The Natural History of Congenital Scoliosis

A Study of Two Hundred and Fifty-one Patients

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ABSTRACT: **The cases of 251 patients with untreated congenital scoliosis were studied, and 216 patients were followed without treatment for an average of 5.1 years. Most of the curves were first seen when the patient was either in the first two years of life or at puberty, when there was an increased rate of deterioration. An early onset carried a bad prognosis. Of the 143 patients who were last seen, without treatment, after the age of ten years, fifty-one (36 per cent) had a curve of 40 to 60 degrees and forty (28 per cent) had a curve of more than 60 degrees. We concluded that the prognosis in such patients is even worse because sixty-seven patients in our series required treatment at or before the age of ten years due to the severity of the curve: twenty of them had a curve of 40 to 60 degrees and thirty-nine had a curve of more than 60 degrees.**

Radiographically, 90 per cent of the curves could be classified into five groups. The rate of deterioration and the ultimate severity of the curve were found to depend on both the type of anomaly and the site at which it occurred. Increasingly severe and progressive scoliosis, regardless of the area of the spine affected, developed when there was a block vertebra, a wedge vertebra, a single hemivertebra, two unilateral hemivertebrae, a unilateral unsegmented bar, or, most severe, a unilateral unsegmented bar with contralateral hemivertebrae at the same level. For each of these types of anomaly, the rate of deterioration was usually less severe if the abnormality was in the upper thoracic region, more severe in the thoracic region, and most severe in the thoracolumbar region. The median yearly rate of deterioration for each type and site of curve without treatment before and after the patient was ten years old was evaluated. Secondary problems due to tilting of the head, elevation of the shoulder line, decompensation of the trunk, pelvic obliquity, and the formation of large secondary structural curves also occurred, and contributed significantly to the over-all disability and deformity.

Congenital scoliosis is a lateral curve of the spine that is due to the presence of vertebral anomalies that cause an imbalance in the longitudinal growth of the spine. These vertebral anomalies develop during the first six weeks of intrauterine life, when the anatomical pattern of the spine is formed in mesenchyme. Once the mesenchymal mold is established, the cartilaginous and osseous stages follow that pattern. The vertebral abnormality is present at birth, but the clinical deformity may not become evident until later in childhood when a scoliosis develops and the diagnosis can be made radiographically. Some anomalies cause so little deformity that they remain undetected, so the true incidence of congenital scoliosis in the general population remains unknown. Wynne-Davies found that multiple vertebral anomalies were often hereditary in origin, but the occurrence of an isolated single anomaly was usually sporadic.

The radiographic appearance of the vertebral anomalies varies considerably and, as a result, congenital scoliosis was for many years thought to be unpredictable in its behavior. Some thought that it seldom required treatment[1,3]. In 1952, Kuhns and Hormell reviewed the cases of 165 children and concluded, as did many physicians at that time, that a congenital scoliosis usually is relatively benign and progresses slowly, if at all. It was not until 1968 that Winter et al., in a study involving 234 children, firmly established the much more serious prognosis for certain types of congenital scoliosis. Apart from these two papers, there have been very few reports of the natural history of congenital scoliosis in large numbers of unselected patients[6].

A congenital scoliosis often is rigid and correction can be difficult. It therefore is important to be able to anticipate when a congenital scoliosis is at risk for rapid deterioration and to initiate treatment when the curve is small rather than to attempt the dangerous surgical salvage procedures that are necessary when the deformity is severe. Planning such a prophylactic course of treatment requires a more thorough knowledge of the natural history of all types of congenital scoliosis than is presently available.

The purpose of this study was to investigate the natural history of congenital scoliosis in a large number of patients. All came from one area and were seen at the Edinburgh Scoliosis Clinic, and most were followed for long periods. This clinic is the only referral point for patients with congenital scoliosis from a large population. It was hoped that this paper would provide indications as to the necessity for treatment of the various types of congenital scoliosis.

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Classification

We classified the types of vertebral anomaly causing congenital scoliosis in the patients in this study according to the system of Winter et al. We added one subcategory, as described by Nasca et al., because it has a particularly severe prognosis. Under the category of simple anomalies, we included failure of segmentation, which consists of: (1) a unilateral unsegmented bar, (2) unilateral failure of segmentation with contralateral hemivertebrae at the same level[6], and (3) bilateral failure of segmentation (block vertebrae).

The other simple anomaly in our series was unilateral failure of formation of a vertebral body, which was either: (1) complete (hemivertebra) or (2) partial (wedge vertebra).

Complex (unclassifiable) anomalies consisted of a jumble of vertebral anomalies that were too varied for separate classification. In some patients included in this group it was impossible to identify the type of anomaly because the severity of the curve was so great.

Occasionally other congenital vertebral anomalies were present at levels in the spine other than those involving the scoliotic curve, but if they did not contribute to the scoliosis they were ignored in the classification. Anomalies of the ribs were very common, but they too were ignored because they did not cause a scoliosis by themselves.

In addition to determining the type of vertebral anomaly, the scoliosis was also classified according to the site of the apex of the curve, which frequently coincided with the site of the anomaly. A scoliosis whose apex lay between the second and sixth thoracic vertebrae, inclusive, was termed an *upper thoracic curve*. A scoliosis whose apex was located between the seventh and eleventh thoracic vertebrae, inclusive, was a *lower thoracic curve*. A *thoracolumbar scoliosis* was one with its apex at the twelfth thoracic or first lumbar vertebra. The apex of a *lumbar scoliosis* was between the second and fourth lumbar vertebrae, inclusive, and the apex of a *lumbosacral scoliosis* was at the fifth lumbar vertebra[8].

A patient with a single congenital scoliosis was considered to have only one structural curve due to the presence of congenital vertebral anomalies. Several patients were classified as having a single congenital scoliosis even though a second structural scoliosis developed either above or below the primary congenital curve. This second curve was not classified as a congenital scoliosis because it did not result from a congenital vertebral anomaly.

A patient was considered to have a double congenital scoliosis if there were two opposing structural curves, each due to separate congenital vertebral anomalies that might or might not be similar. A few patients had more than two structural curves because of separate congenital vertebral anomalies.

Skeletal maturity was determined radiographically when there was complete ossification and fusion of the iliac apophyses[11].

Materials and Methods

A study was made of all patients seen at the Edinburgh Scoliosis Clinic between 1958 and 1981 who were diagnosed as having congenital scoliosis. The majority of these patients were seen after 1965. Patients who had myelomeningocele, Klippel-Feil syndrome, or congenital kyphosis or kyphoscoliosis in which the kyphosis was the predominant deformity were excluded because the natural history of those deformities was very different from that of the ones under study. There were 251 patients who had a true congenital scoliosis that was diagnosed radiographically; that is, each had a lateral curve of the spine due to the presence of congenital vertebral anomalies. Radiographs made elsewhere, prior to referral, were often available, and sometimes provided evidence of progression of the deformity.

There were 179 female and seventy-two male patients, whose ages at diagnosis ranged from birth to twenty-two years (Fig. 1). They had a total of 269 curves (Table I).

Two hundred and sixteen patients were followed for a mean of 5.1 years (range, six months to seventeen years). At their last visit to the clinic, 108 patients had not been treated, and forty-six of them had reached skeletal maturity. The mean length of follow-up for these patients was 7.3 years. The sixty-two patients who were skeletally immature were still attending our clinic, without treatment, after a mean of 5.1 years. The remaining 108 patients had been followed without treatment for a mean of 4.2 years before eventually being treated either with a Milwaukee brace or with spine fusion; the data on these patients relate to the pretreatment interval.

No longitudinal data could be obtained on the natural history of the remaining thirty-five patients, since twenty-three of them received immediate treatment when they were first seen because of the severity of the deformity and the other twelve were skeletally mature when first seen. The findings in these patients are included in some of the analyses in this study because prior to attending our clinic they had been untreated, and therefore contributed data on the natural history of congenital scoliosis.

The sites and types of congenital scoliosis were diagnosed by reviewing the radiographs of the spine, made with the patient standing. Using the Cobb method, care was taken to remeasure from the same levels that were used on the earliest radiograph. Pelvic obliquity was measured on the radiographs of the spine by noting the angle subtended between a line drawn across the tops of the iliac crests and the bottom of the radiograph, which was horizontal when the patient was standing. Decompensation or listing of the trunk and tilting of the head were assessed on the basis of the spine radiographs and the clinical photographs made with the patient standing.

Results

The incidences of the various sites and types of congenital scoliosis are shown in Table I.

TABLE I

SITES AND TYPES OF CONGENITAL SCOLIOSIS (TWO HUNDRED AND FIFTY-ONE PATIENTS)

Site of Curve	Type of Congenital Vertebral Anomaly (*No. of Curves*)					
	Unilateral Unsegmented Bar	Unilateral Unsegmented Bar with Contralateral Hemivertebrae	Block Vertebra	Hemivertebra	Wedge Vertebra	Unclassifiable
Upper thoracic	40	7	7	23	1	5
Lower thoracic	39	11	2	21	4	13
Thoracolumbar	17	10	3	13	3	8
Lumbar	6	0	1	19	1	3
Lumbosacral	0	0	0	12	0	0

A single congenital curve occurred in 238 patients: 216 of the curves were due to simple anomalies and twenty-two, to complex anomalies.

Two or more congenital curves occurred in thirteen patients. In ten of the patients the curves were due to simple anomalies and in three the anomalies were complex.

The ages of the patients and the sizes of the congenital curves at the initial visit are shown in Figure 1. In patients with two or more congenital curves, only the largest curve was used for tabulation. Most of our patients were diagnosed radiographically either in the first two years of life (seventy-seven patients) or between the ages of nine and fourteen years (sixty-eight patients). The degree of severity of the curve did not correlate with the age at presentation. Severe curves of more than 80 degrees were seen not only in older children but also at birth and in the first year of life. Minor curves were seen in patients in all age groups.

Severity of the Curve and Rate of Deterioration

Of the 251 patients in this study, 173 were seen, untreated, at or before the age of ten years. Thirty-eight patients (22 per cent) had a curve of 20 degrees or less, forty-seven (27 per cent) had a curve of 21 to 39 degrees, forty-one (24 per cent) had a curve of 40 to 60 degrees, and forty-seven (27 per cent) had a curve of more than 60 degrees (range, 62 to 148 degrees).

