Being a young adult with Duchenne's Muscular Dystrophy: wellbeing and priorities for an adult life

Kevanne Sanger

Thesis submitted in partial fulfilment of the requirements of Staffordshire University for the degree of Doctorate in Clinical Psychology

October 2019

Total word count: 18,494

THESIS PORTFOLIO: CANDIDATE DECLARATION

Title of degree programme	Professional Doctorate in Clinical Psychology
Candidate name	Kevanne Sanger
Registration number	16025080
Initial date of registration	September 2016

Declaration and signature of candidate
I confirm that the thesis submitted is the outcome of work that I have undertaken during my programme of study, and except where explicitly stated, it is all my own work. I confirm that the decision to submit this thesis is my own. I confirm that except where explicitly stated, the work has not been submitted for another academic award. I confirm that the work has been conducted ethically and that I have maintained the anonymity of research participants at all times within the thesis. Signed: Date:

Acknowledgments

Thank you to my supervisors for their guidance, wisdom, and patience. You all have such unique skills, and without each of your contributions this would not have come together.

Thank you to the amazing MD Team and R&D department at RJAH, and the wonderful people that work at the NMC. Your experience and professionalism kept the service user at the heart of this work, and my passion for it alive.

Thank you to all those people who agreed to be research participants, both in my own study and those that I have referenced. I hope I have done your voices justice.

Mi Amor, I wouldn't have survived the last three years without you. Let's dance through life together.

Preface

Chapter One is written using the American Psychological Association (APA) formatting style, which has been adopted by the Journal of Disability & Society (IF – 1.613) where this article will be submitted for publication. Chapter Two follows the Vancouver formatting method, to align with the style of Neuromuscular Disorders (IF – 2.969) where the article will be submitted for publication. The guidelines for authors will be included in the relevant Appendices sections.

These journals are standardly accessed and submitted to by NHS Hospital Trust that collaborated with the thesis author on the empirical research study.

Paper 1: Literature review: 7,992 (abstract - 112)

Paper 2: Empirical paper: 7,992 (abstract - 241)

Executive summary: 2,018

Thesis abstract: 330

Total word count: 18,685

Thesis Contents

Thesis Abstract

This thesis was completed as part of the Doctorate in Clinical Psychology at Staffordshire University, completed by the first author. The topic originated from the personal experiences of the first author, and an interest in the relationship between physical and mental health, particularly at developmentally significant times like the transition from adolescence to adulthood. Muscular Dystrophy (MD) is a degenerative condition that causes progressive levels of disability and limits life expectancy. Chapter One systematically evaluated the current literature on the wellbeing of adolescents and emerging adults with MD. Nine papers were critically reviewed, and results suggested that the wellbeing of this population is reliant on supportive relationships, autonomy, and a maturing sense of self-acceptance, and they are at an increased risk of mental health difficulties. While the quality of the research varied and conclusions were interpreted with caution, the review highlighted a need for earlier psychological support with a focus on systemic, collaborative working. Chapter Two investigated what emerging adults with Duchenne's MD (DMD: the most common form of this genetic condition), primary carers, and healthcare professionals value for a positive and autonomous adult life with DMD. A Q-method study was used to explore whether these three expert stakeholder groups prioritise similar goals when engaged in transition-based care. Results of the factor analysis found two different views of how to facilitate a positive adult life in society; one that valued taking on a more adult role within their existing system, and another that believed adulthood was enabled by breaking away and accessing new experiences. The mix of expert stakeholders within factor one, and combination of one emerging adult and three healthcare staff in factor two, suggests that transition planning should be held with these different value-bases in mind; potentially guiding exploratory conversations about how to best support the adult life valued by the individual. Chapter Three is a summary of the empirical research, for dissemination to young people with DMD and their families, as well as healthcare staff.

Chapter 1: Literature Review

Psychological wellbeing in adolescents and emerging adults with muscular dystrophy: A review of the literature

Word count: 7,992

Contents

Psychological wellbeing in adolescents and emerging adults with muscular dystrophy: A review of the literature

Abstract

A review of the current literature investigating psychological wellbeing in adolescents (10-19 years) and emerging adults (18-29 years) living with muscular dystrophy (MD) was undertaken. Nine papers were identified, with various forms of MD and cultural contexts, but common themes included autonomy, relationships, the gradual process of self-acceptance, and heightened risk for mental health difficulties. Recommendations are made to replicate and expand research evidence, which currently covers a wide spectrum of disorders and age cohorts with few studies per sub-group to support conclusions. In terms of maximizing the wellbeing of young people living with MD, systemic support, co-production of health care planning, and preventative psycho-education for younger adolescents and professionals is advised.

Introduction

Muscular dystrophy (MD) consists of a group of genetic, degenerative muscle-wasting conditions that affect one in 1,000 live births (Muscular Dystrophy UK, n.d). New treatments, including corticosteroids and assistive breathing apparatus have extended life expectancy in life-threatening forms of the condition by delaying heart and respiratory failure. These recent advances have led patients and families to readjust their life expectations, now anticipating to live into their 30s or 40s. However, the psychological needs of this adult cohort are still unclear, leaving medical and psychological practitioners to provide support without a clear rationale or guidance for best practice. Research evidence of how young adults with MD experience their life while managing a decline in physical health is growing. However, many studies include small samples, and are heterogeneous, spanning various forms of MD and cultural populations. As a rare condition, with over 30 specific diseases (NINDS, 2011), larger trials are difficult and timely to orchestrate. However, exploring adult transition for people with MD within healthcare services is increasing, which should lead to more robust studies and generalisible conclusions.

Most forms of MD are diagnosed in childhood, with symptoms of deterioration first occurring in adolescence or emerging adulthood (Muscular Dystrophy UK, 2017). Adolescence is defined by the World Health Organisation (WHO) as a person aged 10-19 years, and emerging adulthood as the stage between 18-29 years (Arnett, 2000); terms often synonymous in literature with ‘young adults’. At a point when most people are developing a sense of autonomy, self-identity, and adult role taking within their communities (Arnett, 2000; Erickson, 1982), many people with MD are making adjustments for their physical abilities, and accepting more reliance on other people, and assistive technologies like wheelchairs or breathing apparatus. Declining physical health is likely to restrict this population’s options for exploring the emerging self outside of the family context. However developments in social media and specialist support groups provide avenues for self-exploration. The Trailblazers young person’s network within Muscular Dystrophy UK is one such example, campaigning for social inclusion, facilitating networking events, and highlighting positive role models with MD. Nevertheless, adolescence and emerging adulthood is a critical time in developing inner fidelity (Arnett, 2000; Erickson, 1982), which may be restricted by declining physical health in MD conditions. This could impact negatively on the psychological wellbeing of young adults living with MD. As a

construct, wellbeing has been defined in various ways, but this paper will refer to the six-factor model (Ryff, 1989; Ryff & Keyes, 1995), which involves the perceived satisfaction of autonomy, environmental mastery, personal growth, positive relationships, purpose in life, and self-acceptance (Ryff, 1989; Ryff & Keyes, 1995). Little research to date has investigated the impact of MD on well-being, however the wider health literature has shown that chronic health difficulties reduce opportunities for developing environmental mastery (Mangelli, Gribbin, Buchi, Allard, & Sensky; 2002), and that well-being is reduced in those with long-term health conditions (Pusswald et al., 2012).

Reduced opportunities and increased social barriers during adolescent and emergent adulthood can affect identity formation, self-efficacy, and mental health (Yodder, 2000). This in turn can increase burden on local support systems, and communities could lose valuable contributors (Economic and Social Research Council [ESRC, n.d]). Asking how young people with MD are psychologically coping with transition through adolescence and into adulthood amidst declining physical functioning, and its impact on wellbeing could help services to specifically target their interventions. Developing more person-centred guidance for supportive practices could optimize the wellbeing and resiliency of these young adults, and improve the lives of service users and their families, and their wider communities.

Study Aim

This paper will evaluate and synthesize the available literature on the psychological wellbeing of adolescents and emerging adults living with MD, to highlight the psychological impact of transitioning into adulthood within the confines of a life-limiting condition. This in turn could inform support strategies used by families, carers, local authorities, and governmental bodies, encouraging more evidence-based preventative initiatives and secondary interventions.

Research Question

- What does research suggest about the current state of psychological wellbeing in adolescents and emerging adults living with MD?

Method

A systematic review of published research using key terms selected by the authors took place in May 2018. No date restrictions were placed on literature searches, so as to investigate any historical cohort effects within the literature.

Inclusion and exclusion criteria

The inclusion and exclusion criteria applied to the literature search are stipulated in Table 1.

Table 1: Inclusion and exclusion criteria for review

Inclusion Criteria	Exclusion Criteria
• First person report from MD participants • Data from adolescents as defined by the World Health Organisation (WHO [10-19 years]) • Data from emerging adults as defined by Arnett (2000 [18-29 years]) • Studies of MD service user wellbeing or mental health • Peer reviewed (incl. student theses)	• Not available in English • Studies where the target age populations aren't distinguishable • Studies only measuring cognitive performance • Intervention or review articles

Search strategy and article selection

Key words were refined by the authors and used to search five Psychology/Sociology and Health Sciences databases (MEDLINE, CINAHL Plus, SPORTDiscus, PsycINFO, PsycARTICLES) through EBSCO, and a systematic search through the Google Scholar database. The key terms used were "muscular dystrophy", "young adults", "adolesc*", "wellbeing", "depression", and "anxiety". Figure 1 illustrates the process of the literature search, refining the generated article list into a final selection of nine studies for review. The nine papers included three qualitative studies, two mixed-method designs, three quantitative cohort studies, and one quantitative case-control design.

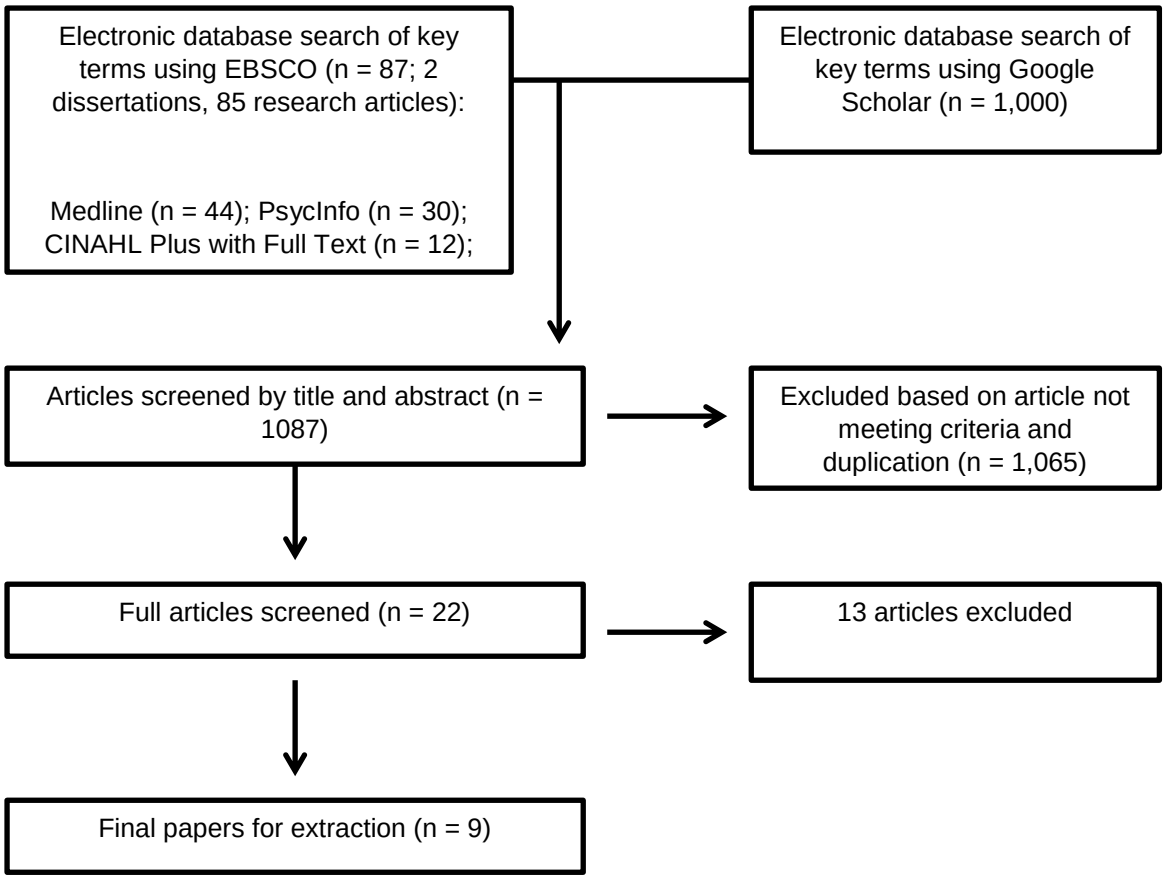


Figure 1: Flowchart of research literature selection for review

Quality assessment

In line with the Health and Care Professions Council (HCPC) standards of proficiency (HCPC, 2015), and the recommendations of the National Collaborating Centre for Methods and Tools (Ciliska, Thomas, & Buffett, 2008), papers were evaluated using the Critical Appraisal Skills Programme (CASP) tools. The CASP tools used in the current review were the case-control checklist, the cohort study checklist, and the qualitative checklist (see Appendices I-III for details). These appraisal guides ask 10-12 critical questions depending on the study design, from which the quality of a given study can be determined. In line with previous relevant systematic reviews (Kerr et al., 2017; Johnston, Jindal-Snape, & Pringle, 2016), the first author scored the resultant nine papers on 10-12 criteria points, allocating studies 0 (not addressed), 1 (somewhat addressed), or 2 (fully addressed). Full details of the CASP scoring in the current review can be seen in Appendix IV. All papers were included in the subsequent synthesis however ‘low quality’ studies that scored 0-9 points were noted

with caution, in keeping with similar systematic reviews (Kerr et al., 2017; Johnston, Jindal-Snape, & Pringle, 2016).

Results

Nine papers were included in the final review, summarised in Table 2. Each study will be outlined, and discussed in terms of their methodology and key findings, before critically appraising the research using the CASP tools.

Literature Summary

Aho, Hultsjo, & Hjelm (2015) – A qualitative study with 14 participants (18-30 years) with recessive limb-girdle muscular dystrophy (LGMD2). Semi-structured interviews were based on the Sense of Coherence (SoC) questionnaire (Antonovsky, 1987) and analysed using salutogenic-informed content analysis.

Aho, Hultsjo, & Hjelm (2018) – A qualitative study with 14 participants (18-30 years) with LGMD2 and 19 parents. Semi-structured interviews were based on the SoC questionnaire (Antonovsky, 1987) and analysed using salutogenic-informed phenomenographic analysis.

Conway, Mathews, Paramsothy, Oleszek, Trout, Zhang, & Romitti (2015) – Medical record abstraction from a large cohort of 857 males from 765 families. The final data abstraction point was at 17 years.

Elsenbruch, Schmid, Lutz, Geers, & Schara (2013) – Quantitative cross-sectional study of 50 young people (aged 8-23 years) with Duchenne's muscular dystrophy (DMD), using standardised measures of depression and health-related quality of life (HRQoL).

Huismann, Sheldon, Yashar, Amburgey, Dowling, & Petty (2012) – quantitative cross-sectional, comparison study. Participants included 24 young people with congenital myopathies and muscular dystrophies (CM/CMDs), aged 18-29 years, and 93 age-matched controls. The study reported markers of emerging adulthood using standardised measures.

Hunt, Carter, Abbott, Parker, Spinty, & deGoede (2016) – Mixed methods study with 12 young men with DMD (11-21 years), and their parents, utilising standardised measures of pain and pain coping, supported by semi-structured interviews.

Jacobs, Willekens, Die-Smulders, Frijns, & Steyaert (2017) – Mixed methods study with 27 participants with juvenile Myotonic Dystrophy type-1 (DM-1), aged 16-25 years. Standardised measures of functioning and life satisfaction were collected, and psychiatric interviews were assessed against the Operational Criteria Checklist for psychotic and affective disorders (OPCRIT).

Miladi, Bourguignon, & Hentati (1999) – Cross-sectional study with 16 young people with Severe Childhood Autosomal Recessive Muscular Dystrophy (SCARMD) or LGMD, aged 10-22 years. Markers of emotional distress were assessed using the Roberts Apperception Test for Children (RATC: McArthur & Roberts, 1982).

Read, Simonds, Kinali, Muntoni, & Garralda (2010) –10 young men with various muscular dystrophies (12-25 years) and their parents completed standardised measures of sleep quality, wellbeing, and depression. These were supported by semi-structured interviews with three families.

Aims

All studies included elements of psychological wellbeing in young people living with MD, but their focus varied. Three studies explored broader elements of the young person's experience; with Aho et al. (2015) inviting participants to explain what it is like living with LGMD2, while Hunt et al. (2016) and Read et al. (2010) focused on the experiences of young men and their families. Two studies focused on people's experience of transition; with Aho et al. (2018) interviewing participants with LGMD2 about their journey from diagnosis to requiring support with daily activities, and Huismann et al. (2012) investigated young people's developing sense of autonomy. Four papers explored levels of emotional distress in young people living with MD. Conway et al. (2015) reported on levels of depression and treatment sought by young adults with various child-onset MDs, Elsenbruch et al. (2013) investigated the impact of DMD on HR QoL and depression in children and young adults, Jacobs et al. (2017) conducted an initial investigation into the frequency of psychotic symptoms in young people with DM-1, and Miladi et al. (1999) looked at various emotional factors that might impact on the wellbeing of young people living with SCARMD or LGMD. Additionally, three studies aimed to gather pilot data for future projects - Jacobs et al. (2017), Hunt et al. (2016), and Read et al. (2010).

Samples and Recruitment

Many studies focused recruitment on one form of MD (Aho et al. (2018; 2015; Jacobs et al., 2017; Hunt et al., 2016; & Elsenbruch et al., 2013). However, four studies recruited participants with various MD conditions (Read et al., 2010; Conway et al., 2015; & Huismann et al., 2012; Miladi et al., 1999). Most studies recruited a broad age range, with five of the nine papers covering the entire adolescent and emerging adult spectrum (Hunt et al., 2016; Conway et al., 2015; Elsenbruch et al., 2013; Read et al., 2010 & Miladi et al., 1999). However, the other four papers focused on older adolescent and emerging adult cohorts (Aho et al., 2015; 2018; Huismann et al., 2012; and Jacobs et al., 2017). Most studies remained small, with seven of the nine papers having less than 30 participants with a MD condition. This limited the statistical power and generalisability of the four quantitative or mixed methods studies, however two are described as pilot projects (Jacobs et al., 2017; Hunt et al., 2016; Huismann et al., 2012 & Read et al., 2010). The three remaining small samples were from qualitative studies, with Aho et al. (2018; 2015) reflecting on the variety of experiences across their sample in order to reach data saturation, and Miladi et al. (1999) reported recruiting all available MD participants that met their inclusion criteria. The exceptions were Elsenbruch et al. (2013) with 50 young men with DMD, and Conway et al. (2015) with 857 males. All nine studies invited participants or gathered post-hoc data from hospitals. Additionally, Hunt et al. (2016) recruited from a local hospice, Aho et al. (2015; 2018) used a national neurological association and an unspecified web-based group for people with disabilities, and Huismann et al. (2012) utilised the Muscular Dystrophy Association's online research noticeboard to widen people's access to study participation.

Methodology

The included studies were equally split into three qualitative (Aho et al., 2018; 2015; & Miladi et al, 1999), three quantitative (Conway et al., 2015; Elsenbruch et al., 2013; & Huismann et al., 2012) and three mixed methods (Jacobs et al., 2017; Hunt et al., 2016; & Read et al., 2010) designs.

Two of the qualitative studies used semi-structured interviews, adopting a salutogenic approach (Antonovsky, 1987) that seeks to capture participants' health perceptions and experiences of living well despite adversity (Aho et al., 2018; 2015).

The generated themes were based on the SoC framework that was used to develop the interview schedule. This framework explains “coherence” as an individual’s perceived ‘comprehensability’ (whether there is a logical narrative to the event), ‘managability’ (perceived resources to cope), and ‘meaningfulness’ (belief that personal efforts are worthwhile or not). Miladi et al. (1999) used the RATC (McArthur & Roberts, 1982), where participants create stories from picture cards scored by a clinician for indicators of emotional distress. The qualitative methods used by two mixed methods papers were not clearly detailed, but included semi-structured interviews with young men and their families (Hunt et al., 2016 & Read et al., 2010). The qualitative aspect of Jacobs et al. (2017) was the subjective quality of the psychiatric interview, guided by the OPCRIT and Diagnostic and Statistical Manual version four (DSM-IV) criteria, and verified by a study-blinded clinician. The quantitative papers used a range of statistical methods. Jacobs et al. (2017), Conway et al. (2015), and Read et al. (2010) used descriptive means and percentages to define the wellbeing of participants. Read et al. (2010) employed correlation analysis, and the small-sample pilot studies applied non-parametric testing (Hunt et al., 2016 & Read et al., 2010). Hunt et al. (2016), Elsenbruch et al. (2013), and Huismann et al. (2012) used between group analysis of variance, as well as within group correlation analyses.

All of the quantitative and mixed-methods studies employed a cross-sectional design. Three of the papers compared young people with MD to post-hoc normative sample data (Conway et al., 2015; Elsenbruch et al., 2013; & Read et al., 2010), while Huismann et al. (2012) recruited 93 age-matched comparison participants from the local university. These student participants were recruited from the commuter student cohort, to control for participants with MD living at home. However the disproportionately large comparison group used in this study seems superfluous unless to increasing statistical power while studying a rare health condition. The final two studies (Jacobs et al., 2017 and Hunt et al., 2016) used within group analysis.

Key Findings

Relationships and communication

Four of the nine papers reported findings related to the importance of intimate connections with family and friends, highlighting it as a pertinent issue for this

population's sense of belonging and security. Several participant groups discussed the security and comfort that came with having a trusted group of longstanding friends and/or family that already knew and understood their physical support needs, and could be relied upon to help both physically and emotionally day-to-day (Aho et al., 2018; 2015; Hunt et al., 2016; Miladi et al., 1999).

However, many of the young people interviewed by Hunt et al. (2016) and Aho et al. (2015) struggled to communicate their needs even to close family, finding it mentally strenuous and requiring patience that, at times, was beyond their psychological resources. Hunt et al. (2016)'s participants for example, explained their resistance to external support (people, technology, medication) for pain management. Rather than disclose their discomfort, the young men would "battle through it" or distract themselves with reading, TV, or conversation as an independent means of taking action. Their decision to manage pain this way highlights the conflict that people can sometimes experience between elements of wellbeing, for example positive relationships and personal autonomy. The emotional burden of pain was also managed by shouting or swearing at loved ones, or socially withdrawing, however these coping mechanisms could magnify distress and isolate them from their support network. Young people's reluctance to share their experience may have contributed to the discrepant findings of Hunt et al. (2016), where self-reported daily pain had no association with the young person's QoL, but there was an observed negative correlation between parent-reported pain and QoL.

Another noted barrier to meaningful relationships was technology, and while some modern developments, like Skype, could increase social connectivity between young people (Aho et al., 2015), other assistive devices, like using a wheelchair, were seen as a barrier to forming new relationships (Aho et al., 2018). This appears to be a significant social barrier, and it is unfortunate that the other reviewed studies did not explore its contribution to wellbeing.

Autonomy

Similar levels of personal autonomy were expressed by MD and healthy comparison participants in the Huismann et al. (2012) study when assessing responses to the Adolescent Autonomy Questionnaire (Noom et al. 2001). However, MD participants'

sense of freedom, optimism, and self-responsibility as measured by the Inventory of the Dimensions of Emerging Adulthood (IDEA; Reifman et al. 2007) was reduced. Participants with MD also identified autonomy of decision-making and goal setting as playing a larger role in perceived QoL than the comparison group, likely because this element of independence was taken for granted by healthy controls. In-group correlation analysis found that disease impact was significantly associated with MD participants' level of uncertainty (positive correlation) and autonomy (negative correlation).

Two cohorts of participants with LGMD described beginning to use a wheelchair as mentally challenging but ultimately led to more freedom, and the ability to engage in additional activities independently (Aho et al., 2018; 2015). Many of the young adults had previously participated in various sports and activities, but as their capabilities deteriorated it became necessary to find new ways of expressing themselves (Aho et al., 2015). Assistive ventilation technology was also celebrated for its beneficial impact on wellbeing (Read et al., 2010), with participants adjusting to its use relatively quickly, despite its negative impact on sleep. Read et al. (2010) suggested that participants' perception of the ventilation system was driven by its contribution to physical health and vitality, which outweighed any impact on sleep.

Uncertainty and acceptance

Five of the nine papers documented how individuals fought against their condition in terms of identity and role, and the gradual acceptance of themselves by integrating their MD diagnosis into their personal narrative. Moreover, self-acceptance of what cannot be changed appeared to play a central role in the maintenance or resolution of distress (Aho et al., 2018; Hunt et al., 2016; Aho et al., 2015; Huismann et al., 2012; Miladi et al., 1999), another key factor for wellbeing as represented in the six-factor model.

In Aho et al.'s (2015) study, participants described leaving work was emotionally difficult, but finding hope through new goals defined by personal identity and a future despite the illness. Meaningful activities that energized and fulfilled people were highlighted as a priority, like socialising, becoming politically active, or pursing further education. Participants stressed that these activities were important when mood and

energy levels were low. They also discussed the continuous need to learn and adjust to their body's level of ability, of which living in the present moment helped, particularly when anxious thoughts of the future arose. It was acknowledged that this skill took time to develop, but was helpful in managing mood and adaptive functioning. Participants in Aho et al. (2018) further described their illness as bringing them a new perspective on life, teaching them to take risks and develop resiliency. Many initially resisted the support of assistive technology due to embarrassment, with wheelchairs, for example, only being used where they would not be recognized. However, as participants matured and their symptoms progressed, many became more open about their condition and emphasised the importance of not being ashamed. In the Elsenbruch et al. (2013) sample, younger adolescents found managing uncertainty particularly challenging, while older participants reported having developed skills in emotion regulation and cognitive flexibility to cope with declining physical health.

Low mood

Several of the studies identified depression as a risk factor for young adults living with MD, spanning various forms of MD and age groups (Conway et al., 2015; Elsenbruch et al., 2013; Huismann et al., 2012). For example, Conway et al. (2015), analysed data from the American dataset MD STARnet, and found that depressed mood was elevated compared to average US estimates. Merikangas et al. (2010) reported mood disorders in 14.3% of US adolescents between 13-18 years, compared to 19% found in Conway et al. (2015). The study also emphasied a potential underestimation of depressed mood in those with dystrophinopathies (a spectrum of X-linked MD conditions including Duchenne's, Becker's, and Duchenne-associated dilated cardiomyopathy) in studies not including older adolescent participants who are more likely to openly seek support. One explanation for these elevated levels is the impact of uncertainty on perceived environmental mastery and self-acceptance. For example, some participants described low mood and suppression of disease-related thoughts that they did not understand or want to process (Aho et al., 2018). Huismann et al. (2012) reported that participants' level of uncertainty around disease progression lead to increased feelings of negativity or depression.

Miladi et al. (1999) found that young people were particularly influenced by familial expressed emotion, for example, feeling guilt for not achieving what they believed their parent's expected of them. These young people were more likely to have a poor self-image, compared to a matched sample recruited from an out-patient mental health service. It was concluded that within this context, young people living with LGMD are more likely to rely on the opinions of close family, potentially due to a higher reliance on family support compared to the control group.

Other mental health difficulties

Elevated levels of psychosis were reported by Jacobs et al. (2017), where 19% qualified for a psychotic disorder compared to the average lifetime prevalence of 3% (Perala, 2007). Participants experiencing psychotic symptoms also reported significantly worse mean Global Assessment of Functioning (GAF) and social satisfaction scores, compared to MD participants not experiencing psychosis. This highlights the importance of positive relationships for good psychological wellbeing.

Two studies identified difficulties with anxiety (Aho et al., 2015; & Miladi et al., 1999). However, a cohort of young men with DMD reported only mildly lower vitality and mental health scores versus the general population (14% and 11%, respectively). Moreover, these participants did not show increased risk for anxiety or depression, and even depleted sleep quality did not negatively impact self-reported wellbeing (Read et al., 2010).

Critical Appraisal

Strengths

Five of the nine papers reviewed satisfied at least 70% of the CASP criteria (Aho et al., 2018; 2015; Conway et al., 2015; Elsenbruch et al., 2013; & Huismann et al., 2012), indicating that although research into the psychological wellbeing in young adults with MD is limited, the emergent studies are relatively robust and contribute to the evidence-base. There was a variety of aims, participant groups, and designs within the selected studies, but within the quantitative studies there was consideration for using appropriate measures with a normative dataset (Elsenbruch et al., 2013; Hunt et al., 2016; Read et al., 2010), or a large matched comparison

group (Huismann et al., 2012) was recruited. Additionally, Elsenbruch et al. (2013) study, which achieved 83% of the CASP criteria, adjusted self-report responses for age, and reduced the risk of demand characteristics by requesting that parents not voice opinions during data collection.

Conway et al. (2015), by accessing national medical records, were able to analyse data from a larger number of participants than would otherwise be achievable with such a rare condition. Moreover, the objective distance of researchers from study participants contributed to this study achieving 70% of the CASP scoring criteria. The quantitative aspects of the Hunt et al. (2016) study was a strength of the pilot; it was thoroughly described, and gave a clear rationale for the use of standard measures of pain that can be easily understood by medical professionals. Their recommendation to co-produce health management plans appear justified, with Huismann et al. (2012) similarly reporting that autonomy of decision-making plays a large role in perceived QoL for those living with early-onset MDs.

The use of mixed-method designs can aid new areas of research, with generalizable quantitative findings and richer qualitative data. Jacobs et al. (2017) for example, was internally reliable, satisfying 79% of the CASP criteria and documented a well-controlled quantitative design with complementary clinical interviews checked by a study-blinded clinician.

