Understanding anxiety in Duchenne Muscular Dystrophy:
Starting with the perspectives of boys with the condition and parents

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Thesis declaration form

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

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Date: 17.09.19
Overview

This thesis focuses on anxiety experienced by males with Duchenne Muscular Dystrophy (DMD), a chronic and life-limiting genetic neuromuscular condition. **Part 1** reviews the research literature on the prevalence, characteristics, causes and impact of anxiety within this population. A high prevalence of anxiety amongst this population is indicated. The causes of anxiety remain unclear, however, organic explanatory frameworks and the adjustment literature offer models for understanding its presence. The review highlights the absence of literature investigating the characteristics and impact of anxiety within this population and the need for further research into these areas.

**Part 2** reports on a qualitative study applying Framework Analysis to perspectives about anxiety elicited from eight boys aged seven to 18 with a diagnosis of DMD and 14 parents of boys aged 18 and under with a diagnosis of DMD. Analysis of focus group transcripts highlights six characteristics of anxiety recurrently reported by participants. Factors impacted by anxiety and reciprocally influential over anxiety are identified across individual, family, social and environmental contexts. A model is provided outlining a set of hypotheses about relationships between anxiety and contextual factors. Clinical implications are discussed, including the importance of psychological assessment, formulation, and intervention for anxiety as part of a comprehensive DMD care approach. Suggestions for future research are made.

**Part 3** offers reflections on several key influences and observations involved in the research processes of this study. These include more detailed considerations of possible causes and consequences of a dominant organic model of anxiety within this population. Reflections on the influence of a Liberation Psychology framework on the researcher’s theoretical orientation, analysis and write-up are shared. Finally,
the challenges and priorities of creating a model of anxiety in DMD are discussed, with a particular focus on the researcher’s attempts to privilege the voices of participants throughout this process.
Impact statement

Duchenne Muscular Dystrophy (DMD) is a complex and life-limiting genetic neuromuscular condition affecting approximately one in 3500-5000 live male births. The primary feature of the condition is progressive muscle wasting, however, this is often accompanied by a constellation of other difficulties such as cognitive, social, emotional, and behavioural problems. Anxiety has received comparatively little research attention in comparison to other aspects of the condition and has not been the focus of any research studies to date, despite estimations that it reaches a clinical threshold for 6.6% - 29.3% of individuals with DMD. Efforts to find a cure for Duchenne Muscular Dystrophy and increase the length of life for individuals with this condition may have diverted research attention away from many of the commonly associated cognitive, social, emotional, and behavioural difficulties, including anxiety.

While a handful of studies have investigated the prevalence of anxiety amongst individuals with DMD, this is the first study to explore the characteristics and impact of anxiety amongst this population. The study's findings offer new insights into the complex and nuanced characteristics of anxiety experienced by individuals with DMD, providing vital information for effective assessment and formulation of this need across research and clinical settings. The pervasive and impactful nature of anxiety evidenced by this study highlights the need for routine psychological assessment within clinical settings alongside psychological formulation and intervention where indicated, as part of comprehensive DMD care approach. The pertinence of this recommendation is increased by the high prevalence of dissatisfaction expressed by parent participants in this study with respect to the support they have received in relation to the anxiety experienced by their sons.
The identification of reciprocally influential pathways between anxiety and individual, family, social and environmental contexts within this population widens the framework for understanding this difficulty beyond the organic explanatory framework that has dominated the literature, whilst not negating the likelihood of an organic contribution. The model of anxiety proposed by this study offers an integrative framework to researchers, clinicians and parents for making sense of this complex, and often confusing experience amongst some boys with DMD. The model also widens potential targets for effective intervention across different systemic levels, guided by robust formulation.

This is the first known study to publish the qualitative perspectives of boys with DMD under the age of 16 and therefore provides an example to the research community of the achievability and value of drawing on their expertise and experiences for research purposes. The study demonstrates the utility of focus groups for generating multiple perspectives from parents of children with DMD and boys with DMD.

The study highlights the lack of research in this area and the need for further research investigating the anxiety experienced by this population, particularly with respect to establishing effective measures for capturing the complexities and nuances of this anxiety for research and clinical purposes.
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Part 1: Literature Review

Anxiety in Duchenne Muscular Dystrophy
Abstract

Aims

This paper explores anxiety amongst males with Duchenne Muscular Dystrophy (DMD). The paper includes a narrative literature review with a focus on the following questions:

1. What is the prevalence of anxiety in DMD?
2. What are the characteristics of anxiety in DMD?
3. What is the impact of anxiety on males with DMD and their families?
4. What causes and maintains anxiety in DMD?

Method

A three-step process was conducted involving consultation with experts in the field, the study of a recent meta-analysis, and a comprehensive literature search of the Ovid Medline database. Relevance criteria were deliberately kept broad to avoid missing papers of interest.

Results

Ninety-one papers identified as relevant to the questions of the paper were reviewed and discussed.

Conclusions

Anxiety experienced by individuals with DMD is an understudied research area, however, a high prevalence of anxiety amongst this population is suggested. An organic contribution to anxiety appears likely but remains undefined. A small number of studies consider anxiety as part of a secondary adjustment response to this condition. The characteristics and impact of anxiety amongst individuals with
DMD are not addressed by extant literature and represent costly gaps in knowledge. These questions will be the focus of a subsequent empirical study.
Introduction

This review aims to explore anxiety amongst males with Duchenne Muscular Dystrophy (DMD). DMD is a complex genetic neuromuscular condition predominantly affecting males. The condition is characterised by progressive muscle wasting, however, this is often accompanied by a constellation of other difficulties such as cognitive, social, emotional, and behavioural problems (Hendriksen et al., 2015; Ricotti et al., 2016; Snow, Anderson, & Jakobson, 2013). Problems such as anxiety and depression (often referred to as internalising problems), are estimated to be present in around a quarter of individuals with DMD (Banihani et al., 2015; Ricotti et al., 2016), although anecdotal evidence from clinicians and parents suggests this is likely to be an underestimate for anxiety. While advancements in medical research and treatment over the last two decades have considerably increased quality of life and life expectancy for individuals with DMD (Bushby et al., 2010a, 2010b), the psychosocial, emotional and behavioural aspects of the condition have received less attention, despite indicators of their prominence.

Due to the complexity of features that often comprise DMD, alongside the demands and stresses of living with a chronic and fatal condition, anxiety in this population is likely to present with differences to the anxiety found in typically developing individuals (Hendriksen et al., 2009; Perrin, Stein, & Drotar, 1991). Developing a robust understanding of anxiety experienced by males with DMD and its’ relationship to other aspects of the condition is therefore essential for parents,

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1 The terms DMD and Duchenne will be used interchangeably throughout this conceptual introduction to describe Duchenne Muscular Dystrophy.
2 The term ‘males’ or ‘individuals’ will be used rather than ‘boys’ throughout this paper to reflect the fact that many individuals with DMD live well into adulthood. The term ‘boys’ will be used only to describe males aged 18 and under.
caregivers, teachers, clinicians, and males with the condition, as well as the academic community.

**Key concepts**

**Duchenne Muscular Dystrophy**

Duchenne muscular dystrophy (DMD) is an X-linked genetic disorder affecting approximately one in 3500-5000 live male births (Emery, 1991; Ricotti et al., 2016). The condition is characterised by progressive muscle wasting and premature death resulting from mutations to the dystrophin gene. The dystrophin gene is one of the longest in the human genome and is found on the X-chromosome. Females have two X-chromosomes whereas males have one X-chromosome and one Y-chromosome. This means that females have two copies of the dystrophin gene and males have only one. If a female has a mutation to the dystrophin gene, the second copy of the dystrophin gene on their other X-chromosome can usually compensate and code for sufficient dystrophin protein to be made. In males, there is no ‘back-up’ X-chromosome, so when a mutation occurs to the dystrophin gene, the production of dystrophin protein is impaired. This is the reason why the condition predominantly affects males.

In healthy individuals, the dystrophin gene provides the necessary instructions for making the protein dystrophin. Dystrophin protein maintains the structural integrity of muscle membranes throughout the body and is also found in particular areas of the brain (Hoffman, Brown, & Kunkel, 1987; Muntoni, Torelli, & Ferlini, 2003). In males with DMD, mutations to the dystrophin gene stop the production of dystrophin protein almost completely, causing muscle fibres to suffer progressive damage and scarring across the body (Perronnet & Vaillend, 2010). In the brain, much remains unknown about the function of dystrophin protein. In typically
developing individuals, dystrophin is thought to play an important role in early brain development and ongoing brain functioning (Anderson, Head, Rae, & Morley, 2002; Mehler, 2000; Perronnet et al., 2012). It is also known to help facilitate signaling between nerve cells. What this means for individuals with DMD requires further investigation (Perronnet & Vaillend, 2010).

Figure 1. The DMD gene, dystrophin protein isoforms and potential mutation sites. Note. Potential mutation sites are indicated by red ‘x’ markers with corresponding letters. The figure shows the location of different dystrophin isoforms. The table shows the dystrophin isoforms that would be lost because of mutations at each site. Adapted from Maresh, K., & Muntoni, F. (2018). Why Some Children with DMD Have Learning and Behaviour Difficulties. In A Guide to Duchenne Muscular Dystrophy (pp. 22-41). London, UK. Copyright (2018) by Jessica Kingsley Publishers.

The dystrophin gene codes for several different forms of dystrophin protein, called isoforms, which are found in specific areas of the body (Muntoni et al., 2003). For example, isoform Dp427m is found in the skeletal and heart muscle, whereas isoform Dp140 is found in the brain, retinas and kidneys. Due to variation in isoform size, the site of the mutation affects which dystrophin isoforms are lost and which remain intact (Figure 1). For example, a mutation near to 5’ end of the dystrophin gene (close to exon 1) will only cause the largest isoform (Dp427) to be lost, whereas a mutation near to the 3’ end of the gene (closer to exon 63) will cause
nearly all isoforms to be lost. As such, the site of the mutation affects the nature of
an individual’s condition. In general, the closer the mutation to the 3’ end of the
gene, the more severe the functional consequences for the individual.

Symptoms of DMD are usually first noticed when boys are aged between
three and five. Loss of ambulation usually occurs by age 13 (Bushby et al., 2010a).
Scoliosis, contractures, and respiratory and cardiac impairment are some of the
common physical features of the condition (D’Amario et al., 2018; Walter & Reilich,
2017) (Figure 2). Premature death is usually caused by cardiac and pulmonary
complications when males are in their 30s (Snow et al., 2013), although
advancements in symptomatic and surgical treatment have significantly increased
average life expectancy over the last two decades (Bushby et al., 2010a; Bushby et
al., 2010b).

<table>
<thead>
<tr>
<th>Age (years)</th>
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<tbody>
<tr>
<td>0</td>
</tr>
<tr>
<td>---</td>
</tr>
<tr>
<td>Minor symptoms</td>
</tr>
<tr>
<td>First problems in walking and climbing stairs</td>
</tr>
<tr>
<td>Wheelchair and severe skeletal deformities</td>
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<tr>
<td>Impaired arm function</td>
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<tr>
<td>Nighttime ventilation</td>
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<tr>
<td>24h ventilation</td>
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<tr>
<td>High mortality</td>
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Figure 2. Typical course and transition in DMD

Alongside the physical components of the condition, males with DMD
commonly present with a wide range of cognitive, social, emotional, and behavioural
problems. Preliminary results from a recent meta-analysis suggest that males with
DMD have an approximate 10-fold increased risk of intellectual disability, 8-fold risk
of autism spectrum disorder (ASD), 7-fold risk of anxiety/depression, 5-fold risk of epilepsy, 3-fold risk of attention deficit hyperactivity disorder (ADHD), and double the risk of behavioural problems compared to the general population (Maresh & Muntoni, 2017) (Figure 3). It is worth noting that while the prevalence of these difficulties is substantially higher than the general population, for each diagnosis, the majority of individuals with DMD do not reach a diagnostic threshold.

**Figure 3.** (adapted from Maresh & Muntoni, 2017): The percentage prevalence of CNS co-morbidities in DMD compared to the normal population.

Within the general population, functional bidirectional relationships are widely agreed to exist between neurodevelopmental, cognitive, emotional and behavioural difficulties, meaning the presence of one problem or disorder increases the chance of further disorders (Cohen, Barwick, Horodezky, Vallance, & Im, 1998; Hesdorffer, 2016; Simonoff et al., 2008). For example, the rate of cognitive difficulties amongst children and young people seen within child and adolescent mental health services (CAMHS) is approximately nine times higher than in the general population (Humphrey, 2006). Around 40% of young people with ASD meet criteria for at least one DSM-IV anxiety disorder (van Steensel, Bögels, & Perrin, 2011), compared with
8.1% of children aged 19 and under in the general population (NHS Digital, 2018). A similar phenomenon is seen amongst males with DMD. In a study of 87 males with DMD, 37% were found to have no emotional, behavioural or neurodevelopmental problems, 26% had a diagnosis of one of these problems, and the remaining 37% had two or more of these problems (Ricotti et al., 2016). A further study found that children with DMD who were identified as having significant behavioural concerns were more likely to have learning disabilities (Darke, Bushby, Couteur, & McConachie, 2006).

As with the overlap seen between neurodevelopmental, cognitive, emotional and behavioural difficulties in the general population, an interaction of genetic and environmental factors is likely to contribute towards the pattern of multiple disorders commonly seen in DMD (Figure 4).

Figure 4. Functional bidirectional relationships between Duchenne muscular dystrophy, neurodevelopmental, cognitive, and emotional and behavioural problems.
Correlation between mutation site and average IQ has been identified, providing strong evidence for a dystrophin-related contribution to intellectual ability (Colombo et al., 2017; Maresh & Muntoni, 2017; Taylor et al., 2010), however, the basis of emotional and behavioural problems such as anxiety is less well understood (Banihani et al., 2015; Ricotti et al., 2016).

**Anxiety**

Anxiety is a universally experienced phenomenon and widely understood as driven by the fear-based adaptive response of the autonomic system. The primary purpose of the autonomic system is to rapidly equip the body for surviving in the presence of a threat to physical safety (Öhman, 2000). Whilst theoretical debate continues regarding the relationship between fear and anxiety, a generally agreed distinction is the future-oriented nature of anxiety and the present-oriented nature of fear (Sylvers, Lilienfeld, & LaPrairie, 2011). Clark and Beck (2011) argue that fear is the primitive, basic process underlying anxiety, and anxiety is a more complex and enduring state associated with threat appraisals. Evolutionary psychologists contend that anxiety is the product of a primal threat-focused part of the brain acting alongside more highly evolved brain regions capable of more complex cognitive processes, such as rumination, imagining the worst outcome, and self-criticism. These more highly evolved cognitive processes are proposed to keep the more primal threat response going for longer (Gilbert, 1993).

Although anxiety is a universal experience, the ways in which it manifests are not universal. Anxiety presents differently in different populations, influenced by a multitude of factors including cultural and social norms and expectations, care-giving relationships, life experiences, and genetics (Creswell, Murray, Stacey, & Cooper, 2011; Hofmann & Hinton, 2014). Whilst most humans feel anxious from time to time, anxiety can have wide-ranging and life limiting consequences for those who experience it on a regular basis. The most recent national survey of adult mental
health indicates that 4.7% of adults in the UK have anxiety problems (Bebbington, Brugha, Jenkins, McManus, & Meltzer, 2009). The most recent survey of the mental health of children and young people in England indicates that 7.2% of 5-19 year old children have an anxiety disorder (NHS Digital, 2018). Persistent anxiety can severely reduce quality of life and level of functioning across environments such as home, school, and work (Mental Health Foundation, 2009). It can impact negatively upon relationships, school attendance, activities of daily living, attainment, productivity, and physical health (Carr, 2016; Mental Health Foundation, 2014; Hagell, Coleman, & Brooks, 2013; Hill, Waite, & Creswell, 2016). These consequences also have implications for those around an individual experiencing anxiety, such as their family members, partners, friends and colleagues (Mental Health Foundation, 2009; Senaratne, Van Ameringen, Mancini, & Patterson, 2010).

Considering these factors within the context of DMD, which presents many additional challenges, it is of vital importance to better understand the ways in which anxiety manifests and affects the lives of this population and their families, for both research and clinical purposes.

**Literature Review Questions**

In this review I will be focusing on anxiety experienced by males with DMD, with a particular interest in the following questions:

1. What is the prevalence of anxiety in DMD?
2. What are the characteristics of anxiety in DMD?
3. What is the impact of anxiety on males with DMD and their families?
4. What causes and maintains anxiety in DMD?
Method

Process

I have addressed the questions of this review by looking at the existing literature. Whilst this is not a systematic review, a three-step process was used to maximise the probability that all relevant literature was identified. Firstly, I consulted with experts in the field, identifying the most relevant papers and reviews, and looking through bibliographies. Secondly, I consulted a recent meta-analysis which I was a contributor of alongside other members of the research team. Thirdly, I conducted a literature search (August 31, 2018) using Medline (Ovid) with the following search terms: (((Anxiety OR fear OR neuropsychological OR psychosocial OR emotional OR "mental health") AND ("Duchenne Muscular Dystrophy" OR DMD OR Duchenne)) NOT smile). Relevance criteria were deliberately kept broad to avoid missing papers of interest, particularly due to my observation during the first two steps that anxiety is explored from many different angles within the literature and considered under a wide variety of terms. Papers containing the word “smile” were excluded due to an unrelated body of research exploring the “Duchenne Smile”\(^3\).

209 results were returned from my Medline search. 118 papers were not considered relevant to the focus of this review, leaving 91 remaining studies. I ensured all papers identified through consultation with experts in the field, bibliographies from key texts, and papers reviewed in the aforementioned meta-analysis were included. Papers were then screened by hand to identify those deemed relevant to addressing the questions of this review.

\(^3\) A phenomenon unrelated to DMD. The “Duchene Smile” is a type of smile resulting from the contraction of muscles surrounding the eye as well as muscles around the mouth.
Results

What is the prevalence of anxiety in DMD?

From my review of the literature, establishing the prevalence of anxiety in DMD is difficult, largely because anxiety is rarely assessed as a discrete entity and more commonly researched under broader categories such as ‘internalising difficulties’ or ‘anxiety/depression’. A handful of papers calculate the prevalence of anxiety specifically, with prevalence rates published of 6.6% - 29.3% (Banihani et al., 2015; Hendriksen, Vles, Aalbers, Chin, & Hendriksen, 2018; Latimer et al., 2017; Lee, Buckingham, Kauer, & Mathews, 2018; Pangalila et al., 2015). Studies investigating the prevalence rate of ‘internalising problems’ (defined as symptoms of anxiety and depression) report prevalence rates of 23.4% - 36.4% (Colombo et al., 2017; Donald, Mathema, Thomas, & Wilmshurst, 2011; Ricotti et al., 2016). A recent meta-analysis of the literature found a pooled prevalence rate of 18.6% for the category of ‘anxiety/depression’ (Maresh & Muntoni, 2017).

The literature is therefore suggestive that anxiety is highly prevalent amongst males with DMD and substantially more prevalent than anxiety observed in the general population, where the most recent survey publishes a 5.4% prevalence rate of anxiety disorders across 5-19 year old males in England (NHS Digital, 2018). Prevalence rates of anxiety experienced by males with DMD must be considered with caution however, due to the small number of studies upon which they are based, the wide range of methods for gathering data, and the different thresholds at which anxiety is considered ‘present’. Amongst the studies identified, anxiety was measured using the following methods: the Hospital Anxiety and Depression Scale (HADS) (Zigmond & Snaith, 1983) (e.g. Pangalila et al., 2015)), the Child Behavior Checklist (CBCL) (Achenbach & Ruffle, 2000) (e.g. Colombo et al., 2017; Donald et al., 2011; Ricotti et al., 2016), the Diagnostic and Statistical Manual of Mental
Disorders (DSM-IV) (American Psychiatric Association, 1994) (e.g. Banihani et al., 2015), a retrospective study of medical notes (e.g. Lee et al., 2018), and parent or patient report about anxiety disorder diagnoses known to have been previously given (e.g. Hendriksen et al., 2018; Latimer et al., 2017). Amongst these methods, a mixture of self-rated, parent/carer-rated, and clinician-rated tools were used. Variation in measurement tool makes it difficult to compare and review findings across studies, an issue highlighted by the aforementioned meta-analysis where only two studies met the inclusion criteria for assessing the prevalence of ‘anxiety/depression’ (Maresh & Muntoni, 2017). As each measurement tool has its own threshold for considering anxiety to be ‘present’, each is likely to yield different results regarding prevalence.

Another issue is the sensitivity of generic measurement tools for identifying anxiety in the DMD population, something that has not been tested to date for any of the aforementioned standardised scales. The CBCL has been criticised for an overreliance on physical symptoms to meet thresholds for anxiety, risking a high rate of false positives amongst children with chronic illnesses such as DMD (Perrin, Stein, & Drotar, 1991). The CBCL measure consequently risks detecting illness-related variables from a child’s condition or the side effects of treatment, rather than the behavioural or psychological difficulties it seeks to measure (Perrin et al., 1991).

Items within generic measurement tools rely on symptoms of anxiety commonly observed amongst typically developing children, rather than children with DMD. For example, items such as “Runs away from home” (Item 67 of CBCL for ages 6-18) or “Often complains of headaches, stomach-aches or sickness” (Item 3 of parent-report SDQ for ages 4-17), do not account for the physical symptoms and limitations of living with DMD. Anxiety may present in alternative ways amongst the DMD population and remain undetected as a consequence.

Stress and emotional reactions are widely considered a normal part of adjustment to a chronic and fatal illness (Eiser, 1990), but are not accounted for by
all measures, risking the pathologising of normal coping behaviour. For example, answers to items such as "Fears going to school" (Item 30 of CBCL for ages 6-18) could, for an individual with DMD, simply reflect a normal adjustment challenge such as adapting to using a wheelchair for the first time in a school environment, rather than an indication of an anxiety disorder.

The Personal Adjustment and Role Skills Scale, 3rd edition (PARS-III) (Klein Walker, Stein, Perrin, & Jones Jessop, 1990), specifically developed to measure psychosocial adjustment in children with chronic physical, has proved reliable and valid for assessment of psychosocial adjustment in males with DMD (Hendriksen et al., 2009). However, this measure was not found within any of the studies identified by this review, beyond the validation study. Whilst its use as a clinical screening tool is recommended (Birnkrant et al., 2018), anecdotal evidence suggests this is not commonplace within clinical settings in the UK.

What are the characteristics of anxiety in DMD?

No papers were found dedicated to the investigation of anxiety characteristics in DMD. This appears to be a reflection of the emphasis within extant literature on anxiety prevalence, which constrains exploration of anxiety characteristics in DMD to the items contained within generic measures.

A small number of papers propose social anxiety as a common characteristic of anxiety in DMD (Harper, 1983; Siapno & Carter, 2013). Beyond these studies, DMD literature in the area of social behaviour focuses either on the prevalence of neurodevelopmental social communication difficulties, or the presence of unspecified ‘social problems’ (e.g. Donders & Taneja, 2009; Hinton, Nereo, Fee, & Cyrulnik, 2006; Snow et al., 2013), rather than the characteristics of social anxiety.

