

## Workshop Report

# Workshop report: Workshop on psychiatric prescribing and psychology testing and intervention in children and adults with Duchenne muscular dystrophy

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## Abstract

This workshop aimed at summarising knowledge and key issues in psychiatric prescribing and psychological testing in children and adults with Duchenne muscular dystrophy (DMD). It comprised clinicians and patient representatives from the UK and the Netherlands. The following topics were discussed: a model for capturing the range of non-motor problems in the domains of cognition, learning, emotion and behaviour; psychosocial screening tools for use with children and adults; assessing neurocognitive functioning in children and adults; parent and teacher perspectives on psychosocial needs; and psychopharmacological treatment for affective disorders, anxiety disorders, obsessive

compulsive disorder, attention deficit hyperactivity disorder (ADHD) and insomnia. Some key considerations included: the need for tools used to assess behavioural and psychosocial functioning to consider motor aspects in DMD; to understand more about working memory performance; the need for early interventions for automatised problems, which affect reading and arithmetic; appropriate selection of tests for neuropsychology assessments; in schools, acknowledging the range of psychosocial risks and gathering evidence of psychosocial needs; the suitability of selective serotonin reuptake inhibitors for mood and anxiety disorders; the use of stimulant medications for ADHD; melatonin use for insomnia; the cautious use of benzodiazepines; and the need for improving pathways for psychosocial care.

## Keywords

Duchenne muscular dystrophy, psychosocial, psychiatry, prescribing, neuropsychology

## Introduction

On 31 January 2023, a workshop on prescribing in psychiatric disorders and psychology testing in Duchenne muscular dystrophy (DMD) was convened by the 'Psychosocial Project Working Group' funded by DMD Care UK. The workshop brought together patient representatives, neuropsychologists, neuropsychiatrists, and neuromuscular, cardiology, respiratory, pharmacology and endocrine clinicians from neuromuscular centres in London and Newcastle, UK and experts from the Center of Neurological Learning Disabilities, Kempenhaeghe, the Netherlands. The aim was to summarise knowledge and key issues in psychiatric prescribing and psychological testing in children and adults with DMD and to highlight areas needing further attention.

## The need for improvements in DMD psychosocial care

The reported experience of many patients with DMD and their families is that they are rarely asked about their psychosocial needs, beyond questions about mood which often occur only as an appointment is ending. Clinicians often report being aware of psychosocial needs, but do not know where to refer the patient or how to best offer support. A common barrier experienced is around prescribing medication for psychiatric comorbidity, for example, for anxiety or depression. Neuromuscular clinicians do not have the confidence in making these prescriptions. Psychiatry specialists are similarly affected as few have experience of working with this patient group. This has been described as a double jeopardy, and patients caught in this situation can find themselves bounced between different clinicians and their GP with no clear route to the care they need.

Beyond medical diagnosis and treatment is the need to improve the psychological and social care of boys and men with DMD across the lifespan to help improve their quality of life, as indicated in the Standards of Care (SoC) (Birnkrant et al. 2018). Care for DMD

needs to be holistic and adopt a biopsychosocial approach rather than a solely medical perspective (Morrow 2004).

## **Behaviour and learning issues in DMD**

Dr Jos Hendriksen reported on the steady increase in the number of scientific articles on the non-motor problems such as learning, behaviour and cognitive problems in DMD in the last 25 years. In clinical practice, the main concerns reported by parents are learning and behaviour issues (Hendriksen et al. 2020).

## **No consensus in assessment of non-motor problems**

Dr Hendriksen described the current difficulty with knowing which mental health assessment tools to use in the DMD population. Currently, there is no consensus on which tools are most appropriate for assessing non-motor problems in DMD. A review of tools used for assessing behavioural and psychosocial functioning across all ages in DMD and Becker muscular dystrophy (BMD) found that 61 instruments were used across 54 studies (Hellebrekers et al. 2019). However, the most commonly used instruments are not designed with DMD in mind and do not take into consideration the motor and physical aspects found in boys with DMD. This can result in false negative assessment results for attention deficit hyperactivity disorder (ADHD) for example, with comorbidities therefore being overlooked and under-reported.

## **Brain involvement**

The interest in the impact of dystrophinopathies on the brain is a burgeoning area. The original work of the French neurologist, Duchenne de Boulogne, who first described DMD, already made reference to neurocognitive comorbidities, such as low intelligence, epilepsy and obsessive behaviour. Despite this early recognition, treatments for DMD and clinical trials have been directed at skeletal muscle, the heart and other organs, and have tended to neglect brain-related comorbidities. However, there is now growing interest and research into brain involvement in the dystrophinopathies (Hendriksen et al. 2020). An EU multicentre study is currently underway looking specifically at brain-related comorbidities in DMD (BIND study; [www.bindproject.eu](http://www.bindproject.eu)). Part of this work will be to develop questionnaires likely to be helpful in future for screening for many non-motor comorbidities.

