

Clinical Characteristics and Thoracic factors in patients with Idiopathic and Syndromic Scoliosis Associated with Pectus Excavatum

Ryoji Tauchi¹⁾, Yoshitaka Suzuki²⁾, Taichi Tsuji¹⁾, Tetsuya Ohara¹⁾, Toshiki Saito¹⁾, Ayato Nohara³⁾, Kazuaki Morishita¹⁾, Ippei Yamauchi¹⁾ and Noriaki Kawakami¹⁾

1) Department of Orthopedic Surgery, Meijo Hospital, Aichi, Japan

2) Department of Orthopedic Surgery, Nagoya Daini Red Cross Hospital, Aichi, Japan

3) Department of Orthopedic Surgery, JCHO Tokyo Shinjuku Medical Center, Tokyo, Japan

Abstract:

Introduction: The purpose of this study is to demonstrate the clinical characteristics and thoracic factors such as sternal tilt angle and Haller index in patients with idiopathic or syndromic scoliosis associated with pectus excavatum.

Methods: We performed a retrospective review on a cohort of 70 patients (37 males and 33 females) diagnosed with idiopathic and syndromic scoliosis associated with pectus excavatum between 1985 and 2014. We investigated age, location and Cobb angle of the main curve, and thoracic factors including sternal deviation and tilting angle and Haller index using radiographs and computed tomography of the chest.

Results: Patients' mean age at the first visit to our hospital was 10.3 years (1-18 years old). There were 41 patients with idiopathic scoliosis and 29 with syndromic scoliosis. Main curve locations were thoracic in 52 patients, thoracolumbar in 10, and lumbar in 8. The mean Cobb angle of the main curve was 45.0 degrees (11-109 degrees). The sternum was displaced on the left side in 72% of patients, central in 23%, and right in 5%. Mean sternal tilt angle was 12.4 degrees (2.3-34 degrees), and mean Haller index score was 4.9 (2.9-9.2). There was no significant correlation between Cobb angle and sternal tilt angle/Haller index. However, a significant difference was found between sternal tilt angle and Haller index.

Conclusion: Most patients with both scoliosis and pectus excavatum have left side deviated sternum and a higher Haller index score; therefore this can negatively impact cardiac function. Prone positioning and the corrective force applied during scoliosis surgery as well as thoracic compression during cast or brace treatment may have a negative effect on cardiac function in these patients.

Keywords:

scoliosis, pectus excavatum, sternum, Haller index

Spine Surg Relat Res 2018; 2(1): 37-41
dx.doi.org/10.22603/ssrr.2017-0027

Introduction

Pectus excavatum is the most common morphological chest wall abnormality with a reported incidence of 0.1-0.3%^{1,2)}. The pathogenesis of pectus excavatum is unknown, but it appears to be polygenetic following autosomal dominant, autosomal recessive, X-linked, or sporadic patterns of inheritance^{3,4)}. Several studies on pectus excavatum patients evaluated the coexistence of scoliosis and concluded that the rate of scoliosis in patients with pectus excavatum was higher than the rate in the general population⁵⁻⁹⁾. Gurnett CA

et al. found that adolescent idiopathic scoliosis and pectus excavatum may have a similar genetic etiology, and the prevalence of scoliosis among pectus excavatum patients is higher than the prevalence within the general population¹⁰⁾. Furthermore, pectus excavatum is frequently associated with heritable disorders of the connective tissue, such as Marfan syndrome in which almost half the patients have pectus excavatum and a large number have scoliosis¹¹⁻¹⁵⁾. However, to our knowledge, although a few studies have reported on the clinical characteristics of patients having both scoliosis and pectus excavatum, they do not give a detailed discussion and

Corresponding author: Noriaki Kawakami, nupriver@gmail.com

Received: April 6, 2017, Accepted: June 23, 2017

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the degree of scoliosis is not severe^{6,8,9,16,17}. The purpose of this study is to demonstrate clinical features and thoracic factors, such as sternal tilt angle and Haller index, in patients that have idiopathic or syndromic scoliosis associated with pectus excavatum.

Methods

Patient Selection and Clinical Investigation

From April 1985 to February 2014, 113 patients with both scoliosis and pectus excavatum were admitted to our hospital. We excluded adult scoliosis (age more than 20 years), congenital scoliosis, neuromuscular scoliosis, neurofibromatosis type 1, scoliosis of less than 10 degrees, and patients with incomplete clinical data or images. A total of 70 patients (37 male and 33 female) that had idiopathic or syndromic scoliosis associated with pectus excavatum were evaluated. We investigated age, location and Cobb angle of the main curve, thoracic factors including the deviation and tilting angle of the sternum, and the Haller index using radiographs and computed tomography of the chest. We also evaluated whether patients received surgical correction of their pectus excavatum.

