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. It has long been recognised that 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 Self-Detection Program for Scoliosis. Central to this program is the self-detection brochure which explains in simple terms the outward signs of scoliosis. If after reading the brochure a girl, or her parents, thinks she has a curvature then follow up with the family doctor is recommended. Further, whenever an adolescent girl consults the family doctor for whatever reason, it is recommended that he/she carries out The Forward Bend Test as detailed herein. It will be 30 seconds well spent.

In July/August each year all government and non-government schools in Australia will be requested to download the self-detection brochure from http://www.scoliosis-australia.org/ and to distribute it to girls in Years 6 and 8 (10 and 12 years of age in most and territories). The internet makes this a practical approach and Year 8 girls can be readily included.

The aims of the program are first, to raise public awareness of scoliosis and to have adolescent girls accept a measure of responsibility for detection. Second, to have family doctors manage minor curvatures and this is sensible. An Australian directory of spinal specialists who have a particular interest in spinal deformity is available.

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 the simple technical details.

The National Self-Detection Program for Scoliosis 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 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 examination of siblings in early adolescence. It is quite rare for more than one child in a family to have a curve which requires treatment.

Sex Incidence

While the incidence of minor curvatures (< 10 degrees) is not too dissimilar in boys and girls, the ratio is 1·8:10 respectively for curves requiring treatment. Curve progression tends to occur much later in boys. 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 the coronal and sagittal planes, with concomitant rotation in the long axis. The bodies rotate to the convex side of the curve, and the spinous processes to the concave aspect.

Loss of the normal thoracic kyphosis is a feature of most thoracic curves. A true thoracic kypho-scoliosis should alert the doctor to diagnostic possibilities other than AIS.

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 two to three percent for curves 10° or more; for curves greater than 40° it is <0.1 percent.

School Screening

Where properly conducted screening programs are carried out these should be supported and participation encouraged. Screening is best performed in girls in Years 6 and 8. The Spine Society of Australia recommends that if screening is to be restricted on budgetary grounds, it should be limited to Year 6.

A two-tier screening process is recommended, the first being school nurses trained in the forward-bend test. Confirmation of a structural scoliosis by a doctor 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).

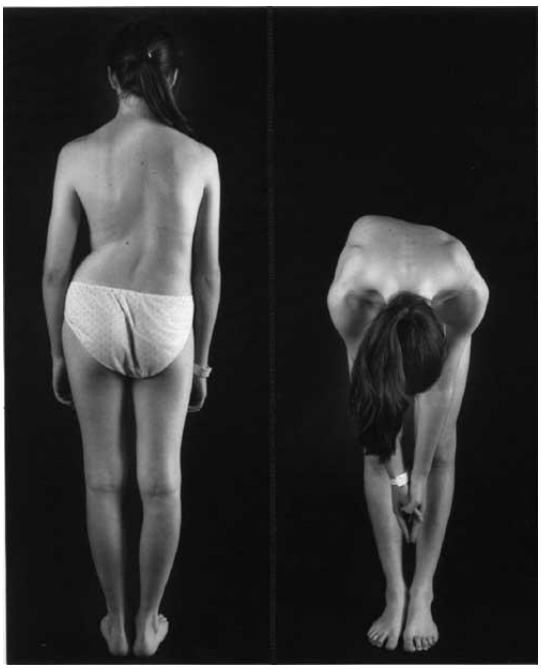

Figure 1. *Left*. An adolescent girl with a right thoracolumbar scoliosis. The right scapula is prominent as is the left hip and her trunk is decompensated to the right. The gaps between the dependent arms and the trunk are asymmetrical. The spine is obviously curved.

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, though this effect may be negated if a proximal compensatory curve to the left has developed. As the ribs on the right rotate backwards, they are also elevated. The right scapula moves laterally and its medial margin may have a sharper profile than its counterpart. The gap between the dependent arms of the trunk will be asymmetrical. If the right shoulder is elevated, the left arm will appear longer.

Asymmetrical flank creases appear in moderate curves and are more marked in an overweight subject (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. 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, though rarely is this very marked. A girl may be aware of breast asymmetry.

