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ORIGINAL ARTICLE

Vertebral, rib, and intraspinal anomalies in congenital scoliosis: a study on 202 Caucasians

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Abstract

Purpose To investigate vertebral, rib and intraspinal anomalies in patients with congenital scoliosis and their association with each other

Methods Clinical data and preoperative imaging studies of 202 Caucasians with congenital scoliosis operated on at an educational hospital within 6 years were reviewed for vertebral, rib, and intraspinal anomalies.

Results Rib and intraspinal anomalies were present in 57.4 and 21.8 % of patients, respectively. Most vertebral anomalies were located in the middle–lower thorax. Being the most common vertebral defect (53.5 %), failure of segmentation was significantly more common in males, whereas mixed defects were more frequent in females. Formation and mixed defects were associated with rib changes. Vertebral anomalies were more extensive in males than in females. The presence of multiple hemivertebrae was associated with rib deformity and intraspinal anomaly. Location of the vertebral anomalies varied with gender and rib involvement. Majority of rib changes were of simple type (70.7 %), significantly more common in

males. Conversely, females had significantly more fused and bifid ribs. Two most common intraspinal anomalies were diastematomyelia (36.4 %) and syringomyelia (18.2 %). Intraspinal anomalies were located most frequently in the upper and lower thoracic regions. Syringomyelia and low conus were associated with female gender, and patients with rib changes suffered from intraspinal anomalies more frequently. No significant association was found between vertebral and intraspinal anomalies.

Conclusions The incidences of rib and intraspinal anomalies were 57.4 and 21.8 % in surgical Caucasians with congenital scoliosis, respectively. Unlike vertebral and intraspinal anomalies, rib and intraspinal anomalies were significantly associated. Male gender and intraspinal anomaly were associated with some previously suggested risk factors of curve progression.

Keyword Congenital scoliosis · Vertebral anomaly · Rib deformity · Intraspinal anomaly

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Introduction

Congenital scoliosis occurs due to abnormal development of the vertebrae in utero at 4–6 weeks of gestation, leading to a curvature of the spine [1]. Although presence of anomalous vertebra is the characteristic feature of congenital scoliosis, this condition is often accompanied with abnormal changes in the ribs and intraspinal anomalies, maybe as a result of the closely related embryonic development of the spine and ribs from the mesoderm [2–5].

It has been suggested that knowledge of the natural history of congenital scoliosis at early stages of the disease is necessary for planning prophylactic approaches.

Synchronous occurrence of vertebral, rib and intraspinal anomalies in patients with congenital scoliosis provides substantial clues for better understanding of the natural history in scoliotic patients [6]. This is particularly important with regard to the intraspinal anomalies, because they are usually occult [7] and at the same time clinically important problems in congenital scoliosis [3].

However, there are scarce studies in this regard in the literature that are limited to certain ethnic groups [7–9].

This study sought to investigate incidence, characteristics and association of vertebral, rib and intraspinal anomalies in a group of Caucasian patients with congenital scoliosis.

Materials and methods

In this retrospective study, medical records of 202 consecutive Caucasian patients with congenital scoliosis operated on at Shafa Orthopedic Teaching Hospital in Tehran from February 2008 to September 2013 were reviewed.

Patients with secondary or adult onset scoliosis, with a positive history of spinal and/or costal trauma/operation, or with incomplete required information were not included.

The required information was collected from each patient's clinical and hospital chart, including preoperative plain radiographs, computerized tomographic scans with three-dimensional reconstructions, and magnetic resonance (MR) images of the whole spine.

Medical records of 183 consecutive patients with polytrauma examined at emergency department in the same hospital served as controls. These patients were previously healthy, nonscoliotic (with no vertebral anomalies) Caucasian subjects with no noticeable surgical alterations affecting the rib cage.

Two scoliosis surgeons with over 10 years of experience reviewed the results of imaging studies independently, with a third scoliosis surgeon arbitrating disagreements.

The ethics committee of the Iran University of Medical Sciences confirmed this study.

Vertebral anomaly

Vertebral anomalies were classified according to the criteria proposed by Hedequist and Emans [10], as caused by failure of formation including hemivertebra (Fig. 1a) and wedge vertebra (Fig. 1b), failure of segmentation including unilateral unsegmented bar (Fig. 1c) and block vertebra (Fig. 1d), or both failures of formation and segmentation (mixed type) [11].

The number of involved vertebrae was regarded as the extent of anomaly.

Locations of the vertebral anomalies were also documented. The thoracic region was divided as upper thoracic (T1–T4), middle thoracic (T5–T8), and lower thoracic (T9–T12) [8].

Rib anomaly

Rib anomalies with variation in number (increased/missing) or structural changes (fused ribs, Fig. 2a, bifid ribs, Fig. 2b, and widened/irregular ribs, Fig. 2c) were classified as either simple or complex following the criteria described by Tsirikos and McMaster [12].

Accordingly, simple rib deformities were reported when one of these abnormalities was present: (a) localized fusion or bifurcation of two or three ribs, (b) small chest wall defect, and (c) increased or decreased number of rib(s).

A complex rib deformity was reported when multiple extensive rib fusions and/or bifurcations were present along with an adjacent chest wall defect due to absence or division of the ribs.

Locations of the rib anomalies were also documented.

