

Early Onset Scoliosis

A patient guide

In brief: Early onset scoliosis is the term used for any sideways curvature of the spine that begins before the age of ten, whatever its underlying cause. The age cut-off is not arbitrary. It marks out the period in which the lungs and the chest are still being built, and that single fact is what sets early onset scoliosis apart from the more familiar curves that appear in teenagers. In a teenager, scoliosis is largely a problem of shape and of the spine itself. In a young child, a curve that distorts the chest can interfere with the growth of the lungs at the very time those lungs are forming, so the condition is as much about breathing and growth as it is about the spine. This dual character, a deformity that is also a developmental problem of the chest and lungs, runs through everything that follows.

What early onset scoliosis is (and why age matters so much)

Scoliosis is a curvature of the spine to the side, usually combined with a twist, measured on an X-ray as the Cobb angle. A curve is generally counted as scoliosis once it exceeds ten degrees. The phrase early onset scoliosis simply gathers together all such curves that appear before a child's tenth birthday, regardless of the reason behind them. It is therefore an umbrella term rather than a single diagnosis, covering a wide spectrum of children whose curves differ enormously in cause, in behaviour and in outlook.

The choice of age ten as the dividing line reflects biology rather than convenience. The origins of the modern concept lie in the 1980s. In 1985 James (Ian) Dickson argued that juvenile idiopathic scoliosis was not a truly distinct biological entity, and proposed instead grouping affected children into early-onset scoliosis, from birth to about five years, and late-onset scoliosis, after about five years. This way of thinking helped shape the modern early-onset concept, before the practical boundary later shifted towards about ten years. That later revision was reached by international consensus, by the Scoliosis Research Society and the Pediatric Orthopaedic Society of North America, drawing the line at ten years. The reason is that the first decade of life is the time when the chest and the lungs do most of their growing, and a spinal curve that arrives during this window has the potential to interfere with that growth in a way that a curve appearing later does not.

This is the central idea that distinguishes early onset scoliosis from adolescent scoliosis. Adolescent idiopathic scoliosis, the common curve of the teenage years, generally appears at or after the age of ten, by which time the lungs are largely formed and the chest has taken much of its final shape. In that setting scoliosis is principally a question of the spine, of posture and appearance and sometimes of back discomfort, and it rarely threatens the lungs. Early onset scoliosis is different precisely because it occurs while the lungs are still being made. The same number of degrees of curve carries a different meaning depending on when in a child's growth it appears.

The growing lung and chest: why timing is everything

To understand early onset scoliosis, it helps to understand how the lungs and chest develop, because this is the engine that drives the whole condition. The lungs are not finished at birth. The great majority of the tiny air sacs that do the work of breathing, the alveoli, are formed after a baby is born. A newborn has in the region of twenty million alveoli, and this number multiplies many times over to reach something close to three hundred million by around the age of seven or eight. After that age the lungs grow mainly by getting larger rather than by adding many new air sacs.

The chest grows on a similar timetable. The volume of the thorax reaches roughly a third of its adult size by the age of five and about half by the age of ten, with the remainder added during the adolescent growth spurt. The spine itself grows quickly in the early years and then more slowly through middle childhood before accelerating again at puberty.

The consequence of all this is a window of vulnerability. A spinal curve that distorts the chest during the years when the alveoli are still multiplying can limit the space available for the lungs to develop, and the lung tissue lost during that window may never be fully recovered. This is the reason a curve in a young child is taken so seriously. It also explains a principle that has reshaped thinking in this field over recent decades, namely that a short, straight spine produced by stopping growth early is no longer regarded as preferable to a longer, gently curved spine that has been allowed to keep growing. Preserving growth, and with it the room for the lungs, has become as important an aim as correcting the curve.

How early onset scoliosis differs from its neighbours

Early onset scoliosis is best understood by placing it alongside the conditions it is most often confused with, because the differences are instructive.

