THE ROLE OF THE FAMILY DOCTOR IN THE DETECTION AND MANAGEMENT OF ADOLESCENT IDIOPATHIC SCOLIOSIS

A health preventative program recommended by the Spine Society of Australia and endorsed by the Paediatrics & Child Health Division of the Royal Australasian College of Physicians

from

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Adolescent idiopathic scoliosis (AIS) is an important and relatively common spinal problem in the female. Early detection and treatment where indicated, result in better outcomes. This was the basis of school screening because in the early stages of curve development there are no symptoms only the signs of the deformity and these may go unnoticed. School screening, which is effective if properly conducted, has been largely abandoned in Australian government schools because of lack of funding for the programs and it is safe to assume that it will not be re-instituted. This has led to the development of The National Scoliosis Detection Program. Central to this program is the Scoliosis Detection Fact Sheet which explains in simple terms the outward signs of scoliosis. If after reading the Fact Sheet a girl, or her parents, thinks she has a curvature then follow up with the family doctor is recommended. Whenever an adolescent girl consults the family doctor, it is recommended that he/she carries out The Forward Bend Test. It will be 30 seconds well spent.

Each Spring all government and non-government schools in Australia are requested to download the Scoliosis Detection Fact Sheet from https://www.scoliosis-australia.org/ and to distribute it to girls in Years 5 and 7 (10 and 12 years of age in most and territories).

The aims of the program are to raise public awareness of scoliosis and to have adolescent girls accept a measure of responsibility for detection. Secondarily, to have family doctors manage minor curvatures (curves <20 degrees), and this is sensible. An Australian directory of spinal specialists who have a particular interest in spinal deformity is available on the website and all major children's hospitals in Australia have scoliosis clinic referrals.

Central to the success of the program will be the standardisation of curve measurement and the reporting on films by radiologists. To this end, an education program for radiologists has been developed. It is strongly recommended that general practitioners also read this paper to familiarise themselves with some of the simple technical details.

The National Scoliosis Detection Program is strongly recommended by the Spine Society of Australia. The program is endorsed by the Paediatrics & Child Health Division of the Royal Australasian College of Physicians. This paper reviews relevant aspects of Adolescent Idiopathic Scoliosis (AIS) for the family doctor, particularly physical diagnosis, and sets down the broad principles of management.

Aetiology

AIS arises spontaneously in otherwise healthy adolescents. The term idiopathic (Gk idios = own) is somewhat misleading. Family studies have shown that either multi-factorial or dominant inheritance are largely responsible. The highest incidence is in first degree female relatives (12 percent). The detection of a curve is an indication for regular examination of siblings in early adolescence. It is unusual for more than one child in a family to have a curve which requires treatment.

Sex Incidence

While the incidence of non-structural curvatures (< 10 degrees) is not too dissimilar in boys and girls, the ratio for curves requiring treatment (>20 degrees) is much higher in girls (1·8:10 respectively). Curve progression is usually associated with growth and tends to occur later in boys (13-17yo). No substantial case has ever been made for routine screening of boys.

The Scoliotic Deformity

AIS develops at or about 10 years of age in a previously normal spine. It deforms in coronal and sagittal planes, and has concomitant rotation in the long axis. The bodies rotate to the convex side of the curve, and the spinous processes to the concave aspect. This leads to the pathognomonic features of truncal rotation and asymmetry.

Typical AIS is flat from the side, with loss of the normal thoracic kyphosis being a feature of most thoracic curves. A kypho-scoliosis, with increased rounding in the sagittal or side plane, should alert the doctor to diagnosis other than AIS. Kyphosis is a red-flag.

Curve Patterns

A curve is described according to the direction of its convexity and the location of the most rotated vertebra (apex) as seen on a plain radiograph. The four most common curve patterns are:

Curve Patterns	Apex
Thoracic	T8-9
Thoracolumbar	T11-12
Lumbar	L1-2
Double major (right thoracic, left lumbar)	Variable

A right thoracic curve is by far the most common curve pattern, followed by a left lumbar one. Most idiopathic curves are based on six to eight vertebrae.

