KYPHOSCOLIOSIS

J. I. P. JAMES, LONDON, ENGLAND

From the Institute of Orthopaedics and the Royal National Orthopaedic Hospital

In ordinary structural scoliosis there is rotation of the vertebral bodies but no alteration of the vertebral column in an antero-posterior direction. At operation the appearance of the full length of the primary curve in approximately 150 patients confirms this statement. Only when the vertebral position is thus visualised is it possible to estimate the effect of rib rotation, which so frequently gives the impression of a kyphosis as well as lateral curvature, especially when rotation of the vertebral bodies approaches 90 degrees.

The term kyphoscoliosis has been used frequently but loosely to describe an effect due to rib rotation without determining whether a kyphosis was in fact present. The purpose of this paper is to describe a number of cases of varying etiology in which there is true vertebral kyphosis and lateral curvature with rotation—kyphoscoliosis in a true anatomical sense.

Kyphosis associated with structural lateral curvature as the equal or even dominant deformity is not discussed in the English literature. Bingold (1953) in a recent paper described three cases of congenital kyphosis at the thoraco-lumbar junction. These are comparable to several of the cases discussed but were not severe deformities and were not associated with scoliosis. The French literature has but a few reports. However, in Germany the condition has been the subject of a number of lengthy and beautifully illustrated papers.

Thirty-one patients seen at the Royal National Orthopaedic Hospital are available for review. Two further cases are included which are acknowledged later.

**Differential diagnosis of kyphoscoliosis.** Kyphosis—True kyphosis in the age group under discussion is commonly due to tuberculosis or Scheuermann’s osteochondritis. Tuberculosis characteristically produces kyphosis; there may be lateral angulation, but there is neither true lateral curvature nor rotation. A number of the cases to be described had previously been diagnosed as tuberculosis.

**TABLE I**

<table>
<thead>
<tr>
<th>Etiology of Kyphoscoliosis</th>
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<tbody>
<tr>
<td>Type</td>
</tr>
<tr>
<td>------------------------------</td>
</tr>
<tr>
<td>Congenital</td>
</tr>
<tr>
<td>Confirmed</td>
</tr>
<tr>
<td>Suspected</td>
</tr>
<tr>
<td>Infantile idiopathic scoliosis with kyphosis</td>
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<tr>
<td>Neurofibromatosis</td>
</tr>
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Adolescent kyphosis is characterised by a long gradual kyphosis; there may be a slight degree of scoliosis, usually with two structural curves. In true kyphoscoliosis the area of kyphosis is very short and in no way comparable to an epiphysitis.

Scoliosis—The lateral radiograph of a scoliotic spine of any etiology may show apparent antero-posterior deformity. Somerville (1952) demonstrated that there might be an initial lordosis. When rotation is gross a lateral radiograph shows an oblique view of the vertebral
column and a long kyphosis may seem to be present. The apex of a true kyphoscoliosis, however, rarely involves more than two or three vertebrae, and is angular.

The combined deformities of kyphosis, lateral curvature, and rotation produce overlap with resultant ill defined bone detail. When such vertebral "confusion" is seen in an antero-posterior radiograph in which the ribs and pelvis have normal definition, kyphoscoliosis should be suspected. Scoliosis shows a smooth curve; when this becomes angular or "squared," and if there is one clear disc space, kyphoscoliosis should be suspected (Figs. 5, 14, 17).

**Etiology of kyphoscoliosis**—The cases reported fall into three etiological groups, as shown in Table I.

**CONGENITAL KYPHOSCOLIOSIS**

Congenital skeletal abnormality is the cause, and the diagnosis cannot be accepted without radiological proof of this. Congenital wedging, vertebral fusion, hemivertebrae, absent vertebral bodies, spina bifida and rib anomalies were present. A number of patients were seen as adults with gross deformity; early radiographs were not available, and congenital anomaly could not be demonstrated in the hopelessly confused vertebral pattern shown in recent radiographs. They were in all other respects the same as the proven cases.

Congenital kyphoscoliosis occurred at two levels, thoraco-lumbar and cervico-thoracic. Paraplegia occurred at both levels (Table II).

