

# HEMIVERTEBRA AS A CAUSE OF SCOLIOSIS

## A STUDY OF 104 PATIENTS

MICHAEL J. McMASTER, CHARLES V. DAVID

*From the Edinburgh Scoliosis Unit*

**We studied 104 patients with a total of 154 hemivertebrae which had produced scoliotic curves. Of the hemivertebrae 65% were of a fully segmented (non-incarcerated) type, 22% were semi-segmented and 12% were incarcerated.**

**We found that the degree of scoliosis produced depended on four factors: first, the type of the hemivertebra; secondly, its site; thirdly, the number of hemivertebrae and their relationship to each other; and finally, the age of the patient. Semi-segmented and incarcerated hemivertebrae usually do not require treatment. Fully segmented non-incarcerated hemivertebrae may require prophylactic treatment to prevent significant deformity.**

A hemivertebra due to complete unilateral failure of formation may result in scoliosis, but its severity varies greatly and there is controversy regarding treatment. Opinions range from total excision to total neglect of the hemivertebra (Moe et al. 1978). To rationalise management a better understanding of the natural history of the different types and sites of hemivertebrae is needed.

This paper does not aim to recommend any specific treatment but to provide an indication of the need for and the timing of prophylactic treatment before there is significant deformity, thereby helping to avoid unnecessary treatment.

### MATERIAL AND METHODS

All patients with radiographic evidence of hemivertebrae seen at the Edinburgh Scoliosis Clinic between 1958 and 1984 were reviewed and those with one or more scoliotic curves, each directly attributable to one or more hemivertebrae, were studied. Patients with hemivertebrae associated with a unilateral unsegmented bar (Nasca, Stelling and Steel 1975) were excluded as their prognosis relates more to the bar. Patients whose hemivertebra caused a kyphosis or a kyphoscoliosis in which kyphosis was the predominant deformity were also excluded because their natural history is very different from those

in which there is scoliosis alone. This left 104 patients with hemivertebrae producing a true congenital scoliosis. There were 66 females and 38 males those ages at diagnosis ranged from birth to 20 years. When first seen, four patients were skeletally mature; the remaining 100 were followed without treatment for a mean of 5 years (range 6 months to 15 years 2 months). At their last clinic visit, 73 patients had had no treatment and 33 of them had reached skeletal maturity. The remaining 31 patients had been followed without treatment for a mean of 3 years 8 months before eventually being treated either in a brace or by spinal fusion.

The types and sites of the hemivertebrae were diagnosed by reviewing the anteroposterior radiographs. The scoliotic curves were measured using the Cobb method, on radiographs taken with the patient standing, taking care that measurements were made from the same spinal levels on all the serial radiographs of each patient. Skeletal maturity was diagnosed when there was complete ossification and fusion of the iliac apophyses. Pelvic obliquity, decompensation or listing of the trunk, elevation of the shoulder and tilting of the head were assessed on the basis of the spinal radiographs and clinical photographs made with the patient standing.

### Sites and types of hemivertebra

The 104 patients had a total of 154 hemivertebrae which were divided into three groups based on the pathological anatomy of the hemivertebra: fully segmented (non-incarcerated), semi-segmented and incarcerated. For each patient, the vertebrae in the thoracic and lumbar regions were numbered on the radiograph in a cranial to caudal direction, counting each vertebra and each hemivertebra as one unit. A thoracic vertebra or hemivertebra was defined by the attachment of a rib or ribs. Using this

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M. J. McMaster, MD, FRCS, Consultant Orthopaedic Surgeon  
Princess Margaret Rose Orthopaedic Hospital, Fairmilehead,  
Edinburgh EH10 7ED, Scotland.

C. V. David, FRCS, Lecturer  
Department of Orthopaedic Surgery, University of Malaysia, Kuala  
Lumpur, Malaysia.

Requests for reprints should be sent to Mr M. J. McMaster.

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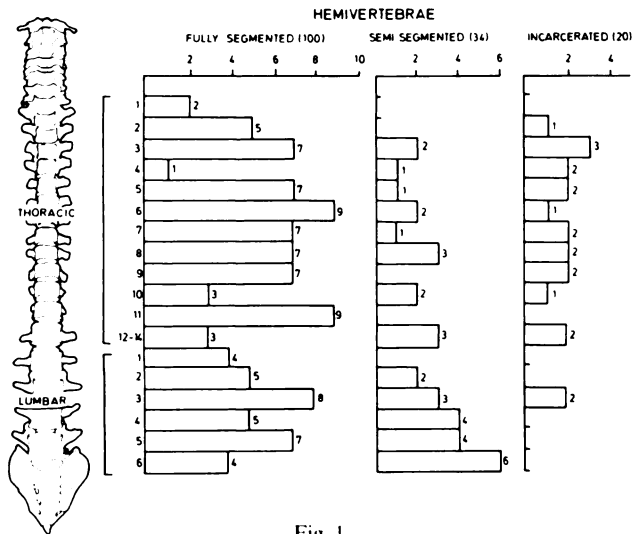


Fig. 1  
The sites and types of 154 hemivertebral in 104 patients.

method 34 patients had 13 thoracic vertebrae and six patients had 14, while 34 patients had six lumbar vertebrae. The sites of the various types of hemivertebral are shown in Figure 1. In all, 70 patients (67%) had a single hemivertebral, 28 (27%) had two hemivertebral and six patients (6%) had more than two hemivertebral. Multiple hemivertebral were never adjacent, always being separated by at least one relatively normal vertebra.

