



Surgical correction of spinal deformity

This information sheet is intended to:

- ▶ help you understand what scoliosis is and the reasons for spinal surgery being considered;
- ▶ help you appreciate what needs to be taken into account before a decision is made to go ahead with surgery.

Please read the following information carefully and discuss it with relatives, friends and your family doctor, if you wish. If there is anything that is not clear, or if you have any further questions, please do not hesitate to ask.

What is scoliosis?

Scoliosis means an abnormal sideways or twisting curvature of the spine and affects about 3-4 children in every 1,000. There are a variety of reasons for a person to develop scoliosis. Scoliosis develops more often in people with muscular dystrophy and some of the other neuromuscular disorders because the spinal muscles are weak and unable to support the spinal column in the upright position, either standing or sitting.

When does scoliosis develop?

Scoliosis can be already present at birth in rare conditions, for example in some children with specific congenital muscular dystrophy (CMD) variants or Spinal Muscular atrophy (SMA) type I.

Scoliosis can develop in the first decade of life, very commonly in children with SMA type II and congenital muscular dystrophy variant such as Ullrich CMD and Rigid Spine syndrome (RSMD1). Some children with SMA III can also develop scoliosis in the first decade of life.

Scoliosis develops very frequently in the early teens and adolescence in Duchenne muscular dystrophy (DMD) and in young people with SMA III in whom walking is not possible any more. Scoliosis can also develop in other conditions such as some congenital myopathies and in Charcot-Marie-Tooth disease.



How does scoliosis develop?

Children grow and change in height and weight, which adds greater demands to their already weak muscles. For some children with neuromuscular conditions, as they get older they become less mobile and spend much of the time sitting. Scoliosis develops and tends to gradually increase more rapidly in children who are unable to stand and walk. It is unclear which additional risk factors could contribute to the onset and progression of the scoliosis. It is not known whether operating the electric wheelchair with the dominant hand will influence the side the scoliosis curve develops for example. Initially most of the scoliosis is quite mobile. However, even a flexible scoliosis may over time become fixed reducing the correction which can be achieved.

How might scoliosis affect my child?

As time goes on (whether or not the muscle weakness progresses) and as the scoliosis increases, there is progressive tilting of the pelvis (pelvic obliquity). This causes increasing difficulty in sitting, as your child's weight is no longer distributed evenly between the buttocks. Scoliosis may then make sitting less comfortable for your child.

Scoliosis can often alter the shape of the chest and restrict the capacity of the lungs and so breathing can become less efficient. Scoliosis may cause your child to have to use their arms to support their body through the armrests of his/her chair.

What can be done?

The following are some management approaches that may be offered to slow down the progression of the scoliosis in your child:

- ▶ Standing and walking have a protective effect on the development of scoliosis. It will benefit children who are able to walk (supported or unsupported) to keep mobile and encourage symmetrical supported standing (for example with standing frames) in those who can stand for as long as possible.
- ▶ In some muscular dystrophies steroid medication is used to help prolong walking. There is increasing evidence that steroid medication in DMD may reduce the development of scoliosis and even the need for surgery.
- ▶ Those children who walk well but who have an increasing scoliosis (an uncommon situation) may benefit from wearing a plastic spinal support called a brace. For those who have significant difficulties in walking, a brace may make it more so. An operation may then be the best solution.
- ▶ For young children who are unable to walk, a spinal brace may be recommended to delay the progress of scoliosis until most of the growth of the spine is completed and allow optimal sitting position of your child. This decision is based upon scoliosis X-rays taken when the child is sitting. However a brace is unlikely to completely prevent such a curve from progressing.
- ▶ A brace may be the management of choice if your child develops a scoliosis late, commonly after the age of 14 years, if the curvature remains mild and does not deteriorate.



- ▶ If your child is sufficiently well grown, or if the curve is progressing quickly or the breathing or heart function deteriorates an operation to correct the scoliosis and stabilise the spine is likely to be the treatment of choice.
- ▶ Modified seating arrangements for their electric wheelchairs might be an alternative solution for those children who are overweight or where surgery is declined or not an option in view of severe respiratory or cardiac problems.

When should surgical correction be considered?

- ▶ Intermediate severity (type II) SMA and Congenital Myopathies

Most children (90%) with type II SMA develop progressive scoliosis, often associated with a pelvis which is not level (pelvic tilt). These children are never able to walk independently and scoliosis shows a continuous progression, often also after spinal growth is completed.

