UK GUIDE TO PHYSIOTHERAPY & OCCUPATIONAL THERAPY FOR DMD

These guidelines have been endorsed by the paediatric and adult North Star Networks collectively. Many of the Network physiotherapists formed part of the Therapy Working Group and all paediatric North Star Network physiotherapists were provided with the opportunity to input the guidelines, and their content.

These guidelines are also endorsed by the Neuromuscular Group of Association of Paediatric Chartered Physiotherapists

14th October 2024
Version 1.0

CONTENTS

INTRODUCTION

This guide outlines the role of **physiotherapy and occupational therapy in Duchenne muscular dystrophy** (DMD) and specific details of these roles at different stages of the disease. Two similar versions of the guide exist. **This document is designed specifically for specialist and community-based physiotherapists and occupational therapists.** The other is a family guide and for those living with DMD – *hereafter referred to as 'individuals'*.

The purpose is to provide more UK-centric and detailed information based on the current international standards of care to improve the quality and consistency of care for those living with DMD (1–3) in the UK. These guidelines are informed by North Star Network input, the Scottish Muscle Network Physiotherapy Management profile as well as the Adult DMD guidelines (4) and DMD Care UK respiratory guidelines (5). Where the overlap is significant a link is provided to avoid excessive repetition.

The guide is also informed by information gathered from questionnaires filled in by therapists and individuals and families living with DMD on their experiences of current delivery of standards of care (SoC) and suggestions for how this could be improved. See Appendix A. There is evidence that this type of feedback can benefit delivery of services (6). An alphabetical list of contributors can be found in Appendix B.

The guide is presented as follows:

- Role of Therapists in specialist clinics and what to expect.
- Role of Community Therapy Team and what to expect.
- A guide to **different stages of DMD** with a description of the stage, details of suitable assessments, summary of physiotherapy and occupational therapy expectations as well as any additional stage-specific management strategies such as orthotics, mobility aids and other team members who may be involved.
- We refer to therapists as physiotherapists (PT) and occupational therapists (OT).

The stages addressed are:

PRE-AMBULATION/EARLY DIAGNOSIS

EARLY AMBULANT

LATE AMBULANT

EARLY NON-AMBULANT

LATE NON-AMBULANT

Additional Resources. Here you will find an overview of therapy strategies.

ADDITIONAL RESOURCES

Getting best use of these guidelines

You can use the additional resources section to learn about key management principles and find resources on how best to apply these.

If you wish to gain more detail about a particular stage, you can use the stage specific sections which provide links to the key management principles.

Expectations for families and individuals attending specialist clinics and community

What should individuals expect when they come to clinic regardless of age or stage?

They should expect to receive regular invitations to attend clinic to see members of a multi-disciplinary team including a specialist PT. The frequency of these visits is discussed later in this document.

What to assess when individuals attend a specialist clinic or community appointment and why?

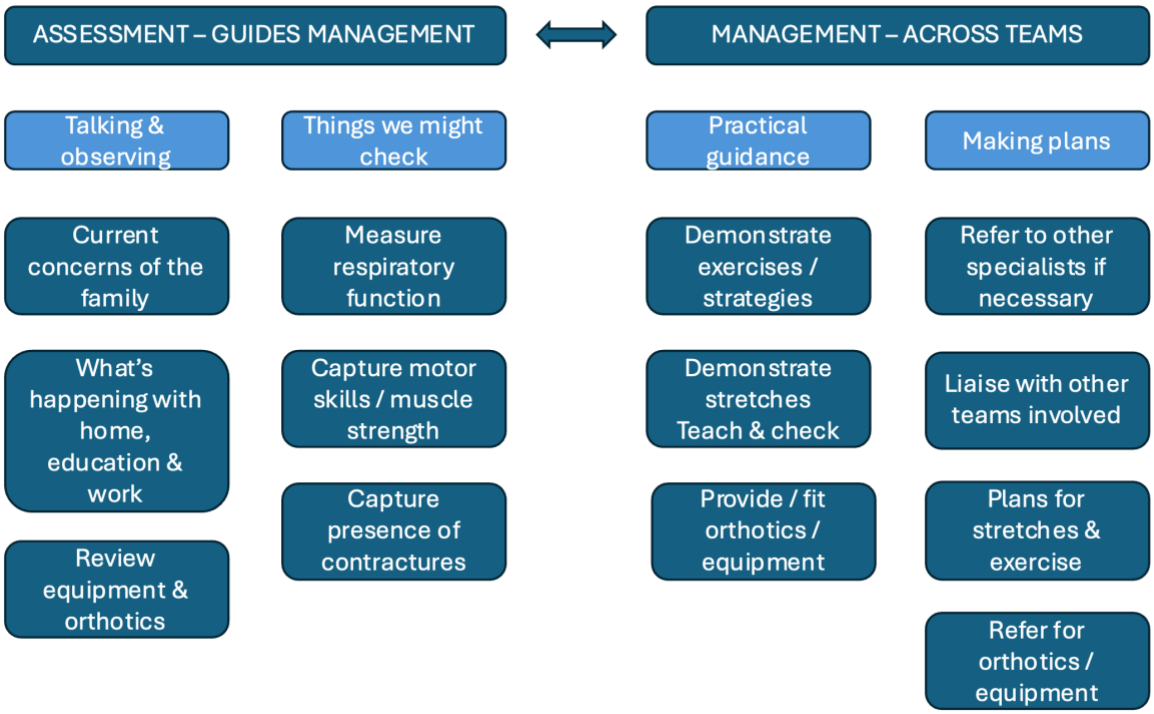
As outlined in the SoC document (2) a reliable assessment is key to ensuring effective and timely management. There are key components to that assessment regardless of age or stage of DMD. Here we outline these assessments. Assessments may be very different in the community when compared to specialist clinics as the focus is different. Specialist clinics wish to monitor disease progression. Community teams wish to evaluate the need or impact of an intervention.

It is important to remember that not every area outlined below is covered at every appointment. An appointment may have a specific focus for the individual and their family or may aim to evaluate a particular area of concern that needs addressing. Other issues may be addressed at another time. Specialist and community appointments may have a very different focus or aim.

Assessments to be carried out at an appointment (Figure 1):

- Ask about what is happening at home and **what has changed since the last appointment**. What issues need to be discussed today to ensure best care.
- Ensure you are aware of any learning disability or processing issues prior to assessment.
- You may ask the individual to fill in some questionnaires beforehand or ask some questions when they arrive. These "patient reported outcome measures" (PROMs) aim to capture real life and not just a snapshot of a single appointment, as both are valuable.
- Specifically **enquire about education or work and social activities**. Include travel to and from and moving safely in all environments. Ask for details of any equipment used or needed.
- Ask about any **falls, trips or accidents** even if the individual is not walking. There may be risks around transfers and safety in a wheelchair due to the prolonged use of corticosteroids which leads to osteoporosis.
- Some measure or **review of the muscles of breathing** should take place in all but the youngest of children. Standard assessments in the UK consist of Forced Vital Capacity (FVC) in a sitting position and an assessment of cough, Peak Cough Flow (PCF). These are done in clinic (neuromuscular or specialist respiratory clinics) and are used to monitor the muscles of breathing over time. For a full assessment to be conducted, you will need to know the individual's current height. This can be done whilst standing (if possible) or by checking arm span or ulna length in a sitting position. More information on measuring height can be found here.
- **A measure of what an individual can do in terms of function** should be performed. This captures both muscle strength and the impact of joint tightness. Importantly this helps evaluate what has changed since the last assessment. This may involve walking and/or how the individual is able to transfer and/or what they are able to do with their arms.
- Check for any **joint tightness** due to shortening of muscles. The number of joints tested varies depending on individual need, from just a few, to many joints. You may do this using a goniometer or by observing.
- **Check any equipment brought in with the individual**. This may include a wheelchair or buggy. Posture and suitability should be checked. Review any orthotics such as ankle splints. If they have not been brought in, discuss and plan a follow-up review if necessary. Ask about transfers in and out of equipment or on and off a bed. This may include respiratory equipment.
- **Check the individual's back and particularly the posture and movement/flexibility of the spine**. This can be done in a wheelchair by lifting clothing or in a standing position with a clear view of the back and chest.
- Ask about **pain and discomfort.**
- Ask how the individual **manages energy** through the day.
- Provide **advice and management** – details of what this might cover are included in the stage specific sections below.

Figure 1. Overview of what might be covered at a specialist appointment*



**Not all areas will be covered at every appointment*

Community assessment may focus on just one or two key areas outlined in Figure 1. Individuals may be seen at home, at school or nursery or a community therapy centre or in their workplace.

Some areas have complex care teams who operate separately to local community teams. They may have their own PTs and OTs and may be useful in delivering care.

Will an occupational therapist be involved?

An OT may work alone or with the PT. Areas that OTs might assess are detailed in the stage specific sections below. Historically in the UK, OTs have not always been present in specialist clinics. Therefore, who performs different parts of the assessment or management varies between centres.

Community based assessments - what are these for?

Individuals should have access to physiotherapy and occupational therapy in the community (7).This is more readily available for children, but you can ask the specialist clinical team about accessing community therapy for adults. The aim is to ensure appropriate and proactive management of changes as well as offering support to individuals as they face new challenges whilst anticipating future needs.

Community teams often focus on assessing how people manage in their home, education, or work environment. They ask about what individuals are able to do and what they might need help with. In many cases they will assess and advise on stretches and exercise. They should be in regular communication with the specialist team, and it is essential that these teams work together to provide consistent advice.

How often do individuals attend specialist clinics?

In the UK, as recommended by the SoC (2) all specialist paediatric centres should plan to see people living with DMD every six months. This happens for the majority. However, you may need to see an individual more frequently. Decreasing frequency of appointments does also occur for several reasons but is not part of SoC.

Increasing frequency of clinic visits	Decreasing frequency of clinic visits
Starting or monitoring change in steroids	Personal choice of family or individual
After a fracture	Clinic/staffing pressure at centre
Loss of ambulation or significant deterioration	Alternative provision of video or telephone clinics
Requiring support with respiratory issues, managing secretions, bowels or heart issues	
Other concerns	

How often do individuals need to be seen in the community?

In an ideal world, individuals would have regular access to treatment and support from physiotherapy and occupational therapy as identified in SoC (2). At the very least they need to be able to contact someone easily and quickly if they need support or advice. Work with families to at least have contact details so that a new referral back into the system is not needed every time. The specialist team should be able to signpost individuals to where they can access appropriate help.

What will management consist of and why?

As outlined in the SoC document (2), clinic assessment is key to ensuring effective and timely management. This is not just about plotting change but identifying ways in which you can help promote function and independence as well as support and improve quality of life. Management may happen in the specialist clinic but is more often appropriately delivered locally, which tends to be more convenient, reducing travel time.

Despite the defined therapy management requirements outlined in the SoC, much of the burden of delivering management, such as stretches, falls on the family. The impact of this should not be underestimated and community teams should aim to deliver face-to-face therapy and updated instruction. Therapists should support families trying to overcome barriers to obtaining the correct care (8). No family or person should be left to do their own therapy without guidance. Best results are seen with face-to-face therapy and home delivery (9).

What is important to families and individuals when interacting with therapy staff?

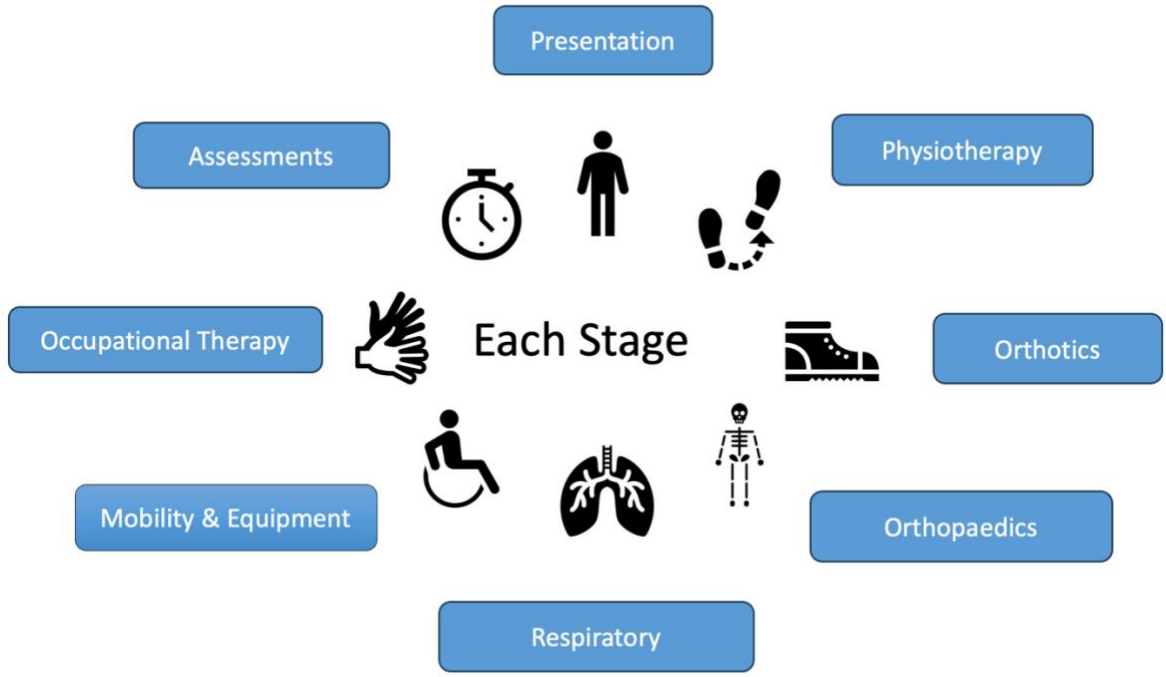
- Key to good management is evidence of effective communication between community and specialist centre teams. It should be and that it is clear to the family how these lines of communication work.
- Seeing the same therapist or a small therapy team promotes confidence.
- In younger children, the morning may be the best time for therapy appointments; in older individuals, a later appointment can be more acceptable, and you may find them less tired. Getting ready in the morning to leave the house can take a long time for some less able people (hoisting, dressing, eating etc).
- If applicable, a strong Transition Team to support the process of transition from child to adult health and social care.
- Having access to a hoist if needed when attending clinic so assessments can be conducted properly.

Key summary points

- Access to therapy should happen as soon as a diagnosis is made even if, in the early stages, actual input is limited (2).
- Therapy interventions vary according to the setting in which care is provided. The members of the team may differ and will depend on resources available.
- Communication between the team around the person living with DMD is key to effective and consistent management.
- All aspects of therapy (stretches, activity, exercise, orthotics, pain management and equipment) aim to minimise secondary complications and therefore maximise function and independence.
- There are significant differences between what is available to children compared to adults which can be influenced by where they live, but limitations occur across the board. Individuals and families should be offered support and advice on how to access what is needed in a timely fashion.
- This guide is designed to work alongside the published adult therapy guide and respiratory guide. Other guides relevant to therapists are in production.
- Other areas where therapists can support individuals may not be covered in this guide but found in additional guides. This includes emotional and psychological support.

There are parts of management which are particular to different stages within DMD. In the next sections we outline the key features of different stages of DMD and key principles of assessment and management (4,5).

Figure 2. Structure for each stage of how guidance is laid out. Key information on management strategies can be found in additional resources.



PRE-AMBULATION/EARLY DIAGNOSIS

Presentation

Diagnosis: These infants may have an early diagnosis because mum is a known carrier, one or more brothers have been diagnosed with DMD, or a child with developmental delay is found to have a high creatine kinase (CK). Children with marked autistic spectrum disorder (ASD) may also be found to have DMD. See pod.nmd.org for details about early signs and symptoms of DMD.

Delay in skills: Infants diagnosed with DMD may have motor or global developmental delay and/or behavioural issues within the ASD spectrum. They frequently have speech delay. Some may never crawl, find moving between positions difficult and may be late walkers (10).

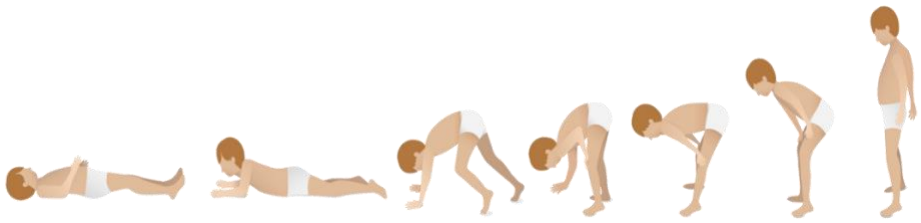
Joints: Some children will develop contractures (tightness in a joint) from a young age. Long finger flexors, hip flexors and ankle tightness can be found before they start to walk. A child who crawls or weight bears on bent fingers or fists may have tight long finger flexors. If they have an increased lordosis from first learning to stand they can already have hip tightness. A child who persistently stands on their toes can develop tightness in gastrocnemius muscle complex. Toe walking is an early sign. This can be related to muscle tightening, and often worsen when the child is tired, but also be impacted by the ASD behaviour and sensory problems.

