

Pectus excavatum

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In pectus excavatum, a developmental chest-wall deformity that affects approximately one in 400 births, the sternum is depressed inward and the ribs protrude anteriorly. This deformity can compromise pulmonary and/or cardiac function. It can also produce a characteristic radiographic appearance on frontal radiographs, which may be mistaken for right-middle-lobe opacification from pneumonia or atelectasis. I present a case of pectus excavatum in a teenage female with characteristic imaging findings, and the disposition of the case.

Case report

A 13-year-old female presented to Pediatrics with a complaint of pectus excavatum. She was concerned that it was embarrassing when others saw it. She and her parents were concerned that it would grow worse as she grew older. She had no complaints of breathing difficulty or chest pain. There is a family history of pectus excavatum in her father, who has remained asymptomatic. She was initially evaluated with chest x-ray, posteroanterior (PA) and lateral.

The PA radiograph revealed symmetric lung volumes with an apparent increased density overlying the right anteromedial aspect of the right middle lobe (Fig. 1A). The heart and mediastinum were displaced to the left. The remainder of the lungs were clear, with sharp pleural margins and an otherwise unremarkable cardiomedial silhouette. A mild dextroconvex curvature of the thoracic spine was present. The lateral radiograph revealed an opacity (the sternum) that filled the retrosternal air space, creating an edge that overlay a portion of the cardia (Fig. 1B). The sternum was displaced inwardly, with ribs seen anterior to the sternum, confirming the diagnosis of pectus excavatum.

The patient was referred to a pediatric cardiothoracic surgeon; before the surgical evaluation, she was seen by a pediatric cardiologist and evaluated with echocardiography. The cardiologist obtained a history that the patient experienced sharp chest pains that occurred 3 to 4 times per day and lasted only a few seconds. The patient also complained of dull chest pains that occurred less frequently but could last a few minutes to an hour. She complained of increasing shortness of breath and fatigue while playing sports. The echocardiogram revealed that the pectus deformity was compressing the right ventricle, although it did not obstruct inflow or outflow of the ventricle. The cardiologist recommended that the pectus excavatum be repaired. The cardiothoracic surgeon also obtained pulmonary function testing (PFT), and CT of the chest. The results of the PFT were consistent with a mild restrictive process. The CT of the chest revealed a Haller index (see Discussion below for information on this index) of 3.90 (Fig. 2). It is of note that this CT was obtained approximately 11 months after the chest films. This adolescent/pediatric patient's deformity likely progressed over this time interval; this is also suggested by the symptoms that the patient reported to the pediatric cardiologist.

This noncontrast CT of the chest also suggested that the inward displacement of the sternum was compressing the right ventricle, as noted on the cardiologist's echocardiography report. The right ventricle is seen directly subjacent to the sternum.

At the time of this case report, the patient is approximately 3 weeks postoperative from the Nuss procedure. The pediatric cardiothoracic surgeon determined that she could benefit from the Nuss procedure for correction of pectus excavatum. It is of note that this evaluation took two years to reach the point of surgery, as it was complicated by other medical issues. During the two years of this evalua-

