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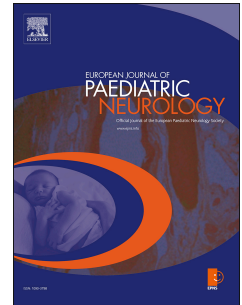
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# Accepted Manuscript

Pain experience, expression and coping in boys and young men with Duchenne muscular dystrophy – a pilot study using mixed methods

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# Pain experience, expression and coping in boys and young men with Duchenne muscular dystrophy

## Mixed methods study

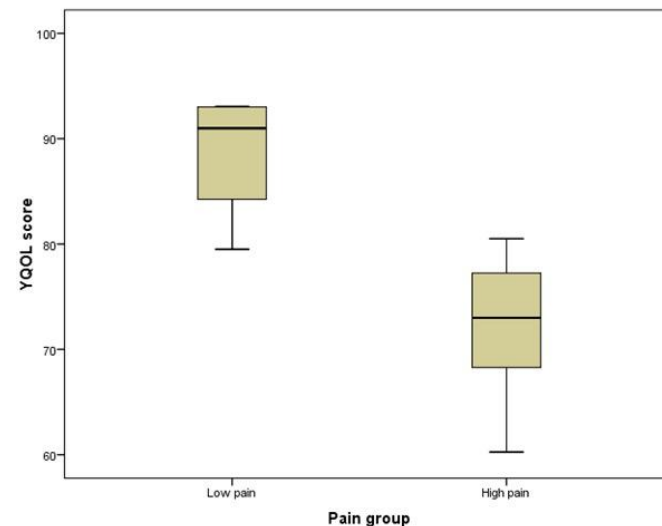
Parents' survey & instruments

Young men's interview & instruments

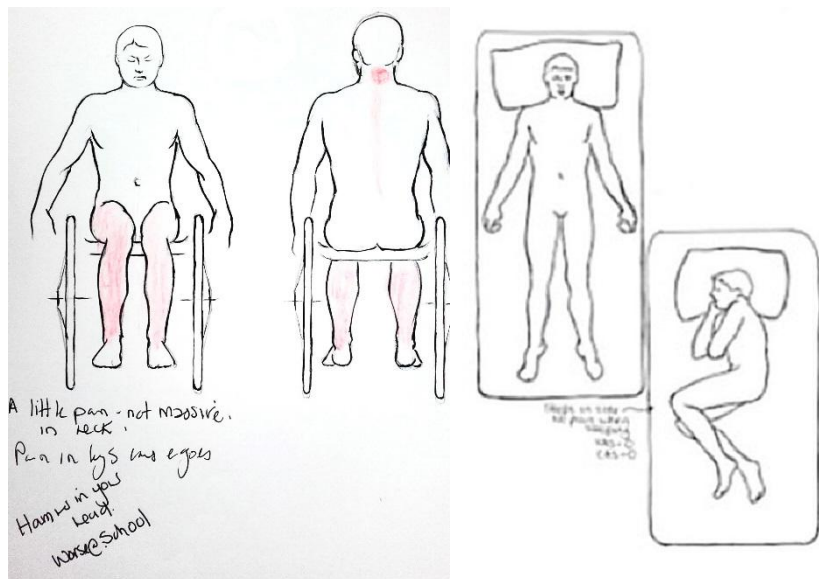
## Qualitative findings

- Hurting in different places
- Telling : holding it in & letting it out
- Acting to relieve pain

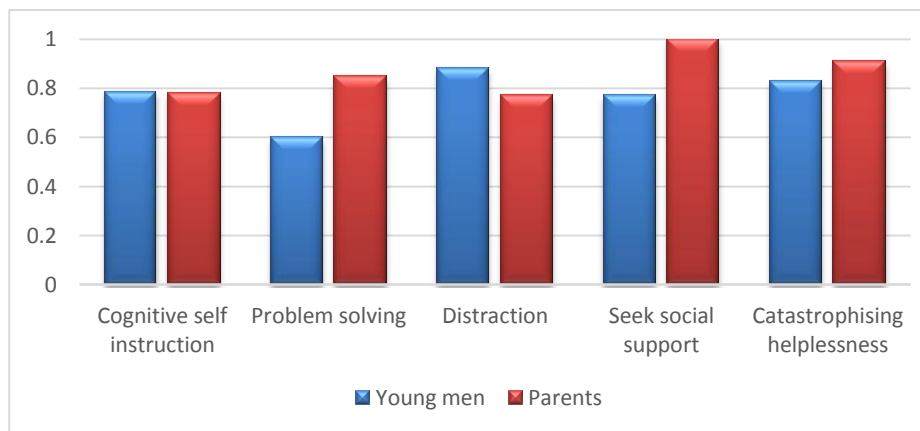
## Impact of pain on quality of life



## Pain body map, young man (aged 11 years)



## Ways of coping with pain (from PPCI)



Pain experience, expression and coping in boys and young men with Duchenne muscular dystrophy – a pilot study using mixed methods