Radiologic Evaluation

CT scans were used to determine sternal deviation (left, center, right), sternal tilt (left or right inclination and angle), and Haller index. We evaluated sternal deviation and tilt in 53 patients, but we were only able to estimate sternal tilt angle and Haller index in 29 patients because some cases did not have an image of the entire chest wall and others did not have CT images taken prior to corrective surgery for the pectus excavatum. We defined sternal tilt as the angle generated by the sternum and the axis of the body at the most collapsed plane of the sternum in axial CT scans (Fig. 1). Position of the sternum was defined as right, center, or left based on the position of the center of the sternum relative to

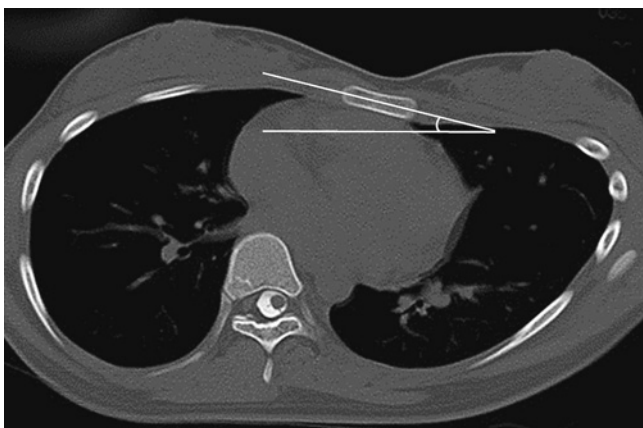


Figure 1. Sternal tilt angle. Axial CT scan showing the angle generated by the sternum and the axis of the body at the most depressed plane of the sternum.

the vertebral body on axial CT scan.

We calculated the Haller index as the inner transverse thoracic diameter divided by the anteroposterior distance between the anterior thoracic wall and the spine at the narrowest point (Fig. 2). In normal individuals, the average Haller index is 2.5, whereas with a significant pectus deformity it is 3.25 or greater¹⁸. In patients with scoliosis and pectus excavatum, since the sternum tends to be deviated unilaterally, we modified the method for calculating the Haller index (Modified Haller index; Fig. 2). Two senior spine surgeons (R.T., Y.S.) measured each parameter twice, and the average score was used for analysis. The intraobserver correlation coefficients were 0.909 (95% confidence interval, 0.830-0.952) and 0.991 (95% confidence interval, 0.983-0.996), and the interobserver coefficient was 0.965 (95% confidence interval, 0.934-0.982).

Statistical Analysis

Data were analyzed using SPSS (version 20.0, IBM Corp, New York, NY). We used Pearson's correlation coefficient to analyze thoracic factors (sternal tilt angle and Haller index) and Cobb angle. The threshold for significance was a *p* value of <0.05.

Results

Patients' mean age at presentation to our hospital was 10.3 years old (1-19 years old). There were 41 patients with idiopathic scoliosis, 29 patients with syndromic scoliosis, including 17 Marfan syndrome, 6 marfanoid types, 1 Prader-Willi syndrome, 1 Sotos syndrome, 1 Down syndrome, and 3 others. The main areas of spinal curvature were thoracic in 52 patients, thoracolumbar in 10, and lumbar in 8. The mean Cobb angle of the main curve was 45.0 degrees (11-109 degrees) (Table 1). The sternum was displaced to the left side in 72% of patients, central in 23%, and to the right in 5%. As for the sternal inclination, 23 patients had a right

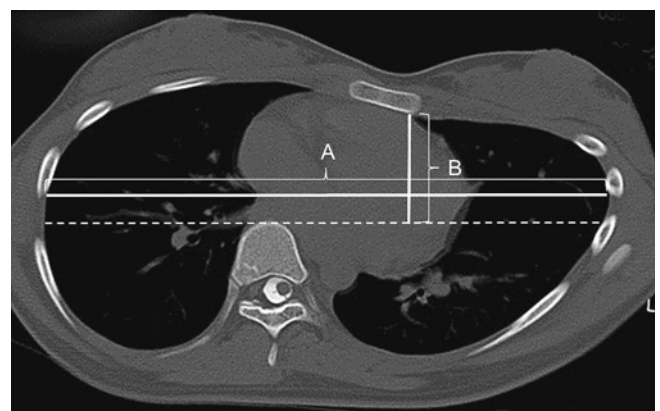


Figure 2. Modified Haller index. Axial CT scan showing positions for measuring. A = distance of the maximal inner transverse thoracic diameter. B = distance between the parallel line on the vertebra to the deepest point of the sternum at the most depressed plane of the sternum.

