

Infantile Idiopathic Scoliosis

A patient guide

In brief: Infantile idiopathic scoliosis is a sideways curvature of the spine that arises in very young children, by definition before the third birthday, with no identifiable cause and with normally formed vertebrae. It occupies an unusual place among the scolioses. On the one hand, the majority of these curves are gentle and resolve entirely on their own, leaving no trace; on the other, a minority are progressive and, if left to run their course, can become severe and rigid at an age when the lungs and chest are still developing. Because the same name covers two such different destinies, the central task in infantile scoliosis is not simply to measure the curve but to work out, as early and as reliably as possible, which path a given curve is on. This is what most distinguishes the infantile form from the scolioses of later childhood and adolescence.

What infantile idiopathic scoliosis is, and how it is classified

Scoliosis is a curvature of the spine in the side-to-side plane, usually accompanied by a degree of rotation, so that the spine twists as it bends. The word idiopathic means that no cause can be identified: the vertebrae are normally formed, there is no underlying nerve or muscle disorder, and no syndrome explains the curve. It is, in essence, a diagnosis of exclusion.

The idiopathic scolioses are traditionally divided by the age at which they appear. The framework most often cited dates from James in 1954 and remains in everyday use. A curve that appears before the age of three is called infantile; one appearing between three and ten years is called juvenile; and one appearing after the age of ten, up to the end of growth, is called adolescent. These age boundaries may look arbitrary, but they carry real prognostic weight, because the behaviour of an idiopathic curve, and the company it keeps, differ markedly depending on when in childhood it first declares itself.

A broader and more modern term, early onset scoliosis, gathers together every scoliosis that begins before the age of ten, whatever its cause. Infantile idiopathic scoliosis is therefore the idiopathic member of the early onset family, occupying its youngest end. The early onset concept matters because curves that develop in the first years of life do so while the lungs are still multiplying their air sacs and the chest is still growing. A severe curve at this stage threatens not only the shape of the back but the development of the lungs themselves, which is why early onset scoliosis is regarded as a different and more serious problem than a curve of similar size appearing in a teenager.

Infantile idiopathic scoliosis is uncommon, accounting for only a small proportion of all idiopathic scoliosis, historically more frequently seen in the United Kingdom and Europe than in North America. Its rarity, combined with its capacity to either vanish or progress dramatically, gives it a reputation out of proportion to its numbers.

How it differs from its neighbours

The single most useful way to understand infantile idiopathic scoliosis is to set it beside the conditions it can be confused with, because in almost every respect it behaves differently.

It differs first from **congenital scoliosis**. A congenital curve is caused by a vertebra that did not form properly before birth, whether through a half-formed vertebra, two vertebrae fused together, or a bony bar tethering one side of the spine. In infantile idiopathic scoliosis, by contrast, the vertebrae are normally shaped; the curve arises in a structurally normal spine. This distinction is fundamental, because it changes both the cause and the management entirely, and it is one of the first things imaging is used to settle.

It differs, second, from the **juvenile and adolescent idiopathic curves** that follow it in childhood. The infantile form shows a male predominance, whereas the adolescent form predominantly affects girls. The infantile curve is usually a left-sided thoracic curve, whereas the classic adolescent curve bends to the right. And, most strikingly, the infantile curve carries a genuine possibility of spontaneous resolution, a disappearance of the curve without any treatment, which is something the established adolescent curve does not do. Once an adolescent curve is present it may stay the same or worsen, but it does not melt away; many infantile curves do exactly that.

It belongs, third, within the **early onset scoliosis** umbrella already described, sharing with the rest of that group the central concern for the growing lungs and chest. What sets the idiopathic infantile curve apart even within this group is precisely its unpredictability, its split between curves that will resolve and curves that will progress, and the long-established tools that exist to tell them apart.

The resolving curve and the progressive curve

If there is one idea that defines infantile idiopathic scoliosis, it is the division between curves that resolve and curves that progress. This division is the reason the condition is approached so differently from later scoliosis.

The reassuring fact, established in the older natural history studies and confirmed since, is that the great majority of infantile idiopathic curves resolve spontaneously. Figures in the literature vary with how the children were referred and studied, but resolution rates in the region of eighty to ninety per cent are commonly quoted. In these children the curve, often noticed in the first months of life, gradually straightens of its own accord as the child grows, and leaves no lasting deformity. No treatment alters this natural improvement; it simply happens.

