‘Understanding the lived experience and psychosocial needs of children and young people with Juvenile Dermatomyositis’

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Declaration

I, Polly Livermore, 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.
Abstract

Background

Whilst causing profound muscle weakness and skin rashes, with no cure and a risk of mortality, the rare autoimmune disease ‘Juvenile Dermatomyositis’ can also have severe psychosocial implications.

Aim

To determine the psychosocial needs of children and young people with Juvenile Dermatomyositis and understand how these can be addressed more fully in the future.

Method

An exploratory, sequential design employed four phases. Hermeneutic phenomenology guided interviews with children and young people over the age of eight (Phase 1). A suite of questionnaires sought to establish resonance from early findings with children from 15 centres across the United Kingdom (Phase 2) and clinical teams completed a survey about their services and delivery of psychosocial care (Phase 3). A dissemination and intervention workshop was the final phase (Phase 4).

Results

The lived experience of Juvenile Dermatomyositis, described across 15 interviews was encapsulated in the metaphor of a ‘rollercoaster’. Data from 123 questionnaire respondents revealed that the population reported lower emotional distress than a normative population, with 81% feeling supported. Where emotional distress was reported it was correlated with uncertainty and a perception of burden. Concerns about school and lack of peer support emerged as new findings. All 40 professionals surveyed described doing their best to consider psychosocial needs, although services were limited in all centres. At the dissemination workshop the 33 participants agreed that school support and peer support must be prioritised.

Conclusion

The study offers new insights from a range of perspectives, built incrementally over four phases, refining the rollercoaster model and detailing where psychosocial care would be best placed to support children and young people. The mnemonic SAFE was introduced to support professionals in implementing these findings: Support, Ask, Friends, Education, reinforcing the importance of asking children and young people how they are coping, whilst living with Juvenile Dermatomyositis.
There is very little known about the psychosocial needs of children and young people with Juvenile Dermatomyositis. This rare, autoimmune condition has many known biological ramifications on the body, but what about the psychological and social aspects? In clinical practice, healthcare professionals, have witnessed the ongoing struggles of children and young people across the United Kingdom with this debilitating disease and its impact on psychosocial well-being. Reports from family members concurred with this perception. This study has sought to establish the nature of these concerns, giving preference to the voice of children and young people.

The significant amount of data presented is complex and in places, contradictory, although this serves to highlight the depth and breadth of this study. Children and young people with Juvenile Dermatomyositis around the UK report lower emotional distress than a healthy, normative population and lower levels of uncertainty than a sample of children with rheumatic disease. A further 81% of children and young people responded positively to having enough support and 49% of health professionals believed their centre was providing excellent or close to an excellent psychosocial service. There are still some areas of concern, including the 40% of our cohort who scored over the recommended clinical cut off for emotional distress. This 40% reported a higher perception of burden and more uncertainty (<0.001***), than others with Juvenile Dermatomyositis.

Children, young people and their families are key to benefit from this work. Recommendations proposed evidence the importance of involvement and always asking children and young people how they are coping. The mnemonic SAFE: Support, Ask, Friends, Education was developed to help professionals address some of these issues, for example by finding out how supportive school and peers are being. The visual image of the rollercoaster model will aid discussions with families, about the potentially turbulent pathway ahead. Other resources, such as the poetry outputs, could encourage supportive conversations within the family.

Health care professionals will benefit from this work. Certainly, from the presentations at meetings and conferences, both nationally and internationally,
Impact statement

health professionals have openly declared how the study findings have informed their practice. Presenting at such events has elevated the importance of considering psychosocial needs and not just accepting them as part of a chronic, life-threatening condition. Increasing this awareness is vital, as with any rare condition, and should lead to earlier diagnosis and prioritisation of psychosocial needs. Two manuscripts have been published in peer-reviewed journals, and further papers are planned, to evidence the importance of unmet psychosocial needs.

Key stakeholders will benefit from this work. The booklet for teachers will be the first JDM teachers’ resource available worldwide. Relationships will continue with pivotal charity groups nationally and internationally and extend into subsequent planned research collaborations. Maintaining networks with colleagues in Canada will ensure the development of a peer mentoring programme becomes a reality. Producing a ‘Standard of Care’ for those with rare, orphan diseases, will be key to aiding stakeholders to fight for further psychological support and challenge the current disparity of care.
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# Table of contents

<table>
<thead>
<tr>
<th>Section</th>
<th>Page</th>
</tr>
</thead>
<tbody>
<tr>
<td>DECLARATION</td>
<td>2</td>
</tr>
<tr>
<td>ABSTRACT</td>
<td>3</td>
</tr>
<tr>
<td>IMPACT STATEMENT</td>
<td>4</td>
</tr>
<tr>
<td>ACKNOWLEDGMENTS</td>
<td>6</td>
</tr>
<tr>
<td>TABLE OF CONTENTS</td>
<td>7</td>
</tr>
<tr>
<td>LIST OF TABLES</td>
<td>17</td>
</tr>
<tr>
<td>LIST OF FIGURES</td>
<td>19</td>
</tr>
<tr>
<td>LIST OF ABBREVIATIONS</td>
<td>21</td>
</tr>
<tr>
<td>CHAPTER 1: BACKGROUND AND LITERATURE REVIEW “MAPPING THE JOURNEY AND PLANNING THE ROUTE”</td>
<td>23</td>
</tr>
<tr>
<td>1.1 OVERVIEW OF INTRODUCTION CHAPTER</td>
<td>24</td>
</tr>
<tr>
<td>1.2 INTRODUCTION TO THE RESEARCH</td>
<td>24</td>
</tr>
<tr>
<td>1.2.1 Why now, why me, why here?</td>
<td>24</td>
</tr>
<tr>
<td>1.2.2 The population: Juvenile Dermatomyositis (JDM)</td>
<td>25</td>
</tr>
<tr>
<td>1.2.3 The context: Juvenile Dermatomyositis Cohort Biomarker Study (JDCBS)</td>
<td>26</td>
</tr>
<tr>
<td>1.2.4 The priority: Why not ask the children?</td>
<td>27</td>
</tr>
<tr>
<td>1.3 MAPPING THE JOURNEY - WHAT TO BEGIN WITH?</td>
<td>29</td>
</tr>
<tr>
<td>1.3.1 Reflecting on the reflexivity</td>
<td>30</td>
</tr>
<tr>
<td>1.3.2 My journey: my pre-suppositions interview</td>
<td>31</td>
</tr>
<tr>
<td>1.4 REVIEWING THE LITERATURE WITHIN THE QUALITATIVE RESEARCH PARADIGM</td>
<td>33</td>
</tr>
<tr>
<td>1.4.1 Scoping review</td>
<td>35</td>
</tr>
<tr>
<td>1.4.2 Scoping findings</td>
<td>44</td>
</tr>
<tr>
<td>1.4.3 Advantages of the scoping review for this study</td>
<td>51</td>
</tr>
<tr>
<td>1.4.4 Limitations of the scoping review</td>
<td>51</td>
</tr>
<tr>
<td>1.4.5 Conclusion of the scoping review</td>
<td>52</td>
</tr>
<tr>
<td>1.5 TERMINOLOGY USED IN THE THESIS</td>
<td>52</td>
</tr>
<tr>
<td>1.6 ORGANISATION OF THE THESIS</td>
<td>54</td>
</tr>
<tr>
<td>1.7 CONCLUSION</td>
<td>57</td>
</tr>
<tr>
<td>1.8 THE NEXT CHAPTER</td>
<td>57</td>
</tr>
</tbody>
</table>
Table of contents

CHAPTER 2: METHODOLOGY “THE BEST METHOD OF TRANSPORTATION” ................................................................. 59

2.1 OVERVIEW OF CHAPTER .......................................................................................................................... 59
2.2 INTRODUCTION ........................................................................................................................................ 59
2.3 MIXED METHODS ..................................................................................................................................... 60
  2.3.1 Why undertake mixed methods research? ......................................................................................... 60
  2.3.2 What does this mixed methods study look like? ................................................................................ 62
    2.3.2.1 Priority ......................................................................................................................................... 63
    2.3.2.2 Implementation ............................................................................................................................. 63
    2.3.2.3 Integration ................................................................................................................................... 66
  2.3.3 What are the theoretical perspectives underpinning this study? ...................................................... 67
    2.3.3.1 The a-pragmatic stance ............................................................................................................... 69
    2.3.3.2 The multiple paradigm ................................................................................................................ 69
    2.3.3.3 The single paradigm .................................................................................................................... 69
2.4 THIS STUDY .............................................................................................................................................. 70
  2.4.1 Research Question/s. .......................................................................................................................... 71
  2.4.2 Research Aims ..................................................................................................................................... 71
2.5 STUDY DESIGN ....................................................................................................................................... 72
2.6 METHODOLOGY ...................................................................................................................................... 73
  2.6.1 An introduction to phenomenology .................................................................................................. 73
  2.6.2 Phenomenology philosophical perspectives ....................................................................................... 74
  2.6.3 Appropriateness of hermeneutic phenomenology for this research ............................................. 77
  2.6.4 Rigour in hermeneutic phenomenology ............................................................................................ 78
2.7 METHODS ............................................................................................................................................... 80
  2.7.1 Interviews for phase one .................................................................................................................... 80
  2.7.2 Questionnaires for phase two ........................................................................................................... 81
  2.7.3 Surveys for phase three ....................................................................................................................... 82
  2.7.4 Workshop for phase four ................................................................................................................... 83
  2.7.5 Juvenile Dermatomyositis Cohort Biomarker Study (JDCBS) ......................................................... 84
  2.7.6 Patient and public involvement and engagement (PPIE) .................................................................... 84
2.8 CONCLUSION ............................................................................................................................................. 86
2.9 NEXT CHAPTER ....................................................................................................................................... 86

CHAPTER 3: PHASE 1 “BEGINNING THE JOURNEY” ................................................................................. 88

3.1 INTRODUCTION ....................................................................................................................................... 88
3.2 CONTEXT ................................................................................................................................................... 88
3.3 RESEARCH QUESTION ............................................................................................................................ 89
3.4 AIM ......................................................................................................................................................... 89
3.5 METHODS ............................................................................................................................................... 89
3.5.1 Setting .................................................................................................................. 89
3.5.2 Ethical procedure ................................................................................................. 89
3.5.3 Participant population ......................................................................................... 90
  3.5.3.1 Inclusion criteria ................................................................. 90
  3.5.3.2 Exclusion criteria ................................................................. 90
3.5.4 Participant recruitment ....................................................................................... 90
  3.5.4.1 Preparation ........................................................................ 90
  3.5.4.2 Identify and approaching ..................................................... 90
  3.5.4.3 Consenting ......................................................................... 91
  3.5.4.4 Sample size ....................................................................... 91
3.5.5 Interview procedure .......................................................................................... 91
3.5.6 Creative methods ............................................................................................... 92
  3.5.6.1 Choosing the methods ......................................................... 93
  3.5.6.2 Chosen Methods .................................................................. 93
    3.5.6.2.1 Icebreaker ................................................................. 93
    3.5.6.2.2 The main phase .......................................................... 94
    3.5.6.2.3 The concluding phase ................................................ 96
3.5.7 Action in event of distress ............................................................................... 96

3.6 ANALYTICAL APPROACH ............................................................................... 97
  3.6.1 Data analysis in hermeneutic phenomenology .............................................. 97
  3.6.2 The process ................................................................................................. 98

3.7 RESULTS ............................................................................................................. 100
  3.7.1 Participants ................................................................................. 100
  3.7.2 Sample demographics .............................................................................. 100
  3.7.3 Data collection ....................................................................................... 101
  3.7.4 Data analysis ....................................................................................... 103
    3.7.4.1 Considerations throughout the analysis ................................... 104
  3.7.5 Early findings ...................................................................................... 104
  3.7.6 Working with the data ............................................................................ 104
  3.7.7 ‘Being-on-the-JDM-rollercoaster’ ......................................................... 107
  3.7.8 Emerging themes .................................................................................. 109
  3.7.9 Themes examined ............................................................................... 109
    3.7.9.1 Being-confused ................................................................. 110
    3.7.9.2 Being-different .................................................................. 112
    3.7.9.3 Being-sick, steroidal and scared ........................................ 114
    3.7.9.4 Being-uncertain ................................................................. 115
    3.7.9.5 Being-accepting ................................................................. 117

3.8 YOUNG PEOPLE’S REFERENCE GROUP WORKSHOP .................................. 118
3.9 DISCUSSION ..................................................................................................... 123
3.10 STRENGTHS .................................................................................................. 126
3.11 LIMITATIONS ................................................................................................. 127
3.12 IMPLICATIONS FOR PRACTICE ................................................................. 127
**Table of contents**

3.13 CONCLUSION ........................................................................................................... 128
3.14 NEXT CHAPTER ...................................................................................................... 128

**CHAPTER 4: POETRY “CREATIVE DETOUR” ......................................................... 130**

4.1 INTRODUCTION .................................................................................................... 130
4.2 THE FIRST POEM ................................................................................................. 131
4.3 THE ART OF CREATING ‘RESEARCH POEMS’ ..................................................... 133
4.4 THE PROCESS OF CREATING ‘RESEARCH POEMS’ ............................................. 136
4.5 THE ONGOING PROCESS OF CREATING MORE .................................................. 140
4.6 SHARING WITH YOUNG PEOPLE ....................................................................... 143
4.7 SHARING WITH HEALTH CARE PROFESSIONALS ............................................. 145
4.8 PERSONAL REFLECTION OF MY JOURNEY WITH POETRY .................................. 146
4.9 STRENGTHS .......................................................................................................... 146
4.10 LIMITATIONS ...................................................................................................... 147
4.11 CONCLUSION ....................................................................................................... 148
4.12 NEXT CHAPTER .................................................................................................... 148

**CHAPTER 5: PHASE 2 “PICKING UP MORE PASSENGERS” ......................... 150**

5.1 INTRODUCTION .................................................................................................... 150
5.2 BACKGROUND ....................................................................................................... 150
5.3 DESIGN .................................................................................................................. 151
  5.3.1 Creating the theory ......................................................................................... 151
  5.3.2 Questionnaire selection ................................................................................. 154
5.4 RESEARCH QUESTION ....................................................................................... 155
5.5 AIM ......................................................................................................................... 156
5.6 HYPOTHESES ....................................................................................................... 156
5.7 METHODS .............................................................................................................. 157
  5.7.1 Setting ............................................................................................................. 157
  5.7.2 Ethical procedure ........................................................................................... 157
  5.7.3 Specific considerations .................................................................................. 157
  5.7.4 Participant population .................................................................................... 158
  5.7.4.1 Inclusion criteria ....................................................................................... 158
  5.7.4.2 Exclusion criteria ..................................................................................... 158
  5.7.5 Participant recruitment .................................................................................. 158
    5.7.5.1 Preparation .............................................................................................. 158
    5.7.5.2 Identifying and approaching ................................................................. 159
    5.7.5.3 Recruitment .......................................................................................... 160
    5.7.5.4 Sample size ........................................................................................... 160
5.7.6 Questionnaire delivery ................................................................. 160
5.7.7 The suite of questionnaires ............................................................ 161
5.7.7.1 Demographic and clinical data collected ....................................... 161
5.7.7.2 Validated measures employed ....................................................... 162
  5.7.7.2.1 The Pediatric Quality of Life Inventory (PedsQL) ....................... 163
  5.7.7.2.2 The Pediatric Quality of Life Rheumatology Module .................. 163
  5.7.7.2.3 Paediatric Index of Emotional Distress (PI-ED) ......................... 164
  5.7.7.2.4 Child Uncertainty in Illness Scale (CUIS) ................................ 164
  5.7.7.2.5 Benefit and Burden Scale for Children (BBSC) ....................... 165
5.7.7.3 JDM experience questionnaire ................................................... 166
5.7.8 Action in event of raised PI-ED score ............................................ 167

5.8  ANALYTICAL APPROACH .................................................................. 168
5.8.1 Analysis of the validated questionnaires .......................................... 168
  5.8.1.1 Exploratory and descriptive analysis ........................................... 168
  5.8.1.2 Univariate analysis .................................................................. 169
  5.8.1.3 Multivariable analysis ............................................................... 170
5.8.2 Analysis of the free-text response in the JDM experience questionnaire .. 171

5.9  RESULTS ......................................................................................... 172
5.9.1 Target population and response ...................................................... 172
5.9.2 Sample response by centre ............................................................. 174
5.9.3 Demographic and clinical characteristics, including comparison of responders and non-responders .......................................................... 175
5.9.4 Missing data .................................................................................. 176
5.9.5 Outcomes data ............................................................................... 177
5.9.6 Comparison of study data with previously published reference data .... 178
  5.9.6.1 Pediatric Quality of Life (PedsQL) published data ....................... 179
  5.9.6.2 Rheumatology Module published data ......................................... 180
  5.9.6.3 Paediatric Index of Emotional Distress (PI-ED) normative data .... 181
  5.9.6.4 Childhood Uncertainty in Illness Scale (CUIS) previously published data .......................................................... 182
  5.9.6.5 Benefit and Burden Scale for Children (BBSC) published data .... 182
5.9.7 Raised Paediatric Index of Emotional Distress (PI-ED) scores .......... 183
5.9.8 Univariate analysis ........................................................................ 184
  5.9.8.1 Analysis of continuous demographical data and clinical variables .... 185
  5.9.8.2 Analysis of categorical; clinical, demographic and experience data .......................................................... 187
    5.9.8.2.1 Demographic variables ......................................................... 190
    5.9.8.2.2 Clinical variables ............................................................... 190
    5.9.8.2.3 Experience variables ........................................................... 190
  5.9.8.3 Relationships between outcomes and independent variables .......... 194
5.9.9 Multivariable analysis .................................................................... 198
  5.9.9.1 PedsQL – TOTAL Score ............................................................. 199
  5.9.9.2 Pain domain in the Rheumatology Module .................................. 199
  5.9.9.3 Worry domain in the Rheumatology Module ............................... 200
  5.9.9.4 Communication domain in the Rheumatology Module ................. 201
  5.9.9.5 Treatment domain in the Rheumatology Module ......................... 202
CHAPTER 6: PHASE 3 “THE VIEW FROM OTHERS ON THE JOURNEY” 234

6.1 INTRODUCTION .................................................................................................................. 234
6.2 CONTEXT ............................................................................................................................... 235
6.3 RESEARCH QUESTION ....................................................................................................... 236
6.4 AIM .................................................................................................................................... 236
6.5 ETHICAL PROCEDURE ...................................................................................................... 236
6.6 METHODS ........................................................................................................................... 237

6.6.1 Setting ............................................................................................................................ 237
6.6.2 Participant population .................................................................................................... 237
6.6.3 Participant recruitment ................................................................................................. 238
Table of contents

6.6.4 Surveys .............................................................................................................. 238
6.6.4.1 What survey mode(s) will be used to ask the questions? ......................... 238
6.6.4.2 Is this question being repeated from another survey, and/or will answers be compared to previously collected data? ................................................................. 239
6.6.5 Will respondents be willing and motivated to answer accurately? ................... 239
6.6.6 What type of information is the question asking for? ....................................... 239
6.6.7 Survey set up ...................................................................................................... 240
6.6.8 Piloting the surveys ............................................................................................ 244

6.7 ANALYTICAL APPROACH .............................................................................. 244

6.8 RESULTS .............................................................................................................. 244
6.8.1 Response rate to the surveys .......................................................................... 245
6.8.1.1 Medical professionals .............................................................................. 245
6.8.1.2 Nurse specialists .................................................................................... 245
6.8.1.3 Clinical psychologists ............................................................................ 245
6.8.2 Site data (Q1 and Q2 for the medical professionals) ...................................... 245
6.8.3 Clinical psychology provision ........................................................................... 246
6.8.3.1 Medical professional responses (Q3 – Q9) .............................................. 247
6.8.3.2 Nurse specialists (Q1 – Q7) .................................................................. 248
6.8.3.3 Clinical psychologists (Q1 – Q7) ............................................................ 250
6.8.4 Role specific questions ..................................................................................... 252
6.8.4.1 Medical professionals (Q10) ................................................................ 252
6.8.4.2 Nurse specialists (Q8 and Q9) ............................................................... 253
6.8.4.3 Clinical psychologists (Q8 and Q9) ....................................................... 254
6.8.5 School liaison / Peer-2-Peer opportunities .................................................... 255
6.8.6 Challenges (Q13 for medical professionals, Q12 for nurse specialists and Q10 for clinical psychologists) ............................................................. 257
6.8.7 Rating of service (Q14 for medical professionals, Q13 for nurse specialists and Q11 for clinical psychologists) ......................................................... 258
6.8.7.1 Medical Professionals .......................................................................... 258
6.8.7.2 Nurse specialists ................................................................................... 259
6.8.7.3 Clinical psychologists .......................................................................... 259
6.8.8 Triangulation of results .................................................................................... 259
6.8.9 Any other comments? (Q15 for medical professionals, Q14 for nurse specialists and Q12 for clinical psychologists) ......................................................... 261

6.9 DISCUSSION .................................................................................................... 262

6.10 IMPLICATIONS FOR PRACTICE .................................................................. 265

6.11 STRENGTHS ..................................................................................................... 266

6.12 LIMITATIONS ................................................................................................... 266

6.13 CONCLUSION .................................................................................................. 267

6.14 NEXT CHAPTER ............................................................................................... 267
CHAPTER 7: PHASE 4 “A REFLECTION OF WHERE WE’VE BEEN AND WHERE STILL TO GO” .................................................................................................269

7.1 INTRODUCTION ...........................................................................269
7.2 WHY DO FAMILIES ENGAGE IN RESEARCH? ............................270
7.3 PATIENT AND PUBLIC INVOLVEMENT AND ENGAGEMENT (PPIE) .........................................................................................270
  7.3.1 Prior to phase four ..................................................................270
7.4 PHASE FOUR: DISSEMINATION AND INTERVENTION WORKSHOP ..........................................................................................273
7.5 WHY USE A WORKSHOP MADE UP OF TWO FOCUS GROUPS? ..............................................................................................274
7.6 RESEARCH QUESTION ..................................................................275
7.7 AIMS ............................................................................................275
7.8 ETHICAL PROCEDURE ..................................................................276
7.9 FOCUS GROUP ONE ......................................................................276
  7.9.1 Evidence for the need to advise schools ..................................276
7.10 FOCUS GROUP TWO .....................................................................277
  7.10.1 Evidence for the need for peer mentoring ..............................277
7.11 METHODS ................................................................................277
  7.11.1 Workshop Participants ..........................................................277
  7.11.2 Setting of the workshop ..........................................................278
  7.11.3 Procedure for the workshop ....................................................279
  7.11.4 The materials and topics for the focus groups .........................280
    7.11.4.1 Dissemination presentation ................................................280
    7.11.4.2 Materials discussed ..........................................................280
7.12 RESULTS ..................................................................................281
  7.12.1 Participants who attended the workshop ...............................281
  7.12.2 Focus group one .................................................................281
  7.12.2.1 Children and young people group ......................................281
  7.12.2.2 Parent Group ...................................................................282
  7.12.2.3 Both groups ......................................................................283
  7.12.3 Focus group two .................................................................285
7.13 DISCUSSION ..............................................................................288
7.14 OUTPUTS: SCHOOL BOOKLET ....................................................288
7.15 OUTPUT: PEER MENTORING ......................................................290
7.16 IMPLICATIONS FOR PRACTICE ..................................................292
7.17 STRENGTHS ..............................................................................292
7.18 LIMITATIONS ............................................................................293
7.19 CONCLUSION ...........................................................................293
7.20 NEXT CHAPTER ..........................................................................293
CHAPTER 8: DISCUSSION, INTEGRATION, CONTRIBUTION AND REFLECTIONS “ARE WE NEARLY THERE YET?” ...........................................295

8.1 INTRODUCTION ................................................................................................................................. 295
8.2 SUMMARY OF THE STUDY .................................................................................................................. 295
  8.2.1 Phase one .................................................................................................................................... 296
  8.2.2 Phase two .................................................................................................................................... 296
  8.2.3 Phase three ................................................................................................................................... 297
  8.2.4 Phase four ................................................................................................................................... 297
8.3 INTEGRATION ..................................................................................................................................... 298
  8.3.1 Following the thread .................................................................................................................... 299
  8.3.2 Adapting the rollercoaster in light of the new threads and new knowledge ......................... 300
8.4 WHAT IS THE CONTRIBUTION FROM THIS RESEARCH? ............................................................... 302
8.5 RECOMMENDATIONS FOR CLINICAL PRACTICE ......................................................................... 305
8.6 RECOMMENDATIONS FOR FUTURE RESEARCH .......................................................................... 307
8.7 STRENGTHS OF THIS RESEARCH .................................................................................................. 307
8.8 LIMITATIONS ..................................................................................................................................... 308
8.9 DISSEMINATION ................................................................................................................................. 309
  8.9.1 With patients and families .......................................................................................................... 310
  8.9.2 With health care professionals .................................................................................................. 311
  8.9.3 With key stakeholders .................................................................................................................. 311
8.10 REFLEXIVE PERSPECTIVE ............................................................................................................. 312
8.11 CONCLUSION ................................................................................................................................. 314

REFERENCE LIST ....................................................................................................................................... 317

APPENDIX 1 – AN EXAMPLE OF THE REFLEXIVE DIARY ................................................................. 356
APPENDIX 2 – A SECTION OF THE ‘CREATIVE METHODS CRITIQUE’ ...........................................359
APPENDIX 3 – ETHICS APPROVAL LETTER FOR PHASE 1 .............................................................. 369
APPENDIX 4 – 8-12 PHASE 1 INFORMATION SHEET .......................................................................... 371
APPENDIX 5 - 13 – 19 PHASE 1 INFORMATION SHEET ..................................................................... 372
APPENDIX 6 - QUESTIONS CONSIDERED FOR PHASE 1 ............................................................... 375
APPENDIX 7 – SECTION OF INTERVIEW PROTOCOL ....................................................................... 377
APPENDIX 8 – INTERVIEW SCHEDULE ............................................................................................... 384
Table of contents

APPENDIX 9 – THE SHARED STORIES.................................................................386
APPENDIX 10 - COMMENTS IN RESPONSE TO POEMS ..................................405
APPENDIX 11 - ETHICS APPROVAL LETTER FOR PHASE 2 .......................406
APPENDIX 12 – QUESTIONS/THOUGHTS CONSIDERED FOR PHASE 2 ......408
APPENDIX 13 - FLOW SHEET FOR LOCAL NURSES ....................................414
APPENDIX 14 - POSTCARD SENT TO ALL IN JDCBS .....................................415
APPENDIX 15 - 8-12 PHASE 2 INFORMATION SHEET ..................................416
APPENDIX 16 - 13-19 PHASE 2 INFORMATION SHEET ...................................417
APPENDIX 17 - VALIDATED QUESTIONNAIRE SUMMARY .........................419
APPENDIX 18 - MISSING DATA .................................................................420
APPENDIX 19 – SCATTERPLOTS OF OUTCOME MEASURES .......................422
APPENDIX 20 - SCATTERPLOTS OF RHEUMATOLOGY MODULE .................423
APPENDIX 21 - COMMENTS TO Q1 .............................................................424
APPENDIX 22 - COMMENTS IN RESPONSE TO Q8 .................................427
APPENDIX 23 - COMMENTS IN RESPONSE TO Q9 .....................................440
APPENDIX 24 - PSYCHOLOGY TOOLS MENTIONED ...................................443
APPENDIX 25 - HEALTH CARE PROFESSIONALS CHALLENGES ............444
APPENDIX 26 - PROMPT QUESTIONS - SCHOOL FOCUS GROUP ............447
APPENDIX 27 - PROMPT QUESTIONS - MENTORING FOCUS GROUP ..........448
APPENDIX 28 - PRESENTATIONS GIVEN .......................................................449
APPENDIX 29 - PEDIATRIC RHEUMATOLOGY PUBLICATION ..................453
APPENDIX 30 – JOURNAL OF POETRY THERAPY PUBLICATION ...............454
List of tables

Table 1-1 Scoping review results
Table 1-2 Type of study identified from scoping review
Table 2-1 Showing the disparity between the two major paradigms
Table 2-2 Comparative terminology to determine rigour/trustworthiness
Table 3-1 Reasons which influenced the selection of creative methods
Table 3-2 Data analysis steps undertaken
Table 3-3 Demographic details of the 15 participants
Table 5-1 Categorical data of responders/non-responders - Chi-squared test
Table 5-2 Continuous data of responders/non-responders using T test
Table 5-3 Mean score and ranges for all respondents for each measure
Table 5-4 Comparisons of this study data set with published PedsQL data
Table 5-5 Comparisons with Rheumatology Module published data
Table 5-6 Comparisons of this study data with PI-ED normative data
Table 5-7 Comparisons of this study data with CUIS published data
Table 5-8 Comparison with BBSC published data
Table 5-9 Those that scored over cut off threshold on PI-ED measure
Table 5-10 Pearson correlation coefficient for the continuous data
Table 5-11 Comparisons of categorical binary data and QOL questionnaires
Table 5-12 Categorical binary data and validated questionnaires
Table 5-13 Experience questionnaire compared with QOL questionnaires
Table 5-14 Experience questionnaire compared to validated questionnaires
Table 5-15 Correlations between outcomes and patient perception variables
Table 5-16 Correlation with the CUIS and perception of burden and benefit
Table 5-17 PI-ED clinical cut-off scores for QOL
Table 5-18 PIED clinical cut-off scores for CUIS, Benefit and Burden
Table 5-19 Analysis of PedsQL total score for all 123 respondents
Table 5-20 Analysis of the Pain domain for all 123 respondents
Table 5-21 Analysis of the Worry domain for all 116 respondents
Table 5-22 Analysis of the Communication domain for all 123 respondents
Table 5-23 Analysis of the Treatment domain for 116 respondents
Table 5-24 Analysis of the ADL domain for 120 respondents
Table 5-25 Analysis of the PI-ED of 123 respondents
List of tables

Table 5-26 Results from Q1 ..............................................................206
Table 5-27 Results from Q2 ..............................................................207
Table 5-28 Results from Q3 ..............................................................208
Table 5-29 Results from Q4 ..............................................................210
Table 5-30 Results from Q5 ..............................................................212
Table 5-31 Results from Q6 ..............................................................212
Table 5-32 Results from Q5 and Q6 combined .........................213
Table 5-33 Coding results of Q7 .......................................................215
Table 5-34 Primary code allocated for each of the comments given for Q8 ....217
Table 5-35 Each hypothesis in turn, accepted or not and evidence for this ....222
Table 6-1 Bespoke questions asked of the three health care professionals ...241
Table 6-2 Medical professional responses to Q1 and Q2 ................246
Table 6-3 Information given to schools ........................................255
Table 6-4 Social interaction opportunities ....................................256
Table 6-5 The main challenge as declared by each professional ..........257
Table 6-6 Rating summary provided by the 40 respondents ........258
Table 6-7 Presenting the rating per centre for each professional ......260
Table 7-1 Children and young people’s school experiences ..........282
Table 7-2 Schooling questions asked of both groups and comments ....283
Table 7-3 Mentorship questions for both groups and comments answered ...285
Table 8-1 Recommendations for clinical practice from each phase ....306
Table 8-2 Overall limitations ...........................................................309
List of figures

Figure 1-1 PRISMA diagram showing the scoping inclusion............................37
Figure 1-2 Newspaper article from 11th January 2016..................................50
Figure 1-3 Defining the three domains of health..........................................53
Figure 1-4 Presenting the road map plan of this thesis.................................56
Figure 2-1 Summarising considerations for research design.........................62
Figure 2-2 Mixed methods research designs..............................................64
Figure 2-3 Mixed methods design of this study..........................................72
Figure 1-1 A drawing representing a participants experience of JDM.............102
Figure 3-2 Mind-map of interview text.....................................................105
Figure 3-3 Themes emerging as stories are plotted next to each other...........105
Figure 3-4 Initial Rollercoaster drawing..................................................107
Figure 3-5 The JDM rollercoaster (Livermore et al., 2019)..........................108
Figure 3-6 Spider chart from one participant.............................................120
Figure 3-7 One young person’s thoughts on the rollercoaster.......................121
Figure 3-8 Another young person’s thoughts on the rollercoaster...............121
Figure 5-1 Re-considering the rollercoaster.............................................152
Figure 5-2 Our Juvenile Dermatomyositis Psychosocial Model....................153
Figure 5-3 Flow diagram of patients included or excluded in the study...........174
Figure 5-4 Percentage response rate from each of the 15 centres...............175
Figure 5-5 Radial list to present coding to each qualitative comment of Q1...206
Figure 5-6 Radial list to present coding to each comment for Q3.................209
Figure 5-7 Radial list to present coding allocated to each comment to Q4.....211
Figure 5-8 Radial list to present coding to each comment in Q6.................214
Figure 5-9 Word cloud presenting the worst thing about JDM...................219
Figure 6-1 Medical professionals responses to the five binary questions......247
Figure 6-2 Nurse specialist binary questions asked....................................249
Figure 6-3 Psychologists answer to patients being screened in advance......251
Figure 6-4 Medical professional response to the question about enough time253
Figure 6-5 Nurse specialist responses to binary questions; Q8 and Q9........254
Figure 7-1 Illustrating patient and public involvement so far.......................271
Figure 7-2 Newsletter sent to all JDCBS participants..................................272
Figure 7-3 Invite and agenda for the morning..........................................279
List of figures

Figure 8-1 Following the threads through this study ........................................300
Figure 8-2 Adapted rollercoaster in light of new knowledge..........................302
Figure 8-3 Dissemination strategy .................................................................310
Figure 8-4 Completed roadmap (for now!) ......................................................315
List of abbreviations

ADL Activities of Daily Living
ANOVA Analysis of Variance
ARMA Arthritis and Musculoskeletal Alliance
BBSC Benefit/Burden Scale for Children
BFSC Benefit Finding Scale for Children
BSPAR British Society of Paediatric and Adolescent Rheumatology
BSR British Society of Rheumatology
CAMHS Child and Adolescent Mental Health Services
CATIS Child Attitude Toward Illness Scale
CDI Children’s Depression Inventory
C-HAQ Childhood Health Assessment Questionnaire
CHQ Child Health Questionnaire
CI Confidence Interval
Com Communication
CUIIS Childhood Uncertainty in Illness Scale
CYP Children and young people
EMG Electromyography
HEE Health Education England
HRQOL Health Related Quality of Life
JDCBS Juvenile Dermatomyositis Cohort Biomarker Study
JDM Juvenile Dermatomyositis
JIA Juvenile Idiopathic Arthritis
JRD Juvenile Rheumatic Disease
JSLE Juvenile Systemic Lupus Erythematous
MeSH Medical Subject Headings
MUIS Mishels Uncertainty in Illness scale
MREC Multicentre Research Ethics Committees
NHS National Health Service
NA Not Applicable
NIHR National Institute of Health Research
PE Physical Education
PedsQL Pediatric Quality of Life Inventory
Phy Physical
Physio Physiotherapy
PI Principal Investigator
PI-ED Paediatric Index of Emotional Distress
PPIE Patient and Public Involvement and Engagement
PRQL Pediatric Rheumatology Quality of Life
Psych Psychological
QOL Quality of Life
R&D Research and Development
SD Standard Deviation
SENCo Special Educational Needs Coordinator
SF-36 Short Form 36
SLE Systemic Lupus Erythematous
SOES Stages of Exercise Scale
SPSS Statistical Package for the Social Sciences
UK United Kingdom
US United States
VAS Visual Analogue Scale
WTE Whole Time Equivalent
“Why me?”

Why me?

It’s so unfair,

Why me?

It hurts everywhere,

Why me?

It hurts to walk,

Why me?

I don’t wanna talk,

Why me?

I miss my life before,

Why me?

My life before I got sore.
Chapter 1: Background and literature review

“Mapping the journey and planning the route”

“Pediatricians and psychiatrists long have recognized that many psychosocial problems facing children with long-term physical disorders can lead to social disability far more serious to an individual than the direct effects of his physical ailment.” (Mattsson, 1972, p. 801).

Being diagnosed with a chronic health condition as a child often has sequelae way beyond that of the physical ramifications, as the quote above from nearly fifty years ago highlights. However, even after all this time, it remains unclear how much we understand and how far we address these psychosocial problems; or are they simply an anticipated and accepted consequence of physical illness? This research aimed to explore the psychosocial implications from one such chronic health condition, Juvenile Dermatomyositis, in order to expand our understanding of its impact in order to describe fully the support needs of children and young people with this disease.

In undertaking this research, and navigating the complexity of the methodological journey that was taken, what has resulted is a different kind of thesis, in style and content. The end-result is enhanced by the arts, underpinned by qualitative and quantitative data. Research poems are interspersed throughout the chapters (these are discussed further in Chapter four), a visual roadmap illustrates the journey and there are frequent additions of narrative from personal reflections. Whilst this research is set in the context of a sequential exploratory mixed methods study, the reader will identify a ‘leaning’ towards the science of phenomena, the use of phenomenology to study ‘experience’. The initial phase was a phenomenological study, and as discussed later, the difficulties of interconnecting phenomenology with more conventional methodologies and methods was encountered on numerous occasions. Despite these challenges, these early methodological decisions have influenced the conduct and approach to this research that has placed the experience of children and young people with Juvenile Dermatomyositis centre stage.
Background and literature review

"Mapping the journey and planning the route"

1.1 Overview of introduction chapter

This chapter presents the current body of understanding. It begins with an introduction to the research to demonstrate the clinical origins of the research question, the context in which it was undertaken and the rationale for the research. Next, an exploration of literature reviewing practices for qualitative studies is described. This is followed by the results from a scoping review, which sought to ask what is known about the psychosocial needs of children and young people with Juvenile Dermatomyositis. This review of the literature leads to the research question and aims, which shape the thesis and terminology used throughout. Finally, this chapter concludes with an overview of the thesis and presents a unique visual roadmap to orientate readers to what is to follow.

1.2 Introduction to the research

This research is unique. It asked children and young people to recount their experience of Juvenile Dermatomyositis, and it used creative methods so that we could listen to them, include them, and then act upon what they told us. It also offered the opportunity for all children and young people aged eight years and upwards throughout the United Kingdom (UK), to share stories of their JDM. This has not been done before. The strength of this research is the emphasis on listening to their voices throughout the research.

1.2.1 Why now, why me, why here?

The idea for this research came out of years of extensive clinical practice: beginning as a junior nurse working on a paediatric rheumatology ward, through many subsequent roles and more recently as a Matron overseeing clinical care. I undertook numerous periods of study, beginning with a diploma, followed by a degree, then nurse prescribing and a Masters, and through each of these, I increased my research knowledge, capabilities and interest. It was after my Masters study that I decided to pursue my research idea to ultimately improve clinical care. Further personal reasons for this are explored in detail later in this chapter during a pre-suppositions interview as prescribed by phenomenological
Background and literature review

“Mapping the journey and planning the route”

studies. Within my clinical practice, I have worked closely with a Professor in Paediatric Rheumatology, who is the Chief Investigator for a research group and repository study, which collects data on the patient group of interest. Inviting me to join the research group and have access to the Juvenile Dermatomyositis Cohort Biomarker Study (JDCBS) has made this research possible (more detail on the JDCBS, later in this chapter).

1.2.2 The population: Juvenile Dermatomyositis (JDM)

This research involves children and young people who have a diagnosis of Juvenile Dermatomyositis. Juvenile Dermatomyositis, is the most common idiopathic inflammatory myopathy of childhood, however, it still remains very rare, with an annual incidence in the UK of between 0.8-4.1 per million children per year (Symmons, Sills and Davis, 1995; Feldman et al., 2008; Martin, Li and Wedderburn, 2012; Almeida et al., 2015). Classic cutaneous manifestations include: 1) Gottrons papules, often over knuckles, elbow and knees, 2) heliotrope rash over eyelids, 3) facial malar rash, and 4) nailbed capillary changes (Kim et al., 2017). Patients frequently present with proximal muscle weakness, although in rare cases it can present without (amyopathic dermatomyositis) (Lowry and Pilkington, 2009). Juvenile Dermatomyositis may also involve other muscular systems, such as, the heart, lungs and gastrointestinal tract (Gowdie et al., 2013); especially when treatment is delayed (Feldman et al., 2008).

Diagnostic criteria are historically defined by Bohan and Peter (Bohan and Peter, 1975a, 1975b) as presence of one of the defined characteristic rashes, combined with three of either: symmetrical proximal muscle weakness, raised serum muscle enzymes, abnormal findings on muscle biopsy, and electromyography (EMG) for a definite Juvenile Dermatomyositis diagnosis. Two of these and a typical rash result in a probable Juvenile Dermatomyositis diagnosis (Feldman et al., 2008; Wedderburn and Rider, 2009). This autoimmune disease of childhood has a mean age of onset of seven years (Batthish and Feldman, 2011; Martin, Li and Wedderburn, 2012), a male-to-female ratio of 1:5 (Lowry and Pilkington, 2009) and is associated with significant morbidity and mortality (McCann et al., 2015). Whilst mortality rates are reported to have decreased dramatically over the last
three decades and are now between 2-3% (Huber et al., 2000; Ravelli et al., 2010; Patwardhan et al., 2012), many children and young people continue to have active disease and irreversible damage from their disease and/or treatment into adulthood (Ravelli et al., 2010). As a study by Stringer, Singh-Grewal and Feldman, (2008) found, only 50% of their patients with Juvenile Dermatomyositis achieved remission after 4.5 years despite early, aggressive treatment with immunosuppressive medications. From here on in, Juvenile Dermatomyositis will be abbreviated to JDM for ease of reading.

1.2.3 The context: Juvenile Dermatomyositis Cohort Biomarker Study (JDCBS)

This research is set within the context of the Juvenile Dermatomyositis Cohort Biomarker Study (JDCBS). This is a longitudinal, inception cohort study of children diagnosed with JDM, collecting clinical and biological data, with linked samples from diagnosis and serially over time (McCann et al., 2006; Martin et al., 2011). The JDCBS, established in the year 2000 in London, collects new and ongoing patient data stored in an electronic database. Progress in understanding the causes and basic underlying autoimmune mechanisms of JDM is hampered by the small numbers of patients in any one centre at a given time. A network of clinical and research centres, such as the JDCBS, collaborating and combining their patient numbers, will facilitate advances more easily.

The JDCBS aims to recruit all reported cases of JDM in children and young people (n=628 as of 10th March 2020) around the UK. At the time of writing, this study has recorded over 6,000 clinic visits, over 2000 blood samples and over 150 muscle biopsies, from seventeen contributing tertiary paediatric rheumatology departments in National Health Service (NHS) hospitals. When this PhD study began, there were 15 centres in the JDCBS and it was with these that I initially engaged with, this has now risen to 17 at time of thesis completion. It is also important to note here, that whilst the JDCBS aims to collect data on every child and young person diagnosed with JDM across the UK, there will be some families who are not in the registry. There will be those who either did not consent
to join, or whom were diagnosed in a centre outside of one of the main tertiary centres – these numbers are not known, but anticipated to be small.

The aims of the JDCBS are to:

1. Determine the demographics of JDM.
2. Define disease presentation, activity, damage and response to current medication use.
3. Determine prognostic biomarkers.
5. Develop a cohort of patients suitable for recruitment to clinical interventional trials.
6. Facilitate international collaborative trials in JDM.
7. Create a consented repository of data and sample collection to investigate the immunological and genetic abnormalities of JDM with a view to identifying future therapeutic targets.

This psychological study therefore, contributes a novel and additional perspective, namely a focus on the psychosocial well-being of those with JDM and how those needs might be better met, in clinical practice.

Each site within the JDCBS has Multicentre Research Ethics Committee (MREC) approval. Each has its own local Research and Development approval (R&D), each has a Principal Investigator (PI) and most have a named research nurse working on the study.

1.2.4 The priority: Why not ask the children?

As Bray, Kirk and Callery, (2014) draw attention to, there is a lack of research exploring children and young people’s experiences of chronic illness. Children with JDM are no exception, and have generally been excluded from all research attempting to examine psychosocial need. The research evidence, as will be discussed later in this chapter, is minimal, and what exists, is directed towards the parents. However, there is a wealth of evidence that proxy reporting by
parents is not the same as asking young people themselves (Theunissen et al., 1998; Waters, Stewart-Brown and Fitzpatrick, 2003; Shaw, Southwood and McDonagh, 2006; Davis, Mackinnon and Waters, 2011; Lal et al., 2011; Sattoe, van Staa and Moll, 2012). The research to date, tells us that parents generally underestimate the impact of health-related quality of life compared to their children. In a study by Waters, Stewart-Brown and Fitzpatrick (2003) adolescents were found to be much less optimistic about their health and well-being than their parents and more likely to be sensitive to pain, mental health problems and the impact of their health on family activities. Asking for proxy reporting can also be confounded by the parents psychological state, with depression being found to negatively correlate with parent proxy health reports (Davis, Mackinnon and Waters, 2011).

Over at least the last two decades, key legislation has helped propel children’s rights into the spotlight. The United Nations Convention on the Rights of the Child (United Nations Convention on the Rights of the Child, 1989), the Children Act 1989 (HMSO, 1989) and the Children Act 2004 (HMSO, 2004) in England and Wales and counterparts in other countries, have set up a legal requirement to consult children and young people when assessing their needs (Farrell, 2005; Greig, Taylor and MacKay, 2007). For example, the United Nations Convention on the Rights of The Child, article 12, states that children in accordance with their age and maturity, have the right to express their views freely in all matters affecting them (United Nations Convention on the Rights of the Child, 1989). Children and young people have a valuable contribution to research, but need to be given the opportunity and the methods to do so (Greig, Taylor and MacKay, 2007). Research with children is different to research with adults for a number of important reasons:

- There is a power imbalance between adult researcher and child participant.
- Children may lack the language to explain their point of view.
- Children are often protected by gatekeepers.
- They may need ‘tools’ and artefacts to tell their stories.
Background and literature review  “Mapping the journey and planning the route”

- Adults often think they know what is important to children and young people without the need to ask them.
  
  (Gallacher and Gallagher, 2008; Hunleth, 2011; Clark et al., 2014).

Children and young people have critical and unique perspectives on their health care experiences, but their voices are too often masked by adults speaking for, and about them (Carter et al., 2014). Therefore, doing research ‘for’ and ‘with’ children, not ‘on’ or ‘about’ them, means listening to what they have to say (Punch, 2002b; Gallacher and Gallagher, 2008), to ensure that:

"Views of children are actively received and acknowledged as valuable contributions to decision-making affecting the children’s lives"  (Murray, 2019, p. 1)

So, research can provide an opportunity for children to be heard, but are they listened to? (Hill, 2015). Listening is defined as:

"An active process of receiving, interpreting and responding to communication"  

(Clark, 2004, p. 1)

Listening is therefore an ‘active emotion’ where the listener makes meaning in an interpretive process (Clark and Moss, 2011). The listener needs to be open to the children’s voices, rather than assuming we already know (Clark and Moss, 2011; Clark et al., 2014). There must continue to be increased effort in this area to use developmentally appropriate methods to ensure children and young people can be heard in consultation and research in an appropriate and informed way (Bray, 2007).

This research provided opportunity for children to express their thoughts and feelings regarding their experience of JDM to reveal what they require in terms of their psychosocial well-being.

1.3  Mapping the journey - what to begin with?

Given the complex, multi-method approaches taken in this research this chapter has been challenging to construct. I have therefore decided to start at the
Background and literature review  “Mapping the journey and planning the route”

beginning - introduce myself, and illustrate for you: the reader, the journey I have taken so far.

1.3.1 Reflecting on the reflexivity

I have already introduced some of my personal motivations for completing this research, however, this section documents some of these reflections in further detail.

The art of reflectivity has long been acknowledged as one of the key concepts of effective person-centred nursing care (Powell, 1989; Jarvis, 1992; Bulman and Schutz, 2013). In fact, writing reflective accounts is essential for nurses as a prerequisite to their three yearly revalidation to remain on the nursing register (Nursing and Midwifery Council, 2019). Reflexivity conversely, considers the role of the researcher, being explicit about our own motivations, beliefs and thoughts throughout the study process (Dowling, 2006; Jootun, McGhee and Marland, 2009; Clancy, 2013). Reflecting on the reflexive stance of the researcher is a skill required by nursing doctoral students, throughout their entire study, but more importantly, to focus on why their study is necessary in the first place.

A constructionist epistemology heavily influenced this research, particularly in the first phase. Here, hermeneutic phenomenology is the methodology of choice. Phenomenology is an approach to qualitative research that seeks to uncover the phenomena of a given experience through a continual deep and reflexive process, beginning with that of the researcher (detailed in further depth in Chapter 2). The art of reflexivity in phenomenology is vital (Clancy, 2013), defined as: the critical self-reflection of how the researcher’s background, assumptions, positioning and behaviour impacts on the whole research process that is crucial when telling the story of the research (Finlay, 2008).

Often within phenomenology texts, an introductory chapter will present the background to the author. This chapter would present the personal pre-understandings, ‘what I bring to the inquiry’, of which there is no right or wrong answer. It is therefore important in phenomenology to highlight when I am speaking with the pronouns ‘I’ and ‘my’ as ‘I’ am part of the interpretive process,
the dialectical way I work with my data, throughout this inquiry (Crowther, 2019). The pronouns ‘I’ and ‘my’ will therefore be used throughout this thesis, and especially when working with the early data, in Chapter 3. As this research is a mixed method, multi phased study, with hermeneutic phenomenology only specifically guiding the first phase; a smaller introduction to the author is presented here, through the discussion of the pre-suppositions interview.

1.3.2 My journey: my pre-suppositions interview

Prior understanding or ‘fore-structure’ as one of the forefathers of phenomenology – Heidegger, termed it, is an essential part of the research and situates the researcher as Being-in-the-world of the participant (MacKey, 2005; McConnell-Henry, Chapman and Francis, 2009). This fore-structure cannot be bracketed out as it is part of us, but instead deep introspection reflexivity is encouraged and acknowledged (Johnston et al., 2017). Hermeneutic phenomenological research recognises that researchers have concerns and matters that they care about and have led them to their study question (Crowther, Smythe and Spence, 2015). It is important to reveal our preunderstandings, as this transparency about our backgrounds, which help the study unfold, ensures readers of the research are clear about the study’s context and possible influencing factors (Tuohy et al., 2013).

One way of sharing our preunderstandings, is to be interviewed by a fellow hermeneutic phenomenologist in a ‘pre-suppositions interview’, which asks questions such as “Tell me how you got interested in this topic?” Doing such an interview, is recognised as the hallmark of a good hermeneutic phenomenological study, after all, researchers cannot free themselves from their own unique pre understandings which will always bias their thinking (Smythe, 2011). Undertaking a pre-suppositions interview can be a difficult and challenging process but can assist to surface prior and evolving understandings in relation to the topic under study (Spence, 2016).
Background and literature review  “Mapping the journey and planning the route”

A senior hermeneutic phenomenologist at the University of Lancashire interviewed me on the 22nd May 2017. I was questioned in depth about my prior experience of nursing children with JDM:

- What happened?
- When?
- Why did it happen?
- Why was that important?
- How did that feel?
- How have you felt since?
- Where is your thinking now?

Some of the preunderstandings that emerged from this reflective experience stemmed from my experience as a newly qualified staff nurse over twenty years ago, from caring for two young children with JDM. Both of these young children unfortunately passed away. Being newly qualified I had a strong desire to provide the best nursing care possible, with a primary focus of helping people recover from illness. I had not experienced death in a patient that I was caring for before and found that my role in this situation changed. I was not the parent, but still wanted to grieve the loss of their young lives. The fact that two patients, both with the same rare disease, both around the same age, both died within a few months of each other, was extremely hard for me. I was the junior nurse on the ward at this point and was unsure about how I should feel or even, behave.

During the majority of my shifts, I cared for these two young children and felt a bond with both of them. In fact, both children I would ask to nurse each shift as I enjoyed caring for them. The first young child with JDM was quiet and withdrawn, but his family were amazing at coping with his illness. They tried hard to be positive and joyful around him, and this would have an effect on me as I nursed him. I was quite shy and quiet myself, and in my eagerness to ‘nurse’ would often be focused on the task in hand, but the family would spend time talking to me about my life and would bring in small treats for me. I felt they looked after me in a way, in return for me looking after their child. They were kind and caring towards me, which extended for many years post their child’s death. The second young
Background and literature review “Mapping the journey and planning the route”

child did not speak English and was very quiet and withdrawn most of the time. I could justify this to myself as she was unwell, in pain and in a foreign country, where she had to endure scary, painful procedures, but I felt a bond with her.

Since this time, I have nursed many other children and young people with JDM and seen a range of coping mechanisms utilised by young people. Due to the chronic nature of this condition, the children and their families are frequently reviewed in the hospital over many years. During my time nursing them, I would become familiar with parents’ first names, sibling’s names and interests, pets they had at home, what year they were in at school and so forth. At certain times, for example at disease flares, I became attuned to spotting changes in their affect and as my own confidence, experience and knowledge has continued to grow I have felt that we could be better at supporting these patients to live with their condition. This reflective engagement provided an insight into my own preunderstandings and helped illustrate how these had brought me to this research area and subsequently, this present study.

I am the British Society of Paediatric and Adolescent Rheumatology (BSPAR) lead nurse across the UK, and as part of this role, I manage a lively email group. Prior to beginning my PhD journey, I emailed these nurses to understand whether my anxieties were shared with others in other centres. The comments that were returned, confirmed that I was not alone in my concerns about the need to provide enhanced psychosocial care, with one nurse commenting that two of their patients with JDM had recently tried to take their own lives. My interest in this area, led me to partake in a quick scan of the literature, which confirmed a lack of published research examining psychosocial needs in JDM. My empathy for, and strong desire to improve psychosocial care of JDM has led me to consider how best to do this.

1.4 Reviewing the literature within the qualitative research paradigm

Once a topic for research has been selected, a literature review helps to refine ideas, improve understanding of the problem and consider appropriate research
Background and literature review  "Mapping the journey and planning the route"  

design (Walker, 2007). In the quantitative paradigm it is usual to complete a structured, rigorous, systematic evaluation to appraise and synthesize the literature results with the aim of developing a hypothesis (Fry, Scammell and Barker, 2017; Munn et al., 2018). In qualitative paradigms conversely, the literature review provides a theoretical context to determine current knowledge of a topic of interest (Klopper, 2008).

In phenomenology however, a review of the literature is recognised as being very different from both quantitative and traditional qualitative reviews, instead recommending engaging hermeneutically with a wide range of reading (Crowther, Smythe and Spence, 2014). There has also historically been much debate over the timing of this identification of current knowledge. Smythe, (2011) asserts that the literature review should be conducted after reporting the phenomena through a ‘new’ hermeneutic lens. With few rules to follow, the key purpose of a literature review here, is to provide context to the found phenomena, provoke thinking and portray the taken-for-granted meanings that make up the knowing of practice (Smythe, 2011; Smythe and Spence, 2012). Delaying the literature review until after data collection and analysis ensures the researcher does not phrase questions or look for themes which they have found in the literature (Hamill and Sinclair, 2010). The researcher can then demonstrate the attempt not to influence the data analysis and thus introduce bias (Chan, Fung and Chien, 2013). Conducted later on through reading wider literature with new understandings learnt throughout the study will reveal agreement, difference and open up new possibilities of thinking (Smythe, 2011).

This is a particular area, which I have grappled with throughout this thesis. Whilst wanting to ‘abide’ by the recommended way of conducting phenomenologically sound research and staying true to the epistemological principles of phenomenology, this is not purely a phenomenological study, but instead a mixed methods research study. I also considered whether it is possible or appropriate to leave the review of the literature until technically the end of the study. Morse (2012) also queries this process of researchers starting research studies without reviewing the literature as they risk not only limiting the rigour and quality of their
inquiries, but also the dangers of replication of a previous study (Fry, Scammell and Barker, 2017).

Hermeneutic phenomenology is based on the premise that the researcher cannot be separated from their study and therefore they naturally bring their own subjective personal knowledge and judgement, and thus already an understanding of the available literature (Walker, 2007). Researchers also are required to submit an ethical application when planning a study, which inherently requests a review of current literature (Fry, Scammell and Barker, 2017). One way round this, suggested by Chan, Fung and Chien, (2013) is to undertake some ground work to answer the question, do we understand the topic area enough to justify our research proposal and ensure our study has not been previously performed, whilst still maintaining our curiosity? A scoping review methodology emerged as the answer to this question, and is used here to scope existing published literature in a comprehensive and systematic way in order to identify key concepts and gaps (Davison et al., 2020). Scoping reviews are different to systematic reviews in that they do not appraise the evidence and consequently cannot determine whether particular studies provide robust or generalizable findings, but their aim is to ‘map’ what is known (Arksey and O’Malley, 2005).

1.4.1 Scoping review

Benefits of a scoping review include the ability to examine the breadth and depth of existing evidence from a diverse range of sources and refine the research question and search strategy (Munn et al., 2018; Davison et al., 2020).

This scoping review was carried out during 2017, prior to starting phase one. This review followed the five stages recommended by Arksey and O’Malley, (2005): Stage 1, identifying the research question, Stage 2, identifying relevant studies, Stage 3, Study selection, Stage 4, Charting the data and Stage 5, Collating, summarising and reporting the results. The optional sixth stage of consultation with external stakeholders was not undertaken for this study, as it is only instigated when it would add to the study design. As the purpose of this review was to site the planned research in the current literature arena, it was not deemed
necessary. These five stages are thus described in detail in relation to this study and the process undertaken:

Stage 1: Identifying the research question

For the purpose of the scoping review, the question was ‘What is known from the existing literature about the psychosocial well-being of children and young people with JDM?’

Stage 2: Identifying relevant studies

The selection of databases used when carrying out literature searching can have a significant impact upon the number of records retrieved. While wishing to identify all the available relevant studies to minimise potential bias, researchers also want to minimise the number of irrelevant records retrieved so the choice of databases is a key decision (Wright, Golder and Lewis-Light, 2015). Three large databases were chosen as they provided comprehensive cover between biomedical (Medline), nursing (CINAHL) and multi-disciplinary (Scopus). The search strategies were developed with the assistance of a subject specialist librarian and included : “dermatomyositis”, “Juvenile Dermatomyositis”, “JDM”, “child”, “adolescent”, “child*”, “juvenile”, “adolescent*”, “teen*”), “psychosocial”, “wellbeing” and “well-being”. The search strategy included Medical Subject Headings (MeSH) and key terms. There were no other limitations such as date or language, applied at this stage. The citation lists of all the included papers were hand searched so that any non-identified papers could be included. As in keeping with phenomenology, any other literature such as newspaper reports, letters to the editor, textbooks or policy documents were also included at this stage.
Stage 3: Study selection

It was anticipated that there would be limited research around this topic area; therefore, it was decided to not limit the search to peer-reviewed journals or to either qualitative or quantitative studies. The aim of this search strategy was to be as inclusive as possible.

The criteria decided upon were as follows:

**Inclusion criteria:**

1. Reporting on JDM.
2. Published in English.

**Exclusion criteria:**

1. Reporting on conditions other than JDM.
2. Reporting on purely adults with Dermatomyositis.

As a first step, titles were screened and all irrelevant papers excluded. Figure 1-1 illustrates the study selection further.
Stage 4: Charting the data

Electronic searches initially revealed a total of 37 articles and six texts from other sources. After removing duplicates and applying the search criteria, 19 were left for the final review. Arksey and O’Malley (2005) suggest that the author, year of publication, study location, study design, sample, aims and findings should be presented in tabular format. Table 1-1 presents this data for this scoping review.
### Table 1-1 Scoping review results

<table>
<thead>
<tr>
<th>Authors / date / Study location</th>
<th>Study design / method</th>
<th>Sample</th>
<th>Aim</th>
<th>Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Almeida et al., 2015 England</td>
<td>Review paper</td>
<td>No sample</td>
<td>A review of epidemiology, pathology, clinical presentation, diagnosis and management</td>
<td>Under the heading multidisciplinary approach, the text says that children with JDM may require input from a psychologist. No other discussion</td>
</tr>
<tr>
<td>Lowry and Pilkington, 2009 England</td>
<td>Review paper</td>
<td>No sample</td>
<td>Review of extra muscular manifestations of JDM and their relevance to prognosis</td>
<td>A paragraph highlights that patients with JDM can experience significant mood and psychological problems</td>
</tr>
<tr>
<td>Luca and Feldman, 2014 Canada</td>
<td>Review paper</td>
<td>No sample</td>
<td>To discuss health outcome measures for paediatric rheumatic diseases</td>
<td>Further work is required to determine health outcome measures and approach for eliciting the patients perceptions of their health state</td>
</tr>
<tr>
<td>Rider et al., 2011 USA</td>
<td>Review paper</td>
<td>No sample</td>
<td>To discuss disease core set measures on disease activity, quality of life and disease damage in JDM and adult dermatomyositis patients</td>
<td>Discussion of how to obtain, administer, score and interpret questionnaire tools including the Childhood Health Assessment Questionnaire (CHAQ)</td>
</tr>
<tr>
<td>Ruperto et al., 2011 Italy</td>
<td>Review paper</td>
<td>No sample</td>
<td>To identify and validate criteria for the evaluation of response to therapy in those with Juvenile Idiopathic Arthritis (JIA), Systemic Lupus Erythematosus (SLE) and JDM</td>
<td>Proposed core sets and definitions are proposed for clinical trials</td>
</tr>
</tbody>
</table>
### Table 1-1 Scoping review results

<table>
<thead>
<tr>
<th>Authors / date / Study location</th>
<th>Study design / method</th>
<th>Sample</th>
<th>Aim</th>
<th>Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wu, Wedderburn and McCann, 2017 England</td>
<td>Review paper</td>
<td>No sample</td>
<td>A review of pathogenesis, treatment and clinical monitoring</td>
<td>There is a paragraph on psychological interventions whereby the paper highlights that JDM patients, siblings and parents are vulnerable to psychological distress and could benefit from assessment/ intervention</td>
</tr>
<tr>
<td>Apaz et al., 2009 Italy</td>
<td>Quantitative – Prospective cohort study</td>
<td>272 patients with JDM and 2288 healthy children</td>
<td>To investigate the health-related quality of life change over time.</td>
<td>Patients with JDM have a significant impairment in health related quality of life (HRQOL) compared with healthy peers determined by the proxy health-related quality of life measure the Child Health Questionnaire (CHQ)</td>
</tr>
<tr>
<td>Butbul-Aviel et al., 2011 Israel</td>
<td>Quantitative – Cross sectional prospective cohort study</td>
<td>155 patients with rheumatic disease, 40 with JDM and 115 JIA (eight-16 years)</td>
<td>To determine and compare the prevalence of disturbed sleep in JIA and JDM and the relationship of sleep disturbance to pain, function, disease activity and medicines</td>
<td>Sleep abnormalities were equivalent in JIA and JDM with poor reported sleep, highly correlated with PedsQL fatigue and fatigue highly negatively correlated with quality of life</td>
</tr>
<tr>
<td>Ramsey et al., 2013 United States (US)</td>
<td>Quantitative - Cross sectional prospective cohort study</td>
<td>30 patients with rheumatic disease (7-18 years) two with JDM, 22 had JIA, two SLE, two Ankylosing Spondylitis and two other</td>
<td>To examine the association between illness-specific appraisals and depressive symptoms in those with rheumatic disease over one year</td>
<td>Findings that children’s negative illness attitudes were associated with increased child depressive symptoms. No specific discussion of JDM findings</td>
</tr>
</tbody>
</table>
### Table 1-1 Scoping review results

<table>
<thead>
<tr>
<th>Authors / date / Study location</th>
<th>Study design / method</th>
<th>Sample</th>
<th>Aim</th>
<th>Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ruperto et al., 2008, Italy</td>
<td>Quantitative – Prospective validation study</td>
<td>294 patients with JDM from 36 countries</td>
<td>To validate a core set of outcome measures for the evaluation of response to treatment in JDM</td>
<td>A core set criteria were proposed for the evaluation of response to therapy that is clinically relevant and statistically validated including the CHQ to measure psychosocial well-being</td>
</tr>
<tr>
<td>Omori et al., 2012, Brazil</td>
<td>Quantitative – prospective longitudinal study – physiotherapy focus</td>
<td>10 patients with JDM (7-17 years)</td>
<td>To prospectively investigate the efficacy and safety of a supervised exercise training program for JDM</td>
<td>Both the child self-report and parent proxy-report PedsQL scores were improved after the intervention</td>
</tr>
<tr>
<td>Pinto et al., 2016, Brazil</td>
<td>Quantitative – cross sectional cohort study – physiotherapy focus</td>
<td>19 patients with JDM (8-22 years)</td>
<td>To measure physical activity in a cohort of JDM patients</td>
<td>Physical capacity and HRQOL were reduced in JDM patients when compared with controls matched by physical activity suggesting that the disease or treatment may adversely affect overall health in JDM. Patients completed the PedsQL and parents completed proxy measures</td>
</tr>
<tr>
<td>Stephens et al., 2016, Canada</td>
<td>Quantitative – cross sectional cohort study – physiotherapy focus</td>
<td>67 patients with rheumatic disease, 15 had JDM, 39 had JIA and 13 had cystic fibrosis</td>
<td>To determine the face, content and construct validity of the stages of exercise scale in children with rheumatic conditions</td>
<td>Scores from the psychosocial measures significantly differed across the groups with the JIA and JDM group supporting good construct validity of the tool. The children completed their own CHAQ, SOES (stages of exercise scale) and Visual Analogue Scale (VAS)</td>
</tr>
</tbody>
</table>
### Table 1-1 Scoping review results

<table>
<thead>
<tr>
<th>Authors / date / Study location</th>
<th>Study design / method</th>
<th>Sample</th>
<th>Aim</th>
<th>Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Takken <em>et al.</em>, 2003 Utrecht</td>
<td>Quantitative surveys – physiotherapy focus</td>
<td>15 patients with JDM (5-14 years)</td>
<td>To study the relationships of muscle function with standard questionnaires as indicators of functioning</td>
<td>A higher body mass score was correlated with lower psychosocial functioning, but it did not correlate with muscle strength. Both HRQOL measures were by proxy: the CHQ and the CHAQ</td>
</tr>
<tr>
<td>Kountz-Edwards <em>et al.</em>, 2017 US</td>
<td>Mixed methods study with questionnaires and interviews</td>
<td>36 mothers &amp; three fathers of JDM patients</td>
<td>To better understand the functioning of families with children with JDM</td>
<td>Families of children with JDM reported difficulties in family functioning, communication problems and an increased number of conflicts</td>
</tr>
<tr>
<td>Davis, 2016 England</td>
<td>Newspaper report</td>
<td>One 10 year old with JDM and her family</td>
<td>To highlight a rare condition and present current research</td>
<td>JDM is a rare condition with no known cure and research is ongoing</td>
</tr>
<tr>
<td>Quiteles, 2017 US</td>
<td>Webpage</td>
<td>One young adult</td>
<td>To share her experience of JDM</td>
<td>One young adult describes how she always felt a burden and wants to encourage other young people to speak out</td>
</tr>
<tr>
<td>Rider <em>et al.</em>, 2007 US</td>
<td>Textbook</td>
<td>No sample</td>
<td>A chapter dedicated to coping with myositis as a family</td>
<td>The chapter describes feelings that children with JDM have and provide suggestions for decreasing the stress</td>
</tr>
<tr>
<td>Uziel <em>et al.</em>, 2002 Israel</td>
<td>Letter to the editor</td>
<td>Case study of one eight year old boy with JDM</td>
<td>To share the joint medical and psychosocial approach to his care</td>
<td>A multidisciplinary team approach and psychosocial interventions are crucial and should be an integral part of treatment</td>
</tr>
</tbody>
</table>
Stage 5: Collating, summarising and reporting the results

Quantitative research studies dominated the literature with eight studies identified. There was also one mixed methods paper, six reviews and four ‘other’ texts. There were no purely qualitative studies exploring any aspect of psychosocial needs in JDM identified. This data is summarised in Table 1-2.

<table>
<thead>
<tr>
<th>Type of study</th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Review papers</td>
<td>6</td>
</tr>
<tr>
<td>Quantitative</td>
<td></td>
</tr>
<tr>
<td>Physiotherapy led cohort studies</td>
<td>4</td>
</tr>
<tr>
<td>Prospective</td>
<td></td>
</tr>
<tr>
<td>Cohort studies</td>
<td>3</td>
</tr>
<tr>
<td>Validation studies</td>
<td>1</td>
</tr>
<tr>
<td>Mixed methods</td>
<td>1</td>
</tr>
<tr>
<td>Other literature</td>
<td></td>
</tr>
<tr>
<td>Newspaper report</td>
<td>1</td>
</tr>
<tr>
<td>Webpage ‘The Mighty’</td>
<td>1</td>
</tr>
<tr>
<td>Textbook ‘Myositis and you’</td>
<td>1</td>
</tr>
<tr>
<td>Letter to the editor</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>19</td>
</tr>
</tbody>
</table>

Psychosocial consequences of JDM is given much less attention in the literature than the physical manifestations or outcome measures and therefore identifying themes though content analysis was not possible. Instead, the scoping review results were organised according to the type of study. The final scoping review is therefore presented here in a narrative summary of the literature identified by study design, as this groups the literature together based on the research approach:

1. Reviews.
2. Quantitative studies in physiotherapy.
3. Prospective quantitative studies.
1.4.2 Scoping findings

Six review papers were identified. None of these sought to present purely psychosocial issues; however all had some mention, however minor. Three of the papers had generic discussions of JDM, including such headings as epidemiology, clinical presentation, diagnosis and management. Within this, the inclusion of psychosocial needs from JDM warranted from one sentence in one paper, to a maximum of a paragraph of text in another. For instance, Almeida et al., (2015) whilst listing the team professionals which are required to care for a child with JDM, commented that a psychologist may be required, with no further discussion as to why this may be so. Two other papers both included a paragraph on psychological concerns. Firstly, Wu, Wedderburn and McCann, (2017) state that children with JDM and their siblings and parents are vulnerable to psychological distress, including a discussion of the effects from corticosteroid therapies, school impact and poor family functioning. Secondly, Lowry and Pilkington, (2009) in three sentences in a paragraph entitled ‘Psychological’, confirm that patients with JDM can experience significant mood and psychological problems, especially with irritability and low mood, but with no references to support these claims.

Three of the review papers all set out to discuss health outcome measures, two in paediatric rheumatology (Ruperto et al., 2011; Luca and Feldman, 2014) and one in adult Dermatomyositis and JDM (Rider et al., 2011). As there are not any currently available psychosocial JDM disease measures, Rider et al., (2011) highlight generic measures such as the Childhood Health Assessment Questionnaire (CHAQ) and the Short Form 36 (SF-36) which should be used, with little discussion other than the practical application and psychometric information. The paper by Ruperto et al., (2011) aims to identify and validate criteria for the evaluation of response to therapy in clinical trials and daily clinical practice, with a sentence that highlights that a quality of life measure is
considered, through either the Child Health Questionnaire (CHQ) or the Paediatric Rheumatology Quality of Life Scale. Lastly in this section, the review paper by Luca and Feldman, (2014) state that health measures should include biological, physical and psychosocial dimensions of health, specifically stating that there are health-related quality of life measures validated in paediatric rheumatology such as the Pediatric Quality of Life Inventory (PedsQL), the Child Health Questionnaire (CHQ), the Pediatric Rheumatology Quality of Life (PRQL) scale and The Quality of My Life Questionnaire. However, they conclude that whilst there are these measures available, more work is required to understand and measure the values that are most important to children and young people with rheumatic conditions. In summary, these review papers all give minimal attention to psychosocial needs in JDM, but do highlight the recent attention given to disease measures.

2. Quantitative studies in physiotherapy

Four papers identified all had a specific physiotherapy focus. As JDM predominantly affects the muscles, one can see why there is a need for targeted physiotherapy research, however, when they included the key words psychosocial or well-being, they were picked up in the scoping review. All papers identified were quantitative papers, three out of the four only including children with JDM. In these three papers there was a relationship found with poorer psychosocial functioning: in one paper, poorer psychosocial functioning was correlated with higher body mass (Takken et al., 2003), in another quality of life scores were reduced in the 10 JDM patients compared to controls (Pinto et al., 2016) and thirdly, child quality of life scores improved from baseline after an exercise intervention (Omori et al., 2012). The fourth study was validating an exercise scale in 54 children with rheumatic disease (15 had JDM) and 13 with cystic fibrosis (Stephens et al., 2016). They found that psychosocial measures differed across the groups, but with little further discussion of this. In total, these four cohort studies only enrolled 59 patients with JDM, and all used a range of different outcome measures (CHQ, CHAQ, PedsQL generic...
and VAS). Of note, they also varied in their target audience. The paper by Takken et al., (2003) asked the parents to complete proxy measures (CHQ) and the papers by both Omori et al., (2012) and Pinto et al., (2016) compared both proxy and self-report measures of the PedsQL generic measure. The only paper that asked children and young people alone to complete measures, was that by Stephens et al. (2016) however, the measures they call psychosocial, are not standard psychosocial / health related quality of life measures, instead they used the stages of exercise scale (SOES), self-efficacy scale, process of change scale and decisional balance scale – which all predominantly assess decision making. There was limited psychosocial discussion in any of the papers.

3. Prospective quantitative studies

There were four prospective studies: one validation study and three cohort studies that were identified in the search. The validation study presented a core set of outcome measures for the evaluation of response to treatment in JDM. During the discussion, the CHQ was recommended to measure psychosocial well-being in this cohort of children as it captures aspects of disease not captured by other measures (Ruperto et al., 2008).

The three quantitative cohort studies identified, all add most so far to the identification of psychosocial well-being in JDM: one looking at sleep, one examining depressive symptoms and one focusing on quality of life.

Butbul-Aviel et al., (2011) focused on sleep and fatigue, and their relationship to pain, disease activity and quality of life. Sleep quality and quantity can be affected by psychosocial stressors in our lives, and sleep itself (or more specifically, lack of it) can be responsible for enhancing psychosocial stressors. They asked 155 patients with rheumatic disease, (40 had JDM) to complete two questionnaires evaluating sleep, one examining quality of life and some visual analogue scores for disease activity assessment. This study concluded that sleep disturbance and fatigue as measured through their
Background and literature review  “Mapping the journey and planning the route”

questionnaires, are prevalent amongst children with different rheumatic diseases and were strongly associated with increased pain and decreased quality of life. Fatigue is a particularly well recognised symptom of JDM, but this study failed to find noticeable differences between JIA and JDM. Whilst a particularly narrow focus, this study shows the relevance of asking children and young people themselves about the wider effects having a chronic disease like JDM can have on all aspects of psychosocial functioning (Butbul-Aviel et al., 2011).

Similarly the study by Ramsey et al., (2013) examined attitude toward illness and depressive symptoms in youth with juvenile rheumatic disease. Thirty patients were included (two had JDM) and asked to complete the Child Attitude Toward Illness Scale (CATIS) and the Children’s Depression Inventory (CDI), on two occasions one year apart. Whilst there is no specific mention of the two patients with JDM, the authors conclude that children’s negative attitudes towards illness were associated with increased depressive symptoms. Despite small numbers again, this paper highlights the importance of asking children and young people how they are coping, rather than making assumptions from their disease severity.

The most widely referenced paper in this scoping review was by Apaz et al., (2009), who adopted the Multinational Quality of Life Cohort Study to examine 272 children with JDM from 37 countries. They examined children’s muscle strength, global assessments and Child Health Questionnaire (CHQ) scores (completed by parents/carers). They found that compared with 2,288 healthy controls, children with JDM had a significantly lower health-related quality of life with a poorer physical and psychological well-being. They found a greater impairment in physical well-being correlated to the level of physical disability, but could not identify determinants for psychosocial well-being (Apaz et al., 2009). Patients with JDM had quality of life scores two standard deviations below the mean of healthy children for emotional behavioural and social limitations, bodily pain, parent emotional impact and family activities. The biggest limitation to this study is the lack of child-reported evaluation.
4. Mixed methods studies

The only paper that was identified throughout this review that included a qualitative element was the one by Kountz-Edwards et al., (2017). This study asked 39 parents (36 mothers and 3 fathers) of children with JDM to complete two health assessment measures (the PedsQL family impact module and the Profile of Mood States) and undergo a semi-structured interview. They found their results were split, with those who had a child in remission showing improved family functioning, whilst having a child with active disease negatively affected a parent’s mood. Parents reported that having a child diagnosed with JDM made them more compassionate and empathic, changing their worldview (Kountz-Edwards et al., 2017). This paper only included proxy reporting rather than child self-report, which limits the findings. This study is described as the family impact of having a child with JDM, but one could argue that as the child’s perspective, nor that of any siblings is included, then this cannot present the full family impact, but merely a parental perspective.

