JUVENILE IDIOPATHIC SCOLIOSIS

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A series of 98 patients with juvenile idiopathic scoliosis have been analysed. This showed that between the ages of four and six there was a higher incidence in boys whereas between seven and nine years of age, the proportion of girls was higher. Regardless of sex and age the majority of the curves were convex to the right and the single thoracic curve was the commonest pattern. Spontaneous resolution occurred in seven patients: in four the curves resolved within two years; in the three others the curves resolved in three, four and five years respectively. Forty-four per cent of all patients were managed conservatively and in 56 per cent spinal fusion was carried out.

The characteristics of an idiopathic scoliosis first appearing between four and nine years of age and designated “juvenile idiopathic scoliosis” are not dissimilar from those of scoliosis of later onset but, because of the different prognosis, separation from the other groups of “infantile” and “adolescent” is now generally accepted (James 1954; Wynne-Davies 1968; James 1976). Despite the great progress made in the study of scoliosis over the past 20 years, little has been said of juvenile idiopathic scoliosis as an entity. The series previously published differ regarding the sex incidence, the area and the side of the curves.

Ponseti and Friedman (1950), in a review of 335 cases, found 13 per cent under the age of 10. James (1954), in a series of 134 patients, described 16 with juvenile idiopathic scoliosis, a majority with severe curves. Moe and Kettleson (1970), analysing 169 patients with idiopathic scoliosis, noted that 26 (1 boy and 25 girls) were in the juvenile group. Keiser and Shufflebarger (1976) reported 20 patients with juvenile idiopathic scoliosis out of 123 patients. There may be differences in the incidence in Great Britain and in North America.

The series presented here covers exclusively the juvenile age range.

MATERIALS AND METHODS

From 114 patients with an onset of idiopathic scoliosis between the ages of four and nine, seen between 1951 and 1979 at the Princess Margaret Rose Orthopaedic Hospital, Edinburgh, 98 were available for this study. The average follow-up period was 4 years 10 months (ranging from 1 year to 25 years 3 months). Clinical notes and radiographs were reviewed and several features peculiar to this group identified.

According to the progress of the curve and type of treatment carried out, the patients were divided for convenience into two groups. In Group I were the 27 patients who were merely observed, for an average of 5 years 3 months (ranging from 1 to 12 years); these were subdivided into Group IA, patients whose curves remained stationary or showed slight progression (up to 10 degrees), and Group IB, patients whose curves resolved. Group II comprised the patients who required treatment: Group IIA were patients who were managed by bracing only, and Group IIB the 55 patients who had operative treatment, with or without previous bracing or localiser jacket. The brace used in Edinburgh is a modification of the Milwaukee brace (Blount and Moe 1973): having no lateral pads pressing on the curves, the brace relies on distraction between the two girdles (McMaster and Macnicol 1979). Of the patients who were operated upon, those seen most recently received Harrington rod instrumentation to supplement fusion by the interfacial technique (Moe 1958).

The curves were measured by Cobb’s method (Cobb 1960) and were classified, according to the location of the apex of the major curve (also defined by Cobb), as thoracic, thoracolumbar (with the apex at T12), lumbar, and double (when there were two structural curves). The rotation seen on clinical examination was considered to be of great importance (James 1954) and was measured by the technique of Nash and Moe (1969). The rib–vertebra angle difference (RVAD) was also measured (Mehta 1972).

RESULTS

Out of the 98 patients reviewed, 32 were male and 66 female (Fig. 1), an overall ratio of 1:2. However, the proportion of these children presenting between the ages of four and six was higher among boys whereas

![Figure 1](image)

Number of children with juvenile idiopathic scoliosis seen at the Princess Margaret Rose Orthopaedic Hospital between 1951 and 1979.
between the ages of seven and nine, the frequency in girls was higher (Fig. 2), there being a significant difference between the two age groups. The curves were right sided in 77.5 per cent of all cases, with no relationship to sex or age (Fig. 3).

Four patients had curves which resolved within two years; in three others, the curves resolved in three, four and five years respectively.