Nevertheless, the reviewed qualitative studies included the widest breadth of findings, as participant responses were not restricted to the pre-selected measures. Miladi et al. (1999) explored the broadest range of emotions, and the use of one subjective measure for all participants provided easier comparison between participants. However the appropriateness of one measure across the age range is questionable and will be reflected on later. The salutogenic approach of the Aho et al. qualitative studies appear to have gathered more empowering voices from their sample of emerging adults with LGMD, as the approach aims to acknowledge the full life experience despite adversity. It may be that coming from a salutogenic rather than an illness perspective is necessary in order to explore all elements of wellbeing, and enable participants to speak more openly. The 2015 and 2018 studies achieved CASP ratings of 85% and 75% respectively, and the earlier study particularly outlined

a clear rationale for the approach and controlled for the interviewer's personal biases.

Limitations

All the included studies used a cross-sectional design, meaning that causation cannot be determined and findings are limited to a contextual snapshot. This is important to highlight, given participants' reflections on how their wellbeing and mental adjustment to having a degenerative condition changed with maturity (Aho et al., 2018; 2015).

This review included two pilot projects (Hunt et al., 2016 & Read et al., 2010), however they satisfied 65% or less of the CASP criteria. There was limited detail regarding the qualitative methodology used within the Hunt et al. (2016) trial, which restricts others ability to critically evaluate or replicate the study. There was also no distinction made between older and younger adolescents within the sample, so one cannot determine whether age impacted on the employed pain management strategy. Read et al. (2010) acknowledged that their findings were exploratory, with a small sample size covering several MD conditions, as well as less sophisticated statistical analyses. Moreover, the qualitative arm of this study recruited only three families, again including limited details of the method, and did not consider the impact of the interviewer. Therefore, while the findings of Read et al. (2010) are encouraging in how assistive breathing apparatus can positively impact the wellbeing of young adults and their families, replication with a larger sample is needed. This is particularly necessary as Read et al. (2010) somewhat contradicts other findings that living with MD can increase risks of developing mental health difficulties during young adulthood.

The brevity of the Miladi et al. (1999) paper limited the review's ability to assess it thoroughly, resulting in it only satisfying 45% of the CASP criteria. While this study offered the broadest range of explored psychological difficulties, the validity of the primary measure was questionable. The Roberts Apperception Test for Children requires participants to create stories from example picture cards, with each story then being scored for the frequency of indicators of, for example, anxiety, aggression, and self-esteem. Whilst the measure allows an accessible way for

children to communicate emotions, its validity with a sample aged 10-22 years is less justified. A single measure allows for uniformity across participants, but a comparable, adult scale for older participants may have resulted in more reliable findings. The study's conclusion that young people living with LGMD have a lesser sense of personal autonomy and wellbeing should therefore be acknowledged with caution. Furthermore, there was no clear rationale for the recruitment of a psychiatric comparison group rather than a healthy age-matched sample. It may be that those accessing mental health services are already seeking support for distressing emotions like guilt, and therefore reported an artificially low baseline for comparison.

Appropriate comparison group recruitment was also a concern within Huismann et al. (2012). While a sample of 93 participants added to the power of the study, it is questionable why such a significant number were used in comparison to 24 participants living with CM/CMDs. Continuing to recruit past the necessary 30 participants to ensure a normal distribution may have skewed mean responses, and a clearer rationale for this disparity would have been informative. Moreover, while p-values and effect sizes were stated, the paper did not report on the variance around the mean, reducing the reader's ability to determine the internal reliability. Increased variance in one group compared to the other may have been impacted by difference in sample size for example. Other studies compensated for the rarity of MD by broadening the inclusion criteria to several forms of MD (Conway et al., 2015; Huismann et al., 2012; & Read et al., 2010). However, as each condition has its own challenges and prognosis this can make generalized findings unrepresentative of any particular population.

There were also some issues that derived from specific methodological designs. The post-hoc abstraction conducted by Conway et al. (2015), for example, is as likely to show the variation in record keeping between medical practices, as significant mental health needs within the MD community. Additionally, the number of participants reporting psychological distress is likely to be a conservative estimate, as it is based on those who accessed professional support. While qualitative studies offer richer data, their format can also undermine the reliability of findings if not controlled for. For example, the results of Jacobs et al. (2017) are relatively robust, however clinical interviews and psychosis disclosures were made while parents were present. This may have altered the researcher-participant dynamic and reduced disclosures. The

age of participants (16-25 years) also calls into question why parental presence was necessary, however the progressive breathing difficulties experienced by this population could have made interview participation challenging, and carers may have provided practical support. As with all the studies reviewed here, the interview data of Aho et al. (2018; 2015) relied on participant recall of past experiences, but there is an additional concern that the salutogenic approach could have led participants into a positive narrative of their health journey. Similarly, Read et al. (2010) considered that participants' positive view of the assistive ventilation could be driven by its contribution to physical health and vitality, in comparison to the prior symptoms of degeneration that participants' had experienced. These optimistic self-reports gathered during semi-structured interviews must be viewed with caution, in light of findings by Jacobs et al. (2017) where emerging adults and their parents overestimated young people's functional abilities. The function behind young adults and their parents over-estimating health could be as a protective wellbeing strategy, or a demand characteristic imposed by the study, either of which could be controlled for with clearer participant instructions or bracketing of researcher bias.

Table 2: Literature review summary table

Authors, publication year, and country	Population	Aims	Design	Key Findings	Strengths	Limitations
Aho, A. C., Hultsjo, S., & Hjelm, K. 2015 Sweden	14 participants with Recessive Limb Girdle Muscular Dystrophy (LGMD2), 8 female. Aged 18-30 years.	To describe young people's experience of living with LGMD2.	Qualitative. Semi-structured interviews based on the Sense of Coherence (SoC) questionnaire (Antonovsky, 1987). Salutogenic content analysis. Themes based on the SoC questionnaire.	Themes generated: Comprehensability; Managability; Meaningfulness.	Outlined a clear rationale for the approach and bracketed the interviewer's personal experiences.	No consideration of author's personal bias.
Aho, A. C., Hultsjo, S., & Hjelm, K. 2018 Sweden	14 participants with LGMD2, 8 female. Aged 18-30 years. 19 parents, 13 female.	To describe young people's experience of transitioning from diagnosis to requiring physical support for activities of daily living.	Qualitative. Semi-structured interviews based on the SoC questionnaire (Antonovsky, 1987). Salutogenic phenomenographic analysis conducted by two coders to check validity of emerging themes. Themes based on the SoC questionnaire.	Themes generated: Difficult time around diagnosis; Time before using a wheelchair; New ways of living; Concerns about disease progression; Factors facilitating everyday life.	Empowering approach with a population that can be marginalized in terms of their disability and physical health status.	Retrospective, risking recall bias. Results were part of a larger interview, so removed from its broader context. Limited consideration of interviewer bias.
Conway, K. C. et al. 2015	857 males from 765 families. Aged 1-29 years (final abstraction time	To capture the frequencies of neurobehavioral concerns among males with	Quantitative. Retrospectively taken from the MD STARnet database.	Depressed mood was elevated (19% of sample), compared to typical population	Large population study. Less chance of researcher bias.	Only disclosed mental health difficulties recorded.

USA	point at 17 years).	childhood-onset dystrophinopathies.	Search terms incl: ADHD (attention deficit hyperactivity disorder), behavior problems, or depressed mood, as well as medication and counseling. Relevant data abstracted by medically trained analysts, and summarized using descriptive and frequency statistics.	estimates. The hazard ratio (HR) for depressed mood in corticosteroid users was 3.5 times higher for those using a mobility device.	Highlights potential underestimation of depression in this population.	Data vulnerable to differences in record keeping between health practices.
Elsenbruch, S. et al. 2013 Germany	50 males with Duchenne's Muscular Dystrophy (DMD). Aged 8-12 years (n=15); 13-16 years (n=11); 17-23 years (n=24). Normative, age-matched comparison data provided by authors for the standardized measures.	Discover the impact of DMD on child, adolescent, and adult health-related quality of life (HR QoL) and depression.	Quantitative. HR QoL measured using the DISABKIDS self-report questionnaire for adolescents, and the German version of the SF-36 for young adults. Depressions-Inventar für Kinder und Jugendliche (DIKJ) measured mood in adolescents while the German version of the Beck Depression Inventory (BDI) was used for those 17+. The Vignos Scale measured mobility. Split analysis into children (8-12 years), adolescents (13-16 years), and young adults (17-23 years). Correlations between Vignos Scale and QoL and depression scores. One-way t-tests assessed HR QoL in DMD participants against normative data.	Children with DMD self-reported substantial reductions in virtually all aspects of self-reported HRQoL, while older groups did not significantly differ from normative samples.	Valid self-report measures that were adjusted for age. Researchers took steps to ensure that respondents were not influenced by their parents' suggestions. Appropriate statistical analysis used throughout.	Small sample per age group. Did not assess anxiety, social relationships etc so other psychological impacts may have been missed.
Huismann, D. J. et al. 2012 USA	24 participants with congenital myopathies and congenital muscular dystrophies	To investigate whether young people living with CM/CMDs experience less markers of	Quantitative. QoL: Adolescent Version (Raphael et al. 1996). Individualized Neuromuscular QoL Measures (INQoL: Vincent et al. 2007). Inventory of the Dimensions of Emerging	Group differences in Identity Exploration and Self-Focused subscales of the IDEA questionnaire. No overall group	Appropriate standardized measures and statistical analysis. Large matched	Measured autonomy through self-report, which may not reflect objective group

	(CM/CMDs) and other early-onset neuromuscular conditions, 15 female. Aged 18-29 years. 93 participants in the age-matched comparison group, 55 female.	emerging adulthood. And whether there are specific relationships between health status and autonomy, emerging adulthood, and QoL.	Adulthood (IDEA: Reifman et al. 2007). Adolescent Autonomy Questionnaire (Noom et al. 2001). Also measured Family Loyalty Autonomy with a subscale from the Worthington Autonomy Scale (WAS: Anderson et al. 1994). ANCOVA and MANCOVA analysis between groups. Correlations identified associations between other measures with General and HR QoL, using age as a covariate.	differences in autonomy. Within group correlations - Greater impact of disease negatively correlated with QoL. Greater disease impact correlated with more uncertainty and lower attitudinal, functional, and familial autonomy.	control group.	differences. Strong p-values reported but without reporting confidence intervals or error rates.
Hunt, A. et al. 2016 UK	12 males with DMD. Aged 11-21 years. Also recruited a parent / guardian for each young male participant.	To assess the validity of a mixed methods approach to investigating pain coping. To provide pilot data for a larger study.	Mixed methods. Pain body maps, Colour Analogue Scale with numerical rating scale (CAS) and the Faces Pain Scale (FPS_R: Hicks et al., 2001). The Paediatric Pain Coping Inventory child and parent versions (PPCI: Varni et al., 1996). The Youth QoL Scale (YQoL: Edwards et al., 2002). The Muscular Dystrophy Functional Rating Scale (MDFRS: Lue et al., 2006). Parent pain perception questionnaire. Semi-structured interviews with DMD participants explored pain coping. Themes generated inductively by two researchers, but qualitative approach was not reported. Correlations between pain, coping, QoL and function. Mann Whitney U tests for YQoL in those reporting greater and lesser pain.	Overall self-reported YQoL was good, however poorer YQoL was found in young men with parent-reported daily pain of moderate or worse severity. Qualitative themes generated: Holding it in and letting it go; Acting to relieve pain.	Rationale for use of mixed methods design. Valid self-report measures and appropriate statistical analysis.	Little detail on recruitment of participants. Unsophisticated method of replacing missing quantitative data. Limited detail of the qualitative approach used.
Jacobs, D. et	27 participants with juvenile	To investigate rates of psychotic	Mixed methods.	Five participants met	Unique initial study	Parents'

al. 2017 Belgium	Myotonic Dystrophy type-1 (DM-1), 14 female. 16-25 years.	symptoms in young people living with DM-1.	Psychiatric interviews used the Operational Criteria Checklist for psychotic and affective disorders (OPCRIT: Craddock et al., 1996) and the General Assessment of Functioning (GAF) checklist. Achenbach System of Empirically Based Assessment (ASEBA: Achenbach and Rescorla, 2001 & 2003). Interviews were verified by two clinicians and subsequent diagnoses checked against Diagnostic and Statistical Manual (DSM-IV). Reported means and percentages.	criteria for a Delusional Disorder and five for a Psychotic Disorder not otherwise specified (all aged 19+ years). Of these ten, two participants described their social life as satisfactory, in comparison to 12 of the 17 participants who did not qualify for a psychotic disorder.	into novel phenomenon. Well controlled quantitative design with complementary clinical interviews.	presence during interviews may have impacted results. Cross-sectional design does not allow for causation. No control group.
Miladi, N., Bourguignon, J., & Hentati, F. 1999 Tunisia	16 participants with Severe Childhood Autosomal Recessive Muscular Dystrophy (SCARMD) or LGMD, 9 female. 10-22 years. Comparison group recruited from a local psychiatric out-patient clinic, matched for age and gender.	To define the cognitive and psychological profile of Tunisian young people living with SCARMD or LGMD.	Qualitative. Measure of relevance – The Roberts Apperception Test for Children. Each 'story' was scored separately on the frequency of indicators for: depression, anxiety, aggression, culpability, perceived rejection, isolation, and self-esteem. Raw scores assessed against outpatient comparison group.	Young people with SCARMD / LGMD showed higher rates of sadness, guilt, and anxiety, self-image was also poorer, and they were more likely to internalize distress. Family was a supportive factor, but also a source of internalized guilt for SCARM / LGMD participants.	Creative method of gathering self-report data, and one uniform measure for all participants. Wide range of emotional impacts explored.	Short report with limited detail. Wide age range, and questionable whether one measure was appropriate for full age spectrum. No rationale for the choice of comparison group.
Read, J. et al.	10 males: DMD (n=6), Congenital MD (1 with Ullrich,	To investigate the quality of sleep and	Mixed methods. Self-reported sleep quality using the Pittsburgh Sleep Quality Index (PSQI: Buysse et al.,	PSQI sleep quality was poor, however sleep satisfaction was rated	Valid self-report measures to assess sleep	Large age range within small sample.

2010 UK	and 1 with merosin positive), Spinal Muscular Atrophy II (n=1) and Emery Dreifuss MD (n=1). Aged 10-25 years. 10 primary carers (8 mothers, 2 fathers). Aged 48-56 years. Normative, age-matched data sets used for standardized measures.	wellbeing in young people living with MD and using assistive ventilation.	1989). The SF-36 and the Hospital Anxiety and Depression Scale (HADS) assessed general well-being in patients and carers. Semi-structured interviews with three families to explore their experience of ventilation treatment. Qualitative approach not reported. Data analysis based on percentages and non-parametric correlation analysis to examine associations between sleep quality and illness, well-being, and family variables. Interview themes were generated into a coding framework.	highly by participants. No other results differed from comparison norms. Interviewees reported feeling physically better for ventilation treatment.	quality and wellbeing with large normative comparison samples.	Unsophisticated quantitative analysis, but acknowledged as a limitation. Risk of recall bias. Only three families included in interviews, with limited details of the qualitative method.

Discussion and Future Directions

This review aimed to evaluate the current state of psychological wellbeing for adolescents and emerging adults living with MD. The systematic literature search produced nine relevant papers, and it is apparent that the field of study is still in its infancy. The variants of this rare neuromuscular, degenerative disease increase the difficulty of researching the psychological needs of young adults living with MD, as there may be specific challenges that come with each condition. However, the current review found several commonalities reported by young people living with various forms of MD, as well as between the developmental stages classed as adolescence (WHO) and emerging adulthood (Arnett, 2000).

Together these studies suggest that young adults living with MD do experience more difficulties with autonomy and cultivating adult relationships. These are important developmental processes for adult identity formation (Erickson, 1982) and more efforts should be made to facilitate this. This could be done through co-produced care planning, or connecting younger adolescents with older peers with MD who could act as mentors, for example. One study of young men with DMD reported viewing autonomy over their physical experience as more important than relieving pain (Hunt et al., 2016), and encouraging a more collaborative approach to pain management, like choices of equipment, positioning, and types and routes of medication could be an effective way to increase wellbeing. Positive relationships were an important factor influencing levels of wellbeing across age and MD condition cohorts (Aho et al., 2018; Hunt et al., 2016; Aho et al., 2015; Elsenbruch et al., 2013; Miladi et al., 1999). The security provided by people that already understood the young person's support needs were universally appreciated, although it was accepted that communicating fluctuations in health, for example, pain, was still necessary and could be a point of tension within close relationships (Hunt et al., 2016). Further exploration with young people living with MD and their families could elicit preferred pain management strategies and provide person-centred ideas for professional input including psychology, physiotherapy, occupational therapy, and social care services. Younger adolescent participants in particular reported repressing negative emotions, socially withdrawing rather than communicating need, and channelling intolerable distress through challenging or aggressive behaviour (Hunt et al., 2016; Aho et al., 2015; Elsenbruch et al., 2013). Studies also found that young people with MD are more vulnerable where close, supportive relationships are

absent (Elsenbruch et al., 2013; Miladi et al, 1999). In order to optimise psychological wellbeing, especially in younger adolescents, professionals could operate systemically, including the individual's extended network to facilitate open and mutually agreeable working relationships. Recommending psychosocial or family support rather than psychiatric intervention in the first instance would also be in line with National Institute for Health and Care Excellence (NICE, 2005) guidelines, when working with mental health difficulties in younger people.

Self-acceptance was another important factor impacting wellbeing, which participants described as developing over time and maturity. The qualitative work of Aho et al. (2018; 2015) document a process of initial resistance to declining physical health, gradually leading to acceptance as service users enter their emerging adult years. This maps onto the psychosocial developmental stages described by Erikson (1982), as people age through identity exploration in adolescence towards more concrete role formation by the end of the emergent adult years. Therefore, this difference in self-acceptance between younger adolescent and emerging adult cohorts with MD is unsurprising, and arguably should not be medicalized as a problem necessitating intervention as it is a natural part of the child-adult transition. However, relational tensions with professionals or carers might be alleviated by an understanding that younger adolescents are likely to experience this increased level of emotional resistance to their diagnosis (Aho et al., 2018; 2015; Huismann et al., 2012), which they will struggle to communicate, and instead use a combination of repression, diverted attention, and aggression in order to cope (Aho et al., 2018; Hunt et al., 2016). This would be particularly beneficially given the evidence that relational difficulties have a significant negative impact on psychological wellbeing (Aho et al., 2018; Hunt et al., 2016; Aho et al. 2015; Miladi et al., 1999).

The majority of studies reviewed highlighted mental health concerns in adolescents and emerging adults living with MD, outlining slightly elevated levels of depression, anxiety, emotional suppression, and psychosis, compared to matched controls or population averages. Read et al. (2010) found no increased risk of depression or anxiety in young people with various forms of MD; however several studies reported that particularly younger adolescents do not communicate emotional distress (Hunt et al., 2016; Miladi et al., 1999) and the anomalous finding of Read et al. (2010)

could have been due to reduced self-disclosure. Future research and medical professionals working with young people living with MD should remain vigilant to the possibility of undisclosed mental distress, and create safe environments that allow young people to communicate any additional needs they may have. Low-level preventative strategies like psycho-education, or mental health friendly environments within physical health settings, for example displaying leaflets on common mental health difficulties in chronic conditions, might help facilitate this process. The reviewed studies would suggest targeting younger adolescent service users for this kind of primary intervention, to limit emotional suffering and accelerate the process of self-acceptance that many older emerging adults with MD develop (Aho et al., 2018).

Limitations

While the literature search criteria and evaluation process was conducted systematically, there is always the risk of relevant papers failing to be revealed. Therefore the search strategy has been clearly documented in the hope that others can replicate and verify its effectiveness. Likewise the choice of CASP tools and current appraisal included a subjective element, which readers should take into consideration.

Conclusions

The current review aimed to synthesize and evaluate the available literature regarding the psychological profile of young people transitioning from adolescence to emerging adulthood while living with MD. This could then inform future research questions, and make recommendations for those supporting the psychological wellbeing of people living with MD during these transition years. Most studies to date have been small, and cover a range of MD variants, cultural contexts, and outcome measures. There is also a wide spectrum of research quality, with studies satisfying between 90% - 45% of the relevant CASP criteria. However, there were common findings, such as a desire for autonomy, and social-orientated values. These results would suggest that more investigation into the emotional experience of living with MD is necessary, with larger and more rigorous studies to replicate previous findings and extend understanding to different MD conditions. It would also be advisable to

include alternatives to the standard individual interview or questionnaire methodologies, to improve participation access for those who are not otherwise comfortable disclosing personal difficulty, particularly younger adolescents.

The current evidence would indicate that young people living with MD often require physical assistance to access additional services, like secondary mental health or social care. Young adults have reported their frustration at needing to request additional support from carers, and this could further reduce individual's sense of autonomy and increase tension with informal carers. However, the participants also voiced that emotional support from their close social network is highly valued, and that they are more likely to communicate openly with those they already know and trust (Aho et al., 2018). Therefore, based on the existing literature, systemically orientated support, co-production of health care planning, and preventative psycho-education for younger adolescents and healthcare professionals would appear the most suitable interventions, rather than involving additional secondary services.

References

Achenbach, T. M., & Rescorla, L. (2001). *ASEBA school-age forms & profiles: Child Behaviour Checklist for ages 6-18: teacher's report and youth self-report.* 99-135. ASEBA.

Achenbach, T. M., & Rescorla, L. (2003). *Manual for the ASEBA adult forms & profiles: for ages 18-59: adult self-report and adult behavior checklist*. ASEBA.

Aho, A. C., Hultsjö, S., & Hjelm, K. (2015). Young adults' experiences of living with recessive limb-girdle muscular dystrophy from a salutogenic orientation: An interview study. *Disability and Rehabilitation*, *37*(22), 2083-2091.

Aho, A. C., Hultsjö, S., & Hjelm, K. (2018). Perceptions of the transition from receiving the diagnosis recessive limb-girdle muscular dystrophy to becoming in need of human support and using a wheelchair: an interview study. *Disability and Rehabilitation*, 1-10.

Anderson, R. A., Worthington, L., Anderson, W. T., & Jennings, G. (1994). The development of an autonomy scale. *Contemporary family therapy*, *16*(4), 329-345.

Antonovsky, A. (1987). Health promoting factors at work: the sense of coherence. *Psychosocial factors at work and their relation to health*, 153-167.

Antonovsky, A. (1987). Unraveling The Mystery of Health - How People Manage Stress and Stay Well, San Francisco: Jossey-Bass Publishers.

Arnett, J. J. (2000). Emerging adulthood: A theory of development from the late teens through the twenties. *American Psychologist, 55*(5), 469-480.

Buysse, D. J., Reynolds, C. F., Monk, T. H., Berman, S. R., & Kupfer, D. J. (1989). The Pittsburgh Sleep Quality Index: a new instrument for psychiatric practice and research. *Psychiatry research*, *28*(2), 193-213.

Ciliska. D, Thomas. H, Buffett. C (2008) *A Compendium of Critical Appraisal Tools for Public Health Practice. National Collaborating Centre for Methods and Tools*, Canada.

Conway, K. C., Mathews, K. D., Paramsothy, P., Oleszek, J., Trout, C., Zhang, Y., & Romitti, P. A. (2015). Neurobehavioral concerns among males with dystrophinopathy using population-based surveillance data from the muscular dystrophy surveillance, tracking, and research network. *Journal of Developmental and Behavioral Pediatrics: JDBP*, *36*(6), 455.

Craddock, M., Asherson, P., Owen, M. J., Williams, J., McGuffin, P., & Farmer, A. E. (1996). Concurrent validity of the OPCRIT diagnostic system. Comparison of OPCRIT diagnoses with consensus best-estimate lifetime diagnoses. *The British Journal of Psychiatry*, *169*(1), 58-63.

Edwards, T. C., Huebner, C. E., Connell, F. A., & Patrick, D. L. (2002). Adolescent quality of life, part I: conceptual and measurement model. *Journal of adolescence*, *25*(3), 275-286.

Elsenbruch, S., Schmid, J., Lutz, S., Geers, B., & Schara, U. (2013). Self-reported quality of life and depressive symptoms in children, adolescents, and adults with Duchenne

muscular dystrophy: a cross-sectional survey study. *Neuropediatrics*, *44*(5), 257-264.

Erikson E. H . (1982). *The life cycle completed*. New York, NY: Norton.

ESRC – Retrieved on 23.06.2018 https://esrc.ukri.org/about-us/strategy-and-priorities/mental-health/

Health and Care Professions Council. (2015). *Standards of Proficiency: Practitioner psychologists.* Park House, London. http://www.hcpc-uk.org/publications/standards/index.asp?id=198

Hicks, C. L., von Baeyer, C. L., Spafford, P. A., van Korlaar, I., & Goodenough, B. (2001). The Faces Pain Scale–Revised: toward a common metric in pediatric pain measurement. *Pain*, *93*(2), 173-183.

Huismann, D. J., Sheldon, J. P., Yashar, B. M., Amburgey, K., Dowling, J. J., & Petty, E. M. (2012). Quality of life and autonomy in emerging adults with early-onset neuromuscular disorders. *Journal of Genetic Counseling*, *21*(5), 713-725.

Hunt, A., Carter, B., Abbott, J., Parker, A., & Spinty, S. (2016). Pain experience, expression and coping in boys and young men with Duchenne Muscular Dystrophy–A pilot study using mixed methods. *European Journal of Paediatric Neurology*, *20*(4), 630-638.

Jacobs, D., Willekens, D., de Die- Smulders, C., Frijns, J. P., & Steyaert, J. (2017). Delusional and psychotic disorders in juvenile myotonic dystrophy type- 1. *American Journal of Medical Genetics Part B: Neuropsychiatric Genetics*, *174*(4), 359-366.

Johnston, B., Jindal-Snape, D., & Pringle, J. (2016). Life transitions of adolescents and young adults with life-limiting conditions. *International journal of palliative nursing*, *22*(12), 608-617.

Kerr, H., Price, J., Nicholl, H., & O'Halloran, P. (2017). Transition from children's to adult services for young adults with life-limiting conditions: A realist review of the literature. *International journal of nursing studies*, *76*, 1-27.

Lue, Y. J., Su, C. Y., Yang, R. C., Su, W. L., Lu, Y. M., Lin, R. F., & Chen, S. S. (2006). Development and validation of a muscular dystrophy-specific functional rating scale. *Clinical rehabilitation*, *20*(9), 804-817.

Mangelli, L., Gribbin, N., Büchi, S., Allard, S., & Sensky, T. (2002). Psychological well-being in rheumatoid arthritis: Relationship to 'disease' variables and affective disturbance. *Psychotherapy and Psychosomatics*, *71*(2), 112-116.

McArthur, D. S., & Roberts, G. E. (1982). Roberts Apperception Test for Children manual. Los Angeles, CA Western Psychological Services.

Miladi, N., Bourguignon, J. P., & Hentati, F. (1999). Cognitive and psychological profile of a Tunisian population of limb girdle muscular dystrophy. *Neuromuscular Disorders*, *9*(5), 352-354.

Muscular Dystrophy UK – Retrieved on 23.06.2018 https://www.musculardystrophyuk.org/about-muscle-wasting-conditions/

Muscular Dystrophy UK. (January 2017). Diagnositic Tests Factsheet. Retrieved on

30.07.2018

National Institute of Neurological Disorders and Stroke (NINDS). (2011). *Muscular dystrophy: Hope through research.* Retrieved May 25, 2012, from ttps://www.ninds.nih.gov/Disorders/Patient-Caregiver-Education/Hope-Through-Research/Muscular-Dystrophy-Hope-Through-Research

NICE. (2005). Clinical Guideline: Depression in children and young people: identification and management. Published 26 September 2005. nice.org.uk/guidance/cg28

Noom, M. J., Deković, M., & Meeus, W. (2001). Conceptual analysis and measurement of adolescent autonomy. *Journal of Youth and Adolescence*, *30*(5), 577-595.

Perälä, J., Suvisaari, J., Saarni, S. I., Kuoppasalmi, K., Isometsä, E., Pirkola, S., ... & Härkänen, T. (2007). Lifetime prevalence of psychotic and bipolar I disorders in a general population. *Archives of general psychiatry*, *64*(1), 19-28.

Pusswald, G., Fleck, M., Lehrner, J., Haubenberger, D., Weber, G., & Auff, E. (2012). The "Sense of Coherence" and the coping capacity of patients with Parkinson disease. *International Psychogeriatrics*, *24*(12), 1972-1979.

Raphael, D., Rukholm, E., Brown, I., Hill-Bailey, P., & Donato, E. (1996). The quality of life profile—adolescent version: Background, description, and initial validation. *Journal of Adolescent Health*, *19*(5), 366-375.

Read, J., Simonds, A., Kinali, M., Muntoni, F., & Garralda, M. E. (2010). Sleep and well-being in young men with neuromuscular disorders receiving non-invasive ventilation and their carers. *Neuromuscular Disorders*, *20*(7), 458-463.

Reifman, A., Arnett, J. J., & Colwell, M. J. (2007). Emerging adulthood: Theory, assessment and application. *Journal of Youth Development*, *2*(1), 37-48.

Ryff, C. D. (1989). Happiness is everything, or is it? Explorations on the meaning of psychological well-being. *Journal of personality and social psychology*, *57*(6), 1069.

Ryff, C. D., & Keyes, C. L. M. (1995). The structure of psychological well-being revisited. *Journal of personality and social psychology*, *69*(4), 719.

Trailblazers Network – Retrieved on 23.06.2018 https://www.musculardystrophyuk.org/campaign-for-independent-living/trailblazers/

Varni J. W., Waldron. S. A., Gragg. R. A., Rapoff. M. A., Bernstein. B. H., Lindsley. C. B., & Newcomb. M. D. (1996). Development of the Waldron/ Varni pediatric pain coping inventory. *Pain, 67*, 141e50.

Vincent, K. A., Carr, A. J., Walburn, J., Scott, D. L., & Rose, M. R. (2007). Construction and validation of a quality of life questionnaire for neuromuscular disease (INQoL). *Neurology*, *68*(13), 1051-1057.

WHO definition of adolescence – Retrieved on 23.06.2018 http://apps.who.int/adolescent/second-decade/section2/page1/recognizing-adolescence.html

Yoder, A. E. (2000). Barriers to ego identity status formation: A contextual qualification of Marcia's identity status paradigm. *Journal of adolescence*. *23*(1), 95-106.

