but not in siblings, as such further research is needed in this area.

10.4. Thesis Strengths & Limitations

This thesis provides an insight into the experiences of siblings with a brother or sister with a chronic illness or condition within the family context from their own perspective using robust qualitative methodologies. It also adds to the existing literature as it combines qualitative and quantitative evidence in the first use of mixed-methods analysis in this subject area. Strength and limitations for have also been addressed in several of the previously chapters. However, there are several strengths and limitations that should be considered at a whole thesis level.

A key advantage of this work is that it is first of its kind. This work has greatly enhanced the understanding of the impact on siblings through the use of a robust mixed methods analysis of a multi-respondent data set. This has allowed aspects of sibling's lives that may have previously gone unnoticed and unrecognised to be pinpointed and has helped to highlight to clinicians and others involved in the care of those with CF the importance of identifying and understand the needs of siblings.

The importance of utilising techniques to enhance the reliability and credibility of qualitative research has been emphasised (Nowell et al., 2017), and therefore multiple techniques to do so, such as member checking, were applied within the qualitative section of this thesis; this along with

transparency in the approach taken to the analysis (Braun & Clarke, 2006) enhanced the strength of the work. Furthermore, the use of mixed methods in research, as in the analysis presented in Chapter 9, has previously been encouraged, particularly in areas where using quantitative measures alone may not be sufficiently sensitive (Hartling et al., 2014). Using a mixed methods approach can help to support or enhance the findings from qualitative or quantitative analysis only (White et al., 2012), and as such provides further depth to the findings presented in the Chapters 4 to 8. The inclusion of multiple-respondent in both the qualitative research and the mixed methods analysis also enhanced the findings presented in this thesis as it increased the breadth and depth of the findings (Johnson et al., 2007). The use of multiple-respondents was also particularly beneficial to this thesis as issues of parent-proxy measures have been highlighted before (Barlow & Ellard, 2006; Mazaheri et al., 2013; Sleeman et al., 2010) therefore it was important to take self-reported measures from sibling. Family experiences tend to be strongly interrelated (Ma et al., 2017) and to fully understand the experiences of siblings it was important to consider the perspectives of their parents and brother or sister also. Incorporating each of these perspectives into the analysis allowed a well-rounded picture of the sibling experience to be established.

Further, the use of CF as a case study within this thesis developed and progressed on the current understanding of sibling impact by considering a condition with a high treatment burden and mortality rate. Being able to isolate the factor of high morbidity and mortality, which was previously identified as potentially at an increased risk (Vermaes et al., 2012), allowed other potentially influential factors and the experiences of this group to be investigated.

Despite the strengths of this thesis and the analyses included within it there are also several limitations that should be considered. In particular there is a risk to the generalisability of the findings. For instance, the meta-analysis presented in Chapter 2 has a smaller sample size and is subject to a high level of heterogeneity which implies that the results should be considered carefully. While the use of CF as a case study brings some strengths to the work, the results may not necessarily be applicable to other siblings with a brother or sister with a different chronic illness or condition diagnosis, although several of the key findings from this work including the use of avoidance coping and sensitivity around critical periods have been previously been found in siblings of children with schizophrenia (Stålberg, Ekerwald, & Hultman, 2004) and cancer (Kaplan, Kaal, Bradley, & Alderfer, 2013).

The qualitative project was disadvantaged by a lack of specific questions about siblings in the interview schedules for the young person, adult, and parent interviews. It was thus reliant on either the interviewer or the participant to raise the topic. This led to a lack of direct consideration of the sibling experience in several interviews, which was the overall objective of this thesis. Additionally, no

measure of the family composition was included in the demographics taken from families, and therefore information on which families had siblings was missing in several cases.

This work is also restricted in its consideration of certain factors that were drawn out in previous research for instance, culture (Lauderdale-Littin & Blacher, 2017; Long et al., 2013; Smith & Elder, 2010; Vermaes et al., 2012) and the number of siblings in the family (Walton, 2016). All participants included in the qualitative analysis and therefore the mixed methods analysis identified themselves as white, and therefore there was no option to assess differences by ethnicity. The lack of recruitment of full families in all cases limited the numbers and variables available for consideration in the mixed methods analysis in particular.

There were key flaws in chapter 9 that should be re-considered in reference to the overall objective of this thesis. Chapter 9 was an initial tentative exploration into how mixed methods analysis could be used to investigate trends between the experiences of siblings and the QoL of the sibling and their immediate family. The qualitative research presented in chapter 4 attempted to consider the nuanced experiences of siblings with a brother or sister with CF. The themes were formed of sections of text (or references) from interview transcripts. In the analysis presented in chapter 9 it is not possible to consider influences of the interviewer on the transcripts or the nature of the references, e.g. if they were particularly negative or positive. Future advances of this work could include the collection of a greater number of full families, which would allow for more robust methodology including those suggested for quantifying qualitative research in a more systematic manor (Chi, M. T. ,1997).

10.5. Future Directions for Research

From the above discussion of the findings from this thesis there are several recommendations for future research that can be given. The number of sibling studies being completed continues to increase and evidently from the suggestions from SIREN (Figures 10.1 & 10.2) there are a variety of views on how research can be improved.

- The first recommendation that can be drawn from this thesis, and in particular from the systematic review and meta-analysis is that consideration should be given to the methodologies used in siblings research, including encouraging greater consistency in measures use.
- Utilising UK population level data to investigate factors that may change the level of impact siblings
 experience would help to begin to clarify how the complex interactions in families living with a
 chronic condition affect siblings. Before these factors are established, any segregation of sibling
 populations should be well justified using the previously published data.
- It would also be particularly beneficial to consider longitudinal investigations of the impact felt by siblings as this could help to draw out periods of sensitivity or changes over time and age of the sibling.

- Longitudinal research on siblings may help to inform the use of appropriately sensitive measures
 in siblings, as from the findings of this thesis it is anticipated that the level of sensitivity needed to
 measure impact in siblings will fluctuate over time.
- The assumption of future caregiving by siblings was not greatly considered in this thesis and while
 there did not appear to be much of a burden in siblings with a brother or sister with CF this may
 not be consistent across diagnoses and needs to be further investigated.
- Further evaluations of The Sibling Embedded Systems Theory (Kovshoff et al., 2017) are needed,
 which may also be enhanced by the identification of factors using population level data.
- Future sibling research should move away from its current reliance on parental proxy measures
 and use self-reported measures, but it should continue to include the perspective of the sibling's
 immediate family given the interconnectedness of their experiences and outcomes.
- The use of coping skills, particularly avoidant coping, and their implication for outcomes in siblings, particularly in times of stress would be highly beneficial for understanding how to approach and support this group.
- Finally, there is a need to continue to engage siblings in research and in particular engaging siblings
 in the development of a support system that is appropriate, delivered in an acceptable way, and
 has a basis in theory.

10.6. Concluding Remarks

In Chapter 1 several gaps in the literature were identified and the work presented in this thesis aimed to address many of these areas. For instance, Chapter 2 aimed to synthesise the evidence on the effectiveness of support services to help address the problem of a lack of consensus and clarity in how siblings should be supported without applying unfounded arbitrary segregations, which previously lacked. Furthermore, the use of qualitative and mixed-methods analysis attempted to address the many instances of divergent findings found in the quantitative literature. The inclusion of multiple respondents throughout the thesis allowed the work presented to not be subject to a potential source of bias that has been reported previously and help to form a well-rounded image of the sibling experience within the family context. By addressing these gaps in the existing literature and this thesis was able to enhance the current understanding of siblings with a brother or sister with a chronic illness or condition and propose ways in which research and support for siblings should be developed.

The work in this thesis emphasises the need to continue to engage siblings in research but to recognise that they may not always wish to focus on the negative aspects of their brother's or sister's condition or think about it at all. Respecting the sibling as the expert in their experiences and engaging with them directly rather than using parental proxy was a key component of this work. Sibling research needs to develop and encourage the use of consistent and reliable methodologies. Support given to

siblings needs to be reconsidered and potentially targeted at specific times of stress. In conclusion, the sibling is an often under recognised member of the family when a child has a chronic illness or condition, yet their outcomes and experiences are highly related to those of the rest of their immediate family and it may be the use of an avoidance coping technique has resulted in previous research being unable to consistently identify a cause for concern. It may also be a lack of consideration of other factors, such as family dynamics during a period of change of stress, which has limited previous research and support. Supporting siblings as a member of the family is important but further research is needed to ensure that this is done in the most clinically and cost-effective way.

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Appendices

Appendix 2.1. Published Paper of Findings Presented in Chapter 2

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Impact of Well-being Interventions for Siblings of Children and Young People with a Chronic Physical or Mental Health Condition: A Systematic Review and Meta-Analysis

Mhairi McKenzie Smith¹ Snehal Pinto Pereira Lynette Chan¹ Charlotte Rose¹ Roz Shafran¹

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Abstract

Siblings of children and young people with a chronic illness are at increased risk of poor psychological functioning. A number of studies have attempted to implement and evaluate interventions targeting the psychological well-being of this at-risk group. This systematic review summarises the evidence regarding psychological functioning of siblings following an intervention targeting their well-being. The meta-analysis considered behaviour and knowledge, two of the most frequently studied outcomes. The following databases were used: PsycINFO, EMBASE, CINAHL, PubMed, Scopus and Web of Science. Seventeen studies were eligible to be included in the systematic review and eight in the meta-analysis. Results from the systematic review reflected the inconsistency of intervention evaluations in this area with a high level of heterogeneity and a total of 23 outcomes considered across the 17 included studies. The meta-analysis estimated effect sizes using a standardised mean difference (SMD) approach. Pre-post analysis suggested significant improvement in behavioural outcomes and knowledge of their sibling's health conditions with a SMD of -0.44 [95% CI -0.6, -0.29); p = 0.000] and 0.69 [(95% CI =0.42, 0.96); p = 0.000], respectively. The SMD was not significant for behavioural outcomes when considering treatment–control studies. In conclusion, the findings suggest interventions for well-being have a positive effect on the psychological functioning of siblings of children and young people with a chronic illness, but their specificity needs to be established. There is a need for further, more methodologically robust research in this area.

Keywords Siblings · Chronic · Psychological · Intervention

Background

It is estimated that anywhere between 13 and 32% of children and young people (0–19 years) suffer from a chronic or life-limiting condition (Fraser et al. 2012; Van Cleave et al. 2010; Wijlaars et al. 2016). A chronic childhood illness can be defined as one that occurs between the ages of 0 and 18 years, is medically diagnosed and reproducible using valid methods or instruments, has been present for longer than 3 months or has occurred three or more times in the

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- Mhairi McKenzie Smith mhairi.mckenzie.16@ucl.ac.uk
- UCL Great Ormond Street Institute of Child Health, 30 Guilford Street, London WC1N 1EH, UK



past year and is likely to reoccur, and is not (yet) curable or is highly resistant to treatment (including mental health conditions) (Mokkink et al. 2008). This definition encompasses both physical and mental health disorders. The World Health Organisation defines a mental health disorder as "generally characterised by a combination of abnormal thoughts, perceptions, emotions, behaviour and relationships with others". Mental Health disorders include depression, bipolar affective disorder, schizophrenia, dementia, intellectual disabilities and developmental disorders including autism (WHO 2017). Hence, for the purposes of this review, developmental disorders including autism are included within the category of mental health disorders. It is estimated that around 54.6%

OECD-32 average proportion of households with two or more children is reported as 54.6%, which is multiplied by our estimated prevalence estimates (13–32%) to achieve our estimate of children that have a sibling with a chronic health condition. "Children" in this instance are generally defined here as dependent resident children under 25 and include both biological children and step- or adopted

of families have two or more children (OECD 2016), which means that approximately 7–17% of children have a sibling with a chronic illness.

Consequences

Previous studies that have investigated the impact of having a sibling with a chronic illness have produced inconsistent results. Some literature suggests an elevation in psychological distress and mental health disorders in siblings (Cadman et al. 1988), and other studies suggest such siblings do not warrant further investigation (Bischoff and Tingstrom 1991). Much of the research has adopted a disease-/disorderspecific approach; some have taken a "non-categorical" or "broad" (Stein et al. 1993; Stein and Jessop 1982) approach, in which they do not differentiate based on the chronic illness (Cadman et al. 1988). Having a sibling with a chronic illness in the family can result in an imbalance of resources, such as time spent with their parents. It can also pose social challenges (Bluebond-Langner 1996) and has been noted to have a negative impact on educational attainment (Breining 2014). Family dynamics can be stressed and can regularly result in tension between parents, along with a lack of socialising outside of the family (Kvist et al. 2013; Mailick Seltzer et al. 2001).

A meta-analysis of 51 studies looking at the psychological impact of having a sibling with a chronic illness found a significant overall negative impact and specifically a significant negative impact on psychological functioning, peer activities and cognitive development (Sharpe and Rossiter 2002). Sharpe and Rossiter's (2002) meta-analysis only included studies which considered the siblings of children with a chronic physical health condition. Their findings are consistent with the findings in a more recent meta-analysis, which included 13 additional studies (Vermaes et al. 2012) and found a significant, although small, negative effect on psychological functioning. Prior to their 2002 meta-analysis Rossiter and Sharpe also published a meta-analysis which considered siblings of children with mental retardation (Rossiter and Sharpe 2001). Their findings indicated that these siblings also have a significantly lower psychological

Psychological function is defined as "an individual's ability to achieve their goals, both within themselves and in the external environment. This includes their emotions, behaviour (both internalising and externalising behaviours), social skills and their overall mental health" (Preedy and Watson 2010). Behaviour is typically measured using tools such as

Footnote 1 (continued)

children or any other children in the household, though exact definitions do vary across countries.

the Childhood Behavioural Checklist (CBCL) (Achenbach 1991) or the Strengths and Difficulties Questionnaire (SDQ) (Goodman 1997). These self-reported tools measure internalising behaviours, e.g. emotional symptoms, and externalising behaviours, e.g. conduct problems, along with a total score of behavioural problems. Both the SDQ and the CBCL have been shown to be able to distinguish between psychiatric and non-psychiatric cases (Goodman 1997; Seligman et al. 2004). It has been noted that there is the potential for a greater impact on internalising behaviours in siblings. An increase in internalising behaviours, including anxiety and depression, have been observed both in studies that adopt a broad approach (e.g. Cadman et al. 1988) and those using a more disease-specific approach (Cadman et al. 1988; Fisman et al. 1996; Hastings 2003; Verté et al. 2003), and this is supported by previous meta-analyses (Rossiter and Sharpe 2001; Sharpe and Rossiter 2002; Vermaes et al. 2012). While evaluating a camp intervention for siblings of children and young people with a chronic illness, Sidhu et al. (2006) found that one-quarter of their sample suffered from psychological distress within the clinical range, and these were more internalising in nature. Suggestions as to why this may be include children not wishing to burden parents further (Sidhu et al. 2006), the quality of the family environment (Verté et al. 2003) and factors relating to the child themselves, e.g. age, sex (Hastings 2003).

Several studies have proposed a link between psychological functioning, e.g. anxiety, and a lack of understanding of a sibling's chronic condition (Carpenter et al. 1990; Houtzager et al. 2001; Sidhu et al. 2006). It has been suggested that a limited understanding of their sibling's illness can lead to poor adaption (Evans et al. 2001). It may also be that the lack of knowledge about their sibling's illness negatively impacts the sibling relationship (Roeyers and Mycke 1995). Improving the child or young person's understanding of the sibling's condition has been linked to reduced anxiety levels (Houtzager et al. 2001). Strategies that adopt an educational approach therefore may help improve the mental health of siblings of children and young people with a chronic illness. In this way, knowledge of a sibling's condition may be considered part of their psychological functioning.

Along with the potential negative psychological impacts noted, the literature exists that suggests positive effects of having a sibling with a long-term condition. For instance, within Rossiter and Sharpe's original meta-analysis, sibling relationship was found to positively moderate the level of psychological distress in siblings of children with mental retardation (Rossiter and Sharpe 2001); however, in their subsequent meta-analysis this relationship was insignificant (Sharpe and Rossiter 2002). Other suggested positive impacts include an increase in maturity (Grossman 1972), warmth and understanding towards their sibling (Fisman et al. 1996), and prosocial behaviour (Ferrari 1984; Lobato



et al. 1988). There has been little investigation into these positive findings, but it has been suggested that they may act as protective factors for mental health outcomes in children and young people (Fisman et al. 1996). Parents may be unaware of the positive interactions occurring between the siblings, perhaps due to the salience of negative interactions (Rivers and Stoneman 2003), and such lack of awareness could potentially influence parental reports which are often used in evaluations.

Predictive Factors

Identifying siblings of children and young people with a chronic illness that are at greatest risk of poor psychological functioning could help to target interventions. Targeting interventions at those who are in greatest need is imperative to resource-limited services. Family-related factors have been suggested that may help identify those at risk. For instance, Daniels et al. (1987) found that less family cohesion and expressiveness were related to an increased psychological risk in siblings. Positive family functioning has also been noted as a potential protective factor in children with siblings with Down's syndrome, yet not in those with pervasive developmental disorder (Fisman et al. 1996). It has also been found that siblings of children receiving treatment for mental health problems were more likely to live in poorly functioning families (Barnett and Hunter 2012). When considering potential predictive factors in their meta-analysis, Vermaes et al. (2012) reported that gender, birth order or diagnosis was not significantly associated with behavioural problems. They did, however, find that when the child has a chronic condition that is associated with a higher mortality rate and more intrusive treatment, the siblings were significantly more likely to have greater internalising and externalising problems, along with less positive self-attributes.

Well-being Interventions

The definition of well-being is continually developing. It is suggested that high well-being is positively related to good mental health and can be made up of the following ten components: competence, emotional stability, engagement, meaning, optimism, positive emotion, positive relationships, resilience, self-esteem, and vitality (Huppert and So 2013). Well-being interventions have been suggested to help improve psychological outcomes (including anxiety, depression, stress, self-esteem and coping) of siblings of children and young people with a chronic illness. These interventions have taken various forms, including group interventions (Heiney et al. 1990; Houtzager et al. 2001; Lobato and Tlaker 1985; Smith and Perry 2005), sibling training (Ferraioli et al. 2012), camps (Kiernan et al. 2004; Sidhu et al. 2006) and family-based support (Besier et al. 2010);

Giallo and Gavidia-Payne 2008). A range of populations have been targeted; some have been disease-specific (Dolgin et al. 1997), while others have taken a broad approach (Cadman et al. 1988). The content and duration of the interventions are highly varied. Camp interventions are typically formulated from the concept of therapeutic recreation (Fine and Fine 1996), which focuses on enjoyment and freedom in recreation, while other studies, particularly those involving a group interventions, have utilised more psychoeducational components (Giallo and Gavidia-Payne 2008; Granat et al. 2012; Lobato and Kao 2002).

Evaluating Interventions

Evaluations of interventions are limited and typically associated with methodological issues including small sample sizes (Marszalek et al. 2011), a lack of intervention integrity tracking (Kryzak et al. 2015), and large heterogeneity (Ali et al. 2014). A previous systematic review that considered interventions for siblings of children with a chronic illness or disability included articles published between 1985 and 2008, adopted a broad approach and included 14 papers (Hartling et al. 2014), although the definition of chronic illness or disability in this review was unclear. Hartling et al.'s review found a large inconsistency in treatment effects on behavioural and emotional outcomes and highlighted the importance of the sensitivity of the measures used as several of the included studies reported the child to be within the "normal" range of mental health prior to the intervention. It is suggested that this may cause a ceiling effect on results as their scores are unlikely to continue to improve beyond their current point. A more recent review by Tudor and Lerner (2015) included 16 papers looking at interventions for psychological functioning targeted specifically at siblings of children with developmental disabilities (DD). Tudor and Lerner initially argued that the experience of typically developing siblings of children with DD was distinguishable from siblings of children or young people with physical disabilities, yet within their conclusion they acknowledged that the best services for siblings may not make that distinction. Therefore, due to a lack of clarity in the advantages of interventions for siblings of individuals with either a physical or psychological disorder, this systematic review and meta-analysis includes both populations.

In summary, previous systematic reviews have suggested that there is a need for interventions to improve psychological well-being in siblings of children and young people with a chronic illness, but limited evidence has been provided about the effectiveness of interventions that are offered to siblings of children with either a physical or mental health condition. When considering this subject it is important to remember that children with a physical health condition have an increased likelihood of having a mental health condition



(Lavigne and Faier-Routman 1992) and that there is a close relationship between physical and mental health. There have been calls for physical and mental health to be more closely integrated (Prince et al. 2007; Scott et al. 2016). In general, there is an absence of evidence on the effectiveness of interventions for siblings and a lack of clarity regarding the requirement for interventions to distinguish between siblings of children with a physical or mental health condition. There is value in investigating the effectiveness of the interventions regarding the psychological well-being of siblings of children and young people with either a chronic physical or mental illness (or both). Additionally, no meta-analysis has been conducted on the effectiveness of psychological interventions on siblings of children and young people with chronic illness. The aims of this review are to:

- Conduct a systematic review to synthesise the literature that evaluates well-being interventions offered to siblings of children and young people with a chronic illness.
- Conduct a meta-analysis to quantitatively evaluate the impact of the evaluations included in the systematic review.