Conversely – some can present with early hypermobility, and this can add to developmental delay due to hip and hindfoot instability.

Sometimes there is evidence of a general low tone or even hypotonia. Sometimes described as "slip through my fingers" when you try and pick them up.

Weakness can be evident from a young age. Hip extensors and knee extensors are the major muscle groups affected causing difficulty with activities such as rising from the floor, from squatting into standing and when climbing. The "Gowers' Manoeuvre" (Figure 3) may be seen from a young age and is not normal for children. It is not exclusive to DMD but added to other symptoms is a warning sign. The soleus muscle in the calf is affected so jumping or hopping may not be effective (or the child may not be developmentally mature enough to hop or jump – depends on age). Weakness especially around the hips may result in a waddling or abnormal gait (wide base especially on speeding up).
Weak neck flexors are an early sign of weakness and are why they roll to the side or to prone in order to sit up or stand up.

Figure 3. Schematic of a Gowers' manoeuvre.



1. Rolls prone. 2. Puts hand to floor and walks them back. 3. Widens feet. 4. Uses hands on legs to push up into standing. Completing any of these movements when getting to standing constitute a Gowers' sign.

Calf hypertrophy (Figure 4) – when you observe the calf muscles, they appear enlarged. This is not evidence of a stronger muscle but pseudohypertrophy, where muscle tissue is replaced with other tissue.

Figure 4. Child with DMD demonstrating calf hypertrophy

Assessments

Joint range: Hips, tendoachillies (TA) and long finger flexors (LFF) need to be measured as tightness can often be evident from very young.
Hypermobility should be noted as it can impact on gross motor function.

Developmental scales for children such as the Bayley Scales of Infant Development (11) or Alberta Infant Motor Scale (AIMS) (12) could be used as most scales specific to DMD will not include the very young or not yet walking child. You can use the North Star Ambulatory Assessment (NSAA) (13–15) as part assessment and part parent report. In younger boys aged 3 – 4 they may not be able to achieve items due to their developmental stage rather than their diagnosis (16). See NSAA worksheet with notes on when skills are achieved in typically developing children.

A **general assessment of muscle power can be made through observation of play**, looking at those groups which are seen to have antigravity power and those which do not (e.g. neck flexors, hip extensors) through performing a pull to sit action and play over a roll or peanut ball.

Physiotherapy

Physiotherapy will be directed at helping progress motor development, improving motor skills, facilitating transitions, standing, cruising, and walking.

Exercise and activity should be done through **play and fun activities.** Specific exercise programmes at this age are not usually given but families may be given a specific goal, such as promoting a particular action. This stage is ideal for introducing water-based play, which will keep muscles and joints supple (make sure the pool is warm enough as cold muscles do not work so well). Starting hydrotherapy early also promotes its use as they get older if it's something they enjoy. Keeping active must be balanced with **pacing advice** to use energy in the best way. For example, it may be better to push a child in a buggy to the park, so he has the energy to play when he gets there.

Avoid eccentric muscle activity such as trampolining. This is because excessive eccentric muscle activity causes damage to the muscle cell membrane and can lead to myoglobinuria (dark coloured urine). If this occurs inform the neuromuscular medical team as soon as possible.

Stretches. Advice should be offered if any joints are tight and on how families can help with improving range of motion. Short duration stretches, either passively or actively, or long duration stretches using ankle-foot orthoses (AFOs) may be a helpful adjunct to the management programme if they are proving difficult to manage.

	Footwear. Spending time barefoot is important for development of the small muscles of the foot. When shoes are needed keep the footwear light. Heavy boots are not usually recommended. Only use boots for those where they are not yet understanding what their feet are for and need to practice "weighted" standing. NB – if mum is a manifesting carrier, she may well have weakness and difficulties (including some learning issues). This needs to be considered when making recommendations, programmes and when teaching any stretches.
Orthotics	Many neuromuscular physiotherapists and centres advocate the use of night time splints and research has shown that they are worn regularly while ambulant (but not whilst they are walking) will have better foot posture when they lose ambulation (17). While this cohort are young, it may be of value in some children to consider them before they lose range of motion as it is easier to maintain range than get it back once lost. In young children early introduction of night splints may result in better compliance. For some foot variations such as pes cavus or pes planus it may be useful to arrange assessment and supply of insoles or inlays.
Equipment	One would not normally suggest equipment at a young age, but the younger boys can be very fidgety when sitting and advice on a good seat may be helpful for parents, playgroups, or childcare. Hand activity and increasing dexterity through games and activity is helpful. A buggy may be sufficient for mobility needs in a young child who cannot walk far and needs his energy for playing, not just travelling. As they grow, a manual wheelchair may be more appropriate.
Orthopaedic considerations	It is very rare, but not unheard of, that the children continually stand on their toes even before they start to walk independently. This can lead to tightness of the Achilles tendon and may need orthopaedic or specific therapy intervention such as serial casting or early ankle release. Serial casting is usually done under the direction of the orthotic team and physiotherapist and can provide a prolonged ankle stretch. This is different protocol to casting for cerebral palsy. Usually more frequent change of cast over a shorter period. Early ankle release should only be considered under the guidance of the multidisciplinary team (MDT) and orthopaedic consultant with experience and knowledge of DMD.
Specialist Services	Children who are diagnosed due to global developmental delay will frequently need input from a speech and language therapist. OT - Occupational therapy input in the community setting may be important around activities of daily living such as dressing and bathing. Other areas of support can be provided by Occupational therapy. Genetic counselling may be appropriate if the family need support around family planning and risks for subsequent children. All children must be followed up by a paediatric neurologist with interest in neuromuscular diseases or a neuromuscular specialist, such as a genetic nurse.
Nursery/play	A community visit to nursery or playgroup is recommended to give advice to the staff on ways to assist if development is delayed or if fatigue is an issue. Classroom assistance may be required, especially if significant management procedures are in place such as stretches or specialist equipment.

EARLY AMBULANT

Presentation

Muscle weakness becomes more noticeable with difficulty getting up off the floor. See Gowers' manoeuvre Figure 3. Children may not run, jump, or hop, and falls are likely. You may observe difficulty with stairs (sitting down to negotiate them) and getting out of a chair (needs help or needs a high seat to manage it independently). When families compare to their peers or siblings, they may be slower to gain skills and be less adventurous in the playground.
There is an increasing risk of tightness in some joints, usually ankles, hips, and wrists because of long finger flexor tightness. Tightness in gastrocnemius muscle complex can reduce ability to manage a step, or stairs, or stand up from a chair.

Calf hypertrophy: The muscles at back of leg/calf may appear to be bulky or large (Figure 4).

When walking, you may observe they have a significant curve in the lower back (lumbar lordosis) and walking is perhaps clumsy and involves swaying side to side (waddling). As walking difficulties increase, any gait analysis must take into consideration, upper body impacts and compensations (18).

They may experience pain and muscle cramps after activity which may be more troublesome at night.

Assessments

The **North Star Ambulatory Assessment** (NSAA) is typically used in specialist clinics to capture ability to walk, jump, run etc. It is usually completed every six months (more frequently if starting steroids) and takes 10 – 15 minutes to perform. It is designed to be used for ambulant children but may not be possible to perform in very young individuals who can't follow instructions (see pre-ambulation/early diagnosis section) or in children with severe learning difficulties and/or behavioural issues. You may observe a child at play and ask the family about what he is able to do.

Ask about or observe an individual ascending and descending the stairs.

Monitor and check **range of motion** (ROM) in some key joints such as ankles, knees, iliotibial band. It is important to use a standard technique to measure joint ranges (19). We know that reduced ROM can impact motor performance (20–22). Check symmetry and mobility of the spine. Issues with the spine are unusual at this stage but need monitoring so changes will be picked up in a timely manner.
Ask if pain or cramps are an issue and observe posture and movement.

Specialist clinics may perform the 6-minute walk test (6MWT) (23–26) but this needs space and time.

Physiotherapy

It is very important that the specialist neuromuscular team liaise with the therapy team local to the family. It is vital that management is consistent across all teams and that when a plan is updated this is communicated to everyone involved.

Stretches are essential where tightness impacts the ability to do tasks (27). At this stage these are most commonly for the ankles, but other joints may be impacted and need care. These stretches need to be made part of a daily routine from diagnosis without days off for good behaviour – just like brushing teeth.

	Activity and exercise advice is essential at this stage. Encourage and support play-based activity and participation with peers in /out of school. Swimming/fun in the pool will keep muscles and joints supple (make sure the pool is warm enough as cold muscles do not work so well). Keeping active must be balanced with **pacing advice** to use energy in the best way. For example, it may be better to push a child in a buggy to the park, so he has the energy to play when he gets there. Here is a Resource on managing fatigue. Some forms of exercise should be limited or avoided. **Avoid eccentric and heavy concentric loading exercise such as repetitive trampolining** (28). Ensure school is fully aware of abilities; what to encourage and what to expect. Direct to involvement of Special Educational Needs Coordinator (SENCO) and educational psychology if appropriate. If required, offer pain management. Muscle cramps may be helped by a warm bath or locally applied heat. In cold weather some children will want to wear additional leggings or football skins under their trousers. This can help with muscle pain. Gentle massage or stretches may help the affected muscles or joints. Seek additional medical support as required.
Orthotics	Most common orthotic at this stage is a night-time use AFO to improve ankle range of motion. Other options may be offered depending on your geographical location. All orthoses should be used in conjunction with regular stretches. Tightness in the ankles is difficult to manage. An increasing loss of movement can occur even when stretches have been performed because muscle growth is typically slower than bone, which means that during the growth phase, the muscles take a bit longer to catch up and regular assessment is essential to keep on top of any joint tightness. Insoles or inlays in footwear to correct foot position may be useful and should be assessed for on an individual basis.
Equipment	Specialist seating/equipment may be beneficial in school to ensure a symmetrical sitting position is promoted. However, children should be encouraged to sit on the floor with their peers, if able, as this is a key area of participation.
Wheelchair Provision	A manual wheelchair may be considered to support independent mobility and manage energy expenditure.
Orthopaedic considerations	Not usually required at this stage however please refer to the orthopaedics guidelines for advice on surgical intervention and management of long bone fractures section. The most likely risk is for long bone fractures.
Specialist Services	**Respiratory issues at this stage are not likely.** However, it is important to encourage activity to promote lung function such as swimming/playing in the water. Baseline lung function tests should be done in the specialist clinic such as Forced Vital Capacity (FVC). See Respiratory guidelines. One benefit of FVC at this stage is practice and getting used to the assessment. It is common for it to improve as they grow, and their technique gets better. Ensure that immunisations and vaccinations are up to date to protect lung health. Teams that may be involved: Dietician: Dietary advice is important, especially if the children are on steroids.

	Genetic counselling may be appropriate if the family need support around family planning and risks for subsequent children. Speak to specialist nurse or medical consultant. Psychologist/Educational psychologist if required. Cardiology, as cardiac issues can appear from an early stage (Cardiac guidelines). Endocrine/Bone Health – all individuals with DMD are at risk of osteoporosis however, this risk is increased when on steroids – See endocrine guidelines. Care adviser or specialist nurse for support around the family and sometimes Social Services. Occupational therapy: A home/school assessment may be needed to evaluate rooms and tasks for planning of any necessary future housing/school environment adaptations.
Education	Ensure school are fully informed about DMD and understand the future implications (access and support for Physical Education (PE), exams and participation). Specific advice is available here inclusive PE at school. Additional support for learning may be appropriate. Education, Health and Care Plans (EHCP) should be considered for all children. See here: Another useful resource is DECIPHA. The Learning and behaviour toolkit maybe useful at this stage as may support for sensory processing.

For information around the recently proposed see Transfer Stage.

LATE AMBULANT

Presentation

Muscle weakness leads to a widened base of support in standing with or without asymmetry and they may have difficulty standing with heels down. If the heels are down, then upper body posture often has to adapt e.g. lower back will be more arched or bottom sticks out as is not aligned with the shoulders. The **lumbar lordosis** is often very prominent. Weakness makes it difficult to get out of a chair. They need to move feet apart and/or use arms to push up. Some will need to turn prone to face the chair to achieve standing. They usually require furniture to assist rise from floor or are unable to get up. Climbing stairs is very effortful or too difficult to manage at all.

They may **fall or trip** more often as they get weaker or may fall less as they avoid walking in certain circumstances. They may need to rest after a short duration of walking. This reduced balance in standing or when moving will impact on the individual's ability to navigate through busy environments e.g. playgrounds and moving between classrooms at school.

Reduced core stability occurs as trunk muscles become weaker, which can lead to difficulty sitting up or a tendency to lean forward when doing activities for prolonged periods of time.

Upper limb weakness is more evident. It is harder for them to lift heavy items or reach up high.

As well as lower limb **contractures**, they may have limited range of motion in arms e.g. supination (difficulty turning palm to face upwards) and long finger flexors (stretching fingers out flat).

They may tire easily with physical activity so may need to **reduce walking** outside and use their wheelchair for longer distances.

Pain may be an issue with calf pain possible in standing at this stage. Look out for back pain, this may be due to significant lordotic posture but be aware of the increased risk of vertebral fractures at this stage.

Assessments

NSAA may still be used but as functional abilities decline, consider using the **Transition Assessment North Star** (TANS). This assessment avoids the harder items on the NSAA and includes transfers. It provides a link to the Egen Klassifikation Scale (EK2) (29–31) used in the non-ambulant population.

Contractures may be more of an issue at this stage with tight ankles sometimes being the reason **for reduced ambulation** rather than just weakness. Most likely muscle groups impacted are: ankle plantar flexors, hip flexors, abductors, wrist flexors, supinators and finger flexors (32). Check range of movement in all joints and measure those with a significant loss.

Performance of Upper Limb (PUL 2.0) (33–35). The Entry item provides a baseline measure of upper limb function to capture progression if there is not time or equipment to perform the whole scale.

Review the individual's **mobility at home and in their education setting.** This should include a **falls risk assessment** as they are at increased risk of falls (36). If falls and trips are an issue, then capturing the number of falls/trips with a **fall's diary** can inform management but should not create too much of a burden (37). It can be difficult to establish why falls are happening. Some examples are sudden turns, hazards on the floor or knees giving way.

	Assess how they move between positions e.g., from a bed to a chair or from the toilet to the shower. In this way we can support people with alternative strategies or equipment to maximise their function. It is important to monitor the **spine and its flexibility**. This should be done in sitting and standing. If asymmetry is noted, discuss this with the medical team and complete a referral to spinal services as necessary. Look at sitting posture in the wheelchair if possible and ensure symmetry is achieved. **Visual analogue pain scale** (VAS) /ruler may help capture pain as well as recording, where, when, intensity and how much it impacts movement or function (38). VAS may not be useful in younger children or those with cognitive issues (39,40). Be particularly vigilant of low back pain as vertebral fractures are more common at this stage. **Fatigue scale** Borg rating of perceived exertion 6 to 20 – using (41). Using this scale may help the individual and their family manage their fatigue by understanding the perception of exertion.
Physiotherapy	Keeping active is still important at this stage. We know that strength training alone or combined with other exercise may improve upper limb function, static and dynamic balance as well as gait and well-being in muscular dystrophy despite no evidence of structural changes (42). E. g, mild to moderate strengthening exercises may be of benefit and should be supervised and monitored by a physiotherapist (43). **Promote symmetrical activities** and avoiding asymmetric activities such as a scooter or long periods sitting in an asymmetric position Page 309 - (32). **Teach** stretches and review the stretching programme at home and at school. This programme may include upper limb stretches (particularly supination and long finger flexors) which can become tight. **Standing** is important and heel wedges for when an individual is on a standing frame may be used to support this if ankles or one ankle is tight. Consider a standing frame in discussion with the young person and family as this can provide a safe and supportive environment when walking becomes difficult. Also, KAFOs for walking/standing. Careful consideration of appropriate standing frame provision is vital relative to their functional ability as they may require a wider foot base. This will need monitoring and adapting as their function changes. **Advice on pacing**, dosing activity and fatigue management is important at this stage as is managing the loss of ambulation. Some families say they are very keen to maintain ambulation as long as possible, whereas the individual can be ready to stop walking as his function and independence are better in a wheelchair. Ensuring the whole team around an individual understands what is happening, and advising on the best balance between walking and wheelchair use is important. Some situations require us to give permission to "stop walking". **Hydrotherapy** is a useful and a safe place to exercise and walk without worrying about falls (44). **Manual handling and safety advice should be** given to the family and the whole team around the child. Making sure that a **falls risk assessment** has occurred, and everyone is clear about reducing the risks of falls and managing them if they occur. Pain and cramps may be eased with warm baths and gentle leg massage. If a fall has occurred, there should be a low threshold for seeking medical advice.

