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CONGENITAL FUNCTIONAL SCOLIOSIS

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Congenital functional scoliosis is a fairly rare and relatively unknown type of scoliosis. It has not previously been mentioned in Scandinavian literature. The object of this paper is to describe the appearances of congenital functional scoliosis and to elucidate the differential diagnostic problems in distinguishing it from other types of scoliosis occurring during the first year of life.

Congenital functional scoliosis is taken to mean scoliosis of unknown origin, appearing during the first year of life and presumably present at birth, and which is possibly due to a faulty intrauterine position. It is a characteristic of this type that it is benign and disappears spontaneously.

This benign scoliosis is called resolving infantile idiopathic scoliosis by Anglo-Saxon writers. Some authors (*James* 1951, *Scott* 1956) divide resolving infantile idiopathic scoliosis into 2 sub-groups: the resolving type and the postural type. According to *Scott* (1956) the postural type will subside slowly when the infant begins to stand, the standing posture stimulating its disappearance, whereas the resolving type does not disappear in the erect position. This latter type may even progress slightly after the infant starts standing in order to resolve spontaneously at the age of 4–5 years. One author (*Browne* 1956) calls all forms of infantile idiopathic scoliosis postural, and in his opinion they all progress if untreated.

In contrast to this benign type, there is the malignant one: progressive infantile idiopathic scoliosis (Figures 4 and 5), also called "the progressive type" which usually sets in during the 2nd or 3rd year of life (*James* 1951, *Scott* 1956), although it may be observed as early as the first year. This type is characterized by steady progression throughout the period of growth. The earlier the scoliosis sets in, the greater

is the tendency to progression. This tendency is greater in boys than in girls, as their growth period is longer than the girls'.

Other types of scoliosis that may occur during the first year of life: structural scoliosis due to congenital malformations (hemivertebrae, etc.) and other types of scoliosis of known aetiology (paralytic, static, etc.) differ clearly from the two types mentioned above and thus fall without the scope of the present study.

PRESENT MATERIAL

During the period 1936 to 1959 a total of 90 patients with congenital functional scoliosis were treated in the Orthopaedic Hospital, Aarhus. No distinction was made between the resolving and the postural type. Out of these 90 patients only 76 are included in the material, 14 being left out as the diagnosis was based exclusively upon the clinical examination without X-ray films. Presumably, these patients did have congenital functional scoliosis, since in all cases the scoliosis remained stationary or disappeared during the period that the patients were observed in the Department. Out of the remaining 76 patients 67 were seen by the author in a follow-up which included clinical as well as X-ray examination. The average follow-up period was 12 years (5½–25½ years). The distribution by length of follow-up period is shown in Table 1.

Table 1. Distribution by length of follow-up period.

Follow-up period	No. of pts.
5–9 years	31
10–14 years	20
15–19 years	10
20–25 years	6

Of the remaining 9 patients 4 stated in letters that the back was straight and normal. One had gone abroad, and all attempts at contact have been in vain. The last 4 patients failed to respond to repeated requests to present themselves for a follow-up examination. This makes a follow-up rate of 88 per cent (67 out of 76 patients), as only those patients who had clinical and radiological examination were considered to have been sufficiently followed.

Table 2 shows the age at which the scoliosis was discovered by the parents, health visitor, or general practitioner at the prophylactic in-

fant examination as well as the age at which the patients were first seen in the Orthopaedic Hospital, Aarhus.

Table 2. Age distribution at detection of scoliosis and at 1st examination at the Orthopaedic Hospital, Aarhus.