**Fully segmented (non-incarcerated) hemivertebral.** Of the 154 hemivertebral, 100 (65%) were fully segmented and non-incarcerated, and were fairly evenly distributed throughout the spine (Fig. 1) with equal numbers on each side.

Fully segmented hemivertebral were usually triangular in shape with the disc spaces above and below appearing to be relatively normal. The hemivertebral was wedged between two relatively normal vertebrae which

often became slightly wedge-shaped during growth but the disc spaces were preserved (Fig. 2). The lateral margin of the hemivertebral was equal or nearly equal in height to the adjacent vertebrae and 23 of these 100 hemivertebral extended across the midline. All had a single pedicle and in the thoracic region they were associated with a rib; this could result in an extra rib on one side of the spine. A single unilateral hemivertebral always lay at the apex of a definite scoliosis and a characteristic of this type was that, as the scoliosis deteriorated, the body of the hemivertebral tended to protrude slightly from the lateral margin of the spine (Fig. 2).

**Semi-segmented (non-incarcerated) hemivertebral.** Thirty-four (22%) of the hemivertebral were semi-segmented. They were commonest in the lumbar region (Fig. 1) and tended to be single.

Semi-segmented hemivertebral were similar in shape to the fully segmented type but the body of the hemivertebral was synostosed with one of its neighbouring vertebra, with no intervening disc space (see Fig. 17). The other disc space either above or below the hemivertebral was relatively normal. Of these 34 hemivertebral, 19 were synostosed with the vertebra above and 16 with the vertebra below. Three cases of hemivertebral, diagnosed in the first year, initially appeared to be fully segmented but during later ossification of the spine it became apparent that they were semi-segmented.

**Incarcerated hemivertebral.** The 20 incarcerated hemivertebral (13%) formed the least common type. They were most frequent in the thoracic region (Fig. 1) and tended to be single.

An incarcerated hemivertebral was usually more ovoid in shape and smaller than a fully segmented one. It was tucked into the spine and set in a niche scalloped out of the adjacent vertebrae (see Fig. 18). The vertebrae above and below were shaped in such a way that they tended to compensate for the hemivertebral and as a



Fig. 2



Fig. 3



Fig. 4

Case 1. Figure 2 - A boy aged 11 years with a 23° right thoracolumbar scoliosis due to a fully segmented (non-incarcerated) hemivertebral at L1. Figure 3 - No treatment was given and by the age of 20 years the patient was skeletally mature and the curve had deteriorated to 52°. Figure 4 - Despite the severity of the curve the cosmetic deformity is relatively mild.

result the general alignment of the spine remained straight with minimal scoliosis. The disc spaces above and below the incarcerated hemivertebra were often narrow and sometimes poorly formed. The lateral margin of the hemivertebra was usually two-thirds or less than the height of the adjacent relatively normal vertebrae and the hemivertebra did not extend across the midline. All the incarcerated hemivertebrae had a single pedicle and in the thoracic region they were all associated with a rib.

**Rib and chest anomalies.** Congenital rib fusions were present in 10 of the 104 patients and were unilateral or bilateral, affecting two or three adjacent ribs at a site distant from the vertebral column. They were not thought to affect the development of the scoliosis; the site of the fusion was not specifically related to the site or side of the hemivertebra.

A chest wall defect was present in four patients and was due to the absence of several adjacent ribs, always on the convex side of the scoliosis and just below the level of the hemivertebra. Congenital elevation of the scapula (Sprengel's shoulder) was present in two patients who had hemivertebrae at T1 or T2. Unfortunately, in both patients, both abnormalities were on the same side and combined to increase the deformity produced by the elevated shoulder.

**Other congenital anomalies.** Occult intraspinal anomalies were diagnosed by myelogram in seven patients (7%); six had diastematomyelia and one a unilateral absence of lumbar nerve roots. Four of these patients had hemivertebrae at the lumbosacral junction.

Other congenital anomalies were also common: Klippel Feil syndrome and heart anomalies were each seen in six patients; thumb deformity and foot deformity each in five patients; rectal atresia, urinary anomalies, radial club hand, or congenital dislocation or subluxation of the hip were seen in four patients. Goldenhar syndrome was seen in three patients; facial asymmetry and tracheo-oesophageal fistula in two patients each; while cleft palate and reduction of the lower limb were each present in one patient of the series.

## RESULTS

Of the 104 patients, 87 had a single congenital scoliosis; 70 were due to a single hemivertebra (67%) and 17 to two hemivertebrae on the same side (16%). Eleven patients had two congenital curves (11%) each of which was due to a single hemivertebra on opposite sides of the spine. Six patients had multiple congenital curves (6%) associated with three or more hemivertebrae which alternated on either side of the spine.