5. Grey literature

Throughout the scoping review and wider reading as recommended in phenomenology studies, other ‘texts’ identified added to the literature when considering psychosocial well-being. For example, ‘A letter to the editor’ in a highly respected rheumatology, peer reviewed journal entitled ‘JDM: Medical and psychosocial approach’ details the experiences of one eight year old boy who became bedridden and seriously unwell from his JDM, and in parallel developed a ‘depressed mood’ (Uziel et al., 2002). A psychiatrist diagnosed two major interacting problems: the physical diagnosis of JDM and a pre-existing grief reaction due to a recent loss of his grandmother. Both of these were felt to have brought about a post-traumatic stress disorder resulting in loss of interest, severe social withdrawal, frequent anger outbursts, listlessness, marked regression and sleep disturbance. Uziel et al., (2002)
Background and literature review

“Mapping the journey and planning the route”

continues to list the interventions that were instigated: daily supportive talks, intensive physiotherapy program, visit from his favourite football star and above else, they say a cooperative team approach in treating not only his body, but soul too. They conclude that psychosocial interventions are crucial and must be an integral part of treatment (Uziel et al., 2002).

“When I was diagnosed with JDM at the age of 15, I always felt as if I was a burden to people around me – my friends, family, teachers and peers. I felt as if I was only paid attention to when I was in a hospital bed, truth be told. I hated it. I was always in pain from walking up a step, speed walking to my next class and even laying down. But yet I never told anyone because I was always scared” (Quiteles, 2017, webpage).

This text is an extract from a webpage called ‘The Mighty’ where a young adult talks about her experience with JDM and her fear about admitting how she really felt (Quiteles, 2017). This young adolescent does not provide further discussion about whether she was ever asked how she felt, however, I would hope that if she had been asked and reported feeling a burden, then someone would have discussed this with her and helped her with her emotions and ability to manage her disease.

‘Myositis and You’ is a textbook published as a guide to JDM for patients and families. Page 110 presents a parent’s perspective of caring for a child with JDM, which include:

- Seeing your child unhappy or in pain and not able to take that pain away is one of the most stressful and painful experiences of a parent.
- Children have a limited understanding of the potential long-term impact of the disease, but as a parent, you might be concerned about the long-term implications and worry about the long-term future.
- Parents have to understand the illness and treatment and educate all around them, whilst supporting their child.
- Parents have a duty to ensure their child receives appropriate treatment.
Worrying about financial costs such as time away from work, travel to hospital appointments, time spent in hospital away from other family members, can cause depression and other mental health problems.

Caring for a sick child results in less time for care for other children in the family, leading to frustration, guilt and anger.

(Rider et al., 2007)

These concerns listed above are unique to a parent or carer. Most of these concerns will not be shared by the child or young person. This is really important, because as shown through this scoping review, if only the parents are asked for proxy reports, their concerns are very different to those of the child or young person.

Finally, a newspaper article from 2016 (Davis, 2016) featuring JDM had the following headline, Figure 1-2:

![Newspaper article from 11th January 2016](Image)

Described in the article is a sudden onset of a disease that rendered a previously fit, healthy and active ten-year-old child to one who could not move, was accompanied by emotional and psychological consequences, for her and her family. However, these emotional and psychological consequences of JDM are rarely mentioned in the literature, as illustrated through the scoping review results presented in this chapter.
1.4.3 Advantages of the scoping review for this study

This scoping review has followed a structured, rigorous and transparent method for mapping a complex, sparse area of literature. In keeping with phenomenological studies this review has widened the searching, to include ‘grey’ literature; textbooks, newspaper reports, web pages and letters and whilst this evidence may not be seen to be as rigorous as others, through using a clearly defined approach, it is hoped that the reader can see the method adopted achieves in-depth and broad results. After all, one of the aims of a scoping review is to include all relevant literature regardless of study design or quality (Arksey and O’Malley, 2005).

1.4.4 Limitations of the scoping review

This scoping review was conducted in 2017, and so the evidence and literature presented was prior to this time frame. The scoping review did not adopt a clear definition of ‘psychosocial’ due to the variability of terms and definitions used within the literature, which would have had an impacted upon the findings, with a range of definitions being included. As Gibson et al., (2017) highlights, scoping reviews can miss papers during the searching process, especially as only a limited number of databases are ever utilised. For this study, three databases were selected to increase breadth, especially into multidisciplinary fields. The search terms used in this study were kept purposefully narrow, but again this would have had an implication on the studies returned. For instance, we know that there is a wealth of literature examining psychosocial needs in children’s oncology care and some of this could offer insight into this study. However, the study question sought to identify what is known specifically about JDM and therefore for these purposes, we were only interested in the JDM literature. Scoping reviews also do not aim to judge quality of studies and thus the findings need to be interpreted with some caution.
1.4.5 Conclusion of the scoping review

The literature that has been identified from this scoping review is very limited. The majority sought to discuss JDM broadly, with only passing reference to the psychosocial issues of these children and young people. For the limited discussions we did find, the studies were not inclusive to children and young people. The two studies in particular that did examine quality of life and family impact purely of JDM (Apaz et al., 2009; Kountz-Edwards et al., 2017), did not ask the children and young people for their involvement. This review has illustrated how proxy reporting is not adequate when aiming to identify and understand children and young people’s experiences.

From examining newspaper reports, online media first-hand accounts, textbooks and published literature together, we can conclude that there is a large gap in what is currently known. The minimal amount of research identified has profound implications for health care professionals who have a duty of care to ensure all young people’s emotional and psychological well-being is fully supported. There were also no qualitative studies identified, supporting the argument that we have never asked children what their needs are, but instead we do it in passing or assume we already know. The aim of this scoping review was to identify the gaps when considering the psychosocial needs of children and young people with JDM.

1.5 Terminology used in the thesis

At the outset, it is important to clarify terminology, as two terms in particular can be interpreted in a variety of ways. Children and young people when used in this thesis specifically refers to an individual up to nineteen years of age, although for the actual study phases only children and young people between the ages of eight and nineteen years of age were included for practical and ethical reasons, discussed later (See 2.7.6).

‘Psychosocial needs’ are also defined here in terms of their particular reference to this study. Measuring health is complex; there is no one universal
term. It is a combination of biological, social and psychological factors with health in the centre. See Figure 1-3.

Due to the wealth of literature already examining the biological aspects of JDM on the health of children and young people (Wedderburn and Rider, 2009; Nistala and Wedderburn, 2013; Ruperto et al., 2016; Papadopoulou and Wedderburn, 2017; Huber, 2018) this study is particularly interested in the impact of the social and psychological factors combined to form the ‘psychosocial needs’ of children and young people with JDM.

The term ‘psychosocial’ is defined as pertaining to the influence of social factors on an individual’s mental health and behaviour (Vizzotto et al., 2013). ‘Psychosocial needs’ throughout this thesis will be used to represent someone’s mental, social, cultural, spiritual and developmental needs arising from emotional responses to their diagnosis, limitations, loss of abilities, complexities of treatment and impact of their diagnosis on extended family and friends (Frost, Brueggen and Mangan, 1997).
1.6 Organisation of the thesis

The metaphor of a journey is used throughout this work. The ‘journey’ signifies the path I took myself as researcher, but also the rollercoaster of JDM for the children and young people and therefore it seemed appropriate to present this thesis as a journey:

- **Chapter 1** – Background and Review of Literature “Mapping the journey and Planning the route”
  This chapter has presented an introduction to the study, specifically highlighting why this study, why now and what makes it unique. The chapter has introduced myself as researcher and the orientation I bring. A scoping review has explored a wide range of current existing literature and highlighted a gap in what is currently known.

- **Chapter 2** – Methodology “The best method of transportation”
  In Chapter two, the methodology that has guided the research from beginning to end is presented. A more detailed explanation of the epistemological underpinnings and justification of the methods used, the role of phenomenology and why a mixed methods approach was adopted is presented.

- **Chapter 3** – Phase 1 “Beginning the journey”
  In Chapter three, the work from phase one; including the background, methods, results and analysis from the qualitative interviews is explored and discussed in detail. This work was crucial to underpin the subsequent stages and direction of the research.

- **Chapter 4** – Poetry “Creative detour”
  Chapter four presents an unplanned addition to the study - the creation of poems from the children and young people’s interview transcripts. This chapter highlights the work undertaken supported by available literature, with poetry examples.
• **Chapter 5** – Phase 2 “Picking up more passengers”
  In Chapter five; the methods, results and analysis of phase two is discussed. During this phase questionnaires were sent to all children and young people with JDM across the UK and as the results are examined, current available literature is explored.

• **Chapter 6** – Phase 3 “The view from others on the journey”
  Chapter six outlines phase three, the survey of health care professionals around the UK. The survey results are presented both quantitatively and qualitatively.

• **Chapter 7** – Phase 4 “A reflection of where we’ve been and where still to go”
  The final phase is presented in Chapter seven. Phase four was a dissemination and future intervention planning workshop, made up of two focus groups of children, young people, parents, charity groups and staff.

• **Chapter 8** – Discussion and Conclusion “Are we nearly there yet?”
  Chapter eight draws together the findings from all of the phases and discusses the limitations of the study as a whole before making recommendations for practice and future research. It is hoped that the results from this research will add to the body of currently limited evidence, to prove the justification for further psychological support for children and young people with JDM.

The planned journey is illustrated visually in Figure 1-4.
Background and literature review

“Mapping the journey and planning the route”

Figure 1-4 Presenting the road map plan of this thesis.
1.7 Conclusion

This chapter has set the scene for the rest of the thesis, including why there was a need for the research, detailing personal motives and the ‘gap’ in the literature and presented the structure of the thesis to guide the reader.

1.8 The next chapter

The next chapter will present the methodology “The best method of transportation”. The aim of this is to explain the epistemological stance of the researcher, detail the mixed methods study design and explain the theoretical perspectives.
“What is it?”

Is it psoriasis? Is it eczema? Is it scarlet fever?

Why are you such non-believers?

Is it lupus? Is it growing pains?

Please help me, use your brains?

Shall we google it?

There’s got to be something that fits?

We’re made to feel we’re wasting their time,

The examining couch too high, I can’t even climb,

My face bright red,

The weight too heavy to lift my head,

I now am very weak,

An answer I seek,

Help me get a name,

Please help me, stop the pain.
Chapter 2: Methodology “The best method of transportation”

“Understanding of children, their lives and their development requires a multiplicity of methodological approaches.” (Greene and Hill, 2012 p. 4).

2.1 Overview of Chapter

The focus of this chapter is a description of the methodology and the relationship with the research question, highlighting throughout how this has shaped the study design. Opening sections introduce relevant terminology and the background to mixed methods, with specific attention given to the multiplicity of methodological approaches as Greene and Hill, (2012) highlight above. Following on from this, the theoretical perspective including the epistemological stance adopted is presented, with particular reference made to the philosophical underpinnings of this mixed methods approach. The second part of this chapter returns to the research question and describes this four-phased mixed method, exploratory sequential approach in further detail. An introduction to the methodological approach and methods employed, for all four phases of the study, are presented. The sequential nature of this study is captured here, but each phase also has a discrete chapter. Hence, Chapter two, is in essence setting the scene, in terms of the methodology, for what follows, and the methods used.

2.2 Introduction

Methodology is distinct from methods, often referred to as the principles guiding the researcher’s choice of strategy and methods for undertaking the research (Fochtman, 2008; Dyson and Norrie, 2013). The methodological approach is based on preferred research philosophy; positivist methodologies are usually quantitative in attitude, interpretivist methodologies are generally qualitative in nature and mixed methods are often a combination of the two (Dyson and Norrie, 2013). These differences will be considered in more detail
The best method of transportation”

later in this chapter. Important key terms which feature throughout this chapter, need to be defined as these guide our understanding, and include:

- Ontology – the study of being (existing).
- Epistemology – the philosophy of knowledge and how it is acquired.
- Phenomenology – the study of phenomena (Annells, 1996).

Mixed methods research has been termed the third methodological movement, taking its place alongside the traditional quantitative and qualitative research approaches (Tashakkori and Teddlie, 2003, 2010). This methodology has gained in popularity and momentum over the last twenty years as a way of using the strengths of both the standard approaches to generate new knowledge. Mixed methods research joins these two together, resulting in one combined study. Mixed methods is defined as:

> “Research in which the investigator collects and analyses data, integrates the findings, and draws inferences using both qualitative and quantitative approaches or methods in a single study or a program of inquiry” (Tashakkori & Creswell, 2007, p. 4).

The underlying principle is that neither qualitative nor quantitative approaches alone are adequate to capture the complexities under study, but when put together, they enable the research question to be answered. This process is not without challenges, but through following a structured and rigorous well-described process for applying mixed methods one can ensure that a study benefits from the application of both approaches.

2.3 Mixed methods

2.3.1 Why undertake mixed methods research?

Through combining quantitative and qualitative research components, knowledge and validity are heightened, leading to reinforced conclusions (Schoonenboom and Johnson, 2017). Greene, Caracelli, & Graham’s (1989) now thirty year old influential seminal classification devised from a
Methodology “The best method of transportation”

comprehensive empirical review, is often used to summarise five different purposes for mixed method research:

1. Triangulation: looking for convergence or corroboration of results from different methods.
2. Complementarity: uses results to seek clarification, elaboration or illustration from one method with the results from the other.
3. Development: seeks to use the results from one method to help develop or inform the other method.
4. Initiation: using different methods to look for contradictions or the discovery of new perspectives.
5. Expansion: seeks to extend the range and breadth of inquiry through different methods for different components.

In Greene, Caracelli and Graham's (1989) empirical review, complementarity was the primary purpose identified in over half of the studies, however a quarter had no purpose described (or one that could be inferred) for the mixed methods designs. Whilst some studies may have a primary and secondary purpose, conceptualising and describing this would enhance the validity of inquiry (Greene, Caracelli and Graham, 1989; Schoonenboom and Johnson, 2017). Over more recent years, this classification has attracted a lot of attention with researchers refining it. Bryman (2006) for one, has added additional rationales, such as: enhanced credibility from two approaches, instrument development, sampling and diversity of views; totalling eighteen reasons why a researcher might combine qualitative and quantitative elements. Adopting a mixed methods approach can be challenging. Reasons for doing so may be diverse, but frequent returning to the research question and considering why, what, when and how, will ensure the opportunity to bring the two sets of findings together in a meaningful integration that addresses the questions at the heart of the study (Bryman, 2007; Plano Clark, 2019).

This research set out to explore in-depth psychosocial needs from a small group of interviewees in the first study and to then examine for resonance with a larger population of young people with the same condition: JDM in the subsequent study. From the above classification therefore, the primary
Methodology “The best method of transportation”

purpose of this study was ‘Development’, explained by using the results from the interviews to help develop and inform the second method, and ‘Complementarity’ for its secondary purpose, seeking clarification from the results of the interviews with the results from the following method (Bryman, 2006). By utilising both qualitative and quantitative methods this study produced enriched understanding of what the psychosocial needs are for children and young people with JDM, which would not have been obtained had a single method been used.

2.3.2 What does this mixed methods study look like?

When designing a mixed methods research study, if consideration is given at the outset to four criteria that are implicit within all research designs: priority, implementation, integration and theoretical perspective, then the rigour of the study can be conveyed (Creswell et al., 2003). These are summarised in Figure 2-1 adapted from Creswell et al., (2003 p. 218).

<table>
<thead>
<tr>
<th>Implementation</th>
<th>Priority</th>
<th>Integration</th>
<th>Theoretical Perspective</th>
</tr>
</thead>
<tbody>
<tr>
<td>No sequence Concurrent</td>
<td>Equal</td>
<td>At data collection</td>
<td>Explicit</td>
</tr>
<tr>
<td>Sequential – Qualitative first</td>
<td>Qualitative</td>
<td>At data analysis</td>
<td>Implicit</td>
</tr>
<tr>
<td>Sequential – Quantitative first</td>
<td>Quantitative</td>
<td>At data interpretation</td>
<td>With some combination</td>
</tr>
</tbody>
</table>

Figure 2-1 Summarising considerations for research design
Each of these criteria will be presented in turn in the following section and discussed in relation to this current study.

### 2.3.2.1 Priority

Priority refers to the importance of the qualitative and quantitative methods; whether equal priority, or greater emphasis on either of the methods is placed with the other used in a secondary role (Creswell and Plano Clark, 2018). For this study, the priority dilemma can easily be answered by referring back to the research question, ‘What are the psychosocial needs of children and young people with JDM?’ (See Section 2.4). Due to a lack of available literature, the aim is to explore psychosocial concerns in this cohort of children and young people, therefore the emphasis was on the initial qualitative methods. Another seminal paper, Morse (1991), proposes a simple notation system to help represent the possible designs, with uppercase letters representing greater weight, so for this study, the notation $\text{QUAL} \rightarrow \text{quan}$ would be used. This signifies a sequential inductive project with a qualitative foundation, followed by a quantitative element (Morse, 1991).

### 2.3.2.2 Implementation

Implementation refers to whether the qualitative data and quantitative data collection and analysis occur in stages or concurrently. For this research, returning to the research question highlights the importance of attaining and analysing the qualitative interview data, to identify what factors would be examined during the larger sample by the quantitative method in the later study. The choice of implementation strategy should also include plans for data analysis and reporting. If two phases of data collection occur, then typically each phase should be analysed and reported separately, with the researcher understanding the importance on the interrelationship between the qualitative and quantitative phases of data collection (Creswell et al., 2003). The appropriate design of the study should clearly be dictated by the research question to be addressed, and with this question in mind, then identify which design is appropriate. Mixed methods research designs have been proposed by many researchers, however, the four most widely cited are;
Methodology “The best method of transportation”

1. The convergent parallel design – quantitative and qualitative data are collected simultaneously with equal emphasis.

2. The explanatory sequential design – quantitative data is collected first, followed by qualitative data that can explain the findings from the quantitative data.

3. The exploratory sequential design – qualitative data is collected first, followed by quantitative data.

4. The embedded design – both quantitative and qualitative data are collected and analysed within a traditional quantitative or qualitative design, often with one methodology added on for just a part (Creswell and Plano Clark, 2018; Taguchi, 2018). Figure 2-2 is taken from Creswell & Plano Clark, (2011) Pg 69.

![Diagram of mixed methods research designs]

Figure 2-2 Mixed methods research designs
For this current study, an exploratory sequential design was chosen. The primary purpose of this design is to generalise qualitative findings based on a small sample size, from the first to the second phase with a larger sample (Creswell and Plano Clark, 2011). The advantages of this design are that from the first exploratory phase, the findings would provide a steer towards the questions that needed to be asked in the next phase. This design is often used when there is limited data available about a particular phenomenon or area under study, and so time can be taken in the first phase to identify salient relevant factors, again, as in the case of JDM.

The major steps of the exploratory design are summarised by Creswell & Plano Clark (2011 p. 87) as:

- The researcher and the research problem are more qualitatively oriented.
- The researcher does not know what constructs are important to study.
- The relevant quantitative instruments are not available.
- There is enough time for the researcher to conduct two large phases.
- The researcher identifies new emergent research questions based on the qualitative results that cannot be answered with qualitative data alone and needs a subsequent quantitative approach.

A particular appeal of this design was the exploratory aspect: it begins in a deserted place of little available evidence, but as it builds upon the qualitative findings, the study grows in size as it then critiques its own findings throughout the quantitative phase, to integrate it back together again. In this study, the first exploratory phase was key to the selection of the questionnaires for phase two. Phase three also brought a predominantly quantitative focus when surveying health care professionals and phase four brought all phases together in the integration and dissemination of all work completed to date.

This design was not easy: it took time to conduct the initial two sequential phases, and for this reason ethical approval was sought on two separate occasions. However, the separate phases make the study logical and
Methodology “The best method of transportation”

straightforward to describe, implement and report, and the design was useful to study the findings from the first phase (Creswell and Plano Clark, 2011).

2.3.2.3 Integration

Integration refers to the combination of the qualitative and quantitative research at a defined stage of inquiry, whether during the research question, data collection, within data analysis or in interpretation of the findings (Creswell et al., 2003). These points of integration are vital, as it is at these time points that the components are mixed, hence the label 'mixed methods'. However, the term mixing can be confusing as mixing in itself is not enough, but careful integration is required (Schoonenboom and Johnson, 2017). This integration is the defining feature of mixed methods research and is what separates it from a study that just presents qualitative and quantitative information (Plano Clark, 2019).

Morse (1991) explains that good integration is about utilising the results from each phase into a cohesive and coherent outcome, achieved by being aware of and adhering to the rules and assumptions inherent in each method, the purpose of each method and contribution of the results to the overall research question. Bryman (2007) adds a more recent perspective, highlighting how true integration can be incredibly difficult, referring to some of the barriers and pitfalls, such as the researcher having greater familiarity with one of the research methods in particular or writing up the two methodologies for different audiences.

Plano-Clark (2019) advise that researchers who explicitly plan for integration at the outset have a better chance of achieving insights that arise from both the qualitative and quantitative aspects of their study. Due to following the exploratory sequential design, the results from the first phase were integrated into the planning and design of the quantitative phase, which was critical to the success of this current study. The issues and concerns that were found in phase one (which had never been described before in children and young people with JDM), directly dictated the questions to be asked in phase two and phase three, making this study unique in that the patient voice was woven
throughout the fabric of the study. Then the findings from phase one, two and three were further integrated as they were reflected upon and discussed in the dissemination workshop in phase four.

2.3.3 **What are the theoretical perspectives underpinning this study?**

The approach taken by qualitative researchers is to present in-depth insight into views to interpret and understand, whereas in contrast quantitative researchers focus on the belief that reality can be observed and measured. Creswell & Plano Clark (2011) state that it is only natural that since the exploratory design begins qualitatively, the research problem and purpose will naturally have a greater qualitative priority. The researcher will work initially from constructivist principles, only to later shift to positivism as the quantitative phase gets under way, thus asking the researcher to adopt multiple worldviews (Creswell and Plano Clark, 2011). However, appearing as polar opposites, it may be questioned whether these two are compatible. Considerations of these questions and the theoretical perspective are vital as mixed methods researchers need to consider the appropriate philosophical underpinnings to their research, to understand how they are approaching their research question. The word paradigm is often used to mean a world view, one which covers an individual’s thoughts and beliefs, the two most prominent being; constructivism or positivism. As described by Bowling (2006), paradigms consist of a set of assumptions on which the research questions are based, and these assumptions encompass stances comprising ontology, epistemology, axiology and methodology.

The two major paradigms can be further divided according to these baseline assumptions (Polit and Hungler, 1999) as detailed in Table 2-1.
Table 2-1 Showing the disparity between the two major paradigms

<table>
<thead>
<tr>
<th>Assumption</th>
<th>Positivist Paradigm Quantitative</th>
<th>Naturalistic or Constructivist Paradigm Qualitative</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ontologic (what is the nature of reality and truth? ‘The study of being’)</td>
<td>- Reality exists; there is a real world driven by natural causes</td>
<td>- Reality is multiple and subjective, mentally constructed by individuals</td>
</tr>
<tr>
<td>Epistemologic (How is the inquirer related to those being researched? ‘The theory and nature of knowledge’)</td>
<td>- Inquirer is independent from those being researched; the findings are not influenced by the researcher</td>
<td>- The inquirer interacts with those being researched; findings are the creation of the interactive process</td>
</tr>
<tr>
<td>Axiologic (What is the role of values in the inquiry? ‘The philosophical study of values’)</td>
<td>- Values and biases are to be held in check; objectivity is sought</td>
<td>- Subjectivity and values are inevitable and desirable</td>
</tr>
<tr>
<td>Methodologic (How is knowledge obtained? ‘The generation of knowledge’)</td>
<td>- Deductive process</td>
<td>- Inductive process</td>
</tr>
<tr>
<td></td>
<td>- Emphasis on discrete, specific concepts</td>
<td>- Emphasis on entirety of some phenomenon, holistic</td>
</tr>
<tr>
<td></td>
<td>- Fixed design</td>
<td>- Flexible design</td>
</tr>
<tr>
<td></td>
<td>- Tight controls over context</td>
<td>- Context bound</td>
</tr>
<tr>
<td></td>
<td>- Emphasis on measured, quantitative information; statistical analysis</td>
<td>- Emphasis on narrative information; qualitative analysis</td>
</tr>
<tr>
<td></td>
<td>- Seeks generalizations</td>
<td>- Seeks patterns- Emerging interpretations grounded in participants’ experiences</td>
</tr>
<tr>
<td></td>
<td>- Verification of researcher’s hunches</td>
<td></td>
</tr>
</tbody>
</table>

It has been said that the mixed method approach largely arose in “solution” to the intense “paradigm wars” of the 1970s and 1980’s where the positivist paradigm of quantitative research was attacked from social scientists supporting qualitative studies (Hall, 2013). Some scholars coined the term ‘incompatibility thesis’ to state that the qualitative and quantitative approaches, due to their different paradigms and opposing epistemological and ontological positions, are incompatible (Greene, 2015). If not compatible, then some scholars argued that research methods and paradigms should remain separate and not be mixed. Therefore, to proceed with this methodology, a unification of methods was needed. Six approaches were originally proposed.
Methodology “The best method of transportation”

by Teddlie & Tashakkori (2003) and later condensed to three by Hall (2013) to highlight the compatibility and show that combining paradigms is allowed and in fact, required for some studies. These are now discussed in turn.

2.3.3.1 The a-pragmatic stance

The a-pragmatic stance ignores the issue by claiming that methodology is independent of the epistemology. This approach is often disregarded as most authors believe that epistemology and methodology are related, as the position adopted inherently affects the type of data collected and interpreted (Patton, 2002; Teddlie and Tashakkori, 2003; Hall, 2013).

2.3.3.2 The multiple paradigm

The multiple paradigm claims that one or more paradigms can be used, maintaining that the mixed methods design determines the appropriateness of paradigm choice. This seems to fit with the opening statement by Creswell & Plano Clark (2011) at the start of this section. However, Hall (2013) states that the problem with this approach is that it is not always made clear when paradigms are mixed, how they are mixed and would not always be appropriate if there is antagonism between the approaches.

2.3.3.3 The single paradigm

The single paradigm approach encompasses both quantitative and qualitative approaches (Hall, 2013). Pragmatism is one of the most frequently cited individual paradigms for mixed methods research, as this approach focuses on the primary importance of the question asked and on the need for multiple methods of data collection to answer the research question (Creswell and Plano Clark, 2011). There remains debate about whether the pragmatic approach is as straightforward as described by some scholars and in fact underestimates the philosophical assumptions. However the pragmatist approach does not ignore the relevance of philosophical concepts, but it does reject the top down privileging of ontological assumptions as too narrow an approach in the philosophy of knowledge, instead they concentrate on methodology that connects issues at the abstract level of epistemology and
Methodology “The best method of transportation”

the mechanical level of actual methods (Morgan, 2007). Paradigmatic assumptions can be used as lenses for viewing the world, revealing phenomena and generating insights that would be difficult when using only one methodological background (Tashakkori and Teddlie, 2010). This linkage between pragmatism and mixed methods research has been described previously (Tashakkori and Teddlie, 2003) and summarised further by Creswell & Plano Clark, (2011):

- Both qualitative and quantitative methods may be used in one single study.
- The research question should be of primary importance, more so than the philosophical worldview or underlying methods used.
- The forced dichotomy between positivism and constructivism should be discarded.
- The use of metaphysical concepts such as “truth” and “reality” should be abandoned.
- A practical and applied research philosophy should guide methodological choices.

When considering these statements, it seems that this study does align with those listed above, in particular, the research question is focused on determining if there are unmet psychosocial needs and this did guide the methods in each phase of the study. Therefore, positioning this research within the pragmatic paradigm means the importance of the research question is elevated above that of philosophical differences adopting the “common-sense” approach to this study.

2.4 **This study**

The aforementioned considerations of methodology ultimately led to a four-phased mixed methods, exploratory sequential design with a pragmatic approach.
2.4.1 Research Question/s

The overarching research question for this study was:

“What are the psychosocial needs of children and young people with JDM?”

To answer this question, four subsidiary questions, one for each phase of the study are introduced:

1. “What is the lived experience of JDM for children and young people?”
2. “What variables are most important from phase one and are there any other psychosocial needs expressed as important in a larger cohort of children and young people with JDM?”
3. “What psychosocial support is provided in local centres and what are their perceived challenges?”
4. “How can we take these findings forward from this research so far, and improve psychosocial care even further in the future?”

2.4.2 Research Aims

This research question was guided by four specific aims, one for each phase of the study:

1. To describe and understand the lived experience of children and young people with JDM to identify their unmet psychosocial needs.
2. To investigate resonance with the themes found in phase one and to identify whether there are any other psychosocial well-being needs, in a large cross-sectional sample of children and young people with JDM throughout the UK.
3. To triangulate views on psychosocial provision from health care professionals to provide a comprehensive picture of care provision in the UK and identify any particular challenges or best practice.
4. To share findings with children and young people, parents, health professionals and key stakeholders to discuss the development of
interventions to address unmet needs and target service gaps in the future.

2.5 Study Design

Central to this four-phased mixed methods, exploratory sequential design was that each phase was informed by the previous one and subsequently enlightened the next phase, this provided a robust data set for practical integration and interpretation (See Figure 2-3).

Figure 2-3 Mixed methods design of this study
2.6 Methodology

The methodology of this study is a mixed methods design, built upon the phenomenological philosophy. This is an interpretive qualitative methodology that specifically directed the focus of the first phase, but which ultimately shaped the entire research. This next section will therefore spend some time presenting phenomenology, particularly what it brings to this study.

2.6.1 An introduction to phenomenology

Health service delivery and the research that informs it, is predominantly informed by positivists and rational measurable models, yet we do not know enough about the experience of health related events and what it is actually like to live the experience of ill health and health care provision (Spence, 2016). As highlighted in the preceding chapter, the majority of research examining psychosocial well-being in children and young people with JDM has asked parents or carers what the experience is like for the young child or young person. However, everyone’s experience is individual and only the individual can report what it is like for them. This view is in keeping with that of phenomenologists. Phenomenological research aims to understand how individuals make sense of their ‘lived experience’ (Standing, 2009) and is both a philosophy and research methodology (Lowes and Prowse, 2001; Tuohy et al., 2013; Wilson, 2015).

There are many different phenomenological perspectives with significant differences in philosophical assumptions which take time to understand, and more essentially, are important to understand. As Wilson, (2014) highlights, there has historically been criticism related to the ontological and epistemological status of phenomenology research studies, predominantly from Crotty, (1998) and Paley, (1998, 2005). Both of which highlight that without understanding the philosophical basis a robust and confident research design cannot be achieved (Wilson, 2015; Zahavi and Martiny, 2019). A comprehensive understanding of the philosophy’s basic concepts and ideas is therefore crucial to correctly adopt the research methodology, with solid philosophical assumptions underpinning the study and philosophical positions.
Methodology “The best method of transportation”

adopted by the authors clearly stated (Lowes and Prowse, 2001; Paley, 2005; Austgard, 2012).

2.6.2 Phenomenology philosophical perspectives

Phenomenology originated in the 20th century at a time when human phenomena were explored independently of the people experiencing the phenomena (Matua and Van Der Wal, 2015). Edmund Husserl (1859-1938) a German Philosopher, now often referred to as the ‘founder of phenomenology’, deliberately set out to develop phenomenology as an alternative approach that was different from the rigid methods that currently existed (Earle, 2010; Miles et al., 2013; Tuohy et al., 2013; Matua, 2015). Husserl’s concepts developed in phenomenology are important to introduce as these have been referred to and built upon by later philosophers. Husserl coined the term ‘lifeworld’ (lebenswelt) to describe human lived experience, that is what individuals experience pre-reflectively, without resorting to interpretations (Dowling, 2007). The life world is those things which are unquestioned, such as preconscious perceptions, with the aim of phenomenology to return to taken for granted experiences and to re-examine them in an intentional manner that brings to light the essence of human experience (Fochtman, 2008).

A key feature of Husserl’s Descriptive phenomenology, is to gain a pure rich description of a phenomenon without making judgements (Matua and Van Der Wal, 2015). This ‘suspension of all judgements’, Husserl called ‘epoche of natural attitude’ and required all researchers to shed their prior knowledge about the phenomena being investigated to prevent bias and preconceptions influencing the findings (Matua, 2015). To do this, the researcher suspends their preconceptions and beliefs in the outer world by disconnecting from the things around them using the metaphor of ‘bracketing’ as a means of phenomenological reduction (Koch, 1995; Sorsa, Kiikkala and Åstedt-Kurki, 2015). This idea is further explained by Husserl himself with the following comment:
Bracketing is the distinguishing characteristic of Husserlian Phenomenology and often described as a means of demonstrating the validity of the data and thus ensuring scientific rigour (Flood, 2010; Dowling and Cooney, 2012). The actual way to perform ‘bracketing’ is the topic of much debate and in practice, difficult to accomplish (LeVasseur, 2003; McConnell-Henry, Chapman and Francis, 2009; Dowling and Cooney, 2012; Zahavi and Martiny, 2019). Bracketing is an etic approach, (the view from the outside), and contrasts with Heideggerian phenomenology where the world of the participants is fused with that of the researcher in an emic approach, (an insider's view), in an attempt to co-construct reality (Hamill and Sinclair, 2010).

Martin Heidegger (1889-1976), also a German philosopher and Husserl's academic assistant, later a fellow professor, rejected Husserl's notion of phenomenological reduction. He instead insisted that we are unable to completely bracket prior conceptions and knowledge (LeVasseur, 2003). Heidegger developed his own tradition in phenomenology, now termed Interpretive phenomenology, which focuses on what humans experience rather than know (Fochtman, 2008). The reason bracketing is not congruent with the philosophical underpinning of Interpretive phenomenology, is in fact because researchers need to come to an awareness of their pre-existing beliefs and bring a “critical self-awareness” of their own subjectivity, interests and assumptions, so as to separate out what belongs to the researcher rather than the researched (Finlay, 2009).

The major distinction between the two main phenomenological research traditions – Descriptive and Interpretive, is how understanding the phenomenon is approached. Whilst Husserl focused more on the epistemological question of the relationship between the knower and object of study, Heidegger moved on to the ontological question of reality and ‘Being’ in the world (Laverty, 2017). Heidegger was interested in moving from description to interpretation, deriving meaning from being, with the researcher as a
Methodology “The best method of transportation”

legitimate part of the research, as *Being-in-the-world* of the participant (McConnell-Henry, Chapman and Francis, 2009). A significant question for Heidegger which is at the heart of his notions, is that of ‘being’. As he says himself at the beginning of his major work ‘Being and Time’:

“For manifestly you have long been aware of what you mean when you use the expression “being”. We, however, who used to think we understood it, have now become perplexed”

(Heidegger, 1962, translated by Macquarrie and Robinson, p. 1)

This is further clarified much later by an explanation by Crowther (2020). ‘Being’ and ‘being’ are used to signal a distinction between the ‘being’ of entities, such as a ‘table’ and ‘Being’ as existential. The capitalisation does not signify a higher order, but merely a way of highlighting difference. The question of the meaning of Being is about what it is that makes beings intelligible as beings (Crowther, 2019). The situated meaning of a human being-in-the-world or *dasein* as Heidegger termed it, (which translated means *being there*), is being capable of wondering about one’s own existence (MacKey, 2005). Heidegger uses the phrase *Being-in-the-world* or sometimes termed “thrownness” to refer to the way human beings are involved in the world, constructed from our own background and experiences. Pre-understanding is not something a person can step outside of, as nothing can be encountered without reference to a person’s background understanding (Laverty, 2017). Therefore, any description either retold or recorded cannot take place without interpretation by the person doing the description and the person doing the listening (Heidegger, 1962; Koch, 1996; Fochtman, 2008).

Interpretive phenomenology is often termed as ‘hermeneutics’ or ‘hermeneutic phenomenology’ (Dowling, 2004; Tuohy et al., 2013). Hermeneutics means to uncover, interpret and understand a phenomenon through language. Hermeneutics interprets the meaning of phenomena through understanding the participants perspective in the phenomena (Fochtman, 2008). Going beyond descriptive accounts of the lived experience as Descriptive Phenomenology does, but instead searching for meanings by interpreting layers of language and rich texts (Philipsen et al., 2019). This interpretation is critical to the process of understanding (Laverty, 2017).
Heidegger proposed that all beings exist hermeneutically, deriving significance in whatever is experienced in the world (McConnell-Henry, Chapman and Francis, 2009). The prior knowledge a researcher brings to a hermeneutic inquiry is essential to ensure the questions that follow are relevant, and are constantly revisited. This back and forth movement of questioning and then re-examining the texts results in an ever expanding circle of ideas, understanding the parts to understand the whole, and is termed the *hermeneutic circle* (Fochtman, 2008; McConnell-Henry, Chapman and Francis, 2009). The basic tenet of hermeneutic phenomenology is that the researcher cannot remove themselves from the meanings extracted from the text, and thus becomes part of the phenomenon (Reiners, 2012). The researcher is legitimately Being-in-the-world of the participant (McConnell-Henry, Chapman and Francis, 2009).

To generate the best ever interpretation of a phenomenon, hermeneutic phenomenologists believe the hermeneutic circle should always be implemented (Kafle, 2011). The final meanings arrived at are a blend of those articulated by participants and researcher in a process Heidegger termed co-constitutionality (Flood, 2010).

In summary, Husserl’s phenomenology is a descriptive phenomenology that requires the researcher to suspend all they know in order to deeply describe an everyday conscious experience in the lifeworld, with the aim of raising awareness. Heidegger’s hermeneutic phenomenology though, places the emphasis on the researcher as part of the process, bringing their own presuppositions and prior experience, reflected upon deeply through the hermeneutic circle to understand and interpret a phenomenon.

### 2.6.3 Appropriateness of hermeneutic phenomenology for this research

As discussed in Chapter 1.4.2, there is very little currently known about children and young people’s experiences of living with JDM, therefore the methodology chosen needs to be able to; explore, describe, identify and understand the phenomena. Phenomenology and nursing are also congruous; both ontologically subjective, both interested in an individual’s experience and both concerned about, and care for the nature of being (Dowling and Cooney,
Methodology “The best method of transportation”

2012; Moxham and Patterson, 2017). Hermeneutic phenomenology is a perfect fit for this study, which explores the phenomenon of JDM through the rich stories of children and young people with JDM and hermeneutic in that it set out to understand the experience of JDM through these interviews.

2.6.4 Rigour in hermeneutic phenomenology

Whilst it is widely acknowledged that maintaining the quality of the research process is the most crucial aspect of any research study, there is much discussion about the best ways to achieve this in a hermeneutic phenomenology study (Koch, 1994, 1996; Kafle, 2011). In the 1980’s, Guba and Lincoln introduced criteria to determine the trustworthiness of qualitative research, replacing positivist ontology and epistemology terminology for achieving rigour, such as reliability, validity, and generalizability with constructivist terminology; dependability, credibility, confirmability and transferability (Guba and Lincoln, 1989; Morse, 2015). See Table 2-2.

<table>
<thead>
<tr>
<th>Positivist terms to determine rigour</th>
<th>Constructivist terms to determine trustworthiness</th>
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<tbody>
<tr>
<td>Reliability</td>
<td>Dependability or Auditability</td>
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<tr>
<td>Validity (internal validity)</td>
<td>Credibility</td>
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<tr>
<td>Generalizability (external validity)</td>
<td>Transferability or Fittingness</td>
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<tr>
<td>Objectivity</td>
<td>Confirmability</td>
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However, whilst credibility and confirmability are often cited as expressions of trustworthiness for qualitative studies, philosophical inconsistencies prevent these from being appropriate for hermeneutic phenomenology (De Witt and Ploeg, 2006). For example, an underlying assumption of credibility is finding the epistemological single truth, whereas there is not one single truth in phenomenology (Sandelowski, 1986; De Witt and Ploeg, 2006). Similarly, confirmability, as parallel to objectivity assuring that data and outcomes are free from bias, the findings of interpretive phenomenology studies are not bias
free, but instead biases are identified and embraced (Sandelowski, 1986; Guba and Lincoln, 1989; De Witt and Ploeg, 2006).

According to Johnston et al., (2017) there are two ways to enhance methodological rigour in phenomenological studies, firstly to explore and adhere to the philosophical base, for example, the ways data are collected, experiences reflected upon and interpretations arrived at. Secondly, through the writings of the study; verbatim participant quotes and clear audit trails allow the readers to interact with the data and audit the authors’ interpretation (Johnston et al., 2017). There are other suggestions within the qualitative literature of how hermeneutic phenomenologists can establish rigour (Koch, 1996; Morse, 2015; Dibley, Williams and Young, 2020) these were implemented for this study and are listed here:

- Being open and honest about the methodological base, using philosophical terminology and referring back to the theory throughout study design, data collection, data analysis and presentation of findings.
- Keeping a reflexive diary throughout this phase of the study to aid self-awareness. Appendix 1 shows an example of the reflexive diary.
- Being open and honest about study inclusion and exclusion criteria and final recruited participants.
- Repeated visits to the data.
- Maintaining a clear audit trail of any decisions reached and how they were implemented to increase dependability.
- Holding regular team discussions.
- Having other members of the supervisory team read the transcripts and aid with data analysis for transferability.
- Presenting verbatim participant quotes throughout.
- Having a presuppositions interview at the start of the study to articulate the pre-understandings of the researcher to make bias explicit and thus aid confirmability (See Chapter 1).

The quest for hermeneutic phenomenology is not to prove or disprove, or to provide irrefutable evidence but rather to provoke thinking towards the mystery
Methodology “The best method of transportation”

of what ‘is’, and therefore thinking is ‘my’ interpretation of coming to understanding (Smythe et al., 2008). As Smythe et al., (2008) continues:

“From the very outset we put aside any claim that our research will produce objective, simplified, scientific concepts of truth” (Smythe et al., 2008 p. 1391)

The findings presented are always simply the impressions gained, an offering of thinking to engage others in their own thinking (Smythe et al., 2008). It is not possible for phenomenology to claim to reveal everything there is to know about a phenomenon (Crowther and Smythe, 2016), and so the data leads, not tells, after all, life is only ever lived as ‘my experience’ (Smythe, 2011).

2.7 Methods

So far this chapter has presented the rationale for using a mixed methods design, deliberated the particulars of this research design and presented the research question and aims. This section considers the methods selected for each phase in further detail; interviews in phase one, questionnaires in phase two, surveys in phase three and workshops in phase four.

2.7.1 Interviews for phase one

Whilst phenomenological research can use a variety of methods to obtain experiences; interviewing is typically the method of choice, with the aim of eliciting a life story or narrative about an experience, usually in a low-structured format with little or no prompting (Clarke and Iphofen, 2006). The skill is to use minimum structure and maximum depth to gain the personal perspective from the participant. Therefore, most researchers use a continuum from unstructured to semi-structured interviews, however, there is very little research about conducting phenomenological interviews with children and young people. Therefore, by adding a creative method into the interview, some of the limitations of interviewing children, such as having a limited vocabulary compared to adults (Punch, 2002b) was overcome. Extensive scoping work was performed to support the rationale for using creative participatory methods, examining the literature on a variety of creative methods and their
advantages and disadvantages; this is documented in Appendix 2, as evidence of how design decisions were reached. This process is described in further detail, including the creative methods chosen, in Chapter three.

2.7.2 Questionnaires for phase two

The aim for phase two was to identify resonance with the themes found in phase one in a large cross-sectional study of children and young people with JDM across the UK. A sub-goal was to identify if there were any other psychosocial well-being needs that had not been identified in phase one. If the early findings were synonymous with other children and young people with JDM, then the phenomena identified could be shared with health professionals and strategies to improve the issues could be discussed. However, if the findings were specific to the small number of individuals interviewed, then why was this so, and what other issues were being identified as important? These kinds of questions can only be answered by asking a larger and more representative sample of children and young people with JDM, using an approach that would enable similarities and differences to be statistically examined.

Phase two aimed to capture as many children and young people as possible and therefore needed to employ a primarily quantitative, closed-question methodology to be able to look for differences and similarities amongst participants from all over the UK. One of the aims of this study was to be inclusive to all children and young people with JDM as much as possible and provide all with the option to tell their own stories, so qualitative comments were also encouraged in places. The methodology for phase two was therefore a mixed approach, moving from the exploratory phase, now looking for resonance through the use of widely distributed questionnaires.

Questionnaires and surveys are not the same. A survey is a general methodology which can take many forms, (one of which uses questionnaires), to describe and explain information from samples to construct a quantitative description of a population. Questionnaires conversely, refer to specific tools for gathering information, often a series of questions on a narrow topic, usually
Methodology “The best method of transportation”

self-administered and are often used to collect qualitative and quantitative data (Slattery et al., 2011). Phase two was therefore approached through a series of questionnaire measures which put together as a package captured information on the items of interest from phase one. This process is described in further detail in Chapter five.

2.7.3 Surveys for phase three

The aim here was to scope current psychosocial provision for children and young people with JDM across the UK. This care at the time was provided by fifteen paediatric and adolescent rheumatology centres in the JDCBS. The survey was designed to capture the views of healthcare professionals. As in phase two, the aim of this phase was to include a large number of respondents and therefore, whilst open fields were included in the survey, the primary purpose was to collect quantitative data regarding support in each of the main 15 paediatric rheumatology centres across the UK. The aim was to provide a comprehensive picture of psychosocial care and triangulate responses from a medical professional, a nurse specialist and a clinical psychologist, in each of the centres.

Surveys collect information by asking a sample of respondents from a well-defined population to answer set questions. Surveys have many advantages over other forms of data collection:

1. They can capture a large geographical population.
2. They are usually cost effective to design and administer.
3. They can be administered through a variety of modalities.
4. They can involve complex questions and visual aids.

As with any research method, there are limitations however, such as:

1. The often low completion rates.
2. The lack of control of response situation.
3. Potential bias from targeted group.
4. General lack of engagement and rapport with sample population.
Careful planning of the three main stages; developing the questionnaire, selecting the sample and collecting the data, are crucial to the success of surveys (Blair, Czaja and Blair, 2014).

Dillman, Smyth, & Christian (2009) present four issues that need to be considered for each question added to a survey:

1. What survey mode(s) will be used to ask the questions?
2. Is this question being repeated from another survey, and/or will answers be compared to previously collected data?
3. Will respondents be willing and motivated to answer accurately?
4. What type of information is the question asking for?

These questions proved to be helpful when working through each question, especially considering the relevance of each question and the response required, for example whether to ask open or closed questions, both of which have their own drawbacks. This process will be discussed further in Chapter six.

2.7.4 Workshop for phase four

The aim of phase four was to share findings with children and young people, siblings, parents, health professionals and key charity groups, with a sub aim of discussing the development of interventions to address unmet needs and target service gaps in the future. Primarily for this phase, the focus was on dissemination of the results from the earlier phases, to share them with interested individuals. A workshop was planned with two focus groups, one for children and young people and one for their parents and carers. Charity personnel and staff mixed between the two groups and helped to facilitate discussion. The data was captured qualitatively, with lengthy discussions about future interventions.

As Troya et al., (2019) highlights, capturing lay perspectives is essential to ensure research findings inform clinical practice and this can be achieved though workshops to discuss dissemination of findings and future pathways for engagement. The workshop consisted of two focus groups, each
Methodology “The best method of transportation”

discussing a potential focus for future interventions. The focus groups were, with one for the children and young people and the other being for their parents and carers. Focus groups are often used to collect information by generating a discussion around a product, service or idea and always with real users. Thereby they produce information that is often difficult to obtain without the group dynamics stimulating further conversations and ideas. Focus groups can be held with any number of participants, but generally between six and fifteen and are often of short duration (Bowling, 2006).

2.7.5 Juvenile Dermatomyositis Cohort Biomarker Study (JDCBS)

All of the patients described in this thesis were recruited into the JDCBS (see Chapter 1.2.3). The specifics for recruitment for each phase will be detailed in each relevant chapter.

2.7.6 Patient and public involvement and engagement (PPIE)

Good, quality research needs to be inclusive and informed by a diversity of public experience and insight, so that it leads to services and treatments which reflect these needs (UK Public Involvement Standards Development Partnership, 2019). This was one of the driving forces for this study, with children and young people involved throughout the whole research journey. Their immersion from start to finish has helped to shape and improve the whole study. Initially ten young people and their families admitted to our hospital ward, were asked what one thing should be researched as important to inform future care? Addressing anxiety, low mood and other psychosocial issues were mentioned by all, and therefore a clear research priority was set. As one young person commented:

"the psychological impact of being normal one day, and then bed bound the next is massive, and no one really acknowledges this".

This research project was also discussed with the national JDM young people and Parent Advisory groups, which is made up of 13 young people (aged 14-24) and 15 parents, and some of these young people offered to be lay advisors
Methodology “The best method of transportation”

for the duration of the study. A further six young people aged 16 - 24 and six parents, discussed the project after reviewing the lay summary and agreed that psychosocial issues were a problem and needed to be given more attention as a research priority.

One of their comments included:

“Please make the age range of the project as wide as possible to include those newly diagnosed children through into adolescence and beyond as psychological needs change over time, both due to developmental reasons and issues relating to disease duration”.

All parents and young people in the room agreed with this request.

Therefore, the age range for the study was set at eight to nineteen years of age. Matza et al., (2013) highlights that at eight years of age and upwards, reliability and validity of child-self report improves. These recommendations are in keeping with reported reliability and validity data of child-report measures, which are often from ages eight years and upwards (McCann et al., 2015). This study also did not want to cause upset to young children through either receiving questionnaires asking about their emotional state blindly through the post, but also taking into account their cognitive ability (to interpret the questions), reading ability (to be able to read the questions) previous reliability (the reproducibility and consistency for the age range tested) and validation data from these tools (to ensure they are meaningful in the age ranges previously tested in). The exclusion of children below the age of eight years of age is therefore a limitation to this research.

The upper age range was more tenuous and was a topic of much discussion in the planning phases. Whilst the study wanted to be inclusive and include as many young people as possible, it also needed to be tenable. The United Nations defines adolescence as those between ten and nineteen years of age (UNICEF, 2011) and it was this definition that was finally adopted for the upper age range. Increasing the study age much further into young adulthood could produce very different psychosocial needs, support needs, emotional well-
Methodology “The best method of transportation”

being and understanding and would require adult questionnaires (which are often quite different) during later stages.

2.8 Conclusion

This chapter has presented the rationale for choosing a mixed methods approach for this study and presented the theoretical assumptions. The methodology underpinning the study has been explored, justifying why the exploratory interpretive approach of hermeneutic phenomenology was compatible with the research question and aims. The methods for each phase have been discussed in detail, demonstrating why they were suitable and highlighting key decisions made.

2.9 Next chapter

The next chapter presents phase one in further detail, beginning with the aim of phase one and how the design of this first study, set the scene for the rest of the research to follow.
“The day my JDM came to town”

My life was turned upside down,

The day my JDM came to town,

It makes me tired, it makes me weak,

I have rashes on my cheek,

There is no cure,

No one knows how I’ll be for sure,

It’s hard to explain,

But I can tell you, nothing since has been the same.

No one understands it, No one gets it,

But I have to live with it,

It makes me tired, it makes me hurt,

It makes me wanna go berserk,

There is no cure, there is no guarantee it will go away,

There is no clue how I will be on any other day.
Chapter 3: Phase 1 “Beginning the journey”

“When one walks down the hall to the drinking fountain, one experiences the activity of going to get a drink of water rather than the movements of the feet and legs, the swing of the arms, and the carriage of the torso for a defined distance on the way to getting a drink. However, in breakdown such as chronic illness, when the body is no longer able to function in the habitual way, activities are experienced differently and the nature of the lived or habitual body changes, requiring new understandings of living a life with a chronic illness” (Ironside et al., 2003 p. 176)

3.1 Introduction

This chapter takes the reader through phase one. The methodology has already been presented in chapter two, with a brief introduction to the interview methods which are prominent in this study. Here, there is a reminder of the aim of this phase in the study, a description of the methods, data collection, analysis, results and discussion. The results are presented through child and young person quotes which are critically analysed through the discussion, ending the chapter with a presentation of the key findings and implications from phase one.

3.2 Context

As the above quote highlights, chronic illness modifies day-to-day activities and how they are experienced. There is nothing magical about the term ‘lived experience’. As van Manen (2017) reaffirms, lived experience is just the name for everyday life experience – until that is, we take up the phenomenological questioning and ask “What is this lived experience like?” This is what phase one attempts to do, to find out what it is like to live with JDM, and this can only be done by listening to the voices of those who have JDM.
3.3 Research question

The research question phase one sought to address was:

- “What is the lived experience of JDM for children and young people?”

3.4 Aim

The overall aim of phase one of the study was:

- To describe and understand the lived experience of children and young people with JDM to identify their unmet psychosocial needs.

3.5 Methods

3.5.1 Setting

Children and young people who were already enrolled in the JDCBS, were approached to take part in the study. These children and young people were attending rheumatology clinic appointments and hospital visits at a Specialist Tertiary Hospital in London.

3.5.2 Ethical procedure

Phase one proceeded by obtaining a substantial amendment to the JDCBS approval (North Yorkshire REC, ref 1/3/22 Appendix 3). Age appropriate information sheets (Appendix 4 and Appendix 5), parental information sheets, consent and assent forms were all submitted for ratification. Approval was gained with no changes or clarifications required. An application was also submitted to the JDCBS Steering Group for their approval and granted with no alterations.
3.5.3 Participant population

3.5.3.1 Inclusion criteria

The inclusion criteria was kept as minimal as possible to be as inclusive as possible:

- Diagnosis of JDM.
- Recruited and consented to the JDCBS.
- Eight - 19 years of age at time of interview.
- Ability to give informed consent if over 16 years of age, or assent and parental/carer consent if under 16 years of age.

3.5.3.2 Exclusion criteria

The exclusion criteria were:

- Those with significant other co-morbidities, such as diagnoses which may change their psychosocial needs.

It was anticipated that children or young people with co-morbidities, such as severe diabetes or a diagnosed mental health illness for example, might have different psychosocial needs to their peers with JDM without such a condition.

3.5.4 Participant recruitment

3.5.4.1 Preparation

The study was discussed in depth with the clinical team, who kindly agreed to initially approach the families and discuss the study with them.

3.5.4.2 Identify and approaching

Purposeful sampling was undertaken, with children and young people, identified in advance from reviewing clinic lists and prospective ward admissions. If they matched the inclusion criteria, then they were contacted in advance and the study discussed with them. Children were recruited either when they were admitted to the inpatient ward, day unit or when they attended
Phase 1 “Beginning the journey”

The interviews took place between August 2017 and the end of November 2017.

Where possible families were contacted in advance and the study discussed by their known nurse specialist, without any coercion to participate. Information sheets were given, and the study discussed in detail, they were all offered the opportunity to return on another day or have a home interview at a convenient time.

3.5.4.3 Consent

Each young person if less than sixteen years of age after having the study discussed again, completed an assent form, and their parents/carers signed consent forms; for those over sixteen years of age, the young person completed a consent form. Consent included specific agreement to audiotaping of interviews and permission to use anonymised young people’s words and drawings in dissemination.

3.5.4.4 Sample size

Generally studies using phenomenology focus on quality rather than quantity with participant numbers often between four - fifteen (Crist and Tanner, 2003; Corney, 2008; Wilson, 2015). Data saturation is not possible in phenomenological research, but instead when adequate convincing interpretations are comprehensive, explicit and visible, further interviews become redundant (Crowther, Smythe and Spence, 2014).

3.5.5 Interview procedure

Children and young people were offered a choice of a traditional unstructured interview, or a choice of creative methods to help them to share their experiences. The rationale behind the creative methods chosen, their applicability to this study and how they were employed, are presented here.
As Ford et al., (2017) explain, if children have to rely solely on the use of words, it can be difficult for them to articulate their experiences. Therefore, it was critical to spend some time deciding on the best methods to help young people tell their story. Five reasons emerged from the literature and were considered significant for this current study (Table 3-1).

<table>
<thead>
<tr>
<th>Reason</th>
<th>Description</th>
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<tbody>
<tr>
<td>1</td>
<td>The methods encourage the active participation of children and young people in the research process, rather than simply imposing researchers view on children and young people. Allowing children and young people to be active research collaborators, having their say using methods meaningful to them, and thus reduce some of the power imbalance between the researcher and researched (Coad, Plumridge and Metcalfe Alison, 2009; White et al., 2010; Angell, Alexander and Hunt, 2015; Nomakhwezi Mayaba and Wood, 2015; Horgan, 2017).</td>
</tr>
<tr>
<td>2</td>
<td>They are congruent with hermeneutic phenomenology, allowing children and young people to be able to freely express their perceptions of their lived experience (McTavish, Streelasky and Coles, 2012). In hermeneutic phenomenology the interview serves a very specific purpose; as a vehicle by which to develop a conversational relationship with the participant about the meaning of an experience, and allowing participants to share their stories in their own words (Johnson, 2000; Ajawì and Higgs, 2007). Combining a creative method with more traditional interviewing has been shown to be advantageous in the gathering of data for hermeneutic phenomenological nursing research, by providing contrasting and complimentary data (Clarke and Iphofen, 2006).</td>
</tr>
<tr>
<td>3</td>
<td>They are fun, creative, portable and positive. Talking about sensitive issues such as the experience of a chronic condition warrants a technique which will be enjoyable to the children and young people without causing distress (Parry, Thomson and Fowkes, 1999; Elden, 2013; Wells, Ritchie and Mcpherson, 2013; Einberg et al., 2015; Wilson, 2015).</td>
</tr>
<tr>
<td>4</td>
<td>They are not restricted by age, but give choice to the children and young people to choose a method which they are comfortable with. Chronological age is not the sole controlling factor when deciding on the methods to use, the preferences of individuals, their experiences and the research theme should also guide techniques (Aldiss et al., 2009; White et al., 2010; Pimlott-Wilson, 2012; Hill, 2015). Using multiple modalities alongside traditional interviewing generates multi-layered, richly textured information which offers variety so that different children and young people would be likely to engage effectively with some of them (Punch, 2002a; Horgan, 2017).</td>
</tr>
<tr>
<td>5</td>
<td>They are novel and relevant to current technological climate (Punch, 2002a; Bengry-Howell et al., 2011). Incorporating visual and digital technologies in research with children and young people can enable them to connect in ways that resonate well with children’s lives in today’s society, moving away from adult-centric ways of knowing and remixing ideas from technologies to produce methods that resonate better with children (Yamada-Rice, 2017).</td>
</tr>
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3.5.6.1 Choosing the methods

As illustrated by the extensive search undertaken in Appendix two, there were many creative methods to consider. As I began to decide upon measures, the opportunity arose to ‘pilot’ some at a Charity family day conference, at which I had been invited to present at. A selection of potential creative methods were discussed and piloted on this day, with some of these not being used as they were either not feasible or realistic, whereas others were agreed upon. For example we discussed whether asking the children and young people to create a Lego model about their JDM would be possible or whether writing a letter to their JDM would be a realistic activity: both of these were agreed to be ‘too abstract’ and not realistic during an interview situation. However, the body mapping and comic book ideas discussed were positively received and were enjoyed on the day by the children, young people and their families, as further discussed through the reflective diary extract shown in Appendix one. This was also the driver to identify an appropriate electronic method. At this time point, there were also many questions that arose throughout this process which needed consideration when choosing the creative methods, as shown in Appendix six. A protocol was developed which would guide this phase, detailing each step, including approaching of patients and etiquette for home interviews. This protocol also explained the rationale for the methods chosen, and therefore this section of the protocol is included as Appendix seven to illustrate further reading and thinking.

3.5.6.2 Chosen Methods

The interviews can be divided into three phases: the icebreaker, the main phase and the concluding phase. The opportunity to have a ‘traditional’ interview with no creative methods was also offered to all children and young people. These will be briefly presented here.

3.5.6.2.1 Icebreaker

Ice breakers are designed to put children and young people at ease and build rapport, and confirm again that they are aware of the purpose of the study and still happy to participate (Cross and Warwick-Booth, 2015; Einberg et al.,
Phase 1 “Beginning the journey”

Initial introductions to each other, including the first name of the researcher and child or young person’s preferred name, and time for any questions that they may have, are just as key as they would be when interviewing adults (Horstman et al., 2008). An introduction to the audio recording equipment, including the instructions for how to stop it if they want at any stage, are also important (Sartain, Clarke and Heyman, 2000). All children and young people were asked to come to the interview with an item that they relate to their JDM, examples were given such as a toy, photo, and hospital letter. This time spent ‘chatting’ about an object of choice of the children and young people is important to convey to them that their individual thoughts and feelings are the priority, likewise that the interview can move forward at their pace (Horstman et al., 2008).

3.5.6.2.2 The main phase

This phase presented four options to the children and young people and these were offered to children of all ages giving them the flexibility to select a method which they wished to engage with. This approach is similar to that adopted by Bray, Kirk and Callery, (2014) where a choice of writing and drawing activities were used to engage the children and young people with a creative and flexible approach, responsive to their preferences and abilities, not to be interpreted as data in this study, but to encourage the discussion of their experiences.

- Electronic Timeline

Using a timeline in the context of an interview can open up participants interpretations of questions and allow a creative way of interviewing that is responsive to participants own meaning and inform interviewing (Bagnoli, 2009; Kolar et al., 2015). Timelines may also increase respondent’s degree of control over the disclosure of sensitive issues and feelings as the children and young people can decide how much to reveal. This study planned to use an electronic app to further engage the young people. The free app, called ‘Timeline’ allows the user to construct their own personal timeline, adding dates, comments and pictures. Using technology has the advantage of
Phase 1 “Beginning the journey”

attempting to bridge the gap between researcher and participant and engage young people through a medium that the majority are comfortable and confident with. As they add their significant events onto the timeline, the researcher further asks for clarification, and thus the individuals lived experience can be discussed. Any identifying data were removed straight after the interview.

- **Comic book drawing of their life; past, present and future**

Alongside the app, a more conventional ‘draw, design and discuss’ comic book approach was offered. Again the advantages of using drawings in interviews with children and young people include; an innovative and developmentally appropriate method to visualise health and to reveal how children and young people understand illness and communicate their experience. Hobday and Ollier, (1998) in their textbook of creative therapy present ‘writing a book’ together approach to encourage children and young people to think about specific issues. Asking the child or young person to design their own comic book about their lived experiences, and within the interview structure further question their creations as they draw and write, should produce similar results to the electronic timeline, but provides a different creative outlet for the young person to choose.

- **Body-mapping**

Body maps have been used predominantly to illustrate location of pain (Crivello, Camfield and Woodhead, 2009; Von Baeyer et al., 2011). However, as Crivello, Camfield and Woodhead (2009) highlight; ‘Body-mapping’ is multifaceted and relatively easy to use with older children and can be a starting point for exploring key events. The aim of body mapping for this project was as a creative ‘tool’, with blank outlines of bodies prepared in advance on coloured card, ready for the child or young person to draw or write their interpretation of how their JDM affects them.
Phase 1 “Beginning the journey”

- Traditional interview

There is evidence that not all young people like creative methods and may prefer to talk about their experiences in a face to face interview (Harden *et al.*, 2000; Carter and Ford, 2013). Unstructured interviews are the method of choice for phenomenological studies as they give maximum scope to the ideas generated by the participant and allow the conversation to follow naturally (Greig, Taylor and MacKay, 2007). Nonetheless, having some interview prompt questions prepared in reserve was seen as useful in case the conversation struggled to flow. See Appendix eight for the interview schedule questions that were ratified as part of ethical approval.

3.5.6.2.3 The concluding phase

For this study, it was particularly important to conclude the interviews with some time to reflect on the interview and how the young person found the overall experience. Being aware that talking about the effects of their JDM could be upsetting for some, the importance of having time to ask questions and debrief from the experience was vital.

3.5.7 Action in event of distress

Asking children and young people to talk openly about their ill health and how this might make them feel, could cause distress. This was therefore discussed with the supervisory team prior to the interviews commencing and plans were put in place to mitigate against this. Prior to each interview, the child or young person was told they could stop at any point and shown how to turn off the audio recording equipment should they so wish. If they became upset in the interviews, they were encouraged to stop, and parents were informed. The two psychologists both provided clinical supervision to myself, especially to manage any distress that may have occurred from listening to any upset reported from the participants. A reflexive diary was also maintained throughout this phase to encourage reflection.
Phase 1 “Beginning the journey”

3.6 Analytical approach

Audio-recordings were transcribed verbatim including any vocal intonations; crying, laughter, ungrammatical expressions such as ‘umm’ and ‘huh’ or other such nuances. At the end of each interview, I transcribed these recordings as soon as possible to allow immersion in the data. As Balls (2009) suggests, transcription in phenomenology should not be delegated to someone else, the researcher needs to continue to live the data, stay familiar with it and be immersed in it.

3.6.1 Data analysis in hermeneutic phenomenology

“A simple thematic analysis is not enough, yet in itself it is an appropriate methodological approach. It just needs to be named as ‘Descriptive interpretive research’ rather than carry the label of phenomenology. To choose to do hermeneutic interpretive phenomenology one needs to understand and demonstrate skills of interpretation” (Smythe, 2011, p. 44).

Whilst this quote clearly emphasises the importance of being clear with data analysis methods, as Crist and Tanner (2003) discuss, many researchers only give brief descriptions of their data analysis, often with no reference to the philosophical underpinnings influencing their research. This leads to a myriad of data analysis ‘tools’ used and a plethora of end results in phenomenological studies. As Spence (2016) articulates, students feel more comfortable following a sequential, step by step clear process, one which is seen as rigorous and more likely to get published, but this does not fit with the hermeneutic phenomenological attitude.

The aim of hermeneutic phenomenology is to describe, understand and interpret experience from those experiencing it (Tuohy et al., 2013). This can be achieved through a circular continuous process of reading, writing and talking which can be never ending. Rather than following a step by step linear list of instructions, it is an attentive attunement to thinking and listening to how the text speaks (Smythe et al., 2008; Tuohy et al., 2013). This moving between the parts and the whole, is illustrated in the metaphor of the hermeneutic circle and is key to the hermeneutic analysis of interpreting the text:
“’In the circle is hidden a positive possibility of the most primordial kind of knowing’ (Heidegger, 1962, translated by Macquarrie and Robinson, p. 195).”

This statement taken from the original source, illustrates the value of the circle in interpreting and understanding.

3.6.2 The process

The actual correct ‘what to do’ when you have the transcripts available, is not clearly defined in the magnitude of published hermeneutic phenomenology papers, and so returning to key texts and papers written by eminent phenomenology researchers is necessary. The paper by Smythe (2011) entitled ‘How to do hermeneutic interpretive phenomenology’, ended up being a ‘recipe book’, with paragraphs on trusting the process, living the method and writing and re-writing, providing some comfort that the process being undertaken was indeed the correct one. From the hermeneutic phenomenology course attended early in 2017, I had been moved by listening to ‘stories’ crafted from interview transcripts from midwives and wanted to present this research data through the same crafting of phenomenological stories. As found when the researcher has done a good job, the human experience shared, resonates with the listener, often leading to a quiet almost stunned reflection (Benner, 1985; Crowther et al., 2017).

Whilst earlier the rationale for not using a structured step-by-step process was argued, still the route taken to analyse the transcripts must be clearly evidenced to show the data trail. This process of identifying and documenting decisions made throughout the data analysis stage and showing the participants perspective as transparently as possible is vital to establish trustworthiness and credibility (McConnell-Henry, Chapman and Francis, 2009). Therefore, the process of how to ‘craft’ or ‘derive’ narratives from transcripts described by Caelli (2001), was the process followed for this study. See Table 3-2. Each original transcript and the steps taken during analysis have been documented, reviewed by the supervisory team and kept securely by the researcher for trustworthiness, providing a clear audit trail.
The only step not undertaken as suggested by Caelli (2001) was the final stage of returning the end text to the participant. For this study, returning the texts to the participants was inappropriate as many of the interviews were full of emotion. Specifically, when working with children and young people, any harm caused by research, such as the potential for them to become upset when reading and reflecting on their thoughts of living with a chronic condition, should be avoided. In support of this stance, Crowther et al., (2017) highlights; returning the text, or member checking is questionable in hermeneutic phenomenology because human understanding is evolving and therefore open to new revision and interpretation. This is supported by McConnell-Henry, Chapman and Francis, (2011) who state that the researcher-participant relationship provides its own credibility, thus negating the need to confirm the understanding, and thus member checking to confirm the accuracy of the researchers interpretations is contradictory to hermeneutic phenomenology.
3.7 Results

3.7.1 Participants

Seventeen children and young people were approached to take part in the study. Parents of two of these, did not want to take part when approached (due to time constraints), but said they might consider their child taking part later. However, upon a follow up phone call, they did not want to commit at that time. As previously discussed, data saturation is not possible in phenomenology studies, however after 15 interviews, similar experiences were being captured and thus data collection was halted. As Dibley, Williams and Young (2020) confirm, the intention is to gather rich data to present a credible interpretation that represents the breadth of participant experience.

3.7.2 Sample demographics

In hermeneutic phenomenology, recruitment is not dependent on demographic diversity, instead the purpose is to bring various voices to the study, not to compare difference (Crowther, Smythe and Spence, 2015). However, being aware of the sample characteristics is interesting when considering the backgrounds individuals bring with them to the interview. Therefore, basic demographic details are presented here in Table 3-3.

<table>
<thead>
<tr>
<th>Table 3-3 Demographic details of the 15 participants</th>
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<tr>
<td>Gender</td>
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<td>Ethnicity</td>
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<td>Age at interview</td>
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Phase 1 “Beginning the journey”

Whilst the study did not aim to be representative, it did include a wide range of experiences of those living with JDM with diseases duration from three weeks to 16 years of disease duration.

3.7.3 Data collection

Most interviews took place in the designated clinic room familiar to the child and young person. Three interviews were requested by families to be carried out in their home, and despite one of these being in Wales, all requests were accommodated. The interviews ranged from 18 to 130 minutes and were all audio-taped with written consent and again verbal consent on the day. A mother was present throughout two of the 15 interviews, once asked for by her adolescent daughter and the other requested herself to sit in, with her child saying they did not mind. All interviews were carried out by myself. Of note, I was known by five of the fifteen children and young people from my previous clinical role in the department, but had not been involved in their clinical care for the last two years and was not currently involved in their clinical care.

Two of those interviewed, brought soft toys as their reminder of their JDM to help initiate conversations. These toys were given to both when they were first admitted into hospital some years ago and accompanied a young educational book on JDM from the Myositis UK charity. One girl chose to bring her diary from when she was an inpatient and read selected passages out throughout the interview.

One of the first young people, interviewed asked to use the Timeline App and enjoyed doing so. He wanted to use it for longer but was unfortunately told by his mother that they needed to go home. The youngest participant drew some pictures (flowers and rainbows); she was offered the book to keep after the interview.

Three children all had a go at drawing the effects of their disease upon the body-map outline, see Figure 3-1 for an example:
The majority of interviewees though said they preferred to just talk rather than be creative, and interestingly these interviews were longer in duration than those who used a creative method and had significantly less interviewer questions and prompts. However, this difference appeared to have been related to the age of the participants, with those choosing creative methods being the five youngest participants in the study (eight – 11 years of age). When young people had an interview with no creative methods, the interview style was an open-ended, unstructured interview, beginning with ‘tell me about your JDM’. The interview schedule (Appendix eight) was rarely used, and only as a guide. As Balls (2009) highlights, researchers should not be surprised when ethics committees request an interview schedule, however, it is not necessary as hermeneutic phenomenology aims to clarify, illustrate and further explore, not to ask questions. Therefore priority was given to the interviewees to lead the conversation with the researcher encouraging, affirming and being open to the direction of play (Clarke and Iphofen, 2006). Two young people (and one mother) did become upset during their interviews, but both did not want to stop, despite being encouraged too. When the interviews had
Phase 1 “Beginning the journey”

concluded, both expressed a feeling of relief that they had been allowed to talk openly about how they truly feel and one commented, “I feel like a weight has been lifted”. All participants were given a certificate and a monetary voucher in gratitude of their effort and time given.

3.7.4 Data analysis

A series of fifteen transcripts were produced and all used in data analysis. The average interview length was an hour, with the longest being 130 minutes, which yielded 26 pages of transcript text. The visual methods were checked alongside the transcribed interviews to ensure all written/drawn creations were also mentioned verbally. There were three body maps, one timeline and one comic book in total (which had been photographed as the young person had kept it). For example, when a young person would draw red over the shoulder, if this was not mentioned as she was drawing it, the interviewer would ask for clarification; “What is that you have drawn?” to which the young person would reply; “that’s where it hurts”. If during the process of the analysis there was no mention where the redness was, the notes were checked to see where this corresponded to on the body-map, and then during the analysis process, the words shoulder was added into ‘that’s where it hurts in my shoulder’. There were very few instances when extra detail was needed to be added, as mostly clear clarification had been sought during the interview for the purpose of the audio recorder.

The process as discussed by Caelli (2001) (See 3.6.2), reduced each transcript to a ‘story’. This procedure was time consuming but rewarding as 15 stories were produced from 15 interviews – one per transcript. The advantages of crafting stories is to bring the phenomenon into clearer focus with a more concise and readable format reducing the need for many pages of interview transcripts, to ‘show’ what the researcher is noticing and interpreting whilst remaining true to the data (Crowther et al., 2017). The final stories ranged in length from one page to six pages (unfortunately none of these are shown, as preserving patient confidentiality in such a small group of participants with a rare condition, is paramount).
3.7.4.1 Considerations throughout the analysis

During this data analysis process of crafting stories from the data, there were two issues that arose that were important to consider at the outset. These were more of the practical, how to do issues. For example, a couple of the young people had talked about the enormity of being referred to a specialist centre in London, rather than their own local hospital. For some this made them realise how poorly they were and led to heightened anxiety, especially after “seeing all the adverts on the telly”. Therefore, whilst trying to preserve confidentiality, this did need to be acknowledged as this added gravity to the severity of their disease and would have had an impact on their understanding. It also served to highlight the confusion with diagnosis and often explained the long journey to get diagnosed.

A parent sat in on two interviews, each time with the promise of not adding, and each time had started joining in the conversation fairly quickly. Whilst this voice was only in two out of 15 interviews, the researcher felt it would be wrong to ignore it, so after discussions with supervisors, the parental voice was woven through the text where it added clarity, but any repetition or irrelevant details were removed in keeping with the analysis steps presented above.

3.7.5 Early findings

When arriving at the 15 individual story stage through the process described earlier in 3.6.2 by Caelli, (2001), it was apparent that data analysis was only half way through and there was much more work still to be done.

3.7.6 Working with the data

The stories were read and read, over and over again, and played as a continuous record, so much so that each one could almost be recited word for word. The reflective diary was vital at this stage to capture every ‘flyaway thought’ and helpful later to ponder on in further detail. Creative mind mapping became an important activity here in making sense of the many parts to the stories. In the beginning, each story was given a title using the child or young person’s own words from what felt like an important part of their story. For
example, one talked extensively about the little garden she was made to go into during school break times to protect her from the rough and tumble of the playground, thus her story became known as ‘The little garden’. Each story was then plotted next to the others in a huge mind-map of pertinent text that had ‘jumped out’ when reading each story, See Figure 3-2 (only showing a small proportion to preserve confidentiality).

Figure 3-2 Mind-map of interview text

Any text that was similar between the stories was shaded to imply resemblance. Following on from this in a dialectical play between discussions with supervisors, colleagues, family and friends, reviewing the concurrent literature and devising more mind maps on interpretations and thoughts, shared themes began to emerge. Figure 3-3 shows this in more detail.

Figure 3-3 Themes emerging as stories are plotted next to each other
At this point, there was clear overlap in the 15 stories. For example, on first impressions, eleven of the stories talked about the negative implications from feeling different to others, an equally high number commented on their medications and some about whether others can understand what they are experiencing. Some of the stories were clearly different to others with some children and young people having had more family and peer understanding and school support. Also, due to the length of time since diagnosis in some, they could not remember a time without their JDM and commented that their JDM is now very much part of them and does not affect them as much now. Compared to others who were more recently diagnosed their stories at times were quite different, talking about a lack of understanding and a lack of support. This led to questions about the phenomenon that was being sought and how could this tie up these different perspectives.

In further dwelling on the analysis and what was emerging, whilst walking to a supervisory meeting over Waterloo Bridge in the sunshine, I was struck by how magnificently JDM affects the equilibrium of the child or young person. To a young child or adolescent who is leading a normal, healthy, active life; to be suddenly knocked off balance almost by something out of nowhere, which potentially renders you unable to move, is huge, and was being verbalised in these stories. Further pulling this apart, (now sat on a cold bench with pen and notebook at the ready), it was more than just knocking them over, it was doing it again and again, taking them up, and dropping them down. Seeing this as a metaphor of a rollercoaster helped illustrate this visually, and suddenly all of the 15 stories could be plotted using this metaphor. The very recently diagnosed hospitalised youngest child in the study had currently just got on the rollercoaster and was on the first bottom dip, whereas the young person of 18 years who had been diagnosed at two years of age, had come so far on the rollercoaster that now she could not remember the track in the beginning. Pen and paper at the ready, the first drawing of the rollercoaster was crafted, (See Figure 3-4).
3.7.7 ‘Being-on-the-JDM-rollercoaster’

This was a turning point as the metaphor not only represented all the children and young people, but was easily explainable to children, (it is a known image that they can visualise) and equally importantly it had a positive message – there will be an up bit of track coming and you should feel a bit better, and so many before you have ridden this rollercoaster, you are not alone.

