

Cerebral Palsy and the Spine

A patient guide

In brief: Cerebral palsy is a non-progressive disorder of movement and posture caused by an injury to the developing brain, usually before, during or shortly after birth. Although the brain injury itself does not change over time, its consequences for the growing body very much do, and the spine is one of the places where those consequences are seen most clearly. The disordered muscle tone, weakness and loss of trunk control that characterise cerebral palsy can, over the years of growth, allow the spine to bend sideways into a curve. This is a neuromuscular scoliosis, and it is one of the commonest and most significant orthopaedic problems associated with cerebral palsy, particularly in those who are most severely affected and least able to walk. The curve in cerebral palsy tends to be long and sweeping, frequently dragging the pelvis out of level with it, and it can interfere with the ability to sit comfortably, to be cared for, and in severe cases with breathing and nutrition. This page describes cerebral palsy entirely from the standpoint of the spine: why the curve develops, how it behaves, how it affects sitting and care, how it relates to the hips, and what the evidence shows about the large operations sometimes used to address it.

Cerebral palsy seen from the spine

Cerebral palsy is, at its root, a problem of the brain rather than the spine. An early injury to the developing brain disturbs the signals that control muscle tone, balance and posture, and the result is a lasting disorder of movement that does not itself get worse over time. The spine, however, sits at the centre of the body's posture, and it depends for its straightness on a balanced pull of the muscles of the trunk and on the ability to hold oneself upright. When that balance and control are disturbed, the spine is left vulnerable to deformity as the child grows.

The link between cerebral palsy and the spine is, therefore, an indirect one. The brain injury does not bend the spine directly. Instead, it removes the steady, symmetrical muscular support that keeps a growing spine straight, and it is this loss of control, acting over years of growth, that allows a curve to develop. This is why the spinal deformity of cerebral palsy is described as neuromuscular in origin, to distinguish it from the idiopathic scoliosis of otherwise healthy adolescents, whose cause is unknown and whose spine has normal underlying muscular control.

Seeing cerebral palsy from the standpoint of the spine means recognising that the severity of the spinal problem tracks closely with the severity of the underlying condition. The children whose movement is least affected, who walk independently, are at relatively low risk of a significant curve. Those at the other end of the spectrum, who cannot sit or stand unsupported, are at much higher risk, and it is in this group that the spine becomes a central concern.

How severity is described: the GMFCS

To understand the relationship between cerebral palsy and the spine, it helps to have a way of describing how severely a person's movement is affected. The system used most widely for this is the Gross Motor Function Classification System, usually shortened to the GMFCS. It was developed by Palisano and colleagues and sorts people with cerebral palsy into five levels according to what they can do, rather than according to the type of cerebral palsy they have.

At level one, a person walks without restriction and is limited only in more demanding physical tasks. At level two, walking is possible but with some limitation. At level three, walking requires a handheld mobility aid. At level four, self-mobility is limited and the person is largely transported or uses powered mobility. At level five, the person is transported in a manual wheelchair and has very limited ability to control their own posture against

gravity. The system is reliable and widely used, and it has become the standard language for describing severity in this field.

The reason the GMFCS matters so much to the spine is that the risk and severity of scoliosis rise steeply across these levels. The non-ambulant levels, four and five, and level five in particular, carry by far the greatest risk of a severe spinal curve. Much of what follows on this page applies most strongly to those at these levels, because it is there that neuromuscular scoliosis is most common, most severe and most likely to require consideration of surgery.

Why the spine curves: the mechanism of neuromuscular scoliosis

The development of a spinal curve in cerebral palsy follows from the loss of balanced muscular control over a growing skeleton. A healthy spine is held straight not by its bones alone but by a continuous, symmetrical tug of the muscles of the trunk and the constant small adjustments of posture that an intact nervous system makes without conscious thought. In cerebral palsy these mechanisms are disturbed. Muscle tone may be increased on one side and reduced on the other, trunk muscles may be too weak to hold the body upright, and the automatic corrections that keep the trunk centred over the pelvis are impaired.