Compared with **adolescent idiopathic scoliosis**, the contrast is one of timing and of stakes. Adolescent curves appear after the lungs are essentially complete and behave mainly as a deformity of the spine. Early onset curves appear during lung growth and can, in their severe forms, lead to restrictive lung disease, a state in which the chest cannot fully expand. Difficulty with breathing and, at the extreme, strain on the heart are recognised in early onset disease but are not features of the adolescent form. The timing changes the entire character of the problem.

Compared with **infantile scoliosis**, the relationship is one of part to whole. Infantile idiopathic scoliosis, with onset in the first three years, is simply the youngest subgroup within early onset scoliosis. It has one striking feature that sets it apart from teenage curves: a proportion of infantile curves resolve on their own as the child grows, a behaviour almost never seen in adolescent scoliosis. This is why tools were developed specifically to tell which infantile curves are likely to disappear and which are likely to progress, a distinction described below.

Compared with **congenital scoliosis**, the difference lies in the nature of the bones themselves. Congenital scoliosis is caused by vertebrae that formed abnormally before birth, such as a half-formed wedge of bone, a hemivertebra, or a bony bar that tethers one side of the spine. Here the curve is built into the skeleton rather than being a bending of otherwise normal bones, which is why congenital curves tend to respond less well to braces and casts, and why they are more often accompanied by abnormalities of the heart, kidneys or spinal cord that warrant looking for. Congenital scoliosis is one of the recognised categories within early onset scoliosis, alongside idiopathic, neuromuscular and syndromic types.

The four families: idiopathic, congenital, neuromuscular, syndromic

Because early onset scoliosis is an umbrella, it is divided according to cause into four broad families, and this grouping shapes how each curve is understood.

Idiopathic early onset scoliosis is a curve with no identifiable cause in an otherwise healthy child with normally formed bones. The infantile idiopathic curves, including those that resolve, sit here.

Congenital early onset scoliosis arises from vertebrae that are malformed from birth, through a failure of formation, a failure of separation, or a mixture of the two. The most relentlessly progressive pattern is a bar joining the vertebrae on one side together with a hemivertebra on the other, which drives the curve in one direction with each year of growth.

Neuromuscular early onset scoliosis develops on the background of a condition affecting the nerves or muscles, such as cerebral palsy, spinal muscular atrophy or a muscular dystrophy. The curve reflects the underlying imbalance of muscle control acting on a growing spine.

Syndromic early onset scoliosis occurs as one feature of a wider syndrome affecting connective tissue or the skeleton more generally. These children often have other features of their syndrome that influence how the spine behaves and how it is managed.

These four families form the backbone of the formal classification described next, and they matter because a congenital curve, an idiopathic curve and a neuromuscular curve of the same number of degrees may behave very differently.

Measuring the curve: Cobb angle and the Mehta angle

Two measurements recur throughout the description of early onset scoliosis, and both are worth understanding.

The **Cobb angle** is the standard way of expressing how large a curve is. It is measured on a front-to-back X-ray as the angle between the most tilted vertebrae at the top and bottom of the curve. It is the number that follows a curve over time, and broadly speaking the larger the Cobb angle in a growing child, the greater the concern. Lung function tends to become measurably affected once curves pass somewhere in the region of fifty to sixty degrees, and very large curves beyond about ninety degrees are strongly associated with strain on the heart and lungs in the growing child.

The **rib-vertebra angle difference**, often called the Mehta angle after the surgeon who described it, is a more specialised measurement used chiefly in the youngest children. Described by Mehta in 1972, it compares the angle that the ribs make with the spine on the inner and outer sides of the curve at its apex. Its purpose is to predict, in infantile scoliosis, which curves are likely to resolve and which are likely to progress. The much-quoted rule of thumb is that a difference below twenty degrees is associated with a curve that tends to resolve, while a difference of twenty degrees or more is associated with a curve that tends to progress, the so-called eighty-twenty pattern. A particular appearance of the rib head, overlapping the vertebra, is taken as a further sign that a curve is more likely to advance. None of this is a prediction for any individual child; it is a description of how these measurements behave across groups.

