

# Neuromuscular scoliosis

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## What is a scoliosis?

Scoliosis is a medical condition that affects the spine. The term depicts a sideways curve of the spine. The vertebrae (back bones) tilt and rotate, and the spine commonly adopts an S or C shape.

## Why are children with neuromuscular conditions at risk of a scoliosis?

Neuromuscular scoliosis is the second most common type of spinal condition after adolescent idiopathic scoliosis (AIS). We don't know the exact cause of AIS but neuromuscular scoliosis occurs as a result of weakened muscles around the spine.

As children grow their body increases in size and weight. Usually the additional forces acting on the growing spine are balanced by symmetrical muscle activity. In neuromuscular conditions these muscles are often weak or have unbalanced action, so struggle to support the additional weight. The trunk of the child is seen to slouch since the spine cannot resist new forces against gravity. With continued spine growth (as part of the overall growing child) further unbalanced force is placed on the spine and instead of growing straight, it may curve sideways. Spinal curves that develop earlier in life, and those present in children more affected by their neurological condition, have a higher risk of progressing to a large size.

Early in the development of the scoliosis the spinal curve is flexible, so when the child is lifted their trunk (main body) straightens, but with time it becomes increasingly rigid and maintains a fixed position.

Many conditions can result in a neuromuscular scoliosis since any condition that affects muscle function/action can cause a scoliosis.

Common conditions causing a neuromuscular scoliosis are those that directly affect muscle function at an early age (such as Duchenne muscular dystrophy and spinal muscular atrophy) or indirectly affect muscle function at an early age such as following injury to the brain (cerebral palsy), spinal cord (spina bifida/trauma) or progressive nervous system damage (Friedreich's Ataxia). All

these conditions affect muscle functioning and thus risk development of a scoliosis.

## How will the scoliosis affect my child?

Children with neuromuscular conditions are affected differently. On one side of the clinical spectrum the child may be only slightly affected, but on the other end, the child can be significantly disabled needing total daily care. The likelihood of developing a scoliosis, and then risk of progression (getting worse), increases with severity of the affecting condition.

In many instances the scoliosis will not affect the child significantly and surgical intervention may not be required. At first, when the spinal curve is flexible and the child can be easily positioned, few problems are encountered. As both the child and curve increase in size it may become difficult for the child to be positioned. They may lean, and despite best seating modification, sit with a tilted pelvis. This seating position may cause pain and pressure sores. General care can become difficult. It is when the curve is seen to be progressing and/or causing difficulties or upset to the child, spinal surgery should be considered.

## Common neuromuscular conditions causing neuromuscular scoliosis

### Cerebral Palsy

This is the most common neuromuscular condition seen by orthopaedic surgeons. Cerebral palsy results from an injury to the immature brain (before the age of 2 years). It leads to muscle imbalance with mixture of muscle weakness and spasticity. The more severely a child is affected, the higher the risk for developing a scoliosis, and progression of that scoliosis. At the most severe end of the spectrum, a child with cerebral palsy who cannot sit independently may have a risk as high as 90% for developing a scoliosis, compared to a child with cerebral palsy who walks where the risk of developing a scoliosis is much less. The scoliosis often develops between 6 and 10 years of age. It is flexible initially but tends to become stiffer with time.



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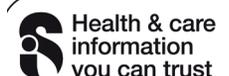
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The Information Standard Certified Member

Edition number: 1  
Issue date: Autumn 2018  
Planned date of review: Autumn 2021

## Spina Bifida

This is a diagnosis which includes a group of spinal birth defects. Before birth, some of the bone and nerve tissue that should cover the spinal cord (thoracic levels) or lower spinal nerve roots (lumbar levels) will not have fully formed (in some cases, not at all).

It can lead to varying degrees of muscle weakness or paralysis. The scoliosis may occur as a result of muscle weakness or from specific vertebra (back bone) abnormalities. This risk of scoliosis is linked to the level of paralysis and severity. A higher level of paralysis increases the risk. A thoracic level spina bifida has a 100% risk of scoliosis. Spinal cord tethering may also increase risk of scoliosis and its progression. Children with spina bifida may also get a forward bend (kyphosis) curve which may need surgery to help with their sitting.

## Duchenne's muscular dystrophy (DMD)

It is the most common of the dystrophies (a group of muscle diseases that results in increasing weakening and breakdown of skeletal muscles over time). DMD causes progressive muscle wasting and is often noticed early in child development with difficulty walking and delayed motor milestones. Children are often wheelchair dependent by age 10.

Once the child loses the muscle strength to walk and becomes wheelchair dependent, a scoliosis often occurs. Spinal surgery has been the treatment of choice when the curve enlarges and patients are unable to be comfortably managed.

Many children with Duchenne's muscular dystrophy will develop weakened hearts and lung function. This may make them unfit for major spinal surgery at an older age. Close monitoring of children with muscular dystrophies is required since we may suggest an earlier timing of surgery to offset the later increased surgical risks from weakened lung and heart function.