Age in months	Scoliosis detected. No. of pts.	Seen for the first time in the Orthopaedic Hospital, Aarhus. No. of pts.
1	3	0
2	7	0
3	11	3
4	12	7
5	11	8
6	9	15
7	5	13
8	2	7
9	1	3
10	4	3
11	2	8
12	0	0

The great majority of cases were detected within the first 6 months of life (53 out of 67 or 79 per cent). This is in keeping with statements in the literature. *James et al.* (1959) reported 4th to 10th month, *Scott & Morgan* (1955) 5½ months, range 1st to 9th month. *Harrenstein* (1936) stated the 3rd to the 7th month, but mentioned at the same time that the scoliosis is seldom observed before he 2nd month. The progressive type is not diagnosed until much later, according to *Scott & Morgan* (1955) at an average age of 15 months. *James* (1959) reported that out of 111 scolioses of the progressive type 33 were diagnosed during the 2nd or 3rd year of life. Incidentally, it is apparent from the table that as a rule about 3 months elapse from the time that the scoliosis is detected until the diagnosis is definitely established.

Table 3. Sex ratio and number of left- and right-sided scolioses.

	Girls	Boys	Total
	29	38	67
Right-sided scoliosis	8	13	21
Left-sided scoliosis	21	25	46

Table 3 gives the sex ratio which shows a considerable male preponderance, viz. 38/29. In this respect too the findings are in agreement with the literature. *James et al.* (1959) found a ratio boys/girls of 50/27, *Lloyd-Roberts & Pilcher* (1965) 67/33. Within the progressive type too *James et al.* (1959) found a preponderance of boys 65/46, while *Scott & Morgan* (1955) found the condition to be equally common in both sexes.

At follow-up 2 patients had complaints in the form of mild pain when overstraining the back, and another 2 had more severe pain. All were found to have weak, insufficient back muscles, and the pain was considered of muscular nature. All 4 had a curvature of less than 10°, 3 of them of less than 5°.

In 9 cases there was clinically very slight scoliosis, merely discernible in 6 of the cases. In 18 cases there was torsion prominence, just discernible in 9, while the other 9 had mild, but distinct torsion.

Table 3 shows, moreover, that left-sided scoliosis was more than twice as common as right-sided. There was no sex difference in this respect. *James et al.* (1959) as well as *Scott & Morgan* (1955) also found left-sided scolioses to be more common, and this applies also the progressive type.

Table 4. Angle of scoliosis at 1st examination and at follow-up.

Angle	When first seen. No. of pts.	At follow-up. No. of pts.
0– 5°	0	47
6–10°	15	17
11–15°	25	3
16–20°	20	0
21–25°	4	0
26–30°	3	0
	67	67

The degree of scoliosis was measured on the primary X-ray films as well as on the follow-up films. The curve was measured as the angle between the upper plate of the cranial and the lower plate of the caudal vertebra involved in the torsion. The primary angle averaged 15° (7–29°). At follow-up the corresponding angle was less than 5° in 47 patients and, as is apparent from Table 4, did not exceed 15° in any case.

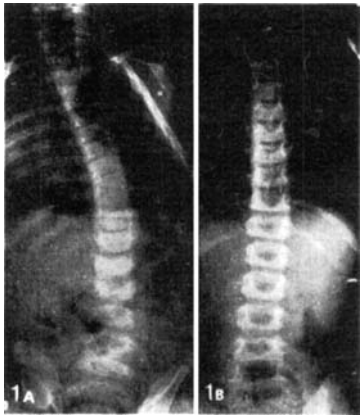

Figure 1. Congenital functional scoliosis (male). A. Age 5½ months. B. Age 6 years.

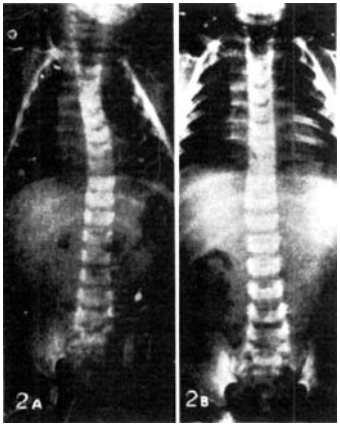

Figure 2. Congenital functional scoliosis (male). A. Age 5 months. B. Age 14 months (not included in the material).