Sixty-seven of these 173 patients were eventually treated at or before the age of ten years because of the severity of the deformity. Before treatment, eight had a

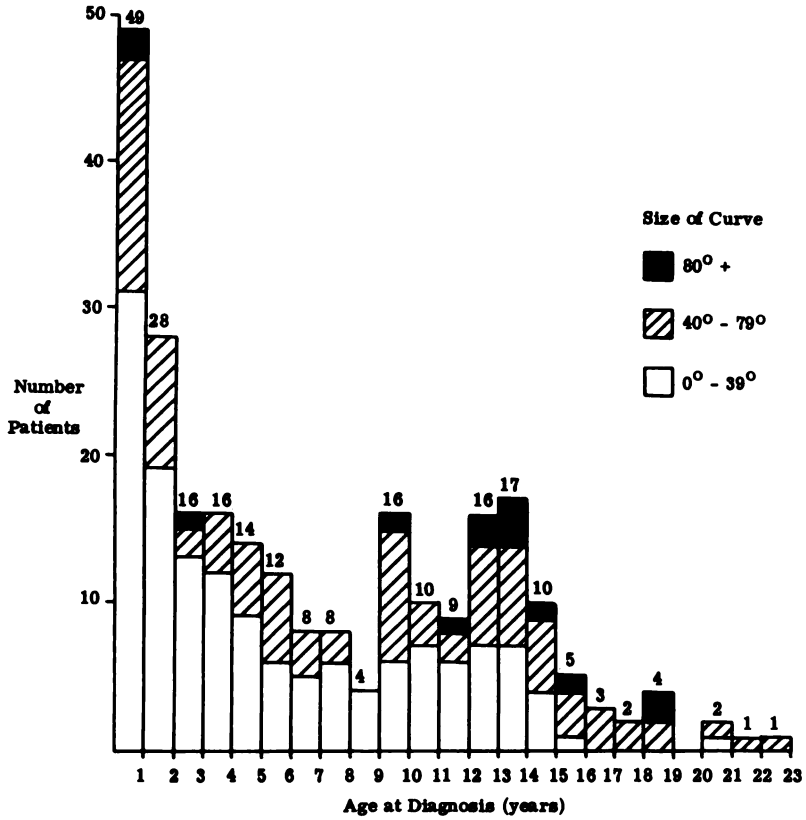

FIG. 1

Ages of the patients and severity of the congenital scolioses at diagnosis.

curve of 20 to 39 degrees, twenty had a curve of 40 to 60 degrees, and thirty-nine had a curve of more than 60 degrees.

One hundred and forty-three patients were last seen, without treatment, after they were ten years old. Fifteen of them (10 per cent) had a curve of 20 degrees or less, thirty-seven (26 per cent) had a curve of 21 to 39 degrees, fifty-one (36 per cent) had a curve of 40 to 60 degrees, and forty (28 per cent) had a curve of more than 60 degrees (range, 64 to 156 degrees).

Fifty-eight patients reached skeletal maturity without treatment. Of these patients, six (10 per cent) had a curve of 20 degrees or less, twenty-two (38 per cent) had a curve of 21 to 39 degrees, nineteen (33 per cent) had a curve of 40 to 60 degrees, and eleven (19 per cent) had a curve of more than 60 degrees (range, 66 to 156 degrees).

All of the large curves that were followed without treatment should have been treated at a much earlier stage, regardless of the patient's age.

We found, as did Winter et al., that the rate of deterioration and the ultimate severity of a congenital scoliosis depended on both the type of vertebral anomaly and the site of the curve. In addition, we found that the rate of deterioration was not constant but tended to increase after the age of ten years.

In order to obtain a more complete understanding of the natural history of all types of congenital scoliosis, we studied the rate of deterioration in degrees per year and the severity of the curve, both before and after the age of ten years, for each type of vertebral anomaly producing a scoliosis in each region of the spine (Tables II through VI). The degree of scoliosis was followed serially and remained fairly constant until between the ages of nine and eleven years, when it tended to increase, especially in patients with unilateral failure of segmentation. The rate of deterioration before and after the age of ten years was estimated by subtracting the initial angle from the final angle of the curve on the routine spine radiographs made with the patient standing, and dividing the increment by the number of years and months. We calculated the median rate of deterioration rather than the mean for each specific type of congenital scoliosis occurring at each specific site, and this value was found to be more representative of each subgroup as a whole and less distorted by values that were greatly different from those of the majority (Fig. 8).

Multiple Congenital Curves

Before analyzing the single curves, we studied the cases of the thirteen patients with two or more congenital curves due to simple (Table II) and complex anomalies.

Three patients (Cases 1, 2, and 3) had double curves due to unilateral unsegmented bars on opposite sides and in different regions of the spine. These double curves showed a high rate of deterioration and became large and unbalanced, causing a list of the trunk that necessitated correction and fusion soon after the patient was ten years old.

Four patients had two hemivertebrae, but on opposite sides and at different levels of the spine. In two of these patients (Cases 4 and 5) the hemivertebrae occurred in the thoracic region, within one or two segments of each other and on opposite sides of the spine. These hemivertebrae caused two small kinks that never became large and that balanced each other, producing a minimum deformity at maturity. Two other patients (Cases 6 and 7) had opposing hemivertebrae that were much more widely separated, on opposite sides, and in different regions of the spine. These hemivertebrae produced bigger curves that were unbalanced, causing a list of the trunk and requiring correction and fusion at the ages of ten and fourteen years.

Two patients (Cases 8 and 10) had three hemivertebrae and one patient (Case 9) had four hemivertebrae which alternated on either side of the spine in the thoracic and thoracolumbar regions. The resulting curves tended to balance each other, but in one patient (Case 8) the upper hemivertebra was at the first thoracic level and this caused an elevation of the shoulder line, requiring correction and fusion of the upper curve at the age of eight years.

Three patients had multiple small curves due to complex anomalies. These curves were closely associated with one another and tended to balance each other, causing little deformity other than stunting of the spine. One patient, however, had an elevated shoulder line that necessitated correction and fusion of the upper thoracic curve at the age of nine years.

Single Congenital Curves

For the purpose of analysis of the natural history of the curves, each of the six groups was divided into three subgroups. Subgroup A comprised those patients for whom serial observations had not been made because they were either treated immediately or were first seen, untreated, at skeletal maturity. Subgroup B contained those patients who had been followed for variable periods before the age of ten years. Subgroup C contained those patients who had been followed at or after the age of ten years. There was some overlap between Subgroups B and C.

Unilateral Failure of Segmentation (Unilateral Unsegmented Bar) (Group 1)

A unilateral unsegmented bar was the cause of a congenital scoliosis in ninety-nine patients and was always present on the concavity of the curve. Ninety-six patients had a single congenital scoliosis and three (Table II, Cases 1, 2, and 3) had two separate and opposing unbalanced congenital curves, each due to a unilateral unsegmented bar but on opposite sides and at different levels of the spine. The unsegmented bars in the ninety-nine patients did not demonstrate a preference for side but were seen more frequently in the thoracic region (seventy-nine patients) than elsewhere. In three patients the unilateral unsegmented bar was not recognized in infancy and was only diagnosed retrospectively, between the ages of three and four years, when the bar began to ossify and appeared on

TABLE

NATURAL HISTORY OF TEN PATIENTS WITH MORE

Case	Sex	Vertebral Anomaly		Curve		Age (Yrs. + Mos.) When First Seen	When Last Seen	Length of Follow-up (Yrs. + Mos.)
		Type	Site	Side	Extent			
1	F	Unseg. bar	T4-T9	L	T3-T12	3 + 0	13 + 6	10 + 6
		Unseg. bar	L2-L4	R	L1-L5			
2	F	Unseg. bar	T2-T5	L	T2-T5	0 + 2	2 + 3	2 + 1
		Unseg. bar	T11-L1	R	T11-L3			
3	F	Unseg. bar	T3-T5	R	T1-T7	8 + 2	11 + 0	1 + 8
		Unseg. bar	L1-L3	L	T7-L3			
4	F	Hemivert.	T3	L	T2-T4	2 + 3	17 + 0	14 + 9
		Hemivert.	T6	R	T5-T7			
5	F	Hemivert.	T2	R	T1-T3	3 + 1	4 + 4	1 + 3
		Hemivert.	T5	L	T4-T6			
6	F	Hemivert.	T7	L	T2-T12	3 + 0	10 + 0	7 + 0
		Hemivert.	L2	R	T12-L5			
7	F	Hemivert.	L1	L	T11-L4	13 + 11		None
		Hemivert.	L6	R	L4-S1			
8	M	Hemivert.	T1	R	C7-T3			
		Hemivert.	T10	R	T8-T11	6 + 3	8 + 1	1 + 10
		Hemivert.	L1	L	T11-L4			
9	F	Hemivert.	T6	L	T4-T8			
		Hemivert.	T9	R	T8-T11	20 + 0		None
		Hemivert.	T12	L	T11-L1			
		Hemivert.	L4	R	L3-L5			
10	F	Hemivert.	T5	L	T3-T6			
		Hemivert.	T8	R	T7-T9	0 + 5	3 + 6	3 + 0
		Hemivert.	L4	L	L1-L5			

radiographs. The rate of progression of these three curves did not differ significantly from that of the other curves in the same regions. The prognosis for congenital scoliosis due to a unilateral unsegmented bar depended mainly on the site of the curve and partly on the extent of the bar. The extent ranged from two to eight vertebrae. However, there was not always a direct relationship between the length of the bar and the rate of progression of the curve. The longer unsegmented bars all tended to produce the larger curves in a specific region, but occasionally a shorter bar produced an equally large curve in the same region.

The seventy-two patients who were followed without treatment were considered to be in Groups 1B and 1C (Table III). The twenty-four patients for whom serial observations had not been made were in Group 1A and were excluded from further analysis.

Upper Thoracic Curves

Of the thirty-eight patients who had an upper thoracic curve, ten were in Group 1A. Nine of them were treated immediately when they were first seen, between the ages of two and fifteen years, and had curves ranging from 26 to 85 degrees. One patient was seen at skeletal maturity, with a curve of 68 degrees.

The remaining twenty-eight patients with an upper thoracic curve were in Groups 1B and 1C (Table III) and were followed without treatment for a mean of 4.7 years (range, nine months to fourteen years and two months). Before they reached the age of ten years, the median rate of deterioration was 2 degrees per year. By the age of ten years, ten of them had a curve of more than 30 degrees. Twelve patients were seen, untreated, at the age of ten years, when the mean curve was 33 degrees (range, 13 to 76 degrees). Two patients had a more rapid rate of deterioration before the age of ten years. One of them, who had a bar involving seven vertebrae, had a 5-degree curve at birth which deteriorated at a rate of 6 degrees per year until it was 62 degrees at the age of ten years (Figs. 3-A and 3-B). The second patient had a bar involving three vertebrae which produced a 24-degree curve at the age of three years and deteriorated at a rate of 5 degrees per year until it was 53 degrees at the age of nine years. The case of this patient demonstrated that even a relatively short unsegmented bar can produce a large curve.

