PROGRESSIVE AND RESOLVING INFANTILE IDIOPATHIC SCOLIOSIS

The Differential Diagnosis

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From the time of the initial definition of infantile idiopathic scoliosis as an entity (James 1951) it has been appreciated that in many infants with this scoliosis spontaneous resolution occurs in the first years of life. There is, however, a smaller but much more important group in which seemingly identical curves progress relentlessly, perhaps to 150 degrees, causing one of the severest forms of scoliosis and frequently premature death from cor pulmonale (Scott and Morgan 1955).

FIG. 1

The radiograph at 5 months of a typical resolving curve, initially 33 degrees with an angle difference of 16 degrees. The rib head on the convex side is clear of the body of the apical vertebra (Mehta's Phase I) (*left-hand side*). The radiograph eighteen months later showing normal appearances (*right-hand side*).

From this first paper and from subsequent reports on infantile idiopathic scoliosis (James 1954, 1971; James, Lloyd-Roberts and Pilcher 1959; Scott 1959; Lloyd-Roberts and Pilcher 1965) the difficulties of distinguishing the resolving and progressive forms have been stressed, for in their earliest stage they are clinically identical.

The best differential diagnosis previously available has been limited to an appreciation that a curve already severe will inevitably progress; the greatest curve reported to resolve was 37 degrees (James *et al.* 1959). The other feature thought to indicate a progressive scoliosis was the development of compensatory curves above and below the primary curve. When

FIG. 2

The radiograph at 4 years of a progressive curve with early compensatory curvatures, and angle difference of 22 degrees, and the rib head obscured by the body of the apical vertebra on the convex side (Mehta's Phase II) (*left-hand side*). The adjacent radiograph five years later shows that the curve has been only slowly progressive.

these could be seen in the radiograph, progression had seemed inevitable, though this was essentially an impression. Obviously neither a small angle of curvature nor the absence of compensatory curves is useful in the earliest cases (James 1971).

The prognosis of these two types of infantile idiopathic scoliosis is now well established. In the important paper by Lloyd-Roberts and Pilcher (1965) it was reported that 90 per cent of the idiopathic structural scolioses seen in infants in the first year of life disappeared spontaneously (Fig. 1), whereas 10 per cent progressed (Fig. 2). A curve starting in the second or third year of life is much more inclined to progress; indeed the majority of such curves remain and increase.

An early opportunity by one of the authors (J. I. P. J.) to read the 1968 Robert Jones Prize Essay by Miss M. H. Mehta, since published in part (Mehta 1972), led to a review of her findings in our series of infantile idiopathic scoliosis of both types.

In 132 cases of infantile idiopathic scoliosis we have reviewed the first radiograph to ascertain the significance of the angle of curvature, the presence or absence of compensatory

curves, and the important original observations of Mehta on the enclosed angles between the apical vertebral body and its two ribs. Mehta found the difference between the rib-vertebra angles on the concavity and the convexity to be small in those curves which later resolved, but in progressive curves the initial difference was found to be large. In addition she described a difference in the position of the rib head and the vertebra in resolving and in progressive curves, and a difference in the obliquity of the twelfth rib in double primary curves.

FIG. 3

A tracing from a radiograph to show measurement of the rib-vertebra angles, and the rib-vertebra angle difference. The rib at the apex of the convexity shows the position characteristic of progressive curves. The rib head is behind the apical vertebra.

In our series we have found the correlation with her observations to be excellent, and indeed we have confirmed her findings in a higher percentage than she herself reported. A retrospective study using the four criteria—the greatest angle of curvature, the presence of compensatory curves, the rib-vertebra angle difference and the relationship of the head of the rib to the vertebral body—has allowed a satisfactory differentiation between progressive and resolving curves even in the earliest stages. To be able to reassure parents that a curve is likely to disappear is invaluable and avoids much unnecessary treatment and anxiety. No less important is the early recognition of those progressive curves which by the use of plaster and a Milwaukee brace can be controlled before they become catastrophic.

