IDIOPATHIC SCOLIOSIS

The prognosis, diagnosis, and operative indications related to curve patterns and the age at onset

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The true understanding of any disease is based upon a knowledge of the etiology and natural history. In scoliosis we have little knowledge of the former; therefore the latter is of even greater significance to our comprehension and management of this still very obscure disease.

In 1950 Ponseti and Friedman presented a classification of idiopathic scoliosis based on the site and number of primary curves. Their conception is simple, and fundamental as a guide to prognosis. In this review the importance of Ponseti's views are confirmed. However, it will be seen that by considering not only the site of the primary curve but also the age at onset of the curve, an even greater accuracy in prognosis is possible. Each pattern of scoliosis has its characteristic age or ages of onset and, as would be expected, the earlier the onset the worse the final curvature. The age of onset is, however, subordinate to the site of the primary curve in the evolution of a curve.

FIG. 1

The iliac apophyses have reached the posterior superior spine. This indicates with reasonable certainty that an idiopathic curve will not increase.

Idiopathic scoliosis begins at all ages of childhood but there are three peak periods of onset—under the age of three, from five to eight, and from ten until the end of growth. For convenience I have called these age groups, infantile, juvenile and adolescent.

Two hundred and forty-one mature cases of idiopathic scoliosis whose curves will not now increase, and sixty-seven immature curves have been reviewed. A mature curve is one where spinal growth and curvature have ceased. Risser and Ferguson's (1936) observation that this coincides with completion of growth in the iliac apophyses round to the posterior superior spine has been confirmed (Fig. 1). The immature cases are discussed where relevant but not in statistics relating to the final curvature.

The severity of a curve has been based on radiographic criteria only. Although radiography gives us a true assessment of the amount of curvature, it does not indicate the

effect upon the child's appearance, for this varies greatly in each curve pattern, though the angle of curvature be the same. Radiographically, curves up to 69 degrees are classified as mild, those from 70 to 99 degrees as severe, and those greater than 100 degrees as very severe.

The types of idiopathic scoliosis, with the number of mature cases seen, are shown in Table I. A few in each series were not mature.

It will be seen that the percentage of each pattern in the two series varies considerably. One possible explanation is that Ponseti and Friedman recognised the primary curve

TABLE I
IDIOPATHIC SCOLIOSIS: CLASSIFICATION AND INCIDENCE BY TYPE

	Number of cases	
	Ponseti and Friedman	James
Lumbar	93	79
Thoraco-lumbar	63	26
Thoracic	87	Adolescent 66 Juvenile 16 Infantile 52
Combined lumbar and thoracic	146	69
Cervico-thoracic	5	0

essentially by radiological criteria, whereas in this series the clinical examination has been regarded as of greater importance. Fixed rotation on forward flexion has been interpreted as indicating a primary curve. In most cases the physical signs and the radiographic appearance lead to similar conclusions. It is in the differentiation of intermediate types that difficulty arises and this may explain the differences in the two series.

The essential feature of each pattern may now be briefly reviewed together with the degree of disability to be expected.

LUMBAR IDIOPATHIC SCOLIOSIS

Seventy-nine mature cases are reviewed. Except for one curve appearing at nine, the curvature invariably developed from ten years of age until the end of growth, the onset being equally scattered through these years of growth. The lumbar curve was unique in this onset during the years of adolescence.

There were seventy female patients and nine male. Unlike the other curves to be studied the proportion of right and left curves was almost equal. The apex of a lumbar curve was most commonly at Lumbar 1 but it was not rare at Lumbar 2 or 3. No significant difference in prognosis could be found in the variation of apex, site or length of this essentially homogeneous pattern. The degree of curvature found in this group at the end of growth confirms Ponseti and Friedman's view that it is benign (Fig. 2).

It is in this group that the radiological criteria of severity are particularly harsh, for a "severe" curve of over 70 degrees is not in fact ugly. Even so, only seven patients, or 9 per

FIG. 2
Lumbar idiopathic scoliosis. The final curvature in seventy-nine mature patients.

cent, had a curvature of over 70 degrees. The minimum final curvature was 12 degrees, the maximum 90 degrees (Fig. 3).

This curve pattern is benign not only because the degree of curvature remains small, but because no ribs are involved in the rotation, and the shoulders remain level.