After the patients were ten years old, the median rate of deterioration increased to 4 degrees per year, and sixteen patients required a spine fusion at a mean age of thirteen years (range, eleven to fifteen years), at which time the mean curve was 59 degrees (range, 30 to 98 degrees). The most rapid rates of deterioration occurred in four of these patients, who were followed untreated prior to spine fusion for periods ranging from nine months to two years and six months. A bar involving three to five vertebrae was present in three of these curves, which deteriorated at a rate of 8 degrees per year. A bar involving seven vertebrae was present in one curve, which deteriorated at a rate of more than 10 degrees per year and measured 90 degrees at the age of twelve. Not all of the upper thoracic curves deteriorated at the same rate, however, and five of the ten curves that were followed, untreated, to maturity then

II

THAN ONE CURVE DUE TO SIMPLE ANOMALIES

Size of Curve (*Degrees*)				
When First Seen	When Last Seen	Total Change	Rate of Change per Year	Comments
22	122	100	9.5	Unbalanced; upper curve corrected and fused at 13 yrs. and 6 mos. old
0	75	75	7	
8	20	12	6	Unbalanced; Milwaukee brace applied at 2 yrs. and 3 mos. old
7	40	33	16	
25	40	15	9	Unbalanced; required correction and fusion at 11 yrs. old
32	53	21	13	
22	45	23	1.5	Balanced
33	52	19	1	
28	29	1	1	Balanced
27	28	1	1	
39	54	15	2	Unbalanced; required correction and fusion at 10 yrs. old
44	61	17	2	
76				Unbalanced; required correction and fusion at 13 yrs. and 11 mos. old
42				
34	34	0	0	High shoulder and upper curve required correction and fusion at 8 yrs. and 1 mo. old
35	35	0	0	
41	45	4	2	
32				
38				Balanced and skeletally mature
32				
17				
16	16	0	0	Balanced
15	16	1	0	
18	18	0	0	

measured 30 to 40 degrees. The largest untreated curves at maturity measured 68 and 88 degrees and were due to bars involving three and five vertebrae, respectively.

Upper thoracic curves, especially those whose apex lay at the second, third, or fourth thoracic vertebra, produced a significant cosmetic deformity due to elevation of the shoulder line on the convex side of the curve. Tilting of the head toward the concavity also occurred in those patients whose curves extended beyond the cervicothoracic junction; the tilting resulted because a satisfactory compensatory curve failed to develop above the congenital curve (Figs. 2-A and 2-B).

In thirteen patients with an upper thoracic curve, with the apex at the fourth, fifth, or sixth thoracic vertebra, an additional long structural curve developed in the lower thoracic or thoracolumbar region (Figs. 3-A and 3-B). Why such a curve should develop in some patients and not in others is unknown, as it was not always associated with the larger upper thoracic curves. This initially compensatory curve, which involved no congenital anomalies, appeared secondarily; while it initially was compensatory and therefore could be corrected, later it became fixed and deteriorated even more rapidly, and it was soon more severe than the primary curve. The major deformity then was caused by the secondary structural curve, which was much more rotated than the primary congenital curve and produced a large rib hump. Five of the patients with this condition were in Group 1A; two were skeletally mature and three required immediate treatment of the secondary structural curve between the ages of fourteen and sixteen years. The remaining eight patients were followed without treatment for a mean of 4.8 years (range, nine months to fourteen years). The median rate of deterioration for the five patients followed before the age of ten years was 6 degrees per year (range, 2 to 8 degrees) for the secondary structural curves and 3 degrees per year (range, 1 to 4 degrees) for the primary congenital curves. Five patients were followed after the age of ten years and then the median rate of deterioration was 7 degrees per year (range, 3 to 10 degrees) for the secondary structural curves and 4 degrees per year (range, 2 to 8 degrees) for the primary congenital curves. The structural compensatory curve usually deteriorated at approximately twice the rate of the primary congenital curve. Six patients were treated between the ages of twelve and sixteen years, when the upper thoracic congenital curves were a mean of 56 degrees (range, 30 to 85 degrees) and the lower secondary structural curves were a mean of 79 degrees (range, 53 to 104 degrees). Three patients reached skeletal maturity without treatment, at which time the upper thoracic congenital curves were 40, 59, and 68 degrees and the lower secondary structural curves were 78, 75, and 82 degrees, respectively.

Lower Thoracic Curves

Of the thirty-eight patients who had a lower thoracic curve, nine were in Group 1A. Six of them were treated immediately, between the ages of eleven and fourteen years, with curves ranging from 41 to 96 degrees (mean, 62 degrees). Three were seen at skeletal maturity, with curves measuring from 66 to 89 degrees.

TABLE

UNTREATED SINGLE CONGENITAL SCOLIOSIS DUE

Site of Curve	No. of Curves	Group 1B (Patients Seen before the Age of Ten Years)												
		No.	Age When First Seen* (Yrs.)	Size of Curve (Degrees)* When First Seen	When Last Seen before 10 Yrs. Old	Rate of Deterioration per Year (Degrees) 1	2	3	4	5	6	7	8	9	10
Upper thoracic	28	15	4 (0-9)	29 (0-60)	35 (13-76)	6	3	4	1	1					
Lower thoracic	29	21	2 (0-9)	40 (12-60)	61 (25-84)	3	3	2	1	2	1	5	4		
Thoracolumbar	12	11	3 (0-9)	35 (15-75)	58 (35-82)		1	1		3	5	1			
Lumbar	3	2	(0-1)	(8-62)	(53-82)					1					1

* Median, with range in parentheses.

† The number of patients who were followed untreated until after the age of ten years but who were first seen before the age of ten years, and who are also included in Group 1B, is in parentheses.

The remaining twenty-nine patients with a lower thoracic curve were in Groups 1B and 1C (Table III) and were followed without treatment for a mean of 3.7 years (range, six months to eleven years and seven months). Before the patients reached the age of ten years, many of the curves deteriorated rapidly (at a median rate of 5 degrees per year) and nine curves deteriorated at a rate of 7 or 8 degrees per year. Ten patients received treatment before the age of ten years. By the age of ten, sixteen of twenty-one curves were more than 40 degrees, and twelve of these were greater than 60 degrees (Figs. 4-A and 4-B). Ten patients were seen, untreated, at the age of ten years, at

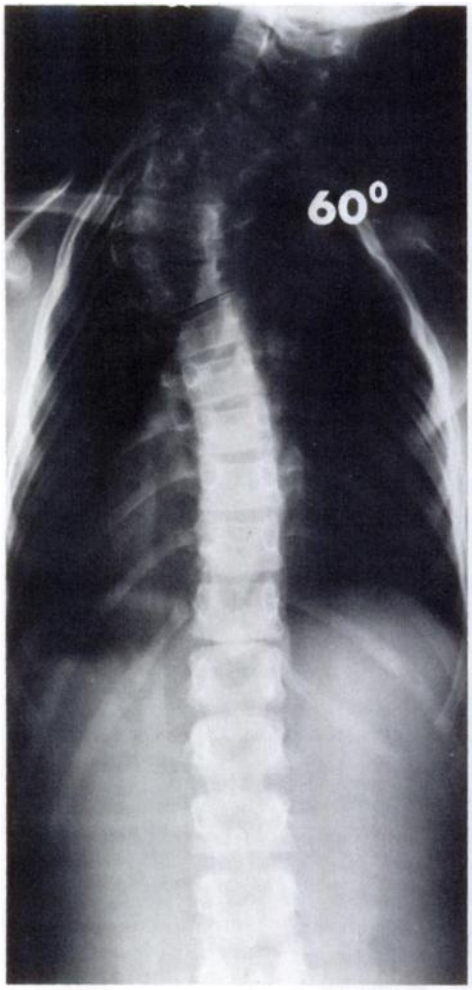

FIG. 2-A

FIG. 2-B

A girl, thirteen years old, who had a unilateral unsegmented bar on the right, extending from the second to the fifth thoracic vertebra and producing a right upper thoracic scoliosis measuring 60 degrees. She had a major cosmetic deformity due to elevation of the left shoulder line and slight tilting of the head to the right.

III

TO A UNILATERAL UNSEGMENTED BAR

Group 1C (Patients Seen at or after the Age of Ten Years)											
	Size of Curve (*Degrees*)*			Rate of Deterioration per Year (*Degrees*)								
No.†	When First Seen at or after 10 Yrs. Old	When Last Seen or at Maturity	Age When Last Seen* (*Yrs.*)	2	3	4	5	6	7	8	9	10
20 (7)	32 (17-81)	47 (20-98)	15 (11-19)	5	2	5	2	2		3		1
14 (6)	43 (25-73)	65 (33-96)	14 (11-18)	1	4	1	1	1				6
6 (5)	58 (35-80)	78 (45-93)	12 (11-17)						1	2		3
1 (0)	20	37	14				1					

which time the mean curve was 48 degrees (range, 25 to 68 degrees). Three of these curves were fused when the patients were ten years old.

After the patients were ten years old, the median rate of deterioration increased to 6.5 degrees per year. Ten patients received treatment at a mean age of thirteen years (range, eleven to sixteen years), at which time the mean curve was 65 degrees (range, 40 to 96 degrees). Three patients reached skeletal maturity without treatment; at that time the curves were 47, 60, and 66 degrees.

The most extensive unsegmented bars in this region occurred in three patients and involved five to six vertebrae. In one of these patients a 45-degree curve was diagnosed at birth which, without treatment, became 77 degrees at the age of four years. The other two patients both were first seen at the age of fourteen years with curves of 57 and 50 degrees, which deteriorated rapidly to 70 and 96 degrees in the patients' fifteenth and sixteenth years, respectively.

Thoracolumbar Curves

Of the fifteen patients who had a thoracolumbar curve, three were in Group 1A. Two of them were treated immediately, at two and thirteen years old, with curves measuring 35 and 85 degrees, respectively. One was seen at skeletal maturity, with a curve of 81 degrees.

FIG. 3-A

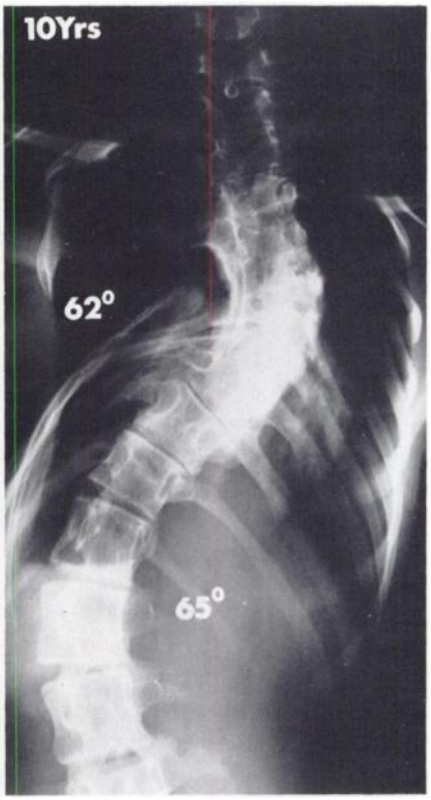

FIG. 3-B

Fig. 3-A: A girl, one month old, who had a radiograph made because of asymmetry of the thoracic rib cage. The radiograph shows absent ribs and a unilateral unsegmented bar extending from the third to the seventh thoracic vertebra on the left.