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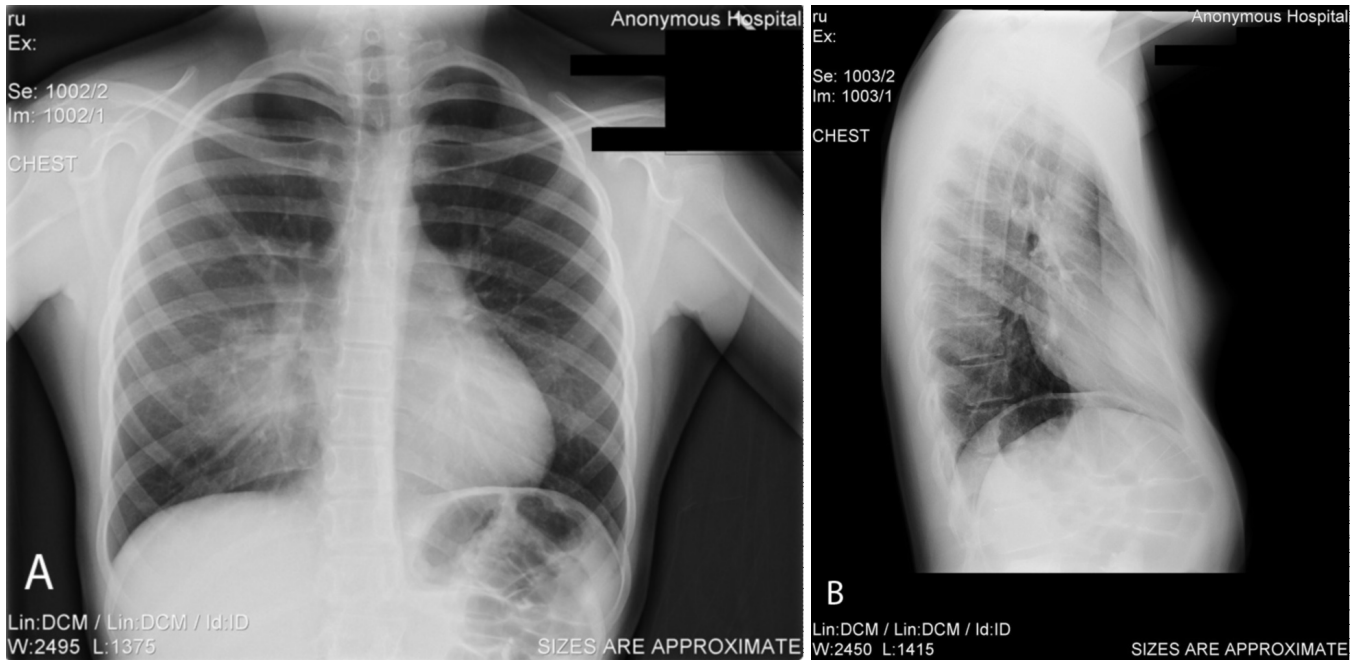


Figure 1. 13-year-old female with pectus excavatum. A. Posteroanterior radiograph. B. Lateral chest radiograph.

tion, the severity of the deformity likely increased and became symptomatic, although the patient was asymptomatic when first evaluated by Pediatrics.

Discussion

Pectus excavatum, the most common anterior-chest-wall deformity, is characterized by inward displacement of the sternum, creating a depression in the chest. It is found in one in 400 live births. It has a male predominance, with a ratio of 3:1 male to female (1). A genetic predisposition to pectus excavatum has never been defined, although there is a clear familial prevalence. Pectus excavatum often acts in an autosomal dominant pattern, but it also occurs randomly (2). It is most commonly associated with scoliosis (15%), and is also associated with Marfan syndrome, congenital heart disease (typically mitral prolapse), rickets, Poland's syndrome, and osteogenesis imperfecta (3). In this case, the patient does have scoliosis, which is suggested by the dextroconvex curvature of the thoracic spine in Fig. 1A. Cardiac evaluation did not reveal mitral valve prolapse or any other congenital cardiac defects. Physical exam did not reveal any Marfanoid characteristics.

Pectus excavatum is categorized according to severity (mild, moderate, or severe) (4). It is evaluated on a case-by-case basis to determine if it is severe enough to justify corrective surgery. Evaluation of pectus excavatum includes CT scan of the chest, PFT, and cardiology evaluation. The CT scan is used to ascertain the severity of the deformity, by determining the Haller index: a ratio of the measure of the transverse diameter of the chest, divided by the sagittal measure of the distance from the sternum to the vertebral

body. The Haller index should be obtained at the deepest point of the deformity. If the deepest point of the anterior chest wall is not in the same sagittal plane as the anterior aspect of the spine, then the Haller index should be measured using two parallel lines drawn tangentially to the ante-

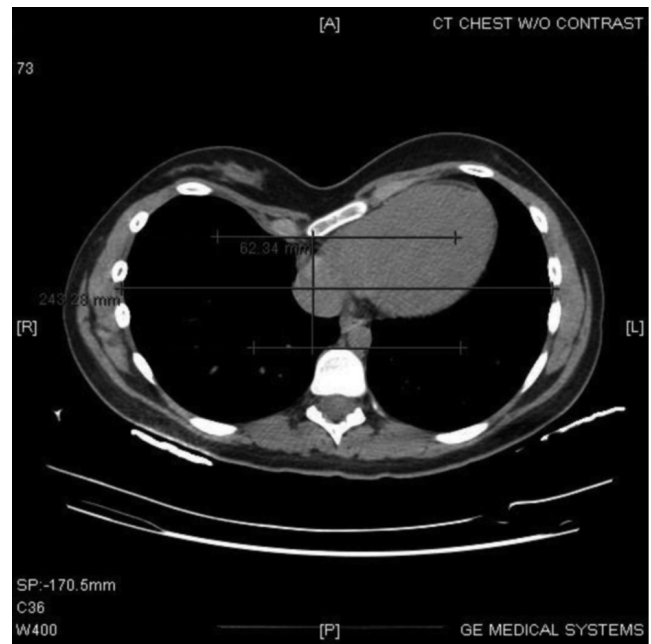


Figure 2. 13-year-old female with pectus excavatum. Non-contrast CT of the chest, with measurements for Haller index.

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rior angle of the spine and across the posterior aspect of the deepest part of the sternum (Fig. 2). The distance between these two lines is the sagittal measurement. A normal chest has a Haller index of 2 or less. A Haller index between 2 and 3.2 is considered a mild deformity; between 3.2 and 3.5, moderate.; 3.5 or greater, a severe deformity. Both moderate and severe deformities can be considered for corrective surgery (4). This patient had a Haller index of 3.90 (Fig. 2). The severity of this index, in the setting of PFTs consistent with restrictive pulmonary process, with an echocardiogram that reveals right ventricular impingement, in a patient with shortness of breath and decreased exercise tolerance—all combine to provide a strong case for corrective surgery.

Chuang et al. states that the use of a Haller index of 3.2 or greater as an indication for surgery is controversial, considering the lack of objective data to correlate the degree of deformity with physiologic dysfunction (5). However, a more recent study by Chu et al. does reveal positive correlations between severity of sternal deformity and cardiac rotation, as well as clinically significant symptoms (6).

Two corrective surgeries are available for pectus excavatum. The highly modified Ravitch technique involves a vertical incision in the midchest with resection of anterior cartilage, and placement of two stainless steel struts to support the sternum. The struts are wired to the ribs on both sides and removed after two years. The Nuss procedure, typically used on adolescent patients, uses two small incisions on either side of the chest to guide a curved steel bar (Lorenz pectus bar) into place. The bar pushes out the sternum from within and is fixed to the ribs on each side; it stays in place for at least two years. Complications from surgical repair could include pneumothorax, bleeding, pleural effusion, infection, displacement of hardware, and recurrence of pectus excavatum (7).

The PA findings in this case are characteristic of pectus excavatum; the heart is displaced to the left and there is an opacity where the right heart border would normally be found (Fig. 1A). This opacity is caused by the compression of soft tissues of the anterior chest wall, as well as the more vertical orientation of the anterior ribs where they articulate with the sternum (3). Thus it appears that the right heart border has been obscured by an opacity, which is the classic appearance of right-middle-lobe atelectasis or pneumonia. Actually, the right heart border has been displaced to the left, and the opacity is neither pneumonia nor atelectasis. Radiologists must be prepared for this appearance, and correlate findings with lateral images, to avoid mistakenly diagnosing right-middle-lobe pneumonias.

The radiologist, an important part of the evaluation of pectus excavatum, should be able to recommend any appropriate followup, including possible CT of the chest, and accurately measure a Haller index.

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