INFANTILE STRUCTURAL SCOLIOSIS

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We are reporting the outcome of structural scoliosis of unknown etiology found in 212 infants before the age of three years. One hundred and fifty-seven infants were from the scoliosis clinic at the Royal National Orthopaedic Hospital and fifty-five were from the Hospital for Sick Children, Great Ormond Street.

In 1936 Harrenstein found a variable, but generally good, prognosis in the forty-six patients with infantile scoliosis that he described; he did not know the etiology but did not classify the scoliosis as idiopathic. In 1951 James described thirty-three patients with infantile idiopathic scoliosis. The term "idiopathic" was then used deliberately as there was no reason to believe the condition to be different from idiopathic scoliosis arising later in childhood. The prognosis was invariably poor.

Since 1951 it has become apparent that there are two types of infantile structural scoliosis, progressive and resolving. The former develops rapidly and relentlessly, causing the severest kind of orthopaedic cripple with dreadful deformity, marked dwarfing and shortening of life. Scott and Morgan (1955) reviewed the prognosis of this progressive type of curve in twenty-eight patients. The resolving type is at first essentially the same, but disappears spontaneously in the first few years of life. Nine such cases were reported at the combined meeting of the Orthopaedic Associations of the English-speaking World by James in 1952.

To confirm the diagnosis of infantile idiopathic scoliosis it is essential to exclude congenital anomalies, either at operation or by seeing an early radiograph before distortion and deformity make the distinction impossible. We have excluded from this series those patients without such confirmation.

### TABLE I

**INFANTILE STRUCTURAL SCOLIOSIS IN 212 PATIENTS**

<table>
<thead>
<tr>
<th>Patients</th>
<th>77</th>
</tr>
</thead>
<tbody>
<tr>
<td>Resolving structural type</td>
<td></td>
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<tr>
<td>Progressive idiopathic type</td>
<td></td>
</tr>
<tr>
<td>Thoracic curves</td>
<td>111</td>
</tr>
<tr>
<td>Double primary curves</td>
<td>13</td>
</tr>
<tr>
<td>Thoraco-lumbar curves</td>
<td>8</td>
</tr>
<tr>
<td>Lumbar curves</td>
<td>3</td>
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<tr>
<td>Total</td>
<td>135</td>
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In all our patients there has been a structural curve with fixed posterior rotation persisting on attempting forward flexion, and in many of the Great Ormond Street patients the radiographs were taken in suspension; by these methods we have, we believe, excluded those postural curves that can be found in any baby (Table I).
Infantile idiopathic progressive scoliosis. A boy with the usual left thoracic curve. The radiograph with a curve of 27 degrees, taken ten years earlier, shows no congenital anomaly and, as can be appreciated, without this the distortion in the present radiograph, with 119 degrees of curvature, would make the exclusion of a congenital scoliosis impossible.
Infantile idiopathic progressive scoliosis. A boy of twelve with a right-sided but otherwise characteristic thoracic scoliosis. The radiograph showing 22 degrees of curvature was taken at eighteen months, and shows no congenital bony anomaly. The radiograph taken ten years later is like those in most of these children at this age if not treated in a Milwaukee jacket, and shows 118 degrees of curvature.
Infantile idiopathic progressive scoliosis. This curve is less common in girls than boys. The initial curve of 14 degrees would be difficult to distinguish from a resolving scoliosis, but very early on the progressive character was apparent, and fifteen years later the curve was 103 degrees.
Infantile idiopathic progressive scoliosis. A further example of a left thoracic scoliosis. This girl is almost mature and was not seen until this age, but an early radiograph showed a curve of 45 degrees; fourteen years later it was 148 degrees. The only two proved cases first seen when mature had curves of over 150 degrees.
Infantile idiopathic progressive scoliosis. These figures show the vicious nature of this deformity. In spite of the innocence of the original radiographs and photographs the condition developed relentlessly into this calamitous deformity. This boy, who has reached maturity, was never treated.
INFANTILE PROGRESSIVE IDIOPATHIC SCOLIOSIS

Thoracic curves—These are the commonest, and were found in 111 patients.

The preponderance in the male, noted by James in 1951, is confirmed, there being sixty-five boys to forty-six girls. As is the remarkable preponderance of ninety left-sided curves (81 per cent) to twenty-one right-sided. This reversal of the side of curvature compared to those with onset in adolescence, where 80 per cent are right-sided, is totally inexplicable.

Five curves were diagnosed at birth, seventy-three appeared in the first year of life, twenty-seven in the second and six in the third. Facial moulding or plagiocephaly on the same side was seen in nineteen patients.

The commonest site of primary curvature, and found in twenty-two patients, was between the fifth and twelfth thoracic vertebrae. The remainder varied from the third and twelfth thoracic vertebrae above to the eighth thoracic and the first lumbar vertebrae below. The number of vertebrae in the primary curve varied between five and eleven, with eight the usual.