Sixteen relevant qualitative studies were identified in the literature review. Qualitative approaches tend to focus on experiences of coping with the physical aspects of DMD, rather than conceptualising anxiety as an integral aspect of DMD
also requiring coping responses. Nevertheless, these studies put forward an important mechanism to consider as part of understanding anxiety amongst this population. The term ‘anxiety’ is rarely mentioned explicitly within the identified qualitative literature, however, attention is given to various challenges of living with DMD or living with someone who has DMD. For example, qualitative studies explore experiences of diagnosis and learning about the condition (Fujino et al., 2016), the impact on siblings of having a family member with DMD (Read, Kinali, Muntoni, Weaver, & Garralda, 2011), coping experiences of parents (Webb, 2005), experiences of transitioning to adult services (Hamdani, Mistry, & Gibson, 2015; Lindsay, McAdam, & Mahendiran, 2017), and experiences of participating in clinical trials (Franson, Kinnett, & Cripe, 2018). These studies bring attention to the secondary consequences of DMD, all of which could be conceived as capable of influencing anxiety, though not explicitly addressed.

One study presents a first-hand account of several anxiety-provoking aspects of adult life with DMD, highlighting how the nature of worries and fears changes as the disease progresses with time (Spies, Schipper, Nollet, & Abma, 2010). First-hand reflections from one of the authors describes his worries and fears relating to the end of life, the possibility of his ventilator stopping working, his emergency alarm not being heard, and the challenges of adjusting to life in a residential home, including receiving intimate personal care from professionals.

**What is the impact of anxiety on males with DMD and their families?**

Little information was found in the literature regarding the impact of anxiety on males with DMD and their family members, with greater attention given to the impact of physical aspects of DMD.
Quality of life

A body of literature explores the impact of DMD as a whole on quality of life, both for individuals with the condition and family members. Quality of life papers were reviewed due to evidence within wider literature that symptoms of anxiety in people with physical health problems strongly predict quality of life (Chachamovich et al., 2010; Lukas et al., 2009). ‘Quality of life’ (QoL) is defined by the World Health Organization (WHO) as “individuals' perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns. It is a broad ranging concept, incorporating in a complex way individuals' physical health, psychological state, level of independence, social relationships, personal beliefs and their relationships to salient features of the environment” (The WHOQOL Group, 1995, p. 1405).

Amongst identified studies, quality of life was usually measured using the PedsQL (Varni, Seid, & Kurtin, 2001), a standardised measure enabling the consideration of social and emotional functioning in the context of a health condition. The PedsQL contains subscales of Physical Functioning, Emotional Functioning, Social Functioning, and School Functioning, as well as an Overall Quality of Life score. A review of quality of life in children with DMD found that health-related quality of life in boys with DMD is generally worse than for healthy peers but also worse than children with other chronic illnesses, especially in the physical domains (Wei, Speechley, & Campbell, 2015). Boys at a more severe stage of the disease reported worse physical health-related quality of life, but not necessarily worse psychosocial health-related quality of life than boys at a less severe stage in this study. As quality of life in a health context is concerned with the impact of the health condition as a whole, it is not possible to use this literature to separate out which aspects of DMD affect quality of life, and more relevantly to this review, the relationship between anxiety and quality of life. Quality of life, as measured at
present, offers little towards understanding the specific impact of anxiety on males with DMD and their family members. Whether anxiety impacts upon quality of life as an intrinsic component of DMD, or as a secondary consequence of living with DMD is a question that is not addressed and remains unclear. It is also worth holding in mind that the PedsQL has been criticised by some authors for its strong focus on functional aspects of quality of life when used with chronically ill children (Fayed et al., 2012; Ravens-Sieberer et al., 2006).

**Adjustment literature**

The literature investigating 'adjustment' was also reviewed to consider the question of the impact of anxiety on males with DMD and their families, however, similar to quality of life literature, adjustment literature generally investigates DMD as a whole rather than separating out the impact of different aspects of the condition. Adjustment can be defined as the adaptive task of managing upsetting feelings and frustrations aroused by illness, and preserving an emotional balance (Moos & Tsu, 1977). Wider literature proposes that a normal adjustment process involves a range of cognitive, emotional and behavioural responses to stressors (Dekker & de Groot, 2018; Eiser, 1990), and that anxiety symptoms can be indicative of the level of psychosocial adjustment (Hendriksen et al., 2009; Klein Walker et al., 1990).

The aforementioned study validating the PARS III adjustment measure for use with the DMD population reports the subscale of 'anxiety/depression' as having significant correlation with the subscales of *peer relations* (.29), *dependency* (.43), *hostility* (.54), *productivity* (.39) and *withdrawal* (.42). This analysis highlights a pattern of relationships existing between anxiety and other factors amongst males with DMD (Hendriksen et al., 2009), however, the direction of effect is not clear, and the distinct roles of anxiety and depression cannot be separated. Within the adjustment subsection of the literature, anxiety is implicitly considered as a part of a
secondary coping response to the condition rather than an intrinsic part of the condition. The literature does not provide insight into how anxiety may itself impact upon adjustment processes in DMD. However, adjustment literature provides an important angle from which to consider several domains that may influence and be influenced by the anxiety experienced by individuals with DMD.

Several papers explore the impact of DMD as a whole on parents and siblings of an individual with DMD (Magliano & Politano, 2016; Peay et al., 2016; Read et al., 2011). Similarly, within this subsection of the literature the impact of anxiety specifically on family members does not receive attention. This is with the exception of one study which found that the anxiety experienced by boys with DMD negatively impacts upon caregiver burden (Pangalila et al., 2012). A further study found that behavioural problems presented by boys with DMD increases stress in their mothers (Nereo, Fee, & Hinton, 2003) although the extent to which “behavioural problems” encompasses anxiety is not discussed.

**What causes and maintains anxiety in DMD?**

A range of factors are likely to influence the presence and nature of anxiety in DMD, however this has not been the focus of any studies to date. Causal models of anxiety in the context of DMD broadly fall under two main models within current literature.⁴ The first, and more dominant model, is that anxiety is an organic and biologically-driven consequence of disordered architecture in the central nervous system, caused by the absence of dystrophin (e.g. Colombo et al., 2017; Ricotti et al., 2016). The second, and less explicitly documented conceptualisation, considers the influence on psychosocial wellbeing of responding to the secondary challenges

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⁴ Due to fact that anxiety is rarely mentioned explicitly within the literature, the models through which it is understood are often communicated implicitly. For example, papers investigating “brain-based comorbidities in DMD” implicitly understand anxiety as a product of disordered brain architecture.
and limitations presented by DMD (e.g. Hendriksen et al., 2009). An even smaller number of studies consider the impact of the wider family system on the general wellbeing of someone with DMD. No research was found addressing how these factors may interact with respect to anxiety, nor how the responses of systems wider than the family, such as social circles, school cultures, healthcare provision and political attitudes may influence and interact with anxiety.

**Evidence for a brain-based model of anxiety**

Some of the strongest evidence in the literature for a brain-based model of anxiety comes from mouse models. It is well regarded that the hippocampus and amygdala are involved in emotional learning and the regulation of emotions such as anxiety (Etkin & Wager, 2007; Marschner, Kalisch, Vervliet, Vansteenhoven, & Buchel, 2008; McDonald & Mott, 2017). Dystrophin is known to be present in these areas of a neurotypical brain (Lidov, 1996). A body of literature demonstrates that the amygdala and hippocampi of mice with DMD (mdx mouse) are affected by dystrophin inhibition. These mice have increased stress reactivity, fearfulness, and altered social behaviour (Goyenvalle et al., 2015; Sekiguchi et al., 2009).

Intellectual disability amongst males with DMD has an emerging brain-based evidence base due to the correlation found between IQ and mutation site (Maresh & Muntoni, 2017; Taylor et al., 2010). Anxiety is often considered within this same brain-based model despite not demonstrating the same reliable correlation. One paper found significantly higher "anxious/depressed" CBCL subscale scores amongst DMD patients with distal mutations than those with proximal mutations (Colombo et al., 2017), however, this finding has not been replicated (Banihani et al., 2015; Ricotti et al., 2016). In the general population, anxiety is higher amongst individuals with an intellectual disability than those without (Emerson & Hatton, 2007) and intellectual disability is approximately 10 times more prevalent amongst individuals with DMD than in the general population (Maresh & Muntoni, 2017).
high co-occurrence of intellectual disability and anxiety may suggest a brain-based link, but this cannot be confirmed due to the potential impact of environmental responses to people with intellectual disabilities, which may increase the likelihood of anxiety, and are not accounted for. The organic relationship between DMD and anxiety therefore remains unclear.

Anxiety is more commonly found amongst individuals with neurological health conditions than individuals with non-neurological health conditions (Hesdorffer, 2016; Mash & Barkley, 2003). Considering the neurological component of DMD due to altered brain dystrophin, this could further suggest that anxiety has a neurological basis in DMD. Additionally, anxiety is higher amongst individuals with ASD (van Steensel et al., 2011), and this neurodevelopmental disorder is more commonly diagnosed amongst individuals with DMD than in the general population. Individuals with DMD are also more likely to have emotional and behavioural problems compared to unaffected siblings (Hinton, De Vivo, Nereo, Goldstein, & Stern, 2000).

**Evidence for psychosocial model of anxiety**

It is unlikely that a brain-based model tells the whole story of anxiety experienced by males with DMD. The aforementioned findings offer support for a biological model of anxiety in DMD, however, they do not confirm this as the only possible influencing mechanism. A psychosocial model is interested in both the responses of an individual with DMD to the consequences and challenges of their condition and the responses of others to their condition. Much is known about the ways humans are affected by the social, political and material world around them which encourages consideration of a wider range of factors when thinking about anxiety (Bronfenbrenner, 1979; Dahlgren & Whitehead, 1991; Williams, Neighbors, & Jackson, 2008). A psychosocial model of anxiety in DMD considers the ways in which social, political, and material contexts respond differently to an individual with DMD than they do to an unaffected child. It is also interested in the ways these
different contexts may interact with one another and the child’s individual responses to their condition.

As individuals with DMD have had their condition from birth, pre-diagnostic information is not available, making it difficult to investigate personal factors that may be predictive of anxiety in the same way as studies have explored in the general population (e.g. Kagan, 1989). Nevertheless, the life limiting and progressive nature of DMD, its physical restrictions, health complications, regular hospital visits, and complex medical regimens can be hypothesised as likely to elicit consequences relevant to the consideration of anxiety. These could include changes to one’s level of independence and roles, changes to social relationships, altered educational or vocational opportunities, a sense of vulnerability, a sense of change happening beyond one’s control, and uncertainty about the progression of the condition. These are all experiences commonly faced by people suffering from anxiety and provide examples of how the secondary consequences of DMD and personal and social responses could influence the anxiety experienced by males with this condition.

Due to the progressive nature of DMD, the challenges faced by those with the condition continually change themselves. This continual adjustment presents a challenge in itself. One study found that adjustment within the DMD population was poorest amongst boys aged 8-10, the age at which ambulation is usually lost (Hendriksen et al., 2009). Another study found that a trend of compromised verbal skills when individuals with DMD are younger influenced higher levels of social difficulties during this stage of life, whereas older children were more likely to struggle with problems arising from their progressing physical disabilities and medical complications (Poysky, 2007). A further study found that anxiety was most pronounced amongst individuals with DMD during transition periods, such as the transition to using a wheelchair or a respirator (Fujino et al., 2016). These studies suggest that anxiety may change over time in response to the changing physical
and neurodevelopmental aspects of the condition. Considering advancements in medical care which continue to prolong life expectancy, individuals with DMD now encounter new psychosocial challenges as an adult which are capable of impacting upon anxiety in new ways. These challenges may relate to independence, romantic relationships, employment, and end of life planning (Hendriksen et al., 2009). The earlier mentioned study (Spies et al., 2010) describes several anxieties experienced by an adult with DMD in relation to some of these issues.

Several studies found an effect of family member stress on the emotional wellbeing of their child with DMD and add evidence to a psychosocial model of anxiety in DMD. These studies include a finding that the combined score of a measure of parent stress about preserving their emotional wellbeing with a measure of ‘parent distress’ accounted for 26% of the variance in parent-reported internalising behaviour problems presented by their child with DMD (Thompson, Zeman, Fanurik, & Sirotkin-Roses, 1992). Another study published results indicating that the level of stress experienced by the family predicted the degree of psychosocial adjustment in adolescents with DMD (Reid & Renwick, 2001). A previously mentioned study which found higher levels of “problem behaviours” amongst boys with DMD who had more highly stressed mothers (Nereo et al., 2003), also found maternal stress to be reciprocally mediated by “problem behaviours”. These studies highlight the role of family responses in the emotional experience of an individual with DMD, although more studies are needed to better understand these relationships and increase the conclusiveness of the findings.

Whilst providing some helpful indications, this literature does not offer robust or conclusive evidence about the relationships between psychosocial factors and the anxiety experienced individuals with DMD. Consideration of wider systemic influences beyond the family on the experience of anxiety amongst individuals with DMD is not evident within the literature, despite common consideration of these factors in the literature regarding anxiety and psychological distress in the general
population. For example, research shows that the level of discrimination experienced by individuals in the general population is positively associated with the prevalence of generalised anxiety (Kessler, Mickelson, & Williams, 1999). Further research highlights the extent of discrimination experienced by people with disabilities (Dixon, Smith, & Touchet, 2018). Positive associations have also been identified between stigma and psychological distress amongst people with intellectual disabilities (Ali, King, Strydom, & Hassiotis, 2015). Physical disability and intellectual disabilities are common components of DMD, however, the impact of social attitudes has not been considered within the literature regarding the emotional wellbeing of this population. Similarly, DMD literature does not consider the impact of environmental factors such as wheelchair accessibility, or a lack of this, on anxiety. One would hypothesise that anxiety would be higher for boy with DMD in a non-wheelchair accessible school than for a boy with DMD in a fully wheelchair accessible school, for example, despite them sharing the same genetic mutation.

One study found evidence supporting the impact of wider social influences on quality of life (Otto et al., 2017). Using the KIDSCREEN-10 index (Ravens-Sieberer et al., 2014), "health-related quality of life" amongst individuals with DMD was associated with household income, the frequency of attending a clinic with specialised staff, the number of days spent outside home, and the attitude of the local community. No significant association with age was found. When the same authors used the generic PedsQL (Varni et al., 2001) and Neuromuscular Module of the PedsQL (Davis et al., 2010), quality of life was positively associated with age as well as by the country of residence and the disease stage. As the study investigates quality of life as a whole, the causal influences of wider social influences on anxiety are not possible to tease apart. The difference in findings between quality of life measures highlights the current problems with reliable measurement in DMD. Despite these issues, the study provides some important indications of the influence of wider social factors on the presence anxiety in DMD.
Integrating available models: a biopsychosocial approach

Considering the literature identified in the review as a whole, it is likely that a set of complex interactions takes place between the biologically determined presentation of DMD and the personal, social, and environmental responses to the condition. How anxiety experienced by this population is conceptualised is likely to have implications for how it is identified and responded to, and the expectations attached to what can create change. For example, a purely biological conceptualisation may drive an expectation of change attached only to biological interventions. Conceptualising anxiety in DMD as a complex interaction of biological, psychological, social and environmental factors, may widen markers for identification and targets for intervention.

Further research

Before the various contributions of biological, psychological, social and environmental factors can be tested and the effectiveness of anxiety interventions can be measured, a robust understanding of how anxiety presents in DMD is required. Without knowing what to look for or enquire about, anxiety cannot be confidently screened for or measured. The literature review provides direction on the areas needing to be explored as a starting place. The review also demonstrates the need for an open and exploratory approach to anxiety in DMD, rather than one constrained by generic anxiety measures.

Considering the wide-ranging and life limiting consequences of anxiety on individuals in the general population and those around them, it is also of vital importance that further research investigates the impact of anxiety on the lives of individuals with DMD and their family members. Some evidence suggests that measuring the impact of emotional and behavioural difficulties is more effective than measuring symptoms for distinguishing between community and psychiatric
samples (Goodman, 1999), highlighting the importance of better understanding the impact of anxiety within this population for research and clinical purposes.

Focus groups are well suited to the open and exploratory approach required for such an investigation. Eliciting multiple perspectives from parents of children with DMD and young people with DMD using semi-structured interview schedules would enable existing literature to direct discussion topics, whilst also allowing new information about the characteristics of anxiety and its impact to emerge.

Framework analysis (Ritchie & Spencer, 1994) is a five-step process used to analyse qualitative data such as focus group transcripts. Benefits of framework analysis include that whilst it is fundamentally driven by the original accounts and observations of participants (inductive), it can also incorporate priori codes (deductive). This suits a study investigating the anxiety experienced by males with DMD due to the balance required between allowing for unexpected themes to emerge, considering the originality of the research, whilst also drawing on themes identified in current anxiety measures, anxiety theory, and DMD literature. The framework approach also enables the researcher to look across the data at the frequency of themes in the sample and analyse single cases if desired. This suits a study which would explore two main participant groups (children and parents), as it allows for the possibility of separate or combined analysis to remain open.

Framework analysis is well suited to research that has specific pre-decided questions, a limited time frame and a pre-designed sample. Although framework analysis may generate theories, the main concern of this type of analysis is to describe and interpret what is happening in a particular setting (Ritchie & Spencer, 1994). The well-defined process involved in framework analysis enables the reworking and reconsideration of earlier ideas due to the traceable nature of the process. These features enable transparency of the researcher’s analytic process.
The five key stages of framework analysis are:

1. **Familiarisation** – The researcher gains an overview of the data by listening to tapes and reading transcripts multiple times.

2. **Identifying a thematic framework** – The researcher highlights interesting and recurrent parts of the data and attempts to identify key issues, concepts and themes relevant to the research questions. Codes or labels are used to describe the themes, which are organised into a framework in discussion with consultees.

3. **Indexing** – The framework is systematically applied to the data. Iterative cycles of refining and reapplying the framework are informed by feedback from consultees and observations of the researcher. The framework continues to be adapted until it can be applied to the data without new codes being generated.

4. **Charting** – The indexed data is summarised into a chart format. Data is abstracted from the transcripts and each participants’ contributions within each category are summarised.

5. **Mapping and interpretation** – The researcher pulls together key characteristics of the data set by reviewing charts and finding patterns. The researcher articulates their own sense-making of the data, in light of the research questions, and sometimes creates a visual map of findings.

Parent and child transcripts could be analysed separately, however, if substantial overlap between the themes in each group was observed, the findings from the separate analyses could be subsequently integrated into a final coding framework.
A focus group study analysed with framework analysis would not be designed to test the causes of anxiety in DMD, but it is hoped that the findings would yield insights in this area. The study would hope to improve the understanding of anxiety in DMD and thus improve the identification and monitoring of anxiety for both research and clinical purposes. It would be hoped that this, in turn, would promote routine screening and inform better clinical management. Such a study would also hope to raise awareness amongst healthcare professionals, teachers, and others working with these families about the importance of considering anxiety as part of a comprehensive and integrated DMD care approach.

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Part 2: Empirical Paper

Understanding anxiety in Duchenne Muscular Dystrophy: Starting with the perspectives of boys with the condition and parents
Abstract

Aims

This qualitative study explores the characteristics of anxiety experienced by individuals with Duchenne Muscular Dystrophy (DMD) and the impact of this anxiety on males with the condition and their families.

Method

Eight boys aged between seven and 18 with a diagnosis of DMD and 14 parents of children aged 18 and under with a diagnosis of DMD were recruited to the study. Separate parent and child focus groups were used to elicit perspectives about anxiety. Qualitative data was analysed using a framework analysis approach. Standardised measures of emotional, behavioural, and social communication difficulties were used to situate the sample.

Results

The framework analysis yielded six characteristics of anxiety recurrently reported by participants. Four further themes described contextual factors relevant to understanding the anxiety experienced by this population. These themes were Individual responses, Family responses, Social responses, and Environmental responses.

Conclusions

Anxiety appears to be a pervasive and impactful issue amongst the DMD population. Recurrently reported anxiety characteristics are suggestive of a DMD-specific anxiety phenotype. A network of interacting factors across family, social and environmental contexts are relevant considerations for psychological assessment, formulation and intervention in this area. A model is proposed outlining a set of
hypotheses about the relationships existing between anxiety and contextual factors. Future research could investigate these relationships further and identify measures capable of detecting the nuanced characteristics of anxiety identified by the study.
Introduction

Duchenne Muscular Dystrophy

DMD is a complex and fatal genetic neuromuscular condition predominantly affecting males. The primary feature of the condition is progressive muscle wasting, however, this is often accompanied by a constellation of other difficulties such as cognitive, social, emotional, and behavioural problems (Hendriksen et al., 2015; Ricotti et al., 2016; Snow, Anderson, & Jakobson, 2013). Individuals with DMD have an approximate 10-fold increased risk of intellectual disability, 8-fold risk of autism spectrum disorder (ASD), 7-fold risk of anxiety/depression, 5-fold risk of epilepsy, 3-fold risk of attention deficit hyperactivity disorder (ADHD), and double the risk of behavioural problems in comparison to the general population (Maresh & Muntoni, 2017).

The lack of dystrophin produced by individuals with DMD causes muscle fibres to suffer progressive damage and scarring across the body. Loss of ambulation usually occurs around age 12, and premature death occurs, on average, when males are in their 30s (Snow et al., 2013). Dystrophin is found in the brain of typically developing individuals and is thought to play a role in early brain development and ongoing brain functioning (Anderson, Head, Rae, & Morley, 2002; Mehler, 2000; Perronnet et al., 2012). Much remains unknown about the consequences of a lack of dystrophin in the brain of an individual with DMD (Perronnet & Vaillend, 2010).

Within the literature, a brain-based dystrophin link is the dominant model for understanding the high prevalence of neurodevelopmental, emotional, and behavioural problems within the population. IQ has shown correlation with mutation site amongst males with DMD (Colombo et al., 2017; Maresh & Muntoni, 2017; Taylor et al., 2010), providing emerging evidence for a dystrophin-related
contribution to intellectual ability. The basis of emotional and behavioural problems such as anxiety amongst individuals with DMD has received comparatively less research attention and remains poorly understood (Banihani et al., 2015; Ricotti et al., 2016).

**Anxiety and DMD**

Available literature suggests a high prevalence of anxiety within the DMD population. Prevalence rates are reported of 6.6% - 29.3% (Banihani et al., 2015; Hendriksen, Vles, Aalbers, Chin, & Hendriksen, 2018; Latimer et al., 2017; Lee, Buckingham, Kauer, & Mathews, 2018; Pangalila et al., 2015), however, anecdotal evidence from parents and clinicians suggests these figures underestimate the true prevalence of anxiety within this population. Prevalence rates may be affected by the reliability and validity of generic measures for use within the DMD population, which remain untested for most measures used within published research.