## Spinal Muscular Atrophy (SMA)

SMA is a condition that causes progressive muscle weakness and loss of movement due to muscle wasting (atrophy). There are several types of SMA and there is significant variability in the disease. Scoliosis is often seen early (aged 2-3 years of age).

New disease modifying treatments are being trialed for the treatment of SMA but are all in the early stages of development. Surgery in

children with SMA may be considered early.

## Friedreich's ataxia

Friedreich's ataxia is a genetic condition that affects the nervous system. Those affected have muscle weakness, among other symptoms. The condition normally becomes apparent between 7 years and 25 years of age. Symptoms are slowly progressive. On average after 10-15 years with the disease patients require use of a wheelchair and require assistance with activities.

Children who develop symptoms of Friedreich's ataxia aged less than 10 years of age and those who develop a scoliosis aged less than 15 years of age have a higher risk of spinal curve progression.

## Treatment of neuromuscular scoliosis

### Multidisciplinary team approach

Children with neuromuscular conditions are often treated by several medical practitioners, each addressing a different element of their condition. All healthcare professionals involved in your child's care will be aware of risk for scoliosis development (and progression) and so will be continually monitoring your child over time. Interval X-rays are commonly performed to record spinal curve change. The scoliosis may affect other aspects of your child's condition, and vice versa, therefore care is generally coordinated among a number of subspecialties.

Not all children with a neuromuscular condition will develop a scoliosis, and not all children who develop a scoliosis require spinal surgery. Treatment of neuromuscular scoliosis is mostly supportive.

### Physiotherapy & wheelchair services

Physiotherapy is important. Maintaining walking and standing in children will offer some protection to curve progression. Standing, and better still walking, will maintain and strengthen core trunk muscles, which supports and protects the spine. Muscle and tendon stretches are important. This helps avoid unbalanced pulling on the body and allows more comfortable sitting.

In wheelchair dependent children, particularly in those who are actively growing, there is the continued challenge of adapting their seat to accommodate a progressive curve. Continuous adjustments are needed such as chest and side supports, shoulder and waist belts and head and foot rests. Occasionally a moulded seat will be required.



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## Bracing

Bracing is largely of limited value in neuromuscular scoliosis and mostly not performed, although exceptions exist. Bracing will not prevent progression of neuromuscular scoliosis and it may contribute or cause other problems such as allowing trunk muscles to become weaker, risk related skin problems (pressure sores/sweat rashes) or increase risk of chest related problems and feeding difficulties. These problems often outweigh benefits. In children with a neuromuscular condition and a scoliosis who can walk, a brace can make it harder for them to walk and put them at risk of falls.

Surgical treatment is largely the only effective treatment in children with a severe neuromuscular scoliosis.

## Does a large curve always mean surgery?

Larger curves will generally cause more problems but there is no specific cut-off. You will see some articles quote 50 degrees. I think it is very reasonable for a spinal surgeon to meet any child with a curve around 50 degrees, but that is not to say they will suggest surgery. The overall curve size at presentation is one of many factors considered.

Some children with large spinal curves may have no significant curve related problems, are seen to be doing well and appear happy with a good quality of life. It may be hard to justify high risk spinal surgery in such situations unless it is judged they are likely to get problems soon.

There are occasions when it's appropriate to consider early surgery on smaller spinal curves such as in neuromuscular conditions which additionally affect heart and lung function. These children may become unfit for major spinal surgery at a later age.

There are also children with large spinal curves who are judged medically unfit for surgery or that surgery is unlikely to offer enough benefit to justify the surgical risks.

Deciding on surgery is a risk balance judgement. The spinal curve, the medical co-morbidities and social issues are all important to consider when making such a decision.

## What does the surgery involve?

Spinal fusion surgery is the most common surgical procedure used to treat neuromuscular scoliosis.

This involves an operation that exposes the curved spine from behind, screws are inserted into the vertebrae and releases made to allow correction of the curve. Sometimes the surgeon will need to fix into the pelvis to allow for a better spinal correction and improved seating position. Rods are inserted to align and rebalance the spine. Very occasionally, if the spinal curve is very stiff, an operation going through the side of the chest or abdomen is needed to further release the spine from the front, before fixing from behind. Your surgeon will discuss the type of surgery planned on an individual basis.

## Deciding on surgery

Advances in the medical management of children with a neuromuscular condition are resulting in improved survival, with many children living into adulthood. Treatment of severe spinal conditions has therefore become increasingly considered with the view of improving their quality of life both for now, and in later life. Deciding, and consenting for surgery requires shared decision making amongst the medical teams, patient/family and carers. A mutual understanding of the child's current and possibly future needs is required.

