

Report on the Psycho-Social Needs and Support in

Duchenne Muscular Dystrophy

The 'Cinderella' of DMD Care

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**University of
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**Duchenne
UK**

I. Introduction

It has been established that children with the genetic muscle wasting impairment Duchenne Muscular Dystrophy (DMD) are at high risk of various psycho-social needs. These can be neuro-developmental such as Dyslexia, Autism Spectrum Disorder and Attention Deficit and Hyperactivity Disorder, or can present as mental health difficulties, in particular, Anxiety, Obsessive Compulsive Disorder, and Depression (Banihani 2015; Caspers- Conway et al 2015; Ricotti et al 2016b) . Furthermore, there can be challenges for both people with DMD and their families with regard to adjustment to living with a physically deteriorating condition (Gocheva et al 2019). In 2018 the updated International Care Considerations for DMD identified different overlapping psycho-social needs and gave advice on best practice intervention (Birnkrant et al 2018). However, these international guidelines were led by clinicians from the United States and focus on supporting young people with DMD and their families in a North American context. In January 2021, over 160 families affected by DMD in the UK completed Duchenne UK's Standards of Care survey reporting that the area of psycho-social needs is the most poorly supported in DMD. Duchenne UK is currently adapting the International Care Considerations through its programme DMD Care UK so that they reflect experiences of families and support available in the UK.

This small-scale project provides insight into the psycho-social needs faced by children and young people (CYP) and adults with DMD and their families in the UK. It explores support mechanisms and barriers to resources that families and adults with DMD experience. It will help to ensure that the DMD Care UK Guidelines for psycho-social needs are relevant and useful and reflect the needs and experiences of people with DMD and their families.

Aims of this project

1. To explore the psycho-social challenges that children and adults with DMD experience
2. To explore barriers and solutions facing children and adults with DMD who experience additional learning and behavioural difficulties
3. To explore mechanisms for receiving support and resources to help additional needs in DMD.

II. Procedure and Participant Characteristics

Ethical approval was provided by the University of East London to conduct a small-scale mixed methods study with parents of children and adults with DMD (n=29), adults with DMD (n=2), and neuro-muscular clinicians (n=4). Recruitment of parents and adults with DMD took place via email through Duchenne UK's and Decipha CIC 's CRM systems and via their social media platforms. Clinicians were approached through Duchenne UK's clinical network. All carers/ adults with DMD participants completed an online questionnaire and took part in online focus groups which were organised according to the age of the young person/adult they cared for, or their own age in the case of adults with DMD. Four clinicians agreed to take part in individual online interviews.

Key participant information from the questionnaire and focus groups:

31 participants completed the questionnaire: 27 parents/carers of children and young people (CYP), two parents of adults with DMD, and 2 adults with DMD who were over 40 years. All lived in the UK and all parents cared for at least one child/young person with DMD

Ethnicity:

84% of participants identified as White

16 % of participants identified as Asian or British Asian.

Geography:

2 participants from Wales, 1 participant from Ireland;1 participant from Scotland; 26 participants from England

Highest Level of Education in household:

87% of participants had a diploma in Higher Education or above, and out of these, 74% were educated to undergraduate degree level. 13% of participants reported being educated to GCSE level or below.

Age of people with DMD and organisation of Online Focus Groups:

Focus Groups were organised by age of the person with DMD and took place online via Microsoft Teams. All groups followed an agreed schedule of questions and took between sixty and ninety minutes and all were transcribed verbatim.

