

## Types and Associated Anomalies of Congenital Scoliosis

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### – Abstract –

**Study design :** Analysis was based on radiographic appearance of 57 cases of congenital scoliosis and associated anomaly

**Purpose :** The aim of the present study was to assess the incidence, morphology and the associated anomalies of the congenital spinal scoliosis.

**Summary of Literature Review :** Hemivertebra is the most common type of congenital scoliosis and urogenital, musculoskeletal and cardiac anomalies are strongly associated.

**Materials and Methods :** The authors analysed the morphology and the associated anomalies of 57 cases of congenital scoliosis from 1994 to 2000.

**Results :** It was more common in male(32 males and 25 females). The bony anomalies were classified as failure of formation(40cases, 70.2%), failure of segmentation(11cases, 19.3%) and mixed type(6cases, 10.5%). Of the failure of formation, there were 36 cases(63.2%) of hemivertebra, 2 cases of posterior quadrant vertebra and 2 cases of wedge vertebra. We found associated anomalies in 26 patients(45.6%). Associated cardiac anomalies were 2 dextrocardia, ventricular septal defect, atrial septal defect and patent ductus arteriosus. Associated musculoskeletal anomalies were 5 rib fusion, 2 developmental dysplastic hip, 3 Klippel- Feil syndrome, Achondroplasia, Arnold- Chiari malformation, spinal dysraphism with sacral hair patch, cleft palate with congenital anklyloglossia. Associated neurogenic anomalies were 2 cases of syringomyelia and 3 mental retardation. There were unilateral renal agenesis and undescended testicle in urogenital anomalies.

**Conclusion :** Common type of the congenital scoliosis was hemivertebra(63.2%) caused by the failure of formation(70.2%). Associated anomalies were found in 26 patients(45.6%).

**Key Words :** Congenital scoliosis, Morphology, Associated anomalies

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\*

1998

16

21

, 12

1,6)

가 1.

57 23

84 percentile(73~90 percentile)

14 ( ~48 )

가 6 , 가 3

가 32 (56.1%) 25 (43.9%)

1,4,6), 11), 1,5), 10)

57

2.

57 26 (45.6%)

5 2 , 1

1 , 1 , 1

14 5 ,

(arthrogryposis)

2 , Klippel-Feil

3 (Fig. 4B), 1 , Arnold-Chiari

1 ,

가 1 , 1

30 가 11 3 2 ,

3 ,

Cobb 1 (Table 1).

