

CLINICAL REVIEW

Adolescent idiopathic scoliosis

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Scoliosis is a three dimensional deformity of the spine defined as a lateral curvature of the spine in the coronal plane of more than 10°. It can be categorised into three major types—congenital, syndromic, and idiopathic. Congenital scoliosis refers to spinal deformity caused by abnormally formed vertebrae. Syndromic scoliosis is associated with a disorder of the neuromuscular, skeletal, or connective tissue systems; neurofibromatosis; or other important medical condition. Idiopathic scoliosis has no known cause and can be subdivided based on the age of onset—infantile idiopathic scoliosis includes patients aged 0-3 years, juvenile idiopathic scoliosis includes patients aged 4-10 years, and adolescent idiopathic scoliosis affects people aged >10 years.

Adolescent idiopathic scoliosis (AIS) is the most common spinal deformity seen by primary care physicians, paediatricians, and spinal surgeons.² This review is focused on AIS and reviews the diagnosis, management, and controversies surrounding this condition based on the available literature.

What causes adolescent idiopathic scoliosis?

The diagnosis of AIS is one of exclusion, and is made only when other causes of scoliosis, such as vertebral malformations, neuromuscular disorders, and other syndromes have been ruled out. According to epidemiological studies, 1-3% of children aged 10-16 years will have some degree of spinal curvature, although most curves will not require surgical intervention.^{3 4}

Suggested causes of AIS include mechanical, metabolic, hormonal, neuromuscular, growth, and genetic abnormalities.^{5 6} These factors are not yet well accepted as a direct cause for this condition. The current view is that AIS is a multifactorial disease with genetic predisposing factors.

What is the natural course of adolescent idiopathic scoliosis?

The natural course of scoliosis was studied in a prospective case series of 133 patients. The patients were followed for an average of 40.5 years (range 31-53 years), and 68% of adolescent idiopathic curvatures were found to progress beyond skeletal maturity. Thoracic curvatures greater than 50° progressed at an

average of 1° a year, thoracolumbar curves progressed at 0.5° a year, and lumbar curves progressed at 0.24° a year. Thoracic curvatures of less than 30° did not progress.⁷

Previous long term retrospective observational studies of idiopathic scoliosis presented a poor prognosis (respiratory failure, cardiovascular risk, and mortality).⁸ This has created a misinterpretation that all types of idiopathic scoliosis inevitably lead to disability from back pain and serious cardiopulmonary compromise. These studies included patients with mixed diagnoses, which could explain the poor outcomes reported. In a more recent prospective case-control study describing the 50 year natural course of untreated idiopathic scoliosis, there was no evidence linking untreated AIS with increased rates of mortality in general, and cardiopulmonary compromise in particular.⁹

Progressive scoliosis can result in the development of a worsening deformity and cosmesis.¹⁰ The physical deformities seen include the development of chest wall abnormality, rib prominences, asymmetry in shoulder height, and truncal shift.

How does adolescent idiopathic scoliosis present?

Patients with AIS most often present with unlevel shoulders, waist line asymmetry (one hip “sticking out” more than the other), or a rib prominence. This is usually first identified by the patient, family member, general practitioner, or a school nurse.

Back pain is sometimes the presenting complaint. The association between scoliosis and back pain has been demonstrated in a retrospective study of 2442 patients with idiopathic scoliosis,¹¹ which found that 23% of patients with AIS had back pain at initial presentation, and another 9% developed back pain during the study. An underlying pathological condition was identified in 9% (48/560) of the patients with back pain, mainly spondylolysis and spondylolisthesis and only one case of an intraspinal tumour.¹¹