The infant with thoracic progressive idiopathic scoliosis is usually a boy with a left thoracic curve. When first seen the curve is likely to exceed 20 degrees and it rapidly becomes worse during the first few years of life (Figs. 1 to 8), and later dwarfing is noticeable. Eleven children had severe interference with growth.

The prognosis is difficult to establish absolutely because these children are now treated with Milwaukee braces and spinal fusions. There are early and late records of only two children who have reached skeletal maturity without operation; they had curves of over 150 degrees (Figs. 9 and 10). The remaining 109 children have either not reached maturity or have had major treatment; thirty had Milwaukee jackets applied, and thirty-one were subjected to operation because they were deteriorating so rapidly that effective treatment was indicated to halt the natural development of the curve.

We have attempted to establish the prognosis from the greatest angle recorded in these children before treatment was started, or, in the youngest, by noting the present angle. We shall never again see this deformity develop unaltered—as we have done in the past few years—because of the lack of treatment during the war and later because of our inability to affect the course of the deformity.

PROGNOSIS OF INFANTILE THORACIC CURVES

The prognosis of infantile thoracic scoliosis by age groups.

![Graph showing the prognosis of infantile thoracic curves by age groups.](image)
The antero-posterior radiographs show the development over seven years of a typical infantile idiopathic progressive thoracic scoliosis. The lateral radiographs show the not uncommon development of a marked kyphosis, which is not seen in other varieties of idiopathic scoliosis.
Figure 11 shows the prognosis of infantile progressive idiopathic thoracic scoliosis. Forty-seven children were aged five years or less; four of them had developed curves of more than 100 degrees, twenty-three between 70 and 99 degrees and only twenty children still had curvature of less than 70 degrees.

In the group aged six to ten years there were thirty-seven children; fourteen had curves of more than 100 degrees, thirteen between 70 and 99 degrees and only ten children still had a curvature of less than 70 degrees.

In the group aged eleven years to maturity there were twenty-three children—the smallest group—because few reached this age without surgery or bracing; twelve had curvatures of more than 100 degrees, nine had curves of 70 to 99 degrees and only two children had reached the age of eleven with curves of less than 70 degrees. In one child the curve increased only 1 degree in six years.

Thus it is shown that the almost inevitable result of infantile progressive idiopathic thoracic scoliosis is the development of a curvature of more than 70 degrees, but most curves, if left untreated, will progress to more than 100 degrees.

In a lateral radiograph kyphosis is often apparent; it is appreciated that this may not always be a true kyphosis because the extreme vertebral rotation gives an antero-posterior view in the lateral projection, but the patients do appear hump-backed (Figs. 12 and 13).

**Combined thoracic and lumbar curves**—In thirteen patients two primary curves were found. Thoracic curves appearing from the age of one and a half to three years which develop slowly should be watched; a lumbar primary curve may appear later but both may be there initially (Fig. 14). The prognosis is considerably better than that of the single thoracic curve.
A father and son with similar curves. The age of onset in the father is uncertain, and, if it is really an infantile scoliosis, his curve is uniquely benign.
Thoraco-lumbar and lumbar curves—In ten years we have seen eight thoraco-lumbar and three lumbar curves. The series is small, and none is very mature but all are progressing. It would seem likely that even at this lower level during the years in which growth occurs there will develop a very serious curvature. One patient (Figs. 15 and 16) presented recently with a progressive lumbar curve. His father then stated that he himself had had a curve since infancy, the precise age at onset being unknown. This had remained slight, and may not be of the same type. In no other patient is there a family history of scoliosis.

INFANTILE STRUCTURAL RESOLVING SCOLIOSIS

In this series there were seventy-seven cases of infantile structural resolving scoliosis. Its true frequency is, we believe, even greater than seventy-seven out of 212, for 77 per cent of the patients from Great Ormond Street had this curvature. The scoliosis clinic at the Royal National Orthopaedic Hospital has many of the more serious curvatures referred by other surgeons. Children with small curves treated by splints have been excluded.

The main characteristic of this curve is its spontaneous disappearance without treatment, usually in the first few years of life. The other features are that there are more boys than girls, in the ratio of fifty to twenty-seven; there were sixty-five left-sided curves and twelve right-sided. Seventy-two of the curves were thoracic.

Only one of seventy-seven curves was noticed at birth, but seventy-three occurred in the first year of life, with the highest incidence between the fourth and tenth months. It was...
Fig. 18
Infantile structural resolving scoliosis. This curve, although structural clinically, is radiologically more like a postural curve although rotation can be seen. Note the absence of compensatory curves, a fact common to all resolving scolioses.