The meetings that followed with the supervisory team, led to cementing this further. Thoughts were continuous day and night, analysing every little bit of the rollercoaster and what this might mean for the young person. The final rollercoaster is illustrated here, See Figure 3-5.
Phase 1 “Beginning the journey”

The notion of ‘Being-in-the-world’ is a fundamental concept of Heidegger’s (Heidegger, 1962) as discussed earlier in this chapter, and presents the idea that our “Being” is constructed by the world we know and live in (Fochtman, 2008). Therefore, in keeping with hermeneutic phenomenology, the term ‘Being’ and the dashes between words, signify how the ‘Being-in-the-world’ has been distorted from the known, and the journey now is shaped by ‘Being-on-the-JDM-rollercoaster’. For the children and young people, they could not always specify the start of their illness, but the recognition over time that they were unwell had changed their ‘Being-in-the-world’. As Madjar and Walton (2005) highlight, both illness and the treatment that comes with it, affect the whole of one’s Being.

It became apparent that whilst the overarching construct of the rollercoaster was perfect for describing the journey of JDM, there were ‘themes’ within it, almost encapsulating each step along the way. With further reading and discussions, the themes expressed as emotions described by the children and young people included confusion, difference, ‘sick, steroidal and scared’, uncertainty and acceptance. Taking this a step further when brainstorming with supervisors, it was suggested to consider ‘crafting a story’ from all the stories combined to illustrate each of these five themes and this served as a perfect way to integrate the body of these stories into a meaningful whole. Immersion in the transcripts, allowing the stories to be recrafted, with threads throughout the transcripts pulling it together, using the young people’s own words, editing
Phase 1 “Beginning the journey”

grammar and punctuation where necessary (Flood et al., 2019), finishing with five unique, but shared stories, all shared stories are presented in Appendix nine.

3.7.8 Emerging themes

Again, paying attention to the ‘Being’ within the themes was essential to view the phenomena through ‘different coloured glasses’ and refocus the attention on the young person (Madjar and Walton, 2005). Considering their journey to get them to their current point and considering their past experiences and influences from people around them was vital. Different themes bothered the young people to different degrees at different time points and this diversity helped enrich the rollercoaster with the lived experience of JDM. If we returned to these young people on a different day, we would get another story, one which may bring different themes, however, the quotes here, are the quotes from today and the stories shared from today. The themes were finalised as; Being-confused, Being-different, Being-sick, steroidal and scared, Being-uncertain and Being-accepting.

3.7.9 Themes examined

In this section each of the five themes is presented in turn illustrated with some direct quotes from the children and young people in italics, speech marks and coloured text for easy reference. Unlike other types of qualitative research, the quotes are not attributed to one named individual as anonymity is essential for families when sharing the sensitive words from a small group of young people with a rare disease.

With each theme there is explanation of why this was felt to be a theme and where it sits within the metaphor of the rollercoaster.
The first theme of *being-confused* sits very clearly at the beginning of the rollercoaster. Young people talked about confusion for many different reasons, the confusion that no one knows what is wrong with them, the confusion that they may be poorly, but equally may just be lazy, the confusion health care professionals have in diagnosing them and the confusion their family have in trying to support them. One of the participants in the study eloquently talked about her confusion in the beginning:

> “At this point I was so frustrated because I was thinking to myself, ‘There’s something wrong with me,’ and I felt embarrassed as well because no one could see it, and only I could feel it. The only reason I could still function was because it had come on so slowly, I’d found ways round everything. As I’d got weaker and weaker, I’d even slowly changed the way in which I get out of bed so that before I could even get the energy to stand up, it felt like I’d done an entire workout just to sit up. At this point, my parents were a bit like, ‘What? What is so difficult?’ and I remember being so frustrated because they could see something wasn’t right, but as a parent, they didn’t want something to be wrong with their child. There were so many times when I was just at the point of trying not to cry because I was like, ‘There’s something wrong with me, and nobody can see it, and nobody can feel it’. I described it as having your entire body covered in weights. It’s not even just like having weights around your ankles, it’s like having weights round your ankle, round your calf, on your knee, on your thigh. Just everything was so heavy, especially my head. My shoulders hurt from the weight of my head”.

This text clearly highlights how confusing JDM can be for the young person experiencing it. Another young person described when he realised something serious was wrong:
Phase 1 “Beginning the journey”

“I thought that I had a bug as I couldn’t get dressed, I couldn’t hold the toothbrush, I couldn’t reach for my hair, but then I started to realize that it was definitely more than a bug when I couldn’t stand up by myself in school, but I didn’t know what was wrong”.

The fear from having a rare, often unknown condition which needed referrals to other specialist centres was mentioned by the majority of young people, as this quote demonstrates:

“We went to see the doctors about my redness, and they all said it was eczema but my mum didn’t believe it was. Eventually they diagnosed me with scarlet fever. Then we saw this other guy, I remember waiting outside while my mum and he were talking, and again I got referred to another hospital. I was then really, really scared”.

One of the young people talked about the speed of onset, for her in particular, especially the impact the JDM had on her muscle strength. She had been rock climbing for her birthday only a month ago, and now she pronounced that she could not scratch an itch on her nose, here is a comment from her and highlights this point further:

“At the beginning, I just started as a normal 10 year old being able to do like cross country with my class and sports and then suddenly I woke up and I wasn’t able to do anything and I think it’s how quickly it happens” (Livermore et al., 2019 p.4)

Some children and young people were able to reflect back on how they had felt at particularly low times and how that confusion had played a part:
Phase 1 “Beginning the journey”

“I didn’t talk, and I remember mum was forcing me to talk because she didn’t really understand how I felt, and it was just so uncomfortable I couldn’t talk, I felt so low on energy. I felt really miserable as well from my physio\(^1\) sessions, because I was so weak and I think the people I was with were stronger than I was at that time, because I couldn’t lift my leg without the physio lifting it, I felt as though everyone was looking at me. I was so confused, I didn’t know when I was going home, I couldn’t really believe what had just happened to me”.

As confusion was most often discussed when talking about diagnosis and early interventions, the confusion was situated at the beginning of the rollercoaster.

3.7.9.2 Being-different

“It just is different, but then I was used to it being different”

The second theme of being-different was very prominent when reading the 15 separate stories. Whilst difference spanned the whole rollercoaster of having JDM, there were times when it was particularly a concern, especially in the beginning when the young people developed the facial rash as described in these extracts from the shared stories:

“In the beginning you look obviously different from everyone else”

“I felt quite insecure about my face, because I had a terrible rash on my cheeks”

There were other obvious differences that the children and young people talked about, firstly with schooling:

“In PE\(^2\) lessons I was always sat on the side, it just made me feel even more isolated and again the odd one out”

And then those that required aids felt evidently different:

\(^1\) Physio = Physiotherapy
\(^2\) PE = Physical Education
Phase 1 “Beginning the journey”

“I have a wheelchair which I hate and although I know it helps I hardly use it because it makes me feel self-conscious again and I just feel weak when I’m in it because everyone can see I can’t do what other people can”

Another interesting point which some of the young people raised was whilst the disease has quite noticeable differences in the beginning, such as the vivid facial rash and gottrons papules on the knuckles and knees; as the disease progresses it becomes more difficult to see. The majority of the young people thought this was a negative thing as they have to keep explaining what is wrong with them as shown through these two quotes:

“JDM is hidden, it’s more inside the body, it’s not like a visible disability with a wheelchair all the time and stuff, so just because you can’t see it, it doesn’t mean it’s not there, so that’s what’s frustrating. With my disease, not many people think there is anything wrong with me, because I do look completely normal now, it’s not obvious I have an illness” (Livermore et al., 2019 p. 5).

“It definitely makes it worse not being able to see it, because at school, I kind of feel bad because it looks like I’m faking to some of the teachers and it means that no one understands. People don’t know what it’s like and they’ve never heard of the condition before, so I have to explain the same thing 24/7 and it’s really irritating to explain all the time and for no one to understand at all” (Livermore et al., 2019 p. 5).

Within this theme, during the initial mind map brain storming phase, being different wasn’t only discussed as a negative state, but there were some advantages from having JDM and being ‘different’. Some of these included tangible benefits, such as sweets and nominations for famous theme parks, others though, were more subtle, such as the following:
**Phase 1 “Beginning the journey”**

“So in a way I’m kind of happier to be here, because while it’s not a good thing to have and I’d rather be normal, in a way, I still like to have JDM to be not normal, because it’s kind of being an individual is better than following a crowd”

### 3.7.9.3 Being-sick, steroidal and scared

“It’s really not nice, having to take medicines every day, no normal 15 year old has to take medicines every day”

The effects of the medications were mentioned by all but one participant, and generally always in a negative manner. Predominantly methotrexate and the all-encompassing side effects was the medication most talked about, as it seemed all the children and young people hated their methotrexate:

“The methotrexate was probably my worst memory that stemmed from fear, it just wasn’t nice”

One adolescent spent most of her interview talking very openly about her negative relationship with methotrexate, and the effect it has on her, here is an extract from her story:

“It’s just so emotionally and physically draining because it’s just not being able to predict exactly what’s going to happen, whether I will be sick or not and how I will feel. It’s just so difficult to overcome that mental kind of barrier and that’s definitely been the hardest bit for me I think throughout everything, the methotrexate has forever been a battle that I’ve had. It’s just frustrating that the fact that I know there’s a really good chance that it could make me better, to get rid of it would be, I don’t even have the words, but I can’t do it. It’s just the fact that it’s always a maybe, that with the medication, it’s like well this might work, or this, there is the uncertainty there’s no cure, they can only try and it doesn’t and then I’ve gone through all of that which is really difficult for me and then I’m still where I was a few months later”

This theme is entitled *sick, steroidal and scared* as the children and young people mentioned all three of these. For example, feeling scared of the medications, both unpredictable side effects and from the subcutaneous needles themselves, is illustrated by the following:
Phase 1 “Beginning the journey”

“My mum would bring it in, I’d sit there being like, ‘No,’ shaking, and she’d swipe my leg, and then I’d literally be grabbing onto her arm, and she’d have to just do it as quickly as she could, and I’d just cry. So this became a routine thing on a Friday, which is obviously not the best way to spend your Friday nights. All my friends are out, and I’m sitting in bed crying. It was that I’d sense her getting closer and closer to my leg, I couldn’t let her and I finally had a bit of a meltdown about it” (Livermore et al., 2019 p. 6)

The term “steroidal” specifically relates to the corticosteroids, or simply ‘steroids’, which are often used in the beginning to reduce inflammation and to gain quick control of disease flares. However, they are known to cause unpleasant side effects, such as stretch marks, weight gain and a cushingoid face. The huge psychosocial impact this caused to the young people, especially the adolescents, both male and female were frequently highlighted in their accounts:

“I just remember feeling so self-conscious because I was a really good size before and I was skinny but I was healthy, and then I was suddenly on all of these steroids and it felt like I was a guinea pig or a hamster”

“The effect the steroids had on my weight was one of the biggest things. I always used to be really self-conscious about it, I never used to feel nice about it, I remember wanting to lose it, because I just didn’t feel comfortable”.

3.7.9.4 Being-uncertain

“I remember before I was diagnosed, people used to say, what’s wrong with your face and I used to say, I don’t know and at the time I didn’t really know”

Uncertainty was an obvious theme during data analysis, not only their own uncertainty, but also others uncertainty:

“The teachers told them I’m allergic to the sun which isn’t actually true, because she doesn’t understand either, I don’t think any of the school teachers know about it” (Livermore et al., 2019 p. 6)

There was uncertainty about why they were unwell and uncertainty about the effects JDM has had on them, such as these two quotes highlight:
Phase 1 “Beginning the journey”

“I think getting ill again might be connected to my mocks, the exam stress, maybe”.

“Maybe without it I probably would still be playing football now, rather than having to stop”

There was also uncertainty about the remitting and relapsing nature of their disease and uncertainty about their future, both long term and short term. One adolescent sums this up:

“Even though you’re doing well, doesn’t mean that you’re going to continue, because everything can change in a month and I don’t really know what’s going to happen in the future. I don’t really have control over my condition, I can’t control if I’m going to have a flare up. I can do physio, I can take my treatment but apart from that, you can’t do anything”.

One of the younger participants in the study, discussed how because there is no cure, no one can know what the future holds and how he is reluctant to have this conversation with his mother as not to upset her, this highlights the significance of this uncertainty.

The Being-different, Being-sick, steroidal and scared and Being-uncertain were themes which spanned the whole rollercoaster, therefore not seen as a particular issue at one time more than another, but would be a concern and then would recede into the background, before coming back into focus again, depending on what else was happening to the young person at certain time points. There was also some overlap between all three of these themes; for example, a statement on Being different due to taking medications, could fit into Being-different, or Being-sick, steroidal and scared.
3.7.9.5 Being-accepting

“We get better, once you’ve learnt to accept, this is what I have, this is what it means, this is how I’m going to deal with it and it does become easier to manage.”

The final theme Being-accepting stood out in the children and young people’s stories about living with JDM, as there is a certain degree of needing to accept JDM and carry on with life. This theme had all but the chronically ill young child who was newly diagnosed within it, as one could argue that she is not yet at this place on the rollercoaster. There were some young people that talked about how they feel their JDM has made them a different person, and due to these voices, this theme had firstly been called ‘metamorphosis’ (as can be seen from the original rollercoaster drawing, see Figure 3-4). However, the term ‘acceptance’ was seen to fit better, although there does remain an element of the JDM changing their personality or ‘being’, as these two quotes illustrate:

“The person I am now is a nice, kind person. I think that’s what the JDM has made me.”

“My JDM has made me a better person, I used to be very negative and now I’m positive. I embrace my JDM and I feel more powerful that I have it, I just feel good that I had it as it has taught me a lot of things” (Livermore et al., 2019, p. 6)

Both of these quotes also highlight a perception of benefit due to having JDM. One adolescent had spent most of his interview telling me that he was special due to his JDM:

“Cus you know its really rare and hardly any people get it, so you feel like Wow, I’m kind of special you know”

This benefit overlaps with both difference and acceptance, for example:
Acceptance was not just about accepting the disease itself, but also, accepting the treatments and the consequences from the disease:

“Yes, it can be annoying sometimes. But most of the time its fine, I put my sun-cream on and then just get back to playing outside with my friends and doing what I want to do and forget about it”

Quite a few of the young people were unsure about whether they still have their JDM. For example, these two young people (who are both still receiving weekly methotrexate, so perhaps their disease is better described as “controlled”), reported:

“I kind of still have JDM, but not really”

“I still have my JDM, but you can’t tell, it’s quite weak inside, I think it’s in remission”

This theme showed the resilience that most of the young people have built up. That whilst they can openly talk about how things have been hard, they equally wanted to share the importance of not giving up and getting on with life:

“I think the main thing is, I’m okay. Things hurt and things are hard sometimes, but I still enjoy them. I’m not dead yet. It didn’t kill me, and it’s not going to”

It was clear from the interviews that the older participants were more often accepting of their JDM:

“I think I’ve just grown up. I think it’s really hard, but you have to live with it”

Or at least, that is, they spoke about it more than the pre-teenagers did.

### 3.8 Young people’s reference group workshop

As described earlier, it was felt to be inappropriate to ask the young people to member check their own stories for concern about causing upset, however, a number of the children and young people had asked to stay involved with the
Phase 1 “Beginning the journey”

A project. Therefore, as a ‘Young Person’s Advisory Reference Group’ the participants were invited back to discuss the general findings and see if the metaphor of a rollercoaster resonated with them. Prior to the workshop a protocol was developed in conjunction with the supervisory team, regarding the aim and processes that would need to be followed. The meeting happened in the spring of 2018 and five young people who had been interviewed all agreed to take part (three of these were attending clinic on this date, and two agreed to attend specifically), two siblings also joined the group, and there were two staff members – myself and a member of the wider rheumatology research team. His role was to take notes and assist with any group work that would occur. All participants were given a certificate of attendance and a monetary voucher as a token for their time.

The afternoon began with lunch, introducing ourselves and then I gave some background to the study. The children and young people were asked to say what they thought of when they said the word “rollercoaster”. Their answers included “nervous”, “fast”, “terrifying”, “ups and downs”, “nauseous”, “harsh”, “sharp turns” “amazing” and “scary”. In fact, all of the young people at the workshop, aged from eight years to 16 years of age, could relate to the word “rollercoaster” and their responses mirrored our phenomenon of “Being-on-the-JDM-Rollercoaster”. We then gave the children and young people some mind map charts with each of the themes in the middle, one piece of A4 paper for each theme and asked them to think about their JDM, and what they thought of when they considered that theme. One of these is shown here in Figure 3-6.
They were then asked to stick green tick stickers or red cross stickers on a A3 copy of the rollercoaster to say whether they concurred or disagreed with the statements on it. Of note, we developed a modified version of the rollercoaster for this activity as we did not want to upset any of the participants and so some of the more powerful texts such as “There is no cure” was removed. A copy of two of these rollercoasters are shown here Figure 3-7 and Figure 3-8.
This activity was an amazing form of member checking, as it highlighted how the rollercoaster resonated with all of the children and young people in the room. There were many more green ticks on all of the charts than red crosses,
Phase 1 “Beginning the journey”

with some added comments, such as on the top left of the above rollercoaster, the young person had written “can move, but not very well”. We had also asked the young people whether they wanted to add any extra text onto the rollercoasters if they felt something was missing, but none did.

During our discussions with the children and young people some other interesting comments were mentioned. All of the children and young people in the room could remember their muscle biopsy from their diagnostic workup, even if they had been very young when this occurred and were now questioning why they underwent this procedure, resonating with the confusion we had identified. All agreed that not being able to visually see JDM was a “good” and a “bad” thing, because whilst “no one can tell”, some people think you are “lazy” or “faking it”. This conversation about the ‘visibility versus invisibility’ of JDM involved all the young people and continued for at least 15 minutes, highlighting how this was an important area of concern. One of the young participants said that none of her friends knows she has JDM and this was concurred by another young person in the room. Both were in the first year of secondary school, and despite being there for the last eight months, it highlighted the complexities around schooling and disclosure of information.

This process highlighted the benefits of peer support from individuals who are going through similar issues. The young people in the room all joined in with their experiences, almost trying to out-do the person who had spoken before them, and from this, seemed to develop a ‘group comradery’. This process appeared beneficial and therapeutic as at the end of the session, most of the young people in the room were reflecting that maybe their JDM had not of been as bad as it was for some others. The children and young people talked openly about whether they had met anyone else with JDM and how this had been for them. For some, it had been incredibly useful and developed long lasting friendships, for others, maybe, they just had not found the right ‘other’ yet. For those that had not met anyone else, they openly asked how this process could happen, and whether it would be possible for them. Another reflection was that of the siblings and their participation in the workshop. One younger sibling for instance kept trying to answer questions for her older sister with JDM, much to the annoyance of the patient. The adolescent with JDM had to keep asking her
Phase 1 “Beginning the journey”
to be quiet and telling us all “that she didn’t know anything”, whereas to me, it seemed that she knew quite a lot! These issues regarding the visibility of JDM, the peer-to-peer support and the effect upon siblings, could be looked at further in future work.

3.9 Discussion

Fifteen young people kindly and bravely, shared their experience of living with JDM. The disease severities, illness durations and experiences were all unique, although there were noticeable similarities in the stories told. This shared experience has been summarised through the phenomenon of a rollercoaster, with five themes identified within it. There is obvious clear overlap between the themes, in both definition and timing. Some themes spanned the whole of the rollercoaster, such as ‘feeling different’, ‘sick, steroidal and scared’ and having ‘uncertainty’, whereas ‘confusion’ and ‘acceptance’ were particularly prominent at certain times through the disease trajectory, with the former being more obvious at diagnosis, and the later further on through the disease journey. A discussion point has been whether the rollercoaster is a linear process ending in acceptance and if so, are we advocating acceptance as the result to successful functioning with JDM? The children and young people who were interviewed and who later attended the follow up Young people’s reference group workshop to discuss the rollercoaster metaphor and themes, agreed that acceptance of the limitations and challenges of JDM is a helpful place to get too, but may not be appropriate or sustainable for all.

The ups and downs described by the voices of children and young people with JDM, have commonalities with other chronic illnesses. Interestingly previous literature has also used the rollercoaster metaphor to describe other chronic illnesses and health events; such as becoming a live liver donor ‘Rollercoaster marathon’ (Cabello and Smolowitz, 2008), becoming the parent of a deaf child ‘The roller-coaster of experiences’ (Bosteels, van Hove and Vandenbroeck, 2012), acquiring a physical impairment in youth ‘The Rollercoaster Ride’ (Cynthia deBono, 2017), and ‘Rollercoaster Asthma’ (Rietveld and van Beest, 2006). Other papers use similar wording, such as Recovery in Bipolar disease
Phase 1 “Beginning the journey”

‘Toward caring for oneself in a life of intense Ups and Downs’ (Veseth et al., 2012) and The Ebb and Flow of the Kidney Transplant (Kerr, Soulière and Bell, 2018). When specifically considering other similar paediatric auto-immune conditions, again parent’s journeys have been described as analogous to the recurring ups and downs of rollercoaster rides due to the underlying anxiety experienced when caring for children with juvenile arthritis (Gómez-Ramírez et al., 2016). The rollercoaster metaphor is not new, or indeed does not have a complex imagery to visualise. It was arrived at by chance when considering all of the various voices and the journey children and young people open and honestly shared with me. I began to visualise this as a rollercoaster, and used it to offer a framework in which to consider the experiences of those living with a chronic health condition that was so clearly a rollercoaster of a journey.

As a limited scoping review was undertaken in the outset of this study in keeping with hermeneutic phenomenological philosophical methods, the metaphor of a rollercoaster was not considered in the beginning. However, whilst finding parallels with other diseases can be disappointing to have not found a distinct JDM unique phenomenon, in fact it provides some reassurance. Reassurance to know that the ups and downs which illustrated the journey children and young people with JDM talked about, has obvious commonalities with other chronic illnesses. A further review of the literature looking at young people’s experiences with JIA, also found that young people have reported equivalent themes to our results, such as striving for normality, aversion to being different, managing treatment and suspension in uncertainty (Tong et al., 2012). Whilst it is not yet possible to directly compare the psychological impact of JDM compared to other chronic paediatric illnesses, this study has provided a starting point of JDM specific factors which do have similarities with other conditions. However, by putting these together they represent the unique experience of JDM described by these young people. As presented through the direct quotes, these include; the difficulty in diagnosis due to its rarity, the speed of onset, the impact of losing muscle strength and potential inability to move, the obvious differences to their peers, the constant need to explain what the condition is, the invisibility over time, the remitting nature and lack of a cure (Livermore et al., 2019).
Juvenile Dermatomyositis is a rare disease managed in specialist centres. Not all of these centres may have access to a psychology service or specialist nurses to undertake the counselling and support role, therefore everyone having an understanding of the psychosocial effects of JDM is a step towards providing holistic care. The uncertainty described by children and young people in this study for example could be lessened by providing regular developmentally appropriate information and opportunities for young people to share their anxieties. Providing strategies to help manage the inevitable uncertainty of living with an unpredictable chronic condition and assisting young people how to explain their condition in a clear and concise way to family members and peers would be beneficial. This is especially important as the children and young people specifically mentioned a lack of knowledge from their teachers in school about JDM, and we believe there is no specific JDM school resource available worldwide.

It needs to be acknowledged that there is extensive progress made when considering psychosocial issues in other diseases of childhood such as oncology, but often due to the nature of the conditions, findings are not always comparable. However, whilst there is limited research with JDM, there is some research in other paediatric rheumatic conditions and some of these recommendations could be adopted for JDM. For example the importance of screening for psychosocial concerns such as anxiety and low mood in young people with SLE has been proposed (Knight et al., 2018) and one-to-one counselling and peer support disseminated through a peer mentoring system for JIA (Kohut et al., 2018).

Returning to the notion of the Being-in-the-world, it is important to consider how those with JDM view their lived experience. For example, JDM can cause loss of mobility. This radical change can remove the affected person from the realm of familiar, predictable and well understood experience, especially children and young people who do not always have the necessary life skills and understanding to build upon (Carel, 2016). Many of these young people could clearly articulate their participation in exercise activities prior to becoming unwell, whether rock climbing, playing weekly football or just doing physical education at school, to slowly become weak enough to abstain from
their regular activity, and in some, causing complete lack of mobility, one can see how this would have an effect on other aspects of their ‘Being’—regressing to need help with dressing, feeding and toileting. As one young adolescent said, “I’d found ways round everything”, even in how she had got out of bed. Therefore, JDM does disclose finitude, disability and alienation from one’s body as an extreme mode of being (Carel, 2016).

This chapter opened with a quote describing how chronic illness changes the way one thinks and feels, how the easy everyday tasks, might not be so easy anymore and how one becomes aware of each movement in a taken for granted task. As this chapter draws to an end, one quote from a young child concurs with the starting quote by Ironside et al. (2003), as she says:

“The only time I figured out how to stand up was when I got out of my bed and tried to grab the commode, and I managed to stand up a little bit, just for a tiny second, and I was really pleased”.

This quote reflects the new challenges that JDM can bring to a young child experiencing the implications from such a rare, chronic, debilitating disease.

3.10 Strengths

Children and young people wanted to tell their stories and get their voices heard. For some of these, it was painful, but all said after the interviews how beneficial they had found it. The final phenomena of the rollercoaster was understood and resonated with all the young people who attended the reference group, and the five themes give clear direction to consider when thinking about psychosocial well-being in future clinical care. This study is the first to ask children and young people what it is like to live with JDM. The children and young people were open about their experiences and how these have shaped their lives to date. The findings from this study provide a starting point which health care professionals can ensure they address with their patients and provide the beginnings for the next steps of this research.
Phase 1 “Beginning the journey”

3.11 Limitations

Qualitative studies are always small in number, however rich in depth. These children and young people may well have different stories to tell from those from other institutions, other countries and those who experience different treatment pathways or processes. In phenomenology studies, the purpose is to bring various voices to the study, not to compare difference and so demographic diversity is not prioritised (Crowther, Smythe and Spence, 2015). However, through chance rather than planned intervention, for this study there was a wide range of disease severities, disease durations, treatments and gender and age of participants. Whilst this qualitative phase did not aim to be representative it is important to acknowledge that this phase was the first stage of a larger study, with the latter three phases more inclusive of diversity through sampling from more than one clinical setting.

The creative methods were more challenging than expected. The timeline – partly due to its electronic nature, but also due to the concentration to get the facts recorded and spelt correctly, was a hindrance to free speech. The drawing book, was also a distraction for the young child who used it, who was free to draw, but not necessarily related to her current situation in her hospital bed – or if it was intentional, it was a positive respite from thinking about her JDM. On reflection, maybe this freedom to be creative in whatever way she wanted was a helpful thing for her at that point and thus whilst not yielding much description about what JDM was like for her, it helped her to have some space from her JDM. Two parents were involved in the interviews and thus had an input into the data collection; however, both added points that the young person may not have offered, had they not been there. Also, whilst the rollercoaster metaphor is likely to be adaptive to changes over time, unfortunately due to the cross-sectional design of the study, this cannot be explored further within this current work.

3.12 Implications for practice

The rollercoaster metaphor is one that can be used clinically to highlight to young people the turbulent nature of their disease and path to come, whilst
Phase 1 “Beginning the journey”
scaffolding the specific requirements for that young person to get them to the
top of the track, thus reducing some of the inherent confusion around JDM. A
young child who may struggle to discuss their feelings and emotions, may be
able to point to the rollercoaster without the need to articulate where they feel
they are. Similarly, it may help all health care professionals to discuss
psychosocial needs at appropriate time points, for example not discussing
acceptance when they may feel confused and overawed with their JDM.

3.13 Conclusion

This chapter has presented the background to phase one. Interview data
through direct quotes with 15 children and young people with JDM have been
discussed and data analysis has been explained in detail and demonstrated in
an open, honest and clear approach. The phenomena of ‘Being-on-a-
rollercoaster’ has been presented and the five themes identified. The themes
have then been considered in the context of the wider literature,
complemented by the rollercoaster metaphor.

3.14 Next chapter

The next chapter presents some additional work which came out of this phase
but was not originally predicted in the initial stages. This work used the
transcripts from the interviews to construct research poems, which add to the
experience of living with JDM.
"Won’t stop me trying”

I can’t remember life before JDM, it feels normal to me,
The hospital appointments, the blood tests, the medicines, the
doctors that I see,
I don’t remember a time before, or without it,
Its normal, JDM and me, we fit,
I’ve always had flare ups, constant for 16 years,
I am the odd one out, different to my peers,
When I’m ill and off school and stuff,
Can’t see my friends, they think I’m making a fuss,
They are either constantly messaging me,
Messages I have to see,
“What’s wrong are you coming back? You’re missing this and missing that”
Or, there’s nothing, complete silence,
No idea if they’re thinking about me, no guidance,
I’ve always found the balance hard,
School work or look after myself, which takes charge?
When it came to revision, I worked night and day,
But my health worsened, and I had to pay,
It took such a toll on me,
Physically and mentally,
I had to miss 3 exams in the end,
But the exam board gave me special compensation, the rules they bend,
So I got my grades, but with disappointment,
Frustration that I didn’t do them, due to all the appointments,
Due to my JDM, due to the things it does to me,
But it won’t stop me trying, just you wait and see!
Chapter 4: Poetry “Creative detour”

“Poetry, perhaps more than any other approach or discipline, gets to the essence of qualitative methodology. It presents, and is a catalyst for, a window into the heart of human experience.” (McCulliss, 2013, p. 83).

4.1 Introduction

Working with qualitative data is always exciting, more so when the unexpected happens. In this chapter, it is this element of surprise that will be detailed. This chapter presents work which occurred throughout phase one of the study, as a result of unexpected outputs when transcribing the interviews (see Chapter three). Children and young people have a way of telling their stories and describing their thoughts and feelings in raw, personal and expressive language which when shared with others, can provoke imagery and stir emotions. The language that young people use can be unrefined, immature even, but powerful. It was these words that were crafted into poems. This chapter will present the theoretical body of evidence supporting the creation and utilisation of poetry in research. This process is presented in its own chapter as it brought depth and richness to the entire study and an enjoyable element to the researcher.

Poems are presented both throughout this chapter and throughout this thesis, to ensure the creativity that helped shape this study is woven throughout the presentation of the lived experience of JDM. As the above quote perfectly explicates, the poems became a catalyst for a window into the heart of living with JDM. This chapter will be written in the first person as in keeping with hermeneutic phenomenology it is impossible to separate myself from this process. I made the poems from rearranging the young person’s words, and therefore the process of what I did and why I did what I did will be presented. In this chapter, poetry is introduced through the offering of the first research poem created. To aid understanding and justification for the poetry creations, the growing solid evidence base is presented through ‘the art of creating research poems’. Next ‘the process of creating research poems will be presented’ and discussion of further poetry. A particularly moving experience
came from sharing some of the poems with young people themselves and this process of ‘going full circle’, taking the research findings back to the participants will be discussed further, along with the process of ‘sharing with health care professionals’. The chapter will conclude with a discussion of limitations and plans for future work.

A total of 15 poems have so far been formed, each from only one participant’s transcript. Some of the interviews were lengthy and had two poems created from the same interview transcripts, some interviews did not produce any poems. All children and young people completed assent and consent forms before their interviews (see Chapter three), to give their permission for their words to be used anonymously when reporting the study.

4.2 The first poem

Realising the potential of ‘words’, occurred after the first few interviews had been completed and before data analysis had begun. Certain words and phrases from the interviews stayed with me. I could hear these comments repeatedly and I kept replaying them as I carried on with daily life. One of the interviews had been particularly moving and throughout, the young person and mother (who had sat in on the interview at her daughter’s request) had frequently become upset. This experience affected me greatly and the words used stayed ‘on repeat’. Then the unexpected happened. As I relived the words from this particular interview on a daily basis, they rearranged themselves within my mind, to create a rhyming poem.
Although only ten lines in length, I felt the final result, captured the key essence of that interview, which had taken over an hour and had resulted in 18 pages of transcribed text. This adolescent talked very bravely and openly to me about the bullying she had experienced since her JDM diagnosis, behaviour which she attributed primarily due to the side effects from her corticosteroid medication. Using the actual words used by her peers was very powerful and gave an insight into the suffering she had experienced. As she shared this with me, she had become very emotional. She talked about how this had made her feel different to others and how she viewed herself at this time. Corticosteroids (shortened to steroids) are often given for JDM at diagnosis to gain quick control and also later on for disease flares. Steroid therapy can result in weight gain through an insatiable increase in appetite and fluid retention, and can also cause a round face, often termed “cushingoid”. One of the developmental tasks for adolescence is reassessment of body image, especially in comparison to their peers (Christie and Viner, 2005), and therefore these particular side effects can be very distressing to an adolescent, as illustrated in this poem.

According to Pinquart (2013) in the aptly named ‘Body Image’ journal, negative effects of visible diseases may be more important in adolescence rather than childhood due to negative reactions from peers. In this poem, the language “Tomato face” is powerful and conjures up a picture including both redness and roundness. From using the direct terms her peers used about her, we can...
emphasise with this adolescent about the impact this would have had on her through her teenage years. Obvious differences or deformities that are highly visible to strangers, are known to cause more distress than those that are hidden (Bolton, Lobben and Stern, 2010), as this poem highlights, you can’t get any more visible than your face. If you lose your hair, you can wear a scarf or hat, if your arms change appearance you can wear a long-sleeved top, but how can you cover your face? Another important aspect again related to body image in this poem, was the redness of the rash, not only the visibility of it but also the subtle changes in the redness. As someone suffering from the condition and living with it daily, you notice everything and worry about all the changes, more so than those who do not have the condition.

A recent qualitative study asking young women to discuss body image and self-image in SLE shares many parallels with this present study. One of the participants reflecting on her time receiving steroids said:

“Just…my face was like HUGE!” (Hale et al., 2015 p 1223).

This study confers with three aspects eluded to in the poem ‘Tomato face’. Firstly that coping with comments from others while feeling sad inside is inevitable, secondly the lupus study found that participants generally felt that their health care providers did not give enough consideration to their concerns over the outward appearance effects of the disease and thirdly, individuals self-perception of body image is vitally important, as if unchecked can lead to social isolation and a serious impact on psychosocial well-being (Hale, Radvanski and Hassett, 2015). This poem ‘Tomato face’ speaks volumes in only a few lines.

4.3 The art of creating ‘Research poems’

After creating the first poem, I began to study the literature and found a wealth of support for using poems in research. There are three main ways poetry can be used, summarised by Faulkner, (2009) as:

1. Poems presented to research participants for critique.
2. Poems created from research transcripts.
Poetry “Creative detour”

3. Poetry to present themes noted by the researcher.

In this study, poems were created from research transcripts, as a powerful method of data presentation. This has many ‘labels’ in the literature; ‘Poetic transcriptions’ (Glesne, 1997), ‘Research poems’ (Langer and Furman, 2004; Furman, Lietz and Langer, 2006; Koelsch, 2015), ‘Poetic representation’ (Shinebourne, 2012) and ‘Found poems’ (Gabriel, Lee and Taylor, 2018). All of these have the same rules in place: the words can come from anywhere in the transcript and be juxtaposed, but enough need to be kept together to represent the speaking rhythm and not change the essence (Glesne, 1997). The participant’s voice and meaning must be clear within the poem grounded in the data (Koelsch, 2015).

In Chapter one, I highlighted the lack of literature involving and listening to children and young people with JDM. As Bray (2007) reminds us, to understand children and young people’s experiences it is vital we consult with them. Therefore, incorporating children’s and young people’s voices whilst living with JDM is one of the key contributions which this study brings to the current lack of literature. The poetry discussed in this chapter captures the words of young people and presents their thoughts, feelings and emotions in their own language. Qualitative researchers and poets typically share a heightened sense of language and have a skill for creating images in their work (McCulliss, 2013). Hunter (2002) expands on this by explaining that the experienced poet magnifies nuances of rich and heartfelt language to represent more than the whole of an experience. Neilsen (2004) advises that we must listen under the words, feel, hear and taste what someone is saying. Listening to young people’s voices, and specifically the language they use is a critical component of this study.

As introduced in chapter three, phase one of the study is based upon the methodology of hermeneutic phenomenology which as discussed, interprets layers of language (Philipsen et al., 2019). Byrne (2001) explains that the two assumptions of hermeneutics are that language provides both understanding and knowledge, and that humans experience the world through language. Engagement in the language used from research participants is where the
world is disclosed to us (McManus Holroyd, 2007). The language the young people used in their interviews, taken from the transcripts and presented in the poems gives further insight into their culture, their understanding and their lived experience (Clarke and Iphofen, 2006). Through all the poems presented within this thesis, I have used repetition, similes, imagery, pauses and rhyme (Öhlen, 2003) but the words spoken are purely from the young people themselves.

One of the main differences between traditional poetry and a research poem is the position of the author to the data (McCulliss, 2013). Research poems, re-presenting a perspective or experience of the interviewee are filtered through the researcher (Glesne, 1997). The emphasis is often on emotions, with the text reduced to its smallest element (Roberts, Brasel, & Crawford, 2014). The primary purpose of research poems is to evoke an emotional response from the individual reading the poem (Koelsch, 2015). The emotional intensity that poetry can bring compared to the full transcript, is as Langer and Furman, (2004) describe as:

"the poem may more accurately express the intensity of emotions conveyed that may be lost in a longer narrative. The clarity and conciseness of the poem was rather like blowing away the husks on wheat and leaving just the kernel"


Research poems use the participant’s language which can encourage the reader to see themselves as undergoing the experience (Koelsch, 2015). Through this, the lived experience of others is presented in an effective way (Glesne, 1997) and the researcher can often gain a new perspective which would not otherwise have been seen without the creation of poetry (Brown, 2019). Poetry through its intense and thoughtful use of language, reveals and shapes the lived experience of human beings. There is no doubt that the use of contemplating and reflecting upon poetry, can benefit phenomenological studies, and in fact, Heidegger himself was interested in poetry, as this quote highlights:
Emotions and passion are central features of poetic work but are often considered incompatible with science, as these go against the positivist paradigms and objective epistemology (Darmer, 2006). Avoiding first person constructs, the importance of objectivity and for those who have been indoctrinated with the experimental method approach (Miller, 2018) engaging with creative arts based approach is difficult to understand and accept. However, poetry is a much bigger part of people’s lives than we realise, it is all around us, from adverts to sports fans chants, and verse has been part of our lives for thousands of years (McCulliss, 2013). Due to the similarities between poetry and the art of nursing, poetry is an excellent medium for enhancing nursing knowledge (Hunter, 2002). As discussed, poetry mirrors the aim of hermeneutics, attentiveness to language and gain understanding of texts (McCulliss, 2013), and that of phenomenology which is to be open to the experience as it is told. Poetry has the potential to transform the way we perform research and the ability to present our findings in a more captivating, evocative and insightful way (McCulliss, 2013).

4.4 The process of creating ‘Research poems’

There is not one way to create research poems: each reflects the way it was done on that occasion (Glesne, 1997). However, some researchers have described their process when creating research poems. For example (Miller, 2018) describes the five steps they took:

1. Deep data immersion.
2. Arranging words to create the poem.
3. Critical reflection on the quality of each poem.
4. Researcher reflexivity and member checking.
5. Engagement and dissemination.
These steps were all intuitively carried out in this study, apart from the member checking, (member checking can be defined as the process of asking the participants to review the end result to verify if it accurately portrays what was said and helps verify a rigorous process). For this study, I was concerned about upsetting the children and young people through returning to them and again discussing what they had previously said, however it was carefully conducted with two adolescents, and this will be discussed later in this chapter. Of note, in both instances the member checking did verify the content of the poems as their own words and the correct context for their words.

There was reams of data from each interview, sentence after sentence that was read over and over again. I looked for clear messages, strong emotion, repetitive phrases and especially an essence of that young person’s personality, a snapshot of who they are with JDM. Within some transcripts the process was easier than in others (maybe due to the amount of data, or to the language of the participant) and led to two poems being crafted, in others, it took some time to find the poem. In two transcripts despite hours of immersion, I could find no poem. I felt frustrated at this point and that it was an injustice to those two interviews. However, through reflection with my supervisory team I was led to believe this was alright and in fact the beauty of this unbounded process. The data cannot be manipulated into what it is not. In all qualitative research the researcher sifts through the data and presents it in a manner that is meaningful to them, and different excerpts will always tell different stories (McCulliss, 2013). This process of moving text around and shaping for the audience is no different. Words are deleted or unused in the final offering, becoming part of the ‘compression’ process, through which, as an essential tool in poetry, allows other words to become more powerful, encouraging the reader to focus on the essence of the work (Langer and Furman, 2004). For rigour, the process that was undertaken is illustrated through the following example, shown in the text box below. The text in the box is taken from one of the interviews (the italic two words were said by the researcher) and the bold text illustrates the words that were lifted out to make the below poem entitled ‘This is the life you’ve got’, shown below.
“.....And everything just came, but then I... you get over it, because you don’t have a choice, that’s the thing that I learnt fairly quickly, that it’s fine to be sad about it, but if you’re going to sit there and be sad about it, you’re not, you’re not going to live your life, you’re going to let it stop you living your life, because it’s not going to go away if your sad about it, it’s not going to go away if you’re happy about it either, but either way ...

*Change it?*

...exactly, so if it’s going to be there, regardless you might as well, get on with it, and, and make the most of what you’ve got, kind of thing because I know it could be, it could be and could have been along the way a hell of a lot worse, even with JDM, I’ve been relatively lucky, not to have had ergh, I was never in hospital with it for a particular, I’ve only ever had like infusions that have taken a day, that’s the only hospital stay that I’ve had cus of JDM, so I know that I’ve been lucky along the way compared to some people, this is the life you’ve got, but, in at certain times it’s very difficult to look at the bigger picture and realise and when you’re, when you have just had an infusion and you feel awful, it’s very difficult to think ‘no I’m not, everyone else is worse off than me, I’m fine, this is fine, I can do this’ because you feel as though you’re the unluckiest person in the world, it’s your picture and its, it’s awful and no one, nothing anyone can say can make it better, but then eventually, it’s, I feel like it’s just with anything, if you have a down day you then, you get over it, it’s just a matter of putting things into perspective which sometimes in the moment is very hard to do, but as soon as you do it, it’s then kind of, up, onwards and upwards sort of thing, you have to just, just carry on, onwards and upwards”.
Poetry “Creative detour”

“This is the life you’ve got”
It’s fine to be sad about it,
But if you’re going to sit there and be sad about it,
You’re not going to live your life,
You’re going to let it stop you living your life,
It’s not going to go away if you’re happy about it,
It’s not going to go away if you’re sad about it,
At certain times it’s very difficult to look at the bigger picture,
You’re the unluckiest person in the world, it’s your picture,
But eventually, you just have to carry on, this is the life you’ve got,
Get on with it and make the most of what you’ve got,
Onwards and upwards,
Onwards and upwards.

Earlier in this chapter, I discussed the importance of textual interpretation. This poem portrays some tension between that which may truly be experienced and that which the young person may feel they should portray. “At certain times it’s very difficult to look at the bigger picture” compared to the positive stance of “Onwards and upwards,” which acknowledges the inherent difficulties from living with a chronic condition. One of the themes that came out of phase one was that of acceptance. Acceptance, as opposed to resignation, is defined as the ability to take possession of one’s condition (Kae Kintner, 2007) and we know that acceptance of illness plays an important role in the daily mood of adolescents (Casier et al., 2013). The language used in the phrase ‘this is the life you’ve got’ shows a level of acceptance and moving on through adversity.

A random selection of poems created were checked for validity by two members of the supervisory team, who analysed the transcripts and the poems created to ensure that no words had been added and that the text was true to the context in which it had been spoken. Constant team discussion occurred throughout this process and a detailed log of the process undertaken was retained.

The poems that were written were not specifically used further in the analysis of the interview transcripts, as this is not a prescribed action of data analysis using Caelli’s steps of data analysis for hermeneutic phenomenology.
Poetry “Creative detour”

presented in Chapter three (Caelli, 2001). The poems were standalone pieces of work that are separate to the interview transcript analysis. However the sheer volume of data collected from in depth qualitative interviews can be overwhelming and the richness of data can easily be lost when staring at pages of transcripts (McCulliss, 2013) leaving the researcher at a loss to know where to start. Reflecting on the research poem can help with this, as Roberts, Brasel and Crawford, (2014) declare, the research poem offers a unique forum to give voice to the participants and thus is inevitable a lens for understanding the lived experience. The poems couldn’t not affect myself as researcher, as they offer the ability to view the data from a more creative and empathetic stance (McCulliss, 2013).

4.5 The ongoing process of creating more

As I began to work with the transcripts, there were often certain key messages in each which the children and young people had focused on and returned to throughout the interview. Through moving words around it was possible to highlight these messages in short texts, however it was essential to only use the words verbalised by the children and young people, and not add any as this would change the context of the message.

One of the later interviews ran for 130 minutes and yielded 26 pages of interview transcript. A key concern for this older adolescent had been a long-standing battle with one of the medications taken for JDM: methotrexate. Methotrexate is a medication often used as a cancer treatment as well as for autoimmune conditions and is known for causing sickness. The doses used in rheumatology are less than those used when treating cancers, however, the sickness and vomiting are still frequently described in rheumatology patients (Ramanan, Whitworth and Baildam, 2003; Bulatovic Calasan et al., 2013; Mulligan et al., 2013; Livermore, 2014; Patil et al., 2014; Ferrara et al., 2018). The poem that was constructed from this adolescents words, is shared here.
“Methotrexate”
You were ok in the start,
I was too naïve to pull it apart,
Chalky horrible, disgusting thing, sticking in my throat,
Hiding in my chocolate mousse, never going to eat that again! (make a note)
Can’t swallow any tablets now, (unfortunately)
Dad doesn’t get it, I know I’m 18, but it’s not my fault, it’s what you’ve done to me,
I don’t want you to win,
Why can’t I beat you? Take it on the chin,
Whole day sick with nerves, all day afflicted,
Mum went to such trouble to get the liquid,
Now I have guilt on top of the fear as I shake my head,
I don’t want it I said!
I physically can’t, it’s more than I don’t want to,
It’s deeper, that’s so true,
I want to say I can do it each week and that you don’t scare me,
But you do.
I know it’s a mental thing,
I haven’t even taken you yet and I’m retching,
I’m scared, what happens if I’m sick in the night, off school again tomorrow,
Another meal regurgitated down to you, smell forever etched on my brain, this
time risotto,
A dream combination so they say,
You and Azathioprine,
Hip hip hooray,
Not in my dreams, that’s for sure,
My parents don’t understand,
I don’t understand, it’s really getting out of hand,
It’s so difficult to explain,
Tablets, liquids, injections all the same,
I wanted you for my skin, is that vain?
If you make me feel worse on a daily basis then what’s the point?
Why do I disappoint?
I try so hard, I psych myself up, but I can’t do it,
I don’t want you to win, not even a bit,
Why can’t I beat you? I wish I knew,
If I can’t beat you, will I ever get better? I wish I knew.

Trying to succinctly capture the depth of this adolescent’s ‘battle’ with methotrexate throughout an interview that lasted for over two hours is
challenging. However, when distilling it down to key phrases of language used, the message comes across clearly. The sickness from the methotrexate was huge, and whilst this adolescent is not alone, it is the extent of this nausea and vomiting which affected every part of day-to-day life, which needs to be acknowledged. This poem above is all about methotrexate sickness and vomiting, but does not use the words ‘nausea’ or ‘vomiting’ and only once uses ‘sick’ in relation to actually being sick. However, the word ‘sick’ used in the phrase ‘Whole day sick with nerves’, conjures up someone who is made unwell due to the anxiety they are feeling.

One study comparing methotrexate related nausea between adult and paediatric rheumatic patients found that 76% of the adolescents (13-19 year olds) they surveyed reported nausea, compared to only 35% of adult patients (Patil et al., 2014). This is a large number of young people to share the experience of one side effect. Recently, the literature adds weight to the notion that the ‘anticipatory nausea’ often experienced prior to taking the weekly methotrexate is also a huge issue for many young people and can cause young people to feel sick before they even take the medication (Howie and Livermore, 2016). This anticipatory nausea has also been reported in a recent qualitative study resulting in a theme being assigned to it ‘Talking about it sometimes makes me feel sick’ (Khan et al., 2019). Through participating in this published study, methotrexate sickness was triggered, with half of the participants feeling nauseous during their interviews, just from talking about it (Khan et al., 2019). For the adolescent I interviewed, they had some understanding that the methotrexate did not just exert physical effects ‘I know it’s a mental thing, I haven’t even taken you yet and I’m retching’, but the acknowledged classic Pavlovian conditioned response from cancer therapies such as this one (Khan et al., 2019), had not been explained previously. This is not recognised enough by health care professionals, who see the disease benefit from the medication and want young people to persevere, without really understanding the emotional consequences. As Mulligan et al., (2015) clearly demonstrate, the side effects from methotrexate in some young people can be so distressing that it is associated with a poorer HRQOL.
Through the choice of language used, this adolescent clearly felt alone in their battle with methotrexate. There are many phrases throughout the poem which highlight this, for example: ‘I physically can’t, it’s more than I don’t want to’, ‘It’s so difficult to explain’ and ‘my dad doesn’t get it’. If families are given more support for early recognition of methotrexate intolerance (Khan et al., 2019), then young people hopefully will not feel so alone in their management of methotrexate in their daily lives.

From talking in depth to this adolescent, there were multiple areas that methotrexate had a huge impact upon life and caused significant upset: few of these are acknowledged in the literature. These include:

- The lack of recognition in the beginning that the side effects experienced were from the weekly tablets.
- Parents hiding it in food and the realisation of this as the sickness would occur after eating that food.
- The effect it had on the ability to swallow tablets.
- The effort the mother had gone to in sourcing the ‘unlicensed’ (at the time) liquid and the struggle to swallow this, but now with added guilt.
- The constant fear from not wanting to be sick.
- Humiliation felt when being sick in public.
- Vomiting after eating favourite meals cooked as a treat to help, but then never being able to eat them again.

This poem encapsulates the depth of emotion that this young person had suffered for many years and the psychological turmoil in trying to make sense of all of it.

### 4.6 Sharing with young people

As discussed in Chapter three, sometimes in hermeneutic phenomenology, the data is returned to the participants for “member checking”. However, a large concern was not wanting to cause distress by sharing the interview transcripts with children and young people who may have moved on from their
Poetry “Creative detour”

experiences they shared at that time. This would not be ethical. The adolescent who had struggled with methotrexate, had repeated many times throughout the interview that other people could not understand how they felt. I therefore, after discussion with my supervisory team chose to offer this poem to them to see if this would be useful. I discussed this openly with them, giving them the choice if they wanted to read it or not, and then upon their request, sent the above poem (whilst consent had been gained to use their words during dissemination, during my communication, I again asked for their permission to use the poem during dissemination of this work). This was the reply:

“The poem is beautiful and really does show how I feel, thank you so much for taking the time to write it! Of course you can use it, I think it’s perfect!”

Later that day I received another email:

“My parents just read it and it made them both cry! It really is beautifully written! Mum wanted me to ask if she can share it on her JDM page on Facebook?”

These comments illustrate the power behind the poems. Whilst stripped away from all the buffering text, each word is there to carry meaning, and knowing that the poem had captured what this adolescent had been trying to say for so long, helping them share this with their parents, really made the process worthwhile. The poem was shared on a closed Facebook page and some of the responses are anonymously presented in Appendix 10, again showing the resonance that young people felt when reading the poem.

One of the other poems was also shared during a clinic appointment with a mother who had been present for her adolescent’s interview. This again was not decided upon lightly, but after much careful consideration upon the benefits of doing so. This adolescent was having a difficult time and felt very alone throughout their journey with JDM. I therefore discussed it with her mother before approaching the young person. The mother was very keen for me to share the poem with her daughter, and so I did. The adolescent was overcome with emotion as she gave me a hug and said “finally someone is listening to me”.
4.7 Sharing with health care professionals

Poetry can be a powerful method of sharing findings to a non-specialist audience (Macdonald, 2017) and was witnessed when presenting the data to a multidisciplinary meeting. The audience was predominantly medical professionals with a smaller number of nursing and allied health care professionals. I decided to open with a poem and end with the same poem. I was not sure how the “creative” element would be received, but had used the poem to support some of the discussion throughout the presentation, clearly from the young person’s perspective using their words. Throughout the presentation it was clear to see that some members of the audience were quite emotional and even after many questions, I had health professionals and charity groups come up to tell me how much the poetry had affected them. A significant moment for me was when one senior medical professional came up to share that he would change his practice from now on and now talk to all young people about their psychosocial needs regardless of their age. This poem was entitled “Worry”.

“Worry”
You give me a name that I can’t say,
But I have to explain what it is every day,
I am ten now, but I worry about how I will be,
What is my JDM doing to me?
It’s not easy to see,
But I know it’s there inside me”.
(Livermore, Wedderburn and Gibson, 2020)

This study is based on the hypothesis that children and young people with JDM often have unmet psychosocial needs. From personal experience I have witnessed children and young people with their rare diseases becoming withdrawn and lose eye contact. One can therefore see how medical and nursing professionals would not direct questions or information to a young child during a short clinic appointment for fear of upsetting them or from concern about their ability to engage and converse. However, as this poem clearly highlights, these young people still have worries and thought processes that we cannot understand or address, unless we make the effort to talk to them,
and find out what their worries are. From adding this young child’s age into the poem, it offers further insight into the individual speaking the words. This child was the only one of two young people to discuss their age in the actual interview and this addition adds strength to the words spoken. In a few lines, the effect of living with a chronic, invisible, serious illness comes across very clearly.

4.8 Personal reflection of my journey with poetry

Poetry was never a planned element of this research study. In fact, I have never written poetry before. Developing my poetic voice has been hugely enlightening and enjoyable, but also concerning at times. Whilst there is a supportive evidence base for using poetry in research (Glesne, 1997; Langer and Furman, 2004; Furman, Lietz and Langer, 2006; Prendergast, 2009; McCulliss, 2013; Roberts, Brasel and Crawford, 2014; Livermore, Wedderburn and Gibson, 2020), there is also the acknowledgement that poetry creation and utilisation goes against the ‘usual, rigorous, testable’ way of doing research and will be open to scepticism and criticism from positivist researchers. Being able to defend the poems through discussion of the meticulous, methodical and transparent process to create them is half of the argument; and a readers open mind and attunement to listening to the power of the poetry, the other. It is this unique, imaginative and visionary process, which inspires and excites qualitative researchers, and can win over those with a positivist lens.

4.9 Strengths

This creative detour, although not planned, was enlightening for myself as researcher and has added a different dimension to the overall research study. Sharing two poems so far with the young people as discussed has been therapeutic for them and their families and provided a deeper understanding of their experience of JDM. It is hoped that the poems can be used in further dissemination in the future, specifically sharing the JDM journey for other children and young people, to aid understanding, and limit some of the
uncertainty and confusion found in phase one of this study. Whilst being incredibly personal, I am sure many of the issues raised within the poems would be shared with other JDM sufferers, and if they helped to resonate some of the emotions so young people do not feel so isolated and alone, then this must be a positive outcome.

### 4.10 Limitations

The findings presented are always simply the impressions gained, an offering of thinking to engage others in their own thinking (Smythe et al., 2008) and thus open to interpretation. Crafting short poems from reams of transcript carries risk of bias. A different researcher may well have used different text to end up with a different final poem (Prendergast, 2006). As researcher on this occasion I was in charge of selecting the text to become the poem, and that which I was not going to include, and this in itself alters the perception of the data (Glesne, 1997). However, this is also true of hermeneutic phenomenology, that it is defined by the interpretation, it is “my experience” of what the data is telling me (Smythe, 2011), there is no single correct interpretation as we are constantly open to new information (McCulliss, 2013).

The credibility of a qualitative study is based on the overall internal validity and methodological rigour, which has been demonstrated within this chapter through a clear process to follow, discussion of data inspection by the supervisor team, frequent team discussion, plus support and a retained reflexive journal of the process undertaken.

On the reverse side of using poems to present the powerful emotional response, this too can be upsetting for some. I saw this myself in the conference where I presented one poem, and whilst not a reason to avoid using them, I feel it is imperative to consider this fully. Warning an audience or at least preparing them for what might follow is good ethical practice. A further limitation offered by McCulliss (2013) is that of potential anonymity and confidentiality issues when presenting findings. This is the primary reason of why you will not generally find the gender and/or age of the majority of the poems throughout this thesis. If one is mentioned, then the other most definitely is not.
4.11 Conclusion

Fifteen poems have been created, each representing a different perspective of the lived experience of these young participants. Most of these have been presented between the chapters, to showcase different elements of the experience of living with JDM. Whilst not planned from the beginning, as the poems took shape, it was clear to see the benefits they could bring during dissemination of this work to multi-disciplinary audiences. The aim of hermeneutic phenomenology is to resonate, to provoke thought and to share an experience with others, using poetry, has helped achieve this. Sharing young people’s words to present the lived experience of JDM has been a powerful, emotional journey. As McCulliss (2013) affirms, poetry presents a window into the heart of human experience.

4.12 Next chapter

The next chapter (Chapter five) presents the work in phase two which asked all children and young people around the UK to complete questionnaire measures to examine if the findings found so far, resonate with a larger group of children and young people with JDM.
“Lazy teenager”

My only symptom was weakness,

No rash, just weakness,

I was at the beach when I first realised something was up,

I was lying down, then I couldn’t sit up,

That night, for a few seconds, I managed a plank,

A month later, couldn’t even hold a plank!

Last year I’d broken school sports day record for triple jump,

This year thought I’d practice in my garden, couldn’t even hop,

no way could I jump,

For so long I’d been told “You’re lazy” and I’d been saying I

need to go to the doctors,

Now I don’t want to go, don’t want to go to the doctors,

As we left, I had been referred to the local hospital as a

paediatric emergency,

With a special ticket to all hours a&e,

Before a diagnosis, it’s just embarrassment, confusion and

humiliation,

Embarrassment, confusion and humiliation.
Chapter 5: Phase 2 “Picking up more passengers”

5.1 Introduction

“Families of patients with inflammatory myositis rated overall quality of life as the variable with the highest average importance” (Tory et al., 2018 p. 3).

Chapter four was a bonus; a creative detour on the journey. Revisiting the roadmap and considering the route undertaken so far, Chapter three presented phase one of the research. The exploratory, qualitative design of the first phase facilitated a small group of children and young people with JDM to discuss what they felt their psychosocial well-being needs were. Phase two of this research explores these results further to establish whether these findings are applicable to a larger and more diverse population of children and young people with JDM. It is this phase that is presented here in Chapter five.

This chapter presents the design, hypothesis, aims, methods, and analysis. The results, discussion, limitations and future relevance of the phase two study are also presented, but first the background.

The quote that opens this chapter was taken from a recent study that asked 194 parents to rank the importance of 19 items related to aspects of healthcare measurement. Quality of life for families was more important than all the other 18 variables which include such themes as: resolution of pain, ability for self-care, medication tolerance, school attendance, and minimisation of hospital visits (Tory et al., 2018). These results are quite staggering, but serve to highlight the importance of addressing quality of life in children and young people with JDM. This quote has significance for what follows in this chapter.

5.2 Background

The role of phase two, was to integrate and expand the findings from phase one. The priority was to place the voice of children and young people with JDM at the core of this study, to let their words be the starting point and direct the focus of inquiry going forwards. The five themes from phase one were key to
Phase 2 “Picking up more passengers”

this process: confusion, feeling different, medication effects, uncertainty and acceptance. Two options presented themselves in the planning of this phase: either to undertake a period of instrument development to produce a measure that would focus on these themes, or search for validated instruments that were the best fit. Instrument development, and the need to develop, test and validate scientifically was not a feasible option, therefore other questionnaires were sought that would capture the themes of interest. In terms of the design for this phase of the study, selecting instruments to best fit was the starting point.

5.3 Design

5.3.1 Creating the theory

Using a theoretical model to drive forward the enquiry in this study would have been easy. A range of models were considered in depth for this, guided by the two psychologists on my supervisory team. For example ‘The Chronic Illness Model’ proposes that background factors such as quality of life and availability of social support influence how people respond and adapt to the various illness stressors with the end result returning to equilibrium through the process of adjustment (Moss-Morris, 2013). This model was initially developed to understand adjustment in adult Multiple Sclerosis and whilst it offers some areas for consideration, with the overarching emphasis on adjustment, it was deemed not to be applicable to the findings from phase one. Further models also at this time point were considered and these included:

- The Pediatric Psychosocial Preventative Health Model (Kazak, 2006).
  What is it - A 3-tier model based on a public health orientation.
  Disadvantage - needs to include all the family
- The Biomedical Model (Farre and Rapley, 2017).
  What is it- emphasises disease and disability
  Disadvantage – less emphasis on social and cultural responses
- The Illness Perception Model (Hale, Treharne and Kitas, 2007).
  What is it – The perception of a health threat, leads to defensive behaviour
Phase 2 “Picking up more passengers”

Disadvantage – For those with active disease states only
- The Corbin and Strauss Chronic Illness Trajectory Model (Corbin and Strauss, 1991).

What is it – A nursing model to guide chronic illness care
Disadvantage – Structured around delivery of care in certain health domains, rather than from the patients perspective

As we considered models in detail, generated from a range of theoretical perspectives and disciplines, it became obvious that we were trying ‘to make a model fit’. This was never the intention for this phase of the research, and in fact it was what made it unique: the qualitative findings from the voices of the young people had to guide the approach in the forthcoming phases, and we kept returning to this point. When re-examining the phenomenon of the rollercoaster and anchoring the key text to each of the themes, the impact each of these had on overarching quality of life and emotional distress was apparent (See Figure 5-1).

![Figure 5-1 Re-considering the rollercoaster](image)

At this point in our deliberations, the focus moved to the practical issues of developing our own JDM model of psychosocial well-being. As quality of life and emotional distress were impacted upon by all of our identified themes, it was agreed that they would become the outputs in our model. It is worth
Phase 2 “Picking up more passengers”

highlighting that whilst emotional distress could be identified as an aspect of quality of life itself, as it came out so strongly in the themes, a priority was to examine how important this was in the overarching outcomes.

Uncertainty was a particular concern from phase one, being mentioned frequently throughout all interviews and therefore was key to consider during this study phase. However, uncertainty, confusion and feeling different all have clear overlaps and consequently if a questionnaire could be identified for use with children and young people, which measured any of these, it might be appropriate for all three.

The acceptance of JDM was an interesting finding from phase one, and one that is not discussed widely in the literature. At this point I returned to the rollercoaster and literature and considered the findings again to understand if acceptance was the most appropriate label to capture this theme, or whether there was a more generic term which would have more applicability. Whilst the children and young people had talked about accepting their JDM, they also talked clearly about the benefits it brought them. According to Sawyer and Aroni, (2005) acceptance of chronic illness in adolescents, follows efforts to find meaning out of having a chronic illness. Therefore, if children and young people could identify some meaning, or potentially benefit from their disease, did this then have an impact upon their perception of their quality of life and their overall emotional distress? It became clear there was significant overlap. The final model developed involved two variables; ‘uncertainty’ and perceived ‘acceptance/benefit’ and two outcomes; ‘quality of life’ and ‘emotional distress’, this is represented in Figure 5-2.

![Figure 5-2 Our Juvenile Dermatomyositis Psychosocial Model](image-url)
Phase 2 “Picking up more passengers”

This model was kept very simple as the purpose was not to build and test a model, but consider the relationships between the variables and outcomes.

Collecting information on medications specifically for this study was identified as being a difficult issue, as not all children and young people in the JDCBS would be receiving medications. Therefore, it was decided to identify a quality of life tool that would capture medication administration, but not make it a clear focus.

5.3.2 Questionnaire selection

Questionnaire selection was key and this process took time, with relevance and utility crucial to the selection process. The literature was searched for questionnaires to measure and mirror the themes added into our JDM model. To be included, measures needed to be for child self-report (ages eight years and upwards), validated and ideally, previously used in paediatric rheumatology. During the scoping review, there were a number of HRQOL tools identified which have previously been used in paediatric rheumatology. Each of these was considered in detail, examining their previous use in research, validity information, ease of use, ease of access, cost and applicability to this study. There was one generic quality of life questionnaire which has been used more extensively in paediatric rheumatology and deemed the best fit and a second, previously developed and validated only for children and young people with rheumatic disease, which also asked medication questions. Using both a generic and disease specific questionnaire is recommended in the literature to provide an enhanced, comprehensive picture (Varni et al., 2002). A paediatric UK developed questionnaire which captured emotional distress was also found which was brief but added a clinically relevant cut-off, which was beneficial to provide a safety net to capture those which may be struggling. An acceptance measure could not be found, but a questionnaire examining ‘perception of benefit’ was identified and included questions which covered a perception of acceptance, such as “My illness has taught me to be happy and enjoy the good things when they happen”. This questionnaire was paediatric specific and believed to be the
Phase 2 “Picking up more passengers”

‘best fit’. As the questionnaires were to be posted to young people, a priority was for them to be easily understood and not too long in length.

After much discussion with the psychologists on my supervisory team, five validated measures were decided upon, the detail of each is presented in 5.7.7.

Whilst the five validated questionnaires captured the majority of the findings from phase one, there were two points that had been raised by a number of the young people which they did not capture; “JDM is invisible over time”, and, “I have not met anyone else with JDM’. These concepts were not coded into their own themes as they appeared in most of the others. For example, the fact that JDM can become invisible can add to confusion either for the young people themselves, or others around them who do not understand, and this can also make them feel different to their peers, whilst struggling with ‘appearing to’ look the same. Not meeting anyone else with JDM, can contribute to young people’s confusion about their own condition, feeling different to their healthy peers and possibly having an impact on their own ability to accept their condition.

A key strength to this study was offering all children and young people between the ages of eight and 19 years of age, with JDM across the UK in the JDCBS an opportunity to voice their concerns from living with JDM to describe what their own psychosocial needs are. It was therefore important to offer some open comment boxes to capture this. Hence, a questionnaire was designed to capture these outstanding issues, and included in the pack sent to young people in this phase two.

5.4 Research Question

The research question this phase sought to address was:

- “What variables are most important from phase one and are there any other psychosocial needs expressed as important in a larger cohort of children and young people with JDM?”
Phase 2 “Picking up more passengers”

5.5 Aim

The aim of phase two was to:

- Investigate resonance with the themes found in phase one and to identify whether there are any other psychosocial well-being needs, in a large cross-sectional sample of children and young people with JDM throughout the UK.

5.6 Hypotheses

In order to explore relationships between the validated measures and the responses from the bespoke questionnaire, (from here on referred to as the ‘JDM experience questionnaire’) and the clinical variables, a number of hypotheses have been proposed:

1. Children and young people with JDM would report lower quality of life and/or more emotional distress than their healthy counterparts.
2. Children and young people with JDM who had higher levels of uncertainty would have lower reported quality of life and higher emotional distress, than those with lower levels of uncertainty.
3. Children and young people who had perceived acceptance and/or benefit from their JDM, would report better quality of life and lower emotional distress than those who did not report perceived acceptance.
4. If children had active JDM (assessed by the clinical variables that were extracted from the JDCBS) they would have lower quality of life and higher emotional distress, than those who did not have active JDM.
5. Children and young people who believe they still have JDM, will score a lower quality of life and more emotional distress than their counterparts who believe their JDM is currently inactive or gone away.
Phase 2 “Picking up more passengers”

Whilst the hypotheses were kept deliberately broad, no causality could be suggested as the relationships could be bidirectional, with either the higher uncertainty causing the lower quality of life, the lower quality of life causing the higher uncertainty, or both might be associated but not causally linked.

5.7 Methods

5.7.1 Setting

For this study, the priority was to gain breadth and depth by including all children and young people with JDM in the JDCBS, within the target age group.

5.7.2 Ethical procedure

Phase two proceeded by obtaining a substantial amendment to the JDCBS approval (North Yorkshire REC, ref 1/3/22 Appendix 11). Approval was gained with no changes or clarifications required. An application was also submitted to the JDCBS Steering Group for their approval of the study, and granted with no alterations. University ethical approval was also obtained along with the Host Trusts R&D ethical approval. When a child or young person is entered into the JDCBS, their assent/consent and accompanying parental consent includes permission to be contacted regarding additional studies. This phase of work used questionnaires, return of which was taken as consenting to participate and this was explained in the letter with the questionnaire pack, and this was approved by the North Yorkshire REC, as part of this substantial amendment.

5.7.3 Specific considerations

As planning for this phase got under way, it quickly became apparent that there were many questions that needed consideration, such as how to track the respondents and whether to send packs to the parents or the child/young person, Appendix 12 summarises these and the decisions made.
5.7.4 Participant population

5.7.4.1 Inclusion criteria

The inclusion criteria for this study were kept as minimal as possible to maximise inclusivity;

- Diagnosis of JDM.
- Recruited and consented to the JDCBS.
- Eight - 19 years of age at time of questionnaire packs being sent out.

5.7.4.2 Exclusion criteria

The exclusion criteria were;

- Children and young people who did not have a current known address.
- Any children and young people that the local centre asked us not to contact.

The exclusion criteria were kept minimal as the study wanted to target as many children and young people with JDM as possible. This was also influenced by the fact that it was not possible to know all other co-morbid conditions of the children and young people in the JDCBS, for example previously reported mental health diagnoses, were not known due to the reliance on the data entry in the 15 local sites and over a time period of up to 18 years previously. First language was also not easily obtainable and therefore, not added as an exclusion.

5.7.5 Participant recruitment

5.7.5.1 Preparation

The JDCBS database does not contain patient names or home addresses. Therefore, the local research nurses needed to send out the study paperwork. A teleconference was organised to discuss the requirements requested from
Phase 2 “Picking up more passengers”

each centre and this was followed up with a video link to all to discuss the practicalities of the study. A flowsheet (Appendix 13) was developed to help the local nurse know what their responsibilities were, along with a personalised letter for each nurse, thanking them for their support.

A brightly coloured postcard was produced to introduce the study and provide an opportunity for families to opt out via an email address should they not wish to receive a questionnaire pack (See Appendix 14). These were sent out by the local nurses ten days prior to the questionnaire packs being posted. Each pack consisted of invitation letter, age appropriate information sheet, (See Appendix 15 and 16), parental information sheet, age appropriate questionnaire (versions for 8-12 years or 13-19 years) and freepost addressed return envelope. Each pack was addressed to the ‘child and the parents/carer of…’ as whilst the questionnaires were for the young person, it was also important for the parent/carer to be involved and aware of the study. The documents were printed on 15 different Trust headed notepaper and sent out via recorded post to each centre.

Each pack had the patients four-digit unique code on it, necessary for the local centre to establish that particular young person’s postal address. A logistical reason of clearly linking each code to each patient was to ensure they received the right age specific pack for either those under or over 13 years of age as the packs were specific with an age appropriate information sheet, accurate footers and the correct age specific version of the PedsQL™ generic questionnaire and PedsQL™ Rheumatology Module questionnaire (see 5.7.6.2).

5.7.5.2 Identifying and approaching

A query was run in the JDCBS database on an agreed date and this dataset generated the group of patients to be approached. Through discussion with the local teams, it became apparent that there were a small proportion of children and young people who could not be approached - for the majority their address was not currently known and in two there were child-protection concerns, these were therefore excluded at the request of the local team.
5.7.5.3 Recruitment

It was decided not to approach children and young people while they attended clinics, as it was more practical and time efficient to collect data remotely, especially considering the large number of sites taking part in the study across the length of the UK. Therefore, every participant in the JDCBS who met the inclusion criteria received a pack in the post.

5.7.5.4 Sample size

No formal power calculation was conducted as the aim was to include every eligible participant in the JDCBS where possible. Also, a further concern was that there is not normative data available for all of the tools used, nor data from paediatric rheumatology patients and therefore comparisons to inform a power calculation would be complex.

However, using a rule-of-thumb calculation (also termed ‘heuristics’) provided some statistical guidance. These are based on power analyses for tests of hypotheses of both multiple and partial correlations (Green, 1991; Dattalo, 2018). A rule-of-thumb for sample size requirements for a multiple regression is 50+8k where k is the number of independent variables in the model (Tabachnick and Fidell, 2012). This indicates that if we had eight independent variables (including demographic, clinical data and patient perceptions) for each outcome, we would need a sample size of 114 for a regression analysis. This is discussed further in 5.8.1.3.

5.7.6 Questionnaire delivery

The rapid adoption of smart devices into children’s daily lives has increased exponentially, with children as young as two, spending up to two hours a day on devices (Dirani, Crowston and Wong, 2019). It was therefore decided to offer an internet completion option, and a paper version, so young people could decide which was most convenient for them, and to cover those who do not have access to technology at home. Each invitation letter clearly stated the choice of completion and provided a web link to the correct age-related version of the questionnaires. The electronic questionnaires were populated using
specific approved university survey software (Opinio®). Details of the software were provided in the parental information sheet, especially with regards to data protection. Using a software system such as this meant it was quicker and easier to complete and return, for those that had access to such technology. After the survey had finished, data from paper responses were entered into the online system, so that all the data could be safely and securely stored in one location.

The paper copies were designed to make them look identical to the electronic ones, to avoid any bias, however, the electronic versions could be set up in such a way that each box needed to be completed before you could move through the questionnaire, and this was not possible with the paper version.

To encourage participation, the surveys offered a competition to win an online shopping voucher for one random participant after the study had closed in a study prize draw. The questionnaires in this phase were collected between October 2018 and the end of December 2018.

5.7.7 The suite of questionnaires

5.7.7.1 Demographic and clinical data collected

The first page of the questionnaire asked the young person to add their own unique code, primarily so that if they had a high score in the emotional distress tool they could be highlighted to their clinical team for further support (See 5.7.8), but it also allowed respondents to be identified for the post study prize draw. They were also asked for their gender and day and month of birth. These questions were added to serve as an extra measure for identification in case the individual did not add their own code. It was decided to not ask children or young people for any clinical data, as it was anticipated that we would be able to access this information from the JDCBS. Some clinical variables were collected from the JDCBS, (although the participants were asked about their perception of their JDM in the experience questionnaire) after the study collection period had closed, including;

- Current age.
Phase 2 “Picking up more passengers”

- Years since diagnosis.
- Length of time since last clinic visit.
- Whether they had received the disease modifying medication methotrexate at any time.
- Whether they had rash or weakness when they were last clinically seen.

No other demographic data such as ethnicity were collected as these are not always completed within the JDCBS.

5.7.7.2 Validated measures employed

The patient validated measures included:

1) The PedsQL™ 4.0 Generic Core Scale (Varni, Seid and Rode, 1999; Varni, Seid and Kurtin, 2001; Varni et al., 2003). Permission granted to use this tool gained on 28th June 2018 (the PedsQL™ will be referred to as PedsQL from here onwards).


3) The Paediatric Index of Emotional Distress (PI-ED) (O’Connor et al., 2016). Tool and manual purchased, and permission granted on 8th October 2018.

4) The Childhood Uncertainty in Illness Scale (CUIS) (Mullins and Hartman, 1995; Pai et al., 2007). Permission to use this tool gained on 15th June 2018.


A brief summary of the measures is provided in Appendix 17 detailing the country of origin, domains, range of score and scoring implication. Unfortunately, the whole questionnaires cannot be shown for copyright reasons.
5.7.7.2.1 The Pediatric Quality of Life Inventory (PedsQL)

The Pediatric Quality Of Life Inventory PedsQL 1.0 was originally developed in the US from a paediatric cancer database (Varni et al., 1998). A more recent version; the PedsQL 4.0 generic core scale is a generic 23 item instrument, available in different formats (parent/carer proxy, and three age-specific related child/young person self-report aimed to capture all ages), and with the advantage of brevity and a UK English version being widely available (Varni et al., 2003; Upton et al., 2005). Collecting HRQOL in the four domains of physical, emotional, social and school functioning, all measured on a five point Likert scale, from zero (never), to four (almost always). Scores are added and transformed on a scale from 0-100, with higher scores indicating better health related quality of life. This can lead to a physical functioning scale score, a psychosocial health summary total score, (comprised of the individual emotional, social and school functioning scores), and an overall total scale score. The PedsQL is developmentally appropriate (with the difference in wording between the different age versions being for child or teenager e.g. ‘other children tease me’ or ‘other teenagers tease me’). However, the similarity of wording and content across the parallel forms facilitates the evaluation of differences in health related quality of life across and between the age groups (Degotardi, 2003). In a reliability and validity study the PedsQL discriminated between healthy children and children with rheumatic diseases as a group (Varni et al., 2002). The UK English version was used in this study.