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**ABSTRACT**

There is limited research exploring the pain experience of boys and young men with Duchenne Muscular Dystrophy. Methods: We conducted a mixed-methods pilot study to assess the feasibility of using particular measures of pain, pain coping and quality of life within semi-structured interviews with boys and young men with Duchenne Muscular Dystrophy and a postal survey of their parents. Non-probability, convenience sampling was used. Results: Twelve young men aged 11 to 21 years (median 15 years), three of whom were still ambulant, and their parents / guardians were recruited. The measures used were acceptable to the young men and demonstrated potential to provide useful data. Two-thirds of young men suffered from significant daily pain which was associated with reduced quality of life. Pain complaints were largely kept within the family. Young men's pain-coping strategies were limited by their restricted physical abilities. Statistical power based on these preliminary results suggests a study of approximately 50 boys/young men which appears feasible. Conclusions: Further study is needed to explore acceptable and effective methods of pain management in this population and ways of enhancing pain-coping strategies. In clinical practice, assessment of pains and discomfort should form part of all routine consultations.

Keywords: child, neuromuscular disease, Duchenne Muscular Dystrophy, pain measurement, pain assessment, adaptive behaviour

## 1.0 Background

Duchenne Muscular Dystrophy (DMD) affects approximately 1 in every 3,500 live males births<sup>1</sup>, and, at any one time, approximately 2,500 boys and young men in the UK. Whilst management of the condition has improved dramatically<sup>2-6</sup>, management of pain has not always appeared to be a clinical or research priority<sup>7,8</sup>. The many and varied complications of DMD, including muscle weakness and lack of mobility<sup>9-11</sup>, spinal deformity<sup>12</sup>, constipation<sup>13</sup>, delay in gastric emptying<sup>14</sup>, cardiomyopathy<sup>15</sup> and osteopenia / osteoporosis<sup>16</sup>, increase the risk to experience many types of pain throughout the progression of the disorder. There is increasing evidence that pain is a significant issue for boys and young men with Duchenne Muscular Dystrophy and should not be ignored<sup>17</sup>.

A bio-behavioural model of children's pain has been proposed whereby coping strategies are vital intervening factors in predicting the variability in paediatric pain perception, pain behaviour, and functional status<sup>18-20</sup>. In turn, this model is enlarged to incorporate a relationship between pain, pain coping and quality of life<sup>21</sup>. Turk and Okifuji (1999) stress the importance of assessing patients' individual coping strategies, attitudes, psychological distress, and the effect of pain on their lives<sup>22</sup>. Engel et al., (2005) suggest that boys and young men with DMD might be at high risk for developing maladaptive pain coping strategies placing additional stress on already stressed families<sup>7</sup>.

Clinical anecdotes and communications with involved clinicians suggest that young men with DMD seldom volunteer that they have pain (Personal communication). Instead, they tend to have a relatively constrained repertoire of verbal pain communication (e.g. 'ouch'ing during movements), non-verbal expression (e.g., grimacing), and seem reticent about taking pain medication. It is unclear whether this implies that pain is not a major issue for them, or whether aspects of gender or social norms and / or other constraints tend to limit the boys' expression of pain. Without a clearer understanding of these issues optimal pain management for boys and young men with DMD may not be achieved.

## 2.0 AIMS

The aims of this study were to 1) assess the feasibility of using validated instruments measuring pain, pain coping and quality of life within semi-structured interviews, 2) provide preliminary quantitative data that would inform the design of a larger study and 3) provide qualitative findings to support understanding of pain in this group of young people.

## 3.0 METHODS

### 3.1 Ethical issues

Conducting this study with young men and their parents required care and forethought. Pain, in particular, can be a sensitive area, especially for children and young people with life-limiting conditions and their families and our aim was to approach the study sensitively, ensuring we took care not to create anxieties about pain and to manage any situations that arose conscientiously. The study

design gained ethical approval from Leeds (East) NHS Research Ethics Committee (08/H1306/165).

### **3.2 Participants**

A convenience sample of young men was recruited through two NHS neuromuscular clinics and two children's hospices in the UK. Prospective participants were aged 11 to 21 years. They did not need to have pain to participate. In describing the study we use the term 'young men' to include all boys and young men participating in the study. The term 'parents' is used to include young men's parents and guardians.

### **3.3 Procedures**

Using a mixed methods approach<sup>23</sup> we conducted interviews with young men alongside administration of a number of validated assessment instruments which provided the structure for the interviews. We gave particular attention to choosing instruments that would be acceptable to the young men and their parents, for instance, the Quality of Life measure we used does not focus on physical function. Parents' questionnaires were administered as postal questionnaires or face to face.

#### Instruments:

Data were collected on the following

3.3.1 Pain. Body maps developed specifically for this study (Figure 1), a Colour Analogue Scale backed by a numerical rating scale (CAS)<sup>24</sup> and the Faces Pain Scale\_Revised (FPS\_R)<sup>25</sup>.