## **Heuristic framework**

There is a range of neurocognitive, neurobehavioural and neuropsychiatric difficulties associated with DMD. Alongside these difficulties, there are also cognitive strengths, such as visual processing and receptive vocabulary. Dr Hendriksen presented a heuristic model – the 'Big 10' – to capture this complex overlap of different brain-related diagnoses. He and his team identified 10 areas of difficulty considered important in understanding learning and behaviour functioning in DMD (Hendriksen et al. 2020). The model categorises problems in

four domains: cognition (automatisation, working memory, and attention), learning (dyslexia and dyscalculia), emotion (anxiety and depression) and behaviour [autism, ADHD and obsessive compulsive disorder (OCD)] (Figure 1). Whilst the domains are described separately, in clinical practice there is a complex overlap amongst the 10 areas and it is rare to see an individual presenting with just one difficulty. The model can help clinicians to consider hidden areas of seemingly minor problems, which may co-exist with the more obvious major problems. As such, it was proposed that the Big 10 model may help as a tool to better define and characterise the nature of problems to be considered. Dr Hendriksen is working with Duchenne Parent Project to operationalise this further.

Fig. 1

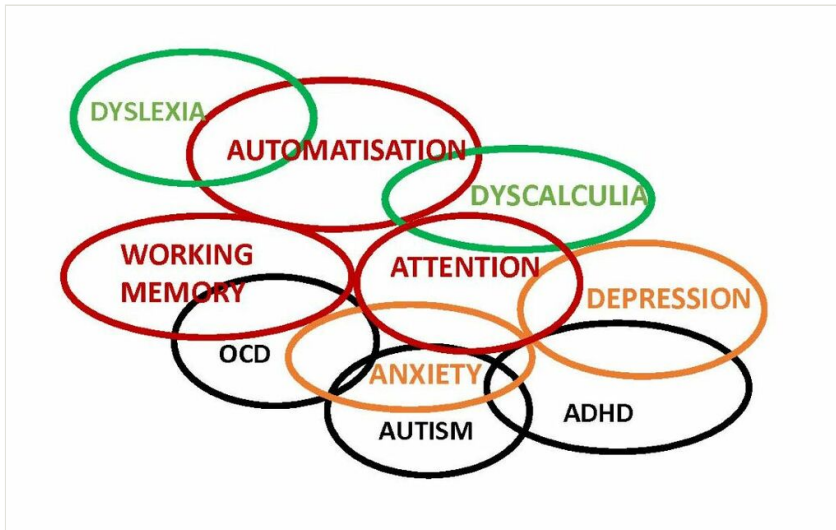


Figure 1. [doi](#)

The Big 10 model of non-motor problems in DMD (kindly provided by Dr Jos Hendriksen).

## Psychosocial adjustment

Dr Hendriksen described psychosocial adjustment as being about coping and stress, which is related to perception. He gave an example of the differing perspectives boys with DMD may have on obtaining their first powered wheelchair: some may perceive it as a loss of function, whereas others may perceive it liberating, enabling them to participate in more activities. People with DMD face a range of physical losses across their lifespan, such as the loss of walking, arm function and breathing. Fatigue, poor endurance and bodily pain are common daily experiences. They are also faced with psychological stresses, such as learning to ask for help and having to wait for help. These are important psychological skills to develop when they are young to support their psychological adjustment.

An easy-to-administer instrument that has been validated in DMD populations to screen for psychosocial adjustment from childhood into adulthood is the Personal Adjustment and

Role Skills Scale (PARS), which is recommended in the SoC for parents of children aged 5-17 years (Birnkranz et al. 2018). The validity and reliability of the tool for use in boys with DMD was reported 14 years ago (PARS-III) (Hendriksen et al. 2008). Since then, as part of the BIND study, the instrument has been developed for use with adults (PARS-A) (Weerkamp et al. 2022a).

Dr Hendriksen presented unpublished data on using the PARS-III to look at psychosocial adjustment over age. As boys with DMD grow older, he found that their adjustment improved. Psychological adjustment levels were the lowest at ages 6 to 10 years possibly because this is the phase during which they must accept and adjust to the diagnosis and its consequences.

Dr Hendriksen highlighted another important psychosocial issue to consider: learned helplessness, which may lead to ineffectiveness, apathy and not doing anything. Boys with DMD are at risk of learned helplessness because they may put a lot of effort into doing something without achieving a tangible result. He gave the example of having intensive therapy with leg braces which did not lead to improvement. As the boys do not experience tangible results from their performed actions, it is important to create an 'action-effect-affect chain', whereby you do something, which has an effect, which then results in an affect or emotion. This approach can be useful to help build self-esteem and prevent learned helplessness.

## Neurocognitive functioning

Dr Hendriksen discussed what is known about neurocognitive functioning in DMD, in the areas of intelligence, working memory and automatisations.

A review of 32 studies between 1960 and 1999, comprising 1224 boys with DMD, found that intelligence levels fell within the low average range (FSIQ of 80) (Cotton et al. 2001). However, in this review, eight different instruments for measuring IQ were clustered together and analysed. Dr Hendriksen and colleagues re-analysed all the data published on intelligence testing in adults and boys, but looked only at those studies using the Wechsler intelligence scales, which are the most used intelligence tests from childhood to adulthood. Data analysed from 43 studies, with 1427 boys and men, from 1960 to 2022, confirmed that the average IQ is below average (FSIQ of 84) (Weerkamp et al. 2022b).

