THE MANAGEMENT OF PROGRESSIVE INFANTILE IDIOPATHIC SCOLIOSIS

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Twenty-seven children with progressive infantile idiopathic scoliosis have been reviewed after long-term management. Twenty-two children had single thoracic curves which were diagnosed at an early age and treated in a modified Milwaukee brace until the age of ten years when the spine was corrected and fused. The mean correction after operation was 40 per cent of the initial degree of curvature seen in early childhood before treatment. Solid spinal fusion led to a further moderate loss in correction due to bending of the fusion mass before the spine became stable several years later. Five children had double structural curves and were treated only in a brace. This provided less satisfactory control of these curves but because of the minimal cosmetic deformity, extensive spinal fusion was avoided.

Infantile idiopathic scoliosis is a structural curve appearing without evident cause in a child before the age of three years (James 1951). Although rare in North America it is not uncommon in Great Britain; in a series of 200 consecutive patients presenting with idiopathic scoliosis at the Edinburgh Scoliosis Clinic between 1968 and 1972, eighty-two were of the infantile variety. Lloyd-Roberts and Pilcher (1965), in a study of 100 infants with infantile idiopathic scoliosis presenting in the first year of life, found that 90 per cent resolved spontaneously within two years; but in an earlier series of 212 children diagnosed before the age of three years and referred to a scoliosis clinic, only 77 per cent resolved (James, Lloyd-Roberts and Pilcher 1959). In the early stage both resolving and progressive types of infantile idiopathic scoliosis appear as virtually identical mild structural thoracic curves. However, with growth and if left untreated, progressive curves deteriorate to become one of the most severe forms of scoliosis (Figs. 1, 2 and 3). At skeletal maturity the majority of thoracic curves will measure over 100 degrees and the associated thoracic cage deformity results in cor pulmonale and probably in death in early adult life (Scott and Morgan 1955). A small minority of progressive thoracic curves that do not deteriorate so rapidly may later develop a second structural curve to the opposite side in the lumbar region; a very few patients may have both these curves present from the onset. The prognosis for these double structural curves is considerably better than that for a single thoracic curve.

The Edinburgh policy in the management of progressive infantile idiopathic scoliosis is to recognise the progressive curve early and start immediate conservative treatment. This management continues, if possible, until the child is ten years old, when the spine is corrected and fused posteriorly. This policy applies to all patients with single thoracic curves but not necessarily to double structural curves in which the thoracic and lumbar components are usually well balanced and the cosmetic deformity is not great. Correction and fusion of double curves produces little cosmetic improvement and the resulting extensive spinal rigidity can be disabling. These curves are therefore managed conservatively until skeletal maturity unless the thoracic component deteriorates to the stage where future cardiothoracic complications become likely.

The purpose of this paper is to report the Edinburgh experience in the long-term management of progressive infantile idiopathic scoliosis diagnosed at an early age and treated by a combination of brace therapy and spinal fusion.

MATERIALS AND METHODS

A retrospective study has been made of twenty-seven children with progressive infantile idiopathic scoliosis who were first diagnosed at an early age at the Edinburgh Scoliosis Clinic and have undergone long-term treatment between 1964 and 1977. The many children who were treated elsewhere before being referred to Edinburgh at a later stage for further treatment have been excluded. In the earlier patients the diagnosis of a progressive as distinct from a resolving scoliosis was made by noting a radiological progression of the curve. Since 1968 a much earlier diagnosis has been made possible by estimating the rib-vertebra angle difference at the apex of the curve on the first spinal radiograph (Mehta 1972). Once the diagnosis of a progressive curve has been made, the likely prognosis and plan of treatment is discussed with the parents. The need for measures to prevent deterioration is emphasised and the child is fitted with a brace. The parents are informed that single thoracic curves will require correction and fusion at the age of ten years as part of the planned course of treatment and not because of the failure of the brace to control the deformity.