3.3.2. Pain coping. The Pediatric Pain Coping Inventory (PPCI)<sup>26</sup> assesses children's pain coping strategies. It comprises four open questions followed by a list of 41 coping strategies each scored on a three point 0 – 2 scale as used 'never', 'sometimes' or 'often'. In addition to a children's version, adolescent (teen) and parent versions are available. We used the original domains 'Cognitive self-instruction', 'Problem solving', 'Distraction', 'Seeks social support' and 'Catastrophising \ helplessness'. Higher scores on each domain indicate more use of the strategy. The PPCI has been used to assess pain coping in children with rheumatologic disease<sup>26 21</sup>, adults with cerebral palsy<sup>27</sup> and for parents' report of pain coping in children with developmental disability<sup>28</sup>. Here we used the teen / adolescent version for the young men and parent (proxy) version for their parents.

3.3.3 Quality of life. The Youth Quality of Life Scale (YQOL)<sup>29,30</sup> was developed by Edwards et al (2002) who used a Grounded Theory approach to identify four domains, these being 'Sense of Self', 'Social Relationships', 'Culture and Community' and 'General Quality of Life'. Further analysis by Patrick et al (2002) supported an overall score across domains. Forty-one perceptual items (known to the young person alone) are scored on an 11 point scale from 0 ("not at all") to 10 ("A great deal" or "Completely"). After summing these 41 responses they are transformed to a 0-100 score, with higher scores indicating better quality of life. In addition to the perceptual scale the instrument includes 15 individual contextual items. These 15 items do not form part of the scale but are individual measures to assess the context of the individual's life<sup>30</sup>.



**3.3.4. Functional assessment.** The Muscular Dystrophy Functional Rating Scale (MDFRS)<sup>31</sup> was designed to measure the functional impact of muscular dystrophy. Four aspects are assessed – 'Mobility' (implying change in location); 'Basic activities of daily living'; 'Arm function' and 'Impairment' (including limb contractures, neck and trunk strength, and use of ventilator). Each item is scored on a scale of 1 to 4 with 1 being total dependence and 4 being independence. Studies have included up to 121 boys and young men with DMD and Becker muscular dystrophy (mean age 20.0 SD 12.4 years; range 7 – 61 years). Of these 65% had DMD. Developers concluded that the instrument was simple to administer and score and suitable for patients with muscular dystrophy aged 6 and above. In the current study the MDFRS forms were completed by the young men's clinical team.

**3.3.5. Parent questionnaire.** The parent questionnaire, posted to parents prior to the young man's interview, was based on one designed for studies of pain in non-verbal children with neurological impairments<sup>32</sup>. Good quality data and a 50% response rate were achieved in those studies, which was thought satisfactory given the stress and work load of these parents. The questionnaire provides space for parents to describe up to three pains suffered by their son, the severity and frequency of pains and to assess the extent to which pain interferes with daily activities. They are asked how they know their son has pain, to list treatments and therapies received by the young person and to indicate if any of these are given for pain.

**3.3.6. Interviews:** Since some young men with DMD might be limited in their capacity to independently complete a written form, face-to-face interviews were chosen as an approach to use consistently across all participants providing the opportunity to clarify questions and explore responses as needed.

Two investigators (AH and AP) visited the young men's homes to conduct the interviews with them and collect survey data from parents if this had not been received prior to the visit. Prior to interviews informed signed consent / assent was provided by the young men, and signed consent of their parents for those aged less than 16 years. Interviews were digitally recorded.

### **3.4 Data analysis**

**3.4.1 Quantitative data:** Numerical data from the pain, pain coping, quality of life and function measures were analyzed using SPSS (version 22). Correlations between pain, pain coping, quality of life and function were calculated using Kendall's Tau statistic. Differences in perceptual YQOL score between those with moderate to severe pain daily (a high pain group), and those with less frequent pain (a low pain group) were examined using the Mann Whitney test. We calculated from the correlation between pain score and quality of life the number of participants that would be required to reject the null hypothesis of no correlation, with  $p < 0.05$ . Where there was occasional missing data this was replaced with the average for the sample. Where there was a failure to respond to an item by more than 30% the item was omitted. The data set was examined for areas of missing data that might inform the design of further studies.



**3.4.2. Qualitative data:** Digital recordings of the interviews were transcribed and transcripts examined manually by two investigators for themes that described the young men's pain experiences and coping methods. Themes / codes were identified inductively from the transcripts. Themes and quotes describing them were entered into a spreadsheet matrix. Links between pain, pain coping and quality of daily life were explored. A third investigator drafted the findings and these were confirmed as accurate by the investigators who had carried out and coded the interviews.

## 4.0 RESULTS

**4.1 Participants:** After initial largely unsuccessful attempts to recruit young men and their parents/carers through postal invitations, a more personal approach to recruit was used via their paediatric NHS consultants either by phone, letter or in clinic. Fifteen young men and their parents/carers were recruited. Twelve young men and their parents/carers provided full sets of interview and questionnaire data and these results are presented.

Young men were aged from 11 to 21 years (median 15 years 6 months) (Table 1). All were interviewed in their home. Interviews lasted from 30 to 45 minutes. All young men asked for their parent/s or other main family carer (e.g. grandparent) to be present during the interview. Occasional individuals appeared anxious and were reassured. With the young men able to see the forms, questions were read by the interviewer sitting next to him and most had no difficulty in providing their ratings either through speech or by pointing to a score. They were able to engage effectively in the dialogue and respond to prompts to elaborate on their responses. Contributions by parents to the conversations were not discouraged although the young man's view was checked, as appropriate. Item 21 in YQOL states 'I feel comfortable with my sexual feelings and behaviours'. Five (42%) young men asked not to respond to this question and for all participants the YQOL score was calculated as a percentage omitting this item.