For the purpose of this study the scoliotic curves were grouped according to the type of hemivertebra and then subdivided by the level of the hemivertebra or, if there were two unilateral hemivertebrae, the apex of the curve. The rate of deterioration of the untreated scoliotic

Tables I, II, III and IV shows the mean yearly rate of deterioration in degrees of scoliotic curves in untreated patients due to different types of hemivertebra occurring in different regions of the spine, both before and after 10 years of age.

**Table I.** Scoliosis due to a single fully segmented (non-incarcerated) hemivertebra

Site of hemivertebra	Number of patients	Rate of deterioration (degrees per year)	
		Before 10 years	After 10 years
T1 T4	4	1	1.5
T5 L1	18	1.4	3
L2 L4	9	0.7	1.7
L5	9		

**Table II.** Scoliosis due to two unilateral fully segmented (non-incarcerated) hemivertebrae

Apex of scoliosis	Number of patients	Rate of deterioration (degrees per year)	
		Before 10 years	After 10 years
T1 T4	4	1.5	2.25
T5 L1	13	3	3.2

**Table III.** Scoliosis due to a single semi-segmented hemivertebra

Site of hemivertebra	Number of patients	Rate of deterioration (degrees per year)	
		Before 10 years	After 10 years
T1 T4	2	0	0
T5 L1	6	0	< 1
L2 L4	6	0	1
L5	5		

**Table IV.** Scoliosis due to a single incarcerated hemivertebra

Site of hemivertebra	Number of patients	Rate of deterioration (degrees per year)	
		Before 10 years	After 10 years
T1 T4	3	0	0
T5 L1	7	0	< 1
L2 L4	1	0	< 1

curves was calculated for each type of hemivertebra and site of curve, both before the age of 10 years and also after 10 years, during the adolescent growth spurt (Tables I, II, III and IV).

### Fully segmented (non-incarcerated) hemivertebrae

**Single hemivertebra.** Forty patients had a single vertebra of this type producing a single congenital scoliosis. These hemivertebrae were considered in four regions of the spine each of which gave different problems and prognoses. Follow-up without treatment was for a mean of 5 years 10 months (range 1 to 13 years).

*T1 to T4.* Four patients had hemivertebrae in this region; three at T3 and one at T2. Three of these patients were seen before the age of 10 years when the mean rate of deterioration of their curves was 1° per year (range 0.7 to 1.1°). At the age of 10 years their mean curvature was 32° (range 28° to 39°). Two of these patients and a third, diagnosed later, were followed after the age of 10 years; the rate of deterioration increased slightly to 1.5° per year (range 1° to 2°). Although these curves deteriorated relatively slowly and never became very large, they caused a cosmetic deformity by elevation of one shoulder and in one patient tilting of the head to the opposite side. To control deformity, two patients had spinal fusion at the ages of 11 and 15 years when their curves measured 32° and 35° respectively, while the third patient reached skeletal maturity untreated with a 48° curve.

*T5 to L1.* Of 18 patients with hemivertebrae in this region of the spine (Figs 2, 3 and 4), eight were seen before the age of 10 years and followed without treatment; their mean rate of deterioration was 1.4° per year (range 1° to 2.3°). Four of these patients were last reviewed at a mean age of 6 years 1 month (range 3 years 4 months to 7 years 2 months), when their mean curvature was 32° (range 23° to 37°). The other four patients were followed to 10 years when their mean curvature was 41° (range 31° to 47°).

After the age of 10 years, 12 patients were followed without treatment and the mean rate of deterioration of the curves increased to 3° per year (range 2.5° to 4.5°). Three patients eventually had spinal fusion at a mean age of 12 years 10 months (range 12 to 15 years) when their mean curvature was 51° (range 44° to 58°). Nine patients reached skeletal maturity without treatment when their mean curvature was 46° (range 35° to 58°).

These lower thoracic and thoracolumbar curves produced only mild to moderate cosmetic deformity because the vertebrae were not significantly rotated and there was only a small rib hump (Fig. 4). Only two of the 18 curves were decompensated, causing the patient to list slightly to one side. An additional problem associated with two of the curves due to hemivertebrae at T6 was the development of a long compensatory thoracolumbar curve which later became fixed. This *secondary structural curve*, containing no congenital anomaly, was much more rotated than the primary congenital curve; it produced a rib hump which was the main cosmetic deformity. Both of these curves were first diagnosed after the age of 10 years and followed to skeletal maturity without treatment, when the thoracic congenital curves measured 44° and 50° and the structural compensatory curves 38° and 52° respectively.