Prevalence

In most western societies the prevalence in the at-risk population (children 10 to 16 years of age) is three percent for curves 10° or more; for curves greater than 40° it is <0.1 percent.

School Screening

When properly conducted screening programs are carried out, these should be supported and participation encouraged. Screening is best performed in girls in Years 5 and 7. The Spine Society of Australia recommends that if screening is to be restricted on budgetary grounds, it should be limited to Year 7.

A two-tier screening process is recommended; the first being school nurses trained in the forward-bend test followed by confirmation of a structural scoliosis by a doctor. This should take place before a family is notified. In this way, over-diagnosis is avoided. The notification rate should be less than three percent (cf. Prevalence data).

The physical signs of AIS

The signs are readily seen when the trunk is viewed from behind with the subject standing erect (Figure 1).

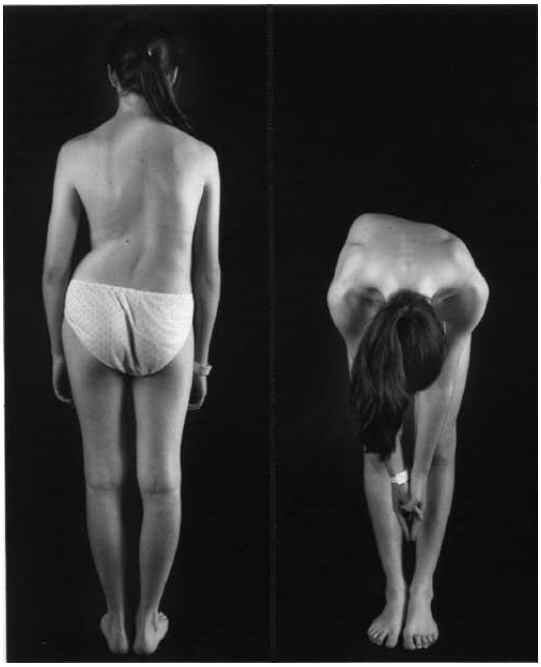

Figure 1. *Left.* An adolescent girl with a right thoracolumbar scoliosis. The right scapula is prominent as is the left hip and her trunk is decompensated to the right. The gaps between the dependent arms and the trunk are asymmetrical. The spine is obviously curved.

Right. Forward bend test of the girl on the left. The thoracolumbar curve is best viewed from in front of the patient and the angular rib deformity is clearly shown.

Palpation of tips of the spinous processes may be misleading because they are rotated towards the concavity of the curve, and can appear to be in a fairly straight line. In a typical right thoracic curve, the right shoulder will be elevated in comparison with the left. This effect may be negated if a proximal compensatory curve to the left has developed, as this balances the shoulders. As the ribs on the right rotate backwards, they are also elevated with pronounced rotation through the chest wall. Hence, the right scapula moves laterally with protraction of the whole shoulder girdle, and the medial scapular margin may have a sharper scapula prominence. The gap between the dependent arms of the trunk will be asymmetrical. If the right shoulder is elevated, the left arm will appear longer. Because of the vertebral rotation with the ribs following the spine, the left side of the anterior thoracic cage is more prominent than the right, with some minor costal margin asymmetry from the front. A girl may be aware of breast asymmetry.

Asymmetrical flank creases appear in moderate curves, with a bit of shift or imbalance, or because of curves in lower areas of the spine. (Figure 2). The left iliac crest (hip) will be more prominent than the right, especially if there is a lateral shift of the trunk (decompensation). A curve is decompensated (out of balance) if the head is not centred over the pelvis. Most thoracic curves are lordo-scoliotic, and best seen viewed from the side.

With thoracolumbar and lumbar curves the 'hip' prominence tends to be more marked, and may be the presenting complaint. The erector spinae on the convex side become

more prominent on forward flexion - the 'bolster' (elongated round pillow) sign. It is remarkable how much outward deformity and shift even moderate lumbar curves produce, especially if poor truncal control / imbalance is a feature..