## 4.2 Quantitative results

### 4.2.1 Pain severity and frequency

Two thirds of young men and their parents reported more than one site or source of regular pain. Pain sites young men reported most often were legs in nine (including thighs, ankles, knees and feet), and back in eight. Other commonly reported pains were gastric pain and neuropathic type pains including numbness, burning, and pins and needles. Pain scores are reported in Table 1. Correlation between CAS and FPS-R was 0.655;  $p=0.007$  (Kendall tau-2 tailed). There was no correlation between pain scores and age.

In their survey, parents described their sons' worst pains as 'mild' in one case, 'moderate' in seven, and 'severe' in three cases. One young man was reported not to have pain. Eight had pain daily; in all of these, pain was reported to be of moderate to severe intensity. Two were described as having pain 'all the time' and six as having pain 'some time each day'. Pain was reported as occurring less than daily in four young men. Parents reported that pain 'often or always interfered' with the young man's sleep and lying in six and sitting and moving (or being

moved) in five. Pain interfered less often ('not at all' or 'occasionally') with enjoying time with the family, favourite pastimes and attendance at school or college.

In the parent survey, 8 of 12 young men were reported to receive analgesics as required (paracetamol, and ibuprofen either orally or as topical gel). Only one individual under the care of a pain clinic, received regular treatment for pain administered via gastrostomy, this being paracetamol four-hourly, tramadol four-hourly, amitriptylline morning and night. Eight young men received laxatives (mainly movicol) and two received omeprazole for gastric pain. Eight were receiving corticosteroids.

#### 4.2.2. Function

Scores for the MDFRS and its domains are included in Table 1 (MDFRS score was unavailable for one young man). Each of the domain scores was highly correlated with the total MDFRS score. There was a significant correlation between the total MDFRS score and age ( $p=0.046$ ). The relationships between MDFRS domains and pain were not significant.

#### 4.2.3. Pain coping

Scores on pain coping domains are included in Table 1. Differences between parent and young men's ratings on PPCI domains of coping were not significant (Figure 2). Neither young men's nor parents' evaluations of pain coping strategies correlated with pain frequency, pain severity or quality of life.

#### 4.2.4. Quality of life

Perceptual YQOL scores (calculated as a percentage excluding item 21) are reported in Table 1.

Higher parent-reported pain frequency and severity each correlated with lower young men's-reported perceptual YQOL ( $p=0.03$  and  $p=0.043$  respectively. Kendall's Tau statistic). Eight young men, reported by their parents to have moderate to severe pain daily (a 'high pain' group) had significantly lower perceptual YQOL scores (mean 72.2 SD 6.5) than did those with less frequent and less severe pain (a 'low pain' group) (mean 88.6 SD 6.4) (MW  $p=0.008$ ) (Figure 3). This was the case for all domains as well as the total score.

A power calculation based on correlation between the VAS pain score and YQOL score demonstrated that (assuming a magnitude of 0.4 using a 5% test with 80% power) would require 47 participants.

### **4.3 Qualitative findings**

Three main themes were identified from the qualitative analysis: 'Hurting in different places', 'Telling: Holding it in and letting it out' and 'Acting to relieve pain'.

#### 4.3.1. Hurting in different places

The qualitative findings resonated with the quantitative reports of the pain sites (legs, back, head) and sources (musculoskeletal, reflux, falls), the nature of pain (e.g. neuropathic) and pain severity (mild through to intense) often occurring with everyday activities such as sitting, lying and change of position.

One young man explained his pain by saying "It's like hurting ... It sometimes gets worse ... Sometimes it's in different places" (YM6). Another described his hip pain as being "tingling, dull, ...just like an irritation... I kind of get nerve pain". He went on to describe his back pain as related to "tension in my shoulders" and "in [my] legs when in chair. It's up my side, [...] and ankles" (YM9).

Young men used figurative language such as "crack", and "plunk" and "bullet" to describe the sensations associated with their pains. One young man, explained: "When I'm in bed, when I sit up, my back cracks. It just feels like it's going to crack and then I sit up and it goes ["crack"] (YM4). Another described how his headaches, which occurred mainly at school, feel like there is "a hammer banging in my head". (YM2).

Pains were often associated with lying down or sitting up, as illustrated by YM4 explained his pain was "mainly in the muscles in my knee when I've been in the chair for a while" (YM4). Young men's descriptions were often supplemented by their parents, YM4's father adding "when you wake him up in the morning, his back, it's mainly his back when you sit him up. Also your hands ache when playing the play-station, don't they?" Another young man reported feeling "more uncomfortable when I'm lying flat on my back" (YM7).