Against this background, the spine of a growing child is left without the even support it needs to grow straight. A small asymmetry of tone or posture tends to be reinforced rather than corrected, and over the years of growth the spine settles progressively into a sideways curve. Because the deforming forces act along the whole length of the trunk rather than at one segment, the resulting curve tends to be long and sweeping, often taking the form of a single broad C that runs through much of the spine, rather than the shorter, more angular curves seen in idiopathic scoliosis.

A defining feature of this neuromuscular curve is its tendency to involve the pelvis. As the curve runs down towards its base, it frequently carries on into the pelvis itself, tilting it out of the horizontal so that one side sits higher than the other. This is called pelvic obliquity, and it is one of the most important characteristics of cerebral palsy scoliosis. An oblique pelvis throws the weight of sitting onto one buttock, undermines a level seated base, and links the spinal deformity intimately to the position of the hips below.

The shape of the curve: long C-shaped curves and pelvic obliquity

The curve of cerebral palsy has a recognisable character that sets it apart from other forms of scoliosis. Where the idiopathic scoliosis of healthy adolescents tends to produce a curve confined largely to one region of the spine, often the chest, with the body compensating above and below to keep the head and pelvis level, the neuromuscular curve of cerebral palsy is typically longer, more sweeping and less compensated.

The classic pattern is a long C-shaped curve that extends across the thoracic and lumbar regions together, frequently reaching down to the pelvis. Because the muscular control that would normally produce compensating curves is itself disturbed, the body is less able to balance the deformity, and the trunk can come to lean markedly to one side. The curve tends to be collapsing in nature, in the sense that it folds the trunk down towards the pelvis rather than holding a balanced posture.

The involvement of the pelvis is central to this picture. Pelvic obliquity, the tilting of the pelvis out of the horizontal as the curve carries into it, is both common and clinically important in cerebral palsy. It is the feature that most directly disturbs sitting, because a tilted pelvis cannot provide the level, symmetrical base that comfortable, stable sitting requires. The combination of a long collapsing curve above and an oblique pelvis below is, in many ways, the signature of the spinal deformity seen in severe cerebral palsy, and much of the surgical thinking in this field is directed at correcting both together.

How the curve behaves over time

The spinal curve of cerebral palsy is not static. Where the brain injury itself is non-progressive, the spinal deformity it gives rise to is anything but, and the natural tendency of these curves, particularly the larger ones in the most severely affected children, is to increase over time.

Curves often begin to appear during childhood and tend to progress through the growth spurt of adolescence, when the rapid lengthening of the spine offers the deforming forces their greatest opportunity. The likelihood of developing a significant curve, and the speed at which it grows, both rise with the severity of the underlying cerebral palsy, so that the steepest progression is seen in non-ambulant children at the higher GMFCS levels.

An important feature, and one that distinguishes neuromuscular scoliosis from the idiopathic scoliosis of healthy adolescents, is that the curve does not necessarily stop progressing once growth is complete. In idiopathic scoliosis, smaller curves tend to stabilise at skeletal maturity. In cerebral palsy, by contrast, the larger curves can continue to increase into adult life, because the underlying loss of muscular control that drives them does not resolve with maturity. Systematic reviews of progression after skeletal maturity describe continued increase in a substantial proportion of patients, particularly those whose curves were already large at the end of adolescence and those most severely affected, with the curve advancing by a degree or more each year. This continuing tendency to worsen is one of the reasons the deformity is taken so seriously.

Effects on sitting, seating and care

For a person who cannot stand or walk, the most important consequence of a spinal curve is its effect on sitting. Sitting is the posture in which much of daily life is conducted, and a stable, comfortable, level seated position is fundamental to being able to interact, to eat, to use the hands, and to be cared for. A long collapsing curve with an oblique pelvis directly undermines this.