	Be aware that fractures, especially vertebral fractures (which can occur at this stage), can be "silent" i.e. not associated with any pain or a particular incident. Also **be aware of the risk of fat embolism even after a minor fall without fracture**. The risk is most high in the 48 hours after a fall.
Orthotics	An **AFO** may be useful at this stage if not already being used. AFO's should not be worn for walking. Heavy boots are not advised. They may improve a heel down posture, but the excess weight makes walking difficult and tiring. For those who are finding walking more difficult and would like to continue ambulating, discuss KAFOs suitability with tertiary centre. See Orthotics for more details.
Occupational Therapy/ Equipment	Ensure referral for adaptations at home is underway. **The home environment** should be assessed with a proactive approach to future needs to ensure adaptations are in place prior to them being necessary. It is important to do this early because if the environment is not suitable for adaptation, then the family need to move house. Anticipate that in a growing child, a larger electric wheelchair may be required, but as they are bulky, this can further impact accessibility. The need of other devices (e.g. ventilatory support) in the future should also be considered when planning extension and adapted bedrooms as they require space and power outlets. A profiling bed/special mattress/bed that provides movement may be needed at home. Consider a suitable form of static seating for home use such as rise recliner chair, although many children will prefer their powered wheelchair as they can easily move about the home. Avoid sitting on office type chairs with wheels as although the child can use their feet a little to move around the room, foot posture is poor and can lead to unmanageable foot deformity making positioning and the wearing of shoes more difficult. Ensure access to a hoist in clinic as well as at home and in the education or work setting. Consider provision of disposable slings in hospital setting or ask the family to bring their own sling to appointments. This would need to be compatible with the hoist. A spreader bar may be needed (if not already provided) to accommodate everyone in clinic. It is a good time to assess for **environmental controls and technology** to maintain independence for the individual as their weakness further impacts activities of daily living (ADLs). Consider a mobile powered arm support assessment if there are difficulties with eating. Consider resting splints for hands and upper limb activities to improve function. Wheelchair: See wheelchair guide. Use thoracic supports if required. Feet should be supported and aligned as best as possible in neutral (helps stability and helps manage ankle contractures). If a powered wheelchair assessment has not yet occurred, this needs to be prioritised. A power assist manual wheelchair could be considered before an electric wheelchair to maintain upper limb function. If the wheelchair is used on transport, then a head support is vital. Make sure they have access to a tray or height adjustable table so the elbows can be supported to improve arm function and ADLs. With shoulders and upper back muscles being weak supporting the arms at rest takes excess pressure off the proximal joint structure.
Respiratory	Routine spirometry of FVC and possible PCF is performed in specialist clinics. Important to promote physical activity and therefore effective expiration. See Respiratory Guidelines (5,45).

Orthopaedic considerations	Monitor spinal posture in sitting and standing (we are looking for any evidence of scoliosis or asymmetry). Ensure additional postural support in sitting if necessary. Serial casting or surgical TA release may be necessary to facilitate KAFOs or use of a standing frame. Any orthopaedic consideration needs to be done through liaison with cardiac and respiratory teams especially with an anaesthetic risk and post operative risks. Be aware of risk of long bone fracture. N.B. Risk of fat embolism Syndrome (5).
Specialist Services	Referrals which may be beneficial: • As well as those mentioned in previous sections (respiratory, dietician, endocrine). • Family Care Advisor, peer support or counsellor input for emotional support (46). • Consider psychology/child and adolescent mental health services (CAMHS) referral for other issues such as fear and anxiety related to physical changes. • Consider mental health support e.g. Headspace app, school counsellor. • Motability services re: vehicle to take powered wheelchair out and about. • Ensure referral to Social Services occupational therapist re: home adaptations. • A dedicated electronic assistive technologies team if available.
Education	Education settings and places of work need to be aware of increased risk of falls and fatigue. The need for postural support in a chair in a classroom/office needs to be checked. Adjustments may be needed to ensure access to physical education/exercise. This could include provision of chairs inside/outside to allow the individual to sit/rest. Avoid floor sitting, as it will be difficult to get up from this position. Review distances walked and advise on pacing, dosing activity and consider transport needs to and from school. Check risk assessments, equipment and staff training for support. Advise on any plans for school residentials to promote inclusion. Ensure back support is available on all chairs, avoid use of stools e.g. in science labs. Ensure access is available to all areas with appropriate lift access available at school or at work. To complete school or college work effectively consider alternative methods of recording work such as laptop provision, extra time, rest breaks in exams and a scribe. Drag and dictate software or similar may be helpful. Moving and handling training should be made available to staff as required and issues around personal care addressed. This includes making sure independence is maintained for the “need to pee” as long as possible using a simple bottle for example rather than resorting to hoisting for such a task. Proactive planning is important especially when place of education changes are anticipated. Each individual should have an individualised educational plan, with multidisciplinary input.

EARLY NON-AMBULANT	
Presentation	**Taking steps** becomes more difficult and may only be over short distances, with support or in the home. **Standing balance** is usually reduced and requires an individual to be placed into hyper lumbar lordosis or they may be unable to stand at all. Due to **advancing muscle weakness and decreasing mobility**, there is an increased risk of scoliosis (47,48). Sitting balance may remain good but dynamic balance may be impaired (difficulty leaning outwards and upwards or bending forwards). This may be exacerbated if the individual is going through puberty and growing and sitting for prolonged periods. Note that people with DMD and on corticosteroids often experience delayed puberty and may be treated by their clinical team with testosterone to induce and maintain puberty (49). They may be able to perform an **assisted standing transfer** or sliding transfer with aids. Often able to self-propel a manual wheelchair but be aware of fatigue and repetitive strain on the shoulder joints. They may have **difficulty raising their arms** above their head and may still be at risk of falls during transfers or from their wheelchair. **Contractures** may be more apparent in the upper limbs. **Pain** may be associated with increased periods of sitting. Be aware they are still at risk of vertebral fractures even though they are not walking. These may or may not present as painful and may or may not be related to a specific event or injury. **Respiratory muscle involvement** becomes more likely at this stage and should be monitored (Link to respiratory guidelines). This can be a complicated period especially if it coincides with puberty and individuals see peers gaining greater independence. For further details see the Adult Therapy Guidelines for DMD (4).
Assessments	The **TANS** could still be used in stronger individuals to capture transfer and standing ability. It is more likely that the Egen Klassifikation (**EK2**) is appropriate, it is useful for assessing function in non-ambulant individuals. The **PUL 2.0** (33,50) including the entry item, is appropriate to capture the progression of upper limb weakness. You could evaluate hand grip strength using myometer and this may be compared to normative data or previous evaluations. It remains important to **capture ROM** especially in the **upper limbs. For those using a wheelchair** for significant amounts of time, monitor ankles as a good appropriate foot position in the wheelchair is important in preventing further foot deformity. Review the amount of abduction and external rotation at the hips which can further cause foot posture to deteriorate. Continue to monitor neck ROM to ensure appropriate range for function. A measure of fatigue may be useful as well as a Quality of life (QoL) measure (51,52) which captures something of relationships and sex. For example, the Quality of Life for slowly progressive NMDs (QoLNMD) (53,54). Other QoL measures exist which are used to map health economics across all ages (55,56).

Physiotherapy	As highlighted in the previous section, this phase needs anticipating. When step taking becomes difficult, participation usually diminishes unless an appropriate wheelchair is put in place. Therapy includes active assisted exercise and **passive movements** and stretches (57). The key to this is 24-hour postural management. Pay particular attention to the **wheelchair** and **moving and handling**. If surgery does occur, then **pre- and post-surgical monitoring is important**. This is about pre- and post-operative assessment, the importance of stretches and exercise to prevent disuse muscle atrophy where possible and anticipating the need for equipment. Consider sleep systems and mattress assessment to promote good posture and to minimise any pain and discomfort due to poor positioning. Function can benefit from specific forms of exercise such as an arm ergometer (cycling with the arms) (58) and/or trunk exercises (59). These should be set up and monitored by a physiotherapist. Pain management is key in this group of individuals and as a therapist you may need the involvement of a specialist pain team or a palliative care team. Be aware of the link between lack of movement and pain and therefore the importance of safe stretching and activity. Pain characteristics do vary according to clinical stage (60). **Standing can be of benefit** and a variety of techniques may be used to promote comfortable standing such a standing frame. A 'sit to stand' type stander may be better tolerated and allow for better manual handling as the condition progresses. For standing, plantar grade ROM at the ankle is preferable, sufficient hip and knee neutrality must be achieved and the young person must want to stand and have the support from carers and professionals to do so regularly (61). Use of a stander should be discontinued if young person complains of pain and/or contractures at hip/knees are problematic. It maybe that the young person just does not want to stand anymore, and this is perfectly acceptable. Falls Management remains important in this group. There is a risk of falling from a chair or during a transfer. Ensure a seatbelt is in situ to assist with good pelvic positioning. It is key to link in with a joint approach with an occupational therapist for how ADL take place in the home. Professionals involved in bed/mattress provision need to be involved.
Respiratory	Both FVC and PCF are conducted in specialist clinics (neuromuscular or respiratory clinic). It may be necessary to introduce lung volume recruitment techniques such as a breath stacking device/manually assisted cough prior to a home ventilation referral for non-invasive ventilation and in-exsufflation device (62). Parents/carers need to be taught secretion mobilisation and chest clearing techniques. This should be anticipated and not taught only when the individual has a chest infection. Ensure early use of antibiotics if a respiratory infection is suspected. For key triggers and more information see Respiratory Guidelines (5)

Table 1. Key reasons for ensuring respiratory input based on respiratory assessments and recommendations for onward referral (5).

Triggers	Recommended onward referral
PCF of <270l/min in an adult or a decline in recorded values in children or adults	Prompt referral to specialist respiratory team and consideration of airway clearance support
FVC < 50% of predicted value per age and height indicates a higher risk of decompensation and possible need for ventilatory support	Trigger referral to respiratory team, even in the absence of symptoms
FVC < 30% increases the risk of requiring ventilatory support	Trigger **urgent referral** to respiratory team if specialised monitoring is not already in place
Any symptoms of sleep-disordered breathing regardless of FVC	Prompt onward referral to respiratory team
Unable to cooperate with spirometry, significant scoliosis and/or Brooke of >3	Prompt onward referral to respiratory team

**Assessments conducted as part of specialist clinic. Important that community work & specialist services work together.*

Orthotics	AFOs are still recommended when sitting in a wheelchair. Further supportive devices may be needed to prevent external hip rotation and abduction which can compromise foot posture. You may consider KAFOs if they are able to stand with assistance for functional rehabilitation. This requires significant input from the family and local therapy teams. See Orthotics section for more detail on prescription and application of these. There should be consideration for a resting wrist/hand splint to prevent significant hand deformity. This is still useful even if wrist and finger tightness has already occurred as it may help prevent further deformity. Maintaining hand function is imperative for the young person as for many, gaming is their window to the world as they form global friendships online.
Occupational Therapy	The home environment needs to be reviewed regularly and the provision of environmental controls and technology can have a significant impact on independence. If the young person is unable to change position in bed you may wish to introduce **postural management for sleep**. Mattress and specialist bed provision, such as a profiling bed, may be required. Where a hoist is being used, assess for appropriate sling with head support if required in collaboration with occupational therapist. A separate toileting sling is important. A height adjustable table for work/mealtimes may be helpful. A high table can help maintain independent arm function for eating and drinking. Consider mobile arm supports, although they can take a lot of work and adjustment to be beneficial (63). Attention to urine bottles and uri-sheath if appropriate to ensure independence in toileting. Ensure any wheelchair supports are easily removable so the young person can use a bottle comfortably. Adapted clothing advice can be useful. An appropriate powered wheelchair should be in place, with tilt, head rest and postural support as needed.
Orthopaedic considerations	Regular review within specialist neuromuscular clinics with referral to an orthopaedic team as indicated.

	Tenotomies may improve sitting posture if ankle deformity is problematic. Surgical TA lengthening procedures are generally percutaneous and, in most cases, require a general anaesthetic. Guidelines inform any use of anaesthetic in DMD (5). This is usually followed by two weeks in plaster. Bilateral AFOs after removal of plasters are required and therefore planning for home discharge is key to post-operative management. Fracture management may be necessary if young person sustains a fracture or has spinal fractures due to secondary osteoporosis. See bone health section Spinal surgery may be required at this stage to correct scoliosis. However, it can improve survival alongside respiratory support (64). Spinal surgery is less likely with the more widespread use of daily corticosteroids (65) and is less likely to be required if they are past puberty and have stopped growing. Hip and shoulder monitoring for subluxation and attention to postural management in the wheelchair are necessary.
Specialist Services	The individual may need: • Dietician; They may feel that eating takes too long and fatigue issues emerge. Feeding ability may be compromised and he may not want to ask for help. Be aware of changes in eating pattern and monitor weight. Useful advice can be found from Pathfinders neuromuscular alliance. They can provide advice on Vitamin D and managing weight for example. • Endocrinology to review bone health and spinal fractures. • Speech and language therapy assessment for chewing and swallowing may be indicated. • Introduction of care agency to assist with areas of personal care. • Continence care services. • Respite care and befriending if necessary. • Psychological evaluation if indicated. • Assessments for housing adaptations if not already in place. • Equipment needs for ADLs/personal care. • Ongoing review with wheelchair services to ensure chair continues to be adapted to changing needs. • Cardiac-specialist involvement. • Possible respiratory team involvement.
Education	Secondary School/college access assessment. Educational psychologist/Family counselling may be useful at any stage. Education, Health and Care Plan (EHCP) – or equivalent in other countries - is pivotal E.G. Scottish Child Plans.

LATE NON-AMBULANT

Presentation	This phase can be lengthy and particularly impacted by **decreased hand function** and difficulty with feeding themselves. This loss of hand function can make driving a power wheelchair with standard joystick and using standard gaming control/laptop difficult. **Muscle weakness leads to reduced head and neck control** and can make it hard to find a comfortable position in bed or in a wheelchair, which is why it is important that there are ways to adjust posture using the equipment. **Contractures.** There is often reduced ROM throughout – particularly noticeable in hands and feet, with difficulty positioning in wheelchair, with the hips abducted and externally rotated. Knee contractures can make it hard to find a comfortable position in bed and when performing ADL. **Reduced respiratory function** is likely with an increased likelihood of needing non-invasive ventilation during the night, or even during the day. Most will have a reduced ability to cough and will require cough augmentation. It is possible that the swallow function is reduced. There is an increased vulnerability to skin breakdown and possible risk of oedema in the limbs. **Swallowing difficulties** can also occur during this stage leading to weight loss.
Assessments	Capture motor performance linked to ADLs using **PUL 2.0 to assess** upper limb function **and** the **EK2 –** which is useful for assessing home functioning and identifying swallowing and eating difficulties. Muscle strength at this stage is captured using these scales. For **ROM** focus on hands and ability to function. Neck movement and head control are important skills for communicating. Ensure regular review of skin pressure areas. Postural assessment in the wheelchair and bed are important and are often better captured by a home visit and the community team. If you are not seeing a person face-to-face you can use the DMD functional ability self-assessment tool (DMD SAT) (66) and the EK2 over the phone or via video call. To capture PROMs consider the PROM-Upper – a measure of ADL and upper limb function (67) and Quality of Life measure for slowly progressive neuromuscular disease (QOLNMD): (54,68) The DMDQOL may be appropriate (55,56). Ensure that you or one of the MDT pay special attention to weight bearing areas of skin and skin creases to ensure good tissue health.
Physiotherapy	Physiotherapy in adults and late-stage mobility involves a highly integrated care team across specialist and community services. As at every stage you must review orthotic support, exercises and stretches alongside range of motion. This includes 24-hour positioning advice where the home and work environment will need to be reviewed. The focus is promoting bodily ability, sociability, content and independence (69).