**TABLE II**

**CONGENITAL KYPHOSCOLIOSIS (TWENTY-ONE CASES)**

<table>
<thead>
<tr>
<th>Level of deformity</th>
<th>Number of cases</th>
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</thead>
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<tr>
<td>Thoraco-lumbar</td>
<td></td>
</tr>
<tr>
<td>Congenital</td>
<td>13</td>
</tr>
<tr>
<td>Congenital suspected</td>
<td>3</td>
</tr>
<tr>
<td>Cervico-thoracic</td>
<td></td>
</tr>
<tr>
<td>Congenital</td>
<td>3</td>
</tr>
<tr>
<td>Congenital suspected</td>
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</tr>
<tr>
<td>Total</td>
<td>21</td>
</tr>
<tr>
<td>Thoraco-lumbar kyphoscoliosis with paraplegia</td>
<td>2</td>
</tr>
<tr>
<td>Cervico-thoracic kyphoscoliosis with paraplegia</td>
<td>3</td>
</tr>
</tbody>
</table>

**CONGENITAL THORACO-LUMBAR KYPHOSCOLIOSIS**

**Pathology**—The congenital anomalies which lead to this deformity are obscure, as few patients have adequate early radiographs. There is first of all the curious predilection for the low thoracic and thoraco-lumbar region. In several cases it seemed that two or three vertebrae were fused together; absence of the vertebral body epiphyses with deficient anterior growth may then have produced the kyphosis. This may be true because the major development of the deformity occurred during the period of rapid growth in the pre-puberty years. At operation one patient was found to have unilateral spina bifida, with fusion of the laminae on the other side; in other patients anomalies of the laminae were seen at operation.

**Clinical examination**—The condition may simulate kyphosis or scoliosis, particularly the latter. The photographs show the typical appearance, and with practice this can be recognised. The antero-posterior radiograph may give the first hint that there is kyphosis as well as scoliosis.
Prognosis—The prognosis must be considered from two aspects. Congenital lesions of the thoraco-lumbar region are not uncommon, whereas kyphoscoliosis is rare; it is not yet possible to appreciate the significant prognostic factors in the development of kyphosis. When kyphoscoliosis has developed, it seems that there is every likelihood of a severe and progressive deformity. Twelve patients have a kyphosis of more than 90 degrees, three have only small deformities (Fig. 1). A baby with an absent first lumbar vertebral body has already developed a severe kyphosis (Fig. 2). The degree of deformity can be remarkable, the two limbs of the kyphotic spine becoming almost parallel (Case 1, Fig. 5). The possibility of paraplegia will be discussed later.

Treatment—Treatment of these deformities has always in the first instance been conservative; it was not of great value. Kyphoscoliosis is a difficult deformity to correct because of the three planes in which deformity occurs. The Risser jacket has not proved effective; the distracting plaster jacket is better but still the kyphosis is obstinate. Most of these patients have been seen late, but it has been thought worth while to correct and fuse the spines of five of them for deformity. In view of the difficulty of correction and, in this series, the severity of the untreated deformity, it would seem wise to fuse the spine earlier. Early fusion of the spine in the treatment of Pott’s disease and scoliosis is theoretically unwise because it may produce lordosis by arresting growth posteriorly; in kyphoscoliosis it would be advantageous if this did occur.

The degree of kyphosis is measured by drawing lines parallel to the anterior borders of the vertebral bodies of each limb and measuring the angle.

Case Reports

Case 1—Curvature developed in a girl of twelve years, but radiographs then showed a barely noticeable kyphosis (Fig. 3). Eighteen months later there was a gross left thoraco-lumbar
Case 1. Figure 3—Kyphosis of 68 degrees. Figure 4—Photograph of patient eighteen months later.

Case 1. The left scoliotic curve is not smooth but angular; the kyphosis is extreme. Bone detail is characteristically confused at the deformity although elsewhere clear definition is achieved.
Case 2. Age five years. Congenital defects are seen at T.11-12.

Case 2. Figure 7—Photograph of the patient at age seventeen. The lateral radiograph (Fig. 8) shows a kyphosis of 125 degrees. There was a scoliosis to the right of 70 degrees.
kyphoscoliosis (Figs. 4 and 5). The lateral radiograph showed the kyphosis to be so gross, measuring 150 degrees, that the two limbs of the vertebral column were almost touching. Despite this there was no involvement of the cord. The apex of the kyphosis lay between T.9 and T.12. After attempted correction in an antero-posterior Risser jacket, fusion was performed. It was then seen that the two laminae above the apex were fused together and likewise those below. On the left, parts of the laminae were missing and a meningocele was present. The deformity has remained unchanged since operation.

Case 2.—A girl was noticed to have a curvature of the spine at the age of ten days. Subsequently she spent eight years in plaster beds (Fig. 6) but despite this the curvature became worse. When seen again at seventeen she had a severe right thoraco-lumbar kyphoscoliosis (Figs. 7 and 8). There were no cord changes. As she was fully grown no intervention was necessary, correction being impossible and no deterioration being expected.