The minority, perhaps one in ten to one in five, follow the opposite course. Their curves progress, sometimes slowly and sometimes relentlessly, and it is this progressive group that gives infantile scoliosis its serious reputation. The natural history of an untreated progressive curve is sobering: a curve still progressing at the age of five has, in many series, a better than even chance of exceeding seventy degrees, and severe early curves can become rigid and difficult to correct.

Because the two groups look similar at the outset but diverge so widely, the entire early approach to infantile scoliosis is built around distinguishing them. This is done partly by measurement at the first visit and partly by watching the trend over a series of films, because a curve that is shrinking on successive radiographs is behaving as a resolving curve, while one that is steadily enlarging is declaring itself progressive.

Mehta's rib-vertebra angle difference, and the phase of the rib head

The most enduring tool for separating resolving from progressive infantile curves was introduced by Min Mehta in 1972, and it remains central today. It rests on a simple observation: in the chest, the ribs and the vertebrae move together, and the angle at which a rib meets its vertebra changes as the spine rotates. By measuring this angle on both the inner, concave side of the curve and the outer, convex side at the apex, and taking the difference between them, Mehta derived a single number, the rib-vertebra angle difference, abbreviated to RVAD.

The value of this measurement lies in what is sometimes called the eighty-twenty rule. In Mehta's original study of one hundred and thirty-eight children, about eighty per cent of curves with a rib-vertebra angle difference below twenty degrees resolved on their own, while about eighty per cent of those with a difference above twenty degrees went on to progress. A low difference therefore points towards resolution and a high one towards progression, though neither is a certainty, and the trend across serial films carries as much weight as any single reading. More recent work has suggested the dividing line may sit a little below the classic twenty degrees, but the principle is unchanged.

Mehta added a second, related observation about the rib head at the apex of the curve. In what she called phase one, the head of the rib on the convex side does not overlap the body of the apical vertebra; a clear gap remains between them, indicating relatively little rotation. In phase two, the rib head has come to overlap the vertebral body, a sign of greater rotation and a more established, rotated curve. A phase two appearance is regarded as a

marker of a progressive curve in its own right. Together, the rib–vertebra angle difference and the phase of the rib head give a structured way of reading an early film and forming an impression of which way the curve is likely to go.

Why the spine may be scanned to look beyond the curve

The word idiopathic carries an important caveat in the very young, and it is one of the features that sets infantile scoliosis apart from the adolescent form. A curve that looks idiopathic can occasionally be the outward sign of an abnormality within the spinal canal or the lower brain, where the nervous system itself is at fault rather than the bones.

The relevant abnormalities include a low-lying part of the brain at the base of the skull, known as a Chiari malformation, a fluid-filled cavity within the spinal cord called a syringomyelia or syrinx, a cord that is split or tethered, and, rarely, a tumour. These neural axis abnormalities are found far more often in early onset and infantile curves than in adolescent ones. Series of young children with apparently idiopathic scoliosis have reported such findings in a substantial minority, in some large studies approaching one in five, with Chiari malformation and syrinx together making up the majority of them.

Certain features raise the likelihood of an underlying abnormality and are recognised in the literature as reasons that magnetic resonance imaging of the whole spine is often advocated: very early onset, male sex, a curve that bends in the atypical left thoracic direction, an unusually marked forward or backward component to the curve, rapid progression, and any abnormality on neurological examination. The purpose of looking is straightforward. If a Chiari malformation, a syrinx or a tethered cord is present, the curve is not truly idiopathic, and the underlying problem becomes the focus, which changes the whole direction of care. This is descriptive of why such scans feature in the assessment of young children with scoliosis, not a recommendation directed at any individual child.

The outlook of an untreated progressive curve

The reason progressive infantile scoliosis is taken so seriously is that its untreated natural history reaches well beyond the appearance of the back. A curve that develops and worsens in the first years of life does so during the window in which the lungs are still forming their air sacs and the chest is still enlarging. A severe, rigid curve at this age can prevent the chest from supporting normal lung growth, a state described as thoracic insufficiency syndrome, in which the thorax is simply unable to do its job.

The consequences of this, documented in long-term follow-up studies of untreated early onset curves, can include restricted lung function, strain on the right side of the heart, raised pressure in the lung circulation, and, in the most severe and longstanding cases, a shortened life from cardiopulmonary failure in later adulthood. This is a markedly worse natural history than that of the adolescent curve, in which important effects on heart and lung function generally appear only with very large curves and in which life expectancy is far less affected. The earlier in life a severe curve becomes established, the greater the threat to the lungs. It is this difference, more than any other, that justifies treating progressive infantile curves early and in a way that protects growth, and it is the backdrop against which all the treatments below are understood.

