



Guidelines for exercise and orthoses in children with neuromuscular disorders

These guidelines were drawn following a workshop held in Newcastle 2002. Several experts from different disciplines including physiotherapists, scientists, a rehabilitation doctor and family care officers (now known as MDC Care Advisors) from the Muscular Dystrophy Campaign met for a day to review the existing research and to share their experience. A report was published in *Neuromuscular Disorders* and these guidelines have been produced as a summary of the findings.

The research evidence was found to be limited particularly in the prescription of exercise regimes, physiotherapy for adults and rarer neuromuscular conditions. Where there was evidence this was considered alongside the collective experience of the workshop participants.

These guidelines are not a fully comprehensive guide for each individual. Not all children are the same and there may be individual circumstances where the guidelines are not appropriate so it is important that a qualified physiotherapist examines your child, shows you how to do the exercises and regularly reviews the exercise regime.

Duchenne muscular dystrophy

There has been concern particularly in Duchenne muscular dystrophy that over activity may cause more harm than good. Some of the widely held views on this are based on evidence from studies with the animal model of Duchenne muscular dystrophy. Care must be taken in extrapolating from exercise studies in mice to predicting effects in people with Duchenne muscular dystrophy because the disease in mice is different to that in boys. However, it is generally thought that exercising with weights or over activity is not to be encouraged. There are also concerns that not doing enough may cause the muscle to become weaker simply because of disuse. It is known for example that long periods of bed rest or immobilisation following a fracture weakens the muscle. Clearly it is difficult in a child to say how much is too much but normal activity that a child does voluntarily without becoming too tired is thought to be positive rather than negative.



Children who are able to walk

A daily regime of exercises is to be encouraged. These should as far as possible fit in with the all the other things a family does. Often after a bath or at bed time is a quieter and more realistic time rather than during the hectic morning routine. However it does not really matter when they are done. Getting a routine going is the most important thing so that doing the exercises should become as familiar as brushing teeth. The exercises should never be painful but stretching exercises will cause a feeling in the muscle that the child will need to become accustomed to. Some children will be able to do self-stretching exercises alongside exercises that you do for your child and your physiotherapist will be able to show you how.





Recommendations for ambulant children

1. Daily stretches to the achilles tendons, hip flexors and iliotibial band (see pictures).
2. Encourage voluntary active exercise such as swimming or hydrotherapy and cycling (may be motor assisted or a trike).
3. Symmetry to be promoted in posture, exercises and activities.
4. Eccentric activities such as running downhill and excessive walking downstairs to be avoided.



Non-ambulant children with Duchenne muscular dystrophy

Children who use a wheelchair all or most of the time should also do stretches for the arms as well as the legs. This is because sitting in a wheelchair or in one position most of the time encourages contractures to develop. For older children who may have already developed contractures, passive movements or active assisted exercises may be preferred instead of or as well as stretches. Your physiotherapist will be able to guide you and show you how to do these exercises. The main purpose of these exercises is to maintain and promote symmetry and comfort. These may be land based programmes or in water if preferred.



ELBOW STRETCH



Spinal muscular atrophy type II and III, (SMA), congenital muscular dystrophy, myopathy and fascioscapulohumeral muscular dystrophy (FSH) of childhood

These are relatively rare conditions so few therapists have significant experience other than those in specialist centers. The role of exercise, stretching and orthoses is not well established in the literature for the congenital muscular dystrophies, FSH or spinal muscular atrophy. Hydrotherapy is the treatment of choice by many therapists but the only available evidence is uncontrolled and observational. Therapists therefore must make individual recommendations for treatment protocols based on regular assessment and review.

Recommendations for exercise in FSH, congenital muscular dystrophy and spinal muscular atrophy

A precise diagnosis should be obtained where possible so that the predicted complications or features of the disease can be assessed and addressed. However there can be great variability in the sort of problems that these children have and so have very different physiotherapy requirements. Until there is research evidence for treatment protocols the following recommendations are made:

1. The physiotherapist should conduct a full assessment prior to prescribing a treatment regime. This should include measurement of respiratory function, power, range of movement and function.
2. Muscle groups that are thought to be prone to contractures should be targeted for stretching exercises and if there is antigravity power in the opposing muscle group, low resistance exercises may be considered. Some specific conditions such as congenital muscular dystrophy are particularly prone to contractures and therefore may require night-time orthoses as well as stretching exercises.
3. Activity programmes to include advice on swimming or hydrotherapy, cycling and riding where appropriate.
4. A qualified paediatric physiotherapist with knowledge of the development of children should be involved in the supervision and prescription of exercises.

Guidelines for the use of orthoses in neuromuscular disease

Orthoses are devices often called 'splints' that are made by an orthotist and they are usually used in people with neuromuscular disorders to maintain or improve range of movement, to prevent deformity or to improve function. Different devices have different names and they are often abbreviated. The most common ones used in neuromuscular disease are described below:

- AFO – ankle-foot-orthoses, can be used during sleep or during the day
- KAFO – knee-ankle-foot-orthosis, usually used to help walking or to stand. May also be called a caliper.



If at all possible you should see an Orthotist alongside your physiotherapist as team work usually gives the best results. It is worth looking for an orthotist with experience of neuromuscular disorders as they will understand the specific problems your child has.

There is almost no research evidence for the use of orthoses in congenital myopathies or the rarer neuromuscular disorders. This is an area that needs to be researched since contractures are a primary problem in some neuromuscular disorders.

Recommendations on orthoses

- ▶ Night-time AFOs in addition to stretching daily are recommended for ambulant children with Duchenne muscular dystrophy to maintain the length of the gastrocnemius-soleus complex. (also generally referred to as the tendo achilles). There is no research evidence to help us decide when to supply night splints but it is recommended that this be when there is loss of dorsiflexion. Your physiotherapist or doctor will be able to tell you when this has happened.
- ▶ Daytime AFOs are not recommended for ambulant children with Duchenne muscular dystrophy as these compromise their ability to walk by preventing the characteristic equinus gait. Children with Duchenne muscular dystrophy need to walk on their toes once the quadriceps muscle is weak. They should not be prevented from doing this by the use of special shoes or AFOs but extra care should be taken to prevent a contracture (fixed loss of range) by night splints and stretches. In ambulant children with other neuromuscular disorders careful assessment is essential to ensure that walking is not compromised.
- ▶ Clinical experience suggests that daytime AFOs should be supplied once ambulation is lost to prevent painful contractures and foot deformity. If tenotomies are performed in the non-ambulant child AFOs should be worn during the day.
- ▶ KAFOs can be used to prolong ambulation for approximately two years in Duchenne muscular dystrophy. They can also help delay the onset of lower limb contractures and there is weaker evidence to suggest that prolonging ambulation beyond 13 years may delay the onset of scoliosis. They should be supplied at the time of loss of ambulation by an orthotist with experience in neuromuscular disorders.
- ▶ In spinal muscular dystrophy II, KAFOs may be used to promote ambulation and there is limited evidence to suggest delayed onset scoliosis and contractures.
- ▶ The benefit of regular standing in the non-ambulant child to control contractures is logical but not evidenced. Standing frames or swivel walkers may be used in children with neuromuscular disorders.



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.

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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@muscular dystrophyuk.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.muscular dystrophyuk.org