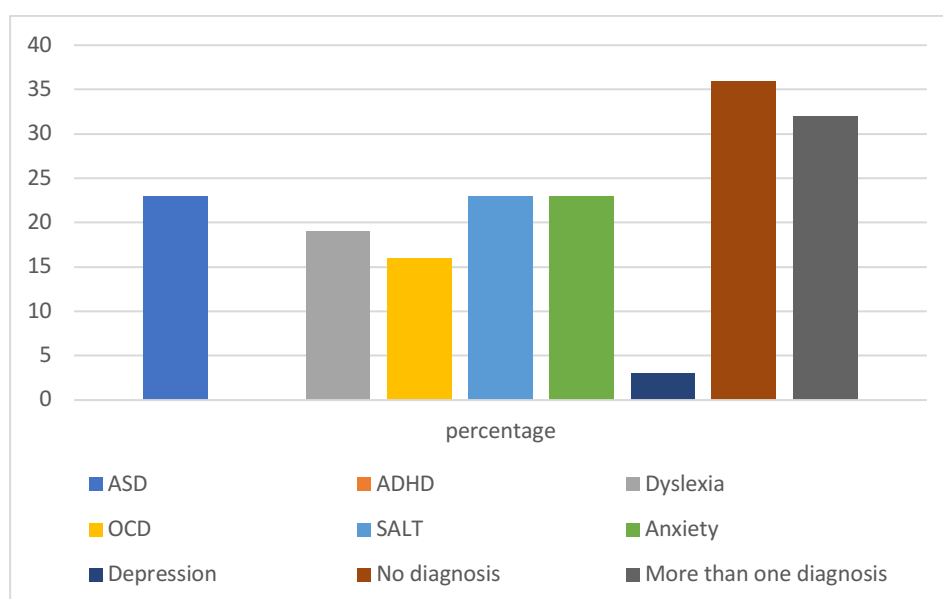
Group 1 aged 0-6 years (n=5)	16%
Group 2 (n=8) and Group 3 (n=10) aged 7 – 12 years	58%
Group 4 aged 13 – 19 years (n=4)	13%
Group 5 aged 20 – 52years (n=4)	13%

Key Information regarding Clinician participants:

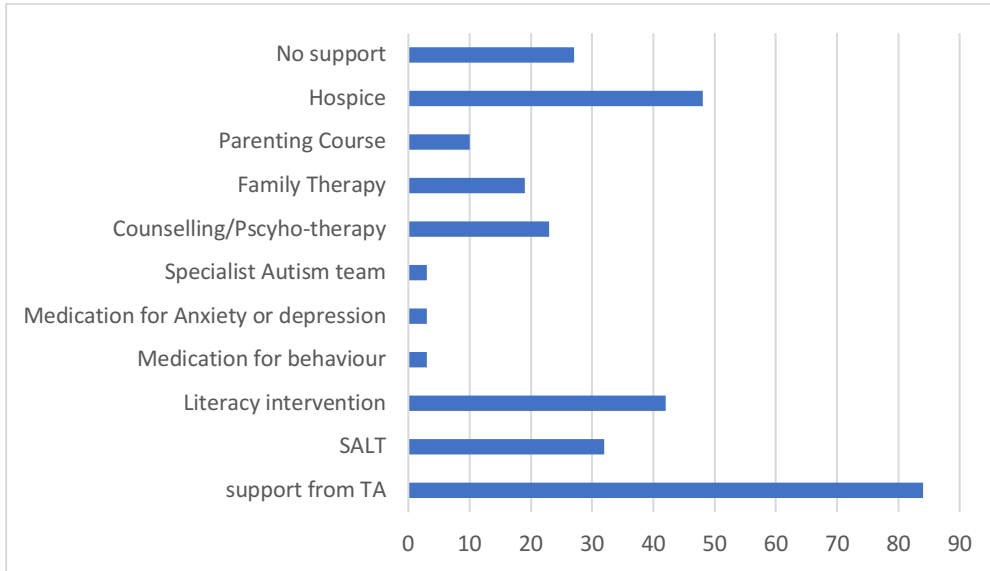
The four participants were Paediatric Consultants at four Neuromuscular Centres of Excellence in England.

III. Key Statistical Findings from Questionnaire

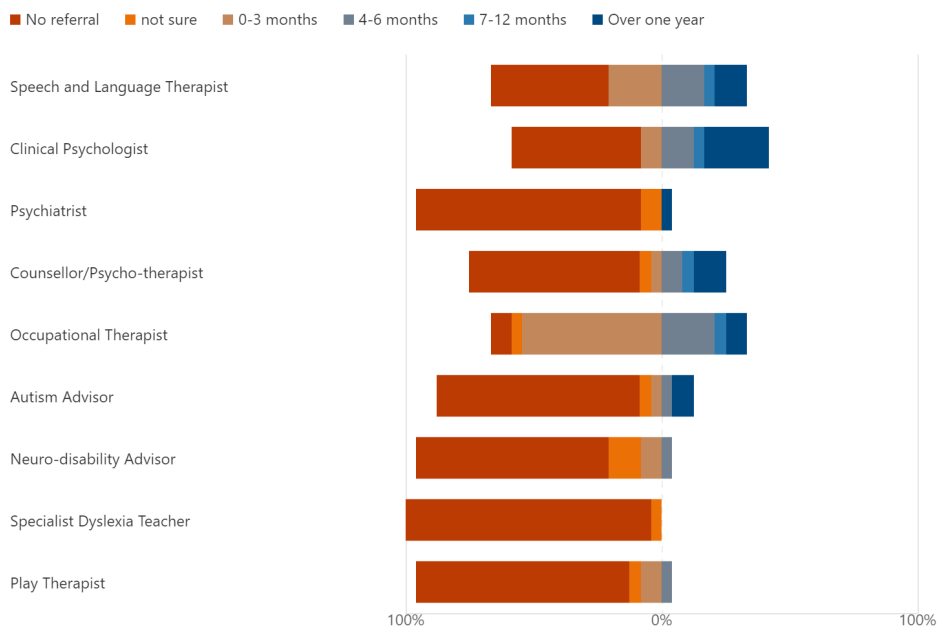
1. Reported Psycho-social diagnoses by percentage (n=31)