Intraspinal anomaly

Intraspinal anomalies were classified as diastematomyelia (Fig. 3a, b), syringomyelia (Fig. 3c), tethered cord (Fig. 3d), low conus (Fig. 3d), or intraspinal tumors (Fig. 3e, f) [9, 13].

The Cobb's method was used to measure the spinal curvature on anteroposterior radiographs [14].

Statistical analysis

The SPSS software version 16.0 (SPSS Inc., Chicago, IL, USA) was used for statistical analysis. Pearson Chi-square test, Fisher's exact test, and independent samples t test were used, where appropriate. The differences with a p value ≤ 0.05 were considered as statistically significant.

Results

A total of 202 patients including 90 males (44.6 %) and 112 females (55.4 %) with a mean age of 13.5 ± 6.6 years (range 2–38) at the time of operation were studied.

The mean Cobb angle was $71.2 \pm 24.9°$ (range 30–140).

The control group consisted 73 males (39.9 %) and 110 females (60.1 %) with a mean age of 12.7 ± 5.1 years (range 2–38). Patients and their healthy counterparts were comparable in terms of sex ($p = 0.36$) and age ($p = 0.89$).

Vertebral, rib and intraspinal anomalies in scoliotic patients are set out in Table 1.
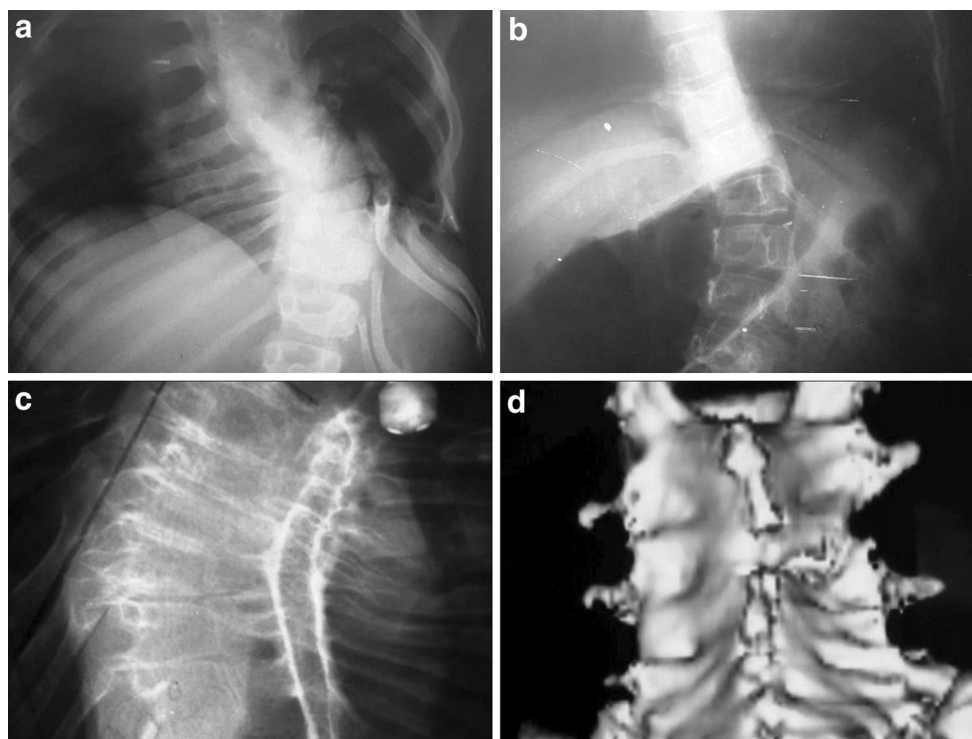
Fig. 1 Vertebral anomalies in patients with congenital scoliosis: a 13-year-old female with hemivertebrae (**a**), a 12-year-old female with wedge vertebrae (**b**), a 14-year-old female with unilateral unsegmented *bar* (**c**), and a 11-year-old female with block vertebrae (**d**)
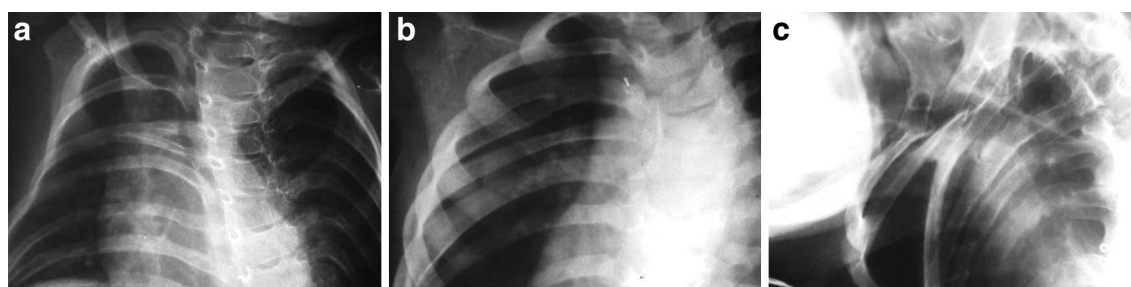

Fig. 2 Rib anomalies in patients with congenital scoliosis: a 10-year-old male with fused ribs (**a**), a 13-year-old female with bifid ribs (**b**), and a 8-year-old male with irregular ribs (c)

Failure in segmentation, formation, and both segmentation and formation was observed in 31.7, 53.5, and 14.9 % of cases, respectively. They comprised hemivertebra (53.5 %), unilateral unsegmented bar (43.6 %), wedge vertebra (14.9 %), and block vertebra (4 %), located most commonly in the thoracic region (Table 1).

