SCOLIOSIS: A REVIEW

School screening of adolescents reveals a high prevalence of mild rotational deformity. The objective of screening is to prevent serious deformity by regular review of these children, early recognition of progression and provision of spinal bracing for curves greater than 25°. Two per cent of students screened in the age range 11-13 years have curves greater than 10° but only two per thousand screened require active treatment. A programme of exercise combined with bracing until skeletal maturity is reached, obviates the need for major surgery. Indications for follow-up and treatment are reviewed, and some current concepts of the aetiology of idiopathic scoliosis are examined with particular emphasis on the relationship between scoliosis and growth.

JAMES R TAYLOR

James Taylor M B , Ch B , Dip Trop Med , Ph D , is Senior Lecturer in Medical Anatomy and Human Biology at the University of Western Australia

COLLEEN B. LISTON

Colleen Liston, A U A , is a part-time lecturer at the Western Australian Institute of Technology and part-time Co-ordinator of the Spinal Deformities Clinic, and Posture and Seating Clinic at the Princess Margaret Hospital for Children

LANCE T TWOMEY

Lance Twomey B Sc. (Hons), B App Sc , Ph D , is Head of the School of Physiotherapy at the Western Australian Institute of Technology and National President of the Australian Physiotherapy Association

Over the last two decades, appreciation of the prevalence of scoliosis has increased dramatically as a result of numerous screening programmes in many countries The effectiveness of prevention and treatment of the deformity have also greatly improved. Despite some advance in our understanding of the familial or genetic nature of idiopathic scoliosis, its aetiology remains obscure (Nachemson and Sahlstrand 1977).

Definition and Classification

Scoliosis is a lateral curvature of the spine. When confronted with a case of scoliosis there are three basic questions which should be answered to adequately define it:

1. Is it (a) structural, or (b) non-structural (postural)?
2. If it is structural, is the cause known or unknown?
3. (a) What region or regions of the spine does it involve,
   (b) what is the side (direction) of the convexity of the curve(s), and
   (c) what is the angle of curvature?

1. (a) **Structural** scoliosis is a lateral curvature which does not correct completely on lateral bending. It is almost always accompanied by a *rotational deformity* and in practice, this rotational deformity causes a 'rib hump' or 'lumbar hump' which can be readily recognised in screening programmes In structural scoliosis there is an asymmetry in the vertebrae, muscles or ligaments of the spine itself.

(b) **Postural or non-structural scoliosis** is a lateral curvature which can fully correct on lateral bending, which is *not accompanied by a fixed rotational deformity* of the spine, and which is either inconstant or is due to some asymmetry of the supporting appendicular skeleton, *eg* leg length inequality.

2. **Types of structural scoliosis**

In the vase majority of scoliotic curvatures, the cause is unknown.

Idiopathic scoliosis is a familial condition but its pathogenesis remains unexplained Idiopathic scoliosis is seen and described in three age groups. infants, juveniles and adolescents. The adolescent form is the most common, worldwide, and the infantile form, though not uncommon in Europe, is rare in North America and Australia (James 1976)

The small minority of scoliotic curvatures with a known 'cause' include **congenital scoliosis** when a congenital vertebral anomaly such as a hemivertebra causes a short sharply angled curve; 'paralytic' scoliosis *eg* following spina bifida or muscular dystrophy where paralysis of vertebral musculature leads to 'collapse' of the column into scoliosis, a few rare syndromes such as Marfan's syndrome, where abnormal laxity of supporting connective tissues contributes to structural scoliosis (Winter 1975).