Methods

Sources and Search Strategy

A systematic search was conducted, following the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines (Moher et al. 2009) and Cochrane recommendations (Higgins and Green 2011). Electronic database searches were completed along with reference list and citation hand searches, and grey literature searches. The following databases were used: PsycINFO, EMBASE, CINAHL, PubMed, Scopus and Web of Science, and PsycExtra was used to search for grey literature. The search strategy was piloted in November 2016, and following review was re-run in January of 2017, by three independent researchers (MMS, CR and LC), to include all literature up to the end of 2016. The search strategy was built using the Participant, Intervention, Comparator, Outcome (PICO) framework, as suggested in PRISMA guidelines (Shamseer et al. 2015). The broad themes included in the search strategy were sibling, chronic condition, intervention and mental health. The search strategy was adapted to each database. The full search strategy can be found in Online Resource 1.

Study Selection

Studies were included if they evaluated an intervention offered to siblings of children and young people with a chronic health condition, as defined previously. It was required that the two children live together (or were of an age where it is assumed they would still live together, i.e. below 18 years of age). The sibling must be considered "healthy" themselves and not a donor for the ill child or young person.

The intervention could take any form, provided it aimed to improve the psychological well-being of the sibling, and reported an outcome that is related to the mental health of the sibling including direct psychological outcomes, e.g. anxiety, depression and stress, as well as related factors, e.g. knowledge, social support, self-esteem, relationships, coping and adjustment. Family-level interventions were not included unless there were sufficient (at least one) sibling-specific outcomes, as described above, reported.

Included studies could be mixed methods if they report the result of at least one quantitative measure. Any form of trial was accepted if it evaluated the effectiveness of the intervention; this included pre-post design trials.

Studies were excluded if unavailable in English or French. Studies that involved bereaved siblings and studies that looked specifically at sibling donors were also excluded.

Risk of Bias Assessment

The Effective Public Health Practice Project Quality Assessment Tool (EPHPP) (Thomas et al. 2004) was used to evaluate the quality of all papers included in this review. This tool was chosen as it has been shown to have a higher inter-rater reliability relative to the Chronic Collaboration Risk of Bias Tool (Armijo-Olivo et al. 2012) and is appropriate for use across different study designs compared to other tools such as the ROBINS-I, which is only appropriate for non-randomised trials (Sterne et al. 2016). This allowed confidence in the consistency and reliability of the assessments.

The EPHPP evaluates studies on eight components: selection bias, study design, confounders, blinding, data collection methods, withdrawals and dropouts, intervention integrity and analyses. The ratings for all but intervention integrity and analyses are combined to give the study an overall rating of strong, moderate or weak.

Searches, study selection, quality assessment and data extraction were completed by three independent researchers: MMS, CR and LC. Any discrepancies were dealt with through discussion, and if a consensus could not be reached, the opinion of an additional independent researcher was sought.

Data Extraction and Analysis

A data extraction form was created, using the Effective Practice and Organisation of Care (EPOC) data collection form (EPOC 2013) as a base, to ensure sufficient data were



collected from each study included in the systematic review and meta-analysis.

It was expected that there would be large heterogeneity in outcome measure used across studies; therefore, a random-effects meta-analysis was conducted. A standardised mean difference (SMD) and restricted maximum likelihood (REML) technique was used to estimate effect sizes and weights in STATA 14 (StataCorp 2015). The SMD was estimated using Hedge's g technique, allowing for a smaller sample size relative to Cohen's d method, which is typically used in meta-analyses in this subject area (Cuijpers 2016).

The SMD technique allows the combination of different scales that are measuring the same outcome. For instance, the Strengths and Difficulties Questionnaire (SDQ) (Goodman 1997) and the Child Behaviour Checklist (CBCL) (Achenbach 1991) both measure behavioural outcomes and have been noted to have highly correlated scores (Goodman and Scott 1999).

This review was registered on PROSPERO (International Prospective Register of Systematic Reviews), registration number CRD42017056740.

Results

Search Results

A total of 1536 papers were identified from the initial searches. After removal of duplicates 980 records were screened. Of that, 913 were excluded based on title and abstract (n = 904), format (n = 7) and language (n = 2). Sixty-seven full-text articles were assessed for eligibility, and 17 were included in the qualitative synthesis. Eight studies were included in the meta-analysis. Five of the papers included reported an outcome measure of behaviour, and five reported on knowledge change following intervention. The nine papers not included in the meta-analysis either did not report sufficient data, used a different study design, or did not use either a behaviour or knowledge outcome measure. The flow of papers through the process of eligibility can be seen in Fig. 1. In the initial search, ten reviews were identified including three systematic reviews (Hartling et al. 2014; Prchal and Landolt 2009; Tudor and Lerner 2015). Rather than including the reviews, as there were discrepancies with the inclusion criteria, it was decided that the individual papers from each should be reviewed against the eligibility criteria.

Baseline Characteristics

Across the 17 included studies, there were 1264 participants. Age of participants ranged from 6 to 15 years, with an average of 10.47 years.² There was a relatively even gender balance in the overall sample, with 53% of participants being female. Further demographic information can be found in Table 1.

Quality Assessment

Of the included studies, eight (47%) were considered of weak quality (Besier et al. 2010; D'Arcy et al. 2005; Evans et al. 2001; Giallo and Gavidia-Payne 2008; Heiney et al. 1990; Houtzager et al. 2001; Kiernan et al. 2004; McLinden et al. 1991), seven (41%) were rated as moderate (Cebula 2012; Dolgin et al. 1997; Granat et al. 2012; Kryzak et al. 2015; Lobato and Kao 2002; Phillips 1999; Williams et al. 2003) and only two (12%) were rated as strong (Sidhu et al. 2006; Smith and Perry 2005). The two strong studies were both rated strong in the confounders, data collection methods, and withdrawals and dropouts components of the EPHPP, and moderate in the remaining three components. The one RCT study included in the review was rated weak overall (Giallo and Gavidia-Payne 2008). One paper was scored N/A for withdrawals and dropouts as it had one time point only, and therefore, quality assessment in this area was irrelevant for this paper (Cebula 2012). A table of quality assessment results can be found in Online Resource 2.

Interventions

Nine of the 17 studies included in this review were group-based interventions (53%); the next most frequent form of intervention was camp-based interventions (18%). The studies were conducted in mainly high-income, predominately Caucasian countries including Germany, UK, The Republic of Ireland, Australia, Canada, the Netherlands and Sweden. The largest number of studies coming from one location was four, which were all based in the USA. The duration of the interventions ran from 4 days (Sidhu et al. 2006) to 96 months (Cebula 2012), with a median and mode duration of 6 days. There was little consensus in the approach taken in the interventions, even between studies that used similar designs. Further details about the included interventions can be found in Table 2.

Of the 17 papers included, six focused on physical illnesses, four focused on mental health conditions, and the remaining seven focused on a combination of physical and mental health conditions. Several studies focused on specific

² From the twelve studies which reported average age.

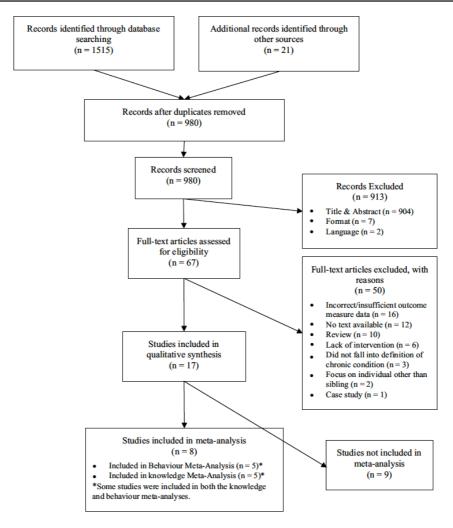


Fig. 1 PRISMA flow diagram

conditions; for instance, four of the 17 studies offered interventions to siblings of children with cancer. Of the six papers that focused on physical illnesses, much of their samples were made up of siblings of children with cancer (minimum 47.9%). A breakdown of the type of intervention offered by physical, mental or combined studies is shown in Fig. 2.

Systematic Review

Physical Conditions

Of the six papers that focused entirely on chronic physical illnesses, four of them focused exclusively on cancer diagnoses, while the remaining two incorporated congenital heart disease, cystic fibrosis and other haematological-related illnesses. Across these six studies, there were nine different outcomes considered and 13 different measures used.

There was significant improvement in self-esteem (Kiernan et al. 2004; Sidhu et al. 2006), behaviour (Besier et al. 2010), knowledge, attitude and feeling, mood (Dolgin et al. 1997), and anxiety (Houtzager et al. 2001; Sidhu et al. 2006). Three studies examined quality of life (QoL), and all found significant improvements (Besier et al. 2010; Kiernan et al. 2004; Sidhu et al. 2006). There was no significant change in "coping" (Heiney et al. 1990) and affect (Kiernan et al. 2004).



Learning disabilities and associated challenging behaviour Additional diagnoses: ABA (16%); ABA control (13%); post-ABA (15%); post-ABA control (15%) Other (Asperger's, ASD, HFA); ABA (20%); ABA Control (27%); post-ABA (12%); post-ABA control (15%) Autism: ABA (80%); ABA control (73%); post-ABA (88%); post-ABA control (85%) Disability (a physical or intellectual; disability, or a combination of both) Haematological-related illness (21.7%) Acute lymphoblastic leukaemia (65%) Congenital heart disease (31.7%) Acute myeloid leukaemia (7.6%) Mild mental retardation Moderate mental retardation Osteogenic sarcoma (3.8%) Neuroblastomas (7.6%) Hepatoblastoma (3.8%) Cystic fibrosis (20.5%) Brain tumours (7.6%) Not reported (26.1%) Ependymoma (3.8%) Siblings condition Cancer (100%) Cancer (100%) Cancer (52.2%) Cancer (100%) Cancer (49%) ASD (100%) Post-ABA control: 46% Gender (% Female) ABA control: 60% Post-ABA: 42% ABA: 53% 57.1% 45.6% 63% 54% 52% 40% Post-ABA control: 9.7 years (3.1) Age, ye ars me an (SD) or range ABA Control: 9.3 years (3.3) Post-ABA: 9.9 years (2.3) 26 (post-ABA and post- ABA: 9.1 years (2.4) ABA control) 11.3 years (3.1) 11.5 years (2.4) 10.6 years (2.1) 8.6 ye ars (3.3) 11.7 years (3) 9-15 years 6-14 years 9-12 years 6-12 years 8-13 years Other n (specified) N/A N/A N/A N/A N/A N/A N/A Control n N/A N/A N/A N/A N/A N/A N/A N/A N/A 8 45 Year Totaln Experimentaln 23 7 24 119 259 26 15 16 28 26 45 259 23 14 24 119 180 132 56 15 56 16 28 Table 1 Summary of demographics 2015 2012 2010 1997 1990 2001 2004 1999 2005 2006 2001 2005 Physical health conditions Mental health conditions Smith and Perry Houtzager et al. Kiernan et al. Kryzak et al. Dolgin et al. Heiney et al. D'Arcy et al. Besieret al. Evans et al. Sidhu et al. Combined Author(s) Phillips Cebula



Author(s) Year Total in Experimental in Control in Other in (specified) Age (fiallo and Gavidia-Payne) 2008 21 12 9 N/A Intition Granat et al. 2012 54 54 N/A N/A N/A 8- Lobato and Kao 2002 54 54 N/A N/A N/A 8- McLinden et al. 1991 11 6 5 N/A 98	Other n (specified) N/A			
2012 54 54 N/A N/A N/A 2002 54 54 N/A N/A N/A N/A 1991 111 6 5 N/A		Age, years mean (SD) or range	Gender (% Female)	Siblings condition
2012 54 54 N/A N/A N/A 2002 54 54 N/A N/A N/A N/A 1991 11 6 5 N/A		Intervention: 11.75 years (2.9)	57.1%	Down syndrome (19%)
2012 54 54 N/A N/A N/A 2002 54 54 N/A N/A N/A N/A 1991 11 6 5 N/A				Autism (23.8%)
2012 54 54 NVA NVA 2002 54 54 NVA NVA 1991 11 6 5 NVA				ADHD (4.8%)
2012 S4 S4 N/A N/A N/A 2002 S4 S4 N/A				Polymicrogyria (4.8%)
2012 54 54 NVA NVA 2002 54 54 NVA NVA 1991 11 6 5 NVA				Multiple disabilities (14.3%)
2012 54 54 NVA NVA 2002 54 54 NVA NVA 1991 11 6 5 NVA		Control: 11.00 years (2.3)		Cystic fibrosis (4.8%)
2012 54 54 N/A N/A N/A 2002 54 54 N/A N/A N/A N/A 1991 11 6 5 N/A				Congenital heart disorder (9.5%)
2012 54 54 NVA NVA 2002 54 54 NVA NVA 1991 11 6 5 NVA				Multiple illnesses (4.8%)
2012 54 54 N/A N/A 2002 54 54 N/A N/A N/A 1991 11 6 5 N/A				Williams syndrome (9.5%)
2002 54 54 N/A N/A N/A 1991 11 6 5 N/A		8-12 years	%19	ADHD (16.7%)
2002 54 54 N/A N/A N/A 1991 11 6 5 N/A				Asperger syndrome (13%)
2002 54 54 N/A N/A N/A 1991 11 6 5 N/A				Physical disability (14.8%)
2002 54 54 N/A N/A N/A 1991 11 6 5 N/A				Intellectual disability (31.5%)
2002 54 54 N/A N/A 1/A 1/B 1991 11 6 5 N/A				Autism (24.1%)
1991 11 6 5 N/A		8-13 years	26%	Physical disabilities (26%)
1991 11 6 5 N/A				ASD (23%)
1991 11 6 S N/A				Mental retardation (21%)
1991 11 6 5 N/A				Medical disorders (17%)
1991 11 6 5 N/A				Combined psychiatric and learning disorders (13%)
		9.8 years (2.2)	64%	Mentally retarded (45%)
				Physically handicapped (9%)
				Multiply handicapped (45%)
Williams 2003 252 79 102 Partial treatment: 71 Int		Intervention: 11.1 years (2.2)	20%	Cystic fibrosis (4.4%)
				Diabetes (34.9%)
Pa		Partial: 11 years (2.5)		Spina B ifida (9.5%)
				Cancer (8.7%)
တိ		Control: 11.2 years (2.5)		Developmental disabilities (42.5%)



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Author(s)	Year	Name of Intervention	Who delive red intervention	How often/how many sessions were involved	Sessions	Protocol available	Protocol adherence recorded	Intervention offered to other members of family
Physical health conditions								
Besier et al.	2010	Family-oriented rehabilitation programme	Psychosocial team	4-week programme. Session offered 1–3 times per week	Psychoeducational group, exercise, relaxation, sup- portive/psychotherapy, parent-child sessions	Individually arranged treatment protocols	N/A	III child admitted for rehabilitation. Parents also treated according to individually arranged protocols
Dolgin et al.	1997	Structured group intervention	Clinical social worker, a child life special- ist and a supervising psychologist	Six group sessions were held on consecutive weeks	In addition to group discussions concerning their experience of the illness and its impact, subjects took part in arts and crafts and other creative activities in order to encourage interaction among participants and to promote non-verbal expression of relevant fedings and themes	Detailed structure available	Unclear	^Q
Heiney et al.	1990	Sibling support group	Co-therapists: a fellow in child psychiatry, and a pediatric oncology nurse	Seven 1-h sessions	The group was organised so that each session focused on a specific topic: introduction and one ntation, diagnosis, treatment, school, coping, family relationships, and the future	°Q	°N	Concurrent parent group
Houzageret al.	2001	Support group for siblings Led by two well-trained psychologists	Led by two well-trained psychologists	Five weekly sessions	First session: getting to know each other, second session: changes, third session: emotions related to illness, fourth session: paediatric oncologist invited to talk, final session: siblings visit the oncology ward	°Z	S _o	N _O
Kiernan et al.	2004	The Barretstown gang camp	Unclear/camp Staff	10-day sessions	Core activities: music, theatre, photography, arts and cards, wordsmith, woodwork, canoeing, fishing, horse-riding, adventure, archery and camping. Periphery Activities: hangout, and evening activities. Social Activities: Cottage char, rest hour and the opportunity to meet other from different	No (more info: https://www.barretstown.norg/)	[©]	Camp for children with life threatening illnesses and their siblings



Intervention offered to other members of family Support for Child with ASD Intensive home-based intervention for child with autism. å ŝ ŝ ° å Protocol adherence recorded Unclear Š ŝ å å Š ŝ No—based on the model developed by Meyer and Vadasy (1994) Manual (soon to be published at point of paper publication) Protocol available å Š Š Š ŝ associated with living with a brother or sister who has a Had a problem-solving focus. The majority of activities were of an educational and Group discussions, recreation and homework assistance encourage the expression of Spectrum Disorders (ASD) Exercises, games, and activities that were fun and appropriate information on gies to enhance adjustment informative nature. Leisure sions on autism and related feelings; and impart strate-Focused on developing a network of peers who face energy activities, inter-spersed with discussion about disability and each sibling's experiences competencies; provide age to the family stressors in a promoted group cohesion, providing information sessimilar family challenges, cancer, treatment and its impact on all the family; activities were also used leve lopmental disability to feelings and attitudes to develop peer support facilitate activities, that disorders, and facilitatprovide an opportunity learning about Autism ing discussion relating Consists of high and low and coping strategies The program aimed to networks and social safe environment Sessions N/A How often/how many sessions were involved evenings. A final day at a local theme park Weekly for 8 consecutive 2-96 months/5-40 h per week Three consecutive full days, and then on a weekly basis for six 15-week, after-school (3-5:30 pm) every Once a month for four consecutive months Seven 2 h sessions weekday weeks 4-day Facing the challenge' multi-disciplinary team. Six team leaders (commu-nity centre staff), and Treatment, Research, and Education for Autism and Developmental Disorders (TRE-ADD) staff Mother/partner, outside agency, or parents and outside agency Group facilitators (under-Special education teacher, took pre-camp training workshop) Who delivered intervenschool councillor, volunteers, a psychology doctoral student Comprised of nurses, Unclear. Clinical Psychologist conducted a psychologist and outreach workers seven volunteers interviews tion Applied Behavior Analy-Sibling support groups Sibling support groups The support and skills Name of Intervention After-school program program (SSP) Camp onwards sis (ABA) SibShops 2015 2012 2006 2001 1999 2005 2005 Year Mental health conditions Table 2 (continued) Smith and Perry Kryzak et al. D'Arcy et al. Evans et al. Sidhu et al. Combined Author(s) Phillips Cebula



lable 2 (continued)								
Author(s)	Year	Year Name of Intervention	Who delivered intervention	How often/how many ses- Sessions sions were involved	Sessions	Protocol available	Protocol adherence recorded	Intervention offered to other members of family
Giallo and Gavidia-Payne 2008	2008	Sibstars	A clinician with post- graduate psychology training	Weekly telephone support offered to sibling and parent for 6 weeks	After the first face-to-face session, each week families were required to read an information booklet and complete the practice activities provided	Yes	Programme adherence checklist was used	Involved Parents
Granat et al.	2012	Sibling group intervention Clinical staff from an outpatient rehabilitic centre	Clinical staff from an outpatient rehabilitation centre	A 2-h session every week for 6 weeks	Content intended to increase knowledge and problemsolving skills	A manual (in Swedish) for clinical practice was compiled	No	Separate education groups being provided for parents
Lobato and Kao	2002	Sib link	Doctoral kvel trainees in psychology or psychiatry	Six 90-min æssions	Activities alternated between explicitly focused "main events" and other more social-recreational activities	Manuals available on request	No	Parent group
McLinden et al.	1991	Sibling support group	School psychologists	Six weeks, 1-h per week	Focused on developing participants' acceptance of both regative and positive feelings about their siblings. Information was provided and numerous activities were utilized.	No—based on Lobato (1985)	Ŷ	Ŷ
Williams	2003	Intervention for Siblings: EXPERUENCE Enhance ment (ISEE)	Pediatric nurse clinicians	5-days	Structured teaching about the brother sister's illness, psychosocial session, a 5-day residential summer camp, and wo booster sibling session and pare nt sessions	Brief Protocol Available	^Q	Parent sessions



Mental Health Conditions

Four of the 17 papers focused solely on children with a sibling with a chronic mental health condition. Two focused on autism spectrum disorders (Kryzak et al. 2015; Smith and Perry 2005), one on learning disabilities (Evans et al. 2001) and one on mental retardation (Phillips 1999). Across these four papers, ten outcomes were considered, and 12 different measures used.

There were significant improvements in self-esteem (Evans et al. 2001), sibling involvement (Evans et al. 2001), social support (Phillips 1999), anxiety and depression (Kryzak et al. 2015; Phillips 1999). There was no significant improvement in sibling interaction (Kryzak et al. 2015),³ sibling relationship, family functioning (Phillips 1999) or coping and adjustment (Smith and Perry 2005). Mixed results were noted from knowledge tests: Kryzak et al. (2015) found no significant improvement in Autism Sibling Knowledge, while a significant improvement in Autism knowledge was noted by Smith and Perry (2005). Evans et al. (2001) reported an improvement in knowledge about their siblings learning disorder but provided no statistical evidence.

Both Physical and Mental Health Conditions

Seven of the 17 papers did not specifically look at either physical or mental chronic illnesses. The study by Cebula (2012) evaluated the effectiveness of an intervention for siblings with autism, but noted that between 13 and 16% of these siblings had additional diagnoses of physical health conditions; therefore, their study is included in this section, rather than in the mental health focused section. Prevalence studies suggest that children with a chronic physical illness are more likely to have emotional/behavioural problems and psychiatric diagnoses (Hysing et al. 2007). The same is true of young people with autism where co morbidity is regularly found, including psychological difficulties and physical conditions (Matson and Goldin 2013). None of the included studies that examined only a mental or physical chronic health condition reported comorbidity, and they were therefore unable to consider how comorbidities may influence siblings psychological functioning.