Assessing general intelligence in isolation may underestimate a patient's potential. Rather, there are cognitive strengths and weaknesses. In clinical practice, Dr Hendriksen noted many patients have strong visuospatial performance despite their motor handicap. Visuospatial functioning and spatial thinking are often areas of cognitive strength in DMD, as well as receptive vocabulary.

Working memory is a core neuropsychological function. The bottleneck theory of working memory states that all information must be transferred via working memory from short- to long-term memory. It is involved in storing information efficiently and being able to retrieve it. In boys with DMD, working memory is thought to be a core cognitive weakness. This

may present as difficulty in retelling a story and difficulty remembering multiple instructions. Teachers often report that the boys are not motivated, while in fact they are not able to quickly switch their attention to move on to the next task if they have working memory problems.

Dr Hendriksen's research has shown there may be a decline in working memory performance over time (Hellebrekers et al. 2020). However, he noted that this is an area that needs more attention, especially in relation to the genotype and mutations which are linked to working memory. The BIND study may shed light on this question.

An important question is whether training can help improve working memory problems. Dr Hendriksen summarised work from his group which showed that computer training can result in an improvement in working memory and in speed of information processing (Hellebrekers et al. 2022). The intervention involved 30 minutes of training across five days a week, over six weeks. This is an area that needs more attention and more research.

Automatisation is the quick and automatic decoding of signs into sounds and is required for reading and arithmetic. Boys with DMD can have problems with automatisation, which can present as dyslexia and reading problems. Higher rates of dyslexia are found in boys with DMD (27%) compared to the general population (3%) (Hendriksen and Vles 2006). It is important to provide early interventions for dyslexia and arithmetic capabilities so that boys can continue to develop their vocabulary, learn to ask questions, and develop literacy and numeracy skills that are important for adult independence. Tests that can help identify problems with automatisation include those measuring the speed of reading (e.g. word lists) and the speed of naming numbers (e.g. dot counting).

## **Neuropsychiatric comorbidity**

Where the presence of one condition interferes with the recognition of others, it is known as diagnostic overshadowing. DMD can easily overshadow the Big 10 problems. In a recent meta-analysis on five neuropsychiatric disorders (of which five are included in the Big 10: ADHD, autism, OCD, depression, anxiety), there was great variance in prevalence rates of each condition, which could indicate that some of these conditions are being overlooked (Pascual-Morena et al. 2022).

## **Parent and teacher perspectives on psychosocial needs in DMD**

Dr Janet Hoskin, Associate Professor of Education, presented findings from a questionnaire and focus groups involving 29 parents of children and young people with DMD and two adults with DMD, as well as interviews with four neuromuscular clinicians (Hoskin 2023).

Questionnaire findings showed a range of psychosocial need: 36% of participants reported no psychosocial diagnoses, whereas 23% self-reported autism, 23% anxiety, 23% speech and language difficulties, 19% dyslexia, 16% OCD, 3% depression, and 32% reported

more than one diagnosis. There were no reported diagnoses of ADHD, suggesting that ADHD may be under-diagnosed, given the reported prevalence rates elsewhere.

Several key findings emerged from the focus groups and interviews. Firstly, parents reported that school would be improved for their sons if psychosocial risks were better acknowledged. Clinicians suggested that one reason for this was the lack of appropriate assessment tools. Secondly, parents reported that their sons' mental health deteriorated as they became teenagers and were not included in friendship groups or able to do the same activities as their friends. Moreover, clinicians highlighted how comparison with non-disabled peers strongly affected the mental health of parents. Thirdly, a need for a 'psychosocial road map' was identified. The crisis points in the trajectory of young people and families with DMD are well-known (Porteous et al. 2021), yet support is often offered too late, if at all. Fourthly, inflexible systems and structures mean that referrals to psychosocial provision for assessment and support either do not happen or are very delayed. Both parents and clinicians reported that some neuromuscular teams have excellent psychological departments within their own trusts to where children with other chronic conditions, such as diabetes or cystic fibrosis, are referred. Furthermore, parents reported very poor experiences with Child and Adolescent Mental Health Services (CAMHS). Finally, successful psychosocial strategies and interventions were identified such as supporting young people's interests. Men with DMD reported that the most effective strategy for good mental health was to have something meaningful to do with their time, such as employment or volunteering. This highlights the importance of early intervention to support psychosocial needs, such as literacy and numeracy skill development.

In addition, Dr Hoskin reported on the work of Decipha CIC during 2022 which is an organisation that supports the Special Educational Needs (SEND) of children with DMD. In particular, Decipha supports families and schools to obtain Education Health and Care Plans (EHCPs). These are statutory documents that all children with DMD should be given and include provision to meet the needs of children with Special Educational Needs at school and college. Obtaining a plan can sometimes be contentious, and much of Decipha's work involves supporting families to attend tribunals with Local Authorities. Letters from neuromuscular clinicians carry a lot of weight in the EHCP process and it was suggested that these should reflect the four areas of the SEND Code of Practice (Department for Education and Department for Health and Social Care 2015). The four categories of SEND are: communication and interaction; cognition and learning; social, emotional and mental health; and sensory and/or physical. In schools, the sensory and physical issues tend to be well addressed, whereas difficulties in the other three areas are more likely to be overlooked or not referred to specifically in clinician letters. Sharing the established risks in DMD would be helpful for schools to gather evidence of need and to refer children for appropriate assessment.