4 가

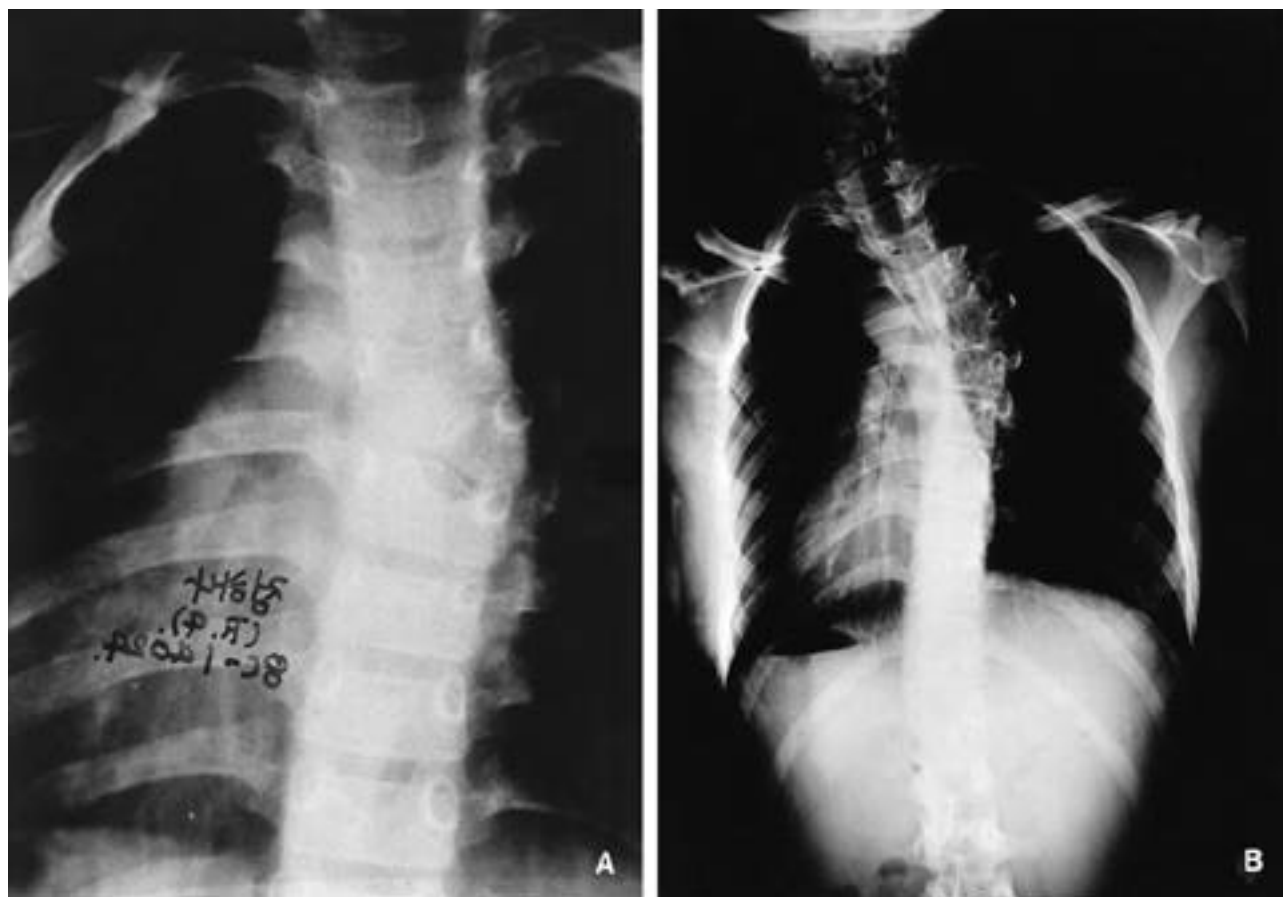
SPSS , Klippel-Feil 가 1 ,

가 가 1 가

**Table 1.** Associated anomalies in 26 patients with congenital scoliosis.

<b>Cardiac anomalies(No. of patient)</b>	
Dextrocardia(2)	Ventricular septal defect(1)
Atrioseptal defect(1)	Patent ductus arteriosus(1)
<b>Musculoskeletal anomalies(No. of patient)</b>	
Rib fusion(5)	Developmental dysplastic hip(2)
Klippel-Feil syndrome(3)	Arnold-Chiari malformation(1)
Spinal dysraphism & sacral hair patch(1)	Achondroplasia(1)
Cleft palate & congenital anklyloglossia(1)	
<b>Urogenital anomalies(No. of patient)</b>	
Unilateral renal agenesis(1)	Undescended testicle(1)
<b>Neurogenic anomalies(No. of patient)</b>	
Mental retardation(3)	Syringomyelia(2)

가 1 . (nonsegmented) 2 . 11  
(hemimetameric shift)가 5 (Fig. 2),  
가 가 2 (Fig. 5),  
가 가 2 ,  
가 2 .  
가 2 , 2 .  
2)  
가 11 (Fig. 3).  
3)  
가 가 3 ,  
가 1  
가 1  
가 1  
6 (10.5%) (Table 2).  
3.  
57 40 (70.2%) 가  
11 (19.3%),  
6 (10.5%)  
(kyphoscoliosis) 5 , (lordoscoliosis)  
1 .  
1)  
가 36 (63.2%) : 12:24  
가 25 ,  
가 11 .  
(unincarcerated hemivertebra) 25 ,  
(fully segmented) 12 (Fig. 1A, B),  
(semisegmented) 11 (Fig. 4A) 8  
, 3 ,



**Fig. 1-A.** Plain radiograph in 1988 shows right T6 hemivertebra fully segmented.  
**B.** 10 years later(1998), the Cobb's angle was increased to 39 degrees.



**Fig. 2.** Whole spine anteriorposterior view shows hemimetameric shift.



**Fig. 3.** 21-year-old-female with unilateral bar from T3-T11, the Cobb's angle was 122 degrees.

4. , (neural tube) , , , 12) , 가 14) . Wynne-Davies 337 16) 5~10% , Connor 2) 51 , 12가 , Peterson Peterson<sup>9)</sup> 가 30~60% 47 4,6) . Bernard 1) 62.5%

23 11 가 20 가 가 32.2 (10~88 ) , 55.4 (26~130 ) (Fig. 6), 39 (19~88 ) , 24.7 (10~43 ) (Table 3).

node) , 가 (proliferating cell) (primitive (invagination) (notochord) 47



**Fig. 4-A.** Whole spine anteriorposterior shows right L2 unincarcerated hemivertebra partially segmented.  
**B.** Cervical spine lateral view shows fusion of C1-2 and C4-6 spine.

가, 2 (3.5%) Drvaric <sup>4)</sup> 37%  
 Klippel-Feil 가 .  
 4 , 32 가  
 7 (21.8%) . 21  
 , , , 25 5  
 8 9% ,  
 . Prahinski <sup>10)</sup> 29 1975 Reckles <sup>11)</sup> 10% 1985 Bernard <sup>1)</sup>  
 30% 42 3  
 MRI가 , Drvaric <sup>4)</sup> 14 5 가  
 37% Klippel-Feil 3  
 가 . Winter <sup>15)</sup> Bernard <sup>1)</sup> 7% Prahinski <sup>10)</sup>  
 , Hensinger <sup>5)</sup> Klippel-Feil 60% 30% .  
 57 26 (45.6%) 1985 <sup>13)</sup>  
 , , 392 75 19.1%  
 가 34 가 2000  
 Jaskwhich <sup>6)</sup> 가 30%,



Fig. 5. MRI shows left T12 hemivertebra with T7, 8 butterfly vertebra.

