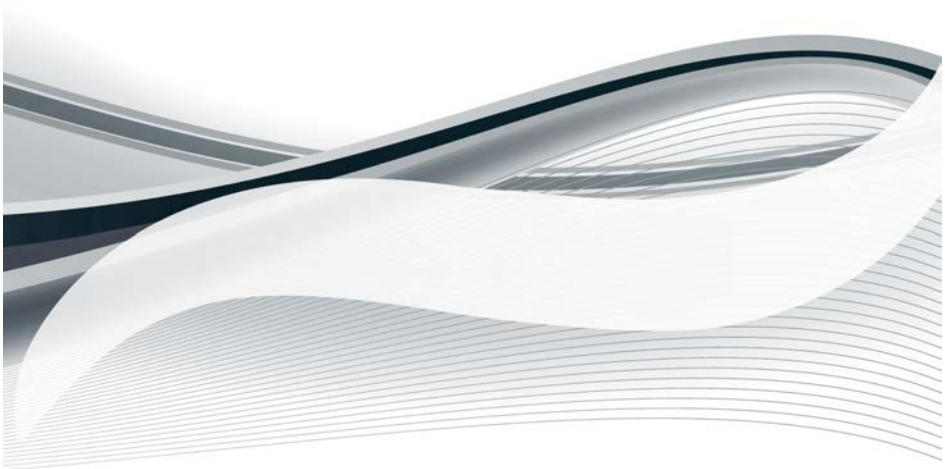


Vertebral Fractures in Children with Duchenne Muscular Dystrophy (DMD)



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Introduction

We tend to think of Duchenne Muscular Dystrophy (DMD) as a muscle wasting condition. While this is one symptom of the condition, there are also other symptoms that affect different parts of the body.

For example, we monitor the heart and lungs regularly. In the clinic, we also check for health issues like nutrition, stomach, and bowel or bladder problems. In the same way, we also need to monitor bones and check that bone health is also being cared for in DMD.

Boys with DMD, generally have slightly weak bones even when they are very young. As the child becomes older and is able to walk less, their bones become weaker. You may hear the term “osteoporosis” being used. Osteoporosis is a condition that causes the bones to become weaker to the extent that they are more easily broken (fractured).

In DMD, there are a number of reasons why this can happen:

- Strong muscles help bones develop during childhood by the “pull” of muscles on the bone as we go through our normal daily activities. In children with weaker muscles, there is less stimulation, or stress, on the bone which would normally help bones develop. As a result of weak muscles, the long bones (for example the bones in the legs and arms) tend to be thinner. A fracture of the long bones of the thigh and leg of boys with DMD are very common after a minor fall.
- The current gold standard treatment for DMD is steroid medication (like Prednisolone or Deflazacort). Whilst we know this helps keep boys with DMD on their feet longer, a very common side-effect of steroid medication is that bones become weaker. Steroid medication, especially when used for many years, leads to low bone density, particularly in the spine in the back. Fractures of the spine bones in the back (vertebral fractures) are very common in steroid treated boys with DMD.

- Other very common side effects of long term steroid medication are that it causes poor growth and also delays puberty from starting in adolescence. Growth and puberty are very important drivers for bones to become stronger in growing children by making bones bigger and thicker. Therefore, steroids also lead to weaker bones because of their impact on growth and puberty.

What is a vertebral fracture?

The spine in the back is made up of many small bones called vertebrae. A fracture in the vertebra is less obvious compared to a fracture in the arms or legs. However, it can be quite painful and may require pain relief medicines. Children may feel very localised pain over the spine, as opposed to broader back pain, associated with muscle weakness and posture. A vertebral fracture can also cause ongoing back pain.

In some children, vertebral fractures can be associated with little or no pain and are usually first discovered on an x-ray of the spine.

On an x-ray, the affected bones in the spine will have a different shape to healthy bones. Since 2015, we have been monitoring boys with DMD with regular checks of the back bones with x-rays once a year. This is to allow us to detect the earliest signs of vertebral fracture, before any back pain, to enable bone strengthening medicines to start. Vertebral fractures are very common in steroid treated boys with DMD, so do not be too alarmed if the x-ray tests identify these fractures, as there are good medicines to help stabilise these fractures.

Some families worry when they hear that their son has vertebral fractures, as they are worried that there could be an impact on the spinal cord or nerves in the back. This is not something that will happen, and with regular monitoring with annual x-rays in the clinic, we hope to detect the earliest changes of these fractures.

How do we treat this type of fracture?

The aims of treatment are to:

- Keep the spine stable
- Restore function
- Reduce or eliminate pain

Once a vertebral fracture is diagnosed in a boy with DMD, the abnormal vertebra does not “reshape”. Reshaping of a vertebral fracture does not happen in adults or children on certain medication, as it can only occur with good growth. In boys with DMD, as growth is generally very poor on steroids, the vertebra on x-ray, does not reshape. However, with treatment using medicines and physical therapy as advised by the physiotherapists, we can achieve the three aims listed above which can improve quality of life.