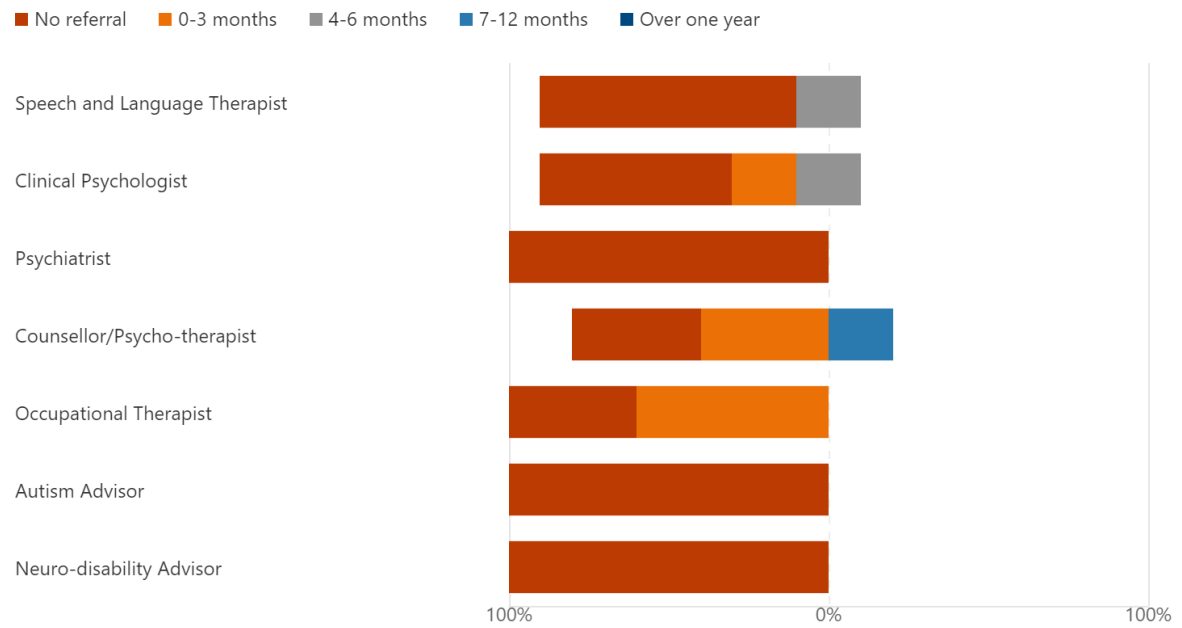
2. Support /Interventions offered by percentage (n=31)



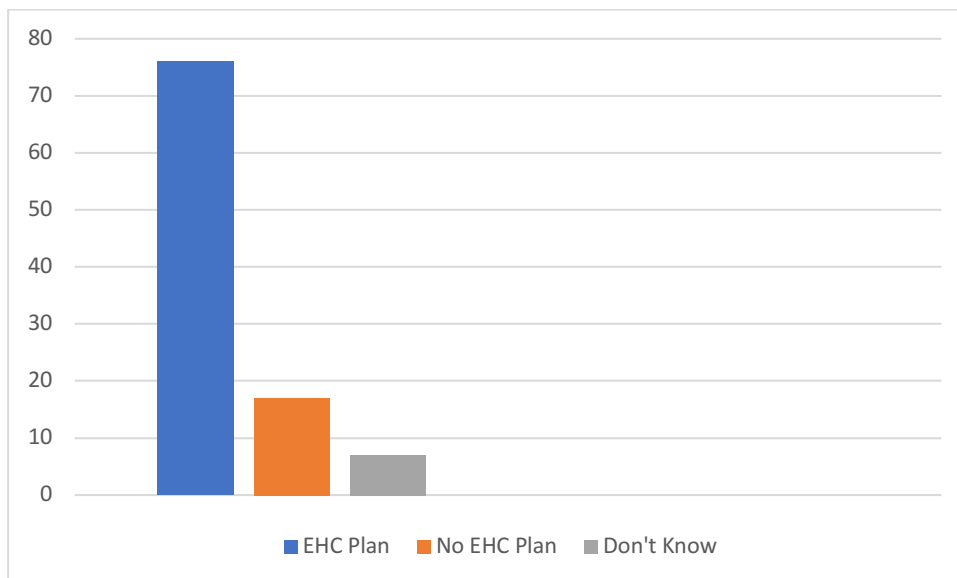
3. Waiting Times for psycho-social referrals in paediatric services by percentage (n=24)