5.7.7.2.2 The Pediatric Quality of Life Rheumatology Module

The PedsQL 3.0 Rheumatology Module was designed to measure HRQOL specifically tailored for paediatric rheumatology as this is known to enhance measurement sensitivity for health domains of a specified condition (Varni et al., 2002). The 22 item PedsQL 3.0 Rheumatology Module measures disease specific HRQOL, with domains for pain and hurt, daily activities, treatment, worry and communication (Varni et al., 2002). There is a five point Likert scale from zero (never), to four (almost always), with scores added together, divided by the total number and transformed on a scale from 0-100, with higher scores indicating better Rheumatology related quality of life. This tool has been validated and again comes in different formats for different aged participants.
Phase 2 “Picking up more passengers”

The PedsQL Rheumatology Module has been used in research studies which include children and young people with JDM (Trapanotto et al., 2009). This will be termed as the Rheumatology Module from this point forward.

5.7.7.2.3 Paediatric Index of Emotional Distress (PI-ED)

The Paediatric Index of Emotional Distress (PI-ED) is a questionnaire developed in the UK to screen for emotional distress in children and young people (O’Connor et al., 2016). It is based on the Hospital Anxiety and Depression Scale (HADS) and uses appropriate language for young people ages eight years and upwards. It comprises a 14-item self-report measure of emotional distress rated on a four-point Likert scale, with two summated subscales assessing depression and anxiety (Canning, Bunton and Robinson, 2014). Whilst these two subscales can be identified O’Connor et al., (2016) recommends that a simple summed score of cothymia (general emotional distress) is assessed primarily by the PI-ED. The PI-ED is valid and reliable, with the attainment of a global score, the higher the score the greater distress with cut-offs indicating clinical significance (See 5.7.8) and suggesting further clinical assessment (O’Connor et al., 2010, 2016). An advantage of the PI-ED however, is that it can be used with children who have a physical illness, as it will not confound physical symptoms of distress with those of a physical condition (Dorris et al., 2017). The PI-ED is not purely a research tool and is used across the UK as a screening tool in clinical practice.

5.7.7.2.4 Child Uncertainty in Illness Scale (CUIS)

Uncertainty is described as the inability to determine the meaning of illness-related events or to predict an outcome accurately and can be influenced by many illness-specific and individual factors (Mishel, 1988; Stewart, Lynn and Mishel, 2005). Uncertainty results when a sufficient cognitive schema cannot be formed with which to interpret the meaning of illness-related events. Uncertainty can lead to psychological distress if coping responses are insufficient to resolve the uncertainty. Mishel’s Uncertainty in Illness theory led to the creation of the Mishels Uncertainty in Illness scale (MUIS) (Stewart, Lynn and Mishel, 2010). Mullins and Hartman (1995) adapted the Child Uncertainty in Illness Scale (CUIS) from the MUIS for use with children and young people.
The CUIS is a 23-item self-report measure used to address four components of children’s perceived uncertainty regarding their illness: ambiguity, lack of clarity, lack of information, and unpredictability. Participants’ rate items on a 1-5 Likert scale, ranging from one (very true) to five (very false). A total score is calculated with higher scores (ranging from 23 to 115) reflecting greater illness uncertainty. The content of the items on the child version is based on the original adult version but contains modified wording to make it more developmentally appropriate for children and adolescents. This measure has previously been used for children between the ages of eight and 18 years (White et al., 2005; Pai et al., 2007). The CUIS has demonstrated good internal consistency reliabilities across a number of chronic illness conditions and has been used in youth with rheumatic disease, including those with JDM (White et al., 2005).

5.7.7.2.5 Benefit and Burden Scale for Children (BBSC)

The Benefit and Burden Scale for Children (BBSC) is a 20-item self-report instrument originally developed for a paediatric cancer study adapted from the 10 question Benefit Finding Scale for Children (BFSC) (Phipps, Long and Ogden, 2007). The Benefit measure initially raised concern as the instrument only included positively worded items, which were felt to possible encourage a socially desirable response leading to inflation of the estimates of benefit. Through adding in both positive and negative questions it was felt to provide a more valid measure of benefit (Currier, Hermes and Phipps, 2009). Therefore, although this questionnaire was initially selected as it served to encompass some of the ‘acceptance’ issues that had arisen from phase 1, the combined BBSC was regarded as a more robust measure than the BFSC alone. The resulting BBSC questionnaire has ten potential benefit and ten potential burden of illness questions answered on a five-point Likert scale ranging from “not at all true for me” to “very true for me”. Scores range from 10 – 50, with a final separate benefit and burden score. The BBSC was initially developed and validated in a paediatric cancer population aged eight years and upwards (Currier, Hermes and Phipps, 2009). More recently it has been converted into Dutch and validated in a study looking at post traumatic stress of cancer.
survivors (Maurice-Stam et al., 2011). No studies that have used it with children with rheumatic conditions could be found.

5.7.7.3 JDM experience questionnaire

The bespoke JDM experience questionnaire was always envisaged to be a much smaller element than the validated questionnaires, primarily addressing factors raised by children and young people in phase one, not represented in any of the validated questionnaires. But also importantly to give all children and young people an opportunity to have their say. Due to the length of the other five questionnaires when combined, this questionnaire was planned not to exceed ten questions. The topic areas that the questionnaire needed to focus on were discussed with the supervisory team and a list of priorities were collated.

Due to the personal and subjective nature of some of the questions, it was important to offer young people a chance to add their own thoughts and feelings, therefore, most questions had a comments option. The JDM experience questionnaire asked the following nine questions;

1. Do you think you still have JDM? (Yes/No/Don’t Know) (Any comment?).
2. If yes, how bad do you think it is? (Not bad at all, Ok, Bad, Very bad).
3. Can people see you have JDM by looking at you? (Yes/No/Don’t Know) (Any comment?).
4. Is it a good or a bad thing for people to see you have JDM or for them not to see it? (Good/Bad) (Why?).
5. Have you met anyone else with JDM? (Yes/No).
6. Would you like to meet someone else with JDM? (Yes/No/Don’t Know) (Any comment?).
7. Do you think you get enough support and help with your JDM? (free text).
8. What is the worst thing about having JDM? (free text).
9. Anything else you want to tell us? (free text).
Phase 2 “Picking up more passengers”

Unfortunately, the questionnaire was not piloted, this will be discussed further in the limitations section. The questionnaire was developed in Word® for the paper versions and an identical version was made in the electronic format. It was decided not to make the text fields compulsory for the JDM experience questionnaire in case the young people did not want to talk about personal aspects of their JDM.

For ease of reporting results responses to these nine questions, each will be simply referred to as Q1, Q2, etc. All direct quotes remain in italics and blue font.

5.7.8 Action in event of raised PI-ED score

As recommended by the manual for the P-IED tool (O’Connor 2010):

“Because the PI-ED measures emotional distress on a scale of severity, a higher score is indicative of greater levels of distress. For this reason, the PI-ED can be used as an index of therapeutic change and as a clinical outcome measure. If a child or young person scores above the cut-off value on the PI-ED, further assessment should follow” (O’Connor et al., 2010 p. 10)

There is no variation in cut-off value recommended by age, however there is a difference in gender, with a score of 10 or more for boys and a score of 11 or more for girls, indicating greater levels of distress. Therefore, it was important to examine the study sample to describe how many scored over this threshold, as this would give a clinical indication for further action. It is important to highlight that since this manual was written, a subsequent paper suggests that a cut-off score of 20 or more was established as an initial screen (O’Connor et al., 2016). However, for this study, the PI-ED manual was purchased at the same time that permission was granted to use the tool, and as this recommends the lower cut-off values it was felt important to abide by these to ensure children and young people were reviewed at potentially a cautious low threshold, ensuring any issues could be picked up early and safely.
5.8 Analytical approach

5.8.1 Analysis of the validated questionnaires

Data were uploaded and analysed using the Statistical Package for the Social Sciences (SPSS® version 21, manufacturer IBM). This phase was guided by the support and expertise of a senior statistician within the University, who was an invaluable member of the study team.

5.8.1.1 Exploratory and descriptive analysis

Initially exploratory work was undertaken, plotting histograms, getting a sense of the distributions, checking for missing data and comparing respondents with non-respondents. Descriptive analysis checked for missing data, examined published comparable data on the validated questionnaires and considered differences between centre responses.

For all outcome measures, histograms were plotted to identify the distribution of variables in order to evaluate whether parametric or non-parametric methods would be appropriate. Where variables were not normally distributed, both Pearson correlation coefficient and Spearman tests were conducted between independent and dependent variables. In general, results were very similar between both types of methods, therefore the results of the parametric method - the Pearson correlation coefficient are presented. This represents the relationship between the two variables without controlling for the effects of other variables.

Descriptive analysis was conducted to examine for missing data. If there was more than 25% missing data for any participant from the validated questionnaires, then their questionnaires were excluded from the analysis, as discussed further in the results section.

The respondents were compared with the non-respondents. Study data were compared with published reference data either from normative populations, or if not available, then from a comparable chronic illness. These results are presented using one sample T tests (comparing mean scores for our JDM
sample with published reference for a continuous variable) and a Chi-square test (to explore the relationship between two categorical variables). Means, confidence intervals (CI), standard deviations (SD) and P-values will be presented throughout.

Highlighting differences between the centres across the UK was kept to a minimum and as such differences between centres was not entered as a variable in the analysis. This was partly to avoid any patient identification (especially important given some centres had only a few JDM patients), but also as the study centres are different in terms of numbers of JDM patients and staff resource and support. The overall response rate and numbers of those who scored in the PI-ED’s is shown, to provide the reader with some background and relevant information, especially when considering patient representation and potential bias. Of note, all centres were coded to remain anonymous throughout this reporting.

5.8.1.2 Univariate analysis

For this study results were accepted as statistically significant if the probability p-value was ≤0.05 and are expressed throughout as p<0.001***, p<0.01**, p<0.05*.

Univariate analyses were performed to examine relationships between independent and each dependent variable separately. In the case of categorical independent variables, differences between categories in psychological distress and quality of life were analysed using independent two samples t-test for variables with two categories (e.g. gender) or analysis of variance (ANOVA) for independent variables with more than two categories e.g. ‘Can people see your JDM?’, (No, yes or don’t know). Associations between continuous variables were analysed using Pearson correlation calculation.

Pearson’s correlation coefficient was used to describe the strength and direction of a linear relationship between two continuous variables, identified as positive or negative with the size of the absolute value providing an indication of the strength of the relationship from -1 to +1 (Pallant, 2016).
Where correlations are highlighted, the direction of the relationship is further explained as some tools had higher scores for better clinical outcomes, e.g. the higher the PedsQL score the better the quality of life (this is also true for the Rheumatology Module and perception of benefit questionnaire), whereas for the PI-ED, CUIS, burden, the higher the score, the greater the emotional distress, level of uncertainty and perception of burden.

The patient-reported measures were separated into those that were outcomes; the PedsQL, the Rheumatology Module and the PI-ED, and the three patient perceptions as the independent variables; uncertainty, burden and benefit. The JDM experience questionnaire items were added into the analysis as independent variables where the data were reported numerically.

Where possible both the psychosocial and physical scores for the PedsQL are presented, but where no difference was found the total score is presented.

5.8.1.3 Multivariable analysis

Multiple regression analyses were conducted to examine the extent to which demographic, clinical and participant perception variables could help to explain variation in emotional distress and quality of life. All variables in the univariate analyses were considered, and independent variables that were significantly associated with the outcomes (*p<0.05) were entered in the multivariable linear regression(s). All models were adjusted for age and gender. No corrections were made for multiple comparison, as this is predominantly a descriptive study, however multiple comparisons were considered during data interpretation.

Regression analysis is a method of estimating and quantifying the numerical relationship between multiple variables to establish how well one variable can predict another (Bland, 1995). Linear regression was carried out for this study as it allowed a parsimonious approach, ‘building up, rather than trimming down’. This study has a relatively small sample size of 123 and as the rule of thumb discussed in Section 5.7.5 allowed for a maximum of eight variables, therefore for this study to prevent overfitting, it was decided to have a simpler model with fewer variables.
Adding multiple variables can be problematic due to multicollinearity, when two variables have a bivariate correlation of 0.7 or higher in the same analysis (Pallant, 2016). Therefore, independent variables which had a correlation of 0.7 or more were not included together.

All assumptions for standard regression analyses were checked, specifically inspecting the normal probability plot of the regression standardised residual, (looking for points in a reasonably straight diagonal line from bottom left to top right, suggesting no major deviations from normality, and a roughly rectangular distribution in the scatterplot of residuals with most of the scores concentrated in the centre).

The adjusted $R^2$ was examined for each multivariable analysis examining how much variance is explained by the model, with the higher the percentage the better the model fit. The standardized beta coefficient was studied during this multivariable analysis as this allowed us to compare the effect of each variable on outcome on a standardised scale. The larger the absolute value, the bigger the effect and the sign indicates the direction of the effect (the same as the actual coefficient). The standardised coefficients express the effect size in standard deviation units and this is preferable when exploring independent variables measured on different scales. Analysis was not specifically aimed at developing a predictive model, but to understand which factors were related to each outcome.

Where a categorical independent variable with more than two categories was significant in the univariate analysis such as “Do you still think you have JDM?” Yes, no or don’t know, it was necessary to create dummy variables to enter into the regression analyses in SPSS. The purpose of this is was to allow for multiple comparisons to be made for example, between those who said yes and no, and yes and don’t know and so forth.

5.8.2 Analysis of the free-text response in the JDM experience questionnaire

Intertwining the results from both the validated measures and the JDM experience questionnaire serve to present a comprehensive picture of JDM
from children and young people around the UK. The qualitative results are presented after the statistical analysis, as a content analysis approach with a systematic process of coding all responses was adopted (Vaismoradi, Turunen and Bondas, 2013). Content analysis is a general term that uses a systematic coding and categorizing approach for large amounts of textual information, looking for patterns and trends and frequency of words used (Vaismoradi, Turunen and Bondas, 2013). The content analysis is presented where possible in a simple radial list showing how many themes were identified in response to each question. For the penultimate question where participants were asked to highlight the worst thing about having JDM, their responses were analysed using content analysis looking for themes in the data and then presented as a ‘Word Cloud’ using freely available computer software. Free text comments are presented in this chapter where necessary to illustrate themes, however, all comments are presented in the appendices with further description of the coding for transparency and rigour.

5.9 Results

This section will begin by presenting the demographics, the characteristics of this cohort and the response rate. Following this, comparison of survey responders and non-responders will be presented in further detail to look for bias in the final sample. The demographics of the respondents will be presented, highlighting mean scores for each questionnaire. After presenting this descriptive data, the study sample including any clinical data collected will then be compared against available normative data. The relationships between the outcomes and variables are then presented, followed by the regression analysis. Each question of the JDM experience questionnaire is presented in turn, with the qualitative comments supporting the numerical data.

5.9.1 Target population and response

On the 19th September 2018, 533 children and young people were identified in the JDCBS. From these, 32 were excluded as they were below eight years of age, and 230 as they were older than 19 years of age, on this date, leaving 271 suitable for inclusion within the target age range. Of these, 26 were
Phase 2 “Picking up more passengers”

excluded as there was not a current address available or they could not be approached from local centre request due to child protection concerns. No emails were received from families requesting to opt out, therefore questionnaire packs were sent. One family requested a questionnaire pack after hearing about the study from another parent, but were not actually enrolled in the JDCBS. The team decided to send them a questionnaire pack for inclusivity and openness for the family, but it was decided in advance to not include their response as they did not meet the inclusion criteria of being in the JDCBS and thus would not have signed initial consent forms. Therefore 246 packs were sent out. Of the 246, 127 responded (52%). Of these, four could not be included; one was unreadable due to water spillage, one only had demographic data completed, one was later found to not have JDM (had been initially diagnosed with JDM, but over time this diagnosis got changed to SLE) and the one not in the JDCBS was excluded as discussed. This left a sample of 123 responders and 119 non-responders. These data are summarised in Figure 5-3.
Phase 2 “Picking up more passengers”

Of the 123 responders, paper responses were completed and returned for 91 (74%) children and young people, with only 32 (26%) being completed electronically.

5.9.2 Sample response by centre

Data were collected from 15 separate sites around the UK. Whilst this phase of the study was not to compare patient responses from one centre to another,

Figure 5-3 Flow diagram of patients who were included or excluded in the study.
Phase 2 “Picking up more passengers”

the percentages of those who responded in each centre are presented in Figure 5-4. These results are expressed as a percentage from each of the sites to provide an understanding of the representation of UK centres. All centres are coded, so remain anonymous.

The percentage response rate ranged from 0% in one centre (this site only had one potential responder), to 83% in a site which had five out of six young people who responded. The mean response rate across centres was 48%.

5.9.3 Demographic and clinical characteristics, including comparison of responders and non-responders

The demographic and clinical data are summarised in Table 5-1 and Table 5-2 including clinical variables and tests for significant differences between responder and non-responder groups.

The responder group had a mean age of 13.42 (SD 3.38), ranging from eight years and two months to 18 years and 10 months. Eighty two (67%) of the responders were female and 115 (97%) of these had received methotrexate at some point.
Phase 2 “Picking up more passengers”

The responders did not differ from the non-responders in terms of gender, medication usage, current age, year of age when diagnosed, or years since diagnosed. The only weakly significant result related to the number of years since last clinic visit, with there being a statistically significant difference of $P < 0.05^*$ with those respondents who completed the questionnaires having been seen more recently.

Table 5-1 Categorical data of responders/non-responders - Chi-squared test

<table>
<thead>
<tr>
<th></th>
<th>Non-responder (n = 119)</th>
<th>Responder (n = 123)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Female n (%)</td>
<td>74 (62)</td>
<td>82 (67)</td>
<td>0.47</td>
</tr>
<tr>
<td>Male n (%)</td>
<td>45 (38)</td>
<td>41 (33)</td>
<td></td>
</tr>
<tr>
<td>Received methotrexate at any point n (%)</td>
<td>109/117 (93)</td>
<td>115/119 (97)</td>
<td>0.22</td>
</tr>
</tbody>
</table>

Table 5-2 Continuous data of responders/non-responders using T test

<table>
<thead>
<tr>
<th></th>
<th>Non-responder (n=119) Mean (SD)</th>
<th>Responder (n=123) Mean (SD)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>13.36 (3.38)</td>
<td>13.42 (3.32)</td>
<td>0.89</td>
</tr>
<tr>
<td>Year of age when diagnosed</td>
<td>7.06 (3.38)</td>
<td>7.59 (3.73)</td>
<td>0.25</td>
</tr>
<tr>
<td>Years since diagnosed</td>
<td>7.17 (3.70)</td>
<td>6.58 (3.93)</td>
<td>0.24</td>
</tr>
<tr>
<td>Years since last visit</td>
<td>1.80 (2.54)</td>
<td>1.11 (1.73)</td>
<td>&lt;0.05*</td>
</tr>
</tbody>
</table>

Therefore, in conclusion, the responders are representative of the population under study and the results taken from this group can be used to generalise to the UK population of those aged eight - 19 year old children and young people who are in the JDCBS.

5.9.4 Missing data

Appendix 18 summarises the missing data. The completion rate ranged from 81-100%, with the JDM experience questionnaire having more missing data
Phase 2 “Picking up more passengers”

than the validated questionnaires, which were fully completed. As the proportions were so high for those that responded, it was not necessary to explore patterns of missing data. Therefore, only the one questionnaire already discussed in 5.9.1 was therefore excluded due to a significant amount of missing data.

5.9.5 Outcomes data

This section presents the means for each outcome assessed. Table 5-3 displays the summary data for each questionnaire across the whole sample of 123 responders. Each questionnaire is shown, with the mean, SD, and minimum and maximum scores presented.
Table 5-3 Mean score and ranges for all respondents for each measure

<table>
<thead>
<tr>
<th>Questionnaire</th>
<th>Total score / domain score</th>
<th>Scoring range</th>
<th>Mean</th>
<th>SD</th>
<th>Min</th>
<th>Max</th>
</tr>
</thead>
<tbody>
<tr>
<td>PedsQL Physical total score</td>
<td>0-100</td>
<td>75.6</td>
<td>24.7</td>
<td>6</td>
<td>100</td>
<td></td>
</tr>
<tr>
<td>PedsQL Emotional domain</td>
<td>0-100</td>
<td>73.6</td>
<td>23.1</td>
<td>20</td>
<td>100</td>
<td></td>
</tr>
<tr>
<td>PedsQL Social domain</td>
<td>0-100</td>
<td>83.5</td>
<td>20.8</td>
<td>25</td>
<td>100</td>
<td></td>
</tr>
<tr>
<td>PedsQL School domain</td>
<td>0-100</td>
<td>73.6</td>
<td>20.0</td>
<td>20</td>
<td>100</td>
<td></td>
</tr>
<tr>
<td>PedsQL Psychosocial total score</td>
<td>0-100</td>
<td>76.9</td>
<td>18.6</td>
<td>22</td>
<td>100</td>
<td></td>
</tr>
<tr>
<td>PedsQL Total PedsQL</td>
<td>0-100</td>
<td>76.5</td>
<td>19.7</td>
<td>16</td>
<td>100</td>
<td></td>
</tr>
<tr>
<td>Rheumatology Module Pain domain</td>
<td>0-100</td>
<td>73.6</td>
<td>25.9</td>
<td>0</td>
<td>100</td>
<td></td>
</tr>
<tr>
<td>Rheumatology Module Activities of Daily Living (ADL) domain</td>
<td>0-100</td>
<td>93.2</td>
<td>13.2</td>
<td>40</td>
<td>100</td>
<td></td>
</tr>
<tr>
<td>Rheumatology Module Treatment Domain</td>
<td>0-100</td>
<td>77.8</td>
<td>21.8</td>
<td>18</td>
<td>100</td>
<td></td>
</tr>
<tr>
<td>Rheumatology Module Worry domain</td>
<td>0-100</td>
<td>73.6</td>
<td>28.8</td>
<td>0</td>
<td>100</td>
<td></td>
</tr>
<tr>
<td>Rheumatology Module Communication domain</td>
<td>0-100</td>
<td>70.3</td>
<td>27.7</td>
<td>0</td>
<td>100</td>
<td></td>
</tr>
<tr>
<td>PI-ED PI-ED total score</td>
<td>0-42</td>
<td>10.6</td>
<td>7.5</td>
<td>0</td>
<td>29</td>
<td></td>
</tr>
<tr>
<td>CUIS CUIS total score</td>
<td>23-115</td>
<td>52.7</td>
<td>16.88</td>
<td>24</td>
<td>88</td>
<td></td>
</tr>
<tr>
<td>Benefit Benefit total score</td>
<td>10-50</td>
<td>32.1</td>
<td>9.1</td>
<td>10</td>
<td>50</td>
<td></td>
</tr>
<tr>
<td>Burden Burden total score</td>
<td>10-50</td>
<td>19.8</td>
<td>9.5</td>
<td>10</td>
<td>48</td>
<td></td>
</tr>
</tbody>
</table>

The study data means of all the validated questionnaires were studied using descriptive statistics.

5.9.6 Comparison of study data with previously published reference data

Although this study did not itself include a control or ‘healthy’ comparator group of children and young people, the data were compared with normative data, where available to address hypothesis one (see 5.6). Our study data is also compared with published data from chronically ill comparator populations,
Phase 2 “Picking up more passengers”
especially where normative data is not available. For clarity, when referring to
data from this phase two study, the terminology ‘study data’ will be used from
this point forwards.

5.9.6.1 Pediatric Quality of Life (PedsQL) published data

Table 5-4 summarises the PedsQL measures compared by one sample T test
to published norms in the healthy UK population of young people aged five -
18 years (Upton et al., 2005) and a JDM previously published population (Varni
et al., 2002). Of note, each published study is compared to the study sample
tested and the associated p-values are shown.

<table>
<thead>
<tr>
<th></th>
<th>Study data (n = 123)</th>
<th>95% CI</th>
<th>Normative healthy UK data (n = 1033) Mean (SD)</th>
<th>P-value</th>
<th>JDM published data n = 14 (Varni et al., 2002) Mean (SD)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Physical summary</td>
<td>75.6 (24.5) (71.3-80.0)</td>
<td>88.5 (11.6) n= 1032</td>
<td>&lt;0.001***</td>
<td>74.1 (20.1)</td>
<td>0.49</td>
<td></td>
</tr>
<tr>
<td>Emotional</td>
<td>73.6 (23.0) (69.5-77.7)</td>
<td>78.5 (17.9)</td>
<td>&lt;0.05*</td>
<td>78.9 (16.3)</td>
<td>&lt;0.01**</td>
<td></td>
</tr>
<tr>
<td>Social</td>
<td>83.5 (20.8) (79.8-87.2)</td>
<td>87.6 (16.5)</td>
<td>&lt;0.05*</td>
<td>74.3 (23.8)</td>
<td>&lt;0.001 ***</td>
<td></td>
</tr>
<tr>
<td>School</td>
<td>73.6 (20.0) (70.0-77.2)</td>
<td>78.9 (15.9)</td>
<td>&lt;0.01**</td>
<td>79.3 (17.2)</td>
<td>&lt;0.01**</td>
<td></td>
</tr>
<tr>
<td>Psychosocial summary</td>
<td>76.9 (18.6) (73.6-80.2)</td>
<td>81.6 (13.2)</td>
<td>&lt;0.01**</td>
<td>77.5 (17.6)</td>
<td>0.71</td>
<td></td>
</tr>
<tr>
<td>Total score</td>
<td>76.5 (19.7) (73.0-78.0)</td>
<td>83.9 (11.8)</td>
<td>&lt;0.001***</td>
<td>76.3 (17.7)</td>
<td>0.93</td>
<td></td>
</tr>
</tbody>
</table>

Compared to the Upton et al., (2005) UK study of healthy school children,
patients scored significantly lower in all domains of quality of life, as can be
seen from the normative means which consistently fell outside the study data
confidence interval. Compared to the small JDM published data of 14
participants in the US (Varni et al., 2002), the participants in the current study
Phase 2 “Picking up more passengers” had statistically significant lower scores for emotional and schooling indicating worse quality of life in these domains, but significantly better quality of life in the social domain than the published data. Whilst it is acknowledged that it is not ideal to include such a small number as the published data on the 14 JDM participants, unfortunately as previously discussed there is limited research on this area in the literature.

5.9.6.2 Rheumatology Module published data

Table 5-5 presents means and SD for the study data for all domains of the Rheumatology Module and results from Varni et al., (2002) focusing on quality of life in paediatric rheumatic disease, assessed by one sample T test. The number of subjects varies for the Varni et al., (2002) paper, so these are shown in brackets, however the JDM reference data is for the subset of 14 participants in this paper.

<table>
<thead>
<tr>
<th>Domain</th>
<th>Study data (n = 123) Mean (SD)</th>
<th>95 % CI</th>
<th>Children with Rheumatic disease (Varni et al., 2002) Mean (SD)</th>
<th>P-value</th>
<th>JDM published data n=14 patients (Varni et al., 2002) Mean (SD)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pain</td>
<td>73.6 (25.9)</td>
<td>(69.0, 78.3)</td>
<td>61.9 (28.3)</td>
<td>&lt;0.001***</td>
<td>77.7 (16.8)</td>
<td>0.086</td>
</tr>
<tr>
<td>Activities of Daily Living</td>
<td>93.2 (13.2)</td>
<td>(90.9, 95.6)</td>
<td>90.1 (15.0)</td>
<td>&lt;0.01**</td>
<td>95.0 (7.8)</td>
<td>0.14</td>
</tr>
<tr>
<td>Treatment</td>
<td>77.8 (21.8)</td>
<td>(73.9, 81.7)</td>
<td>78.0 (19.6)</td>
<td>0.93</td>
<td>84.1 (15.1)</td>
<td>&lt;0.01**</td>
</tr>
<tr>
<td>Worry</td>
<td>73.6 (28.8)</td>
<td>(68.5, 78.8)</td>
<td>74.2 (25.2)</td>
<td>0.84</td>
<td>74.0 (27.1)</td>
<td>0.88</td>
</tr>
<tr>
<td>Communication</td>
<td>70.3 (27.7)</td>
<td>(65.3, 75.2)</td>
<td>70.5 (26.9) (n=231)</td>
<td>0.91</td>
<td>75.6 (20.3)</td>
<td>&lt;0.05*</td>
</tr>
</tbody>
</table>

Compared to Varni et al., (2002) data from the US, patients in the current study scored significantly higher in the pain and activities of daily living domains indicating better quality of life in those two domains. However, the US study
Phase 2 “Picking up more passengers”

included children and young people with fibromyalgia which as highlighted by the authors, skewed the data, especially in the pain domain, therefore making comparisons complicated. Compared to the subset of JDM in this paper, the study data, whilst small numbers, the UK participants scored lower on the treatment and communication domains of the Rheumatology Module indicating poorer quality of life on those domains.

5.9.6.3 Paediatric Index of Emotional Distress (PI-ED) normative data

Normative data for the PI-ED from the development phase based on a sample of school children between seven – 17 years of age, are presented in Table 5-6 in comparison to our findings from the PI-ED, assessed by one sample T test.

<table>
<thead>
<tr>
<th></th>
<th>Study data (n = 123)</th>
<th>School based sample (n = 1026) (O’Connor et al., 2016)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Mean (SD)</td>
<td>Mean (SD)</td>
<td></td>
</tr>
<tr>
<td>Female</td>
<td>10.6 (10.6)</td>
<td>12.8 (6.2)</td>
<td>&lt;0.01**</td>
</tr>
<tr>
<td>Male</td>
<td>11.2 (7.6)</td>
<td>13.8 (6.6)</td>
<td></td>
</tr>
<tr>
<td>Mean (SD)</td>
<td>9.3 (7.4)</td>
<td>11.9 (5.8)</td>
<td></td>
</tr>
</tbody>
</table>

Compared to the UK O’Connor et al., (2016) data, this study population reported less emotional distress than the normative sample of 1026 healthy school children, with the mean score falling outside of the confidence interval of the current study data. Even when breaking down by gender, to allow for the fact that JDM gender distribution is predominantly female this study data reported less emotional distress than the published data.
5.9.6.4 Childhood Uncertainty in Illness Scale (CUIS) previously published data

As this questionnaire examines perception of illness uncertainty, there is no normative data from a healthy population. However, this study data had significantly lower uncertainty scores than the small US sample of Juvenile Rheumatic Diseases (JRD) in 50 children aged five - 17 years old, including seven patients with JDM, as shown in Table 5-7, as assessed by the one sample T test. The uncertainty score measured by the CUIS, has a range of 23-115, with the lower the score representing a lower level of perceived uncertainty (White et al., 2005).

<table>
<thead>
<tr>
<th></th>
<th>Study data (n = 123)</th>
<th>JRD published data (n = 50), including 7 JDM (White et al., 2005)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Mean (SD)</td>
<td>Mean (SD)</td>
<td></td>
</tr>
<tr>
<td><strong>CUIS</strong></td>
<td>52.67 (16.88)</td>
<td>67.02 (18.50)</td>
<td>&lt;0.001***</td>
</tr>
<tr>
<td>95% CI</td>
<td>(49.65-55.68)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

The implications of this lower uncertainty finding in the UK JDM cohort will be explored further in the discussion chapter.

5.9.6.5 Benefit and Burden Scale for Children (BBSC) published data

Limited data of the BBSC questionnaire were available for comparison. Again, there is no healthy child data to compare to, as this questionnaire measures benefit and burden in children and young people with a health condition. Table 5-8 presents perception of benefit and burden results compared against (Currier, Hermes and Phipps, 2009) data of 78 American children with a variety of oncological diagnoses, ranging from eight - 18 years of age, by one sample T test.
As can be seen from these data, the study population had a lower perception of benefit, than the oncology population. Whereas the burden was comparable, with the published mean falling within the study confidence interval range.

### 5.9.7 Raised Paediatric Index of Emotional Distress (PI-ED) scores

As raised PI-ED scores had to be flagged with each individual centre, the data are presented here by centre, to inform the reader of how many children and young people needed to be reviewed and potential further action taken across the UK.

The results are summarised in Table 5-9, presenting the number that responded and those that scored (all centres are coded).
Phase 2 “Picking up more passengers”

Table 5-9 Those that scored over cut off threshold on PI-ED measure

<table>
<thead>
<tr>
<th>Centre number</th>
<th>Number of responders (%)</th>
<th>Number of raised PI-ED’s (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>62/112 (55%)</td>
<td>25/62 (39%)</td>
</tr>
<tr>
<td>2</td>
<td>6/15 (40%)</td>
<td>2/6 (33%)</td>
</tr>
<tr>
<td>3</td>
<td>9/15 (60%)</td>
<td>1/9 (11%)</td>
</tr>
<tr>
<td>4</td>
<td>4/12 (33%)</td>
<td>1/4 (25%)</td>
</tr>
<tr>
<td>5</td>
<td>6/14 (43%)</td>
<td>2/6 (33%)</td>
</tr>
<tr>
<td>6</td>
<td>5/13 (38%)</td>
<td>5/5 (100%)</td>
</tr>
<tr>
<td>7</td>
<td>10/26 (38%)</td>
<td>5/10 (50%)</td>
</tr>
<tr>
<td>8</td>
<td>3/5 (60%)</td>
<td>0/3 (0%)</td>
</tr>
<tr>
<td>9</td>
<td>2/5 (40%)</td>
<td>1/2 (50%)</td>
</tr>
<tr>
<td>10</td>
<td>3/8 (38%)</td>
<td>1/3 (33%)</td>
</tr>
<tr>
<td>11</td>
<td>3/4 (75%)</td>
<td>1/3 (33%)</td>
</tr>
<tr>
<td>12</td>
<td>5/6 (83%)</td>
<td>4/5 (80%)</td>
</tr>
<tr>
<td>13</td>
<td>0/1 (0%)</td>
<td>0/0 (0%)</td>
</tr>
<tr>
<td>14</td>
<td>3/5 (60%)</td>
<td>1/3 (33%)</td>
</tr>
<tr>
<td>15</td>
<td>2/4 (50%)</td>
<td>0/2 (0%)</td>
</tr>
<tr>
<td>TOTAL</td>
<td>123/245 (50%)</td>
<td>49/123 (40%)</td>
</tr>
</tbody>
</table>

Of note, the total in this table is 245, as the one sent a separate questionnaire pack was not enrolled in one of the centres.

A total of 49 (40%) of participants who completed the PI-ED scored above the recommended cut-off for emotional distress and therefore needed highlighting to their local team, with further assessment to follow.

5.9.8 Univariate analysis

Following the descriptive analyses above, univariate analysis was performed as explained in Section 5.8.1.2.
Phase 2 “Picking up more passengers”

The JDM experience questionnaire was important as it was designed to capture some of the reoccurring concerns from the interviews in phase one which were not captured in any of the validated measures. Therefore, when considering the psychosocial well-being of the children and young people, this JDM experience questionnaire had the potential to add some further insight regarding the day to day lives of children and young people with JDM. The four questions (Q1, Q3, Q5 and Q6) which had a numerical score were added into the univariate analysis to consider their relationship with the variables. Q2 was a filter question, so was only a small proportion of the full sample, so this is not included and nor was Q4 as this was an ambiguous question yielding confused results (as discussed later). Q7, Q8 and Q9 were free text only, these are discussed in Section 5.9.10.

5.9.8.1 Analysis of continuous demographical data and clinical variables

Initially the relationship between the demographic and clinical variables, (such as age, years since diagnosis and length of time since last visit) were examined across all the validated measures, in order to explore hypothesis four (See 5.6). Table 5-10 summarises the results using Pearson correlation coefficient.
Table 5-10 Pearson correlation coefficient for the continuous data

<table>
<thead>
<tr>
<th></th>
<th>PedsQL</th>
<th>Rheumatology Module</th>
<th>PI-ED</th>
<th>CUIS</th>
<th>Benefit</th>
<th>Burden</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Psych</td>
<td>Phy</td>
<td>Total</td>
<td>Pain</td>
<td>ADL</td>
<td>Treat</td>
</tr>
<tr>
<td>Current age</td>
<td>-0.08</td>
<td>-0.12</td>
<td>-0.10</td>
<td>-0.05</td>
<td>0.04</td>
<td>-0.19 (&lt;0.05̊)</td>
</tr>
<tr>
<td>Years since diagnosis</td>
<td>0.17 (0.60)</td>
<td>0.03 (0.74)</td>
<td>0.09 (0.31)</td>
<td>0.09 (0.33)</td>
<td>0.19 (&lt;0.05̊)</td>
<td>0.24 (&lt;0.01** )</td>
</tr>
<tr>
<td>Length of time since last visit</td>
<td>0.10 (0.31)</td>
<td>0.15 (0.10)</td>
<td>0.13 (0.15)</td>
<td>0.09 (0.32)</td>
<td>0.08 (0.38)</td>
<td>0.208 (&lt;0.05̊)</td>
</tr>
</tbody>
</table>

(Psych = psychosocial, Phy = physical, ADL = Activities of Daily Living, Treat = treatment, Com = communication)
As seen in Table 5-10, the treatment domain on the Rheumatology Module was statistically associated with these three demographic and clinical variables, indicating that older age, longer duration since diagnosis and a longer duration since last clinic visit were associated with better quality of life in the treatment domain. The older age of participant was also statistically significantly associated with a raised PI-ED score, indicating greater emotional distress among older participants.

Only 56 (49%) out of the 114 participants who there was data on had been seen clinically in the last year. There was no date attached to nine participants’ last clinic visit. The length of time since their last visit was a significant finding with a positive correlation of the treatment domain of the Rheumatology Module. This indicated a lower length of time since last visit, was significantly correlated with a worse treatment score.

Older age was significantly associated with a higher score on the burden tool indicating greater perceived illness-related burden among older participants. Length of time since last visit was negatively correlated with uncertainty and benefit, indicating greater perceived uncertainty and greater perceived benefit finding among participants who had more recently been seen in clinic for their JDM.

5.9.8.2 Analysis of categorical; clinical, demographic and experience data

Univariate analysis of all independent variables may help to answer hypotheses two and three, and therefore all variables were next examined in detail. The demographic variable (gender), the experience data (Q5) and the clinical variable data (rash and weakness at last visit) in the JDCBS database were examined and compared using T-tests against the outcomes and independent variables. Results are shown in Table 5-11 and Table 5-12
Phase 2 “Picking up more passengers”

<table>
<thead>
<tr>
<th></th>
<th>Psych</th>
<th>Phy</th>
<th>Total score</th>
<th>Pain</th>
<th>ADL</th>
<th>Treat</th>
<th>Worry</th>
<th>Com</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>M (SD)</td>
<td>P-value</td>
<td>M (SD)</td>
<td>P-value</td>
<td>M (SD)</td>
<td>P-value</td>
<td>M (SD)</td>
<td>P-value</td>
</tr>
<tr>
<td>Gender</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>F (82)</td>
<td>75.5 (19.2)</td>
<td>0.23</td>
<td>72.8 (25.1)</td>
<td>0.06</td>
<td>74.5 (20.4)</td>
<td>0.12</td>
<td>72.2 (25.3)</td>
<td>0.38</td>
</tr>
<tr>
<td>M (41)</td>
<td>79.8 (17.0)</td>
<td></td>
<td>81.4 (22.3)</td>
<td></td>
<td>80.4 (17.7)</td>
<td></td>
<td>76.5 (27.2)</td>
<td></td>
</tr>
<tr>
<td>Have you met anyone else?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>(N=122)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No (62)</td>
<td>79.4 (18.7)</td>
<td>0.12</td>
<td>77.1 (23.6)</td>
<td>0.47</td>
<td>78.6 (19.5)</td>
<td>0.21</td>
<td>75.0 (25.6)</td>
<td>0.49</td>
</tr>
<tr>
<td>Yes (60)</td>
<td>74.1 (18.3)</td>
<td></td>
<td>73.9 (25.5)</td>
<td></td>
<td>74.1 (19.9)</td>
<td></td>
<td>71.8 (26.4)</td>
<td></td>
</tr>
<tr>
<td>Hash at last visit</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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</tr>
<tr>
<td>(N=116)</td>
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<td></td>
<td></td>
</tr>
<tr>
<td>No (87)</td>
<td>80.7 (16.2)</td>
<td>&lt;0.001***</td>
<td>81.0 (20.6)</td>
<td>&lt;0.001***</td>
<td>80.8 (16.6)</td>
<td>&lt;0.001***</td>
<td>78.3 (23.1)</td>
<td>&lt;0.001***</td>
</tr>
<tr>
<td>Yes (29)</td>
<td>67.1 (22.2)</td>
<td></td>
<td>63.3 (28.9)</td>
<td></td>
<td>65.7 (24.1)</td>
<td></td>
<td>60.3 (30.2)</td>
<td></td>
</tr>
<tr>
<td>Weakness at last visit</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
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<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No (104)</td>
<td>79.6 (17.1)</td>
<td>&lt;0.001***</td>
<td>80.9 (19.7)</td>
<td>&lt;0.001***</td>
<td>80.1 (17.1)</td>
<td>&lt;0.001***</td>
<td>77.7 (20.8)</td>
<td>&lt;0.001***</td>
</tr>
<tr>
<td>Yes (12)</td>
<td>57.1 (21.4)</td>
<td></td>
<td>39.1 (26.7)</td>
<td></td>
<td>50.8 (22.7)</td>
<td></td>
<td>40.1 (18.7)</td>
<td></td>
</tr>
</tbody>
</table>

Psych = psychosocial, Phy = Physical, ADL = Activities of Daily Living, Treat = treatment, Com = communication
### Table 5-12 Categorical binary data and validated questionnaires

<table>
<thead>
<tr>
<th></th>
<th>PIED</th>
<th>CUIS</th>
<th>Benefit</th>
<th>Burden</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>M (SD)</td>
<td>M (SD)</td>
<td>M (SD)</td>
<td>M (SD)</td>
</tr>
<tr>
<td>Gender (n=123)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>F (82)</td>
<td>11.2 (7.6)</td>
<td>54.0 (16.0)</td>
<td>32.5 (9.5)</td>
<td>20.8 (9.6)</td>
</tr>
<tr>
<td>M (41)</td>
<td>9.3 (7.4)</td>
<td>50.1 (16.5)</td>
<td>31.4 (10.3)</td>
<td>17.7 (9.1)</td>
</tr>
<tr>
<td>Have you met anyone else?</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No (62)</td>
<td>9.3 (7.2)</td>
<td>50.7 (17.2)</td>
<td>32.2 (8.6)</td>
<td>18.5 (8.8)</td>
</tr>
<tr>
<td>Yes (60)</td>
<td>11.9 (7.7)</td>
<td>54.9 (16.5)</td>
<td>32.3 (9.4)</td>
<td>21.3 (10.2)</td>
</tr>
<tr>
<td>Rash at last visit (n=116)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No (87)</td>
<td>9.95 (7.2)</td>
<td>50.0 (16.3)</td>
<td>31.3 (9.7)</td>
<td>17.7 (7.9)</td>
</tr>
<tr>
<td>Yes (29)</td>
<td>12.8 (8.4)</td>
<td>61.1 (16.8)</td>
<td>34.1 (7.2)</td>
<td>26.2 (11.4)</td>
</tr>
<tr>
<td>Weakness at last visit (n=116)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No (104)</td>
<td>10.0 (7.2)</td>
<td>51.5 (17.1)</td>
<td>31.7 (9.3)</td>
<td>18.8 (8.9)</td>
</tr>
<tr>
<td>Yes (12)</td>
<td>16.5 (8.6)</td>
<td>64.0 (12.8)</td>
<td>34.8 (8.2)</td>
<td>28.8 (10.8)</td>
</tr>
</tbody>
</table>

Note: P-values indicated with asterisks: <0.01**, <0.05*, <0.001***
5.9.8.2.1 Demographic variables

Gender was significant for the communication and worry domains of the Rheumatology Module, with girls reporting poorer quality of life in the both domains.

5.9.8.2.2 Clinical variables

There was only data on 116 participants who had rash at last visit and only data on 116 participants who had weakness at last visit (not all the same 116 were scored for both), with seven individuals having no data in the JDCBS for this. Those who had a rash at their last clinic visit reported poorer quality of life on the PedsQL and all Rheumatology Module domains, apart from the activities of daily living domain. They also reported more uncertainty and a higher perception of burden. Similarly, participants who were experiencing weakness at their last clinic visit reported poorer quality of life on the PedsQL and all Rheumatology Module domains, apart from the communication domain. Respondents who weakness at their last visit also had a higher emotional distress score, more uncertainty and a higher perception of burden. Whilst interesting findings, due to the variability of time elapsed since this data was recorded, these particular two clinical variables were not taken any further through the analysis. This is explained in detail later in this chapter (see 5.15).

5.9.8.2.3 Experience variables

Q5 of the JDM experience questionnaire which asked “Have you met anyone else with JDM?”, was significant with the worry and treatment domains, with those who had met someone else scoring worse quality of life for worry and treatment.

Table 5-13 and Table 5-14 presents the results from the ANOVA analysis for the categorical data of the patient experience questionnaire for Q1, Q3 and Q6 with the validated questionnaires.
Table 5-13 Experience questionnaire compared with QOL questionnaires

<table>
<thead>
<tr>
<th>Question</th>
<th>Psych</th>
<th>Phy</th>
<th>Total score</th>
<th>Pain</th>
<th>ADL</th>
<th>Treat</th>
<th>Worry</th>
<th>Com</th>
</tr>
</thead>
<tbody>
<tr>
<td>Q1: Do you still have JDM?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>88.7</td>
<td>91.9</td>
<td>89.9</td>
<td>89.5</td>
<td>99.6</td>
<td>93.9</td>
<td>94.1</td>
<td>85.1</td>
</tr>
<tr>
<td>Yes</td>
<td>71.8</td>
<td>70.3</td>
<td>66.0</td>
<td>90.1</td>
<td>69.3</td>
<td>66.3</td>
<td>66.3</td>
<td>66.3</td>
</tr>
<tr>
<td>DK</td>
<td>78.3</td>
<td>79.1</td>
<td>77.6</td>
<td>94.8</td>
<td>84.1</td>
<td>71.5</td>
<td>66.3</td>
<td>64.6</td>
</tr>
<tr>
<td>Q3: Can people see it?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>78.4</td>
<td>78.2</td>
<td>75.6</td>
<td>93.2</td>
<td>78.1</td>
<td>75.0</td>
<td>71.8</td>
<td>71.8</td>
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<tr>
<td>Yes</td>
<td>67.7</td>
<td>66.0</td>
<td>59.7</td>
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<td>74.7</td>
<td>58.3</td>
<td>57.6</td>
<td>57.6</td>
</tr>
<tr>
<td>DK</td>
<td>73.7</td>
<td>71.6</td>
<td>68.3</td>
<td>91.5</td>
<td>76.4</td>
<td>74.4</td>
<td>71.8</td>
<td>71.8</td>
</tr>
</tbody>
</table>

Psych = psychosocial, ADL = Activities of Daily Living, Treat = treatment, Com = communication

(Values that differed significantly at <0.05* sharing the same letter. a = Yes and no differ, b = Yes and don’t know differ, c = No and don’t know differ)
Table 5.13 Experience questionnaire compared with QOL questionnaires

<table>
<thead>
<tr>
<th>Q6 Want to meet anyone else? (n=122)</th>
<th>Psych</th>
<th>Phy</th>
<th>Total score</th>
<th>Rheumatology Module</th>
</tr>
</thead>
<tbody>
<tr>
<td>No (16)</td>
<td>80.9 (20.0)</td>
<td>77.5 (27.0)</td>
<td>79.8 (21.7)</td>
<td>0.17</td>
</tr>
<tr>
<td>Yes (73)</td>
<td>74.2 (19.3)</td>
<td>73.5 (24.5)</td>
<td>74.0 (20.3)</td>
<td>0.52</td>
</tr>
<tr>
<td>DK (33)</td>
<td>80.6 (15.6)</td>
<td>79.1 (23.7)</td>
<td>80.1 (17.2)</td>
<td>0.26</td>
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</tbody>
</table>

Psych = psychosocial, ADL = Activities of Daily Living, Treat = treatment, Com = communication

(Values that differed significantly at <0.05* sharing the same letter. a = Yes and no differ, b = Yes and don’t know differ, c = No and don’t know differ)
Table 5-14 Experience questionnaire compared to validated questionnaires

<table>
<thead>
<tr>
<th></th>
<th>PIED</th>
<th></th>
<th>CUIS</th>
<th></th>
<th>Benefit</th>
<th></th>
<th>Burden</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>M (SD)</td>
<td>P-value</td>
<td>M (SD)</td>
<td>P-value</td>
<td>M (SD)</td>
<td>P-value</td>
<td>M (SD)</td>
</tr>
<tr>
<td>Q1</td>
<td>Do you still have JDM? (n=123)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>7.6 (5.6)</td>
<td>0.06</td>
<td>38.0(^a) (13.2)</td>
<td>&lt;0.001***</td>
<td>27.7(^a) (9.6)</td>
<td>&lt;0.05*</td>
<td>15.3(^a) (6.4)</td>
</tr>
<tr>
<td>Yes</td>
<td>11.4 (7.3)</td>
<td></td>
<td>58.5(^a) (14.9)</td>
<td>33.3(^a) (8.4)</td>
<td>21.8(^a) (10.3)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>DK</td>
<td>11.6 (9.4)</td>
<td></td>
<td>52.5(^c) (16.2)</td>
<td>33.7 (9.4)</td>
<td>19.0 (8.7)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Q3</td>
<td>Can people see it? (n=122)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>9.9 (7.5)</td>
<td>0.10</td>
<td>50.2(^c) (16.3)</td>
<td>&lt;0.01**</td>
<td>32.5 (9.1)</td>
<td>0.83</td>
<td>18.5(^a) (8.8)</td>
</tr>
<tr>
<td>Yes</td>
<td>14.6 (8.0)</td>
<td></td>
<td>62.4 (15.3)</td>
<td>32.1 (8.8)</td>
<td>27.6(^a) (12.2)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>DK</td>
<td>12.4 (7.1)</td>
<td></td>
<td>63.5(^c) (17.2)</td>
<td>30.9 (8.8)</td>
<td>23.4 (9.5)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Q6</td>
<td>Want to meet anyone else? (n=122)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>9.6 (8.0)</td>
<td>0.19</td>
<td>49.8 (18.4)</td>
<td>0.75</td>
<td>28.4 (9.8)</td>
<td>0.08</td>
<td>18.9 (8.9)</td>
</tr>
<tr>
<td>Yes</td>
<td>11.6 (7.7)</td>
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<td>53.3 (16.8)</td>
<td>33.6 (8.5)</td>
<td>20.8 (10.2)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>DK</td>
<td>8.8 (6.8)</td>
<td></td>
<td>53.0 (16.9)</td>
<td>31.2 (9.1)</td>
<td>18.2 (8.2)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Values that differed significantly at <0.05* sharing the same letter. \(^a\) = Yes and no differ, \(^b\) = Yes and don’t know differ, \(^c\) = No and don’t know differ
Phase 2 “Picking up more passengers”

Question 1 asked the children and young people whether they thought they still had JDM, if they answered yes, this was significant with a lower PedsQL score in all domains, particularly the physical domain, indicating a worse quality of life. They also scored significantly worse in all Rheumatology Module domains. Answering yes to this question, was also significant with a higher uncertainty score, a greater perception of benefit and a greater perception of burden.

When asked “If people could see their JDM?” Q3, this was only significant with greater uncertainty and greater perceived burden.

The question, “Would you like to meet anyone else with JDM?” Q6, was not significant with any of the validated questionnaires.

5.9.8.3 Relationships between outcomes and independent variables

The next section set out to address hypothesis two and three (See 5.6) examining uncertainty and perception of benefit in further detail. In the model developed here the PedsQL total score, the Rheumatology Module and the PI-ED are the three outcomes. The independent variables consist of; uncertainty, burden and benefit, age, gender, demographic, clinical and JDM experience variables. Appendix 19 presents the Scatterplot matrixes. These graphs show a negative relationship between quality of life score and the PI-ED, CUIS and Burden scores in this study cohort indicating that as emotional distress, uncertainty or perception of burden increase, then quality of life declines. As expected, there is a positive relationship between the PI-ED and the CUIS and burden, as one increases, so does the other. Appendix 20 presents similar scatterplot matrixes for the five domains of the Rheumatology Module.

Pearson correlations were conducted to examine and quantify the relationship between patient perceptions and outcomes measured to look for correlations within domains of the PedsQL and Rheumatology Module and PI-ED. Results are shown in Table 5-15.
This table shows that as uncertainty and burden increase, quality of life decreases, (as shown by the negative correlations with the PedsQL Physical, Psychosocial and Total scores and each of the Rheumatology Module domains). There is a positive correlation between the uncertainty and burden score and the emotional distress score as measured by the PI-ED. Of note, perceived benefit was not significantly correlated with quality of life or emotional distress outcomes.

Also to note, burden and PI-ED were highly correlated at 0.78, which is above the recommended bivariate correlation level of 0.70 as suggested by Pallant, (2016) as discussed in 5.8.1.3. Therefore to avoid multicollinearity when entering the variables in the multiple linear regression analysis, emotional distress as measured by the PI-ED was retained as the outcome and burden as the variable.

Furthermore, when plotted against each other and quantifying the relationship using Pearson’s correlation coefficient, uncertainty and burden are correlated and this correlation is statistically significant, (p<0.001***) with those participants reporting greater illness uncertainty also reporting greater illness burden.
Phase 2 “Picking up more passengers”

Benefit was found to not be significant with any of the outcomes examined. Benefit finding was also not correlated with uncertainty in this study data set (see Table 5-16), nor were benefit and burden correlated.

<table>
<thead>
<tr>
<th></th>
<th>Burden</th>
<th>Benefit</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Pearsons (r)</td>
<td>P-value</td>
</tr>
<tr>
<td>CUIS</td>
<td>0.64</td>
<td>&lt;0.001***</td>
</tr>
<tr>
<td>Benefit</td>
<td>0.03</td>
<td>.77</td>
</tr>
</tbody>
</table>

When starting to address hypothesis two, data from the PI-ED were compared to all validated questionnaires. All independent variables were statistically significantly associated with children and young people scoring over the PI-ED recommended cut-off, as demonstrated in Table 5-17 and Table 5-18, assessed by independent sample T test. However, the JDM experience questions were not significant when compared against the PI-ED over cut off and under cut off score.
### Table 5-17 PI-ED clinical cut-off scores for QOL

<table>
<thead>
<tr>
<th></th>
<th>Psych</th>
<th>Phy</th>
<th>Total score</th>
<th>Pain</th>
<th>ADL</th>
<th>Treat</th>
<th>Worry</th>
<th>Com</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>M (SD)</td>
<td>P-value</td>
<td>M (SD)</td>
<td>P-value</td>
<td>M (SD)</td>
<td>P-value</td>
<td>M (SD)</td>
<td>P-value</td>
</tr>
<tr>
<td>PI-ED <strong>over</strong> cut off (n=49)</td>
<td>62.67 (17.6)</td>
<td>&lt;0.001 ***</td>
<td>61.10 (26.8)</td>
<td>&lt;0.001 ***</td>
<td>62.16 (19.6)</td>
<td>&lt;0.001 ***</td>
<td>55.61 (26.4)</td>
<td>&lt;0.001 ***</td>
</tr>
<tr>
<td>PI-ED <strong>under</strong> cut off (n=74)</td>
<td>86.31 (12.1)</td>
<td>85.26 (17.1)</td>
<td>85.94 (12.9)</td>
<td>85.56 (17.4)</td>
<td>96.49 (9.0)</td>
<td>84.60 (18.3)</td>
<td>84.80 (22.5)</td>
<td>81.08 (21.3)</td>
</tr>
</tbody>
</table>

(Psych = psychosocial, Phy = Physical, ADL = Activities of Daily Living, Treat = treatment, Com = communication)

### Table 5-18 PIED clinical cut-off scores for CUIS, Benefit and Burden

<table>
<thead>
<tr>
<th></th>
<th>CUIS</th>
<th>Benefit</th>
<th>Burden</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>M (SD)</td>
<td>P-value</td>
<td>M (SD)</td>
</tr>
<tr>
<td>PI-ED <strong>over</strong> cut off (n = 49)</td>
<td>62.96 (14.90)</td>
<td>&lt;0.001 ***</td>
<td>31.31 (8.80)</td>
</tr>
<tr>
<td>PI-ED <strong>under</strong> cut off (n=74)</td>
<td>45.85 (14.55)</td>
<td>32.58 (9.36)</td>
<td>15.07 (4.33)</td>
</tr>
</tbody>
</table>
5.9.9 Multivariable analysis

The next step was to enter the independent variables together into a multiple regression analysis to see to what extent they explained the relative importance of variance in outcomes and which remained as independent predictors. The overall goal of the regression analyses was to examine the extent to which the independent variables explained variance in the outcomes; quality of life scores and emotional distress. All models were adjusted for age and gender. As any variable in the univariate regression with a p-value $p<0.05^*$ was included into the multivariate analysis (see 5.8.1.3), some of the demographic and JDM experience questionnaire responses were included. Years since diagnosis and length of time since last visit were collected from the JDCBS. From the bespoke questionnaire Q1 (“Do you still think you have JDM?”) and Q5 (“have you met anyone else?”) from the JDM experience questionnaire were included where there was a significant relationship between them and any of the outcomes. Q1 of the bespoke questionnaire was added into the regressions after it was recoded (See 5.8.1.3) as it was not a binary variable. The rule-of-thumb expressed earlier in Section 5.7.5 indicates that the final sample size of 123 was sufficient for a regression with eight independent variables, the most included during this regression analysis was seven.

Not all of the univariate analysis could be completed on the full sample as for the JDM experience questions, there was some missing data. For the univariate analysis, only those that do not contain all 123 responses are stated, whereas all the others do have the full 123 sample set.

The regression outcomes will be presented in turn, beginning with the PedsQL total score (as the findings were so similar with the psychosocial and physical summary scores, it was decided to adopt those of the total score), then the five domains of the Rheumatology Module and lastly, the PI-ED.
5.9.9.1 PedsQL – TOTAL Score

CUIS, Burden and Q1.'Do you still have JDM?' were significant in the univariate analyses and were therefore entered into the regression along with age and gender. See Table 5-19.

<table>
<thead>
<tr>
<th></th>
<th>Multivariable Coefficient (Unstandardised B)</th>
<th>P-value</th>
<th>(R²) adjusted</th>
<th>Standardized Beta Coefficients</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>N=123</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Age</td>
<td>.11</td>
<td>0.8</td>
<td>.59</td>
<td>-.28</td>
</tr>
<tr>
<td>Gender</td>
<td>.01</td>
<td>0.9</td>
<td></td>
<td>.01</td>
</tr>
<tr>
<td>CUIS</td>
<td>-.20</td>
<td>&lt;0.05*</td>
<td></td>
<td>-.17</td>
</tr>
<tr>
<td>Burden</td>
<td>-1.20</td>
<td>&lt;0.001***</td>
<td></td>
<td>-.58</td>
</tr>
<tr>
<td>Do you still have JDM?</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>-4.16</td>
<td>.17</td>
<td></td>
<td>-.11</td>
</tr>
<tr>
<td>No</td>
<td>3.70</td>
<td>.34</td>
<td></td>
<td>.08</td>
</tr>
</tbody>
</table>

The independent variables entered into the regression explained 59% of variance in PedsQL total score. Burden was the strongest predictor with the PedsQL. 59% of the variance in the PedsQL total score can be explained by uncertainty, burden, Do you still have JDM? age and gender.

These data indicate that children and young people with JDM, with increased feelings of uncertainty about their disease, increased perception of burden and scoring they that still have JDM reported poorer total quality of life scores.

5.9.9.2 Pain domain in the Rheumatology Module

CUIS, burden and Q1. Do you still have JDM? were significant in the univariate analyses and were therefore entered into the regression along with age and gender. Table 5-20 presents the multivariate linear regression analysis of the Pain domain from the Rheumatology Module.
Only CUIS and burden remained significant independent predictors of pain-related quality of life in the multiple regression analysis. These independent variables and age and gender explain 51% of the total variance, with burden again remaining the strongest predictor.

### 5.9.9.3 Worry domain in the Rheumatology Module

Table 5-21 presents the regression analysis of the Worry domain from the Rheumatology Module. CUIS, burden, Q1 (Do you still have JDM?), Q5 (Have you met anyone else with JDM?), and length of time since last visit were significant in the univariate analyses and were therefore entered into the regression along with age and gender. Not all participants responded to Q5 or was data available for all when considering ‘length of time since last visit’ therefore this multivariate analysis was conducted with 116 respondent’s full data set (of note, this data set was representative of the 123, with those missing these questions being from a range of UK sites, ages, genders and disease clinical variables).
Table 5-21 Analysis of the Worry domain for all 116 respondents

<table>
<thead>
<tr>
<th></th>
<th>Multivariable Coefficient (Unstandardised B) (n=116)</th>
<th>P-value</th>
<th>(R^2) adjusted</th>
<th>Standardized Beta Coefficients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>-.93</td>
<td>.18</td>
<td>.48</td>
<td>-.13</td>
</tr>
<tr>
<td>Gender</td>
<td>5.43</td>
<td>.21</td>
<td></td>
<td>.05</td>
</tr>
<tr>
<td>CUIS</td>
<td>-.60</td>
<td>&lt;.001***</td>
<td>-.30</td>
<td></td>
</tr>
<tr>
<td>Burden</td>
<td>-1.05</td>
<td>&lt;.001***</td>
<td>-.34</td>
<td></td>
</tr>
<tr>
<td>Do you still have JDM?</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>-10.84</td>
<td>.99</td>
<td>.00</td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>10.91</td>
<td>.11</td>
<td>.11</td>
<td></td>
</tr>
<tr>
<td>Have you met anyone else?</td>
<td></td>
<td>.10</td>
<td></td>
<td>-.12</td>
</tr>
<tr>
<td>Length of time since last visit (years)</td>
<td>-6.68</td>
<td>.64</td>
<td></td>
<td>.04</td>
</tr>
</tbody>
</table>

Forty eight percent of the overall variance can be explained by these independent variables; uncertainty, burden, still having JDM, having met someone else, length of time since last visit, age and gender in this model. Both perception of burden and uncertainty were the only two variables that remained significant after multivariate analysis.

5.9.9.4 Communication domain in the Rheumatology Module

The variables significant in the univariate analyses were the CUIS, Burden and Do you still have JDM? and were therefore entered into the regression along with age and gender. Table 5-22 presents the regression analysis of the Communication domain from the Rheumatology Module.
Gender remained significant after multiple regression analysis, showing that females had more communication difficulties. Age, although not significant in the univariate analysis, interestingly came out of the multivariate analysis showing that younger age had more affected communication related quality of life issues. 53% of the variance can be explained by this combination of variables; uncertainty, burden, age, gender and Q1.

5.9.9.5 Treatment domain in the Rheumatology Module

The variables significant in the univariate analyses were; CUIS, Burden, Q1, years since diagnosis and length of time since last visit and were therefore entered into the regression along with age and gender.

Table 5-23 presents the regression analysis of the Treatment domain from the Rheumatology Module. There was data on 120 of the participants when collecting data on years since diagnosis and data on 117 children and young people for length of time since last visit, however when combined these made a data set of 116.
Table 5-23 Analysis of the Treatment domain for 116 respondents

<table>
<thead>
<tr>
<th></th>
<th>Multivariable Coefficient (Unstandardised B) (n = 116)</th>
<th>P-value</th>
<th>$(R^2)$ adjusted</th>
<th>Standardized Beta Coefficients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>1.5</td>
<td>&lt;0.01**</td>
<td>.47</td>
<td>.23</td>
</tr>
<tr>
<td>Gender</td>
<td>2.5</td>
<td>.45</td>
<td></td>
<td>.06</td>
</tr>
<tr>
<td>CUIS</td>
<td>-.31</td>
<td>&lt;0.05*</td>
<td></td>
<td>-.25</td>
</tr>
<tr>
<td>Burden</td>
<td>-.81</td>
<td>&lt;0.001***</td>
<td></td>
<td>-.37</td>
</tr>
<tr>
<td>Do you still have JDM?</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Y</td>
<td>-10.1</td>
<td>&lt;0.05*</td>
<td></td>
<td>-.24</td>
</tr>
<tr>
<td>No</td>
<td>1.43</td>
<td>.78</td>
<td></td>
<td>.03</td>
</tr>
<tr>
<td>Years since diagnosis</td>
<td>.42</td>
<td>.37</td>
<td></td>
<td>.08</td>
</tr>
<tr>
<td>Length of time since last visit</td>
<td>-1.5</td>
<td>.15</td>
<td></td>
<td>-.12</td>
</tr>
</tbody>
</table>

Uncertainty, burden, years since diagnosis, age, length of time since last visit and Q1 were all significant on univariate analysis. However, only uncertainty, burden, Q1 and age were significant at a multivariate level, with uncertainty and burden having similar standardised coefficient betas. Interestingly, age was significant at multivariate analysis for treatment, with a positive correlation, meaning that the older participants were more likely to report a better treatment quality of life. The variables explained 47% of the variance in treatment-related quality of life.

5.9.9.6 Activities of daily living (ADL) domain in the Rheumatology Module

Childhood Uncertainty in Illness Scale, Burden, Q1 and years since diagnosis were significant in the univariate analyses and were therefore entered into the regression along with age and gender.

Table 5-24 presents the multivariable analysis of the ADL domain from the Rheumatology Module. There was not a full data on years since diagnosis, with only 120 out of the 123 having data for this question.
Phase 2 “Picking up more passengers”

Table 5-24 Analysis of the ADL domain for 120 respondents

<table>
<thead>
<tr>
<th></th>
<th>Multivariable Coefficient (Unstandardised B) (n=120)</th>
<th>P-value</th>
<th>((R^2)) adjusted</th>
<th>Standardized Beta Coefficients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>.06</td>
<td>.89</td>
<td>.21</td>
<td>.01</td>
</tr>
<tr>
<td>Gender</td>
<td>.28</td>
<td>.91</td>
<td></td>
<td>.01</td>
</tr>
<tr>
<td>CUIS</td>
<td>-.08</td>
<td>.36</td>
<td></td>
<td>-.11</td>
</tr>
<tr>
<td>Burden</td>
<td>-.47</td>
<td>&lt;0.01**</td>
<td></td>
<td>-.34</td>
</tr>
<tr>
<td>Do you still have JDM?</td>
<td>-2.83</td>
<td>.26</td>
<td></td>
<td>-.11</td>
</tr>
<tr>
<td>Years since diagnosis</td>
<td>.53</td>
<td>.10</td>
<td></td>
<td>.16</td>
</tr>
</tbody>
</table>

Burden was the only variable that was still significant in the multiple regression when examining the ADL domain. 21% of the variance can be explained by the model above, which is the lowest percentage so far.

5.9.9.7 PI-ED score

Child Uncertainty in Illness Scale and Burden were significant in the univariate analyses and were therefore entered into the regression along with age and gender for the PI-ED. Table 5-25 shows the regression outcomes for the PI-ED measure.

Table 5-25 Analysis of the PI-ED of 123 respondents

<table>
<thead>
<tr>
<th></th>
<th>Multivariable Coefficient (Unstandardised B)</th>
<th>P-value</th>
<th>((R^2)) adjusted</th>
<th>Standardized Beta Coefficients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>.29</td>
<td>&lt;0.01**</td>
<td>.63</td>
<td>.13</td>
</tr>
<tr>
<td>Gender</td>
<td>.28</td>
<td>.10</td>
<td></td>
<td>.02</td>
</tr>
<tr>
<td>CUIS</td>
<td>.09</td>
<td>&lt;0.01**</td>
<td></td>
<td>.21</td>
</tr>
<tr>
<td>Burden</td>
<td>.49</td>
<td>&lt;0.001***</td>
<td></td>
<td>.62</td>
</tr>
</tbody>
</table>
These results show that Uncertainty, Burden and age were related to the PIED on univariate analysis. The analysis showed that older respondents report greater emotional distress. Multivariable analysis showed that Burden is the most important, with a standardised coefficient beta of 0.62 standard deviations in terms of explaining the variability in emotional distress scores between the patients. The variance is 63% explained by this model.

5.9.9.8 Brief summary of the results found from these validated measures

These analyses have indicated that many factors may contribute to the quality of life and levels of emotional distress experienced by children and young people with JDM. The key conclusions to be drawn from these data are:

- Perceived burden was the most statistically significantly predictor in all of the outcomes.
- Uncertainty is also an important factor when determining poorer quality of life or emotional distress.
- Age and gender had some impact, however, these variables were not consistently important after multiple regression.

5.9.10 Results from JDM experience questionnaire

As text boxes were not compulsory, there was some empty comments free-text boxes. Most young people completed all the questions, with only one young person repeatedly leaving questions blank. This individual added a reason, explaining that JDM was diagnosed when they were four years old, but now, as they are 18 and do not currently have any symptoms and cannot remember anything from this time, the questions are largely not applicable. This point (time since diagnosis) was noted as a factor in several replies.
5.9.10.1 Q1. Do you still have JDM?

All young people answered this question as presented in Table 5-26.

<table>
<thead>
<tr>
<th>Do you think you still have JDM? N (%)</th>
<th>Yes</th>
<th>No</th>
<th>Don’t know</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>71 (58)</td>
<td>28 (23)</td>
<td>24 (20)</td>
<td>123 (100)</td>
<td></td>
</tr>
</tbody>
</table>

There were 59 free text comments, Figure 5-5 presents the coding that was assigned through content analysis to each of these.

Over half (n= 71, 58%) of the young people still thought they had JDM and the majority used their current symptoms to illustrate this, for example, ‘I still get pain’. Some young people did seem confused about whether they had JDM or not, with the majority of these appearing in the ‘Don’t know’ response. For example, one young person said:
Phase 2 “Picking up more passengers”

“I’m at a stage when all the comments from my doctors are positive but they don’t say whether I have the illness anymore so that leaves me unsure and every now and then in myself I don’t feel 100% or I notice something, I don’t know whether that means I’m still affected by my JDM or if that’s just me over reacting and being paranoid.” Respondent number 22, female, 17 years of age

The comments are presented in detail in Appendix 21 with each comment and the coding group allocated. When young people used the term “In remission” that was coded as a theme, however, if they didn’t use this term, then it was coded as “No more” as being in remission is different to not currently having any symptoms. For example one young person, in answer to Q1 said:

“No, because I’m not feeling pain” Respondent number 114, male, 10 years of age

However, it is important to acknowledge that there may be overlap between these two codes. One young person said they have not had any problems for 10 years, so they are most likely in remission, but the clinical data linked to all of the relevant time points were not available to be able to confirm this or not for all participants.

5.9.10.2 Q2. If yes, how bad is it?

The next question asked young people if they had scored yes in question one, then how bad did they think their JDM was? The purpose of this question was to gain some context to some of their answers throughout the survey. 76 people responded to this question, all of those who had scored “Yes” (71) and five who had scored “Don’t know”. This was purely a multi-choice question, with no added open text fields. These responses are presented in Table 5-27.

<table>
<thead>
<tr>
<th>If yes, how bad do you think it is? N (%)</th>
<th>Not bad at all</th>
<th>Ok</th>
<th>Bad</th>
<th>Very bad</th>
<th>Total who responded</th>
<th>Number who did not respond</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>15 (20)</td>
<td>46 (61)</td>
<td>7 (9)</td>
<td>8 (11)</td>
<td>76 (62)</td>
<td>47 (38)</td>
</tr>
</tbody>
</table>
Phase 2 “Picking up more passengers”

Table 5.27 shows that 46 (61%) of young people who responded to this question thought their JDM was currently “Ok”, with 15 (20%) young people thinking their JDM is currently “Bad” or “Very bad”.

5.9.10.3 Q3. Can people see you have JDM by looking at you?

Children and young people were asked whether people can see if they have JDM. In hindsight, this question was not worded clearly enough and caused some confusion with the respondents. Table 5-28 summarises their responses.

<table>
<thead>
<tr>
<th>Can people see you have JDM by looking at you (%)</th>
<th>Yes</th>
<th>No</th>
<th>Don’t know</th>
<th>Total responders</th>
<th>Number who did not respond</th>
</tr>
</thead>
<tbody>
<tr>
<td>11 (9)</td>
<td>98 (80)</td>
<td>13 (11)</td>
<td>122 (99)</td>
<td>1 (1)</td>
<td></td>
</tr>
</tbody>
</table>

The answers in free text provided in response to this question were difficult to split into themes, as some had more than one point in their answers, but generally young people struggled to answer this question.

Figure 5-6 presents the results from coding from each of the 37 qualitative comments. The number of comments is shown in the brackets, and therefore, not all young people added a comment, for example, only 20 out of 98 (20%), added a reason why they indicated “no” for someone to be able to see their JDM.
Phase 2 “Picking up more passengers”

All but one of the young people who scored “yes” for this question added a comment. The majority mentioned their rash as the visible sign for JDM, with one talking about limping. Three young people talked about hiding their JDM; not being able to wear short sleeved tops, cutting hair short after it became thin due to treatment and using makeup to cover the facial rashes.

Lack of awareness from other people was mentioned in both respondents who said “No” people can not see they have JDM and in those that said “Don’t know.” Some young people thought that people might know they are poorly, but would not know what with:

“They can tell I’m ill but not that it’s JDM” Respondent number 68, male, 14 years of age

Whereas this young person who thought people couldn’t see it, implied that other people might not even know they are poorly:

“No one has approached me about it. When I tell someone about it they are very shocked!” Respondent number 121, female, 10 years of age

Some young people rightly questioned how could they know if others can see it or not, and this is a very good point. This is also perhaps more important with a
Phase 2 “Picking up more passengers”

condition like JDM which can fluctuate in outward appearance over the course of the disease.

5.9.10.4 Q4. Is it a good or a bad thing for people to see you have JDM or for them not to see it?

One hundred children and young people responded to this question marking it either positive or negative, however 14 extra respondents added a comment, without scoring it positively or negatively. This may mirror the ambiguity in the question. The responses are presented in Table 5-29.

<table>
<thead>
<tr>
<th>Good</th>
<th>Bad</th>
<th>Total responders</th>
<th>Number who did not respond</th>
</tr>
</thead>
<tbody>
<tr>
<td>Is it a good or bad thing for people to see you have JDM or for them not to see it? N (%)</td>
<td>53 (43)</td>
<td>47 (58)</td>
<td>100 (81)</td>
</tr>
</tbody>
</table>

The question was meant to mirror their first answer, so if they thought people could not see their JDM, was this good or bad for them, however, not all respondents read it like this. For instance, the first young person had answered that people could not see their JDM in question three. Subsequently, for this question they said it was a bad thing for people not to see it. In their comments, they wrote:

“Because I don’t usually show any signs if something is apparent that my condition is worsening” Respondent number 2, female, 18 years of age

This answer implies that if people can see their JDM, then this is a bad sign as it means their disease is active, therefore using whether people can see their JDM as a marker of disease activity. This was also supported by three other respondents. Another example, is that of a young person who responded again that people cannot see their JDM, but then commented that this was ‘bad’ as they don’t want lots of people to know as it is private to them. I think this question had been interpreted by many, as “Is it a good or bad thing for people to see JDM?”,
Phase 2 “Picking up more passengers”

rather than “Is it a good or bad thing for people to see you have JDM or for them not to see it?” in relation to the answer they had previously put. This is different to initially anticipated and makes interpreting their answers in any depth, questionable. There were also many more comments to this question, with 41 comments out of the 53 respondents commenting that it was a good thing, and 36 saying it was bad, and all adding a reason why. Figure 5-7 shows the coding attached to the responses received.

Some young people thought it was bad if people could not see it as they would expect more from them:

“Bad if they can’t see it because they assume I am fully healthy and expect me to perform at the level of an able bodied person” Respondent number 25, female, 18 years of age

Whereas others thought it was bad if they can see it because they don’t want people to know:
Phase 2 “Picking up more passengers”

“I don’t like people seeing me and think she’s ill or has JDM and I don’t want people to question if there’s something wrong with me.” Respondent number 72, female, 15 years of age

This can be summed up by one of the respondents who did not score the question as good or bad, but added a comment:

“It’s not good, nor bad – it’s good that it’s invisible sometimes so I can blend in without the disabled stereotype. However, sometimes it needs to be seen so I can be understood and not challenged”. Respondent number 44, female, 17 years of age

5.9.10.5 Q5. Have you met anyone else with JDM?

All but one individual scored the next question, which asked if they had met anyone else with JDM. There was no free text box provided here and no option for do not know, as it was hoped the respondents would know if they had or not. Table 5-30 presents this data.

<table>
<thead>
<tr>
<th>Have you met anyone else with JDM? N (%)</th>
<th>Yes</th>
<th>No</th>
<th>Total responders</th>
<th>Number who did not respond</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>60 (49)</td>
<td>62 (50)</td>
<td>122 (99)</td>
<td>1 (1)</td>
</tr>
</tbody>
</table>

5.9.10.6 Q6. Would you like to meet someone else with JDM?

122 out of a possible 123 respondents answered the next question, asking them if they would like to meet someone else with JDM? These data are presented in Table 5-31.