Due to the complexity of features that often comprise DMD, alongside the demands and stresses of living with a chronic and fatal condition, anxiety in this population is likely to present with differences to anxiety found in typically developing individuals (Hendriksen et al., 2009; Perrin, Stein, & Drotar, 1991). However, research dedicated to investigating the characteristics of this anxiety has not been conducted to date. Some research suggests that social anxiety may be a common characteristic of anxiety in DMD (Siapno & Carter, 2013), but literature in this area tends to focus on the prevalence of neurodevelopmental social communication difficulties rather than social anxiety characteristics. A small body of research relating to ‘adjustment’ explores the secondary impact of responding to the physical consequences of DMD, both for the individual with DMD and their family members. Although anxiety is not always mentioned explicitly within this subsection of the literature, wider literature considers anxiety to be a common component of an
adjustment responses to a chronic illness (Dekker & de Groot, 2018; Eiser, 1990). A small number of studies make explicit links between the presence of anxiety and the adjustment of individuals with DMD to the physical aspects of their condition (e.g. Fujino et al., 2016; Hendriksen et al., 2009; Poysky, 2007) and highlight that anxiety is likely to change over time as part of adjustment to the frequently changing physical and neurodevelopmental consequences of the condition. One study publishes a finding that anxiety is most pronounced amongst individuals with DMD during transition periods, such as the transition to using a wheelchair or a respirator (Fujino et al., 2016).

Taken together, available literature indicates a complex set of relationships existing between anxiety and direct biologically-driven consequences of DMD, secondary coping responses, and wider systemic involvement with someone who has DMD. However, a limited understanding is currently held around the characteristics of anxiety which has implications for research and clinical services. For example, without knowing what to look for or enquire about, anxiety cannot be confidently screened for or responded to with effective interventions.

Little attention is given to the impact of anxiety on individuals with DMD and their family members, including the adjustment challenges that may be presented by anxiety itself, in addition to the adjustment challenges presented by the physical consequences of DMD. Early indications suggest that anxiety can both influence and be influenced by significant others such as parents (Fujino et al., 2016; Pangalila et al., 2012), however, these relationships are not well understood, and the impact of anxiety on the individual and wider systems remains minimally addressed.

In the general population, anxiety is known to have wide-ranging and life limiting consequences for those who experience it on a regular basis with the potential to affect relationships, school attendance, daily living activities, attainment, productivity, and physical health (Carr, 2016; Hagell, Coleman, & Brooks, 2013; Hill,
Waite, & Creswell, 2016; Mental Health Foundation, 2014). Anxiety can also affect others around the individual, such as family members, partners, friends and colleagues (Mental Health Foundation, 2009; Senaratne, Van Ameringen, Mancini, & Patterson, 2010). Irrespective of whether anxiety is a direct or indirect consequence of DMD, or an interaction between these two, it is of vital importance to better understand the impact of anxiety on individuals with the condition and family members for research and clinical purposes.

This study therefore aims to investigate the following questions:

1. What are the characteristics of anxiety experienced by males with DMD?
2. What is the impact of anxiety on males with DMD and their families?

As this area has not previously been investigated, the study will seek the first hand perspectives of boys with DMD and parents of boys with DMD. This study is not designed to test the various organic and psychosocial contributions to anxiety in DMD, however it is possible that findings may contribute towards hypotheses about the relationships between anxiety and other aspects of the condition, and its relationships with wider systems involved with someone who has DMD.

**Method**

**Overview**

Boys with a diagnosis of DMD aged 18 and under, and parents of boys aged 18 and under with a diagnosis of DMD were invited to participate in separate child and parent focus groups. The focus groups followed semi-structured schedules exploring the characteristics of anxiety in DMD and experiences of its impact on the individual with DMD and their family members (Appendices A-B).
Expert consultation

During the design of this study, Dr James Poysky, a Clinical Psychologist with a special interest in DMD, and father to a son with DMD, served as a voluntary consultant. Dr Poysky contributed towards the design of both parent and child focus groups and the content of the semi-structured interview schedules. The co-researcher, an expert DMD medical clinician and neuromuscular research fellow, also contributed towards the study design.

Ethical Approval

Ethical approval was sought and granted via the UCL Research Committee only, as participants were not recruited via any NHS sites or clinicians (Research code: 12307/001; Appendix C)

Grant funding

The study was awarded a small doctoral grant of £2000 from the UCL Grand Challenges scheme which supports cross-disciplinary research. The study was eligible due the collaboration of researchers from the UCL faculty of Brain Sciences (Rachel Trimmer) and the UCL faculty of Population Health Sciences (Kate Maresh).

Recruitment Procedure

Participants were recruited through a variety of methods. The researchers contacted several charities and asked for the study to be publicised on their social media platforms and at relevant events. Information about the study and sign-up sheets were additionally presented at a conference and book launch event (Appendix D). A database held by the research team of parents consenting to
contact about future research was also used to approach potential participants (Appendix E).

Participants

Participants were 14 parents of children with DMD and eight boys with DMD. Parents were eligible to participate in the study if they had a son with a diagnosis of DMD aged 18 years or younger. Boys were eligible to participate in the study if they were aged 7-18 and had a diagnosis of DMD. A maximum of one parent per family was permitted to take part in the study to maximise the number of families represented. Parents could participate in the research without their sons taking part and boys could take part irrespective of their parents’ participation. Six parent and child pairs took part in the study.

Design

Parent and child focus groups were chosen to facilitate in-depth discussion and elicit multiple perspectives. The use of semi-structured interview schedules enabled existing literature to guide discussion whilst providing a flexible and open approach to allow new information to emerge. Creative methods for engaging the child participants and eliciting their perspectives were devised, drawing on ideas from child focus group literature (e.g. Fargas-Malet, McSherry, Larkin, & Robinson, 2010; Gibson, 2012; Heary & Hennessy, 2002). Methods used included showing child participants a picture of a fictional character with DMD, called ‘Josh’, and asking participants to imagine what Josh might feel worried about in different environments. In Part 2 of the child focus groups, participants were presented with cards containing pictures and labels of different worries, including worries mentioned during Part 1. Participants were asked to sort the cards into ‘big worries’,
<table>
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<th>Parent participant number</th>
<th>Age</th>
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<th>Ethnicity</th>
<th>Child participant number</th>
<th>Age of child with DMD</th>
<th>Autism Spectrum Disorder diagnosis</th>
<th>Intellectual disability diagnosis</th>
<th>SDQ Total difficulties score</th>
<th>SDQ Impact Score</th>
<th>RCADS total anxiety score</th>
<th>T ≥ 65 = borderline clinical threshold</th>
<th>Level of satisfaction with access to support around understanding and managing son's anxiety</th>
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<td>No</td>
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<td>46-50</td>
<td>F</td>
<td>White British</td>
<td>C06</td>
<td>12-18</td>
<td>“Borderline”</td>
<td>No</td>
<td>25</td>
<td>7</td>
<td>46</td>
<td></td>
<td>Very satisfied</td>
</tr>
<tr>
<td>P12</td>
<td>46-50</td>
<td>M</td>
<td>White British</td>
<td></td>
<td>7-11</td>
<td>Yes</td>
<td>No</td>
<td>9</td>
<td>1</td>
<td>38</td>
<td></td>
<td>Very satisfied</td>
</tr>
<tr>
<td>P13</td>
<td>26-30</td>
<td>F</td>
<td>Arab/African</td>
<td></td>
<td>7-11</td>
<td>No</td>
<td>No</td>
<td>18</td>
<td>3</td>
<td>46</td>
<td></td>
<td>Neither satisfied nor dissatisfied</td>
</tr>
<tr>
<td>P14</td>
<td>41-45</td>
<td>F</td>
<td>White British</td>
<td></td>
<td>&lt; 7</td>
<td>No</td>
<td>No</td>
<td>9</td>
<td>2</td>
<td>56</td>
<td></td>
<td>Very satisfied</td>
</tr>
<tr>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>C05</td>
<td>12-18</td>
<td>No</td>
<td>No</td>
<td>26</td>
<td>8</td>
<td>75</td>
<td></td>
<td>Very dissatisfied</td>
</tr>
<tr>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>C08</td>
<td>12-18</td>
<td>Yes</td>
<td>No</td>
<td>29</td>
<td>2</td>
<td>50</td>
<td></td>
<td>Somewhat dissatisfied</td>
</tr>
</tbody>
</table>

*Parent participants with more than one son with DMD  M = male  F = female  SDQ = Strengths and difficulties questionnaire  RCADS = Revised child and anxiety and depression scale
‘middle-sized worries’, and ‘little worries’ as a technique for facilitating discussion (Appendix F).

The size and duration of focus groups was informed by similar studies and focus group literature (e.g. Hoppe, Wells, Morrison, Gillmore, & Wilsdon, 1995). Each parent focus groups aimed to recruit six participants (18 participants total), however, due to last minute cancellations, 14 parent participants attended three parent focus groups in total (n=5, n=5, n=4). Two focus groups for child participants were held with the aim of recruiting four participants to a group for 7-11 aged participants and four participants to a group for 12-18 aged participants. These participant numbers were achieved.

Information about each participant and their family, including routinely used measures of emotional, behavioural, and social communication difficulties were collected on the day of the focus groups to situate the sample.

**Procedure**

Information sheets and a video about the study were provided in the first instance to parents and children who were interested in participating (Appendix G). Information about the study was written in age appropriate language for the 7-11, 12-18, and adult participants respectively (Appendices H-J). Information was also provided to parents whose sons were interested in participating (Appendix K). It was strongly emphasised to these parents that their son should make his own choice about participating. Prospective participants were requested to leave at least 24 hours after receiving information about the study before consenting to take part to allow time for careful consideration of their decision. Parent and child participants were assigned to groups on a ‘first come first served’ basis. A list of reserve child participants was created due to particularly high levels of interest in these groups.

Focus groups took place in a private room at the Institute for Child Health. All groups were audio and video recorded. The researchers met individually with each
participant prior to each focus group to give out consent forms and answer any
questions. A parent was present for these meetings with child participants.
Participants were informed about the recording of the groups, confidentiality, and
their freedom to withdraw from the study at any point. Boys aged 7-15 signed an
assent form (Appendix L) and boys aged 16-18 signed a consent form (Appendix
M). Parents of all child participants signed a form consenting to their son’s
participation (Appendix N). All parent participants signed their own consent form
(Appendix O). At the start of each focus group, participants were reminded about
confidentiality in the group setting and there being no obligation to respond to
questions asked. It was also acknowledged that talking about anxiety and talking in
a group can both be anxiety-provoking experiences in themselves. Participants were
given permission to leave the room at any time if they needed, and to speak to the
researchers at the end of the group if they felt concerned about how they had been
affected.

Parent focus groups lasted 2 hours with a 30 minute break in the middle. The
7-11 child group lasted 1 hour 15 minutes with a 15 minute break after 45 minutes.
The 12-18 child group lasted 1 hour 30 minutes with a 20 minute break after 45
minutes. Light refreshments were provided for participants and accompanying family
members during the break.

Following the end of the focus groups, participants were sent a £20 gift
voucher as a ‘thank you’ for their time. Travel expenses were reimbursed for each
adult participant and each child participant plus one accompanying adult using the
small doctoral grant.

Analytic procedure

All focus groups were transcribed by the author of this thesis. Transcriptions
were analysed using a Framework Analysis approach (Ritchie & Spencer, 1994)
Details of how the five-step process of Framework Analysis was conducted within this particular study are outlined below:

1. **Familiarisation** – The researcher immersed herself in the data by reading the focus group transcripts, listening to the audio recordings, and watching video footage of the focus groups. The focus group transcripts were uploaded to QSR International's NVivo 12 software (QSR, 2018) to highlight, organise and manage the process of analysis more efficiently.

2. **Identifying a thematic framework** – Notes were made about initial ideas for themes. Notes about themes in the parent focus groups and child focus groups were made separately at this stage. Initial themes were discussed alongside the transcripts between the researcher (R.T.) the supervisor (W.M.) and the co-researcher (K.M.). Similarities between parent and child themes were noted and a decision was made to combine these into one overall framework.

3. **Indexing** – An initial framework was devised and applied to the data. Reliability of the coding process was ensured by consultation with four parent participants, two child participants, the co-researcher (K.M.), and an independent qualitative researcher. Parent participants who wished to take part at this stage were sent a summary of the themes, and a transcript section from their particular focus group demonstrating how the framework had been applied to their contributions. A summary of themes was not sent to the two child participants assisting with validity checks due to the sensitivity of many issues described within the framework. Child participants assisting at this stage were sent coded sections of their focus groups. A final
framework was agreed upon following iterative changes informed by the consultations and feedback.

4. **Charting** – A digital spreadsheet was used to index the data into a chart of category summaries. Each framework category was represented by a column and each participant was represented by a row.

5. **Mapping and interpretation** – A table lists the themes and subthemes comprising the final interpretation of key characteristics from the data (Table 2). Example quotes extracted from the focus groups are shown in the results section as evidence of the quality of the analysis. Sections of dialogue between several participants are included at times to demonstrate the group discussion process around a particular theme. A map of themes is also presented to reflect the relationships between themes drawn by participants within the study and the researcher (Figure 1).

**Disclosure of the Researcher’s Perspective**

I am a White British female researcher completing this research as part of my Clinical Psychology Doctorate at UCL in London. I have a background of working with children and families affected by mental health issues in both clinical and research contexts (Support Worker, Assistant Psychologist). I am interested in the interactions that take place between biological, personal, social, cultural, environmental, and political contexts. I draw on a range of models in my clinical work, including systemic models such as Ecological Systems Theory (Bronfenbrenner, 1979) and Coordinated Management of Meaning (CMM) (Cronen & Pearce, 1982) to help me understand the many contexts influencing and reciprocally influenced by the problems people experience and the meanings they, and others, make about them.
On hearing about the project, I was motivated to take on this research role due to my interest in applying an integrative approach to wellbeing. My motivation for improving the support available to boys with DMD and their families has grown throughout the project as a result of meeting many families affected by DMD and hearing about their experiences of anxiety. I have been encouraged to remain reflective about the influence of my perspective on the analysis, but also to own this position and bring my integrative stance to an issue that has historically been investigated through less integrated approaches.

Results

Results from the focus groups using framework analysis

The focus group transcripts were analysed using Framework Analysis (Ritchie & Spencer, 1994). The final thematic framework is outlined in Table 2. Within these five themes, 15 subthemes were identified. Quotes from parent participants are labelled as ‘P’ followed by a unique numerical identifier. Quotes from child participants are labelled as ‘C’ followed by a unique numerical identifier. To preserve confidentiality, unique identifiers (e.g. P01) are not related to those listed in Table 1.
<table>
<thead>
<tr>
<th>Theme</th>
<th>Subtheme</th>
<th>% parents mentioning theme</th>
<th>% boys mentioning theme</th>
<th>No. times mentioned by parents</th>
<th>No. times mentioned by boys</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Characteristics of anxiety</td>
<td>Catastrophic conclusions: “Worrying about the worst situation”</td>
<td>57</td>
<td>100</td>
<td>18</td>
<td>26</td>
</tr>
<tr>
<td></td>
<td>Rigidly held anxieties: “Going round in circles”</td>
<td>57</td>
<td>13</td>
<td>14</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>Extreme distress: “Total meltdown”</td>
<td>93</td>
<td>38</td>
<td>31</td>
<td>7</td>
</tr>
<tr>
<td></td>
<td>The unexpected and unfamiliar: “Not in the plan”</td>
<td>64</td>
<td>50</td>
<td>15</td>
<td>8</td>
</tr>
<tr>
<td></td>
<td>Social anxieties: “Will I fit in?”</td>
<td>57</td>
<td>100</td>
<td>13</td>
<td>38</td>
</tr>
<tr>
<td></td>
<td>Anxieties about Duchenne-related change, needs, and medical procedures: “We’re the most vulnerable”</td>
<td>86</td>
<td>100</td>
<td>33</td>
<td>76</td>
</tr>
<tr>
<td>2. Individual responses</td>
<td>Individual emotional and physical responses</td>
<td>64</td>
<td>75</td>
<td>18</td>
<td>30</td>
</tr>
<tr>
<td></td>
<td>Individual adapting, coping, and responding</td>
<td>86</td>
<td>100</td>
<td>37</td>
<td>74</td>
</tr>
<tr>
<td>3. Family responses</td>
<td>Family emotional and physical responses: “The whole emotional thing shifts”</td>
<td>94</td>
<td>0</td>
<td>84</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>Family adapting, coping, and responding: “Settling into a new normal”</td>
<td>100</td>
<td>0</td>
<td>89</td>
<td>0</td>
</tr>
<tr>
<td>4. Social responses</td>
<td>Stigma, misunderstanding, and unhelpful responses from others: “They don’t understand”</td>
<td>36</td>
<td>88</td>
<td>17</td>
<td>65</td>
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<tr>
<td></td>
<td>Support and solidarity: “You’re not on your own”</td>
<td>64</td>
<td>50</td>
<td>14</td>
<td>8</td>
</tr>
<tr>
<td>5. Environmental responses</td>
<td>Medication</td>
<td>36</td>
<td>13</td>
<td>7</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>Wheelchair accessibility</td>
<td>29</td>
<td>63</td>
<td>4</td>
<td>25</td>
</tr>
<tr>
<td></td>
<td>Access and quality of schooling and healthcare: “A postcode lottery”</td>
<td>79</td>
<td>13</td>
<td>35</td>
<td>1</td>
</tr>
</tbody>
</table>
Characteristics of anxiety. This theme captures the observations and experiences of anxiety shared by both parent and child participants. The first three subthemes describe internal and external features of anxiety, whereas the following three subthemes describe specific types of situation where participants recurrently reported anxiety being experienced.

(1a.) Catastrophic conclusions: “Worrying about the worst situation”. Just over half of parent participants noticed their child with DMD expressing worries about extreme eventualities. These included worries about tsunamis, burglary, terrorist attacks and fatal accidents. Parents noticed that their sons’ worries commonly contained an irrational and imaginative quality, and described their sons frequently asking ‘What if…’ questions, enquiring about
possible future adverse predicaments. Parents described feeling puzzled and often surprised by the content of worries and ideas expressed by their sons.

“Our questions they come out with, you think you’re going to crash the car!” (P03)

Whilst most child participants did not articulate the process of their anxious thinking, all eight shared worries about highly adverse eventualities within the group setting. For example:

e.g. “I always have a feeling I’ll get lost.” (C02)

e.g. “I get worried I will get on the wrong plane.” (C04)

e.g. “…when you go out somewhere and your parents are at home and you think, “what would happen if I’m out here and they die or something?”” (C05)

One child participant shared an awareness of his catastrophic thinking:

“…you start worrying, you get to like extreme conclusions, so like ‘if this happens, this happens, this might happen to me and I might be ill’ and stuff like that, so like jumping to bad conclusions.” (C08)

(1b.) Rigidly held anxieties: “Going round in circles”. Eight parents described their son’s tendency to become “totally fixed” [P09] on particular ideas, questions and worries. This characteristic was often, but not always, present alongside the catastrophic ideation described in the preceding theme. Several parents noticed their son’s ability to hold onto information he had previously heard before expressing a related worry much later on. Parents shared their experiences of cyclical and repetitive conversations with their sons about his worries. They noticed their sons finding it difficult to let go of ideas and think flexibly.
“Once he’s got a worry in his head he’ll just go over and over and over it to the point where I don’t know what else to say to him, but he just can’t shake it out of his head, he just goes on and on and on about it and it’s…there are so many things.” (P01)

Child participants did not reflect on this style of thinking, however, the researchers noticed several conversations within the child focus groups which appeared to demonstrate repetitive thinking patterns and escalating catastrophic ideation. A section of the transcript provides an illustration:

(C05): …someone might start a fire and how are you meant to get out of there?

(C06): What if you’re on your own? Like for like two minutes.

(C08): Yeah

(C06): If your mum’s in the toilet and she doesn’t even know what’s going on or something

(C08): Yeah

(C05): Yeah or they’re asleep and you’re awake

(C08): Yeah and something happens. Someone comes into the house and they’re like, obviously my mum doesn’t know what’s happening and then I’m downstairs and someone comes up to me and something.

(1c.) Extreme distress: “Total meltdown”. 13 of the 14 parent participants described extreme distress reactions presented by their sons, often, but not always, occurring as a sudden and unexpected response to a perceived present threat, rather than a future-oriented worry. Parents reflected on these reactions as
often seeming disproportionate to their trigger (“it’s usually something quite trivial that’s caused this massive meltdown” [P14]). Many parents believed their son found it harder to control his emotional responses than age-comparison peers.

“I think [A] is very emotionally immature, he has meltdowns like crying, screaming almost, and it could be that something happened to the iPad or something like that and I try to say, “You know you’re going to be 10, do you think 10 year old boys scream and cry like this?” You tend to think 10 year olds would know, but he just can’t seem to control that emotion. And he recovers quite quickly, but for that moment, it’s like the world is ending.” (P04)

Parents described a range of behavioural features presenting alongside ‘meltdowns’, including crying, panic, and aggressive behaviour. Sensory triggers causing extreme distress were mentioned by four parents and included crowded places, high noise levels, and the sensation of socks and shoes. Two of these four parents had sons with a diagnosis of ASD and a third parent’s son had been referred for an ASD assessment.

Two child participants (both in the 7-11 group), described extreme fear responses. One participant gave the example of bowls unexpectedly falling out of a cupboard and smashing onto the floor. He described his response in situations like this as, “I go like manic” [C04].

(1d.) The unexpected and unfamiliar: “Not in the plan”. Around two-thirds of parent participants described their sons’ need for predictable and familiar plans, and the anxiety and distress he experienced when this did not happen (often leading to ‘meltdowns’ described in the preceding theme). Several parents shared anecdotes of their son’s anxiety in response to new experiences or a change to his normal routine.
“Yeah even for us, like simple things, like if I pick [J] up from school, and then… “Where are we going now?” “We’re going home, oh no, I need to stop at the shop”, that’s a big deal. You say to him, “I’ll go to the shop, I’ll get back”, like you’re saying, but it’s not in their plan, and it could be something as trivial as you need to go and buy milk or something, but that is distressing for them.” (P04)

Child participants did not spontaneously mention this aspect of their anxiety, however, when shown the cards depicting different types of worry and asked to sort them into the relative sized worries, half of all child participants identified experiencing anxiety in relation to unexpected and unfamiliar situations. This was more commonly identified by 12-18 aged participants than 7-11 aged participants.

e.g. “Going somewhere new is a big worry.” (C05)

e.g. [When there’s a change to my normal routine] “That one’s going on a ‘big worry’.” (C07)

There appeared to be some overlap between anxieties about new situations and social anxieties (captured within theme 1e. Social anxieties: “Will I fit in?” where appropriate). For example, one parent said the following:

“We tried to get him into new social groups, which took a lot of building up to because of the anxiety thing” (P09)

Contributions from parents of older children and child participants in the 12-18 aged focus group provided indications that the frequency and severity of difficulties with unfamiliar and unexpected situations, and the occurrence of ‘meltdowns’, reduced for some children as they got older.
(1e.) Social anxieties: “Will I fit in?” All eight child participants made reference to social anxieties. Less than half of parent participants contributed to this theme. 7-11 aged participants predominantly identified social anxieties in relation to doing things in front of other people (e.g. “Oh I hate that [doing something in front of the whole assembly]…I don’t want to do it again.” [C03]) 12-18 aged participants more commonly referred to social anxieties relating to social comparison, particularly around appearance and friendships.