Appendix I: CASP Case-Control Checklist details

www.casp-uk.net
info@casp-uk.net
Summertown Pavilion, Middle Way Oxford OX2 7LG

CASP Checklist: 11 questions to help you make sense of a Case Control Study[1] How to use this appraisal tool: Three broad issues need to be considered when appraising a case control study:

Are the results of the study valid? (Section A)

What are the results? (Section B)

Will the results help locally? (Section C)

The 11 questions on the following pages are designed to help you think about these issues systematically. The first three questions are screening questions and can be answered quickly. If the answer to both is "yes", it is worth proceeding with the remaining questions. There is some degree of overlap between the questions, you are asked to record a "yes", "no" or "can't tell" to most of the questions. A number of italicised prompts are given after each question. These are designed to remind you why the question is important. Record your reasons for your answers in the spaces provided.

About: These checklists were designed to be used as educational pedagogic tools, as part of a workshop setting, therefore we do not suggest a scoring system. The core CASP checklists (randomised controlled trial & systematic review) were based on JAMA 'Users' guides to the medical literature 1994 (adapted from Guyatt GH, Sackett DL, and Cook DJ), and piloted with health care practitioners.

For each new checklist, a group of experts were assembled to develop and pilot the checklist and the workshop format with which it would be used. Over the years overall adjustments have been made to the format, but a recent survey of checklist users reiterated that the basic format continues to be useful and appropriate.

Referencing: we recommend using the Harvard style citation, i.e.: ***Critical Appraisal Skills Programme (2018). CASP (insert name of checklist i.e. Case Control Study) Checklist. [online] Available at: URL. Accessed: Date Accessed.***

Appendix II: CASP Cohort Study Checklist details

www.casp-uk.net
info@casp-uk.net
Summertown Pavilion, Middle Way Oxford OX2 7LG

CASP Checklist: 12 questions to help you make sense of a Cohort Study[1] How to use this appraisal tool: Three broad issues need to be considered when appraising a cohort study:

Are the results of the study valid? (Section A)

What are the results? (Section B)

Will the results help locally? (Section C)

The 12 questions on the following pages are designed to help you think about these issues systematically. The first two questions are screening questions and can be answered quickly. If the answer to both is "yes", it is worth proceeding with the remaining questions. There is some degree of overlap between the questions, you are asked to record a "yes", "no" or "can't tell" to most of the questions. A number of italicised prompts are given after each question. These are designed to remind you why the question is important. Record your reasons for your answers in the spaces provided.

About: These checklists were designed to be used as educational pedagogic tools, as part of a workshop setting, therefore we do not suggest a scoring system. The core CASP checklists (randomised controlled trial & systematic review) were based on JAMA 'Users' guides to the medical literature 1994 (adapted from Guyatt GH, Sackett DL, and Cook DJ), and piloted with health care practitioners.

For each new checklist, a group of experts were assembled to develop and pilot the checklist and the workshop format with which it would be used. Over the years overall adjustments have been made to the format, but a recent survey of checklist users reiterated that the basic format continues to be useful and appropriate.

Referencing: we recommend using the Harvard style citation, i.e.: ***Critical Appraisal Skills Programme (2018). CASP (insert name of checklist i.e. Cohort Study) Checklist. [online] Available at: URL. Accessed: Date Accessed.***

Appendix III: CASP Qualitative Study Checklist details

www.casp-uk.net
info@casp-uk.net
Summertown Pavilion, Middle Way Oxford OX2 7LG

CASP Checklist: 10 questions to help you make sense of a Qualitative research[1] How to use this appraisal tool: Three broad issues need to be considered when appraising a qualitative study:

Are the results of the study valid? (Section A)

What are the results? (Section B)

Will the results help locally? (Section C)

The 10 questions on the following pages are designed to help you think about these issues systematically. The first two questions are screening questions and can be answered quickly. If the answer to both is "yes", it is worth proceeding with the remaining questions. There is some degree of overlap between the questions, you are asked to record a "yes", "no" or "can't tell" to most of the questions. A number of italicised prompts are given after each question. These are designed to remind you why the question is important. Record your reasons for your answers in the spaces provided.

About: These checklists were designed to be used as educational pedagogic tools, as part of a workshop setting, therefore we do not suggest a scoring system. The core CASP checklists (randomised controlled trial & systematic review) were based on JAMA 'Users' guides to the medical literature 1994 (adapted from Guyatt GH, Sackett DL, and Cook DJ), and piloted with health care practitioners.

For each new checklist, a group of experts were assembled to develop and pilot the checklist and the workshop format with which it would be used. Over the years overall adjustments have been made to the format, but a recent survey of checklist users reiterated that the basic format continues to be useful and appropriate.

Referencing: we recommend using the Harvard style citation, i.e.: ***Critical Appraisal Skills Programme (2018). CASP (insert name of checklist i.e. Qualitative) Checklist. [online] Available at: URL. Accessed: Date Accessed.***

Appendix IV: Critical Appraisal Outcomes

Reference	Q1	Q2	Q3	Q4	Q5	Q6	Q7	Q8	Q9	Q10	Q11	Q12	Total (score)	Total (%)
Aho et al. (2015)	2	2	2	2	2	1	1	1	2	2	/	/	17 / 20	85
Aho et al. (2018)	2	2	1	2	2	0	1	1	2	2	/	/	15 / 20	75
Conway et al. (2015)	2	2	1	1	0	2	2	1	1	2	1	2	17 / 24	70
Elsenbruch et al. (2013)	2	2	2	1	2	0	2	2	1	2	2	2	20 / 24	83
Huismann et al. (2012)	2	2	2	2	2	2	2	1	1	2	2	/	20 / 22	91
Hunt et al. (2016)	2	2	1	1	1	0	2	0	2	2	/	/	13 / 20	65
Jacobs et al. (2017)	2	2	2	1	1	0	2	1	2	2	/	2	17 / 22	77
Miladi et al. (1999)	2	1	1	1	1	0	0	0	2	1	/	/	9 / 20	45
Read et al. (2010)	2	1	1	2	2	1	1	1	0	0	1	/	12 / 22	55

Appendix V: 'Disability and Society' journal guide for authors

Aims and scope

2018 Impact Factor: 1.613

Ranking: 42/98 (Social Sciences, Interdisciplinary), 40/69 (Rehabilitation)

2017 Citescore 1.43 - values from Scopus

Editor, Professor Michele Moore, *Northumbria University*

Disability & Society is an international journal providing a focus for debate about such issues as human rights, discrimination, definitions, policy and practices. It appears against a background of change in the ways in which disability is viewed and responded to.

Definitions of disability are more readily acknowledged to be relative; segregated approaches are seen as inadequate and unacceptable - placing greater emphasis on community care and inclusion. However, policy intentions may not have the desired effects on the realities of everyday practice and policy changes themselves may be merely cosmetic, or appropriate but unfounded.

The journal publishes articles that represent a wide range of perspectives including the importance of the voices of disabled people.

There is an established well-informed international audience for the journal. Authors are expected to consider this wide readership and to exhibit knowledge of previously-published articles when submitting their work for consideration.

About The Journal

Disability & Society accepts the following types of article:

- Article
- Current Issues
- Student Perspectives
- Doctoral Theses

Peer Review

Taylor & Francis is committed to peer-review integrity and upholding the highest standards of review. Once your paper has been assessed for suitability by the editor, it will then be double blind peer reviewed by independent, anonymous expert referees.

Preparing Your Paper

Articles

- Should be written with the following elements in the following order: title page; abstract; keywords; points of interest; main text introduction, research process, results, discussion; acknowledgments; declaration of interest statement; references; appendices (as appropriate); table(s) with caption(s) (on individual pages); figures; figure captions (as a list)
- Should be no more than 8,000 words (excluding references).
- Should contain an unstructured abstract of 150 words.
- Should contain Points of Interest 100 to 150 words (maximum) describing in plain English the importance of your work for lay readers in 4 or 5 bullet points.
- Have between 2 and 6 keywords.

Chapter 2: Empirical Paper

Facilitating positive adult transition in young people with Duchenne's muscular dystrophy: A Q-sort study of the priorities for service users, carers, and health professionals.

Word count: 7,992

Contents

Title: Facilitating positive adult transition in young people with Duchenne's muscular dystrophy: A Q-sort study of the priorities for service users, carers, and health professionals.

Authors and Affiliations:

Dr. Kevanne Sanger – Staffordshire University

Dr. Helen Scott – Staffordshire University

Dr. Yvette Easthope-Mowatt - Robert Jones & Agnes Hunt Orthopaedic Hospital NHS Foundation Trust

Dr. Helen Combes – Staffordshire University

Corresponding Author:

Dr. Kevanne Sanger

Present / Permanent Address:

s025080g@student.staffs.acuk

Doctorate in Clinical Psychology Programme,
Science Centre,
Leek Road,
Stoke-on-Trent,
ST4 2DF

Word count: 7,992

1. Abstract

Medical advances in the last few decades have improved the life expectancy of men with Duchene's Muscular Dystrophy (DMD); however, this poses new questions about how healthcare services can best support this population to thrive into adulthood. This study sought to explore what factors are perceived to be helpful or unhelpful in facilitating emerging adults with DMD to develop a sense of adult identity, social role, confidence, and autonomy within their community. Thirteen participants, including emerging adult service users with DMD (aged 18-29 years), a primary carer, and Muscular Dystrophy Clinic healthcare professionals, were recruited from a West Midlands Hospital Trust, using Q-methodology to gather their perspectives. Results indicated that priorities for adult transition were shared across expert populations, with two factor groups emerging; one that valued developing an adult role within their established network, and another that focused on independent exploration. Factor one included the full range of expert stakeholders, while factor two represented the voices of three healthcare professionals and one emerging adult with DMD. The mix of participants across factor one, and some variation in factor two would suggest that priorities for an adult life with DMD are not defined by whether someone is a healthcare service user, or paid healthcare employee. However, the emergence of two factors with different values highlights the importance of a personalised transition process that respects the individual's priorities for adulthood. Further research is needed to confirm and expand on these initial findings.

2. Introduction

2.1. Duchenne's Muscular Dystrophy

Duchenne's Muscular Dystrophy (DMD) is the most common form of childhood muscular dystrophy (MD), a group of genetic, degenerative muscle-wasting conditions that affects around 2,500 people in the UK [1]. DMD is an X chromosome-linked condition so while women can be carriers, DMD only develops in men [2]. It is a life-limiting condition, but medical advances in the past two decades have increased the life expectancy from late adolescence to around 30 years of age [3, 4]. However, this has inspired new challenges and questions for researchers and healthcare providers around what psychosocial, mental health, and transition support would maximise quality of life for this population.

2.2. Becoming an adult with DMD

Expert meeting groups [5, 6] have met to discuss the challenges of transitioning to adulthood faced by young men with DMD, like the shift in life expectations that young adults and their families had not planned for. These panels aimed to achieve some consensus on how to reduce the challenges and social barriers that are often experienced due to increased levels of disability, previously highlighted in the documentary "DMD with a Future – The power to live" [7]. By transition to adulthood, Schrans et al. [6] refer to a sense of autonomy, adult self-image, and adult role-taking within wider society, which men with DMD can feel inhibited from developing [7]. These meeting groups specifically highlighted the importance of facilitating greater autonomy for clients classed as 'emerging adults', a stage of life described as bridging the gap between adolescence and adulthood spanning 18-29 years of age [5, 6]. Emerging adulthood is a life stage characterised by change and the exploration of possible life directions, as well as a period when people take on more socially perceived adult roles, like increased autonomy of decision making or responsibility [8]. Developing on from these meeting groups' conclusions, specific models of good practice have advised healthcare services to promote good mental health, autonomy, and self-confidence in young men with DMD through annual assessments, psycho-education, and skills training interventions. These frameworks advise that clinical psychologists have the appropriate skill set to implement such recommendations [9]. Moreover, this literature advocates that services should provide emerging adults with support to achieve some of the classic markers of adulthood [9, 5, 6]. These include the consolidation of an adult identity,

achieving greater levels of independence (with appropriate support), having opportunities to access work, education or training, and access to the full range of adult friendships and intimate relationships. The transition to adulthood can be mentally, logistically, and relationally challenging for young men with DMD and their families [10], and despite the healthcare recommendations in place, for example the Duchenne's Care Considerations [9], the quality of transition support is diverse and often lacking evaluation [10]. This is likely due to the inevitable delay between medical advances that have increased people's lifespan, and the necessary research in how to effectively support this new adult population. Nevertheless expedient investigation to answer this question, and evaluate models of practice, are needed, if emerging adults with DMD are to be optimally supported in living a full adult life.

2.3. From the young persons' perspective

The published literature investigating the psychological impact of living as an emerging adult with DMD has mainly used qualitative interview methods [e.g. 11, 12, 13, 14, 15], only capturing the voices of either young adults themselves, or family carers. These studies highlight that emerging adults with DMD often feel let down or ignored by society; having limited access to activities, training, or work placements after they finish school. The transition to adult healthcare services has also been perceived as a reduction in support, where their reduced life-span is seen to make them a low priority [13, 16]. The majority of young men with DMD measure their 'adultness' in comparison with able-bodied peers, feeling that it would represent a negative attitude if they associated themselves with other life-limited young people or adjusted their expectations based on disability [14]. The eleven young adults interviewed by Gibson et al. [14] had the same aspirations as most young men but particularly focused on academic achievements, paid employment, and personal qualities that others would look up to, like resilience and optimism. Other developmental markers like living away from family, travelling, and forming intimate relationships were present, but deferred for the future. Together, these self-report studies suggest that young adults with DMD often feel marginalised, despite their aspirations being the same as most emerging adults. Investigating service users' views of what specific factors would maximise their 'adultness' could benefit healthcare services charged with delivering a positive transition, and more effectively direct professional frameworks.

2.4. From the carers' perspective

Abbot and colleagues [13, 10] have discussed the incidental benefits of having primary carers present for support during interview studies with young adults with DMD, which subsequently stimulated new conversations between family members. For example, parents have been surprised to discover the distress that wheelchair use can cause their child, or that their son would like more information about their condition, treatment options, and end of life provision [13, 10]. Primary care-givers have also been asked about their changing role as the young person develops into adulthood, and what factors they think are most important for a positive transition [12]. 14 interviewed parents highlighted themes relating to emotional support, and independence, like encouraging physical support to pass to the young adult themselves or paid professionals, as they get older. However, there is much contention in the literature surrounding the carers' role as gate-keeper; and parents can feel conflicted between wanting to shield their child from distress, and wanting them to have an informed choice [12, 13]. Together, these findings would suggest that young adult service users and primary care-givers have different experiences of living with DMD, and that this difference can go unspoken. As mentioned above, Abbot [10] reflected on how his research had given many families the opportunity to broach sensitive topics, improve communication, and gain a greater understanding of each other's viewpoint. This highlights the need for research that encourages new conversations, and explores the experience of DMD from multiple perspectives.

2.5. From the professionals' perspective

No studies appear to have reported on what healthcare professionals think would improve client autonomy and positive adult transition, despite expert panels acknowledgment of the influential role they play in developing this [5, 6]. Including healthcare staff in research could give valuable insight into their views of what most impacts positive adult transition. This would reveal where they currently focus their efforts in facilitating adult transition, and if there are gaps between their priorities and those of the young people they support.

2.6. Aims

Previous studies have captured some of the relevant voices in addressing how to best support adult transition for emerging adults with DMD, but the views of

healthcare professionals are yet to be researched. There is also some acknowledgement that qualitative interviews may be unsuitable for service users, given some patients' breathing difficulties, as well as their sense of powerlessness to speak out [10], suggesting that alternative research methods should be explored. This study sought to investigate the specific priorities of young adults with DMD, their primary care-givers, and healthcare professionals working within a Muscular Dystrophy clinic, for an autonomous and positive adult transition. Q-methodology has been an effective way of studying people's attitudes in health promotion and education research [17], and offers a unique opportunity to explore the range, similarities, and differences in values between these three expert stakeholder groups. It is only by highlighting areas of consensus and difference that gaps can be bridged, and shared understanding be reinforced. This could then lead to evidence-based improvements in personalised care during the transition period, and thus improve the psychological well-being and sense of self-efficacy for people affected by DMD.

3. Method

3.1. Ethics

This study was approved by the Health Research Authority (HRA) and Health and Care Research Wales (REC Ref No. 19/LO/0214). It was subject to scientific review by the Staffordshire University Faculty of Health Sciences Research Ethics Committee, and permission of access was granted by the participating hospital trust R&D department. No ethical concerns were reported during the conduct of this study.

3.2. Participants and recruitment

Primary carers and service users with DMD aged 18-29 years old (the defined age range for emerging adulthood [8]) registered with the participating West Midlands Hospital Trust MD Clinic were eligible to take part, offering a potential participant sample of 16 young person-carer dyads at the time of recruitment. Participating healthcare professionals were all current members of staff within the MD Team with at least 6-months experience, to ensure their understanding of the issue. Participants needed to be fluent English speakers, as the study was not able to provide an interpreter. An original recruitment target of eight emerging adults with DMD, eight carers, and eight healthcare professionals was set in consultation with

the clinical research supervisor. However the final sample of eight healthcare staff, four service users, and one primary carer was the result of a limited recruitment period, difficulty adjusting appointments around participant's health, and the limited availability of carer participants. Further study to recruit more emerging adults and care-givers is recommended, however the final sample population was similar to other relevant Q-studies [e.g. 18, 19]. No financial incentive was offered to participants for taking part.

The participating MD clinic operates a life-long model of care, meaning that patients do not transition to specific adult or palliative care teams once they reach adult maturity. This context will be taken into consideration when discussing the results of the study.

Emerging adults and carers were approached about the study during routine appointments at the participating MD Clinic. Named healthcare professionals offered potential participants an information pack, which included a notification of interest slip (Appendix VI) and information sheet (Appendix VII). The information clearly stated that choosing to participate in the study or not, would not impact their care support. A link to the information sheet and notification of interest slip was also posted on the MD Clinic's social media platforms, in order to maximise awareness of the study to carers and emerging adults. The principle investigator (PI) recruited healthcare staff participants, advertising the study around the MD Clinic setting (Appendix V), providing information packs, and offering information at team meetings. All notification of interest slips were returned to the PI, after-which a convenient appointment time was made on healthcare premises. Informed consent was taken prior to the sorts being completed (Appendix VIII). No participants requested to withdraw from the study.

The demographics of the final sample can be seen in Table 1. The four participants with DMD were in the middle of the emerging adult 18-29 years age bracket (mean = 22.25, range = 20-26). One primary carer was recruited into the study; this was the mother of one of the participating emerging adults with DMD. No participants had a comorbid learning disability. There was a mix of professions within the healthcare staff participants.

Table 1: Participant demographics

Variable	Category	Frequency	Percentage (%)
Participant group	Healthcare staff	8	62
	Young adults with DMD	4	31
	Primary carers	1	8
Gender	Male	5	38
	Female	8	62
Age	18-29	5	38
	40-49	6	46
	50-59	2	15
Healthcare Staff Job Role	Clinical Psychologist	1	12.5
	Specialist Doctor	2	25
	Specialist Nurse	4	50
	Physiotherapist	1	12.5

3.3. Q-methodology

Q-methodology has previously been used to gather different perspectives in health research [17], and could offer an alternative exploratory research method to individual interviews that can be difficult for some young adults with DMD and primary care-givers to access. While traditional qualitative methods can be demanding in terms of time, emotional resources, and physical effort, Q-methodology can be completed within 30-60 minutes and does not require extended conversation, which could be challenging for young adults with DMD using assistive breathing technology.

As a method of scientific study, Q-method was first designed by a British physicist and psychologist, William Stephenson in 1935 [20]. The method aims to cross the divide between the qualitative richness of individual experience, and the quantitative focus on specific construction variables. Brown [21] describes the value of Q-method in comparison to standard quantitative approaches as:

"In the case of R methodology something is done to the person, as when we take

blood pressure or measure height: this is the objective mode and the person's stance relative to measurement is passive. In the case of Q the person actively does something, i.e. measures or scales a population of measurable material: this is the subjective mode insofar as measurement is from the person's standpoint."

In a Q-method study participants are required to rank order a set of statements relevant to a specific question, placing statements in a semi-normal distribution (see Figure 1). The number of participants is not as important as the diversity of opinions and number of Q-sample statements; the method simply requires there to be enough respondents to identify significant differences of opinion for the factor loadings [21]. Factor analysis is then applied to participants' finished Q-sorts (ranked statements), resulting in similarly ranked Q-sorts being grouped in a factor together, which illustrates consensus within and differences between participant groupings. Participants are asked to reflect upon their ranked Q-sort, to provide a richer context to data analysis. This qualitative aspect of the method provides a sample of participant quotations that can describe findings more directly than the researcher's second-hand interpretation [22].

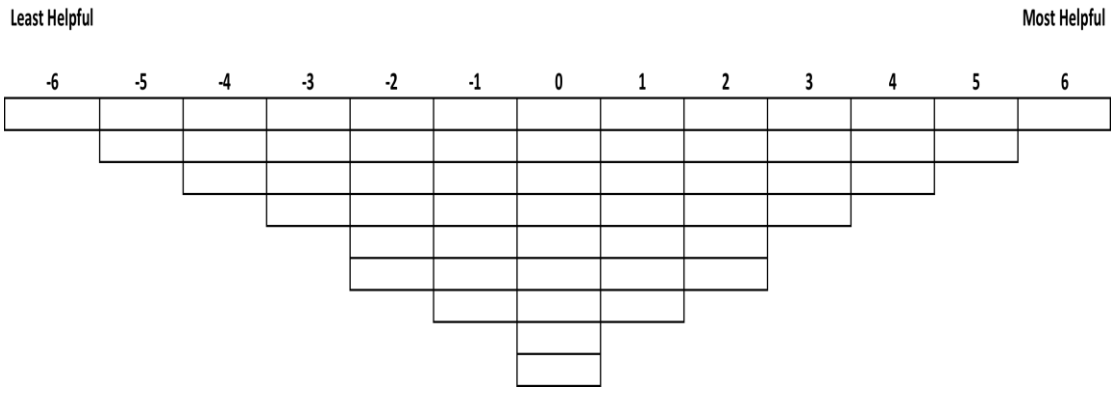


Figure 1: Distribution Q-grid

3.4. Design and Materials

This was a cross-sectional study that recruited participants to complete one Q-sort each, between April – June 2019.

The PI generated a concourse of statements (Appendix X) for participants to rank, using inductive content analysis. This involved drawing upon the previous literature on traditional adult development as well as the literature on development in young people with a disability or physical health condition. In addition, themes discussed in various media sources, including online forums from the MDUK Youth network 'Trailblazers' blog page [23] and CarersUK [24], and news articles involving the three participant groups were used. This captured a wide range of possible themes that related to the facilitation of positive adult transition for emerging adults with DMD. Themes were then scanned for uniqueness, similarity, or duplication, resulting in a final concourse of 55 statements. Two clinical psychologists supervising the project (one who works with young people with MD, and the other a residential expert in Q-methodology at Staffordshire University) verified these statements, as well as two employees with lived experience of MD who work at a neuromuscular charity organisation. The question participants were asked to consider, within the context of growing up with DMD, was *"What helps you most to develop into an adult in society?"* and participants rank ordered the statements from most – least helpful (see Figure 1). Based on past literature 40-60 concourse statements was sufficient for a Q-method investigation [e.g. 25, 26, 27]. After completing the Q-sort, participants were asked to give a description of their rankings and anonymised quotations were used to conceptualise the final factor groupings.

3.5. Procedure

Setting

The sorts were completed in a quiet room on healthcare premises, with all participants recruited through a West Midlands NHS hospital trust. The lone working policies set by the PI's employing NHS Trust and the participating West Midlands Trust were adhered to, with MD Clinic staff always aware of study appointments and present nearby if support was needed.

Data collection

During the study appointment, the PI described the procedure and provided time for questions before taking informed consent. Each participant completed their

individual Q-sort, ranking the concourse of 55 statements (Appendix X) in order of most – least helpful in terms of "*what helps you most to develop into an adult in society?"* The young adult participants were able to have carers present if they wished (two young adults requested that a carer be present), but the PI asked that the ranking itself was solely the views of the participant. The PI was also available to support participants in physically completing the Q-sort, by moving statements around the grid matrix or reading statements aloud if required. Participation in the study took around one hour. After participants had completed the Q-sort they were asked if they would like to describe their final rankings or any particular reflections they had upon completing the Q-sort. These reflections were dictated by the participant and written down by the researcher. Participants were then given a debrief form (Appendix IX) and another opportunity to ask any questions before leaving the study appointment.

3.6. Data Analysis

Data management

Photographs were taken of each participant's completed Q-sort, with ID number and date logged. Qualitative data was labelled in the same way. Electronic data, including photographs, records of demographic information, and linking sheet connecting participants' name and ID number, were stored on a secure desktop computer at the participating MD clinic, and any paperwork, namely consent forms and qualitative descriptions, were kept in a securely locked cabinet at the hospital base. Any identifying information was stored separately from research data, and electronic versions were password protected.

Data Analysis

The data was analysed using a standard Q-method analysis programme, called Ken-Q Analysis software (version 1.04 which is freely available at https://shawnbanasick.github.io/ken-q-analysis/). This first identified the co-correlations within the data, ran the factor analysis, and divided participants into groups with similarly ranked Q-sorts. The PI, utilising the qualitative feedback provided by participants for context, then evaluated the Ken-Q output file (Appendix XI) and composite factor arrays of each group (Appendix XII). Factors were labelled based on the different statements that participants in that factor group valued as

helpful or not to supporting a positive adult transition. The demographics of each factor grouping were checked, to see if results had separated participants out into service user, carer, and healthcare professionals, or whether consensus and differences in viewpoint spanned across these expert groups.

4. Results

4.1. Correlations

Pairwise correlations were examined to determine the strength of relationship between participants' Q-sorts, shown in Table 2. There was a variable amount of agreement between variables, with each Q-sort significantly correlating with between three and eleven others.

Table 2: Correlation Matrix

Participant	001	002	003	004	005	006	007	008	009	010	011	012	013
001	100	34	20	40	10	24	53	50	18	45	32	59	40
002		100	27	23	20	11	40	38	5	40	37	38	32
003			100	27	30	24	27	30	-2	15	19	20	27
004				100	38	33	18	41	19	21	19	37	34
005					100	26	16	34	28	13	30	33	44
006						100	26	21	17	33	-1	16	17
007							100	31	26	25	32	30	23
008								100	-9	42	35	47	40
009									100	-8	2	9	26
010										100	15	48	35
011											100	38	41
012												100	60
013													100

Significant correlations are highlighted in grey, calculated as ≥ 26 using the Brown (1980) formula at significance level p < .05: 1.96 x (1/ √ No. of Q-set statements). Strong correlations are underlined (r= ≥ 50, [28]).

4.2. Factor Analysis

Factor analysis is applied to data to account for common variance. The approach assumes a finite amount of difference in participant responding, so it identifies patterns of similar responses, and groups these together into specific factors [29]. It is described as a data reduction technique, with fewer resultant factors than raw Q-sorts. Centroid factor analysis was used, allowing for a deeper examination of the dataset before determining the number of factors to extract [22]. Brown [21] recommends setting seven potential factors as a default, which is shown in Table 3 with respective eigenvalues (squared factor loading of all Q-sorts in the factor), and the amount of common variance each potential factor would explain.

Table 3: Unrotated centroid factor loadings

Potential Factors	Eigenvalues	% Explained Variance
1	3.8449	30
2	0.7269	6
3	0.15	1
4	0.6709	5
5	0.553	4
6	0.2182	2
7	0.4973	4

The Kaiser-Guttman criterion [30, 31, 32] suggests that factors with an eigenvalue < 1 do not have a significant amount of explanatory power to be extracted and contribute to the final factor solution. However, Brown [21] warns that this criterion should not be applied blindly in Q-techniques, which could result in the loss of qualitatively meaningful and different voices. Inspecting the unrotated factor matrix (see Appendix XI), two participants loaded as high or higher on factor 2 than factor 1, suggesting that excluding factor 2 could result in the loss of an equal share or more of these participants' viewpoint. Humphrey's Rule was applied, where a factor can be considered significant if the cross-product of the two highest loadings of the factor exceed the standard error [21]. This indicated that a two-factor solution was acceptable (see Appendix XI).

Varimax orthogonal rotation was then applied to factors 1 and 2, in order to maximise the apparent differences between groups. Table 4 shows the specific loadings of the Q-sorts onto each factor, with significant loadings highlighted. The amount of common variance explained by each factor is also shown, indicating a factor solution that explained 36% of the variance.

Table 4: Q-sort factor loadings and explained variance

Q sort	Factor 1	Factor 2
001	0.7423	0.1324
002	0.5964	0.0881
003	0.2967	0.295
004	0.3262	0.509
005	0.1613	0.6733
006	0.197	0.3946
007	0.5222	0.2098
008	0.6144	0.2335
009	-0.0118	0.3743
010	0.5579	0.0816
011	0.4654	0.1465
012	0.6607	0.2838
013	0.5279	0.4299
% Explained Variance	**24**	**12**

Significant factor loading was automatically flagged by Ken Q software, highlighted in grey. Brown's (1980) p< .05: 1.96 x (1/ √ no of statements in the Q-set).

4.3. Factor Interpretation

Appendix XII illustrates the two composite factor Q-sorts represented by the final model, highlighting areas of significant difference (positive or negative) between the groups in how they ranked Q-statements. Inspection of these ideal factor arrays was combined with immersion in the qualitative feedback given by participants in each group, using their words to inform the researcher's interpretation of the final factors. There was a mix of demographics and expert stakeholders across the

factors. In the following description of Factor One and Factor Two, statements that were ranked significantly differently between the factors will be represented by * to indicate $p < .05$, and ** where the difference was significant to $p < .001$.

Factor One: Becoming an adult within an established system

Nine participants loaded significantly onto factor one (five healthcare professionals, three young adults with DMD, and one primary carer). It had an eigenvalue of 3.84 and explained 24% of the variance.