Children with congenital myopathies might be able to walk independently without obvious progression of their muscle weakness. However they often show progression of scoliosis and this can be accompanied by significant respiratory muscle weakness. A decision to perform spinal surgery might be taken if the progression of the scoliosis is significant.

For very young children when years of rapid spinal growth lie ahead bracing is often considered unless scoliosis is severe. In later childhood or the teenage years, surgery is usually advised if scoliosis is increasing and the child's health (and especially respiratory function) is sufficiently good.

- ▶ Duchenne muscular dystrophy and other forms of muscular dystrophy

Between 68-90% of boys with Duchenne muscular dystrophy (DMD) will develop scoliosis after having lost the ability to walk, starting between 11-16 years, typically 1-2 years after starting to use their wheelchairs continuously. Rarely boys with DMD might develop scoliosis whilst still walking.

Although scoliosis is a very common problem in DMD, it is also not an inevitable complication. The reasons why a boy with DMD will not develop severe scoliosis are still not completely understood. When the child is no longer on his feet, the scoliosis may start to progress. At this stage discussions may begin between you and the surgeons about spinal surgery to correct the curve. Regular monitoring of scoliosis is very helpful in planning the right time to consider spinal surgery. In DMD consideration of timing that reflects, cardio- respiratory sufficiency and nutrition is very important in planning spinal surgery.

When is the right time for surgery?

The right time to undertake surgery needs to be decided in consultation between the young person, the surgeon, the family and the rest of the multidisciplinary team (see section entitled 'The decision'). There are a variety of factors that should be considered and discussed (see below).



Ideally children/young people who are considered suitable for surgery will have a pre-operative assessment. This approach gives the child/young person and the family a chance to meet people who will be involved in their care when they do come into hospital for surgery.

It can be a helpful experience for children/young people and families to ask any questions they may have. This assessment may also contribute to the final decision regarding surgery. It may be helpful for families and children/young people to consult with another family whose child/young person has had scoliosis surgery.

What factors should be discussed before surgery is considered?

- ▶ How your child feels about the surgery: spinal surgery is a big decision and one that your child should feel confident and optimistic about.
- ▶ Your child's age: ideally surgery is considered when most of a child's growing is complete. In some instances however the progression of scoliosis does not allow waiting and surgery in these cases can be done earlier, in the first decade of life.
- ▶ What your child weighs: spinal surgery is very demanding on the body so having a good weight and nutrition will help in your child's recovery. Some children can lose up to 10% of body weight during surgery and recovery. Being too thin and light puts extra demands on your child's ability to recover.
- ▶ Your child's breathing: this will be discussed with you by the anaesthetist. Broad guidelines would suggest that your child's breathing should be above 30% of normal though there are differences between different conditions. Your child will have a sleep study to measure the breathing efficiency. If needed your child may be referred to a respiratory specialist to assess the breathing and the need of non-invasive breathing support at night around the time of surgery (see MDC factsheet *Making breathing easier*).
- ▶ Your child's heart: if your child's heart is affected as part of his/her condition surgery can still be considered, unless the severity of the cardiac involvement poses significant concern. It is common practice for every child having scoliosis surgery to have their heart function assessed with an echocardiogram (if you are not sure, ask your child's doctor). The surgery puts heavy demands on your child's heart and it is important to consider whether your child's heart is strong enough to cope with this.
- ▶ Your home adaptations: once spinal surgery has been carried out moving and handling your child will need to be carried out using a hoist. This is particularly the case if your child is a constant wheelchair user. Home adaptations are therefore essential for managing your child's needs after the operation.

What does the surgery involve?

There are now several orthopaedic surgeons in the UK performing spinal surgery on patients with neuromuscular disorders and they all have slightly different procedures and approaches to this, which reflect local expertise and different school of training. The following is a guide only.

There are two possible approaches:

- ▶ a posterior approach where the surgeon does the operation through an incision (cut) down the length of the spine;



- ▶ a two stage anterior and posterior approach. The first stage is an anterior approach, where the surgeon makes an incision in the chest to release the spine from the front. The second stage takes place about a week later when the surgeon makes an incision down the length of the spine to insert the rods and fuse the vertebrae (spinal bones).

Which procedure is used depends on your child's condition and is a decision made between the surgeon and the anaesthetist. They will consider safety and your child's specific situation and underlying diagnosis.

The rods used to support the spine in an improved straighter position are metal and fixed in place with screws, but can also be fixed with wires and hooks. There are various designs available. Each surgeon will have his/her individual preference.

It is therefore very helpful for each family to discuss the nature of the surgery with their surgeon and ask for detailed information regarding the exact nature and duration of surgery and the risks involved.