Fig. 19
A characteristic structural resolving scoliosis.
Infantile structural resolving scoliosis. During resolution it is the lateral curvature which disappears first, leaving the rotation as seen in the radiograph on the left. Gradually this rotation also disappears.

This demonstrates the difficulty of distinguishing between the two groups because the radiograph on the left with a curve of 17 degrees fulfills the characteristic criteria of the resolving curves. The radiograph on the right was taken at the age of four, and shows a rapid deterioration.
difficult to know the age when the curve disappeared, but twelve did so in the second year of life and twenty-eight out of thirty recorded had disappeared by five years of age.

The largest angle of curvature was 37 degrees and slightly more than half had a maximum curve between 11 and 20 degrees. Thirty patients showed facial moulding or plagiocephaly of the skull and face of the same side.

The characteristic structural resolving curve therefore appears in the first year of life but not at birth, the angle is usually between 10 and 30 degrees and is left thoracic in type. Compensatory curves were never seen (Figs. 17 to 19).

The lateral curvature disappears first, leaving rotation without lateral curvature: this always disappears later but may be slow to do so (Fig. 20).

**DIFFERENTIAL DIAGNOSIS**

How are we to differentiate the earliest stages of these two curves, the one so innocent, the other so malignant? There is so far no absolute distinction, but one can often prognosticate correctly. The typical resolving curve is single and always without compensatory curves, whereas only seven progressive curves were initially seen with no compensation (Figs. 21 and 22). The presence of compensatory curves seems to be an absolute indication of permanency. It has been suggested that the resolving curve is usually longer than the progressive, but although our longest curve was resolving so was our shortest.

In summary, long curves of less than 35 degrees and without compensation will probably resolve, but it must be emphasised that there is no absolute distinction and there is much to suggest that initially the two types are identical.
Figure 23—The macroscopic appearance of the spine of a child who died at the age of eleven months, with a normal spine alongside. Figure 24—The radiographs of the two spines in Figure 23. There is no congenital anomaly in the affected spine.

Figure 25
Radiographs of sections of the eighth thoracic vertebrae from the spines shown in Figure 23 showing no abnormalities of epiphysial or bone structure.
A girl with 118 degrees of curvature at the age of six and a half years was put into a Milwaukee jacket. Nearly four years later—just before operation—the curve measured 80 degrees.

**ETIOLOGY AND PATHOLOGY**

Until recently no pathological material had been examined and radiographs were the only method of excluding a congenital scoliosis.

We have investigated the spine of a child aged eleven months who died and who was known to have developed scoliosis at eight weeks which had got worse. The curve was shown in a radiograph in suspension taken at eight weeks of age.

Macroscopic examination showed no evidence of congenital anomaly (Fig. 23) and the radiograph of the resected vertebral column showed rotation only (Fig. 24). Microscopically, sections of the vertebral epiphyses were reported as normal by Dr H. A. Sissons. Radiographs of the sections are shown in Figure 25.

If we could establish that the resolving and progressive curves were initially the same, discovery of the cause of resolution might lead to an understanding of the etiology of idiopathic scoliosis.

**TREATMENT**

It is not our purpose to discuss treatment in detail, but a brief outline is worth while. Operative correction and fusion are delayed until ten years of age; almost all require it. Until the age of ten a Milwaukee jacket has been found an excellent method of preventing deterioration (Fig. 26).

In the earliest stages various methods of arresting or correcting the curve have been tried, including the Mürk Jansen lateral plaster bed and plaster jackets applied after correction. In clearly defined progressive cases there was always a deterioration. At present the method under trial is Risser jacket correction followed by the application of a plaster jacket until the child is old enough to wear a Milwaukee jacket, at about the age of
three and a half years. Before this age the lack of cooperation from the child and the absence of a waist makes this jacket impracticable. Although it might be expected that in infants these small curves might be fully correctable, our experience so far is that in none of them were we able to get rid of the curve completely. Whether this method of a Risser jacket followed by plaster jacket is adequate to prevent deterioration is still undetermined.

In a number of children with this type of progressive curve, who were treated by a Milwaukee jacket between the ages of three and a half and ten years, there has in no instance been any deterioration. In most there has been some correction ranging from 40 to 10 degrees. The Milwaukee jacket has, over the last five years, proved a very satisfactory method of preventing deterioration of these curves for anything up to five years and of enabling operation to be delayed until the child is older.

It is not proposed to analyse the results of treatment now, but it may be said that there are many disappointments and that no type of scoliosis has proved more resistant to treatment than this group. Operation has been attempted only twice before the age of ten and in both cases it was unsuccessful. However, a number of patients treated at the age of ten have gained good results, and it is believed that the age limit for operation could safely be lowered.

We would like to acknowledge the excellent photography of Mr R. J. Whitley, Principal Photographer, the Institute of Orthopaedics.

REFERENCES