Fig. 3-B: No treatment was given, and by the age of ten years the unilateral unsegmented bar had produced a 62-degree right upper thoracic scoliosis. The major deformity, however, was due to a very severe secondary left structural thoracolumbar scoliosis that involved no congenital anomalies.

The remaining twelve patients with a thoracolumbar curve were in Groups 1B and 1C (Table III); they were all seen in the first decade of life and were followed without treatment for a mean of five years (range, six months to sixteen years). Before they reached the age of ten years, the median rate of deterioration of the curves was 6 degrees per year. Six patients were treated between the ages of two and seven years, at which time the mean curve was 64 degrees (range, 40 to 82 degrees). The remaining six curves had a mean measurement of 59 degrees (range, 35 to 80 degrees) at the age of ten years.

After the patients were ten years old, the median rate of deterioration increased to 9 degrees per year. Five of six patients received treatment at a mean age of 11.8 years (range, eleven to twelve years), at which time the mean curve was 78 degrees (range, 45 to 90 degrees).

The most extensive bars in this region occurred in three patients and involved six to eight vertebrae. In one patient a 15-degree curve was diagnosed in the first year of life, and at skeletal maturity the untreated curve measured 93 degrees. The second patient was first seen and treated at thirteen years old with an 85-degree curve, and the third was first seen at skeletal maturity with an 80-degree curve.

Pelvic obliquity or listing of the trunk to the side of the unsegmented bar, or both, occurred in twelve of the fifteen patients with a thoracolumbar curve. The degree of pelvic obliquity was severe (more than 25 degrees) in four (range, 26 to 30 degrees), moderate (10 to 25 degrees) in four, mild (less than 10 degrees) in two, and absent in five. The listing was severe in one, moderate in two, mild in five, and absent in seven.

Lumbar Curves

Of the five patients with a lumbar curve, two were treated immediately and were in Group 1A. These two patients had bars involving four and eight vertebrae, producing curves of 64 and 100 degrees at the ages of sixteen and nine years, respectively.

The remaining three patients with a lumbar curve were in Groups 1B and 1C (Table III) and were followed without treatment for a mean of four years (range, one to eight years). One patient had a bar involving three vertebrae which produced a 62-degree curve at the age of one year; the curve had deteriorated to 82 degrees in the second year of life, when it was treated. The second patient also had a bar involving three vertebrae but this produced only an 8-degree curve in the first year of life; the curve deteriorated to 53 degrees at the age of ten years. The third patient had a bar that involved two vertebrae and the curve deteriorated slowly. The curve measured 20 degrees when the patient was eleven years old and then deteriorated more rapidly, to reach 37 degrees when the patient was fourteen years old.

Pelvic obliquity and listing of the trunk occurred in all five of the patients in this group and was directly proportional to the severity of the curve. In the larger curves the pelvic obliquity was the major deforming factor. The severe disability was then due to an apparent shortening of one lower limb.

Unilateral Failure of Segmentation (Unilateral Unsegmented Bar with Contralateral Hemivertebrae) (Group 2)

Twenty-eight patients had a single congenital scoliosis due to a unilateral unsegmented bar on the concavity of the curve with one or more hemivertebrae on the convexity at the same level. Twelve curves were convex to the right and sixteen, to the left. These curves were classified separately from those that were due to a unilateral unsegmented bar alone because they had an even more severe prognosis. By the age of five years, all of these curves became very severe. The rate of deterioration depended mainly on the site of the anomalies and partly on the extent of the unsegmented bar. The bar involved from two to eight vertebrae. The number of hemivertebrae ranged from one to seven. This type of anomaly occurred most frequently in the thoracic region (eighteen patients). The hemivertebrae were most often opposite the bar and not separated by a number of normal vertebrae, as was usually the case when two unilateral hemivertebrae occurred in the absence of a bar. The anomaly was most easily recognized radiographically in the first few years of life (Fig. 5-A), but as the curve rapidly increased the hemivertebrae became obscured (Fig. 5-B) and were difficult to distinguish from a unilateral unsegmented bar alone. The three patients for whom serial observations were not made were in Group 2A and were not included in this part of the analysis. The twenty-five patients who were followed without treatment were in Group 2B (Table IV).

Upper Thoracic Curves

The seven patients with an upper thoracic curve were in Group 2B (Table IV) and were followed without treatment for a mean of 4.6 years (range, six months to twelve years). Before they reached the age of ten years, the median rate of deterioration of the curves was 5 degrees per year (range, 3 to 8 degrees). Four patients were treated between the ages of three and five years. One of these patients, with an unsegmented bar involving eight vertebrae with seven contralateral hemivertebrae, had an 82-degree curve at birth which deteriorated at a rate of 8 degrees per year to become 108 degrees at the age of four years. The other three patients had unsegmented bars whose extent ranged from three to five vertebrae with one or two contralateral hemivertebrae, and the curves measured 35, 52, and 54 degrees prior to treatment. One patient with a bar involving three vertebrae and one contralateral hemivertebra had a 27-degree curve at the age of three months, and was untreated at the age of two years when the curve had increased to 35 degrees.

Two patients reached the age of ten years without treatment. One, with a bar involving six vertebrae with two contralateral hemivertebrae, had a 45-degree curve at one year old, which deteriorated at a rate of 5 degrees per

TABLE IV

UNTREATED SINGLE CONGENITAL SCOLIOSIS DUE TO A UNILATERAL UNSEGMENTED BAR WITH CONTRALATERAL HEMIVERTEBRAE

		Group 2B (Patients Seen before the Age of Ten Years)														
				Size of Curve (*Degrees*)*		Rate of Deterioration per Year (*Degrees*)										
Site of Curve	No. of Curves	No.	Age when First Seen* (*Yrs.*)	When First Seen	When Last Seen before 10 Yrs. Old	1	2	3	4	5	6	7	8	9	10	>10
Upper thoracic	7	7	1 (0-5)	45 (12-82)	54 (35-108)			1	1	3			2			
Lower thoracic	10	10	1 (0-6)	58 (23-76)	84 (27-108)				1	1	5				1	2
Thoracolumbar	8	8	1 (0-2)	58 (41-86)	79 (54-148)						1	1		1		5

* Median, with range in parentheses.

year to become 83 degrees by the age of ten years. The second patient, with a bar involving three vertebrae with one contralateral hemivertebra, had a 40-degree curve at the age of five years old, which deteriorated at a rate of 4 degrees per year to become 59 degrees at the age of ten years. After these patients reached ten years old, the rate of deterioration increased to 5 and 7 degrees per year, and both patients were treated with spine fusion at the ages of eleven and thirteen years old when the curves were 88 and 82 degrees, respectively.

Elevation of the shoulder line or tilting of the head, or both, occurred in all of these patients and, as with other types of upper thoracic congenital scoliosis, caused a significant cosmetic deformity. In addition, in four patients in whom the apex of the curve was at the fourth, fifth, or sixth thoracic vertebra a secondary structural lower thoracic or thoracolumbar scoliosis developed, but did not result from congenital anomalies. These curves deteriorated at a rate of more than 5 degrees per year and measured between 59 and 103 degrees in the three patients who were treated between the ages of three and five years. In one patient, who was treated at the age of thirteen years, the curve measured 72 degrees.

Lower Thoracic Curves

Of the eleven patients who had a lower thoracic curve, one was in Group 2A. This patient, with a 60-degree curve, was treated immediately at the age of ten years.

The remaining ten patients with a lower thoracic curve were in Group 2B (Table IV), and were followed without treatment for a mean of 5.5 years (range, one year to ten years and six months). Because of the severity of the deformity, in eight patients the scoliosis was diagnosed in the first two years of life when the mean curve was 50 degrees (range, 23 to 76 degrees). After the initial diagnosis,

FIG. 4-A

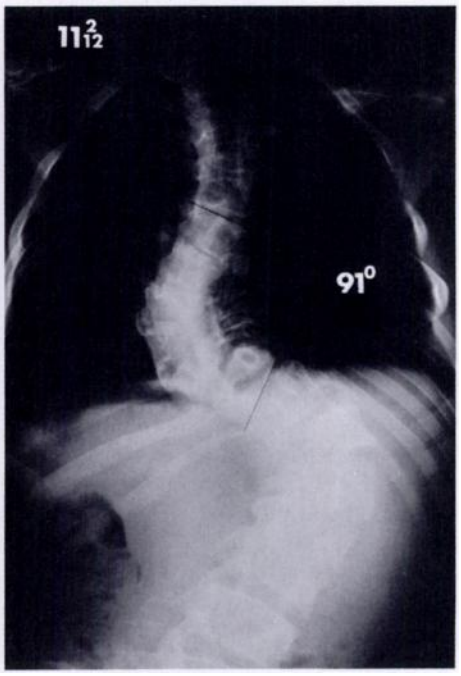

FIG. 4-B

Fig. 4-A: A girl, five years and one month old, with a unilateral unsegmented bar on the right, extending from the eighth to the tenth thoracic vertebra and producing a left lower thoracic scoliosis measuring 61 degrees.

Fig. 4-B: No treatment was given, and by the age of eleven years and two months the lower thoracic curve had deteriorated to 91 degrees.

the median rate of deterioration, without treatment, was 6 degrees per year (range, 4 to 11 degrees) and all but one of the patients received treatment before the age of ten years. Three patients were treated at the age of nine years, with curves of 72, 99, and 106 degrees.

The most extensive vertebral anomalies in this region occurred in four patients who had unsegmented bars involving from seven to nine vertebrae with three to six contralateral hemivertebrae. When the patients were three years old, all of these curves measured more than 70 degrees (range, 76 to 102 degrees). One patient was followed without treatment and at the age of ten years the curve measured 108 degrees. The rate of deterioration then increased, and when the patient was thirteen years old the curve measured 130 degrees.

Thoracolumbar Curves

Of the ten patients who had a thoracolumbar curve, two were in Group 2A. Both of these patients required treatment in the first year of life, with curves of 68 and 75 degrees.

The remaining eight patients with a thoracolumbar curve were in Group 2B (Table IV) and were followed without treatment for a mean of 3.8 years (range, six months to ten years and eight months). These curves were found to have the worst prognosis of any type of congenital scoliosis occurring at any site. All of the scolioses were diagnosed in the first two years of life, when the median curve was 64 degrees (range, 41 to 86 degrees). The curves deteriorated at a median rate of more than 10 degrees per year. All but three of the patients received treatment before the age of five years, when the median curve was 70 degrees (range, 64 to 125 degrees). Two of the three patients who were followed, untreated, for five years received treatment at the age of seven years, with curves of 90 and 148 degrees. The third patient was untreated at ten years old, with a 98-degree curve, which then deteriorated at a rate of 10 degrees per year to become 124 degrees at the age of twelve years and eight months (Figs. 5-A, 5-B, and 5-C).