(C07): Yeah, about doing stuff that you sort of have to do in front of other people, like maybe in the swimming pool, being hoisted in and then there’s a…

(C08): Everyone looking at you.

(C07): Yeah.

(C05): Or like getting out of your chair and looking a bit weird because you can’t get up yourself.

Anxiety relating to visible differences was mentioned most often by boys in the 12-18 aged group and parents of boys aged 12 or over.

“…my son hates any signs of disability, he really struggles with it… and that is a source of anxiety for him (P06).

Conversely, the less visible nature of DMD for younger children appeared to provoke anxiety for some of the 7-11 aged participants.

e.g. “Meeting new people [is a worry] because they don’t know what’s wrong with you.” (C05)
e.g. “If he [Josh, fictional character] goes in [to hospital], he knows he has something, but then when he goes in, he’s like...people look at him like he’s not got anything wrong with him.” (C04)

(1f.) Anxieties about Duchenne-related change, needs, and medical procedures: “We’re the most vulnerable.” All eight child participants and 12 of the 14 parent participants talked about anxieties relating to growing older as an individual with DMD. Nine of the 22 participants mentioned needles or blood from medical appointments or clinical trials as anxiety triggers. One parent believed that a clinical trial had been “the start of anxiety” [P06] for her son due to the blood tests involved.

12-18 aged participants spoke about anxiety in relation to accessibility (e.g. “going to an event you haven’t been to before and you’re worried about will it have access, will there be accessible toilets?” [C07]) and causing harm to others with their wheelchairs (e.g. “I worry about running someone over” [C05]). Links were made by participants between anxiety about using a wheelchair, the responses of other people, and the wheelchair accessibility of the built environment. These links are discussed under themes (4) Social responses, and 5b. Wheelchair accessibility.

Four parents spoke about their sons’ anxiety about his physical ability in comparison to other children.

“He is afraid that some classmates will run into him because he has a lot of accidents happen, that they push him, he can’t stay properly because we know his muscle is not like ours.” (P10)

12-18 aged participants shared anxieties about their physical vulnerability in several hypothetical scenarios such as being in a lift that breaks down, escaping a terrorist attack or fire, being robbed, and being attacked by a bully (e.g. “Yeah, you can’t do anything really can you? So if someone took my phone now, I couldn’t go after
them” [C05]). Several parents said their son had expressed anxieties about his physical decline and the implications of this for his future, including worries relating to death.

“*My eldest talks about dying and stuff and he doesn’t want to die, he wants to live to 100 and all this and gets really anxious about it…when he [other son with DMD] did the North Star score (an assessment of ambulation), last time I think he was about 15, this time he was about 9. This time he couldn’t do half of the things they wanted him to do and he got a bit anxious over it because he knows what’s coming which is really difficult.*” (P13)

Child participants did not mention physical decline or death. One child worried about the implications of his physical ability on future employment.

“*Like what are you going to do for your job because we’ve got weak muscles and we can’t do jobs that other people do.*” (C05)

All 12-18 aged participants spoke about anxiety in relation to school work and exams, reflecting a typical anxiety experienced by many adolescents without DMD. Academic anxieties were not mentioned by 7-11 aged participants. Only one parent mentioned academic anxiety.

Many parents commented on how their son worried less about DMD than they expected, although many parents also commented that they find it hard to know how much their son worries about his condition.

(2) Individual context. Parent and child participants identified a wide range of individual factors existing in relationship with anxiety. Some of these factors reflected the impact of anxiety on the individual, whereas other individual factors appeared to mediate anxiety. Individual factors were often connected by participants
to other subthemes, such as family responses, social responses, and environmental responses.

(2a.) Individual emotional and physical responses. Child participants talked about the physical impact of feeling anxious including “butterflies in your tummy” [C04], aching muscles, headaches, and “feeling puffed out” [C05] (described as a combination of physical and emotional fatigue). Other physical functions reported as impacted by anxiety were sleep, toileting, and vomiting. These physical affects appeared to reciprocally mediate anxiety for some children. For example, some parents spoke about anxiety negatively affecting their son’s sleep and how greater levels of tiredness were then likely to further increase his anxiety. One parent described her son vomiting every morning in the car on the way to school due to his anxieties about school.

Several parent and child participants made connections between anxiety and anger. Four child participants described anxiety, (and other feelings such as guilt), being triggered by anger-driven behaviours (“Angry leads to being afraid.” [C04]).

(C07): I’ve destroyed a lot of things

(C08): It’s like you don’t stop, you just carry on going and then after you’ve done it or after your parents start crying you’re like ‘oh, what have I done?’

(C05): Yeah

(C07): And then you feel awful about it often

(C08): Yeah

(C05): Yeah
While some participants described anger-driven behaviours triggering anxiety, other participants described these behaviours as triggered by anxiety (discussed further under theme 2b. Individual adapting, coping and responding).

(2b.) Individual adapting, coping and responding. Both parent and child participants shared diverse ways that boys with DMD adapt, cope and respond to anxiety and its effects. Participants identified responses they considered more and less helpful to their wellbeing. Responses considered unhelpful were more commonly shared by parent participants, whereas child participants were more likely to share ways of coping they considered helpful.

Several child and parent participants described behaviours often associated with anger, enacted in response to anxiety.

“He bottles it all up inside [the anxiety] and then it comes out in anger or he mucks around.” (P13)

Child participants identified “taking it out on the ones you love” [C08], shouting and swearing at others, and throwing objects in response to feeling anxious.

Several parents commented on their sons’ avoidance of talking about or thinking about his condition as a response to his anxiety. Strategies such as avoiding information about DMD, withdrawing from social situations, sabotaging potentially exposing physical activities, and hiding signs of his condition were all mentioned.

“…even when my parents are over, he had to take the meds in another room, he doesn’t want them to see it. And when he had his biopsy scars, he was going a week later on a camp, I had to go in and talk to them, it was good friends he was sharing a room with, and say, “He’s just had some little cuts on his arms, don’t mention it.” So he’s happy that they know, but he doesn’t want
to have to talk to them about it and he does not want to talk about Duchenne.”

(P06)

12-18 aged participants spoke about practical strategies they used to cope with anxiety and anxiety-provoking situations. Strategies included the use of humour, playing computer games to calm down, focusing on the present rather than the future, finding thoughtful ways of confronting others when they stare or ask personal questions, cultivating acceptance around what they can and can’t do, and “instead of thinking about what you don’t have, thinking about what you do have” [C05].

“I don’t really find it a worry [not being able to do things that other people can do], because I understand that sometimes I’m not going to be able to do everything, that sometimes it’s ok to let people do stuff that you can’t do.”

(C07)

Several parents acknowledged the adaptive abilities of their sons to “take it in their stride” [P11]. One parent felt it was a shame these adaptive and resilient qualities were not captured by anxiety questionnaires and thought more attention should be given to strengths and skills.

(3) Family context. Participants identified a wide range of family factors related to the anxiety experienced by the child with DMD. Some of these factors described the impact of this anxiety on family members, whereas other family factors appeared to mediate the anxiety experienced by the child.

(3a.) Family emotional and physical responses. Parents shared the emotional impact of their son’s anxiety and its consequences on family members. They described feeling helpless, and finding his frequent questioning and difficulties with spontaneity “wearing at times” [P14].
“You feel helpless, you can’t switch it off, you can’t tell him to stop, “Stop it, you’re being silly”, you can’t do that, because he’s not going to go, “Oh yeah, I’m being silly, I’ll stop.” It’s out of control, it’s not something you can control, so as a parent, we already can’t control the fact that our kids have Duchenne, it’s just out of control. It’s something else that is out of control as well as all the stuff with Duchenne. (P0)

The impact of the wider condition of DMD on family members was also relevant to the anxiety experienced by the child with DMD. For example, some parents considered the possibility that their own feelings about their son’s condition, such as feelings of guilt, helplessness, feeling ‘heartbroken’, upset, anxious, and physical and emotional exhaustion, could all impact upon his wellbeing, including on his anxiety.

“…it’s hard not to sometimes let them know that you get fed up with it and that obviously has an impact because I think we’re anxious as well about some things.” (P1)

Parents also described the impact of an anxious family environment on all family members, including the child with DMD.

“Duchenne contaminates everything and then the anxiety I feel contaminates everything as well. It’s pervasive, even when it’s not spoken about and sometimes it’s more tangible than others, but it’s always there. In our house there’s a low level of anxiety every minute of every day and every night. It’s just…I think it’s probably toxic really.” (P3)

(3b.) Family adapting, coping, and responding. Parents described the impact of their son’s anxiety on family activities and described this as a daily issue for the family to adapt and respond to.
(P01): …the anxiety is more of a disruption day-to-day, more of a worry, not a worry, no, not a worry, nothing’s worse than Duchenne but…

(P03): It’s obvious, more obvious

(P01): It’s more obvious, it’s more disruptive to our day-to-day life than Duchenne because there’s so many things…

(P09): It’s every day.

Parents spoke about the different ways in which they draw on their strengths and skills to respond to their son’s anxiety and the dilemmas and uncertainties they face when trying to work out “the right thing to do” [P08]. The impact of anxiety on family relationships was reported to include arguments between parents about different styles of approaching their son’s anxiety, and managing the frustrations of siblings who received less parental attention. Parents noticed that the same strategies for responding to anxiety experienced by their other children were not as effective when applied to their son with DMD.

“And I think with the others (siblings) I would have just walked away and… ‘this is what’s happening, we’ve talked about it’ and it would have been fine, whereas it just seems to go on [with B], we don’t get to do it.” (P08)

Ways of responding and adapting to the anxiety varied between families. Some parents noticed that their child was less anxious when they made detailed plans about activities in advance, whereas other parents reduced their son’s anxiety by being “vague” about plans and waiting until the last minute to share more details. Other parenting responses to anxiety included ignoring the anxiety, (parents generally found this more effective with younger children), having conversations with their son about his anxiety, encouraging their son to face his fears, avoiding
situations that could make their son feel anxious, “picking battles” [P01], finding creative ways to increase their son’s resilience, and hiding their own emotional responses (e.g. “he’d fall over and sometimes we wouldn’t even say anything, because we didn’t want to make him anxious” [P03]).

Many parents made links between the impact of their own feelings on their responses to the anxiety, and the circular relationships between these responses and their son’s anxiety. For example, one parent shared an example of talking to their son about his condition whilst trying not to convey their own parental worry so as to reduce the son’s anxiety.

“I wonder if it felt much more relaxed at that point of understanding that there was a difference, there was an explanation for it [DMD], that we [parents] seemed to understand what it was, we didn’t seem overly worried about it, you know, you try and get that across without making him worried. I guess, parental anxiety as well, that sort of comes into all of this and how you engage with your son.” (P11)

(4) Social Context. Participants spoke about how the responses of other people beyond the nuclear family affected their son’s anxiety through direct and indirect pathways.

(4a.) Stigma, misunderstanding and unhelpful responses from others: “They don’t understand.” Being bullied was mentioned by six of the eight child participants. “Bullying” was not mentioned by any parent participants, although one parent mentioned their child being “picked on” [P01], and two parents spoke about their son not being invited to sleepovers anymore, which they believed had negatively impacted his emotional wellbeing. 12-18 aged participants connected current social anxieties (captured within theme 1e. Social anxieties), to past experiences of bullying. Experiences of bullying shared by participants included
people throwing things out of their reach, turning their wheelchair on as a prank, throwing things in front of their wheelchair, offering them money to drive their wheelchair down stairs, and threatening them.

(C05): And people say ‘if you run me over then I’ll pull you out the chair’

(C08): Yeah, or ‘I’ll call the police on you’.

Parent and child participants shared experiences of unsupportive and stigmatising responses about DMD from friends, family members, clinicians, and members of the public. Three parents talked about “losing” friends and family members following their son’s diagnosis. Parents spoke about the negative impact on their own emotional wellbeing of people who labelled their son’s anxious behavioural responses as “stubborn” or “naughty”.

(4b.) Support and solidarity: “You’re not on your own.” Parent and 12-18 aged participants spoke about supportive others and the positive impact this had on their emotional wellbeing. Supportive others were not mentioned by 7-11 aged participants.

12-18 aged participants described supportive others as people who help them, people who include them, and people who understand their condition. One participant shared how the support he had gained from the focus group itself would make a situation he previously found anxiety-provoking easier in the future.

“I normally get a bit scared when like someone comes up to me and asks me questions [about my condition], but today everyone’s made me feel like I don’t need to do that, I can just tell them exactly what it is.” (C05)
Parent participants spoke about the positive impact of supportive and understanding responses from teachers, strangers, fellow DMD parents, and other children. They noticed how such responses could reduce their son’s anxiety, for example, in relation to his school work.

“I think things like deadlines and just things like, there’s lots of questions about school work and stuff, I think there’s just more so than other kids, I mean other kids are scared of doing tests aren’t they but I just think that he has had to have a lot more support to get to those deadlines and not feel completely terrible about them, but I suppose they’re just managed by people being aware of his condition.” (P14)

Parent participants recognised that supportive responses from other people, including online communities, improved their own wellbeing. Some parents connected their own wellbeing to their ability to cope with their son’s anxiety. Parents said that despite “losing” some friends and family members, they had also “gained” relationships as a result of their son’s condition, particularly with other DMD parents.

(5) Environmental context. Participants made connections between several material and public service environments and their son’s anxiety.

(5a.) Medication. The impact of steroid and sleep medication was mentioned by five parents. Sleep medication was not explicitly linked to anxiety by participants, however, some parents believed sleep problems were a symptom of their son’s anxiety. Some parents noted the emotional and behavioural impact of steroid medication on their sons and how they adapted their parenting responses based on this medication. According to parent participants, steroid medication, for some children, seemed to increase several of the emotions and behaviours that had
been linked to anxiety in other parts of the focus groups, such as extreme distress, physical aggression, and irritability.

“When he is on [steroids], you can’t say anything. You must be aware of what you talk about with him, how you talk with him because he is very…he starts to cry and he makes…. you know…and we know that he is on steroids. “Sshh! Zip it!” We are much quieter…We do not say “you cannot go”, because he will get angry and say “ooh why not?” And the school, they notice as well. When he is on [steroids], he is… “I want to do that, I want to do that, I want to do that.” When he is not on [steroids], “ahhh, no, no”, he is very quiet.” (P10)

(5b.) Wheelchair accessibility. The wheelchair accessibility of different environments was mentioned in relation to anxiety by five child participants (including all 12-18 aged participants), and four parents. The low number of parent contributors to this theme may have been influenced by there being only three parents within the sample with children over the age of 12 using wheelchairs on a full-time basis.

Participants made links between wheelchair-related anxieties (captured within theme 1f. Anxieties about Duchenne-related change, needs, and medical procedures), and wheelchair accessibility. The wheelchair accessibility of different environments appeared capable of increasing or decreasing the anxiety experienced by boys in wheelchairs and parents. For example, participants described poor experiences of wheelchair accessibility affecting the anxiety they subsequently experienced when visiting somewhere new. 12-18 aged participants described poor wheelchair accessibility in busy places increasing their risk of accidentally hitting people, which linked to worries they had mentioned captured within theme 1f. Anxieties about Duchenne-related change, needs, and medical procedures. Wheelchair accessibility also appeared capable of influencing parental
anxiety (captured within theme 3a. *Family emotional and physical responses*), which some parents linked to their son’s anxiety.

“I took P to Nottingham on my own earlier in the week and it was really stressful because you’re going to a hotel, you don’t know what it’s going to be like, you don’t know what there’s…and in fact, it was a bit of a nightmare for me because it was a lot of physical work and he’s anxious about being hoisted and everything in the new place, but yeah, so I can understand how anxious you’d be, all that kind of stuff, and it’s hard as a parent because you can’t go “well it’s all going to be fine” because when we get there you don’t know, because it isn’t like that, and you can’t kind of ring up and make someone give you a virtual tour of your hotel room or whatever, and their idea like someone was saying about accessibility is very different.” (P14)

Participants noted the high prevalence of non-wheelchair accessible environments, and highlighted shortcomings in definitions of ‘accessibility’. Both parent and child participants mentioned the accessibility challenges of transport options, such as buses without enough space for wheelchairs when a pushchair was present, trains where the ramps were not called ahead for, and family cars that needed to fit a wheelchair.

(5c.) *Access and quality of schooling and healthcare “A postcode lottery.”* This theme captures contributions from 11 of the 14 parent participants and one child participant about the quality and availability of school and healthcare services for families, and the relationships between these factors and the anxiety experienced by individuals with DMD.

Parents shared both positive and negative experiences of accessing high quality educational and healthcare services. One parent reflected on the culture of inclusivity at their son’s school and how this had enabled their son to enjoy school without problematic levels of anxiety. One parent shared their experience of a
school meeting arranged in response to their son’s anxiety in the playground, which decreased after a plan from this meeting was implemented. Three parents shared experiences of struggling to get therapeutic support for their son at school, and noticed that “[schools] listen to professionals, they don’t listen to parents” [P13].

Four parents had accessed therapeutic services for their son to support his emotional wellbeing. Therapeutic services had been accessed through NHS paediatric services, NHS CAMHS (Child and Adolescent Mental Health Service), a hospice, school, a charitable trust, and private therapy. Some parents experienced therapeutic services as easy to access and helpful. Other parents described having “the opposite experience” [P09], and having to “push” [P01] to receive therapeutic support for their child. Data collected prior to the focus groups indicated that over half of parents who had wanted support around understanding and managing their son’s anxiety had felt some level of dissatisfaction with what they had been received. One parent described the therapeutic input on offer (one family therapy session every 6 months) as “a drop in the ocean” [P01] in comparison to the family’s emotional needs and her experience of “treading water” [P01] as a result. Three parents spoke about the lack of professional therapeutic support available for parents and the negative impact this had on their own wellbeing, which in turn could affect their ability to support their son. Several parents agreed there should be “mandatory counselling for parents at diagnosis” [P06].

“I’m not a big anxiety person but X has brought more anxieties out…I think there’s no support for parents in terms of helping you deal with your own anxieties to then help the boys. We all deal with it in our own way which internally we try and get on with.” (P09)
Respondent validity

Four parent participants and two child participants provided validity checks. Examples of how their feedback influenced the analysis include the following: One parent highlighted the importance of reflecting the strengths, abilities and resilience of individuals with DMD and their family members through the language used in the thematic framework. The language of ‘adapting, coping, and responding’ was therefore used within two of the subtheme titles. Another parent suggested naming ‘school’ and ‘healthcare’ as specific environments relevant to understanding anxiety. The previously named subtheme, ‘Structural and material factors’, was consequently replaced with ‘Access and quality of schooling and healthcare’. One of the child participants said that unhelpful responses from other people were often unintentional in nature and best captured with the word, ‘misunderstanding’. This word was subsequently included into the relevant subtheme title.

Discussion

To our knowledge, this is the first study dedicated to exploring the anxiety experienced by males with DMD. The study aimed to gain a rich account of: i) the characteristics of anxiety experienced by males with DMD ii) the impact of this anxiety on males with DMD and their families. According to the participants in this study, anxiety is a prevalent and important issue amongst the DMD population. Despite individual differences in anxiety presentation, particular anxiety characteristics were recurrently reported by study participants, suggesting a DMD-specific anxiety phenotype. The impact of anxiety is experienced in equally diverse ways by individuals with DMD and their family members. Anxiety appears to impact a network of interacting factors across family, social and environmental contexts, but also appears impacted by this network of factors. The responses of family, social and environmental contexts to the individual's anxiety and their wider condition are
important considerations for understanding the anxiety experienced by this population. Discussion of these select findings aims to map characteristics of anxiety in DMD and its relationships to other contexts. A model is provided outlining a set of hypotheses for understanding the ways in which anxiety relates to these wider contexts.

**A prevalent and important issue in DMD**

Whilst the research design of this study was not devised to assess anxiety prevalence, the data were highly suggestive of anxiety as a pervasive and impactful issue within this population. Every parent participant identified observations and challenges relating to their son’s anxiety and its consequences. Every child participant identified first-hand experiences of anxiety. This is in line with current research that has found a high prevalence of anxiety amongst DMD boys (Banihani et al., 2015; Colombo et al., 2017; Ricotti et al., 2016).

In contrast to our observation of widespread anxiety amongst the child participants and children of parents who participated in the study, it is notable that few scored above the clinical threshold on quantitative parent-rated anxiety measures. Studies using standardised measures within this population may therefore underestimate the prevalence and impact of anxiety; a notion consistent with anecdotal evidence available prior to the study. Standardised tools measuring the effectiveness of interventions targeting anxiety may also provide inaccurate markers of the success of these interventions. Further investigation is therefore warranted into the sensitivity of commonly used screening tools for measuring anxiety within this population. Quantitative research investigating anxiety in this population must carefully consider the tools used to measure this.
The possibility of a DMD-specific anxiety phenotype

The presence and experience of anxiety varied between individuals and families, however, the recurrent reporting of particular anxiety characteristics in the study points towards the possibility of a DMD-specific anxiety phenotype. This finding offers one possible explanation as to why anxiety in this population may not always be effectively captured by standardised measures.

Anxiety characteristics recurrently mentioned were: the presence of catastrophic conclusions, rigidly held anxieties, extreme distress reactions, anxiety about unexpected and unfamiliar situations, social anxieties, and anxiety relating to Duchenne-related change, needs, and medical procedures. Parents noticed differences between the anxiety experienced by their son with DMD and the anxiety experienced by other children, including siblings, adding to the possibility that anxiety experienced by males with DMD has particular hallmarks.

Several of these anxiety characteristics share similarities with traits common to individuals with ASD, such as an inflexible adherence to routines, extreme distress at small changes, rigid thinking patterns, and high levels of social anxiety (American Psychiatric Association, 2013; Spain, Sin, Linder, McMahon, & Happé, 2018). Reports of these characteristics were not limited to children with ASD diagnoses or parents of children with ASD diagnoses. Indeed, quantitative data collected by the researchers found that 56% (n=9) of children for which data was gathered reached a clinical cut-off on the Social Communication Disorders Checklist (SCDC) (Skuse et al., 2009), whereas only 25% (n =4) of these children had an ASD diagnosis (38% if one “borderline” diagnosis and one child referred for assessment are included). This observation fits with research which suggests that although most children with DMD do not reach a diagnostic threshold for ASD, many have traits associated with ASD (Darke, Bushby, Couteur, Le, & McConachie, 2006).
The trend of inflexibility noted across many of the identified anxiety characteristics may be influenced by impairments in executive functioning associated with DMD (Snow et al., 2013). However, difficulties with cognitive and behavioural flexibility, and the relationship between these difficulties and anxiety have not been the focus of any research studies to date in this population. This is an area that would benefit from further research, including the use of executive functioning measures as part of investigations.

The prominence of social anxieties within the data supports extant literature reporting high rates of social problems amongst individuals with DMD (Donders & Taneja, 2009; Harper, 1983; Hinton, Nereo, Fee, & Cyrulnik, 2006; Siapno & Carter, 2013; Snow et al., 2013). Links have been made between social anxiety and the lack of dystrophin isoforms in the central nervous system (Hinton et al., 2006), and between increasing physical limitations and social participation (Harper, 1983). However, social anxiety is one of several areas within the study where the data highlight the need to recognise the influence of personal, social and environmental responses to individuals with DMD on the presence and nature of social anxiety they experience.