Linked to positioning, both medical (e.g. splints, shower chairs) and leisure equipment (Playstation) could exacerbate or otherwise influence their pain. One young man described how using his Playstation caused him to "get pains here [hands] now" (YM4). A swivel bathing chair was "hated" by YM7, and his mother who reported the chair was "very, very uncomfortable" and "terrible because it was pulling his back and his legs" and her son was emphatic saying "Don't remind me of that. I hated it!" Interventions aimed at relieving pain and disability could also cause pain. Splints worn by YM7 were discarded for "when he took them off in the morning he couldn't walk at all and it took hours and hours to get [moving]" (mother of YM7). YM9 had had significant pain following insertion of spinal rods, however, after 6 months "he never looked back" (mother) and the young man agreed saying "I was straight and I could breathe". Cold weather could precipitate circulation problems and exacerbate pain as YM4 and his mother explained "... he gets cramp a lot in winter in his legs. YM4 added "Sometimes I don't know if they are cold or not".

#### 4.3.2 Telling: holding it in and letting it out

As seen in the previous theme it was clear that the young men could locate and describe their pain; it was equally apparent that they were selective in who they talked to about their pain, who they asked for help, and somewhat cautious in what they directly revealed, although their emotional response (withdrawing, anger, crying, loss of temper) was often indirectly indicative of pain.

Most of the young men only confided in and talked about their pain or discomfort with their parents though even these discussions might need prompting, as the mother of YM5 explained: "He usually tells me if he is in any pain at home. Sometimes though, I have to ask him if he is ok or in pain". Another mother explained that her son (YM11) "will only tell [her] if it [pain] gets bad". Whilst,

talking about pain at home was not easy, talking about it at school was even more challenging, and although YM12 was able to "tell one of the teachers"; other young men did not feel able to do this. As the mother of YM5 explained in her survey, "at school he will not say if he is in pain, but often tells me as soon as he reaches home". It is interesting to note that few of the young men or their parents reported telling a doctor about pain and would only do so "if pain was very severe" (YM2). This may be explained by the limited contact that the families have with doctors they trust. The mother of YM5 explains "My son sees his consultant twice a year. Apart from when he sees his consultant I have no confidence in any other medical professional".

There was a sense that pain was an expected part of life; a "norm" albeit one that could have a major impact on the young men's emotional and behavioural state. YM9 and his mother only fully realised he had been in pain when the pain was taken away through successful intervention via a pain clinic. They had since "discussed and reflected" and agreed that he should not delay in telling her about future pain and that he should not "leave [telling her about] it".

When the young men choose not to directly tell other people about their pain, they are dependent on their parents and carers correctly picking up on their behavioural and emotional indications of pain. Broadly, the young men either held in their feelings by becoming withdrawn or let go of their feelings through emotional outbursts. YM2's mother explained that often her son "will cry and seem depressed if he is in pain" and that "sometimes he wants to be left alone". For others there is a release through 'letting go', being "awkward", "shouting and swearing". The mother of YM12 explained that her son's emotional response of grumpiness and anger was a result of him feeling that "his body is letting him down". YM5's mother said her son "will often lose his temper and shout at anyone who goes near" and YM4 explained that when he was in pain "I swear all the time"; his mother agreed saying "If you sit him up in the morning and his legs are hurting, he will say 'You stupid cow, you've hurt me'".

#### 4.3.3. Acting to relieve pain

The inability of most of the young men to move themselves makes them vulnerable to unrelieved pain. They can be totally dependent on their parents or carers for change of position, or, for instance, easing their pain through rubbing and massaging. The reluctance of the young men to talk about their pain may be linked to this reliance on other people to take action on their behalf. Positioning and repositioning required "patience" and "time", as YM1's mother explained: "he wakes 3 to 4 times per night and needs repositioning". Massage was helpful for some young men in dealing with cramp-related pain; YM4 explained that someone "rub[bing] my ankle.... eventually [helps]" the pain that sometimes "feels like a bullet". For others, swimming was beneficial, though was not always available, as YM10's mother says "water therapy is the best but is not provided here [in this locality]".

Although dependent on others for repositioning and massage, some young men drew on distraction as a more independent means of taking action. YM2 was reported by his parents as using a lot of distraction including "TV, books, games,

hugs and talks about it". However, distraction was not always a successful strategy, as YM9 explained "I do try to do things to distract myself but it doesn't really work".

Young men were ambivalent about using medication to manage pain. Some medications were difficult to swallow whilst others had side effects such as becoming "gassy" from Meloxicam (YM9). This young man, who was on regular medication for his pains administered via a gastrostomy, also had haemophilia and was keen to explain how the experience of pain with each condition differed, and how the difference between them led him to feel differently. He explained "I don't like that pain (from bleeds) as much because I know it won't get better as quickly as if I was to have medicines - I have to wait for the Factor 8 to work". Being able to predict timely relief from his prescribed pain killers helped to alleviate his anxiety.

YM14 explained that he might take medication when his pain "gets to a certain point" although "I don't like taking medication". His mother explained that he had said in the past "If I take tablets I feel like I'm giving up". He agreed with his mother when she said "... you're a battler aren't you; you'd rather battle against it than take painkillers". The mother of YM9 reported that he had "mentioned... that it's almost like it's not macho if you have to take a painkiller".