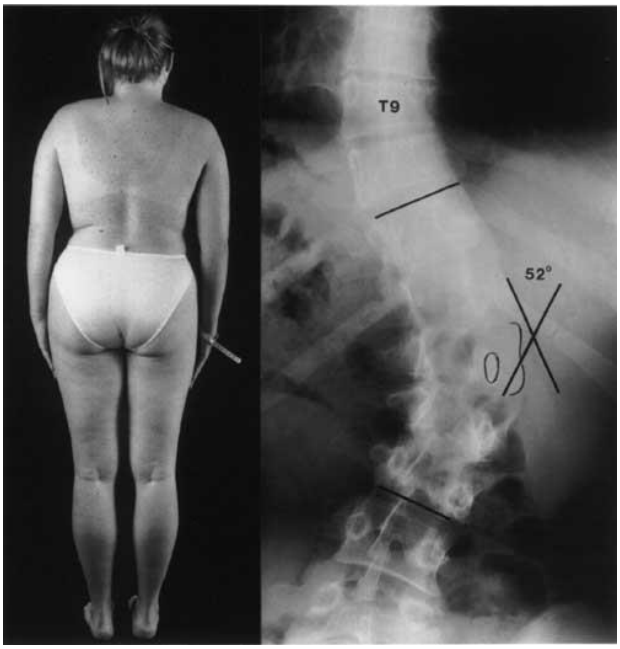

Figure 2. Clinical photograph and corresponding x-rays of a teenager with a 52° R lumbar curve. The apex is at L1. There is little outward deformity, but the flank creases are asymmetrical, and the left shoulder is high.

The Forward Bend Test

This is the pathognomic sign which indicates fixed, structural rotation in the vertebral column.

The subject stands with the feet parallel, and together, and bends forward to the level of the knees. The arms fall forward, palms touching each other, pointed between the great toes. Forward flexion brings the rib prominence (hump is an impolite term best avoided) or 'bolster' into clear profile (Figure 1).

Thoracic curves are best defined with the examiner standing behind the subject. Thoracolumbar and lumbar curves are best visualised with the examiner standing in front of the subject.

If a girl an adolescent has a positive forward-bend test, with the difference in the height of the two sides of the torso being less than 1cm, it is unlikely a significant scoliosis is present. If no other signs of a scoliosis are evident, then it is probable that simple torso asymmetry accounts for the false positive test.

Torso asymmetry as a manifestation of skeletal dimorphism

We all understand there is a difference in the size and shape of the two sides of the body, although the mechanism is not well-understood. This variation in normal growth and development may be evident as facial asymmetry or leg-length discrepancy. Torso asymmetry is the commonest reason for a false positive forward-bend test. It is found to a varying degree in up to 40% of normal schoolgirls and is of no clinical significance. The human eye is very sensitive at detecting asymmetry!

False positive tests brought school screening programs a measure of disrepute, causing parental concern and unnecessary x-rays. Torso asymmetry usually affects the whole of the side of the trunk, in contrast to a structural scoliosis. Minor degrees of asymmetry are readily appreciated by gently running the hand from side to side across the mid-line. When torso asymmetry is present, other features of skeletal dimorphism are usually in evidence. If a doctor is having difficulty in interpreting the forward-bend test, then it is almost certain that an adolescent does not have a curve warranting referral.

Torso asymmetry is most often found in the right thoracic region. The relationship between torso asymmetry and AIS is being investigated in longitudinal studies. The link between the two may be on a genetic basis. It is common for a girl with AIS to have a mother with right torso asymmetry.

X-ray examination

If a structural curve is diagnosed, a baseline AP radiograph of the **erect** thoracolumbar spine should be taken to include the iliac crests (figure 3). In nine out of ten adolescent girls the thoracolumbar spine can be fully visualised on a 43 x 35 cm film. The 91 cm long films should be avoided wherever possible, to reduce exposure of the thyroid gland and the gonads.