3. **Angle and Position of Curve**

When a structural curvature first appears, the vertebral column both curves and rotates to the same side. The curvature is labelled right or left according to the side of its convexity This is most commonly to the right side in the thoracic region (Moe and Kettleson 1970) and is called the primary curve. To maintain normal body alignment a compensatory

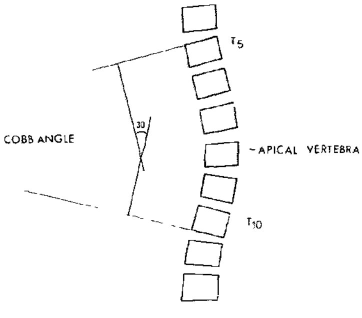

Figure 1: Cobb method of measurement

(or secondary) curve is formed in the opposite direction, usually in the lumbar region If in both curves the vertebral bodies are rotated in the same direction as the lateral curvatures, both curves may be regarded as structural curvatures developing at the same time and be classified as double primary (or double major) curves.

The lateral curvature is usually measured by the Cobb method (Figure 1). This is the angle between the upper surface of the vertebral body at the top of the curve and the lower surface of the vertebral body at the bottom end of the curve. The apical vertebra of the curve is that vertebra which is most rotated and deviated from the median plane. A curvature is referred to as thoracic or lumbar according to the position of the apical vertebra It is designated thoraco-lumbar if the apical vertebra is T12 or L1.

In some clinics a measure of the rotational deformity is made using an inclinometer placed across the rib hump with the patient in the forward bending position, or using Moiré topography (Figure 2) in the erect posture.

Prevalence of Scoliosis

Scoliosis is the most common of all spinal deformities (Winter 1975). Its reported prevalence varies widely between different races and in different environments, but even in similar communities it varies greatly according to the different criteria for diagnosis. Adolescent idiopathic scoliosis develops typically between ten and fourteen years of age, and school screening programmes are directed to its early diagnosis in this age group. In a rapid screening programme, adolescent idiopathic scoliosis cannot be clearly distinguished from other spinal pathology producing structural scoliosis, such as congenital hemivertebra or Scheuerman's disease, and postural scoliosis associated with leg length inequality may not be initially distinguished from structural scoliosis (Taylor and Slinger 1980). Another difficulty in comparing prevalence figures from various centres is the inconstant 'cut-off point' below which different authors ignore minor degrees of spinal asymmetry Thus some groups include all curves of 5° or more while others ignore all curves less than 10°. Kane (1977) points out that the prevalence falls from 7.7 per cent for curves of 5° or more, to 2.3 per cent for curves of 10° or more and 0.5 per cent for curves of 20° or more. The 'scoliosis prevalence' varies at different stages of the screening procedure. Typically, about 10 per cent of girls and about 7 per cent of boys are referred for further investigation, because of the presence of a rib or lumbar hump, visible on forward bending. However, when postural curves and spinal deformities other than scoliosis are excluded, the prevalence of structural scoliosis falls to about 6 per cent in girls and 4 per cent in boys, and when small insignificant curves are excluded the prevalence falls even further. Only about two per thousand adolescents screened will require active treatment, and females predominate over males in this group in the ratio of 5 to 1. (Brooks et al 1975, Golomb and Taylor 1975, Lonstein 1977, Rogala et al 1978, Taylor and Slinger 1980, Liston 1981).

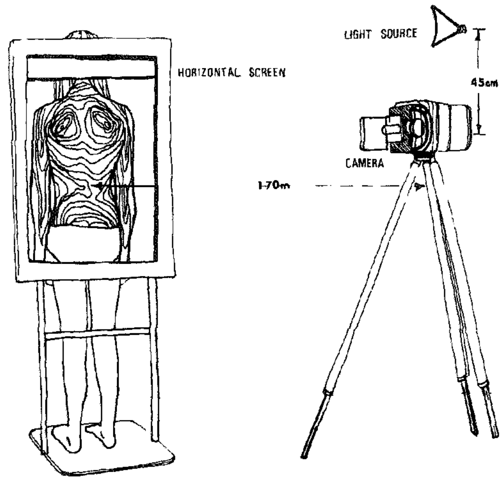

Figure 2: Moiré topography

Screening

Optimum Age for Screening

The incidence of rib or lumbar hump is similar in the 11-12 year and 13-14 year groups. Since maximum growth rate for the average female thoraco-lumbar spine is achieved between 9 and 13 years and progression is related to rate of growth, eleven years is probably the best age at which to screen girls. Boys should ideally be screened two years later since the growth spurt is two years later in boys than in girls (Rogala et al 1978, Taylor and Slinger 1980).