As the trunk leans and the pelvis tilts, the even base that comfortable sitting depends on is lost. Weight is thrown disproportionately onto one buttock, which raises the risk of pressure problems over the skin in that area. The trunk may need to be propped or supported to prevent it folding to one side, and the hands, which might otherwise be free for use, can become occupied in holding the body up. Head control, already often limited, can be further compromised when the trunk beneath is unstable. In short, a deformity of the spine translates directly into a loss of seated function.

There is a knock-on effect on care. A trunk that has collapsed to one side, with a tilted pelvis and uneven pressure, is harder to seat, harder to position, and harder to keep comfortable. Specialised seating systems, with moulded supports, tilt and pressure-relieving cushions, are widely used to accommodate the deformity and to maintain as level and as comfortable a posture as possible, and these form an important part of non-surgical management. The aim of much of the care surrounding cerebral palsy scoliosis, whether through seating or through surgery, is ultimately to preserve or restore the ability to sit comfortably and to be cared for with dignity.

The hip-spine relationship

The spine in cerebral palsy cannot be considered in isolation from the hips, because the two are linked through the pelvis that sits between them. The pelvis is the base on which the spine stands and the platform from which the hips hang, and a deformity in one region tends to be reflected in the others.

The hips in severe cerebral palsy are themselves at high risk, the imbalanced muscle pull tending to draw the head of the femur progressively out of its socket in a process of subluxation and, ultimately, dislocation. A related pattern is the windswept deformity, in which both legs are drawn over to the same side, one hip turning inwards and the other outwards, so that the legs appear swept across as if by a wind. These hip deformities are intimately connected to the position of the pelvis and, through it, to the spine.

Studies of the patterns of deformity in non-ambulant cerebral palsy have shown that the direction of the spinal curve, the direction of the pelvic obliquity, the direction of the windswept hips and the side of hip dislocation are not independent of one another but tend to follow a consistent, linked pattern. A common sequence described is

dislocation of a hip, followed by tilting of the pelvis, followed in turn by curvature of the spine, with the convexity of the spinal curve typically lying opposite to the direction of the windsweeping. The practical importance of this is that the spine, the pelvis and the hips form a single chain. A correction or a problem at one level inevitably has implications for the others, and the assessment of cerebral palsy scoliosis is, in reality, an assessment of the whole hip-pelvis-spine complex.

Surgical considerations: long fusions to the pelvis

When a curve in cerebral palsy becomes severe, progressive and disabling, the surgical option that is most often considered is a long posterior spinal fusion, frequently extending down to include the pelvis. The aim of such an operation is to straighten the spine as far as is safe, to level the pelvis, and to fuse the corrected position permanently into a single solid block of bone, so that the deformity can no longer progress and a balanced, level seated posture is restored.

The reason these operations are so extensive is the nature of the curve. Because the deformity is long and collapsing and frequently carries into the pelvis, a short fusion of a few segments cannot address it. Instead, the instrumentation, typically a construct of screws and rods running down each side of the spine, must usually span most of the length of the spine and anchor into the pelvis itself, in order to control the whole curve and to correct the pelvic obliquity that a fusion stopping short of the pelvis would leave uncorrected. Published series using modern segmental screw techniques describe substantial corrections of both the spinal curve and the pelvic obliquity, frequently of the order of three-quarters of the deformity, with the goal of producing a level, balanced and comfortable sitting position.

The decision to undertake such surgery is not taken on the basis of the spine alone. Because the operation is large and the patients are often medically fragile, the whole person must be weighed in the balance: their general health, their nutrition, their chest and breathing, the state of their hips, and, above all, what the family and carers hope the operation will achieve for daily life. This is reconstructive surgery aimed at function and comfort rather than at producing a normal spine, and the literature consistently frames it as a major undertaking with both substantial potential benefits and substantial risks.

The balance of benefit and risk

Surgery for cerebral palsy scoliosis sits at one of the extremes of spinal surgery in terms of both the size of the potential benefit and the size of the potential risk, and an honest account has to hold the two together.