Across these seven studies there were 15 outcomes considered, and 26 different measures used. Positive significant findings were found in the three studies that used a measure of "intervention impact" (Cebula 2012; McLinden et al. 1991; Williams et al. 2003), in the two that evaluated coping and adjustment (Giallo and Gavidia-Payne 2008; Lobato and Kao 2002), and in the studies that looked at stress, family

functioning (Giallo and Gavidia-Payne 2008), mood (Williams et al. 2003) and connectedness (Lobato and Kao 2002). Non-significant findings were found in the two studies that considered parent-related variables (Cebula 2012; Giallo and Gavidia-Payne 2008). Of the seven papers, five considered behaviour (Cebula 2012; Giallo and Gavidia-Payne 2008; Lobato and Kao 2002; McLinden et al. 1991; Williams et al. 2003), four evaluated the impact on selfesteem (Cebula 2012; D'Arcy et al. 2005; McLinden et al. 1991; Williams et al. 2003) and knowledge (Granat et al. 2012; Lobato and Kao 2002; McLinden et al. 1991; Williams et al. 2003), while two looked at attitude and feelings (McLinden et al. 1991; Williams et al. 2003) and sibling relationship (Cebula 2012; McLinden et al. 1991), and one considered social support (Cebula 2012), all of which produced mixed evidence.

Comparison

Coping and adjustment, knowledge and self-esteem were the only outcomes considered in all three categories (mental health, physical health and combined). Both Giallo and Gavidia-Payne (2008) and Lobato and Kao (2002) looked at a combination of health conditions and found a significant improvement in coping and adjustment, whereas Smith and Perry (2005) and Heiney et al. (1990), who considered mental health and physical health, respectively, found no significant improvement. Both of the studies that found significant improvements in coping and adjustment involved the parents of the sibling, which may indicate that, although both studies were combined studies, this finding may be explained by factors other than the consideration of combined physical and mental health conditions. The results for knowledge were spread across the types of study, and there appeared to be no clear divide across physical, mental health or combined studies. Self-esteem was considered in nine papers, of which six found significant improvements following intervention (Evans et al. 2001; Kiernan et al. 2004; Phillips 1999; Sidhu et al. 2006; Smith and Perry 2005; Williams et al. 2003), and three did not (Cebula 2012; D'Arcy et al. 2005; McLinden et al. 1991). The three that found no significant associations were all combined studies, and only one of the six significant results was a combined study (Williams et al. 2003). It is unclear whether this is due to study design. In the six papers that found a significant association, three were camps (Kiernan et al. 2004; Sidhu et al. 2006; Williams et al. 2003), and three were group support (Evans et al. 2001; Phillips 1999; Smith and Perry 2005). Two of the papers that did not find significant associations were also a group support evaluation (D'Arcy et al. 2005; McLinden et al. 1991), and one was a service for the ill child or young person (Cebula 2012).



³ As determined through behavioural observations of sibling dyads.

Efficacy and Effectiveness

Due to the large heterogeneity in outcomes, small sample sizes and, in some instances, poor study design, it is challenging to compare the efficacy and effectiveness of the interventions. The only RCT included in this review (Giallo and Gavidia-Payne 2008) had a sample size of only 21 across both treatment (12) and control (9). They offered a family-based psychoeducational-based intervention called SibStars. Using seven outcome measures (including an evaluation of the intervention), they found a significant improvement in stress, coping and adjustment, the emotional symptoms subscale of the SDQ (behaviour) and family functioning.

As 11 of the 17 studies (65%) adopted a within-subjects pre-post design, without the use of a control, the results of these papers should be carefully interpreted and no assumptions of causality can be made. Within the discussion of two of the studies, consideration was given to the value of time spent together between the child and parent as a by-product of the intervention, but this was not accounted for this in their analysis (Houtzager et al. 2001; Williams et al. 2003).

Meta-Analysis

There were eight papers that could be included in the metaanalysis since they reported on the same outcomes using the same study groups and time points (Sutton et al. 2000). Of these eight studies, three were group interventions, four were family-based interventions, and one was an intervention for the child with the health condition. Two studies looked at mental health, two looked at physical health, and four looked at a combination of the two. These numbers were too small to conduct a subgroup analysis in this meta-analysis, so the studies were only considered for their results relating to behaviour (internalising, externalising and total score) and knowledge.

Behaviour

The results of the SMD meta-analysis on behaviour was split into three categories: internalising, externalising and total score. Further, these papers were separated by whether they looked at pre-post measures in the treatment group or compared the intervention group post-treatment (Tx) to a control (Cntrl) group. Five papers were included in the analysis. The forest plots for each of the analyses can be found in Figs. 3 and 4. Cebula (2012) reported the SDQ from three different subjects (child, parent and teacher), to be consistent with other studies; only the parent report was included in the meta-analysis.

The pooled SMD estimates in the pre-post analysis (Fig. 3) all indicated improvement in behavioural outcomes on both SDQ and CBCL (as reflected through a reduction in

score, relative to the baseline score). The pooled SMDs were as follows: internalising (SMD = -0.34 [95% CI (-0.50, -0.18); p < 0.001]), externalising (SMD = -0.29[95% CI (-0.45, -0.13); p < 0.001]) and total score (SMD = -0.44 [95% CI (-0.6, -0.28); p < 0.001]). In contrast, the metanalysis of studies comparing a treatment group to a control group resulted in no significant difference in any of the scales of behavioural difficulties considered (Fig. 4).

Knowledge

No two papers used the same measure for knowledge. The results of the meta-analysis that included five studies demonstrated that overall there was a small significant improvement in knowledge following the intervention. Only one paper (McLinden et al. 1991) reported control group results; therefore, this study design was not considered in this meta-analysis; rather, all studies included used a pre-post treatment study design. The results of this analysis can be seen in the forest plot shown in Fig. 5. The pooled SMD estimate for knowledge improvement following intervention is 0.68 [95% CI (0.40, 0.95); p < 0.001].

Bias

Bias in a meta-analysis may be attributed to a range of sources including reporting biases, poor methodological design or chance. Typically, a funnel plot would be used to test for the presence of bias. A funnel plot plots the treatment effect (SMD) against study precision [standard error (se)]. If the funnel plot is not symmetrical within the 95% confidence interval, it is taken as a sign that there is bias present. To ensure that the asymmetry is not attributable to chance, it is recommended to conduct a test for funnel asymmetry, such as Egger's Test (Egger et al. 1997). However, this is not recommended if there are fewer than ten studies included in the analysis, as this is unlikely to distinguish true bias from chance due to low test power (Sterne et al. 2011). Therefore, due to the small number of studies included in this meta-analysis, no formal test for bias was completed.

Discussion

The literature evaluating interventions for siblings of children and young people with a chronic illness is diverse and produces varied results. Studies included in this review involved siblings of children with a range of chronic health conditions, used various techniques to help improve the child's psychological well-being and a range of measures to evaluate several outcomes. Each of these sources of heterogeneity provides significant challenges for conducting a systematic review and meta-analysis. The heterogeneity of



	Camps	Groups	Family-Based	For Child with Chronic Illness
Physical Health	Kieman et al. (2004) Sidhu et al. (2006)	Dolgin et al. (1997) Heiney et al. (1990) Houtzager et al. (2001)	Besier et a1 (2010)	
Mental Health		Evans et al. (2001) Phillips (1999) Smith and Perry (2005)	Kryzak et al. (2015)	Cebula (2011)
Combined	Williams et al. (2003)	D'Arcy et al. (2005) Granat et al. (2012) McLinden et al. (1991)	Giallo and Gavidia- Payne (2008)	

Fig. 2 Types of intervention offered by condition group

interventions and samples makes it particularly challenging to make firm conclusions on the effectiveness of interventions using a pre-post study design. Despite the challenges, the current meta-analysis provided some evidence of effectiveness of interventions for siblings of children and young people with a chronic illness. The analysis of knowledge scores using pre-post measures indicated a small significant positive effect on knowledge following the intervention. Offering a knowledge component in an intervention could help facilitate the child or young person's understanding of their sibling's condition. By increasing understanding, it is possible the sibling will feel they have more control and this may help increase their coping skills (Heiney et al. 1990) although there has been relatively little work in this area.

When the treatment group was considered pre-post treatment, there was a significant improvement in their internalising and externalising behaviours, as well as total score (reflected by a reduction in scores). Yet when we consider treatment group post-intervention relative to a control group, there was no difference in behaviour scores. One suggestion as to why this may be is that the research process increased the salience of the needs of the sibling to parents in the control group which means their outcomes also improved. Data collected in the included studies did not allow for this factor to be considered. Future research should attempt to account for such contamination effects. Further explanations include a lack of difference from control at pre-treatment, low sensitivity of measures, or potentially a bias from parent-reported measures, consideration for each potential explanation will follow.

The sensitivity of the measures used in evaluations of this type should be further considered. For instance, a few of the included studies noted that the children are within the "normal" range before receiving the intervention, and this has the potential to cause a ceiling effect on gains from the intervention (Giallo and Gavidia-Payne 2008; Kiernan et al. 2004; McLinden et al. 1991). As previous meta-analyses (Rossiter and Sharpe 2001; Sharpe and Rossiter 2002; Vermaes et al. 2012) have found a significant negative effect on sibling's psychological functioning, there appears to be a discrepancy between these samples and some of the samples considered in this review. This may be attributed to the



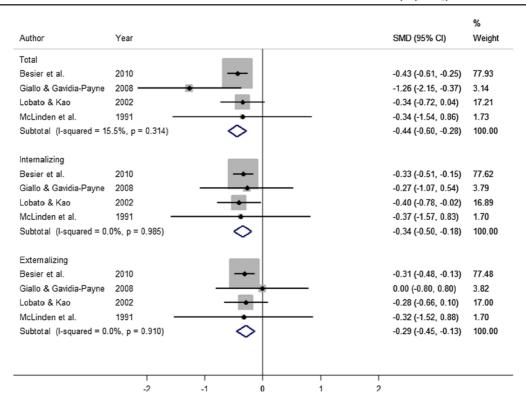


Fig. 3 Forest plot of pre-post behaviour results

samples themselves being unrepresentative or may be related to the measures used in these evaluations.

Another explanation as to why results differed by study design may be that the range in parent or sibling report measures resulted in either a downward (under-reporting) or upward (over-reporting) bias on the results. This singlerater bias has been noted in the literature (Rivers and Stoneman 2003). Within the papers included in this review, three consider the effect of parental reporting bias on results. Cebula (2012) compared the results given to the SDQ by parents, teachers and siblings and found that siblings view themselves significantly more negatively compared to their parents. Sidhu et al. (2006) recognised that parents are only able to report on the externalising behaviours of the sibling, whereas the sibling could report on their internalised problems, and their perceptions of these distresses were generally greater than parents. Finally, Lobato and Kao (2002) found a difference in ratings on the Sibling Perception Questionnaire (SPQ) and bring into question the sensitivity of parentreported results using SPQ.

The positive impact of the interventions on behaviour may operate via a change in the parent-sibling dyad. For instance, parents may spend more time with siblings as a direct result of being involved in the intervention, or it may be that parents gain a higher awareness of the sibling's needs due to the intervention. The increased time spent together between the sibling and parent could have an influence upon the result, but it is challenging to formally record this for it to be considered in the analysis (Houtzager et al. 2001; Williams et al. 2003).

Several of the studies that considered both physical and mental chronic health conditions gave justification as to why they chose to do so (Giallo and Gavidia-Payne 2008; Williams et al. 2003), but there is a lack of evidence as to which approach, disease-specific or broad, produces the optimum results. Evidence put forward by Vermaes et al. (2012) suggests that illnesses with a high morbidity and mortality may act as the largest moderating factors. Therefore, it may be beneficial to focus on siblings of children and young people that have a high impact and high mortality rate condition, regardless of whether the illness is categorised as physical or mental.

Strengths and Limitations

Our study is the first to synthesise the current literature evaluating interventions offered to siblings of children with chronic physical or mental health conditions or both.



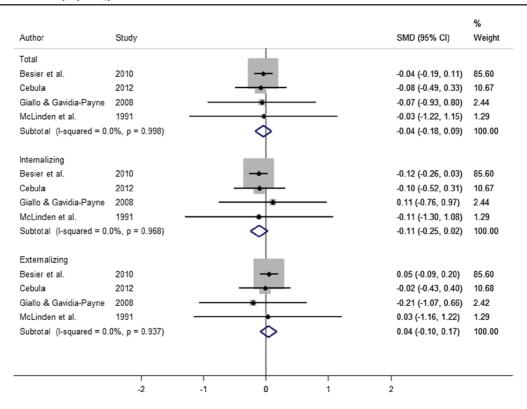
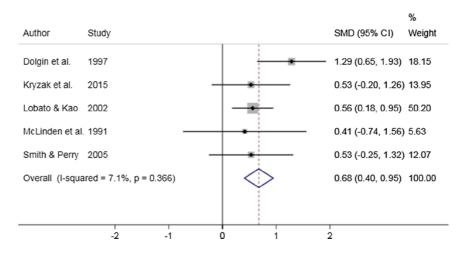


Fig. 4 Forest plot of treatment (Tx)-control (Cntrl) behaviour results

Fig. 5 Forest plot of meta-analysis of knowledge outcomes



Previous reviews have attempted to separate out the two groups, by physical or mental health conditions, and no previous meta-analysis has been completed in this area. The use of the broad approach allows a more complete picture of interventions currently available to siblings, by considering studies that have evaluated interventions focused on siblings of children and young people with a chronic mental

or physical health condition together. It has also highlighted how it may not be the most advantageous approach to consider these groups separately. Consideration has been given to various forms of interventions and has highlighted the importance of more robust and replicable research in this area.



The heterogeneity of the studies included in the review makes it difficult to draw firm conclusions regarding the effectiveness of psychological interventions aimed at siblings of children with chronic conditions. The studies were typically subject to low sample sizes, poor methodology and short follow-up periods; less than a quarter (24%) of the included studies report on a follow-up beyond 1 month postintervention. Although it is advantageous that this analysis has included studies that consider both physical and mental health conditions, this may also have increased the level of heterogeneity and reduced the clinical relevance, relative to reviews that have focused on solely mental or physical health conditions (Tudor and Lerner 2015). It should also be noted that the included studies were all from developed countries, and therefore, the results from this analysis cannot be generalised to those in low- and middle-income countries.

Not many studies were included in the meta-analysis due to a lack of consistent and compatible data. Of the included eight, five used an uncontrolled pre-post study design (Besier et al. 2010; Cebula 2012; Dolgin et al. 1997; Kryzak et al. 2015; Lobato and Kao 2002; Smith and Perry 2005) which makes causality difficult to establish. The four studies included in the meta-analysis that used a treatment–control design used various control groups: One used a waitlist control (Giallo and Gavidia-Payne 2008), one used the participants that refused to participate in the intervention (McLinden et al. 1991), another used a data sample from the general population (Besier et al. 2010), and finally, Cebula (2012) used a retrospective design, and thus, their control are willing subjects who have not previously/not currently using the intervention which limits the generalisability of the findings.

Further limitations of the included studies include a lack of acknowledgement for potential positive impacts of having a sibling with a chronic health condition. There is also a lack of consideration for the influence of parent-reported relative to child-reported outcomes. Cebula (2012) reported measures from parents, siblings and teachers to attempt to deal with this issue. In her analysis, she found that the child or young person reported themselves more negatively on two of the five domains of the SDQ, but they also appeared to have a slightly more positive perception of the sibling relationship, particularly empathy which may be due to the parents' greater attention to negative interactions (Cebula 2012; Rivers and Stoneman 2003). How these change and influence results following intervention may be an important consideration.

Across all studies, there were 23 outcomes considered; sufficient data were reported to combine behaviour and knowledge scores in a meta-analysis. The results of this analysis were limited by the small sample sizes of previous studies, along with methodological problems due to the lack of consistency in measures being used, type and protocol of interventions, and time points being considered. It would

be more beneficial if intervention studies that evaluated the same form of intervention, with the same type of population and using the same outcome measures could be statistically combined; unfortunately, with the current literature this is not possible.

Directions for Future Research

The primary recommendation from this review and metaanalysis is the need for stronger evidence, such as RCTs, which also capture a larger more representative sample of the population. Which tools should be used in evaluations also requires deliberation to help encourage consistency across studies.

Furthermore, studies should be conducted that include siblings of children with both mental and physical health conditions. It may also be important to consider potential moderating factors, including protective factors, which could help target and tailor support services to those most in need. For instance, considering different populations based on moderating factors, i.e. high/low burden, using the same intervention protocol would provide more informative evaluations in this area of research.

Conclusion

This review and meta-analysis improves upon the current literature by combining the existing findings in a systematic and robust manner, providing transparent results and reducing potential sources of bias. It is concluded that psychological well-being interventions for siblings of children and young people with chronic physical and mental health conditions lead to an improvement in illness knowledge and an improvement in externalising and internalising behaviour scores, when using a pre-post one group study design. The findings from the systematic review are mixed and inconsistent, which emphasises the need for additional work that better establishes the needs, appropriate methodologies and evaluation techniques for interventions offered to siblings of children with chronic health conditions.

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Compliance with Ethical Standards

Conflict of interest The authors declare that they have no conflict of interest.



Ethical Approval This article does not contain any studies that involved either human participants or animals performed by any of the authors, therefore ethical approval was not required.

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Online Resource 1: Full Search Strategy

Database	Topic/Headings/Subject/MeSH	Article Title, Abstract, Keywords	All Text/Key terms	Final Search
PsycINFO	1. Siblings		6. sibling* or brother* or sister*	12. 1 or 6
	2. Chronic disease			13. 2 or 7
	Mental Health		ineradicable) AND (disease* or illness* or	14. 3 or 8
	Quality of Life or Quality		condition* or syndrome* or disorder*)	15. 4 or 9
	Adjusted Life Years		8. "mental health" or "psychological wellbeing"	16. 14 or 15
	Social Support		or stress* or anxiety* or depress* or	
			_	18. 12 and 13
			9. "quality of life" or QoL or "quality adjusted	and 16 and
			life year" or QALY or "health related quality	17
			of life" or HRQoL	
			10. intervention*	
			 (support or self-help) and (group* or peer* or 	
			mental-health or social or group* or school*	
			or famil* or early)	
PsycEXTRA	1. Siblings or Brothers or Sisters		7. sibling* or brother* or sister*	13. 1 or 7
	2. Chronic illness		8. (chronic or lifelong or recur* or incurable or	14. 2 or 8
	Mental health or Well Being		ineradicable) AND (disease* or illness* or	15. 3 or 9
	 Quality of Life 		condition* or syndrome* or disorder*)	16. 4 or 10
	Intervention or early		mental health or psychological wellbeing or	17. 15 or 16
	intervention or family		stress* or anxiety* or depress* or	18. 5 or 6 or 11
	intervention or group		psychosocial	or 12
	intervention or school based		10. quality of life or QoL or quality adjusted life	19. 13 and 14
	intervention		year or QALY or health related quality of life	and 17 and
	Support Groups or mental		or HRQoL	18
	health services or self-help		11. intervention*	
	techniques or social support		 (support or self-help) and (group* or peer* or 	
			mental-health or social or group* or school*	
			or famil* or early)	
Embase	1. Sibling		7. sibling* or brother* or sister*	13. 1 or 7
	Chronic disease		 chronic or lifelong or recur* or incurable or 	14. 2 or 8
			meradicable) AND (disease* or illness* or	15. 3 or 9
	well being		condition* or syndrome* or disorder*)	16. 4 or 10
	Quality of life or quality		mental health or psychological wellbeing or	17. 15 or 16

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Mckenzie Smith, M., Pinto Pereira, S., Chan, L., Rose, C., & Shafran, R. (2017). Impact of Wellbeing Interventions for Siblings of Children and Young People with a Chronic Physical or Mental Health Condition. Clinical Child and Family Psychology Review.

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			stress" or anxiety" or depress" or	18. 5 of 6 of 11
	Intervention study or early		psychosocial	or 12
	intervention or early childhood		 quality of life or QoL or quality adjusted life 	19. 13 and 14
	intervention		year or QALY or health related quality of life	and 17 and
	Social support or support group		or HRQoL	18
			12. (support or self-help) and (group* or peer* or	
			mental-health or social or group* or school*	
			or famil* or early)	
PubMed	1. Siblings		01	12. 1 or 6
	2. Chronic disease		7. (chronic or lifelong or recur* or incurable or	13. 2 or 7
	mental health		ineradicable) AND (disease* or illness* or	14. 3 or 8
	4. quality of life or quality		condition* or syndrome* or disorder*)	15. 4 or 9
	adjusted life years		8. "mental health" or "psychological wellbeing"	16. 14 or 15
	5. social support		or stress* or anxiety* or depress* or	17. 5 or 10 or 11
			psychosocial	18. 12 and 13
			9. "quality of life" or QoL or "quality adjusted	and 16 and
			life year" or QALY or "health related quality	17
			of life" or HRQoL	
			10. intervention*	
			11. (support or self-help) and (group* or peer* or	
			mental-health or social or group* or school*	
CINAHIL	 Siblings or brothers or sisters 		 sibling* or brother* or sister* 	13. 1 or 7
	chronic disease or chronic			
	illness or long term conditions		ineradicable) AND (disease* or illness* or	15. 3 or 9
	or chronic conditions		condition* or syndrome* or disorder*)	16. 4 or 10
	mental health or well-being		mental health or psychological wellbeing or	
			stress* or anxiety* or depress* or	18. 5 or 6 or 11
	quality of life		psychosocial	or 12
	5. intervention or early		10. quality of life or QoL or quality adjusted life	19. 13 and 14
	intervention or family		year or QALY or health related quality of life	and 17 and
	intervention or group			18
	intervention or school based			
			 (support or self-help) and (group* or peer* or 	
	support groups or mental health		mental-health or social or group* or school*	
	services or self-help techniques		or famil* or early)	
	or social support			
Scopus		 sibling* or brother* or sister* 		1 and 2 and 3 and 4

Mckenzie Smith, M., Pinto Pereira, S., Chan, L., Rose, C., & Shafran, R. (2017). Impact of Wellbeing Interventions for Siblings of Children and Young People with a Chronic Physical or Mental Health Condition. Clinical Child and Family Psychology Review.