	Abdominal massage for the bowel may be beneficial for managing constipation and needs to be taught in a quiet environment with good access to toileting facilities. Passive movements that increase intra-abdominal pressure can help. See Continence Care Patient Information Sheet from Scottish Muscle Network. If temporomandibular tightness is an issue a jaw stretching device such as the "therabite®" can be beneficial (70). This can help maintain the ability to eat larger items of food that require a wider mouth, improve dental hygiene, and ensure that access is available through the mouth in the event of surgery or an emergency. Basic wheelchair assessment and referral/liaison with local wheelchair service will continue to be essential with possible referral to environmental control teams and services to support access to social activities e.g., gaming (www.specialeffect.org.uk). As the wheelchairs become bigger and more complex, support for funding and repairing these chairs becomes vital. Liaison with occupational therapy colleagues in the community regarding environmental controls, and other postural management is important. Sleep systems/turning beds may be useful at this stage to ensure comfort and prevent excessive turning required through the night. Support around the hip can be important to prevent worsening of frog legging postures. Ensure the young person can call for help as required. Liaison with local communication hubs for communication aids. Families may need support for accessing these hubs or use speech and language therapy advice. Liaisons with tissue viability services for skin management so ensure weight bearing areas and skin creases are monitored. These may be accessed through the GP/district nursing teams or specialist services. Liaison with dietetics and speech therapy colleagues to provide swallowing and nutritional support. Adapted diet and/or assisted feeding might need to be considered in some cases. Liaison with specialist respiratory services to support non-invasive ventilation (NIV) and cough augmentation. Encouraging use of NIV to improve functional quality of life e.g., before completing an exertional task. **For more details refer to supplementary material in the published Adult Therapy Guide (4).**
Respiratory	This section is well covered in the Respiratory Care Guidelines (5). The key at this stage is a proactive approach to management that anticipates changes and the next stage so that crises are avoided. Ensure appropriate cough augmentation and ventilatory support are in place and are delivered by an expert ventilatory team. Support with secretion clearance and involvement in end-of-life care planning and discussion around continuing ventilation may be necessary.
Orthotics	Upper limb splints (orthoses) may be important to maintain hygiene, positioning in wheelchair or for range of movement and comfort. Splinting in the functional resting position of the wrist mechanically influences long finger flexors and may support improved functional use (71,72). Lower limb splints may be suitable to maintain hygiene, positioning in wheelchair, for range of movement and for comfort. Resting AFOs may help preserve foot posture so that feet can still

	be placed in a shoe. Preventing significant foot deformities helps reduce the risk of pain and the need for surgery further down the line. Some individuals may have a spinal brace to support positioning. Monitor fit of the brace for comfort, function, and pressure areas. Liaise with the specialist centre who have provided the brace for adaptations. It is important to always consider the goal of your splint and any detriment to an individual's functional ability.
Occupational Therapy	**Ongoing review of home environment, environmental controls, and technology** Review of current equipment provision and consideration of any additional equipment that may support function and comfort. Including: - Adapted joysticks and adapted games consoles. - Augmented technology to assist communication needs. - Appropriate wheelchair seating. - Appropriate manual handling equipment, considering head support. - Sleep systems (see physiotherapy). - Mobile arm support may suit some individuals but can be cumbersome and tricky to set up and use daily especially if they take a long time to set up from request (73). - Discussion around sex and relationships.
Orthopaedic considerations	Be aware of potential hip and shoulder subluxation and spinal fractures (see bone health). Be aware of scoliosis in individuals who have not had scoliosis correction surgery. Its impact on pressure areas, respiratory, positioning in wheelchair and in bed can be significant and detrimental.
Specialist Services	Wheelchair service – as muscle strength changes more truncal support may be required to maintain function and prevent kyphoscoliosis. Pressure care requirements will change as function reduces. Pain team – Maybe important to manage hip pain due to subluxations and/or back pain due to vertebral fractures. Medical advice is to avoid use of opiate based pain relief (including codeine) because of the risk of significant complications (74). Palliative care team –It is important to involve the team early as they can support the individual and their family well before end of life is imminent. Nutrition support team – required if there is excessive weight loss, repeated infections, skin breakdown or worsening dysphagia. Review management of gastro-oesophageal reflux. Gastroenterology – individuals may present with constipation, flatulence (Especially if on NIV due to air swallowing which can impact ventilation further and led to diverticulitis), and complex feeding discussions. It is important that bowel medications are optimised. Continence nurses – to discuss Farrell valve bags which support with comfort and symptoms of bloating and to support with urinary continence dignity with toileting e.g., convene to support ease of care.

	Cardiology –close monitoring by cardiology is needed. Signs to look out for are dizziness and/or fainting when hoisting, fears of lying down in bed, and palpitations. If there are concerns, link in with the individual's cardiologist. Tissue viability nurses – Look out for any redness or pressure areas. It is likely that if there is a significant loss of weight this increases the risk of pressure areas. Social services – be mindful of carer stress and how much input is required, specifically overnight. Often a need for a carers' assessment. Environmental control team – a review of current controls and whether they are meeting new levels of function. Use of voice activated software. Social services OT – for major adaptations within the home to support independence. Speech and language therapy – management of dysphagia, more frequent choking, more frequent chest infections, weight loss, reduced oral intake. Support with communication aids, voice amplification, eye gaze if required. Psychology – coping with functional loss, discussing fears and future. Peer support if available. Genetic counselling – if considering having a family. Vocational rehabilitation services – supporting access to work and to reassure young men that they can work and have access to support services to gain and maintain employment. Care advisor/Additional professional who can help families secure extra funding for very specialist equipment and advise on what they have a right to receive funding for to avoid unnecessary expenditure.
Education/Work	It is vital to support individuals in their chosen careers (or subject options) and choices around how they spend their time. Communication aids and 1:1 support should be provided where required. It may be helpful to make a referral for vocational support or rehabilitation. You may need to provide supporting information for personal independence payment (PIP) and or employment and support allowance (ESA) and for any reasonable adjustments to maintain equitable access to work/education.

ADDITIONAL RESOURCES

Exercise and activity

Exercise and activity should be done in **an age and stage specific way and advice** is essential (42,75–79) as we recognise that participating in activities of daily life is linked to functional ability (80). For example, swimming/fun in the pool will keep muscles and joints supple (make sure the pool is warm enough as cold muscles do not work so well). Keeping active must be balanced with **pacing advice**, to use energy in the best way. For example, it may be better to push a child in a buggy to the park, so he has the energy to play when he gets there. Here is a Resource on managing fatigue.

Some forms of exercise should be limited or avoided. **Avoid eccentric and heavy concentric loading exercise such as repetitive trampolining** (28)**.** This is because excessive eccentric muscle activity causes damage to the muscle cell membrane and can lead to myoglobinuria (dark coloured urine).

When planning forms of exercise, motivation is key to maintain training routines. It should be balanced with other areas of living family life and most helpfully should create a social arena (81).

Encourage and support play based activity and participation with peers in/out of school/college or work. Ensure school is fully aware of abilities; what to encourage and what to expect. You could refer onto appropriate sport activities such as boccia (UK Boccia) or wheelchair football, both of which are suitable for ambulant and non-ambulant individuals. **Hydrotherapy** is a useful and a safe place to exercise (44).

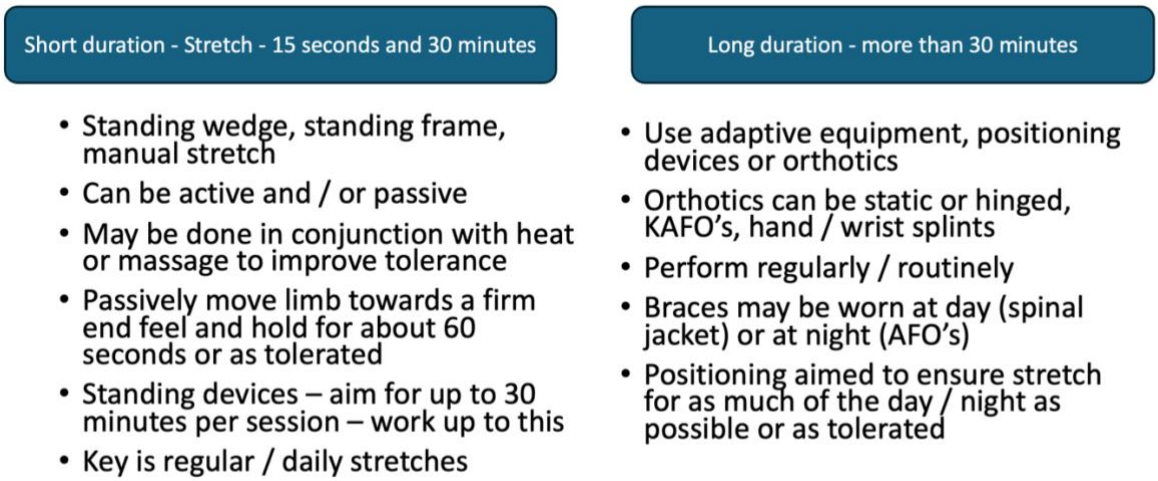
It is important to **promote symmetrical activities** and avoiding asymmetric activities such as a scooter or long periods sitting in an asymmetric position.

Contracture management - Improving & maintaining range of movement

Contractures usually appear alongside muscle weakness when individuals cannot move joints through full range. Muscles that cross multiple joints are at high risk from contractures. As weakness progresses this impacts the biomechanics of walking, and contractures are more likely. In DMD, plantar flexion contractures and anterior pelvic tilt shifts the center of gravity posterior to the hip joint and anterior to the knee and ankle joint. This leads to compensation, inefficient gait, often asymmetrical stance, and excessive lumbar lordosis (82,83). The result is they can stay upright but have a reduced gait speed and stride length. Upper limb contractures usually develop later but may be seen in forearm supinator muscles and long finger flexors earlier if much time is spent on computers and games.

Improving and maintaining range of motion is therefore a key part of management. It can help improve and maintain function and mobility as well as reduce pain. Different methods are available. Contractures may progress suddenly despite good management when growth occurs.

Figure 5. Examples of types of stretch therapy that may be applied - (32)



Stretches

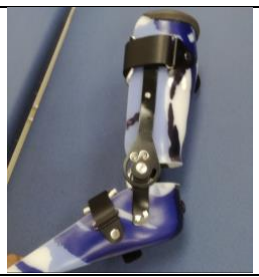
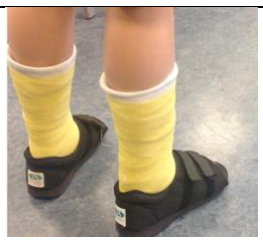
Stretches. Advice should be offered if any joints are tight and how families and individuals can help with improving range of motion. There is limited evidence for effectiveness (84–86) but clinical evidence of response. Stretches can be completed by someone (passive) or by child (active) or assisted by the child (active assisted). Durations of stretch are less important than regularity and have been shown to be effective (87,88). This will depend on their age and understanding. Stretches done in this way are often of a short duration so performing them every day and several times a day and doing them correctly is key. This "gold standard" is hard to achieve for everyone and for every joint. Stretches need to be made part of a daily routine without days off for good behaviour – just like brushing teeth. An easy way to communicate how to stretch a muscle over a joint is by taking a video in a clinic setting of how the therapist does a stretch so this can be shown to all the family and the community team, so management is consistent. Useful links:

- Duchenne UK: Videos of stretches
- MDUK: Stretching Guide
- Guide to passive and active stretches – Action Duchenne
- Parent project: ROM management in DMD
- Scottish Muscle Network: Postural management for adults with DMD

Orthotics

There is no "one" solution for the best type of **ankle orthoses** although centres usually start with provision of an AFO. The best type of ankle orthoses is one that fits well and is used by the individual (89). We strongly recommend that a bespoke night splint is used, not one off the shelf. Ankle contractures can be managed using a nighttime AFO – these are most commonly prescribed (90), serial casting (91,92) or contracture control devices. If tolerated a nighttime gutter/one-piece KAFO can maximise stretch on the muscle.
AFO's are not worn for walking in DMD. They increase the risk of falls and making walking more difficult (93).

Figure 6. Examples of ways of managing tight ankle dorsiflexion.

Daily stretches plus bespoke AFOs worn overnight every night (not when walking). Can be worn if sitting in a wheelchair.	Contracture Control Devices – bespoke and not off the shelf (CCDs). Daily stretches plus CCDs worn 2 hours every day (usually in the evening).	Serial casting. Done under supervision of therapy/orthotics teams. Followed by daily stretching and one of the other two devices.
		

Upper limb contractures are usually more responsive to short duration stretches except perhaps long finger flexors which can benefit from hand splints.

Knee ankle foot orthoses - KAFOs: For those who are finding walking more difficult and would like to continue ambulating, discuss KAFOs suitability with tertiary centre. There is evidence they can prolong assisted walking and standing but limited information on whether they prolong functional walking (94).

General considerations for provision of KAFOs.

1. Is the child and the family motivated to consider continuing to stand and walk and can this be supported at school?
2. Has the child/young person got adequate joint range of movement at ankles, (if more than minus 15° dorsiflexion then balancing in KAFOs is very difficult) if not could this be addressed with serial casting or surgery?
3. Presence of hip and knee contractures – significant contractures may preclude the use of KAFOs.
4. Does the child's specialist centre provide KAFOs or can they be provided elsewhere?

(Association of Paediatric Chartered Physiotherapists (APCP) Guidance for Paediatric Physiotherapists Managing Neuromuscular disorders 2022 provides more guidance p71-73).

Once this has been considered then you can try and mimic KAFOs using a long pair of gaiters with additional support to establish if they can manage the technique required when wearing KAFOs (i.e., hip hitching to gain floor clearance) and their balance. Then liaise with a good specialist neuromuscular orthotist to prescribe the KAFOs followed by rehabilitation afterwards by the physiotherapist and family. Availability varies across tertiary centres.

Footwear

Spending time barefoot is important for development of the small muscles of the foot. When shoes are needed keep the footwear light. Heavy boots are not usually recommended. They may improve a heel down posture, but the excess weight makes walking difficult and tiring. Only use boots for those where they are not yet understanding what their feet are for and need to practice “weighted” standing.

Pain management

Pain is not uncommon so should be a routine part of your assessment (95–97). In younger children muscle cramps may be helped by a warm bath or locally applied heat. In cold weather some children will want to wear additional leggings or football skins under their trousers. This can help with muscle pain. Gentle massage (98) or stretches may help the affected muscles or joints. Analgesia can be used if it’s been approved by the medical team (99). Seek additional medical support as required.

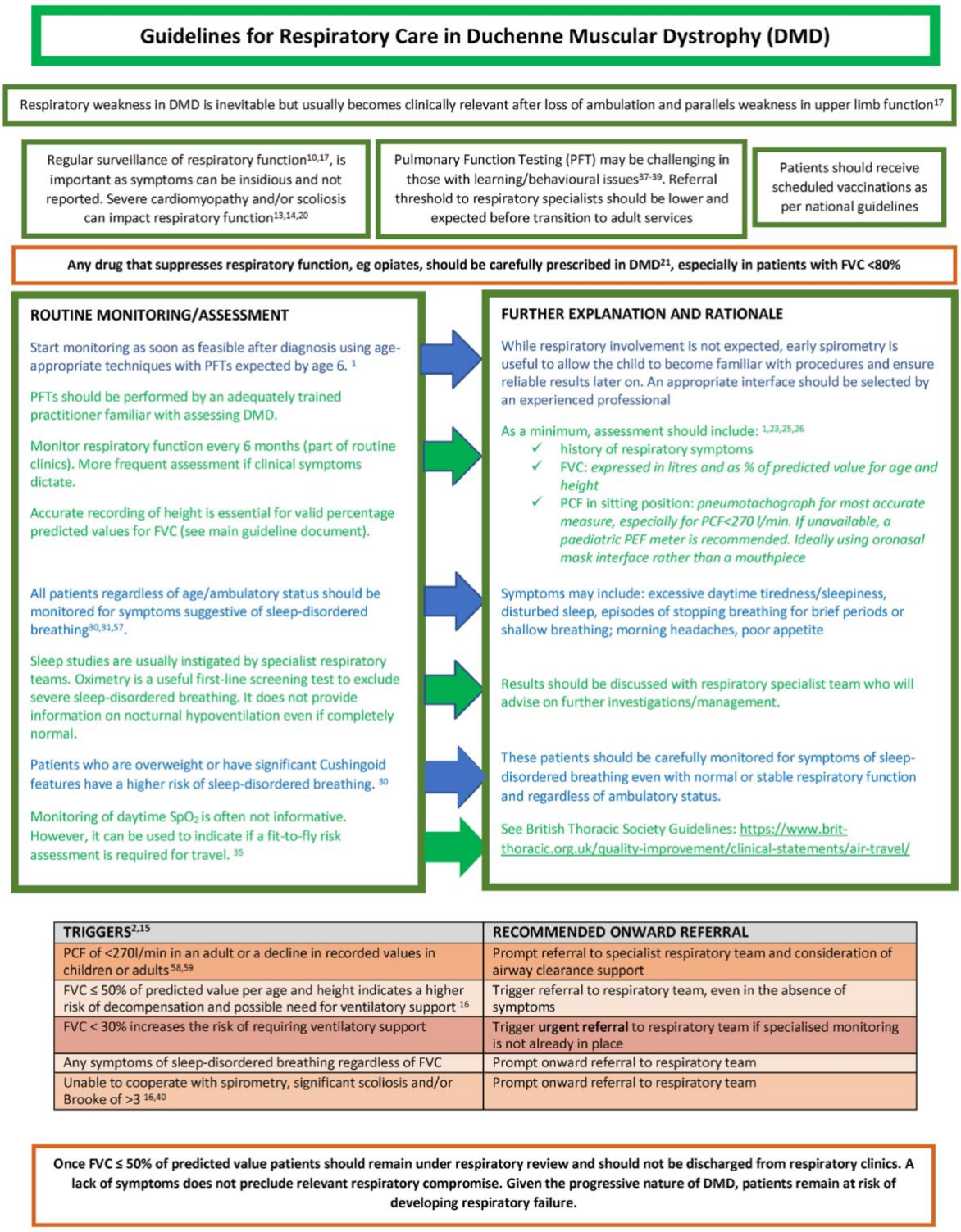
Fatigue management

Keeping active must be balanced with **pacing advice** to use energy in the best way. Saving energy for fun or important things should be considered. Using buggy or wheelchair to get to somewhere and saving walking or upper limb capacity for when you get to the park for example. Here is a resource on managing fatigue.