The brace which has been used in Edinburgh is a modification of

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the Milwaukee brace (Blount, Schmidt and Bidwell 1958) and can be fitted as early as eighteen months of age once the child has developed a waist. If the curve is already greater than 40 degrees, preliminary correction in a Risser turnbuckle jacket is often carried out. The brace used in Edinburgh does not make use of the pads which have been advised by Blount and his associates as a means of applying a lateral corrective force to the site of greatest curvature. Instead, the Edinburgh brace relies only upon controlled distraction between the pelvic girdle and the mandible and occiput. The brace is worn day and night and is removed only for washing and bathing. Regular attendances are made at the Scoliosis Clinic where the brace is distracted and the progression of the curve is assessed radiographically.

Twenty-two children with single thoracic curves were reviewed; all had completed their treatment in a brace and had, at the age of ten years, undergone further correction in either a Risser localiser jacket or by Harrington instrumentation and posterior spinal fusion. Of the remaining five children with double structural curves, two were still being managed in a brace at the time of review, while the other three were skeletal mature and bracing had been discontinued. Erect spinal radiographs of each patient were measured using Cobb’s method (Cobb 1960) and the rib–vertebra angle difference calculated from the earliest radiograph (Mehta 1972). Radiographs taken elsewhere before referral were often available and gave an indication of the rate of progression of the untreated curve.

RESULTS

Single thoracic curves

There were twelve boys and ten girls in this group with a mean rib–vertebra angle difference of 43 degrees (range 22 to 100 degrees) on the first spinal radiograph taken before the age of three years. Mehta (1972) has suggested that rib–vertebra angle difference greater than 20 degrees is indicative of a progressive curve and all of the thoracic curves in this group were seen to progress. The curves extended from an upper limit of the fourth thoracic vertebra to a lower limit of the third lumbar vertebra and varied from seven to eleven vertebrae in length. Left-sided curves were twice as common as right-sided curves.

Single thoracic curves can be managed in three stages but the first stage was not carried out in thirteen patients.

Risser turnbuckle jacket. Nine patients, four boys and five girls, whose scoliosis when first seen was greater than 40 degrees, were corrected initially in a Risser turnbuckle jacket. The age at which the jacket was applied ranged from eighteen months to thirty months, and the mean initial curve of 62 degrees (range 40 to 90 degrees) was decreased by an average of 52 per cent to a mean curve of 29 degrees (range 15 to 40 degrees). After correction those infants who were too young to be fitted with an Edinburgh brace were managed temporarily in a plaster jacket.

Edinburgh brace. All twenty-two children were managed in the Edinburgh modification of the Milwaukee brace which was applied either initially or after preliminary correction in a Risser turnbuckle jacket.
After removal of the Risser jacket there was often some deterioration of the curve before the brace was applied. The mean age at bracing was four years three months (range one year nine months to eight years six months) and twelve out of the twenty-two children (55 per cent) were braced by their fourth birthday. The mean period in the brace was six years five months and 70 per cent of children wore the brace for more than five years.

Table 1. Edinburgh brace treatment of infantile idiopathic progressive thoracic scoliosis (twenty-two patients)

<table>
<thead>
<tr>
<th>Initial curve (degrees)</th>
<th>Number of patients</th>
<th>Brace correction</th>
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<tbody>
<tr>
<td></td>
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<td>Best (per cent)</td>
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<tr>
<td>0–45</td>
<td>6</td>
<td>41</td>
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<tr>
<td>46–75</td>
<td>9</td>
<td>45</td>
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<td>76–105</td>
<td>7</td>
<td>39</td>
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The mean initial curve just before bracing of all children with a single thoracic scoliosis was 63 degrees (range 29 to 105 degrees) and the mean percentage correction after bracing for one year was 38 per cent (Fig. 4). The mean best correction was 43 per cent and this was achieved by one year in eleven patients, by three years in eighteen patients and in all patients by five years. The greatest correction was obtained in those patients whose initial curves ranged from 46 to 75 degrees and the least correction in curves over 76 degrees (Table 1). There was no significant correlation between the correction achieved and the length of the curve or the rib–vertebra angle difference.

After the initial best correction there was then a gradual deterioration of approximately one degree per year. The average progression of untreated single thoracic curves before referral to the Edinburgh Scoliosis Clinic was 12 degrees annually, although this figure varied from 6 to 20 degrees. After wearing the brace for five years the rate of deterioration became greater and the final mean correction in the brace at the age of ten years just before spinal fusion was 11 per cent (Fig. 4); this represents a mean angle of curvature at the end of bracing of 55 degrees (range 20 to 78 degrees). The larger initial curves, however, maintained a better percentage correction than lesser curves (Table 1).