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	sibling* or brother* or sister* (chronic or lifelong or recur* or incurable or ineradicable) AND (disease* or illness* or condition* or syndrome* or disorder*) ("mental health" or "psychological wellbeing" or stress* or anxiety* or depress* or psychosocial) OR ("quality of life" or QoL or "quality adjusted life year" or QALY or "health related quality of life" or HRQoL) Intervention* or ((support or self-help) and (group* or peer* group* or school* or famil* or early))
2. (chronic or lifelong or recur* or incurable or ineradicable) AND (disease* or illness* or condition* or syndrome* or disorder*) 3. ("mental health" or "psychological wellbeing" or stress* or anxiety* or depress* or psychosocial) OR ("quality of life" or QoL or "quality adjusted life year" or QALY or "health related quality of life" or HRQoL) 4. intervention* or ((support or self-help) and (group* or peer* or mental-health or social or group* or sechiel)	
	1 and 2 and 3 and 4



PROSPERO

International prospective register of systematic reviews

Are targeted interventions for healthy siblings of young people with a chronic illness effective?

Mhairi McKenzie, Roz Shafran, Corah Lewis, Charlotte Rose, Lynette Chan

Citation

Mhairi McKenzie, Roz Shafran, Corah Lewis, Charlotte Rose, Lynette Chan. Are targeted interventions for healthy siblings of young people with a chronic illness effective? PROSPERO 2017 CRD42017056740 Available from:

https://www.crd.york.ac.uk/prospero/display record.php?ID=CRD42017056740

Review question

What interventions have been suggested and evaluated for 'healthy' siblings of children/young people with "chronic conditions"? Have they been successful?

How have these interventions targeted the mental health and well being of the 'healthy' siblings? Which outcome measures have they used?

Have any interventions looked in detail at subgroup analysis, such as gender, age, ethnicity, time since diagnosis, and primarily have they compared the interventions in healthy siblings of children/young people with physical, mental, or co-morbid chronic conditions?

What is the suggested 'next step'?

Searches

Databases to be searched: PsycINFO, PsycEXTRA, Embase, CINAHL, PubMed, Scopus and Web of Science

Terms to be used in the searches: sibling*, brothers*, sister*, chronic, illness, mental health, well-being, quality of life, lifelong, recur*, support*, intervention*, peer*, incurable, ineradicable, disease, condition, syndrome, disorder, psychological wellbeing, stress, anxiety, depress, psychosocial, quality adjusted life year, health related quality of life, intervention*, support, mental health services, self-help, social support, early, group*, peer*, school*, famil*.

No restrictions will be put in place on the searches at this point; these will be applied in the elimination stage.

The search strategies were piloted by two independent researches in November 2016, and after necessary changes were made were ran again by three independent researchers to include all research published until the end of 2016.

Types of study to be included

Any trial (randomised or non-randomised) will be accepted, but it is required that pre- and post- measures of some form have been taken in the studies. Studies will be excluded if they do not mention any evaluation of the intervention, or focus on the development only.

Condition or domain being studied

The primary condition to be studied in this review is a "chronic condition" (as defined by each individual study), either physical or mental, in childhood, and its impact on a healthy sibling. Outcomes include mental-health, well-being, adaption, quality of life, and any other outcome which is used across multiple intervention evaluations.

Participants/population

Participants of interest in this study are 'healthy' siblings of a child/young person with a chronic condition.

The participant must have no known long-term condition.

They should be of 18 years of age or less.

The study should not have been conducted after the death of their sibling.

The sibling should not be a donor or caregiver for their sibling.

Intervention(s), exposure(s)

Studies must consider an intervention looking to improve the experience/well-being/mental health/quality of life of a 'healthy' sibling of a child or young person with a "chronic condition". Family-based interventions will

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be considered only where there is sufficient evidence regarding a 'healthy' sibling.

Comparator(s)/control

The ideal control in the included studies would be 'healthy' siblings that have not received the treatment or have been wait-listed. A pre/post repeated measure design will also be included with no control group.

Context

It has been noted that the family of a child with a chronic condition may have needs which continue to go under identified and supported. While there is a fairly large discussion on the impact on parents or caregivers in the literature, the evidence base in regards to siblings is relatively small. While qualitative interviews have facilitated an understanding of the siblings experiences it has a limited usefulness when developing any support for the siblings, or when attempting a comparison of the lives of the sibling and their peers. Siblings may have varying levels of involvement and responsibility when they have a sibling with a chronic condition. This can go from the extremes of potential donor or caregiver to living in a separate household to their sibling. This study is interested in the siblings who have minimal responsibility from their sibling, i.e. no caregiving, but do live in the same household, as living in a separate household may dramatically reduce the impact of having a sibling with a chronic condition.

The unique aspects of this group and their experiences need to be considered and a balanced approach taken to understanding and responding to their needs. Some of the evidence has been brought together to inform interventions targeting this at risk group, but there remains a lack of evaluation, understanding and widespread implementation of these tools.

Main outcome(s)

Any mental-health, well-being, and quality of life measures used both pre- and post-intervention.

Timing and effect measures

A pre- and post- measure must be given. It would be beneficial for the time since intervention to be recorded and for there to be multiple time points post-intervention, although this is not necessary.

Additional outcome(s)

These will be gathered from any scales used across two or more studies, and could include knowledge, adaption, understanding, social skills, or any other related outcome for healthy siblings.

Timing and effect measures

A pre- and post- measure must be given. It would be beneficial for the time since interventions to be recorded and for there to be multiple time points post-intervention, although this is not necessary.

Data extraction (selection and coding)

Data will be extracted using an adapted Effective Practice and Organisation of Care (EPOC) Data collection form

Data will be collected in 9 categories:

- 1. General information;
- 2. Population and setting;
- 3. Methods;
- 4. Participants;
- 5. Intervention groups;
- 6. Outcomes;
- 7. Results;
- 8. Applicability;
- 9. Other.

Risk of bias (quality) assessment

To limit bias, three independent researchers will evaluate the studies for inclusion based on abstracts, and any disagreements will be resolved through discussion.

Following this, two independent researchers will check full texts for eligibility, will extract the data, and assess its quality. Quality will be evaluated using the Effective Public Health Practice Project quality assessment tool for quantitative studies.



Strategy for data synthesis

Quantitative synthesis will be completed where possible to aggregate the findings across studies, and will be weighted to account for the quality of the paper using the EPHPP results.

Where quantitative synthesis is not possible due to heterogeneity of outcomes, a narrative analysis will be used.

Analysis of subgroups or subsets

The key subgroup analysis of interest in this review is a comparison of the interventions and outcomes when the child's sibling has either a mental or a physical chronic condition.

Subgroup analysis will be conducted based on demographic variables where possible.

Contact details for further information

Mhairi McKenzie

mhairi.mckenzie.16@ucl.ac.uk

Organisational affiliation of the review

UCL Great Ormond Street Institute of Child Health

https://www.ucl.ac.uk/ich

Review team members and their organisational affiliations

Miss Mhairi McKenzie. UCL GOSICH

Professor Roz Shafran. UCL GOSICH

Miss Corah Lewis. UCL GOSICH/Manchester University

Miss Charlotte Rose. UCL GOSICH Miss Lynette Chan. UCL GOSICH

Type and method of review

Systematic review

Anticipated or actual start date

01 November 2016

Anticipated completion date

31 May 2017

Funding sources/sponsors

UCL Great Ormond Street Institute of Child Health

Conflicts of interest

None known

Language

English

Country

England

Stage of review

Review Ongoing

Subject index terms status

Subject indexing assigned by CRD

Subject index terms

Adaptation, Psychological; Adolescent; Adolescent Health; Child; Child Health; Chronic Disease; Humans; Life Change Events; Mental Health; Quality of Life; Risk Factors; Siblings; Treatment Outcome

Date of registration in PROSPERO

06 February 2017





Date of publication of this version 06 February 2017

Details of any existing review of the same topic by the same authors

Stage of review at time of this submission

Stage	Started	Completed
Preliminary searches	Yes	Yes
Piloting of the study selection process	Yes	Yes
Formal screening of search results against eligibility criteria	Yes	No
Data extraction	Yes	No
Risk of bias (quality) assessment	No	No
Data analysis	No	No
Variana		

Versions

06 February 2017

PROSPERO

This information has been provided by the named contact for this review. CRD has accepted this information in good faith and registered the review in PROSPERO. The registrant confirms that the information supplied for this submission is accurate and complete. CRD bears no responsibility or liability for the content of this registration record, any associated files or external websites.

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Online Resource 2: Quality Assessment Results

Author	Year	Selection Bias	Study Design	Confounders	Blinding	Data Collection Methods	Withdrawals and Drop-Outs	Overall
Besier et al.	2010	*	*	*	*	**	*	*
Cebula	2012	*	*	***	*	***	N/A	*
D'Arcy et al.	2005	**	*	***	**	*	*	*
Dolgin et al.	1997	**	*	***	*	*	***	*
Evans, Jones & Mansell	2001	*	**	***	*	*	*	*
Giallo & Gavidia-Payne	2008	*	***	***	*	*	*	*
Granat et al.	2012	**	**	**	*	*	* * *	*
Heiney et al.	1990	**	**	*	**	*	*	*
Houtzager, Frootenhuis, & Last	2001	*	*	***	**	*	*	*
Kieman et al.	2004	*	*	***	**	*	*	*
Kryzak et al.	2014	*	*	***	**	***	**	**
Lobato & Kao	2002	*	*	***	**	***	***	*
McLinden et al.	1991	*	*	*	*	*	*	*
Phillips	1999	*	***	***	**	***	***	**
Sidhu et al.	2006	**	*	***	**	***	***	***
Smith & Perry	2005	**	*	***	**	***	***	***
Williams et al.	2003	*	***	***	**	***	***	**

 $\star \star \star = \text{Strong}; \star \star = \text{Moderate}; \star = \text{Weak}$



Professor Roz Shafran Institute of Child Health University College London Guilford Street WC1N 3EH

24 March 2017

Dear Professor Shafran

Qualitative research project Funding confirmation

I am writing to confirm that the Cystic Fibrosis Trust agrees to pay University College London the sum of £37,000 to conduct a qualitative research study among people with cystic fibrosis and their families.

Project details

Project Title (provisional): A study to understand needs, challenges and aspirations of people with cystic fibrosis and their siblings in the UK

The study includes: under 25s with CF; over 25s with CF; siblings of people with CF; questionnaire for parents of children with CF.

The award is made to Professor Roz Shafran and Dr Mandy Bryon as joint Principal investigators, and to Mhairi McKenzie and Anna Coughtrey as co-Investigators.

Any additional funding requirements would be subject to discussion and formal agreement before funding authorisation.

Yours sincerely,

Dr Keith Brownlee

Director of Impact, Cystic Fibrosis Trust

Fighting for a Life Unlimited

Cystic Fibrosis Trust is registered as a charity in Ergiand and Wales (1979046) and Scotland (BC640196) A company limited by guarantee, registered in Ergiand and Wales number 3880213, Registered office:

Cystic Fibrosis Trust 2nd Floor, One Aldgate, London, EC3N 1RE 020 3795 1555 cysticfibrosis.org.uk enquiries@cysticfibrosis.org.uk Patron: His Royal Highness The Prince of Wales, KG, KT, GCB, OM



Professor Roz Shafran Institute of Child Health University College London Guilford Street WC1N 3EH

17 May 2017

Dear Professor Shafran

Qualitative research project – additional funding confirmation (parents extension)

I am writing to confirm that the Cystic Fibrosis Trust agrees to pay University College London the additional sum of £9,994 (on top of the £37,000 already agreed) to conduct an extension of the qualitative research project to include parents of children with cystic fibrosis in the study.

The funding is confirmed until the end of 2018.

Project details

Project Title (provisional): A study to understand needs, challenges and aspirations of people with cystic fibrosis, their siblings and parents in the UK

The study includes: under 25s with CF; over 25s with CF; siblings of people with CF; parents of children with CF.

The award is made to Professor Roz Shafran and Dr Mandy Bryon as joint Principal investigators, and to Mhairi McKenzie and Anna Coughtrey as co-Investigators.

Any additional funding requirements would be subject to discussion and formal agreement before funding authorisation.

Yours sincerely,

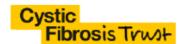
Dr Keith Brownlee

Director of Impact, Cystic Fibrosis Trust

Fighting for a Life Unlimited

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Cystic Fibrosis Trust 2nd Floor, One Aldgate, London, EC3N 1RE 020 3795 1555 cysticfibrosis.org.uk enquiries@cysticfibrosis.org.uk Patron: His Royal Highness The Prince of Wales, KG, KT, GCB, OM







Information about the research for individuals with a sibling who has Cystic Fibrosis.

Study Title: Experiences of Living with Cystic Fibrosis; The impact on children, young people, adults and their families.

We work at The Great Ormond Street Institute of Child Health. We would like to invite you to take part in a research study. Before you decide if you would like to take part, it is important for you to understand why the research is being done and what it will involve for you.

Please read through the following information carefully. Feel free to discuss it with others, or any of our team if you wish, and take your time to decide whether or not you wish to take part.

Please ask us if there is anything that is not clear or if you would like more information.

What is the research and why are we doing it?

We want to find out more about what life is like with a brother or sister with Cystic Fibrosis (CF), as we know this can be challenging at times. The information you give would be used to help the Cystic Fibrosis Trust better understand what it is like to have a brother or sister with CF.



The information you give us would also greatly help the Cystic Fibrosis Trust in developing valuable support services for brothers and sisters of children with CF.

The project looking at siblings is being completed as part of larger study which is investigating the impact of CF on individuals with CF 11yrs and over, and their families.

Why have I been invited?



We are inviting anyone who has a brother or sister with CF, and is between the age of 11 and 25yrs, to take part.

Do I have to take part?



It is up to you to decide if you would like to take part in the study. If you agree to take part, we will then ask you to sign a form, saying you agree to take part. If you are under the age of 16 we will also need your parents to sign aform saying it is ok for you to take part. You can stop taking part in the study at any point even if you said yes at the beginning.

Sibling Information Leaflet 11-16 Date: 23/03/2017 Version No: 2 IRAS ID: 219455

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What will taking part involve?

If you would like to take part in the study, we will arrange a time to speak with you that works for you. We are happy to talk to you on the phone, via skype or face to face. We can meet you at your home, GOSH or at another location that works foryou.



First, we will ask you to complete a short questionnaire. We will then ask you some longer questions about different areas of your life such as your school, friends and home life. Overall, it will take no longer than one hour.

Expenses and Payments

You will receive a £10 Amazon gift voucher for your time. We will also pay for your travelto and from the interview if this is done face to face.

Is there anything to be worried about if I take part?

There should not be any side effects or risks with this study. If you are upset by taking part in the study you can speak to your parents or a member of our team. Our address and phone number are at the end of this sheet.



Will taking part help me?

The study may not directly benefit yourself, however the information we get from this study will help improve the support services available from the Cystic Fibrosis Trust.

Who will know I am taking part in the study?

We would keep your name, address and any other personal information secret. Only members of the research team will know you are taking part. We will write about the answers given in interviews but no names will be used.

All the information you give us will be stored securely either electronically or in paper form at UCL Great Ormond Street Institute of Child Health.

Who has organised the research?

Before any research goes ahead it has to be checked by a Research Ethics Committee. They make sure the research is fair. This project has been checked and approved by the Bromley Research Ethics Committee.

What if there is a problem?

If you have a concern about any aspect of this study, you should ask to speak to the researchers, who will do their best to answer your questions. If you remain unhappy

Sibling Information Leaflet 11-16 Date: 23/03/2017

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and wish to complain formally, you can do this by contacting the PALS Office at Great Ormond Street Hospital (telephone 020 7829 7862 or email pals@gosh.nhs.uk).

In the event that something does go wrong and you are harmed during the research and this is due to someone's negligence then you may have grounds for a legal action for compensation against Great Ormond Street Hospital NHS Trust but you may have to pay your legal costs. The normal National Health Service complaints mechanisms will still be available to you (if appropriate).

What will happen to the results of the study?

We will write to you to let you know the overall findings of the study.

We hope to publish the findings of the study. Some of the findings will also be used by two students for academic projects (BSc Psychology and PhD Child Health Research). No names or other identifiable information will be used in any reports or publications.



Who is funding the research?

This research has been funded by the Cystic Fibrosis Trust.

Contact for further information

If you would like any further information, please contact the research team.



Contact: Mhairi McKenzie

Address: UCL Great Ormond Street Institute of Child

Health 30 Guilford Street, London, WC1N 1EH

Email: ICH.CFstudy@ucl.ac.uk

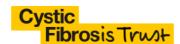
Telephone:



Sibling Information Leaflet 11-16 Date: 23/03/2017 Version No: 2 IRAS ID: 219455

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Information about the research for individuals with a sibling who has Cystic Fibrosis.

Study Title: Experiences of Living with Cystic Fibrosis; The impact on children, young people, adults and their families.

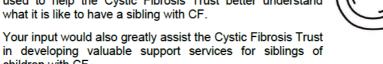
We work at The Great Ormond Street Institute of Child Health. We would like to invite you to take part in a research study. Before you decide if you would like to take part, it is important for you to understand why the research is being done and what it will involve for you.

Please read through the following information carefully. Feel free to discuss it with others, or any of our team if you wish, and take your time to decide whether or not you wish to take part.

Please ask us if there is anything that is not clear or if you would like more information.

What is the research and why are we doing it?

We want to find out more about what life is like with a sibling with Cystic Fibrosis (CF), as we know this can be challenging at times. The information you give would be used to help the Cystic Fibrosis Trust better understand what it is like to have a sibling with CF.





The project looking at siblings is being completed as part of larger study which is investigating the impact of CF on individuals with CF 11yrs and over, and their families.

Why have I been invited?



children with CF.

We are inviting anyone who has a sibling with CF, and is 11yrs of age or over, to take part in the study looking at siblings.

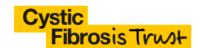
Do I have to take part?



It is up to you to decide if you would like to take part in the study. If you agree to take part, we will then ask you to sign a consent form. this confirms you are happy to take part in the study. You can stop taking part in the study at any point even if you said yes at the beginning.

Sibling Information Leaflet 17+ Date: 23/02/2017 Version No: 2

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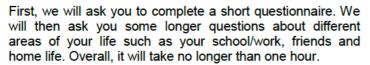






What will taking part involve?

If you give consent to take part in the study, we will arrange a time to speak with you that works for you. We are happy to talk to you on the phone, via skype or face to face. We can meet you at your home, GOSH or at another location convenient for you.





Expenses and Payments

You will receive a £10 Amazon gift voucher for your time. We will also reimburse any travel expenses to and from the interview if this is done face to face.

Is there anything to be worried about if I take part?

There should not be any side effects or risks with this study. If you are upset by taking part in the study you can speak to your parents or a member of our team. Our address and phone number are at the end of this sheet.



Will taking part help me?

The study may not directly benefit yourself, however the information we get from this study will help improve the support services available from the Cystic Fibrosis Trust.

Who will know I am taking part in the study?

We would keep your name, address and any other personal information secret. Only members of the research team will know you are taking part. We will write about the answers given in interviews but no names will be used.

All the information you give us will be stored securely either electronically or in paper form at UCL Great Ormond Street Institute of Child Health.

Who has organised the research?

Before any research goes ahead it has to be checked by a Research Ethics Committee. They make sure the research is fair. This project has been checked and approved by the Bromley Research Ethics Committee.

What if there is a problem?

If you have a concern about any aspect of this study, you should ask to speak to the researchers, who will do their best to answer your questions. If you remain unhappy and wish to complain formally, you can do this by contacting the PALS Office at Great Ormond Street Hospital (telephone 020 7829 7862 or email pals@gosh.nhs.uk).

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In the event that something does go wrong and you are harmed during the research and this is due to someone's negligence then you may have grounds for a legal action for compensation against Great Ormond Street Hospital NHS Trust but you may have to pay your legal costs. The normal National Health Service complaints mechanisms will still be available to you (if appropriate).

What will happen to the results of the study?

We will write to you to let you know the overall findings of the study.

We hope to publish the findings of the study. Some of the findings will also be used by two students for academic projects (BSc Psychology and PhD Child Health Research). No names or other identifiable information will be used in any reports or publications.



Who is funding the research?

This research has been funded by the Cystic Fibrosis Trust.

Contact for further information

If you would like any further information, please contact the research team.



Contact: Mhairi McKenzie

Address: UCL Great Ormond Street Institute of Child

Health 30 Guilford Street, London, WC1N 1EH

Email: ICH.CFstudy@ucl.ac.uk

Telephone:

Sibling Information Leaflet 17+ Date: 23/02/2017 Version No: 2 IRAS ID: 219455

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Information about the research for young people with Cystic Fibrosis aged 11-17 years

Study Title: Experiences of Living with Cystic Fibrosis; The impact on children, young people, adults and their families.

We work at The Great Ormond Street Institute of Child Health. We would like to invite you to take part in a research study. Before you decide if you would like to take part, it is important for you to understand why the research is being done and what it will involve for you.

