Infantile idiopathic scoliosis in the newborn

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Accepted: 5 May 1997

Summary. We have reviewed 7 patients (5 boys and 2 girls) with infantile idiopathic scoliosis which was present at birth and was diagnosed at ages from 1 day to 1 month. The mean follow up was 16 years. The objectives were to discover whether intrauterine forces play a role in the aetiology and to determine whether early treatment of a potentially progressive curve can induce resolution. Two infants were male conjoined twins and were united by skin over the posterior sacrum. Six infants had a rib vertebral angle difference greater than 20° in the first radiograph and 5 had stiff curves. In 2 with flexible curves, the deformity disappeared by the end of the first year. The 5 with stiff curves were treated conservatively for 6 to 24 months. Growth has now been completed in 5; 4 have a straight spine and one developed an adolescent curve of 26°. The 2 who are still growing have no scoliosis. Intrauterine moulding was only demonstrated in the conjoined twins. There was a correlation between the rib vertebral angle difference above 20° and the rigidity of the curve. Early treatment of a potentially progressive curve can lead to resolution.

Résumé. Avec l'objectif de savoir si les forces intra-utérines jouent un rôle important dans l'étiologie de la scoliose infantile et pour assurer si le traitement orthopédique tôt d'une courbure potentiellement progressive peut induire une résolution, nous avons revu 7 malades avec une scoliose infantile présente à la naissance. Cinq d'entre eux étaient des garçons et 2 filles avec un âge au

moment du diagnostique qui oscillait entre 1 jour et 1 mois. Le temps moyen de révision a été de 16 ans (entre 9 et 20 ans). Deux patients masculins étaient des jumeaux pygopagos unis par la peau de la région sacrée. Six patients ont eu une différence des angles costovertébraux (RVAD) supérieur à 20 degrés à la première radiographie et 5 d'entre eux avaient une courbure fixée. Chez 2 malades avec courbure souple, la déformité a disparu d'une façon spontanée après la première année de vie. Cinq malades ont été traités d'une façon conservatrice pendant 6–24 mois. Cinq malades ont achevé la croissance, 4 d'entre eux ont le dos absolument droit et un a développé une courbure de 26° à l'adolescence. Deux patient encore en croissance n'ont pas de scoliose. Conclusions: Les forces modélantes intra-utérines comme facteur initial de scoliose infantile ont été trouvées seulement chez 2 enfants jumeaux pygopagos. Il pourrait exister une relation entre la différence des angles costovertébraux supérieur à 20 degrés et la rigidité des courbures. Le traitement précoce des courbures potentiellement progressives peut induire une correction.

Introduction

Infantile idiopathic scoliosis can be present at birth or at any time up to 3 years [8]. It differs from adolescent idiopathic scoliosis which has a higher incidence in boys, the curve is more frequently convex to the left and the deformity resolves spontaneously in most cases [9, 10, 11]. The number of affected relatives is less in the infantile

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Fig. 1. Radiographs of conjoined twins united by skin over the sacrum

Fig. 2. Anteroposterior radiographs after successful separation. The RVAD Cobb angle was 22° and 34° respectively in one **a** and 32° and 45° in the other **b**

Fig. 3. a A newborn child with a right thoracic curve with a RVAD of 32° and a Cobb angle of 32°. **b** The same child showing almost complete spontaneous resolution of the curve 6 months later. The RVAD was now 6° and the Cobb angle 12°

group [25, 26] and there may be an environmental factor acting in utero or in the first months after birth [1, 6, 18]. It is rare in North America [20] and was considered relatively prevalent in Europe in the 1950s and 1960s. Although some cases have been diagnosed at birth, they have not been precisely described [2, 11, 22].

The purpose of this paper is to report 7 patients with the condition, to determine whether intrauterine moulding forces acting on the growing foetus play a part in the aetiology, and to ascertain whether early corrective treatment in potentially progressive curves will induce resolution, as has been suggested [1, 2, 4, 13, 14, 18, 19].

Fig. 4. RVAD evaluation

Patients and methods

Between 1976 and 1987, 7 infants under the age of 1 month with structural scoliosis were seen in our department. Clinical details of the pregnancy and the history of the birth were recorded. The side of the curve, the presence of moulding deformities, such as plagiocephaly, and the degree of correction which could be obtained were documented.

Curves were classified as flexible when they corrected on passive side bending, and rigid if correction did not occur on application of a considerable force.

Anteroposterior radiographs were taken with the baby suspended by the arms and the rib vertebral angle difference (RVAD), and the Cobb angles were measured and compared with subsequent radiographs.

Conservative treatment was with either a scoliosis tray, braces or serial casts. The final outcome was recorded.

Results

There were 5 boys and 2 girls with an age at diagnosis of from 1 day to 1 month. The mean follow up was 16 years (range 9 to 20 years). All the curves were single and thoracic; 5 were left sided and 2 right. Four had associated anomalies, such as plagiocephaly and facial asymmetry. One had well marked creases on the concave side of the spine. None had hip dysplasia. One infant was diagnosed as hypotonic, but he recovered completely and was walking before his second year.

The mothers had no previous obstetric history or illness while pregnant and they were between 16 and 29 years of age; all the infants were born at over 34 weeks. One case had a family history of scoliosis. There was one breech presentation. Two infants weighed 1730 g and 2020 g, and the rest were within the normal range of birth weight.