This factor was characterised by a focus on young men with DMD developing and maintaining trusting adult relationships within their existing system of health professionals, family, and friends (23**: +6; 22**: +5; 21**: +3 respectively). As one young adult with DMD summarised, *"Having good people around you is the best start"*. This value on reliable, existing relationships is reminiscent of past literature that found young people with various MD conditions, including DMD, found security in being around those who already understood their physical support needs, and could be relied upon physically and emotionally [33, 34, 35]. This sentiment was echoed by one of the healthcare professionals, *"trust in your staff is a relief for physical and mental health"* and the primary care-giver *"trustworthy carers are definitely important"*. Notably however, one young man with DMD highlighted that reinforcing and developing these connections as people grow does not always have to be in person, *"connecting with peers and friends is really important. Xbox Live helps a lot."* The use of online social interaction to maintain friendships within the context of MD and disability has previously been highlighted in qualitative research [35].

The other key value within this factor appeared to be taking responsibility for on-going healthcare and support needs. People highly ranked items that emphasised a shift in ownership over healthcare decisions and action where possible, with one young adult stressing that *"[I] do like to be involved"*. Statements related to this value were ranked significantly more helpful to 'feeling like an adult' by factor one participants, for example, independently managing health needs (10**: +4), being actively involved in healthcare planning (27**: +2), being actively involved in end of life care planning (28**: +1), and talking directly to healthcare professionals (30**:

+4). Moreover, there was an acknowledgment of the social dynamic necessary for this power shift to occur, specifically that professionals include you in all transition decisions (29**: +2). One healthcare professional in particular reflected on the role that others have in facilitating the journey to adulthood, *"it's learning to be an adult but with support to do that, it's about facilitating and offering practical help."* There was also a value placed on managing own finances (19**: +2) and support grants (20**: +2), if the young person was to develop more adult autonomy.

This factor group appeared to value taking on an adult role within a pre-existing collaborative system, including their social network, healthcare system, and financial responsibilities. Two healthcare professionals voiced that *"responsibility is important, it's a mental shift from being closeted as a child"* and that while *"other things are great, practical adult independence needs to come first"*. For this group of staff, service users, and the primary carer, adult development and autonomy should be based within the existing environment, providing a stable base from which the person can flourish; *"I want to stick as I am, with what I can do"* [young adult with DMD].

Factor Two: Becoming an adult by breaking away

Four participants loaded significantly onto factor two (three healthcare professionals and one young adult with DMD). This factor had an eigenvalue of 0.73 and explained 12% of the variance.

In contrast to factor one, the Q-sorts in this factor grouping valued elements of adult life related to independent exploration and stepping away from an established childhood environment. There was a higher focus placed on developing adult connections with peers, for example living with partner / friends / housemates (35**: +3) and being part of a long-term romantic relationship (25**: +2). One healthcare professional explained that for them, it is *"important to have peers to be / live with rather than simply be moulded by your family. Need to make your own way."* Interestingly, previous qualitative research with young men with DMD described their values as presently relating more to the priorities outlined in factor one, while future plans were more similar to the opinions expressed by factor two [14]. Despite this, the current participants in factor two also saw attending to the present-moment as more helpful for adult autonomy and role-taking, compared to Factor One (47**, +3).

This factor identified wanting to distance the adult self from the restrictions of healthcare planning and appointments (43*: -3), as well as reminders of their shortened life expectancy (28**: -5). As the service user included in this factor reflected, *"I'd rather have someone else go or speak for me at hospital appointments and stuff. If it's tests then fine, I need to be there, but discussions are often pointless and just make me depressed."* This view also resonates with previous findings, where young adults with DMD mostly compared themselves to able-bodied peers rather than identifying disability as a characteristic that separated them [14]. Nevertheless, the participants holding this view of what facilitates adult transition did acknowledge the additional needs of emerging adults with DMD, and placed a high value on items that would enable more independence, like assistive technology (38**: +4) and adapted transport (39**: +5), full access to local amenities (1**: +6), and supportive educational environments (3**: +3).

In comparison to factor one, which valued taking on an adult role within the existing system, factor two participants saw support to branch out beyond the world they know as most helpful to facilitating a positive and autonomous adult life. A final interesting point of significant difference between the factors is how the different groups felt about receiving praise from others. Where factor one participants rated it as neutral within the Q-grid (26**: 0), factor two ranked it as significantly less helpful (26**: -2). The qualitative reflections of two young adults with DMD helped to illustrate the difference in perspective, with factor one seeing praise as *"all well and good, but only if you've got the ability to do 'praise-worthy' things – access is more important"*, while the emerging adult loading onto factor two felt that *"praise is difficult to take in big groups or formal settings, it makes me feel self-conscious not grown up."* This view of "it depends what for" seems to underlie factor one's ranking of praise at the neutral point, while factor two appears more actively self-conscious about bringing the attention onto them. This suggests an unmeasured element in how factor two participants assess transition to adulthood, related to achievement being for the self rather than others. This may also relate to those participants within factor two believing it significantly more important to take responsibility for your own actions (54**: +1).

Homogenous statements

Two Q-sort statements were ranked equivalently within factor one and two; living on your own (34: -3), and living with family (36: 0). It seems that all participating stakeholders thought that living with family and maintaining the same living arrangements into adulthood would neither help nor hinder positive adult development within their community, and that living alone would be unhelpful. Several participants commented on these ideas during their qualitative feedback. A factor one service user expressed that they "*wouldn't want to live without family, and other kinds of people would get on my nerves",* the primary care-giver participant felt that they *"wouldn't trust the carers out there"* and the emerging adult with DMD in factor two thought there would be *"too many additional factors if [I] lived alone, it would highlight my disability for me."*

5. Discussion

5.1. Summary of findings

Thirteen participants, including eight healthcare professionals working in a MD Clinic, four emerging adults with DMD, and one primary carer completed Q-sorts ranking 55 statements on how helpful–unhelpful they are in facilitating a positive transition to autonomous adulthood within the wider community. Two distinctive factor groupings emerged from the data: factor one included participants from all three stakeholder groups and explained a larger percentage of the common variance within the model, and factor two included healthcare professionals and one young adult with DMD. The mix of demographics across factor loadings would suggest that different stakeholders affected by DMD do agree on what best facilitates a positive adult transition. However, the existence of two divergent groups does highlight that a universal approach to transition care would not suit the needs or values of everyone. Young people, carers, and healthcare staff should approach conversations about transition with an understanding that they may value different things for an adult life, and could potentially explore the values of everyone involved, informed by these two different factor profiles. This is something that the current participants appreciated, with many of the staff in particular struggling to rank statements in a generalised fashion when they *"work with the individual"*, and the primary carer acknowledged that *"a lot of this depends on what you want out of life"*.

This study consisted of a small sample population from one NHS Trust that operates a life-long model of care. This means that young men with DMD and their carers can expect a consistent relationship with healthcare providers, and the staff participants will have experience working with people with DMD throughout the emerging adult years and beyond. This context does limit the generalisation of findings, but the generated factors do support previous research [33, 34, 35, 14], and the successful use of Q-methodology to investigate such an important exploratory question does show promise for future studies.

5.2. Factor one and the evidence base

Factor one allocated a high value to trusting pre-formed relationships with friends, family, and healthcare professionals. This supports previous qualitative findings by Hunt et al. [33] with young men with DMD and Aho et al. [32, 34] with young people with limb-girdle muscular dystrophy (LGMD). These past studies found that young men appreciated connection with people who knew them well and could offer support without arduous explanation or instruction, as such efforts can highlight difficulty and emphasise their disability. Participants significantly loading onto this factor also felt that more independent ownership of healthcare and financial responsibilities facilitated a sense of adult identity. This is echoed in previous literature, both within the wider context of developmental literature where a greater sense of agency related to stronger self-esteem and internal locus of control [36], and with a mixed cohort of young adult participants with MD reporting higher quality of life when they also felt a stronger sense of autonomy in decision-making and goal setting [37]. The qualitative feedback from young adults spoke of wanting to build autonomy within an existing system; *"I want to stick as I am, with what I can do"*, supported by reliable people that give them the confidence to take on more responsibility for healthcare and financial decisions. Healthcare staff and the primary carer shared this view; however this is likely with a biased lens that emphasised their particular function in enabling more adult role taking. The carer, for example, felt that *"trustworthy carers are definitely important"* while healthcare staff stressed their role in *"offering practical help with how to do that"*. Nevertheless, all showed respect and appreciation for the individual needs of those they support. Healthcare staff recognised that *"staff and medical priorities will be important for staff but this may have less of an impact for the everyday life of patients"*, while the primary carer

conceded that there were *"lots of independence factors that are important… a lot of this depends on what you want."*

5.3. Factor two and the evidence-base

Factor two was defined by a value on things that would support independent exploration, and a distancing of the adult self from health-related contexts. The young adult in this factor explained that *"most of [his] friends are able-bodied, we just have a laugh. My "peers" remind me of things I don't want to think about."* This echoes previous qualitative literature with young men with DMD, whose aim for developing an adult male identity was strongly linked to a desire for normality and to be 'just one of the guys' [14]. The need to feel like, and be seen as, part of the majority group should not be underestimated [38], and has important ramifications for psychological wellbeing [39] and physical health [40]. Interestingly, past developmental research found that relationships to peers did not impact the life satisfaction of emerging adults, rather this was only significant for romantic attachments [41]. However, it might be that in the case of a life-limiting, degenerative condition like DMD, those involved do not see romantic relationships as integral to a full adult experience, instead investing equally or more-so in friendships. The young adult included in factor two reflected that *"Personally, I don't want romantic attachments, porn is easier and less fuss or pressure."* Indeed this reflection aligned greater with the views generally expressed by factor one participants, and therefore the other participating emerging adults with DMD.

The healthcare staff in this factor also put more focus on the independent element of adulthood and the importance of learning to *"make your own way"*, as opposed to the collaborative approach that was valued in factor one. Potentially the healthcare staff and emerging adult included in this factor might have wanted to reduce the burden that continued reliance on primary care-givers can place on family as people with DMD age. Yamaguchi, Sonoda, & Suzuki [42] for example, reported that this can negatively impact carers' anxiety levels and financial burden, as well as increase resentment within families when roles become fixed and restrictive over time. Although the carer in this study loaded onto factor one, their reflection on having not encouraged more academic independence and achievement is poignant here, *"[I] spent the last 20 years not expecting my son to live into adulthood so didn't fight for school education. Regret that now a bit."* It seems that both factor one and

factor two have acknowledged that taking on an adult role involves a shift in power and responsibility, but whether this involves taking ownership of practical and medical obligations, or stepping away from established routines that include a narrative of disability, is not agreed between the two current factor groups.

5.4. Consensus between factors in context

Neither factor one nor two felt that living alone would be beneficial in facilitating the development of a positive, autonomous adult role, and both rated living with family as neutral in its contribution. The narrative behind this appears to relate to anxieties around physical support needs, something that several of the young adults across factor groupings elaborated on; *"home adaptations can be helpful, [but] they are assistive rather than make you independent"* and *"I want to stick as I am, with what I can do… Paid carers are important but they're unreliable and are never there on time."* It seems that the larger social and welfare system provisions including home adaptations and paid care support, which is beyond individual control, are a barrier to these service users, carers, and healthcare staff feeling that living alone could be a positive part of growing up with DMD.

As discussed above, both groups appear to agree that an adult role requires a shift in power to facilitate more autonomy for the emerging adult. This reflects the more general developmental understanding of adolescence, and its psychosocial task of defining identity vs. role confusion [43]. Eriksonian theory describes adolescence and young adulthood as characterised by the generation and resolution of two key questions, "Who am I?" and "What is my place in the world?" [44]. However this inevitably creates tension with those who support the young adult, and the concept of 'emerging adulthood' partly derived from an acknowledgement that the process of disentangling the self may be delayed into late-20s and 30s. It is recognised that most young adults will experience a degree of role confusion, while they experiment with value bases or educational and vocational paths [45], however a lack of resolution can result in self-doubt, loss of meaning in life and an ambivalent view of others [46]. The current study appears to echo this struggle; experimenting with how much autonomy is possible within physical restrictions, and how adult identity is best achieved. So while this journey is normal, the additional factors involved for emerging adults with DMD will make it even more important to

acknowledge the transition difficulties, discuss them openly, and encourage personalised solutions.

5.5. Clinical implications

Q-methodology is a growing method of study within health research [17], but does not seem to have been applied with the current populations. There were some concerns from healthcare staff that young adults with greater physical and mobility difficulties would struggle to complete the Q-sort, but with manual support from the PI or primary carer the participants completed their sorts effectively. The method also comes with a reduced burden on extended conversation, which could become difficult for those using assistive breathing technology. Many participants commented that it was useful and interesting to spend time reflecting and ordering their priorities in this manner, instead of *"working on autopilot."* It also stimulated conversation between the service user and carer dyad, a consequence also noted by Abbot [10], and they ended their appointment by agreeing to explore more options for responsibility sharing and planning new activities. It is interesting to note that these two participants loaded onto factor one, but their discussion led to considering elements of adult exploration more related to factor two priorities, potentially allowing them to consider other aspects of adult identity they had not considered before. It might therefore be useful for transition planning to include prompt cards, to invite carers, young adults, and healthcare professionals to consider a more varied array of options for adult life goals than they might otherwise generate.

Previous lived-experience panel groups have argued that more could be done by health and social care to facilitate young men with DMD to develop a sense of autonomy, adult self-image, and adult role-taking within society [5, 6], as well as remove barriers that can inhibit this from happening [7]. The current study highlighted what specific elements should be facilitated and barriers ought to be reduced, for example creating an environment that welcomes young adults to exercise more agency over healthcare decisions, or helping them to feel secure in exploring new opportunities like travel or living independently. The study also confirms what the carer and several healthcare staff independently reflected; that individual differences in priorities will make the journey to adulthood personal, and therefore support should also be person-centered. The factor solution generated here can begin to make some general conclusions, that at least within this participating NHS Trust, views of

what makes a positive transition to adult life more possible is not split by whether you are a provider of MD healthcare or a service user. Specifically, some participating stakeholders value power sharing within an established system and building confidence upon a foundation of trusted relationships, while for others it will be more important to explore new ways of being and connecting with peers without reminders of being a young adult with a degenerative health condition. However, more research that includes multiple healthcare Trusts and a larger study population would be needed to support these conclusions. Moving forward, these themes for transition priorities could be used as a starting point for conversation between people living with and working with DMD, in order to understand everyone's values or bias before agreeing how to best support the kind of adult life wanted by the individual service user. Recent best practice recommendations [9] have included clinical psychologists as one of the professionals best placed to drive forward such improvements in healthcare support for young people's wellbeing and transition to adulthood. This would appear appropriate, given clinical psychologists competencies around leadership, appreciating individual difference, knowledge of neurodevelopmental stages and needs, and their understanding of systemic principles and how to instigate change within a system. Therefore clinical psychologists working within the field of MD care have a duty to promote the application of, and continue to develop, the evidence base for how to best support adult identity, role taking, and autonomy for emerging adults with DMD; both within the healthcare system through open dialogue, and beyond through psycho-education and skills training.

5.6. Limitations and Future Directions

The current study was conducted within an MD clinic that supplies a life-long service model, rather than a model where transition means moving to a specific adult or palliative care team. This may have contributed to neither factor displaying particularly strong views on having consistent versus adult service support (see Appendix XII), as they have not needed to consider the contrasting options. One healthcare professional reflected that, *"consistency makes transition easier than if you were to change to adult services. They're more stretched and have less time for patients. There's no transition process here so patients might be less aware of this than if it was studied in other areas."* Replicating and expanding on this study by including NHS Trusts that do operate a transition model of care would be helpful in confirming or disproving this assumption, as well as making results more

generalisible. Moreover, while Q-methodology does not require a large number of participants [21], the current study was only able to recruit one primary carer despite efforts to advertise the study at all routine appointments and on the MD Clinic's social media platforms. This difficulty with recruitment was likely due to logistical restraints, as several young adults were seen during respite stays and primary carers were unavailable. In order to give the carer population more of a voice it would be useful to also offer study appointments at the person's home.

The Q-method itself was successfully administered, and even with a small population was able to generate a statistically acceptable factor solution of the priorities for adult transition held by several stakeholder groups affected by DMD. Participants' positive engagement with the method was also encouraging for its on-going application, offering a complimentary method to more commonly used qualitative interview and questionnaire-based studies. This project was intended as a pilot of the model, and the participating MD Clinic will be looking to use these findings to support future research expanding on the sample of participants, honing the Q-statements and ways of presenting the Q-method study, and looking across MD conditions where people's priorities for an adult life might be influenced by differences in disease prognosis.

Staff and young adults both talked about the utility of online resources, and future study could be opened up to a larger population by creating an e-Q-sort that might be more accessible to young adults with DMD and their primary carers. However this may compromise the richness of Q-methodology, as the researcher would be further removed from the participants and elements of the qualitative feedback may be lost without face-to-face interaction. Therefore, piloting an online Q-sort with video-link so that the researcher can still interact with the participant and capture this qualitative feedback in real time is advised. This may improve service user access across a wide geographical area, reduce time demands as people could complete the study from home, work, or day service facilities, and links to study information or sign-up could be passed between emerging adults or primary carers to maximise recruitment.

A limitation of the current study was that while health professionals and those with lived experience of DMD were consulted during the creation of the Q-statements, working alongside DMD stakeholders to co-produce the statements or

holding an initial focus group might have generated a more conclusive list of potential items. Indeed, several participants highlighted statements absent from the Q-concourse, which would be useful to include in future research. The suggestions made by current participants included more items on sex beyond a committed relationship, pets, offering care to others, and engaging in forms of physical activity or hobbies like wheelchair football.

6. Conclusions

Thirteen participants completed a Q-method study ranking a concourse of 55 items that may or may not be helpful in terms of facilitating adult transition for those living with DMD, *"what helps you most to develop into an adult in society?"* Two factor groups emerged from the data, with a mix of those who use services and healthcare professionals in both groups, although factor two only represented one emerging adult voice along with three healthcare professionals. Nevertheless, this suggests a shared understanding and set of priorities between the relevant stakeholder groups. Factor one included five staff, three service users, and one primary carer, who prioritised taking ownership of healthcare and financial responsibilities with support from a trusted social network. Another three healthcare professionals and one emerging adult with DMD loaded significantly onto factor two, who felt that breaking away from the established narrative of healthcare obligations while being practically supported to have new experiences and engage with peers was most helpful in defining an adult identity. Participants reflected that spending time considering their values on this question had been enjoyable and informative, and it stimulated conversation between one emerging adult and his primary carer of where they could further explore his adult experience. An appreciation that young people with DMD, primary carers, and healthcare staff may come from either of these perspectives is therefore advisable when producing personal transition care packages – asking open questions, using prompt cards to explore various facets of adult experience, and being reflective about your own values for an adult life. Moving forward, clinical psychologists are well placed to initiate or supervise the encouragement of adult identity exploration for young people living with DMD, potentially using these two different values profiles to inform healthcare assessments, systemic working practices, and psycho-education.

8. References

[1] Muscular Dystrophy UK. DMD Factsheet, http://www.musculardystrophyuk.org/wp-content/uploads/2015/05/DMD-factsheet.pdf [accessed 17th July 2019].

[2] Muscular Dystrophy Association. https://www.mda.org/disease/duchenne-muscular-dystrophy [accessed 26th July 2019].

[3] Saito T, Kawai M, Kimura E, Ogata K, Takahashi T, Kobayashi M, Takada H, Kuru S, Mikata T, Matsumura T, Yonemoto N. Study of Duchenne muscular dystrophy long-term survivors aged 40 years and older living in specialized institutions in Japan. Neuromuscular Disorders. 2017 Feb 1; 27(2):107-14. https://doi.org/10.1016/j.nmd.2016.11.012

[4] Eagle M, Bourke J, Bullock R, Gibson M, Mehta J, Giddings D, Straub V, Bushby K. Managing Duchenne muscular dystrophy–the additive effect of spinal surgery and home nocturnal ventilation in improving survival. Neuromuscular disorders. 2007 Jun 1; 17(6):470-5. https://doi.org/10.1016/j.nmd.2007.03.002

[5] Schrans DG, Abbott D, Peay HL, Pangalila RF, Vroom E, Goemans N, Vles JS, Aldenkamp AP, Hendriksen JG. Transition in Duchenne muscular dystrophy: an expert meeting report and description of transition needs in an emergent patient population:(parent project muscular dystrophy transition expert meeting 17–18 June 2011, Amsterdam, The Netherlands). Neuromuscular Disorders. 2013 Mar 1; 23(3):283-6. https://doi.org/10.1016/j.nmd.2012.08.009

[6] Bushby K, Finkel R, Birnkrant DJ, Case LE, Clemens PR, Cripe L, Kaul A, Kinnett K, McDonald C, Pandya S, Poysky J. Diagnosis and management of Duchenne muscular dystrophy, part 1: diagnosis, and pharmacological and psychosocial management. The Lancet Neurology. 2010 Jan 1; 9(1):77-93. https://doi.org/10.1016/S1474-4422(09)70271-6

[7] Hendriksen JG, Schrans DG. Jonge de J. Video documentary “DMD with a Future–The power to live. 2011.

[8] Arnett JJ. Emerging adulthood: A theory of development from the late teens through the twenties. American psychologist. 2000 May; 55(5):469. http://dx.doi.org/10.1037/0003-066X.55.5.469

[9] Birnkrant DJ, Bushby K, Bann CM, Apkon SD, Blackwell A, Colvin MK, Cripe L, Herron AR, Kennedy A, Kinnett K, Naprawa J. Diagnosis and management of Duchenne muscular dystrophy, part 3: primary care, emergency management, psychosocial care, and transitions of care across the lifespan. The Lancet Neurology. 2018 May 1; 17(5):445-55. https://doi.org/10.1016/S1474-4422(18)30026-7

[10] Abbott D, Carpenter J, Bushby K. Transition to adulthood for young men with Duchenne muscular dystrophy: research from the UK. Neuromuscular Disorders. 2012 May 1; 22(5):445-6. https://doi.org/10.1016/j.nmd.2012.02.004

[11] Skyrme S. Living with Duchenne Muscular Dystrophy: Relational Autonomy and Decision-Making. Children & Society. 2016 May; 30(3):220-9. https://doi.org/10.1111/chso.12134

[12] Yamaguchi M, Suzuki M. Becoming a back-up carer: parenting sons with Duchenne muscular dystrophy transitioning into adulthood. Neuromuscular disorders. 2015 Jan 1;25(1):85-93. https://doi.org/10.1016/j.nmd.2014.09.001

[13] Abbott D, Carpenter J. ‘Wasting precious time’: young men with Duchenne muscular dystrophy negotiate the transition to adulthood. Disability & Society. 2014 Sep 14;29(8):1192-205. https://doi.org/10.1080/09687599.2014.916607

[14] Gibson BE, Mistry B, Smith B, Yoshida KK, Abbott D, Lindsay S, Hamdani Y. Becoming men: Gender, disability, and transitioning to adulthood. Health, 2014 Jan; 18(1):95-114. https://doi.org/10.1177/1363459313476967

[15] Parkyn H, Coveney J. An exploration of the value of social interaction in a boys' group for adolescents with muscular dystrophy. Child: care, health and development. 2013 Jan; 39(1):81-9. https://doi.org/10.1111/j.1365-2214.2011.01353.x

[16] Beresford B. On the road to nowhere? Young disabled people and transition. Child: care, health and development. 2004 Nov. http://dx.doi.org/10.1111/j.1365-2214.2004.00469.x

[17] Cross RM. Exploring attitudes: the case for Q methodology. Health education research. 2004 Sep 22; 20(2):206-13. https://doi.org/10.1093/her/cyg121

[18] Kelly SE, Moher D, Clifford TJ. Expediting evidence synthesis for healthcare decision-making: exploring attitudes and perceptions towards rapid reviews using Q methodology. PeerJ. 2016 Oct 6; 4:e2522. https://doi.org/10.7717/peerj.2522

[19] Stellefson M, Hanik B, Chaney JD, Tennant B. Analysis of ehealth search perspectives among female college students in the health professions using Q methodology. Journal of medical Internet research. 2012; 14(2):e60. DOI: 10.2196/jmir.1969

[20] Stephenson W. Technique of factor analysis. Nature. 1935; 136: 297. https://doi.org/10.1038/136297b0

[21] Brown SR. Political subjectivity: Applications of Q methodology in political science. Yale University Press; 1980.

[22] Watts S, Stenner P. Doing Q methodological research: Theory, method & interpretation. Sage; 2012 Apr 4.

[23] Trailblaizers blog homehome; http://www.musculardystrophyuk.org/news/blogs/?mdblog_blog_type=trailblazers&post_author=-1&news_year=-1 [accessed 3rd May 2019]

[24] Carers UK discussion homepage; http://www.musculardystrophyuk.org/news/blogs/?mdblog_blog_type=trailblazers&post_author=-1&news_year=-1 [accessed 3rd May 2019]

[25] Johnson BB, Waishwell L. Q method can identify diverse perspectives on 'helpful'information on cancer clusters and inform risk communication generally. Journal of Risk Research. 2014 Oct 21; 17(9):1125-45. https://doi.org/10.1080/13669877.2013.879491

[26] Peter M, Visser M, de Jong MD. Comparing two image research instruments: The Q-sort method versus the Likert attitude questionnaire. Food quality and preference. 2008 Jul 1; 19(5):511-8. https://doi.org/10.1016/j.foodqual.2008.02.007

[27] Rimm-Kaufman SE, Storm MD, Sawyer BE, Pianta RC, LaParo KM. The Teacher Belief Q-Sort: A measure of teachers' priorities in relation to disciplinary practices, teaching practices, and beliefs about children. Journal of School Psychology. 2006 Apr 1; 44(2):141-65. https://doi.org/10.1016/j.jsp.2006.01.003

[28] Cohen J. Statistical power analysis for the behavioral sciences 2nd Edition. New York: Academic press: 1988.

[29] Kline P. An easy guide to factor analysis. Routledge; 2014 Feb 25.

[30] Kaiser HF. The application of electronic computers to factor analysis. Educational and psychological measurement. 1960 Apr; 20(1):141-51. https://doi.org/10.1177/001316446002000116

[31] Kaiser HF. A second generation little jiffy. Psychometrika. 1970 Dec 27; 35(4):401-15. https://doi.org/10.1007/BF02291817

[32] Guttman L. Some necessary conditions for common-factor analysis. Psychometrika. 1954 Jun 1; 19(2):149-61. https://doi.org/10.1007/BF02289162

[33] Aho AC, Hultsjö S, Hjelm K. Perceptions of the transition from receiving the diagnosis recessive limb-girdle muscular dystrophy to becoming in need of human support and using a

wheelchair: an interview study. Disability and rehabilitation. 2019 Sep 11;41 (19):2289-98. https://doi.org/10.1080/09638288.2018.1464602.

[34] Hunt A, Carter B, Abbott J, Parker A, Spinty S, deGoede C. Pain experience, expression and coping in boys and young men with Duchenne Muscular Dystrophy–A pilot study using mixed methods. european journal of paediatric neurology. 2016 Jul 1;20 (4):630-8. https://doi.org/10.1016/j.ejpn.2016.03.002.

[35] Aho AC, Hultsjö S, Hjelm K. Young adults' experiences of living with recessive limb-girdle muscular dystrophy from a salutogenic orientation: an interview study. Disability and rehabilitation. 2015 Oct 23;37 (22):2083-91. https://doi.org/10.3109/09638288.2014.998782.

[36] Schwartz SJ, Côté JE, Arnett JJ. Identity and agency in emerging adulthood: Two developmental routes in the individualization process. Youth & Society. 2005 Dec;37(2):201-29.

[37] Huismann DJ, Sheldon JP, Yashar BM, Amburgey K, Dowling JJ, Petty EM. Quality of life and autonomy in emerging adults with early-onset neuromuscular disorders. Journal of genetic counseling. 2012 Oct 1; 21(5):713-25. https://doi.org/10.1007/s10897-012-9492-z.

[38] Laursen B, Hartl AC. Understanding loneliness during adolescence: Developmental changes that increase the risk of perceived social isolation. Journal of Adolescence. 2013 Dec 1; 36(6):1261-8. https://doi.org/10.1016/j.adolescence.2013.06.003.

[39] Masten CL, Eisenberger NI, Borofsky LA, Pfeifer JH, McNealy K, Mazziotta JC, Dapretto M. Neural correlates of social exclusion during adolescence: understanding the distress of peer rejection. Social cognitive and affective neuroscience. 2009 Jun 1; 4(2):143-57. https://doi.org/10.1093/scan/nsp007.

[40] Brummett BH, Barefoot JC, Siegler IC, Clapp-Channing NE, Lytle BL, Bosworth HB, Williams Jr RB, Mark DB. Characteristics of socially isolated patients with coronary artery disease who are at elevated risk for mortality. Psychosomatic medicine. 2001 Mar 1; 63(2):267-72.

[41] Guarnieri S, Smorti M, Tani F. Attachment relationships and life satisfaction during emerging adulthood. Social Indicators Research. 2015 Apr 1;121(3):833-47.

[42] Yamaguchi M, Sonoda E, Suzuki M. The experience of parents of adult sons with Duchenne muscular dystrophy regarding their prolonged roles as primary caregivers: a serial qualitative study. Disability and rehabilitation. 2019 Mar 27; 41(7):746-52. https://doi.org/10.1080/09638288.2017.1408148.

[43] Erikson EH. Identity: Youth and crisis. WW Norton & company;1968.

[44] McAdams DP, Josselson R, Lieblich A. Identity and story: Creating self in narrative. Psychologia Rozwojowa. 2009;14(1).

[45] Kroger J. Identity in formation. In Hoover K, editor. The future of identity: Centennial reflections on the legacy of Erik Erikson. Lexington Books; 2004 Jun 15.

[46] Bosma HA, Graafsma TLG, Grotevant HD, de Levita DJ, editor. Identity and development: An interdisciplinary approach. Thousand Oaks, CA: SAGE Publications; 1994.

9. Appendices

Appendix I: Staffordshire University ethical approval

INDEPENDENT PEER REVIEW APPROVAL FEEDBACK

Researcher Name	Kevanne Sanger
Title of Study	Facilitating positive adult transition in young people with Duchenne's Muscular Dystrophy
Award Pathway	DClinPsy
Status of approval:	**Approved**

Thank you for forwarding the amendments requested by the Independent Peer Review Panel (IPR)

Action now needed:

You must now apply through the Integrated Research Applications System (IRAS) for approval to conduct your study. You must not commence the study without this second approval. Please note that for the purposes of the IRAS form, the university sponsor is Professor Nachi Chockalingam, N.Chockalingam@staffs.ac.uk .