Please read through the following information carefully. Feel free to discuss it with others, or any of our team if you wish, and take your time to decide whether or not you wish to take part.

Please ask us if there is anything that is not clear or if you would like more information.

What is the research and why are we doing it?

We want to find out more about what life is like with Cystic Fibrosis (CF) as we know this can be challenging at times. The information you give would be used to help the Cystic Fibrosis Trust better understand what it is like to be a young person with CF.



The information you give us would also greatly helpthe Cystic Fibrosis Trust in developing valuable support services for young people with CF.

Why have I been invited?



We are inviting anyone with CF aged 11 or over to take part in the study.

Do I have to take part?



It is up to you to decide if you would like to take part in the study. If you agree to take part, we will then ask you to sign a form saying you agree to take part. If you are under 16 years old, we will also need your parents to sign a form saying it is ok for you to take part.

You can stop taking part in the study at any point even if you said yes at the beginning. This would not affect the standard of care you receive or how quickly you receive care.

Young Person 11-17 Information Leaflet Date: 23/03/2017 Version No: 2 IRAS ID: 219455

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What will taking part involve?

If you would like to take part in the study, we will arrange a time to speak with you that works for you. We are happy to talk to you on the phone, via skype or face to face. We can meet you at your home, GOSH or at another location that works for you.

First, we will ask you to complete a short questionnaire. We will then ask you some longer questions about different areas of your life such as your school, friends and home life. Overall, it will take no longer than one hour.



Expenses and Payments

You will receive a £10 Amazon gift voucher for completing the interview. We will also pay for your travel to and from the interview if this is done face to face.

Is there anything to be worried about if I take part?

There should not be any side effects or risks with this study. If you are upset by taking part in the study, please speak to your parents to begin with. If you would like to speak to someone else our address and phone number are at the end of this sheet. Your treatment will not be changed by taking part.



Will taking part help me?

The study may not directly benefit yourself, however the information we get from this study will help improve the support offered by the Cystic Fibrosis Trust.

Who will know I am taking part in the study?

We would keep your name, address and any other personal information secret. Only members of the research team will know you are taking part. We will write about the answers given in interviews but no names will be used.

All the information you give us will be stored securely either electronically or in paper form at UCL Great Ormond Street Institute of Child Health.

Who has organised the research?

Before any research goes ahead it has to be checked by a Research Ethics Committee. They make sure the research is fair. This project has been checked and approved by the Bromley Research Ethics Committee.

What if there is a problem?

If you have a concern about any aspect of this study, you should ask to speak to the researchers, who will do their best to answer your questions. If you remain unhappy and wish to complain formally, you can do this by contacting the PALS Office at

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Great Ormond Street Hospital (telephone 020 7829 7862 or email pals@gosh.nhs.uk).

In the event that something does go wrong and you are harmed during the research and this is due to someone's negligence then you may have grounds for a legal action for compensation against Great Ormond Street Hospital NHS Trust but you may have to pay your legal costs. The normal National Health Service complaints mechanisms will still be available to you (if appropriate).

What will happen to the results of the study?

We will write to you to let you know the overall findings of the study.

We hope to publish the findings of the study. Some of the findings will also be used by two students for academic projects (BSc Psychology and PhD Child Health Research). No names or other identifiable information will be used in any reports or publications.



Who is funding the research?

This research has been funded by the Cystic Fibrosis Trust.

Contact for further information

If you would like any further information, please contact the research team.

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Contact: Mhairi McKenzie

Address: UCL Great Ormond Street Institute of Child

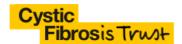
Health 30 Guilford Street, London, WC1N 1EH

Email: ICH.CFstudy@ucl.ac.uk

Telephone:



Young Person 11-17 Information Leaflet Date: 23/03/2017 Version No: 2 IRAS ID: 219455







Information about the research for adults (aged 18 years and over) with Cystic Fibrosis

Study Title: Experiences of Living with Cystic Fibrosis; The impact on children, young people, adults and their families.

We work at The Great Ormond Street Institute of Child Health. We would like to invite you to take part in a research study. Before you decide if you would like to take part, it is important for you to understand why the research is being done and what it will involve for you.

Please read through the following information carefully. Feel free to discuss it with others, or any of our team if you wish, and take your time to decide whether or not you wish to take part.

Please ask us if there is anything that is not clear or if you would like more information.

What is the research and why are we doing it?

We want to find out more about what life is like with Cystic Fibrosis (CF) as we know this can be challenging at times. The information you give would be used to help the Cystic Fibrosis Trust better understand what it is like to be an individual with CF.



Your input would also greatly assist the Cystic Fibrosis Trust in developing valuable support services for individuals with CF.

Why have I been invited?



We are inviting anyone with CF aged 11 or over to take part in the research.

Do I have to take part?

It is up to you to decide to join the study. If you agree to take part, we will then ask you to sign a consent form. You are free to withdraw at any time, without giving a reason. This would not affect the standard of care you receive or how quickly you receive care.

What will taking part involve?

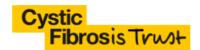
If you would like to take part in the study, we will arrange a time to speak with you that works for you. We are happy to talk to you on the phone, via skype or face to face. We can meet you at your home, GOSH or at another location convenient for you.

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Date: 23/03/2017 Version No: 2 IRAS ID: 219455

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First, we will ask you to complete a short questionnaire. We will then ask you some longer questions about different areas of your life such as your home life, friends and university/work life if applicable. Overall, it will take no longer than one hour.



Expenses and Payments

You will receive a £10 Amazon gift voucher for your time. We will reimburse expenses for travel to and from the interview if you choose to do it in person.

Is there anything to be worried about if I take part?

There are no specific risks to taking part in the study. If you become upset by taking part in the study and you would like to speak to someone our address and phone number are at the end of this sheet. Your treatment will not be changed by taking part.



Will taking part help me?

The study may not directly benefit yourself, however the information we get from this study will help improve the support available to people with CF.

Will my taking part in the study be kept confidential?

We would keep your name, address and any other personal information completely confidential. Only the research team will know you are taking part in the study. We will write about the answers given in interviews but no names will be used.

All electronic data will be stored on the University College London (UCL) SLMS Data Safe Haven. This is a technical environment for receiving, handling and storing sensitive data securely. Any information you give us in paper form will be stored in a secure locked room at UCL Great Ormond Street Institute of Child Health.

Who has organised the research?

Before any research goes ahead it has to be checked by a Research Ethics Committee. They make sure the research is fair. This project has been checked and approved by the Bromley Research Ethics Committee.

What if there is a problem?

If you have a concern about any aspect of this study, you should ask to speak to the researchers, who will do their best to answer your questions. If you remain unhappy and wish to complain formally, you can do this by contacting the PALS Office at Great Ormond Street Hospital (telephone 020 7829 7862 or email pals@gosh.nhs.uk).

Young Person 18+ Information Leaflet

Date: 23/03/2017 Version No: 2 IRAS ID: 219455

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In the event that something does go wrong and you are harmed during the research and this is due to someone's negligence then you may have grounds for a legal action for compensation against Great Ormond Street Hospital NHS Trust but you may have to pay your legal costs. The normal National Health Service complaints mechanisms will still be available to you (if appropriate).

What will happen to the results of the study?

We will write to you to let you know the overall findings of the study.

We hope to publish the findings of the study. Some of the findings will also be used by two students for academic projects (BSc Psychology and PhD Child Health Research). No names or other identifiable information will be used in any reports or publications.



Who is funding the research?

This research has been funded by the Cystic Fibrosis Trust.

Contact for further information

If you would like any further information, please contact the research team.



Contact: Mhairi McKenzie

Address: UCL Great Ormond Street Institute of Child

Health 30 Guilford Street, London, WC1N 1EH

mail: ICH.CFstudy@ucl.ac.uk

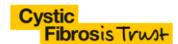
Telephone:



Young Person 18+ Information Leaflet Date: 23/03/2017 Version No: 2 IRAS ID: 219455

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Information about the research for parents

Study Title: Experiences of Living with Cystic Fibrosis; The impact on children, young people, adults and their families.

We work at The Great Ormond Street Institute of Child Health. We would like to invite you and your child(ren) to take part in a research study. Before you decide if you would like to take part, it is important for you to understand why the research is being done and what it will involve for you and your family.

Please read through the following information carefully. Feel free to discuss it with others, or any of our team if you wish, and take your time to decide whether or not you and your child(ren) wish to take part.

Please ask us if there is anything that is not clear or if you would like more information.

What is the research and why are we doing it?

We want to find out more about what life is like with Cystic Fibrosis (CF) as we know this can be challenging at times. We are also interested in what life is like to have a sibling/child with the condition. The information you and your child(ren) give would be used to help the Cystic Fibrosis Trust better understand what it is like to be an individual with CF, or their family.

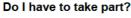


Your input would also greatly assist the Cystic Fibrosis Trust in developing valuable support services for individuals with CF and their families, with CF.



Why have I been invited?

We are inviting anyone with CF and their siblings (over the age of 11) to take part in the study. We are also inviting the parents of individuals with CF to take part.





It is up to you and your child(ren) to decide if you would like to take part in the study. If you agree to take part, we will then ask you to sign a consent form for yourself and another on behalf of your child if they are aged under 16 years. We will also ask your child to sign an assent form. You are free to withdraw at any time, without giving a reason. This would not affect the standard of care your child receives or how quickly they receive care.

Parent/Carer Information Leaflet Date: 18/05/2017 Version No: 3 IRAS ID: 219455

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What will taking part involve?

If you would like to take part in the study, we will arrange a time to speak with you and your child(ren) that works for you. We are happy to talk to you on the phone, via skype or face to face. We can meet you at your home, GOSH or at another location convenient for you.

Participation (whether this be yourself, your child with CF or their sibling) will involve completing one or two short questionnaires, and an interview including questions about different areas such as home life, friends and university/work life if applicable. Overall, each interview will take no longer than one hour.



Expenses and Payments

Each participant will receive a £10 Amazon gift voucher for their time. We will reimburse expenses for travel to and from the interview if you choose to do it in person.

Is there anything to be worried about if I take part?

There are no specific risks to taking part in the study. If you or your child(ren) are upset by taking part in the study and you would like to speak to someone, our address and phone number are at the end of this sheet. Your child's treatment will not be changed by taking part.



Will taking part help me?

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The study may not directly benefit yourself or your child(ren), however the information we get from this study will help improve the support available to people with CF and their families.

Will my taking part in the study be kept confidential?

We would keep your and your child's name, address and any other personal information completely confidential. Only the research team will know you are taking part in the study. We will write about the answers given in interviews but no names will be used.

All electronic data will be stored on the University College London (UCL) SLMS Data Safe Haven. This is a technical environment for receiving, handling and storing sensitive data securely. Any information you give us in paper form will be stored in a secure locked room at UCL Great Ormond Street Institute of Child Health.

Parent/Carer Information Leaflet

Date: 18/05/2017 Version No: 3 IRAS ID: 219455

Page 2 of 3







Who has organised the research?

Before any research goes ahead it must be checked by a Research Ethics Committee. They make sure the research is fair. This project has been checked and approved by the Bromley Research Ethics Committee.

What if there is a problem?

If you or your child(ren) have a concern about any aspect of this study, you should ask to speak to the researchers, who will do their best to answer your questions. If you remain unhappy and wish to complain formally, you can do this by contacting the PALS Office at Great Ormond Street Hospital (telephone 020 7829 7862 or email pals@gosh.nhs.uk).

In the event that something does go wrong and you or your child are harmed during the research and this is due to someone's negligence then you may have grounds for a legal action for compensation against Great Ormond Street Hospital NHS Trust but you may have to pay your legal costs. The normal National Health Service complaints mechanisms will still be available to you (if appropriate).

What will happen to the results of the study?

We will write to you to let you know the overall findings of the study.

We hope to publish the findings of the study. Some of the findings will also be used by two students for academic projects (BSc Psychology and PhD Child Health Research). No names or other identifiable information will be used in any reports, publications or academic work.



Who is funding the research?

This research has been funded by the Cystic Fibrosis Trust.

Contact for further information

If you would like any further information, please contact the research team.



Contact: Mhairi McKenzie

Address: UCL Great Ormond Street Institute of Child

Health 30 Guilford Street, London, WC1N 1EH

Email: ICH.CFstudy@ucl.ac.uk

Telephone:



Parent/Carer Information Leaflet Date: 18/05/2017

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Interview Schedule - Siblings

Introduction

I'm going to ask you a few questions about your experience as a sibling of a child with Cystic Fibrosis. The interview should take no longer than 45minutes, but please tell me if you would like to take a break or stop altogether. If you don't want to answer any of the questions, let me know and we can go on to the next one. Are you happy to continue?

(Would you prefer if your parent stayed with you during the interview?)

Topics

Impact on Day-to-Day life

'Do you believe your sibling's condition has impacted your day-to-day life?'

Prompts.

- Has it influenced your friendships and relationships with others? If so, how?
- Is your school/work life affected? How?
- Does it affect your social life?

Feelings

'How does your siblings condition make you feel?'

Prompts:

- Are you generally a happy person?
- Do you feel different from other children/people your age? (age appropriate)

Awareness of CF

'How much do you think other people know about CF?'

Prompts:

- Do you believe you have a good understanding of the condition?
- Do you think there should be more done to increase understanding of the condition?
- How do you think social media could be used to promote CF?
- How comfortable do you feel talking about your sibling's condition to others?

CF Trust Future Priorities

'What do you think the CF Trust should focus their work on in the future?'

Prompts:

- Research? What would you research?
- Awareness? How would you raise awareness?
- Fund raising? i.e. Event How do you think you would raise more funds?
- Support Services? Who for? What kind of support services?

Sibling Interview Schedule Date: 03/02/2017 Version No: 1 IRAS ID: 219455

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Closing the Interview

Are there any further points you would like to add about the experience of having a sibling with CF?

We've now come to the end of the interview. Thank you for taking the time to answer my questions. I have just a few more things to ask you:

- Would you like a summary of general findings at the end of the study?
- What would be the best way to send that to you?
- Are there any important questions that you think we missed out?
- Would you like to ask any questions or make any comments now that the interview is over?
- If you think of any questions, then please get in touch using the details in the consent form.

Sibling Interview Schedule Date: 03/02/2017 Version No: 1 IRAS ID: 219455

Interview Schedule (11-17yrs)

Introduction

I am going to ask you a few questions about your experience with CF and how it can impact on various aspects on your life. The interview should take no longer than 45 minutes, but please tell me if you would like to take a break or stop altogether. If you don't want to answer any of the questions, let me know and we can go onto the next one. Are you happy to continue?

Questions (with prompts)

Impact on day-to-day life

'How has having CF impacted your everyday life?'

Prompts -

- Do you think it changes your friendships and relationships with others? If so, how?
- Is your school life affected? If so, how?
- Does it affect your time spent with friends?
- What do you find hard?

'How does CF make you feel (about yourself)?'

Prompts -

- Does your CF make you feel isolated?
- Do you think it impacts your confidence? If so, how?
- Are you generally a happy person?

Awareness of CF

'How much do you tell other people about your CF?'

Prompts -

- How much do you think people understand what it is like to have CF?
- How comfortable do you feel talking about your CF to -
 - Friends
 - Family
 - CF team?
- Do you think more should be done to raise awareness of the condition? If so, what?
- Do you use social media to talk to others about CF?

11-17yrs Interview Schedule Date: 03/02/2017 Version No: 1 IRAS ID: 219455

Future priorities

'What are your goals for the future?'

Prompts -

- In terms of work and life?
- Do you think you are ambitious? If so, how?

Becoming Independent

'How do you see your relationship with CF changing as you get older?'

Prompts -

- (If around 18yrs) If you have moved into adult services recently how did you find the transition?
- What do you think would make you feel more confident about managing your CF more by yourself?
- Do you see a big difference between child and adult care?

CF Trust Future Priorities

'What| do you think the CF Trust should focus their work on in the future?'

Prompts -

- Research? What would you research?
- Awareness? How would you raise awareness?
- Fund raising? How do you think you could raise more funds?
- Support services? Who for?

Closing

'Is there anything you would like to add?'

We've now come to the end of the interview. Thank you for taking the time to answer my questions. I have just a few more things to ask you:

- Would you like a summary of general findings at the end of the study?
- What would be the best way to send that to you?
- Are there any important questions that you think we missed out?
- Would you like to ask any questions or make any comments now that the interview is over?

If you think of any questions then please get in touch using the details on the consent form.

11-17yrs Interview Schedule

Date: 03/02/2017 Version No: 1 IRAS ID: 219455

Interview Schedule (18-25yrs)

Introduction

I am going to ask you a few questions about your experience with CF and how it can impact on various aspects on your life. The interview should take no longer than 45 minutes, but please tell me if you would like to take a break or stop altogether. If you don't want to answer any of the questions, let me know and we can go onto the next one. Are you happy to continue?

Questions (with prompts)

Impact on day-to-day life

'How has having CF impacted your everyday life?'

Prompts -

- Do you think it influences your friendships and relationships with others? If so, how?
- Is your school/work life effected? If so, how?
- Does it affect your social life?
- What challenges do you face?

'How does CF make you feel (about yourself)?'

Prompts -

- Does your CF make you feel isolated?
- Do you think it impacts your self-esteem? If so, how?
- Are you generally a happy person?

Awareness of CF

'How much do you tell other people about your CF?'

Prompts –

- How much do you think people understand what it is like to have CF?
- How comfortable do you feel talking about your CF to
 - Friends
 - Family
 - CF team?
- Do you think more should be done to raise awareness of the condition? If so, what?
- Do you use social media to talk to others about CF?

18-25yrs Interview Schedule

Date: 03/02/2017 Version No: 1 IRAS ID: 219455

Future priorities

'What are your goals for the future?'

Prompts -

- In terms of work and life?
- Do you think you are ambitious? If so, how?

Becoming Independent

'How do you see your relationship with CF changing as you get older?'

Prompts -

- (If around 18yrs) If you have moved into adult services recently how did you find the transition?
- (If 19+) How did you find the transition to adult care?
- What do you think would make you feel more confident about managing your CF independently?
- Do you see a big different between child and adult care?

CF Trust Future Priorities

'What do you think the CF Trust should focus their work on in the future?'

Prompts -

- Research? What would you research?
- Awareness? How would you raise awareness?
- Fund raising? How do you think you could raise more funds?
- Support services? Who for?

Closing

'Is there anything you would like to add?'

We've now come to the end of the interview. Thank you for taking the time to answer my questions. I have just a few more things to ask you:

- Would you like a summary of general findings at the end of the study?
- What would be the best way to send that to you?
- Are there any important questions that you think we missed out?
- Would you like to ask any questions or make any comments now that the interview is over?

If you think of any questions then please get in touch using the details on the consent form.

18-25yrs Interview Schedule Date: 03/02/2017 Version No: 1 IRAS ID: 219455

Interview Schedule for adults over 25

Intro:

I am going to ask you a few questions about your experience with CF. Our aim is to better understand how having CF can impact on your life as an adult. The interview should take no longer than 45 minutes, but please tell me if you would like to take a break or stop altogether. If you don't want to answer any of the questions, let me know and we can go onto the next one. Are you happy to continue?

Questions:

Impact of having CF as an adult

- How has CF impacted your life as an adult?
 - ☐ Has the impact differed from when you were a child?
 - What has been the biggest impact?
 - □ Do you think it influences your relationships with others? If so, how?
 - What are the biggest challenges that you face?
 - How have you overcome challenges in the past?
 - ☐ Has it impacted on you work life? If so, how?

If they have children:

- Did you have any concerns about having children of your own?
 - Were you able to discuss these concerns with anyone or get support from any services?
 - Did you seek any guidance from anyone?

Mental Health

- How does having CF make you feel in yourself?
 - ☐ Does your CF make you feel isolated?
 - ☐ Do you have any concerns with body-image?
 - □ Do you think it impacts on your self-esteem? If so, how?
 - ☐ Are you generally a happy person?

Awareness

- How much do you tell other people about your CF?
 - ☐ Has this changed as you got older? If, so how?
 - ☐ Do you think people understand what it is like to have CF?
 - \hookrightarrow Do you think more should be done to raise awareness of the condition? If so, what?
 - ☐ Have you ever used social media to talk about CF or to other with the condition?

Support

- Can you tell me about any support services you know of for adults with CF?
 - □ Prompts if know about existing services –
 - Do you currently/have you in the past used any of these?

25yrs + Interview Schedule Date: 03/02/2017 Version No: 1 IRAS ID: 219455

- Do you feel you benefit from these? How?
- How could these be improved?
- What other support would you like to be available?
- □ Prompts if don't know about existing services –
- Do you think support services would be of benefit? If so, how?
- What would you like these services to offer?

Future

- Looking forward, what support do you feel would be most valuable to you?
 - How would it be different to any help you have had before?
 - How important do you think support will be?
- What do you think the CF Trust should focus their work on in the future?
 - ☐ Research? What would you research?
 - ☐ Awareness? How would you raise awareness?
 - ☐ Fund raising? How do you think you could raise more funds?
 - Support services? Who for?

Closing

That is the end of my questions, is there anything you would like to add? Or do you think I missed out any important questions?

That is now the end of the interview so thank you for taking the time to answer my questions. Just before we go I have a couple of questions about this project,

- Would you like a summary of general findings at the end of the study?
- What would be the best way to send that to you?
- Would you like to ask any questions or make any comments now that the interview is over?

If you think of any questions then please get in touch using the details on the consent form.

25yrs + Interview Schedule Date: 03/02/2017 Version No: 1

IRAS ID: 219455

Interview Schedule (Parents)

Introduction

I am going to ask you a few questions about your and your family's experience with CF and how it can impact on various aspects on your life. The interview should take no longer than 45 minutes, but please tell me if you would like to take a break or stop altogether. If you don't want to answer any of the questions, let me know and we can go onto the next one. Are you happy to continue?