## 5.0 DISCUSSION

This study demonstrated that constructing semi-structured interviews around the administration of validated instruments is feasible and that useful qualitative and quantitative data can be obtained using this mixed-methods approach. The methods allowed for triangulation of findings from numerical scales, qualitative analysis from interviews and data from parent surveys. The study informs our understanding of pain experienced by young men with Duchenne Muscular Dystrophy and the effect it can have on their lives.

This study and the small number of studies investigating pain in this population show that pain can be a characteristic of the disease throughout its course, indeed Douville et al (2005) found that pain (largely due to falls) was greater in the younger boys who were still ambulant<sup>16</sup>. Bushby et al., (2010) describe potential interventions many of which will help prevent or alleviate pain<sup>17,33</sup>.

In this pilot study the majority (two-thirds) of young men (not recruited for the presence of pain) suffered from significant daily pain. Whilst overall self-reported quality of life was good, poorer quality of life was found in young men with parent-reported daily pain of moderate or worse severity. Often young men were limited in their capacity to manage their own pain and dependent on adult caregivers to assist them in pain relief. Pain complaints were largely kept within the family. The findings suggest perhaps a psychosocial problem. How is it possible for young men who require frequent assistance to seek help in a manner acceptable to those who might assist them without being or appearing withdrawn, depressed, awkward or angry? The young men and their parents seem to be 'between a rock and a hard place'. Managing pain in this population requires empathy and acceptance from professionals, and rest and support for parents.



Pain is not only a sensation, it can also be given meaning by the sufferer. Several of the young men describe associations between their pain and bodily function, in that they express (sometimes with the help of their parents) fear that pain indicates a further loss of function, their 'body is letting them down', that taking medication for pain indicates 'giving up', or even loss of manhood "not being macho". Eccleston et al., (2012) caution against viewing children's pain coping and in particular 'catastrophising' (defined as magnifying, rumination and helplessness) as maladaptive, as 'it may be normal for young people to think of painful events as serious, to think about wanting them to stop and to feel helpless to change them'<sup>34</sup>. Given that young men with DMD can be largely dependent for help from parents and professional carers one of the tasks of carers could be to help build the young men's sense of control and participation in finding ways of alleviating their pain, for example, choices of equipment, seating, beds and types and routes of medication.

The World Health Organization (WHO) has issued advice on the pharmacological management of persisting pain in children with medical illnesses<sup>35</sup>. Based on our findings we suggest that more consideration should be given to providing pre-emptive analgesics for incident pain and regular "around the clock", analgesics, including adjuvants such as amitryptiline, gabapentin and carbamazepine for pain that is persistent or recurring and is interfering with activities of daily living. Where opioids are required, laxatives should be prescribed alongside. In the event that the young men are reluctant to take oral medication, finding pharmaceutical preparations that are acceptable to them could be challenging. In this study, topical gel preparations and transcutaneous electrical nerve stimulation (TENS) appeared acceptable and helpful to a number of young men. As also reported by Lager and Kroksmark (2015)<sup>9</sup>, young men and their parents reported that hydrotherapy and swimming were helpful, but pools were not always available, and few of the young men saw a physiotherapist outside of their six monthly or annual follow-up.

We suggest that during all routine clinic or home visits it is important for clinicians to ask the young man and family about the presence of pain and discomforts and take a comprehensive pain history. The use of a body map and pain scale, such as those used in our study may help to focus the conversation and show serious intent. Discussion with the young man and parents may elicit preferred pain management strategies and provide scope for additional ideas for professional input including psychology, physiotherapy, occupational therapy and wheelchair services. Young men with complex persistent or recurring pain should be assessed by an expert in neuromuscular diseases and may benefit from referral to a pain clinic.

Whilst the study was small it is in line with the numbers suggested for pilot studies<sup>36</sup> and is not unusual in qualitative studies. With regards to a larger study, a power calculation based on results here suggests a minimum of 47 participating young men which appears feasible across a number of neuromuscular centres. The study described took a considerable time to complete due to consecutive recruitment of centres to the study, but recruitment improved greatly once families were approached more personally and data collection interviews themselves were not problematic. Only five of 12 Parent Questionnaires were returned by post prior

to the young men's interviews or were ready to collect at the time of the visit; the remaining parents accepted help at the time of the young men's interviews. It did not appear that parents had particular difficulty in completing questions but had put forms aside waiting for available time to complete them. The young men and their parents found the visit, interviews and the chosen instruments acceptable and when asked had no complaints or additional suggestions about the process.

We are aware of only very minor adjustments required to our own data collection procedures. There was no correlation between these assessments and YQOL scores. The young men's assessments on the CAS and Faces pain scales were their assessments of the pains they reported regardless of when they occurred. Analysis would have been enabled by more formal recording of, for instance, worst pain ever and worse pain in the last four weeks. In addition, the measures of function using the MDFS were completed by an individual on the clinical team from memory of the young man and clinical records. In one individual the function measure was not available as the young man had left the service. The functional assessment might be better conducted through observation and dialogue with the young man and parents during the interview visit. Although not designed specifically for this population, the addition of the Gross Motor Function Classification System (GMFCS) Family or Self Report Questionnaire as part of the interview or Parent Survey is a possibility. A central microphone was used to record the interview, but young men spoke quietly and a lapel microphone might have been more appropriate.