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. By-and-large, the more proximal the apex of a curve is in the spine, the greater is the

cosmetic blemish. It is remarkable how little outward deformity even major lumbar curves may produce, especially in overweight girls.

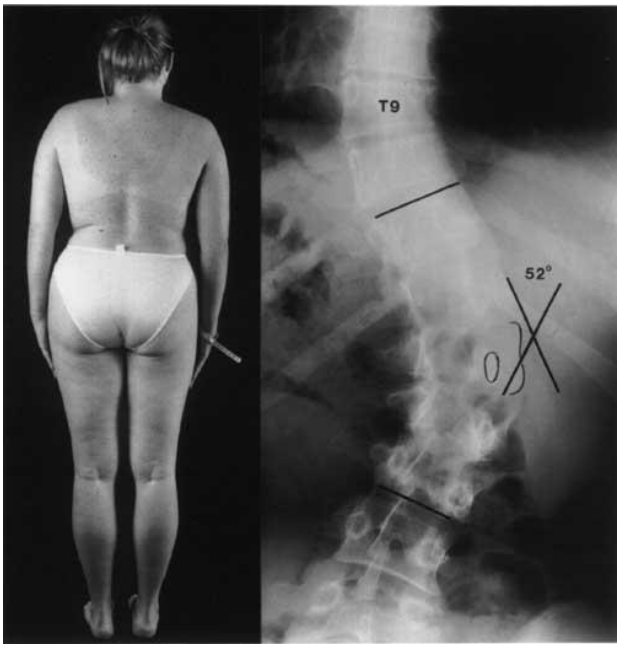

Figure 2. Clinical photograph and corresponding x-rays of an overweight teenager with a 52° R lumbar curve. The apex is at L1. There is little outward deformity but the flank creases are asymmetrical.

The Forward Bend Test

This is the key physical sign and indicates fixed, structural rotation in the vertebral column. The subject stands with the feet parallel, and together, and bends forward as far as he or she can, with outstretched hands, palms facing each other, pointed between the great toes. Forward flexion brings the rib prominence (rib hump is a term best avoided in front of a teenager) 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. A lumber scoliosis based on a leg-length discrepancy is more correctly termed a lumbar tilt, and disappears when the subject is seated. It also disappears in forward flexion of the trunk.

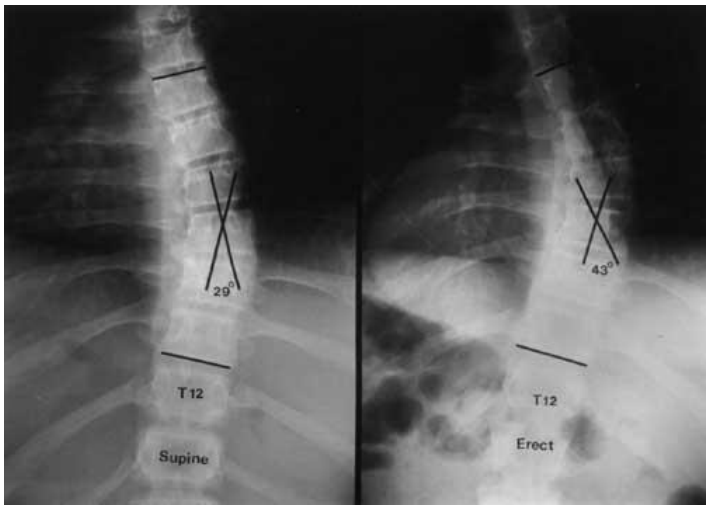

Figure 3. Supine (left) and erect (right) radiographs of an adolescent girl with a typical right thoracic curvature. Curve measurements increase by 14° from the supine to the erect position.

If a girl has a positive forward-bend test with the difference in the height of the two sides of the torso less than 1cm, a significant scoliosis (>20°) is highly unlikely to be 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

Here there is a difference in the size and shape of the two sides of the body. It is not a well-understood variation in normal growth and development. It may be evident as facial asymmetry or leg-length discrepancy. Torso asymmetry on this basis 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 a schoolgirl 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. No other views are needed for the average case at the initial assessment.

Curve Measurement and the Risser sign

Coronal plane measurements are done on plain radiographs of the **erect** thoracolumbar spine and include the iliac crests. The preferred method is the Cobb technique (Figure 4). There may be a difference in curve measurement of up to 10° between supine and erect films. 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.