The most extensive vertebral anomalies in the thoracolumbar region occurred in three patients who had unsegmented bars involving seven to nine vertebrae, with three or four contralateral hemivertebrae. At the age of two years, all of these curves were more than 80 degrees (range, 82 to 103 degrees). One patient was not treated until the age of seven years, when the curve measured 148 degrees.

Pelvic obliquity or listing of the trunk toward the side of the unsegmented bar, or both, occurred in nine of the ten patients with a thoracolumbar curve. The degree of pelvic obliquity was severe (greater than 25 degrees) in two (32 and 50 degrees), moderate (10 to 25 degrees) in four, mild (less than 10 degrees) in two, and absent in two. The listing was severe in four, moderate in one, mild in two, and absent in three.

There were no lumbar curves due to a unilateral unsegmented bar with contralateral hemivertebrae.

Bilateral Failure of Segmentation (Block Vertebrae) (Group 3)

Block vertebrae were present in thirteen patients. Thus, they were not considered to be a common cause of congenital scoliosis. In our series this type of anomaly most frequently occurred in the upper thoracic region (seven patients) and was least common in the lumbar region (one patient). All of these patients had a single congenital scoliosis (eight left and five right) due to a single block of bilaterally unsegmented vertebrae which ranged from two to five segments in length. Four patients had a block of two vertebrae; six, of three vertebrae; one, of four vertebrae; and two had a block of five vertebrae. No patient required treatment and the mean follow-up was 7.8 years (range, two years to sixteen years and six months), with three patients having reached skeletal maturity. There were nine upper thoracic or lower thoracic curves. All of the curves behaved in the same manner and remained 21 degrees or less (range, 10 to 21 degrees). The rate of deterioration was 1 degree or less per year. Of the three curves that were followed to skeletal maturity, none exceeded 21 degrees. There were three thoracolumbar curves that behaved in a slightly different manner. Two of these curves had virtually no deterioration and measured 10 and 14 degrees when the patients were nine and fourteen years old, respectively. The remaining curve, which was first seen when the patient was ten years old, deteriorated a little and reached 35 degrees at skeletal maturity. The one lumbar curve deteriorated very slowly and measured 24 degrees when the patient was ten years old.

Unilateral Complete Failure of Formation (Hemivertebrae) (Group 4)

Hemivertebrae were the cause of a congenital scoliosis in seventy-seven patients. Two of these patients were seen at skeletal maturity and were in Group 4A. The remaining seventy-five patients were followed without treatment and were in Groups 4B and 4C. The hemivertebrae were always on the convexity of the curve. Seventy patients had a single congenital scoliosis (thirty-six left and thirty-four right), of which fifty-six were due to a single hemivertebra (Table V) and fourteen were due to two hemivertebrae on the same side, producing a single congenital scoliosis (Table VI). Seven patients had hemivertebrae on opposite sides but at different levels of the spine, producing two or more opposing congenital curves (Table II, Cases 4 through 10).

Two patients had a hemivertebra that was initially separate but later became synostosed with one of its neighboring vertebrae. Both were in the lumbar region and the resulting curves progressed less rapidly (less than 1 degree per year) than the majority of the curves in this region. Eight patients had an incarcerated hemivertebra; that is, a small, poorly formed segment of bone tucked into a niche between adjacent normal vertebrae. Four of these in-

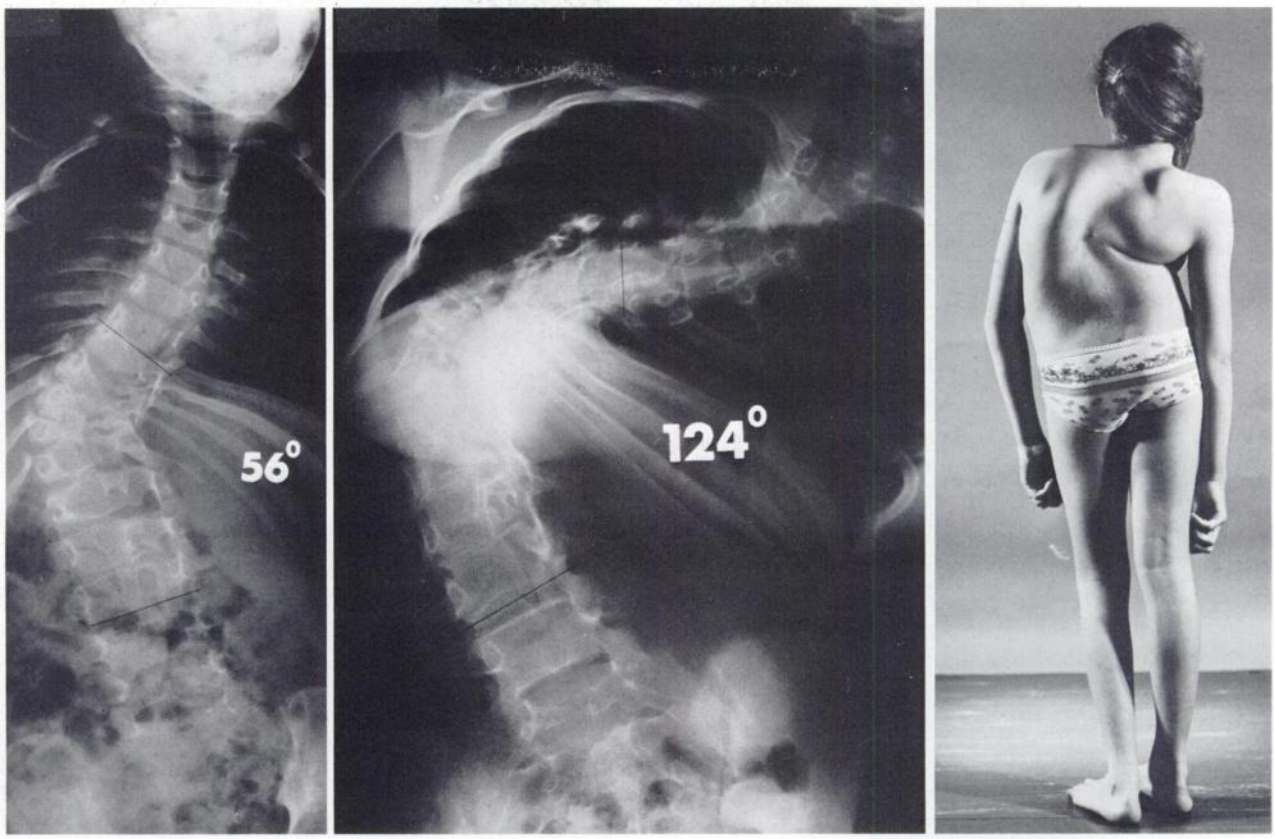

FIG. 5-A FIG. 5-B FIG. 5-C

Fig. 5-A: A girl, two years and ten months old, who had a 56-degree left thoracolumbar scoliosis due to a unilateral unsegmented bar on the right extending from the eighth to the twelfth thoracic vertebra with four adjacent contralateral hemivertebrae at the ninth to the twelfth thoracic levels.

Figs. 5-B and 5-C: No treatment was given, and by the age of twelve years and eight months the scoliosis had increased to 124 degrees and the contralateral hemivertebrae were no longer visible. There was pelvic obliquity, and a severe list of the upper part of the body to the right produced a severe cosmetic deformity.

carcerated hemivertebrae occurred in the upper thoracic region; one, in the lower thoracic region; one, in the thoracolumbar region; and two, in the lumbar region. The resulting curves were all less than 20 degrees, with minimum or no progression.

There was a total of 102 hemivertebrae, which were evenly distributed on either side of the spine (fifty-three on the left and forty-nine on the right) and occurred at any level, although they were seen slightly more frequently in the middle thoracic and lower lumbar regions. Of the patients with two unilateral hemivertebrae producing a single congenital curve, the hemivertebrae were separated by four normal vertebrae in two patients, by three normal vertebrae in three patients, by two normal vertebrae in five patients, and by one normal vertebra in three patients; in only one patient were the hemivertebrae adjacent.

Upper Thoracic Curves

Sixteen patients had a single congenital upper thoracic curve, of which thirteen were due to a single hemivertebra (Table V) and three, to two unilateral hemivertebrae (Table VI). Follow-up without treatment was for a mean of four years (range, one year to nine years and ten months). The median rate of deterioration, without treatment, before the age of ten years for the curves due to a single hemivertebra was 1 degree per year (range, zero to 2 degrees), compared with 2 degrees per year (range, 1 to 2 degrees) in the curves that were due to two unilateral hemivertebrae. By the age of ten years, no curve that was due to a single hemivertebra was greater than 40 degrees, whereas all three patients with two unilateral hemivertebrae had a curve of 40 degrees or more (range, 40 to 47 degrees).

After the patients reached ten years old, the median rate of deterioration doubled (2 degrees per year) in the curves that were due to a single hemivertebra and increased to 2.5 degrees per year (range, 2 to 3 degrees) in those due to two unilateral hemivertebrae. Of six patients with a single hemivertebra, three were followed without treatment to skeletal maturity, at which time the curves measured 25, 39, and 43 degrees, and the remaining three patients had a spine fusion when the curves measured 28, 31, and 44 degrees. Two patients with two unilateral hemivertebrae were followed after the age of ten years and both required a spine fusion during the adolescent growth spurt, when the curves were 43 and 47 degrees.

In general, upper thoracic curves due to one or two unilateral hemivertebrae progressed relatively slowly, and only a few became moderately severe. Although none became very severe, they did, like other congenital upper

TABLE

UNTREATED SINGLE CONGENITAL SCOLIOSIS

Site of Curve	No. of Curves	Group 4B (Patients Seen before the Age of Ten Years)												
		No.	Age When First Seen* (Yrs.)	Size of Curve (Degrees)* When First Seen	Size of Curve (Degrees)* When Last Seen before 10 Yrs. Old	Rate of Deterioration per Year (Degrees) <1	1	2	3	4	5	6	7	8	9	10
Upper thoracic	13	10	2 (0-7)	16 (11-28)	18 (11-36)	5	3	2								
Lower thoracic	9	6	4 (0-9)	34 (17-48)	44 (17-50)	2		4								
Thoracolumbar	5	3	1 (0-3)	26 (20-35)	40 (21-45)		1	2								
Lumbar	15	12	2 (0-9)	24 (0-37)	30 (13-41)	7	3	2								
Lumbosacral	12	8	4 (2-9)	18 (10-32)	19 (10-53)	6	1		1							

* Median, with range in parentheses.