Rib changes were found in 57.4 % of patients, including simple (70.7 %) and complex (29.3 %) anomalies. Rib structural changes were present in 88 cases (43.6 % of all patients, 75.9 % of patients with rib anomalies), including fused ribs (30.7 %), bifid ribs (24.8 %), and widened/irregular ribs (31.7 %).

Rib number was abnormal in 76 cases (37.6 % of all patients, 65.5 % of patients with rib anomalies), including supernumerary (18.4 %) and missing (81.6 %) ribs (Table 1).

Rib anomalies were observed in 6 control subjects (3.3 %). All these cases had numerical changes and no structural abnormalities were found.

Rib anomalies were significantly more common in scoliotic patients than in control group ($p < 0.001$, odds ratio: 39.8 with 95 % confidence interval: 16.8–94.0).

Intraspinal anomalies were found in 21.8 % of patients, including diastematomyelia (14.9 %), syringomyelia (9.9 %), tethered cord (3 %), low conus (3 %), and intraspinal tumors (2 %). The latter included intradural spinal lipoma ($n = 2$) and intradural epidermoid cyst ($n = 2$).
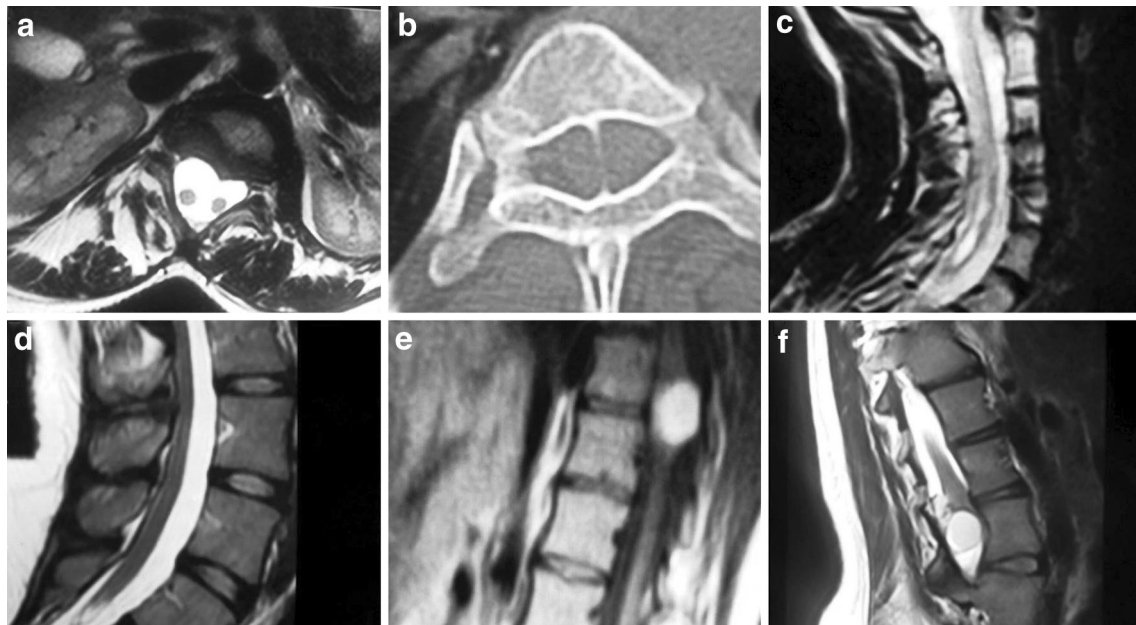
Fig. 3 Intraspinal anomalies in patients with congenital scoliosis: a 22-year-old female with diastematomyelia with fibrous septum (**a**), a 18-year-old female with diastematomyelia with bony septum (**b**), a 10-year-old female with syringomyelia (**c**), a 15-year-old male with tethered cord (L5–S1) and low conus (**d**), a 11-year-old female with intradural spinal lipoma (**e**), and a 25-year-old female with intradural epidermoid cyst (**f**)

In patients with diastematomyelia, pure fibrous septum was present in 20 patients (66.7 %) and bone septum in 8 patients (26.7 %). Bone and fibrous septum coexisted in 2 patients (6.7 %).

The intraspinal anomalies were located mostly in the thoracic region (Table 1).

Neurological problems were present in eight patients (18.2 %) with intraspinal abnormalities and 4 % of all patients, including lower limb weakness in four patients, three with syringomyelia and one with tethered cord; diminished deep tendon reflexes in two cases with syringomyelia; foot drop in one case with tethered cord; and calf muscle atrophy in another case with tethered cord.

Anomalies in the posterior elements were present in 20 cases (9.9 %) including 6 cases (2.9 %) with myelomeningocele and 14 cases (6.9 %) with asymptomatic occult spina bifida. All the patients with myelomeningocele had undergone uneventful operative corrections in early life.

The findings related to vertebral defects, rib changes, and intraspinal anomalies are compared between males and females in Table 2.