Although it is, from the cosmetic point of view, an innocent curve, there is in later life one serious consequence—backache. In this series, thirty-nine patients were over twenty-five years of age and all had backache. Contrast this with thoracic scoliosis where only six patients amongst eighty-two complained of pain. The pain appears to arise in the posterior intervertebral joints, which probably become osteoarthritic because of their extreme rotation (Fig. 4).

FIG. 3

Lumbar idiopathic scoliosis. A curvature of 90 degrees, the most severe lumbar curve in the series. Note that the appearance is not grossly affected.

THORACO-LUMBAR IDIOPATHIC SCOLIOSIS

This curve is not common: twenty-six cases only are available. The onset was in adolescence except for one at three and another at four years of age. Twenty of these twenty-six patients were girls and most curves were convex to the right. The apex always involved the eleventh or twelfth thoracic vertebra (Fig. 5).

As we review the patterns it will be found, in general, that the prognosis becomes worse as the site of the primary curve ascends in the vertebral column. The thoraco-lumbar curves are therefore intermediate between lumbar and thoracic curves in all characteristics. Some ribs are involved in the rotation, the shoulder sometimes drops, and on occasions the hip becomes extremely prominent, so that a slight curve may be very ugly. The incidence of

FIG. 4

A patient of sixty years with a 70-degrees curve. Her appearance is good. The radiograph shows osteoarthritis of the posterior joints, which caused severe pain.

FIG. 5

Thoraco-lumbar idiopathic scoliosis. The photograph and radiograph of a patient with a 60-degrees curve in the thoraco-lumbar region, an average end-result.

severe cases was 31 per cent compared with 9 per cent in the lumbar group (Figs. 6 and 7). Only two of this group have required correction and fusion.

As might be predicted with a curve which is half within the lumbar area, eight adults of the nine complained of pain.

FIG. 6

Thoraco-lumbar idiopathic scoliosis. The final curvature in twenty-six mature patients.

FIG. 7

Thoraco-lumbar idiopathic scoliosis. The greatest thoraco-lumbar curve in the series. 99 degrees.

THORACIC IDIOPATHIC SCOLIOSIS

This large group can be usefully subdivided according to the age of onset: of the 134 mature curves sixty-six began in adolescence, sixteen were classified as juvenile and fifty-two as infantile. In this last subdivision not all the patients were fully grown. Thus it can be seen that the thoracic curve is common: it is also the most important.

It is in thoracic scoliosis that consideration of the age at onset is most important. In many patients thoracic scoliosis commences early, and the earlier it begins, the worse the prognosis (Fig. 8).

Thoracic scoliosis in infants is more common in boys and is to the left in 92 per cent; in adolescence it is to the right in 90 per cent and nearly always affects girls (Figs. 9 and 10). These and other differences have suggested a subdivision of thoracic scoliosis into three groups based on the age of onset. These will now be considered separately.

FIG. 8

Thoracic scoliosis. Percentage of curves greater than 70 degrees related to the age of onset.

FIG. 9

FIG. 10

Figure 9—Sex incidence in infantile and adolescent thoracic scoliosis. Figure 10—Convexity of curve in infantile and adolescent thoracic scoliosis.

The site of the apex from Thoracic 6-10 and the number of vertebrae in the curve did not differ materially between these groups.

Adolescent thoracic idiopathic scoliosis—Sixty-six mature cases were examined. The age of onset was from ten years onwards. The prognosis did not vary whether the onset was in early or late adolescence: the average age of the patients with mild curves at onset was 12.1 years, and of those with very severe curves 12.3 years.

There were fifty-four girls amongst these sixty-six patients. Sixty of the curves were convex to the right, and it is remarkable that only two girls had a curvature to the left, whereas four of the twelve boys had left curves.

The prognosis is unfavourable because, beyond all others, this pattern of curve tends to increase to a gross degree. The percentage of severe or very severe curves was 59 per cent. The smallest mature curve was 24 degrees, the worst 151 degrees (Fig. 11).

FIG. 11

Adolescent thoracic idiopathic scoliosis. The final curvature in sixty-six mature patients.