<table>
<thead>
<tr>
<th>Would you like to meet someone else with JDM? (%)</th>
<th>Yes</th>
<th>No</th>
<th>Don’t know</th>
<th>Total responders</th>
<th>Number who did not respond</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>73 (59)</td>
<td>16 (13)</td>
<td>33 (27)</td>
<td>122 (99)</td>
<td>1 (1)</td>
</tr>
</tbody>
</table>
Phase 2 “Picking up more passengers”

The results showed that 59% of children and young people would like to meet someone else with JDM, with only 13% saying they would not. As the previous question had resulted in a clear split of those that had met someone with JDM before and those that hadn’t, I thought it would be interesting to further separate the results by this. It was expected that if children and young people had met someone previously, then they might say they did not want to meet anybody else, this was not the case as shown by Table 5-32 which shows the data from both of these questions.

<table>
<thead>
<tr>
<th>Table 5-32 Results from Q5 and Q6 combined</th>
</tr>
</thead>
<tbody>
<tr>
<td>Have you met anyone else with JDM? (%)</td>
</tr>
<tr>
<td>Yes</td>
</tr>
<tr>
<td>60 (49)</td>
</tr>
<tr>
<td>No</td>
</tr>
<tr>
<td>62 (50)</td>
</tr>
<tr>
<td>Number who did not respond</td>
</tr>
<tr>
<td>1 (1)</td>
</tr>
<tr>
<td>Would you like to meet someone else with JDM? (%)</td>
</tr>
<tr>
<td>Yes</td>
</tr>
<tr>
<td>36 (29)</td>
</tr>
<tr>
<td>No</td>
</tr>
<tr>
<td>7 (6)</td>
</tr>
<tr>
<td>Don’t know</td>
</tr>
<tr>
<td>17 (14)</td>
</tr>
<tr>
<td>Yes</td>
</tr>
<tr>
<td>37 (30)</td>
</tr>
<tr>
<td>No</td>
</tr>
<tr>
<td>9 (7)</td>
</tr>
<tr>
<td>Don’t know</td>
</tr>
<tr>
<td>16 (13)</td>
</tr>
<tr>
<td>Number who did not respond</td>
</tr>
<tr>
<td>1 (1)</td>
</tr>
</tbody>
</table>

This data illustrates that children and young people would still like to meet someone else with JDM, regardless of whether they have met anybody previously. There were 41 comments received for this question, with most respondents detailing peer support as the reason for meeting someone else, with qualitative comments shown in Figure 5-8.
Some young people clearly articulated the benefits they get from meeting others with JDM, as described by this young person:

“Meeting other people with JDM is so amazing and important because you both just get each other and know what it’s like. I would love to meet someone my own age with it at some point, because in hospital it is usually younger kids who have it. The fact that I have even been able to meet other people is incredible, though, as JDM is so rare.” Respondent number 33, female, 13 years of age

One young person who said they had not met anybody else, was the only one who described an altruistic reason for meeting someone else:

“So I can show my improvement and reassure them everything will be fine”

Respondent number 61, female, 13 years of age

Within the comments received to this question, there were personal comments about how JDM had made the young people feel, as illustrated here:
Phase 2 “Picking up more passengers”

“It would be nice to have a community of teenagers living with a similar condition, and we can help each other. I never knew anyone when I was growing up and going through puberty etc. so at times, I felt very alone and isolated, and like no one would understand.” Respondent number 21, female, 17 years of age

If the children and young people had been able to see each other’s comments for this question, or share these feelings, perhaps they would not feel so alone and isolated.

5.9.10.7 Q7. Do you think you get enough help and support with your JDM?

Question seven had a free text box only, rather than prescribing answers. Five individuals did not add any comments here. The comments were coded into those that were positive and generally said yes they do, or yes they did at the time when they needed it and those that were negative, don’t know or who wrote non applicable. The coding allocated is presented in Table 5-33.

<table>
<thead>
<tr>
<th>Do you think you get enough help and support with your JDM? N (%)</th>
<th>Positive responses</th>
<th>Negative responses</th>
<th>Don’t know</th>
<th>Not applicable</th>
<th>Total responders</th>
</tr>
</thead>
<tbody>
<tr>
<td>100 (81)</td>
<td>13 (11)</td>
<td>2 (2)</td>
<td>2 (2)</td>
<td>117 (95)</td>
<td></td>
</tr>
</tbody>
</table>

The children and young people in their answers that were coded positively, talked mostly about the support they have received from their parents or hospital staff, for example:

“Yes, because the doctors are always very helpful and can explain what is wrong if I feel any pain and my family and friends are there to look out for me.” Respondent number 32, female, 14 years of age

Some young people were intuitive in looking back, thinking about how difficult it was, and now they feel they did have support, but maybe it didn’t feel like it at the time:
Phase 2 “Picking up more passengers”

“I did, though occasionally at the time I thought I didn't.” Respondent number 6, male, 18 years of age

And some added comments about who was supportive, but also, who wasn’t:

“At home, yes. Everywhere else, not so much, there is next to no awareness supporting my condition and every case is different.” Respondent number 44, female, 17 years of age

Young people wrote less to explain why they felt they are not supported, with six, just saying “No” or “Not really”. One young person added that:

“No really - no one talks to me about it - or only every 6 months at appointments.”
Respondent number 53, male, 13 years of age

Generally though, in response to this question, the responses were more positive than negative and showed that most people feel they do get enough support.

5.9.10.8 Q8. What is the worst thing about having JDM?

This question offered an open text box for their answers. 113 young people all offered a comment here, with one commenting ‘Not applicable’. Some young people would list many points in their response, rather than just one, for example:

“The pain, going to the hospital, missing school and my friends, the treatment, it make my head hurt”. Respondent number 82, Female, 9 years of age

After consideration and discussion with the study team it was decided to code all of these responses, rather than just selecting their first point. This could be because that first point, might not be the most important to them, or the young people may feel there are comparable worst things about JDM. In recognition of their time and effort in responding with their worst thing, all were coded, by content analysis, identifying the primary, secondary, tertiary and quaternary themes; data are shown in Appendix 22. Seventeen individuals had up to three themes from their answer and two individuals had four themes assigned. There were 216 codes allocated in total to 21 different themes, the themes are shown ranked in order in Table 5-34 with an example of one comment for each theme.
Phase 2 “Picking up more passengers”

identified. Coding all comments systematically and presenting an example for each, is a characteristic of good, robust data analysis for rigour and transparency.

<table>
<thead>
<tr>
<th>Number of responders who were coded into this theme in total</th>
<th>Themes allocated</th>
<th>An example of one comment for each theme identified</th>
</tr>
</thead>
<tbody>
<tr>
<td>34</td>
<td>Limitations due to JDM</td>
<td>“Not being able to do things that everyone else can do!”</td>
</tr>
<tr>
<td>27</td>
<td>Treatment</td>
<td>“Having weekly injections”</td>
</tr>
<tr>
<td>23</td>
<td>Mental health effects</td>
<td>“It was feeling useless and not being able to do things”</td>
</tr>
<tr>
<td>20</td>
<td>Being different</td>
<td>“Being so obviously different but no one can see it”</td>
</tr>
<tr>
<td>18</td>
<td>Uncertainty</td>
<td>“You don’t really know what is going to happen”</td>
</tr>
<tr>
<td>18</td>
<td>Pain</td>
<td>“The aches in my legs and aches I get really bad and end up in hospital”</td>
</tr>
<tr>
<td>12</td>
<td>Body image</td>
<td>“I become fat!”</td>
</tr>
<tr>
<td>10</td>
<td>Tired</td>
<td>“Getting tired quicker than usual. I love to do things that require a lot of energy and I feel left out if I can’t do anything I like or I used to do”</td>
</tr>
<tr>
<td>6</td>
<td>Lack of understanding</td>
<td>“The worse thing is that there’s not much knowledge about the illness. Not everyone understands what it’s like and how it effects people”</td>
</tr>
<tr>
<td>6</td>
<td>Hospital visits</td>
<td>“All the hospital and doctor appointments”</td>
</tr>
<tr>
<td>6</td>
<td>Lack of stamina</td>
<td>“Not being able to have a high endurance in physical exercise”</td>
</tr>
<tr>
<td>6</td>
<td>Rashes</td>
<td>“Having rashes at certain points that are visible so people think you have something contagious or a serious skin condition and judge you”</td>
</tr>
<tr>
<td>5</td>
<td>Sun protection</td>
<td>“Having to wear a hat in summer because I’m always scared someone will make fun of me”</td>
</tr>
<tr>
<td>4</td>
<td>Methotrexate sickness</td>
<td>“Methotrexate is the worst thing, because I hate the way it burns and it makes me feel sick”</td>
</tr>
</tbody>
</table>
Phase 2 “Picking up more passengers”

<table>
<thead>
<tr>
<th>Number of responders who were coded into this theme in total</th>
<th>Themes allocated</th>
<th>An example of one comment for each theme identified</th>
</tr>
</thead>
<tbody>
<tr>
<td>3</td>
<td>Invisibility</td>
<td>“Because its internal people often don’t believe me, because there’s nothing physically wrong and it puts you down and think bad about yourself, especially when you don’t know why you have it like a cause”</td>
</tr>
<tr>
<td>3</td>
<td>Missing school</td>
<td>“Missing school which had affected my grades”</td>
</tr>
</tbody>
</table>

Table 5 34 Primary code allocated for each of the comments given for Q8

<table>
<thead>
<tr>
<th>Number of responders who were coded into this theme in total</th>
<th>Themes allocated</th>
<th>An example of one comment for each theme identified</th>
</tr>
</thead>
<tbody>
<tr>
<td>3</td>
<td>Blood tests</td>
<td>“The blood tests”</td>
</tr>
<tr>
<td>3</td>
<td>Immunosuppression</td>
<td>“Have to be careful around ill people”</td>
</tr>
<tr>
<td>2</td>
<td>Weakness</td>
<td>“Weakness in muscles”</td>
</tr>
<tr>
<td>2</td>
<td>Physiotherapy</td>
<td>“Aching painful physio”</td>
</tr>
<tr>
<td>2</td>
<td>Don’t know</td>
<td>“I’m not too sure”</td>
</tr>
<tr>
<td>1</td>
<td>Calcinosis</td>
<td>“Calcinosis”</td>
</tr>
<tr>
<td>1</td>
<td>Nothing</td>
<td>“Nothing really”</td>
</tr>
</tbody>
</table>

Limitations due to JDM was identified by children and young people with JDM as the worst thing, and this is represented in the word cloud shown in Figure 5-9:
A word cloud was chosen to depict this data through visualisation of the written responses, as this emerging method has been shown to be advantageous when wanting to increase comprehension and accessibility in qualitative research data (Bletzer, 2015; Sellars, Sherrod and Chappel-Alken, 2018). The word cloud was developed from freely available computer software which digitally examines word frequencies by identification of tags and focuses on a simple visualisation pattern that depicts the words that appear most often in larger font or text size within the cloud (Cidell, 2010). The final graphic portrays patterns of keywords included in the text, which allows the viewers to identify relationships and meaning to provide rapid analysis and clear, visual representation (Sellars, Sherrod and Chappel-Alken, 2018).

Finally, one of the aims of this overall phase was to establish if the findings from phase one resonated with a larger population. There were two young people that added a comment for Q8, which clearly demonstrate some resonance with ‘being different’, ‘being uncertain’, ‘medication issues’, ‘being-confused’ and even hint of the need for ‘acceptance’, all five themes from phase one in one comment, these two are shown here:
Phase 2 “Picking up more passengers”

“In short words, I would probably say that the worst thing is not being able to be as normal as other teenagers. Everyday, I am reminded of JDM, and even when I am home, I still don’t have 100% freedom as I know that I’ll be in hospital the next month with doctors making decisions about me. This then brings in the not knowing and lack of control as to what will happen next, which is really difficult with physio and treatment, probably because I am getting older and feel well. This then also brings JDM into being a mind game – one minute you’re fine the next minute you’re not, or you might currently be fully fine (its all so confusing).” Respondent number 33, female, 13 years of age

“The worst thing about living with JDM is not the pain or the numerous hospital visits, but the not knowing. It is the fear of the unknown. There is not enough knowledge about JDM to know whether you will ever be cured, or whether you will get worse, or whether this treatment will actually work for you. There is no clear-cut pathway to recovery to motivate you to take your medication and to keep doing the physio exercises even though they’re painful. It might all be for nothing. I just want to know what it feels like to go to sleep and then wake up in the morning and actually feel rested. I want to know what it feels like to not have a pain lingering on your bones like a constant background noise. When you get ill from a young age, you live your life constantly grieving the loss of a life you never got the chance to live. Which sounds ridiculous, because how can you grieve something you never experienced? But it is in the times between the bad days, or the fleeting moments of remission, when your mind begins to wonder whether your life could have been so much different had you been dealt a different hand. Ultimately, I think that when you live with a chronic illness, your body is never truly yours. Your body is not your own because it is always being controlled by something else: the disease, the medication, the doctors at the hospital. You feel trapped within this vessel you inhabit”. Respondent number 21, female, 17 years of age

5.9.10.9 Q9. Anything else you want to tell us?

The last question asked whether the children and young people wanted to add any other comments. 33 comments were received, and these were coded depending on the nature of the comment (See Appendix 23).

It was a lovely end to the survey to have so many positive comments. Two individuals had drawn smiley faces and many said thank you.
5.10 Discussion

The discussion will revisit the hypotheses to consider whether the findings from the results are able to accept or reject each in turn. Following on from this, returning to the model proposed at the beginning of this chapter will add structure to the discussion, examining published literature and discussing the implications for clinical practice.

5.10.1 The hypotheses

Table 5-35 presents each hypothesis, whether it has been accepted or rejected, and the evidence for this.
In summary therefore, the findings were not as expected. An increased level of uncertainty (hypothesis two) was the only significant finding from the proposed hypotheses.

Hypothesis one could only be partially accepted, as whilst reporting lower quality of life than a healthy population, our study population also scored significantly lower in emotional distress than a normative sample of healthy school children.
This was a surprising finding. When considering why this may be so, with my supervisory team we questioned the potential positive impact of post traumatic growth and resilience in children and young people who had suffered for many years with a chronic disease diagnosis and recommend that future studies should examine this in more detail: the nature of post-traumatic growth has not been a focus of study in JDM. However, children and young people who triggered above the cut off for the PI-ED, had significantly lower PedsQL and Rheumatology Module scores and higher uncertainty and burden scores than those who did not score. Forty percent of respondents still did score on this measure and did need further local clinical assessment. Of note, the older age of participant was more likely to score on the PI-ED, but less likely on the PedsQL and Rheumatology Module, which is an interesting finding. It is challenging to explain why this might be so: it is possible that the generic questions of the PI-ED (for example ‘I get annoyed easily’) may be more applicable to older teenagers, rather than the health related aspects of the PedsQL and Rheumatology Module (for example ‘I hurt a lot’). This hypothesis would need to be tested in a larger data set to investigate if this effect is robust.

Two hypotheses were rejected. Firstly, increased perceived acceptance/benefit (hypothesis three), did not correlate with any of the outcomes. Secondly, those that thought they had active JDM would have a lower quality of life and higher emotional distress (hypothesis five). However, the qualitative comments proved that children and young people find it hard to determine if they still have JDM with 20% of respondents saying they did not know if they still have it. There was a lot of confusion and uncertainty for the young people, with 80% of the respondents commenting that you could not see their JDM. Having an invisible disease might make it harder to know if it is there, especially when symptoms are quiescent. Hypothesis four could not be accepted or rejected from the data collected in this study and warrants further investigation, this is discussed further in the limitations section 5.15.
5.11 The Juvenile Dermatomyositis Model

The model developed at the beginning of this phase Figure 5-2 postulated that increased ‘uncertainty’ and a lower level of perceived ‘acceptance/benefit’ would have a relationship with a lower quality of life and greater emotional distress.

![Uncertainty vs. Quality of Life and Emotional Distress](image)

Figure 5-2 Juvenile Dermatomyositis Psychosocial Model

Through considering the hypotheses, the data suggest that uncertainty does have a role to play with the outcomes: quality of life and emotional distress. From these results, acceptance and benefit, do not. Therefore, in retrospect the proposed model is an imperfect fit for the study data found, and could have been improved by creating a simpler model by removing the acceptance/benefit arm. The role of acceptance and benefit, will be considered in turn with the available literature.

5.11.1 The role of uncertainty

The results of this phase have proved that there is a correlation between increased uncertainty and lower quality of life, (across four of the five domains of the Rheumatology Module, all except the ADL domain) and greater emotional distress for the children and young people with JDM in this research.

Uncertainty was identified as a theme in phase one and has continued to be a significant finding in phase two. Uncertainty is not a new finding in paediatric rheumatology, in the study by Tong et al., (2012) they coined the phrase ‘suspension in uncertainty’ to explain that experienced by children and young people with JIA. For example, a young person in this study commented that they get disappointed as one minute they are better, and then they are worse again, mirroring the uncertainty that came out of this study. (Tong et al., 2012).
This statement was taken from a paper examining children’s perceived illness uncertainty as a moderator in the parent-child distress relation in JRD, including a small number (n=7) with JDM. In the study by White et al., (2005) whilst they found that uncertainty was not related to child depressive symptoms, they found children’s uncertainty regarding the severity, course, and outcome of JRD may serve as a cognitive vulnerability that provides the opportunity for parent distress to have a greater negative impact on their emotional adjustment. The authors concluded that the unpredictable nature of rheumatic diseases highlights the role of uncertainty in child and parent adjustment to chronic ill health conditions (White et al., 2005).

The current study data, interestingly had lower mean levels of perceived uncertainty than the small US sample of children with JRD in White et al., (2005) study. In the White et al., (2005) the mean CUIS score was 67.02, whereas the current study found a mean of 52.67. The scoring range for the tool is between 23-115 with higher scores indicating greater distress, with this paper reporting a range of 32-107, whereas this UK JDM cohort had a range from 24-88. The US participants had shorter disease duration (0.04-15.73 years) compared to our population who had a longer disease duration (from 0.3-17.3 years) implying that in the study of 50 US participants, they had been more recently diagnosed and this could explain the difference between the two samples.

5.11.2 The role of an increased perception of acceptance/benefit

This research found that ‘acceptance’ was a theme in phase one and therefore explored this further in phase two, through the benefit aspects of the BBSC questionnaire.

The young people interviewed in phase one, could see some benefits of having JDM and had a positive outlook to their quality of life - “I’m special”. Interestingly therefore that actually the benefit results were not significant at all. These results led to revisiting the BBSC questionnaire and questioning whether it indeed
Phase 2 “Picking up more passengers”

addressed ‘acceptance’ adequately from the results from phase one. Whilst the questionnaire produces an overall score, I was interested to look in more detail at specific questions. Out of the ten benefit questions, the top scoring one was “My JDM has helped me know how much I am loved” (with a score of 489 out of 615). This had also been a specific comment from the interview data in phase one. The lowest scoring question was “My JDM has helped me make some new friends” (with a score of 318 out of 615). This was not mentioned by any children or young people in their interviews, in fact the opposite was felt to be true. Therefore, maybe the questionnaire did not fully address the theme of acceptance, but in some parts, was a good fit. Whilst, not adding to the statistical analysis however, it is the trait of a good researcher to revisit research methods and constantly reassess with the overall aims of the research.

5.12 Resonance and new findings

Phase two set out to look for resonance with the findings from phase one, and to look for new findings from a larger more diverse population. This section, presents discussion on both of these aims.

5.12.1 Looking for resonance

One of the aims of this overall phase was to establish if the findings from phase one resonated with a larger population. Whilst it is difficult to quantify amongst so many detailed responses; feeling different, uncertain, confused and struggling with the medication were often commented on by the young people. In fact, in Q7, where the young people all commented what they felt was the worst thing about JDM, for just the primary coding, difficulties with medicines were mentioned 18 times, uncertainty 15 times and being different seven times. One can therefore agree that there is some resonance with the findings from phase one in a larger JDM population.
Phase 2 “Picking up more passengers”

5.12.2 Looking for new findings

One of the advantages of the mixed methods design was that new data can be added in to build upon subsequent findings. Whilst analysing the data from phase two, there were some issues which stood out, specifically: schooling, peer support and perception of burden. These will each be addressed in turn.

5.12.2.1 Schooling

There were only five questions about schooling within the PedsQL, including asking about missing schooling due to being unwell, however issues around schooling featured quite heavily generally throughout the results. The JDM experience questionnaire did not ask specifically about schooling, but there were many qualitative comments regarding it. These comments often referred to young people talking about their grades dropping, or not being able to access school. Some young people added negative comments about lack of school support, including lack of understanding from teachers.

5.12.2.2 Peer-support

When asking if children and young people had met anyone else with JDM, our results were split almost exactly with those that had and those that hadn’t. There were some positive comments discussed from meeting others with JDM, and 60% of our survey said they would like to meet someone else. 27% said they did not know whether they would like to meet someone else with JDM, and the reasons given for this included; being shy, having met a lot already, scared to meet new people and that it might cause bad memories. What was interesting from these findings was that half of those who had met someone else with JDM already, still wanted to meet someone else, and only 6% did not. However, those who had met someone else with JDM had significant scores for treatment and worry on the Rheumatology Module. These are interesting findings which could be explained by these children and young people having more active disease, illustrated by the need for treatment which increases their worry. These results warrant further investigation. Peer support is discussed in further detail in Chapter seven.
Phase 2 “Picking up more passengers”

5.12.2.3 Burden

The burden measure was found to be a significant finding across all statistical tests examined, more so than any of the other measures. These findings are in line with those found in the literature. For example disease related burden has been shown to be strongly associated with almost all psychological outcomes in paediatric cancer patients (Currier, Hermes and Phipps, 2009; Maurice-Stam et al., 2011). Unfortunately, there is no data on burden in paediatric rheumatology to compare our findings to.

The term ‘burden’ did not specifically come out of phase one, but has clear overlap with the other themes found. For example, when young people talked about ‘feeling different to their peers and holding them up when they went out’, this could easily have been classified as ‘burden’. This overlap has made this study challenging, but also worthwhile, especially to see how comprehensively the burden tool captured the feelings from children and young people around the UK. The burden tool was highly statistically significant on every univariate analysis and all but the activities of daily living domain in the multivariate analysis. Of note, the ADL Domain had the lowest $R^2$ with only 21% being explained by regression analysis. This is the smallest amount of explained variance found in the analyses so far, which implies that there are more issues affecting activities of daily living, but that this study has not yet ascertained what these are. Also of note, there was less variability in ADL than in other domains, the mean (SD) was 93.2 (13.2) and ranged from 40 to 100. When considering the questions it can be seen that four out of five of the questions are specifically referring to fine motor skills, such as holding a pencil, using a knife and fork, turning on taps and opening door handles, whereas only one was more generic by asking about difficulty carrying school books. Maybe as JDM primarily affects the proximal muscles, these questions would be less relevant than they would be for other rheumatological conditions, such as JIA for example, where the smaller joints can be inflamed.

As emotional distress could be seen as an aspect of quality of life, one could argue that the PI-ED should have been included as a variable rather than an outcome. However, because the PI-ED and the Burden measure were highly
correlated at 0.78 (above the level for multicollinearity) it made sense to treat PI-ED as a separate outcome and not as an independent variable with burden in the regressions to examine quality of life.

In the website text discussed in the scoping review, the young adult specifically mentioned feeling a burden to others. Admittedly a burden questionnaire was specifically utilised for this phase, but a raised level of burden did correlate with a lower quality of life and higher emotional distress in some children and young people with JDM. The implications from this study are that if young people feel a greater perception of burden, then this needs to be addressed in clinical care before it has an implication on quality of life and emotional distress.

5.13 Implications for practice

This results from phase two have highlighted how important a feeling of uncertainty and a perceived perception of burden are to children and young people with JDM. Also, the importance of school support and peer support, should not be underestimated. Taking the time to address these issues in clinical care may improve young people’s quality of life and emotional distress. Children and young people talked openly about their thoughts and feelings, with 60% of children and young people wanting to meet someone else with JDM. In the clinical setting, offering young people the opportunity to talk to others, may lessen the implications from the limitations they feel.

5.14 Strengths

This is the first ever questionnaire study asking a large number of children and young people with JDM about their quality of life and emotional well-being. The practical advantages of collecting data electronically meant that parents could send the link to children and young people at boarding schools and universities and raised PI-ED scores returned could be scored and dealt with in a timely fashion. Over 50% of the children and young people replied and this was much higher than anticipated, especially for a postal survey to children and young people all around the UK. There was minimal missing data, which really demonstrates the focus given to this from children and young people. Maybe this
Phase 2 “Picking up more passengers”

is due to the research focused ethos of children, young people and families with JDM, who firstly, are often enrolled in the JDCBS with the aim of this registry (to undertake research) being discussed from the outset of diagnosis and secondly from those who have a rare disease with no cure. The high completion rate could also be explained by the promise of entering the prize draw for a voucher - only to those that were 100% completed of the validated questionnaires! Furthermore, there was support from all health care professionals around the UK who took the time to engage with the study and supported it by sending out questionnaire packs, fielding questions and contacting young people with raised emotional distress scores.

5.15 Limitations

Comparison of the data generated from the validated questionnaires was sometimes challenging due to a relative paucity of published comparable data from contemporaneous and geographically matched group of patients. Studies used for comparison were frequently small and/or had been performed in a different time or country so may not be directly comparable.

Unfortunately, due to the incomplete data in the JDCBS it was difficult to compare questionnaire findings with detailed clinical data, for example medication dosage over years, physical disability or current disease activity. Whilst last clinical data could be accessed, 49% of participants had not been seen in the last year. In fact, the range of patients last being seen was zero months – 10.5 years, with a median of 0.957 years and an Inter Quartile Range of 0.586 year to 2.08 years with a long tail out to 10 years. Therefore, it is with caution that we highlight those who had a rash or weakness when they were last seen. However, whilst it was only 41 children or young people who scored as having rash and/or weakness at their last visit, they did score significant findings on the majority of questionnaires. Whilst this data may date some months or even years prior to questionnaire completion, it could be used as a surrogate marker for severity of active disease. Rash and/or weakness are signs of active disease in JDM and could be used by health care professionals to help determine who may need more support. Furthermore, the fact that we found children who had a rash at last visit scored significantly worse on the questionnaire measures, and that those who thought
Phase 2 “Picking up more passengers”

you could see their JDM scored worse for uncertainty and perception of burden, are two factors which could be related and we recommend future studies should examine this in more detail. Even despite the omission of recent clinical data, the questionnaire results themselves are valuable as they are as declared by the child or young person. Declaring emotional distress needs addressing whatever the background circumstances.

As the surveys were sent out in the post, it is impossible to know who had actually completed them. Sometimes a parent would say they had scribed for their child, especially if they were at the lower age of the target range, but it is unknown how often this occurred. It was also not easy to know how much guidance the child or young person might have had in completing the questionnaire, although the majority were obviously completed from the child’s perspective, supported by age appropriate comments (and original spellings).

Time and scope have also been topics of discussion as this phase has progressed. This is a cross sectional study, capturing how young people felt on that day. A longitudinal study would have enhanced the study design, but was not feasible in the time.

The JDM experience questionnaire also had a number of significant limitations. Some of the questions were not clear, some could be interpreted in a number of different ways and some questions were not applicable to everyone. The question asking young people whether it is a good or bad thing for people to see their JDM is a good example of this. This question had specifically been added as children and young people in phase one had talked about the invisibility/visibility of JDM and the implications of this, however, due to the wording of this question it was decided to not take this further through the analysis. The experience questionnaire was not piloted, primarily as all children and young people with JDM were to be included in the study sample and so there was not a clear group who would not be included later to pilot it. On reflection, this was an incorrect decision as had the questionnaire been piloted, some of the issues around construction and language used, would have been resolved.

There is a wealth of literature about what style questions are best to ask, in what order the questions should be presented in and how to visually present surveys
to gain the maximum response rate. Dillman, Smyth and Christian, (2009) recommended that questions are posed in simple and familiar terminology, therefore the wording in this survey was kept very simple, however, now it can be seen that the terminology ‘good’ or ‘bad’ has different connotations to different individuals and thus is accompanied by a large amount of subjective bias, this may have been identified if children and their families had reviewed the questionnaire in a pilot.

5.16 Conclusion

This chapter opened with a quote from families highlighting the importance of assessing quality of life in children and young people with JDM. This study has agreed with this. Throughout phase two we have found a higher level of uncertainty and increased perceived feeling of being a burden was related to a lower expressed quality of life and a higher emotional distress score in this sample of children and young people with JDM.

5.17 Next chapter

The next chapter presents phase three of the research which invites health care professionals to add their contribution about the overall psychosocial needs of children and young people with JDM. The aim was to collect local information on the level of support they have, the challenges they face and any good practice, which could be shared with others. This adds a fuller picture when considering the needs of all children and young people with JDM around the UK.
“Football”

JDM makes your life a bit more exciting,

But you mustn’t give up, you need to keep fighting,

Although it’s a disease, it kind of gives you individuality,

It’s definitely changed my personality!

Having JDM you mature quite quick,

You realise you’re in a hospital, trying not to be sick,

I’m glad I have it in a way,

Makes me feel different to other people every day,

But at the same time, I wish I was the same as my friends,

Being able to play football at the weekends,

I hope my JDM will go away, but I can’t be sure,

If it doesn’t, please can someone find a cure?
Chapter 6: Phase 3 “The view from others on the journey”

“We do see a lot of anxiety. We do see a lot of depression, but I don’t feel like we have the adequate time to really address that” (Knight et al. 2019 p 597).

6.1 Introduction

The quote that introduces this chapter, also sets the scene. It was recently voiced by a social worker in the US who took part in a mixed methods study to identify behavioural health care provider’s perspectives on gaps in mental health for children and young people with rheumatic conditions. Online surveys were returned from 42 social workers and psychologists and semi-structured interviews conducted with 20 of these behavioural care providers. The combined results illustrate a need for interventions addressing depression, anxiety, adjustment/coping, distress, parent/caregiver mental health, as well as peer support services for patients and families (Knight et al., 2019). This research was published after the scoping review had been undertaken, however it is included here, as it provides interesting parallels with this study. One key conclusion from their work is that mental health optimisation requires better utilisation of existing resources (Knight et al., 2019).

In the UK however, we have limited knowledge of what these existing resources are, and this is exactly what this phase set out to establish. This chapter will present phase three of the study: the scoping of current psychosocial provision for children and young people with JDM across the UK being provided by the (then) fifteen Paediatric and Adolescent Rheumatology centres in the JDCBS. The chapter begins with an introduction to the setting, an overview of methods, data analysis and results. Deciding how to present the results was complicated as most questions asked are different for each health professional group asked, but there is also some overlap. Initially, data were reported from each centre, but this could have led to centre identification, so it was then shown by health professional groups, but this made comparisons awkward. Consequently, the results are displayed as a combination of grouping the questions and then presenting in health professional group, with quantitative data and qualitative
data in parallel. The limitations and implications of this work are discussed and ideas for the future presented.

### 6.2 Context

The phase three study was initiated due to the need to establish current psychosocial support provision for children with JDM across the UK and with an aim to determine areas of good practice which could be shared with other centres.

Psychological services for children and young people with chronic health conditions can help at every step of the journey – at diagnosis, during treatment and beyond (Mercer et al., 2015). National standards are now in place acknowledging that psychological services are not simply an “adjunct” to regular medical care but represent an integral component of a healthcare team for children and young people and their families experiencing chronic health conditions. For example, in Facing the Future: Standards for Children with Ongoing Health Needs (Royal College of Paediatrics and Child Health, 2018), standard 8 states that the commissioning and planning of paediatric services needs to:

> “Ensure children have timely access to a range of mental health and psychosocial services that are integrated with children’s health services”. (Royal College of Paediatrics and Child Health, 2018 p. 39)

There is an ever-increasing evidence base to support the clinical effectiveness of psychological interventions for a number of similar, chronic, long-term medical conditions and illnesses in which interventions and early support may result in:

- Better medical outcome (e.g. by increasing levels of adherence)
- Better psychological functioning (anxiety, low mood, distress, anger)
- Better family functioning
- Reduced levels of disability and pain
- Reduced levels of distress around procedures
Phase 3 “The view from others on the journey”

(Pai et al., 2006; Spirito and Kazak, 2006; Edwards and Titman, 2011; Gómez-Ramírez et al., 2016)

An understanding of psychosocial support available for young people with JDM at their major care centres is therefore crucial to establish whether healthcare providers are meeting the needs of young people, and if not, what recommendations can we propose to improve this.

As ‘psychosocial support’ encompass such a large element of psychology (covering mental health, and emotions such as anxiety, stress, worry), this phase was particularly interested in availability of clinical psychology provision in each centre. Whilst psychosocial support, such as coping with a chronic condition and medication adherence advice is provided by all members of a multidisciplinary health care team, involving all members in each centre would not have been possible. Therefore, the three main providers of psychosocial support; the lead consultant, nurse specialist and clinical psychologist, were approached where available.

6.3 Research question

The following question was addressed in this phase:

- “What psychosocial support is provided in local centres and what are their perceived challenges?”

6.4 Aim

- To triangulate views on psychosocial provision from health care professionals to provide a comprehensive picture of care provision in the UK and identify any particular challenges or best practice.

6.5 Ethical procedure

An application was submitted to the JDCBS Steering Group in April 2019, and granted with no amendments suggested; the project was already registered with the JDCBS Sponsors NHS Trust R&D Departments and had received University
ethical approval. The Ethical permission to survey centres who are part of the JDCBS was already in place from the main MREC approval (ref ref 1/3/22 ) for the UK wide study. All participants were alerted with the survey that participation was completely voluntary.

6.6 Methods

6.6.1 Setting

This study was carried out in the context of the JDCBS, previously discussed in chapter 1.2.3. It was with these 15 centres that the surveys were sent out to.

6.6.2 Participant population

Surveys were distributed to three health professionals in each of the 15 centres. The surveys were sent to the Principal Investigator (PI) of each site in the JDCBS (generally a paediatric rheumatology Consultant, from here on termed Medical Professional), the (Paediatric Rheumatology) Nurse specialist and a Clinical psychologist who works with the rheumatology team (if available). The rationale behind this was that triangulation of data in each site would aid a fuller picture of service provision from each of the different disciplines. This could be relevant if for example one health care professional thought they offered a particular psychosocial support, but another could add more information. For example, whilst the team might have psychology provision, the actual waiting list might be too long for the majority of patients to get benefit from that support.

There was a list of PI's for each site, and the 15 nurse specialist’s all belonged to a paediatric rheumatology nurses group. The nurses have a supportive network and it was advantageous that I was active within this network, prior to the study. Of note, this is a different group to those who were instrumental in phase two, as all of the nurses involved in phase two were research nurses rather than nurse specialists. It was more difficult to identify the named psychologist for each centre. An email request was sent to the PI and rheumatology nurse specialist in each hospital asking for the contact details of their clinical psychologist. All but three of the centres could identify either a specific rheumatology psychologist, a
clinical psychologist who works with the rheumatology team, or a Trust-wide psychologist who has worked with the team in the past. Three centres could not identify an individual who would be appropriate to provide a perspective from psychology. This does not mean that they do not have psychology provision, but the numbers cannot be included in the analysis as the arrangements in these centres were unknown. Therefore, for the reporting of data from psychology, percentages are calculated out of 12 centres, rather than 15.

Responses to the surveys in this phase were collected between June 2019 and July 2019.

6.6.3 Participant recruitment

An email was sent to all health care professionals targeted, alerting them to the survey and provided an opportunity to ask questions.

6.6.4 Surveys

Surveys were the method of choice for this study, as discussed in 2.7.3. Returning to the four questions proposed by Dillman, Smyth and Christian, (2009) see section 2.7.3 these are presented in turn in relation to this study.

6.6.4.1 What survey mode(s) will be used to ask the questions?

Considering the mode of survey delivery is important as it has a potential impact on response rate. Postal addresses for the identified staff were not all known and with the inevitable delay of getting surveys through to direct hospital departments and back out again (the responses), and with the additional cost of this, the surveys were delivered electronically. Surveys were distributed using OPINIO® software; freely available through the University, the same software that was used for phase two. The OPINIO software allowed delivery direct to email addresses, and provided reminders at set times. Due to the use of specific email addresses, the surveys were not anonymous, but each individual was informed that their results would not be used in any identifiable way.
Phase 3 “The view from others on the journey”

6.6.4.2 Is this question being repeated from another survey, and/or will answers be compared to previously collected data?

All questions were designed for this survey, and not taken from elsewhere. This phase of the research study was built upon the earlier two phases, with the voices from the children and young people helping shape the questions asked. For instance, in phase one, the participants talked about a lack of knowledge from their teachers in school about JDM. As there is no world-wide JDM resource for teachers, this led to this survey including a question to understand what role medical professionals and nurse specialists play when considering school liaison and information sharing. Phase two had yielded interesting data when children and young people discussed whether they had met anyone else with JDM, and again, in this phase, health professionals were asked what opportunities were there for this to occur in their centre. So, whilst answers are not compared to previously collected data, the concerns of children and young people influenced the questions included.

6.6.5 Will respondents be willing and motivated to answer accurately?

It was anticipated that respondents would be willing and motivated to answer accurately for a number of reasons. Firstly, the results from phase one of the research had been presented at two national meetings and there had been encouraging interest and support from both medical professionals and nurse specialists. Secondly, all of these centres are active, contributing centres to the JDCBS, so already research aware, focused and engaged. Thirdly, the content was generally not on personal practice, but rather the service. If there were obvious discrepancies between centres, it was predicted that these would be due to constraints of that service, rather than a specific individual's practices.

6.6.6 What type of information is the question asking for?

There was a mix of quantitative (Yes/No and Likert style rating) questions and free text qualitative options. The surveys were purposefully kept short to avoid questionnaire fatigue; with 12 questions in the clinical psychologist survey, 14 in the nurse specialist survey and 15 in the medical professional survey. Whilst
asking different questions of each professional group would make analysis challenging, it was felt that some questions needed a bespoke approach. The questions did not aim to reveal differences between same centre responses, but to gain as much information using as few questions as possible. So, for example, there was limited gain in asking all three health professionals if there is a psychology waiting list, as the psychologists would know this answer in preference of the other two disciplines. However, the medical staff were asked if they knew if there was one, as if the centre did not have their own psychologist, it was important to understand if medical staff were aware of a Trust waiting list as this would have implications to their management of psychosocial concerns.

6.6.7 Survey set up

The surveys asked specific questions about six main areas:

- Site data.
- Clinical Psychology Provision.
- Role Specific questions.
- School liaison / Peer-2-Peer opportunities.
- Challenges.
- Rating of service.

Table 6-1 presents the questions asked of each professional group, the question number is shown with each question for each discipline, under the attributed headings, with the answer options in brackets:
### Table 6-1 Bespoke questions asked of the three health care professionals

<table>
<thead>
<tr>
<th>Question</th>
<th>Medical professional</th>
<th>Nurse specialist</th>
<th>Clinical psychologist</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Site data</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Q1. Approximately how many children under 16 years are currently under the care of your whole Paediatric Rheumatology Service? (100-200, 200+)</td>
<td>Not asked of this group</td>
<td>Not asked of this group</td>
<td></td>
</tr>
<tr>
<td>Q2. Approximately how many patients with JDM do you have in total currently in your Center? (0-10, 10-20, 20-30, 30-50, 50+)</td>
<td>Not asked of this group</td>
<td>Not asked of this group</td>
<td></td>
</tr>
<tr>
<td><strong>Clinical Psychology Provision</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Q3. Do you have a named psychologist who provides clinical care to patients with rheumatic conditions including JDM? (No/Yes) (and Freetext options)</td>
<td>Q1. Do you have a named psychologist who provides clinical care to patients with rheumatic conditions including JDM? (No/Yes) (and Freetext options)</td>
<td>Q1. What is the Whole Time Equivalent of the psychology service provided to the Paediatric Rheumatology team in your Hospital, how many individuals make up this total? Please state if no service for Rheumatology. (Freetext options)</td>
<td></td>
</tr>
<tr>
<td>Q4. If you don’t have a named psychologist as part of your team, do you have access to psychology within your Trust and are there any issues/concerns with this? (NA/No/Yes) (Freetext)</td>
<td>Q2. If you don’t have a named psychologist as part of your team, do you have access to psychology within your Trust? (NA/No/Yes) (Freetext)</td>
<td>Q2. How do patients with Rheumatological conditions like JDM get referred to psychology? (Referred by Medical staff, nursing, physio, self referral, all of above, other) (Freetext)</td>
<td></td>
</tr>
<tr>
<td>Q5. Do you ask patients (and parents) routinely whether they are seeing a psychologist / counsellor or other similar in clinic appointments? (No/Yes) (Freetext)</td>
<td>Q3. Do you and your nurse specialist team routinely ask patients and/or parents, whether the young person is seeing a psychologist / counsellor or other in clinical appointments or when talking to them on the phone? (No/Yes) (Freetext)</td>
<td>Q3. Do all new JDM referrals meet the psychology team routinely? (No/Yes) (Freetext)</td>
<td></td>
</tr>
</tbody>
</table>
### Table 6.1 Bespoke questions asked of each of the three health care professionals

<table>
<thead>
<tr>
<th>Medical professional</th>
<th>Nurse specialist</th>
<th>Clinical psychologist</th>
</tr>
</thead>
<tbody>
<tr>
<td>Q6. Does your Rheumatoology Service have a dedicated clinic to which the JDM patients generally attend?</td>
<td>Q4. Do you think (or know) that some of your patients with JDM have unmet psychosocial needs, and if so, what are these?</td>
<td>Q4. Are patients screened for any issues before they come in to clinic to identify needs in advance?</td>
</tr>
<tr>
<td>(No/Yes) (Freetext)</td>
<td>(No/Yes/Don’t know)</td>
<td>(No/Yes/Don’t know)</td>
</tr>
<tr>
<td>Q7. Do you hold any regular psychosocial meetings where children with JDM are discussed?</td>
<td>Q5. Does your rheumatology team hold any regular psychosocial meetings where children with JDM are discussed?</td>
<td>Q5. How many JDM patients have seen/are seeing a psychologist or other similar professional in your Hospital (over the last 2 years)?</td>
</tr>
<tr>
<td>(No/Yes) (Freetext)</td>
<td>(No/Yes) (Freetext)</td>
<td>(Freetext)</td>
</tr>
<tr>
<td>Q8. Do you know if you have a waiting list for psychology input in your Trust (if available)?</td>
<td>Q6. Do you know if you have a waiting list for psychology input (if available)?</td>
<td>Q6. How many JDM patients have been referred to psychology/CAMHS(^3) locally in the last 2 years approximately?</td>
</tr>
<tr>
<td>(NA/Yes - no waiting list/Yes – yes waiting list) (Freetext)</td>
<td>(NA/Yes - no waiting list/Yes – yes waiting list) (Freetext)</td>
<td>(Freetext)</td>
</tr>
<tr>
<td>Q9. Do you know what percentage of JDM patients are currently seeing a psychologist / counsellor or other similar professional, either in your Trust or through a local provider such as CAMHS?</td>
<td>Q7. Do you know how many JDM patients have seen / are seeing a psychologist / counsellor or other similar, either in your Trust or through a local provider such as CAMHS?</td>
<td>Q7. Do you have a waiting list for psychology input?</td>
</tr>
<tr>
<td>(No/Yes) (Freetext)</td>
<td>(No/Yes) (Freetext)</td>
<td>(No/Yes) (Freetext)</td>
</tr>
</tbody>
</table>

### Role Specific Questions

| Q10. Do you think you have enough time to ascertain psychosocial issues with JDM patients in your clinic appointments? | Q8. Do you think you have enough time / experience and/or support to manage any psychosocial concerns that you become aware of? | Q8. Do you attend any regular psychosocial meetings where children with JDM may be discussed? |
| (No/Yes/Sometimes) (Freetext) | (No/Yes) (Freetext) | (No/Yes) (Freetext) |

\(^3\) Child and Adolescent Mental Health Services
### Table 6.1 Bespoke questions asked of each of the three healthcare professionals

<table>
<thead>
<tr>
<th></th>
<th>Medical professional</th>
<th>Nurse specialist</th>
<th>Clinical psychologist</th>
</tr>
</thead>
<tbody>
<tr>
<td>Q9. Do you and your Nursing</td>
<td>Q9. What psychosocial</td>
<td>What psychosocial measures of well-being do you collect? (Freetext)</td>
<td></td>
</tr>
<tr>
<td>team routinely ask whether the</td>
<td>team routinely ask whether the young person is seeing a psychologist / counsellor or other similar in clinical appointments or when talking to them on the phone? (No/Yes) (Freetext)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>young person is seeing a</td>
<td>(Freetext)</td>
<td></td>
<td>(Freetext)</td>
</tr>
<tr>
<td>psychologist / counsellor or</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>other similar in clinical</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>appointments or when talking</td>
<td></td>
<td></td>
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<tr>
<td>to them on the phone?</td>
<td></td>
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</tr>
<tr>
<td>Q10. What information do you</td>
<td>Q10. What information do you give to schools concerning JDM? (None, never been asked, written letters, print off website, direct to someone else, other) (Freetext)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>give to schools concerning</td>
<td></td>
<td></td>
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<tr>
<td>JDM?</td>
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<td></td>
<td></td>
</tr>
<tr>
<td>Q11. Are children and young</td>
<td>Q11. Are children and young people provided with an opportunity for social interaction with other children and young people with JDM or other rheumatological conditions that you know of? (No/Yes) (Freetext)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>people provided with an</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>opportunity for social</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>interaction with other children</td>
<td>Not asked of this group</td>
<td></td>
<td></td>
</tr>
<tr>
<td>and young people with JDM or</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>other rheumatological</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>conditions that you know of?</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Q12. What is your biggest</td>
<td>Q12. What is your biggest challenge when providing psychosocial care? (Freetext)</td>
<td>Q10. What is your biggest challenge when providing psychosocial care? (Freetext)</td>
<td></td>
</tr>
<tr>
<td>challenge when providing</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>psychosocial care?</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Q13. Please rate how you view</td>
<td>Q13. Please rate how you view your psychosocial service for children and young people with JDM? 1 (poor) – 5 (excellent)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>your psychosocial service for</td>
<td>Q14. Please rate how you view your psychosocial service for children and young people with JDM? 1 (poor) – 5 (excellent)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>children and young people with</td>
<td>Q11. Please rate how you view your psychosocial service for children and young people with JDM (1 poor – 5 excellent)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>JDM?</td>
<td>Q15. Do you have any other comments? (Freetext)</td>
<td>Q12. Do you have any other comments? (Freetext)</td>
<td></td>
</tr>
<tr>
<td>Q14. Do you have any other</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>comments? (Freetext)</td>
<td></td>
<td></td>
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</tr>
</tbody>
</table>

**School liaison / Peer-2-Peer opportunities**

- Q11. What information have you given to schools concerning JDM if asked by a family? (None – asked others, written letters, print off website, other) (Freetext)
- Q10. What information do you give to schools concerning JDM? (None, never been asked, written letters, print off website, direct to someone else, other) (Freetext)
- Q12. Are children and young people provided with an opportunity for social interaction with other children and young people with JDM or other rheumatological conditions that you know of? (No/Yes) (Freetext)
- Q11. Are children and young people provided with an opportunity for social interaction with other children and young people with JDM or other rheumatological conditions that you know of? (No/Yes/Don’t know) (Freetext)

**Challenges**

- Q13. What is your biggest challenge when providing psychosocial care? (Freetext)
- Q12. What is your biggest challenge when providing psychosocial care? (Freetext)
- Q10. What is your biggest challenge when providing psychosocial care? (Freetext)

**Rating of service**

- Q14. Please rate how you view your psychosocial service for children and young people with JDM? 1 (poor) – 5 (excellent)
- Q13. Please rate how you view your psychosocial service for children and young people with JDM? 1 (poor) – 5 (excellent)
- Q11. Please rate how you view your psychosocial service for children and young people with JDM (1 poor – 5 excellent)
- Q15. Do you have any other comments? (Freetext)
- Q14. Do you have any other comments? (Freetext)
- Q12. Do you have any other comments? (Freetext)
The answers were not compulsory. However, individuals were informed in the introductory text how many questions there were, and that most were quick, binary options.

### 6.6.8 Piloting the surveys

The surveys were piloted with three individuals from each discipline; these were not included in the final sample (known from previous networking) and were UK wide. There was very little initial feedback from the piloting. Upon chasing feedback, the surveys were described as “clear and easy to complete”. A suggested change was from a senior psychologist, asking to include if psychosocial measures of well-being are routinely collected and to ask whether patients are screened for any psychosocial issues prior to their clinic appointments. These questions were both added.

### 6.7 Analytical approach

The quantitative data are presented simply as descriptive data, (with either numbers of patients in each centre or scoring of their overall psychosocial service provision), presented in tables or graphs where applicable. The free text comments have been added to support specific points, all are in italics and blue font. All data are kept anonymised to prevent site recognition. Where possible the results will show evidence of the triangulation in comparing comments from different healthcare professionals in the same trust, this is discussed further later in this chapter.

### 6.8 Results

First, the response rate is presented. Then the results in turn under each of the six previously identified areas (See 6.6.7) with the results from each professional group under these subheadings, showing the data for each question asked.
6.8.1 Response rate to the surveys

6.8.1.1 Medical professionals

There was a 100% response rate (n=15) with no missing data fields.

6.8.1.2 Nurse specialists

There was 100% response rate (n=15) with only a few fields of missing data. One respondent had missed out a number of the questions, and commented that they did not feel able to answer as new into post. Four respondents had not answered the multi-choice options for the question about having access to psychology within their Trust, but some of these had left qualitative comments.

6.8.1.3 Clinical psychologists

As discussed in 6.6.2 only 12 centres were sent surveys for the clinical psychologists.

A 92% response rate was achieved (n=11) with only one centre not responding to the survey.

There were three fields of missing data: two did not comment on their biggest challenges, and one did not rate their service. The centre that did not respond was sent reminders on three occasions, but no comment was received. Percentages throughout this results section are therefore calculated out of these 12 centres.

6.8.2 Site data (Q1 and Q2 for the medical professionals)

Question 1 and Q2 asked the medical professionals to report the size of their centre and number of JDM patients: this question was only asked of this professional group, as it was anticipated they would have easy access to this information. A range of answers were provided to standardise results. Two of the 15 centres had 100-200 paediatric rheumatology patients and 13 of the 15 had over 200. Within these numbers, seven centres had 10-20 patients with JDM, with other centres having between 0-10 and 50+, see Table 6-2.
6.8.3 Clinical psychology provision

All respondents were asked seven questions about psychology provision. Whilst some questions were different for the medical professionals and nurse specialists, there was also some overlap, asking about psychology provision, psychology waiting lists, psychosocial meetings and whether the number of patients having psychosocial support is known. An additional question asked the nurse specialists whether they thought their patients had any unmet psychosocial needs.

The Clinical Psychologists were asked about the nature of their service, including questions such as: psychology time provided, number of staff members, how JDM patients are referred, whether all new patients are routinely screened for an issues, how many patients with JDM had been seen in the last two years in their Trust and then locally; and whether there was a waiting list for psychological services.

The answers to these questions will now be presented by each health professional group in turn.
Medical professionals were asked seven questions (Q3-Q9) in this section of which five had binary responses. The proportion of Yes/No responses to the five binary questions given by the medical professionals are shown in Figure 6-1.

For Q3, five doctors (33.3%) reported that they did not have a named psychologist as part of their rheumatology team, but did have a Trust psychology service, although the time they offered to rheumatology was very variable, as described in this comment:

“Very limited allocation. Very short staffed and last month announced that they will not be able to see any new referrals that are not inpatients. Totally unsatisfactory”.

One centre had no access to psychology in their Trust, commenting:

“We used to, but they are closed to referrals now due to sickness, maternity leave etc. We are reliant on local services now”.

For Q5 “Do you routinely ask JDM patients or their parents whether they are seeing a psychologist, counsellor or similar?” Nine (60%) medical professionals said no and six (40%) said yes. One who said yes, commented:
Two questions which did not have a binary option, (Q4, Q8) both of which had more answer options:

Question 4 asked “If you don't have a named psychologist as part of your team, do you have access to psychology within your Trust and are there any issues/concerns with this?” Ten participants replied not appropriate (as have Trust psychology), four said yes and one said no.

Question 8 asked “Do you know if you have a waiting list for psychology input in your Trust (if available)?” One replied, NA (as no psychology service), three replied, ‘Yes, I know we don’t have a waiting list’ and eleven replied, ‘yes we do have a waiting list’. Freetext responses noted waiting list of up to five months in two centres, however one individual said:

“We have a waiting list only for brief intervention, they won’t see new referrals”

6.8.3.2 Nurse specialists (Q1 – Q7)

The nurse specialists were asked seven questions in this section, four of them binary choice questions: responses are shown in Figure 6-2.
When asked whether they have a named psychologist as part of their team who provides clinical care to patients with rheumatic conditions including JDM (Q1), eight (53%) nurses replied to say that they did not.

For Q3, one nurse did not answer, leaving seven responding that they do ask if patients are seeing a psychologist, and seven who do not.

For Q5, eight nurses (53%) responded to say their team holds regular psychosocial meetings where children with JDM are discussed.

Question 2, Q4 and Q6 had more than two answer options:

Question 2 asked “If you don't have a named psychologist as part of your team, do you have access to psychology within your Trust and are there any issues/concerns with this?” Four participants replied not appropriate, six said yes and one said no. Four participants did not answer this question. One of those that did not answer the multi-choice question, replied with a comment:

“Very, VERY limited as the trust as a whole has a large short fall in the number of psychologists”

Question 4 asked whether they think that some of their JDM patients have unmet psychosocial needs, 10 (67%) said ‘Yes’, with four (27%) saying ‘don't know’, and
Phase 3 “The view from others on the journey”
only one (7%) saying ‘No’. There were additional comments added here, for example:

“Yes - Body image, disease management, compliance, fatigue, weight gain, lack of friends, falling behind with education”

“Yes – overt skin changes & muscle weakness makes them feel different to peers and a burden on their family”

“Yes – body image as well as issues around compliance to medicines and mental health involvement”.

Question 6 asked “Do you know if you have a waiting list for psychology input in your Trust (if available)?” One replied, ‘No, I don’t know’, two replied, ‘Yes, I know we don’t have a waiting list’. Eleven replied, ‘yes we do have a waiting list’. The longest declared waiting list time was six months, however, the majority commented that they did not know how long it was, but they knew there was one.

6.8.3.3 Clinical psychologists (Q1 – Q7)

There were seven questions asked in this section.

Question 1 asked the clinical psychologists, for the number and hours of psychology provision. Only two (18%) of the sites reported having over 1.0 of a Whole Time Equivalent (WTE) (more than one individual providing psychology service). Of the four who commented that they do not have any dedicated funded rheumatology time, these results are in line with the responses from the nurses, meaning that the medical professionals believe they have a named individual for paediatric rheumatology, however, this may not be a funded service. As one psychologist commented:

“No funded service – there is an informal agreement to accept around 12 referrals/year”.

This concurs with the result that seven (50%) of the 14 centres (including the three who could not identify a psychologist to send the surveys to), do not have a named psychologist as part of the Rheumatology team. This discrepancy
Phase 3 “The view from others on the journey”

between answers from the same centre, was the primary reason for asking for more than one health professional response in each centre.

Question 2 asked “How do patients with rheumatological conditions like JDM get referred to psychology?” There was a range of answers selected, with some individuals selecting multiple choices. Seven respondents replied that patients get referred from all of the selected options, including by medical staff, nurse specialists, physiotherapists, and self-referral or by parent/carer.

In response to Q3 “Do all new JDM referrals meet the psychology team routinely?” 11 respondents selected no.

Responses to Q4 “Are patients screened for any issues before they come in to clinic to identify needs in advance?” are shown in Figure 6-3:

![Figure 6-3 Psychologists answer to patients being screened in advance](image)

This shows that the majority are not screened before they come to clinic. One of the clinical psychologists who recorded yes said:
Question 5, Q6 and Q7, were all free text responses. Q5 asked about the numbers of JDM patients seen in that Trust by a psychologist over the last two years and Q6 asked how many have been seen locally in the last two years. The numbers of JDM patients seen in each site over the last two years and those seen locally, returned limited data with the majority saying they had no way of knowing. The individual who quoted the largest figure of 25 said:

“Based on the numbers from last year, I would imagine that it would be approximately 25 patients”

In response to Q7, nine (82%) of the 11 sites who replied reporting a waiting list, ranging from four to 24 weeks, being at least ten weeks in five of the centres. Two centres said they had no waiting list.

6.8.4 Role specific questions

The role specific questions were added to provide further data to discern differences between the different professionals groups; where/if they existed.

6.8.4.1 Medical professionals (Q10)

Responses to Q10 “Do you think you have enough time to ascertain psychosocial issues with JDM patients in your clinic appointments?” are shown in Figure 6-4. Ten (67%) responded that they do have enough time, and only one (7%) responded that they do not have enough time.
6.8.4.2 Nurse specialists (Q8 and Q9)

The nurse specialists were asked two further questions to establish nursing specific roles. In response to Q8: “Do you think you have enough time and/or experience and/or support to manage any psychosocial concerns?”, six nurses (40%) said yes and 9 (60%) said no. One commented that:

“No, we do not have anywhere near enough time to do what we need to do”.

In response to Q9: “Do you and your nursing team routinely ask whether the young person is seeing a psychologist / counsellor or other similar in clinical appointments or when talking to them on the phone?”, seven nurses (47%) responded ‘yes’ and eight (53%) ‘no’ (see Figure 6-5). Three of those that said they do not ask, commented that they do not have time.
6.8.4.3 Clinical psychologists (Q8 and Q9)

Question 8 and Q9 to psychologists asked role specific questions. Seven (64%) of the clinical psychologists attend regular team meetings where JDM patients are discussed, with four (36%) who don’t, with comments that:

"the biggest challenge is not being integrated into the medical team"

and

"Unable to attend monthly psychosocial meetings as my timetable no longer allows".

One other psychologist commented that:

“We have a very small amount of resource spread across all the paediatric specialties with no designated funding, including rheumatology so we are only able to offer a very brief assessment and intervention service”.

Another said:

“No dedicated psychology time for Rheumatology”.

Question 9 asked what measures of well-being were collected. There were a variety of responses here, with three centres saying ‘none routinely’ and up to
five saying they may use the PedsQL. The range of answers provided are shown in Appendix 24.

6.8.5 School liaison / Peer-2-Peer opportunities

Question 11 for the medical professionals and Q10 for the nurse specialists was focused on school liaison. From some of the comments that were raised in the first phase, it was established that there was no school resource for JDM. This survey therefore sought to establish whether schools are generally told about JDM from healthcare professionals, and if so, what forms did this take. A broadly worded question asked the medical professionals (Q11) and the nurse specialists (Q10) “What information do you give to schools concerning JDM?” The psychologists were not asked this question as it was felt that this is a core responsibility of the medical and nursing professionals who will be providing regular clinical care. Of note, some individuals put a primary and secondary answer, so may in fact write letters and direct to others, but for reporting here, only the primary responses are shown in Table 6-3.

<table>
<thead>
<tr>
<th></th>
<th>Nothing</th>
<th>Written letters upon families request</th>
<th>Directed to others</th>
<th>Always contact the school special educational needs coordinator (SENCo)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Medical professional’s n (%)</td>
<td>5 (33)</td>
<td>10 (67)</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Nurse specialist’s n (%)</td>
<td>2 (13)</td>
<td>8 (53)</td>
<td>3 (20)</td>
<td>2 (13)</td>
</tr>
</tbody>
</table>

Two nurse specialists commented:
Phase 3 “The view from others on the journey”

“We or the therapy team liaise with school SENCo/pastoral care team. If severely affected we may go on school visit”

“It varies depending on the needs of the patient, currently though we would have to write our own letter or use online resources to print off information. Occasionally the Occupational Therapist will write letters also”.

One medical professional commented:

“None - I ask someone else to do it, e.g. the nurses or physiotherapist”.

Question 12 for the medical professionals and Q11 for the nurse specialists asked about Peer-2-Peer opportunities for social interaction with children and young people with JDM or other rheumatological conditions. This question was added from the comments expressed in phase one and phase two from the children and young people. Table 6-4 presents these results:

<table>
<thead>
<tr>
<th>Medical professional’s n (%)</th>
<th>No, there is no opportunity for social interaction</th>
<th>Yes, there is opportunity for social interaction</th>
<th>Don’t know</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 (7)</td>
<td>14 (93)</td>
<td>0 (0)</td>
<td></td>
</tr>
<tr>
<td>Nurse specialist’s n (%)</td>
<td>3 (20)</td>
<td>11 (73)</td>
<td>1 (7)</td>
</tr>
</tbody>
</table>

The numbers suggest that children are generally offered the opportunity for social interaction, with the comments providing further information of what these opportunities are including annual residential activity weekends, family rheumatology days, rheumatology outward bound trips, Scotland wide activities, Versus Arthritis Charity days, and art therapist in clinics. Of note, only two nursing and two medical professionals specifically mentioned JDM specific family days, whilst the majority of events were for all paediatric rheumatology patients. Whilst opportunities may exist, this survey did not question how patients are informed of
Phase 3 “The view from others on the journey”

them, how often they are offered, or what attendance is like, although some did comment on this:

“We’ve run a national rheumatology family day a couple of times now, it’s not been the best attended”

“adolescent activities offered, many do not want to participate”

6.8.6 Challenges (Q13 for medical professionals, Q12 for nurse specialists and Q10 for clinical psychologists)

Each individual responded to a question regarding what they thought was the biggest challenge when providing psychosocial care, these results are shown, coded from the primary answer declared, see Table 6-5. Each row is one centre. The completed answers are shown in appendix 25 for transparency.

### Table 6-5 The main challenge as declared by each professional

<table>
<thead>
<tr>
<th>Medical professionals</th>
<th>Nurse specialists</th>
<th>Clinical psychologists</th>
</tr>
</thead>
<tbody>
<tr>
<td>Time</td>
<td>Time</td>
<td>Buy in by clinical team</td>
</tr>
<tr>
<td>Lack of psychology service</td>
<td>Lack of psychology service</td>
<td>Left blank</td>
</tr>
<tr>
<td>Lack of psychology service</td>
<td>Lack of psychology service</td>
<td>Left blank</td>
</tr>
<tr>
<td>Buy in from family</td>
<td>Buy in from family</td>
<td>Did not reply to survey</td>
</tr>
<tr>
<td>Lack of psychology service</td>
<td>Time</td>
<td>Lack of psychology service</td>
</tr>
<tr>
<td>Lack of psychology service</td>
<td>The JDM itself</td>
<td>Left blank</td>
</tr>
<tr>
<td>Lack of psychology service</td>
<td>Time</td>
<td>Lack of psychology service</td>
</tr>
<tr>
<td>Lack of psychology service</td>
<td>Time</td>
<td>Lack of psychology service</td>
</tr>
<tr>
<td>Lack of psychology service</td>
<td>Lack of psychology service</td>
<td>No Psychologist to ask</td>
</tr>
<tr>
<td>Lack of psychology service</td>
<td>Lack of psychology service</td>
<td>No Psychologist to ask</td>
</tr>
<tr>
<td>Time</td>
<td>Time</td>
<td>Time</td>
</tr>
<tr>
<td>Buy in from family</td>
<td>None</td>
<td>Time</td>
</tr>
<tr>
<td>Space</td>
<td>Time</td>
<td>Time</td>
</tr>
<tr>
<td>Time</td>
<td>Buy in from family</td>
<td>Lack of psychology service</td>
</tr>
<tr>
<td>Lack of psychology service</td>
<td>Lack of psychology service</td>
<td>Time</td>
</tr>
<tr>
<td>Time</td>
<td>Left blank</td>
<td>Time</td>
</tr>
</tbody>
</table>
Phase 3 “The view from others on the journey”
‘Lack of psychology’ is mentioned 16 times and ‘time’ is mentioned 14 times. Four individuals thought ‘buy-in’ from patients and their families was their biggest challenge.

6.8.7 Rating of service (Q14 for medical professionals, Q13 for nurse specialists and Q11 for clinical psychologists)

Each individual was asked to rate their psychosocial service of their paediatric rheumatology department as a whole, these results are summarised in Table 6-6. As one nurse specialist did not score their service, there is a total of 40 replies.

<table>
<thead>
<tr>
<th>Rating</th>
<th>Poor</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5</th>
<th>Total Number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Medical professional n (%)</td>
<td>1 (7)</td>
<td>1 (7)</td>
<td>5 (33)</td>
<td>7 (47)</td>
<td>1 (7)</td>
<td>15 (100)</td>
</tr>
<tr>
<td>Nurse specialist n (%)</td>
<td>0 (0)</td>
<td>2 (14)</td>
<td>7 (50)</td>
<td>4 (29)</td>
<td>1 (7)</td>
<td>14 (93)</td>
</tr>
<tr>
<td>Clinical psychologist n (%)</td>
<td>1 (10)</td>
<td>1 (10)</td>
<td>3 (27)</td>
<td>5 (45)</td>
<td>1 (10)</td>
<td>11 (91)</td>
</tr>
</tbody>
</table>

| Total Number n (%) | 2 (5%) | 4 (10%) | 15 (37%) | 16 (41%) | 3 (8%) | 40/41 (98) |

6.8.7.1 Medical Professionals

The majority, (seven, 47%) of medical professionals scored their service as a four, adding comments that:
Phase 3 “The view from others on the journey”

“Our service is doing its best, but limited by the quantity of psychology provision as a whole”

“Our doctors, nurses and physiotherapists provide psychosocial support as far as possible. When families see Clinical Psychology this is a very good service but the main problem is delay in accessing this”.

6.8.7.2 Nurse specialists

Half of the nurses scored their service as a three. One of the nurses commented that:

“Our psychologist is excellent but part time. To have funding for full time hours would allow more accesses for families in a timely manner”. 

6.8.7.3 Clinical psychologists

Ten out of the eleven psychologists completed this question, with 5 (50%) giving it a rating of four. One commented that:

“We could be doing some more preventative work, at the time of diagnosis-education around adjustment and developmental challenges along the way-preparing parents for conversations about long-term conditions”.

6.8.8 Triangulation of results

It was decided initially to approach three health professionals to answer the surveys as this would enable a more comprehensive picture than only asking one individual in each centre. Whilst the study did not aim to look for differences between individuals, differences have been observed. Whilst rating their overall service, is a subjective question, it is still important to note any obvious differences and similarities. Table 6-7 shows the rating for each individual in each centre.
Phase 3 “The view from others on the journey”

### Table 6-7 Presenting the rating per centre for each professional

<table>
<thead>
<tr>
<th>Medical professional</th>
<th>Nurse specialist</th>
<th>Clinical psychologist</th>
</tr>
</thead>
<tbody>
<tr>
<td>4</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>3</td>
<td>Did not answer the question</td>
<td>No Psychologist</td>
</tr>
<tr>
<td>4</td>
<td>2</td>
<td>4</td>
</tr>
<tr>
<td>4</td>
<td>4</td>
<td>Did not reply to survey</td>
</tr>
<tr>
<td>1</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>4</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>3</td>
<td>3</td>
<td>2</td>
</tr>
<tr>
<td>3</td>
<td>3</td>
<td>No Psychologist</td>
</tr>
<tr>
<td>3</td>
<td>3</td>
<td>No Psychologist</td>
</tr>
<tr>
<td>4</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>4</td>
<td>5</td>
<td>4</td>
</tr>
<tr>
<td>5</td>
<td>4</td>
<td>3</td>
</tr>
<tr>
<td>4</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>3</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>2</td>
<td>3</td>
<td>5</td>
</tr>
</tbody>
</table>

Only two centres had all health professionals in agreement, and both of these were for a grade of ‘4’ in the centre. Three other centres had agreement between the medical professional and nurse specialist, with no clinical psychologist scoring.

One of the aims from this study was to share best practice and good ideas between centres. There was one positive comment sharing good practice which stood out through the many difficult challenges that were discussed in the survey responses. A psychologist stated:
Phase 3 “The view from others on the journey”

“I work in a psychologically minded team where difficulties for the children or the parents are raised in a timely manner. Because I sit in with Consultants during some Rheumatology Clinics this means that we can identify work that can help with psychosocial issues that the rest of the team can also work on without necessarily needing a separate referral to me”.

This integration of the psychologist working with the rheumatology team and making shared decision making is a great result from this centre, but can only be actioned in other centres if psychology support is provided, and with time available to sit in on clinics. This psychologist only scored their service a four though, implying that there is still room for improvement, and whilst this psychologist scored their centre a four, the nurse specialist scored it a two and commented that:

“Our psychology service is very limited, we need more psychology support. The waiting list is extremely long”.

In one of the centres where there is no psychology provision at all, the effect this has on other health professionals could be seen in the answers given and thus it is worth viewing these together. The medical professional knew how many patients had been seen by psychology, always asked in clinics about whether the young person was seeing psychology and holds regular psychosocial meetings. The nurse specialist also checked the phone numerous times a day, always asked children, young people and their families if they were receiving psychosocial support and always contacted the schools in person. Both of these individuals rated their service as a three, see 6.8.7, and this was summed up in the following comment:

“I feel we have a holistic approach and are quick to identify issues and offer as much support as possible, but we struggle to move to the next level. We have no ability to pre-empt issues by involving psychology services from diagnosis”.

6.8.9 Any other comments? (Q15 for medical professionals, Q14 for nurse specialists and Q12 for clinical psychologists)

All health care professionals were offered the opportunity to add any extra free text comments. Five of these are presented here:
Phase 3 “The view from others on the journey”

“We don’t ask enough about mental health in our Connective Tissue Disease patients, too much focus during clinic is on their physical issues”.

“Long waiting times. Big case load means they [the clinical psychology service] do not prioritise diagnosed with children with chronic diseases and are a reactive service. Regional politics also come into it and if they out of our city boundaries they are referred to another team”.

“As the medical professional trying to secure safe, effective, and timely care for their patients, one can feel the frustration from arguing over health care boundaries when dealing with a child’s mental health”.

“Dedicated psychology for rheumatology patients is vital but often comes with unavailable and unacceptable waits”.

6.9 Discussion

The research question that shaped and directed this phase in the beginning of this chapter (See 6.3), is now discussed.

From the answers provided throughout this study, it is clear that most centres do have access to psychology, either through their own named rheumatology clinical psychologist, or through a Trust clinical psychologist giving time to rheumatology. However, the hours of time available for rheumatology patients, is very different in each centre and how integrated the psychologist is, is also different in all centres. Interestingly, in centres where the clinical psychology support is lacking, or very minimal, other members of the team have shown a greater awareness of psychosocial issues and concerns amongst their patients, than perhaps those who do have more psychology availability.

The frustrations among clinical psychologists were apparent from their answers. Thus clinical psychologists voiced frustration regarding, capacity, time, logistics and the limits imposed by not having funded rheumatology time. There are often informal agreements to accept a certain number of patients per year, leading to
the question what happens for the patient who is 13th in line for a psychologist who is only allowed to see a quota of 12 that year? Funding for paediatric psychology provision across different medical specialities is often inconsistent leading to a lack of parity between psychosocial care afforded to those presenting with different long-term health conditions. A number of specialties now formally recognise the importance of truly holistic biopsychosocial care and as such have placed the importance of psychological provision within standards or tariffs for the commissioning of services (e.g. diabetes, oncology). Without this, it seems very challenging to advocate for the importance of dedicated psychology time.

One concern that was raised frequently by all three groups of health care professionals, was that of JDM being a rare disease and patients living far away from treating sites and thus needing long journeys to access care. The issues imposed by working within healthcare boundaries and large geographical areas, is not specific to the UK, with many patients with JDM in the US also having issues when accessing care (Kwa et al., 2018). However, one of the issues specifically highlighted of not knowing who to contact, whether psychologist, Child and Adolescent Mental Health Services (CAMHS), play therapy, health in mind, community councillor or other provider, is important as for rheumatology healthcare professionals without a named individual to refer to, this choice can cause confusion and often delay the referral for even longer as practitioners have to decide who accepts it. This was added as a comment at the end of the survey by a health care professional who said, one can feel the frustration from arguing over health care boundaries when dealing with a child’s mental health.

The result that 50% of the medical professionals scored their service 4/5 is encouraging and mirrors the comments presented of how each individual is trying their best to consider psychosocial care, even if they do not have any psychology support. There were only two centres where all three scored their service the same score. These two centres had the most psychology offered to rheumatology, both with more than one individual being part of the team.

The comments from centres where there is no easy access to psychology, as discussed above, presented a picture where the other team members provide more of this care and support, than centres with psychology support. This may
Phase 3 “The view from others on the journey”

well be appropriate in line with the recent acknowledgement that psychosocial care and provision should be everybody’s business rather than just dedicated psychological practitioner’s responsibility. Also, in centres where there is no easy access to psychology, other team members may become more involved with psychosocial care and supporting patients through their disease trajectory and thus they are more aware of the issues experienced by young people and their families. This is corroborated by the fact that the nurses without psychology support in their centres were those who articulated the issues faced by their young patients, when those who had psychology services did not.

Writing letters can be seen as an administrator task and may contribute to having limited time to provide face to face care. The range of individuals discussed in response to the school question shows the range of healthcare professionals who could be involved in school liaison, such as medical, nursing, physiotherapists, occupational therapist or therapy team. Currently there is no worldwide resource about JDM provided for teachers. A relatively simple solution is the production of a resource for nurseries, schools and universities and may release more time for healthcare professionals to provide psychosocial support.

For JIA, there is a national published ‘Standard of Care’ document, with standard 19 stating:

“In addition to the assessment of disease activity, all aspects of the current physical and psychological health of a child or young person with JIA must be assessed and addressed by members of the MDT”

(Arthritis and Musculoskeletal Alliance, 2010. p. 13)

The document goes on to specify that some families will need help from a clinical psychologist or from mental health services. As this document however, is only for JIA, children and young people with SLE and JDM are not included.

The findings from this survey have relevance not only to JDM, but also to paediatric rheumatology as a whole. Hopefully, this survey can help some centres lobby for more support and services and can use this data to illustrate the variation in practice across the UK, but also the urgent need for more psychology support. As one medical consultant said:
This chapter opened with a quote from a recent study to identify behavioural health care provider’s perspectives on gaps in mental health for children and young people with rheumatic conditions. This study concludes with the statement that optimisation of mental health in paediatric rheumatology is an important and ambitious goal which requires several components to overcome existing barriers (Knight et al., 2019). Integration of proactive mental health, optimising existing mental health resources and providing peer support for children and their families would help to accomplish this goal (Knight et al., 2019). These recommendations are clearly supported by the findings of this study.

6.10 Implications for practice

This study has provided a snapshot of what psychosocial provision for JDM looks like currently across the UK. The data suggest that in some centres, psychology support is absent, or very limited and thus this study will hopefully assist health care professionals to consider what further support they need and lobby for this as a local level.

Despite some novel and interesting results identified in this survey, there is still a great deal of work to be done and could include focus on the following:

1. A standard of care statement or guidance document is required for all rare, chronic paediatric rheumatological conditions. This would help health care providers to have a standard which is endorsed by rheumatological charities, bodies and societies which stipulate the minimal care which should be provided, and this then can be taken to management boards/commissioners to advocate for more psychological assessment and intervention.

2. More preventative work needs to be done early on in the disease trajectory. As one of the psychologists commented “We could be doing some more preventative work, at the time of diagnosis - education around adjustment and developmental challenges along the way- preparing
parents for conversations about long-term conditions”. This can only be possible with more psychology time.

3. More joined up care, especially for those families who cannot travel to the larger centres. As one of the medical professionals commented “Working with groups of patients with JDM is helpful but with small numbers of patients and wide range of ages, issues are different, and benefits of a group are limited. More organised regional/national groups/virtual groups would be helpful”.

6.11 Strengths

There is a paucity of literature on psychological aspects of JDM, and this study provides important information to guide improvement in psychosocial care for this patient population. The strengths of the study include its high recruitment rate and inclusion of three groups of care professionals. This study is the first that we are aware of to ask a UK wide sample about psychosocial provision for children and young people with JDM.

6.12 Limitations

This survey does have some limitations. Firstly, only asking health care providers in the larger centres in the UK may make this less representative to smaller UK sites who see children and young people with JDM. This also has implications when answering some of the questions asked, for example, if you only have a few children with JDM in your centre, you are less likely to have a set clinic for them to attend. Services change rapidly with staff leaving and joining on an ever-rolling basis, so whilst one centre may be struggling this month, provision of care may well have improved by the next month. The survey in itself only asked a very small number of questions and may of missed important aspects of psychosocial care such as, the provision of any intervention groups, such as therapy compliance. The survey also did not ask the wider team their views surrounding psychosocial provision, such as Occupational Therapists, Physiotherapists and School teachers, but their supportive role was often discussed within the medical or nursing responses. In addition, the survey did not gather data about what local primary care services patients were able to access.
6.13 Conclusion

This is the first survey to jointly collate medical, nursing and psychology perspectives across the UK for the provision of psychosocial support in paediatric rheumatology centres for patients with JDM. Through our survey, we found that psychology support is limited in all centres and is determined by time and case load, with one centre having no psychology support and others having very limited. This current study highlights the importance of integrating psychology into paediatric rheumatology teams, sharing knowledge and decision making, to benefit the patients.