† The number of patients who were followed untreated until after the age of ten years but who were first seen before the age of ten years, and who are also included in Group 4B, is in parentheses.

thoracic curves, produce a significant cosmetic deformity due to elevation of the shoulder line on the convex side of the curve, and occasionally they produced tilting of the head. In only one patient, with a single hemivertebra at the fifth thoracic level, did a secondary structural lower thoracolumbar scoliosis develop. At skeletal maturity, this curve measured 52 degrees and the upper thoracic congenital curve measured 43 degrees.

Lower Thoracic Curves

Seventeen patients had a single congenital lower thoracic curve, nine of which were due to a single hemivertebra (Table V) and eight, to two unilateral hemivertebrae (Table VI). These patients were followed without treatment for a mean of 3.6 years (range, one year to nine years and four months), and four reached skeletal maturity. The median rate of deterioration before the patients reached the age of ten years was 2 degrees per year (range, zero to 2 degrees) for the curves that were due to a single hemivertebra and 2 degrees per year (range, 2 to 6 degrees) for the curves that were due to two unilateral hemivertebrae. By the age of ten years, the majority of the patients (four of six) with a single hemivertebra had a curve of between 40 and 50 degrees, whereas all of the six patients with two unilateral hemivertebrae had a curve of 50 degrees or more.

After the patients reached the age of ten years, the median rate of deterioration for the lower thoracic curves due to a single hemivertebra increased to 2.5 degrees per year (range, less than 1 degree to 5 degrees), and in the curves due to two unilateral hemivertebrae the median rate was 3 degrees per year (range, 2 to 4 degrees). Of the six patients with a single hemivertebra who were followed after the age of ten years, three reached skeletal maturity without treatment, at which time the curves measured 37, 38, and 43 degrees; two other curves progressed more rapidly and required spine fusion, measuring 52 degrees when one patient was twelve years old and 60 degrees when the other was fourteen years old; and one patient had a 47-degree untreated curve at the age of eleven years. Of the six patients with two unilateral hemivertebrae who were followed without treatment (Figs. 6-A and 6-B), five required treatment between the ages of twelve and fifteen years, at which time the median curve was 64 degrees (range, 30 to 70 degrees), and one reached skeletal maturity with a 50-degree curve.

Thoracolumbar Curves

Of the ten patients with a thoracolumbar curve, two were in Group 4A. Both of these patients had a single hemivertebra and were seen at skeletal maturity with curves of 52 and 55 degrees. The remaining eight patients with a thoracolumbar curve were followed without treatment for a mean of five years (range, one year and seven months to seven years and seven months).

Five patients had a curve that was due to a single hemivertebra (Table V), and the mean rate of deterioration before the age of ten years was 2 degrees per year (range, 1 to 2 degrees) and 3.5 degrees thereafter (range, 3 to 4 degrees). Before the patients were ten years old, one of three curves was greater than 40 degrees and the two curves that were untreated at skeletal maturity measured 33 and 43 degrees.

Three patients had a single congenital thoracolumbar curve due to two unilateral hemivertebrae (Table VI). These curves were all diagnosed in the first two years of life, when they exceeded 50 degrees. The median rate of deterioration before the age of ten years was 5 degrees per year (range, 4 to 6 degrees) and all three patients required treatment in the third or fourth year of life, when the median curve was 63 degrees.

Lumbar Curves

Fifteen patients had a single congenital lumbar curve, all of which were due to a single hemivertebra at the third or fourth lumbar level (Table V). None of these patients required treatment, and they were followed for a mean of 6.6 years (range, one year to fourteen years and four months). The median rate of deterioration was less than 1 degree per year (range, zero to 2 degrees) before the age of ten years and 1 degree per year thereafter (range, zero to 3 degrees). Before the age of ten years, only one curve was

V

DUE TO A SINGLE HEMIVERTEBRA

Group 4C (Patients Seen at or after the Age of Ten Years)												
	Size of Curve *(Degrees)**			Rate of Deterioration per Year *(Degrees)*										
No.†	When First Seen at or after 10 Yrs. Old	When Last Seen or at Maturity	Age When Last Seen* *(Yrs.)*	<1	1	2	3	4	5	6	7	8	9	10
6 (3)	30 (15-40)	35 (25-44)	16 (11-17)			4	2							
6 (3)	44 (25-50)	45 (37-60)	16 (11-19)	1	1	1	2		1					
2 (0)	30 (21-40)	41 (33-43)	Both mature				1	1						
8 (5)	32 (14-52)	36 (16-55)	17 (13-18)	2	3	2	1							
12 (8)	20 (15-53)	25 (15-83)	15 (11-18)	5	1	2	3			1				

greater than 40 degrees and six patients had curves that had remained virtually unchanged, at less than 30 degrees, from an early age. Five patients were followed, without treatment, to skeletal maturity, when the median curve was 42 degrees (range, 22 to 55 degrees).

In all of the patients with a lumbar curve the trunk remained balanced, but in two there was mild pelvic obliquity.

Lumbosacral Curves

This type of congenital scoliosis occurred only in association with a single hemivertebra at the lumbosacral junction (Table V). There were twelve such patients, who were followed without treatment for a mean of 6.8 years (range, one year to ten years and seven months). In nine patients the hemivertebra was at the fifth lumbar level and in three patients the hemivertebra lay between the fifth lumbar level and the sacrum; in one of these patients there was a failure of segmentation between the hemivertebra and the sacrum. The congenital lumbosacral curves were all very short and extended from the fourth or fifth lumbar level to the sacrum. In all of the patients the pelvis remained level, and as a result the hemivertebra caused the lumbar spine to take off obliquely from the sacrum. To overcome this imbalance, in all of the patients a long secondary thoracolumbar curve developed, extending from the fifth lumbar vertebra to the ninth, tenth, or eleventh thoracic vertebra and soon becoming fixed. In eleven pa-

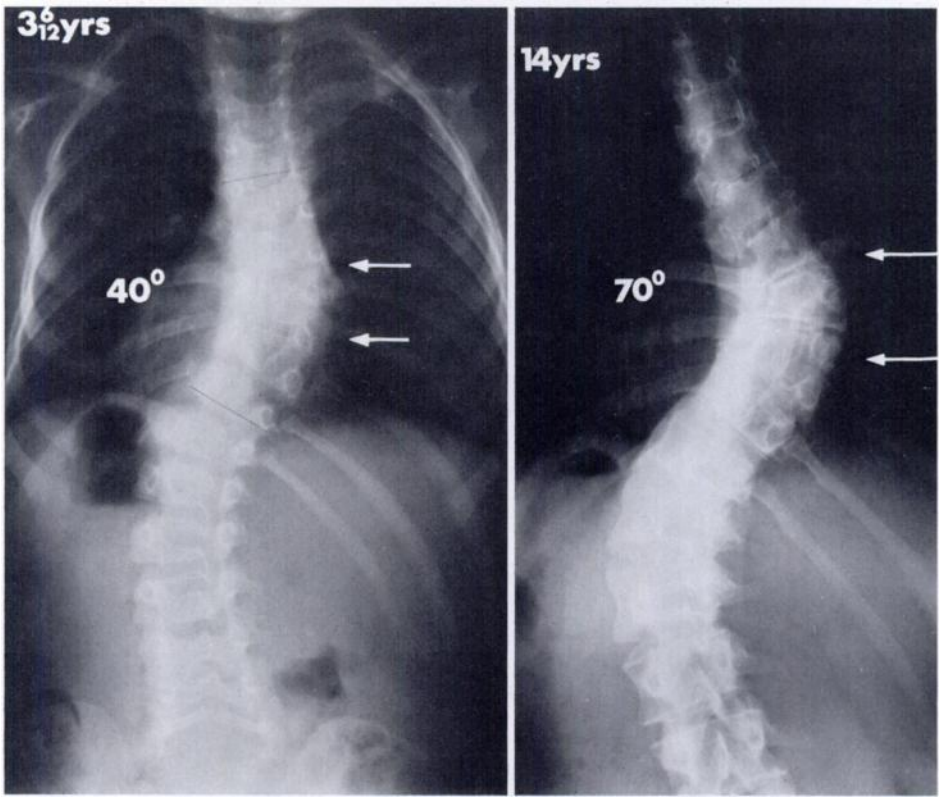

FIG. 6-A FIG. 6-B

Fig. 6-A: A boy, three years and six months old, with a 40-degree right lower thoracic scoliosis due to two unilateral hemivertebrae at the seventh and tenth thoracic levels.

Fig. 6-B: No treatment was given and the curve deteriorated to 70 degrees at the age of fourteen years, just prior to spine fusion.

TABLE

UNTREATED SINGLE CONGENITAL SCOLIOSIS

Site of Curve	No. of Curves	Group 4B (Patients Seen before the Age of Ten Years)												
		No.	Age When First Seen* (Yrs.)	Size of Curve (Degrees)* When First Seen	When Last Seen before 10 Yrs. Old	Rate of Deterioration per Year (Degrees) 1	2	3	4	5	6	7	8	9	10
Upper thoracic	3	3	6 (3-9)	42 (32-44)	44 (40-47)	1	2								
Lower thoracic	8	4	2 (0-4)	40 (30-48)	53 (50-54)		3				1				
Thoracolumbar	3	3	1 (0-2)	55 (50-60)	63 (55-72)				1	1	1				

* Median, with range in parentheses.

† The number of patients who were followed untreated until after the age of ten years but who were first seen before the age of ten years, and who are also included in Group 4B, is in parentheses.

tients this curve did not compensate sufficiently and as a result the upper part of the body listed to a varying degree to the side opposite that of the hemivertebra. In the one patient in whom the body remained balanced, it did so because of a limb-length discrepancy that resulted in a compensatory pelvic obliquity. With time, all of these secondary curves became structural, and three deteriorated to become the major deformity.

Not all of the secondary curves deteriorated at the same rate, and this appeared to depend on the extent of the hemivertebra. Nine patients had a hemivertebra that extended across the midline, and in these patients both the congenital lumbosacral curve and its secondary thoracolumbar curve remained virtually static until the patients reached the age of ten years, at which time neither curve exceeded 25 degrees. After the patients were more than ten years old, the rates of deterioration of both curves increased slightly, as did the tendency of the upper part of the body to list to one side. Five of these patients were followed to skeletal maturity, without treatment, at which time neither the congenital lumbosacral curve nor the secondary thoracolumbar curve exceeded 36 degrees and the degree of listing was not sufficient to require treatment.