Following spinal surgery it is hoped that your child/young person will no longer be required to wear a spinal brace. Again this can vary and some surgeons prefer some kind of support in the form of a spinal brace for a short period following surgery. Families should ask their surgeons about this at the time of surgery.

What are the risks?

This is major surgery. Prior to this surgery, each child/young person needs to undergo a full assessment by the medical, surgical and anaesthetic team to evaluate the risks involved for them.

The risk of anaesthesia requires skilled anaesthetic advice. Certain drugs should be avoided during anaesthesia in order to minimise reactions (see MDC factsheet *Anaesthetics*).

The operation itself is technically demanding and should be carried out by an experienced spinal surgeon. The spinal bones (vertebrae) protect the spinal cord. This surgery exposes the spinal cord during the operation. Careful monitoring of the spinal cord takes place throughout the surgery to detect very early if any stress to the spinal cord is occurring. Any major operation will involve the risk of significant blood loss; this is carefully monitored and managed by the surgeon and anaesthetist. The duration of surgery is an important factor and the longer the procedures, the higher the rate of complications and risks.

Other risks include infection which often requires return to hospital to clean and washout the wound; failure of the metalwork which may require repair at a second operation; further curve progression: prolonged paralysis (bowels stop functioning) which in some patients can prolong the stay in the hospital.



What are the benefits from surgery?

Scoliosis surgery tends to improve sitting posture and trunk length. A straighter trunk will increase the child's/young person's overall height. This should be born in mind if your child/young person travels in an adapted vehicle. It will also prevent further collapse of the spine and the pain and discomfort that this can involve. It will benefit the child's posture, and eliminate the long-term need for a brace or support in his/her wheelchair. It will reduce the effects of a progressing scoliosis on respiratory function.

Are there any disadvantages?

Balancing the risks and benefits of your child/young person prior to having any major operation is essential. Stabilising the spine in a rigid manner, from just below the neck to the pelvis, does restrict the mobility of the trunk. For some children/young people this will make eating more difficult because they cannot lean forward and so their hands can no longer easily reach their mouth. This may be overcome by the use of simple aids such as mobile arm support.

For some a combination of loss of appetite and difficulties of self-feeding can cause some additional weight loss. This is mostly recovered within the first few months following surgery. Spinal surgery restricts further growth of the spine. This is not a problem if the child is close to the end of the growth, but will have an effect on a very young child. As far as possible surgery it is postponed in very young children unless the curve is severe. There are also new procedures which allow the spine to grow even after spinal surgery, although they can require repeated surgical interventions and have increased rate of complications

The decision

The decision as to whether to undertake this operation ideally is made by the parents of young children or by the young person in consultation with the family and an experienced multidisciplinary team. This team may include:

- ▶ a paediatrician;
- ▶ an anaesthetist experienced in neuromuscular disorders;
- ▶ an orthopaedic surgeon experienced in spinal surgery;
- ▶ a nurse specialist;
- ▶ a speech therapist who is experienced in assessing swallowing;
- ▶ a physiotherapist who will talk to you both about the exercises that will help your child recover from the surgery;
- ▶ an occupational therapist who will help you access any equipment your child will need when they have had the surgery and are at home;
- ▶ a dietician who will talk to you and your child about the importance of good nutrition to help cope with the surgery. He/she may also advise on extra nutrition supplements.
- ▶ a social worker who may talk to you about how you will cope at home after the surgery and liaise with local services to help you access what you need;
- ▶ friends or other individuals affected by similar conditions are often the best people to provide a firsthand advice on the pros and cons of spinal surgery. The nursing or medical team may have names of families who are able to help you in this way.



Conclusion

Scoliosis surgery remains the treatment of choice of progressive scoliosis in children/young people with neuromuscular disorders. It should not be viewed as a failure of other management regimes. The choice of performing surgery and the timing depends on the progression of scoliosis and the associated complications. An experienced multidisciplinary team being involved may help in this decision making process. Improved surgical techniques along with careful spinal cord monitoring and skilled anaesthetic cover now make it possible for children/young people with a variety of neuromuscular conditions to have surgical correction of their spinal deformity. For all children/young people with neuromuscular disorders where surgical correction of scoliosis is discussed careful consideration of individual circumstances should be taken into account.

Other relevant publications

- ▶ Anaesthetics
- ▶ Making breathing easier
- ▶ Spinal surgery – questions to ask

If you have feedback about this factsheet or would like to request any references used to produce it, please email info@musculardystrophyuk.org.

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@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

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