Most states have access to modern imaging with a whole spine EOS film available. This allows assessment of the spine without errors in digital stitching and is also ultra-low dose imaging. If an adolescent requires regular assessment of the spine, this is best done with an EOS to minimise the cumulative radiation dose.

http://surgicom.com.au/Surgicom/EOS_Locations.html

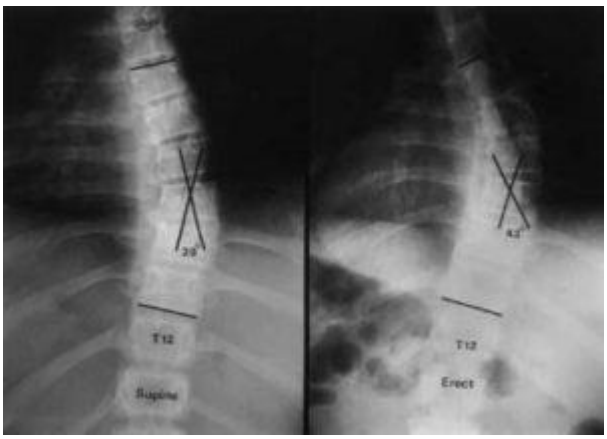

Figure 3. Supine (left) and erect (right) radiographs of an adolescent girl with a typical right thoracic curvature. Curve measurements increase by 15 degrees from the supine to the erect position.

Curve Measurement and the Risser sign

Coronal plane measurements are done on plain radiographs of the **erect** thoracolumbar spine and include the iliac crests. There is a significant difference in curve measurement between supine and erect films. For patients who can stand and walk, the preference is to access the spine standing, as the deformity manifests with gravity.

The preferred method is the Cobb technique (Figure 4). The Cobb technique measures the curve in two planes, whereas the deformity occurs in three planes. Nevertheless, it is a reliable method to monitor progress.

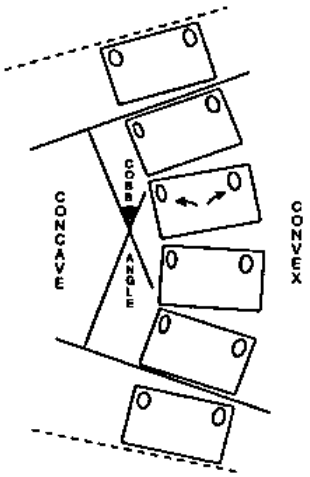

Figure 4. *The Cobb angle.* This is measured at the intersection of perpendiculars drawn to lines parallel to the upper and lower surfaces of the most tilted vertebrae (solid lines). Measurement using the vertebra above and below (dotted lines) would give a false value as these are not as tilted. With a structural scoliosis the vertebra rotates, and the pedicle shadows become asymmetrical / move away from the lateral border of the vertebra (arrows).

The Scoliosis Research Society has set down the criteria for the radiological diagnosis of AIS. **These are 10° as measured by the Cobb technique on a standing upright spinal radiograph (with presence of vertebral rotation).**

Of note a significant structural scoliosis is one that measures more than 20 degrees, with growth remaining. These require referral.

The Risser sign relates to the appearance, medial excursion and fusion of the secondary centres of ossification of the iliac crests. (Figure 5).These appear laterally and, on average, it is two years before they fuse but there is considerable variability in these events. Often there is asymmetry in the degree of excursion and the more immature side should be taken for grading purposes. More than 50 percent of curves will have stopped progressing by the time the crests are half-capped.

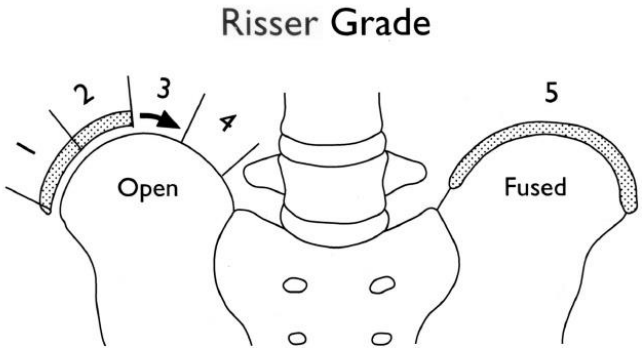

Figure 5. *The Risser Sign*. The degree of medial excursion is graded I-IV. Skeletal maturity occurs when the apophysis fuses (Grade V).