Two infants were male conjoined twins (Fig. 1) and were united by skin over the posterior sacrum; their combined weight was 3730 g. The pregnancy

and vaginal delivery were uneventful. The presentation was cephalic for the first and breech for the second baby. Their mother was 27 years of age and there was no history of twins. Investigation did not reveal any significant abnormality, except for a duplicated ureter in one child. Both had severe rigid curves. Plain radiographs showed spinal curves with opposite convexities without any bony malformation, and no bone union between the sacrum. Successful separation was carried out 36 h after birth and they recovered uneventfully (Fig. 2), leaving hospital on their 34th day of life.

The mean RVAD of the 7 infants at diagnosis was 22° (range 14° to 32°) and in 6 was greater than 20°. The mean Cobb angle was 21° (range 11° to 45°). After 2 to 3 months, the RVAD had increased in 4, decreased in 2 and was the same in one (Fig. 4). Two infants had flexible curves, only one had a RVAD over 20°, and 5 were classed as rigid.

The parents were advised to lay the infants in the prone position and apply a gentle side bending corrective force. Two with flexible curves (RVAD 14° and 32°) improved rapidly and spontaneous regression occurred by the end of the first year (Fig. 3). Five were treated in a Denis Browne splint, spending a minimum of 12 h a day in the tray over 6 to 12 months. In one, the curve corrected completely after 6 months. In the remainder, treatment in a plaster jacket combined with braces was continued for 12 to 24 months until the spine was completely straight.

At the last follow-up, 5 children had finished growing and only one had developed a late benign adolescent scoliosis of 26° at skeletal maturity. Two, aged 9 and 11 years, who are still growing, have straight spines.

Discussion

Infantile structural scoliosis was first described in 1930 [8], and since then two types have been recognised: resolving and progressive [9, 10, 11]. Spontaneous regression occurs within 12 to 18 months of diagnosis in the resolving type; the progressive type increases with growth and, if not treated, can lead to severe deformity, cardio-respiratory compromise and even premature death [5, 12]. The groups are difficult to distinguish at an early age [3, 7, 21]. Mehta reported that when the difference in RVAD was less than 20° the curve was generally resolving, but if the angle was more than 20° the curve was likely to progress [15]. This radiographic measurement must be repeated after 2 or 3 months, if the curve is resolving the RVAD

will be decreased, but in a progressive curve the angle will remain unchanged or increased. Mehta also related clinical flexibility or rigidity to the tendency to progress or resolve; progressive curves being stiff and not correctable [16]. We found some correlation with these observations in our cases. In 5 infants with rigid curves the RVAD was over 20° and was increased or unchanged in the second radiographs. In the 2 infants with flexible curves the RVAD was greater than 20° in one, but had decreased markedly in 3 months; both cases had resolved completely at the end of their first year of life.

The aetiology of infantile scoliosis is unknown, but the deformity seems to result from an interaction of genetic and environmental factors. Wynne Davies confirmed the importance of the former in the aetiology of idiopathic scoliosis, especially of the adolescent type, and recorded that 2.6% of first degree relatives of patients with infantile scoliosis had scoliosis compared with 12% in adolescent scoliosis [25, 26]. Others have agreed that posture in utero could cause scoliosis [1, 6, 11]. Mechanical compression forces acting on the growing foetus could determine the aetiology of the scoliosis and the associated anomalies, the most common being plagiocephaly and unilateral flattening of the face. Nevertheless, scoliosis is rarely apparent at birth. The incidence reported is 6 in 212 patients with infantile scoliosis [11], 1 in 4 [22] and 6 in 134 infants [3]. A low incidence of 0.5% in all idiopathic curves has been recorded in Boston, US, whereas in Edinburgh, Scotland, 50% of all idiopathic curves were infantile [20]. Both populations stem from similar stock so there must be beneficial environmental factors in North America and adverse factors in Scotland. Nursing infants prone, as is done in North America, may prevent or correct any tendency to curvature. Infants in Europe are usually laid on their back or side, and often wrapped tightly in blankets, especially in the winter [14]. A larger number of cases has been reported as developing between October and March [25]. These environmental factors must operate similarly in both types of infantile scoliosis. It has been proposed that although the triggering factor is common to all forms of infantile scoliosis, the subsequent behaviour depends on two factors, namely the magnitude of the deforming forces exerted in utero or immediately after birth, and the genetically determined resistance of the child's tissues to the deformity [18]. In the resolving type correction occurs spontaneously when the moulding forces are removed.

The same forces acting on infants with genetically determined diminished resistance of the tissues to deformity will have a curve which will not correct spontaneously. In our study, intrauterine pressure was only likely to have been the cause in the conjoined twins as they maintained an abnormal foetal posture which produced spinal curves with opposite convexities. Only one case of scoliosis in dizygotic twins has been described [26].

Treatment has been controversial. Browne believed that spontaneous correction would not occur and advised a splint to keep the spine inclined towards the convexity [1]. Mau used a plaster shell in a similar manner [13]. Walker concluded that the splint did not increase the incidence of resolving cases [24]. Thompson suggested that treatment did not influence the outcome of progressive curves and should only be undertaken when the curve was over 40° [23]. Mehta achieved resolution in 21 cases with progressive infantile scoliosis, but no child was referred before the age of one year and their deformities remained untreated for from 13 months to 9 years [17, 18].

If we accept that progressive curves do not resolve, then any child with a rigid curve and a RVAD greater than 20° after 3 months' observation, should be treated conservatively. A period of growth can redress a curved spine into a normal direction. Even small deformities present at, or soon after, birth may get worse in infants whose connective tissues do not have a potential for recovery. Five of our cases were treated conservatively with splints, plaster or braces for from 6 to 24 months.

Intrauterine moulding forces initiating infantile scoliosis were demonstrated in only 2 of our 7 cases. In 5, there was a correlation between rigid curves and a RVAD greater than 20° . Early corrective treatment of potentially progressive curves may result in resolution.

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