6.14 Next chapter

The next chapter presents the work undertaken during phase four of the study. This final phase specifically the dissemination and engagement workshop and discussion for future plans.
“School”

We sat on the floor in assembly,

All my friends and me,

We had just finished singing,

I didn’t know my JDM was beginning,

We had to stand,

But I needed a hand,

I couldn’t get off the floor,

But my friends left through the door,

My parents had to come into school,

My teachers thought I was acting a fool,

They needed to explain,

What JDM is and why it’s such a pain,

JDM makes me different, there is no doubt,

I need lots of sun cream on before I go out,

I get tired real quick,

My medicines can make me feel sick,

Sometimes my legs hurt and ache,

I’m not being a fake,

I have hospital visits and stays,

And I do have some really bad days
Chapter 7: Phase 4 “A reflection of where we’ve been and where still to go”

“Children have aspirations and capacities to participate in promoting their interests as well as the interests of others; that children are active agents.” (Carnevale, 2020, p.3).

7.1 Introduction

In this chapter, the involvement to date of children and young people throughout this research journey will be considered. Their final contribution, and how they came to help shape outputs, and future directions, will be reflected upon. Here I present phase four - the dissemination and intervention workshop, the concept, aims, participants, methods and the discussions that followed.

The idea that children and young people are active agents, having a view on their world and the world of others, as the above quote highlights, is not new. As 1.2.4 emphasised, involving children in research is actively encouraged and provides them with an opportunity to express their views. Actively listening to this voice though is more than listening, it is about empowering and enabling. Taking this a step further and involving children and young people in the results so far and the dissemination of the research they have been a part of, co-creating where to take it next, has taken my research journey full circle.

There is a growing appreciation of the need to report results back to research participants. The National Institute of Health Research INVOLVE published a briefing paper recommending not only reporting back, but sharing information on the progress of the research as well as the final results (INVOLVE, 2012). When carried out in a careful and thoughtful way, to avoid unnecessary upset, it is one possible way of demonstrating respect and gratitude for participants contributions, an approach that may lead to greater trust and engagement in future research (Botkin et al., 2018). For the fourth phase of this study, the priority was to disseminate the work undertaken so far. This was planned as a dedicated phase specifically, as a final engagement with research participants, in recognition of its importance to this study.
7.2 Why do families engage in research?

For families affected by rare conditions, participating in research can be empowering with many patients hoping for a cure. A report by Rare Disease UK found that 80% of patients with a rare disease said they would be interested in participating in research, however, only 20% felt they were given enough information about research to do so (Rare Disease UK, 2019). Research offers potential hope and can make children and young people feel they are giving something back in return for the care they receive, providing a sense of altruism (Paquette et al., 2020).

As discussed already in 1.2.3, patient registries are a key tool for gathering the scarce knowledge relevant to rare diseases in terms of basic and clinical research as well for epidemiological and public health purposes, so as to improve the understanding of these conditions and the treatment available to patients (Aymé and Rodwell, 2014). When considering families understanding of registries, one advantage is that families are often talked to about research at the very beginning of a child’s diagnosis and become “research aware”, as they frequently complete research documentation as part of the longitudinal follow-up. In practice, we have seen the evidence of this when asking children and young people with JDM and their families to engage in patient and public involvement events, and other research studies which would like involvement from those in the registry, as generally interest and engagement is high.

7.3 Patient and Public Involvement and Engagement (PPIE)

7.3.1 Prior to phase four

Patient and public involvement and engagement was a significant feature of this research, even before the study began. This continued throughout all phases with children, young people, their parents, health care professionals and members from relevant charity groups, asking to be involved. A variety of methods were used, some were invited opportunities and others occurred by chance. Involving families throughout the research journey has been at the heart of this study and thus every opportunity that presented itself was grasped, and each one of these
Phase 4 “A reflection of where we’ve been and where still to go”

occurrences, led to more questions, more comments and more invitations. These are summarised in Figure 7-1.

The first PPIE events in 2016 were to establish what children, young people and their parents thought about what was required, whether the study was needed or not. The Charity family day in July 2017 was an ideal opportunity to really engage with families and for them to play a key role in deciding what creative methods would work and what would not work for phase one. Having the children and their parents offering real life experience at this stage, was really valuable. In 2017 and early 2018, the focus shifted to examining the findings from phase one and how this would shape phase two. During the planning of phase two, and the publication of the first paper, this led to some parents contacting me with words of support and encouragement for example:

“I have just read your paper, I love it. It really is exactly what our kids go through, thank you so much for your amazing work. Keep it up!”

In September 2018 and early 2019, the focus of the PPIE involved feeding back and thanking those who had taken the time to respond in phase two. A US JDM family support group who had heard about my work and wanted to establish
further networks also contacted me. In April 2019, a newsletter was sent to all children and young people in the JDCBS, not dependent on whether they had completed questionnaires, see Figure 7-2.

As this newsletter specifically asked families to contact me if they wanted to stay involved in the study, this sparked a flurry of emails, such as these:
Phase 4 “A reflection of where we’ve been and where still to go”

“I received the letter yesterday about the results from the JDM survey and saw the little section at the bottom about getting involved. I would be really interested in getting involved and helping to spread awareness about the illness as I know that when I first fell ill, I felt like no one understood and felt quite alone. Getting us involved is such a great idea and I would love to get involved. Thank you for asking me”

And: “I would love to carry on taking part in your study, I hope it’s all going well!”

During early 2019, the PPIE became more focused, it included key stakeholder groups, such as the major charities associated with JDM. Myositis UK is a small national charity providing information to those affected by myositis and funding research to improve diagnosis and treatment ([https://www.myositis.org.uk/](https://www.myositis.org.uk/)). Versus Arthritis is the UK’s largest charity dedicated to supporting people with arthritis and other musculoskeletal disorders, including JDM ([https://www.versusarthritis.org/](https://www.versusarthritis.org/)). Involving key stakeholder groups was important for a number of reasons:

- To increase collaboration with relevant groups which are working with the population of interest.
- To share research and increase dissemination opportunities.
- To gain support and engagement which may lead to future mutually beneficial prospects.

7.4 Phase four: dissemination and intervention workshop

In the autumn of 2019, children, young people, their siblings, parents, health care professionals and charity groups were invited to spend a morning discussing the findings of the study from the earlier three phases, and help shape ideas for future research. This was the culmination of all the strands of PPIE to date, and all the findings thus far. The planning, preparation and delivery of the workshop and discussion of results are presented here.
7.5 Why use a workshop made up of two focus groups?

As Troya et al., (2019) highlights, capturing lay perspectives is essential to ensure research findings inform clinical practice. Workshops offer a method to achieve this, through group discussion of findings and pathways for future engagement. The session was planned for half a day, and thus the whole event was deemed a ‘JDM workshop’, which was composed of two focus group discussions with two age groups (parents and children/young people).

A focus group is defined as a discussion involving a small number of participants led by a moderator to gain insight into participants experiences and attitudes (Hennessy and Heary, 2005). Focus groups in particular are useful for ensuring service users’ participation in decision making, gaining insight into personal opinions, perceptions, attitudes, thoughts and experiences (Zupančič, Pahor and Kogovšek, 2019). Focus groups are known to have advantages over other research methods including:

- Offering unexpected insights.
- Participants using their own words to express their viewpoints.
- Moderators can ask questions to clarify comments.
- People tend to be less inhibited than in individual interviews.
- Remarks from one individual can stimulate comments from others.
- Focus groups are flexible and can be varied in timings.
- Focus groups usually take less time than individual interviews or written surveys.
- Generally a low cost research method. (Young, 1993).

One of the biggest potential attributes with focus groups is around the group synergy. This chain effect of stimulating participants to remember forgotten details or reflect on new ideas, paves the way from one different interpretation to another and thus produces superior final knowledge (Acocella, 2012).

When considering children and young people in particular, a further advantage is that they can offer safety in a group dynamic, seen as mirroring the type of small group settings that children experience in the daily school environment. Children
and young people can be selective in what they want to disclose and can exercise their right to remain silent if they so wish, and thus this helps to address some of the power imbalance between adult researcher and child participant (Clark et al., 2014).

On the converse, there are concerns with focus groups which need to be considered and mitigated where possible. The speed of conversation, jumping from idea to idea in the earlier described chain effect can leave ideas behind without full discussion and others dwelled upon in minute detail. Other limitations have been discussed by Acocella, (2012) and include:

- The presence of others can inhibit an individual to say what they truly feel.
- Individuals may agree with others when in fact they do not, potentially causing unreliable answers.
- Participants may resist the desired mode of interaction, either remaining silent or only responding when asked direct questions.

7.6 Research question

- “How can we take these findings forward from this research so far, and improve psychosocial care even further in the future?”

7.7 Aims

The aim was to:

- Share findings with children and young people, health professionals and key stakeholders to discuss the development of interventions to address unmet needs and target service gaps in the future.

When considering the findings from the three phases of this study, there were a number of areas one could focus on to share with families and to take forward with the aim of developing interventions. The role of uncertainty had been a prominent finding as discussed in section 5.11.1, however it was felt that there was still significant work needed to fully understand the complexities of
Phase 4 “A reflection of where we’ve been and where still to go”

uncertainty, especially when considering whether the unpredictability or lack of comprehension was most important, before considering an intervention. This is discussed further in section 8.6. More tangible, a lack of a school resource and improving peer support were seen by the wider supervisory team, as more relevant across this population with greater impact for a bigger number of individuals. Timely impact on patient experience could be achieved more easily to move forward with in the near future, and may improve patient experience in a shorter time frame. Therefore, these two ideas were decided upon to address in phase four.

7.8 Ethical procedure

No ethical approvals were necessary. Participation was voluntary and only included those who had expressed previous interest to stay involved.

7.9 Focus Group one

7.9.1 Evidence for the need to advise schools

Throughout this study, it has become apparent that the lack of a school resource was an issue for many children and young people. Time spent frequently explaining to teachers about what JDM is and how it affected them, was seen as frustrating by all children and young people. Parents also commented that they too spent time meeting with school staff and writing letters on behalf of their child. There were many schooling issues discussed in all phases, from the invisibility of the disease, missing school lessons, lack of work sent home, worry over exams, sharing of personal information with peers, bullying by other peers, beliefs and attitudes of teaching staff, attendance at hospital schools, physical limitations imposed by the school environment, limitations from the JDM making some lessons impractical, differences between different schools, support from school nurse and special educational needs team and general lack of understanding from teaching staff. It was these concerns and possible solutions which the focus group sought to capture, in parallel with whether the production of a school resource could address some of these points.
Phase 4 “A reflection of where we’ve been and where still to go”

7.10  Focus Group two

7.10.1 Evidence for the need for peer mentoring

During phase one, children and young people with JDM had articulated that they felt alone, and the majority reported that they had not met anyone else with JDM. This led to phase two asking whether children and young people had met anyone else with JDM, or whether they would like to.

From the results and comments returned, it was clear that the majority had not met anyone else with JDM. For the children and young people who said they had met someone else before, 60% would still like to meet someone else. There were many comments about why young people would like to meet someone else as presented in 5.9.12.6, including: so they would not be so alone and to see how someone else coped with it. Health professionals in phase three discussed a number of networking events for children and young people, but with the knowledge gained from phase two, it is still unclear who is offered these, who they are aimed at, who attends, and how much benefit the children and young people gain from them. This focus group was therefore to discuss whether peer mentoring from another young person with JDM would be helpful and what might be some of the issues to consider.

7.11  Methods

7.11.1 Workshop Participants

From the PPIE events discussed at the opening of this chapter, some children, young people and their families asked to remain involved with this research. This contact occurred through the following avenues:

- Families staying behind after presentations at family days to ask further questions.
- Families who were interviewed in phase one, asking to stay involved.
Phase 4 “A reflection of where we’ve been and where still to go”

- Sending an email in response to receiving the questionnaire pack, highlighting their further interest, or children and young people adding this as a comment to the bespoke questionnaire.
- Emailing in response to the newsletter sent after the phase two study and asking if they could stay involved.
- Comments posted onto the closed Facebook page, which were sent on to me from the parent administrators of this group, who had been incredibly supportive of this study throughout.
- Twitter comments and direct emails in response to the first published paper.
- From contact with the charities who kindly shared and disseminated the earlier results of the research.

Individuals who had sent contact details throughout earlier PPIE events were then emailed to gauge their current interest and commitments and to enquire about potentially attending a workshop in the autumn. Those that responded positively, were sent an email with further details.

An email also went out to the UK-wide paediatric rheumatology nurses group informing them of the workshop and inviting them to participate. Similarly, the supervisory team were all invited. The clinical team in the hospital setting were also invited and two very kindly offered to be involved in the workshop and two others offered to be available to help with logistics.

7.11.2 Setting of the workshop

The date was planned for September 2019 in London. This date was chosen specifically as a JDM Family Day was occurring the same afternoon in the same location. Ideal for families who wanted to attend both. It was also necessary to be on a weekend to maximise children and young people’s attendance when they would not be receiving education. If families asked for help with travel funds to attend, this was accommodated and all young people (those with JDM and siblings) were given a voucher at the end of the morning in gratitude of their time and involvement. Figure 7-3 shows the invite for the event:
7.11.3 Procedure for the workshop

The plan for the workshop consisted of a short presentation and then break out into two focus groups, one consisting of the parents and one for the children and young people. In the groups there was an ice breaker (a round robin fact about yourself), introductions and discussion of a clear set of rules, which included:

- Do not talk over other people.
- Be respectful to everybody’s point of view.
- There is no wrong answer, all viewpoints are valid and important.

In order to reduce the risks with focus groups discussed in 1.4.1, some processes were put in place. There was a moderator allocated to each group, with careful discussion before the workshop to ensure each was confident and capable to maintain group dynamics. The more experienced volunteer from the charity group which works with children and young people on a regular basis, was given
moderation of the children and young person group as she was skilled in ensuring all young people felt able to speak whilst not dominating the group.

At the end of each of the two focus groups there was an opportunity to capture individual ideas and thoughts, written down anonymously on cards. For the school booklet this was seen as particularly useful as it would provide some further notes to take away and collate and consider when finishing off the school resource. This was also useful for the quieter members of the groups, if they had not wanted to share their comments more openly.

7.11.4 The materials and topics for the focus groups

7.11.4.1 Dissemination presentation

The workshop began with a presentation of the research project, detailing all the phases, aims, methods and results. This was all presented in a lay format applicable to children from eight years of age and upwards. The presentation offered the opportunity for comments and questions from the audience, specifically around the rollercoaster phenomena and how well it captured the families’ journeys.

7.11.4.2 Materials discussed

I prepared in advance a draft school booklet of 20 pages and printed one per individual. This was based on other chronic disease resources and included such headings as:

- What is JDM?
- 5 key signs and symptoms.
- Symptom checklist.
- Special considerations for nursery/primary/secondary/college.
- And ten other sections: school environment, PE, fatigue, skin rashes, Sun cream, exercise, diet, infections, medications, exams and top tips.

The participants were encouraged to write on these if they wanted to add any personal comments. There was also copies of other chronic disease school leaflets and booklets provided to help stimulate ideas. A list of prompt questions
was given to the focus group moderator to help provide some direction to the discussion and encourage answers to specific questions (Appendix 26).

For the mentoring session, key prompt questions had also been prepared in advance to cover certain areas, but also free conversation was encouraged (Appendix 27).

7.12 Results

7.12.1 Participants who attended the workshop

The workshop was made up of two focus groups in the morning. According to Bowling (2006), typically focus groups are composed of between six and twenty individuals and a group leader who uses an unstructured guide, with groups lasting between one and two hours in a comfortable environment with refreshments. Initially many individuals had expressed interest and asked to be involved, however, on the day there were four families who were unable to attend.

Thirty-three individuals made up the group on the day with:

- 23 family members (nine parents, two siblings and 12 children and young people with JDM).
- Five charity volunteers (two of these are also parents from the same family).
- Five staff members.

The young people were between the ages of eight and 26 years of age. Families came from all over the UK, with one family attending from Scotland. Due to risk of identification from these small numbers of participation, no further demographic details are presented.

7.12.2 Focus group one

7.12.2.1 Children and young people group

The moderator began by offering the children and young people the opportunity to talk about their school experiences. The majority of these comments centred
Phase 4 “A reflection of where we’ve been and where still to go”

on lack of understanding from teachers, for example getting across the fluctuating nature of JDM, the importance of emailing missed work, needing extra time during exams and other work set during physical education lessons. Further comments are listed in Table 7-1:

<table>
<thead>
<tr>
<th>Table 7-1 Children and young people’s school experiences</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Children and young people’s comments</strong></td>
</tr>
<tr>
<td>“I also had really bad joint stiffness, so writing was really difficult”</td>
</tr>
<tr>
<td>“School nurse emailed teachers after chat with parents to say I needed extended exam time. Teacher asked whether that was right, in the middle of my whole class in front of people and said they did not think it was necessary”</td>
</tr>
<tr>
<td>“My teacher once got angry in front of class because I asked to leave for a walk as I felt stiff, she thought I was time wasting”</td>
</tr>
<tr>
<td>“We need to help teachers to understand how diverse and fluctuating the disease is from day-to-day, I’m not lying about my symptoms they change!!”</td>
</tr>
<tr>
<td>“I’d like greater emotional support in school please”</td>
</tr>
<tr>
<td>“Hospital appointments really disrupt the school day as it makes it very obvious that you have a condition”</td>
</tr>
<tr>
<td>“An understanding at college for when I am off, for them to email me the work I missed that I need to do”</td>
</tr>
<tr>
<td>“Resting area in school as a hall pass, with no questions asked is vital!”</td>
</tr>
<tr>
<td>“Sometimes when the nausea was really bad from methotrexate I was able to leave class straight away, because I had a class pass”</td>
</tr>
<tr>
<td>“For me, health appointments were seen as lack of attendance and I was marked as out of school”</td>
</tr>
<tr>
<td>“In PE [Physical Education] you need to be given something to do, so you can do what you can”</td>
</tr>
<tr>
<td>“Teachers responses are not good, mine didn’t bother to understand”</td>
</tr>
</tbody>
</table>

7.12.2.2 Parent Group

The parent focus group began quietly looking at the prepared booklet and school booklets for other chronic conditions, but after a few minutes of this, they then moved onto answering the prompt questions.
The results combined for both groups from the prompt questions, are shown here in Table 7-2:

### Table 7-2 Schooling questions asked of both groups and comments

<table>
<thead>
<tr>
<th>Questions</th>
<th>Children and young people</th>
<th>Parents</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Should the booklet only target one age group or be for nursery, primary, secondary school and university?</td>
<td>No, I think you need different books&lt;br&gt;I feel like they need one for nursery especially&lt;br&gt;At secondary school they need help understanding rather than giving care&lt;br&gt;I find it harder to differentiate between should ask for help or just get on with it&lt;br&gt;Not one book for all</td>
<td>No, will be too long&lt;br&gt;Nobody reads anything&lt;br&gt;Make a shorter leaflet for teachers&lt;br&gt;Put in contact with charities so they can find their own information&lt;br&gt;Different leaflet for each age group&lt;br&gt;Need to mention calcinosis&lt;br&gt;Need to suggest giving pupils time out cards for when it gets too much&lt;br&gt;Need to keep textbooks in room, not carry around</td>
</tr>
<tr>
<td>2. Should the booklet be thorough and include a lot of information, or better to be less complicated?</td>
<td>Less information, otherwise people skip more&lt;br&gt;Teachers are too busy to read a lot of information&lt;br&gt;Parents can always fill in any gaps, although the school doesn’t always listen to them</td>
<td>Make sure language is easy to read&lt;br&gt;Hard to get across the idea of flare and unpredictability&lt;br&gt;Need to encourage all teachers to read it&lt;br&gt;Maybe need something separate for PE teacher</td>
</tr>
<tr>
<td>3. Should it include medications?</td>
<td>Maybe side effects from medications&lt;br&gt;They don’t need to know the specifics, but just how they affect you</td>
<td>Medication is important&lt;br&gt;They need to understand the importance of pain and flare&lt;br&gt;Similar to arthritis in some, but not all</td>
</tr>
<tr>
<td>Questions</td>
<td>Children and young people</td>
<td>Parents</td>
</tr>
<tr>
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<td>---------------------------</td>
<td>---------</td>
</tr>
<tr>
<td>4</td>
<td>A parent has suggested there should be a tick page in the beginning for quick symptoms the child/young person has, is that a good idea?</td>
<td>Symptoms change, I don’t know how flexible that could be.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>It might make the school aware that there are a lot of symptoms and how they change.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Certain symptoms, such as the facial rash, I don’t feel need to mention in my opinion.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Making them aware of how they need to respond to certain symptoms would be a good idea.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>A description of what it might be like some days maybe.</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>Look at the booklets and tell us any other comments, things that need to be added or taken away?</td>
<td>School could treat it like a serious allergy.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>How to convey to schools that work can be sent home.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>There are so many differences, school to school, disability service, communication and planning.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Counselling services, find that one person that can help you.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Work out how your symptoms affect you, do you need a laptop maybe?</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Tell the school what is needed rather than what the symptoms are.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Tell the school what they can help.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>You should share the diagnosis with the disability support team at university / SENCo team, the leaflet should be given to them.</td>
</tr>
</tbody>
</table>

When comparing answers from both groups to the set questions, they were very similar. Both groups thought a separate booklet should be developed for each age group, it should focus on the essential things you would want a teacher to know and it should mention medications but not in depth. There were most conflicting thoughts for the tick page of symptoms, some believed it was too invasive, whereas others felt it might give information quickly at a glance.
Phase 4 “A reflection of where we’ve been and where still to go”

7.12.3 Focus group two

The prompt questions and responses of both groups in response to the mentorship questions are presented in Table 7-3.

Table 7-3 Mentorship questions for both groups and comments answered

<table>
<thead>
<tr>
<th>Questions</th>
<th>Children and young people</th>
<th>Parents</th>
</tr>
</thead>
<tbody>
<tr>
<td>1  Would it have helped you? / Would it have helped your child?</td>
<td>I hated talking about JDM, it made me different to everyone else</td>
<td>As a parent, it would be great to have known they had someone else to talk too, we have no idea how they feel day to day</td>
</tr>
<tr>
<td></td>
<td>It would have helped to have someone else to support and might give you hope</td>
<td>Wanted to know how I can help as a parent</td>
</tr>
<tr>
<td></td>
<td>I would feel listened to (feeling empathetic)</td>
<td>Parents need to step back, my daughter would have loved it</td>
</tr>
<tr>
<td></td>
<td>Be good to not fully need to explain yourself all the time</td>
<td>We met someone else for the first time after 8 years</td>
</tr>
<tr>
<td></td>
<td>Yes, someone who understands what you’re talking about</td>
<td>Face to face might be stressful for people with anxiety, maybe on technology especially for boys</td>
</tr>
<tr>
<td></td>
<td>More knowledge = more brainstorming</td>
<td>Move away from medical model and talk about coping</td>
</tr>
<tr>
<td></td>
<td>Helps with providing hope</td>
<td>Not all want to speak to someone, not everyone is ready</td>
</tr>
<tr>
<td>2  What age should the mentors be?</td>
<td>There are different stages of illness</td>
<td>Mentee 12-18</td>
</tr>
<tr>
<td></td>
<td>Similar age to the person they are mentoring</td>
<td>Mentor 16-25+</td>
</tr>
<tr>
<td></td>
<td>10-18 is a good age for the younger one</td>
<td>Not a lot of older ones out there</td>
</tr>
<tr>
<td></td>
<td>Younger children might also benefit from having a friend with JDM</td>
<td>Age might not matter but duration of JDM is important for matching</td>
</tr>
<tr>
<td></td>
<td></td>
<td>I wonder what stage of your journey you would think you need it</td>
</tr>
<tr>
<td>3  How often should they meet?</td>
<td>Why can’t they communicate outside of the mentoring times?&quot;</td>
<td>Need to have structured sessions</td>
</tr>
<tr>
<td></td>
<td>I get that there should be no contact outside due to it not being a part of their lives</td>
<td>What happens if you have an issue outside of these times?</td>
</tr>
<tr>
<td></td>
<td>Maybe you could hold onto questions for next call?</td>
<td>Would it feel like a psychology meeting?</td>
</tr>
<tr>
<td></td>
<td>I think you need both structured and non-structured meetings</td>
<td>Also might need informal support</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Should be offered at every clinic appointment by a health care professional</td>
</tr>
</tbody>
</table>
### Phase 4 “A reflection of where we’ve been and where still to go”

#### Table 7.3 Mentorship questions for both groups and comments answered

<table>
<thead>
<tr>
<th>Questions</th>
<th>Children and young people</th>
<th>Parents</th>
</tr>
</thead>
<tbody>
<tr>
<td>4 How long should a meet last?</td>
<td>Maybe 30 minutes</td>
<td>Once a week might be too often, not much happens in a week</td>
</tr>
<tr>
<td></td>
<td>I think little and often</td>
<td>Once a fortnight/monthly might be better</td>
</tr>
<tr>
<td></td>
<td>Kind of varies with each person</td>
<td>Might be better to be less strictly timed</td>
</tr>
<tr>
<td></td>
<td>The time should be decided with the mentor</td>
<td>Probably depends on stage of JDM, e.g. new drug may need more frequently</td>
</tr>
<tr>
<td></td>
<td>Have to be after school</td>
<td>Live chat might work better than skype for some who like meeting</td>
</tr>
<tr>
<td>5 What sort of training should occur for both?</td>
<td>Not feeling like your being spoken at – they need to be a good listener</td>
<td>Training is so important</td>
</tr>
<tr>
<td></td>
<td>They need to care</td>
<td>Training in how to listen and be natural, confidentiality, how to signpost for support, support for mentor too</td>
</tr>
<tr>
<td></td>
<td>Would help if it’s all confidential</td>
<td>How to deal with emotions and respond in a helpful way</td>
</tr>
<tr>
<td></td>
<td>Could advise on where to get help</td>
<td>How to give encouragement</td>
</tr>
<tr>
<td></td>
<td>Reassurance from someone would be good</td>
<td></td>
</tr>
<tr>
<td>6 Do you think they should be paid?</td>
<td>No, because they will care less</td>
<td>No, might get the wrong people saying they will do it</td>
</tr>
<tr>
<td></td>
<td>No, because they’d just be doing it for the money</td>
<td>Compensation might depend on the nature of the involvement</td>
</tr>
<tr>
<td></td>
<td>Paid expenses maybe?</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Depending on the set up of the situation</td>
<td></td>
</tr>
<tr>
<td>7 Should there be a set conversational topic?</td>
<td>No, there might be something else you want to talk about</td>
<td>Need to be really careful about what picture they paint</td>
</tr>
<tr>
<td></td>
<td>What is an appropriate question?</td>
<td>Need someone with same experiences</td>
</tr>
<tr>
<td></td>
<td>No, might want to talk about a bad day</td>
<td>What are the boundaries?</td>
</tr>
<tr>
<td></td>
<td>Freedom is better later on, after diagnosis</td>
<td>Safety?</td>
</tr>
<tr>
<td></td>
<td>Set topics might be better early on in diagnosis or for quieter mentees or early on in the relationship</td>
<td>No topic means you can talk about what the mentee needs</td>
</tr>
<tr>
<td></td>
<td>Needs to be flexible</td>
<td>Talking about medicines and side effects would be very hard, what happens in different experiences</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Could advise on job applications, getting rid for work</td>
</tr>
</tbody>
</table>
Table 7 3 Mentorship questions for both groups and comments answered

<table>
<thead>
<tr>
<th>Questions</th>
<th>Children and young people</th>
<th>Parents</th>
</tr>
</thead>
</table>
| **8** Who should be doing the training?              | Nurses
They should be given real life situations
Training needs to cover that those who have JDM are the experts
Psychologists, they can tell you what to look out for | Needs to be mentioned by a health care professional, not parents
Health care professionals should do the training
People with JDM would need to be involved too
Need training on how to deal with an appropriate question |
| **9** What benefits could come to the mentor?        | More awareness for JDM for both sides
Being able to have a connection with someone who understands
Would want to help someone else
You would be inspiring them                          | Big commitment and responsibility, is it too much responsibility?
Would need signed contracts
Also for the mentor having someone to talk to and feeling better themselves by helping
Make them more responsible                           |

The possibility for the mentor to be a friend was seen as a positive by both groups but the adults were more in favour of structured sessions. Both groups were not in favour of any monetary reward, although a good offer of paying expenses was suggested by the child group and both groups thought the training should occur by health care professionals and involve psychologists wherever possible.

The benefits to both mentor and mentee could be easily seen, however, the parent group did have some reservations, especially around safeguarding and bad experiences. They spent quite a lot of time discussing ‘safeguarding’ and how you would maintain safety of both mentor and mentee, but especially mentor. The adults could see how the older adolescent may come across difficult situations that they might not know how to manage. They also raised the issue about the importance of matching disease severity, whilst being mindful of bad experiences of side effects, for example if one mentor had previously had bad side effects with methotrexate, then you might not want them to mentor a mentee who had not experienced any negative side effects. When considering their own experiences, it was interesting that the majority of the group felt very passionately that this scheme should be offered by a health care professional, rather than
Phase 4 “A reflection of where we’ve been and where still to go”

through the parents. This insight into the dual (often-conflicting) role parents often face between parental ‘support’ and parental ‘health practitioner’ was interesting.

7.13 Discussion

“I’d love to of had the ability to speak to my eight year old self to help them realise it is ok, that they will get through it” (16 year old).

This comment encapsulated the mornings work. There had been a lot of talk about how hard JDM can be, but also much discussion about moving forward and how we could make things better for the future for everyone with JDM. There was a positive energy and buzz in the room. The discussion for this section will be split into the school booklet and then the peer mentoring, with a joint conclusion.

7.14 Outputs: School booklet

“During a focus group, a major concern for many teenagers was the impact their health has on their education”. (Rare Disease UK, 2019 p. 5)

Paediatric health conditions can affect education in many ways, for example frequent absences (due to treatments, flares or hospitalisations) can disrupt education and thus the child/young person begins to lose touch with their peers, resulting in feelings of isolation, struggling to keep up with work and feeling different (Barlow and Ellard, 2006). Despite the widely acknowledged importance of education, there is a dearth of research to examine issues of school absence, engagement, functioning and achievement in children with JDM. When caring for rare diseases, health care professionals become expert in extrapolating data from other comparable conditions. In this instance, looking to other paediatric rheumatology conditions such as JIA and the effects this condition has on schooling may offer some insight, all be it, contradictory. The first study by Sturge et al., (1997) found that school attendance was generally good, with a mean attendance rate of 92% whereas Bouaddi et al., (2013) found that whilst all healthy controls in their study were able to attend school, 33% of patients with JIA were unable to do so. Disease activity and functional impairment were the biggest factors affecting school attendance (Bouaddi et al., 2013).
Factors relevant to JDM were clearly talked about throughout phase one and two of this study and have been presented throughout this thesis. Children talked openly about feeling different to their peers and how absences from school made them feel: even from the side effects (the sickness) of their medications, they worried that they might need to miss school (as shown in the methotrexate poem) and the invisibility of JDM. Some of these findings are backed up in the literature, for example it has been discussed for many years that teachers make more allowances for children with stigmata than those with no visible disabilities (McAnarney et al., 1974). However, the biggest factor that came out of this current study in all phases, was the lack of knowledge that teachers have about JDM and the lack of a world-wide school resource. This latter concern was expressed by health care professionals throughout the study; from the international visit in July 2018 to SickKids Hospital in Toronto and the senior paediatric rheumatology consultant there describing the letters he writes to schools and again when asking the same question of nurses and medical professionals in phase three.

The literature supports the notion that further education and support of school staff is essential. In a study asking 161 parents to complete questionnaires about school experience, fifty-seven per cent of parents did not think that teachers had received sufficient education to be able to accommodate their child’s health need (Notaras et al., 2002). In a further study, school staff were eager to learn more about chronic diseases, and most respondents in this survey of 23 elementary schools in California, had requested additional information on health conditions of their pupils (Taras and Potts-Datema, 2005). Mukherjee, Lightfoot and Sloper, (2000) comments that it is important that school staff are given assistance with obtaining health-related information and ensuring health-related information is passed between and within schools. Children and adolescents with chronic illness therefore need school personnel who are willing to integrate their educational and health needs by collaborating with families and health care professionals (Thies, 1999).

If a school wide resource is to be published, there are areas of specific advice for JDM that can be recommended. Crucially teachers are best place to provide in-depth information about how a child is coping with their condition, after all, how a
child is functioning at school is often an excellent indicator of how they are managing in wider everyday activities and teachers and school nurses can provide clues on both the physical and psychological impact of JDM (Lloyd and McCann, 2013).

The discussions that ensued during the school focus group from both children, young people and the parents was very positive. Helpful ideas were discussed about how the booklet could be structured and what the content should include. There are a wealth of resources available for other health conditions and it would seem sensible to adopt the same format as many of these, rather than to start from scratch. This opportunity to ask the individuals who themselves are living with the condition and their parents who witness the daily struggles and have tips to share with others, was invaluable.

7.15 Output: Peer Mentoring

“I would like to inspire others. I know how hard it is to get through the day. As soon as you are diagnosed, your life changes” (18 year old).

Mentoring is not new, with An and Lipscomb, (2010) acknowledging that it can be traced back to when Odysseus placed his friend Mentor in charge of his son Telemachus while he went to fight in the Trojan War in the late 12th century. Peer mentoring though is a newer concept, with the majority of studies focusing on adult populations (Kohut et al., 2016). Bussey-Jones et al., (2006) define it is a mentoring relationship between individuals equal in age, experience and rank. When individuals share the same disease, peer support can encourage sharing of experiences to increase knowledge and skills of self-management (Sandhu et al., 2013). Peer support models have been used successfully in many adult health conditions including arthritis, cancer, lupus, HIV and diabetes (Sandhu et al., 2013; Williams et al., 2018).

With increased acknowledgement of the benefits peer mentoring can bring to adult health care, there is a new surge of interest in using peer mentoring for paediatrics. Kohut et al., (2018) define it as connecting an adolescent with chronic illness with an older peer with the same chronic illness who is successfully managing their own health condition. Young adult peer mentors may positively
influence health outcomes in adolescents with chronic illness by addressing feelings of isolation and loneliness, help to reinforce disease-management skills and promote a positive outlook (Kohut et al., 2014). Improved self-management is constantly referred to in the literature for the main goal to arise from peer mentorship training programs, such as:

- Enhance self-management of chronic pain in adolescents (Kohut et al., 2016).
- To improve self-management in paediatric Inflammatory Bowel Disease (Mackner, Ruff and Vannatta, 2014).
- Demonstrate higher levels of self-management in those adolescents who had received a liver transplant.
- To reinforce self-management of JIA (Stinson et al., 2016).

One can see therefore how peer mentoring can be beneficial when self-management skills (as well as autonomy and resilience) are essential in the transfer from paediatric to adult rheumatology care (Cai et al., 2019).

The rheumatology speciality is at the forefront in designing and implementing peer mentoring schemes, with a particularly active group in Toronto. Initially developed for chronic pain (Kohut et al., 2016) it was soon offered to adolescents with JIA (Stinson et al., 2016). The iPeer2Peer program provides individualised mentorship to sex matched adolescents, with ten meets over the course of 2-3 months via video call, with an emphasis on the adolescents leading their topics of conversation (Kohut et al., 2018). Full training is provided to the mentors and full support is given to both mentor and mentee. From this literature and discussions with the Toronto research group, questions were devised which were discussed during the focus group.

A limitation of a focus group is the inability to always probe answers further due to the group dynamics, so for example one of the young people had commented that they did not like talking about their JDM as it made them different to everyone else, but one does not know whether they would have liked to of talked about it with someone else who had it, or just whether they did not want to talk about it. Generally the peer mentoring idea was received favourably, but some interesting
Phase 4 “A reflection of where we’ve been and where still to go”

points were raised which will be useful when considering how to take this initiative further in the future.

In phase two we found that those who had met someone else, scored greater in the treatment and worry domains of the Rheumatology Module. This led to questions being asked about whether meeting someone else with JDM could increase an individual’s feeling of worry. However, as there is no formal mentoring scheme currently in the UK, most times when young people have met each other, they would have been sat in clinic waiting rooms, or at the few family days. These meetings would therefore have not have had the planned supportive discussions and focus on practical hints and tips which will be encouraged in safely matched up peer-to-peer sessions, such as those discussed here.

7.16 Implications for practice

This phase of the study concluded with energy and positive practical ideas of how future clinical care for children and young people with JDM, could be improved. Production of a school booklet for teachers would hopefully benefit so many future patients and lessen the anxieties children and young people have described facing at school. Through providing some peer support from a careful implemented mentorship program from others experiencing JDM, this could lessen the experiences of confusion, difference and uncertainty. Until this can be implemented, health care professionals could consider whether they could offer to put similar patients in touch with each other, if they feel this would benefit some.

7.17 Strengths

For this specific activity, the groups ran smoothly. The focus group for the school booklet was incredibly useful to finalise some of the work that had already occurred and for the charity groups to hear for themselves how beneficial this would be. The peer mentoring focus group was initial brainstorming and to understand if children, young people and parents thought this might be an idea that would have helped them, and would be beneficial in the future. The JDM families were keen and eager to participate, and wanted to get their voices heard.
Phase 4 “A reflection of where we’ve been and where still to go”

7.18 Limitations

There are limitations with running any focus group as has already been highlighted. The biggest limitation was the timing restrictions, which meant that conversations had to stop, when they were often in mid flow. A further limitation was the other attendees who had wanted to attend, but then could not. Running only one small workshop is limiting in itself, and had it been possible, being able to have more conversations, would have perhaps produced a different set of data.

7.19 Conclusion

The workshop had good engagement from the children, young people and their families. The charities and staff who attended provided support, assistance with the organisation and practical delivery. The aims were met of disseminating the findings with the wider JDM community and key stakeholders and ideas for the future have been usefully discussed and debated. Whilst there is still significant work left to be done; the school booklet to finish and publish, and further peer mentoring discussions to continue after this thesis, this phase has highlighted some real concerns from children, young people and their parents. Schooling is such a large part of most children’s lives, and for so many to feel such a lack of understanding and empathy, is worrying. Health professionals, parents and stakeholders need to work together to improve this for the current JDM patients, but also for the future. Assisting young people to gain support and understanding from other young people, should lessen the confusion, uncertainty and feeling of difference, which so many people have talked about throughout this research.

7.20 Next chapter

Chapter eight is pivotal in drawing the journey to a close – for now! To reflect upon the path undertaken, the limitations and the strengths of this work as a whole, but most importantly, the implications for children and young people with JDM.
“Invisibility”

Not allowed to do pe as they were cautious of me,

No wall climbing, or strenuous activity,

I can’t do the stuff I want to do,

They don’t understand, they have no clue,

I don’t want them to judge me on my illness,

Leave me behind, in the wilderness,

The same thing 24/7 I have to explain,

Tell them what it is, again and again,

In the beginning, red food colouring spilt all over me,

Red on knuckles, elbows and my knee,

They could see I was different,

My limitations apparent,

Now as times goes on, not visible on the outside,

But hidden away on the inside,

A disease that could kill, that’s secret power is invisibility,

A disease with no cure, but long standing emotional disability.
Chapter 8: Discussion, integration, contribution and reflections “Are we nearly there yet?”

8.1 Introduction

“Integration necessitates more than simply assembling different types of data or empirics and aggregating them together; rather, it requires stitching together a rich tapestry of explanation and in doing so discovering those theoretical connections that bridge diverse empirical insights” (Tunarosa and Glynn, 2017 p. 237).

In this last chapter I will close the hermeneutic cycle of interpretation from this research study, bringing together the four phases which explored the lived experience and psychosocial needs of children and young people with JDM. As the above quote highlights, the aim is to stitch together all the parts of the study, to make the whole and discover the theoretical connections. The integration of these parts and synthesis of findings is core to the mixed methods design. The chapter begins with an overview of the study, with a discussion of the findings from each phase. Following on from this, I will consider the contribution to new knowledge this study brings and as important, the implications for future clinical care and research. The chapter will conclude with the dissemination strategy, the limitations encountered and a personal reflexive perspective.

8.2 Summary of the study

This was a mixed methods exploratory research study, exploring the lived experience of JDM in children and young people between the ages of eight and 19 years of age. The uniqueness of this study stems from the collaboration with children and young people, involving creative work to collect their oral narratives and personal experiences. Whilst there is a wealth of literature examining psychosocial needs in other chronic childhood illnesses, I have demonstrated that there is a paucity of published work in JDM. This is the first study to explore lived experiences qualitatively and quantitatively in a large number of children and young people with JDM using a phenomenological lens to understand and interpret this from their perspectives.
8.2.1 Phase one

Research question:

- “What is the lived experience of JDM for children and young people?”

Fifteen children and young people between the ages of eight and 19 were interviewed using the methodology of hermeneutic phenomenology. This was aided by the use of creative methods. Their rich narratives were crafted into stories, sharing their lived experiences of JDM. From these data, the metaphor of a rollercoaster was developed, with five themes that were significant and present across the voices. Being-on-the-JDM-rollercoaster phenomenon illustrated the journey that the children and young people described, highlighting the difficulties and benefits experienced along the way. This simple and visual representation embraced all the parts of what it is like to live with JDM, and was later discussed and explored with some of the participants.

8.2.2 Phase two

Research question:

- “What variables are most important from phase one and are there any other psychosocial needs expressed as important in a larger cohort of children and young people with JDM?”

For phase two, questionnaire content was chosen based on the themes from phase one, and were supported by a bespoke JDM experience questionnaire. The results from the 123 responses supported impaired quality of life compared to a normative population, but less so, for emotional distress. However, 40% of the respondents did score over the cut off for emotional distress that necessitated further clinical assessment, which was a concern. Uncertainty and a perception of burden were raised by the young people in the study with significant findings when correlated to quality of life and emotional distress. However, acceptance, measured by the benefit questionnaire did not return significant results. Qualitative comments revealed further areas of concern for participants,
Concerns about school, and salient points regarding peer support from others with JDM: whether young people have had any, and whether they would like more.

8.2.3 Phase three

Research question:

- “What psychosocial support is provided in local centres and what are their perceived challenges?”

Phase three; a survey of health professionals from 15 centres across the UK investigated psychosocial support given to children and young people with JDM. Each survey was specific to the professional group; medical, nursing or psychology. The clinical set-up - whether there are regular clinics which JDM patients attend and regular meetings where these patients are discussed, differed between paediatric rheumatology centres. Psychology service provision was found to be related to funding provided to the rheumatology service by the individual Trusts, with one centre having no provision at all, and some only limited overall Trust provision. The level of psychology provision was generally described as the biggest challenge, but despite this, the majority of health professionals (41%) scored their service, four out of five on a Likert scale.

8.2.4 Phase four

Research question:

- “How can we take these findings forward from this research so far, and improve psychosocial care even further in the future?”

A workshop was organised for all interested parties from across the journey of this research to attend. This workshop comprised of two focus groups. The primary aim was to disseminate the findings from the research and to thank participants for their engagement and support. Secondly, the two future research ideas (production of a school resource and peer mentoring) were discussed and some group work informed how these could be taken forward. Both of these future interventions were well received by participants, which included parents
and key members from key charity groups, as well as children and young people with JDM, their siblings, and some staff members.

8.3 Integration

As discussed in 2.3.1 this research used a mixed methods design with one primary purpose – Development, and one secondary purpose – Complementarity, both of which strengthened the study (Bryman, 2006). When considering the integration of the findings for this research study, these both served a specific purpose:

- Development – this study was incrementally built, with the findings from the prior phases shaping the thinking, and the focus of questions for the subsequent phase. This development was pivotal to the design of this study and ensured that the integration featured at every step.
- Complementarity – ensured that the findings from each phase were constantly considered and reflected upon throughout the study. As the study grew richer in participation and engagement, continuous reflection, thinking and discussion, led to new ideas and new concepts which complemented the preceding phases.

The data integration discussed here, used a technique or process Moran-Ellis et al., (2006) termed ‘following a thread’ where datasets are analysed inductively within its paradigmatic parameters, then analytic questions or themes which emerge from this dataset are followed up in others, with the overall aim of interweaving the findings that emerge across all datasets (Moran-Ellis et al., 2006). Using this defined process of integration adds credibility, and ensured that a process to simply mix and match methods was avoided (Dupin and Borglin, 2020).

This integration was therefore two fold, first using the data from one phase to shape another, second to identify key themes requiring further exploration in a previously analysed data set (O’Cathain, Murphy and Nicholl, 2010).
8.3.1 Following the thread

Using this process, the lack of school support and peer support, emerged as the threads to follow. School support was mentioned on many occasions during the interviews. For some participants, school support had been good, and young people described how the school had tried to help them: for example, providing the little garden for break time recreation, encouraging the application of sun cream, providing a chair during assembly. For others, they had voiced their lack of support from school: "the teachers think I am faking it".

Peer support was two-fold; current peer group and those peers also with JDM. Children and young people had shared difficulties with friendships and some had shared personal stories of bullying. Others talked about lack of understanding from their friends and some confided that they were not able to disclose their JDM to their peers. There was a consensus that having good friends, especially when managing the JDM was beneficial and an understanding friend who contacted you when you were absent from school, was indeed a useful ally. Taking this a step further, children and young people talked about feeling alone. As their current peer group could generally not understand what it was like to have JDM, they would like to talk to someone who would understand. Most children and young people had not met anyone else with JDM, and this confounded their feelings of difference, confusion and uncertainty.

These concerns were considered in the findings from phase two and helped shape the survey developed for phase three, asking health professionals about the information they gave to schools and what opportunities they offered for peer support. Both of these threads thus became the focus for the final fourth phase of this research. This process of following the thread, and revisiting early phases is illustrated in Figure 8-1. This depicts the journey taken, showing each phase and the thinking that emerged, and the revisiting of earlier data to pick up earlier threads.
8.3.2 Adapting the rollercoaster in light of the new threads and new knowledge

This research has been shaped and developed as it progressed. Throughout the journey, the path taken has been mapped out from the places visited before. Thus, thinking around the earlier themes and the rollercoaster has also developed. The metaphor of the rollercoaster came from the phenomenology
interviews in phase one. It therefore cannot be changed, but it can be built upon with the new knowledge acquired from subsequent phases.

From reflecting and revisiting earlier data, the new threads of school support and peer support have been woven through the rollercoaster metaphor. Positioning a spotlight on either or both of these, will have implications for the day-to-day lived experience of those with JDM.

Children and young people have also talked about time points when certain support was more beneficial, and this had a clear influence: family in the beginning, peers during the adjustment period and the young person themselves and how they ultimately learn to live with their JDM over many years.

Reflecting on the qualitative comments in the questionnaires, the interaction with patients by health professionals and the views from those who attended the workshop, the rollercoaster predominantly encompasses three time points or stages of a chronic disease process. These have been termed and defined as:

1. Unfolding - time of diagnosis (0-3 months approximately).
2. Adjusting - the period of time after diagnosis when an individual learns to adjust to their 'new normal' as the disease requires (approximately 3 months - 3 years).
3. Living - the time when an individual and the disease are not separable, but living and working together (3 years plus).

Separating the rollercoaster into these time phases allows for clarity and a more useful model to inform practice. The rollercoaster itself remains unchanged, and the themes untouched, but now the spotlight is focused on the three time points and the three main processes that the children and young people throughout the study had referred to. This new adapted rollercoaster is informed by all four of the study phases and provides a novel portrayal of the lived experience of children and young people with JDM (See Figure 8-2).
8.4 What is the contribution from this research?

Within each chapter, there has been a discussion of the findings for that phase and the associated implications. This section ties that discussion together to bring the conversation full circle and conclude with what is perhaps the most important question to answer, what is the contribution this research makes?

The overarching research question which began this journey asked: “What are the psychosocial needs of children and young people with JDM?” This question was borne out of clinical practice, from watching children and young people suffer and from talking to them and their families about their real-life experiences. I expected to find that children and young people with JDM were struggling. However, the results from this research did not corroborate that early perception. The majority of children and young people with JDM around the UK are in fact doing well, with lower emotional distress levels than even a healthy, normative population (5.9.6.3) and lower levels of uncertainty than a sample of children with rheumatic disease (5.11.1). The questionnaire results showed that 81% of children and young people responded positively to having enough help and support with their JDM (5.9.10.7) and the majority of health professionals surveyed, believed their centre was providing close to an excellent psychosocial service (6.8.7).
The unique contribution however, lies in the centrality of the child’s voice. Prior to this study, we did not know how children and young people coped with their JDM, who was doing well and who was not, because we had not asked them directly. This research was the first to ask them, using different methods, then listening to what they wanted to say. We can be confident that for the participants in this study, most are coping well. However, some were struggling, with 40% of the participants surveyed scoring over the clinical cut off, for emotional distress. Indeed, this finding alone supports the need to keep on asking children how they are, and finding out how are they getting on. One of the medical professionals surveyed in phase three declared:

“We don’t ask enough about mental health in our Connective Tissues Disease patients, too much focus during clinic is on their physical issues”

There could be many reasons why we do not ask enough. Time, for example, was declared as the second biggest challenge, from the surveys and this was acknowledged by all three of the health professional respondents. Lack of awareness could be another, as a medical professional said in Chapter four, after listening to the worry poem, he would now be talking to all children and young people with JDM. ‘Fear of opening a can of worms’ could also be a concern in those centres with limited psychology support, as one psychologist commented:

“we are only able to offer a very brief assessment and intervention service”.

If the contribution from this research encourages more health professionals to find the time to ask about psychosocial needs, then this research has been worthwhile. A simple mnemonic may help health care professionals in clinic remember the findings from this study and the importance in asking children and young people directly, rather than their parents. A well-known mnemonic used for adolescent health in transitional clinics, is HEADSS (Homelife, Education, Activities, Affect, Drugs, Sex, Suicide, Sleep), but here, friendships are not asked about in their own right, but instead come under the A for activities (Christie and Viner, 2005). I am suggesting that the word SAFE is more appropriate to the findings from this study and may be particularly useful with younger children, when you would not generally need the ‘D’ for drug use or ‘S’ for sex in HEADSS.
The mnemonic SAFE is specifically relevant to support, and different kinds of support, with each letter being represented:

S  Support?
A  Ask....
F  Friends?
E  Education?

This whole study has illuminated the importance of friends and peers, and their role in helping someone cope with the impact of JDM. If children and young people do not feel able to share their diagnosis with their closest friends, perhaps health care professionals can suggest ways to do so, to increase the support they receive. Maybe, this is where the importance of being introduced to someone else with JDM plays a particular role, in helping those children and young people to feel less confused and different. Likewise, asking about school support may identify areas of lack of support or understanding from teachers, and from talking these through, suggesting useful measures to improve the current situation, may be beneficial. When I asked the two children and young people in this study if their parents were aware that their friends did not know about their JDM, they both believed they did not. Again, this highlights the importance of taking the time to talk to the child or young person.

Taking this a step further and offering up the rollercoaster as a resource to help visually illustrate the JDM journey may also benefit some children and young people. The rollercoaster can help at diagnosis to map the progress of the disease, to highlight feelings that have been expressed, which if acknowledged and addressed, may improve the journey. Whilst metaphors of rollercoasters have been used in other published literature, this rollercoaster is unique. In 3.9, I said that not all children and young people need to reach acceptance, but being able to live with the JDM is an end-point we want each young person to reach.

This study has presented many findings, and each phase of data collection has told a part of the story, but together they make up a fuller picture. By addressing the confusion; the uncertainty; the difference; the side effects; the perception of burden; the lack of school support and peer support; - we can help people live with their JDM.
8.5 Recommendations for clinical practice

This research found that whilst a significant proportion of children and young people with JDM are coping well with their JDM, there is a small proportion who are not. However, to date neither this research nor the wider literature has ascertained a way of predicting which children will struggle, which will do better, or do worse, using the clinically available information available to clinicians. If further studies could generate reliable predictors of the subgroup of children who will struggle to target them for more support, this would be a valuable addition to use in clinical care. This has implications for practice where the absence of psychosocial needs identified during diagnosis, either in hospital or a clinic, should not negate the need for regular assessment and early intervention later on in the disease course. As highlighted by this research, JDM is a disease with periods of flares and remission, and this changeable course means that individuals can face different challenges at different times.

A key finding of this research is that uncertainty can have implications for impaired quality of life and raised levels of emotional distress. In clinical practice, all health professionals would benefit from being equipped to deal with psychosocial care and may be able to support children and young people by providing psychosocial assessment and interventions early to reduce the implications from unaddressed uncertainty. Medical professionals and nurse specialists who may have more contact with these patients than other health professionals must also be aware of the unavoidable uncertainty that is inherent from living with JDM (the unpredictable flares), but can prepare the child or young person for these and recommend coping and behavioural strategies to manage these (e.g. pacing, planning activities in advance and managing exam stress).

The rollercoaster can be a beneficial visual aid for health professionals to use in clinic with children and young people to prepare them for the inevitable ups-and-downs of their chronic disease course. All clinicians are encouraged to identify in partnership with the child or young person before them, where they are on the rollercoaster. It would be futile to be discussing how to independently manage JDM, if the young person is still in a period of adjustment. Further recommendations from each phase are in Table 8-1:
Table 8-1 Recommendations for clinical practice from each phase

<table>
<thead>
<tr>
<th>Phase</th>
<th>Unfolding</th>
<th>Adjusting</th>
<th>Living</th>
</tr>
</thead>
<tbody>
<tr>
<td>Phase one</td>
<td>1. Need to provide clear developmentally appropriate information on JDM from the beginning to lessen some of the confusion.</td>
<td>1. Need to (frequently) offer opportunities for peer interaction, which would lessen some of the feelings of difference, loneliness in dealing with medication side effects and the daily uncertainty.</td>
<td>1. Whilst young people may have had their disease for a number of years, and flares may be less, ongoing support is vital in this stage to aid coping with the unpredictability of JDM.</td>
</tr>
<tr>
<td>Phase two</td>
<td>1. A diagnosis of JDM can have huge implications for young people and profound implications on their perceived quality of life, their perception of their quality of life should be explored with them in a supportive and constructive manner at every opportunity.</td>
<td>1. Young people should be counselled about the visible signs of JDM and the side effects from steroids, right from an early stage. If required a dietician or psychologist should be involved to provide healthy eating advice and to provide coping strategies and an opportunity to disclose any bullying from peers.</td>
<td>1. Young people need to be included in discussions surrounding their condition and whether it is in remission on therapy or in remission off therapy and the implications of this for them. This may help with their adjustment, coping strategies and ability to accept their JDM as they transition into adult care.</td>
</tr>
<tr>
<td>Phase three</td>
<td>1. More psychosocial preventative work, at the time of diagnosis, providing written information about JDM and medications.</td>
<td>1. Provide education around adjustment and developmental challenges along the way to the young person, but also to other health providers with a more united, joined up approach.</td>
<td>1. For all health professionals to continue to ask young people whether they have any psychosocial needs at each opportunity to allow for intervention at an earlier stage.</td>
</tr>
<tr>
<td>Phase four</td>
<td>1. Signposting children, young people and their families to places of information and support would enable families to maximise their support of each other, increase their understanding and thus lessen their confusion.</td>
<td>1. Clear, appropriate, up to date and relevant information needs to be regularly provided to schools with reviews on support provided to ensure young people do not feel negatively different to their peers, but still with understanding and support from teachers.</td>
<td>1. Offering young people a chance to have a peer mentor would increase young people’s understanding and resilience living with JDM, but also offering the opportunity to be a mentee would enable older adolescents to share their life skills and tips of JDM.</td>
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</table>
8.6 Recommendations for Future Research

There are four main future research areas that have emerged through this work:

1. How does current disease activity affect psychosocial needs?
   This research question examining the relationship with clinical variables is important to understand how the current disease state adds into the findings from this study.

2. What is the role of peer support to those with JDM and how can we improve upon it?
   This study has demonstrated the value that children and young people gain from having good friends, but how can we assist them to share their diagnosis and to feel ‘supported’ rather than ‘different’.

3. How well equipped do nursing staff feel when providing psychosocial care to children and young people with JDM when services do not have psychology support?
   Children’s nurses do not receive much specific mental health training during their education, however, as the nurses in this study explained, often they are providing mental health advice. Through an intervention, such as a short module, this could be improved.

4. Is unpredictability of the disease or a lack of comprehension more important whilst living with the uncertainty from JDM?
   Further research into uncertainty and the role of unpredictability of disease compared to uncertainty from lack of knowledge and understanding in JDM, could be explored further.

8.7 Strengths of this research

This research is unique. It asked a population of children and young people about a particular rare disease they were experiencing to share their stories. Specifically, it gave them an opportunity to talk about their psychosocial needs. What emerged from phase one was rich, powerful and emotive stories, from
interviews with children and young people. Creative methods were offered to assist children and young people to tell their stories, but not always required, as some just wanted to talk. The creation of research poems and the visual construction of the rollercoaster add to the understanding of the lives of those with JDM and provide rhyme and visual imagery to help explain the phenomena. Over 100 children and young people responded to the questionnaires, providing a voice from most corners of the UK.

The use of mixed methods broadened the research questions addressed, and deepened the inferences identified. Using techniques drawn from the qualitative and quantitative traditions of research meant that a comprehensive picture of the experience of JDM could be presented. The pragmatic grounding of the study enabled me to use the methods which would best answer the phase specific research questions, to develop a comprehensive analysis of living with JDM. The psychosocial needs of children and young people with JDM, would not have been obtained in such detail had a single method been used. Hermeneutic phenomenology afforded time, space and completely absorption in the interviews to interpret the meaning of the stories being shared. Following this, the predominantly quantitative questionnaires allowed the hypotheses developed from phase one to be tested. The later surveys invited a different perspective and the workshop allowed meaning found in the questionnaires to be critiqued and considered further. This research was comprised of four studies and each demanded new ways of thinking.

8.8 Limitations

There were several challenges in this research, many of which I have already discussed to some degree in previous chapters. Facing challenges was perhaps not surprising since I brought together many different methods to explore experience in different ways.

The biggest limitation of this study however, is the inability to compare the results found with current disease activity. For example, exploring whether our higher levels of uncertainty were found in those children and young people with the most active JDM. Previous research has identified psychosocial difference when
Discussion, integration, contribution and reflections “Are we nearly there yet?”

Comparing family impact of JDM, with those who had a child in remission showing improved family functioning, whilst having a child with active disease negatively affected a parent’s mood (Kountz-Edwards et al., 2017). These results, are understandable when considering those in remission, with currently inactive disease, compared to those who have recently been diagnosed.

Table 8-2 depicts overall limitations:

<table>
<thead>
<tr>
<th>Limitation</th>
<th>Solution for future research</th>
<th>Rationale</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age range of participants</td>
<td>Capture those under eight years of age and over nineteen years of age</td>
<td>To provide a more comprehensive picture of JDM in the UK</td>
</tr>
<tr>
<td>Cross sectional data</td>
<td>Longitudinal data collection</td>
<td>To identify changes due to illness trajectory and life experiences</td>
</tr>
<tr>
<td>Young people interviewed only from one centre</td>
<td>Multi-centre interviews</td>
<td>To detect any differences due to location and services provided</td>
</tr>
<tr>
<td>Use of validated questionnaires</td>
<td>Create a specific measure from these study findings</td>
<td>To ensure accuracy and complete relevance</td>
</tr>
<tr>
<td>Lack of clinical data</td>
<td>Include patient clinical notes for accurate and timely disease status</td>
<td>To be able to draw comparisons between patient reported outcomes and disease status</td>
</tr>
<tr>
<td>Only three types of health professionals included</td>
<td>To include other health professionals such as Occupational therapists and School teachers</td>
<td>To provide a more comprehensive picture of support provided by other professionals to meet needs</td>
</tr>
<tr>
<td>Not including siblings or parents</td>
<td>To include the whole family's perspective at every step of the research</td>
<td>To understand how the needs of the family, and the support they provide, play a part with coping with JDM</td>
</tr>
</tbody>
</table>

8.9 Dissemination

This study has previously, and will continue to, utilise multiple dissemination methods, as presented by the dissemination strategy in Figure 8-3.
8.9.1 With patients and families

Patient engagement with children, young people and their parents has been presented in Chapter seven. Thus, patient dissemination so far has been on a number of levels, through presentation at three JDM Family Days (two in London and one as an invited speaker to the first ever Northern JDM Family Day), through the workshop held and comments sent from parents who discussed this work on a parental closed JDM Facebook group®. I have also presented work at two national charity days in Oxford organised by Myositis UK, attended by children, young people and their families.
8.9.2 With health care professionals

The sharing of the rollercoaster phenomena is essential to increase the awareness of JDM and the impact it has upon the psychosocial well-being of children and young people. The findings from this research have been presented at a number of national and international meetings, with a range of audiences. These oral presentations given to date, whether as an oral lecture or an oral poster presentation, and who the target audience was, is presented in Appendix 28. This work has also been published in *Pediatric Rheumatology* (Livermore et al., 2019) (Appendix 29) and *The Journal of Poetic Therapy* (Livermore, Wedderburn and Gibson, 2020) (Appendix 30) with a further paper ready for submission, and another in draft form.

The poetry work has been presented in poster format by producing voice recorded cards and attaching to a poster. This meant that the audience could open the greeting cards and hear the poems, spoken by children and young people themselves. This turned out to be a very powerful way of sharing the poems and won first prize at the University Open Day and the Students Choice prize award.

8.9.3 With key stakeholders

This work has led to close relationships with a number of key stakeholders. Charity groups have shown interest in this research and been keen to engage. I was invited to a Versus Arthritis Family Day and invited to speak and run workshop sessions at two Myositis UK Family Days. This collaboration has led to discussions regarding the school resource and may lead to the shared publication. I was also invited to present this work at a staff event at the leading specialist medical society for Rheumatology and Musculoskeletal professionals; the British Society of Rheumatology (BSR). This aim of this session was to share paediatric rheumatology research, with a particular focus on the novel and powerful poetry outputs. During this session, we discussed the need for a policy statement for JDM, of which they would be pivotal in helping to implement.

In March 2020, as all the research phases had ended, I was invited to present the findings in a live hour-long webinar, hosted by the Arthritis and
Musculoskeletal Alliance (ARMA). This lecture had over 100 participants in the virtual audience, (including patients, parents, health care professionals and stakeholders) and five months later has already had 383 views on a freely available social media platform.

8.10 Reflexive perspective

The first chapter of this thesis introduced myself and my path taken up until the commencement of this research. Whilst this path spanned over 20 years of clinical nursing, my growth over the last three years has greatly exceeded those years before. It is therefore fitting to end with a reflexive stance, to illustrate this succinctly, to you, the reader.

From the onset of designing the study to the point of analysis a number of challenges had to be overcome. These included: the choice of psychological measures, considering psychology models, learning quantitative methods, integration of qualitative and quantitative data and ensuring rigour in the data analysis. Some of the challenges were unavoidable, while others could have been foreseen.

Throughout design, data collection, analysis and write up, I have kept a reflective journal to understand how I may have influenced this research and I have exercised constant reflexivity to consider this further. My prior understandings, beliefs and way of viewing the world, all contribute to the findings found.

Some parts of this research, I have found harder than others. Three of these in particular were:

1. Considering a psychological model to base the work upon.
2. Adding in the quantitative element.
3. Balancing the phenomenology.

After phase one, conversations with my supervisory team turned to what questionnaire methods to subsequently use. Many psychological models were suggested, all I which I researched and considered, however, none of these
models were a good fit to the findings found so far. Discussions continued about whether I should have used a model from the outset, as then this process would have been much easier. I am not a psychologist, and many of these models were complex to understand. Frequent returning to the aims of the study and the importance of the phenomenological beginnings of starting with a blank canvas and letting the children and young people shape the priorities was vital. This was what made this study unique, and thus no model or previously validated questionnaire would address perfectly what we had found. There needed to be some compromise and find the best-fit model and questionnaires, rather than the perfect questionnaire.

The quantitative element was a very steep learning curve, but one that has ultimately benefitted this study. Mixing the qualitative and quantitative has also been hard at times, and to keep the phenomenology in its place has been a challenge.

Other challenges include: the tools which did not perfectly capture the findings we had found, the enormous emotional impact of listening to young people’s heart felt stories, and such a huge piece of research with so much learning and writing in a time of an international pandemic, whilst trying to home school three adolescent boys!

There have been many positives. The creation of the poetry and watching the reactions of those who listened to them has been a particular highlight for me. Time and space to read thousands of papers, write some of my own and build up a long list of networks and collaborations has been a privilege, as has the opportunity to present this work, specifically at the international conferences: Berlin, Lisbon, Toronto and Atlanta. However, the most important impact on me has been the children and young people and the experiences they have shared.

I began this journey as a novice researcher and have ended as a Clinical Academic. Now in a role as ‘Clinical Academic Lead for Nurses and Allied Health Professionals’, I can help support others to complete their journeys. The thesis road map ends this thesis and visually illustrates my journey, but now with all the additions that happened along the way Figure 8-4.
I would like to finish with one more poem, to bring the work full circle, that is the words of a young person aged 14 years:

“Powerful”

When I first got JDM, I felt really sad about it,
I didn’t understand about it,
I was confused about it.
But now adays, I embrace it,
I feel more powerful that I have it,
I can be very strong, and I didn’t even know it,
It’s a good lesson, because I will be ok in the end, with it.

This poem describes the research that has come out of this study. It illustrates the tough times, but also shows there is light at the end of the tunnel.
Discussion, integration, contribution and reflections “Are we nearly there yet?”

‘Psychosocial Needs in JDM’
Roadmap through the thesis

Research question: What are the psychosocial needs of children and young people with Juvenile Dermatomyositis to identify their unmet psychosocial needs?

Chapter 1 “Mapping the Journey”
- ATTENDANCE AT PHENOMENOLOGY CONFERENCE
- “The best method of transportation” mixed-methods, sequential exploratory design

Chapter 2
- SICK KIDS HOSPITAL VISIT & PRESENTATION, IN TORONTO

Chapter 3 “Beginning the journey”
- YOUNG PEOPLE’S REFERENCE GROUP
- POETRY THERAPY
- “Creative detour” poetry

Chapter 4 “Picking up more passengers”
- 3 MINUTE THESIS COMPETITION – THROUGH TO 2ND ROUND
- POETRY SHOWCASE IN BERLIN AT GECM
- PHASE 3

Chapter 5 “A reflection of where we’ve been and where still to go”
- THE VIEW FROM OTHERS ON THE JOURNEY
- 1ST PRIZE POSTER & STUDENTS’ CHOICE PRIZE

Chapter 6 “We nearly there yet?”
- UNITED SPEAKER AT PHENOMENOLOGY CONFERENCE

Chapter 7
- DISCUSSION & CONCLUSION

Figure 8-4 Completed roadmap (for now!)
“JDM”

Joints, Damage, Muscles

Jolly-well Disappear Musclepain

Jaded, Depressed Moody

Just Disappointing Me

Join Doctors & Mum

Joke, Dumb Mylife

Journey, Diagnosis, Mess ..... 

.....Anything but Juvenile Dermatomyositis


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Reference List


Appendix 1 – An example of the reflexive diary

Sunday X July – Myositis UK Charity day in Oxford – I had been invited to speak

I was very nervous about attending the Myositis UK Family day, not least because I needed to drive there and had offered to drive CD, and I hate driving!! The journey there was fraught and stressful (I had no idea where we were going) and to make matters worse, I really did not know what to expect when we got there. We had been warned that whilst this is a Charity that supports Adults with Dermatomyositis and Paediatrics with Juvenile Dermatomyositis, there are very big differences between the two groups. There are a lot more adults, and because the disease is so very different (especially with the high cancer risk), and most are in wheelchairs. Generally, there are only a few paediatric families who attend as they the parents do not think it is a positive message to give to their children, so there may only be 2 or 3 families there. As it turned out we were welcomed into the group and shown to the room. We had 4 families and 7 children in total. CD gave her talk first and had many questions about autoantibodies, it was an amazing talk, but this only left 20 minutes for my talk (part of me wanted a long time and part of me did not!). I was anxious as the children would be in the room and some listening, but generally I think the talk went ok. Now with Kate’s concerns ringing in my ears I set the scene that the parents could talk to their children over lunch about whether they want to trial any activities, but it was totally up to them. After lunch when we returned, the young adult who had JDM, took over and offered the arts and crafts to the group. While the families were not too sure what to do, I felt it was important that I leave them, rather than guide too much.

One adolescent boy did a good day and bad day body map, but needed lots of encouragement from his father. This was really interesting for me to think about as it was actually his father who was encouraging him to remember a time when he had been in a flare, or been in hospital and how he felt. One of the things that came out for him was different terminology and understand it, e.g. he didn’t know
Appendix 1 – An example of the reflexive diary

what subcut meant, and it seemed a big concern was his medications – although he thought he was now well and had no concerns. The final products had quite a few overlaps on them, for instance on a good day he had put – “Doesn’t bother me”, “could be worse”, “I feel I can do the things I want”, and on a bad day he had written “why me?” “Can’t be bothered to do much”.

A young girl also became very interested with the comics and pretty much drew and wrote her own, with only some help with spelling the medicines needed from her mother. She filled eight pages about “How I got JDM and how it makes me feel”. Generally the text was positive, but she had written “when I have my medicine it feels like I am trapped in a jail cell”.

The session finished on time, and then after tea we went into the joint final session. This ran over and I was anxious about getting home to the family, so CD and I left slightly early. The journey home was awful with the gears going on the car and then the brakes alarming that they had failed the whole way home, I got home a complete nervous wreck!

A very stressful day, but I had some important thoughts after the day:

1. Whilst it was a very stressful day, I was glad I had gone and given up my time. It gave me a chance to talk to families of those with JDM from other centres around the UK, and to listen to the families and to think about what things they all experience. All of the families said they thought psychosocial concerns were a very important overlooked part, apart from psychology help with needle phobia (seemed to be a shared concern), none of them had otherwise seen a psychologist. The family of the boy actually said to me “we work so hard with the physical side, you know the physio, but we’d not thought about the emotional side, we’re not trained to address these issues”. Also, this family, whilst living outside London, really wanted to be part of the project and said that they would love for me to
Appendix 1 – An example of the reflexive diary

Interview their son. They were fed up that they couldn’t be as they are not a patient at our tertiary centre in London. I did say I would try hard to keep them involved in any way I could, and they took my number. Interestingly though, they were very keen, but their son looked less impressed to be involved. I wonder if I will come across this more often.

This family also raised the issues of why can’t we capture this on an app, as this age are really into their technology. Need to see if there are any apps that may be appropriate and easy to bring in to use.

2. The role parents say in coaching, triggering memories and generally encouraging.

3. Whether the tools will bring out the experiences or whether they might be too fun. Both the body maps and comic book did yield some interesting results, is this enough?

4. My training and background as a nurse has always been to turn things around and look at the positives and to try and pick people up and encourage them to see the best things. However, for this project, I need to be able to get young people to talk about their experiences and feelings – good and BAD, can I do this?? Does this go against what I’ve always tried to do? I am not a psychologist, is it wrong to dabble in these areas without the proper training, or is that the whole point of the project?
### Appendix 2 – A section of the ‘Creative methods critique’

<table>
<thead>
<tr>
<th>'Draw, write &amp; tell'</th>
<th>Advantages</th>
<th>Disadvantages</th>
</tr>
</thead>
</table>
| 'Draw, write and tell': a literature review & methodological development on the ‘draw and write’ research method (Angell, Alexander and Hunt, 2015)  
-a literature of studies utilising this method was conducted during the planning stages and noted a range of benefits of ‘draw and write’ in enabling child participation, however it also identified that the method had been used inconsistently and found issues with interpretation of creative work and analysis of data  
-as a result the entitled ‘draw, write & tell’ method was developed in an attempt to provide a more child-orientated & consistent approach to data collection, interpretation & analysis  
-got some good background to CYP research & draw & write background from 1972 Wetton | -the use of both drawing & writing might result in ‘richer’ data  
-enables participation across broad range of ages and abilities, both acceptable & enjoyable, being a non-threatening means of eliciting ideas even when a difficult subject  
-as a means to reduce the power differentials between CYP and adults because it enables CYP to guide the research exercise  
-in this study picture, text & spoken all combined into a ‘written commentary’  
-draw and write as a participatory method is consistent with a belief that CYP are unique, thinking individuals & capable of expressing their thoughts and emotions, as such is congruent with an approach that respects CYP at each stage and  
-hard for CYP (children and young people) to decline participation if conducting in schools  
-CYP might reveal more in their drawings than they want to  
-maintaining confidentiality in the publication or in the dissemination of visual work can be difficult  
-CYP might present information reflecting the dominant discourses on particular issues and be significantly affected by wider cultural issues  
-CYP might tell researchers what they believe to be the ‘right’ opinions or feelings  
-one of the risks is that the content & detail of CYP’s drawings might be regarded as |
### Appendix 2 – A section of the ‘Creative methods critique’

<table>
<thead>
<tr>
<th>Been used in…</th>
<th>Advantages</th>
<th>Disadvantages</th>
</tr>
</thead>
<tbody>
<tr>
<td>- It has been noted that the emphasis has been on methodological techniques and practical and ethical issues at the expense of epistemological &amp; analytical concerns</td>
<td>- their agreement to take part must be central to this and repeatedly sought both explicitly and implicitly.</td>
<td>- literal representations of their thoughts &amp; feelings</td>
</tr>
<tr>
<td>- 'adults who undertake research that involves CYP can perhaps never truly understand the world that they are exploring’</td>
<td>- CYP were asked if their work could be scanned and they were given the original</td>
<td>- draw and write data not always interpreted with reference to CYPs own interpretation leading to incorrect assumptions</td>
</tr>
<tr>
<td></td>
<td>- CYP must have control over the information that is gathered about them</td>
<td>- thematic analysis has often been used of, or in addition to, content analysis</td>
</tr>
<tr>
<td></td>
<td>- the DWT method is regarded as essential to marry up the CYPs interpretation with the content of their drawing &amp; text</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lego duplo, rainbows &amp; clouds &amp; moodboards</td>
<td>Duplo – basing research encounters around play reduces the pressure of a semi-structured interview</td>
<td>- individuals may have preconceived ideas about the age suitability of construction blocks</td>
</tr>
<tr>
<td>- Visualising children’s participation in research: Lego Duplo, rainbows &amp; clouds, and moodboards (2012 Pimlott-Wilson) <strong>Healthy CYP</strong> analysed by thematic analysis</td>
<td>- not just used in CYP, but adults too</td>
<td>- participants may become distracted from the task at hand, choosing to play their own game &amp; moving beyond the remit of the research</td>
</tr>
<tr>
<td>- examines the use of visual methods during research encounters with CYP when investigating their perspectives &amp; feelings towards their home &amp; school life</td>
<td>- some may not have the linguistic abilities or be better at expressing themselves creatively</td>
<td></td>
</tr>
<tr>
<td></td>
<td>- 5-6yr old build their own home &amp; enact roles</td>
<td></td>
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</tbody>
</table>
### Appendix 2 – A section of the ‘Creative methods critique’

<table>
<thead>
<tr>
<th>Been used in….</th>
<th>Advantages</th>
<th>Disadvantages</th>
</tr>
</thead>
<tbody>
<tr>
<td>- acknowledging that CYP are social actors whilst an appreciation that the abilities of CYP are generally different to those of adults</td>
<td>- duplo normalised the process of questioning, reduced reliance on discursive &amp; drawing capabilities, and considered cultural experience</td>
<td>- may be difficult to ascertain where domestic behaviour ends and imaginative play begins</td>
</tr>
<tr>
<td>- attempts are made to make research techniques more participatory, drawing on the cultural experiences of individual CYP</td>
<td>Duplo engages behaviours, abstract concepts &amp; metaphors</td>
<td>- therefore duplo (like other activity-based methods) cannot be used in isolation &amp; continued dialogue with the participant &amp; engagement in the task is necessary to understand the meaning assigned to the creations</td>
</tr>
<tr>
<td>- by using a range of research methods, CYP can relate their experiences to the research question and given the tools, environment and voice to effectively express their views, giving credence to their opinions &amp; providing an insight into their lifeworlds</td>
<td>Rainbows &amp; clouds – a structured d&amp;w activity can help CYP express their opinions about a real life issue even when they have conflicting views</td>
<td>- also, the research materials provided may not correspond with the concepts respondents are trying to express</td>
</tr>
<tr>
<td>- good intro to visual methods &amp; effect of culture</td>
<td>- this method was conductive to expressing both positive &amp; negative perceptions</td>
<td>- also the CYP were concerned that their duplo house did not mirror the one on the packet</td>
</tr>
<tr>
<td>- Does not discuss how they coded the duplo models?</td>
<td>- method can be adapted to the preferences of each participant</td>
<td>Rainbows &amp; clouds – some CYP found it difficult to concurrently write &amp; talk</td>
</tr>
<tr>
<td>- during normative activities such as storytelling, children will rarely display negative feelings</td>
<td>- acted as a method which allows CYP to visualise &amp; articulate contradictions discussing at their own pace, gives greater time, allowing them to make</td>
<td></td>
</tr>
</tbody>
</table>
### Been used in….

