Duchenne muscular dystrophy: changing needs

“As a mother of a son with Duchenne muscular dystrophy, I can’t over-emphasise the importance of correct information. Once I had a good idea of Vivek’s condition and how it would affect him, I was able to do some soul searching and find other ways of doing things. It helped me to think and plan ahead for Vivek’s changing needs.

“We can’t control the future, but Vivek and I have been able to control his environment. That’s been empowering for both of us.”

Manjula Gohil, whose now adult son, Vivek, was diagnosed with Duchenne muscular dystrophy as a child

This factsheet is designed to help you understand the way Duchenne muscular dystrophy progresses and the expected timing of its complications. It is important to note, however, that all age-related timings are approximate and can vary from person to person.

With the introduction of standards of care in Duchenne muscular dystrophy, the natural course of the condition has significantly changed. There are standards of care for corticosteroid treatment, respiratory support and heart drugs, orthopaedic and physiotherapy management. Mobility and quality of life have improved considerably, and children with Duchenne muscular dystrophy now live into adulthood.

Duchenne muscular dystrophy is a severe and progressive muscle-wasting condition. Treatments and proactive interventions are available to help delay the complications of the condition, even though there is currently no cure. There are also several clinical trials currently in progress for potential new drug treatments for the condition.

Ages 7-11
During the junior school years, the child with Duchenne muscular dystrophy will usually experience deterioration of muscle strength and function, especially in the legs. Walking and climbing stairs become more difficult with time: the child tires more easily and tends to fall frequently. They need help to get up from the floor and will likely need a wheelchair for long distances.

Over this period, they might lose the ability to walk independently but the timing of loss of ambulation varies from child to child, and might depend on whether they have been on steroids and the response they have had to them. Walking or standing ability may be prolonged through the treatment with corticosteroids. Physiotherapy and orthotics (splints) are also used to help delay contractures and maintain mobility.
Main priority areas

- Regular stretching to preserve joint mobility and muscle tone.
- Medical/orthopaedic surveillance of contractures and use of orthoses (ankle splints, callipers, etc.).
- Support at school in lessons requiring physical activity.
- Attention to behavioural problems.
- Early consideration of needs for secondary education. These may include school adaptations, adapted toilet, and hoisting.
- Maintenance of independent mobility through provision of a wheelchair, with appropriate seating, to maintain good posture.
- Housing needs addressed so adaptations completed before stair-climbing becomes impossible, to ensure independent mobility in the home.

Ages 12-14
This can be a particularly difficult time for a child with Duchenne muscular dystrophy, as further deterioration of muscle strength can have an impact on mobility and independence. Home and school should have been fully adapted by now to allow maximum independence.

Walking may still be possible, although a wheelchair is usually required at this stage. An electric wheelchair is preferable, as it will enhance independence.

At this stage, the arms may be noticeably weaker. It may be tiring to write for a long time, so support at school is important.

Significant heart and breathing problems are unexpected at this age, although careful medical surveillance is important to ensure early changes are promptly treated. The number of hospital appointments might increase in this period.

Some children, especially those who have not been treated with steroids, may develop curvature of the spine (scoliosis). It is important that this is checked regularly, particularly if they use a wheelchair full-time.

Steroids have many positive effects, however side-effects – such as delayed puberty and growth, both of which can cause frustration – need to be addressed. As muscle function often deteriorates during adolescence, it’s extremely important to ensure the young person’s psychological wellbeing. Sports or social activities with other young people in similar situations can often be helpful.

Main priority areas

- Full access to school in mainstream or special education as chosen by the family. This includes full access to toilets, with appropriate hoisting arrangements as necessary.
- Medical surveillance, especially for spine, heart and lungs.
- Continued regular stretching to control joint contractures and to keep mobility.
Ages 14-16
At this stage, most of the adaptations at home should already be in place to enable maximum independence. Using an electric wheelchair will provide mobility both indoors and outdoors. IT and classroom support can be helpful and can compensate for weakness in the arms.

Heart function and breathing surveillance is essential and, if necessary, start treatment promptly.