The method of measuring the rib-vertebral angle was clearly described by Mehta. The angle is measured on both sides of the apical vertebra. A perpendicular is drawn to a line parallel to the lower border of the apical vertebra (Fig. 3); lines are then drawn through the mid-line of the neck and head of each rib to meet this perpendicular. The angles at which these lines meet the perpendicular are the rib-vertebra angles and the difference between them is the rib-vertebra angle difference. In thoracic curves it is greater on the concavity.

The apical vertebra was always chosen but if the apex of the curve was at an intervertebral space the vertebra above was used. To determine the apical vertebra is important because small variations in measurement are found when the adjacent vertebrae are selected and a

significant difference could arise. For the measurement a radiograph taken in the supine position to give greater clarity was used. No particular difficulty was found in making satisfactory and repeatable measurements, but not quite as easily as might seem from reading Mehta's description. The axis along the middle of the rib is perhaps the hardest to judge.

OBSERVATIONS

The type and the outcome of the 132 cases are shown in Table I.

TABLE I
OUTCOME IN 132 CASES OF IDIOPATHIC INFANTILE SCOLIOSIS

	Number of cases	Per cent
Resolving	68	52
Progressive	40	30
Double primary curves	12	9
Improving or resolved after treatment	12	9

ANGLE OF CURVATURE

Whereas a small angle of curvature clearly implies a probability of spontaneous resolution, there is no question that when these babies are seen very young their curves are small whatever the outcome. Figure 4 illustrates well the danger of thinking a small curve indicates resolution. However, small curves do mostly resolve and large curves usually progress. The largest curve to resolve spontaneously in this series was one of 46 degrees. When such a large curve can

FIG. 4

Radiographs to illustrate the danger if the potential severity of infantile idiopathic scoliosis is not always remembered. By the old criteria applied to the first radiograph this child could easily have had a resolving scoliosis, but within three years it had produced this disastrous deformity.

disappear it is useless to define a maximum angle beyond which resolution never occurs, because nearly all progressive curves are less than this when first seen.

Compensatory curves—When infantile idiopathic thoracic scoliosis is first recognised there is usually a single curve, the primary curve; compensatory curves have not developed. It has been stated already that when compensatory curves develop the primary curve will prove to be progressive. In this series this impression was confirmed. No child with resolving scoliosis developed compensatory curves. However, in early cases the value of this observation is limited because neither progressive nor resolving scolioses at this stage show compensatory curves. Forecasting the outcome by the aid of the initial angle of curvature and the presence or absence of compensatory curves is therefore useful but of limited value in the early stages.

The observation that a curve is increasing suggests progressive scoliosis but in the earliest stages it is not tantamount to proof. Observation over six to twelve months, however, nearly always gives an answer because by this time the curve is either disappearing or has increased.

RIB-VERTEBRA ANGLE DIFFERENCE AND RIB HEAD POSITION

Resolving thoracic scoliosis—In the series of 132 cases sixty-eight curves have proved to be resolving. Fifty-three were observed to completion—that is, they became straight. In the other fifteen, still under observation after a year or more, the curves have all but disappeared or are

TABLE II
SIXTY-EIGHT RESOLVING CURVES

Rib-vertebra angle difference	Number of cases	Angle of curve
Less than 10 degrees	36	Less than 24 degrees
From 11 to 19 degrees	31	From 7 to 46 degrees
20 degrees	1	10 degrees

TABLE III
FORTY PROGRESSIVE CURVES

Rib-vertebra angle difference	Number of cases	Angle of curvature
20 degrees or less	5	Initial angle 15 to 94 degrees
From 21 to 30 degrees	11	
From 31 to 40 degrees	8	
Over 40 degrees	16	

clearly doing so. In sixty-seven of the sixty-eight resolving curves the angle difference was less than 20 degrees. In the one exception the difference was 20 degrees with a 10-degree curve; measurement several months later showed the difference reduced to 16 degrees with a 3-degree curve (Table II).