On the side of benefit, the operation can achieve a large and durable correction of a deformity that would otherwise continue to worsen. It can level the pelvis, restore a balanced seated posture, halt the progression of the curve, and address the problems of sitting, pressure and care that the deformity creates. Studies that have asked carers and families about the results, rather than measuring the spine alone, have frequently reported high levels of satisfaction and meaningful improvements in ease of seating, comfort and care, although it is fair to say that the evidence on quality of life is not uniform, and some studies have found the picture more mixed, with measurable gains harder to demonstrate on formal quality-of-life instruments than the high satisfaction rates would suggest. The benefit, where it is realised, tends to be expressed in terms of sitting, comfort and the practicalities of daily care rather than in the things measured after surgery for idiopathic scoliosis.

On the side of risk, this is among the higher-risk operations in elective orthopaedics, precisely because of the comorbidities the patients carry. Large series document major complication rates that are substantial, with problems of the chest and lungs, wound and deep infection, considerable blood loss, and difficulties related to nutrition and the gut all featuring prominently. A small but real perioperative mortality is reported in the literature, reflecting the fragility of the most severely affected patients. Reoperation, often for problems with the wound or the instrumentation, is not uncommon. None of this argues against the operation, which can transform the situation for the right person, but it explains why the decision is made carefully, by an experienced team, with full and frank discussion of what is being weighed.

Comorbidity: nutrition, the chest and the whole child

A recurring theme in the literature on cerebral palsy and the spine is that the spine cannot be operated on as if it were the only problem. The children and adults who reach the point of needing major spinal surgery are frequently affected in many other systems, and these comorbidities shape both the risk of the operation and the preparation for it.

Nutrition is a central concern. Difficulty with feeding and swallowing is common in severe cerebral palsy, and undernutrition, with low body weight and poor nutritional reserves, is associated with higher rates of wound and other complications after major surgery. Optimising nutritional status before an operation, sometimes with the help of feeding support, is widely regarded in the literature as an important part of reducing risk.

The chest is the other dominant concern. Respiratory function is frequently impaired in severe cerebral palsy, through a combination of weak respiratory muscles, the mechanical effect of the deformity itself on the rib cage and lungs, a tendency to recurrent chest infection, and the risk of aspirating food or secretions. These factors make respiratory complications one of the commonest problems after spinal surgery in this group, and pre-existing lung function is one of the strongest predictors of how a person will fare. Seizure disorders, problems with the gut and bowel, and the general frailty that accompanies severe neurological disability all add to the picture. The consistent message of the literature is that the safe management of cerebral palsy scoliosis is the management of the whole child, with the spine considered as one part of a complex medical whole.

Outlook

The outlook in cerebral palsy and the spine depends above all on the severity of the underlying condition. For those who are mildly affected and able to walk, a significant spinal curve is relatively uncommon, and the spine is unlikely to become a dominant concern. For those at the severe end of the spectrum, who cannot sit or stand unsupported, the spine is one of the central orthopaedic issues, with a high likelihood of a curve that develops in childhood, progresses through adolescence, and may continue to worsen into adult life.

Where surgery is undertaken for a severe, progressive and disabling curve, the realistic aim is a level, balanced and comfortable seated posture and a halt to progression, rather than a normal spine. The reported results, judged in those terms and from the standpoint of families and carers, are often good, with high satisfaction and real gains in seating and ease of care, set against a genuinely high burden of surgical risk that reflects the fragility of the patients rather than any failing of the operation.

The themes that run through the whole subject are consistent. The spinal deformity is a neuromuscular consequence of a non-progressive brain injury, driven by loss of muscular control over a growing skeleton. It produces a characteristic long, collapsing curve with pelvic obliquity, intimately linked to the hips. It matters most for its effect on sitting and care. And its management, whether through seating or through major surgery, is always the management of the whole person rather than of the spine alone. Any individual outlook is a matter for assessment by the treating team, considered in the round.