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Information for patients and families

Bone health in DMD

Duchenne UK

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Bone health in DMD

DMD Care UK has brought together experts in bone health and neuromuscular diseases to agree on the best standards of bone care for people with Duchenne muscular dystrophy (DMD) in the UK. These are the clinical recommendations which are based on the latest evidence, expert-opinions, and patient perspectives.

Full clinical recommendations can be accessed at **dmdcareuk.org/clinical-recommendations**

This leaflet is designed to help DMD parents/caregivers and patients understand the clinical recommendations for bone care.

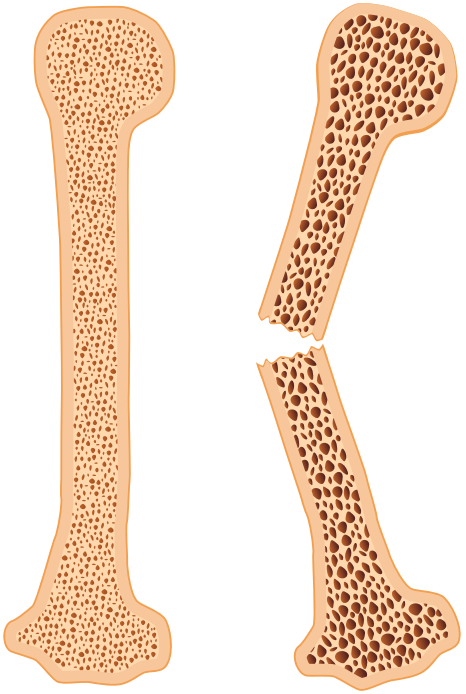
Osteoporosis

Osteoporosis is a condition where bones become weak and fragile. As a result, bones break (or **fracture**) more easily.

People with DMD are at high risk of developing osteoporosis for several reasons:

- Muscle weakness leads to reduced physical activity, which makes bones thinner and weaker.
- Long-term use of steroid medicines, while useful for the muscles, causes thinning of the bones and low bone density.
- Puberty is often delayed in DMD because of steroid use. Puberty is important for bone strength, especially increasing the thickness of bones.
- Vitamin D from good nutrition and sun exposure is important for bones. Some people with DMD may not be out in the sun as often as others.

People with DMD are diagnosed with osteoporosis if they break a long bone, such as the thigh bone or shin bone, without significant trauma (like a car accident), or if they have one or more fractures in the bones in the spine (vertebral fractures).

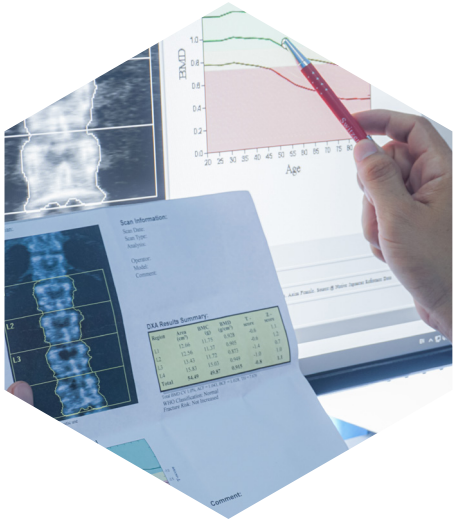


Normal bone Osteoporosis

How is bone health monitored?

Bone health in a person with DMD should be checked regularly to identify the signs of osteoporosis early. This way, doctors can recommend medicines to strengthen bones. The DMD Care UK guidance recommends the following monitoring for all people with DMD:

- Checking the spine for vertebral fractures with an x-ray (some types of DXA scanner can do this instead) once a year.
- Measuring bone density with a DXA scan once a year. This is because changes (or trends) in bone density provide additional information on the health of the bones.
- Checking vitamin D levels once a year through a blood test.



What is a DXA scan?

A DXA scan (or DEXA scan) uses low-dose X-rays to see how dense (or strong) your child's bones are. This scan gives information on the risk of fractures, and repeated measurements are helpful to understand trends.