Bisphosphonates

If your child has been diagnosed with a vertebral fracture, the doctors at the bone clinic will consider treatment with medicines called **Bisphosphonates**. These are medicines which help to strengthen bones by slowing down the breakdown of bones. Usually, these medicines are given as a drip infusion into the arm.

As well as improving bone health, these medicines also help with back-pain as a result of vertebral fractures. However, separate pain relief medicines are sometimes necessary. We usually give Bisphosphonates once every six months for a period of two years and then usually once a year. The bone specialist will give you more information on bisphosphonate therapy.

There are also other new forms of medicines which are being used in clinical trials. The doctor may discuss these with you, if appropriate.

Gradual return to activity

It is important your child remains as active as possible and you plan a gradual return to activities. Most children with DMD do not require bed rest and can continue to go to school as normal. We can give you individual advice on activity at the clinic.

Children who are wheelchair dependent

If your child has a wheelchair, good posture within the chair is important. If there is a “tilt in space” function on the chair, then your child should be encouraged to use this often. Tilting back regularly will help maintain the comfort of his spine.

In the car, try to avoid sudden, jerky moves around corners and roundabouts. Make sure your child is as stable as possible in the car to help keep pain to a minimum.

During moving, handling and transfers, if there is a “recline” function on the chair, moving the back support into a reclined position before the sling is inserted can help. Too much hip flexion when sliding the sling under the leg, can increase back pain. The chair can then be put back into the upright position ready for transfer.

Tracking hoists tend to be a bit smoother as they lift your child directly upwards whereas, mobile hoists will move the child forward and up and can be a little jerky. When helping your child with moving and handling, try to limit any jerky movement to limit pain.

Turning in bed can also be tricky and doing this as one move (log roll) is best. Avoid moving the shoulders before the hips as this corkscrew movement may be more painful and puts more pressure on the spine. Having good support both in the chair and bed are essential and your therapist can advise on their positioning.

Children who are ambulant (able to move about)

For children who are standing to brush teeth etc, try to avoid leaning forward too much. Avoid movements that involve bending, twisting and lifting as this can cause pain while the fracture is healing.

When sitting, we want to make sure your child is as symmetrical as possible. Your child may need a specialist seating assessment from an Occupational Therapist to make sure they are in a good position when at school, at home doing homework, using a games console or other activities.

Whilst walking is good, you may notice your child cannot walk as far, as they may feel a little sorer after a while. Lots of breaks and the opportunity to sit and rest frequently should be encouraged. This is especially important in instances where your child may have a larger curve (lordosis) at the bottom of his spine as this puts additional strain on the back.

We do recommend that your child restricts activities that involve lifting, bending, reaching or twisting. Also avoid activities that may jar the spine such as bumpy car rides. Your child will need a regular movement programme to maintain flexibility, balance and posture.

How can I protect my child's spine?

It helps if your child learns and practices good posture in standing, sitting and lying. We have a separate leaflet on postural management (🌐 www.smn.scot.nhs.uk). Using a standing frame when he is no longer able to stand for long periods of time is a good way to maintain weight through the legs and spine, keeping the back as strong as possible.

If your child has been diagnosed with vertebral fractures you may wish to avoid activities such as amusement park rides, horse-riding, diving and jumping.

Activities such as trampolining put a lot of stress on the spine and we also know that they can damage muscle.

What can school children do to keep their spines healthy?

It is important to wear seatbelts in cars and avoid sitting at the back of the bus where the ride is most bouncy. When possible, have a locker at school or keep a second set of textbooks at home to avoid carrying a heavy bag, back and forth to school. If your child needs to carry a bag, it is best to use a rucksack and carry it on both shoulders. Some rucksacks also have a chest strap and this helps distribute the weight more evenly.

When sitting at school, posture is important. Make sure chairs offer good back support. Feet must be supported and not dangling and this is also important at home too. When feet are dangling, it puts excessive strain on the lower back and the muscles have to work harder to keep the body in an upright position. When the feet are supported, the pelvis is stable and there is less strain on the muscles of the back, chest and pelvis. Your therapist can advise on appropriate equipment for the classroom.

What else can be done to protect the spine?

Exercise is important for your child's muscles and bones. Swimming, hydrotherapy and water-based activities are excellent as they provide good joint movement without stress on the bones. These types of activities are also good for the heart, lung and back muscles.

Walking, going out in the wheelchair and all types of assisted movement are also great. You can further discuss your child's movement and stretching practice with your therapist.

Any other hints?

Anyone who sits a lot at a desk chair or in a wheelchair is best to change position often. If it is possible, it helps to get out of the wheelchair and lie down for a break or stand up for periods throughout the day.

If you have any questions regarding any of the advice, please contact your child's doctor or therapist.

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