While failure of segmentation was more common in males (37.8 vs. 26.8 %), mixed anomalies were observed more frequently in females (19.6 vs. 8.9 %; $p = 0.05$). Vertebral anomalies were significantly more extensive in males than in females (2.8 ± 1.9 vs. 2.1 ± 1.3, $p = 0.003$), including the mean number of hemivertebra (4.1 ± 1.8 vs. 2.8 ± 1.1, $p < 0.001$) and wedge vertebra (1.3 ± 0.5 vs. 1.9 ± 0.8, $p = 0.02$). In addition, anomalies in the upper and lower thorax were more common in females (16.1 vs. 6.7 % and 32.1 vs. 17.8 %, respectively), and the lumbar region and upper and middle thoracic regions concomitantly were involved more frequently in males (17.8 vs. 7.1 % and 24.4 vs. 10.7 %, respectively, $p = 0.002$).

Complex rib anomalies were seen more frequently in females than in males (23.2 vs. 8.9 %, $p = 0.02$).

Among structural changes, both fused and bifid ribs were seen more frequently in females than in males (41.1 vs. 17.8 % and 35.7 vs. 11.1 %, respectively; $p < 0.001$ for both comparisons).

Among different types of intraspinal anomalies, syringomyelia and low conus were seen more frequently in female patients than in males (14.3 vs. 4.4 %, $p = 0.02$ and 5.4 vs. 0 %, $p = 0.03$, respectively).

The findings related to vertebral defects and intraspinal anomalies are compared between patients with and without rib changes in Table 3.

Patients with rib anomalies had higher rates of formation and mixed-type defects compared to those without rib anomaly (58.6 vs. 46.5 % and 19 vs. 9.3 %, respectively; $p = 0.003$).

While hemivertebra and multiple hemivertebrae were seen more frequently in patients with rib changes (62.1 vs.

Table 1 Vertebral, costal and intraspinal anomalies in patients with congenital scoliosis

Anomaly	Datum
Vertebral	202 (100)
Classification	
Segmentation failure	64 (31.7)
Formation failure	108 (53.5)
Mixed type	30 (14.9)
Types	
Single hemivertebra	62 (30.7)
Multiple hemivertebrae	26 (12.9)
Unilateral unsegmented bar	56 (27.7)
Wedge vertebra	18 (8.9)
Block vertebra	8 (4)
Unilateral unsegmented bar and contralateral hemivertebra	20 (9.9)
Unilateral unsegmented bar and wedge vertebra	12 (5.9)
Extent	2.5 ± 1.6 (1–7)
Hemivertebra	3.4 ± 1.6 (2–7)
Unilateral unsegmented bar	2.1 ± 1.6 (1–7)
Wedge vertebra	1.6 ± 0.7 (2–4)
Block vertebra	2.5 ± 0.9 (1–7)
Location	
Upper thoracic	24 (11.9)
Middle thoracic	54 (26.7)
Lower thoracic	52 (25.7)
Lumbar	24 (11.9)
Thoracolumbar	4 (2)
Upper–middle thoracic	34 (16.8)
Middle–lower thoracic	10 (5)
Rib	116 (57.4)
Types	
Simple	82 (70.7)
Complex	34 (29.3)
Side	
Convex	16 (13.8)
Concave	56 (48.3)
Bilateral	44 (37.9)
Structural changes	88 (43.6)
Fused rib	8 (9.1)
Bifid rib	6 (6.8)
Widened/irregular rib	16 (18.2)
Fused and bifid ribs	10 (11.4)
Fused and widened/irregular ribs	14 (15.9)
Bifid and widened/irregular ribs	4 (4.5)
Fused, bifid and widened/irregular ribs	30 (34.1)
Number of the ribs	
Increased	14 (6.9)
Decreased (missing rib)	62 (30.7)
Normal	126 (62.4)

Table 1 continued

Anomaly	Datum
Location	
Upper thoracic	18 (15.5)
Middle thoracic	28 (24.1)
Lower thoracic	26 (22.4)
Upper–middle thoracic	4 (3.4)
Middle–lower thoracic	32 (27.6)
Upper–middle–lower thoracic	8 (6.9)
Intraspinal	44 (21.8)
Types	
Diastematomyelia	16 (36.4)
Syringomyelia	8 (18.2)
Tethered cord	2 (4.5)
Low conus	2 (4.5)
Diastematomyelia and syringomyelia	10 (22.7)
Diastematomyelia, syringomyelia and intraspinal tumor	2 (4.5)
Diastematomyelia, tethered cord and low conus	2 (4.5)
Diastematomyelia, tethered cord, low conus and intraspinal tumor	2 (4.5)
Location	
Cervical	6 (13.6)
Upper thoracic	10 (22.7)
Middle thoracic	6 (13.6)
Lower thoracic	14 (31.8)
Lumbar	4 (9.1)
Middle–lower thoracic	2 (4.5)
Cervical, thoracic and lumbar	2 (4.5)

Data are presented as frequency (%) or mean ± standard deviation (range)

41.9 %, $p = 0.004$ and 17.2 vs. 7 %, $p = 0.03$, respectively), unilateral unsegmented bar was detected more frequently in patients with normal ribs (53.5 vs. 36.2 %, $p = 0.01$).

In comparison with patients with no rib change, those with rib anomalies had higher rate of vertebral defects located in the upper thorax (15.5 vs. 7 %) and lower rate of lumbar involvement (3.4 vs. 23.3 %, $p < 0.001$).