Summary points

- Scoliosis is a lateral curvature of the spine measuring $>10^\circ$ in the coronal plane
- Several different types of scoliosis exist, and idiopathic scoliosis occurs in 0.5-3.0% of the paediatric population
- Initial evaluation should involve a focused history and physical examination. The Adam's forward bend test is particularly useful for detection
- Factors predicting curve progression include maturity (age at diagnosis, menarchal status, and the amount of skeletal growth remaining), curve size, and position of the curve apex
- Bracing is used to treat scoliosis in many European countries, but practice is divided in the UK and US, and elsewhere
- Surgery is recommended in adolescents with a curve of a Cobb angle more than 45° - 50°

Sources and selection criteria

We searched Medline and the Cochrane Library using MeSH terms "adolescent idiopathic scoliosis", and "scoliosis bracing". We included systematic reviews, randomised controlled trials, and good quality prospective observational studies mainly from the past 15 years but did not exclude seminal papers from before this time.

How is adolescent idiopathic scoliosis diagnosed?

On presentation of a patient with scoliosis to primary care, a detailed history, examination, and radiological investigations should be undertaken before referral to a specialist.

The history should include a detailed birth history, developmental milestones, family history of spinal deformity, and assessment of physiological maturity. Difficulties during labour can be associated with a diagnosis of cerebral palsy, which can lead to neuromuscular scoliosis. A history of developmental delay can be indicative of a non-idiopathic cause for the scoliosis.

Assessment of maturity includes inquiry about the growth spurt and the menarchal status in girls, as menarche indicates a point at which the growth starts to decrease over a period of two years from its onset.¹²

The patient's presenting complaint should be elicited, including back pain, neurological symptoms, and any concerns regarding cosmesis. The presence of constant pain, night pain, or radicular pain indicates that further investigations are required to exclude underlying pathology.¹³

When examining a patient with suspected scoliosis, adequate exposure is required to assess the spine appropriately. Boys should be examined in their underwear or shorts; girls should be wearing underwear and a bra. Gait and posture should be evaluated, looking in particular for a short-leg gait due to leg length discrepancy and listing to one side seen in severe curves.

The patient's upright posture should be evaluated from the front, back, and sides. The relative heights of the iliac crests and the shoulders should be observed for any asymmetry that could be indicative of curve severity. The pelvis should be level and any lower limb discrepancy compensated with a lift (a series of wooden blocks may be placed under the short leg until the hips are level). If a curvature of the spine is seen, the location and direction of the curve(s) should be noted. The curve is designated according to the direction of the curve convexity.

The back should be inspected for the presence of café au lait spots, subcutaneous nodules, and axillary freckles, which are seen in neurofibromatosis. The presence of hairy patches or skin dimples over the lower back can be an underlying sign of spinal dysraphism (a constellation of congenital abnormalities including defects of the spinal cord and vertebrae).

The balance of the thorax over the pelvis is assessed by dropping a plumb line from the C7 spinous process, which normally falls within the gluteal cleft. In cases of coronal imbalance the

distance from the plumb line to the gluteal cleft is measured in centimetres and the direction of deviation noted.

The Adam's forward bend test¹⁴ is carried out to assess the degree of rotational deformity associated with the scoliosis. The patient is asked to bend forward at the waist with the knees straight and the palms together (fig 1⇓). The examiner looks down the back for the presence of asymmetry in the rib cage (rib prominence) or deformities along the back indicative of a structural scoliosis. A non-structural curve (postural scoliosis) normally disappears on bending forwards.

A scoliometer is an instrument that is placed on the back and can be used to provide an objective measure of curve rotation.¹⁵ In primary care the use of a scoliometer is not required for the diagnosis of scoliosis, and suspected cases should be referred for specialist opinion on diagnosis.

A detailed neurological examination should be performed testing motor and sensory function and reflexes. Asymmetries in reflexes can be a sign of an intraspinal disorder.¹⁶ The abdominal reflex refers to the neurological reflex stimulated by stroking the abdomen around the umbilicus. This usually involves a contraction of the abdominal muscles, resulting in the umbilicus moving towards the source of the stimulation. An abnormal abdominal reflex may be suggestive of an intraspinal disorder and is often absent on the convex side of the curve.

What imaging is required?