In this group the curves measured from 2 to 46 degrees initially. One could not closely correlate the rib-vertebra angle difference with the angle of curvature: the 46-degree curve that resolved had a 15-degree difference, and the 20 degrees difference occurred in a curve of 10 degrees. It is to be noted, however, that of these sixty-eight resolving curves only two had angles of curvature greater than 30 degrees.

In the sixty-eight resolving curves the relationship of the rib heads clear of the body of the apical vertebra was confirmed as being in Mehta's Phase I (Fig. 3).

Progressive thoracic scoliosis—The findings in forty patients with curves which proved to be progressive again confirmed Mehta's observations. In this group the initial curves varied from 15 to 94 degrees. The 15-degree curve, the smallest, showed an angle difference of 21 degrees, and the 94-degree curve, the largest, had an angle difference of 55 degrees.

Only three of the forty curves had an angle difference less than 20 degrees: these had curves of 27 to 36 degrees. Two of these curves showed an increase of the angle difference to 21 degrees in the first year. One remained at the very low level of 3 to 4 degrees; it then became negative—that is to say, the angle was greater on the convex side, the criterion of the double curve. However, there was no clinical sign of a double curve, and the single curve, after slow deterioration, was fused.

Two of the progressive curves had an initial angle difference of 20 degrees, with curves of 58 degrees and 64 degrees respectively. Both curves on follow-up proved to be very serious, deteriorating quickly to 92 and 112 degrees respectively, and the difference increased to 34 degrees. Both required correction and fusion.

Except for one patient with a 15-degree curve, the remaining thirty-five patients with progressive scoliosis had severe curves and had angle differences varying from 21 to more than 60 degrees. The variation is shown in Table III. The relationship of rib head to vertebra confirmed Mehta's observations in all cases (Fig. 3).

Thus for single primary curves in the thoracic region Mehta's observations have been fully confirmed. It is clear that, as she suggests, 20 degrees is the borderline angle difference; only one of sixty-eight resolving curves had an angle difference of 20 degrees. In thirty-five out of forty progressive curves the difference was more than 20 degrees. In resolving curves the angle difference becomes even less as the curves get smaller; in progressive scoliosis it becomes greater as the angle of curvature increases.

Double primary curves—As has been noted in previous reports on infantile idiopathic scoliosis, double primary curves are not uncommon, particularly in those that begin in the second or third year. It has also been observed that the scoliosis may present and persist for some time as a single primary curve in the thoracic region; on forward bending the child shows no rotation in the lumbar region. Nevertheless, the thoracic curve progresses more slowly than one would expect in a single primary thoracic scoliosis and the observer may be puzzled by this. On a number of occasions the explanation has become apparent only when lumbar rotation appeared, the patient clearly having then a double primary curve. The differentiation therefore between a single and double curve may require a year or two of observation, and the prognosis has to be guarded until this point is clarified.

In this series twelve of the 132 patients were known to have double curves. In these

FIG. 5

The radiograph at 3 years 9 months of a typical double curve with a negative angle difference of 16 degrees, that is, a greater angle on the convexity. The characteristic droop of the eleventh and twelfth ribs on the concave side is well shown.

twelve, retrospective measurement of the earliest radiographs showed either a very low figure for the angle difference or even a negative value, that is, the angle between the rib and apical vertebra was greater on the convex than on the concave side. This proved a reliable and early sign. The obliquity of the twelfth ribs, and occasionally of the eleventh ribs, a curious feature which Mehta noticed to be present in double curves (Fig. 5), was also present in all; indeed its constancy proved remarkable.

It is clear that a very low angle difference may indicate a double curve rather than a resolving single curve. This small difference may become negative with a slowly deteriorating curve which later develops a double curve pattern. The very low or negative angle difference and the oblique twelfth rib were always present from the beginning; thus double curves could be identified from the start. The prognosis of double curves, though they slowly progress, is much better than for single thoracic curves. No double curves resolved.