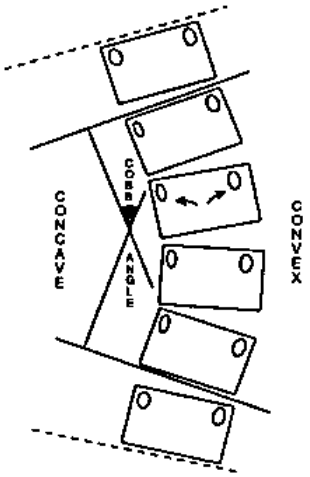

Figure 4. *The Cobb angle.* This is measured at the intersection of perpendiculars drawn to lines parallel to the upper and lower surfaces of the vertebrae which tilt towards the concavity (solid lines). Measurement using the vertebra above and below (dotted lines) would give a false value as these surfaces do not tilt towards the concavity. When a vertebra rotates the pedicle shadows become asymmetrical (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.**

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.

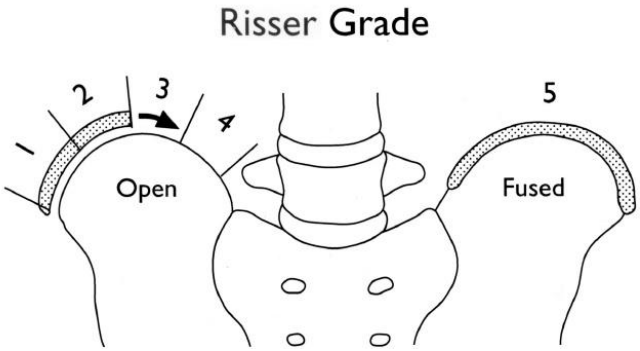

Figure 5. *The Risser Sign*. The degree of medial excursion is graded I-IV. Skeletal maturity occurs when the apophysis fuses (Grade V).

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. Then 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.[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 asks that they come up on their toes on the short side, with the knee held straight. Removable heel inserts are far more acceptable to most teenagers than an addition to the shoe heel, and trainers are difficult to alter in this way.

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. A great deal of new information has come forward in the past decade on the natural history of AIS, 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 by radiographic evidence of progression of more than 5° is a pre-requisite. Many centres have narrowed the indication for bracing to curves in the 30° - 40° range. About one third of curves in this range do not progress either in adolescence or in later life.
- >40° - 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). The onset of the growth spurt is heralded by early breast development. There is evidence that girls with AIS grown more rapidly and earlier than do those without scoliosis, but their adult heights are closely similar. A common pitfall is to relate growth to 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 hand and wrist radiograph is far more dependable.

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 at least 18 years of age. 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.*

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

> *No evidence suggests that they will in any way influence the natural history of scoliosis. Exercise programs are not recommended.*

Will electrical stimulation therapy help? - ***No.*** *Studies show that this putative treatment has been completely discredited.*

Will the presence of scoliosis affect pregnancy? - ***No.*** *This will occur* ***only*** *if curves are severe.* (>65°).

Should any restrictions be placed upon activities? *For curvatures less than 40° the answer is* ***No.***

> *Teenagers should be encouraged to keep fit and trim through regular sport. If a patient is wearing a brace, contact body sports are to be avoided, principally because of the danger of opponents injuring themselves against the brace*

What are the results of treatment? - *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.*

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. Progression beyond 20° is an indication for a specialist opinion. 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.

Brace Management

A detailed discussion of orthotic treatment is beyond the scope of this paper. However, 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 which are

commercially available are used. These are well-disguised in conventional clothing. A brace is worn 23 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 end-point is full medial excursion of the iliac apophyses (Risser grade IV). This is contemporaneous with, but unrelated to, the cessation of vertebral growth. On average, bracing is discontinued some time in the 16th year. Under optimum conditions it is successful in controlling 80% of curves so managed.

Surgical treatment

From the 60s 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 fusion) was less than five percent. However, the distraction moment does not affect rotation and so the rib hump of thoracic curves is not reduced.