Surgical goals are to prevent curve progression, improve sitting balance and tolerance, and reduce child repositioning and associated pain. Thus, specific factors that may make spinal surgery beneficial are large spinal curves which are already impacting the child's health (causing difficulty in sitting or pain) and a scoliosis judged at particular risk of further progression and later causing aforementioned problems.

Nevertheless, deciding to operate on children with neuromuscular scoliosis remains difficult. There is no doubt, in the majority of children undergoing scoliosis correction surgery, it makes a significant improvement to the child's quality of life and feedback from families and carers is consistently positive. But risk exists, and it's because of those risks, surgical specialists are focused on whether surgery will benefit the child enough to justify these potential risks.

The overall risk of complications after surgery is broadly related to the severity of the neuromuscular condition and any associated medical problems. Children more severely affected by their condition and who have other medical difficulties are at greater risk of a complication compared to a child less affected.



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Nearly all of the risks listed below are applicable to all children undergoing spinal surgery and not just children with underlying neuromuscular conditions.

Complications from surgery can be broadly grouped into early and late. Early complications include, but not limited to, those related to large blood loss, spinal fluid leak, infection, breathing/lung difficulties, feeding and bowel related problems, delayed wound healing, early metal-work problems such as bone cut out or metal-work prominence, worsening of lower limb/ bladder and bowel function (if applicable) and incomplete sitting balance correction. Later complications include metal-work related problems, delayed deep wound infection, the child requiring further surgery to further improve sitting position or extend the spinal surgical instrumentation.

### Pre surgical assessment (getting ready for surgery)

Planning care in children with neuromuscular scoliosis is tailored to the individual child. A thorough multidisciplinary pre-operative assessment is made. Children may require several medical investigations such as sleep studies, lung function assessments and heart tests before listing to ensure the child is fit for surgery. Children with low body weight will often be seen by a dietician and surgery will be delayed until the child gains appropriate weight. The importance of good nutrition cannot be over emphasised before spinal surgery. Occasionally a feeding tube is inserted, on a temporary basis, to help the child get the calories needed to gain enough weight for surgery to be safely performed.

Spinal surgery in children with complex medical conditions is high risk and the above tests and procedures will help the anaesthetist and surgeon more accurately discuss the risks with parents and family before a final decision for surgery is made.

### Waiting for surgery

These operative cases are complex and require the input of large specialised medical teams. They are often performed by two consultant spinal surgeons and rely on experienced paediatric anaesthetists and intensive care teams. Some cases are delayed until the summer months in children who are deemed susceptible to respiratory tract illnesses or other seasonal related conditions. For these reasons (and others) dates are planned far in

advance. Unfortunately, despite months of planning, the surgery is at risk of late cancellation because the specialised children centres where these operations take place will face additional pressures on their intensive and high dependency beds from the wider region.

### Admission for surgery

It will have been decided in the pre-operative assessment clinics the best time for admission. Occasionally the child may be admitted a day or two in advance of the operation day to ensure they're optimised for surgery.

On the day of surgery usually only one parent is able to accompany the child into the anaesthetic room and stay until they fall asleep (a shortage of space limits the number of people). Once the child is asleep, a nurse will accompany you back to the ward. Once the child is awake you will be brought by a nurse to the recovery room to be with your child.

### Following surgery

The initial concern in patients who have undergone surgery for neuromuscular scoliosis is their breathing and circulating blood volume. Lots of monitors will be attached. Children with poor lung function may benefit from extended breathing support. This may mean the breathing tube stays in place and they are not immediately awakened after surgery. Sometimes, the breathing tube is removed but they require a tightly fitted breathing mask to help push air into their lungs (they may be too weak to take full deep breaths). They will be receiving fluids into their veins and may require a blood transfusion in the day's following surgery.

The wound is kept covered with dressings. Occasionally the surgeon may place a drain in the wound, which is removed a day or two after surgery. A concern in the days immediately after surgery is contamination of the surgical wound (which increases the risk of infection). Regular checks are needed to ensure dressings are secure.

A physiotherapist will regularly assess the child. Chest physiotherapy, airway clearance and early mobilisation (sitting out) are encouraged.

Some children are slow to start eating after surgery so a temporary feeding tube may be inserted to ensure they get adequate initial nutrition.

Length of stay after surgery varies greatly. Overall, it is probably 2 to 4 weeks in most children



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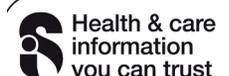
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(assuming no complications).

Your surgeon will review your child until they are confident a spinal fusion (bone forms across the operated length of the spinal curve acting in-tandem with the metalwork to hold the spinal correction) has occurred and no other problems are foreseen.

### Outcome

As stated above, there is no doubt, in most children undergoing scoliosis correction surgery, it makes a significant improvement to the child's quality of life and feedback from families and carers is consistently positive.

Due to the underlying neuromuscular conditions no functional improvements are seen following surgery but patients who had pre-operative pain are found to be in less discomfort following surgery and overall sitting balance in children following surgery is improved generally making them happier and facilitating easier care.



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