Screening Methods

The most frequently used screening method is the forward bending test. The patient is first viewed from behind in the erect posture and inspected for lateral spinal curvature, and then in the forward bending position with the knees straight, from behind, in front, and from the side looking for rib or lumbar hump and for kyphotic deformity (Figure 3). Lumbar hump due to rotational deformity of the spine should be distinguished from a 'high hip' due to leg length inequality with pelvic obliquity. The apparent rotation associated with pelvic obliquity can be distinguished from a true lumbar or rib hump, by repeating the forward bending test with the patient sitting on a firm level surface or following insertion of blocks of appropriate thickness below the 'short leg'. The whole examination takes less than a minute.

Moiré topography has been advocated as an alternative screening method by Adair et al (1977) Willner (1979) and Jones et al (1981). Jones points out that Moiré topography is as effective as the forward bending test when both screening procedures are used on the same population Each of the two screening methods picks up an occasional case of mild scoliosis missed by the other method, so that the two methods may be considered complementary.

In Moiré topography back contour lines are viewed through a special screen. These are the fringe patterns seen when a bright light is shone through the screen onto the patient's back (Figure 2). A rib hump causes asymmetry in the contour lines, which can be recorded photographically. Children can be screened at the rate of 30 to 40 per hour (Adair et al 1977, Jones et al 1981).

In a preliminary report, Cooke et al (1979) advocate thermography as a screening procedure, using a scanning camera sensitive to infrared radiation. In contrast to a normal group, scoliotic patients show greater infrared emission from the back on the concave side of the curvature

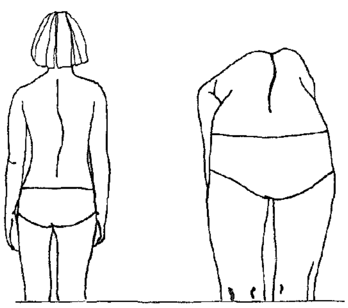

Figure 3: Forward bending test

Clinical examination using the forward bending test remains the simplest and most widely accepted method of screening. Many centres use multiple screening procedures. A primary screening by the school nurse selects a group for secondary screening by the school doctor, a subgroup being referred to an orthopaedic specialist at a 'scoliosis clinic' for final assessment (Drummond et al 1979).

The Effect of Screening Programmes on the Pattern of Treatment

The advent of school screening has greatly increased the number of patients requiring supervision and to a lesser extent the number requiring treatment. It has also reduced the percentage of patients requiring surgery while increasing the use of spinal bracing and exercise in the management of spinal deformity (Torell et al 1981, Liston 1981).

No treatment programmes should be undertaken which cannot be shown to improve the patient's prognosis, preventing deformity which would otherwise occur and thus reducing the likelihood of the complications of serious chest deformity A knowledge of the natural history of the condition is a basic prerequisite for this judgement.

Natural History

A small proportion of adolescent scoliotic curvatures less than 25° may regress spontaneously and the majority of these are non-progressive (Torell et al 1981, Taylor and Slinger 1981), but curves greater than 25° frequently progress (Winter 1975) and may progress to severe deformity during periods of rapid growth Even after skeletal maturity, moderate curves (35°-60°) may progress and severe curves (>60°) usually progress slowly, about 1° each year. Severe thoracic curves are associated with cardiopulmonary complications and a mortality up to twice the rate of the normal population, due to cardiopulmonary disease in most cases Those who survive have a high incidence of disability and back pain (Riseborough and Herndon 1975, Fowles 1978, Swank et al 1981, Weinstein et al 1981)