The major curves
The single thoracic pattern was the most common in this series (62 per cent) followed by double thoracic and lumbar (22 per cent), thoracolumbar (15 per cent) and lumbar (1 per cent) (Fig. 4).

Single thoracic curves. Sixty-one patients (23 male and 38 female) presented with this curve pattern. The average age when first seen was 8 years 6 months. The curve was convex to the right in 47 patients and to the left in 14. The curves variously extended from T2 to L3, the apex occurring at T9 in 41 per cent of patients. The average number of vertebrae in the curves was eight, and the curvature ranged from 10 to 122 degrees. In the severe cases, extreme vertebral rotation gave the patients a humpback appearance (James, Lloyd-Roberts and Pilcher 1959).

The classic textbook picture can be seen in this type of scoliosis, the ugliest of all, with drooping of the shoulder, a prominent hip on the side of the concavity and a humpback (Figs 5 to 9).

Thoracolumbar scoliosis. This type of curve occurred in 15 patients (3 male and 12 female). In nine patients the curve was convex to the right and in the six to the left. The curves variously extended from T4 to L4 with an average of nine vertebrae (range 7 to 11). The apex in all cases was at T12. Eighty per cent of patients had curves under 70 degrees, the average being 27 degrees. A drooping shoulder and, particularly, a prominent hip are obvious though less unsightly than in thoracic scoliosis.

Double thoracic and lumbar curves. The second most frequent pattern in this series, with an incidence of 22 per cent, was a double curve with thoracic and lumbar scoliosis. Of the 21 cases seen, 15 patients were boys and six girls. In every case but one the thoracic curve was convex to the right and the lumbar curve was convex to the left. The most frequent site of the apex in the thoracic curves was T8 and in the lumbar L1 (12 cases each). Usually both thoracic and lumbar curves had the same number of vertebrae, the average being six in each curve. The average curvature when first seen was 39 degrees in the thoracic curve and 34 degrees in the lumbar.

The rib–vertebra angle difference at the apex of the thoracic curve was low or negative: that is, the angle between the rib and the vertebra on the apex was greater on the convex side than on the concave. That a negative RVAD may indicate a double curve has been pointed out by Ferreira and James (1972).
Lumbar curves. There was only one lumbar curve in this series. This was in a girl aged 11 years 7 months with a 17-degree curve convex to the left and extending from T9 to L3 with the apex at L1. She was kept under observation and at the age of 18 years the curve was 35 degrees with very little deformity.

Family history
There is a significant proportion of families in whom idiopathic scoliosis appears among several members and it has been suggested that scoliosis may be produced by genetic factors (Wynne-Davies 1968; MacEwen and Cowell 1970; Riseborough and Wynne-Davies 1973). In this study 13 per cent of all patients gave a history of family involvement.

Associated diseases
Mental deficiency was the most common association observed in this study. There were 12 affected among the 98 patients; four were male and eight female. This is a common association with idiopathic scoliosis but most frequent in those with a juvenile onset.

Epilepsy was found in two per cent of the patients.

Other abnormalities observed were inguinal hernia (three cases), spina bifida occulta (six), pyloric stenosis (two), congenital dislocation of the hip, dextrocardia, metatarsus varus, joint laxity, hemimelia and absent thumb (one case each).

Progression and treatment
As noted already, the patients were divided into two groups. One patient was included in both groups because she developed two curves at different times: the first curve resolved spontaneously and the other, appearing a couple of years after the first, progressed and was treated initially by bracing and later by spinal fusion.

Group I (27 patients). Attendance at the scoliosis clinic
allowed these patients to be observed and their progress assessed clinically and radiologically until skeletal maturation.

Group IA (20 patients). In 8 per cent the curve remained stationary and in 12 per cent the progress was less than 10 degrees throughout growth. Six of them had a thoracic scoliosis. Six girls had thoracolumbar curves, four convex to the right and two to the left. Seven patients (two boys and five girls) had double curves—right thoracic and left lumbar. One girl had a left lumbar scoliosis. The average age of onset was 5 years 6 months.