*L2 to L4.* Nine patients had a single fully segmented hemivertebra in this region (Figs 5 and 6). Eight of these were seen before the age of 10 years when the mean rate of deterioration of their curves was 0.7° per year (range 0° to 1.4°). One patient was last seen at two years of age with a 24° curve, while a second patient, last seen at 6 years 8 months, had a 35° curve. The other six patients

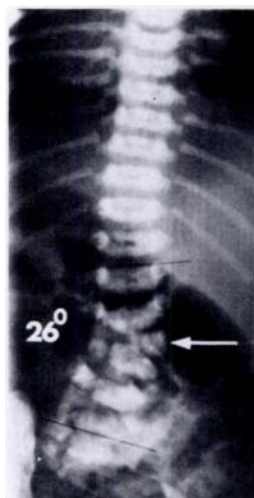


Fig. 5

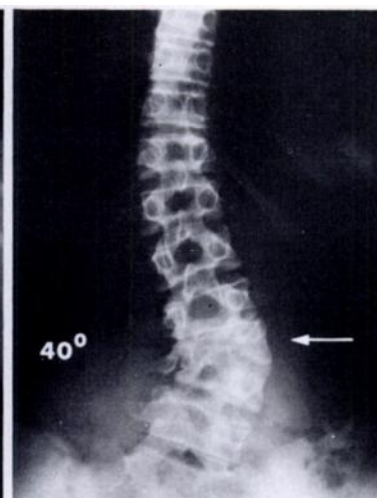


Fig. 6

Case 2. Figure 5 - A one-day-old girl with a 26° right lumbar scoliosis due to a fully segmented (non-incarcerated) hemivertebra at L4. Figure 6 - No treatment was given and by the age of 13 years the curve had deteriorated to 40°.

had been followed untreated to 10 years of age when their mean curvature was 33° (range 22° to 43°). Five of these patients and one other were followed untreated after the age of 10 years when the mean rate of deterioration of their curves increased to 1.7° per year (range 1° to 2.3°). Three patients were last seen between the ages of 11 and 14 years when their mean curvature was 42° (range 40° to 46°). The three patients who had been followed to skeletal maturity had a mean curvature of 45° (range 40° to 55°).

These lumbar curves produced only a relatively mild cosmetic deformity because they were not significantly rotated and there was no rib hump. None of the curves were decompensated and there was no listing of the trunk. The pelvis remained level in all but three patients who had mild leg-length discrepancy.

*Lumbosacral.* Nine patients with a single fully segmented hemivertebra at the lumbosacral junction showed a very short congenital scoliosis from L4 or L5 vertebra to the sacrum (Figs 7, 8 and 9). Three patients had six lumbar vertebrae and two had four lumbar vertebrae, including the hemivertebra.

The lumbosacral hemivertebra caused the lumbar spine to take off obliquely from the sacrum and, in order to compensate, all the patients developed a long secondary thoracolumbar or lumbar curve which was initially mobile but later became fixed. This *secondary structural curve*, which contained no congenital anomalies, extended upwards to T10, T11 or T12 and was always much larger and more rotated than the primary congenital curve. This secondary curve did not fully compensate for the congenital scoliosis and as a result the upper part of the body listed to the side opposite the hemivertebra in the six patients who had a level pelvis (Fig. 9). In three patients a leg-length discrepancy fortunately produced a compensatory pelvic obliquity, allowing the upper body to remain balanced.



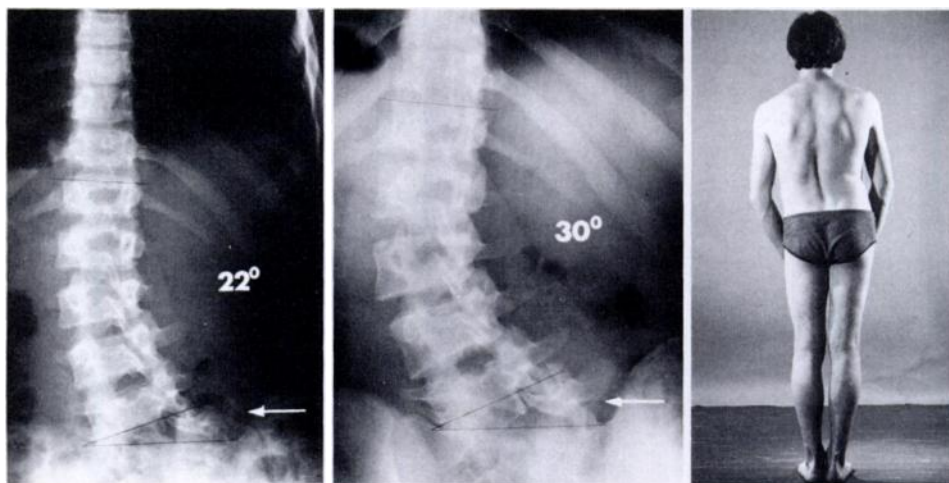


Fig. 7

Fig. 8

Fig. 9

Case 3. Figure 7 A boy aged 12 years 8 months with a single fully segmented (non-incarcerated) hemivertebra at the lumbosacral junction. This causes the lumbar spine to take off obliquely from the sacrum and a long secondary lumbar curve has developed which measures  $22^\circ$  and does not contain any congenital anomalies. Figure 8 No treatment was given and by the age of 17 years the secondary lumbar curve has deteriorated to  $30^\circ$ , and become fixed and rotated. The pelvis remains level. Figure 9 The main cosmetic deformity is due to the secondary lumbar curve and the tendency to list to the left.