Since idiopathic scoliosis begins before lung growth is complete it can lead to permanent reduction in vital capacity Reduction in the volume of the pulmonary circulation leads to pulmonary hypertension and heart failure. Right ventricular hypertrophy is seen in 65 per cent of moderate and severe scoliotics at autopsy (McHardy 1979, Shneerson 1979) The appearance of the deformity in itself, causes the patient distress and cosmesis is one of the principal objectives of patients seeking surgery (Bunnell 1979, Swank et al 1981)

Indications for follow-up

A major problem is the difficulty of forecasting which curves will progress (Schultz 1976) Very few curves of less than 10° at initial examination ever progress, and reassurance is all that is required. Follow-up should concentrate on patients with curves greater than 10°, and these patients are usually seen at three monthly intervals. Clinical examination and Moiré topography may be performed at each visit and photographic records should be kept for reappraisal Radiographic reassessment is only necessary if it appears that deformity is increasing. It is important that all those with curves less than 20°, should be reassured that progression is highly unlikely but that follow-up of curves between 10° and 20° is necessary until spinal growth is complete, so that any progression can be recognised and unsightly deformity prevented. Spinal growth is virtually complete by fifteen years in girls and by seventeen years in boys

Conservative Treatment

A spinal brace is prescribed for all curves between 25° and 45° in otherwise normal immature individuals. In addition, curves with proved progression to more than 20° (5° or more increase in Cobb angle) are usually treated by a brace (Nash 1980). The Boston brace (Hall et al 1975) has superceded the Milwaukee brace in many centres. It is as effective, and is more easily accepted as it is worn beneath the clothes. The older Milwaukee brace caused lower jaw and dental deformities but modifications to the head piece has removed this problem (Mellencamp, Blount and Anderson 1977) The Milwaukee brace relies on both longitudinal distraction and lateral pressure to correct deformity and prevent progression of deformity It has been shown that with the minor and moderate curves (25°-40°) now treated by bracing, the lateral pressure applied by pads attached to the brace is more effective than the longitudinal distraction requiring the Milwaukee 'superstructure' including the head piece. The Boston brace without superstructure is effective in treating lumbar, thoracolumbar and most thoracic curves, and when necessary a superstructure can be attached to it for correcting upper thoracic curves (Watts 1979). The brace must fit well, and pads are applied laterally below the apex of each curve Bracing will prevent further progression and may produce correction of the curve, though this correction may be lost when brace treatment is terminated at skeletal maturity Because of individual variation in maturation, Risser's sign (appearance, excursion and fusion of the iliac crest apophyses) is used to judge individual maturity (Terver et al 1980).

The use of muscle stimulators with either internally or externally applied electrodes, activating post spinal or para spinal muscles, has been advocated as an alternative to brace treatment (Goldberg 1978, Bobechko et al 1979), but bracing remains the conservative treatment of choice in most centres (Bunnell 1979, Torell et al 1981).

Brace and Exercise

In the past exercise programmes have been given to attempt to correct or halt the progression of a scoliosis (Nash 1980). Klapps crawling (Gardiner 1957) and other unilateral exercise regimes (Wale 1961) are excellent for mobility, postural improvement and parent appeasement but it should be stressed that they will not abate curve progression

Exercise programmes are an essential adjunct to brace wearing (Lovell and Winter 1978, Dickens 1979 and Nash 1980). Although a brace may be of value alone, exercises are not (Stone et al 1979).

The brace is worn twenty-three hours a day and exercises are performed in the brace, and out of the brace (Watts 1977). The principles are basically the same with all types of brace.

Aims:
- Postural correction — to assist in *active* correction of abnormal curves and rib deformities.
- Chest mobility to maintain proper respiration.
- To maintain and/or increase spinal flexibility.
- To maintain and/or increase muscle strength in the whole trunk (especially abdominals).
- To maintain range of motion and prevent contractures — especially of the hip flexors.
- To maintain good general physical condition.