Three patients had a hemivertebra that did not extend across the midline, and this produced a greater list and a much larger secondary structural thoracolumbar curve, which was severely rotated and produced a large rib hump. Before the patients were ten years old, both the lumbosacral curve and the secondary thoracolumbar curve deteriorated relatively slowly (at a rate of 1 to 3 degrees per

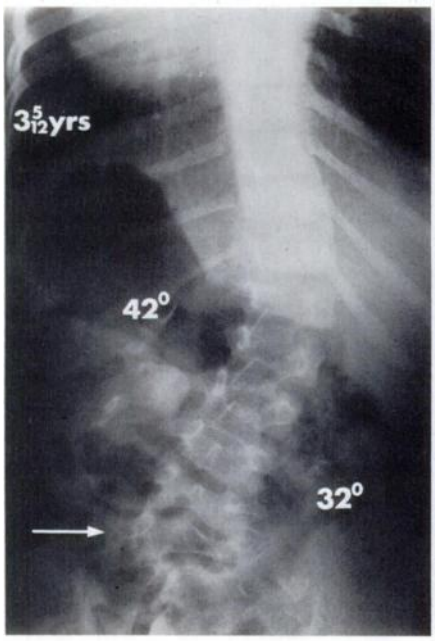

FIG. 7-A

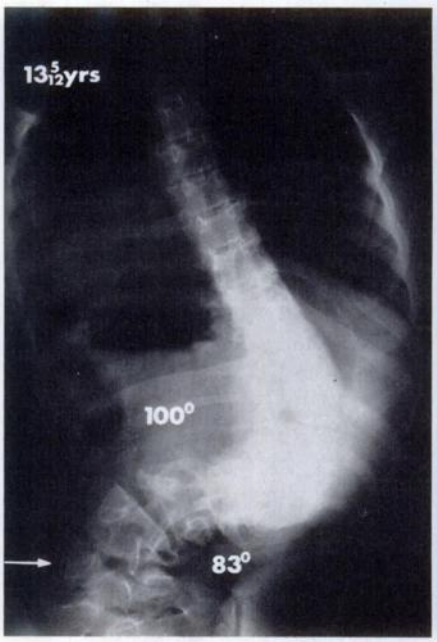

FIG. 7-B

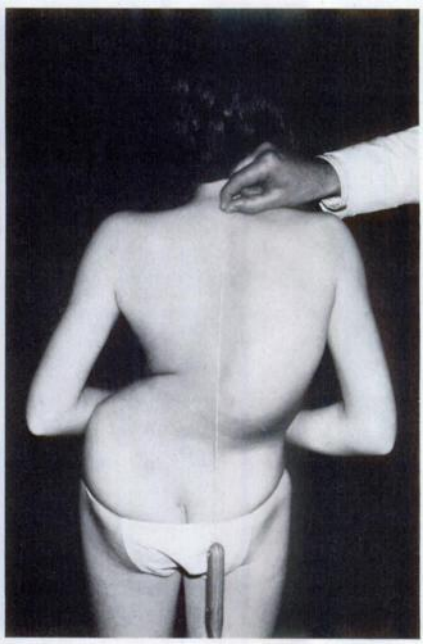

FIG. 7-C

Fig. 7-A: A girl, three years and five months old, with a 32-degree left lumbosacral scoliosis due to a single hemivertebra at the lumbosacral junction. There was also a 42-degree right thoracolumbar scoliosis in which no congenital anomalies were involved. The pelvis was level but the upper part of the body listed to the right.

Figs. 7-B and 7-C: No treatment was given, and by the age of thirteen years and five months the congenital lumbosacral curve had deteriorated to 83 degrees. The thoracolumbar curve measured 100 degrees and had become structural and severely rotated. The list had increased but the pelvis remained level. The major deformity was due to the severe thoracolumbar scoliosis which, despite its size, failed to compensate for the list to the right.

VI

DUE TO TWO UNILATERAL HEMIVERTEBRAE

Group 4C (Patients Seen at or after the Age of Ten Years)										
	Size of Curve *(Degrees)**			Rate of Deterioration per Year *(Degrees)*						
No.†	When First Seen at or after 10 Yrs. Old	When Last Seen or at Maturity	Age When Last Seen* *(Yrs.)*	2	3	4	5	6	7	8	9	10
2 (2)	42 (40-44)	45 (43-47)	12 (11-16)	1	1							
6 (2)	51 (28-54)	62 (30-70)	13 (12-17)	2	2	2						
0 (0)	—	—	—									

year), but after the age of ten years a significant cosmetic deformity developed due to the rib hump and an increasing tendency of the upper part of the body to list to one side. The congenital lumbosacral curves in two of these patients measured 25 and 42 degrees at the age of fourteen years, and the secondary structural thoracolumbar curves measured 40 and 48 degrees, respectively. The most severe deformity occurred in the third patient who, at the age of thirteen years, had an 83-degree lumbosacral curve with a 100-degree secondary structural thoracolumbar curve and a severe list (Figs. 7-A, 7-B, and 7-C).

Unilateral Partial Failure of Formation (Wedge Vertebrae) (Group 5)

Wedge vertebrae occurred in only nine patients. They all had a single congenital scoliosis (five left and four right). The most commonly affected vertebrae were in the thoracic region (six patients). Seven patients had a single wedge vertebra and two patients had a pair of adjacent thoracic vertebrae that were affected on the same side.

Upper Thoracic Curves

There was only one upper thoracic congenital scoliosis that was due to wedging, and it involved the fourth thoracic vertebra. This curve deteriorated at a rate of slightly less than 2 degrees per year, from 23 degrees when the patient was twelve years old to 31 degrees at skeletal maturity.

Lower Thoracic Curves

There were four lower thoracic congenital curves, of which two were due to a single wedge vertebra and two, to a pair of adjacent wedge vertebrae. Three patients were followed, without treatment, for a mean of 6.4 years (range, one year and eight months to nine years and nine months) and required treatment at nine, eleven, and twelve years old, when the curves measured 33, 39, and 41 degrees, respectively. One patient with two wedge vertebrae was first seen at skeletal maturity, when the curve measured 49 degrees.

Thoracolumbar Curves

There were three thoracolumbar congenital curves, which were followed for a mean of 3.4 years (range, one year to seven years and four months). One patient required treatment at the age of five years, when the curve measured 32 degrees. The remaining two patients were untreated, and the curves deteriorated at a rate of 2 degrees per year until they measured 17 degrees, at which time one child was thirteen years old. The other child had a 40-degree curve at skeletal maturity.

Lumbar Curves

Only one lumbar curve was due to wedging, and it involved the fifth lumbar vertebra. This curve deteriorated slowly, at a rate of less than 1 degree per year over three years, and measured 12 degrees when the patient was seven years old.

Complex (Unclassifiable) Anomalies (Group 6)

There were twenty-five patients with complex (unclassifiable) congenital vertebral anomalies causing a congenital scoliosis. Twenty-two of these patients had a single congenital curve (twelve right and ten left) and three had multiple congenital curves.

Nineteen patients had a single congenital scoliosis due to a jumble of vertebral anomalies, and they were followed without treatment for a mean of seven years (range, one to seventeen years). Two patients had an upper thoracic curve; eight patients, a lower thoracic curve; seven patients, a thoracolumbar curve; and two patients, a lumbar curve. All of these curves were much more unpredictable in their behavior than the congenital curves that were due to simple anomalies in the same regions, but in general they tended to progress relatively slowly regardless of their location. Fifteen patients were untreated at the age of ten years, at which time eight curves were less than 20 degrees; three curves, between 20 and 30 degrees; five curves, between 31 and 40 degrees; and only two curves measured more than 40 degrees (51 and 73 degrees). After the age of ten years, the rate of deterioration usually increased. Seven patients reached skeletal maturity without treatment, at which time the curves ranged from 8 to 56 degrees (mean, 31 degrees). The largest untreated curve in the remaining five skeletally immature patients was in the thoracolumbar region and measured 82 degrees at the age of twelve years.

Three patients who were seen at skeletal maturity had a single congenital scoliosis, but the congenital anomaly was partially obscured and could not be accurately

Site of Curvature	Type of Congenital Anomaly					
	Block Vertebra	Wedged Vertebra	Hemivertebra: Single	Hemivertebra: Double	Unilateral Unsegmented Bar	Unilateral Unsegmented Bar and Contralateral Hemivertebrae
Upper thoracic	<1° – 1°	★ – 2°	1° – 2°	2° – 2.5°	2° – 4°	5° – 6°
Lower thoracic	<1° – 1°	2° – 2°	2° – 2.5°	2° – 3°	5° – 6.5°	6° – 7°
Thoraco lumbar	<1° – 1°	1.5° – 2°	2° – 3.5°	5° – ★	6° – 9°	> 10° – ★
Lumbar	<1° – ★	<1° – ★	<1° – 1°	★	> 5° – ★	★
Lumbo sacral	★	★	<1° – 1.5°	★	★	★

No treatment required. May require spinal fusion. Require spinal fusion.

★ Too few or no curves

FIG. 8

Median yearly rate of deterioration (in degrees) without treatment for each type of single congenital scoliosis in each region of the spine (187 patients). The numbers on the left in each column refer to patients who were seen before the age of ten years; the numbers on the right refer to patients who were seen at or after the age of ten years.

classified. All three patients had a thoracic or thoracolumbar curve of more than 100 degrees.

Discussion

The prognosis for a patient with congenital scoliosis can vary considerably. Some patients are first seen with small curves, many of which progress minimally, whereas others are first seen with larger curves that deteriorate rapidly and cause extreme deformity. Of the 251 patients in this study, 143 were last seen, untreated, after the age of ten years, at which time fifty-one (36 per cent) had a curve of 40 to 60 degrees and forty (28 per cent) had a curve of more than 60 degrees. We concluded that the prognosis for untreated curves after the age of ten years generally becomes more unfavorable because in our series most of the other 108 patients (that is, an additional sixty-seven patients) required treatment at or before the age of ten years because of the severity of the curve. Twenty of these curves measured 40 to 60 degrees and thirty-nine, more than 60 degrees. These findings are very different from those of Kuhns and Hormell, who found that only 38 per cent of eighty-five children who were followed to skeletal maturity without treatment had curves of more than 30 degrees. Our findings are more in agreement with those of Winter et al., who found that 84 per cent of thirty-eight children who were followed without treatment beyond the age of ten years had curves of more than 40 degrees.

To understand the variable prognosis for congenital scoliosis, it is necessary to correlate the principles of normal growth of the spine with the pathological anatomy of the various types of congenital vertebral anomalies. Normally, longitudinal growth of the spine is the sum total of the growth occurring at the end-plates on the upper and lower surfaces of the vertebral bodies, which occurs equally on either side of the spine so that the spine remains straight and without a scoliosis[2]. A congenital vertebral anomaly can, however, cause a growth imbalance due to a deficiency in either the number of end-plates or their rate of growth on one side of the spine. The lateral curve that results is of a severity proportional to the degree of the growth imbalance.

