THE MANAGEMENT OF INFANTS WITH SCOLIOSIS

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This is a study of children who first attended as infants with either progressive infantile idiopathic scoliosis or congenital scoliosis. All had a pattern of scoliosis in which early and damaging deterioration is inevitable. The infants were treated from before the age of three, initially by plaster casts and then a Milwaukee brace, followed at about the age of ten by correction and fusion. The cases were then observed to the end of growth or near that point. In the main study there were twelve cases, six of progressive infantile idiopathic scoliosis and six of congenital scoliosis, which were followed through this long period. Only one of the twelve had a curve worse at the end of growth compared with the initial radiograph as an infant; this one curve had increased only 16 degrees in almost as many years. Although small, the series does show that it is nearly always possible to control even the most serious scoliosis in an infant, if it is tackled early and unremittingly. There are supportive studies of children who have partially completed this regime, and interim results in a newer group of children with spina bifida and scoliosis.

Scoliosis still presents some of our most difficult problems, problems that are indeed at times insoluble. This is particularly true of infants under three years of age with progressive scoliosis, for over the next fifteen to eighteen years of growth their curves are likely to progress relentlessly and may lead to an uncomfortable death from cor pulmonale in early adult life. A severe scoliosis causes compression of the lungs with consequent difficulty in aeration of the alveoli, and right heart hypertrophy because of the difficulty in circulating blood through the lungs. There is then a gradual failure of both respiratory and cardiac function—cor pulmonale (Zorab 1973). Curves over 70 degrees if idiopathic, or less if congenital or paralytic, are sufficient to cause cor pulmonale. In Sweden 30 per cent of such patients are granted a disability pension (Bjure and Nachemson 1973), and the mortality is twice that expected of their age group. Disability and death are almost invariably cardiac and respiratory in origin. Few months go by in which one of these tragic late cases is not referred to us, a reminder, if one were needed, of the penalties of doing nothing. On the other hand successful management is not easy. The parents and the child must face together many wearisome years of visits to clinics, the wearing of a clumsy cast or brace, and finally spinal fusion.

Many hundreds of older children whose curves started in infancy have been seen at the scoliosis clinic held initially in London from 1948 to 1958 and subsequently in Edinburgh. The great majority came too late, and it was they who taught us how serious is the result of neglect. It has therefore seemed worth while to review those cases from the Edinburgh clinic who came before the age of three and who have remained under care throughout growth, amply long enough to demonstrate the results that can be achieved by attempts to prevent serious curvature. In our view spinal fusion in the very young is wrong, so this assessment is of our present methods, which include observation, hinging plaster jackets, the Milwaukee brace and spinal fusion at the age of ten or eleven. The number in whom an end-result is available is relatively small because of the many years required for a continuous study from infancy to skeletal maturation.

While scoliosis in the older child may result from any one of fifty different diseases, in the infant there are only a few causes, with a great predominance of the infantile idiopathic and congenital varieties. A few infants have been seen with scoliosis from Morquio’s disease and neurofibromatosis, but they are too few to consider. In congenital scoliosis a significant degree of curvature is frequently delayed until early adolescence; infants with a visible congenital scoliosis before the age of three are uncommon and the curvature usually becomes very severe; this small group with a high risk of early deterioration concerns us here.

INFANTILE IDIOPATHIC SCOLIOSIS

This disorder affects the majority of infants with scoliosis, and indeed its frequency in Great Britain as compared with North America is quite astonishing (Wynne-Davies 1975). A study of 200 cases of idiopathic scoliosis seen between 1968 and 1972 indicates the proportion of cases of early onset (Table 1).

<table>
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<th>TABLE 1</th>
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<td>Two Hundred Consecutive Cases of Idiopathic Scoliosis</td>
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<tr>
<td>-----------------------------------------------</td>
</tr>
<tr>
<td>Infantile (onset before 3 years) ... 82</td>
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<tr>
<td>Juvenile (onset from 4 to 9 years) ... 34</td>
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<tr>
<td>Adolescent (from 10 years to end of growth) ... 84</td>
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In the present series there was a total of 120 cases of infantile idiopathic scoliosis to be considered. Plagiocephaly is present at an early age in all infants with idiopathic scoliosis; the typical facial and cranial deformity is shown in Figure 3. As is now well known, the majority show spontaneous resolution of the structural curve and have so-called resolving infantile scoliosis. In contrast the minority have progressive curves, and if not treated deteriorate steadily, forming a group comprising some of the most badly deformed children we see.

Resolving infantile idiopathic scoliosis
In the last sixteen years ninety cases have been followed through to resolution. In three cases there is no record of when this happened, but in the other eighty-seven the curves had all disappeared by the sixth year (Table II).