**Multiple interacting relationships with anxiety**

The study’s investigation into the characteristics and impact of anxiety experienced by the DMD population found data supporting the presence of multiple interacting factors that were both influenced by and influential over the anxiety being investigated. Boundaries between causes and consequences often appeared blurred as a consequence, and the study’s investigation into the impact of anxiety became inextricable from the consideration of factors affecting anxiety.

Although the term ‘impact’ was used in the original research questions, feedback from parents, alongside the findings of the study and the position of the researcher led to a decision to replace this with ‘responses’ within the titles of some
themes and subthemes. This word was deemed to better reflect the active nature by which individuals, families and wider systems described coping with and adapting to anxiety (irrespective of the helpfulness of these responses), and the circular relationships identified between anxiety and other factors. The consideration of circular rather than linear relationships also positions anxiety as a coping response in itself, fostering exploration into the multiple challenges faced by individuals with this condition beyond the direct consequences of their genetic mutation.

The data suggest that the anxiety experienced by individuals with DMD elicits a wide range of responses from the individual, the family system, and wider social and environmental contexts. These responses appear capable of reciprocally influencing anxiety in this population, although specific influential factors are likely to be highly individualised. For example, a parental response to anxiety, such as making plans with their son in advance, or finding age-appropriate ways to talk about their son’s condition with him, may reduce anxiety for some boys with DMD. Other emotional responses of parents to their son’s anxiety, such as exasperation, anxiety, or emotional and physical fatigue may, at times, present challenges to parents for responding helpfully to their son’s anxiety. The study supports links made by extant literature of reciprocal relationships between child and parent wellbeing in DMD families (Fee & Hinton, 2011; Nereo, Fee, & Hinton, 2003; Thompson, Zeman, Fanurik, & Sirotkin-Roses, 1992). It is worth considering that responses to anxiety, including parental responses, appear themselves to be influenced by a wide range of factors, such as the availability of social and professional support.

The data highlights the influence on anxiety of responses to the wider condition of DMD from the individual, family members, and social and environmental contexts. For example, poor wheelchair accessibility, stigma, bullying and discrimination can all negatively affect anxiety. Conversely, responses such as inclusive school cultures, wheelchair-friendly environments, and people who
understand the condition appear capable of positively affecting anxiety. These issues connect to wider systemic factors beyond the individual with the condition and their family, encouraging consideration of social and cultural values attached to disability and difference. Such associations are supported by research, including findings that increased social networks significantly contribute towards positive child adjustment in DMD (Fee & Hinton, 2011), and that social support is a protective factor for resilience amongst carers of individuals with DMD (Glover, Hendron, Taylor, & Long, 2018). Existing research also shows that people with disabilities are at greater risk of violence than people without disabilities (Dammeyer & Chapman, 2018), however, the researcher is not aware of any large-scale studies to date investigating the relationship between disability-related discrimination or violence and emotional and behavioural difficulties such as anxiety, although a negative impact is hypothesised. Further research investigating relationships between anxiety in DMD and these wider contexts is required to develop a richer understanding of complex systemic influences.

The study provides tentative indications that age plays an influential role in the presence and nature of anxiety in DMD. For example, social anxiety amongst 12-18 aged participants appeared more strongly connected to issues of social comparison than for 7-11 aged participants, particularly with respect to appearance. This trend reflects typical adolescent development and the characteristics of anxiety commonly experienced by adolescents who do not have DMD (Carr, 2016). An interaction between normal developmental processes and the consequences of DMD may therefore take place and be influential over anxiety. Due to the low number of parent participants with children aged 12 or over, and only four child participants aged 12 or over, further research investigating the influence of age on anxiety experienced by individuals with DMD is indicated.
A model of multiple interacting factors

Using the findings of the study alongside extant research, the researcher proposes a model of multiple interacting factors relevant to understanding the anxiety experienced by individuals with DMD (Figure 2). Anxiety may be a biologically determined consequence of DMD, a secondary adjustment response to the condition, a response to familial, social and environmental responses to DMD, or a combination of these factors. Circular interactions are likely to take place between these factors as indicated by the data. As such, the consequences and causes of anxiety are unlikely to be mutually exclusive. The model also highlights the added layer of complexity around measurement. Anxiety may be present, but the way it is measured affects its identification. Whether anxiety is identified or not in turn affects the nature and likelihood of intervention.

**Figure 2. A biopsychosocial model of anxiety in DMD**

The model provides a guiding framework for understanding the anxiety experienced by this population. It represents a set of tentative hypotheses and it is
important to acknowledge that a lot remains unknown about the relationships proposed. The model does not claim to map the full extent of anxiety in DMD and its complex relationships with other factors. Arrows do not reflect fixed pathways of causation but possible pathways of influence.

**Methodological strengths and limitations**

To the author's knowledge, this is first study dedicated to anxiety in this population. It is also the first study to use qualitative data from boys under the age of 16 with DMD. The findings from this thematic interpretation offer an original contribution towards the development of a comprehensive understanding of the characteristics of anxiety commonly experienced by males with DMD and the range of factors influenced by and influencing this experience. The study brings a psychological and systemic approach to this topic where biological approaches have predominated. Focus groups offered an effective way of eliciting this information from participants. For example, the researchers noticed many instances where the contribution of one participant triggered contributions from other participants relevant to the discussion.

Methodological limitations relate to the study sample. Given the exploratory nature of the study and the relatively small sample recruited, it was beyond the scope of this study to map the full extent of anxiety in DMD and its complex relationships with other factors. It is therefore likely that relevant themes and relationships have been missed.

The use of a database containing details of parents interested in taking part in DMD research alongside adverts published by charitable organisations were useful methods for recruiting participants. However, the self-selecting nature of the sample means the prevalence and severity of anxiety raised by participants may have been biased due to the study attracting participants for whom anxiety was a highly
relevant issue. Conversely, the prevalence of social anxiety amongst males with DMD may have created a selection bias amongst child participants. Such a bias may have reduced participation from boys for whom social anxiety was a problem, and consequently biased the data, particularly in relation to social anxiety.

The study also lacked representativeness across a number of demographics. No child participants or children of parent participants had a learning disability, despite the high prevalence of this diagnosis within the population. Amongst parent participants, only one father took part, and only three parents had children aged 12 or older. Considering the developmental differences of older children, added to the changes occurring with age for individuals with DMD, the themes identified may therefore have been skewed towards younger children, and the perspectives of mothers.

**Clinical Implications**

The study highlights a wide range of factors relevant to the consideration of anxiety experienced by this population and supports the recommendation made in recently published international guidelines for mental health and quality of life to be screened at each neuromuscular visit (Birnkrant et al., 2018). Incongruity observed between parent-rated scores on standardised anxiety measures and the prevalence and impact of anxiety indicated by the focus group data suggests the utility of a clinical interview alongside standardised measures as part of a comprehensive assessment, particularly while the SDQ and RCADS remain unvalidated for use within this population. Ideally, a family-based psychological assessment is conducted, considering characteristics identified by this study as common to a DMD-specific anxiety phenotype, and considering the influence of broader contextual factors identified as relevant to understanding anxiety experienced by an individual with DMD, such as their family context, social, healthcare, school, and other relevant contexts.
Findings from the study indicate that the anxiety experienced by individuals with DMD can be impactful upon parents in many ways, in addition to the challenges presented to parents by other aspects of the condition. Considering these findings alongside those already published, such as the positive impact of decreased parental stress levels on child adjustment in DMD (Fee & Hinton, 2011) and links between parental stress and child wellbeing (Nereo et al., 2003; Thompson et al., 1992), an assessment of parental wellbeing and adjustment should be an integral part of a comprehensive anxiety assessment and formulation with this population.

Relationships identified within the data between anxiety and wider contextual factors offer a range of targets amenable to psychological intervention, increasing the possibilities for improvements across domains where medical interventions may be unable to effect change. Interventions may be effective at a variety of systemic levels and should be guided by thorough assessment and formulation. For example, psychological intervention may be most impactful if targeted at school management or parent wellbeing, rather than the individual, if this is indicated by the formulation.

A deficit-focused approach is dominant within DMD research and clinical spheres, yet individuals with this condition and their family members regularly draw on many strengths and skills in ways that positively affect anxiety and their wider wellbeing. Exploration of these factors should be part of any assessment and formulation regarding anxiety. Acknowledging, harnessing, and developing these protective factors is likely to be an important aspect of any subsequent intervention.

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Part 3: Critical Appraisal
Introduction

A fundamental aspect of good quality qualitative research is the researcher’s transparency and reflexivity with respect to methodological decision-making and their relationship with the data (Meyrick, 2006). This appraisal therefore reflects on several influences and observations deemed important to the research process within this study. The appraisal firstly discusses an observation made about the dominant model through which anxiety experienced by individuals with DMD is understood. Possible reasons for the dominance of this model alongside potential implications for DMD families and the evidence base are considered. The appraisal then discusses the influence of a Liberation Psychology framework (Martín Baró, 1996) on the researcher’s theoretical orientation, and the impact this had on the analysis and write-up of the project. The principles of Liberation Psychology are outlined, and the ways in which individuals with DMD and their families may be ‘problematised’, disadvantaged, and marginalised are discussed through this lens. A Liberation Psychology frame is also used to understand the therapeutic quality of the focus groups described by many participants. The appraisal discusses the challenges and priorities of creating a model of anxiety in DMD, and the ways the researcher attempted to privilege the voice of participants throughout this process, whilst also exploring the researcher’s biases and the scope of conclusions.

Observations of a dominant model of anxiety in DMD

The request for this research came from a team exploring genetic, medical, and pharmacological interventions aiming to improve and lengthen life for individuals with DMD. The ultimate aim of this team is to find a genetic cure for the condition and accurate ways of measuring the success of their interventions across a wide range of domains impacted by the condition, including neurodevelopmental, emotional and behavioural concerns. The research team is particularly interested in
developing sensitive parent-report tools as one way of assessing whether their medical interventions are making a difference to psychological difficulties. A decision was made for the study to focus on anxiety to ensure the scope of the project remained realistic in the time frame available.

I became aware that anxiety was being considered as an organically-driven problem within this research team, and while I did not dispute the possibility of an organic contribution to anxiety in DMD, my clinical psychology training encouraged me to think more widely about the contexts and challenges faced by these families, and the many factors that could be involved in an experience of anxiety amongst individuals with a chronic, fatal, and debilitating condition.

I was not familiar with DMD before taking on this research project and so I sought different ways to introduce myself to the condition and the various communities connected to it. Attending a national conference aimed at both the academic and patient community was a good way to do this and I was introduced to families, researchers, clinicians, equipment specialists and charity workers at this event. Around 30 academic posters had been selected for presentation at the conference and I was struck by finding only one poster that mentioned emotional or behavioural difficulties. This poster published prevalence rates of emotional and behavioural difficulties alongside prevalence rates of intellectual disability, ASD and ADHD under the category of ‘Brain-based disorders’. I came to understand that the role of dystrophin in the brain and its implications for individuals with DMD is a relatively infantile area of DMD research in comparison to others, and one that is generating increasing interest amongst DMD communities. I did not find any posters at the conference which explored the experiences or challenges of living with DMD from the perspective of individuals with the condition or their family members.

5 This poster won first prize for posters presented at the conference.
Although parents and some children with DMD were present at the conference, the voice of those directly affected by DMD seemed missing from the research being presented. As I investigated the research further for my literature review, the trend I had observed at the conference appeared reflected in the DMD literature more widely.

A paucity of qualitative research in the field of DMD research may be influenced by the understandable drive to find life-lengthening medical treatments and a biological cure for this condition, however, I wondered whether the experiences and challenges of living with this condition were being eclipsed as a result. The poster exhibition at the conference had not given me a sense of what it was like to live with DMD, and I wondered whether an assumption was held that families living with DMD, and particularly children with DMD, were unable to provide perspectives considered valuable to the priorities of the research community.

During the recruitment phase of the project, I contacted around 200 parents by telephone and email about my study. Parents who I spoke to almost unanimously told me that anxiety was something they recognised as a problem for their son and a challenge for their family. Irrespective of whether they were able to take part, parents welcomed the research and wanted to be kept informed about my findings. Whilst I felt encouraged by these responses, I regularly thought back to the conference poster exhibition and became increasingly aware of a mismatch between the challenge seemingly presented to these families by anxiety and other emotional and behavioural difficulties, and the level of attention being given to these issues within the research. Comments from some parents suggested this mismatch was also reflected in the clinical services available to support them. I had already decided to use focus groups as my research method for exploring anxiety in this population, but it seemed increasingly important to capture the voice of families in my study and to use the research as a platform for their experiences to be heard more widely.
Findings from the empirical study also supported many of my original observations that anxiety amongst this population is given disproportionately little attention. For example, experiences were shared by parent participants of feeling confused about their son’s anxiety in the absence of support to help them make sense of this difficulty. Several parent participants spoke about the “postcode lottery” of trying to access appropriate emotional and behavioural support, including support for their own wellbeing. The range of factors proposed by the study as relevant to anxiety offer many possible intervention targets for improving quality of life within this population. These are interventions that can take place alongside the important pursuit of a cure for the condition and one of my primary reflections from the project is that there is room and need for both. It is important that the outcomes used as markers of progress in this field reach beyond those around halting disease progress, prolonging life expectancy, or finding a cure for the condition and consider broader factors such as quality of life, parental wellbeing, adjustment, relational opportunities, and emotional and behavioural difficulties.

The influence of Liberation Psychology

The analysis and write-up of this project were heavily influenced by my third year specialist clinical placement which had a focus on ‘Liberation Psychology’. I began this placement shortly after completing the focus groups for my study and continued on the placement throughout the analysis and write-up phases of the project. Liberation Psychology describes a collection of practices which developed out of the ideas behind Liberation Theology in the 1960s (Freire, 1973; Martín Baró, 1996). A Liberation Psychology frame assumes that people thrive in the context of being heard and understood, and feeling that they have some agency in being part of the solution to problems they experience (Afuape & Hughes, 2016). Liberation Psychology starts with the assumption that emotional distress and difficulties in
behaviour emerge out of social, cultural and relational contexts that are restricting, harmful or oppressive (Burton, 2013). Rather than responding to people as problems to be fixed, a liberation framework positions people as having important things to say about a ‘problem’ of interest and people who can be part of the solution. Liberation psychology privileges the voices of the most marginalised and disadvantaged members of a system or community, assuming that by starting with the liberation of these members, through reflection and action, the wellbeing of the the whole of society is improved, and things change for the good of all those who are part of it (Martín Baró, 1996). During the project, I became increasingly aware of the ways in which my participant sample could be considered ‘problematised’, disadvantaged, and marginalised members of society. I felt curious about the impact these experiences could be having on wellbeing, including on the anxiety I was investigating.

‘Problematised’

Due to the physical, neurodevelopmental, emotional and behavioural complexities associated with DMD, this is a population who often receive many diagnoses and labels. Available research indicating the high prevalence of neurodevelopmental, emotional and behavioural diagnoses amongst this population, alongside contributions from parents during the focus groups which highlighted their experiences of a constant focus on their son’s problems rather than his strengths and abilities, offer evidence towards the idea that many within the DMD community experience being ‘problematised’. One parent said, “Just from a parent perspective, it’s very depressing sitting here looking at everything that is bad. It would be really nice to have a questionnaire that also had things like ‘my child responds well to X’…rather than ‘these are all the problems’, you know?”

Many of the child participants (and sons of parent participants) had been given diagnoses and labels beyond their diagnosis of DMD, such as scoliosis,
ADHD, dyslexia, and ASD, however, certain diagnoses were under-represented compared to the wider DMD population. For example, while some child participants (and sons of parent participants) had diagnoses of learning difficulties such as dyslexia, none of the participant sample had a diagnosis of intellectual disability, which around one third of the DMD population are given. Additionally, child participants with greater numbers of physical diagnoses, such as those relating to cardiac or ventilation problems, may have felt unable to participate in the study due to the limitations imposed by these problems. In these ways, the study sample may have been less 'problematised' than the wider DMD population, nonetheless, contributions from participants about the focus within their lives on ‘problems’, alongside available literature about the high prevalence of different diagnoses commonly given to this population both point towards experiences of being ‘problematised’.

**Disadvantaged**

Despite legislation aiming to improve equality, such as the Equality Act (2010), ongoing discrimination and societal inequality often place those who live with a physical disability and family members who care for them at a disadvantage (Dixon, Smith, & Touchet, 2018). Many participants shared examples of experiencing disadvantage with respect to educational and employment opportunities, wheelchair access (e.g. buildings, outdoor spaces, transport), and the negative stereotypes held by others about their value and ability. One study has identified significantly greater socioeconomic disadvantage at diagnosis amongst DMD families than the average of the population from which they were drawn (Bushby, Raybould, O'Donnell, & Steele, 2001).
Marginalised

Whilst not a review of all DMD literature, the literature review I conducted identified only 16 studies (out of a total of 210) using qualitative methodology to explore the perspectives of individuals with DMD or their family members. Of these, only six studies shared the direct accounts of individuals with the condition. I am not aware of any qualitative studies that have sought the perspectives of children under the age of 16 with DMD. Within this study's focus groups, child participants also talked about their experiences of social marginalisation, including through being bullied. Parent participants shared experiences about the marginalisation they experienced through services not considering their own wellbeing needs, and their experiences of school staff listening more to professionals than to them as parents. Considered alongside the conference poster exhibition, these observations further suggest that the voices of people directly affected by DMD are marginalised rather than privileged, particularly the voices of children with the condition.

These considerations about the context of my sample using the frame of Liberation Psychology influenced the theoretical stance from which I undertook the analysis of this project. While the focus groups enabled me to hear the voice of children with DMD and their parents, I prioritised the inclusion of their voices throughout the analysis of the study, seeking validity checks from both parent and child participants during this phase.

I also became aware of an assumption I held behind my second research question about the impact of anxiety on individuals and family members. My assumption was that anxiety must only have a negative impact on children with DMD and their families. Increasingly, my theoretical stance encouraged me to attune to the ways in which participants drew on their strengths, resources, and skills in response to the challenges posed by anxiety and the systems around them as I conducted the analysis. I began to question the language of 'the impact of
anxiety’, which I increasingly interpreted as implying a population who are passive recipients of a one-way linear relationship with anxiety. In consultation with participants, the language of ‘impact’ was replaced with language such as ‘adapting’, ‘coping’, and ‘responding’, to better reflect the active nature by which participants described responding to anxiety. As one parent said in their feedback, “this is not just a story of tragedy”. With these thoughts in mind, it was important to acknowledge and highlight the challenges and struggles of these families with respect to anxiety in my write-up, but also to avoid a wholly ‘problematised’ story of their lives and to bring attention to the resourceful and varied ways with which they actively face the challenges presented to them.

The ideas of Liberation Psychology also gave me a helpful perspective from which to make sense of the therapeutic quality of the focus groups, which was reported by many of the participants in their reflections about participating in the study. For example, one child reflected that the discussions in his focus group had supported him in ways that would help him manage anxiety in the future. Several parents also suggested hiring out a room on a regular basis to talk in a similar way together again in the future. I wondered whether these benefits had been influenced by the focus group creating a context where previously marginalised voices had been positioned as expert perspectives from which much could be learnt, and that through being heard and understood, the wellbeing of some participants had been enhanced. The fact that participants had ideas arising from the focus groups, such as connecting together again, seemed to closely mirror the process of liberation described by Freire (1973). Freire proposes that when people critically reflect on their experiences, their experiences become visible, a new critical consciousness emerges, and new possibilities for action arise that are liberating for all participants. Reflections from some participants drew particular attention to the power of the collective aspect of the focus groups. One 12-18 aged participant said, “It’s like,
because we’ve all got the same condition, it feels like we can share anything.” One parent said the following at the end of their focus group:

“That’s my greatest sense of comfort that, that I’ve got you and you and you and you and all the other mums and I just look at you and I know your pain, because your pain is my pain and so is yours and yours and yours and everyone else’s because you’re the mother of that child, you’re the mother of those children and nobody knows what it’s like to love that child as the mother, only the mother, and that’s why I feel such an affinity, such a ‘oh my God, I’ve got your back cos’ you’ve got mine’, and you know how shit it is, because they’re our babies, and nobody else knows that.”

These observations indicate the potential value of peer support groups as a therapeutic intervention that may be beneficial to this population.

**Creating a model of anxiety in DMD**

I was particularly struck by the confusion and frustration expressed by parents in all three parent focus groups arising from trying to make sense of their son’s anxiety in the absence of an explanatory framework or professional support to assist them with this. For some parents, participation in the study appeared motivated by the very possibility of better understanding their son’s anxiety through taking part. Parents regularly wondered aloud during the focus groups about whether their son’s anxiety was caused by a lack of dystrophin in his brain, or other factors, such as autism, the challenges of his condition, or their own parental anxiety. On two occasions, parents directly asked the researchers to explain the causes of anxiety in DMD. A further parent said they would “find solace” in seeing a visual map explaining anxiety in DMD, including common characteristics of this anxiety. The original aim of the study was not to answer the question of why individuals with DMD experience anxiety, although the study was designed in such a way as to yield
insights in this area. Hearing this difficulty expressed by parents, and trying to make sense of it therefore seemed important endeavours as part of the research process.

Humans intrinsically seek concrete explanations and hold an aversion to the unknown (Kruglanski, 1989), however, it is important to consider the possible influence of the medical context within which the DMD population are unavoidably embedded when attempting to make sense of the strong desire for an explanation to anxiety. Medical systems are largely diagnosis-driven and routinely use tests to give definitive answers about the presence, absence or level of a problem. These experiences, which may represent a norm for many of the participants, may encourage parents to expect similarly definitive answers about the anxiety they notice their son experiencing.

It is also important to consider the influence of the medical context on the research and the researcher, considering that the study was conducted within a medically-led team who were predominantly interested in the organic basis of emotional and behavioural difficulties. This environment had the potential to influence or even bias the findings. For example, greater weight being given to organic or ‘pathological’ explanations for the anxiety presented by this population, rather than a more psychological assumption that anxiety is to be expected within the context of the challenges and circumstances faced by this population and that anxiety serves many adaptive functions. Several measures were taken to avoid unhelpful bias. These included regular supervisory and consultative conversations with both medical and psychology colleagues where the study design, data interpretation, and presentation of findings were discussion. Secondly, I prioritised being guided by the voices of the participants throughout the project. The resulting biopsychosocial model of anxiety in DMD evolved through feedback from the participants sample and integrated a recognition of the possible but undefined influence of dystrophin-based changes in the brain on anxiety with psychological thinking about anxiety as part of an adaptive coping response and a response also
open to influence from systemic contextual factors. Parents expressed the importance to them of acknowledging both organic and non-organic components.