## **Psychopharmacological treatment in children and adults with DMD**

### **Clinical and published evidence on psychopharmacological treatment in DMD**

Dr Phillipe Collin, Child and Adolescent Psychiatrist, presented the evidence from a recent scoping review of psychopharmacological treatment in neuromuscular disorders, which reported on five published papers in DMD (Brusa et al. 2022). Two were case reports, two case series and one was an observational study. Most patients were under 18 years, but one study included patients up to age 23.

Included in the review was the first case report of the use of the selective serotonin reuptake inhibitor (SSRI) fluoxetine to treat OCD in a boy with DMD (Hendriksen et al. 2016). They reported a clinically significant and positive effect of fluoxetine in a boy with DMD and co-existent OCD and autism.

A case series of 15 DMD patients, aged 5-23 years, with OCD (Lee et al. 2018) found a clinically significant improvement in 10 of 15 patients using drugs from the SSRI class, including fluoxetine, paroxetine and citalopram. One patient of 15 reported rash, gastrointestinal upset and apathy. Urinary urgency was noted with all SSRIs but paroxetine.

Observational data from 10 boys with DMD and ADHD aged 6-10 years using the stimulant methylphenidate up to a dose of 0.2-0.6 mg/kg/day have been described (Lionarons et al. 2019). In the long term, seven of 10 patients showed clinically significant improvements in inattentive symptoms. There were no reported side effects.

A large retrospective case series reported on 700 DMD patients found high rates of neuropsychiatric symptomatology, primarily emotional/behavioural dysregulation and inattention in more than a third of patients and obsessive-compulsive symptoms in 25% (Darmahkasih et al. 2020). Almost a third required pharmacological intervention and, in this study, the main drug type used was SSRIs, mostly fluoxetine.

Prescribing data from the Center of Neurological Learning Disabilities, Kempenhaeghe was then described. Dr Collin noted that referrals to the centre tend to involve more complex cases due to its national specialism in DMD.

Between 2015 and 2022, the centre treated 35 male DMD patients, all under 18 years. Seven boys were diagnosed with autism (two also with OCD and two with behavioural problems). Fifteen were diagnosed with ADHD, seven with a comorbid behavioural problem, OCD was diagnosed in five patients, and an anxiety disorder in three patients. There were four boys diagnosed with mood disorder, one of them with comorbid OCD and one a comorbid anxiety disorder. One medication was prescribed in 28 cases. Seven patients were given more than one medication, usually to treat comorbidity. The most



commonly prescribed medication was the stimulant methylphenidate (primarily used to treat ADHD), given to 22 patients. In two patients, an alternative stimulant, dexamphetamine or its long-acting pro-drug lisdexamphetamine, was used. The non-stimulant drugs clonidine, guanfacine and atomoxetine, also used to treat ADHD, were used on three occasions. Only methylphenidate showed a significant positive effect, which was observed in 10 of 22 patients where it was prescribed. Drugs from the SSRI antidepressant/anxiolytic class were prescribed on eight occasions, with fluoxetine most commonly used (in six patients). In five of the six patients prescribed fluoxetine and in the one patient prescribed escitalopram, a significant positive clinical improvement was noted. The most commonly prescribed antipsychotic medication was risperidone, primarily used for behavioural disturbance, in six patients. One of those patients showed a significant clinical improvement. Benzodiazepines (alprazolam and oxazepam) were prescribed on three occasions, with significant positive effect in the two patients prescribed alprazolam for anxiety.

Regarding safety aspects of prescribing in DMD, Dr Collin reported that all medication was discussed with the cardiologist treating the patient before it was prescribed. In all cases, the cardiologist was happy for treatment to be started. There were no incidences of cardiac side effects in the 35 cases treated. The side-effect profile of the commonly used medications, for example, SSRIs such as fluoxetine and the stimulant methylphenidate, appears to be similar to general psychiatric patients under 18 years. Notably, risperidone was often associated with weight gain and methylphenidate with aggression in some patients.

Data of psychopharmacological treatment in a larger sample of DMD patients from Kempenhaeghe and Leuven University Medical Centres have been combined and were recently published (Weerkamp et al. 2023).

## **Psychopharmacological treatments: evidence from the general population**

Dr Rory Conn, Consultant in Paediatric Liaison Psychiatry, and Dr Dorothea Bindman, Consultant in Adult Neuropsychiatry, presented evidence on the use of medications commonly prescribed in psychiatric disorders.

### ***Selective serotonin reuptake inhibitors (SSRIs)***

SSRIs are first-line treatment for the majority of affective (mood and anxiety) disorders. In most cases, effectiveness within the SSRI class is broadly similar, but side effects, interactions and half-life vary. Despite the comparable efficacy, there may be some benefit to switching to another SSRI if the first one prescribed is ineffective.