Full length standing posteroanterior and lateral radiographs of the spine are required in order to assess the degree of deformity. These are taken with the patient in a standing position in order to assess the effect of gravity on the deformity. Patients are instructed to remove their shoes, and any lower limb discrepancy is compensated with a shoe lift before the radiograph is taken. Radiographs are taken with the patient looking straight ahead, legs apart for stability and with their hands on clavicles. If a radiograph is normal the patient and family can be reassured that there is no scoliosis. A referral can still be made if there is concern about pain, axial tenderness, or neurological abnormalities. If x ray facilities are not available, the patient may be referred directly to the specialist without radiographs.

On a full length posteroanterior plain radiograph, the magnitude of a scoliosis curvature is determined with the Cobb technique (fig 2⇓). Firstly, it is important to identify the superior and the inferior end vertebrae—the vertebrae with the greatest tilt at the proximal and distal ends of the curve. The angle between them is measured by drawing a line from the top of the superior end vertebra parallel to the upper endplate, and another line from the bottom of the inferior end vertebra parallel to the lower

endplate. Perpendicular lines are then constructed at right angles to the lines along the endplates. The angle formed by the intersection of the perpendicular lines defines the Cobb angle (fig 2).¹⁰

If surgery is considered, films of lateral bending view (full length posteroanterior plain radiographs with patient bending to the right and to the left) are first taken to determine curve flexibility, which is important in the preoperative evaluation and surgical planning.

The presence of a left thoracic curve or an abnormal neurological finding are most predictive of the presence of an underlying disease and warrant referral for further imaging.¹¹ Magnetic resonance imaging is useful for the identification of tumours and other pathological lesions—associated neural axis abnormalities such as syrinx (a fluid filled cavity within the spinal cord) and Arnold-Chiari malformations.¹⁸

What are the risk factors for curve progression?

For decisions about choosing conservative or surgical treatment, the child's maturity and the severity of the curvature are the two most important factors. It is important to evaluate maturity because the younger the child the greater is the likelihood of curve progression, equally the larger the curve magnitude the greater is the risk of progression.⁹

Scoliosis with a high risk for rapid progression must be detected as early as possible. In a retrospective case series of 205 patients (163 girls and 42 boys) with idiopathic scoliosis at skeletal maturity, the surgical risk for a curve of 20° at the onset of puberty was at 16%. This surgical risk increased to 100% for curves ≥30° at the onset of puberty.¹⁹ The table summarises the risk factors for curve progression.

Scoliosis curve progression increases markedly at the time of the adolescent growth spurt in idiopathic curves and markedly slows or ceases at the time of completion of growth.²⁰⁻²² Spinal growth is closely associated with increase in height, but the measurement of height velocity at sequential visits is often associated with inaccuracies. Other maturity markers are therefore often used to measure the growth rate. The use of these maturity markers allows us to determine which curves are at risk of progression. This information allows the clinician to differentiate between curves that require careful regular monitoring and ones that require active treatment.

The total growth spurt has a duration of about 2.5-3.0 years,³ with the mean age for peak height velocity being about 14 years in boys and 12 years in girls.²³

Sexual maturity can be evaluated with the Tanner grading scale,²⁴ which is based on the extent of development of secondary sexual characteristics. It is important to ask about menarche because curve progression is less common after its onset.

Skeletal age is a more accurate marker of maturity. The Risser sign,²⁵ which refers to the appearance of the iliac apophysis of the pelvis, can be used to determine skeletal age. There are six Risser stages, from zero to five, denoting the course of the apophysis from the anterior to the posterior iliac spine, and then the fusion with the iliac bone (fig 3).²³ The incidence of progression of untreated AIS has been correlated with Risser sign and curve magnitude.²⁶ For curves of 20°-29° in a immature child with a Risser sign of 0 or 1, the incidence of progression was 68%. For curves <19° in a mature adolescent with a Risser sign of ≥2, the incidence of progression was 1.6%. For small curves <19° in an immature child (Risser sign 0 or 1), and larger

curves (20°-29°) in a mature child (Risser sign ≥2), the incidence of progression was about the same, at 22% and 23% respectively.²⁶ The disadvantages of the Risser sign are that it correlates with skeletal age differently in boys and girls and it typically appears after the peak height velocity.