Formulation, which is given central importance by psychologists, recognises that when someone is able to make sense of their experiences, this can be "a powerful intervention in its own right" (Division of Clinical Psychology, 2011), often enabling someone to “move forward with a richer understanding of their dilemmas and difficulties” (Johnstone & Dallos, 2014). The model of anxiety developed as part of this study differs from a formulation in many ways, namely that it is a general framework of hypotheses, rather than an idiosyncratic map centred on the personal meanings of an individual’s experiences. However, it shares the aim of offering hypotheses about a problem by linking theory with real life accounts. The model hopes to provide a starting place from which a richer understanding of the dilemmas and difficulties associated with the anxiety experienced by individuals with DMD can emerge. For some people, this may in itself, have a positive impact on their wellbeing.

The principles of Liberation Psychology also helped me attune to the knowledge and expertise shared by participants, which I noticed offered valuable insights into the very question about the causes of anxiety they were asking. Collating and interpreting the insights of participants, and combining them with relevant literature and psychological theory contributed towards the initial model of anxiety experienced by this population. Other researchers using framework analysis (e.g. Parkinson, Eatough, Holmes, Stapley, & Midgley, 2016) have drawn on the proposal of Denzin (1989), who suggests that the test of interpretations lies in how others perceive their usefulness and the interest they provoke. Drawing on this suggestion, it was essential that any model proposed by this study was useful and engaging to families affected by DMD and privileged their voices. Parent participants were therefore consulted on the format and content of the model when they provided validity checks on the qualitative analysis.
The first draft of the model (Figure 1) was undoubtedly influenced by my own bias towards systemic maps of problems, including Bronfenbrenner’s ecological systems theory (Bronfenbrenner, 1979). I wanted to present a model of broadly interacting systems rather than suggesting definitive and linear pathways of causation, which I thought risked oversimplifying the highly complex and idiosyncratic factors pertinent to each individual’s experience of anxiety.

Parent feedback, however, was that the concentric circles of this model were difficult to understand, and the biological aspect of DMD felt sidelined. For people affected by a physical health condition, it made sense, on reflection, that a highly psychosocial model of anxiety could feel invalidating of the physical context which is dominant in their lives. This initial model did not attend to the likely, but undefined organic contribution towards anxiety suggested in the literature. Conversely, a purely biological model risked presenting a narrow explanation of anxiety that

![Figure 1. Draft model of anxiety in DMD](image-url)
ignored the secondary challenges faced by these families and the influential responses of wider social and environmental systems to people with disabilities. A purely biological model also disregarded the research consistently pointing towards a strong environmental component in the etiology of childhood anxiety disorders (Murray, Creswell, & Cooper, 2009), and the data supporting this from the study. A biopsychosocial model helped to bridge this gap. Although I was hesitant about creating a model with boxes and arrows, due to the same fear of suggesting definitive, causative and linear pathways, parent participants fed back that the biopsychosocial model was easier to understand, and it provided them with a useful starting place from which to understand their son's anxiety.

**Scope of the study’s conclusions**

As acknowledged within the discussion of the empirical paper, the study lacked representativeness in several ways, with implications for the scope of findings and conclusions. No child participants or children of parent participants had an intellectual disability, despite around one third of males with DMD being given this diagnosis. Only one father took part, and only three parents had children aged 12 or older. The characteristics of anxiety identified may therefore have been skewed towards younger children and the perspectives of mothers, and may have missed characteristics more common to those with learning disabilities. Conversely, children with ASD were over represented within the sample. The prevalence of ASD within the DMD population is estimated to be around 8.2%, however, four child participants and sons of parent participants had a confirmed ASD diagnosis. If overlap between parent and child participants is considered, this meant that 25% of the 16 children with DMD whose experiences were discussed within the focus groups had a diagnosis of ASD. Although reports of anxiety characteristics that shared traits with ASD were not limited to those with an ASD diagnosis, the higher
rate of ASD within the sample compared to the wider DMD population may have contributed towards an over representation of ASD-type anxiety characteristics amongst the findings. Considering the relatively small sample size and the problems with representativeness, the conclusions of the study and the biopsychosocial model of anxiety presented must be held as a ‘starting place’ for understanding the anxiety experienced by this population and which further research must build upon. Considering the diversity amongst the DMD population across genetic, physical and neurological domains, alongside the differences between family and social systems around an individual with DMD, any proposed trends in anxiety experienced by this population must be considered alongside an individualised approach to assessment, formulation, and treatment.

References


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Appendix A

Child focus group semi-structured interview schedule

Child focus groups semi-structured interview schedule

- 7-11: Part 1: 10.30-11.15 (45 mins) Part 2: 11.30-12.00 (30 mins)

Intro (5 mins)

Warm up game (5 mins)

Exercise (35 mins)
This is Josh. Josh is 8 years old and he has Duchenne Muscular Dystrophy. Josh is feeling worried. What are some of the things he might be feeling worried about? (Picture of “Josh” printed onto piece of A0 paper. One facilitator writes each suggestion into a thought bubble above Josh. The other facilitator writes down each suggestion onto A5 coloured card for card sorting exercise in Part)

Prompts:
- What if Josh is at school. What might he be worried or afraid about there?
- What if Josh is playing with others…
- What if Josh is at home…
- What if Josh is trying to get to sleep…
- What if Josh is at the hospital?

- I wonder if any of you have ever felt worried or afraid in these places? (Prompts: can you tell me more about that. What did you worry would happen?)

- Are your worries similar or different to Josh’s? (Prompts: tell me more about that, can you think of an example? What about at school, do you have worries that are similar or different to Josh’s? What about at home? What about when you’re with your friends? What about at the hospital? What would be the worst thing that could happen? What would be the best thing that could happen? One facilitator to write down any new worries shared on coloured card for later exercise).
Further prompts: How often does that happen? Is that similar or different for others? How worried does it make you feel (use fear thermometer).

  - What happens when you feel worried or afraid? How would other people know you are feeling worried or afraid? *(Can use an example that has come up lots. Use printed A3 body outline and one facilitator write ideas onto it.)*

**Prompts**

- What do you do when you are worried or afraid? How is that different to normal?
- What do you say? Who do you say it to?
- Would they notice any changes in your body? E.g. heart beating faster
- What do you feel like inside when you are anxious?
- What sort of thoughts go through your mind? What things do you think that other people can’t see?

**Exercise (20 mins):**
*(Use worries that have been written on coloured A5 card plus pre-made worry cards.)*

Ask group to put all cards into three different groups, (each with a piece of A3 paper): one for ‘big worries’, one for ‘middle-sized worries’ and one for ‘little worries’. Also ask them to put the cards in an order from most worrying to the least worrying within each category. *(Aim: to stimulate interesting discussions about the different things making the participants feel anxious)* Follow up questions e.g. why did you decide to put X there and not there? Did anybody disagree with that? Why is X a little worry? E.g. because it doesn’t happen very often or because it doesn’t matter very much? How big is the biggest worry? *(Can use fear thermometer again)* Is that similar or different for others? Why is that such a big worry?

**Finishing up (5 mins):**

  - Vouchers as a thank you
  - Any questions for us?
  - What did you like about today? Were there any bits you didn’t like?
  - Reiterate speaking to us if they are feeling worried or upset about anything they have heard today.
Appendix B

Parent focus group semi-structured interview schedule

**Parent focus groups semi-structured interview schedule**

Intro (5 mins)

- Is anxiety something you have ever noticed your son experiencing?

- How do you know when your son is feeling anxious? What do you notice?

  a) **Prompts to put forward if they don’t come up naturally:**
     - What do you notice him doing? What sort of behaviour do you see?
       E.g. becomes shy/quiet, becomes demanding or angry, becomes controlling
     - What does he say (to you/to others)?
     - Do you notice any physical signs in his body or does he complain of any? E.g. trembling, tense, tummy ache, eye contact reduces
     - What do you think is going through his mind? What is worrying him?
     - Are there any behaviours that you understand happening as a result of your child’s anxiety but which you think could be interpreted differently by people who know your child less well (e.g. aggression or attention-seeking behaviour)?

- What does your child feel anxious about? Or worry about?
  a) **Prompts**
     - Unexpected changes
     - Separating from you
     - Being around others/crowded places

- What is often happening around your child when your child becomes anxious? Are there any particular triggers you are aware of?
  a) **Prompts:**
     - Particular places / situations / events
- Times of the day / week
- In relation to particular people
- Any other things that seem to trigger anxiety in your son?

- Are there situations or triggers which you actively avoid because your son would find it very difficult to tolerate due to anxiety?

- How does your son manage his feelings when he is experiencing anxiety?

- What do you do as a parent to try to prevent or avoid him feeling anxious?

- What is it like for you as a parent when your child is anxious?

- Is there an impact of your child’s anxiety on the family?
  
  a) Prompts
  
  - Are there things it stops you or your family doing?
  - Does it impact siblings?
Appendix C

Letter of ethical approval

UCL RESEARCH ETHICS COMMITTEE
OFFICE FOR THE VICE PROVOST RESEARCH

15 December 2017

Professor Francesco Muntoni
Dubowitz Neuromuscular Centre
UCL GOSH Institute of Child Health

Dear Professor Muntoni,

Notification of Ethics Approval with Provisos
Project ID/Title: 12307/001: Understanding and measuring anxiety in Duchenne Muscular Dystrophy
Exploring the perspectives of families and males with the condition

Further to your satisfactory responses to the Committee’s comments, I am pleased to confirm in my capacity as Co-Chair of the UCL Research Ethics Committee (REC) that the data collection element of your study has been ethically approved by the UCL REC until 31st July 2019. This approval also incorporates approval of the following small changes you would like to make to the study:

1. Increasing the duration as well as recording the focus groups using video equipment as well as audio equipment with the explicit permission of research participants.
2. To recruit participants from a ‘subject pool’ held by the Dubowitz Neuromuscular Centre at the Institute of Child Health of individuals with DMS who have expressed an interest in being involved in research.

Ethical approval is also subject to the following conditions.

Notification of Amendments to the Research
You must seek Chair’s approval for proposed amendments (to include extensions to the duration of the project) to the research for which this approval has been given. Ethical approval is specific to this project and must not be treated as applicable to research of a similar nature. Each research project is reviewed separately and if there are significant changes to the research protocol you should seek confirmation of continued ethical approval by completing an ‘Amendment Approval Request Form’ http://ethics.grad.ucl.ac.uk/responsibilities.php

Adverse Event Reporting – Serious and Non-Serious
It is your responsibility to report to the Committee any unanticipated problems or adverse events involving risks to participants or others. The Ethics Committee should be notified of all serious adverse events via the Ethics Committee Administrator (ethics@ucl.ac.uk) immediately the incident occurs. Where the adverse incident is unexpected and serious, the Joint Chairs will decide whether the study should be terminated pending the opinion of an independent expert. For non-serious adverse events the Joint Chairs of the Ethics Committee should again be notified via the Ethics Committee Administrator within ten days of the incident occurring and provide a full written report that should include any amendments to the participant information sheet and study protocol. The Joint Chairs will confirm that the incident is non-serious and report to the Committee at the next meeting. The final view of the Committee will be communicated to you.
Final Report
At the end of the data collection element of your research we ask that you submit a very brief report (1-2 paragraphs will suffice) which includes in particular issues relating to the ethical implications of the research i.e. issues obtaining consent, participants withdrawing from the research, confidentiality, protection of participants from physical and mental harm etc.

In addition, please:

- ensure that you follow all relevant guidance as laid out in UCL’s Code of Conduct for Research: http://www.ucl.ac.uk/srs/governance-and-committees/resgov/code-of-conduct-research
- note that you are required to adhere to all research data/records management and storage procedures agreed as part of your application. This will be expected even after completion of the study.

With best wishes for the research.

Yours sincerely

Dr Lynn Ang
Joint Chair, UCL Research Ethics Committee

Cc: Rachel Trimmer & Kate Maresh
Appendix D

Recruitment advert

Understanding anxiety in Duchenne muscular dystrophy (DMD)

Everybody feels worried or anxious from time to time, but sometimes boys with DMD can have these feelings more often than other children.

We are holding discussion groups with boys with DMD and their parents to explore the perspectives of people living with DMD and how feelings of worry or anxiety might affect them. We hope this study will help us to understand better how to help people with DMD who have these feelings.

We are looking for boys with DMD aged 7-18 years old and parents of children with DMD to take part in these discussion groups. You can join in whether or not you have experience of anxiety or similar feelings.

The study will involve coming to UCL GOS Institute of Child Health in London for one group session (separate for parents and children) during which there will be a number of focussed activities and discussions.

You will be reimbursed for your travel expenses and will receive a voucher as a ‘thank you’.

If you are interested please contact us:

Email: k.mares@ucl.ac.uk or rachell.trimmer.16@ucl.ac.uk

Address: Dubowitz Neuromuscular Centre, UCL Great Ormond Street Institute of Child Health, 30 Guilford Street, London, WC1N 1EH

Study Title: Understanding and measuring anxiety in Duchenne Muscular Dystrophy: exploring the perspectives of families and males with the condition.

Principal Investigator: Professor Francesco Muntoni

This study has been approved by the UCL Research Ethics Committee (Project ID No. 12307/001). All data will be collected and stored in accordance with the Data Protection Act 1998.
Appendix E

Recruitment email to prospective parent participants

Dear parents of X,

I am getting in touch to find out if you would be interested in taking part in a study I am running with my colleague, Dr Kate Maresh, at the Institute of Child Health investigating the profile of anxiety in DMD. I have your contact details from a list held by the Institute of Child Health of parents who have said they are happy to be contacted about upcoming studies and trials for boys with Duchenne.

I have attached more information about this study along with a short video we have made.

In brief, we are running small focus groups with boys with Duchenne and with parents of boys with Duchenne (separate groups) to find out more about the nature of anxiety in Duchenne and the impact it has on families. Little research has investigated this area yet it seems to be a really common and challenging aspect of the condition for many families. The purpose of the focus groups is to find out more from you, the experts, about the anxiety you notice and the impact this has, or in the case of boys, the anxiety being experienced first-hand. We hope this information will improve our understanding of anxiety in Duchenne and better inform those working with families around this important area.

All groups are held at the Institute of Child Health in London. Travel expenses will be reimbursed.

- 28th April morning group: arrival at 9.30. We expect to be finished by 12.30.
- 28th April afternoon group: currently fully booked although I can put you on a reserve list in case of drops out.

We will be running a third group for parents on one of the following days dependent on which date suits parents best:

- Saturday 30th June in the morning
- Saturday 21st July morning or afternoon

If you are interested in participating, let me know about your availability for these dates. Please also get in touch with any questions you have about the study. I would be happy to answer these by email or phone (XXXXXXXXXX), whichever suits you best.

I hope to hear from you soon.

Best wishes,
Rachel Trimmer
Trainee Clinical Psychologist, UCL
Appendix F

Photos of card sorting exercise from child focus groups
Appendix G

Still images from study information video
Appendix H

7-11 participant information sheet

Study Title: Understanding and measuring anxiety in Duchenne Muscular Dystrophy (17NM36)
Information Sheet for children (7-11 years) version 2.0, December 2017

A study about feeling worried and afraid
Information sheet for children (7-11 years)

Everybody feels worried and afraid from time to time but did you know... boys with DMD often feel worried and afraid more than other children?

Our names are Kate Maresh and Rachel Trimmer and we want to find out more about this. We are inviting boys and young men like you, who have a diagnosis of Duchenne Muscular Dystrophy, if they would like to take part in a 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 taking part will involve. Please take time to read the following information carefully. You may want to talk about it with other people such as your parents. Please ask us if there is anything that is not clear or if you would like more information. Take your time to decide if you would like to take part.
Study Title: Understanding and measuring anxiety in Duchenne Muscular Dystrophy (17NM36)  
Information Sheet for children (7-11 years) version 2.0 December 2017

Why is this study being done?

We want to find out what makes you feel worried. How do you know when you are feeling worried? Where do you feel worried? This study is being done because scientists have not asked boys such as yourselves very much about this before. If we learn more from you about what, when, and how you feel worried, this will help adults like doctors, teachers and parents to understand and help other boys and young men with DMD much better.

What will happen if I do the study?

If you agree to take part, you and your parents/guardians will sign a form to say that you are happy to take part.

You will come to the Institute of Child Health in London for a session with 3 other boys. We are holding another group for older boys aged 12-18 years old and a group for parents of boys with DMD. In your group we will do some activities which help us to talk more about feeling worried and afraid. For example, we will talk about how worried you feel in different types of places. We might ask you to bring some photos with you to help with this. The activities won’t be scary and hopefully you will find them fun.

The session will last for 2 hours with a short break in the middle. Snacks will be provided.
Will anything about the research upset me?

Talking about feeling worried and afraid may feel like a difficult or scary thing to do so we are coming up with ways to make this as easy as possible. Firstly, you do not need to talk about anything in the group that you don’t want to talk about. Secondly, everybody in the group will be asked not to tell anyone else what other people have said, including on social media. This means that ‘what is said in the room, stays in the room’. We are also doing our best to find fun and enjoyable ways to help everybody talk about their experiences. We hope all of these things help you to feel safe to talk freely.

If you want to talk more about feeling worried after the session, we will help you to find the best person talk to. This may be your GP or local child and adolescent mental health service (CAMHS). We can write a letter to help you get the support you need.

Will taking part help me at all?

You may enjoy sharing your feelings with other boys in the group and you may find it helpful to talk about things that you haven’t talked about before. You may also enjoy taking part in the study because you know you will be helping other boys with DMD.

As a thank you for taking part, we will give you a voucher to spend. We will also let you and your parents know what we find in the study by writing you a letter.

Will I be recorded and will anyone else see my information?

Kate and Rachel will write down information about you but the information will have a number on it instead of your name. Only Kate
Study Title: Understanding and measuring anxiety in Duchenne Muscular Dystrophy (17NM36)
Information Sheet for children (7-11 years) version 2.0 December 2017

and Rachel will know who you are and be able to see your personal information. If the information goes anywhere else to other people there will be no way of telling who was in the study. The only exception is if during the session we hear anything which makes us worried that someone might be in danger of harm. If this happens, we might need to let other people know about this to keep everyone safe.

We will use an audio and video recorder to record the session. These audio and video recordings will only be used for analysis and for examples in conference presentations and lectures. The recording will not be used in any other way without your permission, and no one outside the project will be allowed access to the original recordings. When the research is finished, we will delete all of the recordings and your personal details unless we ask your permission to keep them.

If you are worried about how information about you will be used, you can ask your parent/guardian to email:

data-protection@ucl.ac.uk.

If you are still unhappy, you can ask your parent/guardian to contact the Information Commissioner’s Office (ICO). The information needed to do this can be found on this website:

The information you tell us may be used in future research about feeling worried and afraid in boys who have DMD, however, this information will have a number next to it rather than your name so no one will be able to tell who it came from.
Study Title: Understanding and measuring anxiety in Duchenne Muscular Dystrophy (17NM38)
Information Sheet for children (7-11 years) version 2.0, December 2017

Do I have to take part in this study?

Remember, being in this study is up to you. You don’t have to be in this study if you don’t want to! Even if your parents think that you should do it, you should decide for yourself if you want to take part. If you decide not to take part, this will not change anything about how you are cared for. Have a chat with your family and friends about taking part in the study if it helps you to decide. If you decide to take part, you can change your mind at any time, even after the session has started and up to 2 weeks after the discussion group. If this happens, we will ask you what you would like to happen to the information you have given up to that point.

Contact us...
If you or your parents/guardians have any further questions please contact us:

Dr Kate Maresh and Rachel Trimmer
Phone: 020 7905 2151
Email: k.maresh@ucl.ac.uk rachel.trimmer.16@ucl.ac.uk
Dubowitz Neuromuscular Centre, UCL Great Ormond Street Institute of Child Health, 30 Guilford Street, London, WC1N 1EH

If you would like to contact the supervisor of this project please contact Dr.Will Mandy: Phone: 020 7679 1897 Email: will.mandy@ucl.ac.uk
Department of Clinical, Educational and Health Psychology, University College London, Gower Street, London, WC1E 6BT

Thank you very much!
Appendix I

12-18 participant information sheet

Study Title: Understanding and measuring anxiety in Duchenne Muscular Dystrophy (17NM36)
Information Sheet for children (12-18 years) version 2.0 December 2017

A study about feeling anxious

Information sheet for young people (12-18 years)

Everybody feels anxious from time to time but did you know... boys with DMD often feel anxious more than other children?

We would like to find out more about this and so we are inviting boys and young men like you, who have a diagnosis of Duchenne Muscular Dystrophy, if they would like to take part in a 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 taking part will involve. Please take time to read the following information carefully. You may want to talk about it with other people such as your parents. Please ask us if there is anything that is not clear or if you would like more information. Take your time to decide if you would like to take part.

Why is this study being done?

We want to find out what feeling anxious is really like for you. For example, what makes you feel anxious and how do other people know when you are feeling anxious?
This study is being done because scientists have not asked boys and young men with DMD very much about this before. If we learn more from you about what, when, and how you feel anxious, this will help other people like doctors, teachers and parents to understand and help other boys and young men better with their anxiety. You can take part in this study whether or not you have noticed feeling anxiety or similar feelings before.

What will happen if I do the study?

If you agree to take part, you and your parents/guardians will sign a form to say that you are happy for this.

You will come to the Institute of Child Health in London for a 2 hour session with 3 other boys. We are holding another group for younger boys aged 7-11 years old and another group for parents of boys with DMD. We will do some activities which help to explore your perspectives on anxiety. For example, we will talk about how worried you feel in different types of places. The activities are not designed to make you feel anxious and hopefully you will find them interesting and fun. We'll also have a short break in the middle and snacks will be provided.

As a thank you, we will give you a gift voucher to spend. We will also let you and your parents know what we find in the study by writing you a letter.
Is there anything I need to be worried about if I take part?

We appreciate that talking about anxiety may feel like a difficult or scary thing to do and so we are thinking carefully about how to make this as easy as possible. Firstly, you do not need to talk about anything in the group that you don’t feel comfortable to share. Secondly, everybody in the group will be asked not to tell anyone else what other people have said, including on social media. This means that ‘what is said in the room stays in the room’. We are also doing our best to find fun and enjoyable ways to help everybody talk about their experiences. We hope these things help you to feel safe to talk freely.

If you want to talk more about feeling anxious after the session, we will help you to find the best person to talk about this with. This may be your GP or local child and adolescent mental health service (CAMHS). We can write a letter to help you get the support you need.

What are the possible benefits of taking part?

You may enjoy sharing your experiences with other people in the group and you may find it helpful to talk about things that you haven’t talked about before. You may also enjoy taking part in the study because you know you will be helping other boys with DMD.

Will anyone else see my information? A notice about data protection privacy:

The session will be run by Kate Maresh and Rachel Trimmer. Kate and Rachel will write down information about you but the information will
have a number on it instead of your name. Only Kate and Rachel will know who you are and be able to see your personal information. If the information goes anywhere else to other people, there will be no way of telling who was in the study. The only exception is if during the session we hear anything which makes us worried that someone might be in danger of harm. If this happened, we might need to let other people know about this to keep everyone safe.

We also ask that after the session you don’t talk about what other people have said, especially on social media, to respect their confidentiality. This means that ‘what is said in the room stays in the room’ and it is to keep the group a safe place where others feel they can talk freely.