The National Institute of Clinical Excellence (NICE), The Royal College of Psychiatrists and the British Association for Psychopharmacology recommend the use of antidepressant medication (initially an SSRI) in adults with depression of at least moderate severity according to ICD-10 criteria or in mild depression of a duration of greater than two years and in anxiety disorders (including Generalised Anxiety Disorder, Panic Disorder and

Social Anxiety Disorder). If first-line therapy fails, there may be some benefit to switching to another SSRI before trying an antidepressant of a different class, such as a serotonin and norepinephrine reuptake inhibitor (SNRI) (e.g. venlafaxine).

SSRIs are also the pharmacological treatment of choice in OCD. In that case, treatment response takes longer to emerge (12 weeks vs. 2-4 weeks) and may not become apparent until the dose is titrated to the British National Formulary (BNF) maximum.

### **Cardiovascular risks of SSRIs**

In citalopram and escitalopram, small decreases in heart rate and systolic blood pressure may be seen. A dose-related increase in QTc interval and Torsade de Pointes have been reported, mainly in overdose. There is no evidence of conduction disturbance. Caution is advised in patients with recent myocardial infarction or uncompensated heart failure, but there is some evidence of safety in patients with cardiovascular disease. There is no clear evidence of increased risk of arrhythmia at any licensed dose.

Sertraline and fluoxetine show minimal effects on heart rate and blood pressure. They have no effect on QTc interval at standard doses and there are no reports of arrhythmia or conduction disturbance with either drug. Sertraline is the drug of choice post myocardial infarction, but formal labelling acknowledges an effect on QT interval and cautions against use in patients with additional risk factors for QTc prolongation.

### **Other risk issues**

Hyponatraemia can occur with all antidepressants. Patients with CYP2D6 enzyme deficiency are at higher risk, as are those co-prescribed drugs such as angiotensin-converting enzyme (ACE) inhibitors.

All serotonergic antidepressants theoretically affect platelet function and thus may increase the risk of bleeding.

### ***ADHD medications***

Stimulant drugs (methylphenidate and dexamphetamine) are the first-line pharmacological treatment in children and adults diagnosed with ADHD. Effect sizes, of up to 0.8, are some of the highest of all medications used in psychiatric disorders. Non-stimulant alternatives are available, including atomoxetine, which in the UK is licensed for adult prescribing, and the  $\alpha$ -agonists guanfacine and clonidine, both of which are licensed for use in children but not in adults.

### **Stimulant medications for ADHD**

In children, methylphenidate appears to be the most efficacious and tolerable of the two stimulants. Dexamphetamine has the highest efficacy and acceptability in adults, its tolerability is equivalent to methylphenidate (Cortese et al. 2018).

NICE recommends either long-acting methylphenidate (e.g. Concerta XL) or lisdexamphetamine (Elvanse) as the first choice in children and adults with ADHD.

The Royal College of Psychiatrists and NICE advise that stimulants are initiated by a specialist in ADHD (Royal College of Psychiatrists 2023). Once titrated to a stable dose over 2-3 months, a formalised Shared Care protocol can be agreed with the patient's GP.

### **Cardiovascular risks**

On initiation of stimulants, small increases in heart rate (5-10 bpm), systolic blood pressure (3-8 mmHg) and diastolic blood pressure (1-14 mmHg) have been described (Mick et al. 2013).

There have been concerns regarding cardiovascular safety with stimulants, specifically ventricular arrhythmia and sudden cardiac death, but recent meta-analyses provide some reassurance. Zhang et al. (2022) showed no significant increase in relative risk of cardiac adverse events for the stimulant class of drugs. In a large population-based cohort, the incidence rate ratio for major cardiovascular and cerebrovascular complications in lisdexamphetamine was 1.10 (Forns et al. 2022). No association was found between stimulants and QT prolongation or Torsade des Pointes (Beach et al. 2018). Of note, patients with a history of inherited Long QT Syndrome were at higher risk of cardiac events (Zhang et al. 2015).

Cardiovascular history, family history, and measurement of blood pressure, heart rate and weight are recommended at baseline. An ECG and cardiology review are advised before starting treatment if there is a family history of structural heart disease or sudden, unexplained death, abnormalities on cardiovascular examination or the patient is on other drugs that cause QT prolongation.

Measurement of blood pressure, heart rate and weight (and height in children) should continue at regular intervals as long as stimulants are prescribed.

### **Other side effects**

Appetite suppression is commonly described in adults. A mean weight loss of 1.8 kg over 52 weeks in adults has been reported. Insomnia is frequently reported by patients when first starting stimulants, but sleep studies have found no change in sleep quality.

There is a very small increase in agitation, aggression and emergence of psychotic symptoms. Caution is advised in patients with a history of mania or psychosis.

### **Non-stimulant medications for ADHD**

An MHRA review on Atomoxetine found similar effects on blood pressure and heart rate to stimulants with the majority experiencing modest increases in heart rate and/or blood pressure (less than 10 beats per minute and 5 mmHg) (MHRA 2012). Increases were more

significant in around 10% of patients, implying a theoretical risk of cardiovascular complications.