Please forward a copy of the letter you receive from the IRAS process to ethics@staffs.ac.uk as soon as possible after you have received approval.

Once you have received approval you can commence your study. You should be sure to do so in consultation with your supervisor.

You should note that any divergence from the approved procedures and research method will invalidate any insurance and liability cover from the University. You should, therefore, notify the Panel of any significant divergence from this approved proposal.

When your study is complete, please send the IPR coordinator (Dr Peter Kevern) an end of study report. A template can be found on the ethics BlackBoard site.

Comments for your consideration:

Signed: Dr Peter Kevern
University IPR coordinator

Date: 4.6.18

Appendix II: Bromley research ethics committee (REC) favourable opinion

NHS
Health Research Authority
London - Bromley Research Ethics Committee
Level 3, Block B
Whitefriars
Lewins Mead
Bristol
BS1 2NT

Telephone: 0207 104 8057

Please note: This is the favourable opinion of the REC only and does not allow you to start your study at NHS sites in England until you receive HRA Approval

08 February 2019

Dr Helen Scott
Clinical Psychology, Science Centre, Staffordshire University
Leek Road
Stoke-on-Trent
ST4 2DE

Dear Dr Scott

Study title:	**Facilitating positive adult transition in young people with Duchenne's muscular dystrophy: A Q-sort study of the priorities for service users, carers, and health professionals.**
REC reference:	**19/LO/0214**
Protocol number:	**N/A**
IRAS project ID:	**251237**

The Proportionate Review Sub-committee of the London - Bromley Research Ethics Committee reviewed the above application on 24 January 2019.

We plan to publish your research summary wording for the above study on the HRA website, together with your contact details. Publication will be no earlier than three months from the date of this favourable opinion letter. The expectation is that this information will be published for all studies that receive an ethical opinion but should you wish to provide a substitute contact point, wish to make a request to defer, or require further information, please contact hra.studyregistration@nhs.net outlining the reasons for your request. Under very limited circumstances (e.g. for student research which has received an unfavourable opinion), it may be possible to grant an exemption to the publication of the study.

Appendix III: NHS health research authority (HRA) ethical approval

Dr Helen Scott
Clinical Psychology, Science Centre, Staffordshire University
Leek Road
Stoke-on-Trent
ST4 2DE

Email: hra.approval@nhs.net
Research-permissions@wales.nhs.uk

11 February 2019

Dear Dr. Scott,

HRA and Health and Care Research Wales (HCRW) Approval Letter

Study title:	**Facilitating positive adult transition in young people with Duchenne's muscular dystrophy: A Q-sort study of the priorities for service users, carers, and health professionals.**
IRAS project ID:	**251237**
Protocol number:	**N/A**
REC reference:	**19/LO/0214**
Sponsor	**Staffordshire University**

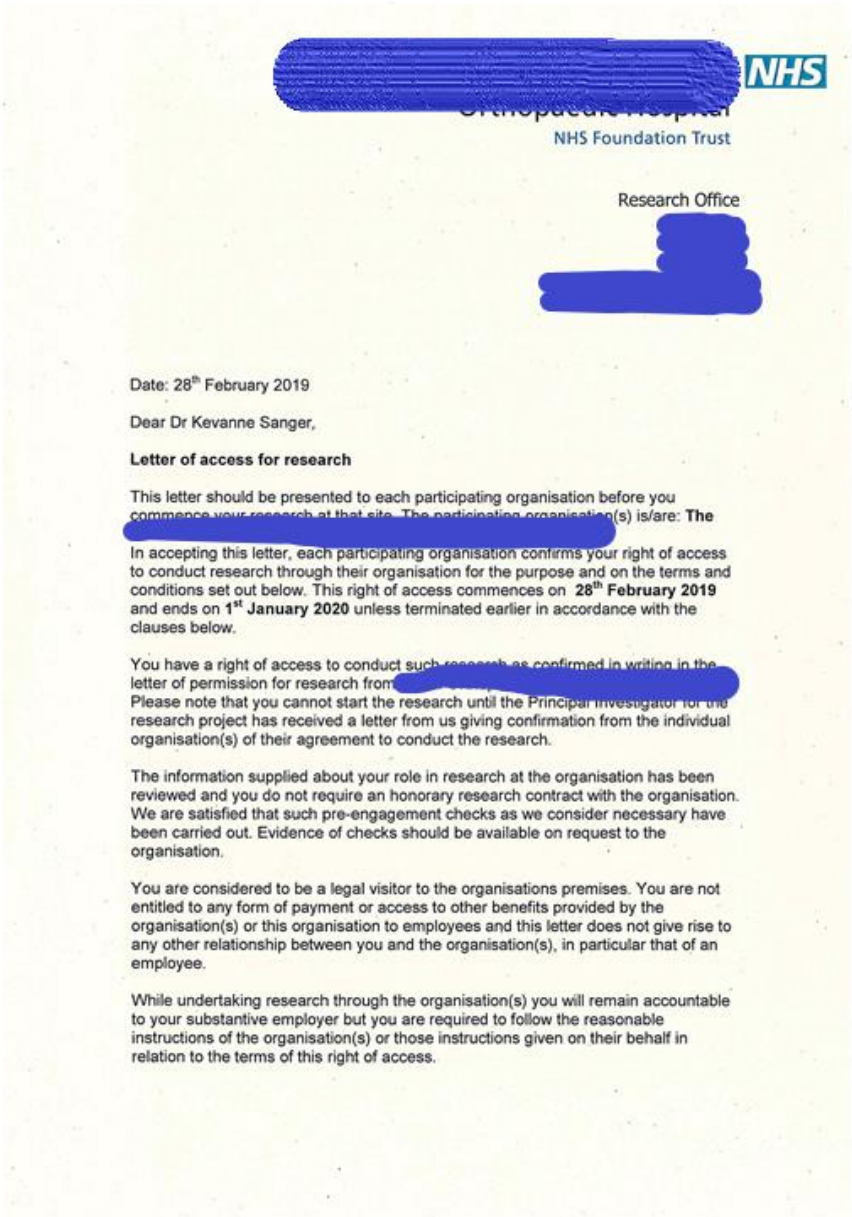
I am pleased to confirm that HRA and Health and Care Research Wales (HCRW) Approval has been given for the above referenced study, on the basis described in the application form, protocol, supporting documentation and any clarifications received. You should not expect to receive anything further relating to this application.

How should I continue to work with participating NHS organisations in England and Wales?
You should now provide a copy of this letter to all participating NHS organisations in England and Wales, as well as any documentation that has been updated as a result of the assessment.

Following the arranging of capacity and capability, participating NHS organisations should **formally confirm** their capacity and capability to undertake the study. How this will be confirmed is detailed in the "*summary of assessment*" section towards the end of this letter.

You should provide, if you have not already done so, detailed instructions to each organisation as to how you will notify them that research activities may commence at site following their confirmation of capacity and capability (e.g. provision by you of a 'green light' email, formal notification following a site initiation visit, activities may commence immediately following confirmation by participating organisation, etc.).

Appendix IV: Research and development letter of access



[redacted] NHS
NHS Foundation Trust

Research Office
[redacted]

Date: 28th February 2019

Dear Dr Kevanne Sanger,

Letter of access for research

This letter should be presented to each participating organisation before you commence your research at that site. The participating organisation(s) is/are: **The** [redacted]

In accepting this letter, each participating organisation confirms your right of access to conduct research through their organisation for the purpose and on the terms and conditions set out below. This right of access commences on **28th February 2019** and ends on **1st January 2020** unless terminated earlier in accordance with the clauses below.

You have a right of access to conduct such research as confirmed in writing in the letter of permission for research from [redacted]
Please note that you cannot start the research until the Principal Investigator for the research project has received a letter from us giving confirmation from the individual organisation(s) of their agreement to conduct the research.

The information supplied about your role in research at the organisation has been reviewed and you do not require an honorary research contract with the organisation. We are satisfied that such pre-engagement checks as we consider necessary have been carried out. Evidence of checks should be available on request to the organisation.

You are considered to be a legal visitor to the organisations premises. You are not entitled to any form of payment or access to other benefits provided by the organisation(s) or this organisation to employees and this letter does not give rise to any other relationship between you and the organisation(s), in particular that of an employee.

While undertaking research through the organisation(s) you will remain accountable to your substantive employer but you are required to follow the reasonable instructions of the organisation(s) or those instructions given on their behalf in relation to the terms of this right of access.

Appendix V:

Healthcare staff advertising poster

STAFFORDSHIRE UNIVERSITY

[redacted] NHS

[redacted] NHS

Research Study Participants Wanted!

Expert views on adult transition in young people with muscular dystrophy

- I am a clinical psychology trainee, and I'm looking for **staff volunteers** involved in the care of people with muscular dystrophy to share their views on what can increase service user independence, and positive transition into adulthood.
  - This is part of a larger study looking at the experience of transition from the perspectives of service users, carers, and healthcare professionals.
- Study participation will be conducted in the Muscular Dystrophy Clinic site within [redacted] NHS Foundations Trust.
- Participating in this **study would take between 30-minutes to 1-hour** of your time.

If you would like to hear more about taking part, please contact:

The clinical supervisor; [redacted]

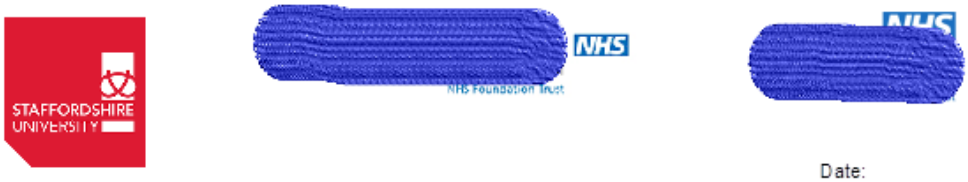
- [redacted]

The Principle Investigator; Kevanne Sanger

- Kevanne.Sanger@combined.nhs.uk / s025080g@student.staffs.ac.uk

Version 1 – 06.05.2018

Appendix VI: Participant invitation letter



Date:

To whom this may concern,

You have been invited to participate in a research study looking into what is important for encouraging independence and positive adult transition in young people with muscular dystrophy.

Participating in this study would involve coming to the Muscular Dystrophy Clinic for a 1-hour study appointment, and rank ordering 40-60 different possible factors that may impact on young people with muscular dystrophy feeling more in control of their life and having a positive adult role in society. You will also be able to offer feedback on how and why you ordered these factors, telling the research team why you think some things are more or less important to this issue.

Please read the attached information sheet for more details about the study and what taking part would mean for you.

If you are interested in taking part, please complete for Notification of Interest slip below and hand it to [redacted] (this can be via your healthcare worker at the Muscular Dystrophy Clinic). Alternatively you can directly e-mail the principle researcher at the address listed below.

Notification of Participant Interest - SU, carer, and health professional views of adult transition in DMD

Name: .....

I am a: Young person with muscular dystrophy ☐

Carer of a young person with muscular dystrophy ☐

Healthcare professional working with people with muscular dystrophy ☐

Contact Telephone Number: .....

Contact E-mail Address: .....

Principle Investigator: Kevanne Sanger, Trainee Clinical Psychologist
Kevanne.Sanger@combined.nhs.uk / s025080g@student.staffs.ac.uk

Appendix VII: Participant information sheet

Participant Information Sheet

Project Title: Facilitating positive adult transition in young people with Duchenne's muscular dystrophy: A Q-sort study of the priorities for service users, carers, and health professionals.

IRAS Project ID Number: 251237

You have been invited to take part in a research study. Please take a few minutes to read through this information carefully. You can talk the information through with family, friends, or healthcare professionals to help you decide if you would like to take part in the study or not. You can also ask the research team any questions that you may have.

Who is involved in this study?

Staffordshire University is the sponsor for this study, based in England. We will be using information from you in order to undertake this study and will act as the data controller for this study. This means that we are responsible for looking after your information and using it properly.

The study is being carried out as part of a Doctoral qualification. The principle investigator (PI), Kevanne Sanger, is studying for a Doctorate in Clinical Psychology at Staffordshire University and this project forms the clinical research component of that training course.

Why are we doing this research?

To explore the factors impacting on transition to adulthood as described by the European Expert Meeting Group – autonomy, adult self-image, and adult role-taking (Schrans et al., 2013). We want to look at what helps transitioning from child to adult healthcare services, but also more broadly within an adult social world, for young and emerging adults with muscular dystrophy (MD) diseases. The aim is to explore this question from the viewpoint of patients themselves, parents and care-givers, and healthcare professionals. This should offer a unique opportunity to explore the range, similarities, and differences in priorities between these expert stakeholder groups. The study hopes to highlight areas of cohesion as well as where increased partnership could improve standards and the well-being of all those effected by MD.

Why is this relevant to me?

As someone who has been invited to take part in this study, you are either someone with a muscular dystrophy condition, the primary care-giver or support person for a young adult with muscular dystrophy, or a healthcare professional specialising in muscular dystrophy care. As one of these groups you should have a direct relationship with the topic of this research, and we hope you will share your views with us about what you find important in maximising positive adult transition in people with MD.

The aim is to increase awareness of what each of these three expert groups prioritise when it comes to developing as an adult with a condition like muscular dystrophy. Having a shared understanding of where everyone is coming from will help professionals, policy

Principle Investigator: Kevanne Sanger, Trainee Clinical Psychologist
s025080g@student.staffs.ac.uk

makers, and individuals (including yourself) work together, and hopefully encourage people to work towards mutually desired goals.

What will participating involve?

Participation may take no longer than 30-minutes, but a quiet room will be booked for 1-hour so that there is no rush during your study appointment. You will be asked to complete an individual Q-sort. This involves the forced choice ranking of 40-60 Q-sample items printed onto statement cards, and arranging them from most to least relevant to you in terms of what maximises autonomy and positive adult transition, into an empty pyramid-shaped grid. The researcher will explain the procedure when you come for your appointment, and give plenty of time to answer any questions. Young adult participants are able to have the researcher or a support person present to help them complete the task, but it is very important that the ranking order of Q-sample statements are only the opinion of the participant. Photographs will be taken of the completed Q-sort, labelled with your unique study ID number.

After you have completed the Q-sort and are happy with the way you have ordered the Q-sample statements, you will be asked if you would like to offer a description of how you ranked the statements, so we can better understand your thinking. The researcher can record your description, or you can write it down on paper provided. This will be used in interpreting the final results when all participants have given their views on least – most important factors for maximising adult transition.

What will happen to my information?

The Muscular Dystrophy Clinic, [redacted] and study PI Kevanne Sanger, will collect information from you for this research study in accordance with General Data Protection Regulations (GDPR) instructions.

The Muscular Dystrophy Clinic will keep your name and contact details confidential and will not pass this information to Staffordshire University. The Muscular Dystrophy Clinic will use this information as needed, to contact you about the research study, and to oversee the quality of the study. Certain individuals from Staffordshire University and regulatory organisations may look at your research records to check the accuracy of the research study. Staffordshire University will only receive information without any identifying information. The people who analyse the information will not be able to identify you, as your data will be given a unique study identification (ID) number. Only the Muscular Dystrophy Clinic and PI Kevanne Sanger will be able to find out your name, or contact details.

In line with GDPR, Staffordshire University will keep identifiable information about you only for the duration of this study (until the end of 2019) but anonymous data will be kept for 10 years after study completion, until 2029. Paper documentation will be stored in a securely locked filing cabinet for the duration of the research project. Participant consent forms will be the only document that show the participant name and ID number together, so these will be kept separate from other documentation in order to maintain data anonymity and confidentiality.

Electronic information e.g. photographs of complete Q-sort grids will be saved in password protected folders and only accessible by the study's PI. This data will be linked to your unique study ID number to maintain anonymity and confidentiality.

Your rights to access, change, or move your information are limited, as we need to manage your information in specific ways in order for the research to be reliable and

accurate. If you withdraw from the study, we will keep the information about you that we have already obtained. To safeguard your rights, we will use the minimum personally-identifiable information possible. You can find out more about how we use your information at http://www.staffs.ac.uk/data-protection/data-protection-policy.jsp

Who has reviewed this study?

All research in the NHS is looked at by an independent group of people, called a Research Ethics Committee to protect your safety, rights, wellbeing and dignity. This study has been reviewed and given approval by HRA and Health and Care Research Wales (REC Ref No. 19/LO/0214). It has also been subject to scientific review by Staffordshire University Faculty of Health Sciences Research Ethics Committee.

What are the possible benefits to taking part?

Participants will receive no direct benefits as a result of taking part in this study. You will be offering your views about this important issue, and adding to our understanding of how to best support emerging adults with muscular dystrophy. The results will be written up for publication and aims to inform academics, healthcare professionals, policy makers, and service users alike.

What are the possible down sides to taking part?

We do not anticipate any risks from participating in this study, but please only offer opinions that you are comfortable with sharing. You are within your rights to refuse participation, or to withdraw from the study at any point without this damaging your healthcare or legal rights.

Will I find out the results of the study?

If you would like to find out the results of the study, please inform the researcher and we will retain your contact details so that a copy of the final research summary can be posted or e-mail to you. This would likely be available by September 2019.

Who do I contact for further information or to make a complaint?

For general enquiries please contact the principle investigator:

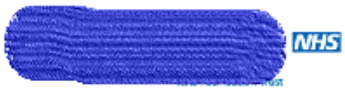
Kevanne Sanger
Trainee Clinical Psychologist
Kevanne.Sanger@northstaffs.nhs.uk / s025080g@student.staffs.ac.uk

To make a complaint please contact the academic supervisor in the first instance, but if you are still unsatisfied, follow the NHS complaints procedure by contacting the Patient Advice and Liaison Service (PALS).

Helen Scott
Senior Lecturer in Clinical Psychology and Registered Clinical Psychologist
H.Scott@staffs.ac.uk

PALS
01782 275031 or 0800 389 9676 (Freephone),
patientexperienceteam@northstaffs.nhs.uk

Appendix VIII: Participant consent form



IRAS ID Number: 251237 **Participant ID Number:**

CONSENT FORM

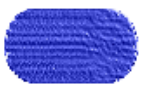
Title of Project: Facilitating positive adult transition in young people with Duchenne's muscular dystrophy: A Q-sort study of the priorities for service users, carers, and health professionals.

Name of Principle Investigator: Kevanne Sanger

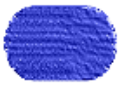
Please initial box

1. I confirm that I have read the information sheet dated..... (V3) for the above study. I have had the opportunity to consider the information, ask questions, and these have been answered satisfactorily. ☐
2. I understand that my participation is voluntary and that I am free to withdraw without giving a reason, without my medical care or legal rights being affected. ☐
3. I understand that the information collected about me will be stored securely, and that my confidentiality and anonymity will be protected. ☐
4. I give permission for anonymised quotations relating to my feedback about my Q-sort to be used within the study write-up. ☐
5. I agree to take part in the above study. ☐

Name of Participant	Date	Signature
_____	_____	_____
Name of Person taking consent	Date	Signature



Principle Investigator: Kevanne Sanger, Trainee Clinical Psychologist
Kevanne.Sanger@combined.nhs.uk / s025080g@student.staffs.ac.uk



Appendix IX: Participant debrief form

Debriefing Form

Project Title

Facilitating positive adult transition in young people with Duchenne's muscular dystrophy: A Q-sort study of the priorities for service users, carers, and health professionals.

Background and Purpose

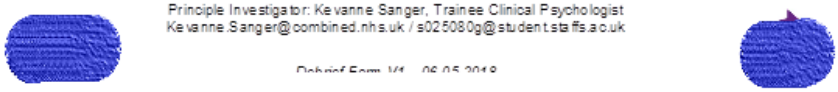
Schrans et al. (2013) held an expert meeting group to look at the transition to adulthood for young men with Duchenne's Muscular Dystrophy (DMD). The meeting group highlighted the importance of encouraging greater independence for clients classed as 'emerging adults', a stage of life described as bridging the gap between adolescence and adulthood and spans 18-29 years of age. It is a time of change and exploration of possible life directions, as well as a period when people take on a more socially perceived adult role (Arnett, 2000). The group argued that services should provide emerging adults with support to achieve some of the 'classic' markers of adulthood, including the consolidation of an adult identity; achieving greater levels of independence (with appropriate support); having opportunities to access work, education, or training; and access to the full range of adult friendships and intimate relationships. However, how these recommendations are to be put in place is left to the individual trusts and there has been no published research to look at the efficacy of interventions so far employed by UK health providers.

This study aimed to explore the factors impacting on emerging adults with DMD in relation to their transition from childhood to adulthood in healthcare settings as well as their wider communities. The objective was to explore what factors impact on emerging adults with DMD being able to develop a sense of adult identity, social role, confidence, and autonomy, from the viewpoint of service users, parents and care-givers (non-professional carers), and healthcare professionals. The study used a Q-sort method so that these groups of individuals could rank order possible factors they feel influence positive adult transition, with the aim of identifying areas of similarity and disagreement between them. A greater understanding of the priorities and values expressed by these three different expert groups will be used to advise researchers and professionals when designing interventions, and guide healthcare policy from a person-centred perspective.

Questions

For general enquiries or to receive a summary of the final research report please contact the principle investigator:

Kevanne Sanger
Trainee Clinical Psychologist
Kevanne.Sanger@combined.nhs.uk / s025080g@student.staffs.ac.uk



Principle Investigator: Kevanne Sanger, Trainee Clinical Psychologist
Kevanne.Sanger@combined.nhs.uk / s025080g@student.staffs.ac.uk

Appendix X: Q-Statements

Q-Sort Statements – What helps you most to develop into an adult in society?

1. Fully accessible local amenities and buildings
2. Joining / participating in local or national groups
3. Campaigning with local or national groups
4. Taking on role / responsibility within groups
5. Keeping a blog / vlog
6. Others asking your advice
7. Connecting with peers with muscular dystrophy
8. Having physical support from paid care workers
9. Having physical support from friends / family / partner
10. Independently managing my health needs e.g. pain, position adjustment
11. Scoring high grades in education
12. Being in a supportive education setting
13. Studying at college or university
14. Leaving formal education
15. Talking to others about career goals and options
16. Additional support to access education or training
17. Paid employment
18. Voluntary work
19. Being responsible for own finances
20. Claiming financial support / grants
21. Friends / partner you can trust
22. Family you can trust
23. Professionals you can trust
24. Forming romantic attachments
25. Being part of a long-term romantic relationship
26. Receiving praise from others
27. Actively involved in healthcare planning
28. Actively involved in end of life care planning
29. Professionals that include you in all ‘transition’ decisions
30. Talking directly to healthcare professionals (not through carers)
31. Having others talk to you “like an adult”
32. Moving to specific adult support services
33. Consistent support – staying with the same known professionals
34. Living on your own
35. Living with partner / friends / housemates
36. Living with family
37. Having house adjustments made to provide more independence
38. Using a wheelchair or other assistive technologies
39. Having adapted transport
40. Advocating for your own needs
41. Disability Discrimination Act (1995)
42. Completing household chores
43. Organising your own medical appointments / treatment
44. Going to appointments / events independently
45. Gathering / reading information on planning for your future
46. Making plans and dreams for your future
47. Focusing on the present, not past or future
48. Travelling to new places
49. Planning and having new experiences
50. Succeeding despite adversity
51. Learning to manage emotional ups and downs
52. Forming own values / opinions
53. ‘Breaking the rules’
54. Taking responsibility for own actions
55. Forming more equal relationship with parents

Appendix XI: Ken-Q Output

Unrotated Factor Matrix

PID	Participant	Factor 1	Factor 2	Factor 3	Factor 4	Factor 5	Factor 6	Factor 7
1	001	0.6905	-0.3029	0.1357	-0.3778	0.2084	0.1561	0.2484
2	002	0.5446	-0.2585	0.0965	0.0534	-0.0402	0.007	-0.1907
3	003	0.4107	0.0801	0.0022	0.0781	-0.1912	0.049	-0.1332
4	004	0.5542	0.2415	0.0511	-0.0147	-0.0986	0.013	0.1094
5	005	0.5085	0.4698	0.3125	0.3356	-0.0148	0.1145	-0.0754
6	006	0.3831	0.2183	0.04	-0.3835	-0.3663	0.3383	-0.045
7	007	0.5507	-0.1161	0.0217	-0.1928	0.0781	0.0282	-0.4319
8	008	0.6405	-0.1477	0.0332	0.088	-0.2227	0.0645	0.1523
9	009	0.1984	0.3176	0.1002	-0.2073	0.3882	0.1581	-0.1302
10	010	0.509	-0.2425	0.0847	-0.0101	-0.175	0.0351	0.1656
11	011	0.4683	-0.1371	0.0289	0.3609	0.2154	0.1694	-0.0888
12	012	0.707	-0.1316	0.0272	0.0428	0.0884	0.0047	0.301
13	013	0.6778	0.0636	0.0008	0.1982	0.1898	0.0614	0.096

Eigenvalues	3.8449	0.7269	0.15	0.6709	0.553	0.2182	0.4973
% Explained Variance	30	6	1	5	4	2	4

Humphrey's Rule

Inclusion Criteria is cross products of factors highest loads > standard error

Standard Error = 1 / √No Statements = 1/√55 = 0.135

Cumulative Communalities Matrix

PID	Participant	Factor 1	Factor 2	Factor 3	Factor 4	Factor 5	Factor 6	Factor 7
1	001	0.4768	0.5685	0.5869	0.7296	0.773	0.7974	0.8591
2	002	0.2966	0.3634	0.3727	0.3756	0.3772	0.3772	0.4136
3	003	0.1687	0.1751	0.1751	0.1812	0.2178	0.2202	0.2379
4	004	0.3071	0.3654	0.368	0.3682	0.3779	0.3781	0.3901
5	005	0.2586	0.4793	0.577	0.6896	0.6898	0.7029	0.7086

6	006	0.1468	0.1945	0.1961	0.3432	0.4774	0.5918	0.5938
7	007	0.3033	0.3168	0.3173	0.3545	0.3606	0.3614	0.5479
8	008	0.4102	0.432	0.4331	0.4408	0.4904	0.4946	0.5178
9	009	0.0394	0.1403	0.1503	0.1933	0.344	0.369	0.386
10	010	0.2591	0.3179	0.3251	0.3252	0.3558	0.357	0.3844
11	011	0.2193	0.2381	0.2389	0.3691	0.4155	0.4442	0.4521
12	012	0.4998	0.5171	0.5178	0.5196	0.5274	0.5274	0.618
13	013	0.4594	0.4634	0.4634	0.5027	0.5387	0.5425	0.5517
Cumulative % Explained Variance		30	36	37	42	46	48	52

Factor Matrix with Defining Sorts Flagged

PID	Q sort	Factor 1		Factor 2	
1	001	0.7423	flagged	0.1324	
2	002	0.5964	flagged	0.0881	
3	003	0.2967	flagged	0.295	
4	004	0.3262		0.509	flagged
5	005	0.1613		0.6733	flagged
6	006	0.197		0.3946	flagged
7	007	0.5222	flagged	0.2098	
8	008	0.6144	flagged	0.2335	
9	009	-0.0118		0.3743	flagged
10	010	0.5579	flagged	0.0816	
11	011	0.4654	flagged	0.1465	
12	012	0.6607	flagged	0.2838	
13	013	0.5279	flagged	0.4299	
% Explained Variance		24		12	

Factor Scores with Corresponding Ranks

Statement Number	Statement	Statement No.	Factor 1	Factor 1	Factor 2	Factor 2
			Z-score	Rank	Z-score	Rank
1	Fully accessible local amenities and buildings	1	1.55	4	2.29	1
2	Joining / participating in local or national groups	2	-0.69	43	-0.01	28
3	Campaigning with local or national groups	3	-1.41	51	-0.06	31
4	Taking on role / responsibility within groups	4	-0.43	37	-0.89	46
5	Keeping a blog / vlog	5	-1.91	54	-2.42	55
6	Others asking your advice	6	0.08	25	-0.62	41
7	Connecting with peers with muscular dystrophy	7	-0.14	30	0.1	26
8	Having physical support from paid care workers	8	1.56	3	0.9	11
9	Having physical support from friends / family / partner	9	1.28	7	1.86	2
10	Independently managing my health needs e.g. pain, position adjustment	10	1.46	5	-0.18	34
11	Scoring high grades in education	11	-1.79	53	-1.43	50
12	Being in a supportive education setting	12	-0.18	31	0.92	10
13	Studying at college or university	13	-0.79	44	0.13	22
14	Leaving formal education	14	-1.02	47	-0.38	36

15	Talking to others about career goals and options	15	0.79	15	0.12	23
16	Additional support to access education or training	16	0.16	23	0.81	12
17	Paid employment	17	1.15	8	0.45	19
18	Voluntary work	18	-0.1	29	-0.71	43
19	Being responsible for own finances	19	0.87	13	-0.04	30
20	Claiming financial support / grants	20	0.83	14	-1.13	49
21	Friends / partner you can trust	21	1.14	10	0.01	27
22	Family you can trust	22	1.65	2	0.46	18
23	Professionals you can trust	23	1.8	1	0.69	13
24	Forming romantic attachments	24	-0.51	39	-0.44	37
25	Being part of a long-term romantic relationship	25	-0.43	38	0.67	14
26	Receiving praise from others	26	0.06	27	-0.79	45
27	Actively involved in healthcare planning	27	1.03	12	-0.66	42
28	Actively involved in end of life care planning	28	0.25	21	-1.8	54
29	Professionals that include you in all ‘transition’ decisions	29	1.04	11	-0.48	39
30	Talking directly to healthcare professionals (not through carers)	30	1.3	6	-0.35	35
31	Having others talk to you “like an adult”	31	0.56	17	1.72	4
32	Moving to specific adult support services	32	-0.58	42	0.11	25