Case 3.—A curvature had been noticed two weeks after birth and it steadily increased in a girl now eight years old. She had some spastic changes in the left leg, not progressive, and ascribed by a neurologist to cerebral cortical changes. She also had congenital absence of the abductor pollicis brevis and opponens pollicis muscles on the left. The apex of the kyphosis was low thoracic, probably T.9. On the left were fifteen ribs, on the right thirteen (Fig. 9).

Because of her age there was reason to expect an increase of the deformity, and, because of the slight doubt as to spinal cord compression, fusion was performed. Correction on this occasion, and since, has been carried out by the distraction jacket. It proved effective within the limits expected when dealing with a congenital bony abnormality.

CONGENITAL CERVICO-THORACIC KYPHOSCOLIOSIS

The site of this deformity is in the upper thoracic spine, but as one limb of the kyphosis is the cervical spine it seems appropriate to use this term. Congenital anomalies are present and have included fusion of vertebral bodies, hemivertebrae and rib anomalies. Three of the five cases had acceptable evidence of congenital anomaly; early radiographs were not available in the other two.
Case 4—Antero-posterior radiograph shows right scoliosis. The lateral film shows kyphosis of 90 degrees. There is fusion of thoracic vertebrae 2-3.

Case 5—Moderate scoliosis; kyphosis 140 degrees.
There is remarkably little visible deformity; even with a 90 degrees' kyphosis there is no more than a marked "dowager's hump." The upper margin of the trapezius may be elevated if scoliosis with rotation is marked.

Prognosis would appear not to differ from that of deformities occurring at the lower level. All five patients had a kyphosis of more than 90 degrees; the scoliosis varied. Three developed paraplegia. Congenital upper thoracic scoliosis without kyphosis is common, and the factor which determines kyphosis is not known; the serious consequences when kyphosis is present are, however, apparent. Treatment by correction in a distraction jacket followed by fusion is suggested.

Case Reports

Case 4—A girl now aged fifteen years developed curvature at two years. She had a severe right-sided high thoracic kyphoscoliosis. Stereoscopic radiographs showed fusion of T.2-3, and there are eleven ribs on the right, twelve on the left (Fig. 10).

Case 5—A girl developed a curvature at one year of age. She now has a kyphosis of 140 degrees (Fig. 11). No congenital defect can be seen but the radiographs are difficult to interpret and no early ones are available.

CONGENITAL KYPHOSCOLIOSIS WITH PARAPLEgia

Among seventeen patients with severe deformities, five had paraplegia, three at the upper level, two at the lower. In four cases the primary abnormality was proved at operation to be a congenital skeletal anomaly; in all four, pressure on the cord arose from an anterior ridge of bone. All of them had a severe kyphoscoliosis, though some patients without paraplegia had worse deformities. In four patients the paraplegia developed in the later years of growth; in the fifth patient the history is not well recalled but from the age of two there was difficulty with walking (Case 6).

Treatment of the paraplegia was not considered necessary in one patient (Case 6). In the other patients transposition of the cord was carried out by a posterior approach (two cases) or by antero-lateral decompression (two cases). The first method gave success, but two years after operation in both there was a recurrence probably due to increasing deformity. In each case a distraction jacket relieved the recurrent paraplegia, and fusion, one posterior, one anterior, has been performed. The antero-lateral approach gave one success, one failure.

Case Reports

Case 6—A woman of thirty-seven stated that since the age of two years she had experienced difficulty in walking, and that this was now becoming worse. She had an upper thoracic kyphosis, clinically not gross, and a spastic left leg with gross adduction and equinus. After investigation it was thought probable that her increasing difficulty was due to the leg deformities rather than to progressive cord pressure (Fig. 12).

Case 7—A young man with a high thoracic scoliosis developed paraplegia at the age of nineteen, with a sensory level at the fourth thoracic segment (Fig. 13). A forward transposition of the cord

Fig. 12 Case 6—This lateral radiograph shows a right-angled kyphosis. There are four or five vertebrae fused in the apical mass.
Case 7—Congenital kyphoscoliosis with paraplegia. Congenital lesions confirmed at operation.

Case 8—Left scoliosis, showing clear disc at apex and "squared" curve. Lateral radiograph shows kyphosis of 130 degrees.
Case 9—Idiopathic kyphoscoliosis. Early radiographs show no skeletal anomaly and no kyphosis.