The Risser sign was used as a marker to assess skeletal maturity / or bone age, as it can be seen on the imaging of the spine. The high variability and poor prognostication mean that sometimes a left-hand AP is taken for assessment of bone age. This is often directed by the surgeon looking after a patient, as knowledge about how much growth is remaining helps with prognosis.

Symptoms

Most adolescents with AIS have no symptoms at presentation. However, non-specific aching discomfort in the spine is not infrequent when curves progress rapidly. Chronic lumbar backache may also occur with thoracolumbar and lumbar curves in late adolescence. It tends to be associated with long hours of study, being overweight, and decreased general fitness. Such backache responds well to appropriate alterations in lifestyle. On the other hand, a painful scoliosis is a clear indication to fully investigate the patient for possible underlying pathology e.g. a spinal tumour. Spinal stiffness is a feature of spinal tumours generally, be they benign or malignant.

Scoliosis of other aetiology

Scoliosis is a physical sign and no more than that. In essence, a diagnosis of AIS is one of exclusion. The detection of a curve is an indication for a full physical examination, particularly a neuromuscular assessment.

Scoliosis may be a manifestation of neuromuscular disorder (syringomyelia, Arnold-Chiari malformation, Charcot-Marie-Tooth disease, Freidreich's ataxia, cerebral palsy, myopathy, etc.); connective tissue disorders (Marfan's syndrome, neurofibromatosis Type 1, etc.); dysmorphic states and various syndromes of a widely diverse nature. The list of conditions in which scoliosis may be present is a long one indeed, but usually the diagnosis of the underlying disorder is evident on clinical grounds.

Congenital scoliosis based on congenital vertebral abnormalities usually comes to attention early in life. Such curves tend to be rigid on side-bending. All patients with congenital scoliosis should have a renal ultrasound to exclude a significant congenital abnormality in the renal tracts.and a whole spine MRI at some point.[2]

True and apparent leg lengths should be carefully measured in all patients with AIS, especially if a curve is decompensated. Spinal balance may be improved by levelling the pelvis with a raise to the heel on the short side (true or apparent). To see if this is so, the doctor sits behind the patient and places blocks/books under the short side, to balance the pelvis. Another alternative is to assess the patient seated, to see if the apparent scoliosis disappears or the imbalance improves. Removable heel inserts are far more acceptable to most teenagers than an addition to the shoe heel.

Management of AIS

Because no treatment will return the scoliotic spine to normal, management is a more appropriate term to use. The only treatments which have been shown to be effective are bracing and surgery. The natural history of AIS has been well described, and this is the basis for current medical advice.

The following are generally accepted guidelines which are strongly influenced by the age, the degree of curve at presentation and the remaining growth potential.

- <20°. Observations through the rapid growth phase on a six-monthly basis, or earlier review if parents think there has been a deterioration.

- 20° - 40°. Bracing may be indicated if radiographic evidence of progression of more than 5° , or if there is 12 months or more of growth remaining. One third of curves in this range do not progress significantly either in adolescence, or in later life.

- >45°. Surgery may be indicated.

The risk of curve progression in AIS during adolescence is known (Table 1). It is important this be made known to the patient and parents at the initial visit. Curves progress most rapidly during peak velocity growth (10 - 12 years) for girls.

The onset of the growth spurt is heralded by early breast development. There is evidence that girls with AIS grow more rapidly and earlier than do those without scoliosis, but their adult heights are closely similar. A common pitfall is to relate growth to onset of menarche (average age 13.5 years). Chronological age and age at menarche are unreliable indicators of spinal growth potential. Skeletal age as determined by a left-hand radiograph is far more dependable. (Sanders score NOT Greulich and Pyle assessment).