Specific exercises are described elsewhere (Liston 1979). They must be supervised at regular intervals by the physiotherapist.

It is recommended that the list of exercises —
- be restricted to three or four at any one time;
- are performed regularly, slowly and correctly with a gradual increase in the number of each exercise to a maximum of 15-20 repetitions for each one;
- be varied from time to time maintaining a balance of exercises to achieve the aims.

Surgery

Surgical intervention is appropriate, to correct the deformity and fuse the affected region of the spine, in patients with curves greater than 50° or in skeletally immature patients with curves from 35° to 50° where bracing treatment has failed (Moe 1980). Posterior spinal fusion, with Harrington instrumentation, or Luque rods, is the usual procedure of choice, and about 50% correction of the curvature can usually be achieved In some scolioses resulting from neuromuscular disease, anterior spinal fusion with excision of intervertebral discs and Dwyer instrumentation may be preferred. Zielke of Turbingen, Germany, has designed a similar but firmer fixation. Correction of the curve will produce height gain of several centimetres. Following surgery the patient wears a body cast for six months and then a spinal brace for six months until spinal fusion is demonstrably complete and stable.

Either procedure involves extensive surgery in a three hour procedure requiring an experienced specialist team and routine blood transfusion. Operative mortality is quoted as 0.5 per cent with an infection rate of 1 per cent or less and a pseudarthrosis rate of 2 per cent or less (Bunnell 1979, Winter 1975).

Progress in Understanding the Aetiology of Idiopathic Scoliosis

Scoliosis was described by the ancient Greeks, but we still do not know how it arises. It has been shown to be a familial condition (Wynne-Davies 1968, Riseborough and Wynne-Davies 1973, McEwan 1973). The inheritance was previously described as a sex-linked dominant mode but it now appears to be polygenic *ie* dependent on mutual reinforcement by multiple genetic characteristics for manifestation of a scoliotic curvature during growth (Czeizel et al 1978). The infantile form of the conditions is also regarded as genetic and it is found in a number of European countries However, infants of the same racial groups do not appear to develop scoliosis in North America and Australia (James 1976). This puzzling phenomenon suggests the operation of environmental factors.

It has also been noted that female adolescents develop progressive forms much more frequently than males. In this context different growth patterns may play an important role (Taylor and Slinger 1980).

The relationship between scoliosis and growth is of particular interest. Scoliosis in infants typically presents as a left thoracic curvature in males (James 1976) but in adolescents a right thoracic curvature in females is the most common pattern (Moe and Kettleson 1970). These fairly constant but different patterns at different stages of growth are seen in both idiopathic and neuromuscular forms of scoliosis. Infantile idiopathic scoliosis usually resolves spontaneously and at adolescence reversal of the direction of the original infantile primary curvature in the same individual has been observed in a number of cases (James 1976, Bjerkreim 1977, Taylor 1980). Spontaneous resolution of adolescent idiopathic scoliosis is relatively uncommon. Normal growth factors may influence the different directions of curvature seen at different stages of growth and the capacity for resolution of infantile scoliosis. In early idiopathic scoliosis, rotation is the most obvious feature and lateral curvature with secondary changes in the vertebral bodies may well follow later (Somerville 1952). Knutsson (1963) suggested that asymmetrical growth at the bilateral growth plates between the vertebral arch and the vertebral body could lead to rotational deformity and scoliosis. Taylor (1980) examined vertebrae from human fetal, infant and juvenile spines and reported that minor degrees of asymmetry in vertebral arches and asymmetrical closure of the neuro-central growth plates were very common. These asymmetries would produce rotational deformity and were such that they could cause left thoracic curves in infants and right thoracic curves in juveniles and adolescents (Figure 4). There was no associated wedging of vertebral bodies The observations of Farkas (1941) and Smola (1975) support these findings The greater tendency of females to develop progressive curves in adolescence may be related to the greater growth velocity in length of the female spine from 9-13 years. Females tend to develop longer narrower vertebral bodies than males and to have less strong muscle supports (Taylor and Slinger 1980, Twomey 1981).