How early onset scoliosis is classified (C-EOS)

For many years early onset scoliosis lacked a shared language, which made it hard to compare children or to study treatments. This was addressed in 2014 with the publication of the Classification of Early Onset Scoliosis, usually shortened to C-EOS, developed by a consortium of surgeons led by Williams and colleagues.

The system records several pieces of information in a compact form. It begins with the child's **age**. It then records the **cause**, using the four families already described: congenital or structural, neuromuscular, syndromic, or idiopathic. It grades the **size of the main curve** into bands, broadly under twenty degrees, twenty to fifty degrees, fifty-one to ninety degrees, and over ninety degrees. It notes the amount of **kyphosis**, the forward curve of the spine seen from the side, as low, normal or high. And it can add an optional **progression** marker describing how quickly the curve is changing each year.

The value of C-EOS is that it brings together the features that actually predict how a curve behaves and how it responds to treatment, in a way that different teams can use consistently. Studies have shown it to be reproducible between observers and have linked its categories to real outcomes, such as the speed at which certain implants loosen and the likelihood of lung problems. It is now a standard reference point in describing these curves, though it is a way of organising information rather than a treatment plan in itself.

Thoracic insufficiency syndrome

At the severe end of the early onset spectrum lies a concept that captures, better than any measurement of the spine alone, what is truly at stake. This is thoracic insufficiency syndrome, a term introduced by Robert Campbell. It is defined as the inability of the chest to support normal breathing or normal lung growth.

The idea behind it is that a severe three-dimensional distortion, involving not only the spine but the ribs and the shape of the whole chest, can shrink the space the lungs have to grow into and stiffen the chest so that it cannot expand and contract properly. In a young child, whose lungs are still multiplying their air sacs, this can limit lung development in a way that has lifelong consequences. Thoracic insufficiency syndrome reframed the whole field, because it shifted the goal of treatment away from simply straightening the spine and towards preserving the volume of the chest and the growth of the lungs. It is also the reason that some of the devices described below are designed to act on the ribs and the chest, not only on the spine. Not every child with early onset scoliosis develops thoracic insufficiency; it is a feature of the more severe and complex curves, particularly those with rib abnormalities.

The natural history of an untreated curve

The course an early onset curve takes if left to progress depends heavily on which family it belongs to and on how it behaves in the early years. The general picture drawn from the literature is that a relentlessly progressive curve in a young child can lead to severe deformity and, in its most serious form, to restrictive lung disease, to raised pressure in the lung circulation and to strain on the heart, with a measurable effect on health and life expectancy. This grave outlook for the most severe untreated curves is the historical reason the condition is regarded so seriously, and it stands in deliberate contrast to the generally benign course of an adolescent curve.

Within this, the families behave differently. Many infantile idiopathic curves, especially those with a low rib–vertebra angle difference, resolve without intervention. Others progress and require attention. Congenital curves vary according to the precise malformation, with the bar-and-hemivertebra combination being the most progressive. Neuromuscular and syndromic curves track the demands of the underlying condition. The aim of describing natural history is not to predict any one child's path but to explain why curves are watched so closely during the years of rapid growth and why the response differs so much from child to child.

What the evidence shows about growth-friendly treatment — impersonal

It is important to state at the outset that the evidence base in early onset scoliosis is built largely on case series, registry data, classification and reliability studies and expert consensus, rather than on large randomised trials, which are difficult to conduct in a condition this varied and this uncommon. The descriptions that follow set out what the literature reports about the available approaches; they are not guidance for any individual child, and decisions in practice are made jointly by families and treating teams on a case-by-case basis. The unifying theme across all of these approaches is the attempt to control the curve while allowing the spine and chest to keep growing, which is why they are collectively called growth-friendly.