Table 1. Probabilities of Progression
Magnitude of curve at initial detection versus age

Curve magnitude at detection (degrees)	Age (years) 10-12	13-15	16
<19	25%	10%	0%
20-29	60%	40%	10%
30-59	90%	70%	30%
>60	100%	90%	70%

(Nachemson, A; Lonstein, J; and Weinstein S: Report of the SRS Prevalence and Natural History Committee 1982. SRS Annual Meeting, Denver, Colorado, September 1982).

Vertical growth all but ceases at about 15 years of age in girls. The growth curve flattens at this time, and a girl on the 50th centile will increase her height by only 2cm between 15 and 18 years of age.

Risk factors for progression include:

- Age and degree of curve at presentation.

- Risser grade at presentation. The lower the grade, the higher the probability of progression. When the grade is 0 - 1, the risk for a curve <20° is 22%, whereas it is 68% if the curve is in the 20° - 29° range. Conversely, in the same curve ranges when the Risser grade is 2 - 4, the risks are 1.6% and 23% respectively.

- Double major curves tend to have a worse prognosis.

A long-term follow-up study has shown that approximately two-thirds of curves followed for more than 40 years, progressed after maturity. This was most marked in curves in the 50° - 75° range, and took place at about 0.75° to 1° per year.

Curves greater than 30° should be followed until the patient is 18 years of age, or two years after they have stopped growing. Larger curves should be followed on a regular basis into the 20s, particularly if there is trunk decompensation in thoracolumbar and lumbar curves.

Advice to Family

It is most important that the family doctor allay anxiety when a scoliosis is detected. Not unexpectedly, the patient and the parents fear the worst. The following are the questions most frequently asked by parents:

Is the curvature related to the carrying of a heavy schoolbag? - ***Categorically No.***

Is scoliosis related to poor posture? - ***Definitely not.***

> *The adolescent slouch is a kyphotic posture which can be actively or passively corrected. This will disappear on lying prone – face down.*

Will physiotherapy or chiropractic treatment correct the curve? - ***No***

> *No evidence suggests that exercise alone will prevent progression of significant structural curves (>20 degrees). Exercise programs are an adjunct.*

Will electrical stimulation therapy help? - ***No.***

> *Studies show that this putative treatment has been completely discredited.*

Will the presence of scoliosis affect pregnancy? - ***Variable.***

> *This will occur **only** if curves are severe*. (>65°).
>
> *Or if the patient is fused below the level of L3.*
>
> *Back pain however, is increased in pregnancy, especially the third trimester.*

Should any restrictions be placed upon activities? ***No.***

> *Teenagers should be encouraged to keep fit and trim through regular sports, and there are no specific restrictions nor recommendations.*
>
> *If a patient is wearing a brace, ideally they would balance their hours by not wearing their brace during sports.*
>
> *Contact body sports specifically: do not wear the brace because of the danger of opponent injuring themselves against the brace.*

What are the results of treatment? ***Excellent.***

In general, the results of modern- day treatment of scoliosis are excellent.

It must be emphasised that brace treatment is aimed at stopping curve progression through the growing years, and bracing after growth is NOT indicated.

Most adolescents and adults with AIS live a normal life.

The prevalence of significant back pain in an adult with AIS is equivalent to that of the rest of the population.

Specialist Referral

There is no good reason why a patient with AIS whose curve is less than 20° should be immediately referred to a spinal surgeon. Such curves can be safely managed by the family doctor along the lines laid down in this paper.

If the curve progresses beyond 20 degrees, there is significant growth remaining, or bracing is warranted, then early referral to a paediatric spinal surgeon or scoliosis service is indicated.

There are scoliosis clinics in all children's hospitals in Australia and in some adult teaching hospitals. Accurate curve measurement on plain radiographs is essential for these decisions and an educational program for radiologists has been developed.

Scoliosis of aetiology other than AIS should be referred for specialist assessment because the natural history of such curves varies greatly, and curve management needs to be integrated with that of the underlying disorder. The management of congenital scoliosis is a complex matter and early specialist opinion should be sought.

If scoliosis is present in a child under the age of 10yo, early referral is indicated as this is NOT AIS.