- **Rainbows & clouds** – A3 sheet of paper with rainbow on & another with cloud, sunshine’s & raindrops cut out separately, CYP wrote on these, sun introduced first.

- **Chronological age is not the sole controlling factor** when deciding on the range of methods to use, the preferences of individuals, their experiences & the research theme should also guide techniques used.

- **Issues of ownership & anonymity** are considerations as with any final output, some retained others didn’t want to.

- **Productions need to be treated sensitively**, as handwriting may be recognised, researchers therefore have a responsibility as with other data to ensure that the information presented will not identify the participant & cause undue distress.

- **The moodboards concept** was introduced to participants showing an example that had already been produced, although the provision of an amendment, gave a visual prompt, placing of suns and clouds important.

### Advantages

- **Moodboards** – like a collage, but not called collage as to highlight the expressive power of images beyond first impressions, can be images, text and samples of objects.

- **Allow participants to express beliefs & abstract ideas** without the requirement to put those feelings into words first.

- **Using images from the media can mean** that participations feel less limited by their own technical abilities.

- **Researcher is able to ask probing questions** and the respondent has time to make amendments to the final production.

- **The CYP said this technique allowed them to express thoughts & perspectives** which standard

### Disadvantages

- **Moodboards – must ensure materials are not inappropriate and provide a wide range of resources**

- **Can be a lengthy task**

- The choice of images available to them was constrained by the media placed before them.

- **CYP may not identify with the age, gender or ethnicity of people depicted in the media**

- **Researchers could decide to use pre-cut images from the media but by doing so, the research would be subject to researcher editing**

- To avoid this, large stacks of complete magazines, brochures and leaflets were used in the research. You could buy lots of the same magazines, but as individuals
<table>
<thead>
<tr>
<th>Been used in….</th>
<th>Advantages</th>
<th>Disadvantages</th>
</tr>
</thead>
<tbody>
<tr>
<td>example could introduce bias into the research encounter, the novelty of the method necessitated the use of an illustration so that respondents felt more comfortable in the task</td>
<td>interview questioning alone would not have elucidated.</td>
<td>ascribe different meanings to the same image, they will find other images suitable</td>
</tr>
<tr>
<td>-permission was asked to use copies/photos of the end results in academic papers</td>
<td></td>
<td>-moodboards also raise issues of authenticity, interpretation &amp; reliability</td>
</tr>
<tr>
<td>-CYP are not less skilled because of their chronological age, yet the socialisation of children in western contexts means few are accustomed to prolonged question &amp; answer sessions with an unfamiliar adult &amp; their linguistic skills may not allow them to effectively express their viewpoint.</td>
<td></td>
<td>-it is pertinent to remain aware of any research encounter may affect the information which participants chose to disclose</td>
</tr>
<tr>
<td>Lego / duplo</td>
<td></td>
<td>-interpretation should be carried out alongside the dialogue of the participant</td>
</tr>
<tr>
<td>-Creative and visual methods for exploring identities (2006 Gauntlett)</td>
<td>-I'm developing research which enables people to communicate in a meaningful way about their identities &amp; experiences through creatively making things themselves</td>
<td>-you should not analyse each creative artefact because that is done by the person that made it</td>
</tr>
<tr>
<td>-Conversation published where Gauntlett defends creative methods (drawing, collage, video or lego model) and esp his work with lego</td>
<td>-the method is empowering</td>
<td></td>
</tr>
</tbody>
</table>
## Appendix 2 – A section of the ‘Creative methods critique’

<table>
<thead>
<tr>
<th>Been used in….</th>
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<th>Disadvantages</th>
</tr>
</thead>
</table>
| - Talks about Merleau-Ponty – body & mind are inseparable  
- talks about drawing together the researchers horizon and the participants horizon | - ‘The therapeutic value of creative activity is well documented, but in these methods discussed here, the possible value for a participant is like a happy side effect’  
- it engages the brain in a different way |  |
| **Puppets** | - using a puppet may reassure CYP that they are the ‘expert’ & their interacting with adults when they have something concrete to focus upon, rather than needing to maintain eye contact  
- there was a sense that play & toys were an essential part of these CYP’s hospital experiences  
- play has many functions in hospital; distraction, can reduce anxiety, relief from boredom, or symbol of familiarity  
- the ‘power imbalance’ between the adult researcher & young participant is reduced as the facilitator needs to have the skills to enable quality evidence to be collated  
- researchers need to employ reflexive techniques to consider their role, their assumptions, research methods & their application when creating the worlds of children  
- undertaking research with CYP who were mainly on treatment was logistically challenging & required understanding & flexibility  
- interviews had to be short for this age group |  |
| What is important to young children who have cancer while in hospital? (2009 Aldiss) CYP with illness used thematic analysis  
- participatory research project exploring CYP’s experiences & views of cancer care services  
- 10 CYP aged 4-5yrs  
- Play & puppets were used to help CYP express their views  
- well recognised that CYP & adults have a different understanding & experience of the world, this can lead to disparities in what is perceived as important |  |
### Appendix 2 – A section of the ‘Creative methods critique’

<table>
<thead>
<tr>
<th>Been used in:...</th>
<th>Advantages</th>
<th>Disadvantages</th>
</tr>
</thead>
</table>
| -requires researchers to enter the world they are familiar with, good intro to creative methods  
- CYP not informed of gift until they had taken part | child is not required to talk directly to the researcher or maintain eye contact  
- parents played an important role in supporting their child, maybe in the future they could become co-researchers to enable CYP to articulate thoughts and feelings | - project wanted pts at different time points in treatment, but was too difficult  
-yp wanted their own puppet but team decided against it as they wanted their own viewpoint rather than a fictional characters viewpoint  
-a few considered the tweenie dolls to be ‘babyish’ and chose not to use them |
| On reflection it was felt that it would have been beneficial to have a number of techniques to aid communication from which each child could select |  |  |

- Reality box  
- Ascertaining the perspectives of young children in care: case studies in the use of reality boxes (2012, Winter) **Healthy CYP**  
- Innovative methods to elicit perspectives of children aged 4-7years in care 14 CYP semistructured interviews 40-90mins  
- 3 main developments underpin all of this work;  
  1. the use of growing range of creative & participatory methods  
- be aware of issues of power, CYP may feel there are demands on them and look to the researcher for confidence, skill, responsiveness, reflexivity, flexibility and creativity to negotiate the way  
- must avoid discriminating against young children on the basis of their age  | -consideration needs to be given to dissemination of research findings where participatory methods have been used with CYP. There is potential for the abuse of power in interpreting the CYP’s work for them rather than with them and in keeping and using their work without their knowledge of permission.  
- need to be sensitive to issues of risk  
- balance between the fact that their work could not be attributed to them personally, |
<table>
<thead>
<tr>
<th>Been used in….</th>
<th>Advantages</th>
<th>Disadvantages</th>
</tr>
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<tbody>
<tr>
<td>2. Increased knowledge about children’s experiences of &amp; relationships with world</td>
<td>- Use of age-appropriate, multimethod research methods that ‘play to children’s strengths rather than weaknesses enables CYP to express their thoughts and feelings, and is creative and interesting for CYP</td>
<td>3. Application of a child’s right-based approach while protecting them from risk of exploitation, exposed them to the risk of feelings ‘pushed to one side’ and silenced</td>
</tr>
<tr>
<td>3. Application of a child’s right-based approach</td>
<td>- adapted the idea from a conference, made reality boxes – outside of box, public person, inside – private person</td>
<td>- This project sees CYP as competent to express their views and promotes their participation rights through the use of participatory methods</td>
</tr>
<tr>
<td>- This project sees CYP as competent to express their views and promotes their participation rights through the use of participatory methods</td>
<td>- Mixed methods – photo elicitation, draw &amp; write techniques &amp; focus groups</td>
<td>- This raises the question about ‘duty to care’ to audiences given the sensitivity of some of the issues that CYP have chosen to explore and express</td>
</tr>
<tr>
<td>- Mixed methods – photo elicitation, draw &amp; write techniques &amp; focus groups</td>
<td>Multimethodology research with boys with severe haemophilia (Khair, 2013)</td>
<td>- Critique by Pimlott-Wilson 2012 paper says that it is potentially limiting for CYP who do not feel they possess necessary artistic talents or imaginative flair to assign meaning to craft materials such as lollipops to stick on their box</td>
</tr>
<tr>
<td>- Study specifically talks about engaging boys in research</td>
<td>- Need to get the process right, eg who takes the photos, who prints them, what about gatekeepers having rights to delete any etc</td>
<td>- CYP take photos that they think we want to see</td>
</tr>
</tbody>
</table>
### Appendix 2 – A section of the ‘Creative methods critique’

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</tr>
</thead>
<tbody>
<tr>
<td>run by participant co-researcher &amp; individual interviews</td>
<td>- Grounded theory, with new theories emerging, 4-16yr olds, 4-7yrs digital photos to use ad discussion prompts 8-12yrs draw or write about their experiences of hae 13-16yrs were invited to a focus group they were asked to draw a picture of himself and having haemophilia, then they were interviewed at home 2 wks later to discuss it. 10boys took photos – pilot 7yr old took 214 photos, 6hrs of printing, - in future parents reimbursed, CYP picked their favourites to discuss children invited to say anything off tape the triangulation of data, allowed the researcher to compare views of life with haem across a wide age range.</td>
<td>- CYP may forget or deny the hardest aspects of illness experience, so this gives them the opportunity to reflect on things they do not usually think about. drawing can enable even very young CYP to express their views about their health &amp; medical care. drawing cab ne seen as non-threatening, making them exert, able to communicate their thoughts, beliefs &amp; knowledge as they describe their drawings focus groups encourage in-depth discussion and produce data that reflect the views of each participant as well as the group.</td>
</tr>
</tbody>
</table>
### Appendix 2 – A section of the ‘Creative methods critique’

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<tr>
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</tr>
</thead>
<tbody>
<tr>
<td>-only 4 attended focus group discussion due to drop outs</td>
<td>-this study demonstrates how multiple research methods can be incorporated into a single study</td>
<td>-did not talk about using the draw data</td>
</tr>
</tbody>
</table>
Appendix 3 – Ethics approval letter for phase 1

Health Research Authority

North East - York Research Ethics Committee

Room 001
Jarrow Business Centre
Rolling Mill Road
Jarrow
Tyne & Wear
NE32 3DT

Tel: 0207 104 8282

Please note: This is the favourable opinion of the REC only and does not allow the amendment to be implemented at NHS sites in England until the outcome of the HRA assessment has been confirmed.

23 June 2017

Dear [Redacted]

Study title: [Redacted]
REC reference: 01/3/022
Amendment number: Substantial Amendment 12 - April 2017
Amendment date: 25 May 2017
IRAS project ID: 229746

The above amendment was reviewed by the Sub-Committee in correspondence.

This amendment is to seek approval to interview up to 20 young people with Juvenile Dermatomyositis who have already enrolled in the Juvenile Dermatomyositis Cohort and Biomarker Study (JDCBS). This is to collect data to assess psychosocial health which is not currently collected.
Appendix 3 – Ethics approval letter for phase 1

Ethical opinion

The members of the Committee taking part in the review gave a favourable ethical opinion of the amendment on the basis described in the notice of amendment form and supporting documentation.

The Sub Committee did not raise any ethical issues.

Approved documents

The documents reviewed and approved at the meeting were:

<table>
<thead>
<tr>
<th>Document</th>
<th>Version</th>
<th>Date</th>
</tr>
</thead>
<tbody>
<tr>
<td>Covering letter on headed paper</td>
<td>Email from Cerise Johnson</td>
<td>31 May 2017</td>
</tr>
<tr>
<td>Non-validated questionnaire [Proposed Interview Questions]</td>
<td>1</td>
<td>30 April 2017</td>
</tr>
<tr>
<td>Notice of Substantial Amendment (non-CTI/MP)</td>
<td>Substantial Amendment 12 - April 2017</td>
<td>25 May 2017</td>
</tr>
<tr>
<td>Other [Summary of changes]</td>
<td>1</td>
<td>30 April 2017</td>
</tr>
<tr>
<td>Participant consent form [Child Assent Form]</td>
<td>1</td>
<td>30 April 2017</td>
</tr>
<tr>
<td>Participant consent form [Young Persons Consent Form]</td>
<td>1</td>
<td>30 April 2017</td>
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<tr>
<td>Participant consent form [Parental Consent]</td>
<td>1</td>
<td>30 April 2017</td>
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<tr>
<td>Participant information sheet (PIS) [Parent Information Sheet - April 2017]</td>
<td>1</td>
<td>30 April 2017</td>
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<tr>
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<tr>
<td>Participant information sheet (PIS) [Child Information Sheet - April 2017]</td>
<td>1</td>
<td>30 April 2017</td>
</tr>
<tr>
<td>Research protocol or project proposal [Protocol (1/3/22) - Version 10 April 2017]</td>
<td>10</td>
<td>30 April 2017</td>
</tr>
</tbody>
</table>

Membership of the Committee

The members of the Committee who took part in the review are listed on the attached sheet.

Working with NHS Care Organisations

Sponsors should ensure that they notify the R&D office for the relevant NHS care organisation of this amendment in line with the terms detailed in the categorisation email issued by the lead nation for the study.

Statement of compliance

The Committee is constituted in accordance with the Governance Arrangements for Research Ethics Committees and complies fully with the Standard Operating Procedures for Research Ethics Committees in the UK.

We are pleased to welcome researchers and R & D staff at our Research Ethics Committee members’ training days – see details at http://www.hra.nhs.uk/hra-training/
Appendix 4 – 8-12 phase 1 information sheet

1. Why are you doing this study?
You are already in a big study looking at how Juvenile Dermatomyositis (JDM) affects your body and what medicines you take. But we want to know how your JDM makes you feel.

2. What will happen?
We want to ask you some questions about your JDM. If you decide to take part, one of the nurses will ask you some questions about your JDM; this is called an interview. If you want to talk, like a toy or picture, and we may play or draw while we talk. If you want an adult to sit in with you too, then that’s OK. We would like to tape record the interview, but only if that is OK with you.

3. When will this happen?
You can have this information from home and talk about it with your family and then come back on another day that is good for you to have your interview, including the weekend. Or if you want, we may be able to do it today. If you prefer, the nurse could come to your house if that would be better for you and your family.

4. What are the good things about saying yes?
This interview gives you a chance to talk about what it feels like to be you, sometimes it feels good to do that. Also, by sharing your story it will help doctors and nurses ask better questions and help children like you in the future.

5. Are there any bad things about saying yes?
Sometimes talking about things can make children feel a bit sad. If you feel sad and want to stop the interview, then it is fine and you will be told how to do this. If the nurse is worried that you feel very upset then she will ask your family and your doctor about this to see what else we can do to help you. We have someone on our team in the hospital who can give you ways to help you with these feelings.

6. Who will know what I say to the nurse?
We won’t use real names when we write about the project or when we talk to other people who want to know about our work. Instead, we will give you a made-up name—that way nobody will be able to guess who said what! The only time we might tell another adult what you told us is if we are worried about your safety.

7. Do I have to do it?
No, it is fine to say no, and no one will mind. And, if you have said yes, it’s still ok to change your mind and say no later.

Do you want to talk about your JDM?

Information sheet for children 8-12 years old

Thank you for reading this

Appendix 4 – 8-12 phase 1 information sheet
Appendix 5 - 13 – 19 phase 1 information sheet

Juvenile Dermatomyositis Cohort Biomarker Study and Repository (UK and Ireland) (JDCBS)

Information sheet for young people aged 13-19 years

Offering Children and Young People the opportunity to talk about their Juvenile Dermatomyositis

What is the purpose of this study?

As you may know, you are already enrolled in the Juvenile Dermatomyositis Cohort and Biomarker Study, which means that your medical team collect information about your condition each time they see you, like what medicines you are on. However, we don’t ask young people how they are feeling and how their Juvenile Dermatomyositis affects them.

Therefore, we want to ask you whether you would like to take part in a smaller part of this big study, to ask you what it is like to have Juvenile Dermatomyositis so that we can help you and young people like you in the future. We want to know what things are important to you and how Juvenile Dermatomyositis makes you feel. What things are good and what things are not so good. We will ask other young people like you to also tell their story and then we can put it all together and work out how we can ask the important questions in the future.

Please take your time to read this information sheet carefully and if you have any questions or anything is not clear, please just ask us.

Do I have to take part?

No, you do not have to take part.

Your current and future care within the health service will not be affected in any way if you decide not to take part. If you say yes now, you are free to stop at any time without giving a reason.
What will I be asked to do if I agree to take part?

We are asking some people to take part in an interview with a nurse. We suggest you take this sheet away and think about whether you want to take part, then if you do we will arrange for you to come back for your interview or the nurse could come and talk to you in your house if you would prefer. We would like you to bring something, maybe a photo, or card, or you could draw a picture or write a story - something that makes you think of your Juvenile Dermatomyositis, then it's easier to start talking. We would like to record the interviews on an audio recorder so that we can remember what you said. Your parent or a friend can sit in with you if you would like.

When you have read this information you will be given the opportunity to ask any questions you may have. You will then be asked to sign an assent form (or consent form if over 18) for this part of the study if you decide to take part. You will be given a copy to keep and the original will be kept as part of your medical records.

What are the advantages about taking part?

You and other young people who agree to take part in this study will have the opportunity to talk about their experience of Juvenile Dermatomyositis in more detail, which may be helpful. Sometimes it helps to talk to someone that is not very close to you, like a family member.

It will also help other people by helping doctors and nurses having a better idea about how young people with Juvenile Dermatomyositis feel and then we can help other people like you, by asking the right questions in the future.

What might be the disadvantages about taking part?

Taking part in this study means giving up some of your time, this might be up to an hour, depending on how much you want to say.

Some young people might get upset talking about their illness. If this happens to you, then we will stop and talk about this with your medical team and your parents or carer to see how we can help you more now.

Some young people find it hard to talk about their illness, but we will try and make the interview relaxed and fun, and you can bring something to talk about if you think it might help.
Appendix 5 - 13 – 19 phase 1 information sheet

What will happen to the results of the study?

You will be invited to a meeting when the study is finished in 2 years’ time to share the findings from all the young people in anonymised forms, no one will know who has said what. Other healthcare professionals from around the UK and other researchers will be invited to the meeting too.

Who is organising and funding this study?

This study is being carried out by Polly Livermore, Senior Nurse and current researcher with the Juvenile Dermatomyositis Research Team at Great Ormond Street Hospital.

The research has been funded by a National Institute of Health Research (NIHR) Clinical Doctoral Research Fellowship.

Investigator is carrying out this project as part of a research doctorate programme supervised (please see her details below).

Like all research studies, ethical approval has been gained for this study from

Who should I contact with any questions?

Contact for further information:

Chief Investigator

Principal Academic Supervisor

Thank you for taking the time to read this information sheet
Appendix 6 - Questions considered for phase 1

- Do we split the methods into expected age ranges to use them?
- Or, one technique across all ages?
- Or, offer a range of techniques for all?
- Is it better to have more or less methods to chose from?
- To have ice breakers and closure activities as well?
- Can I easily access the materials I need?
- Can I travel with the equipment required (eg how portable is it?)
- How costly are the materials?
- Are they cleanable for infection control purposes?
- Confidentiality – would we use the end results? eg drawings, could they be identified?
- Do we offer them for the young people to keep them?
- What happens if they want to write their name on them?
- How to make it positive and fun?
- Has there been research done on them before?
- Make it novel or base on other techniques?
- What are the advantages of each method?
- What are the limitations of each method?
- Are we actually analysing the output, or the verbal explanation of them?
- What happens if they want to use a couple of methods?
Appendix 6 - Questions considered for phase 1

- Use of YouTube videos? Positives vs negatives (might encourage yp to look at other YT videos, some very negative)

- Would all children and young people be able to use a tool like lego, what happens if their hand function is bad or they can’t sit up?

- Would vignettes work?

- Are we going to get the true essence of what it is really like to have JDM?
Appendix 7 – Section of interview protocol

icesbreaker

The icebreaker phase is essential for this study as a time to get to know the CYP. Ice breakers are designed to put the CYP at ease and build rapport, and confirm again that the CYP has signed the assent/consent form and is aware of the purpose of the study (Cross and Warwick-Booth, 2015; Einberg et al., 2015).

Initial introductions to each other, including first name of researcher and CYP’s preferred name, and time for any questions the CYP may have, are just as important as they would be when interviewing adults (Horstman et al., 2008). An introduction to the audio recording equipment, including the instructions for how to stop it and permission to do so if they want at any stage, are equally important in empowering the CYP to take an active role in the interviews (Sartain, Clarke and Heyman, 2000).

All CYP will be asked to come to the interview with an item that they relate to their JDM. This could be a stuffed toy, a photo, a card, a box of tablets, clinic letter etc. This time spent ‘chatting’ about an object of choice of the CYP is important to convey to them that their individual thoughts and feelings are priority, and that their interests are essential, and that the interview can move forward at their pace (Horstman et al., 2008). In Sheridan, Chamberlain and Dupuis, (2011) study, participants were asked to produce a material object which held special meaning from the perspective of the study, this was not for triangulation or reliability, but rather to explore the experience in a variety of ways, and proved to work very well. It is also an important introduction to the researcher about their confidence when speaking to an unknown adult, their willingness to take part in the study and an insight into their JDM.
The theme phase

This phase has three options for the CYP, and these will be offered to all ages giving them the flexibility to select a method which they wish to engage with. There may be time for multiple methods, this will be left to the CYP and researcher to decide upon the day.

1. Electronic Timeline

The use of personal history timelines, time lining or life grids to aid participants to recall and consider important life events has been well documented (Parry, Thomson and Fowkes, 1999; Wilson et al., 2007; Crivello, Camfield and Woodhead, 2009; Sheridan, Chamberlain and Dupuis, 2011; Kolar et al., 2015; Pfister, Vindrola-Padros and Johnson, 2017). Using a timeline in the context of an interview can open up participants interpretations of questions and allow a creative way of interviewing that is responsive to participants own meaning and inform verbal semi-structured interviewing, especially when researching sensitive topics (Bagnoli, 2009; Kolar et al., 2015). Timelines may also increase respondents degree of control over the disclosure of sensitive issues and feelings as the CYP can decide how much to reveal, plus through looking at the timeline, the need for sustained eye contact is averted (Wilson et al., 2007), and as this is one of the rationales for this project (the difficulty communicating, low mood and general loss of eye contact), this method is appropriate.

One of the aims of this research study was to create a positive, therapeutic experience for those involved, whilst being able to explore issues of chronic illness in CYP. One of the ways the timeline can help to facilitate this, is to include hopes and dreams for the future, to aid the young person to see a life past that of their disease, as Bagnoli, (2009) highlight whilst past timelines can help collect information regarding events that have been particularly significant to CYP, future timelines can provide a projection of the events that are expected to structure their lives in the future.
Appendix 7 – Section of interview protocol

Critiques of this method include that recall of dates can be prone to inaccuracy, past experiences can be merged with others (Parry, Thomson and Fowkes, 1999) and can be challenging in individuals who struggle to understand time, such as those with significant mental disabilities (Bagnoli, 2009). However, as their use in this study is to prompt respondent memories of personal experience, rather than obtaining accurate timeline data, and the exclusion criteria rules out co-morbidities, these critiques are superfluous.

All of the literature critiqued to date, uses tradition pen and paper to design the participants timelines. However this study, plans to use a freely available app to further engage the CYP with the process of designing their timeline.

The app, called ‘Timeline’ allows the user to construct their own timeline, adding dates, comments and pictures/photos if needed. The primary advantage of using technology, is that the research attempts to bridge the gap and engage the CYP through a medium which the majority are comfortable and confident with. The naturalization of information technologies in childhood have changed the practices of CYP significantly over the past decade (McTavish, Streelasky and Coles, 2012) At present, current research states that approximately three-quarters of teenagers own a smartphone, 24% of adolescents describe themselves as “constantly connected” to the Internet and 50% report feeling “addicted” to their phones (Reid Chassiakos et al., 2016). Using an electronic device, the previous difficulties as experienced by Wilson et al., (2007) in that some respondents did not like writing and thus did not want to complete a timeline, and from a previous critique that writing is too much like school work should be overcome. Previous studies have shown significant greater compliance when using electronic diaries compared to paper completion (Palermo, Valenzuela and Stork, 2004) and retention and adherence were significantly higher in a study comparing a smartphone app compared to a paper diary for recording weight loss (Carter et al., 2013).

The app is free, and does not require the internet, it is therefore portable and each timeline can be personalised by the young people. As they add their significant events onto the timeline, the researcher PL will further ask for clarification, and thus the individuals lived experience can be discussed.
2. Comic book drawing of their life, past, present and future

Alongside the app, a more conventional ‘draw, design and discuss’ comic book approach will be offered.

Again the advantages of using drawings in interviews with CYP have been extensively discussed and include; an innovative & developmentally appropriate method to visualise health & to reveal how CYP understand illness & communicate their experience, prioritises cognitive ability over verbal ability, provides a child centered way to share the lived experience, taking away focus from adult researcher, can be used across all age groups, is a non-threatening method of collecting data and puts CYP into the role of ‘experts’ & that their thoughts are valued (Nic Gabhainn and Kelleher, 2002; Horstman et al., 2008; Coad, Plumridge and Metcalfe, 2009; Elden, 2013; Literat, 2013; Lima and de Lemos, 2014; Angell, Alexander and Hunt, 2015).

The literature is awash with papers reporting on the ‘draw and write technique’ (Backett-Milburn and McKie, 1999; Sartain, Clarke and Heyman, 2000; Nic Gabhainn and Kelleher, 2002; Horstman et al., 2008; Elden, 2013) however, in parallel with these are numerous critiques. CYP producing data they perceive adults want to see, how much direction and prompting is required, the analysis of drawings can be difficult, who owns the final creations and not all CYP like drawing, to name a few. Angell, Alexander and Hunt, (2015) believe that to overcome some of these difficulties, the ‘draw, write and tell’ method is a more robust way of carrying out such research, but stress the importance of considering the epistemological and analytical concerns.

A Hermeneutic Phenomenological epistemology stance assumes that knowledge making is possible through subjective experience and insights, producing rich textual descriptions of the phenomena to develop deeper understandings (Kafle, 2011). Whilst Phenomenological Research can use a variety of methods to obtain experiences, such as interviews, conversations, focus groups and participant observation, the skill is to use minimum structure and maximum depth to gain the personal perspective from the participant. Most researchers use semi-structured
Appendix 7 – Section of interview protocol

interviews, however, there is very little research about conducting phenomenological interviews with CYP. Therefore, by adding a creative method into the interview, some of the limitations of interviewing CYP, such as having a limited vocabulary compared to adults (Punch, 2002b)

Hobday and Ollier, (1998) in their textbook of creative therapy present ‘writing a book’ together approach to encourage a CYP to think about specific issues, this can either be writing about themselves or a fictional character, and stress the importance of ending on a positive note. Therefore using this approach, a personalised blank comic book easily available, can be used to illustrate their timeline of life before, during and after JDM. Asking the CYP to design their own comic book about their lived experiences, and within the interview structure further question their creations as they draw and write, should produce similar results to the electronic timeline, but provides a different creative outlet for the young person to choose. The comic book approach also can end on thoughts about the future, thus ending on a positive as recommended (Hobday and Ollier, 1998).

3. Standard verbal interview

There is evidence that not all CYP do not like creative methods (after all as many adults remember finding ‘art’ at school a confusing medium, this lead us to question how many children may feel like this now (Harden et al., 2000)), and may prefer to talk about their experiences in a face to face interview (Carter and Ford, 2013). Coyne, Hayes and Gallagher, (2009) stress the importance of offering different methods to suit different children although highlight that individual face-to-face interviews were very effective in their study of hospitalised children aged 7-18 and produced very rich data.

Interviewing is the method of choice for phenomenological studies with the aim of eliciting a life story or narrative about an experience, usually in a low-structured format with little or no prompting (Clarke and Iphofen, 2006).
Appendix 7 – Section of interview protocol

Extras:

Body-mapping.

Bodymaps have been used predominantly to illustrate location of pain (Crivello, Camfield and Woodhead, 2009; Von Baeyer et al., 2011; Anna et al., 2014), however, as Crivello, Camfield and Woodhead (2009) highlight; Body-mapping is multifaceted and relatively easy to use with older children and can be a starting point for exploring key events.

The aim of body mapping for this project is to be an added creative tool, which should be quick to complete but would allow the CYP to draw or write their interpretation of how their JDM affects themselves. This would give the researcher in insight into the CYP’s coping strategies and current active disease state, whilst complementing data regarding the CYP’s experience to date.

The concluding phase

For this study, it is very important to conclude the interviews with some time to reflect on the interview and how the young person found the overall experience. Being aware that talking about the effects of their JDM could be upsetting for some, the importance of having time to ask questions and debrief from the experience may be vital for some of the CYP.

There is also the option to undertake; ‘3 wishes’ / ‘2 things which could make things better for you’ if required.

Coyne, Hayes and Gallagher, (2009) at the end of their interviews asked CYP to write down three wishes on paper. The purpose of this exercise was to allow the CYP another means of expressing what was important to them and used as another means of obtaining data.

For this project, the author feels it is very important to conclude the interviews with a positive activity that leaves the CYP feeling in a good frame of mind, so whilst 3 wishes may seem quite a tall task to ask CYP to name, asking them what 2 things could make things better for them, would seem an ideal compromise.
Appendix 7 – Section of interview protocol

All children and young people will be asked at the end of the interview what they thought of the methods used for their session. This will be an informal, but highly important part of this research project.

Post interviews:

After each interview the researcher will spend some time reflecting on any essential points to note, such as any non verbal additions, or any body language of note. Post the interviews as soon as possible they will be transcribed verbatim. After this point, second researcher on GOS site will listen to the tapes and compare against the transcripts for reliability and transparency. The transcripts will be discussed with the wider team.
Interview schedule

Depending on the age of the young person the number and complexity of the interview questions may vary. The researcher, with knowledge of this patient population, will be flexible, responsive and able to refine the core questions to the individual. The prompts brought to the interview by young people will also help in framing the questions asked.

Core questions:

What is it like living with JDM?

What do you understand about JDM?

How does it affect you?

JDM is not the same for everyone, can you describe your JDM?

Can you give an example of a good/bad day?

(When was that, what happened, how did you feel)

Tell me about the first time you remember that you had an illness

Can you tell me about your first admission to hospital? (if appropriate)

What was your last admission like? (if appropriate)

Tell me about a time when something went well/didn't go so well
Probing questions:

Can you tell me more about X?

What did you mean by X?

What did you do then?

What happened next?

How did you feel when X happened?

Can you tell me more about it?

I noticed you just said X, can you tell me more about that?
Appendix 9 – The shared stories

Shared ‘Being-confused’

Before my JDM I was a normal person, going to school and doing normal things. But then when it started I remember if I walked for a while my legs ached and I struggled to carry my school bag and I was slower than everyone else. Then we went on holiday I didn’t want to get up or do anything, but I didn’t understand why. Looking back now, the symptoms were there months before I was diagnosed. We went to the beach and I tried to sit up and I was like, ‘Oh, a bit embarrassing, I can’t just sit up,’ and I thought I was just a bit weaker than most people my age. That night I held a plank in my room and I could only hold it for a few seconds, that was when I first really noticed it. As time went on I was so frustrated because I was thinking to myself, ‘There’s something wrong with me,’ and I felt embarrassed as well because no one could see it, and only I could feel it. I remember kneeling on the floor and my family just going, ‘Just push on your legs. Push on your legs. What’s so hard about pushing on your legs to stand up?’ I was sitting there and I remember being in tears going, ‘I can’t.’ I’d found ways round everything. As I’d got weaker and weaker, I’d even slowly changed the way in which I get out of bed so that before I could even get the energy to stand up, it felt like I’d done an entire workout just to sit up. I remember being so frustrated because there were so many times when I was just at the point of trying not to cry because I was like, ‘There’s something wrong with me, and nobody can see it, and nobody can feel it’. I described it as having your entire body covered in weights. It’s not even just like having weights around your ankles, it’s like having weights round your ankle, round your calf, on your knee, on your thigh. Just everything was so heavy, especially my head. My shoulders hurt from the weight of my head.

Then my face went bright red, followed by my knees, my hands and my knuckles. We thought maybe it was due to the football match or the cold, or maybe an allergic reaction, but it didn’t go away and kept getting redder and redder. It wasn’t just red though, it was a scary blue kind of colour as well. We went to the GP but he thought it was eczema, then they said it was scarlet fever, some others thought it was Lupus and some said it could be growing pains. We were often made to
feel like we were wasting their time as we kept going back so many times. We saw so many doctors. I remember just being so anxious about what was going on, I didn't know why my body wasn't functioning in the way it should. I thought that I had a bug as I couldn't get dressed, I couldn't hold the toothbrush, I couldn't reach for my hair, but then I started to realize that it was definitely more than a bug when I couldn't stand up by myself in school, but I didn't know what was wrong. I just felt so miserable and so lacking in energy which just made me feel absolutely rubbish. People around me said I should do more exercise and go to the gym. Some people told me to stop being lazy, others told me to stop being silly. By now the weakness was getting worse, I began finding it hard to lift my leg over to get into the bath and get into my dad’s car, I couldn't lift my neck off my pillow, my neck started hurting when I was brushing my teeth and then I began literally crawling up the stairs because I couldn't walk by myself, it was really hard. I just became really, really weak, it kind of happened so quickly, but took so long for them to say what it really was. It took them so long to actually work out what was wrong because it is so rare, no one knows about it.

I was in a way, hoping for them to find something because I had no other explanation. I’d got so concerned, and because so many people turned around to me and said ‘A disease isn’t going to do that to you. An illness isn’t going to do that to you. It’s just you.’ So, I had gone on to the internet and I put in, ‘What can cause muscle weakness?’ to try and find an answer. It is so difficult to explain, and especially when you have no other physical symptoms, it does just look like you’re being a drama queen. I think my entire family were embarrassed about me, I was embarrassed about me, and then I was upset as well because I couldn’t explain it. I think that was one of the hardest things, the fact that you can’t explain it. Before a diagnosis, it is embarrassment and confusion because you don’t know what’s happening to you.

My GP said he didn't know what was happening to me, so he referred me to my local hospital, but they didn’t know what was wrong with me. So I was referred to a specialist hospital, this made it really, really scary and here I stayed for many months, missing school and home. When they started talking about what they thought I had, they took my mum and dad into a separate room to talk to them, but it I was like ‘Oh, what are you talking to them about?’ It takes a long time for
everything to sink in as everything’s just so rushed and you’re so scared. When they said about needing a muscle biopsy, I felt so anxious because I’d never been put under anesthetic before, and then when I woke up, I had a feeding tube in and it felt so uncomfortable.

I remember the awful bits quite well; clearly the muscle biopsy, the MRI scan, daily physio was terrible, the picc line which led to a portacath, the NG tube up my nose - all of it really, it wasn’t a nice experience. I remember the feeling of being scared, scared all the time and I remember that the medicines were horrible. It’s horrible having a tube up your nose, when they put it in it feels like your choking, it just kept on bothering me. It’s also scary not being able to stand up. The only time I figured out how to stand up was when I got out of my bed and tried to grab the commode, and I managed to stand up a little bit, and was really pleased, but then I dropped the pen on the floor and I tried to get it, I got it once, but then the next time I fell over. My mum stayed with me when I was in hospital, she was really upset, she wasn't crying but I saw that she wanted to cry.

I had a lot of pain in my arms, both legs and my ankles, and I would start to cry a lot. At the time I felt quite upset and worried, because I had one of those diseases that I knew was quite dangerous and could kill. You become a lot more nervous about everything than everyone when you have nurses and doctors rushing around you, it’s pretty scary. I felt like I wasn’t really there anymore, I wasn't really myself. I didn’t want to talk, I felt so low on energy, I felt really miserable as well from my physio sessions, because I was so weak and I thought the people I was with were stronger than I was, because I couldn’t lift my leg without the physio lifting it. I felt as though everyone was looking at me. I didn't know when I was going to be able to go home, I couldn't really believe what had happened to me. I kind of literally felt as though I wasn’t there. I was really sad about it, I just really wanted to leave as I didn’t want to be there. I didn't understand anything about it and I was really confused. I was definitely a lot more faded out of my own body, kind of felt as though my condition was defining me, so all I was feeing was pain and weakness. I felt as though I was never going to get out of the hospital, but part of you is scared too. Once I went home for the weekend, because I had to go up some stairs to get to my house, I went so slowly, my mum was behind me, but I was so scared I might fall. After that I didn’t want to go home again.
I felt tired, frustrated, isolated and invaded. My dad barely ever saw me when I was in hospital. I now only see him twice a month, he doesn't really think its JDM, he just thinks its mild arthritis. He doesn't understand what JDM is, because he hasn't looked into it, he hasn't asked any doctors and he doesn't know what I've been through, or what anyone with JDM has and he hasn't ever had anything wrong with him so he wouldn't know what it feels like. My dad kind of knows I struggle, but he’s too busy to realise it. Being in hospital was frustrating, everything I ate or drank was recorded, and as they made me drink about three times the amount I would normally drink, I just constantly needed to go to the toilet. At the time, I couldn't walk so this caused a bit of a problem. I didn't want to ask the nurses because I was too embarrassed. I also felt very cut off, I had social media which was good because I could keep in contact with people from school, but on the other hand it was bad because I'd see my best friend had gone out with other friends and I felt really replaced. I thought it was going to be so great having your friends visit, but a month in I felt like I’d been forgotten. No-one had visited me yet. It was the fact that everyone who I thought had cared about me suddenly seemed like they didn't. Whatever they're up to at school, I'm sat in bed ill, wanting to be at school and thinking about school and them, and what I’m missing, and yet sometimes if feels like they’re too busy to remember that’s where I am. I felt really smothered by everyone in the hospital and abandoned by the people outside. I knew that in the hospital I was safe as I’d been there for so long. The hospital very much became my life, and even moving to patient accommodation was terrifying because the hospital had become a protective shell. Later on, I remember going home for the weekends and wanting to go back because at home, I couldn't just press a nurse bell if I needed something. But in hospital I literally felt that as though I was trapped, the doctors controlling what I was allowed to do and the physios restricting me to my wheelchair, but it feels like they don't really care about what you are feeling. I still don’t know which bit of my illness affects what, it just confuses me, even after all this time, but I know without the medication, I wouldn’t be here today. In the beginning I didn’t know how seriously ill I was, all I wanted to know, was when am I ever going to have a normal life again?
“When I was at primary school, I had to go into like this little garden at break and lunch times, because when I had the hickman line and portacath, the doctors were worried about people running into me. It was like a little sort of cage, it wasn’t really a cage, but it was like a fenced off little play area, so, it was quite apparent that I had something because I was separated from the others. It was a good thing, because they were protecting me, but it was noticeable. It just is different, but then I was used to it being different. I felt a lot more tired, especially in the beginning, so different to others, a bit un-normal to the rest of school. Even now, I just feel like I’m kind of the odd one out. I can’t do as much as my friends, I can’t ride my bike or play football as I struggle a lot; I have pain in my ankles and feet. I find it hard to run, I can’t keep up and so my JDM has stopped me doing sports and it stopped me from being a footballer as I couldn’t go for a year as I wasn’t well enough. It stopped me doing PE in school as they were cautious of me. In PE lessons I was always sat on the side, it just made me feel even more isolated and again the odd one out. On school trips for instance, I wasn’t allowed to do the physical activities mainly because of the central lines, rather than the actual illness. I’d love to go to camp even now, but I can’t go there and take all my drugs, and then obviously I can’t do all the physical activity, although my head still thinks it can, but my body knows it can’t and it’s really disappointing, so that upsets me a lot. Other things like I can’t go travelling, I can’t go to a hot country like my friends because I already have to put sun cream on every day, if I was there I’d have to do it all the time. I do get more illnesses than other people as well and also the illnesses last a lot longer. I also find that if I get poorly I would have to go to hospital when others wouldn’t, like for chicken pox or tummy bugs, mainly because my immune system is quite low.

I have a wheelchair which I hate and although I know it helps I hardly use it because it makes me feel self-conscious again and I just feel weak when I’m in it because everyone can see I can’t do what other people can. People also look at you funny when you are in a wheelchair and then suddenly get out. I feel like I’m faking, so I just feel really guilty all the time. I need to sit in the wheelchair as much as I hate it, and went we went to a theme park I was quite happy with myself for doing it because usually I don’t and then I go through the pain of walking for
Appendix 9 – The shared stories

ages. So I got in the wheelchair as I wanted to see some of the rides, but then when I got out of the wheelchair I felt so bad, like I was faking it to get a pass, that made me not really enjoy the trip as much.

In the beginning, you look obviously different from everyone else. I remember when my skin was really itchy and I couldn't go to sleep, it looked like I had red food colouring spilt all over me and I felt quite insecure about my face, because I had a terrible rash on my cheeks. I hated the way that I looked so I wouldn’t leave the house without make up on. My face was like literally bright red and swollen, and I just looked horrible, like a baboons bum and so I got called names, like ‘tomato face’ and stuff like that. When people call me names, I keep it inside. Like when they called me slowcoach, then when I got home, I feel very frustrated at them, like that’s a horrible thing to say. I normally get very sad pretty quickly, I’ve been depressed and I get quite angry at a lot of things which I just kind of keep to myself. I have loads of cuts where I punch things, I know it’s not great, it’s just the way to release the anger.

JDM is hidden, it’s more inside the body, it’s not like a visible disability with a wheelchair all the time and stuff, so just because you can’t see it, it doesn’t mean it’s not there, so that’s what’s frustrating. With my disease, not many people think there is anything wrong with me, because I do look completely normal now, it’s not obvious I have an illness. They don’t know that other people are different, not everyone is the same, there can be something wrong with another person, they don’t realise it, because they just can’t see it, it’s not on the outside, it’s the inside. If people can’t see an illness by looking at someone then you don’t have to explain all the time to everyone, but then they don’t know you’re poorly and so I think it definitely makes it worse not being able to see it, because at school, I kind of feel bad because it looks like I’m faking to some of the teachers and it means that no one understands. People don’t know what it’s like and they’ve never heard of the condition before, so I have to explain the same thing 24/7 and it’s really irritating to explain all the time and for no one to understand at all. If it was something more common, then they would understand more because they would have heard of it and be like ‘Oh that’s really serious isn’t it?’, but no one’s heard of JDM and they’ll like, ‘What the hell is JDM?’.
After a while I realised I was the odd one out, rather than it being kind of normal, and it got quite difficult with my friends. Missing school, and not being able to go, even now, having to make sure there is shade or having to leave early, and to be constantly covering myself in sun cream. Only my friends that I’ve talked to about it kind of understand and my friends that I’ve known since I was diagnosed, they can be really kind and treat me as if I'm normal. They still remember that I have it, but they just treat me like I never had it, like they don’t mind, but in a good way. They understand when I say I’m a bit tired, they’ll understand you know.

My new friends from going to secondary school don’t really understand, I’ve explained what it is, but it’s rare so they need to be in my shoes to know how I feel and its hard, they expect me to still do stuff that they can do, but I can’t, they just don’t get it. They don’t necessarily immediately think of my illness when they think of me, because they didn’t know me when I got it. Others don’t know about my JDM yet at all, I know I will eventually have to explain, but I don’t want them to see me differently and I don’t want them to judge me on my illness. It just hurts how much I get left behind, because my body can’t keep up with everybody else. It’s frustrating not being like all your friends. When I’m off sick or in hospital, I find they are either constantly messaging me, or being like ‘are you ok, what’s wrong? are you coming back soon?’, or I feel as though they have forgotten about me. So it’s one extreme or the other. It’s hard because no one else can walk in my footsteps. I’m the only one who knows what it all feels like and you can’t explain everything to other people. Coming up to hospital and taking medicines, it just makes you different.

I think one of the few good things that comes out from having a medical condition is you find out who’s really there for you. The other day someone asked, "How come you’re writing with a laptop?" So I replied, "Because I can’t write properly at the moment." She then responded, "Well, you could write properly a while ago, and you have hands, don't you?" So I responded, "Yes, a while ago Stephen Hawking could write, and he still has hands."

I went to school on the first day of September, but I haven’t been since. In year 3 I had 3.5 months off, which was crazy and was really bad for my attendance, I do miss a lot of school, it’s just like I disappeared for a year when I was first poorly.
Appendix 9 – The shared stories

Now I've had my illness for many years, I probably can do everything my friends can do, although I'll probably just get achy legs sooner than them. At times I can't do all the gymnastics they can do, but then at other times I think I can do almost everything they can do and maybe even more, it's really hard as each day can be different. For me, I found the school stopped being the supportive place that it was, it was almost like I became old news. I had to make the decision to drop my A-levels because the school was just not being supportive.

It turned out that having the illness has good moments as well, because it's so rare people took an interest. I got invited to this party for people with bad illnesses, so you do feel kind of special, no one would probably even know what JDM is half the time. It's really good when you don't look any different and feel normal. When we went back home after being in the hospital, there was a big surprise party waiting for me and I also got nominated to go to dream flight to Disneyland in Florida.

I've still got JDM because I have to take the tablets, my skins quite tight and I'm getting rashes all the time. I feel like a 60 year old man, it gets me really down at times. I get upset, it's like a massive weight over me at the moment getting me down, it's not nice and it's frustrating. Emotionally, it just gets you down because you can't do the stuff you want to do. Physically as well, you're not able to do the stuff that others can do.

So in a way I'm kind of happier to be here, because while it's not a good thing to have and I'd rather be normal, in a way, I still like to have JDM to be not normal, because it's kind of being an individual is better than following a crowd. It gives you individuality. I'm kind of glad I have it in a way because it makes you different from the person sitting next to you. I feel a kind of good different, but at the same time I wish I was the same as my friends. My JDM is always on my mind because I'd love to play football in the park like my friends, but I can't without it being sore, so I try my best to fit in because I just want to be in their position, but I know I can't and to know it hasn't got a cure is really scary.”
Appendix 9 – The shared stories

Shared ‘Being-uncertain’

I remember before I was diagnosed, people used to say, what's wrong with your face and I used to say, I don't know and at the time I didn’t really know. Another early memory was after diagnosis, when I was about 7 or 8 years old and I was aware something was wrong with me, but I didn't know what it was. I remember I was at this wedding reception, and I was with my dad, and there was a baby there. All of the kids my age, were going to hug the baby, and I asked my dad, “oh, am I allowed to touch the baby?” and he went “what do you mean?” and I said “well, cuz there’s something wrong with me, I don't know if I am allowed to touch the baby?” he replied “what do you mean?” obviously like when you’re at that age, you think it’s like a disease, don’t you?

In the beginning I was really angry and I didn't know what was happening and I just wanted to go home. I was really confused why it was happening to me. I remember actually saying, ‘Look. I know that I am on the road to recovery but I came in here being able to walk and can’t now after having medication that you told me would make me better.’ I didn’t fully comprehend that I was going to have this forever, until a nurse said it to me. We asked a question about one of the drugs and then it just sort of led on from there and that I have to come off them when I want to have a baby, which made me think, wait, how long am I going to have to be on these for? When I went home from the hospital, my parents moved all my stuff downstairs so that my room was in what was my dad's office. You know the feeling when you're on a holiday but its raining and there isn't much to do and you feel a bit miserable? It was like that because you didn’t know what was happening. Everything had literally just been thrown off balance and it was scary.

My friends didn’t know why I didn't really participate in PE when I was poorly, but then when I came back from the physio, my mum told them a little bit about it. They know I have JDM and they understand that I have problems, you know not being able to do everything they can do, but I'm not sure they'll ever fully get it though. It’s only like 3 in a million or something who get JDM, so it’s not like cancer where it’s very well known. It’s a rare condition, so it’s kind of hard for them to understand how I feel and that really frustrates me. I guess you can never
Appendix 9 – The shared stories

quite get it, unless you experience it yourself. Nobody else would understand, maybe they might feel sorry for me, but I don’t want them to feel sorry for me. I have lots of friends who don’t actually really know about my JDM they just think I have to go to doctor’s appointments, they don’t actually really know something is wrong with me. I just don’t want to tell them. It would be a bit weird if I did. I’ll probably tell them one day, maybe like in a year, or half a year maybe. The teachers told them I’m allergic to the sun which isn’t actually true, because she doesn’t understand either, I don’t think any of the school teachers know about it. I don’t think my younger sister really understands either although she thinks it’s good because we can get into theme parks with a ride access pass so we won’t have to queue for so long.

I feel that you can’t worry about it if you don’t understand it well enough and as I have begun to understand what was happening to me, so as I got older if my legs were aching I knew what that meant, I understood what those signs were and what they could lead too. The methotrexate didn’t have a bad impact on me in the beginning, it’s because I started to understand it all more and I could then make the connection. Obviously as a kid you don’t know that you’ve got a potentially life threatening illness so once you start to understand that then it’s like, ‘ohh, that doesn’t sound good, why is that happening?’ Understanding is the biggest thing, because I do feel when my dad went ‘why can’t you just take a tablet?’ and then feeling them get frustrated and it’s not so much they’re frustrated at me, their frustrated for me, but I feel like I’m then being a burden, then I feel like their annoyed at me. It is really difficult for doctors and others who don’t really understand JDM and don’t understand the whole mental barrier around the methotrexate. I would sit there and literally put my head in my hands, and question why is everything so difficult? There are a lot of times where you just freak out, and think, if I didn’t have myositis, I would have been able to do my higher education exams. I’d be at least slightly more normal. I was really frustrated at myself and about the whole JDM and it really got to me then, because I was like, if I was ok, and if none of this was going on, I could have got those results and been really happy with them. Maybe without it I probably would still be playing football now, rather than having to stop. I was getting really worried and sad and I just didn’t know what to do anymore, like I didn’t think I would go to
school again and be like a normal person. I thought people would think I’m a bit strange. I never thought I could get like an ordinary job and I would just have moments where I think of my future and past and look back at everything and say I will probably never be ordinary, I would just kind of sit there and cry.

I think about it when I have my injections, I’m just like I wonder whether the injections will stop. I have had discussions with mum about whether to stop my drugs a couple of times, especially because I just don’t think they’re really helping that much. I think getting ill again might be connected to my mocks, the exam stress. I am worried that I’m going to fall behind. I am worried about what’s going to happen at university, I’m worried about a lot of stuff, to be honest, but I think once you kind of get to this point, you’ve lived with it, you know what’s happening better, and when you meet a new person, you don’t look disabled, but once you say to them, ‘I have myositis,’ they can go, ‘Oh, what’s that?’ It’s easier now to make a joke out of it. I don’t like going to hospital and sometimes now it feels that I’m wasting my time, because I need to focus on my school work and hopefully get rid of my JDM before I go to University.

My JDM had gone away and then it came back, it was quite shocking because I wasn’t really expecting it to come again. The doctors had said there was only 6% chance it would come back, so it was quite a shock. The constant fear of bad flares is huge, it’s absolutely huge, and I want to know ‘Why did I all of a sudden fall sick in the beginning?’ So many questions, and so many uncertainties. My transition to adult care is not good and I have just been left, my Disability Living Allowance stopped and the relentless physical and mental toll of my methotrexate, I just can’t do it. I think it would be good to have a friend to talk who has JDM as well so then you know they are feeling the same way as you and so you know you can have more of a bond because you have the same condition and they’ve been through the things you’ve been through. I haven’t got any friends with JDM to talk too.

I do get a bit worried about the future of my JDM, like who knows I could be a bit disabled. I remember I couldn’t walk, I could be in a wheelchair, no one really knows. I haven’t told anyone I’m worried, because if I told my mum, she’d be like, don’t say that, that will never happen, but no one can predict the future. Some
have died from it and it’s quite a scary thing to think about, so I just want to make
the most of everything, because you don't know if somethings going to happen
so you just kind of do everything, but I always like to think that I'll be like all my
other friends when I'm older.

It’s there all the time and you have good days and bad days, but you never know
what kind of day it will be. I always feel like a faker and that I’m not really ill,
especially as my JDM seems to be going fast and is a lot easier than it used to
be. I found it so hard that I would always compare myself to my friends, but they
don't get it. There were people calling me attention-seeker. I was like, ‘Yes, you're
exactly right. I attention-seeked my way to hospital. I even attention-seeked my
blood results, and my MRI results.’ You get so frustrated with people, and in a
way, I guess, it’s not their fault entirely because they don’t understand, but at the
same time, they don't make the effort to. When people ask me what I’ve got, they
go, “what, what’s that?” so they don’t know about it, even some doctors, even
physios at my local hospital don’t know what it is and they have to look it up. I
don't really have control over my condition, I can't control if I'm going to have a
flare up, I can do physio, I can take my treatment but apart from that, you can't
do anything. So, I just kind of accept that I'm not going to know what’s going to
happen. Even though you're doing well, doesn't mean that you're going to
continue, because everything can change in a month and I don't really know
what’s going to happen in the future. To be honest I can't ever see myself without
JDM.
Appendix 9 – The shared stories

Shared ‘Being-sick, steroidal and scared’

I think my medicines are too much. I have to have them all the time. They don’t really help me, and some of them taste really horrible, especially when you can’t swallow them in tablets and sometimes I forget to take them so I have to wake up in the middle of the night just to take them. It’s really not nice, having to take medicines every day, no normal 15 year old has to take medicines every day. I used to have two injections at one point, I hated them so much. My dad used to do them, I guess I’ll have to learn one day.

Methotrexate is the worst medicine, it used to take half an hour for me just to take a single syringe of it. The methotrexate injections, were horrible. I now can’t stand cling film, because it reminds me of when it used to hold the numbing cream in place, it kind brings it all back whenever I see it, it’s really weird. It just gradually built up, it wasn’t necessarily the injection that I found difficult it was knowing that it was going to make me feel ill and knowing what it would do to me afterwards that scared me. I used to get really, really nervous of injections. Then I’d feel sick before I even had it, then I started almost throwing up when I was having the injection, and having a panic attack before it because the day afterwards, no matter what I did, I would feel sick, and just wanting to cry because I just felt so horrible. I think I was starting to get a bit anxious about it and then the smell of the alcohol wipes was making me feel really sick. I would feel sick all Friday night, even when I was asleep and I couldn’t do anything on a Saturday. My Saturday was gone. You didn’t know how you would feel and how long it would last. Some days it would hurt and I wouldn’t really feel sick, then other days it wouldn’t hurt at all and I’d feel really sick. Last year I was always sick every single time, physically sick and feeling sick.

My mum would bring it in, I’d sit there being like, ‘No,’ shaking, and she’d swipe my leg, and then I’d literally be grabbing onto her arm, and she’d have to just do it as quickly as she could, and I’d just cry. So this became a routine thing on a Friday, which is obviously not the best way to spend your Friday nights. All my friends are out, and I’m sitting in bed crying. It was that I’d sense her getting closer and closer to my leg, I couldn’t let her and I finally had a bit of a meltdown about it. At this point, we had to stop the injections because I physically couldn’t let her
Appendix 9 – The shared stories

do it. I literally begged them to move me on to the tablet form. So then the first few weeks I was like, ‘This is great,’ and I woke up the next day being like, ‘I have my Saturday back.’ A few weeks later though, I wake up feeling sick again and the longer I was on it the worse it got. It got to the point where I’d literally take a tablet and throw up as soon as I swallowed it. I couldn’t take it, and I started refusing again, and this time my mum couldn’t pin me down. She would sit in my room and go, ‘Take them, take them now,’ and I would have to sit there and take them and then physically throw up all my dinner. So it got to the point where I was almost refusing to have it. I then couldn’t swallow the tablets, my dad still really struggles with understanding that and that I have to have it in liquid form because I can’t swallow a tablet, because they were that horrible chalky type, they got stuck in my throat and the taste was horrible. We tried crushing them in yogurt and crushing them in chocolate mousse, which I now can’t eat. That’s why it is so hard and so frustrating that it’s like why can’t I do it? It is why can’t I beat it?

My mum went through such difficulty trying to get the methotrexate in liquid form and I couldn’t take it, so then I felt awful. It is more than just that I don’t want it, it’s like a deeper I really can’t do it. It’s obviously a mental thing, because I feel ill before I’ve even looked at it, I feel sick, the same sort of sick that I feel after I’ve had it, so I know that it is all in my mind which I think is why it was difficult for my parents to understand. It’s so difficult to try and explain it, I don’t even understand it and its happening to me, all I can say is that I can’t take it. I have a really bad mental barrier with it, I hate it, it’s awful. I feel I can’t justify having what I feel like is such a terrible thing every week. It feels like the methotrexate weekly feels worse than what I’m living with at the moment. I wonder if I’d never of taken the injection, would I of felt sick on the tablets? Also, it does give you slight hair loss because its chemotherapy, like when I’m having a hair wash sometimes more hair comes out, so that’s pretty annoying.

They also gave me steroids infusions in the beginning which hurt, they felt like tiny little razor blades travelling up my arm. Then after about a week of oral steroid medication, I was looking around for food, I was always hungry. The effect the steroids had on my weight was one of the biggest things. For me the steroids were even worse than the methotrexate in some ways, because they make you all swollen and stuff from the drug. I just remember feeling so self-conscious
because I was a really good size before and I was skinny but I was healthy, and then I was suddenly on all of these steroids and it felt like I was a guinea pig or a hamster. I’ve never been really overly self-conscious about my weight, but that was the first time that I really was, because I noticed putting on weight, I noticed it when my jeans got tighter. It’s just not nice, it’s out of your control. My appearance really changed as I noticed my face went very red at the time, I had the round chubby moon face, most of my body put weight on and I became hairier. There were so many mirrors in hospitals. I could always see myself. I was thinking, ‘I’m getting uglier.’ I always used to be really self-conscious about it, I never used to feel nice about it, I remember wanting to lose it, because I just didn’t feel comfortable. It makes you feel kind of out of place, because you try not to eat, but then because the steroids make you hungry you can’t help but eat. It didn’t always bother me it depended on what I was doing, but I do worry about my appearance as I don’t want to put weight on and be fat. I'm off my steroids and now I just feel better. I think being self-conscious definitely is probably the worst part and now that I'm off the steroids and I'm back to my normal size, it’s really annoying because everyone is like ‘Oh you look really well, you've lost weight, oh why are you skinny?’ and surprisingly most of this is from the nurses and I just think they especially should know this.

My biologic infusions, they kind of make me either really like down, I don't want to do anything, or it makes me really energetic. The IVIG I also had made me really, really poorly. As soon as I had it the next morning, I'd just fell like bluuurgh. I'd throw up, I had really bad headaches, my head was booming and I couldn't even look at the television or anything as it would just really hurt my eyes, and then my stomach would really hurt. It is frustrating and often painful to have an injection, I suppose you kind of think about your friends and what they're doing at that moment whilst you're having an injection, but you need to try and remember, it’s going to make you better so there’s often a good side, people just trying to make you better. The drugs are really irritating, but they are just medicines.
Appendix 9 – The shared stories

Shared ‘Being-accepting’

I can’t remember life before JDM, it feels normal to me. I can’t remember a time where I haven’t had to take a medicine every day or every week, it’s been quite a constant thing for the majority of my life. I can’t really remember much about the beginning as I was quite young. I think having JDM makes you kind of mature quite quickly because you realise that you're not in a playground, you're in a hospital trying to stay alive.

I wished that I could do all the things that I could do before, but you’ve just got to carry on with life. It gets better, once you’ve learnt to accept, this is what I have, this is what it means, this is how I’m going to deal with it and it does become easier to manage. I think if you give up that’s not going to help your cause, you need to keep fighting against it, because that’s the only way you're going to beat JDM. It doesn't ruin your life, it makes it a bit more exciting in some ways. You meet a lot of new people through it, but its fun as well, and you can make loads of new friends. I think being in hospital so much actually helped me because it’s given me a lot more opportunities as I learnt a lot more about science and stuff and how diseases work. Having JDM opens up your eyes for a lot more. If someone is hurt, you can think about their position and how they feel or if someone has a disease you can see it from their perspective, but other adults and children would just act like they know about it and say they care, but because of being ill you know what it’s like and can help, which will make you have an intrinsic reward and make you feel happy about yourself. The person I am now is a nice, kind person. I think that’s what the JDM has made me. So in a way it’s a benefit as it’s made me more of a people’s person, I like helping others. My JDM hasn't stopped me, it actually pushes me more because I used to be scared of a lot of things and now a days I just don't care. I will go jumping off boats, I'll go swimming in the sea, I know how much I have been through and that I’m a brave person and then I just push myself past my limits. I’m fitter now and I learn something new every day and its fun living like that. JDM has really helped me become more of a positive, happy, brave person than I was before.

I had to put one foot in front of the other, there was literally no other way. It was a case of, get on with it because you have to. Hospital has become part of my
Appendix 9 – The shared stories

life, so it’s kind of normal. It’s kind of weird to think, wait, these other people in these rooms don’t routinely go to hospital, they don’t have a PICC line or have ever had a cannula. So, it’s nice in hospital to talk to other people who know what you’re talking about, and it’s nice out of hospital to have a normal conversation with people who in a way, have no idea what it is or what it’s like. My portacath was a reminder my illness was always there, whereas when it was taken out I felt a bit more confident that it was under control. I was a bit less reliant on hospitals, they also changed my medicine to oral, "oh I have a tablet", and lots of people have tablets, I mean I hated those tablets, but it felt like a more normal thing to do. I think as well because I don’t have a brother or a sister, I had nothing to compare too. If I had an older sibling, I’d be questioning why am I not more like them? I’ve never known having a mum and dad together. It’s a bit like that, I don’t know any different.

I feel sad that it happened to me, but I believe that everything happens for a reason and now I’m stronger than ever, mentally I’m super brave, I’m not scared of anything and I think that anything is possible. You feel as though you’re the unluckiest person in the world and its awful and nothing anyone can say can make it better but it’s just a matter of putting things into perspective which sometimes in the moment is very hard to do. It’s a bit like, me then and me now, a lot better you know. My JDM has made me a better person I used to be very negative and now I'm positive. I embrace my JDM and I feel more powerful that I have it, I came from nothing and now I'm stronger, can walk by myself and I just feel good that I had it as it has taught me a lot of things. My mum was really supportive and she always told me that everything would be better and look at me now, I'm better! It's just all normal now to me, I mean I used to be very weak and now I'm the strongest out of all of my friends. It doesn't affect me so much now, I've not forgotten about it because it was a massive part of my life, but it’s not in the back of my mind all the time, I feel like a different person, I have changed quite a lot. I feel like I’m glad it’s happened to me, as I feel I can deal with a lot more stuff, as opposed to if it just happened now, I would be like, ‘Oh my god, what’s going on?’

You get over it, because you don’t have a choice, that’s the thing that I learnt fairly quickly, that it’s fine to be sad about it, but if you’re going to sit there and be
Appendix 9 – The shared stories

sad about it, you’re going to let it stop you living your life, because it’s not going to go away. I know it could have been a hell of a lot worse along the way, even with JDM, I’ve been relatively lucky, but, at certain times it’s very difficult. Having JDM helped me find out who my real friends are, I made some good friends in hospital, and you realise that actually a lot of people have a chronic disease and they know how you feel, so you show them it’s going to be alright in the end that you just need to keep on being positive and make sure they don’t focus on their illness. I wouldn’t mind meeting up with someone else with JDM though, because you know what they’re going through and they know what you’re going through.

I can still live a relatively ordinary life, but it’s kind of reduced. I do have to be careful, I have to think about whether or not I’m going to need a stick today, you have to think in advance, but I’ve got better and better over the years so, now I can do most things. I know it will stay in my body for ever, but it won’t be like bad forever. I definitely feel in a different position as to how I used to be, I feel a lot better in myself. It is definitely a 100 times better than before, because I’m kind of more normal now. I still have my JDM, but you can’t tell, it’s quite weak inside, I think it’s in remission. I really feel like I don’t actually have it anymore, and that soon I won’t have to go hospital as I’m improving all the time. I feel like a normal person without any problems other than taking medicines every day. I kind of still have JDM, but not really. When I went down on my methotrexate I flared up, so I had to go back up on the injection, but it’s not affecting me anymore, so I’m not really tired and can still walk, but it’s still there though, they still have to treat it, it’s not really active right now, but like it’s a tiny bit active, it’s not fully turned off yet. Sometimes I feel like there’s nothing wrong with me and that I never had JDM because now I’m recovered I just feel very strong and happy mentally and physically.

You’ll look back on it when you’re older and be like ‘wow I went through that and now I can go through anything’ Yes, it can be annoying sometimes. But most of the time its fine, I put my suncream on and then just get back to playing outside with my friends and doing what I want to do and forget about it. I learnt you have to take the medicines either way so you might as well just let it go faster, so eventually I just got so used to the medicine and let them get on with it. It doesn’t stop you really, but its been hard, I’ve had to work hard, I’ve tried hard and then
Appendix 9 – The shared stories

I did get good results. I think I’ve just grown up. I think it’s really hard, but you have to live with it. I still say to myself that I’m going to get better either way, like it may take longer than other people, may be shorter or just the same, but I know eventually I will get better. I hope I will be like anyone else that hasn’t been ill, ultimately live a happy life, put this all behind me and hopefully not get it again, but who knows? Think about the people that are worse than you, think about all the good things in your life, because like you don’t live forever. Sometimes you’ve just got to try, but it’s hard because you don’t know what’s around the corner and whether things will get worse. I think the main thing is, I’m okay. Things hurt and things are hard sometimes, but I still enjoy them. I’m not dead yet. It didn’t kill me, and it’s not going to. I hope my JDM will go away, but I can’t be sure, I’ll just have to wait, but if it doesn’t, I’d like them to find a cure.
Appendix 10 - Comments in response to poems

It made me cry and [redacted] identified with every word.

I did get a notification you'd posted something last week but then it wasn't there when I came on. It's a horrible, horrible drug. It really does ruin our children's lives even though it's helping them. That sounds extreme but every day and hour was a countdown for [redacted] to when he was having the next injection. Just the word or seeing the sharps bin made him feel sick. Coming off it has helped so much with how he feels about JDM and also seems to have helped with his needle phobia too. Just told him he's having his flu jab Saturday and he didn't even bat an eyelid! Xxx

Hi [redacted]. I didn't see your original post. Reading this brought a tear to my eye as it really strikes a chord... I hate hate hate what our kids have to endure... [redacted] and your family have gone through so much over the years, which comes across very poignantly in this poem. Very moving 😇 xxx

[redacted] is sick straight away after hers and just not herself for at 2-3 days after 😔

So [redacted] - usually a very tough cookie but she wanted me to read the poem and her eyes just started leaking!! I said sorry for reading but she said it made her feel better knowing she's not the only one feeling the battle every day. We have been given your research request as about to embark upon a course of IVIG - just as soon as we have got our heads around the next drama, I'm sure [redacted] will be able to help. I so appreciate your interest in these kids' lives. We are bombarded with "cures" but the reality is it is difficult to feel grateful with the psychological drain that comes with them. So, thank you so much.
Appendix 11 - Ethics approval letter for phase 2

Please note: This is the favourable opinion of the REC only and does not allow the amendment to be implemented at NHS sites in England until the outcome of the HRA assessment has been confirmed.

15 August 2018

Study title:
REC reference: 01/3/022
Amendment number: Substantial Amendment 13, 20 July 2018
Amendment date: 26 July 2018
IRAS project ID: 229746

The above amendment was reviewed by the Sub-Committee in correspondence.

Summary of amendment

This amendment was to introduce questionnaires regarding elements of psychosocial health and send to all those 8-19 year olds already enrolled in the Juvenile Dermatomyositis Cohort and Biomarker Study. All responders to these questionnaires will then go into a prize draw to win a £100 voucher after the study has ended.

Ethical opinion

A Research Ethics Committee established by the Health Research Authority
Appendix 11 - Ethics approval letter for phase 2

The members of the Committee taking part in the review gave a favourable ethical opinion of the amendment on the basis described in the notice of amendment form and supporting documentation.

Approved documents

The documents reviewed and approved at the meeting were:

<table>
<thead>
<tr>
<th>Document</th>
<th>Version</th>
<th>Date</th>
</tr>
</thead>
<tbody>
<tr>
<td>GP/consultant information sheets or letters</td>
<td>6</td>
<td>20 July 2018</td>
</tr>
<tr>
<td>[Physician information sheet]</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Letters of invitation to participant</td>
<td>1</td>
<td>20 July 2018</td>
</tr>
<tr>
<td>[Psychosocial invitation and instruction letter]</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Non-validated questionnaire [Psychosocial 8-12 years Questionnaire Pack]</td>
<td>1</td>
<td>20 July 2018</td>
</tr>
<tr>
<td>Non-validated questionnaire [Psychosocial 13-19 years questionnaire pack]</td>
<td>1</td>
<td>20 July 2018</td>
</tr>
<tr>
<td>Notice of Substantial Amendment (non-CTIMP)</td>
<td></td>
<td>26 July 2018</td>
</tr>
<tr>
<td>[Notice of Substantial Amendment] Substantial Amendment 13, 20 July 2018</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Other [Psychosocial Flowsheet for local centres]</td>
<td>1</td>
<td>20 July 2018</td>
</tr>
<tr>
<td>Other [Psychosocial Pre Study Card]</td>
<td>1</td>
<td>20 July 2018</td>
</tr>
<tr>
<td>Participant consent form [Consent Form - Parent/Guardian]</td>
<td>5</td>
<td>20 July 2018</td>
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<tr>
<td>Participant consent form [Consent - Patient over 16]</td>
<td>5</td>
<td>20 July 2018</td>
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<tr>
<td>Participant information sheet (PIS) [Psychosocial CYP Information sheet 13-19yrs]</td>
<td>1</td>
<td>20 July 2018</td>
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<tr>
<td>Participant information sheet (PIS) [Child Young Person Information Sheet]</td>
<td>8</td>
<td>20 July 2018</td>
</tr>
<tr>
<td>Participant information sheet (PIS) [Parent Information Sheet]</td>
<td>12</td>
<td>20 July 2018</td>
</tr>
<tr>
<td>Participant information sheet (PIS) [Psychosocial YP information sheet]</td>
<td>1</td>
<td>20 July 2018</td>
</tr>
<tr>
<td>Research protocol or project proposal [Protocol]</td>
<td>11</td>
<td>20 July 2018</td>
</tr>
</tbody>
</table>

Membership of the Committee

The members of the Committee who took part in the review are listed on the attached sheet.

Working with NHS Care Organisations

Sponsors should ensure that they notify the R&D office for the relevant NHS care organisation of this amendment in line with the terms detailed in the categorisation email issued by the lead nation for the study.

Statement of compliance

The Committee is constituted in accordance with the Governance Arrangements for Research Ethics Committees and complies fully with the Standard Operating Procedures for Research Ethics Committees in the UK.