Group IB (seven patients). These curves resolved. Four patients (three boys and one girl) had thoracic curves, two being left-sided and two right-sided. Three patients (one boy and two girls) had thoracolumbar scoliosis, one right-sided and two left-sided. The average age of onset was 6 years 1 month. The maximal size of the curves varied from 10 to 35 degrees and the average follow-up was 5 years 11 months (range 2 years to 10 years 2 months). One boy, first seen at the age of 4 years 2 months, with a 35-degree right thoracic curve extending from T5 to L1 and a negative RVAD of 4 degrees,
experienced a decrease in curvature to 10 degrees within two years. It remained at 10 degrees until the end of growth.

One girl had a structural thoracolumbar scoliosis to the left which disappeared spontaneously. Two years later she developed a thoracolumbar curve to the right, exactly at the same level and this progressed. Her case history is now given in detail.

This girl was first seen at the age of 5 years 4 months with a left thoracolumbar scoliosis extending from T8 to L3 and measuring 23 degrees; it had first been noticed at the age of four years (Fig. 10). Her progress was typical of the severe cases. The RVAD was 6 degrees at the apex of the curve yet within 2 years 3 months the curve had resolved completely (Fig. 11). At the age of 9 years 1 month she developed a thoracolumbar curve, convex to the right, extending exactly as before from T8 to L3, measuring 10 degrees and with an RVAD of 10 degrees (Fig. 12). The curve progressed and one year later measured 23 degrees (Fig. 13). She was then fitted with a brace and one year later, after having worn the brace full-time, the curve was 27 degrees with an RVAD of 13 degrees. By that time T7 had been included in the major curve. She wore the brace for 3 years 4 months, by when the curve measured 27 degrees (Fig. 14). At the age of 13 years 8 months her spine was fused from T6 to L3 and the curve corrected to 15 degrees (Fig. 15). At routine revision of the fusion, six months later, no pseudarthrosis was found. Nevertheless, after one year from fusion the curve measured 30 degrees. Ten months after the revision the curve had deteriorated to 40 degrees. Due to this loss of correction there was a suspicion of non-union and a bone scan showed an increased uptake at the level of T11 and T12 which was highly suggestive of a pseudarthrosis. She was re-explored and a bilateral hairline pseudarthrosis was found at the level of T11 and T12. Repair of the pseudarthrosis was carried out and the curve corrected by means of Harrington instrumentation. At the end of growth the curve measured 27 degrees and her appearance was good (Figs 16 and 17).

Group II (72 patients). This group were treated either with a brace alone or by spinal fusion with or without a preliminary brace.

Group IIA (17 patients). These patients were managed in the Edinburgh brace only; six of them still remain under observation as skeletal maturation has not been completed. Treatment began with full-time wearing of the brace, which was removed only for bathing. The patients were seen at regular intervals at the scoliosis clinic and the time in the brace was gradually reduced according to the maintenance of correction. The average age at bracing was 8 years 3 months. The average follow-up was 4 years 1 month (range 1 to 16 years). The mean correction was 4 degrees with an average of 37 degrees before bracing and 34 degrees afterwards. One patient with a 30-degree single right thoracic scoliosis extending from T3 to T11 and with a negative RVAD of 7 degrees before bracing had developed, after two years in the brace, two smaller curves extending from T5 to T11 and from T11 to L4 measuring 28 and 20 degrees respectively.

Group IIB (55 patients). These patients (56 per cent of those reviewed) had operative treatment. Twenty-eight (12 boys and 16 girls) had been managed in the Edinburgh brace before operation. The average age at bracing was 7 years 2 months; the average time of bracing was three years. The patients had curves measuring, on average, 43 degrees before bracing and 34 degrees afterwards, a mean correction of 21 per cent. In the double-curve pattern the mean correction at the end of bracing was 40 per cent for the thoracic and 43 per cent for the lumbar curve. One patient who had a single thoracic curve initially, changed pattern to a double thoracic scoliosis after wearing the brace for 1 year 8 months.