4. Waiting Times for psycho-social referrals in adult services in percentage (n=5)



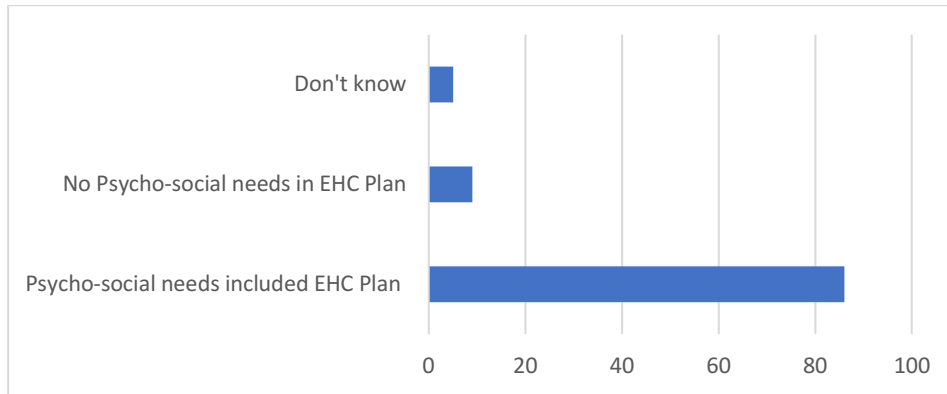
5. Education Health and Care Plans in percentage (n=29) *



*only available in England for 0-25 years, finishes at end of formal education

6. Education Health and Care Plans including Psycho-social needs in percentage

(n=22)



IV. Key themes and sub-themes from Parent and Adults with DMD

focus groups

1. Impact of learning and behaviour (neuro-biological) risks in DMD on young person and family

Lack of acknowledgement of neuro-biological differences and the challenges they can bring

- Parents of CYP with inflexible behaviour/autistic traits report high levels of stress *'Autism issues are more stressful than the Duchenne diagnosis'*. Some CYP experience severe meltdowns and several argue with teachers if they believe they (or anyone else in class) are being treated unfairly. Others present with rigid and inflexible behaviour *'The atmosphere of the house depends on P's mood'*
- Lack of discussion with doctors of neuro-biological risks and little known by teachers and other professionals.
- Difficulties getting school to understand the various difficulties around behaviour – especially in secondary school where there are many teachers/staff. Parents report being *'fed up of being **that** person'* and reported schools thinking *'here she comes again!'*
- There is a lack of acknowledgement in DMD community that CYP with DMD can have a learning disability or global delay.

2. Adjustment to deteriorating nature of DMD and widening gap with non-disabled peers

Increase in isolation and mental health difficulties as CYP with DMD reaches adolescence

- Higher levels of isolation as CYP becomes adolescent from friends due to lack of access to people's houses and inability to keep up with peers. This impacts the wider family as well as the CYP with DMD who also get cut off from wider family / friends events etc
- Issues around inclusion at secondary or special school '*he doesn't fit in anywhere*', reduced friendships and social life, schools' inability to include in lessons or make adjustments in playground.
- Overlap with autistic or OCD traits, for example, some repetitive, inflexible or anxious behaviour worsens as the CYP became an adolescent.
- Increase in mental health problems over time, '*he used to be the life and soul of the party*' but now refuses to leave house.
- A minority of parents of adolescents with DMD report their children mentioning suicide and self-harm with some CYP asking questions like '*What's the point in living?*'
- Several parents report the CYP viewing the wheelchair as a social stigma and would not use it. For one young person this meant leaving college as soon as he was unable to use the stairs as he refused to use the lift.

- Widened perceived gap between CYP with DMD and non-disabled peers as CYP gets older, eg social life/ not driving a car/ having a girlfriend etc.