Skeletal age can also be assessed by evaluating the development of the left hand and wrist on a radiograph: the bones are compared with those of a standard atlas compiled by Greulich and Pyle.²⁷ Sanders found that the scoring of the metacarpals and phalanges more closely related to scoliosis progression than other maturity indicators, including Tanner stage and Risser sign.²³ Dimeglio et al described elbow maturation as being more precise than hand maturation.²⁸

How is adolescent idiopathic scoliosis managed?

Observation for AIS is the most common approach used for patients with mild deformity (such as a Cobb angle measurement <25°). Depending on the degree of skeletal maturity, patients are assessed every four to six months at a specialist clinic to watch for curve progression. The interval of follow-up will be determined on an individual basis, based on the age of the patient, degree of curve, and skeletal maturity. Posteroanterior radiographs only are taken during each follow-up visit in order to minimise the exposure to radiation.

Bracing

Bracing in AIS is controversial, with treatment effectiveness remaining questionable based on available evidence, with most published studies being of low methodological quality. The rationale for the use of braces has been that external forces can guide the growth of the spine. Brace treatment is not necessarily benign in terms of the psychosocial and body image concerns it causes for many patients and their families. Bracing is used for the treatment of scoliosis in many centres in continental Europe, but practice is divided in the UK and US, and elsewhere.

Advocates of bracing quote level 2 evidence based information from prospective controlled studies²⁹⁻³¹ as well as other studies with level 3 and 4 information³²⁻³⁴ in support of bracing efficacy. In a meta-analysis a total of 1910 patients had non-operative treatment for idiopathic scoliosis, with 129 patients managed with observation only.³⁴ The analysis concluded that bracing was effective in altering the natural course of scoliosis. In 1995, a prospective, multicentre, non-randomised, non-blinded study also showed the effectiveness of bracing in girls with curves of 25°-35°.³⁰

Other studies have shown less positive results. A prospective case series of 102 immature patients with idiopathic scoliosis reported that bracing provided curve correction in only 15% of patients, while 42% later became surgical candidates.³⁵

The primary goal of bracing for scoliosis is to halt curve progression. The most widely accepted practice for brace treatment suggests that patients with curves of 25°-45° and in the most rapidly growing stage (Risser stage 0 or 1) should be offered a brace on initial evaluation. Curve progression is defined as an increase in the magnitude of the deformity by more than 5° at consecutive follow-up appointments of between four and six months.

Various factors can hinder successful brace treatment. Poor adherence is common. A meta-analysis reported that a protocol of 23 hours/day was more successful than protocols of 16 hours/day or night time use.³⁴ A multidisciplinary team approach involving the patient's general practitioner, surgeon, orthotist,

physiotherapist, and parents is needed to improve adherence. Families must be counselled that there is a risk that bracing may not be successful, but that the chances of success are improved with discipline and adherence to wearing the brace for the recommended time. Patients who have passed the peak height velocity, are within a year of skeletal maturity, or are a year or more after menarche are unlikely to benefit from use of a brace.

When should surgery be considered?

About 10% of adolescents with idiopathic scoliosis will progress to a level requiring consideration of surgery.³⁶ Surgery is generally indicated to treat a significant clinical deformity or to correct a scoliotic deformity that is likely to progress. Surgery is recommended in adolescents with a curve that has a Cobb angle greater than 45°–50°. This recommendation is derived from studies that have shown that curves >50° tend to progress slowly after maturity.¹¹ The decision to proceed with surgical correction therefore needs to take into consideration the clinical assessment, comorbid conditions, the wishes of the patient, and the effects the scoliosis has on the patient's quality of life. It is not clear that surgery is an effective treatment for back pain associated with scoliosis.