![Diagram of scoliosis](image)

**Figure 1**
To show the method of measuring the rib-vertebra angles at the apex of the curve (Mehta 1972). In this tracing of a radiograph the rib on the concave side forms the greater angle with the vertebra. The rib-vertebra angle difference (R.V.A.D.) is therefore positive for the concave side. Because the difference here is 38 degrees this is a progressive curve. Notice also that the head of the rib is partly behind the apical vertebra on the convexity, another indication of a progressive curve.

There was no other feature of interest to report, but one must stress the importance of Mehta's (1972) observation that the difference in obliquity of the ribs on either side of the apical vertebra is of great value in the differential diagnosis between resolving and progressive scoliosis (Fig. 1). When the rib-vertebra angle difference (RVAD) is less than 20 degrees, resolution will occur (Fig. 2); a difference of more than 20 degrees usually indicates a progressive curve. The rib-vertebra angle is greater on the concavity in single curve scoliosis because the rib on the concave side is less oblique to the vertebra.

**Double structural curves**—Mehta described two radiological features indicating the presence of a second structural curve, the double structural curve scoliosis of infants, before it could be suspected clinically. These are a very oblique eleventh or twelfth rib on the concave side and a rib-vertebra angle now greater on the convexity (Fig. 4). In this series double curves in two infants have undergone resolution, an occurrence not previously reported. There were four other infants with double curves; one has not increased during ten years and the other three, as expected, behaved well but are now in Milwaukee braces and progressing favourably at the ages of eleven, eleven and eight years respectively.
A case of double structural idiopathic scoliosis. In such cases the rib-vertebra angle is greater on the convexity than on the concavity; the difference, usually small, is written as a negative figure to indicate this. Note the obliquity of the eleventh and twelfth ribs on the concave side of the thoracic curve, another strong indication of a double curve.

FIG. 4

A photograph showing plagiocephaly, a characteristic facial deformity associated with infantile idiopathic scoliosis, both resolving and progressive. Seen from above there is hypoplasia of the right cheek and brow. The child would therefore have a curve convex to the right. Note that the left ear is turned forward, a characteristic feature of plagiocephaly.

FIG. 3

Radiographs of a child who had a resolving scoliosis. The R.V.A.D. was 16 degrees. Note that all the rib heads on the convexity were clearly visible, a valuable sign of a resolving scoliosis. Eighteen months later the curve had disappeared.

FIG. 2
without benefit of effective treatment, and there is much previous information from which to gauge the severe prognosis (James 1954; Scott and Morgan 1955). Figure 5 shows a typical case. Figure 6 conveys the expected outcome of progressive curves left untreated. This histogram and the papers just referred to indicate very clearly the poor prognosis of untreated progressive infantile idiopathic scoliosis: by the age of five years 57 per cent have already passed the point of no return, that is a 70 degree curve, with the inevitable cor pulmonale and other irreversible problems.

If an infant is seen with even a very small curve but with a rib-vertebra angle difference exceeding 20 degrees, one must consider this to be progressive. Treatment begins with a hinging plaster jacket until the child is old enough to wear a Milwaukee brace at two to three years of age. A child first seen at two and a half to three years goes straight into a brace. The brace is worn until ten or eleven years, when correction and fusion are usually performed.

There were six children in this series who came before the age of three, had a full attempt at control and were followed to fusion and after, over total periods of more than ten years. The effects on the curvatures after each stage of treatment are recorded in Table III. Only in Case 2 was the final curve after fusion, 70 degrees, greater than that recorded in the first radiograph, 54 degrees, some ten years before. The other five children had smaller measurements after fusion than as infants, which clearly demonstrates that such curves can usually be controlled. Figure 7 illustrates a typical sequence.

Another eight infants treated by plaster and brace, or by brace alone, have not reached the age for fusion. Seven of the curves have stabilised in the brace and from experience are now unlikely to deteriorate (Fig. 8). One curve is worse (Case 6). The details are recorded in Table IV. The first seven children will be maintained in a brace until old enough for fusion, when their curves may be expected to be still further reduced by the corrective procedures undertaken immediately before that operation. Case 8 clearly does not require fusion, though all the evidence suggests that this was a progressive curve.

"Pseudo-cures"

There were ten patients who appeared to have done exceptionally well in plaster and brace, some to the extent of a cure. Now that Mehta's method of classification is available, and with hindsight, these ten would seem to have had resolving curves. The curves were either rather severe and therefore seemed of the progressive type, or more commonly the reason for treatment was a marked increase in the curve.