3. Non-existent Psycho-social Support Services

Lack of support for mental health and anxiety – for CYP and whole family

- Lack of any emotional support at diagnosis for parents and /or siblings.
- Lack of emotional support for young people with DMD . Negative experiences of CAMHS eg not meeting criteria or having cases closed when young person would not engage despite severe difficulties.
- Some families report useful family therapy and temporary counselling / psychotherapy through their neuro-muscular centres but this is not viewed as a permanent arrangement. Comparisons made to other conditions such as cystic fibrosis and cancer where services buy in counselling/psycho-therapy services.
- Some families pay privately for counselling/psycho-therapy, up to £100 per hour.
- Lack of respite for families– children and adults not meeting criteria for hospices as they grow older, and parents tired of ‘fighting’
- Education Health and Care(EHC) plans can work but are dependent on the people who implement them

4. Feelings of Hope

Strategies that work for CYP and family

- Supporting the CYP with DMD to develop their hobbies from a young age, especially those that can still be experienced after loss of ambulation, eg embracing disability sports such as wheelchair football, or computing skills.
- Counselling and Cognitive Behavioural Therapy, although most groups reported a trained psychologist wasn't essential but rather someone who *'gets'* DMD and is a good listener.
- Some breathing and mindfulness activities for parents and children
- Adults with DMD report the importance of keeping busy, finding a job or volunteering. This was particularly so at end of formal education when it can be easy to play computer games all day. Both adult participants reported value of supporting younger CYP with DMD and importance of role models.
- Meeting other parents is viewed as essential and this occurs through user-led organisations and/or social media pages and fora.
- Effective communication between parents, professionals and school (although often difficult to achieve) is key.

V. Key Themes and subthemes from Clinician Interviews

1. The area of Psycho-social needs is '*the biggest unmet need*' in DMD

Psycho-social needs are the '*Cinderella*' of DMD care

- Health teams now support most CYP with DMD to transition to adult services but clinicians want to support CYP's quality of life as well
- There is a need for a standardised assessment for Psycho-social needs with agreed outcome measures– (like the North Star Assessment).
- CYP with DMD can present with a range of different needs and impairments and without appropriate support can reach crisis levels.
- Neuro-muscular consultants want a dialogue with psychiatrists with regard to medications for eg. ADHD and Anxiety and other psycho-social needs

2. Families need support at diagnosis and at other key moments as 'routine' so that psycho-social support is normalised

There is a need for a clear psycho-social roadmap for DMD

- Support at diagnosis is key. Some families find it impossible to move on from the diagnosis
- Clinicians can find it difficult to suggest another diagnosis as well as DMD, especially if CYP is recently diagnosed. Time can also be an issue to discuss this in depth at appointments.

- Families and CYP would be better engaged with the clinical team if mental health support was part of the routine offer for CYP with DMD and their families.
- As well as the need for expert psychological services, all neuro-muscular professionals can be trained to listen and support effectively.

3. Lack of clear and timely referral routes stop CYP and families getting the support they need

An over-complicated and under-resourced system

- The referral route can be extremely complicated and you often need psychological expertise and /or experience to complete
- All clinicians interviewed would prefer to have psycho-social support within their own team and/or within their hospital
- The postcode lottery of support can mean that families who see the same Neuro-muscular team may struggle to get parity of support from local services
- Health trust politics can mean that families may have to be on a waiting list or wait a long time to be seen
- It is acknowledged that Camhs is 'broken' and under-resourced.

VI. Recommendations:

1. Psycho-social difficulties need to be acknowledged by the Neuro-muscular team and discussed with families at all clinic appointments. A standardised screening / assessment tool needs to be used alongside that for physical health.
2. Emotional support needs to be available for parents at diagnosis and for a CYP with DMD at key points of their life eg. transition to primary/ secondary school, when major physical changes occur such as loss of ambulation, so that mental health support is normalised.
3. Mental Health support should also be available for CYP and family as and when they need it from a team that understands DMD, not necessarily CAMHS. The Neuro-muscular team should be able to refer for or offer this rather than families having to fight (or pay) for it.
4. More focus needs to be placed on the experience of CYP reaching adolescence and appropriate intervention, in particular psycho-therapeutic support.
5. Improved and formalised communication between the neuromuscular clinic and school and other local services is necessary for CYP to gain necessary referrals and resources.

6. More opportunities need to be offered for families and adults with DMD to share what works regarding psycho-social needs.

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