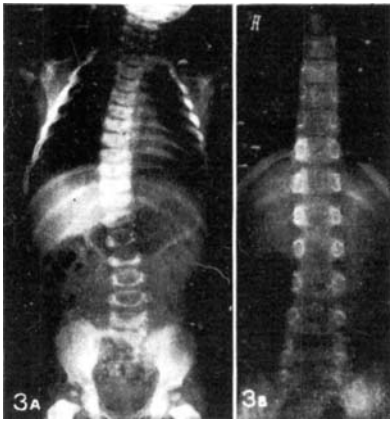

Figure 3. Congenital functional scoliosis (with compensatory curvature) (male). A. Age 8 months. B. Age 7 years.

Radiographically, most patients showed long, soft, C-shaped single curvatures without compensatory curves (Figures 1 and 2). In one case, however, there was a hint of compensation (Figure 3). At the primary physical examination 55 patients had exhibited slight posterior torsion of the ribs on the same side as the convexity of the scoliosis. In the remaining 12 cases no mention is made of a torsion prominence. The number of vertebrae involved in the curve ranged from 6 to 11, average 9 (8.6). The greater part of the curvatures affected the thoracic spine, the peak of the convexity corresponding to the 9th or 10th thoracic vertebra (T_9–T_{10}). Table 5 illustrates the site and extent of the scolioses. In more than half, viz. in 38 out of 67 patients, or in 57 per cent of the scolioses, the uppermost vertebra was T_5–T_7, and the lowermost L_1–L_2.

Table 5. Extent of scoliosis. Uppermost (vertical column) and lowermost (horizontal line) involved vertebra.

	T_{12}	L_1	L_2	L_3	Total
T_4	3	4	3		10
T_5	3	9	8	2	22
T_6	1	8	7	1	17
T_7		1	6		7
T_8		3	4	1	8
T_9			1	2	3

James et al. (1959) found the majority of curvatures to be 11–20°, *Scott & Morgan* (1955) below 20°. Moreover, the latter found torsion to be minimal, while in the progressive type the curvature was seldom below 20° and the torsion more pronounced already at the first examination. In the progressive type, moreover, there is compensatory curvature (Figures 4 and 5) which is rarely present in the functional type (Figures 1 and 2). *Scott & Morgan* (1955) also found the progressive types to involve fewer vertebrae than the functional type, 3–8 vertebrae in the progressive as compared with 5–10 in the functional type.

During the period 1936–1954 44 cases of congenital functional scoliosis were diagnosed in the Orthopaedic Hospital, Aarhus, as compared with 4 cases of the progressive type. This means a frequency of progressive scoliosis of 8.3 per cent, but no doubt this is too high, as some of the functional cases are never seen by an orthopaedic surgeon or

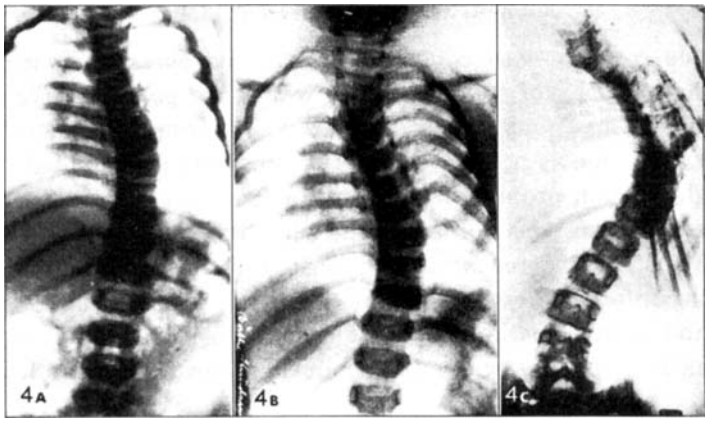

Figure 4. Infantile progressive idiopathic scoliosis (female). A. Age 11 months. B. Age 19 months. C. Age 13 years.

will simply be overlooked and thus never be recorded, whereas the progressive cases are bound to be sent for treatment in an orthopaedic department. *Lloyd-Roberts & Pilcher* (1965) reported a frequency of 8 per cent.

Prior to 1955 12 of the most severe cases of congenital functional scoliosis were treated in a plaster or celastic bed. Two patients were treated in a corrective bed, the trunk being bent to the side of the convexity of the scoliosis. Patients seen after 1955 have received no treatment at all.