This makes the female spine with a mild curvature less able to remain stable in response to axial gravitational stresses which tend to buckle the curved column.

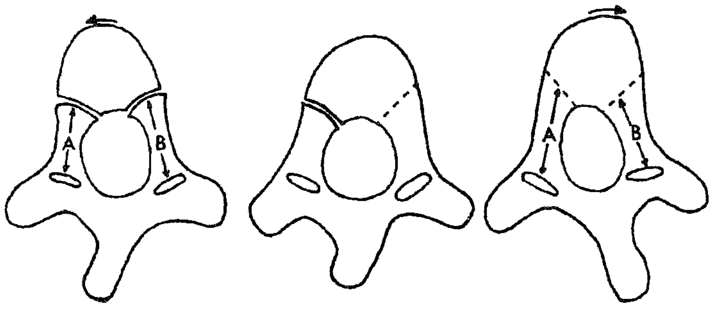

Figure 4: Vertebral bodies' asymmetries

Smola (1975) and Burwell and Dangerfield (1977) claimed that idiopathic scoliosis is part of a general skeletal growth disturbance which Smola termed the 'Length Disharmony Syndrome'. Evidence of associated asymmetrical growth of the skull (James 1976, Ceballos 1979), ribs (Sevastikoglou 1980) and limbs and trunk (Willner 1975b, Burwell and Dangerfield 1977, Taylor and Slinger 1980) supports this view.

The side of skull flattening in plagiocephaly and side of the longer upper limb correspond with the side of convexity of the thoracic scoliotic curvature (Wynne-Davies 1968, Burwell and Dangerfield 1977). While 'short leg' is more common in scoliotics than in the general population, the side of the 'short leg' bears no constant relationship to the direction of the scoliotic curvature (Willner 1975b, Taylor and Slinger 1980) Whether the primary pathology of idiopathic scoliosis is a skeletal growth disturbance, a pathology of the muscle or connective tissue supports of the spine, or a disturbance of their central nervous control remains an unsolved problem.

Anomalies of connective tissue metabolism (Zaleske et al 1980, Uden et al 1980, Clark et al 1980) and of muscle metabolism and histology (Fidler and Jowett 1976, Wong et al 1977, Yarom et al 1978) and disturbances of equilibrium (Sahlstrand et al 1978) are reported in association with idiopathic scoliosis. The connective tissue anomalies are also found in neuropathic (paralytic) scoliosis while the other anomalies are demonstrated in clinically established scoliosis and may be secondary phenomena.

On the other hand, patterns of asymmetrical skeletal growth are evident before birth, and in an apparently normal population of infants and juveniles (Taylor 1980) suggesting that normal growth phenomena contribute to the *patterns* of curvatures seen in infantile and adolescent scoliosis. This may explain the exceedingly common nature of mild scoliosis, and the different sex-related growth patterns already described may explain the higher incidence of curvature of greater magnitude in females.

Progressive forms of scoliosis may well involve other aetiological factors such as greater than average growth velocity with earlier skeletal maturity (Willner 1975a, Taylor and Slinger 1980) or unusually 'square' shaped vertebral bodies (Schultz and Cisewski 1978) which reduce the stability of the vertebral column. Alternatively, reduced efficiency of muscular support of the column due to a neuropathy (Lloyd-Roberts et al 1978, Kaplan et al 1980) or an anomaly of postural control (Sahlstrand et al 1978) may be implicated.

(*Footnote* The nomenclature used is, in the main, that used in 'A Glossary of Scoliosis Terms' (*Spine* 1976, 1, 57-58))

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