33	Consistent support – staying with the same known professionals	33	0.41	20	-0.14	32
34	Living on your own	34	-1.07	48	-1.1	48
35	Living with partner / friends / housemates	35	-0.97	46	1.13	7
36	Living with family	36	0.11	24	0.12	24
37	Having house adjustments made to provide more independence	37	1.15	9	1.36	6
38	Using a wheelchair or other assistive technologies	38	0.58	16	1.45	5
39	Having adapted transport	39	0.44	18	1.76	3
40	Advocating for your own needs	40	0.02	28	0.62	15
41	Disability Discrimination Act (1995)	41	-1.38	50	-0.58	40
42	Completing household chores	42	-2.67	55	-1.78	53
43	Organising your own medical appointments / treatment	43	-0.33	36	-1.08	47
44	Going to appointments / events independently	44	-0.24	33	0.44	20
45	Gathering / reading information on planning for your future	45	-1.1	49	-0.17	33
46	Making plans and dreams for your future	46	0.21	22	0.59	16
47	Focusing on the present, not past or future	47	-0.94	45	1.04	8
48	Travelling to new places	48	-0.23	32	0.98	9
49	Planning and having new experiences	49	-0.26	34	0.53	17
50	Succeeding despite adversity	50	0.07	26	-0.46	38

51	Learning to manage emotional ups and downs	51	-0.52	40	-0.03	29
52	Forming own values / opinions	52	0.44	19	-1.5	51
53	'Breaking the rules'	53	-1.42	52	-1.5	52
54	Taking responsibility for own actions	54	-0.58	41	0.22	21
55	Forming more equal relationship with parents	55	-0.3	35	-0.74	44

Factor score correlations

	Factor 1	**Factor 2**
Factor 1	1	0.4607
Factor 2	0.4607	1

Factor 1	Sorts Weight

Q Sort	Weight
001	10
012	7.09219
008	5.96981
002	5.59884
010	4.8995
013	4.42668
007	4.34285
011	3.59336
003	1.96786

Factor 2	Sorts Weight

Q Sort	Weight
005	10
004	5.57777
006	3.7947
009	3.53415

Factor 1 Sorts Correlations

Q Sort	001	012	008	002	010	013	007	011	003
001	100	59	50	34	45	40	53	32	20
012	59	100	47	38	48	60	30	38	20
008	50	47	100	38	42	40	31	35	30
002	34	38	38	100	40	32	40	37	27
010	45	48	42	40	100	35	25	15	15
013	40	60	40	32	35	100	23	41	27
007	53	30	31	40	25	23	100	32	27
011	32	38	35	37	15	41	32	100	19
003	20	20	30	27	15	27	27	19	100

Factor 2 Sorts Correlations

Q Sort	005	004	006	009
005	100	38	26	28
004	38	100	33	19
006	26	33	100	17
009	28	19	17	100

Factor Scores for factor 1

Statement No.	Statement	Z-score	Sort Value	001	012	008	002	010	013	007	011	003
23	Professionals you can trust	1.801	6	3	4	2	5	3	4	5	2	1
22	Family you can trust	1.645	5	4	5	3	1	2	5	0	3	3
8	Having physical support from paid care workers	1.558	5	2	4	4	5	5	0	0	2	4
1	Fully accessible local amenities and buildings	1.552	4	4	6	4	3	-1	0	4	1	0
10	Independently managing my health needs e.g. pain, position adjustment	1.457	4	2	0	5	2	6	3	2	4	2
30	Talking directly to healthcare professionals (not through carers)	1.301	4	3	1	5	0	1	2	4	3	4
9	Having physical support from friends / family / partner	1.282	3	3	4	4	-1	0	5	-1	3	5
17	Paid employment	1.154	3	6	2	0	1	4	-2	3	0	0
37	Having house adjustments made to provide more independence	1.154	3	0	5	1	1	5	3	1	4	0
21	Friends / partner you can trust	1.139	3	4	3	3	-1	2	4	1	-2	2
29	Professionals that include you in all ‘transition’ decisions	1.043	2	3	0	1	3	1	1	4	5	-2
27	Actively involved in healthcare planning	1.029	2	2	1	2	2	1	-2	6	5	1

19	Being responsible for own finances	0.868	2	5	3	1	0	2	-1	2	-1	-5
20	Claiming financial support / grants	0.83	2	5	1	-1	2	2	0	1	0	-1
15	Talking to others about career goals and options	0.789	2	2	1	6	1	-1	1	1	-1	1
38	Using a wheelchair or other assistive technologies	0.577	2	-1	0	0	4	0	6	-2	6	0
31	Having others talk to you “like an adult”	0.556	1	1	0	2	-1	1	2	5	0	-1
39	Having adapted transport	0.442	1	-1	2	-1	2	-1	3	3	3	-1
52	Forming own values / opinions	0.435	1	1	-2	-1	4	4	2	0	-1	1
33	Consistent support – staying with the same known professionals	0.41	1	2	1	0	3	0	0	1	-1	-4
28	Actively involved in end of life care planning	0.25	1	-1	2	2	0	0	-1	3	1	-3
46	Making plans and dreams for your future	0.206	1	0	-1	1	6	-2	-1	-1	-2	6
16	Additional support to access education or training	0.16	1	-1	-3	3	2	1	1	0	2	0
36	Living with family	0.109	0	0	3	0	0	0	4	-5	-1	-2
6	Others asking your advice	0.083	0	2	3	-1	-3	2	1	-4	0	-4
50	Succeeding despite adversity	0.067	0	1	0	-2	2	0	0	0	-2	2
26	Receiving praise from others	0.061	0	1	1	0	-1	-2	2	-2	1	0
40	Advocating for your own needs	0.018	0	1	-1	0	-4	-2	1	3	1	5
18	Voluntary work	-0.104	0	-1	2	-2	0	2	-1	-4	4	-2

7	Connecting with peers with muscular dystrophy	-0.135	0	-2	2	0	-2	0	3	-2	-1	2
12	Being in a supportive education setting	-0.182	0	-2	-3	3	4	-5	0	0	2	1
48	Travelling to new places	-0.228	0	0	-2	-1	0	3	2	0	-5	-3
44	Going to appointments / events independently	-0.236	-1	-3	-1	0	1	4	-2	-1	0	2
49	Planning and having new experiences	-0.26	-1	0	0	-1	-4	-2	2	1	1	-1
55	Forming more equal relationship with parents	-0.297	-1	1	-1	-4	-1	-3	0	2	0	3
43	Organising your own medical appointments / treatment	-0.325	-1	-1	-1	-1	3	-1	-2	0	-2	-1
4	Taking on role / responsibility within groups	-0.426	-1	-3	2	2	-2	3	-1	-4	-5	1
25	Being part of a long-term romantic relationship	-0.427	-1	-2	-2	1	0	3	-3	-1	-2	0
24	Forming romantic attachments	-0.514	-1	0	-2	2	-3	1	-3	-1	-4	0
51	Learning to manage emotional ups and downs	-0.521	-2	0	-1	-3	0	-3	-4	-1	2	4
54	Taking responsibility for own actions	-0.576	-2	-4	-2	-3	1	1	1	2	-2	2
32	Moving to specific adult support services	-0.578	-2	-1	0	-5	-3	-2	0	2	0	3
2	Joining / participating in local or national groups	-0.694	-2	-2	1	-2	1	-1	-1	-3	-3	-5
13	Studying at college or university	-0.788	-2	0	-3	1	-3	-6	-2	0	1	-2
47	Focusing on the present, not past or future	-0.941	-2	-3	-2	-3	-2	0	-1	-2	0	1
35	Living with partner / friends / housemates	-0.967	-3	1	-4	1	-5	-1	-4	-3	-3	0
14	Leaving formal education	-1.021	-3	-3	0	-2	-2	-2	-4	-2	1	-3

34	Living on your own	-1.069	-3	0	-5	-5	0	0	-6	2	-3	-1
45	Gathering / reading information on planning for your future	-1.102	-3	-4	0	-4	-1	-4	1	-2	-1	-1
41	Disability Discrimination Act (1995)	-1.384	-4	-4	-1	-3	-2	-3	-3	-1	-6	3
3	Campaigning with local or national groups	-1.41	-4	-2	-4	-2	-5	-1	0	-3	-4	-3
53	'Breaking the rules'	-1.418	-4	-5	-3	-2	-1	-3	-3	1	-3	-2
11	Scoring high grades in education	-1.793	-5	-2	-4	-6	-4	-5	-2	-3	0	-4
5	Keeping a blog / vlog	-1.908	-5	-6	-5	0	-2	-4	-5	-6	2	-2
42	Completing household chores	-2.671	-6	-5	-6	-4	-6	-4	-5	-5	-4	-6

Factor Scores for Factor 2

Statement Number	Statement	Z-score	Sort Values	005	004	006	009
1	Fully accessible local amenities and buildings	2.285	6	6	3	1	6
9	Having physical support from friends / family / partner	1.856	5	5	4	-1	4
39	Having adapted transport	1.762	5	4	1	4	5
31	Having others talk to you “like an adult”	1.717	4	4	4	0	4
38	Using a wheelchair or other assistive technologies	1.453	4	5	0	2	2
37	Having house adjustments made to provide more independence	1.359	4	4	1	3	1
35	Living with partner / friends / housemates	1.126	3	2	2	5	0
47	Focusing on the present, not past or future	1.044	3	1	6	-2	3
48	Travelling to new places	0.977	3	2	1	1	4
12	Being in a supportive education setting	0.919	3	3	2	-1	1
8	Having physical support from paid care workers	0.904	2	3	5	0	-5
16	Additional support to access education or training	0.813	2	2	1	0	3
23	Professionals you can trust	0.686	2	1	3	1	0
25	Being part of a long-term romantic relationship	0.669	2	-1	5	5	-2
40	Advocating for your own needs	0.619	2	2	-2	4	1

46	Making plans and dreams for your future	0.592	2	0	4	2	-1
49	Planning and having new experiences	0.525	1	-1	2	4	2
22	Family you can trust	0.455	1	0	3	0	1
17	Paid employment	0.445	1	-1	0	6	2
44	Going to appointments / events independently	0.435	1	3	-2	2	-2
54	Taking responsibility for own actions	0.218	1	1	-4	3	3
13	Studying at college or university	0.125	1	0	1	0	0
15	Talking to others about career goals and options	0.12	1	2	0	-2	-2
36	Living with family	0.12	0	0	1	-1	1
32	Moving to specific adult support services	0.114	0	2	0	-3	-1
7	Connecting with peers with muscular dystrophy	0.098	0	0	2	1	-3
21	Friends / partner you can trust	0.012	0	-2	3	1	0
2	Joining / participating in local or national groups	-0.007	0	1	0	1	-4
51	Learning to manage emotional ups and downs	-0.026	0	1	-2	0	0
19	Being responsible for own finances	-0.035	0	-2	0	3	2
3	Campaigning with local or national groups	-0.06	0	1	-3	2	-1
33	Consistent support – staying with the same known professionals	-0.145	0	-1	2	-2	0
45	Gathering / reading information on planning for your future	-0.171	-1	0	-2	0	1

10	Independently managing my health needs e.g. pain, position adjustment	-0.184	-1	3	-3	-1	-5
30	Talking directly to healthcare professionals (not through carers)	-0.349	-1	-1	1	-2	-1
14	Leaving formal education	-0.375	-1	0	-1	-2	-1
24	Forming romantic attachments	-0.437	-1	-2	2	0	-3
50	Succeeding despite adversity	-0.456	-1	-4	-1	2	5
29	Professionals that include you in all 'transition' decisions	-0.481	-1	-1	-2	-1	1
41	Disability Discrimination Act (1995)	-0.584	-2	1	-5	-4	2
6	Others asking your advice	-0.62	-2	-2	0	-2	0
27	Actively involved in healthcare planning	-0.662	-2	-3	0	2	-2
18	Voluntary work	-0.71	-2	0	-1	-5	-2
55	Forming more equal relationship with parents	-0.739	-2	-2	-1	-1	-1
26	Receiving praise from others	-0.787	-2	-1	0	-1	-6
4	Taking on role / responsibility within groups	-0.887	-3	-2	-3	3	-4
43	Organising your own medical appointments / treatment	-1.08	-3	0	-4	-4	-3
34	Living on your own	-1.097	-3	-4	-1	1	-2
20	Claiming financial support / grants	-1.126	-3	-4	-1	-4	3
11	Scoring high grades in education	-1.43	-4	-3	-2	-5	-1
52	Forming own values / opinions	-1.499	-4	-5	-3	0	0

53	'Breaking the rules'	-1.504	-4	-5	-1	-3	0
42	Completing household chores	-1.778	-5	-3	-6	-6	2
28	Actively involved in end of life care planning	-1.795	-5	-3	-5	-3	-3
5	Keeping a blog / vlog	-2.422	-6	-6	-4	-3	-4

Descending Array of Differences Between Factor 1 and Factor 2

Statement Number	Statement	Factor 1	Factor 2	Difference
28	Actively involved in end of life care planning	0.25	-1.795	2.045
20	Claiming financial support / grants	0.83	-1.126	1.956
52	Forming own values / opinions	0.435	-1.499	1.934
27	Actively involved in healthcare planning	1.029	-0.662	1.691
30	Talking directly to healthcare professionals (not through carers)	1.301	-0.349	1.65
10	Independently managing my health needs e.g. pain, position adjustment	1.457	-0.184	1.641
29	Professionals that include you in all 'transition' decisions	1.043	-0.481	1.524
22	Family you can trust	1.645	0.455	1.19
21	Friends / partner you can trust	1.139	0.012	1.127
23	Professionals you can trust	1.801	0.686	1.115
19	Being responsible for own finances	0.868	-0.035	0.903
26	Receiving praise from others	0.061	-0.787	0.848
43	Organising your own medical appointments / treatment	-0.325	-1.08	0.755
17	Paid employment	1.154	0.445	0.709
6	Others asking your advice	0.083	-0.62	0.703

15	Talking to others about career goals and options	0.789	0.12	0.669
8	Having physical support from paid care workers	1.558	0.904	0.654
18	Voluntary work	-0.104	-0.71	0.606
33	Consistent support – staying with the same known professionals	0.41	-0.145	0.555
50	Succeeding despite adversity	0.067	-0.456	0.523
5	Keeping a blog / vlog	-1.908	-2.422	0.514
4	Taking on role / responsibility within groups	-0.426	-0.887	0.461
55	Forming more equal relationship with parents	-0.297	-0.739	0.442
53	'Breaking the rules'	-1.418	-1.504	0.086
34	Living on your own	-1.069	-1.097	0.028
36	Living with family	0.109	0.12	-0.011
24	Forming romantic attachments	-0.514	-0.437	-0.077
37	Having house adjustments made to provide more independence	1.154	1.359	-0.205
7	Connecting with peers with muscular dystrophy	-0.135	0.098	-0.233
11	Scoring high grades in education	-1.793	-1.43	-0.363
46	Making plans and dreams for your future	0.206	0.592	-0.386
51	Learning to manage emotional ups and downs	-0.521	-0.026	-0.495
9	Having physical support from friends / family / partner	1.282	1.856	-0.574

40	Advocating for your own needs	0.018	0.619	-0.601
14	Leaving formal education	-1.021	-0.375	-0.646
16	Additional support to access education or training	0.16	0.813	-0.653
44	Going to appointments / events independently	-0.236	0.435	-0.671
2	Joining / participating in local or national groups	-0.694	-0.007	-0.687
32	Moving to specific adult support services	-0.578	0.114	-0.692
1	Fully accessible local amenities and buildings	1.552	2.285	-0.733
49	Planning and having new experiences	-0.26	0.525	-0.785
54	Taking responsibility for own actions	-0.576	0.218	-0.794
41	Disability Discrimination Act (1995)	-1.384	-0.584	-0.8
38	Using a wheelchair or other assistive technologies	0.577	1.453	-0.876
42	Completing household chores	-2.671	-1.778	-0.893
13	Studying at college or university	-0.788	0.125	-0.913
45	Gathering / reading information on planning for your future	-1.102	-0.171	-0.931
25	Being part of a long-term romantic relationship	-0.427	0.669	-1.096
12	Being in a supportive education setting	-0.182	0.919	-1.101
31	Having others talk to you “like an adult”	0.556	1.717	-1.161
48	Travelling to new places	-0.228	0.977	-1.205

39	Having adapted transport	0.442	1.762	-1.32
3	Campaigning with local or national groups	-1.41	-0.06	-1.35
47	Focusing on the present, not past or future	-0.941	1.044	-1.985
35	Living with partner / friends / housemates	-0.967	1.126	-2.093

Factor Q-sort Values for Statements sorted by Consensus vs. Disagreement

Statement Number	Statement	Factor 1	Factor 2	Z-Score variance
34	Living on your own	-3	-3	0
36	Living with family	0	0	0
24	Forming romantic attachments	-1	-1	0.001
53	'Breaking the rules'	-4	-4	0.002
37	Having house adjustments made to provide more independence	3	4	0.011
7	Connecting with peers with muscular dystrophy	0	0	0.014
11	Scoring high grades in education	-5	-4	0.033
46	Making plans and dreams for your future	1	2	0.037
55	Forming more equal relationship with parents	-1	-2	0.049
4	Taking on role / responsibility within groups	-1	-3	0.053
51	Learning to manage emotional ups and downs	-2	0	0.061
5	Keeping a blog / vlog	-5	-6	0.066
50	Succeeding despite adversity	0	-1	0.068
33	Consistent support – staying with the same known professionals	1	0	0.077

9	Having physical support from friends / family / partner	3	5	0.082
40	Advocating for your own needs	0	2	0.09
18	Voluntary work	0	-2	0.092
14	Leaving formal education	-3	-1	0.104
8	Having physical support from paid care workers	5	2	0.107
16	Additional support to access education or training	1	2	0.107
15	Talking to others about career goals and options	2	1	0.112
44	Going to appointments / events independently	-1	1	0.113
2	Joining / participating in local or national groups	-2	0	0.118
32	Moving to specific adult support services	-2	0	0.12
6	Others asking your advice	0	-2	0.124
17	Paid employment	3	1	0.126
1	Fully accessible local amenities and buildings	4	6	0.134
43	Organising your own medical appointments / treatment	-1	-3	0.143
49	Planning and having new experiences	-1	1	0.154
54	Taking responsibility for own actions	-2	1	0.158
41	Disability Discrimination Act (1995)	-4	-2	0.16
26	Receiving praise from others	0	-2	0.18

38	Using a wheelchair or other assistive technologies	2	4	0.192
42	Completing household chores	-6	-5	0.199
19	Being responsible for own finances	2	0	0.204
13	Studying at college or university	-2	1	0.208
45	Gathering / reading information on planning for your future	-3	-1	0.217
25	Being part of a long-term romantic relationship	-1	2	0.3
12	Being in a supportive education setting	0	3	0.303
23	Professionals you can trust	6	2	0.311
21	Friends / partner you can trust	3	0	0.318
31	Having others talk to you “like an adult”	1	4	0.337
22	Family you can trust	5	1	0.354
48	Travelling to new places	0	3	0.363
39	Having adapted transport	1	5	0.436
3	Campaigning with local or national groups	-4	0	0.456
29	Professionals that include you in all ‘transition’ decisions	2	-1	0.581
10	Independently managing my health needs e.g. pain, position adjustment	4	-1	0.673
30	Talking directly to healthcare professionals (not through carers)	4	-1	0.681
27	Actively involved in healthcare planning	2	-2	0.715

52	Forming own values / opinions	1	-4	0.935
20	Claiming financial support / grants	2	-3	0.956
47	Focusing on the present, not past or future	-2	3	0.985
28	Actively involved in end of life care planning	1	-5	1.046
35	Living with partner / friends / housemates	-3	3	1.095

Factor Characteristics

	Factor 1	**Factor 2**
No. of Defining Variables	9	4
Avg. Rel. Coef.	0.8	0.8
Composite Reliability	0.973	0.941
S.E. of Factor Z-scores	0.164	0.243

Standard Errors for Differences in Factor Z-scores

	Factor 1	Factor 2
Factor1	0.232	0.293
Factor2	0.293	0.344

Distinguishing Statements for Factor 1

(P < .05: Asterisk (*) Indicates Significance at P < .01)

Both the Factor Q-Sort Value and the Z-Score (Z-SCR) are Shown

Statement Number	Statement	Factor1 Q-SV	Factor1 Z-score	Significance	Factor2 Q-SV	Factor2 Z-score
23	Professionals you can trust	6	1.8	*	2	0.686
22	Family you can trust	5	1.65	*	1	0.455
8	Having physical support from paid care workers	5	1.56		2	0.904
1	Fully accessible local amenities and buildings	4	1.55		6	2.285
10	Independently managing my health needs e.g. pain, position adjustment	4	1.46	*	-1	-0.184
30	Talking directly to healthcare professionals (not through carers)	4	1.3	*	-1	-0.349
17	Paid employment	3	1.15		1	0.445
21	Friends / partner you can trust	3	1.14	*	0	0.012
29	Professionals that include you in all ‘transition’ decisions	2	1.04	*	-1	-0.481
27	Actively involved in healthcare planning	2	1.03	*	-2	-0.662

19	Being responsible for own finances	2	0.87	*	0	-0.035
20	Claiming financial support / grants	2	0.83	*	-3	-1.126
15	Talking to others about career goals and options	2	0.79		1	0.12
38	Using a wheelchair or other assistive technologies	2	0.58	*	4	1.453
31	Having others talk to you “like an adult”	1	0.56	*	4	1.717
39	Having adapted transport	1	0.44	*	5	1.762
52	Forming own values / opinions	1	0.44	*	-4	-1.499
28	Actively involved in end of life care planning	1	0.25	*	-5	-1.795
16	Additional support to access education or training	1	0.16		2	0.813
6	Others asking your advice	0	0.08		-2	-0.62
26	Receiving praise from others	0	0.06	*	-2	-0.787
40	Advocating for your own needs	0	0.02		2	0.619
18	Voluntary work	0	-0.1		-2	-0.71
12	Being in a supportive education setting	0	-0.18	*	3	0.919
48	Travelling to new places	0	-0.23	*	3	0.977
44	Going to appointments / events independently	-1	-0.24		1	0.435
49	Planning and having new experiences	-1	-0.26	*	1	0.525
43	Organising your own medical appointments / treatment	-1	-0.33		-3	-1.08

25	Being part of a long-term romantic relationship	-1	-0.43	*	2	0.669
32	Moving to specific adult support services	-2	-0.58		0	0.114
54	Taking responsibility for own actions	-2	-0.58	*	1	0.218
2	Joining / participating in local or national groups	-2	-0.69		0	-0.007
13	Studying at college or university	-2	-0.79	*	1	0.125
47	Focusing on the present, not past or future	-2	-0.94	*	3	1.044
35	Living with partner / friends / housemates	-3	-0.97	*	3	1.126
14	Leaving formal education	-3	-1.02		-1	-0.375
45	Gathering / reading information on planning for your future	-3	-1.1	*	-1	-0.171
41	Disability Discrimination Act (1995)	-4	-1.38	*	-2	-0.584
3	Campaigning with local or national groups	-4	-1.41	*	0	-0.06
42	Completing household chores	-6	-2.67	*	-5	-1.778

Distinguishing Statements for Factor 2

(P < .05 : Asterisk (*) Indicates Significance at P < .01)

Both the Factor Q-Sort Value and the Z-Score (Z-SCR) are Shown

Statement Number	Statement	Factor1 Q-SV	Factor1 Z-score	Factor2 Q-SV	Factor2 Z-score	Significance
1	Fully accessible local amenities and buildings	4	1.55	6	2.29	
39	Having adapted transport	1	0.44	5	1.76	*
31	Having others talk to you “like an adult”	1	0.56	4	1.72	*
38	Using a wheelchair or other assistive technologies	2	0.58	4	1.45	*
35	Living with partner / friends / housemates	-3	-0.97	3	1.13	*
47	Focusing on the present, not past or future	-2	-0.94	3	1.04	*
48	Travelling to new places	0	-0.23	3	0.98	*
12	Being in a supportive education setting	0	-0.18	3	0.92	*
8	Having physical support from paid care workers	5	1.56	2	0.9	
16	Additional support to access education or training	1	0.16	2	0.81	

23	Professionals you can trust	6	1.8	2	0.69	*
25	Being part of a long-term romantic relationship	-1	-0.43	2	0.67	*
40	Advocating for your own needs	0	0.02	2	0.62	
49	Planning and having new experiences	-1	-0.26	1	0.53	*
22	Family you can trust	5	1.65	1	0.46	*
17	Paid employment	3	1.15	1	0.45	
44	Going to appointments / events independently	-1	-0.24	1	0.44	
54	Taking responsibility for own actions	-2	-0.58	1	0.22	*
13	Studying at college or university	-2	-0.79	1	0.13	*
15	Talking to others about career goals and options	2	0.79	1	0.12	
32	Moving to specific adult support services	-2	-0.58	0	0.11	
21	Friends / partner you can trust	3	1.14	0	0.01	*
2	Joining / participating in local or national groups	-2	-0.69	0	-0.01	
19	Being responsible for own finances	2	0.87	0	-0.04	*
3	Campaigning with local or national groups	-4	-1.41	0	-0.06	*
45	Gathering / reading information on planning for your future	-3	-1.1	-1	-0.17	*
10	Independently managing my health needs e.g. pain, position adjustment	4	1.46	-1	-0.18	*
30	Talking directly to healthcare professionals (not through carers)	4	1.3	-1	-0.35	*

14	Leaving formal education	-3	-1.02	-1	-0.38	
29	Professionals that include you in all 'transition' decisions	2	1.04	-1	-0.48	*
41	Disability Discrimination Act (1995)	-4	-1.38	-2	-0.58	*
6	Others asking your advice	0	0.08	-2	-0.62	
27	Actively involved in healthcare planning	2	1.03	-2	-0.66	*
18	Voluntary work	0	-0.1	-2	-0.71	
26	Receiving praise from others	0	0.06	-2	-0.79	*
43	Organising your own medical appointments / treatment	-1	-0.33	-3	-1.08	
20	Claiming financial support / grants	2	0.83	-3	-1.13	*
52	Forming own values / opinions	1	0.44	-4	-1.5	*
42	Completing household chores	-6	-2.67	-5	-1.78	*
28	Actively involved in end of life care planning	1	0.25	-5	-1.8	*

Consensus Statements -- Those That Do Not Distinguish Between ANY Pair of Factors

All Listed Statements are Non-Significant at P > 0.01, and Those Flagged with an * are also Non-Significant at P > 0.05)

Statement Number	Significance	Statement	Factor1 Q-SV	Factor1 Z-score	Factor2 Q-SV	Factor2 Z-score
1		Fully accessible local amenities and buildings	4	1.55	6	2.29
2		Joining / participating in local or national groups	-2	-0.69	0	-0.01
4	*	Taking on role / responsibility within groups	-1	-0.426	-3	-0.887
5	*	Keeping a blog / vlog	-5	-1.908	-6	-2.422
6		Others asking your advice	0	0.08	-2	-0.62
7	*	Connecting with peers with muscular dystrophy	0	-0.135	0	0.098
8		Having physical support from paid care workers	5	1.56	2	0.9
9	*	Having physical support from friends / family / partner	3	1.282	5	1.856
11	*	Scoring high grades in education	-5	-1.793	-4	-1.43
14		Leaving formal education	-3	-1.02	-1	-0.38
15		Talking to others about career goals and options	2	0.79	1	0.12
16		Additional support to access education or training	1	0.16	2	0.81

17		Paid employment	3	1.15	1	0.45
18		Voluntary work	0	-0.1	-2	-0.71
24	*	Forming romantic attachments	-1	-0.514	-1	-0.437
32		Moving to specific adult support services	-2	-0.58	0	0.11
33	*	Consistent support – staying with the same known professionals	1	0.41	0	-0.145
34	*	Living on your own	-3	-1.069	-3	-1.097
36	*	Living with family	0	0.109	0	0.12
37	*	Having house adjustments made to provide more independence	3	1.154	4	1.359
40		Advocating for your own needs	0	0.02	2	0.62
43		Organising your own medical appointments / treatment	-1	-0.33	-3	-1.08
44		Going to appointments / events independently	-1	-0.24	1	0.44
46	*	Making plans and dreams for your future	1	0.206	2	0.592
50	*	Succeeding despite adversity	0	0.067	-1	-0.456
51	*	Learning to manage emotional ups and downs	-2	-0.521	0	-0.026
53	*	‘Breaking the rules’	-4	-1.418	-4	-1.504
55	*	Forming more equal relationship with parents	-1	-0.297	-2	-0.739

Relative Ranking of Statements in Factor 1

Statement No.	Highest Ranked Statements	Factor 1	Distinguishing / Consensus	Factor 2
23	Professionals you can trust	6	D*	2
	Positive Statements Ranked Higher in factor 1 Array than in Other Factor Arrays			
22	Family you can trust	5	D*	1
8	Having physical support from paid care workers	5	D	2
10	Independently managing my health needs e.g. pain, position adjustment	4	D*	-1
30	Talking directly to healthcare professionals (not through carers)	4	D*	-1
17	Paid employment	3	D	1
21	Friends / partner you can trust	3	D*	0
29	Professionals that include you in all 'transition' decisions	2	D*	-1
27	Actively involved in healthcare planning	2	D*	-2
19	Being responsible for own finances	2	D*	0
20	Claiming financial support / grants	2	D*	-3
15	Talking to others about career goals and options	2	D	1
52	Forming own values / opinions	1	D*	-4
33	Consistent support – staying with the same known professionals	1	C*	0
28	Actively involved in end of life care planning	1	D*	-5

36	Living with family	0	C*	0
6	Others asking your advice	0	D	-2
50	Succeeding despite adversity	0	C*	-1
26	Receiving praise from others	0	D*	-2
18	Voluntary work	0	D	-2
7	Connecting with peers with muscular dystrophy	0	C*	0
	Negative Statements Ranked Lower in factor 1 Array than in Other Factor Arrays			
36	Living with family	0	C*	0
40	Advocating for your own needs	0	D	2
7	Connecting with peers with muscular dystrophy	0	C*	0
12	Being in a supportive education setting	0	D*	3
48	Travelling to new places	0	D*	3
44	Going to appointments / events independently	-1	D	1
49	Planning and having new experiences	-1	D*	1
25	Being part of a long-term romantic relationship	-1	D*	2
24	Forming romantic attachments	-1	C*	-1
51	Learning to manage emotional ups and downs	-2	C*	0
54	Taking responsibility for own actions	-2	D*	1