Respiratory physiotherapy

These are published as separate guidelines created by the respiratory multidisciplinary team. Monitoring respiratory function is imperative, support for breathing is available and can ensure a comfortable breathing experience. Guidelines are accessible here.

Figure 7. Summary schematic of respiratory care in DMD



TRIGGERS[2,15]	RECOMMENDED ONWARD REFERRAL
PCF of <270l/min in an adult or a decline in recorded values in children or adults [58,59]	Prompt referral to specialist respiratory team and consideration of airway clearance support
FVC ≤ 50% of predicted value per age and height indicates a higher risk of decompensation and possible need for ventilatory support [16]	Trigger referral to respiratory team, even in the absence of symptoms
FVC < 30% increases the risk of requiring ventilatory support	Trigger **urgent referral** to respiratory team if specialised monitoring is not already in place
Any symptoms of sleep-disordered breathing regardless of FVC	Prompt onward referral to respiratory team
Unable to cooperate with spirometry, significant scoliosis and/or Brooke of >3 [16,40]	Prompt onward referral to respiratory team

Once FVC ≤ 50% of predicted value patients should remain under respiratory review and should not be discharged from respiratory clinics. A lack of symptoms does not preclude relevant respiratory compromise. Given the progressive nature of DMD, patients remain at risk of developing respiratory failure.

Adult therapy guidelines

Much of the adult guidelines are relevant to younger individuals. This paediatric guide does not seek to replicate what is already in the public domain. Where appropriate we have referenced the guidelines. We suggest you refer to the supplemental material attached to the published article (4) which contains a wealth of information or see pod-nmd.org

Bone health in DMD – Implications for therapists

Individuals living with DMD are at high risk of developing osteoporosis which means they are at a higher risk of fractures (long bones and vertebra). This is because muscle weakness leads to reduced mobility, which reduces bone density but also impairs bone development such that long bones are smaller in size and thinner in diameter. Long-term use of corticosteroids, whilst helpful to maintain muscle function, further adds to the insult on the skeleton by their impact on bone development and reduction in bone density, especially in the spine. Puberty is an important time for bone development - corticosteroids lead to delayed puberty in the majority of boys especially those treated with daily corticosteroids, and therefore delayed puberty (if not appropriately addressed) could further exacerbate the already impaired bone health. Individuals with DMD may spend less time outside, therefore having reduced exposure to the sun which would normally boost vitamin D production – an important component for mineral deposition in bones.

The presence of osteoporosis means they are more likely to fracture without significant trauma. This can be long bone fractures or vertebral fractures. Management by a multidisciplinary team, including endocrinologists, bone specialists and orthopaedic teams for the life span of a person, is key to maintaining function and independence (100).

This guide includes therapeutic considerations for vertebral fractures, long bone fractures and scoliosis management and information around reducing the risk of and managing falls. For more details on bone health and osteoporosis in DMD see here:

Vertebral fractures

Vertebral fractures are extremely common in corticosteroid treated individuals with DMD and have been reported to occur in about 50% of those treated with daily corticosteroid after an average of four years of treatment. They have been detected as early as after six months of corticosteroid treatment. There is usually no history of significant trauma, and these fractures can occur even after handling, such as during a hoist transfer. The current standards of care recommend annual lateral spine imaging to identify vertebral fractures early, to allow introduction of osteoporosis therapy with bisphosphonates. Thus, it is expected that, if identified early, individuals will not experience significant back pain. If they are in pain, an individual should be offered pain relief such as paracetamol. Ibuprofen may be used if advised by a doctor. Intravenous bisphosphonates will also help reduce back pain that is occurring as a result of vertebral fracture.

Assessing for vertebral fractures

- A thorough subjective assessment including questioning of onset, duration and location of pain. Any recent injuries? Unusual or new onset of pain? Prolonged postures? Fatigue? Sleeping position and bed? Wheelchair and seating? Is this pain relieved with painkillers?
- Key pointers to vertebral fracture might include:
    - Back pain that is unusual and new in onset
    - Focal back pain with tenderness to palpation over the spine
    - Pain when turning or rolling in bed
    - Pain experienced when driving over bumps
- Complete a visual analogue pain scale assessment and fill out body chart with the individual.
- An objective assessment including palpation of the spine and surrounding paraspinal musculature.
    - Point tenderness of the central spinous process could be indicative of a vertebral fracture
    - Gentle vibration applied to the spine by the assessor's fist reproducing pain could be indicative of a vertebral fracture
    - Lateral paraspinal muscular tenderness is less likely to be related to vertebral fracture
    - Postural assessment in standing (if appropriate) and in seated position
    - Assess for scoliosis and check Spinal Surgeon reports with x-ray findings of Cobb angle for correlation with pain
- Discuss with Endocrinology for relevance of subjective and objective assessment to most recent lateral spine and DEXA results.
- Repeat lateral spine X-ray if assessment findings indicative of vertebral fracture.

Therapy should:

- Promote good posture (see Scottish Muscle Network leaflet on postural management in Neuromuscular Diseases (NMD)).
- Safe moving and handling including care when turning in bed using the “log rolling” technique. When toileting or moving into a shower chair, avoid too much flexion. When using a hoist, the individual might find it more comfortable to remove any additional strengthening rods in the hoist sling.
- Encourage individuals to keep active, however, overexercising can make the pain worse.
- See also SMN Vertebral Fractures leaflet.

Falls

Following assessment, management aims to reduce the risk of falls/trips either by using specific equipment or aids or by suggesting changes to how an individual gets about. It may include specific visits to the home and/or assessing the risk of falling in certain environments such as school. For more information see pod-nmd.org and the Scottish Muscle Network leaflet on falls in DMD and Vertebral Fractures leaflet.
Consideration should also be given as to how the child or young person gets up off the floor once they fall and a full risk assessment should be carried out and reviewed regularly.

Scoliosis management

Scoliosis prevention. Maintaining ambulatory function for as long as possible with the use of corticosteroids can reduce the risk of the development of scoliosis and can decrease the severity of scoliosis in the long term (47,48,101). For therapists it is important to know that **bracing does not prevent scoliosis**. However, encouraging symmetry and good posture in both sitting and standing is important through all phases of DMD and when the ability to walk is lost, maintaining supported standing can be beneficial. Poor/asymmetrical sitting posture can impact spinal posture which can in turn impair sitting ability. Scoliosis may also adversely affect chest wall and lung compliance which can cause a restrictive lung pattern (102). This further highlights the need to monitor and manage scoliosis.

If surgery is planned for managing pelvic obliquity and spinal deformity, the key aims are to improve sitting balance and prevent scoliosis progression (103).

Principles for therapists are:

Pre-operatively:

- MDT meeting involving families and surgeons so that everyone is aware of benefits and risks.
- This will include respiratory care whilst in hospital based on current status.
- An awareness of the impact of surgery on sitting height and upper limb function (ability to self-feed may be impacted).
- Planning for post-operative mobilisation parameters whilst an in-patient and once discharged.
- Ensuring equipment and care is in place to manage all activities of daily living once home. This includes recline/tilt in space function on a wheelchair for managing pressure care. Does wheelchair need review post-surgery due to postural alterations.

Post-operatively:

- Ensuring local physiotherapists and occupational therapists are aware of discharge and need for evaluation and therapy, including consideration of NIV and secretion management.

Long bone fractures

During what is often a traumatic experience for the whole family, making immediate contact with an individual's neuromuscular team is crucial for advice around any surgery (before, during and after the procedure), and contacting the local physiotherapist and occupational therapist who will work with the specialist team to deliver care afterwards. See podcast links under early ambulant/orthopaedic. For those that are ambulatory there is good evidence that given the right type of surgical fixation and rehabilitation, these individuals can get back on their feet (104). Refer to the DMD Care UK Orthopaedic guidelines for detailed advice (104).

Pre-operative therapy considerations

- Alert community team as post-operative rehabilitation will be imperative.
- Therapy involvement in surgical plan as - for those still walking - fixation which allows rapid post-op mobilisation is key to getting individuals back on their feet. If necessary, discuss this point directly with the surgical team as a surgical plan involving significant periods of immobilisation can lead to unnecessary loss of mobility.
- Involve wheelchair services as the wheelchair may have to be adapted with elevating leg rests to support the fractured leg.
- Ensure adequate liaison with hospital teams take place so that respiratory issues are pre-empted prior to surgery.
- Be aware of hydrocortisone/steroid management plans to avoid adrenal crisis. Remember that people on long-term steroids should be assumed to have adrenal suppression.
- Be aware of the increased risk of fat embolism (5).

Post-operative planning

- Discussion with the family as to how activities of daily living can be achieved in the home and school/workplace environment and what equipment planning might need to be instigated.

Guide to post- operative therapy

- Intensive post-operative therapy is required.
- For those non-weight bearing in hospital this includes exercise for the un-affected leg and static exercises for the affected leg.
- Mobilisation as per the surgeon's instructions. E.g., Sitting out, any early partial weight bearing practice.
- Using a walking frame or parallel bars can be a good starting point and can be replicated at home if discharged early.
- Involving community teams in delivering regular therapy input at home.
- Young people are often frightened of weight-bearing and practicing little and often is more beneficial than only getting seen occasionally.
- Hydrotherapy if the fixation permits can be extremely beneficial.

Fracture – not surgically managed

In some circumstances, surgery may not be possible, and the individual will be discharged home either in a plaster of Paris cast or lightweight splint/gaiter to immobilise the leg. In these circumstances, a rapid response from PT /OT is essential as the young person may be looked after on bed rest until he is able to get into sitting. An OT can provide a bed commode in the first week or so, if pain prohibits the individual from getting onto a regular commode. Moving and handling strategies are imperative with good pain management. Looking after pressure care is a priority, as heels can break down when on bed rest. Many families feel that this is the only way they can manage the young person as the care is the responsibility of the family alone.

Wheelchair services should be alerted as soon as possible as getting back into the wheelchair at the earliest opportunity, whilst managing an immobile leg can take a bit of planning. If bed rest is inevitable, passive movements and stretches are essential to maintain ROM, and gradual sitting over the side of the bed with the foot down will need to be introduced slowly as part of the rehabilitation plan.

PODCASTS: Advice if a child sustains a long bone fracture
Part 1: Immediate care
Part 2: Rehabilitation
See Respiratory Care Guidelines for Emergency Care Imperatives.

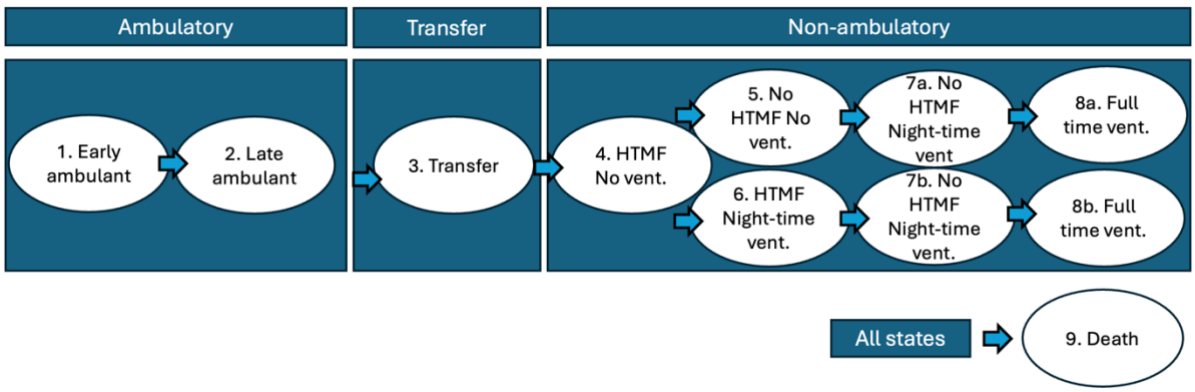
Overlap between physiotherapy and occupational therapy

In the community these two professions often work together and cover similar areas, even overlapping. Often the occupational therapist covers aids, equipment, wheelchairs, and arm function however, their role is extensive. See Occupational Therapy section. Physiotherapists more commonly cover stretches, orthotics, exercise, and activity. Either may visit you in your home, school, or place of work. Not every specialist team has occupational therapists working with them.

Transfer stage

A revised natural history model (NHM) has been developed with input from patients, caregivers and clinicians, to capture health states which reflect clinically important changes in patient care and health-related quality of life (105). Notably, it includes a transfer state, which has not been included in previous NHMs. This study predicts a mean time of approximately 9.5 years in ambulatory states and 1.5 years in the transfer state; a total of approximately 11 years prior to moving into non-ambulatory health states. The 1.5 years spent in the transfer state was consistent with clinical/expert opinion derived from the elicitation exercise.

Figure 8. Developing a Natural History Model for Duchenne Muscular Dystrophy



HTMF – Hand to mouth function. Vent. - Ventilation

The 'transfer stage', which sees the transition from the ambulant to the non-ambulant stage of the disease (not the same as transition from child to adult services), involves a huge change in quality of life for people living with the condition and their carers. This phase is not identified in the current DMD SoC (2). For therapy guidance refer to the late ambulant and early non-ambulant stages.

Suitable measures at this point include the TANS (a clinical synthesis of easier items on the NSAA and useful observed items from the EK2).

Transitioning between paediatric and adult services

This is a huge topic, and a specific working group will aim to tie in all the advice from different working groups to inform families and individuals how best to navigate this phase. New models on making this transition successful are emerging (106). The same plan might not work for all individuals so should be personalised (107).

Guide for therapists preparing for transition to adult services

Transitioning from paediatric medical care to adult medical care can be a challenging process for children with DMD and their families. Here's a guide to preparing for this transition. Not all points are important to the therapy team but understanding the wider considerations is essential to smooth transition.

1. Early Planning: Start planning for the transition early, ideally during adolescence. This allows time to discuss concerns, gather information, and make informed decisions about adult healthcare providers and services.

2. Transition Team: Establish (or know who is responsible for creating) a transition team consisting of paediatric and adult healthcare providers, including neurologists, rehabilitation consultants, pulmonologists, and other specialists involved in DMD care. This team can facilitate coordination and communication between paediatric and adult healthcare settings.

3. Education and Empowerment: Educate the adolescent with DMD about their condition, healthcare needs, and self-management skills. Encourage independence in managing medications, appointments, and health-related decisions to empower them for adulthood.

4. Healthcare Records Transfer: Ensure a smooth transfer of healthcare records from paediatric to adult healthcare providers. This is of particular importance if the young person attends several hospitals in different territorial health board areas, where access to electronic medical records might not be straightforward. The family is advised to keep a copy of all hospital letters as this can help when being asked about dates, medication etc. Health professionals often want to know about medical history, diagnostic tests, treatment plans, and emergency care protocols. Coordinate with healthcare providers to facilitate this process.

5. Adult Healthcare Providers: Research and select adult healthcare providers experienced in treating DMD or neuromuscular conditions. Consider factors such as expertise, accessibility, insurance coverage, and compatibility with the individual's preferences and needs. You may need to find therapy staff willing to learn about DMD and take on care, as services and knowledge of the role of therapy in DMD can be extremely limited.

6. Comprehensive Care Planning: The Transition team should be responsible for developing a comprehensive care plan that addresses the individual's medical, physical, psychological, and social needs. This plan should include strategies for managing DMD-related complications, optimizing quality of life, and promoting independence.

7. Transition Resources and Support: Seek out transition resources and support services available in the community, such as transition clinics, support groups, vocational rehabilitation programs, and advocacy organizations. These resources can provide guidance, peer support, and practical assistance during the transition process.

8. Financial and Legal Planning: Not usually the role of the therapy team.

9. Emotional and Social Support: Address emotional and social aspects of the transition by providing counselling, peer support, and opportunities for social engagement. Encourage the adolescent with DMD to connect with peers facing similar challenges and share experiences.

10. Continuity of Care: Ensure continuity of care by maintaining open communication between paediatric and adult healthcare providers, monitoring health status and treatment adherence. Also address any concerns or challenges that arise during the transition and beyond.

By following these steps and working collaboratively with healthcare providers, families, and support networks, the transition from paediatric to adult medical care can be navigated more effectively, ensuring optimal health outcomes and quality of life for individuals with DMD. The therapy Team play an important role but should not interfere with (only promote) other important spheres of life such as education/work and participation.

Useful resources include:

NICE Quality Standard for Transition

National Confidential Enquiry into Patient Outcome – The inbetweeners (NCEPOD) (2023)

Well Child (2023) 8 Principles for Transition

Wheelchairs

Type of wheelchair: In the first instant a buggy or manual wheelchair may be sufficient. They are usually light and can fit in the back of a car. Once a child is older and finding it harder to self-propel a manual chair or needing more independence, a powered chair may be suitable.
A manual assisted wheelchair may be a suitable interim between these two phases and can promote upper limb function such as e-motion wheels however, they are not usually funded by the NHS so families may need advice and support with accessing funding. Height adjustable wheelchairs are useful for access and for interacting with others.