Treated cases—Amongst 132 patients were twelve children who were thought to have had progressive curves but who did well. (The others reported above have remained severe and already have had or are awaiting fusion.) This view was based on a large angle of curvature, an increasing curvature, or sometimes the presence of compensatory curves. For such reasons these patients were treated either by hinging plaster casts followed by a holding jacket if under two years or by a Milwaukee brace if older.

In three patients the curves disappeared completely after treatment. In retrospective measurement the first of these was found to have a negative angle difference of -9 degrees with a 30-degree curve in the initial radiograph. Treatment had been continued for more than three years before the brace was discarded. After further observation the patient was finally discharged without any scoliosis. This may well have been a case of double curvature.

The second patient had an angle difference of 18 degrees and a curve of 24 degrees which was tending to increase. The holding jacket was maintained for fifteen months, by which time the curve had disappeared. By these new criteria this may well have been a case of resolving scoliosis.

In the third patient there was an initial curve of 28 degrees with an angle difference of 20 degrees. After three years the curve had become so small that it seemed safe to discard the brace. At one stage the curve was 2 degrees with an angle difference of 2 degrees. The curve finally disappeared. It would seem likely, again in retrospect, that with an initial rib-vertebra angle difference of 20 degrees this may have been a resolving scoliosis.

Three other patients showed an initial low angle difference which became negative after treatment with a brace. In all three the curves have remained very small, around 5 degrees; all are now free and progressing favourably. It could be that these were cases of double primary curves.

In the remaining six patients, still under observation, all had an initial angle difference less than 20 degrees. Four are now out of their braces, having done well, and two are still in a brace with curves less than 10 degrees. It may well be that because of their increase in the initial stages these curves were mistakenly regarded as progressive.

SUMMARY

1. Resolving infantile scoliosis is transient and unimportant; progressive infantile idiopathic scoliosis can be catastrophic.
2. To be able to differentiate the two at an early stage is a considerable advance. This is important for many reasons, but particularly for parents who are anxious for the future of an infant with a small curve which looks so innocent but which can be so malignant.
3. With the new observations reported by Mehta on the difference of the angles between the apical vertebra and its two ribs, and on the radiological relationship of these rib heads to the vertebral body, the prognosis is now almost wholly reliable.

4. Our former clinical impression that all cases with compensatory curves are progressive has now been verified.
5. At last an early distinction between progressive and resolving scoliosis can be made with confidence.

REFERENCES

- JAMES, J. I. P. (1951): Two Curve Patterns in Idiopathic Structural Scoliosis. *Journal of Bone and Joint Surgery*, **33-B**, 399.
- JAMES, J. I. P. (1954): Idiopathic Scoliosis. *Journal of Bone and Joint Surgery*, **36-B**, 36.
- JAMES, J. I. P. (1971): Infantile Idiopathic Scoliosis. *Clinical Orthopaedics and Related Research*, **77**, 57.
- JAMES, J. I. P., LLOYD-ROBERTS, G. C., and PILCHER, M. F. (1959): Infantile Structural Scoliosis. *Journal of Bone and Joint Surgery*, **41-B**, 719.
- LLOYD-ROBERTS, G. C., and PILCHER, M. F. (1965): Structural Idiopathic Scoliosis in Infancy. *Journal of Bone and Joint Surgery*, **47-B**, 520.
- MEHTA, M. H. (1972): The Rib-vertebra Angle in the Early Diagnosis between Resolving and Progressive Infantile Scoliosis. *Journal of Bone and Joint Surgery*, **54-B**, 230.
- SCOTT, J. C. (1959): Resolving Scoliosis. *Journal of Bone and Joint Surgery*, **41-B**, 105.
- SCOTT, J. C., and MORGAN, T. H. (1955): The Natural History and Prognosis of Infantile Idiopathic Scoliosis. *Journal of Bone and Joint Surgery*, **37-B**, 400.