32	Moving to specific adult support services	-2	D	0
2	Joining / participating in local or national groups	-2	D	0
13	Studying at college or university	-2	D*	1
47	Focusing on the present, not past or future	-2	D*	3
35	Living with partner / friends / housemates	-3	D*	3
14	Leaving formal education	-3	D	-1
34	Living on your own	-3	C*	-3
45	Gathering / reading information on planning for your future	-3	D*	-1
41	Disability Discrimination Act (1995)	-4	D*	-2
3	Campaigning with local or national groups	-4	D*	0
53	'Breaking the rules'	-4	C*	-4
11	Scoring high grades in education	-5	C*	-4
	Lowest Ranked Statements			
42	Completing household chores	-6	D*	-5

Relative Ranking of Statements in factor 2

Statement No.	Highest Ranked Statements	Factor 2	Distinguishing / Consensus	Factor 1
1	Fully accessible local amenities and buildings	6	D	4
	Positive Statements Ranked Higher in factor 2 Array than in Other Factor Arrays			
9	Having physical support from friends / family / partner	5	C*	3
39	Having adapted transport	5	D*	1
31	Having others talk to you “like an adult”	4	D*	1
38	Using a wheelchair or other assistive technologies	4	D*	2
37	Having house adjustments made to provide more independence	4	C*	3
35	Living with partner / friends / housemates	3	D*	-3
47	Focusing on the present, not past or future	3	D*	-2
48	Travelling to new places	3	D*	0
12	Being in a supportive education setting	3	D*	0
16	Additional support to access education or training	2	D	1
25	Being part of a long-term romantic relationship	2	D*	-1
40	Advocating for your own needs	2	D	0
46	Making plans and dreams for your future	2	C*	1
49	Planning and having new experiences	1	D*	-1

44	Going to appointments / events independently	1	D	-1
54	Taking responsibility for own actions	1	D*	-2
13	Studying at college or university	1	D*	-2
36	Living with family	0	C*	0
32	Moving to specific adult support services	0	D	-2
7	Connecting with peers with muscular dystrophy	0	C*	0
2	Joining / participating in local or national groups	0	D	-2
51	Learning to manage emotional ups and downs	0	C*	-2
3	Campaigning with local or national groups	0	D*	-4
	Negative Statements Ranked Lower in factor 2 Array than in Other Factor Arrays			
36	Living with family	0	C*	0
7	Connecting with peers with muscular dystrophy	0	C*	0
21	Friends / partner you can trust	0	D*	3
19	Being responsible for own finances	0	D*	2
33	Consistent support – staying with the same known professionals	0	C*	1
10	Independently managing my health needs e.g. pain, position adjustment	-1	D*	4
30	Talking directly to healthcare professionals (not through carers)	-1	D*	4
24	Forming romantic attachments	-1	C*	-1

50	Succeeding despite adversity	-1	C*	0
29	Professionals that include you in all 'transition' decisions	-1	D*	2
6	Others asking your advice	-2	D	0
27	Actively involved in healthcare planning	-2	D*	2
18	Voluntary work	-2	D	0
55	Forming more equal relationship with parents	-2	C*	-1
26	Receiving praise from others	-2	D*	0
4	Taking on role / responsibility within groups	-3	C*	-1
43	Organising your own medical appointments / treatment	-3	D	-1
34	Living on your own	-3	C*	-3
20	Claiming financial support / grants	-3	D*	2
52	Forming own values / opinions	-4	D*	1
53	'Breaking the rules'	-4	C*	-4
28	Actively involved in end of life care planning	-5	D*	1
	Lowest Ranked Statements			
5	Keeping a blog / vlog	-6	C*	-5

Appendix XII: Composite Factor Arrays

Composite Q sort for Factor 1

-6	-5	-4	-3	-2	-1	0	1	2	3	4	5	6
✳✳◀ Completing household chores	Scoring high grades in education	✳✳◀ Disability Discrimination Act (1995)	✳✳◀ Living with partner / friends / housemates	Learning to manage emotional ups and downs	✳◀ Going to appointments / events independently	Living with family	✳✳◀ Having others talk to you "like an adult"	✳✳▶ Professionals that include you in all 'transition' decisions	Having physical support from friends / family / partner	✳◀ Fully accessible local amenities and buildings	✳✳▶ Family you can trust	✳✳▶ Professionals you can trust
	Keeping a blog / vlog	✳✳◀ Campaigning with local or national groups	✳◀ Leaving formal education	✳◀ Moving to specific adult support services	✳✳◀ Planning and having new experiences	✳▶ Others asking your advice	✳✳◀ Having adapted transport	✳✳▶ Actively involved in healthcare planning	Having house adjustments made to provide more independence	✳✳▶ Independently managing my health needs e.g. pain, position adjustment	✳▶ Having physical support from paid care workers	
		'Breaking the rules'	Living on your own	✳✳◀ Taking responsibility for own actions	Forming more equal relationship with parents	Succeeding despite adversity	✳✳▶ Forming own values / opinions	✳✳▶ Being responsible for own finances	✳▶ Paid employment	✳✳▶ Talking directly to healthcare professionals (not through carers)		
			✳✳◀ Gathering / reading information on planning for your future	✳◀ Joining / participating in local or national groups	✳▶ Organising your own medical appointments / treatment	✳✳▶ Receiving praise from others	Consistent support – staying with the same known professionals	✳✳▶ Claiming financial support / grants	✳✳▶ Friends / partner you can trust			
				✳✳◀ Studying at college or university	Taking on role / responsibility within groups	✳◀ Advocating for your own needs	✳✳▶ Actively involved in end of life care planning	✳▶ Talking to others about career goals and options				
				✳✳◀ Focusing on the present, not past or future	✳✳◀ Being part of a long-term romantic relationship	✳▶ Voluntary work	Making plans and dreams for your future	✳✳◀ Using a wheelchair or other assistive technologies				
					Forming romantic attachments	Connecting with peers with muscular dystrophy	✳◀ Additional support to access education or training					
						✳✳◀ Being in a supportive education setting						
						✳✳◀ Travelling to new places						

Legend

✳ Distinguishing statement at P< 0.05

✳✳ Distinguishing statement at P< 0.01

► z-Score for the statement is higher than in all the other factors

◀ z-Score for the statement is lower than in all the other factors

Composite Q sort for Factor 2

-6	-5	-4	-3	-2	-1	0	1	2	3	4	5	6
Keeping a blog / vlog	**► Completing household chores	Scoring high grades in education	Taking on role / responsibility within groups	**► Disability Discrimination Act (1995)	**► Gathering / reading information on planning for your future	Living with family	**► Planning and having new experiences	**◄ Having physical support from paid care workers	**► Living with partner / friends / housemates	**► Having others talk to you "like an adult"	Having physical support from friends / family / partner	*► Fully accessible local amenities and buildings
	**◄ Actively involved in end of life care planning	**◄ Forming own values / opinions	*◄ Organising your own medical appointments / treatment	*◄ Others asking your advice	**◄ Independently managing my health needs e.g. pain, position adjustment	*► Moving to specific adult support services	**◄ Family you can trust	*► Additional support to access education or training	**► Focusing on the present, not past or future	**► Using a wheelchair or other assistive technologies	**► Having adapted transport	
		'Breaking the rules'	Living on your own	**◄ Actively involved in healthcare planning	**◄ Talking directly to healthcare professionals (not through carers)	Connecting with peers with muscular dystrophy	*◄ Paid employment	**◄ Professionals you can trust	**► Travelling to new places	Having house adjustments made to provide more independence		
			**◄ Claiming financial support / grants	*◄ Voluntary work	**► Leaving formal education	**◄ Friends / partner you can trust	*► Going to appointments / events independently	**► Being part of a long-term romantic relationship	**► Being in a supportive education setting			
				Forming more equal relationship with parents	Forming romantic attachments	*► Joining / participating in local or national groups	**► Taking responsibility for own actions	*► Advocating for your own needs				
				**◄ Receiving praise from others	Succeeding despite adversity	Learning to manage emotional ups and downs	**► Studying at college or university	Making plans and dreams for your future				
					**◄ Professionals that include you in all 'transition' decisions	**◄ Being responsible for own finances	*◄ Talking to others about career goals and options					
						**► Campaigning with local or national groups						
						Consistent support – staying with the same known professionals						

Legend

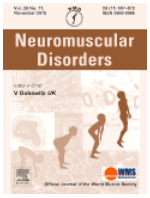
* Distinguishing statement at P< 0.05
** Distinguishing statement at P< 0.01
► z-Score for the statement is higher than in all the other factors
◄ z-Score for the statement is lower than in all the other factors

Appendix XIII: Authors guide for publication

NEUROMUSCULAR DISORDERS
Official Journal of the World Muscle Society

AUTHOR INFORMATION PACK

TABLE OF CONTENTS



ISSN: 0960-8966

DESCRIPTION

This international, multidisciplinary journal covers all aspects of **neuromuscular disorders** in **childhood** and **adult** life (including the muscular dystrophies, spinal muscular atrophies, hereditary neuropathies, congenital myopathies, myasthenias, myotonic syndromes, metabolic myopathies and inflammatory myopathies).

The Editors welcome original articles from all areas of the field:
• Clinical aspects, such as new clinical entities, case studies of interest, treatment, management and rehabilitation (including biomechanics, orthotic design and surgery).
• Basic scientific studies of relevance to the clinical syndromes, including advances in the fields of molecular biology and genetics.
• Studies of animal models relevant to the human diseases.

The journal is aimed at a wide range of clinicians, pathologists, associated paramedical professionals and clinical and basic scientists with an interest in the study of **neuromuscular disorders**.

In addition to original research papers, the journal also publishes reviews and mini-reviews, preliminary short communications and book reviews, and has editorial, correspondence and news sections. Reports on congresses and workshops, taking the form of a digested or very comprehensive commentary, pointing out some of the particular highlights in relation to the contributors and giving some detail of the area covered, important contributions and a list of participants, are also welcome.

The journal is published monthly and aims at rapid publication of high quality papers of scientific merit as well as general interest to a wide readership. There is also a fast track for rapid publication of new material of outstanding scientific merit and importance.

Neuromuscular Disorders is the official journal of the World Muscle Society an international, multidisciplinary, scientific society, dedicated to the advancement and dissemination of knowledge in the field of neuromuscular disorders.

AUDIENCE

Clinicians, pathologists, associated paramedical professionals and clinical and basic scientists with an interest in the study of neuromuscular disorders.

GUIDE FOR AUTHORS

Types of Paper

Research Articles
Regular original research articles should be sent to the main Editorial Office. There is no restriction on length though most articles are between 2500 and 6000 words long. Please contact the Editorial Office if you wish to discuss. The Editor-in-Chief or an appropriate Executive Associate Editor will handle the submission. (For more information on Executive Associate Editors please see Editor's Commentary. Neuromuscular Disorders, Volume 26, Issue 1, January 2016, Pages 1–4.)

Animal Models for Neuromuscular Diseases
Gillian Butler-Browne will be allocated research articles submitted under this section. There is no restriction on length though most articles are between 2500 and 6000 words long. Please contact the Editorial Office if you would like to discuss.

Veterinary Myology
Diane Shelton will be pleased to receive research articles covering clinical or investigative aspects of spontaneously occurring myopathies, neuropathies or disorders of neuromuscular transmission in domestic animals. There is no restriction on length though most articles are between 2500 and 6000 words long. Please contact the Editorial Office if you would like to discuss.

In addition to submitting regular original research articles, letters and meeting reports, we invite readers to submit interesting articles to the special sections listed below. All items should be submitted online in the usual way to the main Editorial Office in London, with the relevant article type selected from the drop-down menu. If you wish to discuss anything with section editors prior to submission please refer to the journal homepage online or the inside front cover of the printed journal for up-to-date contact information of each section editor.

Reviews
Review papers should cover recent, important developments related to diagnosis, pathogenesis or therapy of a neuromuscular disorder. They can be either in-depth and comprehensive, or short, mini-reviews. Please include an abstract and key words. Reviews will be directed to Anders Oldfors who will co-ordinate peer review. There is no upper limit on the length though most articles do not exceed 6000 words. Please contact the Editorial Office if you would like to discuss.

Case Reports
Case reports should be of interest to the multidisciplinary readership of Neuromuscular Disorders and have a neuromuscular component. Topics such as sensory neuropathies and ataxias are of limited interest to our readership. Case reports should not exceed 2000 words and may include up to three tables or figures and a maximum of 25 references. They should take the form of Title, Abstract (up to 150 words), Introduction, Case Report, Discussion, Acknowledgements and References. Please note that key clinical information must be included in the abstract. Case reports will be directed to Beril Talim who will co-ordinate the editorial process.

Picture of the Month
Please send an interesting picture, clinical, pathological or imaging, of clinical challenge or interest. This should be accompanied by a brief case presentation and discussion, highlighting the special features of the picture, in no more than 300 words and up to three references (no abstract is required). The picture should be the main part of the presentation and be of adequate size and good quality.

Clinical Casebook
Contributions will be welcome for this section for cases that show a conflict of interpretation between the clinical and the investigative aspects of a case, with a view to raising questions, promoting thinking and discussion and potentially opening new channels of research to advance our knowledge.

Historical Reports
We welcome articles of historical interest. These can be sent to the Editorial Office in the first instance and will be redirected to the Histor

ENMC Workshop Reports

These submissions will be treated as a report on a workshop, with the convenor(s) listed as corresponding author(s). They will not be subjected to peer review and, after approval by the Editor, will be published in the next available issue of the journal. The workshop report should be concise and follow the agenda of the workshop - it has the nature of a workshop report, not of a review article (setting the stage for future developments).

The length of a report will vary depending on the number of topics discussed. Workshop reports need to be succinct, focusing on the new information. The references should be confined to those directly relevant to the workshop. Up to three tables, figures or photos may be included. No abstract is required.

1. The basic format of the ENMC-based workshop reports will be the same as in the past with a TITLE reflecting the number of the ENMC workshop, the number if appropriate of the topic workshop and the location and date.

2. A full list of all PARTICIPANTS will be included at the end of the report, with their city and country. This list will also include any ENMC representative as appropriate with [ENMC] after their name.

3. Full ACKNOWLEDGEMENT will be given to ENMC and all its sponsoring organisations at the end of the report using the exact wording as requested by ENMC as one of the conditions in their original letter of acceptance of the workshop.

4. In principle, only the workshop organizers will be the author(s) of the workshop report.

The organizers are to make sure that the tasks of all workshop participants regarding the preparation of the meeting report will have been discussed prior to closing the workshop.

All workshop participants will be included in the "ENMC XXXX Workshop Study Group*", so that they can be found in PubMed as co-authors of the workshop report. The workshop participants/report authors will be mentioned in an Appendix under the asterisk. The maximum number of authors for a workshop report (including the "ENMC study group") will be five – so a maximum of four (organizer) names can be used for the workshop report.

The list of authors will be included on the first page of the report, under the title, with a similar format to original papers in the journal. A full but preferably brief address can be included for each author, and the corresponding author for proofs and reprints should also be indicated.

5. As in the past, these reports will not be subjected to any peer review and it will be assumed that the content has the approval of all participants of the workshop. Once approved by the editor, the report will be given priority publication in the next available issue of the journal.

6. Keywords can be provided for reference.

Contact details for submission

Authors may send queries concerning the submission process, manuscript status or journal procedures to the Editorial Office (jane.miller@ucl.ac.uk).

BEFORE YOU BEGIN

Ethics in publishing

Please see our information pages on Ethics in publishing and Ethical guidelines for journal publication.

Description of variants (mutations)

Authors are required to follow the recommendations of the HGVS to describe sequence variants (see http://www.HGVS.org/mutnomen/ for a summary of the current recommendations).

Submission of data to a genetic database

In keeping with the recommendations of the Human Variome Project (Cotton RG et al 207. Nat Genet 39:433 http://www.nature.com/ng/journal/v39/n4/full/ng2024.html) authors submitting a manuscript to *Neuromuscular Disorders* are required to submit all variants and phenotype descriptions to a public database prior to acceptance. Authors must declare the status of database submission in their covering letter upon submission to the journal. In addition, authors should indicate in

their manuscript the database(s) to which they have submitted the variants, and provide the URL. For further information and links to gene variant databases either use GeneSymbol.lovd.nl (e.g. TP53.lovd.nl) or visit the following website: http://www.hgvs.org/dblist/dblist.html.

Declaration of interest

All authors must disclose any financial and personal relationships with other people or organizations that could inappropriately influence (bias) their work. Examples of potential competing interests include employment, consultancies, stock ownership, honoraria, paid expert testimony, patent applications/registrations, and grants or other funding. Authors must disclose any interests in two places: 1. A summary declaration of interest statement in the title page file (if double-blind) or the manuscript file (if single-blind). If there are no interests to declare then please state this: 'Declarations of interest: none'. This summary statement will be ultimately published if the article is accepted. 2. Detailed disclosures as part of a separate Declaration of Interest form, which forms part of the journal's official records. It is important for potential interests to be declared in both places and that the information matches. More information.

Submission declaration and verification

Submission of an article implies that the work described has not been published previously (except in the form of an abstract, a published lecture or academic thesis, see 'Multiple, redundant or concurrent publication' for more information), that it is not under consideration for publication elsewhere, that its publication is approved by all authors and tacitly or explicitly by the responsible authorities where the work was carried out, and that, if accepted, it will not be published elsewhere in the same form, in English or in any other language, including electronically without the written consent of the copyright-holder. To verify originality, your article may be checked by the originality detection service Crossref Similarity Check.

Preprints

Please note that preprints can be shared anywhere at any time, in line with Elsevier's sharing policy. Sharing your preprints e.g. on a preprint server will not count as prior publication (see 'Multiple, redundant or concurrent publication' for more information).

Use of inclusive language

Inclusive language acknowledges diversity, conveys respect to all people, is sensitive to differences, and promotes equal opportunities. Articles should make no assumptions about the beliefs or commitments of any reader, should contain nothing which might imply that one individual is superior to another on the grounds of race, sex, culture or any other characteristic, and should use inclusive language throughout. Authors should ensure that writing is free from bias, for instance by using 'he or she', 'his/her' instead of 'he' or 'his', and by making use of job titles that are free of stereotyping (e.g. 'chairperson' instead of 'chairman' and 'flight attendant' instead of 'stewardess').

Contributors

Each author is required to declare his or her individual contribution to the article: all authors must have materially participated in the research and/or article preparation, so roles for all authors should be described. The statement that all authors have approved the final article should be true and included in the disclosure.

Changes to authorship

Authors are expected to consider carefully the list and order of authors **before** submitting their manuscript and provide the definitive list of authors at the time of the original submission. Any addition, deletion or rearrangement of author names in the authorship list should be made only **before** the manuscript has been accepted and only if approved by the journal Editor. To request such a change, the Editor must receive the following from the **corresponding author**: (a) the reason for the change in author list and (b) written confirmation (e-mail, letter) from all authors that they agree with the addition, removal or rearrangement. In the case of addition or removal of authors, this includes confirmation from the author being added or removed.

Only in exceptional circumstances will the Editor consider the addition, deletion or rearrangement of authors **after** the manuscript has been accepted. While the Editor considers the request, publication of the manuscript will be suspended. If the manuscript has already been published in an online issue, any requests approved by the Editor will result in a corrigendum.

Copyright

Upon acceptance of an article, authors will be asked to complete a 'Journal Publishing Agreement' (see more information on this). An e-mail will be sent to the corresponding author confirming receipt of the manuscript together with a 'Journal Publishing Agreement' form or a link to the online version of this agreement.

Subscribers may reproduce tables of contents or prepare lists of articles including abstracts for internal circulation within their institutions. Permission of the Publisher is required for resale or distribution outside the institution and for all other derivative works, including compilations and translations. If excerpts from other copyrighted works are included, the author(s) must obtain written permission from the copyright owners and credit the source(s) in the article. Elsevier has preprinted forms for use by authors in these cases.

For gold open access articles: Upon acceptance of an article, authors will be asked to complete an 'Exclusive License Agreement' (more information). Permitted third party reuse of gold open access articles is determined by the author's choice of user license.

Author rights

As an author you (or your employer or institution) have certain rights to reuse your work. More information.

Elsevier supports responsible sharing

Find out how you can share your research published in Elsevier journals.

Role of the funding source

You are requested to identify who provided financial support for the conduct of the research and/or preparation of the article and to briefly describe the role of the sponsor(s), if any, in study design; in the collection, analysis and interpretation of data; in the writing of the report; and in the decision to submit the article for publication. If the funding source(s) had no such involvement then this should be stated.

Funding body agreements and policies

Elsevier has established a number of agreements with funding bodies which allow authors to comply with their funder's open access policies. Some funding bodies will reimburse the author for the gold open access publication fee. Details of existing agreements are available online.
After acceptance, open access papers will be published under a noncommercial license. For authors requiring a commercial CC BY license, you can apply after your manuscript is accepted for publication.

Open access

This journal offers authors a choice in publishing their research:

Subscription

• Articles are made available to subscribers as well as developing countries and patient groups through our universal access programs.
• No open access publication fee payable by authors.
• The Author is entitled to post the accepted manuscript in their institution's repository and make this public after an embargo period (known as green Open Access). The published journal article cannot be shared publicly, for example on ResearchGate or Academia.edu, to ensure the sustainability of peer-reviewed research in journal publications. The embargo period for this journal can be found below.

Gold open access

• Articles are freely available to both subscribers and the wider public with permitted reuse.
• A gold open access publication fee is payable by authors or on their behalf, e.g. by their research funder or institution.

Regardless of how you choose to publish your article, the journal will apply the same peer review criteria and acceptance standards.

For gold open access articles, permitted third party (re)use is defined by the following Creative Commons user licenses:

Creative Commons Attribution-NonCommercial-NoDerivs (CC BY-NC-ND)
For non-commercial purposes, lets others distribute and copy the article, and to include in a collective work (such as an anthology), as long as they credit the author(s) and provided they do not alter or modify the article.

The gold open access publication fee for this journal is **USD 3100**, excluding taxes. Learn more about Elsevier's pricing policy: https://www.elsevier.com/openaccesspricing.

Green open access
Authors can share their research in a variety of different ways and Elsevier has a number of green open access options available. We recommend authors see our green open access page for further information. Authors can also self-archive their manuscripts immediately and enable public access from their institution's repository after an embargo period. This is the version that has been accepted for publication and which typically includes author-incorporated changes suggested during submission, peer review and in editor-author communications. Embargo period: For subscription articles, an appropriate amount of time is needed for journals to deliver value to subscribing customers before an article becomes freely available to the public. This is the embargo period and it begins from the date the article is formally published online in its final and fully citable form. Find out more.

This journal has an embargo period of 12 months.

Language (usage and editing services)
Please write your text in good English (American or British usage is accepted, but not a mixture of these). Authors who feel their English language manuscript may require editing to eliminate possible grammatical or spelling errors and to conform to correct scientific English may wish to use the English Language Editing service available from Elsevier's WebShop.

Informed consent and patient details
Studies on patients or volunteers require ethics committee approval and informed consent, which should be documented in the paper. Appropriate consents, permissions and releases must be obtained where an author wishes to include case details or other personal information or images of patients and any other individuals in an Elsevier publication. Written consents must be retained by the author but copies should not be provided to the journal. Only if specifically requested by the journal in exceptional circumstances (for example if a legal issue arises) the author must provide copies of the consents or evidence that such consents have been obtained. For more information, please review the Elsevier Policy on the Use of Images or Personal Information of Patients or other Individuals. Unless you have written permission from the patient (or, where applicable, the next of kin), the personal details of any patient included in any part of the article and in any supplementary materials (including all illustrations and videos) must be removed before submission.

Submission
Our online submission system guides you stepwise through the process of entering your article details and uploading your files. The system converts your article files to a single PDF file used in the peer-review process. Editable files (e.g., Word, LaTeX) are required to typeset your article for final publication. All correspondence, including notification of the Editor's decision and requests for revision, is sent by e-mail.

Submit your article
Please submit your article via EVISE

PREPARATION

Peer review
This journal operates a single blind review process. All contributions will be initially assessed by the editor for suitability for the journal. Papers deemed suitable are then typically sent to a minimum of two independent expert reviewers to assess the scientific quality of the paper. The Editor is responsible for the final decision regarding acceptance or rejection of articles. The Editor's decision is final. More information on types of peer review.

Use of word processing software
It is important that the file be saved in the native format of the word processor used. The text should be in single-column format. Keep the layout of the text as simple as possible. Most formatting codes will be removed and replaced on processing the article. In particular, do not use the word processor's options to justify text or to hyphenate words. However, do use bold face, italics, subscripts, superscripts etc. When preparing tables, if you are using a table grid, use only one grid for each

individual table and not a grid for each row. If no grid is used, use tabs, not spaces, to align columns. The electronic text should be prepared in a way very similar to that of conventional manuscripts (see also the Guide to Publishing with Elsevier). Note that source files of figures, tables and text graphics will be required whether or not you embed your figures in the text. See also the section on Electronic artwork.
To avoid unnecessary errors you are strongly advised to use the 'spell-check' and 'grammar-check' functions of your word processor.

Article structure

Text
Papers should be organized in the following format: Abstract (which must consist of a single paragraph only and no sub-headings), Introduction, Materials (or Patients) and Methods, Results and Discussion. Other descriptive headings and sub-headings may be used if appropriate. Every effort should be made to avoid jargon and non-standard abbreviations. Contents of the study should be presented as clearly and as concisely as possible.

Subdivision - numbered sections
Divide your article into clearly defined and numbered sections. Subsections should be numbered 1.1 (then 1.1.1, 1.1.2, ...), 1.2, etc. (the abstract is not included in section numbering). Use this numbering also for internal cross-referencing: do not just refer to 'the text'. Any subsection may be given a brief heading. Each heading should appear on its own separate line.

Introduction
State the objectives of the work and provide an adequate background, avoiding a detailed literature survey or a summary of the results.

Material and methods
Provide sufficient details to allow the work to be reproduced by an independent researcher. Methods that are already published should be summarized, and indicated by a reference. If quoting directly from a previously published method, use quotation marks and also cite the source. Any modifications to existing methods should also be described.

Results
Results should be clear and concise.

Discussion
This should explore the significance of the results of the work, not repeat them. A combined Results and Discussion section is often appropriate. Avoid extensive citations and discussion of published literature.

Conclusions
The main conclusions of the study may be presented in a short Conclusions section, which may stand alone or form a subsection of a Discussion or Results and Discussion section.

Appendices
If there is more than one appendix, they should be identified as A, B, etc. Formulae and equations in appendices should be given separate numbering: Eq. (A.1), Eq. (A.2), etc.; in a subsequent appendix, Eq. (B.1) and so on. Similarly for tables and figures: Table A.1; Fig. A.1, etc.

Essential title page information

• ***Title.*** Concise and informative. Titles are often used in information-retrieval systems. Avoid abbreviations and formulae where possible.
• ***Author names and affiliations.*** Please clearly indicate the given name(s) and family name(s) of each author and check that all names are accurately spelled. You can add your name between parentheses in your own script behind the English transliteration. Present the authors' affiliation addresses (where the actual work was done) below the names. Indicate all affiliations with a lower-case superscript letter immediately after the author's name and in front of the appropriate address. Provide the full postal address of each affiliation, including the country name and, if available, the e-mail address of each author.
• ***Corresponding author.*** Clearly indicate who will handle correspondence at all stages of refereeing and publication, also post-publication. This responsibility includes answering any future queries about Methodology and Materials. **Ensure that the e-mail address is given and that contact details are kept up to date by the corresponding author.**

• ***Present/permanent address.*** If an author has moved since the work described in the article was done, or was visiting at the time, a 'Present address' (or 'Permanent address') may be indicated as a footnote to that author's name. The address at which the author actually did the work must be retained as the main, affiliation address. Superscript Arabic numerals are used for such footnotes.

Abstract

A concise and factual abstract (up to 200 words for full length articles and 150 words for case reports) is required. The abstract should state briefly the purpose of the research, the principal results and major conclusions. An abstract is often presented separately from the article, so it must be able to stand alone. For this reason, references should be avoided, but if essential, then cite the author(s) and year(s). It should comprise one complete paragraph with no subheadings. Also, non-standard or uncommon abbreviations should be avoided, but if essential they must be defined at their first mention in the abstract itself.

Highlights

Highlights are mandatory for this journal. They consist of a short collection of bullet points that convey the core findings of the article and should be submitted in a separate editable file in the online submission system. Please use 'Highlights' in the file name and include 3 to 5 bullet points (maximum 85 characters, including spaces, per bullet point). You can view example Highlights on our information site.

Keywords

Immediately after the abstract, provide a maximum of 6 keywords, using American spelling and avoiding general and plural terms and multiple concepts (avoid, for example, 'and', 'of'). Be sparing with abbreviations: only abbreviations firmly established in the field may be eligible. These keywords will be used for indexing purposes.

Abbreviations

Define abbreviations that are not standard in this field in a footnote to be placed on the first page of the article. Such abbreviations that are unavoidable in the abstract must be defined at their first mention there, as well as in the footnote. Ensure consistency of abbreviations throughout the article.

Any ambiguous symbols (e.g. the letter 'O' vs the numeral '0', the letter 'l' vs the numeral '1') should be identified. Unnecessary abbreviations should be avoided.
At his discretion the Editor-in-Chief will convert any such abbreviations into their unabbreviated form in order to maintain the flow and sense of the text.

Acknowledgements

Collate acknowledgements in a separate section at the end of the article before the references and do not, therefore, include them on the title page, as a footnote to the title or otherwise. List here those individuals who provided help during the research (e.g., providing language help, writing assistance or proof reading the article, etc.).

Formatting of funding sources

List funding sources in this standard way to facilitate compliance to funder's requirements:

Funding: This work was supported by the National Institutes of Health [grant numbers xxxx, yyyy]; the Bill & Melinda Gates Foundation, Seattle, WA [grant number zzzz]; and the United States Institutes of Peace [grant number aaaa].

It is not necessary to include detailed descriptions on the program or type of grants and awards. When funding is from a block grant or other resources available to a university, college, or other research institution, submit the name of the institute or organization that provided the funding.

If no funding has been provided for the research, please include the following sentence:

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Footnotes

Footnotes should be used sparingly. Number them consecutively throughout the article. Many word processors can build footnotes into the text, and this feature may be used. Otherwise, please indicate the position of footnotes in the text and list the footnotes themselves separately at the end of the article. Do not include footnotes in the Reference list.