Twenty-seven patients (8 boys and 19 girls) were treated surgically without previous bracing. The average age at operation was 11 years 10 months. The earlier cases in the series (39 patients) were corrected in a localiser jacket before spinal fusion. In the later cases (19 patients) Harrington rod instrumentation was used and gave a mean correction of 74 per cent. The final loss of correction in these later patients was 7 degrees as opposed to the ones who did not have instrumentation whose final loss was 17 degrees at the end of growth.

All patients were followed up until skeletal maturation, an average of 7 years 5 months; the mean follow-up after operation was 4 years 10 months.

COMPLICATIONS

With brace treatment. Pressure sores complicated brace treatment in eight patients but responded to adjustment of the brace. It was therefore possible to continue treatment. Protrusion of the teeth was observed in two patients; correction was achieved after appropriate orthodontic treatment, though it also occurs spontaneously when growth potential remains.

After operation. Pressure sores due to plaster jackets occurred in six patients: all healed after trimming or temporary removal of the plaster. Infection was a problem on two occasions, Staphylococcus aureus being isolated from an iliac crest donor site in one patient and in the discharge from a revision wound in the other; both lesions were treated with appropriate chemotherapy and healed.

At routine revision six months after spinal fusion, pseudarthrosis was found in three patients. One had bilateral non-union at the level of T12–L1 which was repaired by grafting. In two other patients bilateral hairline pseudarthroses were found: in one at the level of T12–L1 and in the other where the attempt at fusion had failed at two levels, T8–T9 and T11–T12. A rapid loss of correction of 15 to 20 degrees may be an indication of pseudarthrosis (McMaster and James 1976). A hairline pseudarthrosis cannot be seen radiologically, indeed in one case in this series such non-union was not apparent at the first revision of a spinal fusion.

DISCUSSION

Several special features were noted in 98 children with idiopathic scoliosis occurring in the juvenile years.

The overall sex ratio was one boy to two girls. It is evident, however, that between the ages of four and six there is a higher incidence in boys whereas the
proportion of girls is higher between seven and nine years of age (Fig. 2). Regardless of sex and age the majority of the curves were convex to the right (Fig. 3). Wynne-Davies (1973) in her study of idiopathic scoliosis found that the sex ratio and side of the curve were equal in the middle years of childhood. Tolo and Gillespie (1978) studying 59 patients with juvenile idiopathic scoliosis found that girls were affected more than boys in a proportion of 4.4:1, being equal at the age of four to six; in their series there were no boys between seven and eight, and in the children of nine and 10 the ratio was 8.7:1.

It is interesting to notice that idiopathic scoliosis is virtually absent in the United States of America under the age of six years whereas in Europe, particularly in Britain, the number of patients with infantile and juvenile scoliosis is very high. In Edinburgh it equals or slightly exceeds those with an onset in adolescence.

The single thoracic curve was found in 62 per cent of all cases and was the most common pattern. Double curves were present in 22 per cent of patients. Fifteen per cent had thoracolumbar scoliosis. Only one patient in this series had a lumbar curve. A few patients with single thoracic scoliosis later developed double-curve patterns after wearing the brace. A single thoracic curve when first seen with a low or negative RVAD value should be suspected of being a potential double curve. Cervicothoracic and multiple curves (Ponseti and Friedman 1950; Travaglini 1975) were not seen in this series.

Spontaneous resolution of a structural curve, as described in the infantile group of scoliosis, was observed in seven cases. Bjerkreim (1977) described one case of a 10-year-old girl who, after four years of spontaneous correction of a previous curve, developed a progressive structural scoliosis which took six years to resolve.

Mental deficiency and epilepsy were the commonest diseases associated with the scoliotic patients in this survey.

Forty-four per cent of all patients were managed conservatively. During bracing special exercises were not prescribed though normal activity was encouraged. Fifty-six per cent of the patients had a spinal fusion, a third of them with Harrington instrumentation. It has been a policy in Edinburgh to carry out spinal fusion in all progressive thoracic curves at about the age of 10 years, so avoiding the inconvenience of wearing the brace for a long and uncertain period of time which can be so distressing to both parents and children.

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REFERENCES