In assessing their quality of life, young men were generally very positive about the quality of their family life. The young men had the option of talking to us on their own, but all asked for their parents to be with them. Many looked to their parents for reassurance to talk about their experiences but we did not feel that the veracity of their responses was affected by the presence of their parents. Their relationship with parents appeared positive in our sample, however, as Craig et al.,<sup>37</sup> state, in perceiving a child's pain, parents and other adults normally experience an empathetic distress which in turn and in most circumstances moves them to take measures to relieve the child's pain. The young men are likely to be very vulnerable should relationships be less positive.

## CONCLUSIONS

We found that conducting pain research is feasible and worthwhile in this group of young people. The majority of young men in this study experienced significant daily pain and increased pain frequency and severity was associated with poorer quality of life. The young men do not readily communicate their pain to professionals and are limited in their capacity to manage their pain independently of their caregivers. It is important for family and professional caregivers to ask the young men about their experience of pain and offer physical, medical, psychological and social support for pain coping. Further research in to pain management and pain coping in this group will be helpful.

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Table 1. Descriptive Statistics for Age, Pain (CAS, FPS\_R), Function (MDFRS) and Quality of Life (YQOL)

	N	Minimum	Maximum	Median
Age (years)	12	11.0	21.0	15.0
YM's Pain measures (a)				
CAS	12	3.5	10.0	5.5
FPS_R	12	0.0	10.0	5.0
Mean MDFRS score (b)				
MDFRS Domains				
Mobility	11	1.1	3.0	1.3
Basic ADL	11	1.0	3.3	1.3
Arms	11	1.6	3.3	2.4
Impairment	11	2.3	3.9	3.0
Perceptual YQOL (c)				
YQOL Domains				
Sense of self	12	43.8	79.4	65.0
Social relationships	12	58.6	98.6	78.9
Culture and community	12	65.0	100.0	79.0
General quality of life	12	46.7	100.0	90.0

- a) Colour analogue scale (CAS) and Faces Pain Scale (revised) (FPS\_R): Higher scores indicate worse pain.
- b) YQOL scores transformed to percentages (calculated excluding item 21): Higher scores indicate better quality of life.
- c) Muscular Dystrophy Function Rating Scale (MDFRS): Higher scores indicate worse function.

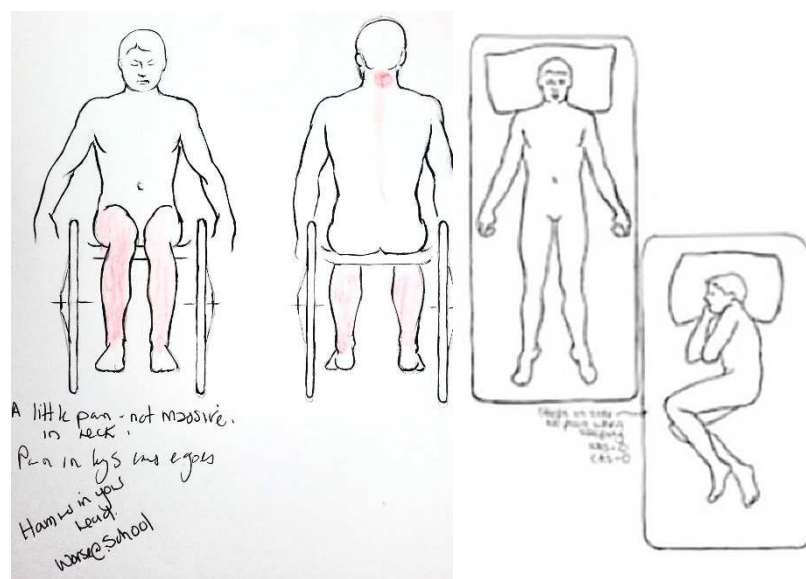


Figure 1.

Body map designed specifically for this study. Distribution of pain described by young man (aged 11).