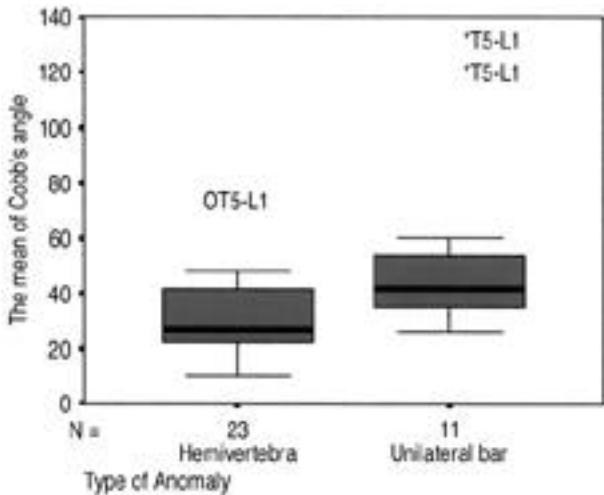


Fig. 6. Distribution of Cobb's angle of hemivertebra and unilateral bar shows 32.1 degrees of the mean Cobb's angle in hemivertebra and 55.4 degrees in unilateral bar.

1960  
가  
6). McMaster Ohtsuka<sup>8)</sup>  
가  
, 1985 Bernard<sup>1)</sup> 75%  
50%  
,  
가 ,  
2  
. McMaster  
75%  
가 가  
,  
. Jaskwhich<sup>6)</sup>  
1 1~2  
10 40  
3~4 가  
10 2~6 가 가  
가 . McMaster  
가 가  
David<sup>7)</sup>  
가  
40 30  
,  
45

40% . 1986 McMaster  
David<sup>7)</sup> 104  
67% , 11% (hemimeta-  
meric shift), 16% , 6%  
3 ,  
가 65%  
가 22%, 가  
12% , ,  
,  
가  
40 (70.2%),  
11 (19.3%), 6 (10.5%) .  
가 36 63.2%  
가 25 가  
69.4% ,  
12 48%, 11  
44% McMaster David<sup>7)</sup>  
가 .

**Table 2.** Summary of vertebral anomalies

Type of anomaly(No. of patient)
<b>Failure of formation(40)</b>
Hemivertebra(36)
Single hemivertebra(25)
unincarcerated fully segmented(12)
unincarcerated semisegmented(11)
fusion to upper vertebra(8)
fusion to lower vertebra(3)
unincarcerated nonsegmented(2)
Multiple hemivertebra(11)
hemimetameric shift(5)
butterfly vertebra+semisegmented hemivertebra(2)
butterfly vertebra+full segmented hemivertebra(2)
ipsilateral multiple hemivertebra(2)
Posterior quadrant vertebra(2)
Wedge vertebra(2)
<b>Failure of segmentation(11)</b>
Lateral unilateral bar with scoliosis(11)
<b>Mixed type(6)</b>
Posterior quadrant vertebra+block vertebra+unilateral bar(1)
Hemivertebra+block vertebra(3)
Hemivertebra+block vertebra+unilateral bar(1)
Hemivertebra+unilateral bar(1)

**Table 3.** The mean of Cobb's angle of hemivertebra and unilateral bar

Type of anomaly	location (No. of patient)	The mean of Cobb's angle
Hemivertebra (full segmented)	T*1-T4(1) T5-L <sup>†</sup> 1(8) L2-L4(3)	40 38.1 41
Hemivertebra (semisegmented)	T1-4(1) T5-L1(5) L2-L4(5)	39 22.2 24.4
Unilateral bar	T1-4(2) T5-L1(7) L2-L4(2)	32.5 66.3 40

T\*: thoracic spine; L<sup>†</sup>: lumbar spine

가 30 1 40 (70.2%), 11 (19.3%) , 가 36  
6 (10.5%) 가 26 ,  
(63.2%) 가 , 2 가 .  
가 . 가  
10 40 가  
3 5 50  
가 , 70  
가 .  
가 , Day 3) 90  
가 .  
가 20 가 32.2 ,  
55.4 ,

39 ,  
24.7 .  
 , ,  
57  
가 36  
26 ,  
가 .  
가 .  
가 .  
가 .

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: 1994 2000 57  
 : 57 32 (56.1%), 25 (43.9%) . 40 (70.2%),  
 11 (19.3%), 6 (10.5%) . 가 36 (63.2%), (post quadrant vertebra)가  
 2 , (wedge vertebra)가 2 . 2 , 1 ,  
 1 , 1 , 5 , 2 , Klippel-Feil  
 3 , 1 , Arnold-Chiari 1 , 가 1 ,  
 1 , 2 , 3 ,  
 1 26 (45.6%) .  
 : 가 가 가 . 26 (45.6%) ,  
 , .