How can you prevent osteoporosis?

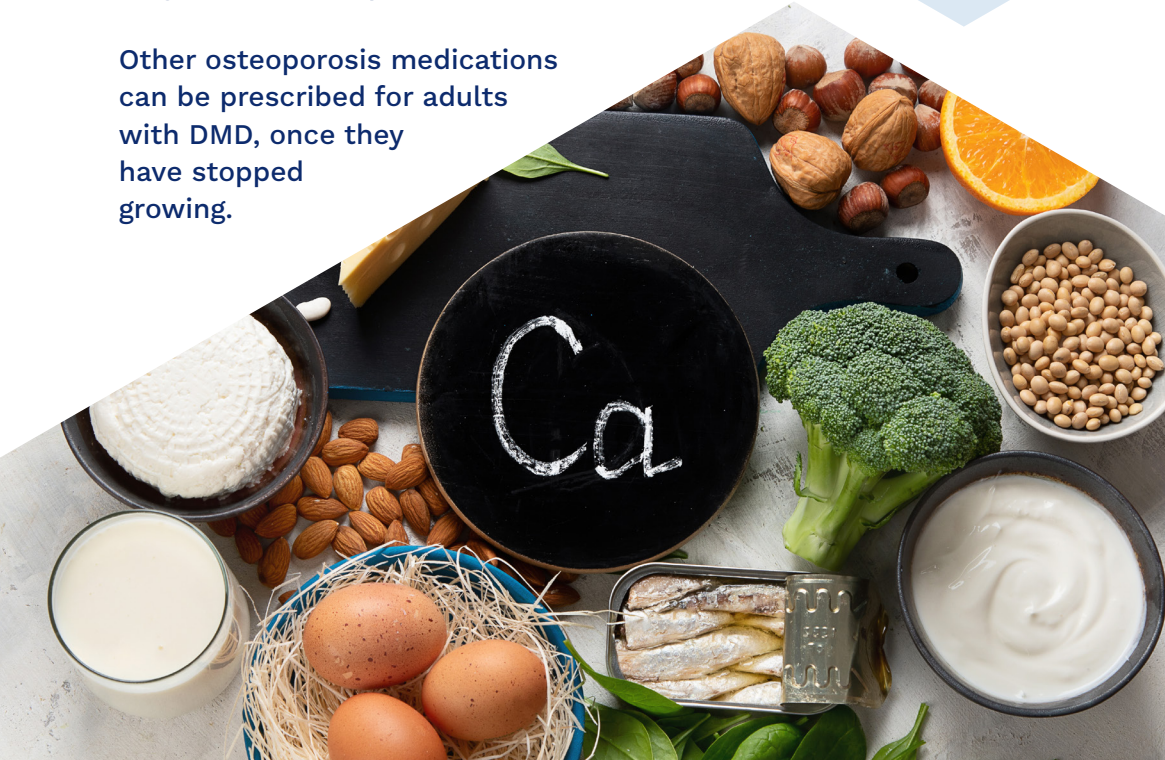
There is currently no medication that can build up bone in growing children, but the following can help keep bones healthy for longer:

- Eating a diet rich in calcium (such as dairy, fortified bread and leafy greens).
- Taking vitamin D supplements as recommended by your doctor.
- Appropriate physical activity as recommended by a physiotherapist.
- Boys should be checked for signs of puberty by age 12. If puberty is delayed, testosterone treatment can help to strengthen bones.

How can you treat osteoporosis?

Medication for treatment of osteoporosis (such as bisphosphonates) may be prescribed if there is a diagnosis of osteoporosis.

Other osteoporosis medications can be prescribed for adults with DMD, once they have stopped growing.