The aims of surgery may be to arrest curve progression by achieving a solid fusion, to correct the deformity, and to improve cosmetic appearance. If the decision is taken to operate, the usual approach in AIS is posterior (fig 1). In this approach a longitudinal posterior midline incision is used. Pedicle screws are inserted into the spine and two metal rods are measured and contoured. Curve correction is achieved as the two metal rods are attached and tightened on to the pedicle screws. An anterior fusion is used in AIS either as the sole approach in thoracolumbar or lumbar curves or in conjunction with posterior fusion in special cases.

Surgical treatment of AIS has a low rate of non-union and other complications. The incidence of neurological complications for spinal deformity surgery has been estimated by the Scoliosis Research Society at <1%.¹⁰ A more recent prospective clinical case series of 1301 patients reported a neurological complication rate of 0.69%.³⁷ A long term case-control study of scoliosis curves fused to the lumbar spine evaluated pain and functional status of AIS patients with a minimum of 10 years' follow-up (average 19 years).³⁸ These patients were compared with a control population matched for work, age, and recreational activities. The two groups did not differ with respect to functional status or pain.

After surgery it is important to check for abnormal neurology and for bowel and bladder symptoms. Back pain after surgery is not uncommon, especially if it is mechanical in nature. In the presence of continuous or night pain, infection or non-union should be considered, and referral to a specialist is advised.

Postoperative follow-up often involves clinical and radiological reviews at six weeks, three months, six months, and one year. These intervals will vary between institutions, but follow-up until completion of growth is common.

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Additional educational resources*Resources for healthcare professionals*

Scoliosis Research Society website. www.srs.org

AAOS American Academy of Orthopaedic Surgeons. Adolescent idiopathic scoliosis: etiology, anatomy, natural history, and bracing. *Instructional Course Lectures* 2005;54:529-36.

Resources for patients

Scoliosis Association United Kingdom (SAUK). www.sauk.org.uk—Provides patient information on the condition and treatments

Scoliosis Research Society. www.srs.org/patient_and_family—Patient and family section provides information on the condition, treatments, and outcome

Tips for non-specialists

Postural scoliosis can be differentiated from structural scoliosis with the Adam's forward bend test: the curvature will disappear on forward bending in postural scoliosis

If scoliosis is seen in a premenarchal female there is a higher risk of curve progression, and early referral to a specialist is advised

Patients undergoing brace treatment for scoliosis must be encouraged to adhere with brace treatment. Patients must be informed that the brace can be removed for washing and swimming

Table

Table 1| Risk factors for curve progression in adolescent idiopathic scoliosis

Risk factor	Comment
Age	The younger the age at diagnosis, the greater potential for curve progression at the onset of adolescent growth spurt
Sex	Progression is more common in girls
Menarche	Progression is least common after menarche
Remaining skeletal growth	More skeletally immature the greater risk of curve progression
Curve pattern	Double curves are more likely to progress than single curves
Curve magnitude	The risk of progression increases with curve magnitude

Figures

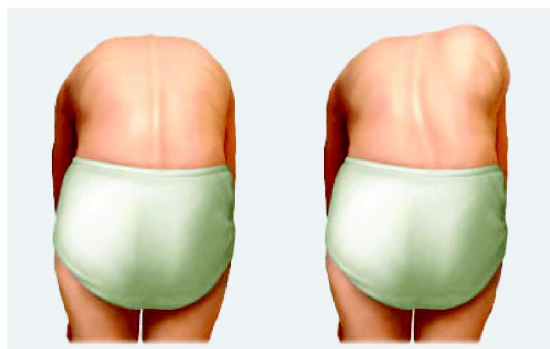


Fig 1 The Adam's forward bend test performed by (left) a patient without scoliosis, and (right) a patient with scoliosis showing a rib prominence