Eight of the nine secondary structural curves behaved in a very similar manner. Only two of these patients were seen before the age of 10 years and neither of their curves exceeded  $40^\circ$ . After the age of 10, the eight secondary structural curves deteriorated at a mean rate of  $3^\circ$  per year (range  $2^\circ$  to  $4^\circ$ ). Five patients reached skeletal maturity without treatment with a mean curvature of  $44^\circ$  (range  $33^\circ$  to  $54^\circ$ ); all had a significant cosmetic deformity (Fig. 9). Two patients had spinal fusion at the age of 13 years 6 months when they both had secondary structural curves measuring  $50^\circ$ . The eighth patient was last seen untreated at 13 years when she had a  $40^\circ$  secondary curve. The ninth patient had a much more malignant deformity; she was first seen at the age of 3 years 5 months with a  $42^\circ$  secondary curve which deteriorated at  $3^\circ$  per year up to the age of 10 years and at  $7^\circ$  per year thereafter. This curve reached  $100^\circ$  at the age of 13 years 5 months resulting in a very severe deformity.

**Double unilateral hemivertebrae.** Seventeen patients had two fully segmented hemivertebrae on the same side of the spine. The two hemivertebrae were separated by from 1 to 4 relatively normal vertebrae (mean 2). The patients were followed without treatment for a mean of 3 years 3 months (range 6 months to 12 years).

**T1 to T4.** Four patients with two unilateral hemivertebrae each had single curves whose apex lay between T1 and T4 inclusive (Figs 10 and 11). Three of these patients were seen before the age of 10 when the mean rate of deterioration was  $1.5^\circ$  per year (range  $1^\circ$  to  $2^\circ$ ). By the age of 7 years 8 months, one patient had developed a  $43^\circ$  curve which caused a significant cosmetic deformity, by elevating the shoulder and tilting the head (Fig. 11), and needed treatment. Two patients reached 10 years without treatment when they had curves of  $47^\circ$  and  $40^\circ$ ; these also caused significant deformity from an elevated shoulder.

After the age of 10 years, the rate of deterioration of these curves increased to  $2^\circ$  and  $2.5^\circ$  per year; both were treated by spinal fusion at the ages of 12 and 13 years for curves measuring  $51^\circ$  and  $48^\circ$  respectively.

**T5 to T12.** Thirteen patients had two unilateral hemivertebrae with a single curve with apex from T5 to T12 (Fig. 12). Before the age of 10 years, 10 of these patients showed a mean rate of deterioration of  $3^\circ$  per year (range  $1.5^\circ$  to  $4^\circ$ ). Seven of these were eventually treated in a brace at a mean age of 3 years 5 months (range 2 years 2 months to 4 years 10 months), when their mean curve was  $53^\circ$  (range  $43^\circ$  to  $74^\circ$ ). The other three patients in this subgroup were last reviewed at a mean age of 9 years 6 months (range 8 to 10 years) when their mean curvature was  $50^\circ$  (range  $43^\circ$  to  $55^\circ$ ). After the age of 10 years,

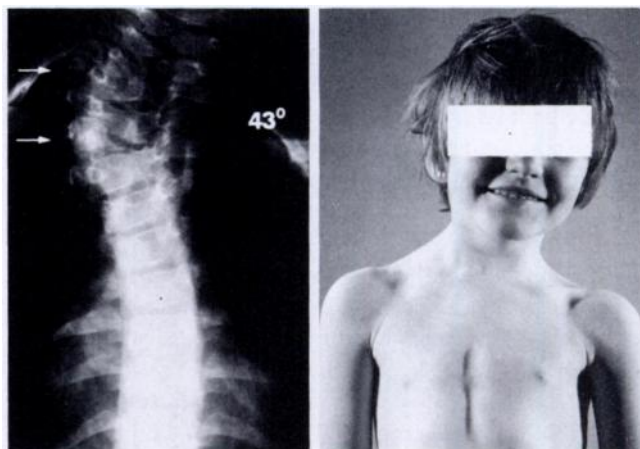


Fig. 10

Fig. 11

Case 4. Figure 10 - A girl aged 7 years 8 months with a  $43^\circ$  right upper thoracic scoliosis due to two unilateral fully segmented (non-incarcerated) hemivertebrae at T2 and T4. Figure 11 Her cosmetic deformity is due to elevation of the right shoulder and slight tilting of the head to the left.

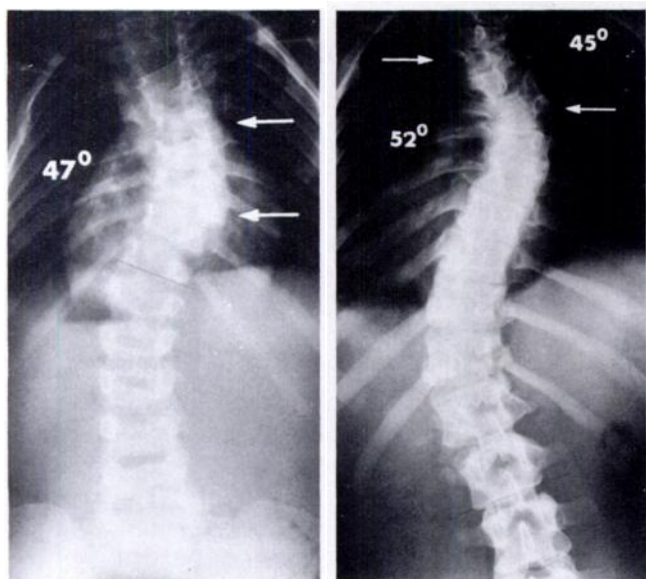


Fig. 12

Fig. 13

Cases 5 and 6. Figure 12 - A boy aged 4 years 10 months with a 47 right thoracic scoliosis due to two unilateral fully segmented (non-incarcerated) hemivertebrae at T6 and T9. Figure 13 - A skeletally mature girl aged 18 years with two opposing hemivertebrae in the same region at T4 and T6. These hemivertebrae produce curves which balance each other and cause minimal cosmetic deformity.