Guanfacine and clonidine can cause hypotension and carry a risk of QT prolongation, but are generally safe in patients with cardiac disease. If stopped abruptly, they can cause a rebound hypertension.

### ***Hypnotics/sedatives***

Melatonin is available as a controlled release preparation in the UK to treat insomnia. In adults, its licence is restricted to patients over 55 years, but it is commonly prescribed off-licence in adults with neurological disorders. There is some evidence that it has cardio-protective effects, particularly in age-related cardiovascular disease (Opie and Lecour 2016).

## **Psychopharmacological treatments in patients with DMD**

### ***SSRIs***

On reviewing the evidence, the group agreed that SSRI antidepressant/anxiolytic medications are likely to be suitable for use in DMD as in the general population, but that review of a baseline ECG and discussion with the treating cardiologist would be recommended for children and adults with DMD before prescribing. Increased risk of gastrointestinal perforation and bleeding in some DMD patients was noted, as was the frequent use of drugs that act on the renin-angiotensin system (increasing hyponatraemia risk). Regular monitoring of blood electrolytes is advised where such drugs are prescribed.

### ***Stimulants and non-stimulants for ADHD***

A small study of the use of stimulants in DMD patients provides reassuring data regarding cardiovascular risk (Lionarons et al. 2019). As with SSRIs, review of cardiovascular status by the treating cardiologist is advised before medication initiation.

The other area of concern is appetite suppression and, in children, reduced growth rate. At present, the data do not suggest significant effects on weight and growth, but in children with DMD the effect of stimulants on growth may be masked by steroids. This may have implications in future as new steroid-like agents without appetite-stimulating properties are developed.

### ***Hypnotics/sedatives***

There is little published evidence regarding the use of these agents in patients with DMD, but based on the clinical experience of the working group, there are no concerns regarding the use of melatonin. Benzodiazepines should be used with caution in unventilated patients and prolonged administration should be avoided. However, once non-invasive ventilation

(NIV) has been initiated, the effects of respiratory depression are reduced if the benzodiazepine administration is timed to coincide with the patient using NIV.

## Psychological testing in adults

Dr Linda Bouquillon, Clinical Psychologist, spoke about the need for psychological testing in adults and provided suggestions for recommended routine care in this area. As referred to in the SoC (Birnkrant et al. 2018), it is important to understand the psychosocial needs across the lifespan and to be able to provide appropriate care for adults which can improve their quality of life. However, compared to children with DMD, there is much less known about the neuropsychological, psychosocial and psychiatric care needs in adults. As males with DMD are living longer, they are presenting with additional care needs and issues that are specific to adults. This may include vocational pursuits, wanting to become more independent, difficulties with community participation, increased physical disability, increased healthcare needs, greater impact on caregivers, non-compliance with more invasive treatments and end-of-life choices. Navigating many of these situations involves complex decision-making, utilising a range of cognitive skills. Hence, when considering the psychosocial needs of adults, it is important to understand the impact of any specific cognitive difficulties so that clinicians can support patients through these stages.

As noted earlier (see 'Heuristic framework'), the Big 10 of psychological/neuropsychiatric comorbidities are well studied in children. However, there are few published studies looking at these possible difficulties in adults. There is variability in the literature and further research is needed to understand more about the trajectory of these difficulties.

There is a similar scarcity of published research on the neurocognitive profile in adults with DMD compared to that in children. When cognition is assessed in adults, the data tend to be combined and analysed with those of children in the same study, which can sometimes cover a broad age range (e.g. age range 2-27 years in Cotton et al. (2001)). A problem with this is that it is impossible to determine whether there are changes in cognition with age. Only one known paper has looked specifically at cognitive skills in adults with DMD (Ueda et al. 2017) and this study involved only 15 people. They found that difficulties in sequentially processing auditory and visual information remained into adulthood, and that verbal comprehension was intact.

Dr Bouquillon suggested that some key outstanding questions for psychology testing in adults are:

- What is the prevalence of neuropsychiatric/neuropsychological difficulties in adults with DMD?
- Do the neurodevelopmental disorders present with similar or different difficulties into adulthood?

- How do some of these difficulties/disorders and cognitive impairments impact on the decision-making and choices in adulthood?
  - For example, clinicians noted that some individuals with autism, intellectual disabilities or OCD may struggle more when adjusting to new medical interventions, such as non-invasive ventilation.
- How do the difficulties and impairments impact on social participation and quality of life?
  - For example, if there are ongoing difficulties with social communication into adulthood, this may impact on participation in social groups.
- How can those people who are most in need of psychosocial support be identified?
- Are there particular instruments that are useful for this identification?
- What might appropriate support look like?