Brace Management

A detailed discussion of orthotic treatment is beyond the scope of this paper. The aim of brace treatment is to control the curve through the rapid growth phase.[1] There is no evidence that patients with curves <30° are in any way disadvantaged in later life, or that they are more subject to disabling back pain.

A brace program is a demanding exercise and depends strongly upon patient compliance and family support. Nowadays, most curves are managed in high-density polyethylene under-arm orthoses. Prefabricated modular braces are also available. These are well-disguised in conventional clothing. A brace is worn 20 hours per day.

The Milwaukee brace, with its conspicuous vertical struts and obvious neck ring, and which the adolescent finds so objectionable, is rarely used these days.

There are many factors to consider when weaning a patient from a brace and this decision is very much an individual one. A widely accepted endpoint is full medial excursion of the iliac apophyses (Risser grade IV), or Sanders 7 on hand bone age. This is contemporaneous with the cessation of vertebral growth. On average, bracing is discontinued some time in the 14th yearf or girls, or 16-17th year for boys. Under optimum conditions it is successful in controlling 80% of curves so managed.

Surgical treatment

From the 1960s to the 80s, Harrington instrumentation and spinal fusion were the standard methods of surgical management. This involved two systems: distraction (the principal corrective force) on the concave side, and compression on the convex side of the curve. This technique was effective and permitted early mobilisation, but with external support such as the plaster of Paris body jacket. The pseudarthrosis rate (failure of bony fusion) was high, requiring revision surgery for bone grafting. However, the distraction moment does not affect rotation and so the rib hump of thoracic curves is not reduced.

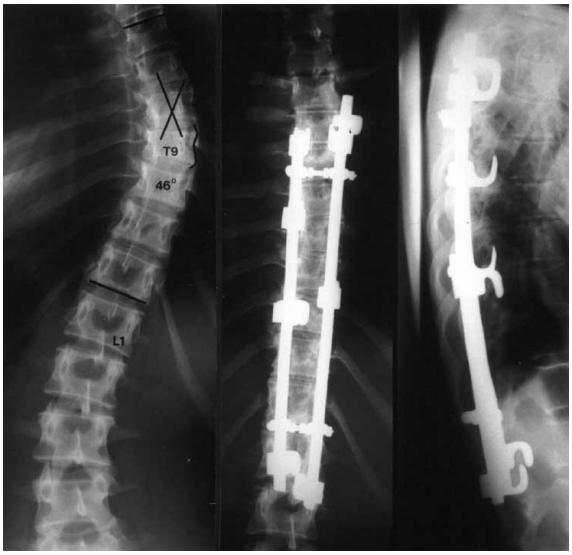

Figure 6. Angular posterior radiograph of a typical right 46° AIS in an adolescent girl. There is trunk decompensation to the right. Good curve correction has been obtained with Cotrel-Dubousset instrumentation. Rib symmetry has been restored together with a thoracic kyphosis.

In the late1980s, the system developed by two French surgeons, Cotrel and Dubousset, largely replaced Harrington instrumentation. The method corrects a curve by a derotation manoeuvre with concomitant coronal translation (lateral movement) of the apical vertebrae (Figure 6). It provides a very effective fixation in the corrected position. No post-operative casting or other protection, is required. Further, the rigidity of the system strongly favours graft healing.

Now there are many instrumentation systems which embody the same biomechanical principles. Although some surgeons, particularly in Europe, use this system for most curve patterns, it is fair to say that it is best suited to the correction of thoracic curves.

Thoracolumbar and lumbar curves are best suited to anterior correction where the intervertebral discs are completely excised and internal fixation is achieved by linked trans-vertebral screws. The first system was introduced by the Australian surgeon, the late Alan Dwyer, who used a cable threaded through the screw heads, rather than a rod. When the cable was tensioned the vertebrae came together like cotton reels on a string. A refinement of this technique entails a rod which can rotate in a screw assembly, thereby producing very effective de-rotation of the vertebral column (Figure 7). Anterior correction allows fewer vertebrae to be fused than when posterior surgery is carried out. There are several commercially available systems which entail this principle.