Intraspinal anomalies were significantly more common in patients with rib changes compared to those with intact ribs (32.8 vs. 7 %, $p < 0.001$).

Compared to patients without rib anomaly, all types of intraspinal anomalies except for intraspinal tumors were significantly more common in patients with rib changes, including diastematomyelia (22.4 vs. 4.7 %, $p < 0.001$), syringomyelia (13.8 vs. 4.7 %, $p = 0.03$), tethered cord (5.2 vs. 0 %, $p = 0.04$) and low conus (5.2 vs. 0 %, $p = 0.04$).

Table 2 Vertebral, costal and intraspinal anomalies in patients with congenital scoliosis stratified by gender

Variable	Males (n = 90)	Females (n = 112)	p value	OR	95 % CI
Vertebral anomaly					
Classification					
Segmentation failure	34 (37.8)	30 (26.8)	0.05	–	–
Formation failure	48 (53.3)	60 (53.6)			
Mixed type	8 (8.9)	22 (19.6)			
Type					
Hemivertebra	42 (46.7)	66 (58.9)	0.08	1.6	0.9–2.9
Unilateral unsegmented bar	38 (42.2)	50 (44.6)	0.73	1.1	0.6–1.9
Wedge vertebra	14 (15.6)	16 (14.3)	0.80	0.9	0.4–2.0
Block vertebra	4 (4.4)	4 (3.6)	0.10*	0.8	0.2–3.3
Multiple hemivertebrae	14 (15.6)	12 (10.7)	0.31	0.7	0.3–1.5
Unilateral unsegmented bar and contralateral hemivertebra	6 (6.7)	14 (12.5)	0.17	0.5	0.2–1.4
Extent	2.8 ± 1.9	2.1 ± 1.3	0.003**	–	–
Hemivertebra	4.1 ± 1.8	2.8 ± 1.1	<0.001**	–	–
Unilateral unsegmented bar	2.3 ± 1.7	1.9 ± 1.5	0.21**	–	–
Wedge vertebra	1.3 ± 0.5	1.9 ± 0.8	0.02**	–	–
Block vertebra	3.0 ± 1.2	2.0 ± 0.0	0.13**	–	–
Location					
Upper thoracic	6 (6.7)	18 (16.1)	0.002	–	–
Middle thoracic	26 (28.9)	28 (25)			
Lower thoracic	16 (17.8)	36 (32.1)			
Lumbar	16 (17.8)	8 (7.1)			
Thoracolumbar	2 (2.2)	2 (1.8)			
Upper–middle thoracic	22 (24.4)	12 (10.7)			
Middle–lower thoracic	2 (2.2)	8 (7.1)			
Rib anomaly	46 (51.1)	70 (62.5)	0.10	1.6	0.9–2.8
Type					
Simple	38 (42.2)	44 (39.3)	0.02	2.8	1.1–6.9
Complex	8 (8.9)	26 (23.2)			
Side					
Convex	10 (11.1)	6 (5.4)	0.13	–	–
Concave	20 (22.2)	36 (32.1)			
Bilateral	16 (17.8)	28 (25)			
Structural changes					
Fused rib	16 (17.8)	46 (41.1)	<0.001	3.2	1.7–6.2
Bifid rib	10 (11.1)	40 (35.7)	<0.001	4.4	2.1–9.5
Widened/irregular rib	28 (31.1)	36 (32.1)	0.88	1.0	0.6–1.9
Number of the ribs					
Increased	8 (8.9)	6 (5.4)	0.39	–	–
Decreased (missing rib)	24 (26.7)	38 (33.9)			
Normal	58 (64.4)	68 (60.7)			
Location					
Upper thoracic	6 (6.7)	12 (10.7)	0.41	–	–
Middle thoracic	10 (11.1)	18 (16.1)			
Lower thoracic	14 (15.6)	12 (10.7)			
>One location	16 (17.8)	28 (25)			

Table 2 continued

Variable	Males (*n* = 90)	Females (*n* = 112)	*p* value	OR	95 % CI
Intraspinal anomaly	16 (17.8)	28 (25)	0.22	1.5	0.8–3.1
Type					
Diastematomyelia	10 (11.1)	20 (17.9)	0.18	1.7	0.8–3.9
Syringomyelia	4 (4.4)	16 (14.3)	*0.02*	3.6	1.2–11.1
Tethered cord	2 (2.2)	4 (3.6)	0.69*	1.6	0.3–9.1
Low conus	0 (0)	6 (5.4)	*0.03**	0.5	0.4–0.6
Intraspinal tumor	0 (0)	4 (3.6)	0.13*	0.5	0.4–0.6
Location					
Thoracic	10 (11.1)	22 (19.6)	0.30*	0.5	0.1–1.8
Extra-thoracic	6 (6.7)	6 (5.4)			

Data are presented as frequency (%) or mean ± standard deviation

CI confidence interval, *OR* odds ratio

Italic *p* value ≤0.05 is statistically significant

Statistical analysis was performed using Chi-square test, * Fisher's exact test or ** independent sample *t* test

The mean age of patients with intraspinal anomalies was 14.14 ± 8.25 vs. 13.33 ± 6.07 years in the group without intraspinal anomalies (independent samples *t* test *p* = 0.55).