We will use an audio and video recorder to record the session. These audio and video recordings will only be used for analysis and for examples in conference presentations and lectures. The recording will not be used in any other way without your permission, and no one outside the project will be allowed access to the original recordings. When the research is finished, we will delete all of the recordings unless we ask your permission to keep them.

All paper notes and consent forms will be stored in the site file, which is kept in a lockable room in the Institute of Child Health. These paper notes and consent forms will be stored for 5 years, after which they will be archived for a minimum of 20 years.

Electronic data will be stored indefinitely on the UCL network and encrypted USB sticks. Personal identifiable data will be stored in the UCL Data Safe Haven. The exception is the original digital audio and video files which will be deleted at the end of the study.
If you are worried about how information about you will be used, you can email data-protection@ucl.ac.uk or ask your parent/guardian to do this on your behalf. If you are still unhappy, you can contact the Information Commissioner’s Office (ICO) or ask your parent/guardian to do this on your behalf. The information needed to do this can be found on this website: https://ico.org.uk/for-organisations/data-protection-reform/overview-of-the-gdpr/individuals-rights/

The anonymous data from this study may be used in future studies investigating ways of understanding and measuring anxiety in DMD.

Do I have to take part in this study?

Being in this study is completely up to you. You do not have to take part if you don’t want to! Even if your parents think that you should do it, you should decide for yourself if you want to take part. If you decide not to take part, this will not change anything about how you are cared for. Have a chat with your family and friends about taking part in the study if it helps you to decide.

What if I decide I don’t want to do the research anymore?

If you decide to take part, you can change your mind at any time, even after the session has started and up to 2 weeks after the session. If this happens, we will ask you what you would like to happen to the information you have given up to that point.
Contact us...
This study is being organised by researchers from the UCL department of Clinical, Educational and Health Psychology and the UCL Institute of Child Health. It is funded by UCL Grand Challenges. If you or your parents/guardians have any further questions, please contact us:

Dr Kate Maresh and Rachel Trimmer
Phone: 020 7905 2151
Email: k.maresh@ucl.ac.uk     rachel.trimmer.16@ucl.ac.uk
Dubowitz Neuromuscular Centre, UCL Great Ormond Street Institute of Child Health, 30 Guilford Street, London, WC1N 1EH

If you would like to contact the supervisor of this project please contact Dr. Will Mandy:
Phone: 020 7679 1897 Email: will.mandy@ucl.ac.uk
Department of Clinical, Educational and Health Psychology, University College London, Gower Street, London, WC1E 6BT

Thank you very much!
Appendix J

Parent participant information sheet

Study Title: Understanding and measuring anxiety in Duchenne Muscular Dystrophy (17NM36)
Information Sheet for adult participants version 2.0 _December 2017

A study about anxiety in Duchenne Muscular Dystrophy

Information sheet for adult participants

Everyone feels anxious from time to time however research suggests that boys with DMD often feel more anxious than other children of the same age.

We want to understand this better and so we are inviting parents and guardians of boys and young men with a diagnosis of Duchenne Muscular Dystrophy if they would like to take part in a study. Before you decide, it is important for you to understand why the research is being done and what participation will involve. Please take time to read the following information carefully and discuss it with others if you wish. Ask us if there is anything that is not clear or if you would like more information. Take your time to decide whether or not you wish to take part.

Why is this study being done?

We want to hear your perspectives on your son’s anxiety and the family’s experience of this. For example, what makes him feel anxious? How do you know when he is feeling anxious? And what is the impact of your son’s anxiety on the family? Few studies have explored the perspectives of parents on anxiety in DMD in this way. We hope that by hearing your views, better ways of identifying and measuring anxiety in DMD can be found so that doctors, teachers and parents can better understand and respond to the needs of both boys with DMD and their families. You can take part in this study whether or not you have noticed anxiety or similar feelings in your son previously.

Please note, to be included in this study, your son needs to have had a confirmed diagnosis of DMD, for example by a genetic test or muscle biopsy.

Approved by the UCL Research Ethics Committee (Project ID No. 12307/001)
Registered with the Joint Research & Development Office, UCL Great Ormond Street Institute of Child Health & Great Ormond Street Hospital for Children NHS Foundation Trust (R&D No. 17NM36)
Study Title: Understanding and measuring anxiety in Duchenne Muscular Dystrophy (17NM36)
Information Sheet for adult participants version 2.0_December 2017

What will happen if I decide to take part in the study?

If you agree to take part, we will ask you to sign a form consenting to the study. The study will take place at the Institute of Child Health in London. It will involve a guided group discussion around what you notice about your son’s anxiety and your experiences of any questionnaires you have been asked you to complete relating to this. There will 6 parents of boys with DMD in the group including yourself. The session will last for 2.5 hours with a break in the middle.

We are conducting similar group discussions with boys who have a diagnosis of DMD to explore their first-hand perspectives on anxiety. We are inviting boys aged 7-11 and 12-18 to participate in two separate discussion groups as part of this study.

Refreshments will be provided and we will reimburse all travel expenses. We will let you know the findings of the research by writing you a letter once the study has finished. We will also give you a gift voucher as a ‘thank you’ for taking part.

Taking part in this study is entirely voluntary. If you decide not to take part, this will not change anything about the support your family receive. You can change your mind about taking part in the study at any time, even after the session has started and up to 2 weeks after the session. If this happens, we will ask you what you would like to happen to the information you have given up to that point.

If issues arise from the focus group either around your own anxiety or your son’s anxiety, we can support you to find the best service to help you with this. This may be your GP or your local adult or child and adolescent mental health service (CAMHS). We are able to write a supporting referral letter to the appropriate service if this is helpful. If any concerns are observed in relation to your health, we will contact your GP.

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Registered with the Joint Research & Development Office, UCL Great Ormond Street Institute of Child Health & Great Ormond Street Hospital for Children NHS Foundation Trust (R&D No. 17NM36)
Study Title: Understanding and measuring anxiety in Duchenne Muscular Dystrophy (17NM36)
Information Sheet for adult participants version 2.0, December 2017

What are the possible disadvantages of taking part?

We appreciate that talking about your son’s anxiety and your family’s experiences may feel anxiety-provoking. We are therefore thinking carefully about how to make the group discussion as easy for participants as possible. Firstly, do not need to share any information in the group that you do not feel comfortable sharing. Secondly, everybody in the group will be asked not to tell anyone outside of the group what other people have said, including on social media. This means that ‘what is said in the room stays in the room’. We hope these things help you to feel safe to talk freely.

If issues arise during the group which you would like further support with, we will help you to find the best person to provide this support. This may be your GP or an appropriate mental health service. We can write a supporting referral letter to help you get the support you need.

What are the possible benefits of taking part?

You may enjoy sharing your experiences with other people in the group and you may find it helpful to talk about things that you haven’t talked about before. You may also enjoy taking part in the study knowing that the study aims to better understand and support the needs of boys with DMD.

Data Protection and Privacy Notice

The session will be run by Dr Kate Maresh (Specialist Registrar in Neurology) and Rachel Trimmer (Trainee Clinical Psychologist). All information from the study will be anonymised so that it will not be possible for you to be identified and the data will be kept strictly confidential. Nobody other than Kate and Rachel will be able to see any personally identifiable information from this research. We will endeavour to minimise the processing of personal data wherever possible. The only exception is if we hear anything during the session which makes us worried that someone might be in danger of harm. If this happens, we may need to let relevant others know about this to keep everyone safe.

We also ask that after the session you don’t talk about what other people have said, especially on social media, to respect their confidentiality. This means

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Study Title: Understanding and measuring anxiety in Duchenne Muscular Dystrophy (17NM36)
Information Sheet for adult participants version 2.0_December 2017

that ‘what is said in the room stays in the room’ and it is to keep the group a safe place where others feel they can talk freely.

We will use an audio and video recorder to record the session. These audio and video recordings will only be used for analysis and for anonymous examples in conference presentations and lectures. The recording will not be used in any other way without your permission, and no one outside the project will be allowed access to the original recordings. When the research is finished, we will delete all of the recordings unless we ask your permission to keep them.

All paper notes and consent forms will be stored in the site file, which is kept in a lockable room in the Institute of Child Health. These paper notes and consent forms will be stored for 5 years, after which they will be archived for a minimum of 20 years.

Electronic data will be stored indefinitely on the UCL network and encrypted USB sticks. Personal identifiable data will be stored in the UCL Data Safe Haven. The exception is the original digital audio and video files which will be deleted at the end of the study.

The data controller for this project will be University College London (UCL). The UCL Data Protection Office provides oversight of UCL activities involving the processing of personal data, and can be contacted at data-protection@ucl.ac.uk. UCL’s Data Protection Officer is Lee Shailer and he can also be contacted at data-protection@ucl.ac.uk.

Your personal data will be processed for the purposes outlined in this notice. The legal basis that would be used to process your personal data will be the provision of your consent. You can provide your consent for the use of your personal data in this project by completing the consent form that has been provided to you.

*Your personal data will be processed so long as it is required for the research project.*

If you are concerned about how your personal data is being processed, please contact UCL in the first instance at data-protection@ucl.ac.uk. If you remain unsatisfied, you may wish to contact the Information Commissioner’s Office (ICO). Contact details, and details of data subject rights, are available on the

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Study Title: Understanding and measuring anxiety in Duchenne Muscular Dystrophy (17NM36)
Information Sheet for adult participants version 2.0 December 2017

ICO website at: https://ico.org.uk/for-organisations/data-protection-reform/overview-of-the-gdpr/individuals-rights/

If future studies are completed investigating ways of understanding and measuring anxiety in DMD then the anonymous data from this study may be used.

Contact us...
This study is being organised by researchers from the UCL department of Clinical, Educational and Health Psychology and the UCL Institute of Child Health. It is funded by UCL Grand Challenges. If you have any further questions about this study or the study with boys and young men with DMD, please contact us:

Dr Kate Maresh and Rachel Trimmer
Phone: 020 7905 2151
Email: k.maresh@ucl.ac.uk rachel.trimmer.16@ucl.ac.uk

Dubowitz Neuromuscular Centre, UCL Great Ormond Street Institute of Child Health, 30 Guilford Street, London, WC1N 1EH

If you would like to contact the supervisor of this project please contact:
Dr. Will Mandy: Phone: 020 7679 1897 Email: will.mandy@ucl.ac.uk
Department of Clinical, Educational and Health Psychology, University College London, Gower Street, London, WC1E 6BT

Thank you very much!

Approved by the UCL Research Ethics Committee (Project ID No. 12307/001)
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Appendix K

Parent of child participants participant information sheet

Study Title: Understanding and measuring anxiety in Duchenne Muscular Dystrophy (17NM38)
Information Sheet for parents/guardians of participants version 2.0, December 2017

A study about anxiety in Duchenne Muscular Dystrophy

Information sheet for parents and guardians of participants

Everyone feels anxious from time to time however research suggests that boys with DMD often feel more anxious than other children of the same age.

We want to understand this better and so we are inviting boys and young men with a diagnosis of Duchenne Muscular Dystrophy if they would like to take part in a study. It is important for you to understand why the research is being done and what your son’s participation will involve. Please take time to read the following information carefully and discuss it with others, including your son, if you wish. Ask us if there is anything that is not clear or if you would like more information. We encourage your son to take time to decide whether or not he wishes to take part.

Why is this study being done?

We want to find out more from boys and young men with DMD about their experiences of anxiety. For example, what makes them feel anxious and how do they and those around them know when they are feeling anxious? Previous studies have not investigated this first-hand account of anxiety in DMD. We hope that by hearing from boys and young men with DMD themselves, better ways of identifying and measuring anxiety can be found for this group so that doctors, teachers and parents, can better understand and respond to their needs. Your son can take part in this study whether or not he has noticed anxiety or similar feelings in the past.

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Registered with the Joint Research & Development Office, UCL Great Ormond Street Institute of Child Health & Great Ormond Street Hospital for Children NHS Foundation Trust (R&D No. 17NM36)
What will happen if my son takes part in the study?

If your son agrees to take part, you will each sign a form consenting to the study. The study will take place at the Institute of Child Health in London. The study will involve a discussion group guided by activities aiming to help the boys share their perspectives on anxiety. These activities are not designed to induce anxiety and will hopefully be fun and interesting for everyone taking part. The session will last for 2 hours with a short break in the middle. We will hold two groups for boys with DMD: one group for younger children aged 7-11 and one group for older children aged 12-18. There will be 4 boys with DMD in each group. Refreshments will be provided.

We are also holding discussion groups for parents of boys with DMD as part of this study. This will involve two groups with 6 parents in each.

As a thank you, we will give your son a gift voucher to spend. We will also reimburse all travel expenses. We will let you and your son know the findings of the research by writing a letter to you both once the study has finished.

If your son wishes to talk more about feeling anxious after the session, we will support him to find the best person talk about this with and discuss this with you if appropriate. This may be your GP or local child and adolescent mental health service (CAMHS). We can write a supporting referral letter to the appropriate service if this is helpful. If any concerns are observed in relation to your son’s physical health, we will contact your GP.

What are the possible disadvantages of taking part in the study?

We appreciate that talking about anxiety, particularly in a group setting, may itself be anxiety-provoking for your son. We are therefore thinking carefully about how to make the group discussion as easy for participants as possible. Firstly, your son will be reminded that he does not need to share any information in the group that he does not feel comfortable sharing. Secondly, everyone in the group will be asked not to share any information outside of the group about what other participants have said, including on social media. This means that ‘what is said in the room stays in the room’. We are also doing our
best to find fun and enjoyable ways to help participants talk about their experiences. We hope these things will help your son to feel safe to talk freely.

What are the possible advantages of taking part in the study?

Your son may enjoy sharing his experiences with other people in the group and he may find it helpful to talk about things that he may not have talked about before. He may also enjoy taking part in the study knowing that it aims to better understand and support the needs of boys with DMD.

Data Protection Privacy Notice

The session will be run by Dr Kate Maresh (Specialist Registrar in Neurology) and Rachel Trimmer (Trainee Clinical Psychologist). All information from the study will be anonymised so that it is not possible for your son to be identified and the data will be kept strictly confidential. Nobody other than Kate and Rachel will be able to see any personally identifiable information from this research. We will endeavour to minimise the processing of personal data wherever possible. The only exception is if we hear anything during the session which makes us worried that someone might be in danger of harm. If this happens, we may need to let relevant others know about this to keep everyone safe.

We also ask that after the session you ensure that the confidentiality of other participants is respected, so your son should be aware that he should not discuss the personal details of others outside the group, especially on social media. This means that ‘what is said in the room stays in the room’ and it is to keep the group a safe place where others feel they can talk freely.

We will use an audio and video recorder to record the session. These audio and video recordings will only be used for analysis and for anonymous examples in conference presentations and lectures. The recording will not be used in any other way without you and your son’s permission, and no one outside the project will be allowed access to the original recordings. When the

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Registered with the Joint Research & Development Office, UCL Great Ormond Street Institute of Child Health & Great Ormond Street Hospital for Children NHS Foundation Trust (R&D No. 17NM36)
Study Title: Understanding and measuring anxiety in Duchenne Muscular Dystrophy (17NM36)
Information Sheet for parents/guardians of participants version 2.0_December 2017

research is finished, we will delete all of the recordings unless we ask for permission from you and your son to keep them.

All paper notes and consent forms will be stored in the site file, which is kept in a lockable room in the Institute of Child Health. These paper notes and consent forms will be stored for 5 years, after which they will be archived for a minimum of 20 years.

Electronic data will be stored indefinitely on the UCL network and encrypted USB sticks. Personal identifiable data will be stored in the UCL Data Safe Haven. The exception is the original digital audio and video files which will be deleted at the end of the study.

The data controller for this project will be University College London (UCL). The UCL Data Protection Office provides oversight of UCL activities involving the processing of personal data, and can be contacted at:
data-protection@ucl.ac.uk
UCL’s Data Protection Officer is Lee Shailer and he can also be contacted via this email.

Your son’s personal data will be processed for the purposes outlined in this notice. The legal basis that would be used to process your personal data will be the provision of your consent and consent from your son. You can provide your consent for the use of your son’s personal data in this project by completing the consent form that has been provided to you.

**Your son’s personal data will be processed so long as it is required for the research project.**

If you are concerned about how your son’s personal data is being processed, please contact UCL in the first instance at data-protection@ucl.ac.uk. If you remain unsatisfied, you may wish to contact the Information Commissioner’s Office (ICO). Contact details, and details of data subject rights, are available on the ICO website at: https://ico.org.uk/for-organisations/data-protection-reform/overview-of-the-gdpr/individuals-rights/

If future studies are completed investigating ways of understanding and measuring anxiety in DMD then the anonymous data from this study may be used.

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Registered with the Joint Research & Development Office, UCL Great Ormond Street Institute of Child Health & Great Ormond Street Hospital for Children NHS Foundation Trust (R&D No. 17NM36)
Study Title: Understanding and measuring anxiety in Duchenne Muscular Dystrophy (17NM36) Information Sheet for parents/guardians of participants version 2.0_December 2017

Does my son have to take part in this study?

No. Although you may bring this study to your son’s attention, the decision for him to take part must be entirely his. If he decides he does not want to take part, this will not change anything about the support your family receive. Your son may wish to chat with you about taking part if this helps him to decide. If your son decides he would like to take part, he can change his mind at any time, even after the session has started and up to 2 weeks after the session. If this happens, we will ask your son what he would like to happen to the information he has given up to that point.

To be included in this study, your son needs to have a definite diagnosis of DMD and is within one of the two age brackets listed.

Contact us...

This study is being organised by researchers from the UCL department of Clinical, Educational and Health Psychology and the UCL Institute of Child Health. It is funded by UCL Grand Challenges. If you have any further questions, please contact us:

Dr Kate Maresh and Rachel Trimmer
Phone: 020 7905 2151
Email: k.maresh@ucl.ac.uk rachel.trimmer.16@ucl.ac.uk

Dubowitz Neuromuscular Centre, UCL Great Ormond Street Institute of Child Health, 30 Guilford Street, London, WC1N 1EH

If you would like to contact the supervisor of this project please contact:
Dr. Will Mandy: Phone: 020 7679 1897 Email: will.mandy@ucl.ac.uk
Department of Clinical, Educational and Health Psychology, University College London, Gower Street, London, WC1E 6BT

Thank you very much!
Appendix L

Assent form

Study Title: Understanding and measuring anxiety in Duchenne Muscular Dystrophy

‘Child Participant’ Assent Form version 2.0 December 2017

Understanding and measuring anxiety in Duchenne Muscular Dystrophy

Assent Form (for participants aged 7-15 years)

Please complete this form after you have read the Information Sheet and/or listened to an explanation about the research.

Title of Study: Understanding and measuring anxiety in Duchenne Muscular Dystrophy

Department: a) Department of Clinical, Educational, and Health Psychology b) Institute of Child Health

Name and Contact Details of the Researchers: Dr Kate Maresh (k.maresh@ucl.ac.uk) and Rachel Trimmer (Rachel.trimmer.16@ucl.ac.uk) Phone: 020 7905 2151

Name and Contact Details of the Principal Researcher: Dr. Will Mandy (will.mandy@ucl.ac.uk) Phone: 020 7679 1897

Name and Contact Details of the UCL Data Protection Officer: Lee Shailer (data-protection@ucl.ac.uk)

This study has been approved by the UCL Research Ethics Committee: Project ID number: 12307/001

Thank you for considering taking part in this research. The person organising the research must explain the project to you before you agree to take part. If you have any questions about the Information Sheet or anything about the study, please ask the researcher before you decide if you want to join in. You will be given a copy of this Assent Form to keep and look at any time.

I confirm that I understand that by putting a tick or my initials in each box below I am consenting to this part of the study. I understand that it will be assumed that boxes without a tick or my initials mean that I DO NOT consent to that part of the study. I understand that by not ticking any of the boxes this may mean I am not be able to take part in the study.
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<tr>
<td>1.</td>
<td>I confirm that I have read the Participant Information Sheet (version 2.0, December 2017) for the above research study and I understand what the study involves.</td>
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<td>2.</td>
<td>I understand that I do not have to take part, and if I decide that I do not want to take part in this project at any time I can stop at any time.</td>
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<td>3.</td>
<td>I agree that the discussion group session can be audio and video-recorded.</td>
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<tr>
<td>4.</td>
<td>I agree that my personal information (such as my name and date of birth) and the information from the audio and video recordings can be used for this research study.</td>
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<tr>
<td>5.</td>
<td>I understand that this information will be kept confidential, so it will only be seen by researchers working on the study.</td>
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<tr>
<td>6.</td>
<td>I agree that I will not share what has been discussed in the group, such as personal details of other participants, outside of the group (including on social media).</td>
<td></td>
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<tr>
<td>7.</td>
<td>I agree that the results from the study can be published and shared with other researchers, but only after they have been fully anonymised, so I won’t be identified.</td>
<td></td>
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<tr>
<td>8.</td>
<td>I agree that this research study has been explained to me to my satisfaction and I agree to take part in this study.</td>
<td></td>
</tr>
</tbody>
</table>

If you would like your contact details to be kept so that you can be contacted in the future by UCL researchers who would like to invite you to take part in follow up studies to this project, or in future studies that are similar, please tick the right box below.

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<tr>
<td>Yes, I would be happy to be contacted in this way</td>
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<tr>
<td>No, I would not like to be contacted</td>
<td></td>
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</tbody>
</table>

Name of participant _______________ Date _______________ Signature _______________

Name of Parent/Guardian _______________ Date _______________ Signature _______________

Researcher _______________ Date _______________ Signature _______________
Appendix M

16-18 participants consent form

Approved by the UCL Research Ethics Committee (Project ID No. 12307/001)
Registered with the Joint Research & Development Office, UCL Great Ormond Street Institute of Child Health & Great Ormond Street Hospital for Children NHS Foundation Trust (R&D No. 17NW36)

‘Child Participant’ Assent Form version 2.0_ December 2017

Consent Form for participants aged 16-18 years

Please complete this form after you have read the Information Sheet and/or listened to an explanation about the research.

Title of Study: Understanding and measuring anxiety in Duchenne Muscular Dystrophy

Department: a) Department of Clinical, Educational, and Health Psychology b) Institute of Child Health

Name and Contact Details of the Researchers: Dr Kate Maresh (k.maresh@ucl.ac.uk) and Rachel Trimmer (Rachel.trimmer.16@ucl.ac.uk) Phone: 020 7905 2151

Name and Contact Details of the Principal Researcher: Dr. Will Mandy (will.mandy@ucl.ac.uk) Phone: 020 7679 1897

Name and Contact Details of the UCL Data Protection Officer: Lee Shailer (data-protection@ucl.ac.uk)

This study has been approved by the UCL Research Ethics Committee: Project ID number: 12307/001

Thank you for considering taking part in this research. The person organising the research must explain the project to you before you agree to take part. If you have any questions arising from the Information Sheet or explanation already given to you, please ask the researcher before you decide whether to join in. You will be given a copy of this Consent Form to keep and refer to at any time.