In addition to facilitating independence and participation, electric powered indoor/outdoor chairs can have a therapeutic benefit. They can help manage pain and weakness. Specialised seating needs careful consideration in supporting progressive muscle weakness and the management of scoliosis. Pain, discomfort, pressure risk, and muscle fatigue may be reduced by use of tilt-in-space (108). The recline function tilts only the back rest and this is a good position for rest. It is also helpful if a gastrostomy is in situ to allow a larger hip angle and can assist with adjusting clothes. The recline and tilt-in space used together are both very useful functions. The more ability the chair has to support changes in position, the better!

It is imperative that posture is reviewed in any wheelchair. Individuals with DMD are more likely to exhibit pelvic obliquity and trunk tilt (109). They tend to abduct their hips due to tightness in the iliotibial band so it's essential to ensure the legs are supported with lateral abduction supports/adductor pads.

There is evidence to suggest that powered standing wheelchairs are of benefit and criteria have been described for successful use of these (ability to stand comfortably for 10 minutes, nor more than 10 degrees loss of ankle ROM and subject motivation to use the standing device) (110,111).

Basic principles:

- The correct size especially for width and leg length (given that short stature is more usual).
- Promotes symmetry.
- Arm rests at the correct height. Arm rests too low – excessive traction is placed on shoulder joint and the child leans which leads to asymmetry and poor spinal posture. Arm rests too high – children either don't use the arm rests and may lean or hunch forward which leads to poor spinal posture.
- Includes foot plates so feet are supported in plantar grade at the ankles.
- Suitable cushion or base.
- Suitable back support – with lateral supports if necessary.
- Adductor pads to promote good lower limb alignment and foot position but which can be swung away for toileting as required.
- Head support if appropriate for dynamic or static use, required if wheelchair is being used in a car.
- Sufficient arm strength is required for manual wheelchair provision to be appropriate.

Timing of provision: Where fatigue appears to be impacting daily routine and/or there is an increasing falls risk, particularly outdoors.

Provision: Your community or neuromuscular physiotherapist can initiate a referral for a wheelchair assessment via the local NHS Wheelchair Service.

www.england.nhs.uk/wheelchair-services/ + Scotland and Wales and Northern Ireland.
Provision via NHS wheelchair services can be made for ages 3+
Adapted buggies can be provided for ages 3 and under where indicated.

Private/partially funded purchase wheelchairs

If NHS provided wheelchairs fail to meet the needs of the child there are several resources which can help with a private purchase.
It is important that chairs provided in this way are maintained and checked (as any chair would be) and that families understand how repairs and servicing will be carried out.

Choosing the right wheelchair
https://www.which.co.uk/reviews/mobility-equipment/article/choosing-the-best-wheelchair-aAfTd2J7ji39
https://livingmadeeasy.org.uk/
Financial support
https://www.whizz-kidz.org.uk/families/application-process
https://www.musculardystrophyuk.org/get-support/adaptations-and-equipment/joseph-patrick-trust-grants

Other considerations:

- Ensure updates to an Education, Health and Care Plan (EHCP) to encompass wheelchair use.
- Consider accessibility of transport – see https://www.gov.uk/financial-help-disabled/vehicles-and-transport for further advice.
- Consider accessibility of the home - liaise with your Occupational Therapist. In the UK this is usually your social services OT.

Behavioural and learning issues

For some children the clinic environment is a tough space to navigate given their behavioural, sensory or autistic spectrum difficulties. DMD has a high rate of cognitive and learning disabilities as well as neurobehavioral disorders, some of which have been associated with disruption of dystrophin isoforms in the brain (112). In this retrospective study of 59 boys the cognitive and neurobehavioral profile of boys was as follows: Full-scale IQ of < 70 was seen in 27%; learning disability in 44%, intellectual disability in 19% (these three values all refer to a learning/intellectual disability); attention-deficit/hyperactivity disorder in 32%; autism spectrum condition 15%; and anxiety in 27%. It is important that the therapy team are aware of any or all of these issues and modify their approach accordingly. It can also be a highly stressful environment for the parents and family. Keep appointments short and minimise the time spent in clinic by combining examinations with the medical team. For children who are unable to cooperate use parent report (such as the NSAA-OR) and use your time watching the child play or move with their parents. Ask parents to direct their child to show you what they can do.

Occupational therapy

Paediatric occupational therapy for children with DMD can greatly enhance care across all stages of the disease.

In the early stages, therapy focuses on maintaining independence in daily activities and optimizing motor skills. As the disease progresses, therapy shifts towards managing muscle weakness, preventing contractures, and adapting environments to support changing abilities. In later stages, occupational therapists assist with mobility devices, adaptive equipment, and strategies to maintain quality of life and participation in activities.

Overall, paediatric occupational therapy for children with DMD is holistic and client-centred, addressing the unique needs and abilities of each child while empowering them to live life to the full. Collaboration with families, caregivers, and other healthcare professionals is essential to provide comprehensive care and support throughout the journey with DMD.

Specialist equipment – OT

Specialist equipment can play a crucial role in promoting function and independence for children with DMD. Here are some examples of specialist equipment commonly used:

1. Mobility Devices: Wheelchairs- manual or powered wheelchairs can provide independent mobility for children with DMD who have difficulty walking or standing for extended periods.

2. Assistive Technology: Environmental Control Systems- remote control systems or voice-activated devices enable children with limited mobility to control electronic devices, such as lights, TVs, and thermostats, within their environment. Communication Devices: Augmentative and alternative communication (AAC) devices, including speech-generating devices, communication boards, or software apps, help children with DMD communicate effectively, especially as speech abilities decline. Computer Access Aids: Adaptive keyboards, mice, switches, and software programs enable children with DMD to access computers, tablets, and other digital devices independently.

3. Orthotic Devices: Hand splints- customized hand splints or braces can support hand function, prevent contractures, and facilitate activities such as grasping, writing, and self-feeding.

4. Adaptive Seating and Positioning: Specialized seating systems- customized seating systems, such as supportive chairs, recliners, or tilt-in-space and/or recline wheelchairs, provide comfort, postural support, and pressure relief for children with DMD who spend extended periods sitting. Cushions and Mattresses: Pressure-relieving cushions, alternating pressure mattresses, and foam positioning aids help prevent pressure ulcers, promote comfort, and support optimal positioning during sleep and rest periods. Some beds provide movement during the night but are not always effective (due to slippage for example).

5. Bathroom and Toileting Aid: toilet supports- toilet frames, raised toilet seats, and grab bars assist children with DMD in safely transferring on and off the toilet and maintaining stability during toileting activities. Bathing Aids: Shower chairs, bath benches, and handheld showerheads enable children with DMD to bathe independently or with minimal assistance while maintaining safety and comfort.

6. Assistive Devices for Daily Living: Adaptive utensils- modified utensils, cups, and feeding devices with built-up handles, angled designs, or specialized grips facilitate independent eating and drinking for children with DMD. Dressing Aids: Button hooks, zipper pulls, elastic shoelaces, and dressing sticks assist children with DMD in managing clothing fasteners and dressing tasks independently.

By incorporating specialist equipment into the daily routines and environments of children with DMD, occupational therapists and healthcare professionals can enhance function, promote independence, and improve overall quality of life. Customization and ongoing assessment are essential to ensure that equipment meets the individual needs and abilities of each child with DMD.

Educational interventions – OT

Occupational therapists play a vital role in educational interventions for children with DMD, focusing on optimizing participation, promoting independence, and ensuring accessibility. Here are some key interventions they may implement:

1. Functional Assessments: Conduct comprehensive assessments to identify the child's strengths, challenges, and specific needs related to DMD. Assessments may include fine and gross motor skills, ADLs, mobility, positioning, and environmental factors.

2. Individualized Education Plans (IEPs): If applicable, collaborate with teachers, parents, and other educational professionals to develop and implement these plans which should be tailored to the child's abilities and educational goals. This may include accommodations, modifications, and assistive technology to support learning and participation in the classroom. Education, Health and Care Plans (EHCP) in England and support plans should be used.

3. Adaptive Equipment and Assistive Technology: Recommend and provide adaptive equipment and assistive technology to facilitate access to educational materials, activities, and environments. This may include specialized seating, mobility devices, computer access aids, and communication devices.

4. Environmental Modifications: Evaluate and make environmental modifications within the school setting to enhance accessibility, safety and promote social interaction for the child with DMD. This may involve adjusting classroom layout, furniture, and equipment to accommodate mobility aids and optimize positioning.

5. Motor Skills Training: Provide motor skills training and interventions to improve or maintain functional abilities, such as fine motor skills, handwriting, self-care tasks, and transitions between activities. Use evidence-based strategies and adaptive techniques to address limitations associated with DMD.

6. Energy Conservation Strategies: Teach energy conservation techniques and pacing strategies to help the child manage fatigue and conserve energy throughout the school day. This may include scheduling rest breaks, prioritizing tasks, and utilizing ergonomic principles to reduce exertion.

7. Social Skills Development: Facilitate social skills development and peer interactions to foster social inclusion and participation in school activities. Provide guidance on effective communication, social cues, and building friendships within the classroom and school community.

8. Transition Planning: Collaborate with the school team and family to develop transition plans for key educational transitions, such as from elementary to middle school or high school to post-secondary education such as university or college. Address concerns related to changes in routines, environments, and support services. This will include advice on accessible living at university and/or liaising with disability services at the university as well as teaching and studying rooms.

9. Educational Advocacy: Advocate for the educational rights and needs of children with DMD, ensuring access to appropriate educational programs, services, and accommodations. Provide education and training to school staff and administrators on DMD-related considerations and best practices.

10. Collaboration and Consultation: Maintain open communication and collaboration with school staff, parents, healthcare providers, and community resources to ensure coordinated support and continuity of care for the child with DMD. Offer consultation and support to address emerging needs and challenges in the school environment.

11. OT can offer support for sensory processing issues. Although there is no research around this in DMD it is reporting by families. See Parent Project Sensory Processing Disorder.

By implementing these interventions and strategies, occupational therapists can support children with DMD in maximizing their educational potential, promoting independence, and fostering meaningful participation in the school setting.

Mental health and well-being – OT

Occupational therapists can play a significant role in promoting mental health and well-being for children with DMD through various interventions and support strategies. Here are some ways occupational therapists can promote **mental health and well-being for children with DMD**:

1. Positive Reinforcement and Self-Esteem Building: Encourage and reinforce the strengths, abilities, and achievements of children with DMD to enhance their self-esteem and self-confidence. Focus on their talents, interests, and accomplishments, and empower them to set realistic goals and pursue meaningful activities that promote a sense of competence and mastery.

2. Social Skills Development: Facilitate social skills development and peer interactions to foster social inclusion, friendship, and support networks for children with DMD. Provide opportunities for socialization, group activities, and participation in school, community, or recreational programs to enhance social connections and reduce feelings of isolation or loneliness.

3. Adaptive Leisure and Recreational Activities: Adapt leisure and recreational activities to accommodate the abilities and interests of children with DMD, ensuring access to enjoyable and fulfilling experiences that promote relaxation, enjoyment, and social engagement. Offer adaptive sports, arts and crafts, music therapy, or other leisure pursuits tailored to the child's preferences and physical capabilities.

4. Relaxation Techniques: Teach relaxation, and stress management techniques to help children with DMD cope with pain, fatigue, and emotional distress. Deep breathing exercises, guided imagery, and relaxation techniques may reduce tension, and improve emotional well-being.

5. Emotional Support: Provide emotional support to children with DMD and their families to address feelings of anxiety, depression, frustration, or grief related to the diagnosis and progression of the condition. Offer a safe space for children to express their emotions, concerns, and fears, and help them develop coping strategies to manage stress and uncertainty.

6. Education and Advocacy: Educate children with DMD, their families, caregivers, and peers about the condition, its impact on physical and emotional health, and strategies for coping and adaptation. Advocate for inclusive practices, accessibility, and accommodations in educational, recreational, and community settings to promote full participation and equal opportunities for children with DMD.

7. Family Support and Education: Provide support, education, and resources to families of children with DMD to help them cope with the challenges of caregiving, navigate the healthcare system, and access available support services. Offer guidance on adaptive parenting strategies, sibling support, and family-centred care approaches to promote resilience and well-being within the family unit.

8. Collaboration with Multidisciplinary Team: Collaborate with a multidisciplinary team of healthcare professionals, including psychologists, social workers, physical therapists, and physicians, to address the holistic needs of children with DMD and ensure comprehensive care that considers physical, emotional, and psychosocial well-being.

By integrating these approaches into their practice, occupational therapists can contribute to the promotion of mental health and well-being for children with DMD, empowering them to lead fulfilling and meaningful lives despite the challenges posed by the condition.

Appendix A – Key recommendations improving SoC for Therapy in DMD

Regular appointments are critical (6 monthly SP NM and more frequent in community) and should be more frequent if necessary*.
Discourage 'as requested appointments' or 'episodes of care' where possible as this results in reactive management rather than proactive management.
*More frequently: New diagnosis, long bone fracture, vertebral fracture, steroid changes, parental concerns significant loss of function, specific therapy input

Assess and manage. Whole team should be aware of the link between assessment and management and therefore if things change how management might need to be updated.
Time spent with individual's should be **driven by changes and issues**
Issues include: Function – upper and lower limbs), mobility including transfers and risk of falls, pain & cramps, contractures, orthotics, spine /posture, activity & exercise, fatigue, equipment, wheelchair, respiratory as well as other forms of support (emotional, learning, participation)

Content of Therapy Appointments
Families should be aware that **not all assessments or forms of management need to happen at every appointment**. This is particularly true in the community where issues often relate to a particular location. E.g. Home or place of education
Standard assessments should be used where possible for a specific issue. E.g. North Star Ambulatory Assessment (NSAA) to measure function and response to steroids or a specific postural assessment for evaluating specialist seating.
Advice should be given even if it is to reassure the family that they should continue with current management. This should include hands on advice and instruction.
Structure of appointments should help families avoid repeating the same verbal information too many times and making use of joint appointments to save time and improve communication. This may be a MDT format but provision should be made for discreet conversations where necessary

Communication.
Families want to know what is being assessed and why and may want to know the scores / results of any tests, but this should not be assumed.
Having reliable methods for families to contact the therapy team is critical, be it a telephone number which takes messages and / or specific or team emails.
Communication could be written advice, photos, videos on family phones, emails, phone calls. Do families know where the teams are based and how to contact them? Have you informed the family of changes.
Use joint working to improve communication and knowledge amongst teams
Use the opportunity when demonstrating therapy to communicate this to the wider team effectively (other family members, community, educational establishment)
The community would benefit from easier ways to share information between teams and families and update these quickly and efficiently.
Make it clearer to families the role of OT in all areas and how to access and source OT support.

Additional recommendations. Younger boys are best seen earlier in the day. Older children / young people often later appointments as getting up and out is a lengthy process
Having access to a hoist if needed when they attend clinic so assessments can be conducted properly.
Deliver a strong Transition Team to support the process of transition from child to adult health and social care

Resources and education
A more detailed guide to care could improve knowledge and care of those with DMD
Community therapists should be offered more opportunities for learning and peer support in specific therapies
Education of therapists should include the learning and behavioural issues

Service provision
Specialist therapy teams and community teams need support on comparing current delivery to SoC and using the information on inadequate provision to commission better services or access to equipment etc.
Business plan support could help specialist teams secure funding for therapy provision

Appendix B - Contributors to this guide

Name	Location
Aleks Pietrusz	Department of Neuromuscular Disease, UCL Queen Square Institute of Neurology
Amy Wolfe	The Dubowitz Neuromuscular Centre, UCL Great Ormond Street, Institute of Child Health, London
Andrea Tobin	Children's Health, Ireland
Anna Mayhew	John Walton Muscular Dystrophy Research Centre, Newcastle University
Charlotte Massey	Department of Neuroscience, University of Sheffield
Denise Wooding	Alder Hey Children's NHS Foundation Trust
Emma Gallagher	University Hospitals Bristol NHS Trust Foundation
Hayley Davis	Cardiff and Vale University Hospital Board
Heather McMurchie	Heartlands Hospital Birmingham
Jassi Sodhi	John Walton Muscular Dystrophy Research Centre, Newcastle University
Lindsey Pallant	Leeds Teaching Hospitals NHS Trust
Marina Di Marco	Royal Hospital for Children, Glasgow
Marion Main	The Dubowitz Neuromuscular Centre, UCL Great Ormond Street, Institute of Child Health, London
Meredith James	John Walton Muscular Dystrophy Research Centre, Newcastle University
Nicola Burnett	Great Ormond Street Hospital
Rosanna Rabb	Heartlands Hospital Birmingham
Sarah Brown	Royal Hospital for Children, Glasgow
Sejal Thakrar	Family Representative
Sinead Warner	Manchester University NHS Foundation Trust
Suni Narayan	University Hospital Southampton

We acknowledge the significant input of other DMD Care UK working groups to this document:

- Orthopaedic working group
- Bone & Endocrine working group
- Education working group
- Psychosocial working group
- Transition working group
- Spinal working group
- Palliative working group

We would like to dedicate these therapy guidelines to Shiv Thakrar [*February 2011 – March 2023*] and acknowledge the support and input Sejal Thakrar has given to DMD Care UK and the Therapy Working Group in particular.