four patients followed without treatment showed an increased mean rate of deterioration of 3.2 per year (range 2.5 to 4). Three of these patients had a spinal fusion at a mean age of 14 years 7 months (range 12 years 3 months to 16 years 2 months), when their mean curve was 60 (range 57 to 62); the fourth was untreated at skeletal maturity when his curve was 72.

All these patients developed a significant cosmetic deformity before the age of five years, due to the severity of their curve and the presence of a rib hump. In addition, 6 of the 13 patients had decompensated curves and listed to one side. Two patients with curves whose apex was at T6 and T7 developed secondary structural curves in the thoracolumbar region. These two patients were untreated at age 16; their congenital curves measured 50 and 60 and their structural compensatory curves were 82 and 77 respectively.

**Double opposing hemivertebrae.** Eleven patients had two fully segmented hemivertebrae on opposite sides of the spine, resulting in double congenital scoliosis. These patients could be divided between three subgroups, depending on the sites and relationship of the hemivertebrae: each had a different prognosis.

1. Four patients had two opposing hemivertebrae occurring in the *same region* within one or two segments of each other, three in the thoracic spine and one in the lumbar region. These pairs of hemivertebrae caused small kinks in the spine which never became large and balanced each other to produce minimal cosmetic deformity at maturity (Fig. 13).

2. Two patients had opposing hemivertebrae that were more widely separated and in *different regions* of the spine. These hemivertebrae produced much bigger

curves, similar to those produced by single hemivertebrae in the same region. The resulting curves were unbalanced and caused listing of the upper trunk to one side. One patient had opposing lower thoracic and lumbar curves which measured 54 and 61 at the age of 10 years and required treatment because of decompensation of the trunk (Figs 14 and 15). The second patient had opposing lower thoracic and thoracolumbar curves which measured 48 and 49 respectively at the age of 12 years 6 months. This patient was balanced by slight shortening of one leg which compensated for the tendency to list.

3. Five patients had the combination of a *lumbosacral hemivertebra with a second hemivertebra on the opposite side in the lumbar region* (L1, L2 and three at L3). These patients were all unbalanced, listing to the side opposite the lumbosacral hemivertebra. Four of the patients were followed without treatment to skeletal maturity. Their lumbar curves measured 50, 54, 55 and 56 and they all had a moderate cosmetic deformity. The fifth patient had a decompensated 64 lumbar curve at the age of 14 years 7 months and was then treated by spinal fusion.

#### Semi-segmented hemivertebrae

Nineteen patients had a single semi-segmented hemivertebra producing a single congenital scoliosis. Seventeen of these patients were followed without treatment for a mean of 6 years 1 month (range 2 to 15 years) and two were first seen untreated at skeletal maturity.

*T1 to T4.* Two patients had a single semi-segmented hemivertebra, both at T3. Neither curve progressed, and at the most recent reviews at the ages of 12 and 13 years the curves measured 24 and 32 respectively.

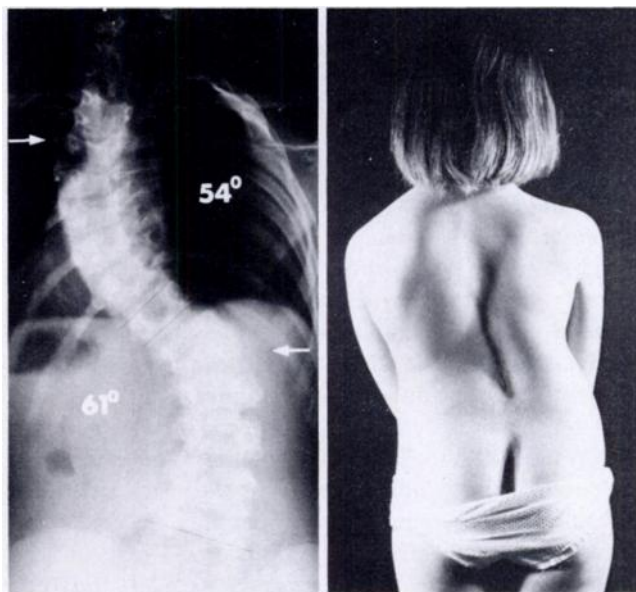


Fig. 14

Fig. 15

Case 7. Figure 14 - A girl aged 10 years with two opposing hemivertebrae occurring in different regions at T5 and L2. These hemivertebrae have produced two curves which are unbalanced. Figure 15 The patient lists to the left and there is a significant cosmetic deformity.