**Spinal fusion.** All single thoracic curves were routinely fused at the age of ten years. In the first fifteen patients correction in a Risser-type of localiser cast and fusion resulted in a mean correction of 40 per cent; this represented an improvement from a mean angle of 71 degrees out of the brace before operation to 42 degrees afterwards. A greater mean correction of 53 per cent was achieved in the last seven patients by using Harrington instrumentation: a mean improvement in the curve from 62 degrees out of the brace before operation to 30 degrees afterwards.

After operation the degree of curvature in twenty out of twenty-two patients was less than that measured on the radiograph taken in infancy and before the application of the Risser turnbuckle jacket or Edinburgh brace. The two patients whose curves were still slightly greater after operation had both been corrected in a Risser localiser jacket. The mean correction after operation for the twenty-two patients was 40 per cent of the degree of curvature when first seen in early childhood before treatment (Fig. 4).

Of the fifteen patients whose curves were treated in a localiser cast three were later found to have pseudarthroses which were repaired at periods varying from nine to thirty-three months after the attempted fusion. All patients have now been followed for an average of seven years (range three to ten years) and in those patients with a solid fusion the mean loss in correction after one year was 6 degrees and at final follow-up was 17 degrees. In the three patients with pseudarthroses the mean loss in correction after one year was 3 degrees and at final follow-up was 27 degrees.

The seven patients whose curves have been corrected by means of Harrington instrumentation have been followed for an average of twenty-one months (range fifteen to forty-two months). All have been surgically re-explored at six months and no pseudarthroses found. The mean loss in correction at one year was 5 degrees and at final follow-up there had been no further loss.

Fig. 4

A histogram showing the mean degree of correction obtained during brace therapy and after spinal fusion of twenty-two single thoracic curves as compared with the degree of curvature when first seen in early childhood and before treatment.

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The following two case reports illustrate the treatment and results obtained in two patients, one with a mild and the other with a severe thoracic scoliosis.

**Case 1.** This boy's mother first noticed that his spine was not straight when he was a few months old. She was reassured by her local doctor but became dissatisfied when at the age of two years and ten months she noted that the deformity had increased. The child was then seen at the Edinburgh Scoliosis Clinic where he was found to be normal except for a mild left thoracic scoliosis with an associated rib hump (Fig. 5) and left plagiocephaly. Radiographs revealed a left thoracic scoliosis extending from the fourth thoracic vertebra to the first lumbar vertebra and measuring 30 degrees with a rib–vertebra angle difference of 37 degrees (Fig. 6).

The child was immediately fitted with an Edinburgh brace which he wore until he was nearly ten years old. The best correction in the brace was achieved at the age of five years and nine months when the curve measured 17 degrees (Fig. 7). Good correction was maintained until the age of eight years and five months when the curve began to deteriorate. At the age of nine years and seven months, when the curve measured 47 degrees in the brace (Fig. 8), a posterior spinal fusion with Harrington instrumentation was performed. The curve measured 51 degrees out of the brace before operation and was corrected to 20 degrees without complication. The child has now been followed for over one year with only a 5 degree loss in correction (Figs 9 and 10).

**Case 2.** This girl was first seen at the Edinburgh Scoliosis Clinic at the age of two years. She had been delivered by Caesarean section and had developed normally until the age of eighteen months when a rib hump was first noted by her mother. On examination the infant was found to be normal except for a severe left thoracic scoliosis with an associated prominent left rib hump and a left-sided plagiocephaly. Radiographs revealed a mild thoracic scoliosis which extended from the fifth thoracic vertebra to the first lumbar vertebra and measured 78 degrees. The rib–vertebra angle difference was 50 degrees (Fig. 11).