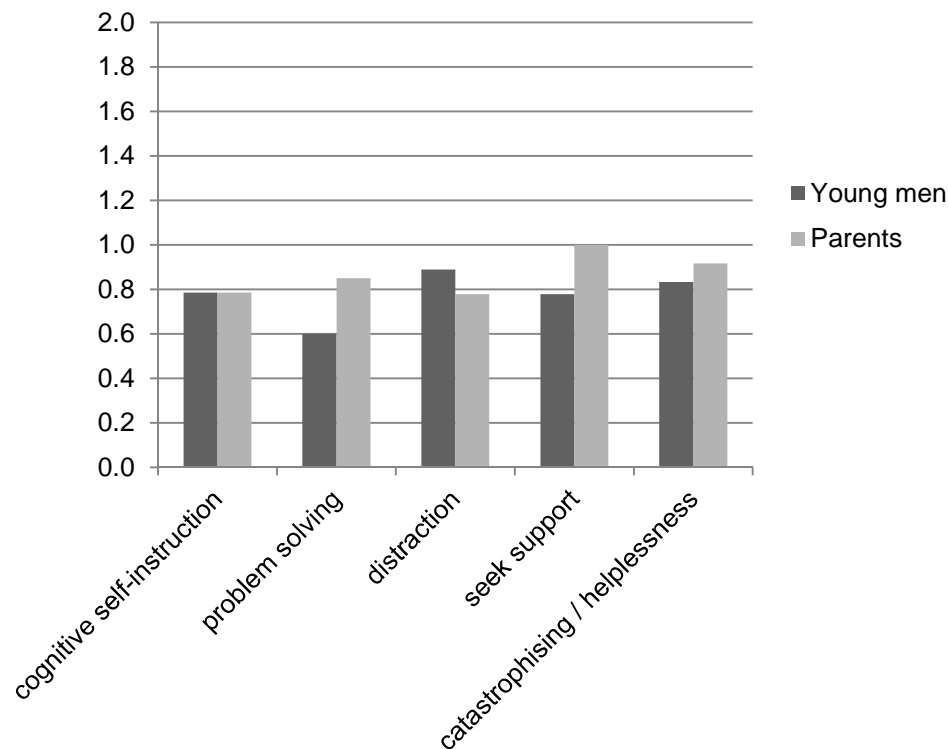


Figure 2. Pain coping strategies of young men with Duchenne Muscular Dystrophy by self and parent report.

Average (median) of young men's and parent's scores on pain coping domains of Pediatric Pain Coping Inventory (PPCI). Differences between young men and parent scores are not statistically significant on any domain.

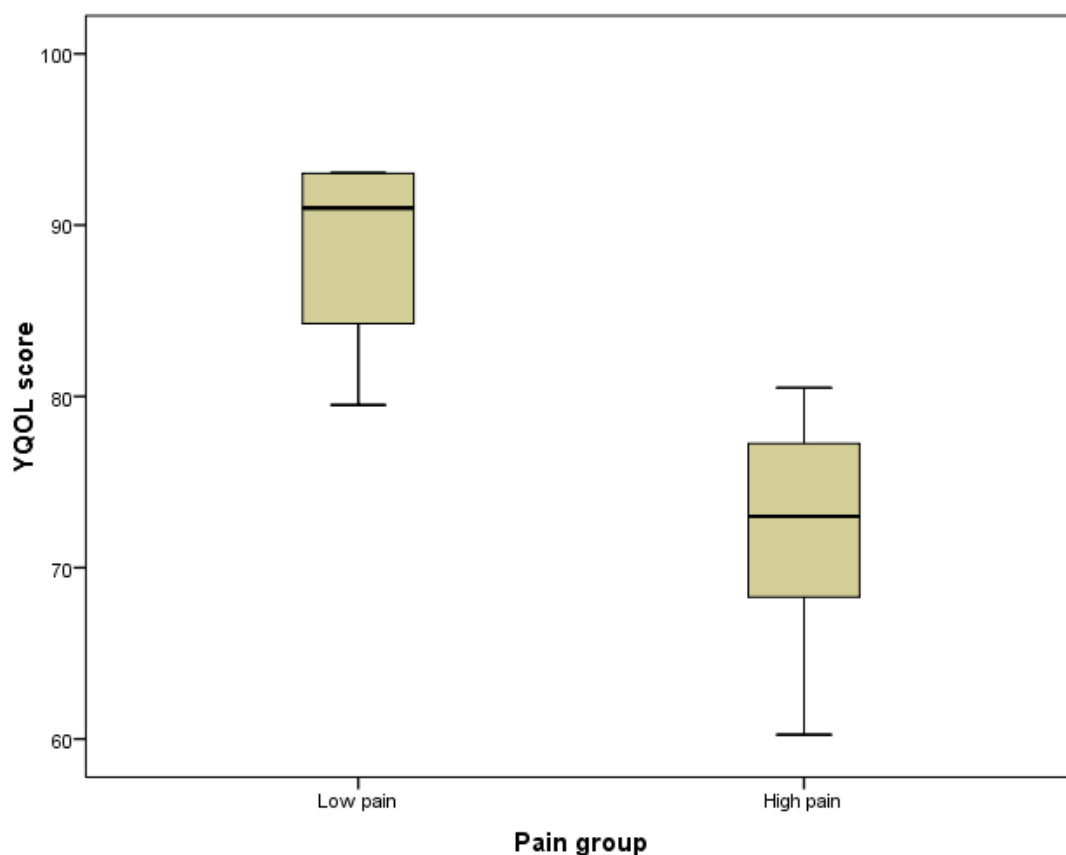


Figure 3. Perceptual YQOL score by pain group.

High pain group had daily pain of moderate or worse severity. Young men who had daily pain of moderate or worse severity (a high pain group  $n=8$ ) had significantly lower quality of life as measured using the Youth Quality of Life Scale (YQOL) than a low pain group ( $n=4$ ).

Perceptual YQOL score is calculated as percentage excluding item 21 which had low level of response.



**Highlights**

- We add evidence of significant pain in young men with Duchenne Muscular Dystrophy
- We provide insights into their pain experience, pain coping and quality of life
- Young men with moderate or worse pain daily had poorer quality of life
- They do not readily report pain outside of the family. Clinicians need to ask them.
- We provide preliminary data to inform power and size for a future study