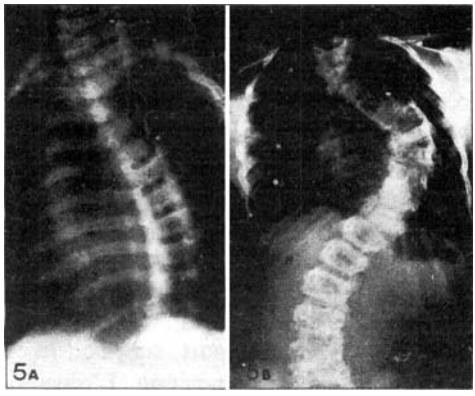

Figure 5. Infantile progressive idiopathic scoliosis (male). A. Age 10 months. B. Age 10 years.

DIAGNOSTIC CRITERIA

The typical patient with congenital functional scoliosis is a healthy, normal-looking boy of 6–7 months in whom the parents have noted a curvature of the spine, stationary through a couple of months. The thoracic spine shows a long, very mild, most often left-sided scoliosis with slight torsion prominence of the ribs, but no compensatory curvature. This scoliosis does not disappear when the baby is held suspended by the arms. X-rays reveal a long, soft, C-shaped, single curve with slight torsion and no compensation. The apex of the scoliosis is at T_9 or T_{10}, and it involves about 9 vertebrae which exhibit no congenital malformations. In most cases the scoliosis is localized within the area of T_5–L_2.

The main problem is to distinguish the benign type of congenital functional scoliosis from malignant infantile idiopathic scoliosis as early as possible. During the first year of life, it is usually impossible to decide with certainty whether a scoliosis is going to progress or to disappear. The functional type involves a larger number of vertebrae and has no compensatory curvature. Besides, it shows a lesser angle and less torsion, and it sets in at an earlier age. The progressive type is far less common than the functional type.

Only close observation can afford the answer. Therefore, the congenital functional scolioses have to be followed until they have disappeared or have remained stationary and very mild for a long time. As a rule, this cannot be decided until the age of 2 years.

SUMMARY

The most important types of scoliosis which may occur during the first year of life are briefly reviewed. A follow-up on a material of 67 patients with congenital functional scoliosis is reported. Average follow-up period was 12 years. At follow-up, practically all the scolioses had straightened spontaneously. The appearances of congenital functional scoliosis are described, and the most important differential diagnostic criteria from infantile progressive idiopathic scoliosis are discussed. It is concluded that a distinction between the two types can be made only by frequent examinations during the first years of life.

RESUME

Les types les plus importants de scolioses qui peuvent se produire pendant les premières années de la vie sont passés en revue. L'obser-

vation de 67 malades atteints de scoliose congénitale fonctionnelle est exposés. La période moyenne d'observation a été de 12 ans. On a constaté au cours de ces observations que pratiquement toutes les scolioses se sont redressées spontanément. Les aspects de la scoliose fonctionnelle congénitale sont décrits et il est discuté des critères les plus importants du diagnostic différentiel de la scoliose idiopathique infantile progressive. Il est conclu que seuls des examens fréquents durant la première année de la vie permettent d'établir une distinction nette entre ces deux types de maladies.

ZUSAMMENFASSUNG

Die wichtigsten Typen von Skoliose, welche im ersten Lebensjahre auftreten können, werden in Kürze durchgegangen. Eine Nachuntersuchung eines Materiales von 67 Patienten mit kongenitaler funktioneller Skoliose wird vorgelegt. Die durchschnittliche Beobachtungszeit war 12 Jahre. Bei der Nachuntersuchung hatten sich fast alle Skoliosen von selbst ausgerichtet.

Das Aussehen der kongenitalen, funktionellen Skoliose wird beschrieben und die wichtigsten differential-diagnostischen Unterschiede von der infantilen progressiven, idiopathischen Skoliose werden besprochen. Man schliesst, dass eine Unterscheidung beider Arten nur mittels häufiger Untersuchung in den ersten Lebensjahren gemacht werden kann.

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