The mean preoperative Cobb angle was 71.43° ± 22.87° in patients with intraspinal anomalies versus 71.20° ± 25.33° in patients without intraspinal anomalies (independent samples *t* test *p* = 0.98).

Types and locations of vertebral defects in association with intraspinal anomalies are summarized in Table 4. Multiple hemivertebrae were significantly more frequent in patients with intraspinal anomaly compared to that in patients without intraspinal anomaly (22.7 vs. 10.1 %, *p* = 0.03). No other significant difference was found between patients with and without intraspinal anomaly.

Discussion

The present study is one of the rare investigations in the literature that examines vertebral, rib, and intraspinal anomalies at the same time in patients with congenital scoliosis.

Vertebral defects in our patients were failure in formation (53.5 %), failure in segmentation (31.7 %), and a mixed form of both (14.9 %).

The rates are in conformity with previous reports [3, 9].

Hemivertebra was the most common type of vertebral defects in this series (53.5 %), followed by unilateral unsegmented bar (43.6 %), wedge vertebra (14.9 %), and block vertebra (4 %).

Hemivertebra was also the most common vertebral anomaly in previous studies [8, 15].

Vertebral defects were most frequently observed in the middle–lower thoracic region in the present work.

Xue et al. [8] also reported the thoracic region as the most common location for vertebral anomalies in Chinese patients with congenital scoliosis.

In this study, rib anomalies were present in 57.4 % of patients, comprising simple (70.7 %) and complex (29.3 %) anomalies.

Rib anomalies were present in 50.3 % of Chinese patients in another study [8], including simple type in 55.2 % and complex type in 44.8 %.

In contrast, the incidence of rib anomalies was considerably lower (19.2 %) in another report by Tsirikos and McMaster [12], possibly because they did not include numerical rib changes in their final tally.

Rib anomalies in the present work consisted both structural (75.9 %) and numerical (65.5 %) changes. Fused, bifid and widened/irregular ribs were observed in 30.7, 24.8, and 31.7 % of patients, respectively. Increased and decreased rib numbers were present in 18.4 and 81.6 % of cases, respectively.

In line with these findings, Xue et al. [8] reported structural and numerical changes in 72.4 and 46.4 % of their patients with rib changes, respectively. Similar to our findings, fused and missing ribs were the most common rib anomalies.

Wattanasirichaigoon et al. [6] also reported similar findings.

In the current study, rib anomalies were located mostly in the middle thoracic region and on the concavity of the scoliosis.

In contrast, they were located most commonly in the lower thoracic region in a series by Xue et al. [8]. Similar

Table 3 Vertebral and intraspinal anomalies in patients with congenital scoliosis stratified by the presence/absence of rib anomaly

Anomaly	Rib anomaly		p value	OR	95 % CI
	No ($n = 86$)	Yes ($n = 116$)			
Vertebral					
Classification					
Segmentation failure	38 (44.2)	26 (22.4)	*0.003*	–	–
Formation failure	40 (46.5)	68 (58.6)			
Mixed type	8 (9.3)	22 (19)			
Type					
Hemivertebra	36 (41.9)	72 (62.1)	*0.004*	2.3	1.3–4.0
Unilateral unsegmented bar	46 (53.5)	42 (36.2)	*0.01*	0.5	0.3–0.9
Wedge vertebra	12 (14)	18 (15.5)	0.76	1.1	0.5–2.5
Block vertebra	2 (2.3)	6 (5.2)	0.47*	2.3	0.5–11.6
Multiple hemivertebrae	6 (7)	20 (17.2)	*0.03*	2.8	1.1–7.3
Unilateral unsegmented bar and contralateral hemivertebra	8 (9.3)	12 (10.3)	0.81	0.9	0.3–2.3
Extent	2.3 ± 1.4	2.6 ± 1.8	0.25**	–	–
Hemivertebra	3.1 ± 1.4	3.7 ± 1.8	0.13**	–	–
Unilateral unsegmented bar	1.8 ± 1.1	2.3 ± 1.8	0.14**	–	–
Wedge vertebra	1.4 ± 0.4	1.9 ± 0.8	0.17**	–	–
Block vertebra	2.0 ± 0.0	2.7 ± 1.0	0.42**	–	–
Location					
Upper thoracic	6 (7)	18 (15.5)	*<0.001*	–	–
Middle thoracic	24 (27.9)	30 (25.9)			
Lower Thoracic	20 (23.3)	32 (27.6)			
Lumbar	20 (23.3)	4 (3.4)			
Thoracolumbar	0 (0)	4 (3.4)			
Upper–middle thoracic	0 (0)	10 (8.6)			
Middle–lower thoracic	16 (18.6)	18 (15.5)			
Intraspinal	6 (7)	38 (32.8)	*<0.001*	6.5	2.6–16.2
Type					
Diastematomyelia	4 (4.7)	26 (22.4)	*<0.001*	5.9	2.0–17.7
Syringomyelia	4 (4.7)	16 (13.8)	*0.03*	3.3	1.0–10.2
Tethered cord	0 (0)	6 (5.2)	*0.04* *	0.6	0.5–0.6
Low conus	0 (0)	6 (5.2)	*0.04* *	0.6	0.5–0.6
Intraspinal tumor	0 (0)	4 (3.4)	0.14*	0.6	0.5–0.6
Location					
Thoracic	6 (7)	26 (22.4)	0.17*	0.8	0.7–1.0
Extra-thoracic	0 (0)	12 (10.3)			

Data are presented as frequency (%) or mean ± standard deviation

CI confidence interval, *OR* odds ratio

Italic *p* value ≤0.05 is statistically significant

Statistical analysis was performed using Chi-square test, * Fisher's exact test or ** independent sample *t* test

to the present work and another study [12], however, they found rib anomalies with the greatest incidence on the concavity of scoliosis [8].