Questions (with prompts)

Impact on day-to-day life

'How has having a child/children with CF impacted your day-to-day life?'

Prompts -

- Can you tell me how your child's treatments fit into your day?
- Is your work life affected?
- Does it affect your social life?
- Do you think it affects your friendships/relationships with others?

Mental health/Chronic stress/Isolation (support from family/friends)

'Do you think that your child's Cystic Fibrosis has had an effect on your mental health – for example, feelings of anxiety or depression? If so, how?'

Prompts -

- Who do you lean on when things get difficult with [child]?
- Do you talk to others much about your child's CF?
- Do you believe your child's CF affects their mental health or change they feel about themselves?
- Does it impact on your health more generally?

Support (from charities/organisations)

'What is your view on the current support available for children with CF and their families?'

How could something like the CF Trust support you in dealing with the challenges of having a child with CF?

Prompts –

- Did you receive any information about CF and the support available when your child was first learned of your child's Cystic Fibrosis?
- Do you perceive an unmet need or do you think the current support options are adequate?

Parent Interview Schedule Date: 18/05/2017 Version No: 1 IDAS ID: 210455 - Looking forwards, what sort of support do you think you will require in the future, both in terms of yourself and your family?

Coping skills/Psychological Support

'Do you believe you have developed certain 'coping skills' to manage the demands of having a child with CF? If so, what are these?'

Prompts -

 Do you feel support in developing/understanding these 'coping skills' may be beneficial?

Impact on the relationship with children

'Do you feel your relationship with your child(ren) is affected by Cystic Fibrosis? If so, how?'

Prompts -

- Does it have an impact on the family dynamics?
- Would you say it has impacted your relationship with [partner] at all?
- Do you and [partner] have similar parenting styles? If not, how do these differ?

Advice

'What advice would you give to a parent who has just found out their child has CF in terms of how to deal with the challenges?'

Prompt -

- Is there anything you would like to have known when you first received the diagnosis?

Closing

'Is there anything I haven't asked you about in terms of what is like to be a parent of a child with CF that you think I should have in order to really understand the support that you would like from the CF Trust

We've now come to the end of the interview. Thank you for taking the time to answer my questions. I have just a few more things to ask you:

- Would you like a summary of general findings at the end of the study?
- What would be the best way to send that to you?
- Are there any important questions that you think we missed out?
- Would you like to ask any questions or make any comments now that the interview is over?

If you think of any questions then please get in touch using the details on the consent form.

Parent Interview Schedule Date: 18/05/2017 Version No: 1



London - Bromley Research Ethics Committee

Level 3, Block B

Whitefriars

Lewins Mead

Bristol

BS1 2NT

Please note: This is the favourable opinion of the REC only and does not allow you to start your study at NHS sites in England until you receive HRA Approval

20 April 2017

Professor Roz Shafran UCL Great Ormond Street Institute of Child Health 30 Guilford Street WC1N 1EH

Dear Professor Shafran

Study title: Experiences of Living with Cystic Fibrosis; The impact

on children, young people, adults and their families

REC reference: 17/LO/0348

Protocol number: 1 IRAS project ID: 219455

Thank you for your letter responding to the Committee's request for further information on the above research and submitting revised documentation.

The further information has been considered on behalf of the Committee by the Chair.

We plan to publish your research summary wording for the above study on the HRA website, together with your contact details. Publication will be no earlier than three months from the date of this opinion letter. Should you wish to provide a substitute contact point, require further information, or wish to make a request to postpone publication, please contact https://doi.org/10.1007/nns.net outlining the reasons for your request.

Confirmation of ethical opinion

On behalf of the Committee, I am pleased to confirm a favourable ethical opinion for the above research on the basis described in the application form, protocol and supporting documentation as revised, subject to the conditions specified below.



Conditions of the favourable opinion

The REC favourable opinion is subject to the following conditions being met prior to the start of the study.

Management permission must be obtained from each host organisation prior to the start of the study at the site concerned.

Management permission should be sought from all NHS organisations involved in the study in accordance with NHS research governance arrangements. Each NHS organisation must confirm through the signing of agreements and/or other documents that it has given permission for the research to proceed (except where explicitly specified otherwise).

Guidance on applying for NHS permission for research is available in the Integrated Research Application System, www.hra.nhs.uk or at http://www.rdforum.nhs.uk.

Where a NHS organisation's role in the study is limited to identifying and referring potential participants to research sites ("participant identification centre"), guidance should be sought from the R&D office on the information it requires to give permission for this activity.

For non-NHS sites, site management permission should be obtained in accordance with the procedures of the relevant host organisation.

Sponsors are not required to notify the Committee of management permissions from host organisations

Registration of Clinical Trials

All clinical trials (defined as the first four categories on the IRAS filter page) must be registered on a publically accessible database within 6 weeks of recruitment of the first participant (for medical device studies, within the timeline determined by the current registration and publication trees).

There is no requirement to separately notify the REC but you should do so at the earliest opportunity e.g. when submitting an amendment. We will audit the registration details as part of the annual progress reporting process.

To ensure transparency in research, we strongly recommend that all research is registered but for non-clinical trials this is not currently mandatory.

If a sponsor wishes to request a deferral for study registration within the required timeframe, they should contact https://doi.org/10.25/. The expectation is that all clinical trials will be registered, however, in exceptional circumstances non registration may be permissible with prior agreement from the HRA. Guidance on where to register is provided on the HRA website.

It is the responsibility of the sponsor to ensure that all the conditions are complied with before the start of the study or its initiation at a particular site (as applicable).



Summary CV for supervisor (student research) [Roz Shafran CV]	1	03 February 2017
Summary CV for supervisor (student research) [Fiona Mitchell Ulph CV]	1	03 February 2017
Validated questionnaire [CFQ-UK Children 11yrs]		
Validated questionnaire [CFQ-UK Children 12-13yrs]		03 February 2017
Validated questionnaire [CFQ-UK Teens and Adults (14+)]	1	03 February 2017
Validated questionnaire [WHOQOL-Bref (for siblings/parents 18yrs+)]	1	03 February 2017
Validated questionnaire [KIDSCREEN-10 (for siblings 11-17yrs)]	1	03 February 2017

Statement of compliance

The Committee is constituted in accordance with the Governance Arrangements for Research Ethics Committees and complies fully with the Standard Operating Procedures for Research Ethics Committees in the UK.

After ethical review

Reporting requirements

The attached document "After ethical review – guidance for researchers" gives detailed guidance on reporting requirements for studies with a favourable opinion, including:

- Notifying substantial amendments
- Adding new sites and investigators
- · Notification of serious breaches of the protocol
- · Progress and safety reports
- · Notifying the end of the study

The HRA website also provides guidance on these topics, which is updated in the light of changes in reporting requirements or procedures.

User Feedback

The Health Research Authority is continually striving to provide a high quality service to all applicants and sponsors. You are invited to give your view of the service you have received and the application procedure. If you wish to make your views known please use the feedback form available on the HRA website:

http://www.hra.nhs.uk/about-the-hra/governance/quality-assurance/

HRA Training

We are pleased to welcome researchers and R&D staff at our training days – see details at http://www.hra.nhs.uk/hra-training/



With the Committee's best wishes for the success of this project.

Yours sincerely

p.

pp Ms Jayne Steadman

Chair

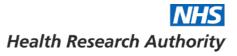
Email:nrescommittee.london-bromley@nhs.net

Enclosures: "After ethical review – guidance for

researchers"

Copy to: Ms Emma Pendleton

Dr Thomas Lewis, Great Ormond Street Hospital



Professor Roz Shafran
UCL Great Ormond Street Institute of Child Health
30 Guilford Street
WC1N 1EH

Email: hra.approval@nhs.net

24 April 2017 Reissued 04 May 2017 to correct document dates

Dear Professor Shafran

Letter of HRA Approval

Study title: Experiences of Living with Cystic Fibrosis; The impact on

children, young people, adults and their families

IRAS project ID: 219455

Protocol number: 1

REC reference: 17/LO/0348

Sponsor UCL Great Ormond Street Institute of Child Health

I am pleased to confirm that <u>HRA Approval</u> has been given for the above referenced study, on the basis described in the application form, protocol, supporting documentation and any clarifications noted in this letter.

Participation of NHS Organisations in England

The sponsor should now provide a copy of this letter to all participating NHS organisations in England.

Appendix B provides important information for sponsors and participating NHS organisations in England for arranging and confirming capacity and capability. Please read Appendix B carefully, in particular the following sections:

- Participating NHS organisations in England this clarifies the types of participating
 organisations in the study and whether or not all organisations will be undertaking the same
 activities
- Confirmation of capacity and capability this confirms whether or not each type of participating
 NHS organisation in England is expected to give formal confirmation of capacity and capability.
 Where formal confirmation is not expected, the section also provides details on the time limit
 given to participating organisations to opt out of the study, or request additional time, before
 their participation is assumed.
- Allocation of responsibilities and rights are agreed and documented (4.1 of HRA assessment criteria) - this provides detail on the form of agreement to be used in the study to confirm capacity and capability, where applicable.

Further information on funding, HR processes, and compliance with HRA criteria and standards is also provided.

It is critical that you involve both the research management function (e.g. R&D office) supporting each organisation and the local research team (where there is one) in setting up your study. Contact details

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IRAS project ID	219455

and further information about working with the research management function for each organisation can be accessed from www.hra.nhs.uk/hra-approval.

Appendices

The HRA Approval letter contains the following appendices:

- · A List of documents reviewed during HRA assessment
- . B Summary of HRA assessment

After HRA Approval

The document "After Ethical Review – guidance for sponsors and investigators", issued with your REC favourable opinion, gives detailed guidance on reporting expectations for studies, including:

- Registration of research
- Notifying amendments
- Notifying the end of the study

The HRA website also provides guidance on these topics, and is updated in the light of changes in reporting expectations or procedures.

In addition to the guidance in the above, please note the following:

- HRA Approval applies for the duration of your REC favourable opinion, unless otherwise notified in writing by the HRA.
- Substantial amendments should be submitted directly to the Research Ethics Committee, as
 detailed in the After Ethical Review document. Non-substantial amendments should be
 submitted for review by the HRA using the form provided on the <u>HRA website</u>, and emailed to
 hra.amendments@nhs.net.
- The HRA will categorise amendments (substantial and non-substantial) and issue confirmation
 of continued HRA Approval. Further details can be found on the HRA website.

Scope

HRA Approval provides an approval for research involving patients or staff in NHS organisations in England.

If your study involves NHS organisations in other countries in the UK, please contact the relevant national coordinating functions for support and advice. Further information can be found at http://www.hra.nhs.uk/resources/applying-for-reviews/nhs-hsc-rd-review/.

If there are participating non-NHS organisations, local agreement should be obtained in accordance with the procedures of the local participating non-NHS organisation.

User Feedback

The Health Research Authority is continually striving to provide a high quality service to all applicants and sponsors. You are invited to give your view of the service you have received and the application procedure. If you wish to make your views known please use the feedback form available on the HRA website: http://www.hra.nhs.uk/about-the-hra/governance/quality-assurance/.

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IRAS project ID	219455
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HRA Training

We are pleased to welcome researchers and research management staff at our training days – see details at http://www.hra.nhs.uk/hra-training/

Your IRAS project ID is 219455. Please quote this on all correspondence.

Yours sincerely

Kevin Ahmed Assessor

Telephone: 0207 104 8171 Email: hra.approval@nhs.net

Copy to: Ms Emma Pendleton, Sponsor Contact, Great Ormond Street Hospital

Dr Thomas Lewis, R&D Contact, Great Ormond Street Hospital
Miss Mhairi McKenzie, Student, Great Ormond Street Hospital
Miss Corah Lewis, Student, Great Ormond Street Hospital

IRAS project ID	210455
INAS project ib	219400

Appendix A - List of Documents

The final document set assessed and approved by HRA Approval is listed below.

Document	Version	Date
Covering letter on headed paper [Cover Letter]	1	09 March 2017
Covering letter on headed paper [Cover Letter]		24 March 2017
Evidence of Sponsor insurance or indemnity (non NHS Sponsors only) [UCL Insurance]	1	03 February 2017
Interview schedules or topic guides for participants [Interview Schedule for YP with CF 11-17yrs (subject to change)]	1	03 February 2017
Interview schedules or topic guides for participants [Interview Schedule for individuals with CF 18-25yrs (subject to change)]	1	03 February 2017
Interview schedules or topic guides for participants [Interview Schedule for individuals with CF 26yrs+ (subject to change)]	1	03 February 2017
Interview schedules or topic guides for participants [Interview Schedule for Healthy Siblings (subject to change)]	1	03 February 2017
IRAS Application Form [IRAS_Form_10032017]		10 March 2017
IRAS Application Form XML file [IRAS_Form_10032017]		10 March 2017
IRAS Checklist XML [Checklist_24032017]		24 March 2017
Letter from funder [CF Trust Funding Confirmation]	1	03 February 2017
Non-validated questionnaire [Self-completion questionnaire for Parents of YP/Children with CF]	2	23 March 2017
Participant consent form [Assent Form for Children <16yrs]	1	03 February 2017
Participant consent form [Consent form for individuals 16+]	1	03 February 2017
Participant consent form [Consent form for Parents (of child <16yrs)]	1	03 February 2017
Participant information sheet (PIS) [PIS for Healthy Siblings 11- 16yrs]		23 March 2017
Participant information sheet (PIS) [PIS for Healthy Siblings 17+]	2	23 March 2017
Participant information sheet (PIS) [PIS for YP (11-17yrs) with CF]		23 March 2017
Participant information sheet (PIS) [PIS for individuals 18yrs+ with CF]		23 March 2017
Participant information sheet (PIS) [PIS for Parents]	2	23 March 2017
Research protocol or project proposal ["Experiences of Living with Cystic Fibrosis": Protocol]	2	23 March 2017
Summary CV for Chief Investigator (CI) [Roz Shafran CV]	1	03 February 2017
Summary CV for student [Mhairi McKenzie CV]	1	03 February 2017
Summary CV for student [Corah Lewis CV]	1	03 February 2017
Summary CV for supervisor (student research) [Roz Shafran CV]	1	03 February 2017
Summary CV for supervisor (student research) [Fiona Mitchell Ulph CV]	1	03 February 2017
Validated questionnaire [CFQ-UK Children 11yrs]		
Validated questionnaire [CFQ-UK Children 12-13yrs]		03 February 2017
Validated questionnaire [CFQ-UK Teens and Adults (14+)]	1	03 February 2017
Validated questionnaire [WHOQOL-Bref (for siblings/parents 18yrs+)]	1	03 February 2017
Validated questionnaire [KIDSCREEN-10 (for siblings 11-17yrs)]	1	03 February 2017
Other [Statement of Activities]	1	24 April 2017

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IRAS project ID	219455

Other [Schedule of Events]	1	24 April 2017
Copies of advertisement materials to research participants [Poster]	2	09 March 2017
17 LO 0348- Further Information Favourable Opinion		20 April 2017

Appendix B - Summary of HRA Assessment

This appendix provides assurance to you, the sponsor and the NHS in England that the study, as reviewed for HRA Approval, is compliant with relevant standards. It also provides information and clarification, where appropriate, to participating NHS organisations in England to assist in assessing and arranging capacity and capability.

For information on how the sponsor should be working with participating NHS organisations in England, please refer to the, participating NHS organisations, capacity and capability and Allocation of responsibilities and rights are agreed and documented (4.1 of HRA assessment criteria) sections in this appendix.

The following person is the sponsor contact for the purpose of addressing participating organisation questions relating to the study:

Name: Ms Emma Pendleton

Tel: 02079052698

Email: research.governance@gosh.nhs.uk

HRA assessment criteria

Section	HRA Assessment Criteria	Compliant with Standards	Comments
1.1	IRAS application completed correctly	Yes	No comments
2.1	Participant information/consent documents and consent process	Yes	No comments
3.1	Protocol assessment	Yes	No comments
4.1	Allocation of responsibilities and rights are agreed and documented	Yes	The sponsor has submitted the HRA Statement of Activities and intends for this to form the agreement between the sponsor and study sites.
			The sponsor is not requesting, and does not require any additional

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IRAS project ID	219455
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Section	HRA Assessment Criteria	Compliant with Standards	Comments
			contracts with study sites.
4.2	Insurance/indemnity arrangements assessed	Yes	Where applicable, independent contractors (e.g. General Practitioners) should ensure that the professional indemnity provided by their medical defence organisation covers the activities expected of them for this research study
4.3	Financial arrangements assessed	Yes	External study funding has been secured from The Cystic Fibrosis Trust. No study funding will be provided to sites, as detailed at Schedule 1 of the Statement of Activities.
5.1	Compliance with the Data	Yes	No comments
5.1	Protection Act and data security issues assessed	Tes	No comments
5.2	CTIMPS – Arrangements for compliance with the Clinical Trials Regulations assessed	Not Applicable	No comments
5.3	Compliance with any applicable laws or regulations	Yes	No comments
6.1	NHS Research Ethics Committee favourable opinion received for applicable studies	Yes	No comments
6.2	CTIMPS – Clinical Trials Authorisation (CTA) letter received	Not Applicable	No comments
6.3	Devices – MHRA notice of no objection received	Not Applicable	No comments
6.4	Other regulatory approvals and authorisations received	Not Applicable	No comments

Participating NHS Organisations in England

This provides detail on the types of participating NHS organisations in the study and a statement as to whether the activities at all organisations are the same or different.

Page 6 of 8

IRAS project ID	219455
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All participating NHS organisations will undertake the same study activities. There is therefore only one study site 'type' involved in the research.

The Chief Investigator or sponsor should share relevant study documents with participating NHS organisations in England in order to put arrangements in place to deliver the study. The documents should be sent to both the local study team, where applicable, and the office providing the research management function at the participating organisation. For NIHR CRN Portfolio studies, the Local LCRN contact should also be copied into this correspondence. For further guidance on working with participating NHS organisations please see the HRA website.

If chief investigators, sponsors or principal investigators are asked to complete site level forms for participating NHS organisations in England which are not provided in IRAS or on the HRA website, the chief investigator, sponsor or principal investigator should notify the HRA immediately at hra.approval@nhs.net. The HRA will work with these organisations to achieve a consistent approach to information provision.

Confirmation of Capacity and Capability

This describes whether formal confirmation of capacity and capability is expected from participating NHS organisations in England.

NHS organisations in England that are participating in the study will be expected to formally confirm their capacity and capability to host this research.

- Following issue of this letter, participating NHS organisations in England may now confirm to
 the sponsor their capacity and capability to host this research, when ready to do so. How
 capacity and capacity will be confirmed is detailed in the Allocation of responsibilities and
 rights are agreed and documented (4.1 of HRA assessment criteria) section of this appendix.
- The Assessing, Arranging, and Confirming document on the HRA website provides further information for the sponsor and NHS organisations on assessing, arranging and confirming capacity and capability.

Principal Investigator Suitability

This confirms whether the sponsor position on whether a PI, LC or neither should be in place is correct for each type of participating NHS organisation in England and the minimum expectations for education, training and experience that PIs should meet (where applicable).

The sponsor has correctly assessed that a Principal Investigator should be appointed at study sites.

GCP training is <u>not</u> a generic training expectation, in line with the <u>HRA statement on training</u> expectations.

HR Good Practice Resource Pack Expectations

This confirms the HR Good Practice Resource Pack expectations for the study and the pre-engagement checks that should and should not be undertaken

Where arrangements are not already in place, network staff (or similar) undertaking any of the research activities listed in A18 or A19 of the IRAS form (except for administration of questionnaires

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1		

or surveys), would be expected to obtain an honorary research contract from one NHS organisation (if university employed), followed by Letters of Access for subsequent organisations. This would be on the basis of a Research Passport (if university employed) or an NHS to NHS confirmation of preengagement checks letter (if NHS employed). These should confirm enhanced DBS checks, including appropriate barred list checks, and occupational health clearance. For research team members only administering questionnaires or surveys, a Letter of Access based on standard DBS checks and occupational health clearance would be appropriate.

Other Information to Aid Study Set-up

This details any other information that may be helpful to sponsors and participating NHS organisations in England to aid study set-up.

The applicant has indicated that they do not intend to apply for inclusion on the NIHR CRN Portfolio.



London - Bromley Research Ethics Committee

Level 3, Block B Whitefriars Lewins Mead Bristol BS1 2NT

Please note: This is the favourable opinion of the REC only and does not allow the amendment to be implemented at NHS sites in England until the outcome of the HRA assessment has been confirmed.

08 June 2017

Professor Roz Shafran UCL Great Ormond Street Institute of Child Health 30 Guilford Street WC1N 1EH

Dear Professor Shafran

Study title: Experiences of Living with Cystic Fibrosis; The impact on

children, young people, adults and their families

REC reference: 17/LO/0348

Protocol number: 1

Amendment number: No. 1 Amendment date: No. 1 18 May 2017

IRAS project ID: 219455

The above amendment was reviewed by the Sub-Committee in correspondence.

Ethical opinion

The members of the Committee taking part in the review gave a favourable ethical opinion of the amendment on the basis described in the notice of amendment form and supporting documentation.