****Serial casting****, often called Mehta casting after the surgeon who refined it, uses a series of plaster jackets applied under anaesthetic and changed every few months, working with the child's rapid early growth to gently guide the spine straighter. Mehta's influential series of one hundred and thirty-six children reported that, when started early and in small flexible curves, many infantile idiopathic curves could be corrected or even resolved, with the best results in those treated before the age of two. The literature describes casting as most effective for flexible idiopathic curves and less so for congenital and syndromic ones, and it notes the burden of repeated anaesthetics as a recognised drawback.

****Bracing**** uses a removable external jacket, and is described in the literature as a means of controlling milder or flexible curves or of delaying surgery, with congenital curves responding less well.

****Traditional growing rods**** are an operative, distraction-based approach in which rods are anchored above and below the curve and lengthened through repeated small operations, classically about every six months, to keep pace with growth. The reported series, including the large multicentre work associated with Akbarnia and colleagues, describe that growth can be maintained and a substantial part of the curve corrected, but at the cost of a high rate of complications that rises with the number of lengthenings, including problems with the anchors, breakage of the rods and infection. The need for repeated open operations is the principal limitation described.

****Active Apex Correction (APC)**** is a growth-guidance growing-rod strategy that takes a different route to the same goal. Rather than relying on repeated distraction at the ends of the construct, it is designed to act directly on the apex of the curve, the most deformed point, applying a corrective force there to draw the curve straighter while still permitting the spine to grow. This contrasts with traditional distraction rods, which are lengthened at intervals through small operations to keep pace with growth. The intention of an apex-directed, growth-guidance approach is to obtain and maintain correction at the part of the spine that drives the deformity, while preserving the growth that protects the developing chest and lungs.

****Definitive spinal fusion****, the permanent joining of the affected vertebrae, is the long-standing operation for adolescent scoliosis but is approached with caution in young children. The literature, including work by Karol and colleagues, documents that fusing the chest portion of the spine early, particularly long fusions and those high in the chest, is associated with reduced lung function in later life, and that a short straight spine is no longer regarded as a desirable end in itself. A related concern in the very young is the crankshaft phenomenon, in which continued growth at the front of a spine fused only at the back produces a recurring twist. For these reasons definitive fusion is generally reserved for when adequate chest and lung growth has been achieved, or for selected localised congenital problems such as removal of a single malformed vertebra. A further recurring theme across all the distraction methods is the so-called law of diminishing returns described by Sankar, whereby the spine gradually stiffens and gains less length with each lengthening, partly through spontaneous fusion of the bones, which is one of the reasons children treated with these systems eventually move on to a final operation.

In summary, the evidence describes a graded toolbox running from casting and bracing in the youngest and mildest curves, through the various growth-friendly implants for progressive curves in the growing child, to definitive fusion once growth is sufficiently complete, all chosen within a framework of individualised, shared decision-making rather than fixed rules.

Outlook

The outlook in early onset scoliosis is as varied as the condition itself, and it cannot be read from a single number. At the favourable end sit the many infantile idiopathic curves that resolve on their own, and the flexible curves caught early and controlled well, where the general picture reported in the literature is encouraging in terms of both the spine and the lungs. At the more demanding end sit the severe, progressive and complex curves, particularly those associated with abnormal ribs or with thoracic insufficiency syndrome, where the central concern is the lungs rather than the spine alone, and where the aim of treatment is to protect lung growth through the critical years.

Across the whole spectrum, several themes are consistent in the literature. The behaviour of the curve depends strongly on its cause and on how early in growth it appears. Preserving the growth of the spine and chest has become as important a goal as correcting the curve. And the modern toolbox of growth-friendly treatments has shifted the emphasis away from early permanent fusion. The variation between children is wide, and any individual outlook is a matter for assessment by the treating team in the light of that particular child's curve, cause and growth, rather than something that can be inferred from a category alone.