More about bisphosphonates

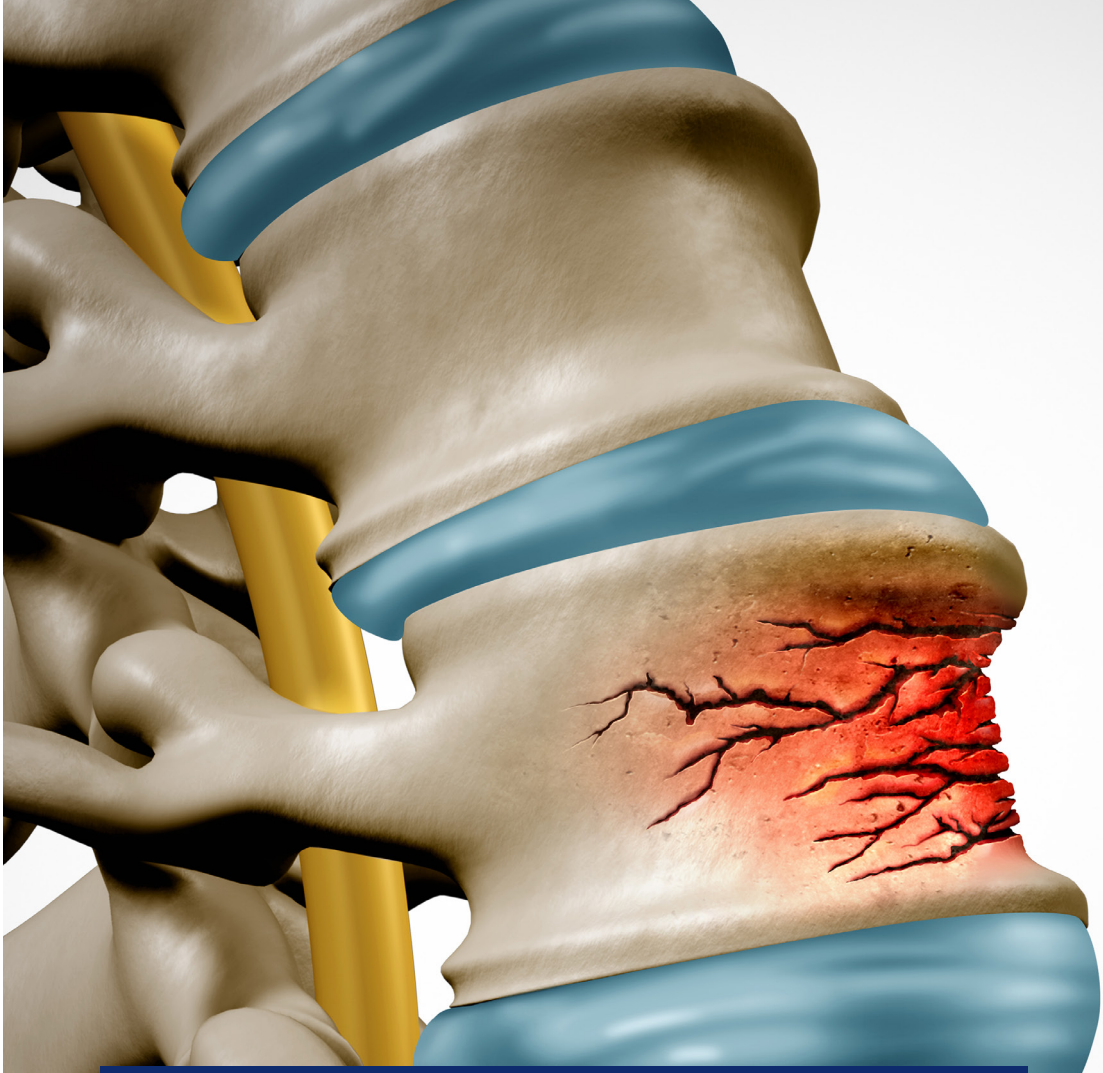
Bisphosphonates are medicines that strengthen bones and can reduce the risk of fractures. They are usually given as an infusion via a drip in the arm. The commonly used bisphosphonate infusions are Zoledronate or Pamidronate.

Initially, bisphosphonate infusions are usually given every four months (Pamidronate) or every six months (Zoledronate). After a few years, the infusions may be given once a year. Bisphosphonates are generally given regularly until the person with DMD stops growing.