Appendix C – Abbreviations

Abbreviation	Full term
6MWT	6-minute walk test
AAC	Augmentative and alternative communication
ADLs	Activities of daily living
AFOs	Ankle foot orthoses
AIMS	Alberta Infant Motor Scale
APCP	Association of Paediatric Chartered Physiotherapists
ASD	Autism spectrum disorder
CAHMS	Child and adolescent mental health services
CK	Creatine kinase
DMD	Duchenne muscular dystrophy
DMD SAT	Duchenne muscular dystrophy functional ability self-assessment tool
E.G.	Example
EHCP	Education, Health and Care Plan(s)
EK2	Egen Klassifikation Scale
ESA	Employment and support allowance
FVC	Forced vital capacity
I.E.	Id est | that is
KAFOs	Knee ankle foot orthoses
LFF	Long finger flexors
MDT	Multidisciplinary Team
N.B.	Nota bene | note well
NHM	Natural history model
NIV	Non-invasive ventilation
NSAA	North Star Ambulatory Assessment
OT	Occupational Therapists
PCF	Peak cough flow
PE	Physical Education
PIP	Personal independence payment
PROMs	Patient reported outcome measures
PT	Physiotherapists
PUL 2.0	Performance of Upper Limb
QoL	Quality of Life
QoLNMD	Quality of Life for slowly progressive neuromuscular disorders
ROM	Range of motion
SENCO	Special educational needs coordinator
SoC	Standards of Care
TA	Tendoachillies
TANS	Transition Assessment North Star
UK	United Kingdom
VAS	Visual analogue pain scale

References

1. Birnkrant DJ, Bushby K, Bann CM, Alman BA, Apkon SD, Blackwell A, et al. Diagnosis and management of Duchenne muscular dystrophy, part 2: respiratory, cardiac, bone health, and orthopaedic management. Vol. 17, The Lancet Neurology. 2018. p. 347–61.
2. Birnkrant DJ, Bushby K, Bann CM, Apkon SD, Blackwell A, Brumbaugh D, et al. Diagnosis and management of Duchenne muscular dystrophy, part 1: diagnosis, and neuromuscular, rehabilitation, endocrine, and gastrointestinal and nutritional management [Internet]. Vol. 17, The Lancet Neurology. Elsevier; 2018 [cited 2018 Feb 1]. p. 211–2. Available from: http://www.thelancet.com/journals/laneur/article/PIIS1474-4422(18)30024-3/fulltext
3. Birnkrant DJ, Bushby K, Bann CM, Apkon SD, Blackwell A, Colvin MK, et al. Diagnosis and management of Duchenne muscular dystrophy, part 3: primary care, emergency management, psychosocial care, and transitions of care across the lifespan [Internet]. Vol. 17, The Lancet Neurology. Elsevier; 2018 [cited 2018 Feb 1]. p. 445–55. Available from: http://www.thelancet.com/journals/laneur/article/PIIS1474-4422(18)30026-7/fulltext
4. Narayan S, Pietrusz A, Allen J, Dimarco M, Docherty K, Emery N, et al. Guidelines Adult North Star Network (ANSN): Consensus Document for Therapists Working with Adults with Duchenne Muscular Dystrophy (DMD)-Therapy Guidelines. J Neuromuscul Dis. 2022;9:365–81.
5. Childs AM, Turner C, Astin R, Bianchi S, Bourke J, Cunningham V, et al. Development of respiratory care guidelines for Duchenne muscular dystrophy in the UK: key recommendations for clinical practice State of the art review. [cited 2024 Jan 23]; Available from: http://thorax.bmj.com/
6. Burnett N, Crook V, Sarkozy A. 53 The use of virtual focus groups to evaluate and develop the new neuromuscular clinical and physiotherapy provision for non-ambulant boys with Duchenne muscular dystrophy. 2021 Dec;A20.1-A20.
7. Kinnett K, Rodger S, Vroom E, Furlong P, Aartsma-Rus A, Bushby K. Imperatives for DUCHENNE MD: A simplified guide to comprehensive care for duchenne muscular dystrophy. PLoS Curr. 2015;7(MUSCULARDYSTROPHY).
8. Heutinck L, Van Gameren M, Verschuuren JJGM, Geurts ACH, Jansen M, De Groot IJM. Clinical Management of Duchenne Muscular Dystrophy in the Netherlands: Barriers to and Proposals for the Implementation of the International Clinical Practice Guidelines. J Neuromuscul Dis. 2021;8(4):503–12.
9. Hernández-Sánchez A, Parra-Sánchez L, Montolio M, Rueda-Ruzafa L, Ortiz-Comino L, Sánchez-Joya MDM. Family Involvement and at-Home Physical Therapy on Duchenne Muscular Dystrophy: A Randomized Controlled Trial. Pediatr Neurol. 2024 Mar 1;152:34–40.
10. van Dommelen P, van Dijk O, de Wilde J, Verkerk P. EP.76Developmental delay in Duchenne muscular dystrophy. Neuromuscular Disorders. 2019 Oct;29:S174.
11. Connolly AM, Florence JM, Cradock MM, Eagle M, Flanigan KM, McDonald CM, et al. One Year Outcome of Boys with Duchenne Muscular Dystrophy Using the Bayley-III Scales of Infant and Toddler Development. Pediatr Neurol. 2014;50(6):557–63.
12. Darrah J, Piper M, Watt MJ. Assessment of gross motor skills of at-risk infants: predictive validity of the Alberta Infant Motor Scale. Dev Med Child Neurol [Internet]. 1998 Jul [cited 2016 Nov 3];40(7):485–91. Available from: http://www.ncbi.nlm.nih.gov/pubmed/9698062
13. Scott E, Eagle M, Mayhew A, Freeman J, Main M, Sheehan J, et al. Development of a Functional Assessment Scale for Ambulatory Boys with Duchenne Muscular Dystrophy. Physiotherapy Research International. 2012;17(2).
14. Mayhew AG, Cano SJ, Scott E, Eagle M, Bushby K, Manzur A, et al. Detecting Meaningful Change Using The North Star Ambulatory Assessment In Duchenne Muscular Dystrophy. Dev Med Child Neurol. 2013;55(11).
15. Mayhew A, Cano S, Scott E, Eagle M. Moving towards meaningful measurement: Rasch analysis of the North Star Ambulatory Assessment in Duchenne muscular dystrophy. Medicine & Child … [Internet]. 2011 [cited 2016 Nov 3]; Available from: http://onlinelibrary.wiley.com/doi/10.1111/j.1469-8749.2011.03939.x/full
16. Mercuri E, Coratti G, Messina S, Ricotti V, Baranello G, D'Amico A, et al. Revised north star ambulatory assessment for young boys with Duchenne muscular dystrophy. PLoS One [Internet]. 2016 [cited 2016 Nov 15];11(8):e0160195. Available from: http://www.ncbi.nlm.nih.gov/pubmed/27494024
17. Scott OM, Hyde SA, Goddard C, Dubowitz V. Prevention of deformity in Duchenne muscular dystrophy. A prospective study of passive stretching and splintage. Physiotherapy [Internet]. 1981 Jun [cited 2017 Dec 7];67(6):177–80. Available from: http://www.ncbi.nlm.nih.gov/pubmed/7301976
18. Romkes J, Viehweger E, Bracht-Schweizer K. Upper body gait deviations in children with Duchenne muscular dystrophy. Gait Posture. 2021 Oct;90:213–4.
19. Gajdosik RL, Bohannon RW. Clinical measurement of range of motion. Review of goniometry emphasizing reliability and validity. Phys Ther [Internet]. 1987 [cited 2024 Jul 3];67(12):1867–72. Available from: https://pubmed.ncbi.nlm.nih.gov/3685114/

20. Akkurt L, Gürbüz IA, Karaduman A, Yilmaz ÖT. Lower limb flexibility in children with Duchenne muscular dystrophy: Effects on functional performance. Pediatr Exerc Sci. 2019 Feb 1;31(1):42–6.
21. Woof AL, Selby K, Harris SR. Ankle contractures and functional motor decline in Duchenne muscular dystrophy. Brain Dev. 2022 Feb 1;44(2):105–13.
22. Duong T, Canbek J, Fernandez-Fernandez A, Henricson E, Birkmeier M, Siener C, et al. Knee Strength and Ankle Range of Motion Impacts on Timed Function Tests in Duchenne Muscular Dystrophy: In the Era of Glucocorticoids. J Neuromuscul Dis. 2022;9(1):147–59.
23. Geiger R, Strasak A, Treml B, Gasser K, Kleinsasser A, Fischer V, et al. Six-Minute Walk Test in Children and Adolescents. Journal of Pediatrics [Internet]. 2007 Apr 1 [cited 2023 Jun 5];150(4):395-399.e2. Available from: http://www.jpeds.com/article/S0022347607000145/fulltext
24. Goemans N, Klingels K, Van Den Hauwe M, Boons S, Verstraete L, Peeters C, et al. Six-minute walk test: Reference values and prediction equation in healthy boys aged 5 to12 years. PLoS One. 2013;
25. Pane M, Mazzone ES, Sormani MP, Messina S, Vita GL, Fanelli L, et al. 6 minute walk test in Duchenne MD patients with different mutations: 12 month changes. PLoS One. 2014;9(1).
26. Lammers AE, A.A. Hislop, Flynn Y, Haworth SG. The 6-minute walk test: normal values for children of 4–11 years of age. Arch Dis Child [Internet]. 2008 Jun 1 [cited 2023 Jun 5];93(6):464–8. Available from: https://adc.bmj.com/content/93/6/464
27. Rose KJ, Burns J, Wheeler DM, North KN. Interventions for increasing ankle range of motion in patients with neuromuscular disease. In: Rose KJ, editor. Cochrane Database of Systematic Reviews [Internet]. Chichester, UK: John Wiley & Sons, Ltd; 2010 [cited 2016 Nov 14]. p. CD006973. Available from: http://www.ncbi.nlm.nih.gov/pubmed/20166090
28. Lovering RM, Brooks S V. Eccentric exercise in aging and diseased skeletal muscle: Good or bad? J Appl Physiol. 2014 Jun 1;116(11):1439–45.
29. Brunherotti MA, Sobreira C, Rodrigues-Júnior AL, Renato de Assis M, Terra Filho J, Baddini Martinez JA. Correlations of Egen Klassifikation and Barthel Index scores with pulmonary function parameters in Duchenne muscular dystrophy. Heart Lung [Internet]. 2007 Mar [cited 2022 Mar 31];36(2):132–9. Available from: https://pubmed.ncbi.nlm.nih.gov/17362794/
30. Lee HN, Sawnani H, Horn PS, Rybalsky I, Relucio L, Wong BL. The Performance of the Upper Limb scores correlate with pulmonary function test measures and Egen Klassifikation scores in Duchenne muscular dystrophy. Neuromuscular Disorders. 2016;
31. Fagoaga J, Girabent-Farres M, Bagur-Calafat C, Steffensen BF. Evolution of functional capacity, assessed with the Egen Klassifikation scale, in the Spanish population with spinal muscular atrophy or Duchenne muscular dystrophy. A three year longitudinal study. Rev Neurol [Internet]. 2015 Oct 16 [cited 2016 Nov 15];61(8):344–8. Available from: http://www.ncbi.nlm.nih.gov/pubmed/26461127
32. Deconinck, Nicolas; Goemans N. Neuromuscular Disorders in Children. London: Mac Keith Press; 2019.
33. Mayhew AG, Coratti G, Mazzone ES, Klingels K, James M, Pane M, et al. Performance of Upper Limb module for Duchenne muscular dystrophy. Dev Med Child Neurol. 2020;62(5).
34. Pane M, Mazzone ES, Fanelli L, De Sanctis R, Bianco F, Sivo S, et al. Reliability of the Performance of Upper Limb assessment in Duchenne muscular dystrophy. Neuromuscular Disorders [Internet]. 2014 [cited 2016 Nov 15];24:201–6. Available from: http://dx.doi.org/10.1016/j.nmd.2013.11.014
35. Mayhew A, Mazzone ES, Eagle M, Duong T, Ash M, Decostre V, et al. Development of the Performance of the Upper Limb module for Duchenne muscular dystrophy. Dev Med Child Neurol. 2013;55(11).
36. Wu KW, Yu CH, Huang TH, Lu SH, Tsai YL, Wang TM, et al. Children with Duchenne muscular dystrophy display specific kinematic strategies during obstacle-crossing. Scientific Reports 2023 13:1 [Internet]. 2023 Oct 10 [cited 2024 Jul 3];13(1):1–16. Available from: https://www.nature.com/articles/s41598-023-44270-9
37. Griffin J, Lall R, Bruce J, Withers E, Finnegan S, Lamb SE, et al. Comparison of alternative falls data collection methods in the Prevention of Falls Injury Trial (PreFIT). J Clin Epidemiol. 2019 Feb 1;106:32–40.
38. Myles PS, Urquhart N. The Linearity of the Visual Analogue Scale in Patients with Severe Acute Pain. https://doi.org/101177/0310057X0503300108 [Internet]. 2005 Feb 1 [cited 2024 Jul 3];33(1):54–8. Available from: https://journals.sagepub.com/doi/abs/10.1177/0310057X0503300108
39. Shields BJ, Cohen DM, Harbeck-Weber C, Powers JD, Smith GA. Pediatric pain measurement using a visual analogue scale: a comparison of two teaching methods. Clin Pediatr (Phila) [Internet]. 2003 [cited 2024 Jul 3];42(3):227–34. Available from: https://pubmed.ncbi.nlm.nih.gov/12739921/
40. Shields BJ, Palermo TM, Powers JD, Grewe SD, Smith GA. Predictors of a child's ability to use a visual analogue scale. Child Care Health Dev [Internet]. 2003 Jul [cited 2024 Jul 3];29(4):281–90. Available from: https://pubmed.ncbi.nlm.nih.gov/12823333/
41. Pfeiffer KA, Pivarnik JM, Womack CJ, Reeves MJ, Malina RM. Reliability and validity of the Borg and OMNI rating of perceived exertion scales in adolescent girls. Med Sci Sports Exerc [Internet]. 2002 Dec 1 [cited 2024 Jul 3];34(12):2057–61. Available from: https://pubmed.ncbi.nlm.nih.gov/12471316/