In this study, the overall incidence of rib anomalies was 3.3 % in healthy subjects, all with abnormal rib numbers but no structural changes similar to those found in scoliotic cases.

The rate of overall rib anomalies in normal pediatric population ranges from 4.5 to 14.2 %, with the incidence of abnormal rib number varying between 4.1 and 8 % [16–19].

Although ethnic differences may explain this variation, it should be noted that the definition of rib anomalies, particularly structural changes, is different between studies.

Table 4 Vertebral versus intraspinal anomalies in patients with congenital scoliosis

Vertebral anomaly	Intraspinal anomaly		*p* value	OR	95 % CI
	No (*n* = 158)	Yes (*n* = 44)			
Failure					
Segmentation	54 (34.2)	10 (22.7)	0.28	–	–
Formation	80 (50.6)	28 (63.6)			
Mixed type	24 (15.2)	6 (13.6)			
Type					
Hemivertebra	80 (50.6)	28 (63.6)	0.13	1.71	0.86–3.40
Unilateral unsegmented bar	72 (45.6)	16 (36.4)	0.28	0.68	0.34–1.36
Wedge vertebra	24 (15.2)	6 (13.6)	0.80	0.88	0.34–2.31
Block vertebra	6 (3.8)	2 (4.5)	0.70*	1.21	0.24–6.20
Multiple hemivertebrae	16 (10.1)	10 (22.7)	*0.03*	2.6	1.1–6.3
Unilateral unsegmented bar and contralateral hemivertebra	14 (8.9)	6 (13.6)	0.39*	0.6	0.2–1.7
Extent	2.4 ± 1.7	2.4 ± 1.5	0.69**	–	–
Hemivertebra	3.3 ± 1.6	3.8 ± 1.4	0.50**	–	–
Unilateral unsegmented bar	2.1 ± 1.6	1.9 ± 1.2	0.55**	–	–
Wedge vertebra	1.6 ± 0.7	1.0 ± 0.0	0.23**	–	–
Block vertebra	2.7 ± 1.0	2.0 ± 0.0	0.42**	–	–
Location					
Thoracic	134 (84.8)	40 (90.9)	0.30	0.56	0.18–1.70
Lumbar	24 (15.2)	4 (9.1)			

Data are presented as frequency (%) or mean ± standard deviation

CI confidence interval, *OR* odds ratio

Italic *p* value ≤0.05 is statistically significant

Statistical analysis was performed using Chi-square test, * Fisher's exact test or ** independent sample *t* test

For example, Merks et al. [17] examined chest radiographs of 881 Caucasian children and reported rib anomalies in 14.2 %. These anomalies were cervical rib anomalies (6.1 %), aplasia of 12th ribs (6.6 %), lumbar ribs (0.9 %), bifurcations (0.7 %), and synostosis bridging (0.3 %).

The incidence of intraspinal anomalies was 21.8 % in the present work, including diastematomyelia (14.9 %), syringomyelia (9.9 %), tethered cord (3 %), low conus (3 %), and intraspinal masses (2 %).

The incidence of such intraspinal anomalies varies largely from 4.9 to 58 % in the literature [2, 3, 7–9, 15, 20–22]. This heterogeneity may be due to small numbers of studied cases [2, 21], ethnic differences, and technical shortcomings [20, 23].

In a Chinese series, in line with our results, diastematomyelia was the most common manifestation of intraspinal anomalies (27.2 %), followed by syringomyelia (20.2 %), low conus (6.5 %), and tethered cord (3.7 %) [8].

Similarly, major manifestations of intraspinal anomalies were diastematomyelia (17.6 %), syringomyelia (5.0 %), tethered cord (4.1 %), and intraspinal mass (1.5 %) in another study [1].

Diastematomyelia was also the most common intraspinal anomaly associated with congenital scoliosis in other studies [15, 22, 24].

In patients with diastematomyelia, fibrous septum, bone septum and a combined form were present in 66.7, 26.7, and 6.7 % of patients in the present work, similar to findings by other studies [8, 22].

In an attempt to clarify the role of gender, it was found that the failure of segmentation was significantly more common in males, and mixed form was seen more frequently in females. In addition, vertebral defects in females were located more frequently in the upper–lower thoracic region, whereas in males they were more common in the upper–middle thoracic and lumbar regions.

Although gender differences have been implicated in scoliotic curve progression [25, 26], it is not known whether they may also contribute to such differences between males and females.

Some studies have suggested a role of genes in differentiation of the vertebral column and ribs [27, 28]. The role of gender-related genes in this regard, however, needs to be elucidated later.

Comparing the two groups with and without coexisting rib anomalies showed that the cases with formation and mixed-type defects were significantly more frequent in the first group. In addition, hemivertebrae were significantly more common in patients with rib anomalies, whereas the incidence of unilateral unsegmented bars was significantly higher in cases without rib changes.