There is a general consensus that psychosocial support should be available to all those who need it (Birnkranz et al. 2018), whether to help improve social participation, quality of life or compliance with treatments. To support this aim, there is a need to understand more about the trajectory of neuropsychiatric and neurocognitive difficulties, alongside psychosocial care needs, into adulthood. With this in mind, the suggested recommendations for routine care for adults with DMD would ideally include: a psychosocial screening tool – both a self-report version and a proxy version for those unable to complete themselves; a neuropsychological assessment, as currently in the UK not everyone has an assessment in childhood; a neuropsychiatry review to be available; access to appropriate support following assessments; and clear pathways for onward referrals, potentially in the form of an algorithm.

Dr Bouquillon described a recently started research project in which she is involved that is seeking to understand more about the psychosocial care needs of adults with DMD. The three-year project, funded by Duchenne Research Fund, aims to profile the neuropsychiatric and neuropsychological disorders in adults and to evaluate the impacts of these difficulties on, for example, patients' quality of life and engagement with medical teams. They plan to do this through a psychosocial assessment with patients during their routine inpatient multidisciplinary (MDT) assessment and with external patients referred via the Adult North Star Network. Psychoeducation on why patients are being asked questions about their mood and psychological well-being will be part of the assessment. They will be made aware of the possible outcomes, such as psychological therapy, group work or a referral for a neuropsychiatry review. Assessments will be with the patient and with their carer. This assessment will include screening measures for ADHD, autism, OCD, depression, anxiety and caregiver stress, as well including instruments to assess quality of life, activities, adjustment and participation. The second phase will involve administering a neurocognitive screen to all patients as part of their inpatient MDT assessment. The neurocognitive screen will assess across the following domains: general intellectual ability (General Ability Index, as Full Scale IQ is not possible due to motor difficulties); reading and literacy; language, including naming and comprehension; memory and learning; visuospatial skills; executive functioning and attention; and information processing speed. Appropriate tests will be selected based on what is known about the individual's level of

intellectual functioning. Assessments will be administered with those with known intellectual disabilities, as it is important to obtain a picture of their strengths and weaknesses. This will be useful in helping the people supporting them and in maximising their participation.

The reasons for recommending psychology testing in adults with DMD are manifold. Some of the key aims are to better understand the neurocognitive, neuropsychiatric and psychosocial factors in adults and to understand the impact of these on adults' psychosocial functioning. Another aim is to be able to provide neuropsychological or neuropsychiatric support at specific times of need, for example, during transition or when adjusting to new medical interventions. A further aim is to inform the clinical care team around the person with DMD on how best to treat and care for the patient. Whilst initially this work may be undertaken in a local service, the plan is to be able to spread the recommendations nationally and to implement better pathways of care to support the psychosocial care needs of all adults with DMD.

## **Psychology testing in children**

Dr Chloe Geagan, Clinical Psychologist, presented on mental health screening tools for use with young people, outlined a comprehensive neuropsychology assessment, and discussed how the assessment findings can inform areas of psychosocial care.

There are multiple areas where neuropsychology or psychology input can be effective for supporting psychosocial needs in children with DMD and their families. These include psychotherapy for patients and families; cognitive/neuropsychology testing; cognitive rehabilitation, which might involve liaison with schools; end-of-life issues, including anticipatory grief; staff support; school liaison; group work, with young people or families; and research and service development.

## **Service mapping**

It is currently unknown how many young people with DMD in the UK have had a neuropsychology assessment. To understand this more, a service mapping initiative has been started. Findings so far show there are some psychologists in neuropsychology services in the UK who are able to see young people with neuromuscular conditions. However, a large number of families in other regions of the UK are unable to access comprehensive neuropsychology assessment due a lack of funding in these services. A further aspect that is being explored is whether educational psychologists are offering assessments for informing EHCPs and, if so, to ensure that the four different areas to which Dr Hoskin referred are included, especially cognition.

## **Psychosocial screening tools**

Dr Geagan proposed screening questionnaires that could be of use in paediatric neuropsychology teams. It is important to have parent/caregiver reports and teacher

reports, as well as self-reports from the young person with DMD. The process of asking the young person to complete self-reports on how they are feeling should start when they are quite young, within the clinical setting.

The proposed screening measures referred to in the SoC, and also being used in the BIND study, are the PARS-III, as discussed by Dr Hendriksen, and the Strengths and Difficulties Questionnaire (SDQ) (Goodman 2001), which looks at emotional symptoms, behavioural difficulties including hyperactivity, peer relationship problems and prosocial behaviour. Dr Geagan shared a provisional pathway for psychological intervention after administering the SDQ, which is modelled on other stepped care models being implemented in paediatric epilepsy services (George et al. 2021).

## Neuropsychology assessment

The current recommendations in the SoC are that “neuropsychology evaluations should be considered within the first year of diagnosis to establish a baseline” and that “re-evaluations should be done every 2-3 years to monitor developmental progress and response to interventions” (Birnkrant et al. 2018). Clinical judgement should be used to decide when to re-evaluate as, for some young people, this might not be necessary until they are transitioning to secondary school or unless specific cognitive concerns are raised by families or teachers.