She was admitted to hospital where a Risser turnbuckle jacket was applied and the deformity corrected to 35 degrees. Two months later a moulded plaster jacket was applied (Fig. 12) and maintained until the age of two years and eight months when she was fitted with an Edinburgh brace. The best correction in the brace was achieved at the age of four years and eleven months when the curve measured 45 degrees (Fig. 13). Good correction was maintained until she was aged eight years six months when the curve slowly began to deteriorate. At the age of ten years when the curve measured 67 degrees in the brace (Fig. 14) she underwent a posterior spinal fusion with Harrington instrumentation. Before operation her curve measured 84 degrees out of the brace and this was corrected to 41 degrees without complication. The patient has now been followed for over one year with only a 3 degree loss in correction (Fig. 15).

**Double curves**

There were three boys and two girls in this group. In all cases the rib–vertebra angle at the apex of the convexity of the thoracic curve was greater than that on the concavity so producing a negative rib–vertebra angle difference; this ranged from −9 to −50 degrees (mean...
-22 degrees). Mehta (1972) found that a negative rib-vertebra angle difference at the apex of the thoracic curve was often indicative of a second structural curve in the lumbar region and this proved so in all these patients. The upper curves extended from the sixth to the twelfth thoracic vertebra and the lower curves from the tenth thoracic to the fifth lumbar vertebra. The mean number of vertebrae in each curve was six. There were three patients with left thoracic and right lumbar curves and two with the reverse pattern. The mean initial thoracic curve measured 45 degrees (range 34 to 55 degrees) and the mean initial lumbar curve measured 48 degrees (range 41 to 60 degrees).

The rate of deterioration of these double structural curves was much less than in the single thoracic curves with the result that bracing was not necessary until a later age. The brace was applied at a mean age of seven years and nine months (range four to thirteen years) and was worn for a mean period of three and a half years.

The mean best correction was achieved by two years in all patients: 14 per cent for thoracic curves and 23 per cent for lumbar curves. Three of these children have now reached skeletal maturity and are out of the brace. Two children are still wearing the brace after eighteen and forty-four months. The mean correction for the thoracic curves when last seen was -1 per cent (range -26 per cent to +17 per cent) and for the lumbar curves 17 per cent (range 2 to 39 per cent).

Case 3. This boy was first seen at the age of four years and seven months. A few months after birth, his mother noticed a slight prominence of one side of his back posteriorly and also an asymmetry of his face but the spinal deformity did not appear to deteriorate until just before his referral to the Scoliosis Clinic. On examination he was found to be normal except for a mild left thoracic scoliosis and mild right lumbar scoliosis both of which showed clinical rotation. Radiographs confirmed a double structural scoliosis with a left thoracic curve extending from the seventh to the twelfth thoracic vertebra measuring 28 degrees and a lumbar curve from the twelfth thoracic to the fifth lumbar vertebra measuring 22 degrees. The rib-vertebra angle difference was -40 degrees (Fig. 16).

No specific treatment was prescribed and the child was seen regularly at the Scoliosis Clinic where the progress of the curve was followed radiographically. There was little deterioration until the age of eight years and eight months when the thoracic curve was found to be 45 degrees and the lumbar curve 41 degrees (Fig. 17). Although the cosmetic deformity was small it was decided to treat the curves in an Edinburgh brace to prevent further deterioration. The best correction in the brace was achieved at the age of ten years and six months when the thoracic curve measured 30 degrees and the lumbar curve 24 degrees (Fig. 18). At present he is still wearing his brace at the age of fourteen years and there has been a slight deterioration of the thoracic curve to 39 degrees and the lumbar curve to 36 degrees (Fig. 19). The cosmetic deformity remains small and it is proposed to continue to use the brace until skeletal maturity if possible.

Complications
Clinically significant orthodontic moulding occurred in eight of the twenty-two patients with single thoracic curves (36 per cent) and in one of the five patients with double structural curves but after final removal of the
brace and with further growth the deformity usually resolved. Pressure sores developed in four patients but responded to removal of the brace for a few days and adjustment of the pelvic girdle.