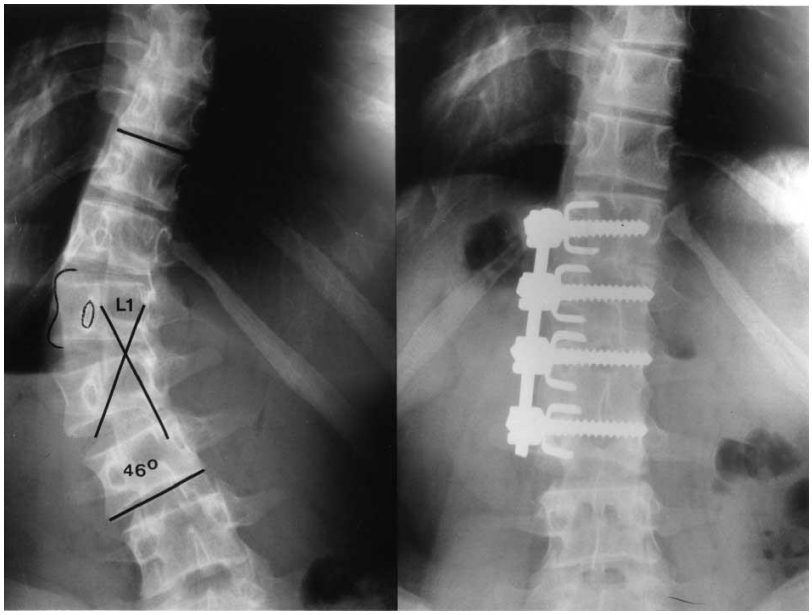

Figure 7. Radiographs showing the correction of a 46° decompensated lumbar idiopathic curvature by the anterior route. Here, derotation has effectively brought the spine back into near normal alignment with equalisation of the forces acting on the remaining mobile lumbar discs.

A number of other advances have made major spinal surgery for scoliosis very safe. There have been vast improvements in anaesthetics in general, and in intensive care supervision when required. Cell-saver technology is important and allows the adolescent patient's own blood to be cleaned, filtered and given back during surgery. Spinal cord monitoring for posterior spinal surgery, with simultaneous motor and sensory-evoked potentials, is now the standard of care. If needed, the use of the reliable "wake-up" test during surgery is performed. Post-operative pain control with either a continuous narcotic infusion or patient-controlled analgesia is offered with confidence for effective pain management in the immediate post-operative phase. Most adolescent patients now leave hospital by 5-7 days after surgery, with early and quicker mobilisation and oral intake key components in this rapid recovery.

This is a far cry from the 1950s when spinal fusion was carried out through a window in a plaster jacket without the benefit of instrumentation and the pseudarthrosis rate was then in the order of 70 percent. In those days, patients were kept recumbent for at least six months post-operation. Witness the extraordinary progress that has been made.

Following successful spinal surgery today, patients return to a full and active life, returning to most team sports at 6 months postoperatively, with some restrictions until the 12 month mark. Usually, the main restriction placed on their activities is avoidance of body contact/collision sports until the spine is fully fused. Patients are advised against occupations entailing heavy physical or labouring work, and activities which are normally carried out in cramped and confined quarters.

Summary

For the foreseeable future, the family doctor will continue to play a central role in the detection of scoliosis and in advising the family accordingly, Further, public awareness needs to be heightened. The program outlined herein will help shift responsibility back into the community. The public hospital system in this country is

faltering and preventative health programs need to be adjusted to meet these changes. These shifts in responsibility should be viewed in a positive light.

Over-diagnosis and unnecessary treatment of AIS must be avoided at all costs. There are tacit responsibilities and obligations for the medical profession.

Examination of the spine and the simple forward-bend test should be part of routine physical examination of the adolescent. **It will be 30 seconds well spent**.

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Links

Scoliosis Research Society BrAIST trial:

https://www.srs.org/UserFiles/file/SRSDolanBrAISTinitiative.pdf