We are pleased to welcome researchers and R & D staff at our Research Ethics Committee members’ training days – see details at http://www.hra.nhs.uk/hra-training/

A Research Ethics Committee established by the Health Research Authority
## Appendix 12 – Questions/thoughts considered for phase 2

<table>
<thead>
<tr>
<th>Questions/thoughts/problems</th>
<th>Current progress/solutions/final decisions</th>
</tr>
</thead>
<tbody>
<tr>
<td>How to track the children and young people (CYP)?</td>
<td>I need to be able to know who has completed for the PIED scoring reasons, so will ask for unique code to be added to electronic ones &amp; paper one (backed up by already written on back of paper ones and envelopes) with the incentive of the prize draw</td>
</tr>
<tr>
<td>Why do I need to track them?</td>
<td>Because of the PIED and prize draw</td>
</tr>
<tr>
<td>Should we include the PIED?</td>
<td>Lots of discussion, final decision, yes</td>
</tr>
<tr>
<td>What happens to CYP who score over a certain score?</td>
<td>Have discussed with sites on TC, on flowsheet and in video – will let sites know within next working day, and they can action as they would normally do so for a CYP in distress</td>
</tr>
<tr>
<td>The ethical issues of offering internet based questionnaires</td>
<td>Many ethical issues, such as the youngest age being only 8 years, and not all CYP having access to internet – so begin by providing paper copy to all and then they can choose how to complete</td>
</tr>
<tr>
<td>The positive issues of offering internet questionnaires</td>
<td>Gives choice, mindful of current technological climate, saves on families having to post back, stores responses, provides some basic data analysis, easy for me to access</td>
</tr>
<tr>
<td>The negative issues of offering internet based questionnaires</td>
<td>Not all have access to internet, is the data safe, do I meet the criteria for electronic surveys from the companies? Would parents be unhappy about encouraging CYP to be on computers more?</td>
</tr>
<tr>
<td>What happens if CYP put wrong code in?</td>
<td>Tried to mitigate by asking also for correct code completion to allow for identification for prize draw, and backed up by asking for gender, day and month of birth, and pre filling in the paper versions, and making the electronic box needing completion before can progress through rest of survey</td>
</tr>
<tr>
<td>Gender – just M &amp; F, or should we put other, or I don’t want to say?</td>
<td>Decided to add in other to allow for options, but aware this could encourage CYP to put other – which may then limit identifiable information</td>
</tr>
</tbody>
</table>

Solved?

| How to track the children and young people (CYP)?                                         | Partial                                                                 |
| Why do I need to track them?                                                              | Yes                                                                   |
| Should we include the PIED?                                                               | Yes                                                                   |
| What happens to CYP who score over a certain score?                                       | Yes                                                                   |
| The ethical issues of offering internet based questionnaires                              | Yes                                                                   |
| The positive issues of offering internet questionnaires                                   | Yes                                                                   |
| The negative issues of offering internet based questionnaires                             | Partial                                                               |
| What happens if CYP put wrong code in?                                                    | Partial                                                               |
| Gender – just M & F, or should we put other, or I don’t want to say?                      | Yes                                                                   |
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<td>What personal information can we collect?</td>
<td>Research Governance Manager at local R&amp;D agreed we can collect gender, day and month of birth (did not want us to ask for year of birth). Also passed through ethics.</td>
<td>Yes</td>
</tr>
<tr>
<td>Could we follow a child from just their day and month of birth?</td>
<td>Hope so!, but if not hopefully code will be correct and gender added too</td>
<td>Yes</td>
</tr>
<tr>
<td>Do people want to receive a free pen, or do they prefer we spent the money on the study?</td>
<td>Mixed opinions, probably won’t include a pen as trying to keep costs down, and if complete online, wouldn’t need a pen anyway!</td>
<td>Yes</td>
</tr>
<tr>
<td>Should I use coloured paper?</td>
<td>Have decided to use some coloured paper to make postcard and cover letter stand out amongst usual family post, the pen ink was also varied for each centre to help me identify centre at a glance</td>
<td>Yes</td>
</tr>
<tr>
<td>Should I put the postcards into envelopes?</td>
<td>Strong views about this, so have decided too, to ensure further anonymity. Would not want anyone to read about someone having JDM if went through wrong door.</td>
<td>Yes</td>
</tr>
<tr>
<td>Should we give an option for them to opt out?</td>
<td>Research Governance Manager at local R&amp;D liked this idea, thought it made the project stronger, plus would make a smaller target audience, so the response rate may be better. Postcards approved by ethics.</td>
<td>Yes</td>
</tr>
<tr>
<td>Should we hand write the addresses on the envelopes?</td>
<td>Yes, if possible</td>
<td>Yes</td>
</tr>
<tr>
<td>How many times can I send reminders to people to complete?</td>
<td>Ethics did agree on the flowsheet, that we can contact those that don’t opt out twice. Whether local centres will have time and resources to do this is unknown.</td>
<td>Yes</td>
</tr>
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</table>
### Questions/thoughts/problems

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<tr>
<td>Do we send the postcards and questionnaires to just the child/parent or both?</td>
<td>Research Governance Manager in local R&amp;D felt strongly that needed to go to parents, rather than children, however, the questionnaire is actually for the child and young person, so compromised by saying we would address to both.</td>
<td>Yes</td>
</tr>
<tr>
<td>Which email address should we put on the postcards for the opting out?</td>
<td>Decided to use the JDRG one rather than my personal one, but will need to check this regularly. Sent test email to ensure this was working and could access the server.</td>
<td>Yes</td>
</tr>
<tr>
<td>Should we include a parent information sheet?</td>
<td>Some felt strongly there should be one explaining the risks and what happens if their CYP scores highly, so one was made and passed through ethics, also includes the families in the project, not secretive!</td>
<td>Yes</td>
</tr>
<tr>
<td>How long do we leave it before we send a reminder/call them?</td>
<td>This is set out for 2 weeks, but may be slightly longer after discussions with others. Will need to use some common sense and be aware of time restraints for local staff.</td>
<td>Yes</td>
</tr>
<tr>
<td>What do we do about each section not starting a fresh page?</td>
<td>Have managed to get around this by retyping the questionnaires into word, rather than using the OPINIO software which would not allow it, but managed to get both copies looking identical (finally)!</td>
<td>Yes</td>
</tr>
<tr>
<td>How do we add a header and footer?</td>
<td>Can do this on the paper ones, have added in a version of it on the electronic ones at the beginning, but took a long time to figure out how to create, including lots of calls to software helpline</td>
<td>Yes</td>
</tr>
<tr>
<td>How do we add logos?</td>
<td>Some of the tools have made this a requirement of using their tool, can do this easily on the paper ones easily, but the electronic ones were harder. Needed to have help from Information Technology experts!</td>
<td>Yes</td>
</tr>
<tr>
<td>Who can access the electronic one?</td>
<td>Those who are sent the link</td>
<td>Yes</td>
</tr>
<tr>
<td>Where is the list of who don’t want to be involved to be stored?</td>
<td>Will need to be stored with codes in UCL, as registry data currently stored in line with data protection rules and regulations.</td>
<td>Yes</td>
</tr>
<tr>
<td>What about the problems with children who change age outside a bracket within the study timeframe?</td>
<td>Will need to really plan this before hand and be aware of those under and over, and those that may change age. Will need to be very organised with this.</td>
<td>Yes</td>
</tr>
</tbody>
</table>
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<tbody>
<tr>
<td>How to make the packs age specific?</td>
<td>Will need to be organised and plan in advance, take time to make the packs up with right information sheets and questionnaire versions. The centres will need to send the right packs to the right centres, so codes will need to be right on them.</td>
<td>Yes</td>
</tr>
<tr>
<td>How do I get the packs to the local centres?</td>
<td>Recorded post to specified name and address or take to BSPAR meeting in Southampton in October.</td>
<td>Yes</td>
</tr>
<tr>
<td>Is the CUIS too hard to complete?</td>
<td>Worries with this tool now, but can justify why we use it, just aware there may be some feedback post study of it.</td>
<td>Yes</td>
</tr>
<tr>
<td>Are they too many pages?</td>
<td>Probably, but again better to be thorough, than miss things? Expect criticism later.</td>
<td>Yes</td>
</tr>
<tr>
<td>Do we encourage CYP to complete with their parents?</td>
<td>Have tried to encourage them to complete alone by wording of documents, but aware this may happen, can't exclude parents, leave open text boxes in case CYP or parent want to add comments.</td>
<td>Yes</td>
</tr>
<tr>
<td>Would a research passport help me to visit the local centres?</td>
<td>Mixed answers, probably not.</td>
<td>Yes</td>
</tr>
<tr>
<td>How long will it take to complete?</td>
<td>Realistically about 30-45 minutes</td>
<td>Yes</td>
</tr>
<tr>
<td>How do I find out how many CYP there are?</td>
<td>The registry. Should be able to get the most current information from the registry</td>
<td>Yes</td>
</tr>
<tr>
<td>Is it safe URL?</td>
<td>Have asked software company, have checked with UCL IT department, using UCL recommended package</td>
<td>Yes</td>
</tr>
<tr>
<td>Do we need to give the questionnaires individual titles?</td>
<td>Can’t easily on the electronic version, and surely both should be the same? Do we need too? Decided not to in the end.</td>
<td>Yes</td>
</tr>
</tbody>
</table>
### Appendix 12 – Questions/thoughts considered for phase 2

<table>
<thead>
<tr>
<th>Questions/thoughts/problems</th>
<th>Current progress/solutions/final decisions</th>
<th>Solved?</th>
</tr>
</thead>
<tbody>
<tr>
<td>Where does the copyright want the logos, front page or on the actual tool?</td>
<td>Will try and put on paper ones, but not so easy with electronic version, need to go with the actual questionnaires. Finally figured out in time and added.</td>
<td>Yes</td>
</tr>
<tr>
<td>How quickly can I let centres know if someone has scored highly on the PIED?</td>
<td>Have told all centres will be next working day, obviously will need to consider my schedule for this also, dependent on when we start sending out, but will need to clear time. Important this is done correctly, also to protect at risk young people.</td>
<td>Yes</td>
</tr>
<tr>
<td>Should we ask for a named individual at each title?</td>
<td>Yes, have done so on the TC, flowsheet and video. One of the first tasks to be completed. Luckily each centre happy to give me a clear name.</td>
<td>Yes</td>
</tr>
<tr>
<td>How do we ensure they all sent out at the same time, e.g. what happens if a research nurse is on leave?</td>
<td>Once receive name at each centre, will need to stay in close contact and ensure they are engaged and on board and disclose any time off coming up – also have back up name where possible, even if local PI. Will need to ask centres to try and address and send out at same time!</td>
<td>Partial</td>
</tr>
<tr>
<td>Parents have asked to advertise it on facebook, is this ok?</td>
<td>Checked with supervisory team. Yes, if they advertise it on my behalf and encourage parents to complete, and email me if any problems. I can’t view the group, so will need to ask for assistance with this.</td>
<td>Yes</td>
</tr>
<tr>
<td>What happens to those that have transitioned or are lost to follow up?</td>
<td>Ideally, if we can still chase them, then would be good too, but do expect to not be able to access all</td>
<td>Yes</td>
</tr>
<tr>
<td>How do I collate all the data and analyse it?</td>
<td>Can use the OPINO software if I type all the paper results in</td>
<td>Yes</td>
</tr>
<tr>
<td>What about the qualitative data obtained, how do we add this in?</td>
<td>Same as above</td>
<td>Yes</td>
</tr>
<tr>
<td>Questions/thoughts/problems</td>
<td>Current progress/solutions-final decisions</td>
<td>Solved?</td>
</tr>
<tr>
<td>-------------------------------------------------------------------------------------------</td>
<td>-------------------------------------------------------------------------------------------------------------</td>
<td>---------</td>
</tr>
<tr>
<td>There’s potentially 250+ CYP in London, that is a lot of work to make packs and address envelopes!</td>
<td>Just need to be organised! Need to start buying paper, envelopes, stamps etc and be organised.</td>
<td>Yes</td>
</tr>
<tr>
<td>What do I do if not many respond?</td>
<td>Keep trying. But have done best to try and encourage local centre engagement, we expected a low response rate, predicted at 35% on original funding proposal.</td>
<td>Partial</td>
</tr>
<tr>
<td>When should we start, sooner or take time and check issues are right</td>
<td>As soon as all above sorted</td>
<td>Partial</td>
</tr>
</tbody>
</table>
Appendix 13 - Flow sheet for local nurses

Flowsheet of Assistance required (in red) by local sites in small sub study ‘Psychosocial needs of Juvenile Dermatomyositis Survey’

Teleconference (TC) to discuss the study with all sites across UK on 12/09/18

Video clip of study emailed out to all centres with this flowsheet of responsibilities post TC

Each centre to provide a name of an individual to be the liaison for this small sub survey

Each centre will be sent an email detailing their number of JDM patients

Each centre be sent a pack of stamped postcards to be kindly addressed and sent out in the post to each of their children and young people with JDM, this postcard highlights the impending study and gives the family the opportunity to opt out via an email address

2 weeks later, each centre will be sent via recorded delivery (or hand delivered) a pack of questionnaires with a covering letter, instructions and SAE for each child and young person with JDM in a stamped envelope. The patient and parent information sheet will need printing onto local headed paper and then the envelopes will need to be addressed and sent out in the post please

3 weeks later, each centre will be contacted and asked to phone a given list of non-respondents to remind them about the survey

6 weeks after first survey sent out, each centre will be sent a repeat pack for the non-responders and asked to please address and send out in the post

As the questionnaire packs get sent back to P.Levermore by post or via the online web address, P.Levermore will assess them to score the Paediatric Index of Emotional Distress (PI-ED) questionnaire. If the results are above the clinical cut off, then P.Levermore will contact the named individual in each centre who can flag this with the Principal Investigator and discuss locally what to do for that young person

After the study, all local personal involved will be invited to a dissemination workshop to share the findings and discuss future interventions

Thank you for your support and help with this study survey

Should you have any further questions or comments please contact P.Levermore, Principal Investigator and Research Nurse for this study

Juvenile Dermatomyositis Cohort and Biomarker Study & Repository 99RU11
Psychosocial needs survey

Version 1. July 2018
Appendix 14 - Postcard sent to all in JDCBS

Thank you for being part of something special - the Juvenile Dermatomyositis Cohort and Biomarker Study (JDCBS). This big study is constantly carrying out research to improve young people’s lives with Juvenile Dermatomyositis (JDM). This might be in finding new blood or muscle markers, or whether to look at how some medicines work.

This postcard is to let you know that we are starting a new UK wide survey, asking children and young people to fill in some questionnaires to tell us what it is like to have JDM and how it makes them feel. Even if their JDM has gone away, we still want to know how they are now.

You will be getting these questionnaires very soon in the post, and your child/young person with JDM has the choice whether to fill in paper copies and send back to us in the envelope we send you, or to fill them in on a special website. There will be a letter explaining this further with your pack.

The original consent form you signed for the JDCBS asked for your permission for us to contact you for future studies, such as this one, but if you don’t want us to send you these questionnaires then that is fine. If so, please email [email protected] and we won’t send you them. Or email this address for any questions you may have.

Thank you very much for reading this and considering our study.
Appendix 15 - 8-12 phase 2 information sheet

Appendix 15 - 8-12 phase 2 information sheet

416


A SURVEY ‘ASSESSING YOUR NEEDS IN JUVENILE DERMATOMYOSITIS (JDM)’

BRIEF SUMMARY
Juvenile Dermatomyositis is often called JDM. We want to know what it is like for YOU to have JDM. If we understand how it makes young people feel, then we can help you and other people like you. Only those with JDM can tell us what it is like and that is why we would like you to fill in the following questions. This study is for all young people around the United Kingdom (UK) and will look at all of the results at the end to see what we can improve upon.

WHAT’S INVOLVED?
From talking to children and young people with JDM, we know there are some thoughts and feelings from having JDM which are shared. We therefore want to give everyone with JDM (between the ages of 8 and 19 years) around the United Kingdom (UK) a chance to say what they think and whether these same things are important to you or felt by you.

WHAT HAPPENS IF I DO WANT TO DO IT?
By doing these questionnaires, you get the chance to say what it is like to have JDM and help us work out how to make things better for you and others like you. Also, everyone who sends back completed questionnaires will be entered into a prize draw to win £100 Amazon vouchers.

WHAT DO I NEED TO DO?
You can choose whether to fill the questionnaires on the computer and then the answers come straight to us, or whether to fill them in on paper and post them to us in the envelope included.
Appendix 16 - 13-19 phase 2 information sheet

Psychosocial needs survey  Child and Young person (13-19yrs) Information sheet  Version 1  July 18

We will not ask for your name, but to know who has sent one back so they can be entered into the prize draw, there will be a code with this pack on your invitation letter, which will need to be entered onto the website. We will also ask for day and month of birth as a back-up system, just in case the code gets entered in wrongly. The website is called Qipio and the personal information that you give for this survey will only be used for the purposes of the survey.

Please also only fill one copy in. If you fill the paper ones in, then you do not need to fill the computer ones in too. Take your time to read the instructions on how to fill them in on the instruction page. It is fine to ask your parent or carer to help explain what things mean to you, but we do want your honest answers.

**DO I HAVE TO DO IT?**

NO, You DO NOT have to fill these in if you don’t want to, it won’t affect your care. Just don’t send them back to us.

**WHAT ARE THE RISKS OF DOING IT?**

Filling in the questionnaires will take up some of your time. There are X pages and from practicing it, we think this would take you about 30-45 minutes if you take your time. We hope answering these questions won’t upset you, but if they do, please talk to your parents, a teacher or one of your nurses or doctors who look after you. You can also speak to______, another of the people who is running the study, her email address is______, _________ and she can speak to your parent or carer, local doctor or nurse to get you some help.

**WHAT WILL HAPPEN TO WHAT I SAY?**

Your answers will be not be shared unless you tell us that you are very unhappy or worried or that we need to help you with anything, if so than we may need to discuss this with your local doctor or nurse and your parent to help you.

**WHAT HAPPENS TO THE RESULTS?**

After the study has finished, you and your family will get a newsletter telling you about everyone’s results. You won’t get your own results back. This newsletter will tell you what we have found and how we want to improve things in the future.

Thank you very much

Juvenile Dermatomyositis Cohort and Biomarker Study & Repository 99RU11
### Appendix 17 - Validated questionnaire summary

<table>
<thead>
<tr>
<th>Tool</th>
<th>Country of Origin</th>
<th>Domains / questions</th>
<th>Range of score</th>
<th>Scoring implication</th>
</tr>
</thead>
<tbody>
<tr>
<td>PedsQL</td>
<td>US</td>
<td>2 main Individual domains:</td>
<td>0-100</td>
<td>Lower scores = worse quality of life</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Physical total Score</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Psychological total score</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>(can be broken down into Emotional/Social/School)</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Or overall Total score</td>
<td></td>
<td></td>
</tr>
<tr>
<td>PedsQL Rheumatology Module</td>
<td>US</td>
<td>5 domains scored individually:</td>
<td>0-100</td>
<td>Lower scores = worse Rheumatology related quality of life</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Pain</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Activities of daily living</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Worry</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Treatment</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Communication</td>
<td></td>
<td></td>
</tr>
<tr>
<td>PI-ED</td>
<td>UK</td>
<td>Overall score</td>
<td>0-42</td>
<td>Higher scores = greater emotional distress</td>
</tr>
<tr>
<td>CUIS</td>
<td>US</td>
<td>Overall score</td>
<td>23-115</td>
<td>Higher scores = greater perception of uncertainty</td>
</tr>
<tr>
<td>BBSC</td>
<td>US</td>
<td>Score for Burden and separate score for Benefit</td>
<td>10-50</td>
<td>Higher scores = greater perceived burden and/or benefit</td>
</tr>
</tbody>
</table>
## Appendix 18 - Missing data

<table>
<thead>
<tr>
<th>Question</th>
<th>N</th>
<th>Missing</th>
<th>% complete response</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Demographic questions</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sex</td>
<td>123</td>
<td>0</td>
<td>100</td>
</tr>
<tr>
<td>Age</td>
<td>123</td>
<td>0</td>
<td>100</td>
</tr>
<tr>
<td>Years since Diagnosis</td>
<td>120</td>
<td>3</td>
<td>98</td>
</tr>
<tr>
<td>Difference in years between final visit and questionnaire completion</td>
<td>117</td>
<td>6</td>
<td>95</td>
</tr>
<tr>
<td><strong>Validated questions</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Physical Score</td>
<td>123</td>
<td>0</td>
<td>100</td>
</tr>
<tr>
<td>Psychosocial score</td>
<td>123</td>
<td>0</td>
<td>100</td>
</tr>
<tr>
<td>Total score</td>
<td>123</td>
<td>0</td>
<td>100</td>
</tr>
<tr>
<td>Pain total</td>
<td>123</td>
<td>0</td>
<td>100</td>
</tr>
<tr>
<td>Worry total</td>
<td>123</td>
<td>0</td>
<td>100</td>
</tr>
<tr>
<td>Communication total</td>
<td>123</td>
<td>0</td>
<td>100</td>
</tr>
<tr>
<td>Treatment total</td>
<td>123</td>
<td>0</td>
<td>100</td>
</tr>
<tr>
<td>ADL total</td>
<td>123</td>
<td>0</td>
<td>100</td>
</tr>
<tr>
<td>Question</td>
<td>N</td>
<td>Missing</td>
<td>% complete response</td>
</tr>
<tr>
<td>----------------------------------</td>
<td>-----</td>
<td>---------</td>
<td>---------------------</td>
</tr>
<tr>
<td>PIED total</td>
<td>123</td>
<td>0</td>
<td>100</td>
</tr>
<tr>
<td>CUIS total</td>
<td>123</td>
<td>0</td>
<td>100</td>
</tr>
<tr>
<td>Burden total</td>
<td>123</td>
<td>0</td>
<td>100</td>
</tr>
<tr>
<td>Benefit total</td>
<td>123</td>
<td>0</td>
<td>100</td>
</tr>
<tr>
<td>Bespoke questions</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Do you still have JDM?</td>
<td>123</td>
<td>0</td>
<td>100</td>
</tr>
<tr>
<td><strong>Filter question</strong> - if yes, how bad is it?</td>
<td>90</td>
<td>33</td>
<td>73</td>
</tr>
<tr>
<td>Have you met anyone else with JDM?</td>
<td>122</td>
<td>1</td>
<td>99</td>
</tr>
<tr>
<td>Would you like too?</td>
<td>122</td>
<td>1</td>
<td>99</td>
</tr>
<tr>
<td>Can people see it?</td>
<td>122</td>
<td>1</td>
<td>99</td>
</tr>
<tr>
<td>Is it good or bad for people to see it?</td>
<td>100</td>
<td>23</td>
<td>81</td>
</tr>
</tbody>
</table>

Range from 81-100% complete response (not including the filter question)
Appendix 19 – Scatterplots of outcome measures

Scatterplot matrix of outcome measures
Appendix 20 - Scatterplots of Rheumatology Module

Scatterplot Matrix for Rheumatology Module Variables

- Pain_total
- ADL_total
- Treatment_total
- Worry_total
- Communication_total
## Question 1 – “Do you think you still have JDM?”

<table>
<thead>
<tr>
<th>Comment</th>
<th>Coding</th>
</tr>
</thead>
<tbody>
<tr>
<td>No</td>
<td>I’m not sure whether I do</td>
</tr>
<tr>
<td>No</td>
<td>Not a facet of my life now</td>
</tr>
<tr>
<td>No</td>
<td>No issues since signed off 10 years ago</td>
</tr>
<tr>
<td>No</td>
<td>No symptoms anymore</td>
</tr>
<tr>
<td>No</td>
<td>Off all meds, no flares, I don’t have JDM</td>
</tr>
<tr>
<td>No</td>
<td>Been OK for past 5 years</td>
</tr>
<tr>
<td>No</td>
<td>No problems for 10 years</td>
</tr>
<tr>
<td>No</td>
<td>Drug and symptom free since I was 5</td>
</tr>
<tr>
<td>No</td>
<td>No longer on any meds, go out with my friends</td>
</tr>
<tr>
<td>No</td>
<td>I was discharged a year ago</td>
</tr>
<tr>
<td>No</td>
<td>No, because I’m not feeling pain</td>
</tr>
<tr>
<td>No</td>
<td>I only had it when I was a baby/toddler</td>
</tr>
<tr>
<td>Yes</td>
<td>Have a rash and drs told me I have it</td>
</tr>
<tr>
<td>Yes</td>
<td>I have new calcinosis lumps appearing</td>
</tr>
<tr>
<td>Yes</td>
<td>It is present on my face</td>
</tr>
<tr>
<td>Yes</td>
<td>I occasionally get joint pains</td>
</tr>
<tr>
<td>Yes</td>
<td>Still get pains</td>
</tr>
<tr>
<td>Yes</td>
<td>I think I do because I get tired and ache a lot and haven’t got the stamina</td>
</tr>
<tr>
<td>Yes</td>
<td>In a way as I don’t feel 100%</td>
</tr>
<tr>
<td>Yes</td>
<td>Abit, because I get tired easily like I did when I was first told</td>
</tr>
<tr>
<td>Yes</td>
<td>Because I still get some aches / pains in my legs and still on medicine!</td>
</tr>
<tr>
<td>Yes</td>
<td>My skin has not gone away yet</td>
</tr>
<tr>
<td>Yes</td>
<td>My papules went away</td>
</tr>
<tr>
<td>Comment</td>
<td>Coding</td>
</tr>
<tr>
<td>------------------------------------------------------------------------</td>
<td>---------------------</td>
</tr>
<tr>
<td>Yes The symptoms link to what the doctors told me</td>
<td>Symptoms</td>
</tr>
<tr>
<td>Yes Relapsed in Nov 17, now back on meds</td>
<td>Medications</td>
</tr>
<tr>
<td>Yes Because I am still on Medicines</td>
<td>Medications</td>
</tr>
<tr>
<td>Yes I am still taking medicines</td>
<td>Medications</td>
</tr>
<tr>
<td>Yes I would like to fully come off Methotrexate to see how it affects me</td>
<td>Medications</td>
</tr>
<tr>
<td>Yes I know I still have it which is why I take the medication</td>
<td>Medications</td>
</tr>
<tr>
<td>Yes Because I still have my injections</td>
<td>Medications</td>
</tr>
<tr>
<td>Yes I have become much better than before, but still on medicines</td>
<td>Medications</td>
</tr>
<tr>
<td>Yes Will have for the rest of my life</td>
<td>Duration</td>
</tr>
<tr>
<td>Yes I will always have it</td>
<td>Duration</td>
</tr>
<tr>
<td>Yes In remission</td>
<td>Duration</td>
</tr>
<tr>
<td>Yes Because I will always have to think about it and worry of it is affecting me more than a normal person</td>
<td>Duration</td>
</tr>
<tr>
<td>Yes But I am in remission and have been for quite a few years</td>
<td>Duration</td>
</tr>
<tr>
<td>Yes As its winter it isn’t as bad but I know that when summer comes it might get bad again</td>
<td>Duration</td>
</tr>
<tr>
<td>Yes I have been in remission since October</td>
<td>Duration</td>
</tr>
<tr>
<td>Yes I know that my condition is life long and there is no cure yet</td>
<td>Duration</td>
</tr>
<tr>
<td>Yes I have hit a plateau, I’m not getting better and I’m not getting worse</td>
<td>Duration</td>
</tr>
<tr>
<td>Yes Went to see my consultant</td>
<td>Other</td>
</tr>
<tr>
<td>Yes I hate JDM</td>
<td>Other</td>
</tr>
<tr>
<td>Yes I know I do</td>
<td>Other</td>
</tr>
<tr>
<td>Yes Because last time I went to the hospital they told me I still have JDM</td>
<td>Other</td>
</tr>
<tr>
<td>Don’t know I’m at a stage when all the comments from my doctors are positive but they don’t say whether I have the illness anymore so that leaves me unsure and every now and then in myself I don’t feel 100% or I notice something, I don’t know whether that means I’m still affected by my JDM or if that’s just me over reacting and being paranoid</td>
<td>Confusion</td>
</tr>
<tr>
<td>Comment</td>
<td>Coding</td>
</tr>
<tr>
<td>---------</td>
<td>--------</td>
</tr>
<tr>
<td>I don’t know because it still plays in the back of my mind and worries me of the chance of coming back</td>
<td>Confusion</td>
</tr>
<tr>
<td>Because I am very well now and don’t feel my JDM that much, it makes me question if I still have it, even though the doctors do still say that I do. Also, when I don’t have my treatment for a while, I still feel really well, which makes me confused and want to do physio and have my infusions less</td>
<td>Confusion</td>
</tr>
<tr>
<td>I have no symptoms but is always in the back of my mind</td>
<td>Confusion</td>
</tr>
<tr>
<td>They say I don’t have to come back for a year. I don’t know?</td>
<td>Confusion</td>
</tr>
<tr>
<td>I am unsure, I still get aches and pains, I’m still tired a lot of the time and I can’t join in many activities so apart of me thinks yes, a part of me thinks no</td>
<td>Confusion</td>
</tr>
<tr>
<td>I know there is a problem but in really good days, I feel like people just exaggerate my JDM</td>
<td>Confusion</td>
</tr>
<tr>
<td>Although I have been taken off the medicine as there is no sign of 'active disease' like before, I still have calcinosis and obvious scars which remind me I’m not back to ‘normal’</td>
<td>Symptoms</td>
</tr>
<tr>
<td>I’m still having treatment but I don’t feel effects from them and I don’t have any problems from JDM</td>
<td>Symptoms</td>
</tr>
<tr>
<td>I still have aches in my muscles and joints</td>
<td>Symptoms</td>
</tr>
<tr>
<td>As the main illness – no, but symptoms re-occur in conjunction with arthritis</td>
<td>Symptoms</td>
</tr>
<tr>
<td>I am off all my medicines but I still get some of my symptoms now and again</td>
<td>Symptoms</td>
</tr>
<tr>
<td>I don’t need medication but sometimes I feel achey and tired</td>
<td>Symptoms</td>
</tr>
<tr>
<td>I am in remission</td>
<td>Remission</td>
</tr>
<tr>
<td>In remission</td>
<td>Remission</td>
</tr>
</tbody>
</table>
### Appendix 22 - Comments in response to Q8

<table>
<thead>
<tr>
<th>Q8. What is the worst thing about having JDM?</th>
<th>Coding Framework</th>
</tr>
</thead>
<tbody>
<tr>
<td>For me, the worst thing is fatigue because whenever I come home from school I'm exhausted, and don't want to go to a friend's house after school and I have to take a rest. Also, when my friends are playing something that involves a lot of exercise, I get tired.</td>
<td>Primary</td>
</tr>
<tr>
<td></td>
<td>Secondary</td>
</tr>
<tr>
<td></td>
<td>Tertiary</td>
</tr>
<tr>
<td></td>
<td>Quarternary</td>
</tr>
<tr>
<td>For me, the worst thing is fatigue because whenever I come home from school I'm exhausted, and don't want to go to a friend's house after school and I have to take a rest. Also, when my friends are playing something that involves a lot of exercise, I get tired.</td>
<td>Tired</td>
</tr>
<tr>
<td></td>
<td>Being different</td>
</tr>
<tr>
<td>getting tired easily</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Tired</td>
</tr>
<tr>
<td>everything</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Mental health effects</td>
</tr>
<tr>
<td>Cant do things that other people can do and having to go to hospital</td>
<td>Being different</td>
</tr>
<tr>
<td></td>
<td>Limitations due to JDM</td>
</tr>
<tr>
<td></td>
<td>Hospital visits</td>
</tr>
<tr>
<td>The pain, going to the hospital, missing school and my friends. The treatment, it make my head hurt.</td>
<td>Pain</td>
</tr>
<tr>
<td></td>
<td>Hospital visits</td>
</tr>
<tr>
<td></td>
<td>Treatment</td>
</tr>
<tr>
<td></td>
<td>Missing school</td>
</tr>
<tr>
<td>Not being able to do things</td>
<td>Limitations due to JDM</td>
</tr>
<tr>
<td>I couldn't sit down properly</td>
<td>Limitations due to JDM</td>
</tr>
<tr>
<td>Calcinosis Weakness in muscles Unable to walk Taking so many medicines</td>
<td>Calcinosis</td>
</tr>
<tr>
<td></td>
<td>Weakness</td>
</tr>
<tr>
<td></td>
<td>Limitations due to JDM</td>
</tr>
<tr>
<td></td>
<td>Treatment</td>
</tr>
<tr>
<td>steroid injections</td>
<td>Treatment</td>
</tr>
<tr>
<td>Q8. What is the worst thing about having JDM?</td>
<td>Primary</td>
</tr>
<tr>
<td>---------------------------------------------</td>
<td>---------</td>
</tr>
<tr>
<td>I do not like doing injection because of the metal taste it puts in my mouth for two days</td>
<td>Treatment</td>
</tr>
<tr>
<td>Being different to others</td>
<td>Being different</td>
</tr>
<tr>
<td>Having Methotrexate</td>
<td>Treatment</td>
</tr>
<tr>
<td>Not knowing if it will get worse for example I have to wait 3 months for an MRI scan and I don't know if it will get worse in those 3 months.</td>
<td>Uncertainty</td>
</tr>
<tr>
<td>My skin and frequently going to the doctor</td>
<td>Rashes</td>
</tr>
<tr>
<td>Having Methotrexate injection Not going out in the sun</td>
<td>Treatment</td>
</tr>
<tr>
<td>Aching painful physio</td>
<td>Physiotherapy</td>
</tr>
<tr>
<td>That I know I can't do what most people can and the pain that hurts a little bit is annoying</td>
<td>Limitations due to JDM</td>
</tr>
<tr>
<td>Sun cream makes you look shiny and you can't go to some places. School can be dangerous when others are sick.</td>
<td>Sun protection</td>
</tr>
<tr>
<td>blood tests have to be careful around ill people</td>
<td>Blood tests</td>
</tr>
<tr>
<td>Having to be stuck on a drip in hospital all day.</td>
<td>Treatment</td>
</tr>
</tbody>
</table>
### Appendix 22 - Comments in response to Q8

<table>
<thead>
<tr>
<th>Q8. What is the worst thing about having JDM?</th>
<th>Primary</th>
<th>Secondary</th>
<th>Tertiary</th>
<th>Quarternary</th>
</tr>
</thead>
<tbody>
<tr>
<td>I remember that I couldn't move properly without support.</td>
<td>Limitations due to JDM</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Being in pain, lack of activities</td>
<td>Pain</td>
<td>Limitations due to JDM</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Taking my medicines</td>
<td>Treatment</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Having some muscle, blood tests, injections and medication</td>
<td>Treatment</td>
<td>Blood tests</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Not being able to physically keep up with people and the aches or pains.</td>
<td>Lack of stamina</td>
<td>Pain</td>
<td>Being different</td>
<td></td>
</tr>
<tr>
<td>The aches and pains I get.</td>
<td>Pain</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Its a waste of life</td>
<td>Mental health effects</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Being limited to certain abilities</td>
<td>Limitations due to JDM</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>METHOTREXATE IS THE WORSE THING, BECAUSE I HATE THE WAY IT BURNS AND IT MAKES ME FEEL SICK. I ALSO REALLY DON'T LIKE PUTTING SUN CREAM ON EVERY DAY. BUT I CAN COPE WITH THIS.</td>
<td>Methotrexate sickness</td>
<td>Treatment</td>
<td>Sun protection</td>
<td></td>
</tr>
<tr>
<td>It is harder to do thing you want to do. Get tired more than others.</td>
<td>Limitations due to JDM</td>
<td>Tired</td>
<td></td>
<td></td>
</tr>
<tr>
<td>? injections</td>
<td>Treatment</td>
<td></td>
<td></td>
<td></td>
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<th>Tertiary</th>
<th>Quarternary</th>
</tr>
</thead>
<tbody>
<tr>
<td>Not having enough stamina.</td>
<td>Lack of stamina</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>The blood tests</td>
<td>Blood tests</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Don't know</td>
<td>Don't know</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Having to wear a hat in summer because I'm always scared someone will make fun of me</td>
<td>Sun protection</td>
<td>Being different</td>
<td>Mental health effects</td>
<td></td>
</tr>
<tr>
<td>that you can't do much and feel lots of pain</td>
<td>Limitations due to JDM</td>
<td>Pain</td>
<td></td>
<td></td>
</tr>
<tr>
<td>You get tired easily and can't do things.</td>
<td>Tired</td>
<td>Limitations due to JDM</td>
<td>Mental health effects</td>
<td></td>
</tr>
<tr>
<td>I can't keep up with my friends and not always nice because of this. Friends see it as an excuse when I can't do something/ have to do something different to the. They don't understand it and think its like being ill for a day.</td>
<td>Being different</td>
<td>Lack of understanding</td>
<td>Mental health effects</td>
<td></td>
</tr>
<tr>
<td>having to take all the tablets</td>
<td>Treatment</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Before treatment I was unable to move and my family had to do everything for me</td>
<td>Limitations due to JDM</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>In the worse times not being able to play like everyone else and being in pain</td>
<td>Limitations due to JDM</td>
<td>Pain</td>
<td></td>
<td></td>
</tr>
<tr>
<td>The effects of losing the ability of doing what you used to do. And the pain in your muscles.</td>
<td>Limitations due to JDM</td>
<td>Pain</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
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<th>Primary</th>
<th>Secondary</th>
<th>Tertiary</th>
<th>Quarternary</th>
</tr>
</thead>
<tbody>
<tr>
<td>At first you don't see the effect it has but it does change and limit what you can do and only after you are diagnosed you realise. You feel sad you can't do the things you used to and uncomfortable questions are sometimes asked.</td>
<td>Limitations due to JDM</td>
<td>Mental health effects</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Probably having aches and pains and staying in the hospital and also the treatment.</td>
<td>Pain</td>
<td>Hospital visits</td>
<td>Treatment</td>
<td></td>
</tr>
<tr>
<td>The fact that you never know when it will act up</td>
<td>Uncertainty</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Getting tired quicker than usual. I love to do things that require a lot of energy and I feel left out if I can't do anything I like or I used to do</td>
<td>Tired</td>
<td>Limitations due to JDM</td>
<td>Being different</td>
<td></td>
</tr>
<tr>
<td>Injections and liquid medicine.</td>
<td>Treatment</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>not be able to do what any other 14 year old girl dose</td>
<td>Limitations due to JDM</td>
<td>Being different</td>
<td></td>
<td></td>
</tr>
<tr>
<td>not being able to have a high endurance in physical exercise</td>
<td>Lack of stamina</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I used to walk nearly everywhere when me and my father were visiting family, friends, or just going to his place to spend time together. Though the walk isn't what I miss, it's more the whole saving money from buses that I liked. Otherwise, the methotrexate injections in the muscle of my thighs usually hurt like hell. Mostly cause I was concentrating on it too much.</td>
<td>Lack of stamina</td>
<td>Treatment</td>
<td>Mental health effects</td>
<td></td>
</tr>
<tr>
<td>Not being able do do as much as i used to. Feeling scared and a burden all the time. Hating how i look after all the medicine.</td>
<td>Limitations due to JDM</td>
<td>Mental health effects</td>
<td>Body image</td>
<td></td>
</tr>
</tbody>
</table>
## Appendix 22 - Comments in response to Q8

<table>
<thead>
<tr>
<th>Q8. What is the worst thing about having JDM?</th>
</tr>
</thead>
<tbody>
<tr>
<td>Not being able to join in all physical activities, having a ‘smaller’ body than everyone else my age</td>
</tr>
<tr>
<td>Lack of stamina</td>
</tr>
<tr>
<td>Being different</td>
</tr>
<tr>
<td>Body image</td>
</tr>
<tr>
<td>Physical disfigurement without a doubt. The mental part stems from this, I just wish I could have it removed</td>
</tr>
<tr>
<td>Lack of stamina</td>
</tr>
<tr>
<td>Being different</td>
</tr>
<tr>
<td>Body image</td>
</tr>
<tr>
<td>Mental health effects</td>
</tr>
<tr>
<td>That I can’t do as much as my friends such as football</td>
</tr>
<tr>
<td>Limitations due to JDM</td>
</tr>
<tr>
<td>Not being sure what I can/can’t do</td>
</tr>
<tr>
<td>Uncertainty</td>
</tr>
<tr>
<td>Not knowing when it can come back.</td>
</tr>
<tr>
<td>Uncertainty</td>
</tr>
<tr>
<td>Not having strength. Losing weight. Rashes on face and body.</td>
</tr>
<tr>
<td>Lack of stamina</td>
</tr>
<tr>
<td>Body image</td>
</tr>
<tr>
<td>Rashes</td>
</tr>
<tr>
<td>Obvious symptoms like redness pigmentation on my eyelids. Also having painful joints.</td>
</tr>
<tr>
<td>Rashes</td>
</tr>
<tr>
<td>Body image</td>
</tr>
<tr>
<td>Pain</td>
</tr>
<tr>
<td>Back then was not being able to move as fast as I wanted</td>
</tr>
<tr>
<td>Limitations due to JDM</td>
</tr>
<tr>
<td>The effect it can have on my family and me. The idea that my body is attacking itself and no-one knows why</td>
</tr>
<tr>
<td>Being different</td>
</tr>
<tr>
<td>Mental health effects</td>
</tr>
<tr>
<td>Not being able to do things that everyone else can do</td>
</tr>
<tr>
<td>Limitations due to JDM</td>
</tr>
<tr>
<td>Being different from other people.</td>
</tr>
<tr>
<td>Being different</td>
</tr>
</tbody>
</table>
**Appendix 22 - Comments in response to Q8**

<table>
<thead>
<tr>
<th>Q8. What is the worst thing about having JDM?</th>
<th>Primary</th>
<th>Secondary</th>
<th>Tertiary</th>
<th>Quarternary</th>
</tr>
</thead>
<tbody>
<tr>
<td>don't want to seem disadvantaged to others, and i don't want it to get worse, therefor affect me in the future</td>
<td>Uncertainty</td>
<td>Mental health effects</td>
<td></td>
<td></td>
</tr>
<tr>
<td>The uncertainty of not knowing if you are ever going to completely recover or if you'll go down hill again. How you can have different days on how you're feeling. Having to explain to others why you're tired or feeling the way you are and when they don't understand.</td>
<td>Uncertainty</td>
<td>Mental health effects</td>
<td>Tired</td>
<td>Lack of understanding</td>
</tr>
<tr>
<td>Having weekly injections</td>
<td>Treatment</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Knowing it will, never go away or that all your treatment is guesswork</td>
<td>Uncertainty</td>
<td>Treatment</td>
<td>Mental health effects</td>
<td></td>
</tr>
<tr>
<td>When I had the treatment made me lose my hair</td>
<td>Treatment</td>
<td>Body image</td>
<td></td>
<td></td>
</tr>
<tr>
<td>The worst thing when I was first trying to get diagnosed was when I couldn't clench my fists properly and when I couldn't run or walk as fast or as much as my friends, and when I couldn't get myself off the floor, so I had to be carried and helped off the floor a lot of the time.</td>
<td>Treatment</td>
<td>Limitations due to JDM</td>
<td>Weakness</td>
<td></td>
</tr>
<tr>
<td>MISSING SCHOOL WHICH HAD AFFECTED MY GRADES. GETTING TIRED EASILY. TAKING AND BEING ON MEDICATION AS SOMETIMES YOU FORGET AND SOMETIMES YOU DON'T WANT TO TAKE IT IN FRONT OF OTHER PEOPLE. AND THE WORST THING IS THAT BECAUSE IT IS VERY NOTICEABLE IT LOWERS MY CONFIDENCE, I'M SELF CONSCIOUS AND I CAN'T WEAR THINGS LIKE V NECKS</td>
<td>Missing school</td>
<td>Tired</td>
<td>Being different</td>
<td>Treatment</td>
</tr>
</tbody>
</table>
Appendix 22 - Comments in response to Q8

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<th>Quarternary</th>
</tr>
</thead>
<tbody>
<tr>
<td>Leg pain and taking foul medicines (methotrexate oral and steroids) and being sick during treatment</td>
<td>Pain</td>
<td>Treatment</td>
<td>Methotrexate sickness</td>
<td></td>
</tr>
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<td>The worst thing when I was first trying to get diagnosed was when I couldn't clench my fists properly and when I couldn't run or walk as fast or as much as my friends, and when I couldn't get myself off the floor, so I had to be carried and helped off the floor a lot of the time.</td>
<td>Limitations due to JDM</td>
<td>Weakness</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Because its internal people often don't believe me, because there's nothing physically wrong and it puts you down and think bad about yourself, especially when you don't know why you have it like a cause</td>
<td>Invisibility</td>
<td>Uncertainty</td>
<td>Mental health effects</td>
<td></td>
</tr>
<tr>
<td>Methotrexate because it makes me feel extremely nauseous and even the ondansetron don't help very much. I am getting used to having the injection, but it's taken me 2 years</td>
<td>Treatment</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Leg pain and taking foul medicines (methotrexate oral and steroids) and being sick during treatment</td>
<td>Pain</td>
<td>Treatment</td>
<td>Methotrexate sickness</td>
<td></td>
</tr>
</tbody>
</table>
Q8. What is the worst thing about having JDM?

<table>
<thead>
<tr>
<th>Under representation and lack of awareness</th>
<th>Lack of understanding</th>
</tr>
</thead>
</table>

The worst thing about living with JDM is not the pain or the numerous hospital visits, but the not knowing. It is the fear of the unknown. There is not enough knowledge about JDM to know whether you will ever be cured, or whether you will get worse, or whether this treatment will actually work for you. There is no clear-cut pathway to recovery to motivate you to take your medication and to keep doing the physio exercises even though they’re painful. It might all be for nothing. I just want to know what it feels like to go to sleep and then wake up in the morning and actually feel rested. I want to know what it feels like to not have a pain lingering on your bones like a constant background noise. When you get ill from a young age, you live your life constantly grieving the loss of a life you never got the chance to live. Which sounds ridiculous, because how can you grieve something you never experienced? But it is in the times between the bad days, or the fleeting moments of remission, when your mind begins to wonder whether your life could have been so much different had you been dealt a different hand. Ultimately, I think that when you live with a chronic illness, your body is never truly yours. Your body is not your own because it is always being controlled by something else: the disease, the medication, the doctors at the hospital. You feel trapped within this vessel you inhabit.
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<tr>
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<th>Tertiary</th>
<th>Quarternary</th>
</tr>
</thead>
<tbody>
<tr>
<td>In short words, I would probably say that the worst thing is not being able to be as normal as other teenagers. Everyday, I am reminded of JDM, and even when I am home, I still don't have 100% freedom as I know that I'll be in hospital the next month with doctors making decisions about me. This then brings in the not knowing and lack of control as to what will happen next, which is really difficult with physio and treatment, probably because I am getting older and feel well. This then also brings JDM into being a mind game - one minute you're fine the next minute you're not, or you might currently be fully fine (its all so confusing).</td>
<td>Being different</td>
<td>Uncertainty</td>
<td>Mental health effects</td>
<td></td>
</tr>
<tr>
<td>Under representation and lack of awareness</td>
<td>Lack of understanding</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Restricted in some areas</td>
<td>Limitations due to JDM</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Probably taking so much medication with bad side effects. It can have a very negative impact on your physical and mental health. Also being immune suppressed means I get ill a lot and therefore miss out on a lot eg school on the past and work now.</td>
<td>Treatment</td>
<td>Mental health effects</td>
<td>Immunosuppression</td>
<td>Missing school</td>
</tr>
<tr>
<td>You don't really know what is going to happen</td>
<td>Uncertainty</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>When you feel helpless and like you have no control of yourself. The fact that no one understands and that your family is affected.</td>
<td>Mental health effects</td>
<td>Lack of understanding</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Not having a prognosis, ie will it reoccur? Will it go away?</td>
<td>Uncertainty</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Aching / Not able to do what others can.</td>
<td>Pain</td>
<td></td>
<td>Limitations due to JDM</td>
<td></td>
</tr>
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<th>Tertiary</th>
<th>Quarternary</th>
</tr>
</thead>
<tbody>
<tr>
<td>Not knowing how it still effects my life and not knowing if I will fall ill again.</td>
<td>Uncertainty</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Having a rash Methotrexate made me sick</td>
<td>Rashes</td>
<td>Methotrexate sickness</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Being so obviously different but no one can see it.</td>
<td>Being different</td>
<td>Invisibility</td>
<td></td>
<td></td>
</tr>
<tr>
<td>The side effects of JDM because when I was younger I was bullied for the way I look and that I couldn't do everything they could do</td>
<td>Treatment</td>
<td>Limitations due to JDM</td>
<td>Body image</td>
<td></td>
</tr>
<tr>
<td>The worse thing is that there's not much knowledge about the illness. Not everyone understands what it's like and how it effects people</td>
<td>Treatment</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>The injections</td>
<td>Treatment</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>all the hospital and doctor appointments</td>
<td>Hospital visits</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hair loss and how the methotrexate makes me feel ill for the day after</td>
<td>Body image</td>
<td>Treatment</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Can't do as much</td>
<td>Limitations due to JDM</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>It is undependable - you could be in pain when you have something very important on.</td>
<td>Uncertainty</td>
<td>Invisibility</td>
<td></td>
<td></td>
</tr>
<tr>
<td>People don't understand what it is and because people can't see JDM they think you are being pathetic or lying. I also get very worried about it coming back and never being able to do anything</td>
<td>Lack of understanding</td>
<td>Uncertainty</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
### Appendix 22 - Comments in response to Q8

<table>
<thead>
<tr>
<th>Q8. What is the worst thing about having JDM?</th>
<th>Primary</th>
<th>Secondary</th>
<th>Tertiary</th>
<th>Quarternary</th>
</tr>
</thead>
<tbody>
<tr>
<td>I become fat!</td>
<td>Body image</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Stiffness in the hands. Medicine makes me sick - methotrexate</td>
<td>Limitations due to JDM</td>
<td>Methotrexate sickness</td>
<td></td>
<td></td>
</tr>
<tr>
<td>I'm not too sure</td>
<td>Don't know</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>it was feeling useless and not being able to do things</td>
<td>Mental health effects</td>
<td>Limitations due to JDM</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Having rashes at certain points that are visible so people think you have something contagious or a serious skin condition and judge you</td>
<td>Rashes</td>
<td>Being different</td>
<td></td>
<td></td>
</tr>
<tr>
<td>1) The inability to see friends while in hospital as the hospital is far away and it often gets lonely in the hospital. 2) It damages the skin cells and it isn't appealing to look at from other point of view.</td>
<td>Hospital visits</td>
<td>Body image</td>
<td></td>
<td></td>
</tr>
<tr>
<td>sometimes you get aches and feel tired.</td>
<td>Pain</td>
<td>Tired</td>
<td></td>
<td></td>
</tr>
<tr>
<td>The unknown - never knowing if a pain is JDM or normal, not having a clear/certain picture of the future/how well I will be. Having to explain to people why I am and look well but still have to go about things differently or not do some things as preventative measures eg not being tired / or going in the sun etc</td>
<td>Uncertainty</td>
<td>Limitations due to JDM</td>
<td>Being different</td>
<td>Sun protection</td>
</tr>
<tr>
<td>Not being able to do normal things by yourself: - go to the toilet - walk up the stairs - pick things up, etc</td>
<td>Limitations due to JDM</td>
<td>Being different</td>
<td></td>
<td></td>
</tr>
<tr>
<td>The fact that you have to take medication and constantly exercise.</td>
<td>Treatment</td>
<td>Physiotherapy</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
## Appendix 22 - Comments in response to Q8

<table>
<thead>
<tr>
<th>Q8. What is the worst thing about having JDM?</th>
<th>Primary</th>
<th>Secondary</th>
<th>Tertiary</th>
<th>Quarternary</th>
</tr>
</thead>
<tbody>
<tr>
<td>taking medicines</td>
<td>Treatment</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Nothing really</td>
<td>Nothing</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>The aches in my legs and aches I get really bad and end up in hospital</td>
<td>Pain</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Worrying about the future.</td>
<td>Uncertainty</td>
<td>Mental health effects</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Not being able to do what my peers are doing. Changing my life plans. Not being able to do simple things.</td>
<td>Limitations due to JDM</td>
<td>Mental health effects</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Not being physically fit my movement is restricted</td>
<td>Limitations due to JDM</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>My rash - its itchy and hurts and looks weird</td>
<td>Rashes</td>
<td>Body image</td>
<td></td>
<td></td>
</tr>
<tr>
<td>The pain</td>
<td>Pain</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>It stops me from doing things I missed out on a lot of stuff</td>
<td>Limitations due to JDM</td>
<td>Being different</td>
<td></td>
<td></td>
</tr>
<tr>
<td>feeling tired sometimes, sometime aching muscles and joints</td>
<td>Tired</td>
<td>Pain</td>
<td></td>
<td></td>
</tr>
<tr>
<td>JDM will never go away. It always sitting at the background.</td>
<td>Uncertainty</td>
<td>Mental health effects</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
### Appendix 23 - Comments in response to Q9

**Question 9 – Anything else you want to tell us?**

<table>
<thead>
<tr>
<th>Comment</th>
<th>Coding</th>
</tr>
</thead>
<tbody>
<tr>
<td>The nurse that would visit every Wednesday to administer said methotrexate injections and I never learned her surname. If she still works at the X and remembers my name X tell her thank you again for me, cause I know she was only ever helping me even if at the time I hated knowing that she was coming.</td>
<td>Thank you</td>
</tr>
<tr>
<td>I have become stronger as a person and thanks to my doctors they make me feel confident.</td>
<td>Thank you</td>
</tr>
<tr>
<td>I really hope that someone works out what causes JDM and can stop it.</td>
<td>Research</td>
</tr>
<tr>
<td>I had JDM when I was 4, back in 2005, so I feel that these questions are largely not applicable to me because I do not currently have the symptoms and I cannot remember anything from when I was diagnosed with the symptoms.</td>
<td>Re survey</td>
</tr>
<tr>
<td>I was wondering whether setting up a support group for younger people with Dermatomyositis would be possible? And if so, would I be able to help?</td>
<td>Alturistic</td>
</tr>
<tr>
<td>I wish more energy, effort and emphasis was put on my physio and mental health so I could continue to take care of myself in adult life</td>
<td>Mental Health comment</td>
</tr>
<tr>
<td>I was diagnosed with JDM when I was 5 yrs old and my treatment was so successful that it has not returned. I am now 15 and have been discharged from all health checks for at least 7 years so many of the questions were not applicable.</td>
<td>Re survey</td>
</tr>
<tr>
<td>I really hope this survey helps with your research</td>
<td>Re survey</td>
</tr>
<tr>
<td>I am very thankful for my hospital to help me get better, and to allow my mum to stay with me whilst I was having treatment so I wasn’t scared. I am also extremely thankful for the doctors and nurses on penguin ward for making me laugh and making me less worried about my illness and for helping me feel unique about it instead of seeing it as a negative thing :)</td>
<td>Thank you</td>
</tr>
<tr>
<td>I think there should be more support for people with mental health issues due to suffering with JDM because it definitely effects mental health.</td>
<td>Mental Health comment</td>
</tr>
<tr>
<td>JDM has made me a lot stronger and I have learnt to deal with problems much more logically. JDM has made me grow so much more mature because I needed to learn how to make sure I was taking care of myself properly.</td>
<td>+ve effects of JDM</td>
</tr>
<tr>
<td>Comment</td>
<td>Coding</td>
</tr>
<tr>
<td>---------</td>
<td>--------</td>
</tr>
<tr>
<td>I also sometimes get annoyed at how naive all the other teens I know are as they haven't seen the other side of life before. However, this makes me feel grateful for JDM as I would be such a different person without it.</td>
<td>+ve effects of JDM</td>
</tr>
<tr>
<td>I think counselling and support groups should be offered to those who have been cured and those still going through treatment as it may improve peoples security within themselves, their understanding and all round mental health.</td>
<td>Mental Health comment</td>
</tr>
<tr>
<td>The struggle is constant, being someone with an invisible disability makes coping so much harder as no one can see it as well as trying to cope everyday.</td>
<td>Mental Health comment</td>
</tr>
<tr>
<td>If it ever comes back then I would fill it out differently because I would understand it more. But I don't want it again, thanks.</td>
<td>Re survey</td>
</tr>
<tr>
<td>Can someone confirm if JDM is a disability? I'm worried about discrimination in the future and feel it we had clarity it would help.</td>
<td>Question</td>
</tr>
<tr>
<td>thank you for everything getting me through it and keeping me up to date, I feel better than ever. I'm expecting my first baby boy.</td>
<td>Thank you</td>
</tr>
<tr>
<td>When will they stop my injections. Will I get better?</td>
<td>Question</td>
</tr>
<tr>
<td>Not helped with my mental health and the change over from Child/Adolescent/adult has been appalling. Feel completely let down and life is a constant struggle. You have helped me lots ideally everyone needs someone like her/you in their life.</td>
<td>Mental Health comment</td>
</tr>
<tr>
<td>that I'm very grateful for the doctors and Rheumatology nurses on pelican and the physios at gosh and that i feel alot better and feel my strength is back to how it was before having JDM</td>
<td>Thank you</td>
</tr>
<tr>
<td>I hate jdm</td>
<td>Mental Health comment</td>
</tr>
<tr>
<td>I want my sickness to go away? I don't like taking the treatment, it make me feel sick</td>
<td>Personal comment</td>
</tr>
<tr>
<td>All the nurses and doctors that are helping or have helped anyone should be proud for being so amazing</td>
<td>Thank you</td>
</tr>
<tr>
<td>Sensitive to sunlight The school got funding to change the lighting in my classrooms year 3 and 4. When I'm in sun or bad lighting I get tired and rash comes up.</td>
<td>Personal comment</td>
</tr>
<tr>
<td>I don't think so except my complicated science and math questions!</td>
<td>Personal comment</td>
</tr>
<tr>
<td>I would like to have another JDM family day sooner. My parents always made me stay positive and it has kept me going.</td>
<td>Personal comment</td>
</tr>
<tr>
<td>Comment</td>
<td>Coding</td>
</tr>
<tr>
<td>--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------</td>
<td>---------------------</td>
</tr>
<tr>
<td>I don't like people treating me different just because I have JDM. I can be and do the same as anybody else. JDM will not stop being who I am.</td>
<td>Personal comment</td>
</tr>
<tr>
<td>Help us get our benefits back. Please x</td>
<td>Personal comment</td>
</tr>
<tr>
<td>JDM makes me feel unique and I am happy. JDM makes me feel special.</td>
<td>Personal comment</td>
</tr>
<tr>
<td>Mum: she is 8 and had to help her answer the questions, most of them she couldn't understand the meaning. Q17. We answer most of them somewhat but her real answer was 'I don't know'. She has a very mild JDM and only muscles most visible</td>
<td>Personal comment</td>
</tr>
<tr>
<td>I have been dancing for 4 yrs now and I am in a competition team as well as performing solos. I am enclosing photos with a hope that other children at the start of their illness could see to give them hope and inspire them. Thank you.</td>
<td>Personal comment</td>
</tr>
<tr>
<td>Would like to know if there is any support groups.</td>
<td>Question</td>
</tr>
<tr>
<td>I have met people who have autoimmune conditions and have spoken to them about how their condition has effected them. It was nice to know that I wasn't the only one who felt the way I did. :-)</td>
<td>Personal comment</td>
</tr>
</tbody>
</table>
Question 8
What psychosocial measures of health/wellbeing do you collect?

<table>
<thead>
<tr>
<th>Text input</th>
</tr>
</thead>
<tbody>
<tr>
<td>None routinely as far as I am aware. If a patient is seen in the psychology service then measures such as the Becks and PedsQL may be used.</td>
</tr>
<tr>
<td>PedsQL (child, parent and Family Impact modules), SDQ</td>
</tr>
<tr>
<td>Depending on patient - PI-ED, GBOM, RCADS, CSES, Peds-QoL</td>
</tr>
<tr>
<td>None routinely</td>
</tr>
<tr>
<td>PEDS QL, RCADS, Peds-QL Family impact and Goal based outcomes. Sometimes disorder specific measures. Sometimes parenting stress index.</td>
</tr>
<tr>
<td>RCADS, PedsQL</td>
</tr>
<tr>
<td>Beck youth inventory (self concept, anx, dep, anger). Likert scales re pain, goal attainment. Any others as indicated.</td>
</tr>
<tr>
<td>We do a Goal Based thermometer to identify progress. At times we will do a RCADS depending on the presenting difficulties.</td>
</tr>
<tr>
<td>SRS and ORS SLDOM</td>
</tr>
<tr>
<td>None routinely. But individual clinicians use a variety of measures as appropriate.</td>
</tr>
</tbody>
</table>
# Appendix 25 - Health care professionals challenges

<table>
<thead>
<tr>
<th>Biggest challenge – medical professionals</th>
<th>Biggest challenge – Nurse Specialists</th>
<th>Biggest challenge – Clinical Psychologists</th>
</tr>
</thead>
<tbody>
<tr>
<td>Time, local input</td>
<td>Undisturbed time slots with the family and the interaction to build that trust in the relationship. Interactions with the families become treatment related or problem solving interactions Clinics do not allow enough time</td>
<td>'Buy in' to biospsychosocial model. Being fully up to date on medical treatment/treatment plan. If children are inpatient being away from normal life and the additional stressors this places on mood/social functioning</td>
</tr>
<tr>
<td>Lack of psychology, locally and within Trust. Dedicated psychology for rheumatology patients is vital but often unavailable/ unacceptable waits</td>
<td>Limited resources available however our patients have access to specialist physiotherapist who seems/is very knowledgeable and experienced at delivering a range of support and signposting patients</td>
<td>NO ANSWER – LEFT BLANK</td>
</tr>
<tr>
<td>I think most services directed towards crisis management and chronic pain. I think more can be done with early intervention if more resource, but overall I think our JDM patients tend not to struggle as much as some other disease groups and benefit from advice on pacing/management of fatigue/fitness/acceptance of disease from other team members as well as psychology</td>
<td>Our psychology service is very limited at the moment. We really need more psychology support. The waiting list is extremely long and many of our patients cannot wait this length of time before being seen</td>
<td>NO ANSWER – LEFT BLANK</td>
</tr>
<tr>
<td>Difficult question, depends on the individual patients and their requirements. We have good access to psychology, art therapy via Teapot trust and Liaison psychiatry when required so access is not the biggest challenge</td>
<td>Admission by family or young person that mental health is an issue that needs addressing</td>
<td>DID NOT REPLY TO SENT SURVEY</td>
</tr>
<tr>
<td>Lack of psychologist on team</td>
<td>Not enough time nor experience, nor access to resources</td>
<td>Not enough psychosocial resource in this area to meet need</td>
</tr>
</tbody>
</table>
### Appendix 25 - Health care professionals challenges

<table>
<thead>
<tr>
<th>Biggest challenge – medical professionals</th>
<th>Biggest challenge – Nurse Specialists</th>
<th>Biggest challenge – Clinical Psychologists</th>
</tr>
</thead>
<tbody>
<tr>
<td>Access to psychology in network clinics, patients have to come to central hospital for input due to pressure on other psychology services</td>
<td>Wide age range for children, chronic nature of condition, not enough psych time and long distances travelled by families</td>
<td>NO ANSWER – LEFT BLANK</td>
</tr>
<tr>
<td>Under-resourced clinical psychologist service which cannot meet demands and are stretched across several different Paediatric subspecialties</td>
<td>Time, resource. Once they see psychology they get a good service the delay is the problem</td>
<td>Not being integrated into the medical team. The medical team adopt a very psychosocial approach to care, but their resources are limited and there is no dedicated psychological provision to the team</td>
</tr>
<tr>
<td>Lack of psychology in the team or named within the psychology department</td>
<td>Not having a psychologist in JDM clinics</td>
<td>NO PSYCHOLOGIST TO ANSWER</td>
</tr>
<tr>
<td>Extremely limited access to “in-house” children’s psychology services</td>
<td>Knowing where we access the support particularly as we are a regional service and patients want this care to be closer to home. It needs to be in the context of the whole family and this can be difficult where families are fragmented and living in differing locations</td>
<td>NO PSYCHOLOGIST TO ANSWER</td>
</tr>
<tr>
<td>Time. However the team are excellent at stepping up. There is a big challenge sometimes re lack of parental engagement with this side of the care we give</td>
<td>Time and space to see the young person</td>
<td>Time – clinical psychologist is very part-time and cannot easily be present during team clinics</td>
</tr>
<tr>
<td>Patients who live a long distance with poor local care or those that don’t engage with psychology input</td>
<td>We offer psychology to all pts who need it as we have a dedicated clinical psychologist on our team</td>
<td>Time! And we have a very large catchment area since we are a Specialist Centre</td>
</tr>
<tr>
<td>Clinic space for psychologist</td>
<td>Time – consistent resources that can cover the whole of the South-west</td>
<td>Logistics, I am part of a regional service, therefore providing equity of psychology provision across the service</td>
</tr>
</tbody>
</table>
### Appendix 25 - Health care professionals challenges

<table>
<thead>
<tr>
<th>Biggest challenge – medical professionals</th>
<th>Biggest challenge – Nurse Specialists</th>
<th>Biggest challenge – Clinical Psychologists</th>
</tr>
</thead>
<tbody>
<tr>
<td>Timely access</td>
<td>Engagement with services or consent to referrals</td>
<td>Resourcing it</td>
</tr>
<tr>
<td>Until recently, lack of psychology, now better</td>
<td>No dedicated psychology time or specific psychologist identified</td>
<td>Not having dedicated time for rheumatology limits the screening / pre-emptive / early intervention work that can be done</td>
</tr>
<tr>
<td>Not enough psychology time, waits to see our psych are getting longer</td>
<td>No answer</td>
<td>Current wait for assessment, and limited psychology time - would be ideal to have the time to be based in clinic more to routinely meet newly diagnosed families</td>
</tr>
</tbody>
</table>
Appendix 26 - Prompt questions - school focus group

- Do you think the booklet should cover nursery/school/Uni. Or just target one age group?
- What are the advantages and disadvantages of including nursery/school/uni?
- Should the booklet include more info or less information?
- Should medications be included?
- Should it include the tick page for symptoms that the child or young person has at that time? This would make it an interactive book, which the parents/child could give to school at the start of each new term/school year?
- Look at the contents page, anything obviously missing/not needed/too much?

Then go through page by page, discussing the text, feel free to scribble on the book

Any experiences that anyone would like to write down for each section – good/bad/informative
Appendix 27 - Prompt questions - mentoring focus group

- Would having a peer mentor helped you/your child/(or if a sibling, would it help you now if you are having a bad day at school?)
- What age would be appropriate for the mentors?
- What age would be appropriate for the mentees?
- How often would you expect the two individuals to talk?
- How long would you expect each meet to last?
- Do you think skype is appropriate? What others way could there be of mentoring?
- What do you think the training of the mentors would need to include?
- Should the mentors be paid?
- Do you think there should be a set list of things that are talked about on each call or should the conversation just flow?
- Any ideas on how do you think this project could continue with training mentors? Eg who could train them?
- What benefits would there be to be a mentor? Do you think it would be good to have a 'mentor identity'?
- Do you think the mentors should have the responsibility to flag concerns?
- Do you think there would be any differences in gender? Would males and females equally like the mentoring idea/talking to someone/need the same training
- Do you know of any other mentoring schemes?
## Appendix 28 - Presentations given

<table>
<thead>
<tr>
<th>Year</th>
<th>Date</th>
<th>Name of Meeting</th>
<th>Lecture / Poster presentation</th>
<th>Target Audience</th>
<th>Location</th>
</tr>
</thead>
<tbody>
<tr>
<td>2017</td>
<td>18th May</td>
<td>Paediatric Rheumatology Nurses Annual Meeting</td>
<td>Lecture</td>
<td>Nurses</td>
<td>Peterborough</td>
</tr>
<tr>
<td></td>
<td>15th June</td>
<td>ORCHID National Study Meeting <em>Putting patients and parents at the centre of research</em></td>
<td>Lecture</td>
<td>Health Professionals</td>
<td>London</td>
</tr>
<tr>
<td></td>
<td>2nd July</td>
<td>Myositis Family Conference</td>
<td>Lecture</td>
<td>Patients and families</td>
<td>Oxford</td>
</tr>
<tr>
<td></td>
<td>9th Sept</td>
<td>JDM Family Day</td>
<td>Lecture</td>
<td>Patients and families</td>
<td>London</td>
</tr>
<tr>
<td></td>
<td>10th Oct</td>
<td>Myonet Meeting (adults and paediatrics)</td>
<td>Lecture</td>
<td>Rheumatology Health Professionals</td>
<td>Guys Hospital</td>
</tr>
<tr>
<td></td>
<td>24th Oct</td>
<td>Adolescent Research Science Meeting</td>
<td>Lecture</td>
<td>Mixed Health Professionals</td>
<td>London</td>
</tr>
<tr>
<td></td>
<td>14th Nov</td>
<td>Health Education England Clinical Academic Careers Meeting</td>
<td>Lecture</td>
<td>Those interested in Clinical Academic Careers</td>
<td>London</td>
</tr>
<tr>
<td></td>
<td>28th Nov</td>
<td>Rheumatology Research Meeting</td>
<td>Lecture</td>
<td>Those interested in Clinical Academic Careers</td>
<td>London</td>
</tr>
<tr>
<td>2018</td>
<td>3rd May</td>
<td>Upgrade presentation</td>
<td>Lecture</td>
<td>Anyone interested in the University</td>
<td>London</td>
</tr>
<tr>
<td></td>
<td>13th July</td>
<td>Advanced Rheumatology Study Day</td>
<td>Lecture</td>
<td>Rheumatology Health Professionals</td>
<td>London</td>
</tr>
<tr>
<td></td>
<td>31st July</td>
<td>Paediatric Academic Meeting</td>
<td>Lecture</td>
<td>Canadian Health professionals</td>
<td>Toronto, Canada</td>
</tr>
</tbody>
</table>
## Appendix 28 - Presentations given

<table>
<thead>
<tr>
<th>Year</th>
<th>Date</th>
<th>Name of Meeting</th>
<th>Lecture / Poster presentation</th>
<th>Target Audience</th>
<th>Location</th>
</tr>
</thead>
<tbody>
<tr>
<td>5th Sept</td>
<td>5th Sept</td>
<td>Paediatric Rheum European Society Meeting (PReS)</td>
<td>Poster presentation</td>
<td>Rheumatology Health Professionals</td>
<td>Lisbon</td>
</tr>
<tr>
<td>17th Sept</td>
<td>17th Sept</td>
<td>ORCHID PhD Student Day</td>
<td>Lecture</td>
<td>ORCHID students and staff</td>
<td>London</td>
</tr>
<tr>
<td>25th Sept</td>
<td>25th Sept</td>
<td>Rheumatology Research Meeting</td>
<td>Lecture</td>
<td>Rheumatology Health Professionals</td>
<td>Alderhey</td>
</tr>
<tr>
<td>10th Oct</td>
<td>10th Oct</td>
<td>Otto Wolf Mental Health Day</td>
<td>Poster presentation</td>
<td>Mental Health Professionals</td>
<td>London</td>
</tr>
<tr>
<td>19th Oct</td>
<td>19th Oct</td>
<td>Presentation at British Society of Paediatric and Adolescent Rheumatology (BSPAR) Research Conference</td>
<td>Lecture</td>
<td>Rheumatology Health Professionals</td>
<td>Southamp ton</td>
</tr>
<tr>
<td>17th Nov</td>
<td>17th Nov</td>
<td>JDM Family Day</td>
<td>Lecture</td>
<td>Patients and families</td>
<td>London</td>
</tr>
<tr>
<td>27th Nov</td>
<td>27th Nov</td>
<td>ORCHID Clinical Academic Careers Evening</td>
<td>Lecture</td>
<td>Those interested in Clinical Academic Careers</td>
<td>London</td>
</tr>
<tr>
<td>2019</td>
<td>31st Jan</td>
<td>UCLH Adolescent Open Day</td>
<td>Poster presentation</td>
<td>Prospective university students</td>
<td>London</td>
</tr>
<tr>
<td>26th Feb</td>
<td>26th Feb</td>
<td>3 Minute Thesis Competition</td>
<td>Lecture</td>
<td>University students and staff</td>
<td>London</td>
</tr>
<tr>
<td>18th Mar</td>
<td>18th Mar</td>
<td>Juvenile Dermatomyositis Research Group Principal Investigator day</td>
<td>Lecture</td>
<td>Rheumatology Health Professionals from around UK</td>
<td>London</td>
</tr>
<tr>
<td>27th Mar</td>
<td>27th Mar</td>
<td>Global Conference on Myositis (adults and paediatrics)</td>
<td>Poster presentation</td>
<td>Rheumatology Health Professionals</td>
<td>Berlin</td>
</tr>
<tr>
<td>9th Apr</td>
<td>9th Apr</td>
<td>3 Minute Thesis Competition 2nd round</td>
<td>Lecture</td>
<td>University students / staff</td>
<td>London</td>
</tr>
</tbody>
</table>
### Appendix 28 - Presentations given

<table>
<thead>
<tr>
<th>Year</th>
<th>Date</th>
<th>Name of Meeting</th>
<th>Lecture / Poster presentation</th>
<th>Target Audience</th>
<th>Location</th>
</tr>
</thead>
<tbody>
<tr>
<td>16\textsuperscript{th} May</td>
<td>16\textsuperscript{th} May</td>
<td>Paediatric Rheumatology Nurses Annual Meeting</td>
<td>Lecture</td>
<td>Nurses</td>
<td>Edinburgh</td>
</tr>
<tr>
<td>6\textsuperscript{th} June</td>
<td>6\textsuperscript{th} June</td>
<td>Clinical Academic Careers Roundtable Event</td>
<td>Lecture</td>
<td>Clinical Academics and students</td>
<td>Surrey University</td>
</tr>
<tr>
<td>23\textsuperscript{rd} June</td>
<td>23\textsuperscript{rd} June</td>
<td>Myositis Family Conference</td>
<td>Lecture</td>
<td>Patients and families</td>
<td>Oxford</td>
</tr>
<tr>
<td>25\textsuperscript{th} June</td>
<td>25\textsuperscript{th} June</td>
<td>Rheumatology research meeting</td>
<td>Lecture</td>
<td>Rheumatology Health Professionals</td>
<td>GOSH – London</td>
</tr>
<tr>
<td>28\textsuperscript{th} June</td>
<td>28\textsuperscript{th} June</td>
<td>Royal College of Nursing Rheumatology Workshop</td>
<td>Lecture</td>
<td>Nurses</td>
<td>London</td>
</tr>
<tr>
<td>12\textsuperscript{th} July</td>
<td>12\textsuperscript{th} July</td>
<td>Keynote Speaker at Hermeneutic Phenomenology Symposium</td>
<td>Lecture</td>
<td>Phenomenologists</td>
<td>Lancashire University</td>
</tr>
<tr>
<td>19\textsuperscript{th} Aug</td>
<td>19\textsuperscript{th} Aug</td>
<td>Invited Speaker to first Northern JDM Family Day</td>
<td>Lecture</td>
<td>Patients and families and Northern Health Professionals</td>
<td>Manchester</td>
</tr>
<tr>
<td>2\textsuperscript{nd} Sept</td>
<td>2\textsuperscript{nd} Sept</td>
<td>BRC PPIE Meeting</td>
<td>Lecture</td>
<td>BRC staff</td>
<td>London</td>
</tr>
<tr>
<td>5\textsuperscript{th} Sept</td>
<td>5\textsuperscript{th} Sept</td>
<td>Royal College of Nurses International Research Conference</td>
<td>Lecture</td>
<td>Nurses</td>
<td>Sheffield</td>
</tr>
<tr>
<td>14\textsuperscript{th} Sept</td>
<td>14\textsuperscript{th} Sept</td>
<td>JDM Family Day</td>
<td>Lecture</td>
<td>Patients and families</td>
<td>London</td>
</tr>
<tr>
<td>30\textsuperscript{th} Sept</td>
<td>30\textsuperscript{th} Sept</td>
<td>ORCHID PhD Student day</td>
<td>Lecture</td>
<td>ORCHID students and staff</td>
<td>London</td>
</tr>
<tr>
<td>11\textsuperscript{th} Nov</td>
<td>11\textsuperscript{th} Nov</td>
<td>American College of Rheumatology Meeting</td>
<td>Lecture</td>
<td>Rheumatology Health Professionals</td>
<td>Atlanta - US</td>
</tr>
<tr>
<td>22\textsuperscript{nd} Nov</td>
<td>22\textsuperscript{nd} Nov</td>
<td>GOSH Nursing Conference</td>
<td>Poster presentation</td>
<td>Nurses</td>
<td>London</td>
</tr>
<tr>
<td>4\textsuperscript{th} Dec</td>
<td>4\textsuperscript{th} Dec</td>
<td>Oral presentation to BSR Staff Meeting</td>
<td>Lecture</td>
<td>Medical society staff</td>
<td>London</td>
</tr>
<tr>
<td>Year</td>
<td>Date</td>
<td>Name of Meeting</td>
<td>Lecture / Poster presentation</td>
<td>Target Audience</td>
<td>Location</td>
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<tr>
<td>9th Dec</td>
<td>Oral presentation re CAC at Clinical Academic Roles and Career Pathways Implementation Network Meeting (CARIN)</td>
<td>Lecture</td>
<td>Clinical Academics</td>
<td>Coventry Hospital Trust</td>
<td></td>
</tr>
<tr>
<td>2020</td>
<td>27th March</td>
<td>Invited to present an hour long Arthritis and Musculoskeletal Alliance (ARMA) Webinar on this study findings</td>
<td>Webinar</td>
<td>Everyone</td>
<td>On line and now freely available on the internet</td>
</tr>
</tbody>
</table>
Appendix 29 - Pediatric Rheumatology publication

Being on the juvenile dermatomyositis rollercoaster: a qualitative study

Polly Livermore12,9,11,12, Suzanne Gray2, Kathleen Mulligan4,5, Jennifer N. Stimson6,7, Lucy R. Wedderburn16,8,9 and Faith Gibson2,10,11

Abstract

Background: Juvenile Dermatomyositis is a rare, potentially life-threatening condition with no known cure. There is no published literature capturing how children and young people feel about their condition, from their perspective. This study was therefore unique in that it asked children and young people what it is like to live with Juvenile Dermatomyositis.

Methods: Data were obtained from fifteen young people with Juvenile Dermatomyositis, between eight and nineteen years of age from one Paediatric Rheumatology department using audio-recorded interpretive phenomenology interviews. Data were analyzed phenomenologically, using a process that derives narratives from transcripts resulting in a collective composite of participants shared experiences, called a ‘phenomenon’.

Results: The overarching metaphor of a rollercoaster captures the phenomenon of living with Juvenile Dermatomyositis as a young person, with the ups and downs at different time points clearly described by those interviewed. The five themes plotted on the rollercoaster began with confusion; followed by feeling different, being sick, steroidal and scared from the medications; uncertainty; and then ended with acceptance of the disease over time.

Conclusion: Young people were able to talk about their experiences about having Juvenile Dermatomyositis. Our findings will aid clinicians in their practice by gaining a deeper understanding of what daily life is like and highlighting ways to enhance psychosocial functioning. Hopefully, this study and any further resulting studies, will raise understanding of Juvenile Dermatomyositis worldwide and will encourage health care professionals to better assess psychosocial needs in the future.

Keywords: juvenile dermatomyositis, Qualitative research, Phenomenology, Uncertainty, Psychosocial needs

Background

Juvenile Dermatomyositis (JDM) is a rare, potentially life-threatening, systemic condition of unknown origin, characterized by weakness in proximal muscles and skin rashes, often involving other systems [1–4]. Weakness is progressive, which can first become evident with having difficulty climbing stairs and can become profound, with children progressing to becoming bed bound, unable to sit up or roll over and for some, they may even require nutritional support through feeding tubes [5–7]. In severe cases, this debilitating condition involves children and young people (CYP) requiring multiple hospitalizations, experiencing extreme muscle pain, requiring daily physiotherapy and cytotoxic medications to gain symptom control, and as a result they miss months of schooling [8–10].

The term ‘psychosocial’, pertaining to both psychological and social, encompasses a multitude of variables, such as social and emotional support, mood and anxiety. Research into psychosocial needs in CYP with JDM is sparse. One of only two published studies examining Quality of Life (QoL) in JDM found significantly lower health related QoL in this group compared to controls.
You give me a name that I can’t say, but I have to explain what it is every day: the power of poetry to share stories from young people with a rare disease

P. Livermore, L. R. Wedderburn and F. Gibson

*Infection, Immunity and Inflammation Research and Teaching Department, University College London Great Ormond Street Institute of Child Health, London, UK; Centre for Outcomes and Experience Research in Children’s Health, Illness and Disability (ORCHE), Great Ormond Street Hospital for Children NHS Foundation Trust, London, UK; NIHR Biomedical Research Centre at Great Ormond Street Hospital, London, UK; Centre for Adolescent Rheumatology Venous Arthritis at UCL UCLH and GOSH, London, UK; School of Health Sciences, University of Surrey, Guildford, UK

ABSTRACT
Qualitative research is about people’s lives; their stories, their thoughts, their feelings, and their experiences. Researchers continue to reflect on the best way to present other people’s stories in a way that stays true to their accounts and delivers the message in a manner which resonates. This article presents one such way, discovered through serendipity, which allowed the researcher to share findings from young people’s stories eloquently and passionately to audiences of healthcare professionals from different disciplines. The silence and raw emotion witnessed after the poetry delivery was akin to that often seen during childbirth when those around are stunned into thoughtful awe. The poetry, which was crafted entirely from young participants’ interview transcripts had two benefits: (1) sharing young people’s stories using their own words and (2) using a presentation format that demanded attention; so that people took notice, listened and reflected on the words and experiences being expressed.

Introduction
Qualitative research aims to understand people’s thoughts, experiences and emotions (McCulliss, 2013). Whilst there are various methodologies and methods within the overarching umbrella term of “qualitative”, the researcher always undertakes to sift through, organise and present the data in a meaningful way to arouse an empathetic reaction in the consumer of research; hopefully leading to a deep, personal understanding of the individual or topic under study (Lietz, Furman, & Langer, 2006; McCulliss, 2013). Phenomenology is one such methodology which aims to slow the researcher down and hold his or her gaze as they bring the phenomena or lived experience of some focus under study to the front (Willis, 2002), but by doing so, reams of “data” are gathered along the journey. In this paper, I discuss a research project, where I discovered the potential power of poetry, an

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