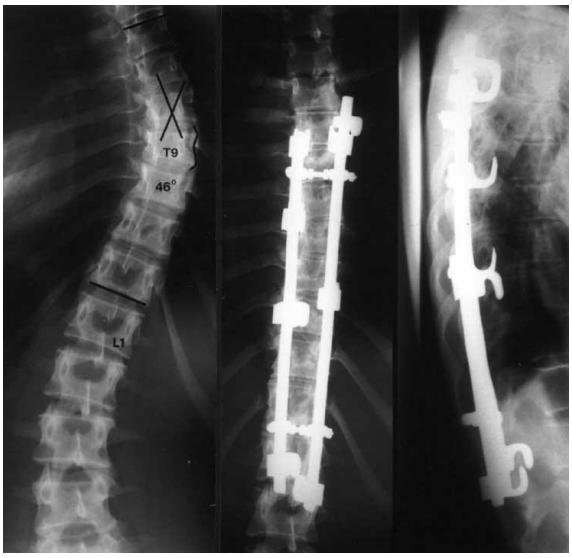

Figure 6. Angular posterior radiograph of a typical right 46° AIS in an adolescent girl. There is trunk decompensation to the right. Good curve correction has been obtained with Cotrel-Dubousset instrumentation. Rib symmetry has been restored together with a thoracic kyphosis.

In the late 80s, 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 look-alike 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 a number of commercially available systems which entail this principle.

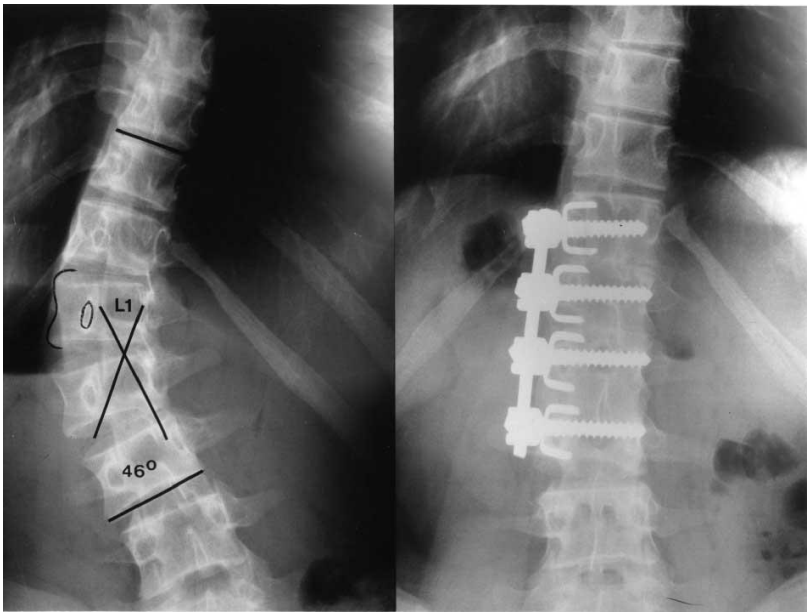

Figure 7. Radiographs showing the correction of a 46° decompensated lumbar idiopathic curvature by the anterior route. Here, derotation has effectively brought the spine back into near normal alignment with equalisation of the forces acting on the remaining mobile lumbar discs.

There are a number of other advances which have made major spinal surgery of this type very safe. There have been vast improvements both in anaesthetics in general, and in intensive care supervision when required. Autologous blood transfusion and cell-saver technology are important for the adolescent female. Spinal cord monitoring for posterior spinal surgery, with simultaneous motor and sensory-evoked potentials, is now routine in some centres. Others use the reliable "wake-up" test. Post-operative pain control with either a continuous narcotic infusion or patient-controlled analgesia can now be offered with confidence for effective pain management in the immediate post-operative phase. Most adolescent patients now leave hospital at about one week post-surgery. This is a far cry from the 50s 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, at or about 8 to 12 months post-operation. Usually, the only restriction placed on their activities is avoidance of body contact/collision sports. Patients are advised against

occupations entailing heavy physical work, and activities which are normally carried out in cramped and confined quarters. Repetitive bending and twisting movements should be avoided.

Summary

It is clear that in the immediate future the family doctor will have to play a more 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. Remember - only three per 1000 adolescent girls have a curve which requires active treatment.

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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