After the first infusion, side-effects such as fever, flu-like symptoms and tummy upset for the first few days are common. The treating doctor will advise that extra steroid medicines (a sick day steroid stress dose) should be given with the first infusion. If your child becomes very unwell, such as with drowsiness or vomiting after bisphosphonate infusion, they will need a hydrocortisone injection and to be reviewed in the emergency department.

Please refer to the DMD Care UK Adrenal Insufficiency Leaflet (https://tinyurl.com/Steroid-Stress-Dose)

Make sure the person with DMD drinks plenty of fluids after the infusion. If urine becomes very dark and does not clear with extra fluid, take them to the hospital as they could be at risk of rhabdomyolysis, a serious condition that causes kidney damage.



What is a vertebral fracture?

A vertebral fracture is a broken bone in the spine, which is different to a fracture of your arm or leg. The spine is made up of 33 small bones called vertebrae. Most vertebral fractures are compression (crush) fractures, which means that one or more vertebrae collapses. This may be painless or very painful. If vertebral fractures are identified, this means that the bones are fragile, and bisphosphonate treatment may be recommended.

How can vertebral fractures be managed?

If your child has back pain from vertebral fractures, they should be offered pain relief. Initially, this could be paracetamol. You could also use ibuprofen if advised by your doctor. If prescribed, bisphosphonates may also help reduce bone pain.

Good posture can help with back pain. Remaining active can help to strengthen bones. However, intensive exercise may make back pain worse. A neuromuscular-specialist-physiotherapist can advise on ways to exercise and improve posture.

How should long bone fractures be treated?

A long bone fracture is a crack or break in one of the body's long bones, such as the hips, arms or legs. The orthopaedic doctor will decide on the best way of managing fractures which may require putting on a cast or surgery.

If the person with DMD is still walking and breaks a bone, surgery (rather than casting) to manage the fracture should be considered, if possible. Extra steroids should be given at the time of surgery. A specialist physiotherapy team should advise on a rehabilitation programme to help recovery.

Fat embolism syndrome (FES) is a rare complication that can happen in people with DMD following a fall or fracture, sometimes even if the fall is mild and a fracture is not obvious. FES causes fat to leak from the bone marrow and enter the bloodstream, travelling to different parts of the body like the brain, lungs and skin. Although rare, FES can be life-threatening so needs to be treated quickly.

Symptoms usually occur 1-3 days after the injury, and can include drowsiness, breathing difficulties, a purple rash and a major change in behaviour.

If you notice these symptoms, call 999 and tell the emergency department that your child has DMD and is showing signs of FES. Extra steroid medicines (a sick day steroid stress dose) should be given.

What to do in an emergency

DMD Care UK is developing tools and information to support you and inform medical professionals in an emergency. You can find the latest information and resources here:

dmdcareuk.org/emergency-support

About DMD Care UK

DMD Care UK is a nationwide initiative to ensure every person living with Duchenne muscular dystrophy (DMD) in the UK has access to the best care.

This project is funded by Duchenne UK, Joining Jack and the Duchenne Research Fund. They work closely with the John Walton Muscular Dystrophy Research Centre in Newcastle and in collaboration with the North Star Network, funded by MDUK.

DMD Care UK has produced a series of information resources for UK DMD patients, families and other non-specialists on the recommended standards of care for DMD.

Find out more at **dmdcareuk.org**

Do you have questions or feedback about this booklet? Get in touch with **support@duchenneuk.org**

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Notes

You may find it useful to jot down questions, thoughts or useful information here. We have included a few suggestions below.

You can also find a glossary of terms here:
dmdcareuk.org/glossary

Name and contact details of endocrinologist:

Details of next appointment/s:

This booklet has been developed by DMD Care UK and reviewed by clinicians and the family focus group within the project. It is based on the DMD Care UK bone and endocrine recommendations endorsed by the British Society of Paediatric Endocrinology and Diabetes.

This booklet is for informational and educational purposes only. You should always discuss your medical care with your clinical team.

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