42. Leone E, Pandyan A, Rogers A, Kulshrestha R, Hill J, Philp F. Effectiveness of conservative non-pharmacological interventions in people with muscular dystrophies: a systematic review and meta-analysis. J Neurol Neurosurg Psychiatry. 2023 Dec 12;jnnp-2023-331988.
43. Lott DJ, Taivassalo T, Cooke KD, Park H, Moslemi Z, Batra A, et al. Safety, feasibility, and efficacy of strengthening exercise in Duchenne muscular dystrophy. Muscle Nerve. 2021 Mar 1;63(3):320–6.
44. Hind D, Parkin J, Whitworth V, Rex S, Young T, Hampson L, et al. Aquatic therapy for children with duchenne muscular dystrophy: A pilot feasibility randomised controlled trial and mixed-methods process evaluation. Health Technol Assess (Rockv). 2017;21(27):1–120.
45. Mayer OH, Finkel RS, Rummey C, Benton MJ, Glanzman AM, Flickinger J, et al. Characterization of pulmonary function in Duchenne Muscular Dystrophy. Pediatr Pulmonol. 2015;50(5):487–94.
46. Hiscock A. The role of the neuromuscular care advisor in Duchenne muscular dystrophy. Paediatrics and Child Health (United Kingdom). 2017 Oct 1;27(10):481–3.
47. Alman BA, Raza SN, Biggar WD. Steroid treatment and the development of scoliosis in males with duchenne muscular dystrophy. J Bone Joint Surg Am [Internet]. 2004 [cited 2024 Jul 3];86(3):519–24. Available from: https://pubmed.ncbi.nlm.nih.gov/14996877/
48. Lebel DE, Corston JA, McAdam LC, Biggar WD, Alman BA. Glucocorticoid treatment for the prevention of scoliosis in children with Duchenne muscular dystrophy: Long-term follow-up. Journal of Bone and Joint Surgery. 2013 Jun 19;95(12):1057–61.
49. Wood C, Cheetham T, Guglieri M, Bushby K, Owen C, Johnstone H, et al. Testosterone Treatment of Pubertal Delay in Duchenne Muscular Dystrophy. Neuropediatrics [Internet]. 2015 Sep 26 [cited 2017 Nov 11];46(06):371–6. Available from: http://www.thieme-connect.de/DOI/DOI?10.1055/s-0035-1563696
50. Voet NBM, van der Kooi EL, van Engelen BGM, Geurts ACH. Strength training and aerobic exercise training for muscle disease. Cochrane Database of Systematic Reviews. 2019.
51. Uttley L, Carlton J, Woods HB, Brazier J. A review of quality of life themes in Duchenne muscular dystrophy for patients and carers. Health Qual Life Outcomes. 2018 Dec 19;16(1).
52. Philip PA, Carlton J, Woods HB, Mazzone P. Measuring quality of life in Duchenne muscular dystrophy: A systematic review of the content and structural validity of commonly used instruments. Health Qual Life Outcomes. 2020 Aug 3;18(1).
53. Dany A, Rapin A, Lavrard B, Saoût V, Réveillère C, Bassez G, et al. Quality of Life in Patients with Genetic Neuromuscular Disease Questionnaire: Rasch validation of the French Version (QoL-gNMD). Muscle Nerve [Internet]. 2017; Available from: http://doi.wiley.com/10.1002/mus.25598
54. Dany A, Barbe C, Rapin A, Réveillère C, Hardouin JB, Morrone I, et al. Construction of a Quality of Life Questionnaire for slowly progressive neuromuscular disease. Quality of Life Research. 2015;24(11):2615–23.
55. Powell PA, Carlton J, Rowen D, Brazier JE. Producing a preference-based quality of life measure for people with Duchenne muscular dystrophy: A mixed-methods study protocol. BMJ Open. 2019 Mar 1;9(3).
56. Powell PA, Carlton J, Rowen D, Chandler F, Guglieri M, Brazier JE. Development of a New Quality of Life Measure for Duchenne Muscular Dystrophy Using Mixed Methods: The DMD-QoL. Neurology. 2021 May 11;96(19):E2438–50.
57. Jansen M, Van Alfen N, Geurts ACH, De Groot IJM. Assisted bicycle training delays functional deterioration in boys with Duchenne muscular dystrophy: The randomized controlled trial “no use is disuse.” Neurorehabil Neural Repair. 2013 Nov;27(9):816–27.
58. Alemdaroğlu I, Karaduman A, Yilmaz ÖT, Topaloğlu H. Different types of upper extremity exercise training in Duchenne muscular dystrophy: Effects on functional performance, strength, endurance, and ambulation. Muscle Nerve. 2015 May 1;51(5):697–705.
59. Güneş Gencer GY, Yilmaz Ö. The effect of trunk training on trunk control, upper extremity, and pulmonary function in children with Duchenne muscular dystrophy: A randomized clinical trial. Clin Rehabil. 2022 Mar 1;36(3):369–78.
60. Kim A, Park M, Shin HI. Pain characteristics among individuals with Duchenne muscular dystrophy according to their clinical stage. BMC Musculoskelet Disord. 2022 Dec 1;23(1).
61. Bonarrigo K, McGuire M, Dorich JM, Bolger A, Lambert J, Horn PS, et al. Use of supported standing in males with Duchenne muscular dystrophy: Individual and family perspectives. J Pediatr Rehabil Med. 2023 Sep 19;16(3):553–69.
62. Jenkins HM, Stocki A, Kriellaars D, Pasterkamp H. Breath stacking in children with neuromuscular disorders. Pediatr Pulmonol. 2014;49(6):544–53.
63. Gandolla M, Antonietti A, Longatelli V, Pedrocchi A. The Effectiveness of Wearable Upper Limb Assistive Devices in Degenerative Neuromuscular Diseases: A Systematic Review and Meta-Analysis. Front Bioeng Biotechnol. 2020 Jan 24;7:488700.
64. Eagle M, Bourke J, Bullock R, Gibson M, Mehta J, Giddings D, et al. Managing Duchenne muscular dystrophy - The additive effect of spinal surgery and home nocturnal ventilation in improving survival. https://eprints.ncl.ac.uk. 2007 Jun;17(6):470–5.
65. Schiava M, Lofra RM, Bourke JP, Díaz-Manera J, James MK, Elseed MA, et al. Functional abilities, respiratory and cardiac function in a large cohort of adults with Duchenne muscular dystrophy treated with glucocorticoids. Eur J Neurol [Internet]. 2024 Mar 31 [cited 2024 Apr 26]; Available from: https://onlinelibrary.wiley.com/doi/10.1111/ene.16267

66. Landfeldt E, Mayhew A, Eagle M, Lindgren P, Bell CF, Guglieri M, et al. Development and psychometric analysis of the Duchenne muscular dystrophy Functional Ability Self-Assessment Tool (DMDSAT). Neuromuscular Disorders. 2015;25(12).
67. Klingels K, Mayhew AG, Mazzone ES, Duong T, Decostre V, Werlauff U, et al. Development of a patient-reported outcome measure for upper limb function in Duchenne muscular dystrophy: DMD Upper Limb PROM. Dev Med Child Neurol [Internet]. 2016;1–8. Available from: http://doi.wiley.com/10.1111/dmcn.13277
68. Dany A, Rapin A, Réveillère C, Calmus A, Tiffreau V, Morrone I, et al. Exploring quality of life in people with slowly-progressive neuromuscular disease. Disabil Rehabil. 2017;39(13):1262–70.
69. Handberg C, Werlauff U, Højberg AL. Perspectives on Everyday Life Challenges of Danish Young People With Duchenne Muscular Dystrophy (DMD) on Corticosteroids. Glob Qual Nurs Res. 2022;9.
70. Morris EHL, Estilow T, Glanzman AM, Cusack SV, Yum SW. Improving temporomandibular range of motion in people with duchenne muscular dystrophy and spinal muscular atrophy. American Journal of Occupational Therapy. 2020 Apr 1;74(2).
71. Weichbrodt J, Eriksson BM, Kroksmark AK. Evaluation of hand orthoses in Duchenne muscular dystrophy. Disabil Rehabil. 2018 Nov 6;40(23):2824–32.
72. Houwen-Van Opstal SLS, Van Den Elzen YMEM, Jansen M, Willemsen MAAP, Cup EHC, De Groot IJM. Facilitators and Barriers to Wearing Hand Orthoses by Adults with Duchenne Muscular Dystrophy: A Mixed Methods Study Design. J Neuromuscul Dis. 2020;7(4):467–75.
73. Cruz A, Callaway L, Randall M, Ryan M. Mobile arm supports in Duchenne muscular dystrophy: a pilot study of user experience and outcomes. Disabil Rehabil Assist Technol. 2021;16(8):880–9.
74. Marini Bettolo C, Guglieri M, van Ruiten H, Straub V, Bushby K, Lochmüller H. Cautionary tale in Duchenne muscular dystrophy – Opioids in neuromuscular disorders. Neuromuscular Disorders [Internet]. 2015 Oct 1 [cited 2024 Jul 5];25:S200. Available from: http://www.nmd-journal.com/article/S0960896615002412/fulltext
75. Anziska Y, Inan S. Exercise in neuromuscular disease. Semin Neurol. 2014;34(5).
76. Hammer S, Toussaint M, Vollsæter M, Tvedt MN, Røksund OD, Reychler G, et al. EXERCISE TRAINING IN DUCHENNE MUSCULAR DYSTROPHY: A SYSTEMATIC REVIEW AND META-ANALYSIS. J Rehabil Med. 2022;54.
77. Markert CD, Case LE, Carter GT, Furlong PA, Grange RW. Exercise and Duchenne muscular dystrophy: where we have been and where we need to go. Muscle Nerve [Internet]. 2012 May [cited 2023 Jun 12];45(5):746–51. Available from: https://pubmed.ncbi.nlm.nih.gov/22499105/
78. Alemdaroğlu I, Karaduman A, Yilmaz ÖT, Topaloğlu H. Different types of upper extremity exercise training in Duchenne muscular dystrophy: effects on functional performance, strength, endurance, and ambulation. Muscle Nerve [Internet]. 2015 May [cited 2017 Jan 27];51(5):697–705. Available from: http://doi.wiley.com/10.1002/mus.24451
79. Scott OM, Hyde SA, Goddard C, Jones R, Dubowitz V. Effect of exercise in Duchenne muscular dystrophy. A controlled six-month feasibility study of the effects of two different regimes of exercises in children with Duchenne muscular dystrophy. Physiotherapy. 1981;67(6):174–6.
80. Bendixen RM, Lott DJ, Senesac C, Mathur S, Vandenborne K. Participation in daily life activities and its relationship to strength and functional measures in boys with Duchenne muscular dystrophy. Disabil Rehabil. 2014 Oct 1;36(22):1918–23.
81. Aune SD, Schaufel MA, Andersen TM, Hammer S, Magnussen LH. Experiences of individually tailored physical exercise in boys with Duchenne muscular dystrophy: a qualitative study. Physiother Theory Pract [Internet]. 2024 Feb 29 [cited 2024 Apr 26];1–10. Available from: http://www.ncbi.nlm.nih.gov/pubmed/38420935
82. D'Angelo MG, Berti M, Piccinini L, Romei M, Guglieri M, Bonato S, et al. Gait pattern in Duchenne muscular dystrophy. Gait Posture. 2009 Jan 1;29(1):36–41.
83. Barrett R, Hyde SA, Scott OM, Dubowitz V. Changes in center of gravity in boys with Duchenne muscular dystrophy. Muscle Nerve [Internet]. 1988 [cited 2024 Jul 3];11(11):1157–63. Available from: https://pubmed.ncbi.nlm.nih.gov/3226433/
84. Rose KJ, Burns J, Wheeler DM, North KN. Interventions for increasing ankle range of motion in patients with neuromuscular disease. Cochrane Database Syst Rev [Internet]. 2010 Feb 17 [cited 2024 Jul 3];(2). Available from: https://pubmed.ncbi.nlm.nih.gov/20166090/
85. Young R, Nix S, Wholohan A, Bradhurst R, Reed L. Interventions for increasing ankle joint dorsiflexion: a systematic review and meta-analysis. J Foot Ankle Res [Internet]. 2013 Nov 14 [cited 2024 Jul 3];6(1):46. Available from: /pmc/articles/PMC4176290/
86. Harvey LA, Katalinic OM, Herbert RD, Moseley AM, Lannin NA, Schurr K. Stretch for the treatment and prevention of contractures. Cochrane Database of Systematic Reviews [Internet]. 2017 Jan 9 [cited 2024 Jul 3];2017(1). Available from: https://www.cochranelibrary.com/cdsr/doi/10.1002/14651858.CD007455.pub3/full
87. Craig J, Hilderman C, Wilson G, Misovic R. Effectiveness of stretch interventions for children with neuromuscular disabilities: Evidence-based recommendations. Pediatric Physical Therapy. 2016;28(3):262–75.
88. Ayala F, De Baranda AndúJar PS. Effect of 3 different active stretch durations on hip flexion range of motion. J Strength Cond Res [Internet]. 2010 Feb [cited 2024 Jul 3];24(2):430–6. Available from: https://pubmed.ncbi.nlm.nih.gov/20072058/

89. Moat D, McCallum M, Muni-Lofra R, Wong K, Michell-Sodhi J, James M, et al. FP.25 Contracture management in ambulant boys with Duchenne muscular dystrophy (DMD). Neuromuscular Disorders. 2022 Oct;32:S97–8.
90. Kern V, Wicklund M, Haulman A, McDermott MP, Martens WB, Griggs RC, et al. Ankle bracing practices in ambulatory, corticosteroid-naive boys with Duchenne muscular dystrophy. Muscle Nerve. 2020 Jan 1;61(1):52–7.
91. Carroll K, de Valle K, Kornberg A, Ryan M, Kennedy R. Evaluation of Serial Casting for Boys with Duchenne Muscular Dystrophy: A Case Report. Phys Occup Ther Pediatr. 2018 Jan 1;38(1):88–96.
92. Glanzman AM, Flickinger JM, Dholakia KH, Bönnemann CG, Finkel RS. Serial Casting for the Management of Ankle Contracture in Duchenne Muscular Dystrophy. Pediatr Phys Ther [Internet]. 2011 Sep [cited 2024 Jul 3];23(3):275. Available from: /pmc/articles/PMC5210185/
93. Townsend EL, Tamhane H, Gross KD. Effects of AFO use on walking in boys with duchenne muscular dystrophy: A pilot study. Pediatric Physical Therapy. 2015 Mar 1;27(1):24–9.
94. Bakker JPJ, de Groot IJM, Beckerman H, de Jong BA, Lankhorst GJ. The effects of knee-ankle-foot orthoses in the treatment of Duchenne muscular dystrophy: review of the literature. Clin Rehabil [Internet]. 2000 [cited 2024 Jul 3];14(4):343–59. Available from: https://www.ncbi.nlm.nih.gov/books/NBK68027/
95. Zebracki K, Drotar D. Pain and activity limitations in children with Duchenne or Becker muscular dystrophy. Dev Med Child Neurol. 2008 Jul;50(7):546–52.
96. Kim A, Park M, Shin HI. Pain characteristics among individuals with Duchenne muscular dystrophy according to their clinical stage. BMC Musculoskelet Disord [Internet]. 2022 Dec 1 [cited 2024 Jul 5];23(1):1–8. Available from: https://bmcmusculoskeletdisord.biomedcentral.com/articles/10.1186/s12891-022-05504-5
97. 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. Eur J Paediatr Neurol. 2016 Jul 1;20(4):630–8.
98. Carroll K, Yiu EM, Ryan MM, Kennedy RA, de Valle K. The effects of calf massage in boys with Duchenne muscular dystrophy: a prospective interventional study. Disabil Rehabil. 2021;43(26):3803–9.
99. Mburu B, Severn Hospice DW. 155 The role of palliative medicine in end stage duchenne muscular dystrophy. BMJ Support Palliat Care [Internet]. 2019 Mar 1 [cited 2024 Jul 5];9(Suppl 1):A65–A65. Available from: https://spcare.bmj.com/content/9/Suppl_1/A65.1
100. Apkon SD, Alman B, Birnkrant DJ, Fitch R, Lark R, MacKenzie W, et al. Orthopedic and surgical management of the patient with Duchenne muscular dystrophy. Pediatrics. 2018 Oct 1;142:S82–9.
101. Matthews E, Brassington R, Kuntzer T, Jichi F, Manzur AY. Corticosteroids for the treatment of Duchenne muscular dystrophy. Cochrane Database of Systematic Reviews. 2016 May 5;2016(5).
102. Mauro A Lo, Aliverti A. Physiology of respiratory disturbances in muscular dystrophies. Breathe [Internet]. 2016 [cited 2024 Jul 3];12(4):318. Available from: /pmc/articles/PMC5297947/
103. Garg S. Management of scoliosis in patients with Duchenne muscular dystrophy and spinal muscular atrophy: A literature review. J Pediatr Rehabil Med. 2016 Feb 27;9(1):23–9.
104. McCormick A, Wong B, McGuire M. P.7.12 Recovery of ambulation and functional mobility in boys with Duchenne Muscular Dystrophy following femoral fractures. Neuromuscular Disorders. 2013 Oct;23(9–10):777.
105. Broomfield J, Hill M, Chandler F, Crowther MJ, Godfrey J, Guglieri M, et al. Developing a Natural History Model for Duchenne Muscular Dystrophy. Pharmacoecon Open [Internet]. 2024 Jan 1 [cited 2024 Jul 4];8(1):79–89. Available from: https://link.springer.com/article/10.1007/s41669-023-00450-x
106. Longford E, Smith L, Curtis A, Guerrero J. P-42 Transitioning to adult care – a new project for young adults with duchenne muscular dystrophy. 2022 Nov;A26.2-A26.
107. Lupu M, Ioghen M, Perjoc R Ștefan, Scarlat AM, Vladâcenco OA, Roza E, et al. The Importance of Implementing a Transition Strategy for Patients with Muscular Dystrophy: From Child to Adult—Insights from a Tertiary Centre for Rare Neurological Diseases. Children. 2023 Jun 1;10(6).
108. Frank AO, De Souza LH. Clinical features of children and adults with a muscular dystrophy using powered indoor/outdoor wheelchairs: disease features, comorbidities and complications of disability*. Disabil Rehabil. 2018 Apr 24;40(9):1007–13.
109. Pellegrini N. Seated postural in wheelchair in NMD. Ann Phys Rehabil Med. 2016 Sep;59:e82.
110. Young H, Bray P, McKinnon K, Burns J, Bundy A. Everyday Life Participation Using Powered Wheelchair Standing Devices by Boys With DMD. OTJR (Thorofare N J). 2021 Jul 1;41(3):175–84.
111. Schofield C, Evans K, Young H, Paguinto SG, Carroll K, Townsend E, et al. The development of a consensus statement for the prescription of powered wheelchair standing devices in Duchenne muscular dystrophy. Disabil Rehabil. 2022;44(10):1889–97.
112. Banihani R, Smile S, Yoon G, Dupuis A, Mosleh M, Snider A, et al. Cognitive and Neurobehavioral Profile in Boys With Duchenne Muscular Dystrophy. J Child Neurol [Internet]. 2015 Oct 22 [cited 2024 Jul 3];30(11):1472–82. Available from: https://pubmed.ncbi.nlm.nih.gov/25660133/
113. Duchenne Muscular Dystrophy in Females: A Rare Genetic Disorder and Occupat...: EBSCOhost [Internet]. [cited 2024 Apr 26]. Available from: https://web-p-ebscohost-com.libproxy.ncl.ac.uk/ehost/detail/detail?vid=24&sid=2c035cdb-df41-458a-9691-7f3412eb06c5%40redis&bdata=JnNpdGU9ZWhvc3QtbGl2ZQ%3d%3d#AN=27639093&db=cin20