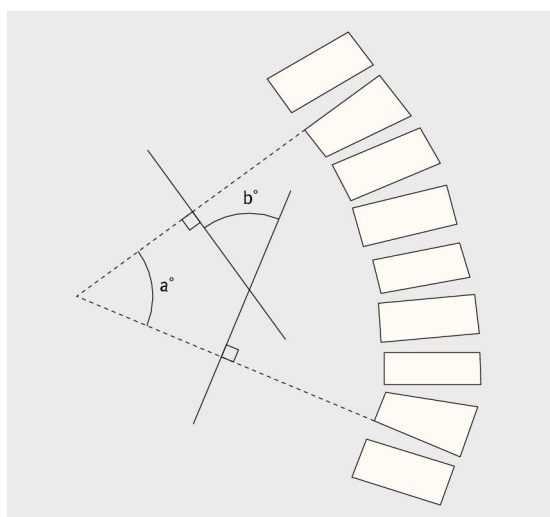


Fig 2 Cobb technique for determining size of a scoliosis curvature. On a posteroanterior view of the spine, tangents (dashed-dotted lines) are drawn along the superior endplate of the superior end vertebra and the inferior endplate of the inferior end vertebra. The angle formed (angle a) by the intersection of these two lines is the Cobb angle. This is more conveniently measured as the angle (b) formed by the intersection of two lines drawn perpendicular to the tangents. Adapted from Kim et al¹⁷

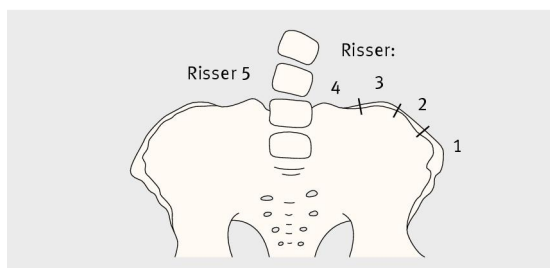


Fig 3 Illustration of the six Risser stages of skeletal age, from 0 to 5, denoting the course of the apophysis from the anterior to the posterior iliac spine, and then the fusion with the iliac bone

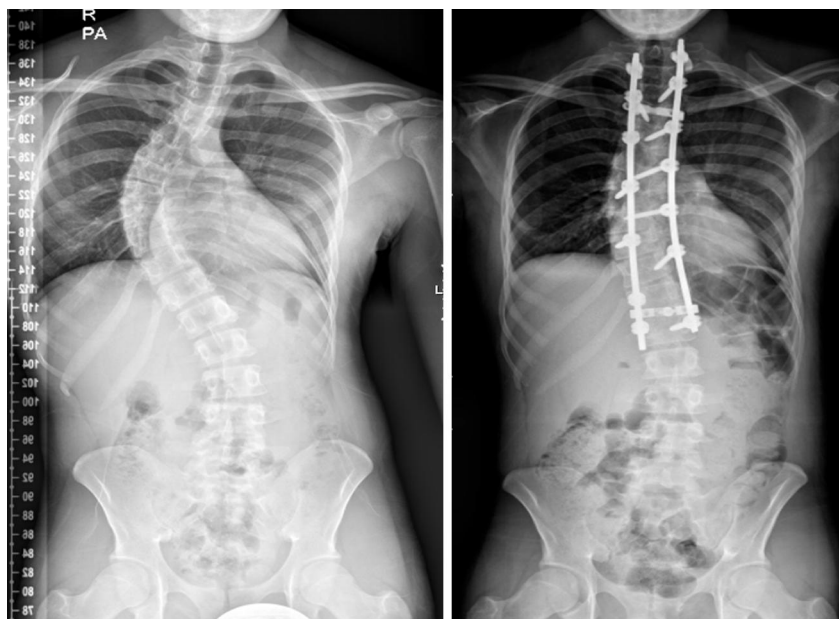


Fig 4 Preoperative (left) and postoperative (right) radiographs of an adolescent boy with idiopathic scoliosis, showing correction of the scoliosis by posterior instrumented fusion of the spine