CONGENITAL SCOLIOSIS

Obviously the vertebral anomalies which cause congenital scoliosis are present in infancy. However, most of the affected infants do not present with curvature in the
Case 3—Infantile idiopathic thoracic scoliosis measuring 61 degrees and with an R.V.A.D. of 45 degrees. The curvature being clearly progressive, the child was put into a hinging plaster jacket and the curve was reduced to 27 degrees eighteen months later. Six years later still, and just before fusion, the curve measured 40 degrees in the Milwaukee brace. Two years after correction and fusion it was 39 degrees.

This child came rather late with a curve of 73 degrees and an R.V.A.D. of 71 degrees. The photograph shows the amount of rotation when he first presented; this will persist and remain very ugly and is a major reason for early treatment. A brace was immediately applied because he was old enough for this. As expected, the correction to 55 degrees is poor but will be maintained at about this level until fusion.
first three years because the irregular growth has not had time to deform the spine. Indeed, congenital anomalies which cause a visible curve of the spine in the first three years usually have an exceptionally poor prognosis, which is very evident from the terrible problems seen in un-

reached the end of growth (Fig. 9). One case needed fusion in adolescence because of late deterioration.

Only six patients have had treatment from infancy to near skeletal maturation, but the results are very instructive (Table V). The prognosis in congenital scoli-

Fig. 9
A case of congenital scoliosis. In 1965 this child had a curve measuring 20 degrees, but seven years later, without treatment, it had reduced to 8 degrees.

Fig. 10
A case of very severe congenital scoliosis, 97 degrees when first seen in 1959, and 111 degrees two years later when the child was old enough for a brace. Some six years later still, the curve, then reduced to 100 degrees, was corrected and fused, with little change as expected. Two years after fusion it measured 95 degrees, only 2 degrees less than some thirteen years earlier but much better than if nothing had been done.

treated cases and which has been very well documented recently (Winter, Moe and Eilers 1968). In this series there were forty-nine such cases to be considered. Nineteen required no treatment during the period of observation, which was usually many years and in some cases osis is not so well defined as in other varieties, but one would have expected all of these six curvatures to get much worse, and none did so (Fig. 10).

As in the idiopathic group, a number of children are under treatment by brace and are apparently stabilised.
A case of high thoracic congenital scoliosis. The first radiograph shows the typical congenital deformities and a curve of 24 degrees, which increased to 53 degrees six years later. The child came to us at this stage showing a very ugly deformity with elevation of the shoulder-neck line from the vertebral rotation.

Fusion is to be performed quite soon.
but not yet old enough for fusion. Most of them have been in a brace for five years or more. All of the eleven such cases have improved (Fig. 11).

The very ugly high thoracic curve is a special case; it must never be allowed to increase over 30 degrees. The child shown in Figure 12 came to us late and has the typical and irreversible raised neck-shoulder line from rotation of the vertebrae.

Kyphoscoliosis with its risk of paraplegia is a special and very important problem which it is not proposed to consider here.

**TABLE V**

<table>
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<tr>
<th>Case number</th>
<th>Initial curve (degrees)</th>
<th>After plaster (degrees)</th>
<th>After brace (degrees)</th>
<th>After fusion (degrees)</th>
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<td>76</td>
<td>59</td>
<td>62</td>
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<td>6</td>
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**SPINA BIFIDA WITH SCOLIOSIS**

In this final group, children with primarily a spina bifida problem are beginning to present with developing scoliosis. Those with paraplegia often develop severe curves, whereas others may have severe but flexible curves. There were thirteen children with a major spina bifida and scoliosis under long enough study to be considered here. Six have progressed slowly and are receiving no treatment; seven are wearing a Milwaukee brace. This brace presents two difficult problems—pressure sores may develop over anaesthetic skin, and the wearing of both a brace and calipers effectively stops walking. A compromise under trial is to use the brace at night and calipers during the day. At present we seem to be as successful as in the other groups, but the difficulties with the brace may well defeat our aims.

**COMMENT**

Despite the arduous demands on all concerned, there is no doubt that one can hold both progressive infantile idiopathic and congenital curves from infancy onwards, and that in the few that do show an increase (in this series only two) the increase is very much less than in untreated cases. These excellent results must encourage all of us to act more vigorously. The alternative may be disaster.

The series reported here is small for obvious reasons, but it has been remarkably and unexpectedly successful. Exceptional cases of failure are bound to occur in both the idiopathic and the congenital groups, and then earlier correction and fusion may have to be performed.

I am grateful to many orthopaedic colleagues who continue to refer these cases, and to Miss R. Wynne-Davies who compiled the figures given in Tables I and II.

**REFERENCES**