Approved documents

The documents reviewed and approved at the meeting were:

Document	Version	Date
Interview schedules or topic guides for participants [Parent Interview Schedule.docx]	1	18 May 2017
Notice of Substantial Amendment (non-CTIMP) [AmendmentForm_ReadyForSubmission.pdf]	No. 1	18 May 2017
Participant consent form [CF Parental consent form for child under	2	18 May 2017



16 - track changed.docx]		
Participant consent form [CF Parent consent form v2 - track changed.docx]		18 May 2017
Participant consent form [CF Parent consent form v2- clean.docx]	2	18 May 2017
Participant consent form [CF Parental consent form for child under 16 - clean.docx]	2	18 May 2017
Participant information sheet (PIS) [PIS Parent v3 - track changed.docx]	3	18 May 2017
Participant information sheet (PIS) [PIS Parent v3- clean.docx]	3	18 May 2017
Research protocol or project proposal [CF Protocol v3 - clean.docx]	3	18 May 2017
Research protocol or project proposal [CF Protocol v3 - tracked.docx]	3	18 May 2017

Membership of the Committee

The members of the Committee who took part in the review are listed on the attached sheet.

Working with NHS Care Organisations

Sponsors should ensure that they notify the R&D office for the relevant NHS care organisation of this amendment in line with the terms detailed in the categorisation email issued by the lead nation for the study.

Statement of compliance

The Committee is constituted in accordance with the Governance Arrangements for Research Ethics Committees and complies fully with the Standard Operating Procedures for Research Ethics Committees in the UK.

We are pleased to welcome researchers and R & D staff at our Research Ethics Committee members' training days – see details at http://www.hra.nhs.uk/hra-training/

17/LO/0348: Please quote this number on all correspondence

Yours sincerely



Ms Jayne Steadman Chair

E-mail: nrescommittee.london-bromley@nhs.net

Enclosures: List of names and professions of members who took part in the

review

Copy to: Dr Thomas Lewis, Great Ormond Street Hospital

Ms Emma Pendleton



London - Bromley Research Ethics Committee

Attendance at Sub-Committee of the REC meeting on 02 June 2017

Committee Members:

Name	Profession	Present	Notes
Dr Angela Orunta	Consultant Anaesthetist	Yes	
Ms Jayne Steadman (Chair and Meeting Chair)	Consultant Physiotherapist	Yes	

Also in attendance:

Name	Position (or reason for attending)
Ms Helen Sivey	REC Manager

From: AMENDMENTS, Hra (HEALTH RESEARCH AUTHORITY) [mailto:hra.amendments@nhs.net]

Sent: 08 June 2017 15:34

To: Shafran, Roz < r.shafran@ucl.ac.uk >; research.governance@gosh.nhs.uk;

Cc: Thomas.Lewis@gosh.nhs.uk

Subject: Amendment 17/LO/0348/AM01, IRAS Project ID: 219455 - Confirmation of HRA Approval

Dear Professor Shafran,

Further to the below, I am pleased to confirm that HRA Approval has been issued for the referenced amendment, following assessment against the HRA criteria and standards.

The sponsor should now work collaboratively with participating NHS organisations in England to implement the amendment as per the below categorisation information. This email may be provided by the sponsor to participating organisations in England to evidence that the amendment has HRA Approval.

Please contact hra.amendments@nhs.net for any queries relating to the assessment of this amendment.

Kind regards

Ali Hussain Amendments Co-ordinator



Health Research Authority
HRA, Ground Floor, Skipton House, 80 London Road, London, SE1 6LH
E: hra.amendments@nhs.net: T: 020 7972 2649
www.hra.nhs.uk

The HRA is keen to know your views on the service you received – our short feedback form is available <u>here</u>

Dear Dr Bryon,

Project Title	A study to understand needs, challenges and aspirations of people with cystic fibrosis and their siblings in the UK	
IRAS Number:	219455	
REC Reference	17/LO/0345	
R&D Reference	16PP30	
Date of REC Favourable Opinion	20th April 2017	
Protocol version	V1 5 th May 2016	
HRA Approval Notification Date	24th April 2017 reissued 4th May 2017	
Statement of Activities/Issues	Any external researchers need access arrangements in place to participate in the research at GOSH. Please contact Elliot.dickens@gosh.nhs.uk for further advice if necessary.	
	As this is single site/sponsor study no statement of activities or schedule of events documents are required.	

This research project has been reviewed locally at Great Ormond Street Hospital NHS Foundation Trust for capacity and capability. After review we can confirm a **Notification of No Objection** to this research being conducted at GOSH.

Please contact us using the contact details below if you require any further information.

We wish you all the best with your research.

Kind regards

Dr Thomas Lewis



Great Ormond Street
Hospital for Children
Mrs Foundation Trust

Joint Research and Development Office Division of Research and Innovation Dr Thomas Lewis | Research Management and Governance Officer Division of Research & Innovation | Joint R&D Office for GOSH/ICH I UCL Institute of Child Health | 30 Guilford Street | London, WCIN IEH

www.gosh.nhs.uk/research-and-innovation +44 (0) 207 905 2249 | thomas.lewis@gosh.nhs.uk

Monday, September 23, 2019 at 8:49:46 PM British Summer Time

Subject: RE: Qualitative research - young people / older adults

Date: Friday, 21 July 2017 at 14:26:12 British Summer Time

From: T.Caumont@rbht.nhs.uk

To: N.Simmonds@rbht.nhs.uk

CC: McKenzie, Mhairi, Lewis, Corah

Dear All,

We confirm that the Royal Brompton and Harefield NHS Foundation Trust is now an active Participant Identification Centre for the named research project.

Kind regards,

Tom Caumont

Non-Commercial Research Coordinator Royal Brompton & Harefield NHS Foundation Trust Research Office, Chelsea Wing Sydney Street SW3 6NP

Tel: 0207 351 8574 Fax: 0207 351 8578

Int: http://www2.rbht.nhs.uk/services/research/ Ext: http://www.rbht.nhs.uk/research/

Appendix 3.7. Confirmation of Capacity and Capability (cont.)

Appendix 3.7.3. Cardiff & Vale University Health Board



Ysbyty Athrofaol Cymru University Hospital of Wales

Heath Park, Cardiff, CF14 4XW Phone 029 2074 7747 Fax 029 2074 3838 Minicom 029 2074 3632 Parc Y Mynydd Bychan, Caerdydd, CF14 4XW Ffôn 029 2074 7747 Ffacs 029 20743838 Minicom 029 2074 3632

Tel: 029 20746986

CAV research.development@wales.nhs.uk

Professor C Fegan R&D Director R&D Office, 2nd Floor, TB2 University Hospital of Wales Cardiff CF14 4XW

14 September 2017

Professor Roz Shafran
Professor of Translational Psychology and Honorary Consultant Clinical Psychologist
UCL Great Ormond Street Institute of Child Health
30 Guilford Street
London
WC1N 1EH

Dear Professor Shafran,

Study title	Experiences of Living with Cystic Fibrosis; The impact on children, young people, adults and their families
Cardiff and Vale UHB reference	17/JUL/7005
IRAS reference	219455

Thank you for your recent communication informing us about the above research project.

Where the only involvement of an NHS organisation in a project is as a Participant Identification Centre, NHS R&D approval is not required. I am pleased to confirm that Cardiff and Vale University Local Health Board has no objections to acting as a Participant Identification Centre for this project subject to the following:

 Before asking the CF clinic to undertake PIC activities, the clinic manager must first have agreed to participate.

Please ensure that you supply the appropriate research site/s with a copy of this letter.

May I take this opportunity to wish you success with the project and remind you that this letter relates only to Cardiff and Vale UHB undertaking the role of Participant

Bwidd feefyd Prifysgol Caerdydd a'r Fro yw enw gweithredol Bwyrdd loehyd Lleol Prifysgol Caerdydd a'r Fro Cardrff and Vale University Health Board is the operational name of Cardrff and Vale University Local Health Boar





Identification Centre for this study. You must inform the Health and Care Research Wales Permissions Service and the UHB R&D Office if:

- You wish to seek approval for Cardiff and Vale UHB to undertake any participantrelated research procedures specified in the study protocol (including taking informed consent for participation) and therefore become a research site for this study
- You make any amendments relating to the protocol, including personnel changes and amendments to the actual or anticipated start / end dates
- You would like Cardiff and Vale UHB to refer potential participants to any research sites in this study other than those listed above.
- You would like Cardiff and Vale UHB to act as a Participant Identification Centre for any research other than the above-named project.

Procedures for participant identification must comply with the Data Protection Act 1998. For further advice and information please contact the UHB Data Protection Manager, Ann Morgan (Ann.Morgan4@Wales.nhs.uk).

Yours sincerely,

Professor Christopher Fegan
R&D Director / Chair of the Cardiff and Vale Research Review Service (CaRRS)

CC R&D Lead, Jamie Duckers
Finance, Anthony Williams
Clinical Board Assistant Head of Finance, Medicine
Sponsor contact, Emma Pendleton
Academic supervisor Dr Fiona Ulph

Monday, September 23, 2019 at 9:02:59 PM British Summer Time

Subject: IRAS 219455 STUDY Confirmation of Capacity and Capability at Manchester University NHS

Foundation Trust

Date: Thursday, 12 October 2017 at 13:28:45 British Summer Time
From: Fridrihsone Zane (ROA) Manchester University NHS FT

To: McKenzie, Mhairi

CC: 'alexander.horsley@manchester.ac.uk', Aziz Azad (ROA) Manchester University NHS FT,

Macnaughton Cassandra (ROA) Manchester University NHS FT

Attachments: image001.png, hra-statement-activities-CF-Trust Manchester CF 12-Oct-17.pdf

Dear Mhairi,

RE: IRAS 219455 Confirmation of Capacity and Capability at Manchester University NHS Foundation Trust

Full Study Title: Experiences of Living with Cystic Fibrosis; The impact on children, young people, adults and their families

This email confirms that Manchester University NHS Foundation Trust has agreed to act as a PIC for the above referenced study. Please find attached our agreed Statement of Activities as confirmation.

We can start identifying patients when the Sponsor gives green light to begin.

If you wish to discuss further, please do not hesitate to contact me.

Kind regards

Zane Fridrihsone

Senior Clinical Trials Administrator

NIHR Manchester Clinical Research Facility
North West Lung Research Centre (Charles Blackley Ward)



Manchester University

NHS Foundation Trust

Southmoor Road, Manchester, M23 9LT

T: 0161 291 4168 F: 0161 291 2941

E: zane.fridrihsone@mft.nhs.uk

www.mft.nhs.uk

On 1 October 2017 Central Manchester University Hospitals NHS Foundation Trust and University Hospital of South Manchester NHS Foundation Trust joined together as a single organisation, called Manchester University NHS Foundation Trust (MFT). Please note that there are currently no changes to research processes and contacts at the different hospital sites.

Monday, September 23, 2019 at 9:01:29 PM British Summer Time

Subject: IRAS 219455 (MFT PIN R04762) Confirmation of Capacity and Capability at MFT

Date: Monday, 6 November 2017 at 11:03:01 Greenwich Mean Time

From: Fox Hannah (ROA) Manchester University NHS FT

To: 'research.governance@gosh.nhs.uk', Robinson Alison (ROA) Manchester University NHS FT
CC: McKenzie, Mhairi, Browne Claire (ROA) Manchester University NHS FT, Maitra Anirban

(ROA) Manchester University NHS FT

Attachments: image001.jpg, 219455 Statement of Activities v1 24042017 MFT ready for CCC.pdf

IRAS: 219455 MFT PIN: R04762

Full Study Title: Experiences of Living with Cystic Fibrosis; The impact on children, young people, adults and their families

Dear Ms Pendleton.

This email confirms that Manchester University NHS Foundation Trust has the Capacity and Capability to act as a Patient Identification Centre for the above referenced study. Please find attached our agreed Statement of Activities as confirmation.

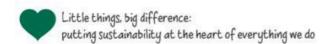
If you wish to discuss further, please do not hesitate to contact me.

Kind regards Hannah

Mrs Hannah Mckeown (nee Fox) Research Support Officer

Research Office
Manchester University NHS Foundation Trust
1st Floor NOWGEN Centre
29 Grafton Street
Manchester M13 9WU

Telephone 0161 276 8016 Hannah.Fox@mft.nhs.uk



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Administrator: Mrs Elaine O'Neill Telephone Number: 0141 232 1815 E-Mail: elaine.o'neill2@ggc.scot.nhs.uk Website: www.nhsggc.org.uk/r&d R&D Management Office West Glasgow ACH Dalnair Street Glasgow, G3 8SJ

19 October 2017

Prof Roz Shafran UCL, Great Ormond Street Ins of Child Health 30 Guilford Street London WC1N 1EH

NHS GG&C Board Approval Participant Identification Centre

Dear Prof R Shafran,

Study Title: Experiences of Living with Cystic Fibrosis; The impact on children, young

people, adults and their families

GG&C HB site Queen Elizabeth University Hospital

Sponsor UCL Institute of Child Health

R&D reference: GN17RM561 REC reference: 17/LO/0348 Protocol no: V3; 18/05/17

I am pleased to confirm that **Approval** has been granted for NHS Greater Glasgow & Clyde to act a **Patient Identification Centre (PIC)** for the above study.

Conditions of Approval

- 1. During the life span of the study, GG&C R&D Management Office requires the following information:
 - Any amendments Substantial or Non Substantial that may change the workload of GG&C as a PIC.
 - ii. Notification of trial/study end.

Please add this approval to your study file as it may be subject to audit and monitoring.

Your personal information will be held on a secure national web-based NHS database.

I wish you every success with this research study

Page 1 of 2

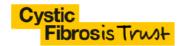
PIC Approval_GN17RM561

Yours sincerely,



Mrs Elaine O'Neill Senior Research Administrator

Cc: Emma Pendleton (UCL Great Ormond Street Ins)



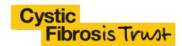




	CONSEN	IT FORM		
Project title: Experience adults and their familie		Fibrosis; The impact on children, young	people,	
Name of Chef Investigat	tor: Professor Roz Shafran			
		Please in	nitial all bo	
	had the opportunity to con	nformation sheet dated XXX vX for the sider the information, ask questions and		
•		and that I am free to withdraw at any time care or legal rights being affected.		
 In the event that I decide to stop taking part in the study, I agree that the information I have previously provided being used in the analysis of the study results. 				
I understand that the information I provide may be used for future academic projects and that no names or identifiable information will be used.				
 I agree to allow personal data about myself to be entered on a confidential computer database held at the Institute of Child Health, University College London. 				
6. I agree to the intervi	ew being audio recorded.			
7. I agree to take part in the above study.				
Name of participant	Date	Signature		
Name of person Date taking consent		Signature		

Young Person (16+) Consent Form Date: 03/02/2017 Version No: 1 IRAS ID: 219455

Page 1 of 1







Dage 1 of 1

Participant Identification Number for this project: CONSENT FORM Project title: Experiences of Living with Cystic Fibrosis; The impact on children, young people, adults and their families. Name of Researcher: Professor Roz Shafran Please initial all boxes 1. I confirm that I have read and understand the information sheet dated XXX version X for the above study. My child and I have had the opportunity to consider the information, ask questions and have had these answered satisfactorily. 2. I understand that my child's participation is voluntary and that they are free to withdraw at any time without giving any reason, without my child's medical care or legal rights being affected. 3. In the event that my child, or myself on behalf of my child, decide to stop taking part in the study, I agree that the information theyhave previously provided being used in the analysis of the study results. 4. I understand that the information my child provides may be used for academic projects and that no names or identifiable information will be used. 5. I agree to the interview with my child being audio recorded. 6. I agree to my child taking part in the above study. Name of participant Signature of parent Name of person Date Signature An original copy of the participant information sheet and completed informed consent form is to be given to the participant, in addition to the original copy that is filed in the investigator file. Parent/carer consent form for child under 16 years Date: 18/05/17 Version number: 2

IDAC ID: 2104EE







		CONSENT FO	RM	
	oject title: Experiences ults and their families.	of Living with Cystic Fibros	is; The impact on children, young	people,
Na	me of Researcher: Profes	ssor Roz Shafran		
			Please in	nitial all bo
1.	3 for the above study. I	d and understand the informa have had the opportunity to c these answered satisfactorily	•	
2.	, ,	ticipation is voluntary and than, without my medical care or	t I am free to withdraw at any time legal rights being affected.	
3.	In the event that I decide to stop taking part in the study, I agree that the information I have previously provided may still be used			
 I understand that the information I provide may be used for academic projects and that no names or identifiable information will be used. 				
I agree to the interview being audio recorded.				
6.	I agree to taking part in	the above study.		
Nar	me of participant	Date	Signature	
	me of person	 Date	 Signature	

An original copy of the participant information sheet and completed informed consent form is to be given to the participant, in addition to the original copy that is filed in the investigator file.

Parent/carer consent form Date: 18/05/17: Version number: 2 IRAS ID: 219455

Page 1 of 1







Assent Form

Assent Form			
Project title: Experiences of Living with Cystic Fibrosis; The impact on children, young people, adults and their families.			
Please circle all you agree with:			
Has somebody explained this project to you?	Yes/No		
Do you understand what this project is about?	Yes/No		
Have you asked all the questions you want to?	Yes/No		
Has someone answered these questions and do you understand the answers?	Yes/No		
Do you understand it's OK to stop taking part at any time?	Yes/No		
Are you happy to take part?	Yes/No		
If your answers are "yes" and you do want to take part, please write you Your name	ir name below.		
Date			
The person who explained this project to you needs to sign too below: Print Name (Person taking consent)			
,			
Sign			
Date			

An original copy of the participant information sheet and completed informed consent form is to be given to the participant, in addition to the original copy that is filed in the investigator file.

Child Assent Form (under 16) Date: 03/02/2017 Version No: 1 IRAS ID: 219455

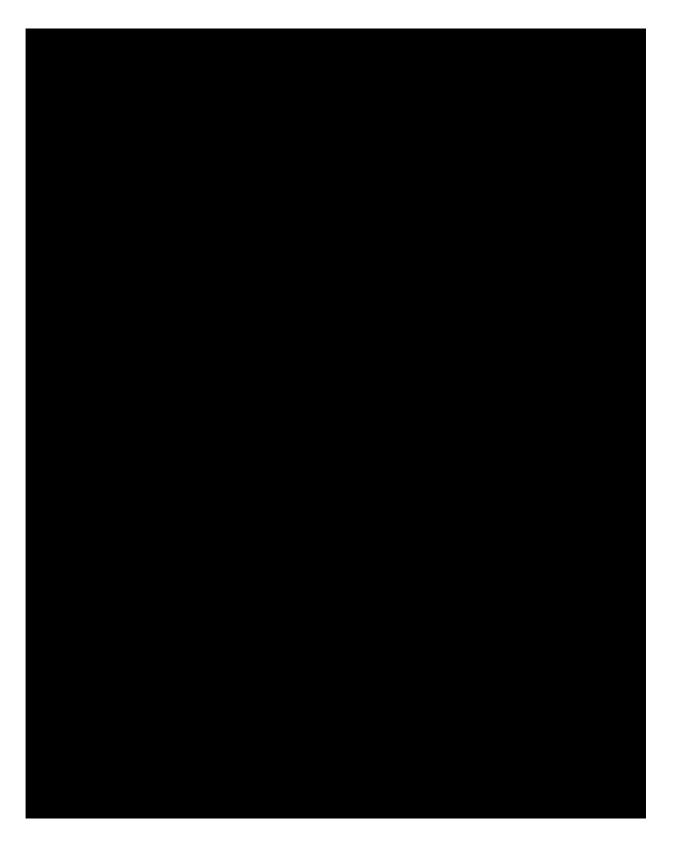
Page 1 of 1



KIDSCREEN-10 Index Health Questionnaire for Children and Young People

Child and Adolescent Version 8 to 18 Years English (UK)





© The KIDSCREEN Group, 2004; EC Grant Number: QLG-CT-2000- 00751 KIDSCREEN-10 Index, Child and Adolescent Version Page 2 of 2

THE WORLD HEALTH ORGANIZATION QUALITY OF LIFE (WHOQOL) -BREF

The World Health Organization Quality of Life (WHOQOL)-BREF

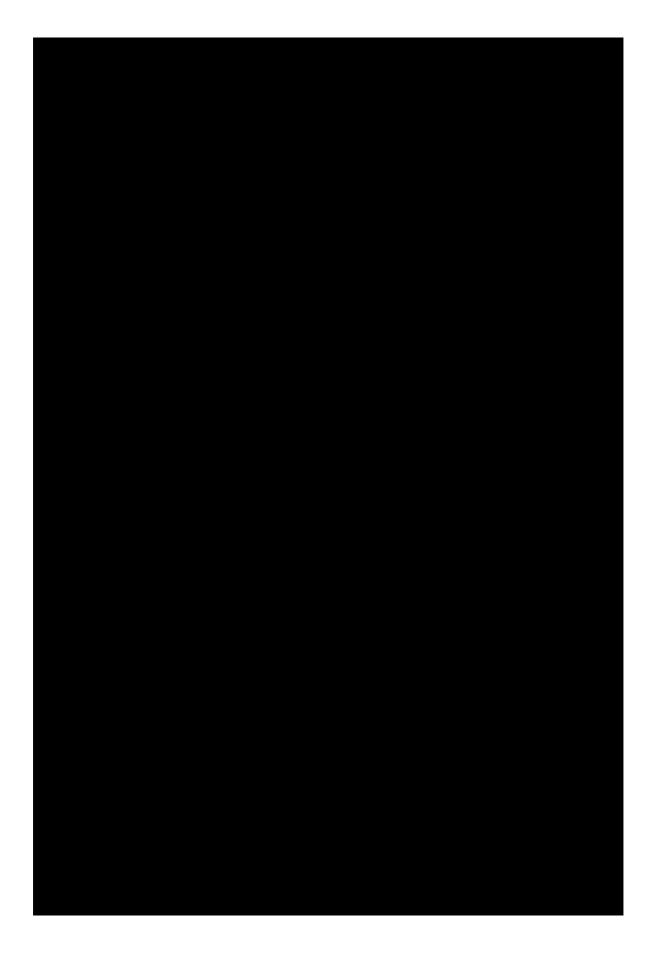
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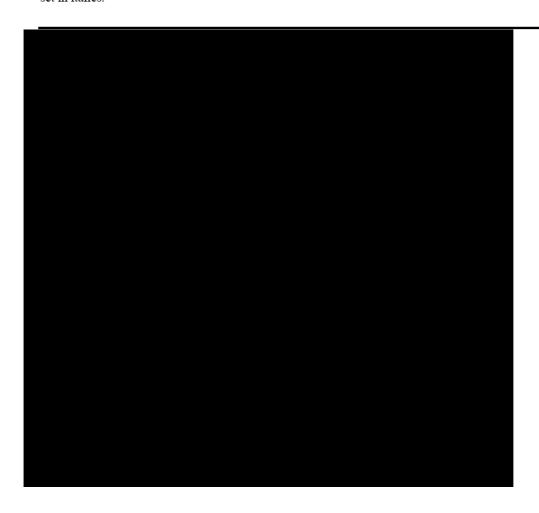




Children Ages 6 to 11

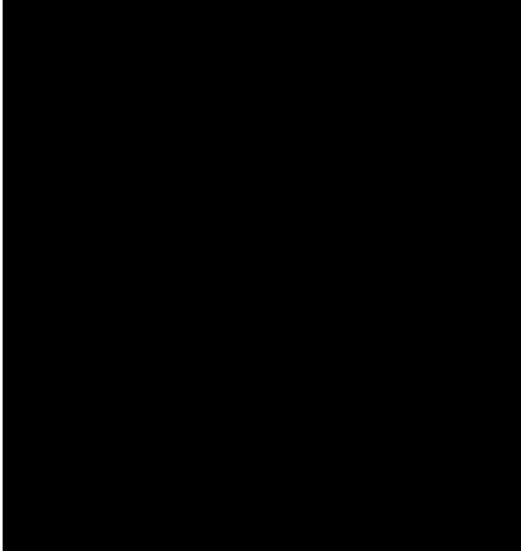
This questionnaire is formatted for use by an interviewer. Please use this format for younger children. For older children who seem able to read and answer the questions on their own, such as 12 and 13 year olds, use this questionnaire in its self-report format.