Table 1. Patient's Characteristics.

Case	n=70
Age (1 st visit)	10.3 (1 ~ 19)
Sex (male/female)	37/33
Types of scoliosis	
Idiopathic	41
syndromic	29
Main curve	
thoracic	52
thoracolumbar	10
lumbar	8
Cobb angle	Ave. 45.0° (11 ~ 109°)

Table 2. Thoracic Parameters in Patients with Both Scoliosis and Pectus Excavatum.

Thoracic parameters	
Cases	n=43
Location of sternum	left 31 central 10 right 2
Sternal inclination	right 21 left 22
Sternal tilt angle	Ave. 12.4° (1.8 ~ 34°)*
Haller index	Ave. 4.9 (2.9 ~ 9.2)*

*29 cases which could be evaluated about all parameters by CT scan of the thorax.

inclination, and 22 had a left inclination among 45 patients who could be evaluated by CT scan. The mean sternal tilt angle was 12.4 degrees (1.8-34 degrees) and mean Haller index was 4.9 (2.9-9.2) among 29 patients who had their parameters evaluated by a CT scan of the whole chest area. (Table 2).

There was no significant correlation between Cobb angle and thoracic factors (sternal tilt angle and Haller index); p-values were 0.371 and 0.159, respectively. However, with regard to the severity of the pectus excavatum, a significant difference was found between sternal tilt angle and Haller index ($p = 0.009$) (Table 3). Twenty-four patients (34%) received surgical correction of their pectus excavatum. In the scoliosis surgery group (27 patients), 16 patients had surgical correction of their pectus excavatum prior to scoliosis surgery, and 2 of the 14 patients in the scoliosis cast/brace treatment group had pectus excavatum surgery prior to applying the cast or brace (Fig. 3). The Nuss procedure using a chest bar or open repair procedure such as Ravitch procedure were performed for the pectus excavatum. Regardless of whether pectus excavatum surgery was done before scoliosis treatment, none of the patients experienced severe hypotension or cardiopulmonary dysfunction during scoliosis correction surgery or cast or brace treatment.

Table 3. Correlation between Cobb Angle and Thoracic Parameters.

		Cobb angle	Sternal tilt angle	Haller index
Cobb angle	r	-	0.172	0.269
	P value	-	0.371	0.159
Sternal tilt angle	r	-	-	0.473
	P value	-	-	0.009

Discussion

This study presents the clinical features and thoracic factors, such as sternal tilt angle and Haller index, seen in 70 patients diagnosed with scoliosis associated with pectus excavatum. Most of these patients had a left side deviated sternum and a higher Haller index score. Although the Cobb angle and thoracic parameters did not significantly correlate with each other, with regard to the severity of the pectus excavatum, a significant difference was found between sternal tilt angle and Haller index. Regardless of whether pectus excavatum surgery was performed before scoliosis treatment, none of our patients experienced severe hypotension or cardiopulmonary dysfunction during their scoliosis correction surgery or cast or brace treatment.

Several studies have reported the incidence of scoliosis in association with pectus excavatum. In 1989, Waters P et al. evaluated the incidence of scoliosis in 461 patients with pectus excavatum. They found that 99 of their study's patients (21.5%) had scoliosis with an average Cobb angle of 15 degrees (range 6-78 degrees)⁹. Hong JY et al. examined 248 patients diagnosed with pectus excavatum for the incidence of scoliosis, and the prevalence of idiopathic scoliosis was 22.58%. Furthermore, the incidence of scoliosis was significantly higher in female patients, and the Lenke type 1 predominated in pectus patients (48.2%)⁶. Similarly, Wang Y et al. reported that 25 out of 142 patients (17.6%) with pectus excavatum had scoliosis with a Cobb angle > 10 degrees, and in 80% of the cases the spinal column was bent to the right. They found that adolescents had a higher incidence than pediatric patients, and those with a Cobb angle greater than 10 degrees were mainly within the severe scoliosis group and had Haller indices ≥ 3.5 ⁸.