**DISCUSSION**

Posterior fusion is a well-established method of preventing an increase in spinal deformity but unfortunately cannot be applied to very young children. The fused area will not grow in length and will therefore cause a relative shortening of the spine as the child grows and also a possible increasing lordosis due to continuing growth of the unfused vertebral bodies anteriorly (Ponseti and Friedman 1950). In order to minimise these difficulties James (1967) recommended that posterior spinal fusion be postponed until the age of ten years unless progression of the curve could not be controlled by other means. A long period of conservative treatment is therefore necessary to prevent deterioration of a curve which develops before the age of three years and can only be safely fused after the age of ten years. The most effective means of preventing deterioration is the Milwaukee brace, although if the curve is already severe correction in a Risser turnbuckle jacket may be necessary before applying the brace. The Edinburgh brace is a modification of the Milwaukee brace and depends only on distraction. The modern Milwaukee brace relies less on distraction and more on an axially directed force applied through the ribs to the site of maximum spinal curvature. This is a dynamic force produced by the patient who performs specific exercises against a fixed pad applied over the rib hump. In curves over 40 degrees the axially directed force becomes progressively less effective and distraction becomes more important as the size of the curve increases (Blount and Moe 1973).

In our study many patients were referred late for treatment and the mean degree of curvature for a single thoracic scoliosis immediately before bracing was 63 degrees. The Edinburgh brace produced a mean best correction of 43 per cent in the twenty-two patients with single thoracic curves (Fig. 4) and the greatest percentage correction was obtained in curves measuring
46 to 75 degrees (Table I). The brace, however, was much less effective in gaining correction in the five patients with double structural curves: a mean best correction of 14 per cent for thoracic curves and 23 per cent for lumbar curves.

The best correction of single thoracic curves was obtained in 59 per cent of our patients in two years, but some required a five-year period of bracing. After the initial best correction there was then a slow deterioration of approximately one degree per year, but after five years in the brace the rate of deterioration usually increased and by the age of ten years the overall mean curve correction was 11 per cent. However, larger thoracic curves maintained better correction than lesser curves as might be expected in a pure distraction system. The final degree of correction of single thoracic curves in our patients was less than that achieved at skeletal maturity in juvenile and adolescent idiopathic scoliosis by Moe and Kettleson (24 per cent) and by Keiser and Shufflebarger (26 per cent) but our infantile patients wore their brace on average for twice the period reported in these series.

In Edinburgh it has been the policy to carry out a posterior spinal fusion of all progressive single thoracic curves as a routine procedure at the age of ten years. If spinal fusion were not performed a brace would have to be worn until skeletal maturity which in boys can be as late as eighteen years. By ten years of age 70 per cent of our patients had already worn their brace for over five years and we consider it unfair to subject these children to an even longer period in the brace unless it is absolutely necessary. Immediately after spinal fusion the degree of curvature in all but two patients was better than that before the onset of conservative treatment in early childhood, the overall degree of correction being 40 per cent (Fig. 4). Harrington instrumentation is now routinely used in nearly all curves and had it been applied to these two patients the degree of correction might have been greater. Unfortunately in these young children the development of solid spinal fusion did not necessarily prevent further moderate loss in correction. Those patients with a solid spinal fusion whose scoliosis had been corrected in a localiser cast and who have been followed for an average of seven years after operation have lost an average of 17 degrees of correction. Nearly all of this loss occurred in the first two to three years and is thought to be due to bending of the solid but immature fusion mass before it eventually became stable. Those patients whose spines were corrected by means of Harrington instrumentation all developed a solid spinal fusion, and in an effort to produce a stronger fusion mass all received a massive onlay of autogenous cancellous bone strips after deep decortication at the time of the routine interfacetal fusion. These patients have now been followed for a maximum of three and a half years and there has so far been no loss in the mean correction after the first year. A much longer follow-up is necessary before any definite conclusion can be drawn.

In the much smaller number of children with double structural curves the possible disability resulting from a nearly total spinal fusion at a relatively early age outweighs the advantages, and brace therapy is therefore continued until skeletal maturity. Fortunately even quite large double structural curves are cosmetically acceptable, but should the thoracic curve continue to progress to the stage where cardiorespiratory complications seem likely, Harrington instrumentation and posterior spinal fusion are performed immediately.

We therefore suggest that a planned course of treatment incorporating both brace therapy and spinal fusion is an effective means of managing progressive infantile idiopathic scoliosis.

We would like to thank Professor J. I. P. James for allowing us to review his patients and also Mr M. Devlin for his photographic assistance.

REFERENCES