A potential barrier to accessing a neuropsychology assessment is lack of provision. In some services, neuropsychology services are only accessible for specific departments, such as neurology. The assessment requires specialist skills by a paediatric (neuro)psychologist. A full neuropsychology assessment is more than cognitive testing and has potential to inform other interventions, such as psychotherapy to reduce distress or liaison with schools to help around behavioural issues. The assessment can also identify prognostic and diagnostic indicators: to help understand what may be contributing to a particular presentation; to identify what the young person may be having difficulty with (e.g. working memory problems); and to highlight what is going well for the young person. Information from the assessment can be useful when consulting with primary and secondary care services, including schools. It is important to have functional and meaningful goals in mind, to consider before an assessment how it might be helpful, and also to think practically about what intervention can be offered afterwards. Data collated across neuropsychology assessments will also be helpful in informing practice-based research, such as the neurodevelopmental trajectory and parental/caregiver needs compared to other health conditions.

The suggested areas to include in a neuropsychology assessment are: general abilities (i.e. IQ), mental health and well-being, along with optional tests to assess specific memory skills, attention skills, executive functioning skills and language skills (see Table 1).

Table 1



Table 1.

Proposed neuropsychology tests and questionnaires for assessing functioning, cognition and mental health difficulties.

Domain	Test
General abilities / IQ	Wechsler scales: <ul style="list-style-type: none"> <li>• WPPSI-IV, for <math>\leq 6</math> years</li> <li>• WISC-V, for <math>\geq 6</math> years</li> </ul> Parental and school reports
Daily living skills	Adaptive Behaviour Assessment System (ABAS-3)
Memory	Child and Adolescent Memory Profile (ChAMP)
Attention	Test of Everyday Attention (TEA-Ch)
Executive functioning	Delis-Kaplan Executive Function System (D-KEFS) Behaviour Rating Inventory of Executive Functioning (BRIEF-2)
Language	British Picture Vocabulary Scale (BPVS) Clinical Evaluation of Language Fundamentals (CELF)
Mental health difficulties	Revised Children and Anxiety Scale (RCADS)
Quality of life	PedsQL-Neuromuscular module

There are many tiers within language skills. Some young men with DMD may have good vocabulary, but may struggle to express or understand more complex verbal ideas or concepts. It is helpful to obtain a profile to understand how the young person communicates and what they find difficult, which can inform not only home and school life, but also how clinicians ask them questions. Ideally, speech and language therapy would be involved as early as possible if concerns are present for younger children.

Other considerations when undertaking a neuropsychology assessment are the young person's level of fatigue, impact of any pain and medication. Selection of tests should take into account reduced motor skills with increasing age.

## Following the assessment

Following a neuropsychology assessment, the psychologist needs to support the family to understand what that cognitive profile means and how it might help understand the young person's behaviour better. The psychologist might discuss the cognitive profile and impact of mental health difficulties with clinicians to help them also understand the person's behaviour. Psychologists may suggest other recommendations to support the young person, such as having a break or exercise, or recommending simplified resources in clinic or school. If cognitive difficulties are highlighted, this should be shared with the school and be part of the EHCP and include within it the young person's strengths. The results will help guide further intervention, for example, if a talking therapy is indicated, adaptations may be needed to account for highlighted difficulties. This is especially important for informing other services, such as CAMHS, so that, if there are difficulties engaging the

young person, the service can better understand and take into account the underlying factors. Tracking change over time to help inform the appropriate support into adulthood is important, particularly around the transition to adult services. Having a neuropsychology assessment in the early years can help identify whether a child has a specific learning difficulty (such as dyslexia), an intellectual disability (i.e. significantly impaired intellectual and adaptive functioning) or a neurodevelopmental condition (such as autism) early on, so that appropriate support can be provided in school and referrals to appropriate community teams can be made.

Future aims of the paediatric psychosocial care project include establishing more indirect pathways to psychologically informed care. This could involve upskilling the local team in more psychosocial aspects of care as well as other local teams to be able to support families. The plan is also to demonstrate the effectiveness of psychosocial interventions and to develop guidelines based on practice-based evidence. A further key outcome is to disseminate findings to increase awareness nationally of cognitive strengths and weaknesses in boys with DMD.

## Conclusions

This workshop has highlighted the need for more research concerning the psychosocial, neuropsychiatric and neurocognitive needs in children and adults living with DMD in order to inform the provision of appropriate psychosocial support. Through building on our current understanding of these needs, appropriate interventions can then be identified and provided. This will enable a treatment pathway to be established with the aim of quickly identifying the most appropriate form of support for each individual with DMD, such as accessing psychological or pharmacological intervention.

To develop effective treatment pathways in child and adult services, there is a requirement for screening tools capable of identifying psychosocial and neuropsychiatric needs which can be utilised nationally by all professionals working with DMD. In addition, neurocognitive assessments should be offered to all patients with DMD as a way of addressing cognitive concerns, allowing for early intervention and support and as a baseline to evaluate how or if cognitive function changes across the lifespan.

Understanding the impact of the psychosocial, neuropsychiatric and neurocognitive needs in patients with DMD will aid in the development of national guidelines that will provide a structure for assessment and intervention to help improve the quality of life for all patients.

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## **Disclosures**

In the view of the Corresponding Author, there are no relevant disclosures to declare.

## **Conflicts of interest**

The authors have declared that no competing interests exist.

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