On the other hand, several authors reported severe complications associated with the prone position during surgery in children with scoliosis and pectus excavatum. Alexianu D et al. reported the case of a child with neurofibromatosis, pectus excavatum, and rapidly progressive scoliosis who developed severe hypotension after placement in the prone position during elective posterior spinal fusion. Transesophageal echocardiogram (TEE) revealed compression of the right ventricle by the sternum due to the compression of the transverse bolsters. After repositioning the longitudinal bolsters, the patient remained hemodynamically stable with no compression of the right ventricle evident on the TEE, and the posterior fusion proceeded uneventfully¹⁹. Bafus BT

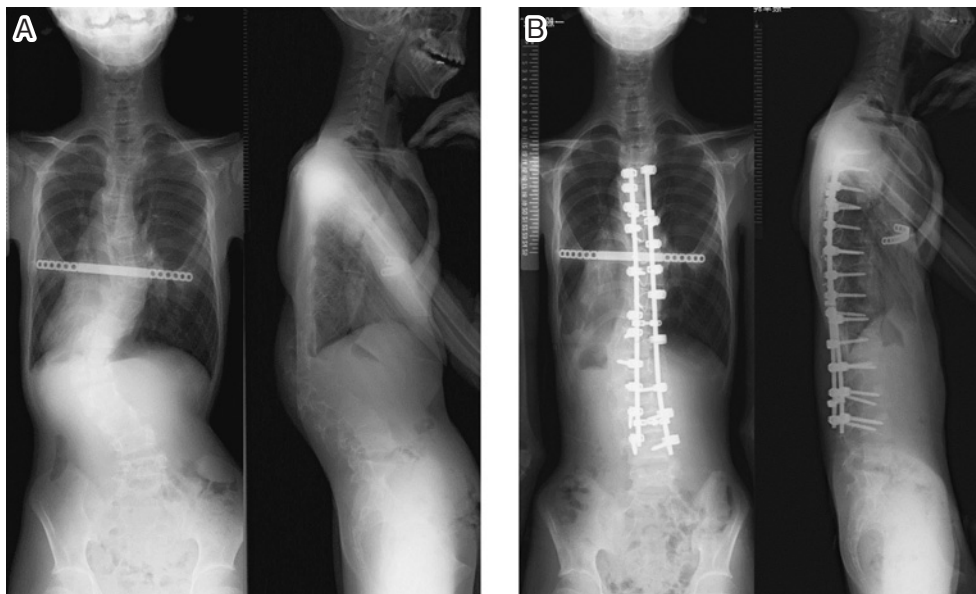


Figure 3. A 13-year-old male patient with syndromic scoliosis (Marfan syndrome) and pectus excavatum. He underwent anterior and posterior fusion surgery (T4/L4) after corrective surgery of the pectus excavatum.

A. Preoperative: thoracic 49 degrees, main thoracolumbar 64 degrees of coronal curvature in A-P and lateral radiographs.

B. Postoperative: thoracic 25 degrees, main thoracolumbar 21 degrees of coronal curvature in A-P and lateral radiographs.

et al. also reported a case of severe hypotension in a child with pectus excavatum who seemed marfanoid undergoing posterior spinal fusion for scoliosis after the patient was placed in the prone position. They accomplished the posterior spinal fusion for scoliosis after performing a modified Ravitch procedure to repair the pectus excavatum²⁰. Similarly, Galas JM et al. also presented a case with pectus excavatum and thoracolumbar scoliosis that developed severe hypotension after placement in the prone position for spinal fusion. A TEE showed the sternum compressing the right heart. After Nuss bar placement to correct the pectus excavatum, the patient underwent posterior spinal fusion uneventfully²¹.

Malek MH et al. performed a meta-analysis on cardiovascular function following surgical repair of pectus excavatum, and they demonstrated that surgical repair of the pectus excavatum significantly improves cardiovascular function²².

There were several limitations in our study. First, this was a retrospective study with a small sample size. We could not collect appropriate numbers of patients with each type of scoliosis. Second, we could not evaluate thoracic parameters in all the cases because some CT scans did not reveal the entire chest wall in the image. On the other hand, to the best of our knowledge, this is the first study to report thoracic factors in patients with both scoliosis and pectus excavatum which included idiopathic or syndromic type and varying degrees of scoliosis from moderate to severe which required treatment such as surgery or cast/brace. Finally, we suggest that the orthopedic surgeons fully assess the thoracic factors in patients with scoliosis and pectus excavatum prior

to performing scoliosis surgery or cast/brace treatment and carefully monitor their patient's general condition during scoliosis treatment.

Conclusion

We evaluated 70 idiopathic or syndromic scoliosis patients who also had pectus excavatum. In most cases, the sternum was located on the left side, and their Haller index scores were higher than the normal population. Prone positioning, the corrective force applied during scoliosis surgery, and the thoracic cage compression that occurs during cast or brace treatment might negatively affect cardiac function in patients with both scoliosis and pectus excavatum.

Conflicts of Interest: The authors declare that there are no conflicts of interest.

Acknowledgment: We sincerely express our appreciation to Ms. S Hiraiwa and Michael Anton Castro for their contribution to this project.

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