In thoracic scoliosis the deformity is made worse by rotation of ribs, dropping of the shoulder, and prominence of the hip. In this group the radiological grading does not sufficiently stress the ugliness of the curve (Figs. 12 to 14).

FIG. 12

Adolescent right thoracic scoliosis of 76 degrees. These curves often need operative correction at 70 degrees but there is considerable individual variation in appearance.

Nineteen corrections and fusions were undertaken in this group, because of either existing or expected deformity. Many curves were incorrigible when first seen.

Juvenile thoracic idiopathic scoliosis—This small group of sixteen with an onset between the fifth and eighth years is possibly not worthy of separation from the preceding group:

FIG. 13

FIG. 14

Adolescent right thoracic scoliosis. At the age of thirteen the curve was 51 degrees (Fig. 13). Three years later the curve had increased to 151 degrees (Fig. 14).

FIG. 15

Infantile thoracic idiopathic scoliosis. A girl of four and three-quarter years with a curvature of 87 degrees. A year later it was 116 degrees.

FIG. 16

FIG. 17

Infantile thoracic idiopathic scoliosis. At the age of one and a half years the curvature was of 40 degrees (Fig. 16). At the age of twelve years it had increased to 136 degrees (Fig. 17).

FIG. 18

Combined lumbar and thoracic scoliosis. A combined curve, each primary being of 66 degrees.

further observation will clarify this detail. Nevertheless the prognosis does seem significantly worse. Fifteen of these sixteen patients were girls; all but two of the curves were right.

There was a majority of severe curves (87 per cent), as might be expected of a curve progressing during many years of active growth. The two mild curves were only classified so by a margin of a few degrees. Of this small group nine have been corrected and fused and three were inoperable.

Infantile idiopathic thoracic scoliosis—Fifty-two cases will be discussed, but nine of these must be considered separately because their curves disappeared spontaneously. Of the remaining forty-three patients only a proportion are fully grown. Adults attending with a history of onset in infancy have been excluded unless an early radiograph showed no bony congenital abnormality, because adult radiographs are so confused by secondary changes that the exact etiology cannot always be established. In all cases the onset has been before three years of age and was most common from the sixth to eighteenth month of life. Boys predominate and left curves have a near monopoly.

TABLE II
INFANTILE IDIOPATHIC THORACIC SCOLIOSIS:
SEVERITY OF CURVE RELATED TO PRESENT AGE

Curvature	Number of patients	Average present age
0°-69°	21	4.8 years
70°-99°	12	8.2 years
over 100°	10	12.3 years

These and other features so separate them from the other thoracic scolioses as to leave small doubt that this is a distinct pattern. There is much evidence to suggest that it is in no way different from the idiopathic scoliosis of later years. No previous suggestion has been found in the literature that an infantile group of idiopathic scoliosis exists but exclusion of all cases with visible bony abnormality leaves this remarkably homogeneous group.

Although many curves are immature, comparison of the average age in children who have at present mild, severe or very severe curves is instructive (Table II).

This analysis suggests that only the very young have mild curves. No child over the age of ten years has escaped a curve of 70 degrees or more, nine children under ten have curves of over 70 degrees and several of these have curves over 100 degrees. It seems likely that in every case the prognosis is serious, and severe crippling, with a curve of over 70 degrees and often over 100 degrees, must be expected. There are now few orthopaedic conditions able to produce so hideous a deformity as this pattern of scoliosis (Figs. 15 to 17).

As already mentioned, there are still to be considered nine cases, all in patients under two years, with curves of less than 20 degrees with radiological and sometimes clinical rotation. In all, the curves disappeared spontaneously by two years of age. I am not fully persuaded that they were all structural curves, although some had a fixed rotation.

CERVICO-THORACIC IDIOPATHIC SCOLIOSIS

Ponseti described five such cases with a good prognosis. No cases have been seen in this series.

COMBINED LUMBAR AND THORACIC IDIOPATHIC SCOLIOSIS

Until now we have reviewed scolioses with three curves, the middle one of which is the primary. This last pattern has four curves, the middle two being primary. It combines characteristics of both lumbar and thoracic curves but the development of the thoracic element seems to be held in check. The shoulders remain level, the hips covered, and with a

posterior rotation on the left and right the back looks flat. The patient looks much better than the radiograph would suggest. The two primary curves generally keep in step (Fig. 18).