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<td>1.</td>
<td>I confirm that I have read and understood the Information Sheet for the above study. I have had an opportunity to think about the information and what will be expected of me. I have also had the opportunity to ask questions and I am happy with the answers I have been given. I would like to take part in a group discussion.</td>
</tr>
<tr>
<td>2.</td>
<td>I understand that I will be able to withdraw my data up to 2 weeks after the discussion group.</td>
</tr>
<tr>
<td>3.</td>
<td>I consent to the processing of my personal information (name, address) for the purposes explained to me. I understand that such information will be handled in accordance with all applicable data protection legislation.</td>
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</tbody>
</table>
| 4. | Use of the information for this project only  
   I understand that all personal information will remain confidential and that all efforts will be made to ensure I cannot be identified.  
   I understand that my data gathered in this study will be stored anonymously and securely. It will not be possible to identify me in any publications. |
| 5. | I understand that my information may be subject to review by responsible individuals from the University (including sponsors and funders) for monitoring and audit purposes. |
| 6. | I understand that my participation is voluntary and that I am free to withdraw at any time without giving a reason and without the care I receive or my legal rights being affected.  
   I understand that if I decide to withdraw, any personal data I have provided up to that point will be deleted unless I agree otherwise. |
| 7. | I understand the potential risks of participating and the support that will be available to me should I become distressed during the course of the research. |
| 8. | I understand the direct/indirect benefits of participating. |
| 9. | I understand that the data will not be made available to any commercial organisations but is solely the responsibility of the researcher(s) undertaking this study. |
| 10. | I agree that my anonymised research data may be used by others for future research. [No one will be able to identify you when this data is shared.] |
| 11. | I understand that the information I have submitted will be published as a report and I wish to receive a copy of it. Yes/No |
| 12. | I understand that I must not talk to others outside of the discussion group about personally identifiable information including posting on social media. |
| 13. | I consent to my interview being audio and video recorded and understand that the recordings will be destroyed immediately following transcription. |
| 14. | I hereby confirm that I understand the inclusion and exclusion criteria as detailed in the Information Sheet and explained to me by the researcher. I do not fall under the exclusion criteria. |
| 15. | I agree that my GP may be contacted if any unexpected results are found in relation to my health. |
| 16. | I am aware of who I should contact if I wish to lodge a complaint. I have informed the researcher of any other research in which I am currently involved or have been involved in during the past 12 months. |
| 17. | I voluntarily agree to take part in this study. I am aware of who I should contact if I wish to lodge a complaint. |
| 18. | Use of information for this project and beyond  
   I would be happy for the data I provide to be stored after completion of the study: I understand that paper notes and consent forms will be stored for 5 years, after which they will be securely archived for a minimum of 20 years. |
Electronic data will be stored indefinitely, with exception of digital audio/video files which will be deleted after processing.
I understand that other authenticated researchers will have access to my anonymised data.
I voluntarily agree to take part in this study.

If you would like your contact details to be retained so that you can be contacted in the future by UCL researchers who would like to invite you to participate in follow up studies to this project, or in future studies of a similar nature, please tick the appropriate box below.

| Yes, I would be happy to be contacted in this way |  |
| No, I would not like to be contacted |  |

Name of participant ___________________________ Date __________ Signature __________

Researcher ___________________________ Date __________ Signature __________
Appendix N

Parent of child participants consent form

Approved by the UCL Research Ethics Committee (Project ID No. 12307/001)
Registered with the Joint Research & Development Office, UCL Great Ormond Street Institute of Child Health & Great Ormond Street Hospital for Children NHS Foundation Trust (R&D No. 17NM336)

Study Title: Understanding and measuring anxiety in Duchenne Muscular Dystrophy
‘Parent of Participant’ Consent Form version 2.0_ December 2017

Consent Form for parents and guardians of participants

Please complete this form after you have read the Information Sheet and/or listened to an explanation about the research.

Title of Study: Understanding and measuring anxiety in Duchenne Muscular Dystrophy

Department: a) Department of Clinical, Educational, and Health Psychology b) Institute of Child Health
Name and Contact Details of the Researchers: Dr Kate Maresh (k.mareshe@ucl.ac.uk) and Rachel Trimmer (rachel.trimmer.16@ucl.ac.uk) Phone: 020 7905 2151
Name and Contact Details of the Principal Researcher: Dr. Will Mandy (will.mandy@ucl.ac.uk) Phone: 020 7679 1897

Name and Contact Details of the UCL Data Protection Officer: Lee Shailer (data-protection@ucl.ac.uk)

This study has been approved by the UCL Research Ethics Committee: Project ID number: 12307/001

Thank you for considering your son taking part in this research. The person organising the research must explain the project to you before you agree for your son to take part. If you have any questions arising from the Information Sheet or explanation already given to you, please ask the researcher before you decide whether you consent to your son joining in. You will be given a copy of this Consent Form to keep and refer to at any time.

I confirm that I understand that by putting a tick or my initials in each box below I am consenting to this part of the study. I understand that it will be assumed that boxes without a tick or my initials mean that I DO NOT consent to that part of the study. I understand that by not giving consent for any one element that my son may not be able to take part in the study.
<p>| | |</p>
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<tbody>
<tr>
<td>1.</td>
<td>I confirm that I have read and understood the Information Sheet for the above study. I have had an opportunity to think about the information and what will be expected of my son. I have also had the opportunity to ask questions and I am happy with the answers I have been given. I agree to my son taking part in a group discussion.</td>
</tr>
<tr>
<td>2.</td>
<td>I understand that I will be able to withdraw my son’s data up to 2 weeks after the discussion group.</td>
</tr>
<tr>
<td>3.</td>
<td>I consent to the processing of my son’s personal information (name, address) for the purposes explained to me. I understand that such information will be handled in accordance with all applicable data protection legislation.</td>
</tr>
</tbody>
</table>
| 4. | **Use of the information for this project only**
I understand that all personal information will remain confidential and that all efforts will be made to ensure my son cannot be identified. I understand that my son’s data gathered in this study will be stored anonymously and securely. It will not be possible to identify my son in any publications. |
| 5. | I understand that my son’s information may be subject to review by responsible individuals from the University (including sponsors and funders) for monitoring and audit purposes. |
| 6. | I understand that my son’s participation is voluntary and that both me and my son are free to withdraw his participation at any time without giving a reason, and without the care either of us receive or our legal rights being affected. I understand that if my son decides to withdraw, any personal data he has provided up to that point will be deleted unless we both agree otherwise. |
| 7. | I understand the potential risks of participating and the support that will be available to my son should he become distressed during the course of the research. |
| 8. | I understand the direct/indirect benefits of participating. |
| 9. | I understand that the data will not be made available to any commercial organisations but is solely the responsibility of the researcher(s) undertaking this study. |
| 10. | I agree that my son’s anonymised research data may be used by others for future research. [No one will be able to identify your son when this data is shared.] |
| 11. | I understand that the information my son has submitted will be published as a report and I wish to receive a copy of it. Yes/No |
| 12. | I understand that my son must not talk to others outside of the discussion group about personally identifiable information including posting on social media. |
| 13. | I consent to my son’s interview being audio and video recorded and understand that the recordings will be destroyed immediately following transcription. |
| 14. | I hereby confirm that I understand the inclusion and exclusion criteria as detailed in the Information Sheet and explained to me by the researcher. My son does not fall under the exclusion criteria. |
| 15. | I agree that my son’s GP may be contacted if any unexpected results are found in relation to his health. |
| 16. | I am aware of who I should contact if I wish to lodge a complaint. I have informed the researcher of any other research in which I am currently involved or have been involved in during the past 12 months. |
| 17. | I voluntarily agree for my son to take part in this study. I am aware of who I should contact if I wish to lodge a complaint. |
| 18. | **Use of information for this project and beyond**
I would be happy for the data my son provides to be stored after completion of the study. I understand that paper notes and consent forms will be stored for 5 years, after which they will be archived for a minimum of 20 years. Electronic |
data will be stored indefinitely, with exception of digital audio/video files which will be deleted after processing.

I understand that other authenticated researchers will have access to my son’s anonymised data.

I voluntarily agree for my son to take part in this study.

If you would like your contact details to be retained so that you can be contacted in the future by UCL researchers who would like to invite you or your son to participate in follow up studies to this project, or in future studies of a similar nature, please tick the appropriate box below.

<table>
<thead>
<tr>
<th>Yes, I would be happy to be contacted in this way</th>
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<td>No, I would not like to be contacted</td>
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<tr>
<th>Name of participant</th>
<th>Date</th>
<th>Signature</th>
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<tr>
<td>Name of Parent/Guardian</td>
<td>Date</td>
<td>Signature</td>
</tr>
<tr>
<td>Researcher</td>
<td>Date</td>
<td>Signature</td>
</tr>
</tbody>
</table>

Approved by the UCL Research Ethics Committee (Project ID No. 12307/001)
Registered with the Joint Research & Development Office, UCL Great Ormond Street Institute of Child Health & Great Ormond Street Hospital for Children NHS Foundation Trust R&D No. 17NM5361
Appendix O

Parent participant consent form

Study Title: Understanding and measuring anxiety in Duchenne Muscular Dystrophy

‘Adult Participant’ Consent Form version 2.0, December 2017

Approved by the UCL Research Ethics Committee [Project ID No. 12307/001]
Registered with the Joint Research & Development Office, UCL Great Ormond Street Institute of Child Health & Great Ormond Street Hospital for Children NHS Foundation Trust (R&D No. 17NM36)

Consent Form for adult participants

Please complete this form after you have read the Information Sheet and/or listened to an explanation about the research.

Title of Study: Understanding and measuring anxiety in Duchenne Muscular Dystrophy

Department: a) Department of Clinical, Educational, and Health Psychology b) Institute of Child Health

Name and Contact Details of the Researchers: Dr Kate Maresh (k.maresh@ucl.ac.uk) and Rachel Trimmer (rachel.trimmer.16@ucl.ac.uk) Phone: 020 7905 2151

Name and Contact Details of the Principal Researcher: Dr. Will Mandy (will.mandy@ucl.ac.uk) Phone: 020 7679 1897

Name and Contact Details of the UCL Data Protection Officer: Lee Shailer (data-protection@ucl.ac.uk)

This study has been approved by the UCL Research Ethics Committee: Project ID number: 12307/001

Thank you for considering taking part in this research. The person organising the research must explain the project to you before you agree to take part. If you have any questions arising from the Information Sheet or explanation already given to you, please ask the researcher before you decide whether to join in. You will be given a copy of this Consent Form to keep and refer to at any time.

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| 10. | I agree that my anonymised research data may be used by others for future research. [No one will be able to identify you when this data is shared.] |   |
| 11. | I understand that the information I have submitted will be published as a report and I wish to receive a copy of it. Yes/No |   |
| 12. | I understand that I must not talk to others outside of the discussion group about personally identifiable information including posting on social media. |   |
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</table>

Name of participant | Date | Signature
Researcher | Date | Signature
### Appendix P

#### Framework analysis example

<table>
<thead>
<tr>
<th>ID No.</th>
<th>Charting</th>
<th>Mapping</th>
</tr>
</thead>
<tbody>
<tr>
<td>C01</td>
<td>//I got worried about choking or something. //I thought if you broke the lock then you could never get out.</td>
<td>2 Catastrophic belief from tale. Physical catastrophe</td>
</tr>
<tr>
<td>C02</td>
<td>//I get scared that there’s someone inaudible. When I turn my light on, I don’t normally turn it back off because I’m scared there’s just going to be a shadow. //That you’re going to fall off the stairs and smash your face.</td>
<td>2 Catastrophic physical injury, horror-type fear</td>
</tr>
<tr>
<td>C03</td>
<td>//I get worried I will get on the wrong plane. //cos it you watch movies with people like that, you might think they’re in your house. //You could, somebody might look like your mum or dad and you might follow them and they might be strangers.</td>
<td>3 Mistakes with catastrophic implications, separation from parents, influence of films</td>
</tr>
<tr>
<td>C04</td>
<td>//It’s not ok for me because I always have a feeling I’ll get lost. //Because if you get lost, your parents might think you’re right next to them when you’re not.</td>
<td>2 Getting lost. Constant feeling of impending catastrophe, separation from parents</td>
</tr>
<tr>
<td>C05</td>
<td>//And you get to like, and you start worrying, you get to like extreme conclusions so like ‘if this happens, this happens, this might happen to me and I might be ill’ and stuff like that so like jumping to bad conclusions. //Like you’re doing new things and like adventurous things and you’re not doing the zip wire and stuff like that and like rock climbing and you’re worried that you’re...like what’s going to happen so if you do this, you’re worrying about the worst situation, so if you’re doing rock climbing and the rope snaps and you fall and you can’t do anything about it and you just break your legs //Like extremist things. So like what’s been on the news lately and stuff like, if you don’t mind me saying, terrorism and stuff like that, it just, going to extremes again thinking about... //Or someone just like breaks into the house or something like that //Yeah and something happens. Someone comes into the house and they’re like, obviously my mum doesn’t know what’s happening and then I’m downstairs and someone’s come up to me and something.</td>
<td>5 Bad conclusions, train of thought. Terrorism. House break in. Influence of media.</td>
</tr>
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## Appendix Q

### Example coded transcript (parent focus group transcript)

<table>
<thead>
<tr>
<th>Parent focus group transcript</th>
<th>Final framework code</th>
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<tbody>
<tr>
<td>P12: Just two things, one which you mentioned was and I just wondered if the other... long before X was diagnosed, something that was really peculiar about him as a child was that he would as he started to learn to walk, he was a little bit late, not terribly, he would put everything to make sure that it was solid and safe and we used to laugh about it, he never had accidents like other toddlers would, he would always put something. We thought this was very funny, that he was so risk averse but now looking back, whether he was even in himself aware, but the other thing also was long before he was diagnosed is that my wife had picked up on he would become very anxious about doing things so if he was somewhere and you said “right ok we’re going now X”, he would have a meltdown whereas if you just give him a few minutes, you know say “in ten minutes we’re going to leave” then invariably you wouldn’t have a problem and it was just constantly, and I know speaking with other parents this is something that does come out a lot, it’s sort of, you know, they’re actually much more adaptable but you can’t just spring surprises because that suddenly... “but this isn’t what I was expecting”, you know, “I’m doing this” but if you give lots of warning then it becomes a lot easier to manage and so now that becomes part of your life doesn’t it, you’re just constantly explaining what’s going to happen, so if we’re going away, he’s probably comfortable about this as long as we’re giving him lots of “right ok, so this will be different, that will be different, but it will be exciting”, and he’ll take that on board but, surprises, no.</td>
<td></td>
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<tr>
<td></td>
<td>1(f): Anxieties about Duchenne-related change, needs, and medical procedures</td>
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<tr>
<td></td>
<td>1(d): The unexpected and unfamiliar</td>
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<tr>
<td></td>
<td>1(c): Extreme distress</td>
</tr>
<tr>
<td></td>
<td>3(b): Family adapting, coping, and responding</td>
</tr>
<tr>
<td>P14: Yeah even for us, like simple things, like if I pick X up from school, and then “where are we going now?” “We’re going home, oh no, I need to stop at the shop”, that’s a big deal. You say to him ?Inaudible?, I’ll go to the shop, I’ll get back, like you’re saying, but it’s not in their plan and it could be something as trivial as you need to go and buy milk or something but that is the distressing for them. His brother would probably moan I but it wouldn’t be distressing for him, he’s going “fine, we’re doing that” but not like, “why didn’t you tell me?”, like almost nagging at you because it’s not been to his... so I’d say that would be the main difference for us anyway.</td>
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<tr>
<td></td>
<td>2(b): Individual adapting, coping, and responding</td>
</tr>
<tr>
<td></td>
<td>1(d): The unexpected and unfamiliar</td>
</tr>
<tr>
<td></td>
<td>1(c): Extreme distress</td>
</tr>
<tr>
<td>P11: I think I saw a big difference with my kids. X (son with Duchenne) is the oldest so again it’s quite tricky to know, not having anyone to compare him with, but then my daughter was just so much more sociable and less worried about things and you could do things spontaneously with her, that was a big thing about lack of spontaneity that I remember when the kids were little, I just used to find that a bit wearing at times to be honest, but yeah, it’s just lots of explaining and also she used to feel that he never spoke to her about how he felt about her, she used to be moaning. “X doesn’t love me” because he just wasn’t interested, and still isn’t really interested in talking about his feelings, but anyway, that’s different from being anxious, I guess but yeah, I definitely saw a difference and my other two were always a lot less worried about... happy to do stuff physically whatever it was.</td>
<td></td>
</tr>
<tr>
<td></td>
<td>1(d): The unexpected and unfamiliar</td>
</tr>
<tr>
<td></td>
<td>1(c): Extreme distress</td>
</tr>
<tr>
<td></td>
<td>3(a): Family emotional and physical responses</td>
</tr>
<tr>
<td>P14: I think X is very emotionally immature, he has meltdowns like crying, screaming almost, and it could be that something happened to the iPad or something like that and I try to say, “you know you’re going to be 10, do you think 10 year old boys scream and cry like this?”</td>
<td></td>
</tr>
<tr>
<td></td>
<td>1(c): Extreme distress</td>
</tr>
<tr>
<td></td>
<td>3(b): Family adapting, coping, and responding</td>
</tr>
</tbody>
</table>
Appendix R

Validit check request email to child participant and transcript excerpt with feedback\(^6,7\)

Hi X,

I hope you are well and you had a good time over Easter. You may remember me from last summer when you came to do a focus group in London with three other boys who have Duchenne and we talked about anxiety. You told us lots of really interesting things. I have been studying everything that was said in the focus groups since last summer and I have been trying to work out what the most important things were so that I can tell other people what we found.

I have come up with some categories to try and describe what you and the other people said, but I want to know what you think about these categories. I want to know if you think they describe what you were talking about or not.

I have typed up everything that was said in the focus groups and I have attached to this email 4 short sections from your focus group with things that you said. Your number is C6 and you will see I have highlighted this in yellow. Other people have different numbers. There are no names so that no one knows who took part apart from me.

If you would like to help me, I would like you to look at the 4 short sections of your focus group and think about the questions I have written for you on the right side of the attachment. There are no right and wrong answers, I am interested in anything you think! Please let me know if I haven't described what you need to do very well.

If you don't want to help that is ok. I asked your mum if I could email you and she said you might be able to help, but it is totally up to you! You can send me an email back with your answers or we can speak on the phone if that is easier. My number is XXXXXXXXXX.

I look forward to hearing from you,

Rachel Trimmer

\(^6\) Child feedback is written in red.
\(^7\) Themes in excerpt do not necessarily represent the final framework due to validity checks occurring at an earlier stage of analysis.
### Section 1:

C6: When I’m irritated or anxious, that’s something, I just play games and then I just forget.

RT: So doing some sort of activity that takes your mind off...

C6: Yeah it’s quite good to make my mood better.

### Section 2:

C7: I think this will be a big worry, ‘worries about not being able to do things that other people can do’, that would be a big worry for me

C6: Yeah

RT: What do other people think about that?

C5: I don’t think about that

RT: That’s not a big worry for you

C5: I just push it out the way and just don’t think about it

C6: I’d say, yeah

C8: Others will do what they have to do, I don’t get involved

RT: That sounds like for some people that’s quite a big worry

C7: Yeah

RT: We can put it up here even though I know that’s not the case for everyone

C6: I don’t really find it a worry because I understand that sometimes I’m not going to be able to do everything, that sometimes it’s ok to let people do stuff that you can’t do because otherwise people could get annoyed with the fact that just because you’re there they sort of have to stop doing stuff they can’t do, that you can’t do

C8: I mean other people feel bad themselves

C6: Yeah

C7: Yeah

C8: They want to involve you but they can’t

C6: But they can’t and sometimes they will say ‘oh why’, sort of like, it’s not a worry if they’re kind of like okay about it cos usually they understand.

### Questions for T

1. T, I have called this blue text, “Adapting and coping” - are you talking about how you adapt and cope here? Or would you describe it differently? Yes it’s about coping.

2. T, in this blue section, are you again talking about ways that you adapt and cope? It’s not ‘coping’ it’s my outlook – I don’t have to be the same as everyone else.

3. I’ve put this orange text into a category called “Support and solidarity” Solidarity is another word to describe people who stick with you when something is difficult. Is this what you were talking about here? Or would you use other words to describe it? Solidarity is a good word – some people understand more than others.
Section 3:

RT: Oh so worrying how he might look to other people?

C7: Yeah

C6: Yeah about doing stuff that you sort of have to do in front of other people, like maybe in the swimming pool, being hoisted in and then there’s a...

C5: Everyone looking at you.

C6: Yeah.

C7: Or like getting out of your chair and looking a bit weird because you can’t get up yourself.

RT: So situations where there might be lots of other people around and they might be...

C7: Yeah

C6: Yeah who don’t kind of understand, especially little children who have a habit of staring at you because they don’t understand.

C7: Yeah.

Section 4:

RT: Is there anything else before we finish for the break is there any other things that haven’t been mentioned that you’ve just held in mind, things that make you feel particularly scared or worried, or things that happen?

C6: Yeah, I get quite worried or anxious if I’m doing something I haven’t done before.

C7: Yeah

C5: Yeah

C6: Like going in the taxi on your own or...

C7: Or going to school on your own, and people like...

C6: Or going to an event you haven’t been to before and you’re worried about will it have access, will there be accessible toilets?

C5: Or just like...

C6: Will the stuff I want to see be accessible?

4. I have put this purple text into a category called “Social anxiety” which means worrying about fitting in and worrying about what other people will think about you.

Is this what you were describing here? Or is it something else? Yes the purple text is social anxiety.

5. I have put this orange text into a category called “Unhelpful responses from others”. Does this describe what you were talking about? Not intentional so not ‘unhelpful responses’ I would call it misinformation and misunderstanding.

6. I have put this purple text into a category called “Anxiety about the unexpected and unfamiliar” Is this what you were talking about here? Or was it something else? Yes I agree it’s definitely about the unexpected.

7. I have put these last 2 bits of purple text into a category called “Age-related change, needs and limitations”. Do you think what you were saying here fits into this category? I have been anxious about going places since I’ve been in a wheelchair – it’s mainly about wheelchair access.
Appendix S

Validity check request email to parent participant

Dear X,

Thank you again for agreeing to help with the analysis stage of the DMD and anxiety study. I have attached two things for you to have a look at if you can. The first is a draft framework which aims to describe the focus groups as a whole. The second is a transcript excerpt from the group that you attended.

Brief information about the framework:

- The draft framework aims to capture the important themes arising from all 5 focus groups (3 parent groups and 2 child groups)
- Each theme in the framework has a title that I have come up with and an alternative title in italics taken directly from focus group text.
- The second page gives you more details about what is being included under each theme.

I’d like to hear your thoughts about the following:

- What do you think about how I have coded the transcript excerpt?
- Is there anything you think could improve how themes in the transcript are captured?
- What do you think about the structure and language of the framework?
- Is it a good representation of:
  a) Your experiences of Duchenne and anxiety in Duchenne in particular
  b) What was discussed in the focus group you attended
- Is there anything that would make the framework better in your opinion?

I’ll call you next Tuesday at HHMM to talk about this briefly on the phone. Let me know what number is best to call you on.

Many thanks,
Rachel Trimmer
*Trainee Clinical Psychologist, UCL*