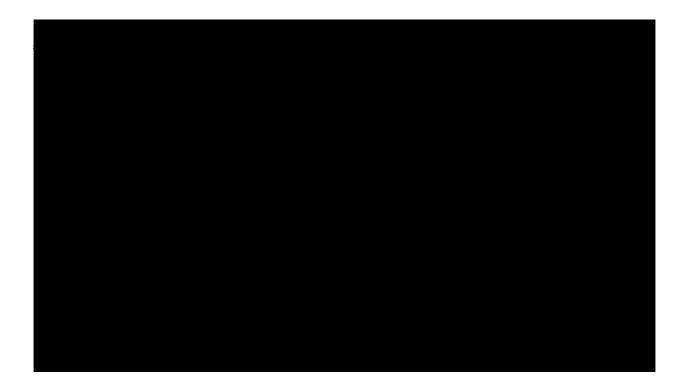
There are directions for the interviewer for each section of the questionnaire. Directions that you should *read* to the child are indicated by quotation marks. Directions that you are to *follow* are underlined and set in italics.

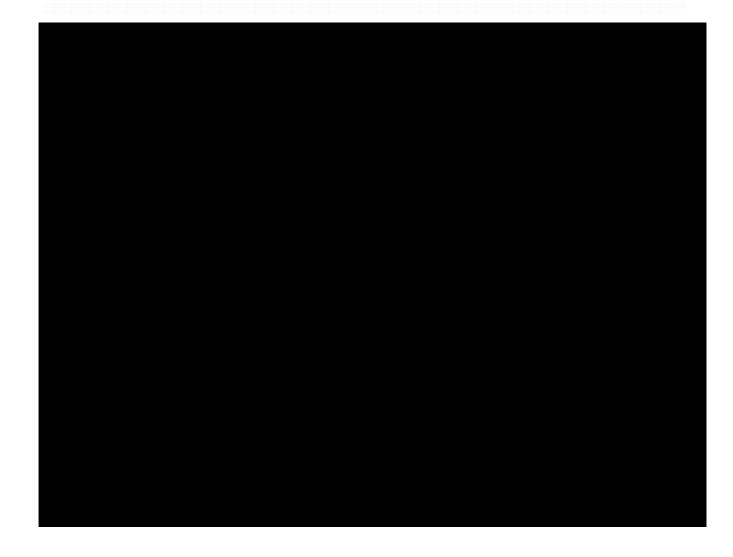




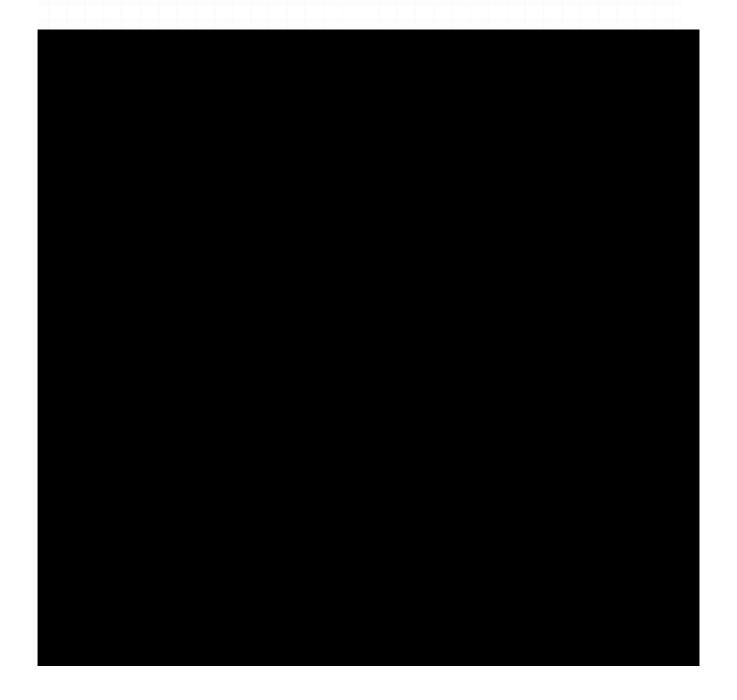
















Thank you for your cooperation

$\pmb{CFQ\text{-}UK}_{\text{ CYSTIC FIBROSIS QUESTIONNAIRE - UK}}$

Children Ages 12 and 13

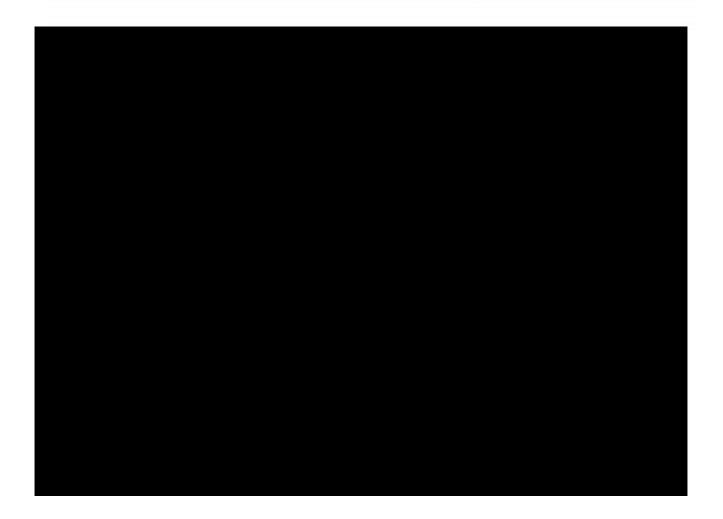
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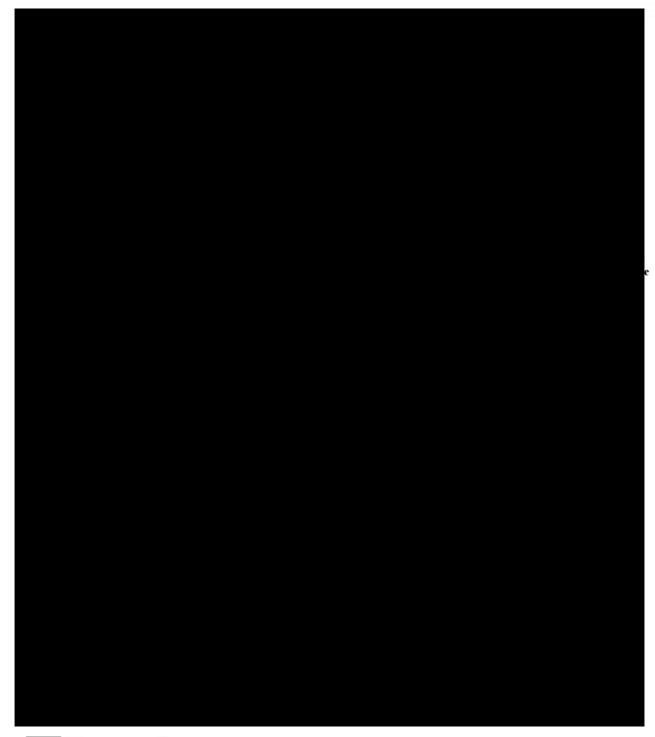




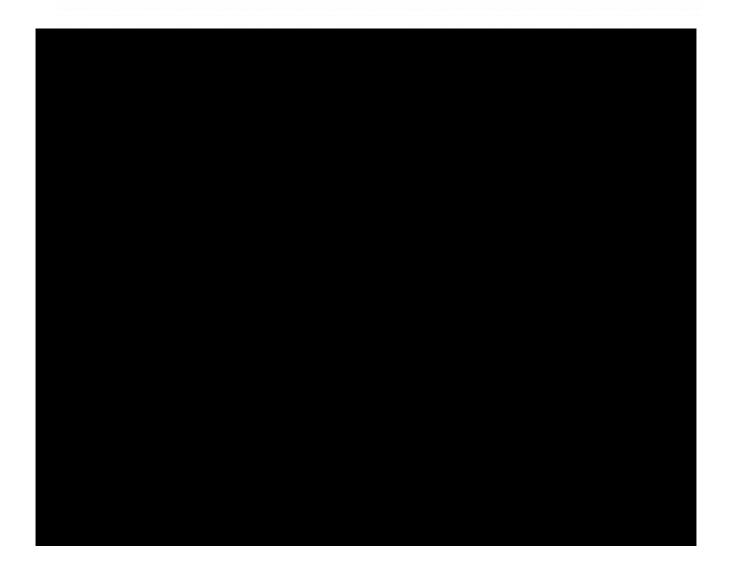






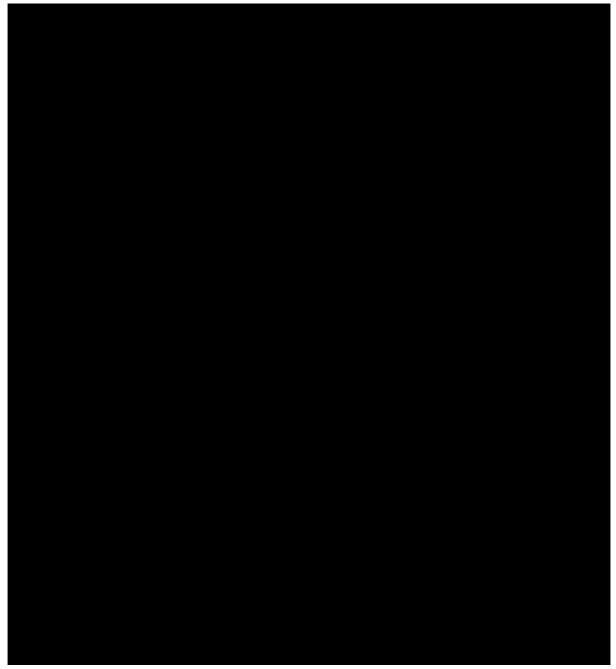


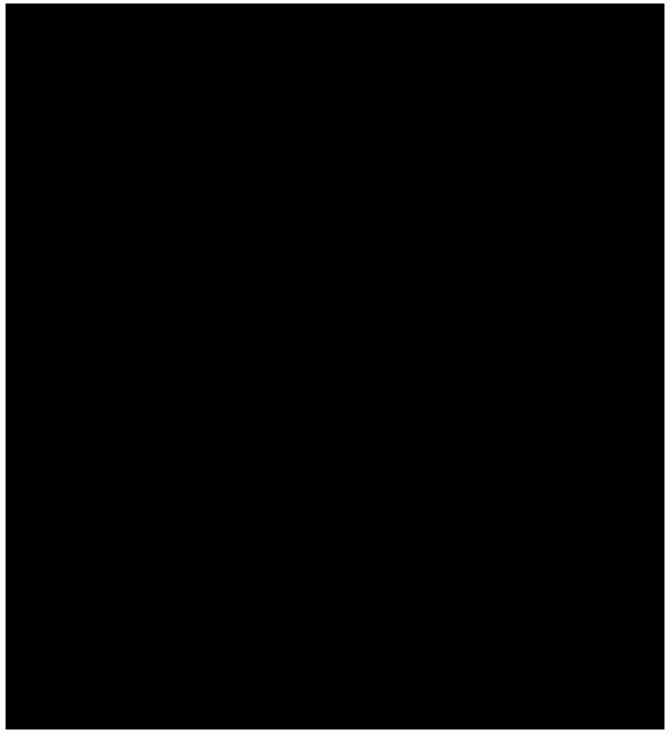


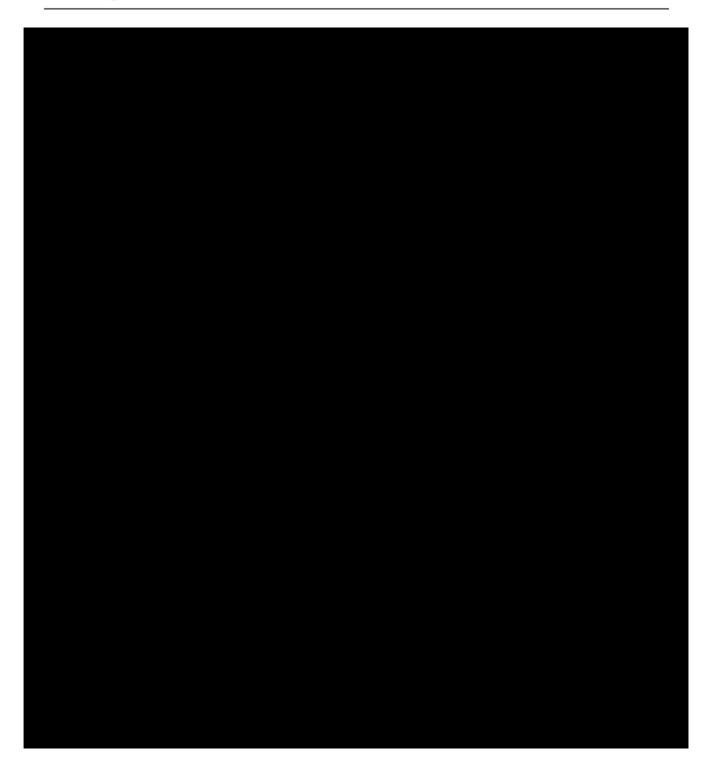


CFQ-UK CYSTIC FIBROSIS QUESTIONNAIRE - UK

Adolescents and Adults















Sibling Research Advisory Group - Sibling group member Feedback - 10.02.2018

For each statement below, please tick a box to indicate how much you agree with the statement from 1 (strongly disagree with the statement) to 5 (strongly agree with the statement). Please add additional comments if you have any.

Particip Focus (pation in the	Strongly Disagree				Strongly agree	Additional comments
rocus (1	2	3	4	5	
1.	The group was lead in a professional manner				/		
2.	The group leaders were effective in helping us talk about issues				J		
3.	The group meeting was interesting				/		
4.	There was enough time to talk about the issues		۰	1			
5.	I felt comfortable sharing my thoughts		99	/			
6.	I feel that joining in with the group today was worthwhile			-		/	
7.	I would like to attend another Sibling Research Advisory Group meeting				/		

What part of the Sit The Qu disscuss	olling Research Advisory Group 112" Part beca and take part	p did you like the most? use every one-co	rud

How was your overall experience this afternoon in the Sibling Research Advisory Group meeting?
1 1000
chance to discuss.
What could we do to make this Sibling Research Advisory Group better?
more interactive activities.
Is there anything else you would like to let us know?
For example about how the session was today, any of the research ideas we talked about, advice
for the future of the group, practical issues etc.
Everything was perfect *
Minutel Committee Committee Charles Charles Bananasala Adula and Carron annula S

Would you take part in the Sibling Research Advisory Group again?

Yes B

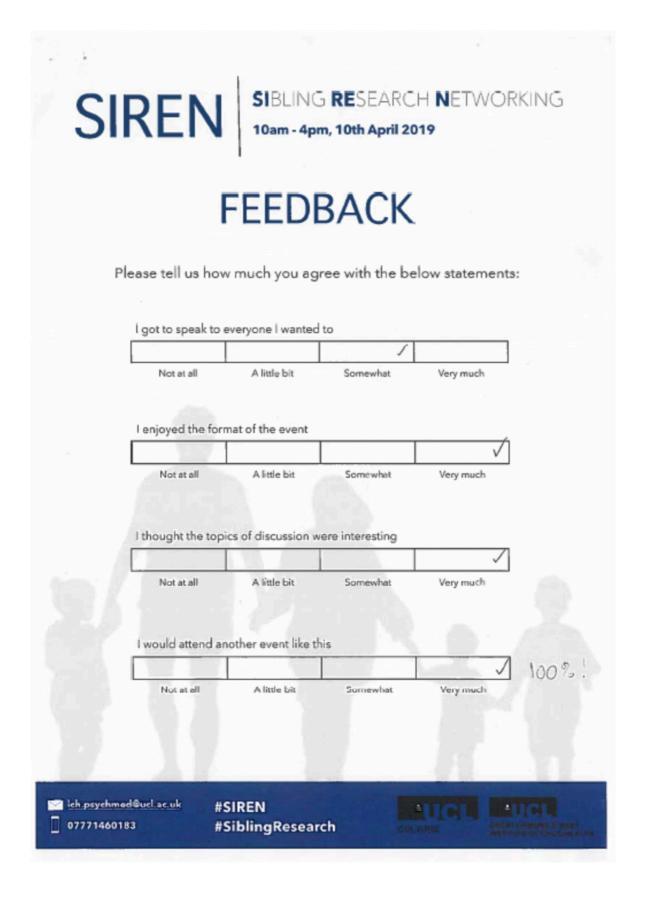
For each statement below, please tick a box to indicate how much you agree with the statement from 1 (strongly disagree with the statement) to 5 (strongly agree with the statement). Please add additional comments if you have any.

Partici Focus (pation in the Group	Strongly Disagree		3		Strongly agree 5	Additional comments
1.	The group was lead in a professional manner					~	
2.	The group leaders were effective in helping us talk about issues				/		. :
3.	The group meeting was interesting					/	
4.	There was enough time to talk about the issues				/		
5.	I felt comfortable sharing my thoughts					/	
6.	I feel that joining in with the group today was worthwhile		_		/		
7.	I would like to attend another Sibling Research Advisory Group meeting			<u></u>			

What part of the Sibling Research Advisory Meeting other people can comb	ing whe people paddeds are making new
friends and learn more somewhat	e- our/other experience.

How was your overall experience this afternoon in the Sibling Research Advisory Group meeting?
It was everyn and calm because I was haver been stressed.
What could we do to make this Sibling Research Advisory Group better?
by soying how was our expiran with our siblings
Is there anything else you would like to let us know? For example about how the session was today, any of the research ideas we talked about, advice for the future of the group, practical issues etc.
Would you take part in the Sibling Research Advisory Group again?

No



SIREN

SIBLING RESEARCH NETWORKING 10am - 4pm, 10th April 2019

FEEDBACK

Please Indicate how much you feel each of the objectives of SIREN were met:

Not at all	A little bit	Somewhat	Very much
Objective 2: Get esearch and sup		work and the futur	re of sibling
			1
Not at all	A little bit	Somewhat	Very much
Objective 3: Mee	t like-minded peo	ple	
Objective 3: Mee	et like-minded peo	ple	
Objective 3: Mee	et like-minded peo	ple	√ Very much



SIREN SIBLING RESEARCH NETWORKING 10am - 4pm, 10th April 2019

FEEDBACK

Please tell us how much you agree with the below statements:

Not at all	A little bit	Somewhat	Very much
enjoyed the for	mat of the event		1000
			-
Nut at all	A little bit	Somewhat	Very much
hought the top	ics of discussion w	rere interesting	
thought the top	ics of discussion w	vere interesting	
thought the top Not at all	ics of discussion w	vere interesting	Very much
			Very much
			Very much
Not at all	Á little bit	Somewhat	Very much
Not at all		Somewhat	Very much
Not at all	Á little bit	Somewhat	Very much





SIBLING RESEARCH NETWORKING 10am - 4pm, 10th April 2019

FEEDBACK

Please Indicate how much you feel each of the objectives of SIREN were met:

Objective 1: Talk with you about current sibling research and support A little bit Not at all Somewhat Very much Objective 2: Get your input on our work and the future of sibling research and support Not at all A little bit Somewhat Very much Objective 3: Meet like-minded people A little bit Not at all Somewhat Very much General Comments: