Double crush to the thorax: Pectus excavatum and kyphoscoliosis

Elizabeth A. Berdan a,*, David J. Nuckley b, David W. Poly Jr. b, Daniel A. Saltzman c

a Department of Surgery, University of Minnesota, MMC 195, 420 Delaware St. S.E., Minneapolis, MN 55455, USA
b Department of Orthopaedic Surgery, University of Minnesota, USA
c Division of Pediatric Surgery, University of Minnesota, USA

Abstract

The relationship between adolescent idiopathic scoliosis (AIS) and pectus excavatum (PE) is not well understood. We hypothesize that the combined thoracic malformations of AIS and PE may lead to a “double crush to the thorax” resulting in greater cardiopulmonary impairment than either thoracic malformation in isolation. We evaluated the Haller index (HI) and thoracic volume changes that occur pre- and post-operatively for kyphoscoliosis and PE by examining the case of a 12-year-old girl who had a “double crush to the thorax.” Using posterior-to-anterior and lateral standing scoliosis films obtained before and after both operations we determined the PE severity index (HI). Using the same radiographs a Blender based computational model was used to approximate the thoracic volumes. With correction of kyphoscoliosis the HI acutely increased from 1.9 to 3.2 with a decrease in thoracic volume of 11% resulting in shortness of breath and tachycardia. With PE repair the HI was returned to normal (HI = 2.4) and the thoracic volume increased 18%, with resolution of her cardiopulmonary symptoms. When severe kyphoscoliosis and PE co-exist consideration should be given to the implication of the “double crush to the thorax” if cardiopulmonary symptoms arise.

© 2014 The Authors. Published by Elsevier Inc. Open access under CC BY-NC-ND license.

Article info

Article history:
Received 12 June 2013
Received in revised form 19 November 2013
Accepted 19 November 2013
Available online 4 December 2013

Key words:
Pectus excavatum
Adolescent idiopathic scoliosis
Thoracic volume

Adolescent idiopathic scoliosis (AIS) severe enough to warrant spinal arthrodesis is relatively common with an estimated prevalence of 0.04% [1,2]. Pectus excavatum (PE), or funnel chest, is an anterior chest wall malformation which characteristically has a decreased anteroposterior diameter of the mediastinum. PE is estimated to occur in 1 out of 400–1000 live births making this the most common congenital malformation of the chest wall [3]. It is estimated that 16–23% of patients with PE have AIS [4–6]; however, there are no studies estimating the prevalence of PE in the AIS population let alone investigating the cardiopulmonary pathology of this patient population. This case report illustrates the acute thoracic volume changes that occur as the result of surgical correction of kyphoscoliosis and pectus excavatum in a young girl.

1. Case report

This is a 12-year-old female with a history of arthrogryposis, congenital short stature, neurologic delay, infantile endocarditis and tracheostomy dependence until the age of 3 years for thoracic dystrophy. She also presented with severe kyphoscoliosis and clinically profound pectus excavatum. Despite these limitations, she was a very active child that fully participated in her activities of daily living without reports of cardiopulmonary insufficiency. She suffered from the “double crush to the thorax” with both thoracic deformities (kyphoscoliosis and pectus excavatum) requiring surgical correction. She developed progressive kyphoscoliosis with a Cobb angle of 31° and kyphosis of 70° as a result of growth hormone injections for the treatment of her congenital short stature. She underwent posterior spinal fusion (PSF) from T3 to L3 with Smith-Peterson osteotomies. After PSF of her kyphoscoliosis she developed severe cardiopulmonary symptoms that resolved with the correction of her PE eight months later using the minimally invasive Nuss repair.

The patient was a 12-year-old girl who presented with progressively severe kyphoscoliosis and clinically evident PE. While her Haller index (HI) was 1.9 clinically her sternum was not correctly positioned due to her kyphosis. The HI is defined by the horizontal distance of the ribcage divided by the shortest distance between the vertebrae and sternum (Fig. 1). In Fig. 1 the sternum is located posterior to her abdominal wall. While the HI was initially defined using computed tomography (CT), 2-dimensional chest radiographs are acceptable in patients that do not otherwise require a chest CT [7,8]. Using posterior-to-anterior and lateral standing
scoliosis films obtained before and after both operations the HI was determined (Figs. 1–3). Using the same radiographs a computational model was utilized to estimate the thoracic volume. The computational model consists of a skeletal deformable body model within Blender 2.63 atm software wherein the spine and ribs may be deformed to match the biplanar radiographs. The 3D deformed spine and thorax are then used to compute the thoracic volume. This methodology has exhibited a mean error of 2.4% when compared with CT reconstructions of the same individual. This methodology minimizes thoracic volume estimation errors.

The pre-operative transthoracic echocardiogram showed trace mitral valve thickening with trace regurgitation. There are no pulmonary function tests available as the patient was unable to complete the study. Once her kyphoscoliosis was repaired the pectus excavatum severity index (HI) was acutely increased from 1.9 to 3.2 (Figs. 1 and 2) which resulted in significant cardiopulmonary symptoms. As determined by computational thoracic modeling, her initial thoracic volume was 2209 cm$^3$. After correction of her kyphoscoliosis the thoracic volume decreased to 1956 cm$^3$, a decrease of 11%. With correction of her pectus excavatum the HI was returned to normal, HI = 2.4 (Fig. 3). The PE repair resulted in a thoracic volume increase to 2312 cm$^3$, which is an 18% increase in thoracic volume after repair of her kyphoscoliosis and a 5% increase in thoracic volume overall.

2. Discussion

This study is the first to describe the volumetric thoracic changes that occur before and after repair of a severe kyphoscoliotic deformity and PE. As a result of the spinal arthrodesis, she sustained an acute decrease in her mediastinal volume identified by the acute increase in her HI (1.9–3.2). The HI is used to define the severity of pectus excavatum with a normal HI = 2.5. A HI of greater than or equal to 3.2 indicates a severe PE deformity that necessitates surgical correction. The HI and was initially described using a chest CT; however, two-view chest radiographs are now utilized [7,8]. Following her spinal fusion she was unable to ascend a flight of stairs without tachycardia or shortness of breath. These symptoms resolved after repair of her pectus excavatum, which returned her to a normal HI of 2.4 (Fig. 3). The patient’s cardiopulmonary symptoms after correction of her kyphosis are likely due to the acute volumetric change of the patient’s mediastinum causing restrictive cardiac pathology. Once the sternum was surgically lifted, space was made available for improved cardiac form and function thus resolving her symptoms.

The literature regarding the pathophysiology of PE and the effect of corrective surgery are controversial given the difficulty with the evaluation tools (assessing the right ventricle with a transthoracic echocardiogram, pulmonary function tests in young children), the small numbers studied and differences in study protocol. Many studies report a restrictive pulmonary pattern in the patient with extreme pectus excavatum deformities [9–12]. Some studies suggest that there is no effect on the heart with a PE deformity or a change in cardiac function with corrective surgery [13]. Still other investigators have found a difference in PE patients with an improvement in cardiac form and function after relief of cardiac compression with the corrective surgery [10,14–17]. Overwhelmingly, patients report resolution of their cardiopulmonary symptoms upon correction of the deformity leading us to conclude that the current assessment tools are not measuring the appropriate variable(s), and perhaps they lack the sensitivity required to characterize the pathophysiology of this complex thoracic deformity.

The dynamic three-dimensional physiology of the chest cavity is difficult to objectively quantitate. The breathing pattern is altered in kyphoscoliosis and pectus excavatum requiring patients to work harder to generate an inspiratory effort that achieves the necessary trans-diaphragmatic pressure for a normal breath. While the goal of surgical correction of severe AIS is to maintain pulmonary function

![Fig. 1. Prior to correction of kyphoscoliosis. Posterior-to-anterior and lateral standing scoliosis radiographs.](image-url)
Fig. 2. After posterior spinal fusion of kyphoscoliosis. Posterior-to-anterior and lateral standing scoliosis radiographs.

Fig. 3. After correction of pectus excavatum. Posterior-to-anterior and lateral standing scoliosis radiographs.
it has been shown that the approach used, specifically violation of the anterior thoracic cage, may decrease pulmonary function [18]. The majority of the pulmonary function data poorly correlates to the two-dimensional radiographic measurements made in AIS [19]. This may be accounted for, in part, by unrecognized anterior chest wall malformations and the resulting complex thoracic kinematics, which have been shown to be altered in malformations of the spine and chest wall [20, 21].

3. Conclusion

The thoracic malformations of scoliosis and pectus excavatum in isolation have complicated and poorly understood effects on the dynamic kinematics of pulmonary and cardiac physiology. We hypothesize that the combined thoracic malformations of AIS and PE may lead to greater cardiopulmonary impairment than either thoracic malformation in isolation, termed the “double crush to the thorax.” Complex thoracic malformations may have significant implications for the clinical and surgical management of this patient population. This is particularly important to consider should cardiopulmonary symptoms arise post-operatively. If the patient becomes symptomatic, consideration should be given to an altered thoracic volume that impacts cardiopulmonary form and function. The combined thoracic malformations of PE and AIS have intrigued the orthopedic and pediatric surgery communities for the past five decades. Awareness that an acute change in mediastinal volumes after repair of kyphoscoliosis may occur is an important consideration when treating a patient who develops cardiopulmonary symptoms post-operatively.

Conflict of interest

No authors have a conflict of interest as it relates to the generation and production of this manuscript.

Source of support

This project was supported by a grant obtained from the Chest Wall and Spine Deformity Research Foundation.

References


