DMD CARE UK
Best care for all

Orthopaedic Guidance

for Patients and Families with Duchenne Muscular Dystrophy (DMD)

Managment of long bone fractures and contractures

The John Walton Muscular Dystrophy Research Centre

DMD CARE UK Best care for all

Duchenne UK

dmdcareuk.org

DMD CARE UK
Best care for all

Patient information:
Orthopaedic Guidance

1. Introduction

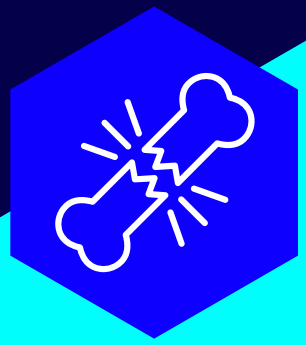
People with DMD are at higher risk of fracturing bones if they fall, stumble or are knocked. This is because bones tend to be weaker than in someone without DMD. You can read more about this in our Bone Health Guide (**https://tinyurl.com/59zdc8jx**).

Keeping mobility and independence, whether this is by walking or using a wheelchair, is very important to people with Duchenne muscular dystrophy (DMD).

Orthopaedic care in DMD focuses on treating bone fractures (broken bones) and manage contractures (restricted joint movement) so that people can maintain mobility and independence without discomfort.

Orthopaedic care works closely with physiotherapy, orthotics (for supportive devices like braces and splints) and occupational therapy teams to achieve this goal together with individuals and their families.

This guidance does not include treatment and management of scoliosis and vertebral fractures, which will be covered separately within the spinal care guidelines.



2. What if I/my child has a possible bone fracture

If someone with DMD has an injury that may have caused a fracture of a bone, it is very important that they are seen at hospital straight away.

It is likely that you will go to your nearest hospital with an accident and emergency (A&E) department by private car or in an ambulance. This may not be your normal hospital and may not have specialists in DMD.

Here are some of the important things the hospital needs to know straight away if you have a fracture:

- Long-term steroid use (prednisolone, deflazacort or vamorolone) causes people to have weaker bones, so fractures are more likely, even after a mild injury.
- If you are taking corticosteroids (prednisolone, deflazacort or vamorolone) **you are at risk of adrenal insufficiency** after an injury, including a fracture. A steroid stress dose and dosing plan will be required. Hydrocortisone should be used for stress dosing in addition to your routine oral corticosteroids. This should be detailed in your stress dosing plan. If in doubt, the extra hydrocortisone dose should be given. Please refer to further detailed guidance here: (https://www.duchenneuk.org/wp-content/uploads/2022/07/DMD-Care-Adrenal-insufficiency-leaflet-digital-updated-July-22.pdf)
- People with DMD are at increased risk of **Fat Embolism Syndrome (FES)** after an injury, even if there is no obvious fracture. The doctors should look out for symptoms which may be subtle and include:
  - new breathing problems
  - sleepiness
  - confusion
  - agitation
  - rash

- It is best not to move someone with a fracture from one hospital to another if it can be avoided. If the hospital where you are first seen has an appropriate level of medical and anaesthetic expertise, they can usually treat you there.
- However, it is very important that they **contact the following specialists** to discuss your fracture and treatment as soon as possible and no later than 24 hours after you have arrived:
    - An **orthopaedic surgeon with expertise in DMD** (or neuromuscular diseases) at a designated children's trauma centre (a list is given in Box 1)
    - Your usual **neuromuscular expert care team**
- Even if you are an adult with DMD, the hospital treating your fracture can still seek advice from the children's orthopaedic teams listed in Box 1.
- If you do need to be transferred to a specialist centre for treatment, this should be completed within 24 hours of the fracture happening. The first hospital you are moving from should get clear instructions from the specialist centre on how to take care of you before and during the transfer.