Case 9—The deformity has increased. The lateral curve is becoming angular and the kyphosis marked.
was carried out; relapse of the paraplegia occurred, but was relieved by incision of the dura two months after the first operation. Three years later he again developed paraplegia, which steadily increased. Radiographs showed the upper limb of the kyphosis to be at a right angle to the lower, and displaced downwards. A distraction jacket completely relieved the paraplegia. Owing to the original extensive laminectomy posterior fusion would have been difficult. A rib graft was therefore placed on the antero-lateral aspect of T.1-4 at the junction of the two limbs of the kyphos.

Case 8—A young man aged nineteen was referred to Mr H. J. Seddon with a "tuberculous paraplegia" of two years' duration. Radiographs showed no evidence of tuberculosis. They did, however, present appearances identical with those already seen (Fig. 14). These radiographs do not demonstrate congenital abnormality, but at operation a massive congenital fusion was found in an antero-lateral approach for transposition of the cord, which resulted in excellent recovery. The cord pressure arose from an anterior ridge of bone.

IDIOPATHIC KYPHOSCOLIOSIS

Because of the interest aroused by these congenital anomalies and by Somerville's work on lordosis in idiopathic scoliosis, a more careful study of the lateral radiographs of all our patients with scoliosis has been undertaken, with the unexpected finding of a number of patients with severe idiopathic kyphoscoliosis.

Examples have been seen only among the group of infantile idiopathic curves. These curves, by definition, commence before the age of three and the radiograph must exclude congenital skeletal anomalies. This pattern of scoliosis is found only in the thoracic region. Some seventy children have been observed with this type of scoliosis, ten of whom have developed additionally a kyphosis, although early radiographs showed no gibbus. Operation has confirmed the presence of kyphosis, and the absence of posterior congenital anomalies. The usual site is mid-thoracic. Clinically suspicion is aroused by the increased deformity, roundness of the back and short stature. Radiologically the bone detail becomes confused, and the curve is no longer the arc of a circle but becomes angular or squared as the two limbs of the spine become more horizontal.
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Kyphosis has only been seen in the severest examples of idiopathic infantile thoracic scoliosis. The prognosis of the whole group is poor, but with the additional deformity it is usual to find a scoliosis and a kyphosis each of 100 degrees or more. Paraplegia has not been seen.

Treatment is similar to that advised for congenital kyphoscoliosis: correction by distraction and fusion. The deformity may be gross as early as five or six years of age. It is then perhaps wise to delay fusion. The Milwaukee jacket devised by Blount has been useful in preventing further deformity.

Case Reports

Case 9—A girl now aged eight developed infantile idiopathic left thoracic scoliosis when aged three months. Early radiographs showed no congenital defect or kyphosis (Fig. 15). At four years of age kyphosis was first noted. The deformity became severe, a scoliosis of 92 degrees and a kyphosis of 130 degrees being present at seven years of age (Fig. 16). Because of the kyphosis correction was carried out by a distraction jacket and fusion. There was no congenital anomaly of the laminae at operation.

Case 10—A boy now aged eleven years developed infantile idiopathic thoracic scoliosis at the age of eight months. Early radiographs showed no evidence of a congenital defect, but kyphosis was noticeable when this boy first attended two years ago. Correction and fusion were carried out; at operation no abnormality of the laminae was noted (Fig. 17).

KYPHOSCOLIOSIS IN NEUROFIBROMATOSIS

One high thoracic and one thoraco-lumbar kyphoscoliosis have been seen in two girls. In both the deformity was severe. The characteristic pigmented skin patches were present. Kyphoscoliosis in neurofibromatosis is well recognised, and is mentioned only to recall its existence when the differential diagnosis is considered.

SUMMARY

1. The true deformity of kyphoscoliosis has received little attention. Twenty-one deformities of congenital origin, ten idiopathic, and two secondary to neurofibromatosis, are discussed. The diagnosis is established and usually first suspected by radiography.

2. The deformity was severe and progressive except in three cases; paraplegia occurred in five congenital cases.

3. Early correction and fusion are advocated in the hope of preventing paraplegia and because correction of the old-established deformity is difficult or impossible.

Mr A. L. Eyre-Brook, Mr D. Lloyd Griffiths and Mr H. J. Seddon have been kind enough to allow me to include three cases under their care. I would like to thank the many surgeons who have referred cases to the scoliosis clinic. I would thank Mr R. J. Whitley for his skilled help with the radiographs which are, from the nature of the deformity, difficult to reproduce.

REFERENCES


Evans, E. Laming (1934): Two cases of Congenital Kyphosis. Proceedings of the Royal Society of Medicine (Section of Orthopaedics), 27, 1,265.