The major medical complication at this stage is the development of spinal curvature, for which regular follow-up is needed. It is important to adapt the wheelchair to ensure good posture. Spinal surgery might be discussed, however it is only occasionally required in young people in this age group, who may have been treated with steroids since childhood.

There will generally be an increasing number of hospital appointments in this period.

Main priority areas
- Attention to post-school education or employment.
- Medical surveillance for heart and breathing function, as well as spinal curvature.
- Adequate support in the wheelchair to ensure good posture.
- Respite care.

Ages 16+
The availability of surgical management for scoliosis and the medical management of cardiac and respiratory failure have allowed some young people with Duchenne muscular dystrophy to live into adulthood. As the young person approaches their late teens, they are susceptible to chest infections and require closer medical surveillance. Treatment with corticosteroids has led to more young people retaining adequate respiratory function and some upper limb function into their late teens, as well as reducing the need for spinal surgery. Respiratory, heart and orthopaedic surveillance remain essential.

Encouraging independence is vital at this stage. Some young people may wish to attend college or university, and choose to live away from home. Care support, for example reviewing a care package, looking at employing more carers (who are not family members) or personal assistants (PAs) to enable this, need to be put in place to fully support their needs.
Growing into adulthood, the young person will have significant weakness in their muscles. It is therefore important that in these teenage years they develop skills and hobbies that can be fulfilling as an adult. Technological advances ensure that accessing the internet and computer use are possible, even for those with very weak muscles.

The Joseph Patrick Trust (JPT), the welfare fund within Muscular Dystrophy UK, provides grants towards the costs of specialist equipment and assistive technology, for children and adults with muscle-wasting conditions.

Most young people with Duchenne muscular dystrophy will, in their 20s and 30s (and sometimes younger), use a breathing machine (ventilator) at night because of weakness in the breathing muscles. This is a very effective treatment that improves quality of life and survival.

Cough assist machines are recommended in the NICE (National Institute for Health and Care Excellence) Accredited Guidelines for Duchenne muscular dystrophy and in NHS England’s neuromuscular service specifications. This specialist equipment can help keep respiratory function as strong as possible and prevent potentially fatal respiratory problems.

Main priority areas

- Independence and provision of care support.
- Attention to tertiary education or employment.
- Constant medical surveillance.
- Respite care.

Young people with Duchenne muscular dystrophy may be interested in joining Trailblazers, the network of young disabled people aged 16 to 35. This network, which is part of Muscular Dystrophy UK, offers young disabled people the opportunity to campaign for change on issues that affect them, make friends, and gain skills and work experience.

If you wish to learn more about the latest research and standards of care, contact our research team at: 020 7803 4813 or research@musculardystrophyuk.org

Other related publications

- Duchenne muscular dystrophy
- Heart check
- Surgical correction of scoliosis
- Making breathing easier
- Education
- Steroids
- Guide to transition (to adult health services)
- Trailblazers: www.musculardystrophyuk.org/trailblazers
- Joseph Patrick Trust: www.musculardystrophyuk.org/jpt
We’re here for you at the point of diagnosis and at every stage thereafter, and can:

- give you accurate and up-to-date information about your or your child’s muscle-wasting condition, and let you know of progress in research
- give you tips and advice about day-to-day life, written by people who know exactly what it’s like to live with a muscle-wasting condition
- put you in touch with other families living with the same muscle-wasting condition, who can tell you about their experiences
- tell you about – and help you get – the services, equipment and support you’re entitled to.

If you would like your GP or other health professional to have more information about Duchenne muscular dystrophy, we have some other relevant materials. We’ve developed an online training module for GPs, as well as one for physiotherapists working with adults with muscle-wasting conditions. Contact our helpline or email us to find out more.

Disclaimer
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Here for you
The friendly staff in the care and support team at the Muscular Dystrophy UK’s London office are available on 0800 652 6352 or info@musculardystrophyuk.org from 8.30am to 6pm Monday to Friday to offer free information and emotional support.

If they can’t help you, they are more than happy to signpost you to specialist services close to you, or to other people who can help.

www.musculardystrophyuk.org