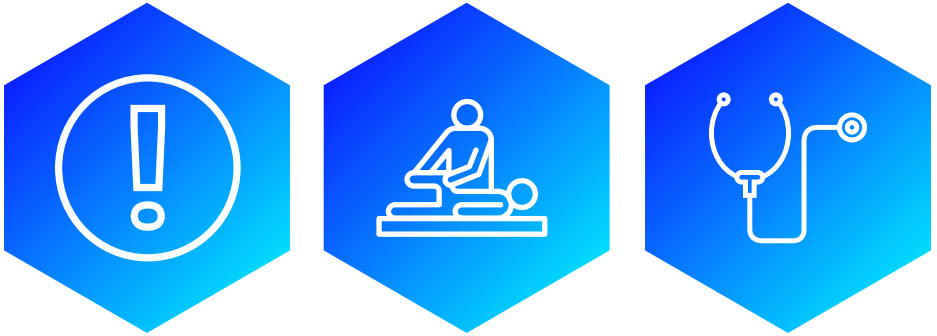
3. How the fracture might be treated

If you were still able to walk and/or use your arms before your injury, it is important that your treatment allows you to still bear weight (e.g. stand) if you have a leg fracture or lift items if it is an arm. This will help regain as much function as possible as soon as possible. Depending on the type of fracture you may need:

- A light-weight brace or cast if your broken bones have not moved out of their usual position
- An operation to realign and fix the broken bones if they are out of place

Whatever treatment you receive, your joints should never be fixed in a way that means they could not be used – e.g. your knee should not be set in a bent position and your ankle should always be set in a 90 degree position.

To make sure you have the best chance of a return to your previous mobility, it is very important that the doctors treating your fracture have spoken to your specialist DMD team, including your neuromuscular physiotherapists.



4. If you need surgery

If your fracture needs an operation to fix it, you will need a general anaesthetic. A senior anaesthetist with experience in managing DMD should be involved. They should also look at your most recent heart and breathing tests and/or repeat them if necessary.

Total Intra Venous Anaesthesia (TIVA) is the recommended anaesthetic technique.

Some anaesthetics should not be used for people with DMD:

- **Suxamethonium should NEVER be used** as there is a risk of the heart stopping (cardiac arrest)
- Volatile agents should be avoided as there is a risk of dangerous muscle breakdown (rhabdomyolysis)

If you take corticosteroids (prednisolone, deflazacort or vamorolone), Hydrocortisone will be needed intravenously (IV/directly into a vein) before an operation, to prevent adrenal crisis.

For any minor or major surgeries, your medical team should follow DMD Care UK and BSPED Adrenal Insufficiency guidance which can be found here:

DMD Care UK - https://tinyurl.com/dmdcareuk-endobone-monitoring

BSPED - https://www.bsped.org.uk/adrenal-insufficiency

- https://www.bsped.org.uk/media/ewaphps5/bsped-adrenal-insufficiency-card.pdf

All surgery comes with risks. These should be discussed with you and your family by the medical team beforehand and you should ask about any concerns or questions that you have.

5. Management of contractures

Management of contractures should be part of the routine care for all patients with DMD. This usually includes a home-stretching programme and regular physiotherapy to try to prevent contractures and keep you as flexible as possible.

You may be offered devices like ankle-foot orthoses (AFOs) to help with inflexible ankles.

You can find the family guide for physiotherapy and occupational therapy here: **https://tinyurl.com/yc2dj3by**

Planned (elective) orthopaedic surgery to treat joint contractures

In some cases, a surgery is recommended to treat joint contractures. For patients who are still walking, planned orthopaedic surgery aims to treat contractures so you can keep walking for longer. If you are using a wheelchair, it may be to make sure your seating position is comfortable and stable and that you can use your arms more easily. If you are receiving a planned orthopaedic operation, you should have this done in a specialist centre.

Before the surgery

- The reasons for surgery and the risks and benefits should be discussed between you, your family and your medical team (physiotherapist, occupational therapist, orthotist, neurologist, community paediatrician, cardiologist, respiratory specialist, anaesthetist and orthopaedic surgeon)
- You should have a full respiratory and cardiac assessment
- Your medical team should put a steroid plan in place for the time just before, during and immediately after surgery

After the surgery:

- A critical care bed should be ready for you, in case you need it
- You should receive regular specialist physiotherapy input
- Regular skin checks should take place to guard against pressure sores

6. Pain relief

Effective pain relief should be provided throughout your treatment if needed, whether you have a fracture, or you require an emergency or planned surgery.

Managing pain can be difficult and often requires a combination of pain relief drugs and physiotherapy input. Opiate drug use should be limited as they can affect breathing in people with DMD and can cause severe constipation.