Fifty-four mature cases were seen. The majority of combined curves started in adolescence. This pattern, which does occasionally commence in infancy, is the most frequent curve arising in the juvenile group. In addition to the fifty-four mature cases whose prognosis is reviewed, there were additional patients, still immature. Altogether nine combined curves started in infancy, but it is much less common than the infantile thoracic curve already discussed. Seventeen curves started between the fifth and eighth years and it is the characteristic curve of the juvenile group. Amongst the nine infantile curves were three males and six left thoracic curves.

The adolescent group consisted largely of girls with right thoracic primary curves. The

FIG. 19

Combined lumbar and thoracic scoliosis. The final curvature in fifty-four mature patients.

apex of the thoracic curve lay between the sixth and eighth thoracic vertebrae and the lower curve at the second lumbar.

The prognosis is good, only 31 per cent being classified as severe or very severe. The three very severe curves of over 100 degrees all started in the juvenile group but there were twelve severe cases in the adolescent group (Fig. 19). Because of the small cosmetic effect no patient has so far been advised to have correction and fusion (Figs. 20 to 22).

DISCUSSION

It is instructive to compare the prognosis and appearance of the four major patterns that have been described (Figs. 23 and 24).

There is no evidence that any form of conservative treatment influences the prognosis; indeed most of the mild curves received no treatment, whereas the severe curves have all received vigorous and varied conservative therapy over many years.

It is realised that the material drawn from a special clinic is selected, with an undue proportion of serious cases referred. This is unlikely to have affected the comparison between the various patterns.

Diagnosis—Idiopathic scoliosis is a three-curve arrangement, or four. In the former case the middle one of the three is primary, in the latter the middle two of four. The primary curves retain rotation on forward flexion, whereas the compensatory curves do not except to a minor degree in some long standing cases. Clinical examination is better than radiography to determine which is the primary curve and rotation is the significant sign in establishing the curve pattern. Occasional intermediate patterns with minimal rotation in the lumbar region offer difficulty in diagnosis between the three- and four-curve patterns.

Indications for operation—The lumbar and perhaps the combined thoracic and lumbar scolioses never need correction, although fusion may be indicated for pain.

FIG. 20

FIG. 21

Combined lumbar and thoracic idiopathic scoliosis in a girl. At the age of eleven the primary curvature was 37 degrees and 30 degrees (Fig. 20). Four years later it was 101 degrees and 112 degrees (Fig. 21).

FIG. 22

Same patient. Photograph to show clinical appearance of curve at age fifteen.

Thoraco-lumbar curves rarely require correction and fusion, and so good is the general prognosis that it is better to await the development of deformity, although a prominent hip may be most conspicuous even when the curve is small.

FIG. 23

Prognosis according to pattern of curve. Incidence of mild, severe and very severe curves in each of the four major curve patterns. Mild curves are represented by light shading, severe curves by dark shading, and very severe curves in black. Compare with Figure 24.

FIG. 24

Photographs of four patients each with the same degree of curvature (70 degrees) but each with a different curve pattern. From left to right the curves are lumbar, thoraco-lumbar, combined thoracic and lumbar, and thoracic.

In thoracic scoliosis the situation is different. When the onset is in adolescence perhaps half of all patients may be expected to develop a severe curve; with a curve of juvenile onset the proportion is over 80 per cent, and with an infantile over 90 per cent.

Severe scoliosis is not in itself an invariable indication for radical treatment, but a curve with a bad prognosis in degree of curvature and appearance obviously demands surgery.

If the curve pattern and age of onset are considered together, accurate prognosis is possible. Correction and fusion may then be confidently advised for the *prevention* of future deformity. Correction of well established curves is difficult and the rib rotation remains. In young patients, before deformity is serious, correction is relatively easy and more complete, but I have hesitated so far to operate before the age of ten years. If spinal fusion in adolescence proves satisfactory, it may be possible to intervene at an earlier age.

SUMMARY

The evolution of an idiopathic scoliosis is determined by the site of the primary curve and by the age of onset. It is significant that thoracic primary curves are commonly severe and the early onset of this curve accentuates this feature. Early operation based on prognosis is practised but sufficient time has not yet elapsed to justify any conclusions.

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