In classifying a congenital scoliosis with regard to its prognosis, the most important feature is, therefore, the growth imbalance caused by the vertebral anomalies that predominate on one side of the spine. The radiographic classification that was used in this study was found to be generally satisfactory in this respect, although not all of the curves progressed as expected and the growth potential could not be exactly predicted radiographically. The vertebral anomalies in 90 per cent of the patients could be classified into one of five specific groups, but some curves could not be classified (Group 6) because they either involved a complex jumble of anomalies or, on occasion, because the severity was so great as to obscure the radiographic characteristics of the anomaly.

Congenital scoliosis occurred significantly more often in girls than in boys in our series. The congenital curves often were present at birth and were most frequently diag-

nosed in either the first few years of life or between the ages of nine and fourteen years, probably because the periods of most rapid growth of the spine are *in utero*, from birth to the age of three years, and at puberty[7]. The scolioses that presented as a clinical deformity in the first year of life had the worst prognosis, as this indicated a marked growth imbalance that would continue until skeletal maturity, resulting in severe deformity.

A single congenital scoliotic curve occurred in 95 per cent of the 251 patients and was much more common than the occurrence of two or more congenital curves (5 per cent of the patients). The commonest type of vertebral anomaly causing the scoliosis was a unilateral unsegmented bar (38 per cent of the 269 congenital curves; Table I), followed by hemivertebrae (33 per cent), complex anomalies (11 per cent), a unilateral unsegmented bar with contralateral hemivertebrae (10 per cent), and a block vertebra (5 per cent); the least common was a wedge vertebra (3 per cent). The commonest site for the congenital scoliosis was the lower thoracic region (33 per cent of the 269 congenital curves; Table I), followed by the upper thoracic (31 per cent), thoracolumbar (20 per cent), and lumbar regions (11 per cent); the least common was the lumbosacral region (5 per cent).

Like Winter et al., we found that the rate of deterioration and the ultimate severity of the congenital scoliosis depended not only on the type of anomaly but also on the site at which it occurred. The site of curvature that had the worst prognosis, for each type of vertebral anomaly, was usually the thoracolumbar region, and the prognosis was only slightly less severe in the lower thoracic region. These findings differ from those of Winter et al., who found that lower thoracic curves had a worse prognosis than thoracolumbar curves. We agree with Winter et al. that lower thoracic and thoracolumbar curves usually have a worse prognosis than do lumbar curves, and that the most benign curves occur in the upper thoracic region.

The type of anomaly causing the most severe scoliosis in each region of the spine was a unilateral unsegmented bar with contralateral hemivertebrae at the same level. This was followed in severity by scoliosis caused by a unilateral unsegmented bar alone, two unilateral hemivertebrae, a single hemivertebra, and a wedge vertebra; the least severe was scoliosis caused by a block vertebra (Tables II through VI).

In addition, we found that the rate of deterioration of the curves was not constant, but if the curve was present before the patient was ten years old it usually increased, in particular during the adolescent growth spurt. Although there was often a relatively wide range in the rate of deterioration, the majority of the curves that were due to radiographically similar anomalies and that occurred in the same region tended to deteriorate at approximately the same rate (Fig. 8).

A unilateral unsegmented bar does not contain growth plates and therefore cannot grow longitudinally, whereas normal or nearly normal growth may occur on the opposite side of the spine. In our series, the longer unsegmented bars tended to produce the larger curves in a specific region, but occasionally a shorter bar produced an equally large curve in the same region. In three children the unsegmented bar was not recognized radiographically until they were three and four years old, when it became more ossified. As a result, these patients were initially misdiagnosed as having infantile idiopathic scoliosis. Because of the severe growth imbalance, all of the curves that were associated with an unsegmented bar and that were present during infancy deteriorated very rapidly, and all became very severe. The mean rate of deterioration in patients who were younger than ten years old ranged from 5 degrees per year for lower thoracic curves to 6 degrees per year for thoracolumbar curves (Table III). Of the thirty-six patients with a lower thoracic, thoracolumbar, or lumbar curve who were last seen without treatment at or before the age of ten years, ten had a curve that measured between 40 and 60 degrees and nineteen had a curve of more than 60 degrees. This type of congenital scoliosis requires treatment as soon as the anomaly is diagnosed.

It is important to recognize the anomaly of a unilateral unsegmented bar with contralateral hemivertebrae, as described by Nasca et al., because it has the worst prognosis of any type of congenital vertebral anomaly. All of these patients were seen before the age of two years in our series. At that age the hemivertebrae could be seen radiographically, but as the deformity progressed the hemivertebrae tended to be obscured because of the severity of the curve, and the anomaly then became indistinguishable from an unsegmented bar alone. The median rate of deterioration before the age of ten years ranged from 5 degrees per year for upper thoracic curves to more than 10 degrees per year for thoracolumbar curves. Sixteen of eighteen lower thoracic and thoracolumbar curves exceeded 50 degrees once the patient was three years old (Table IV). This type of congenital scoliosis requires treatment as soon as the anomaly is diagnosed.

A hemivertebra produces a scoliosis by acting as an enlarging wedge on the affected side of the spine, whereas in patients with a unilateral unsegmented bar there is retarded growth on the affected side. The growth imbalance in patients with hemivertebrae is never as severe as in those with a unilateral unsegmented bar. Not all hemivertebrae, however, produce the same degree of growth imbalance at the same site. Eight patients in our series had an incarcerated hemivertebra[5], which we defined as a small, poorly formed, extra segment of bone tucked into the spine between adjacent normal vertebrae. This type of hemivertebra has no growth potential and the resulting curves were all less than 20 degrees, with minimum or no progression. Two patients had a hemivertebra that became synostosed with a neighboring vertebra (semisegmented), and this also produced a curve that progressed less rapidly than the others in the same region. The majority of single hemivertebrae, however, caused slowly progressive curves. Before the patients were ten years old the median

rate of deterioration for both upper thoracic and lumbar curves due to a single hemivertebra was 1 degree per year and no curve exceeded 40 degrees (Table V). These curves usually do not require treatment. Single hemivertebrae in the lower thoracic and thoracolumbar regions caused more severe deformity. Before the patients were ten years old, the median rate of deterioration was 2 degrees per year, and six of nine curves were between 40 and 50 degrees. These patients often require treatment during the adolescent growth spurt. Two unilateral hemivertebrae produced a greater growth imbalance and caused a much more severe deformity. The median rate of deterioration before the age of ten years ranged from 2 degrees per year for upper thoracic curves to 5 degrees per year for thoracolumbar curves. All seven lower thoracic and thoracolumbar curves were greater than 50 degrees by the time the patient was ten years old. These curves usually require treatment before the patient is five years old.

A wedge-shaped vertebra is due to a unilateral partial failure of formation of a vertebra, with retarded longitudinal growth on the hypoplastic side. In two patients, two adjacent vertebrae were affected, but this did not unduly increase the severity of the scoliosis. The one upper thoracic curve and one lumbar curve in our series deteriorated slowly (less than 2 degrees per year) and did not require treatment, whereas the seven lower thoracic and thoracolumbar curves deteriorated somewhat more rapidly (median rate of increase, 2 degrees per year) and three required treatment during the adolescent growth spurt.

A block vertebra is due to bilateral failure of segmentation, and longitudinal growth is impaired on both sides of the spine but not always symmetrically. The result was a mild degree of curvature that rarely exceeded 20 degrees, and the thirteen patients in our series who were so affected did not require treatment.

The thirteen patients who had two or more congenital curves had a variable prognosis, depending on the site of the opposing anomalies. If the anomalies, either similar or dissimilar, occurred within a few segments of each other and were in the same region, they tended to balance each other and produced little deformity other than a kink in the spine. If, however, they were widely separated and in different regions, the resulting curves tended to be unbalanced, producing decompensation or listing of the trunk that required treatment.

Apart from the rate of deterioration and ultimate severity of a congenital scoliosis, there were also a number of important secondary features that should be emphasized because they contributed significantly to the over-all disability and deformity of the patient.

Upper thoracic curves, especially those that extended cranially beyond the cervicothoracic junction, commonly produced a cosmetic deformity due to elevation of the shoulder or, less frequently, tilting of the head (Figs. 2-A and 2-B). Because congenital scoliosis occurs so frequently in the upper thoracic region (31 per cent of our patients) the deformity is a common one, and the higher the apex of the curve the more severe was the deformity. An elevated shoulder line was most distressing to girls, and a 30-degree curve seemed to be the upper limit that the patients would tolerate.

An additional problem associated with thoracic curves, especially those with the apex at the fifth, sixth, or seventh thoracic vertebra, was the development of a long secondary structural curve in the lower thoracic or thoracolumbar region. This curve, which did not involve any congenital anomalies, initially was compensatory and was correctable, but later it tended to become fixed and to deteriorate even more rapidly than the primary (congenital) curve (Figs. 3-A and 3-B). Moreover, the congenital thoracic curve usually had only a mild degree of rotation, whereas the lower secondary curve often was severely rotated. The large rib hump so produced usually constituted a major deformity. In nineteen of forty-five patients who had a congenital curve due to a unilateral unsegmented bar, with or without contralateral hemivertebrae in the upper thoracic segments, a secondary structural lower thoracic or thoracolumbar curve developed.

In patients with a lower thoracic, thoracolumbar, or lumbar curve, especially those due to a unilateral unsegmented bar with or without contralateral hemivertebrae, a compensatory curve that was adequate to balance the congenital curve often failed to develop because there were too few normal mobile vertebrae between the anomaly and either the upper end of the spine or the sacrum. As a result, in 80 per cent of the patients with a congenital thoracolumbar curve and in all with a lumbar curve in our series some degree of pelvic obliquity and an apparent shortening of one lower limb developed (Figs. 5-B and 5-C). Decompensation or listing of the upper part of the body to one side was also a common finding associated with the more severe lower thoracic and thoracolumbar curves and could result in a very severe deformity.

The twelve lumbosacral curves in our series were all due to a single hemivertebra at the lumbosacral junction but, unlike the situation with congenital lumbar or thoracolumbar curves, the pelvis remained level. The hemivertebra, therefore, caused the lumbar spine to take off obliquely from the sacrum. In an attempt to keep the body balanced, a secondary lumbar or thoracolumbar curve developed, but unfortunately it usually was insufficient to prevent the trunk from listing to the side opposite that of the hemivertebra. In nine patients, both the list and the secondary curve remained small (less than 36 degrees) and produced only a mild to moderate deformity. In three patients, however, the secondary curve became large, fixed, and rotated and caused a major cosmetic deformity due to a large rib hump and decompensation of the trunk (Figs. 7-B and 7-C).

In conclusion, we can state that congenital scoliosis is a potentially serious condition, which can and often does result in severe curvature of the spine and malalignment of the body. We have shown that it is possible to anticipate the course of a congenital scoliosis if the type of vertebral

anomaly and its site are known. Ideally a congenital scoliosis should be diagnosed while the patient is young and the curve is small. At that stage, a curve that is at risk for progression can be recognized and an appropriate prophylactic course of treatment can be planned so as to prevent severe deformity.

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