Non-steroidal anti-inflammatory drugs (NSAIDs) should only be used for short periods because they can harm the stomach which, in DMD, may already have a thin lining because of steroid use. Stomach-protecting medicines should be given at the same time as NSAIDs to reduce this risk.

Key Points to Remember:

Care at Specialist Centres: Planned (elective) surgeries should be done at centres that specialise in DMD care. For serious fractures in children, it's important to consult a children's orthopaedic specialist within 24 hours of the trauma, and the DMD care team should also be notified. Adult orthopaedic care can also benefit from advice from children's specialist centres.

Steroid Medication and Surgery: People with DMD often take corticosteroids which results in adrenal suppression. This needs careful management before, during and after surgery to prevent adrenal crisis which can be life-threatening. For more information please refer to the Adrenal Insufficiency patient information leaflet here: https://www.duchenneuk.org/wp-content/uploads/2022/07/DMD-Care-Adrenal-insufficiency-leaflet-digital-updated-July-22.pdf

Heart and Lung Checks: DMD can affect the heart and lungs, the most recent heart and lung test results need to be checked before any surgery.

Fat Embolism Syndrome (FES): This is a serious condition that can happen after an injury in people with DMD, requiring immediate medical attention.

Surgical and Anaesthetic Considerations: An experienced anaesthetist should handle surgeries for people with DMD, using specific techniques that are safer for them. Some anaesthetics must not be used.

Fracture Management: Many fractures in people with DMD can be treated without surgery, but in some cases, surgery might be needed to help maintain load bearing, mobility and function. Pain relief and careful monitoring are crucial to allowing a good recovery.

Planned Surgery for Contractures: Surgery for contractures (tight muscles or joints) in people with DMD should only be done at specialised centres with the necessary expertise.

This summary highlights the importance of specialised care and careful management in treating fractures or planning surgeries for people with DMD, ensuring they receive the safest and most appropriate care possible.

Important guidelines for orthopaedic surgeons and neuromuscular teams are available. You can direct your medical team to them here:

In Case of Emergency (ICE) App

DMD Care UK has developed an app for people with DMD to help guide their medical care in an emergency or unplanned situation. The app holds data entered by the person with DMD or a parent/carer. It displays important information that is specific to you and your medical care needs that can be very important for doctors or first responders (like ambulance crews). You can find out more and download the app here: **www.duchenneuk.org/dmd-emergency-support/**

Box 1: specialist children's trauma care centres suitable for management of fractures in Duchenne muscular dystrophy patients include:

Aberdeen Royal Infirmary
Addenbrooke's Hospital, Cambridge
Alder Hey Children's Hospital, Liverpool
Birmingham Children's Hospital
Bristol Children's Hospital
University Hospitals, Plymouth
Dundee Ninewells Hospital
Evelina London Children's Hospital
Great Ormond Street Hospital
John Radcliffe Hospital, Oxford
Leeds General Infirmary
Leicester Royal Infirmary
Manchester Children's Hospital
Norfolk and Norwich Hospital
Queens Medial Centre, Nottingham
Royal Belfast Hospital for Sick Children
Royal Hospital for Children and Young People, Edinburgh
Royal Hospital for Children in Glasgow
Royal Stoke Hospital
Royal Sussex County Hospital
Royal Victoria Infirmary, Newcastle upon Tyne
Sheffield Children's Hospital
Southampton Children's Hospital
University Hospital of Wales, Cardiff

DMD CARE UK
Best care for all

Patient information:
Orthopaedic Guidance

In the event of a fracture, the following people from your neuromuscular team should be contacted ASAP:
(Add their names and contact details on this page)

1. Your neuromuscular consultant or your muscle team main contact if your consultant is unavailable

2. Your physiotherapy team

3. Specialist nurses

4. Your clinical trial team (if applicable)

In addition, it might be useful to inform the following people, for long-term care after a fracture:

1. Your endocrinologist/bone health specialist

2. Occupational therapy team

3. Your school/educational establishment

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**

THE John Walton MUSCULAR DYSTROPHY RESEARCH CENTRE

DMD CARE UK
Best care for all

Duchenne UK

Notes

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 recommendations endorsed by the British Thoracic Society.

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

THE John Walton MUSCULAR DYSTROPHY RESEARCH CENTRE

DMD CARE UK
Best care for all

Duchenne UK