Artwork

Electronic artwork

General points

• Make sure you use uniform lettering and sizing of your original artwork.
• Embed the used fonts if the application provides that option.
• Aim to use the following fonts in your illustrations: Arial, Courier, Times New Roman, Symbol, or use fonts that look similar.
• Number the illustrations according to their sequence in the text.
• Use a logical naming convention for your artwork files.
• Provide captions to illustrations separately.
• Size the illustrations close to the desired dimensions of the published version.
• Submit each illustration as a separate file.

A detailed guide on electronic artwork is available.

You are urged to visit this site; some excerpts from the detailed information are given here.

Formats

If your electronic artwork is created in a Microsoft Office application (Word, PowerPoint, Excel) then please supply 'as is' in the native document format.

Regardless of the application used other than Microsoft Office, when your electronic artwork is finalized, please 'Save as' or convert the images to one of the following formats (note the resolution requirements for line drawings, halftones, and line/halftone combinations given below):

EPS (or PDF): Vector drawings, embed all used fonts.

TIFF (or JPEG): Color or grayscale photographs (halftones), keep to a minimum of 300 dpi.

TIFF (or JPEG): Bitmapped (pure black & white pixels) line drawings, keep to a minimum of 1000 dpi.

TIFF (or JPEG): Combinations bitmapped line/half-tone (color or grayscale), keep to a minimum of 500 dpi.

Please do not:

• Supply files that are optimized for screen use (e.g., GIF, BMP, PICT, WPG); these typically have a low number of pixels and limited set of colors;
• Supply files that are too low in resolution;
• Submit graphics that are disproportionately large for the content.

Please note: Figures and tables must be presented in portrait format, or, if landscape, must fit across a portrait page and still be legible for the print journal. Please ensure that any lettering is large enough to read on the journal's print pages.

Colour artwork

Please make sure that artwork files are in an acceptable format (TIFF (or JPEG), EPS (or PDF), or MS Office files) and with the correct resolution. **For colour reproduction in print, you will receive information regarding the costs from Elsevier after receipt of your accepted article. Please note that since the journal Neuromuscular Disorders has a significant print circulation, it is essential that any figures requiring colour should be published in colour in print. The cost for colour reproduction is 200 Euros for the first figure and 100 Euros for each additional figure.** For further information on the preparation of electronic artwork, please see http://www.elsevier.com/artworkinstructions.

Illustration services

Elsevier's WebShop offers Illustration Services to authors preparing to submit a manuscript but concerned about the quality of the images accompanying their article. Elsevier's expert illustrators can produce scientific, technical and medical-style images, as well as a full range of charts, tables and graphs. Image 'polishing' is also available, where our illustrators take your image(s) and improve them to a professional standard. Please visit the website to find out more.

Figure captions

Ensure that each illustration has a caption. Supply captions separately, not attached to the figure. A caption should comprise a brief title (**not** on the figure itself) and a description of the illustration. Keep text in the illustrations themselves to a minimum but explain all symbols and abbreviations used.

If the figure has been published previously a credit line should be included

Tables

Please submit tables as editable text and not as images. Tables can be placed either next to the relevant text in the article, or on separate page(s) at the end. Number tables consecutively in accordance with their appearance in the text and place any table notes below the table body. Be sparing in the use of tables and ensure that the data presented in them do not duplicate results described elsewhere in the article. Please avoid using vertical rules and shading in table cells.

Tables should be submitted online as a separate file and should bear a short descriptive title. If a table must exceed one typewritten page, duplicate all headings on the second sheet. Every column in the table should have an abbreviated heading. Define all abbreviations and indicate the units of measurements for all values. Explain all empty spaces or dashes. Indicate footnotes to the table with the superscript symbols cited in order as you read the table horizontally. Unless tables are unavoidably wide, please present them in portrait format with adequate left and right-hand margins to ensure they do not default to landscape presentation at the typesetters.

References

Citation in text

Please ensure that every reference cited in the text is also present in the reference list (and vice versa). Any references cited in the abstract must be given in full. Unpublished results and personal communications are not recommended in the reference list, but may be mentioned in the text. If these references are included in the reference list they should follow the standard reference style of the journal and should include a substitution of the publication date with either 'Unpublished results' or 'Personal communication'. Citation of a reference as 'in press' implies that the item has been accepted for publication.

Reference links

Increased discoverability of research and high quality peer review are ensured by online links to the sources cited. In order to allow us to create links to abstracting and indexing services, such as Scopus, CrossRef and PubMed, please ensure that data provided in the references are correct. Please note that incorrect surnames, journal/book titles, publication year and pagination may prevent link creation. When copying references, please be careful as they may already contain errors. Use of the DOI is highly encouraged.

A DOI is guaranteed never to change, so you can use it as a permanent link to any electronic article. An example of a citation using DOI for an article not yet in an issue is: VanDecar J.C., Russo R.M., James D.E., Ambeh W.B., Franke M. (2003). Aseismic continuation of the Lesser Antilles slab beneath northeastern Venezuela. Journal of Geophysical Research, https://doi.org/10.1029/2001JB000884. Please note the format of such citations should be in the same style as all other references in the paper.

Web references

As a minimum, the full URL should be given and the date when the reference was last accessed. Any further information, if known (DOI, author names, dates, reference to a source publication, etc.), should also be given. Web references can be listed separately (e.g., after the reference list) under a different heading if desired, or can be included in the reference list.

Data references

This journal encourages you to cite underlying or relevant datasets in your manuscript by citing them in your text and including a data reference in your Reference List. Data references should include the following elements: author name(s), dataset title, data repository, version (where available), year, and global persistent identifier. Add [dataset] immediately before the reference so we can properly identify it as a data reference. The [dataset] identifier will not appear in your published article.

References in a special issue

Please ensure that the words 'this issue' are added to any references in the list (and any citations in the text) to other articles in the same Special Issue.

Reference management software

Most Elsevier journals have their reference template available in many of the most popular reference management software products. These include all products that support Citation Style Language styles, such as Mendeley and Zotero, as well as EndNote. Using the word processor plug-ins from these products, authors only need to select the appropriate journal template when preparing their article, after which citations and bibliographies will be automatically formatted in the journal's style. If no template is yet available for this journal, please follow the format of the sample references

and citations as shown in this Guide. If you use reference management software, please ensure that you remove all field codes before submitting the electronic manuscript. More information on how to remove field codes.

Users of Mendeley Desktop can easily install the reference style for this journal by clicking the following link:
http://open.mendeley.com/use-citation-style/neuromuscular-disorders
When preparing your manuscript, you will then be able to select this style using the Mendeley plug-ins for Microsoft Word or LibreOffice.

Reference style
Text: Indicate references by number(s) in square brackets in line with the text. The actual authors can be referred to, but the reference number(s) must always be given.
List: Number the references (numbers in square brackets) in the list in the order in which they appear in the text.
Examples:
Reference to a journal publication:
[1] Van der Geer J, Hanraads JAJ, Lupton RA. The art of writing a scientific article. J Sci Commun 2010;163:51–9. https://doi.org/10.1016/j.Sc.2010.00372.
Reference to a journal publication with an article number:
[2] Van der Geer J, Hanraads JAJ, Lupton RA. The art of writing a scientific article. Heliyon. 2018;19:e00205. https://doi.org/10.1016/j.heliyon.2018.e00205
Reference to a book:
[3] Strunk Jr W, White EB. The elements of style. 4th ed. New York: Longman; 2000.
Reference to a chapter in an edited book:
[4] Mettam GR, Adams LB. How to prepare an electronic version of your article. In: Jones BS, Smith RZ, editors. Introduction to the electronic age, New York: E-Publishing Inc; 2009, p. 281–304.
Reference to a website:
[5] Cancer Research UK. Cancer statistics reports for the UK, http://www.cancerresearchuk.org/aboutcancer/statistics/cancerstatsreport/; 2003 [accessed 13 March 2003].
Reference to a dataset:
[dataset] [6] Oguro M, Imahiro S, Saito S, Nakashizuka T. Mortality data for Japanese oak wilt disease and surrounding forest compositions, Mendeley Data, v1; 2015. https://doi.org/10.17632/xwj98nb39r.1.
Note shortened form for last page number. e.g., 51–9, and that for more than 6 authors the first 6 should be listed followed by 'et al.' For further details you are referred to 'Uniform Requirements for Manuscripts submitted to Biomedical Journals' (J Am Med Assoc 1997;277:927–34) (see also Samples of Formatted References).

All co-authors in a reference should be included where there are up to six. If there are more than six, include the names of the first six plus 'et al'. Type references double spaced. References cited only in tables or figure legends should be numbered in accordance with a sequence established by the first mention in the text of a particular table or figure. The authors are responsible for the accuracy and completeness of the references.

Journal abbreviations source
Journal names should be abbreviated according to the List of Title Word Abbreviations.

Video

Elsevier accepts video material and animation sequences to support and enhance your scientific research. Authors who have video or animation files that they wish to submit with their article are strongly encouraged to include links to these within the body of the article. This can be done in the same way as a figure or table by referring to the video or animation content and noting in the body text where it should be placed. All submitted files should be properly labeled so that they directly relate to the video file's content. . In order to ensure that your video or animation material is directly usable, please provide the file in one of our recommended file formats with a preferred maximum size of 150 MB per file, 1 GB in total. Video and animation files supplied will be published online in the electronic version of your article in Elsevier Web products, including ScienceDirect. Please supply 'stills' with your files: you can choose any frame from the video or animation or make a separate image. These will be used instead of standard icons and will personalize the link to your video data. For more detailed instructions please visit our video instruction pages. Note: since video and animation cannot be embedded in the print version of the journal, please provide text for both the electronic and the print version for the portions of the article that refer to this content.

Data visualization

Include interactive data visualizations in your publication and let your readers interact and engage more closely with your research. Follow the instructions here to find out about available data visualization options and how to include them with your article.

Supplementary material

Supplementary material such as applications, images and sound clips, can be published with your article to enhance it. Submitted supplementary items are published exactly as they are received (Excel or PowerPoint files will appear as such online). Please submit your material together with the article and supply a concise, descriptive caption for each supplementary file. If you wish to make changes to supplementary material during any stage of the process, please make sure to provide an updated file. Do not annotate any corrections on a previous version. Please switch off the 'Track Changes' option in Microsoft Office files as these will appear in the published version.

Research data

This journal encourages and enables you to share data that supports your research publication where appropriate, and enables you to interlink the data with your published articles. Research data refers to the results of observations or experimentation that validate research findings. To facilitate reproducibility and data reuse, this journal also encourages you to share your software, code, models, algorithms, protocols, methods and other useful materials related to the project.

Below are a number of ways in which you can associate data with your article or make a statement about the availability of your data when submitting your manuscript. If you are sharing data in one of these ways, you are encouraged to cite the data in your manuscript and reference list. Please refer to the "References" section for more information about data citation. For more information on depositing, sharing and using research data and other relevant research materials, visit the research data page.

Data linking

If you have made your research data available in a data repository, you can link your article directly to the dataset. Elsevier collaborates with a number of repositories to link articles on ScienceDirect with relevant repositories, giving readers access to underlying data that gives them a better understanding of the research described.

There are different ways to link your datasets to your article. When available, you can directly link your dataset to your article by providing the relevant information in the submission system. For more information, visit the database linking page.

For supported data repositories a repository banner will automatically appear next to your published article on ScienceDirect.

In addition, you can link to relevant data or entities through identifiers within the text of your manuscript, using the following format: Database: xxxx (e.g., TAIR: AT1G01020; CCDC: 734053; PDB: 1XFN).

Mendeley Data

This journal supports Mendeley Data, enabling you to deposit any research data (including raw and processed data, video, code, software, algorithms, protocols, and methods) associated with your manuscript in a free-to-use, open access repository. During the submission process, after uploading your manuscript, you will have the opportunity to upload your relevant datasets directly to *Mendeley Data*. The datasets will be listed and directly accessible to readers next to your published article online.

For more information, visit the Mendeley Data for journals page.

Data statement

To foster transparency, we encourage you to state the availability of your data in your submission. This may be a requirement of your funding body or institution. If your data is unavailable to access or unsuitable to post, you will have the opportunity to indicate why during the submission process, for example by stating that the research data is confidential. The statement will appear with your published article on ScienceDirect. For more information, visit the Data Statement page.

Submission checklist

The following list will be useful during the final checking of an article prior to sending it to the journal for review. Please also complete the submission checklist and upload this with the files for your submission. For further details of any item please consult this Guide for Authors.

Ensure that the following items are present:

One author has been designated as the corresponding author with contact details:

- E-mail address
- Full postal address
- Phone numbers

All necessary files have been uploaded, and contain:

- Keywords
- Highlights
- All figure captions
- All tables (including title, description, footnotes)

Further considerations

- Manuscript has been 'spell-checked' and 'grammar-checked'
- References are in the correct format for this journal
- All references mentioned in the Reference list are cited in the text, and vice versa
- Permission has been obtained for use of copyrighted material from other sources (including the Web)
- Color figures are clearly marked as being intended for color reproduction
- Each figure or table must be loaded up to the website and labelled individually, and not embedded in the main text.

For any further information please visit our Customer Support site at http://support.elsevier.com.

AFTER ACCEPTANCE

Proofs

One set of page proofs (as PDF files) will be sent by e-mail to the corresponding author (if we do not have an e-mail address then paper proofs will be sent by post) or, a link will be provided in the e-mail so that authors can download the files themselves. Elsevier now provides authors with PDF proofs which can be annotated; for this you will need to download the free Adobe Reader, version 9 (or higher). Instructions on how to annotate PDF files will accompany the proofs (also given online). The exact system requirements are given at the Adobe site.

If you do not wish to use the PDF annotations function, you may list the corrections (including replies to the Query Form) and return them to Elsevier in an e-mail. Please list your corrections quoting line number. If, for any reason, this is not possible, then mark the corrections and any other comments (including replies to the Query Form) on a printout of your proof and scan the pages and return via e-mail. Please use this proof only for checking the typesetting, editing, completeness and correctness of the text, tables and figures. Significant changes to the article as accepted for publication will only be considered at this stage with permission from the Editor. We will do everything possible to get your article published quickly and accurately. It is important to ensure that all corrections are sent back to us in one communication: please check carefully before replying, as inclusion of any subsequent corrections cannot be guaranteed. Proofreading is solely your responsibility.

Offprints

The corresponding author will, at no cost, receive a customized Share Link providing 50 days free access to the final published version of the article on ScienceDirect. The Share Link can be used for sharing the article via any communication channel, including email and social media. For an extra charge, paper offprints can be ordered via the offprint order form which is sent once the article is accepted for publication. Both corresponding and co-authors may order offprints at any time via Elsevier's Webshop. Corresponding authors who have published their article gold open access do not receive a Share Link as their final published version of the article is available open access on ScienceDirect and can be shared through the article DOI link.

AUTHOR INQUIRIES

Visit the Elsevier Support Center to find the answers you need. Here you will find everything from Frequently Asked Questions to ways to get in touch.

You can also check the status of your submitted article or find out when your accepted article will be published.

Chapter 3: Executive Summary

What's helpful when becoming an adult?

Views from emerging adults with Duchenne's Muscular Dystrophy, healthcare professionals, and a primary carer.

Word count: 2,018

What's helpful when becoming an adult?

Views from emerging adults with Duchenne's Muscular Dystrophy, healthcare staff, and a primary carer.

Executive Summary

Dr. Kevanne Sanger

Clinical Psychologist in Training

TERMINOLOGY	
Duchenne's Muscular Dystrophy (DMD)	Most common form of childhood muscular dystrophy. An X-chromosome linked, muscle-wasting condition that affects around 2,500 males living in the UK[1]
Emerging adulthood	Age group between 18-29 years. It is a phase of exploration, and increasing levels of responsibility and independence [2]
DMD lived experience panel	A meeting of people who live with or are affected by DMD.
Transition	When a young person moves from child services to adult services, normally at 18 years of age.
Disability	An umbrella term covering impairments, activity limitations, and participation restrictions. It reflects the interaction between features of a person's body and the society in which they live[3].
Clinical Psychology	Field of study related to people's mental health.
Psycho-education	Teaching knowledge that will support mental health.
Ethics	Guiding rules of conduct governing an individual or group[4].
Ethical committee	Professional board of experts who judge whether a research study upholds ethical standards.
Participant	Someone who consents to participate in research.
Q-method	The format of research conducted in this report.
Q-sort	The way a participant rank orders statements in a Q-methodology study.
Analysis	A detailed examination of something in order to understand its nature: a thorough study[5].
Factor analysis	Statistical method of highlighting agreement and difference between participants' viewpoints.
Significance	A quality of being statistically important, and the

	measured likelihood of occurring by chance.

Background

- Medical advances mean that people with Duchenne's Muscular Dystrophy (DMD) are now living longer into adulthood[6,7].
- However, this poses new questions for people with DMD as well as carers and healthcare staff:

- What do people with DMD want for their adult life, and how do we make sure that happens?
- Are emerging adults with DMD, primary carers, and healthcare staff focusing on the same aspects of an "adult life"?

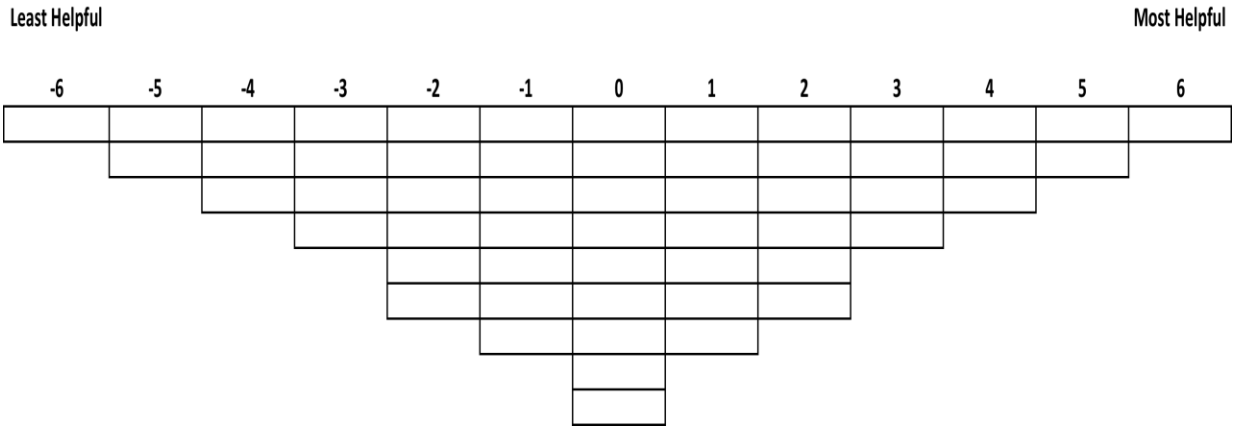
- Previous research suggests that:
  - Emerging adults with DMD want the same things as their able-bodied peers e.g. academic achievement, paid employment, and to form adult relationships[8].
  - Primary carers often feel conflicted between wanting to protect emerging adults with DMD and giving them more independence.
  - No research seems to have asked healthcare staff what they think would most help emerging adults with DMD to experience a full adult life.
  - Emerging adults with DMD can feel unsupported by the healthcare system[9,10] and their wider communities[11] when transitioning to adulthood.
  - Psychological interventions could be helpful in supporting emerging adults' to assert their adult status[12].

Aims of this study

- To find out what emerging adults with DMD, their primary carers, and healthcare staff think is most helpful for emerging adults to experience a full adult life.
- Highlight areas of agreement and difference in people's views.
- Use participants' opinions to guide recommendations for how the healthcare system can best support people with DMD with the kind of adult life they want.

Study methodology

- The study was given ethical approval by Staffordshire University Faculty of Health Sciences Research Ethics Committee and Health Research Authority & Health and Care Research Wales (REC Ref No. 19/LO/0214).
- The study was done in a West Midlands NHS Hospital Trust, and all participants were recruited through them:
    - 4 emerging adults with DMD (aged 18-29 years)
    - 1 primary carer of an emerging adult with DMD
    - 8 employed staff in a Muscular Dystrophy Clinic
- Participants were asked to complete a Q-method study, rank ordering 55 statements from least – most helpful in relation to the question:



"What helps you or someone with DMD most to develop into an adult in society?"

- Participants ordered the 55 statements in an empty grid like the one shown in Figure 1. The researcher was present to help if needed, and to write down people's thoughts about their Q-sort rankings e.g. *"I think adapted transport is very important because it gives me more independence."*

Figure 1: Example Q-Grid

- The 55 statements covered a wide range of themes, and were based on past research of what people believe is important for becoming an adult, as well as social media discussions and consultations with a Muscular Dystrophy charity organisation.

Example statements:

Having adapted transport

Family you can trust

- After collecting the Q-sorts from all participants, the data was analysed using computer software called Ken-Q Analysis.
- This software runs a factor analysis on all the Q-sorts, and groups together participants who ordered the statements in a similar way.
- Two kinds of opinion were found, expressing different values regarding what is helpful or not for becoming an adult in society when living with DMD.

Dissemination of study findings

- Participants were asked to provide an e-mail address if they would like a summary of the research findings sent to them.
- This executive summary will be sent to the NHS Hospital Trust and charity organisation that supported the study, for general dissemination.
- The research report will be submitted for publication in an international academic journal.

Study results

Group One

Becoming an adult within an established system

Included 5 staff, 3 emerging adults with DMD, and 1 primary carer

Things this group ranked significantly more helpful than Group Two:

- ✓ Professionals you can trust (+6)
- ✓ Family you can trust (+5)
- ✓ Friends / partner you can trust (+3)
- ✓ Independently managing healthcare (+4)
- ✓ Talking directly to healthcare professionals (+4)
- ✓ Actively involved in healthcare planning (+2)
- ✓ Responsible for own finances (+2)

Healthcare staff:

"Trust in your staff is a relief for physical and mental health"

Emerging adult:

"Having good people around you is the best start."

Primary carer:

"Trustworthy carers are definitely important."

Group One

Becoming an adult within an established system

Included 5 staff, 3 emerging adults with DMD, and 1 primary carer

Primary carer:

"Nothing you can do to control dating. Whereas groups and keeping friends is more valuable and likely.

Healthcare staff:

"Other things are great, but practical adult independence needs to come first".

Emerging adult:

"I want to stick as I am, with what I can do"

The participants in this group felt that being involved in decisions around healthcare and finances, whilst having support from existing, trusted people was the best way to achieve independence and an adult role.

Group Two

Becoming an adult by breaking away

Included 3 staff, and 1 emerging adult with DMD

Things this group ranked significantly more helpful than Group One:

- ✓ Adapted transport (+5)
- ✓ Having others talking to you like an adult (+4)
- ✓ Using assistive technologies (+4)
- ✓ Living with peers (+3)
- ✓ Focussing on the present (+3)
- ✓ Travelling to new places (+3)
- ✓ Supportive educational environment (+3)
- ✓ Being part of a long-term romantic relationship (+2)

Healthcare staff:

"Important to have peers to be / live with rather than simply be moulded by your family. Need to make your own way."

Emerging adult:

"Adapted transport helps tremendously. I'd be a shut in without it. Planes however should be much more wheelchair

Group Two

Becoming an adult by breaking away

Included 3 staff, and 1 emerging adult with DMD

Emerging adult:

"Praise is difficult to take in big groups or formal settings, it makes me feel self-conscious not grown up."

Healthcare staff:

"End of life decisions and process… It's good to do, but that's so it's done and can be locked away and not looked at again really."

Emerging adult:

"I'd rather have someone else go or speak for me at hospital appointments… If it's tests then fine, I need to be there, but discussions are often pointless and just make me depressed."

This group of participants saw adult life as being helped by exploring new opportunities, associating with peers, and stepping away from an identity and set of responsibilities defined by disability.

Agreement between groups

Two statements were ranked the same by both groups

Living with family

- Ranked as neither helpful nor unhelpful (0) by both groups.
- The majority of emerging adults with DMD live with family, and all participants felt that continuing what is likely to be their existing living arrangements would neither help nor hinder the development of an adult life.

Living on your own

- Ranked as unhelpful (-3) by both groups.
- Living alone was believed by everyone to likely reduce emerging adults' ability to develop an adult role in society.

Primary carer:

"Living alone [ranked] down the bottom - wouldn't trust the carers out there"

Emerging adult:

"Too many additional factors if I lived alone, it would highlight my disability for me."

Discussion of findings and recommendations

- This study highlighted two different sets of opinion regarding what is most helpful and unhelpful for building an adult life and role in society with DMD.
- Group one was not categorised by demographic (young adult, carer, or staff) and group two also represented the voice of one young adult as well as three healthcare staff. This suggests that at least within this NHS Trust, people living with DMD or caring for those with DMD do value and prioritise similar things when thinking about transitioning to adulthood.
- However, two different groups were found. This means that is important for everyone involved in supporting the transition process to know what they value for an adult life, because everyone involved may not prioritise the same things.
- Previous lived experience panels highlighted that healthcare services could be doing more to facilitate young people with DMD's adult development[13,14] and the resulting advice stated that clinical psychologists could be helpful in building emerging adult's confidence to assert their adult status (through annual assessments, psycho-education, and skills training)[12].
    - Annual assessments could be guided by the values highlighted in this study, to see if a young adult with DMD wants more control over health and financial decisions vs. support to access education or travelling for example.

Study limitations and future research

- Small sample of participants
  - A larger study including views from more young adults with DMD and primary carers would explore additional views or confirm the factors found here.
- Life-long vs. transition service model
  - This study was conducted in an NHS Trust using a 'life-long service' model, meaning that people continue to see the same healthcare team throughout life. People supported by or working for a Trust operating a 'transition model', where you change between child and adult services, might value different things for adulthood.
  - Further research to see if values are different between 'life-long' and 'transition' service models, particularly opinions of healthcare support, is recommended.
- Use an online Q-method study tool
  - Many participants said they enjoyed taking part and reflecting on their values in this way.
  - An online Q-sort could help more participants access the study without needing to travel for appointments. However, an online version may lose the personal understanding of face-to-face conversation. Future studies could explore an online Q-sort with video link between the researcher and participant.
- Missing themes and statements
  - Participants suggested including the following statements in future Q-method studies:

1. Offering care to others
2. Owning a pet
3. Sex outside of a committed relationship
4. Physical activities / hobbies

Conclusions

This was the first time a Q-method study had been used to ask people's opinions of what is most helpful or unhelpful for an emerging adult with DMD to take on an adult role and identity in society. Two different groups of values were apparent, although the groups were not split by whether they were an emerging adult, carer, or healthcare staff member, This suggests that the various people affected by DMD are often focusing on the same goals for an adult life. Conducting transition conversations with these two opinions in mind could help young people with DMD, their primary carers, and healthcare staff, all make sure they are effectively supporting the kind of adult life wanted by the emerging adult. This project is an encouraging start, but more research is needed. It is particularly advised that gaining more opinions from young people with DMD and primary carers, and those within NHS Trusts using a 'transition model' of support,

References

1. Muscular Dystrophy UK. http://www.musculardystrophyuk.org/wp-content/uploads/2015/05/DMD-factsheet.pdf
2. Arnett, J. J. (2000). Emerging adulthood: A theory of development from the late teens through the twenties. American Psychologist, 55(5), 469-480.
3. World Health Organisation. https://www.who.int/topics/disabilities/en/
4. Merriam-Webster dictionary. https://www.merriam-webster.com/dictionary/ethic
5. Merriam-Webster dictionary. https://www.merriam-webster.com/dictionary/analysis
6. Saito, T,. Kawai, M., Kimura, E., Ogata. K., Takahashi. T., Kobayashi. M., Takada, H., Kuru, S., Mikata, T., Matsumura, T., & Yonemoto, N. (2017). Study of Duchenne muscular dystrophy long-term survivors aged 40 years and older living in specialized institutions in Japan. Neuromuscular Disorders, 27(2), 107-14.
7. Eagle, M., Bourke, J., Bullock, R., Gibson, M., Mehta, J., Giddings, D., Straub, V., & Bushby, K. (2007). Managing Duchenne muscular dystrophy–the additive effect of spinal surgery and home nocturnal ventilation in improving survival. Neuromuscular disorders, 17(6), 470-5.
8. Gibson, B. E., Mistry, B., Smith, B., Yoshida, K. K., Abbott, D., Lindsay, S., & Hamdani, Y. (2014). Becoming men: Gender, disability, and transitioning to adulthood. Health. 18(1), 95-114.
9. Beresford, B. (2004). On the road to nowhere? Young disabled people and transition. Child: care, health and development.
10. Abbott, D., & Carpenter, J. (2014). 'Wasting precious time': young men with Duchenne muscular dystrophy negotiate the transition to adulthood. Disability & Society, 29(8),1192-205.
11. Hendriksen, J. G., & Schrans, D.G. (2011). Jonge de J. Video documentary "DMD with a Future–The power to live.
12. Birnkrant, D. J., Bushby, K., Bann, C. M., Apkon, S. D., Blackwell, A., Colvin, M. K., Cripe, L., Herron, A. R., Kennedy, A., Kinnett, K., & Naprawa, J. (2018). Diagnosis and management of Duchenne muscular dystrophy, part 3: primary care, emergency management, psychosocial care, and transitions of care across the lifespan. The Lancet Neurology, 17(5), 445-55.

13. Bushby, K., Finkel, R., Birnkrant, D. J., Case, L. E., Clemens, P. R., Cripe, L., Kaul, A., Kinnett, K., McDonald, C., Pandya, S., & Poysky, J. (2010). Diagnosis and management of Duchenne muscular dystrophy, part 1: diagnosis, and pharmacological and psychosocial management. The Lancet Neurology, 9(1), 77-93.
14. Schrans, D. G., Abbott, D., Peay, H. L., Pangalila, R.F., Vroom, E., Goemans, N., Vles, J. S., Aldenkamp, A. P., & Hendriksen, J. G. (2013). Transition in Duchenne muscular dystrophy: an expert meeting report and description of transition needs in an emergent patient population:(parent project muscular dystrophy transition expert meeting 17–18 June 2011, Amsterdam, The Netherlands). Neuromuscular Disorders, 23(3), 283-6.