A close association between embryological development of the vertebrae and ribs justifies the coexistence of these developmental abnormalities in patients with congenital scoliosis [29].

Lateral moieties of the somites are the origins of the ribs. Chondrification begins in the seventh fetal week and by the end of fourth month surrounds most of the thoracic wall [6], a period within which the process of segmentation and resegmentation of the developing somites also occurs. This synchrony may describe the associations of particular types of vertebral anomalies and their location with rib anomalies in congenital scoliosis [8].

A significant association between rib changes and vertebral anomalies, independent of their type, is consistent with this previously suggested hypothesis that thoracic vertebral and/or rib anomalies occur as a result of interaction in developmental time of the independent variables of vulnerabilities of rib and/or vertebral primordial and teratogenic stimulating factors [8].

In our series, rib anomalies were detected more frequently in patients with defective vertebrae located in the upper thorax than those with vertebral anomalies in the lumbar region.

Xue et al. [8] showed that rib anomalies were most common in cases with thoracic/thoracolumbar vertebral changes, but they did not report their exact locations in the thoracic region. The authors also found that females had rib changes more commonly than males.

Although the overall incidence of rib changes did not differ significantly between males and females in our study, complex rib changes and fused/bifid types were significantly more common among females than males.

While the overall incidence of intraspinal anomalies was similar between male and female patients in the current work, frequencies of syringomyelia and low conus were significantly higher among females. We did not also find a significant association between intraspinal anomalies and age of the patients.

These findings are in contrast with the results of a previous study by Liu et al. [1], in which intraspinal anomalies were more likely to be found in females or older patients. Two other Chinese studies [8, 22] also reported higher incidence of intraspinal anomalies in females than in males. These three studies, however, did not examine the frequency of each type of intraspinal anomalies individually between males and females.

In line with a previous study [8], we found that the incidence of intraspinal anomalies was significantly higher in patients with than without rib changes.

In conformity with a previous study [7], we found no significant correlation between the magnitude of scoliosis and the presence of intraspinal anomalies.

In contrast with some previous studies [3, 8, 22], we did not observe any significant connection between the presence of intraspinal anomalies and the type or location of vertebral pathologies. Although ethnic differences may explain these conflicting findings, further studies are needed.

Neurological problems were present in 4 % of our patients, 18.2 % of those with intraspinal anomaly. In a study by McMaster [20], the incidence of neurological abnormalities was 4.8 %. In another study by Rajasekaran et al. [7], 20 % of patients with congenital scoliosis (42.9 % of those with intraspinal anomaly) had clinically detectable neurological abnormalities. Different ages at the time of examination and the severity of other complication(s) could underlie this heterogeneity.

Posterior element deficiencies were present in 9.9 % of our patients including asymptomatic occult spina bifida (6.9 %) and operatively corrected myelomeningocele at early ages (2.9 %).

It has been shown that while understanding the posterior anatomy is essential for planning surgery and avoiding unexpected intraoperative findings, the posterior element anatomy is generally neglected in dealing with congenital scoliosis [30, 31] and the true incidence of posterior element defects is not known.

In a recent study, McMaster [32] found that the extent of unsegmented bar and the presence of unilateral unsegmented bar, multiple (double) hemivertebrae, and unilateral unsegmented bars with contralateral hemivertebra may significantly contribute to curve progression in patients with congenital scoliosis.

Although the present study was not originally designed to examine prognostic factors, due to the clinical importance of the issue these factors were tested, as well.

Accordingly, vertebral anomalies were significantly more extensive in males than in females, but their extent was not associated with rib or intraspinal anomalies.

Unilateral unsegmented bars were significantly more frequent in patients without rib anomalies. In contrast, multiple hemivertebrae were significantly more common in patients with rib anomalies, as well as in patients with intraspinal problems.

Finally, unilateral unsegmented bars with contralateral hemivertebra were not associated with patients' gender, presence or absence of rib anomaly, or the status of the intraspinal problems.

The only unexpected finding in this regard was a significant association between unilateral unsegmented bars and normal ribs. This finding coupled with a significant association between multiple vertebrae and rib anomalies might cancel out any significant role of rib anomaly in curve progression, a finding that was demonstrated previously [32].

This study was carried out on operated cases with congenital scoliosis. Although because of curve progression surgery remains the fundamental treatment of congenital scoliosis and nonoperative management is of limited value in this regard [33], it should be acknowledged that in patients not being operated, the incidence, extent or location of anomalies might differ. It needs to be investigated in future studies.

In summary, this study provided characteristic details of a large number of surgical Caucasian patients with congenital scoliosis. Vertebral, costal and intraspinal involvements were scrutinized in terms of type, extent and location, and their interrelations were examined. According to our results, rib and intraspinal anomalies were present in 57.4 and 21.8 % of patients, respectively. While vertebral and intraspinal anomalies, unlike some previous reports, were not associated, the presence of rib abnormality highly suggested concomitant intraspinal anomalies. Some of vertebral indicators of curve progression, namely the extent of unsegmented bars and the presence of multiple hemivertebrae, were associated with male gender and intraspinal anomalies, indirectly suggesting prognostics roles for the latter parameters.

Conflict of interest None.

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