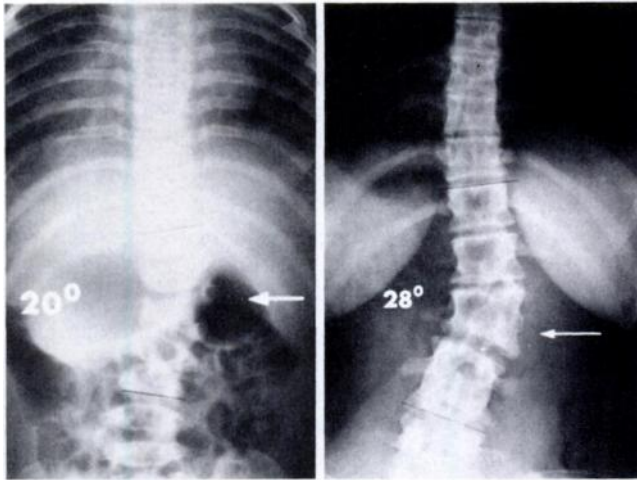


Fig. 16

Fig. 17

Case 8. Figure 16 A girl aged 6 months with a 20° right lumbar scoliosis due to a semi-segmented hemivertebra at L2. The hemivertebra is synostosed with L3. Figure 17 No treatment was given and the curve deteriorated very slowly to measure 28° at 15 years 8 months when the patient was skeletally mature.

*T5 to L1.* Six patients had semi-segmented hemivertebrae in this region. Only two patients were seen before the age of 10 years when their curves were less than 25° and did not deteriorate. The other four patients were followed after the age of 10 years when their curves deteriorated at less than 1° per year. One patient was last seen at age 13 when her curve measured 27°; three patients reached skeletal maturity without treatment with a mean curvature of 34° (range 31° to 39°).

*L2 to L4.* Six patients had a lumbar semi-segmented hemivertebra (Figs 16 and 17). Five of them were seen before the age of 10 years; four of the curves did not progress and one deteriorated at less than 1° per year, giving a mean curvature at 10 years of age of 27° (range 18° to 36°). After the age of 10, four untreated patients were followed and the mean rate of deterioration of their curves increased to 1° per year (range 0° to 1.5°). Two patients last reviewed at 11 and 14 years had curves measuring 18° and 33° respectively, while the other two patients reached skeletal maturity without treatment with curves of 28° and 37°.

*Lumbosacral.* Five patients had a lumbosacral hemivertebra which was synostosed to the sacrum. These patients presented between the ages of 10 and 14 years because of the secondary structural curve above the hemivertebra and the tendency for the upper trunk to list to the side opposite the congenital anomaly. These secondary curves deteriorated at a mean rate of 2° per year (range 1.8° to 2.4°); three of the patients reached skeletal maturity without treatment with curves between 26° and 31°. The remaining two patients received treatment to control their list when they were 14 years old and their secondary structural curves measured 20° and 31°.

#### Incarcerated hemivertebrae

Eleven patients had a single incarcerated hemivertebra and all their curves behaved in the same manner regard-

less of their site (Figs 18 and 19). Eight patients were diagnosed in the first year of life with a mean curvature of 17° (range 11° to 22°). All 11 patients were followed without treatment; before the age of 10 eight curves did not progress and three deteriorated at less than 0.5° per year, so that at 10 years no curve exceeded 28°. Three patients were followed untreated after 10 years and their curves did not deteriorate. One patient had reached skeletal maturity with a 23° curve.

#### Multiple hemivertebrae

Six patients had multiple small curves due to hemivertebrae which alternated on either side of the thoracic and thoracolumbar spine. Two patients had three hemivertebrae, three had four hemivertebrae each, and one patient had six. These were a mixture of all three basic types of hemivertebrae. The resulting curves were closely associated and tended to balance each other, causing little deformity other than stunting of the spine.

#### DISCUSSION

This study has shown that the potential for a hemivertebra to cause a significant scoliosis depends on four factors: first, and most important, the type of hemivertebra; secondly, its site; and thirdly, the number of hemivertebrae and their relationship to each other. Finally, the age of the patient is also important because curves due to hemivertebrae often deteriorate much more rapidly during the adolescent growth spurt (Tables I, II, III and IV). It is only at skeletal maturity, when the vertebral growth plates fuse, that the potential for increasing deformity ceases.