Mehta serial casting, the elongation–derotation–flexion method

For progressive infantile curves, the treatment most closely associated with the condition is serial casting by the elongation–derotation–flexion method. The cast itself was devised in France by Cotrel and Morel, but it was Min Mehta who turned it into a serial, corrective strategy and who showed that, in the youngest children, growth could be harnessed as a force to correct the curve rather than merely to hold it.

The principle is that a custom-moulded cast, applied to the trunk under a brief general anaesthetic on a special frame, applies three corrective forces at once: a lengthening pull along the spine, a derotating twist to unwind the rotation, and a gentle flexion. Windows are cut in the cast to allow the chest and abdomen to expand and to guide the growing spine towards a straighter shape. The cast is changed every two to three months, more often in the

youngest and fastest-growing children, so that as the child grows the correction is repeatedly renewed.

What makes this approach distinctive is its ambition. In a young child with a flexible, moderate curve, serial casting can do more than contain the curve; it can resolve it permanently, so that the child needs no further treatment and grows up with a straight spine. In an older child, or one with a larger and stiffer curve, casting is used instead as a holding measure, buying time and allowing the chest to grow before any decision about surgery. This dual role, curative in the young and flexible curve and temporising in the older one, is characteristic of casting in infantile scoliosis and is reflected throughout the published series.

What the evidence shows about treatment — impersonal

It is worth stating plainly that the evidence base in infantile idiopathic scoliosis is built from cohort studies, case series and natural history reviews rather than large randomised trials, which the rarity of the condition makes difficult to mount. The account that follows describes what the literature reports, not guidance for any individual child.

****Observation**** is the established approach for curves judged likely to resolve, typically the smaller curves with a low rib–vertebra angle difference. The literature supports watching such curves with serial radiographs at intervals of several months, on the understanding that the great majority will improve on their own and that the trend across films, rather than any single measurement, reveals which way a curve is heading.

****Serial elongation–derotation–flexion casting**** is the best-supported active treatment for progressive curves. Mehta's own prospective study of progressive infantile and early onset curves reported full correction in the majority of children, with the curve resolving completely in those treated youngest, at around a year and a half of age and with moderate curves, who then needed no further treatment. Subsequent series have repeatedly found that a large proportion of children treated with derotation casting, often around seven to eight in ten, avoid surgery, or have it substantially delayed while the chest grows. The consistent message of these studies is that younger age, a smaller and more flexible curve, and a genuinely idiopathic cause predict the best results.

****Bracing****, usually with a custom thoracolumbosacral orthosis, features in the literature mainly as a way of maintaining correction after a course of casting, or as an alternative for smaller or less rigid curves. Its evidence as a primary curative treatment is weaker than that for casting, and its long-term effectiveness in this age group is less firmly established.

****Growth-friendly surgery**** is described for progressive curves that cannot be controlled by casting or bracing, or that present already severe. The guiding aim is to control the deformity while allowing the spine and chest to keep growing, because a definitive fusion in a very young child would leave a short trunk and restrict the lungs. The reported techniques include growing rods anchored above and below the curve and lengthened periodically, magnetically controlled growing rods that can be lengthened without repeated operations using an external magnet, and rib-based devices used particularly where the chest wall is involved. Definitive fusion of the spine is generally deferred until the child is more mature. Across all of these, the literature frames the choice as an individualised one, weighing the severity and stiffness of the curve, the age of the child and the state of the lungs.

Outlook

The outlook in infantile idiopathic scoliosis is, more than in almost any other spinal deformity, a tale of two populations. For the large majority whose curves resolve, the long-term picture is excellent: the curve straightens with growth, no treatment is needed, and there is no lasting effect on the back or the lungs. For these children the condition, in retrospect, was a self-limiting phase rather than a disease.

For the minority with progressive curves, the outlook depends heavily on how early the progression is recognised and addressed and on how the chest develops. Where a progressive curve is treated young, while it is still flexible, the published results of serial casting are encouraging, with many curves resolving or being held in check and surgery avoided or postponed. Where progression is severe, longstanding or recognised late, the demands of

treatment are greater and the concern for the lungs more pressing, which is why the progressive group attracts such close attention.

The themes that run through the whole field are consistent. The earlier a progressive curve is identified, the better the prospects; the trend of the curve across serial films is more informative than any single reading; the rib–vertebra angle difference and the phase of the rib head help separate the two destinies; and any individual child's outlook is a matter for assessment by the treating team rather than something that can be read from a category alone.