The commonest pathological type of hemivertebra (65%) is fully segmented and non-incarcerated. Here there is absence of two growth plates on the unformed

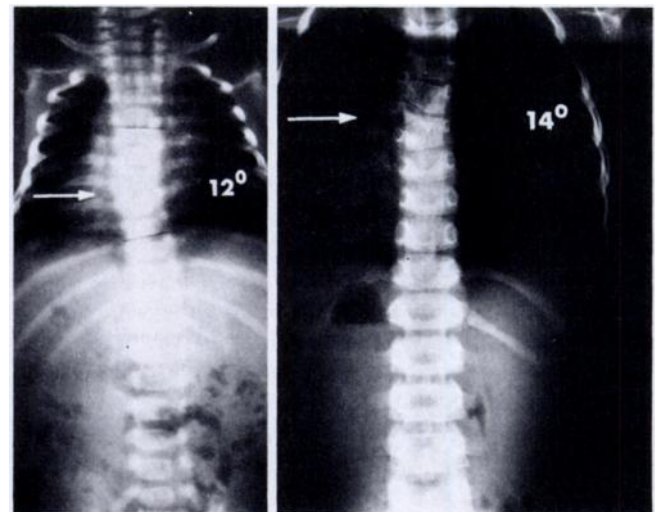


Fig. 18

Fig. 19

Case 9. Figure 18 An infant aged two months with a 12° left thoracic scoliosis due to an incarcerated hemivertebra at T6. Figure 19 No treatment was given and there was virtually no deterioration at 13 years 8 months.

side of the hemivertebra in contrast to the two relatively normal growth plates, one on each surface of the developed part of the vertebra. This means that as the hemivertebra grows it acts as an enlarging wedge.

An incarcerated hemivertebra is the least common type (13%), and although it is fully segmented, it has a poor growth potential. The resulting scoliosis deteriorates at less than 1° per year and these curves, regardless of their site, never exceed 30° at skeletal maturity (Figs 18 and 19) so treatment is not required.

A semi-segmented hemivertebra (22%) is congenitally synostosed to one of its neighbouring vertebra and, as a result, two growth plates are absent on the convexity of the curve. This tends to balance the growth plates on the two sides of the hemivertebra although the hemivertebra itself causes tilting of the spine and can induce a progressive scoliosis (Figs 16 and 17). These curves do not exceed 40° at skeletal maturity and treatment is not usually required except occasionally when the hemivertebra is at the lumbosacral junction.

Fully segmented non-incarcerated hemivertebrae cause most problems. A single such hemivertebra is most common and the prognosis for deterioration of the resulting curve depends on the site of the hemivertebra.

*Upper thoracic curves* (hemivertebrae T1 to T4) deteriorate slowly but can reach 40° by skeletal maturity. Although these curves are not severe, they can cause a cosmetic deformity by elevation of the shoulder; this becomes apparent when the curve exceeds 30° and may be distressing enough in girls to warrant early prophylactic treatment. This is especially true if there is an associated Sprengel's shoulder on the same side as the hemivertebra.

*Lower thoracic and thoracolumbar curves* (hemivertebrae T5 to L1) deteriorate more rapidly and can exceed 45° at skeletal maturity, but these curves are not usually as rotated as an equivalent idiopathic curve and do not produce as severe a cosmetic deformity (Figs 3 and 4).

*Lumbar curves* (hemivertebrae L2 to L4) deteriorate relatively slowly at a rate similar to those in the upper thoracic region. They can reach 45° by skeletal maturity but cause only a relatively mild cosmetic deformity.

*Lumbosacral* hemivertebrae are important because they cause the lumbar spine to take off obliquely from the sacrum, resulting in the development of a large secondary structural thoracolumbar curve. This curve is usually significantly rotated and when combined with the trunk listing to the side opposite the hemivertebra, results in moderate to severe cosmetic deformity (Figs 8 and 9). These patients require early prophylactic treatment before they begin to list and before the secondary curve becomes structural.

Two unilateral fully segmented hemivertebrae are much less common but cause a much greater growth imbalance because four of the growth plates on the concavity of the curve are absent. These curves deteriorate much more rapidly than those due to a single fully segmented hemivertebra (Table II). Upper thoracic curves of this type (apex T1 to T4) usually exceed 40° by 10 years of age, causing significant cosmetic deformity due to shoulder elevation and head tilting (Figs 10 and 11). Lower thoracic and thoracolumbar curves (apex T5 to T12) deteriorate most rapidly of all at 3° per year and frequently exceed 50° by 5 years of age. Without treatment these curves could reach 70° by skeletal maturity and therefore all curves due to two unilateral fully segmented hemivertebrae require prophylactic treatment as soon as they are diagnosed.

If there are two opposing fully segmented hemivertebra the prognosis depends on whether the hemivertebrae are close together or in different regions of the spine. If they are close together they tend to balance each other and only cause two small kinks in the spine with minimal cosmetic deformity (Fig. 13). No treatment is required. If the hemivertebrae are in different regions, the resulting curves are often unbalanced and cause the trunk to list to one side (Figs 14 and 15). When this occurs prophylactic treatment is required.

Multiple hemivertebrae are often a mixture of types but the spine usually remains balanced and there is no deformity apart from some stunting of the trunk. No treatment is required.

In conclusion, it appears that incarcerated and semi-segmented hemivertebrae do not usually require treatment. The fully segmented non-incarcerated hemivertebra may require treatment, but this depends on the site, number and relationships of the anomaly or anomalies. If treatment is necessary, this should be anticipated and carried out at an early age because it is much easier to prevent than it is to correct severe deformity resulting from hemivertebrae. In planning prophylactic treatment, it should be appreciated that it is not possible to create growth on the unformed side of the hemivertebra and treatment should therefore be directed towards preventing the deforming growth of the hemivertebra.

#### REFERENCES

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