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Identifying pectus excavatum in the primary care setting

Abstract: This article reviews pectus excavatum and the role of the primary care provider in correct identification. Historically, pectus excavatum was viewed as a cosmetic concern. Research indicates that severe cases result in cardiopulmonary impairment and physiologic limitations. Evidence demonstrates that surgical repair improves cardiovascular function, exercise tolerance, and body image.

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Primarily care providers possess the unique role of being the first point of contact for patients with a variety of concerns, and chest deformities are no exception. Research indicates that severe cases result in cardiopulmonary impairment and physiologic limitations.¹ In the case of pectus excavatum, patients are often advised that the deformity will improve with age; surgical repair is dangerous, minimally effective, and unnecessary;

and that the malformation produces few symptoms and is primarily a cosmetic problem.² Scientific research, as well as patient testimony, has demonstrated that pectus excavatum is more than a cosmetic concern and can impact all facets of a patient's life. This article provides a brief overview of pectus excavatum, its classic clinical presentation, radiologic findings, and most common surgical interventions to help primary care providers

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diagnose and appropriately refer patients with pectus excavatum.

■ Epidemiology

Pectus excavatum is one of the most common chest wall deformities. It is estimated that pectus excavatum is present in 1 in 300 live births and more often affects males.³ Although the prevalence is significantly higher in Whites, there is no known racial predisposition.³ In 15% to 40% of cases, the patient has a close relative on either side of the family with the same deformity, indicating that there may be a genetic component. At this time, neither the mode of transmission nor the responsible gene is known.³

■ Etiology and pathophysiology

The etiology of pectus excavatum is uncertain; however, the deformity may be attributable to an overgrowth or disturbance of the costal cartilages.^{1,4} Additionally, some researchers theorize that the deformity may be the result of an intrinsic abnormality of the costochondral cartilage because there is a significant occurrence of pectus excavatum among patients with connective tissue disorders, such as Marfan syndrome, Ehlers-Danlos syndrome, and Sprengel deformity.^{1,5} Furthermore, abnormalities have been discovered within the cartilage following its surgical removal, including stress/strain deformation and abnormal collagen content.¹ Most often, pectus excavatum involves the lower sternum near the xiphoid and adjacent costal cartilages. The depressions were frequently described as either focal “cup” shaped depressions or broad, shallow “saucers.” However, an additional group exists,

progressively worsening symptoms and cardiopulmonary function as they age.¹

■ Historical perspectives

Throughout history, recognition of this chest wall deformity was not often reported or recorded. The earliest cases are from an excavated grave site in Hungary in which two pectus excavatum sternums were discovered and dated from the 10th to the 16th centuries.⁵ The earliest patient report was published in 1594 at which time fresh air, breathing exercises, aerobic activities, and lateral pressure were prescribed for the condition.⁵

Prior to the advances made by Dr. Mark M. Ravitch in the 1950s, surgical treatment of pectus excavatum carried a significant risk of death. Initial surgical attempts were made to entirely remove costal cartilage. One surgeon removed a section of the anterior chest wall after which the patient’s heart could be seen pulsating under the muscle flap. Over the next 60 years, other surgeons developed methods for repairing the deformity. In the 1990s, Nuss and associates began using a minimally invasive technique to install internal stainless steel braces to correct the depression.⁵ Both of these techniques have been modified over the past several years and are used in practice today.

■ Clinical presentation

The physical presentation and symptoms vary widely among individuals and in their degree of severity. Physically, pectus excavatum presents as a prominent but variable indentation of the lower sternum and cartilages, often creating an asymmetric appearance of the chest.³ Although the deformity may present in various configurations, the most common is a cup-shaped concavity.¹ As described by Koumbourlis, patients with pectus excavatum tend to be tall, thin, and have a “characteristic posture consisting of ‘slouching’ and tilting of the shoulders to the side of the sternal rotation” when standing.³ As a

result, their arms hang abnormally toward the front of the trunk as opposed to the side. Additionally, although these patients tend to be thin, they have a protuberant abdomen.³

Symptoms often develop in adolescence and adulthood as the skeletal system matures and depressions tend to worsen. One theory surmises that patients experience worsening symptoms as they age because their chest wall is less compliant as compared to being very compliant in infancy.¹ Patients commonly present with exercise intolerance, endurance issues, dyspnea, air hunger, and pain or heaviness in the chest.^{3,6} Decreasing exercise tolerance and endurance are the symptoms that most often prompt a patient to seek further



Pectus excavatum is present at birth, however, the depression becomes more severe during the adolescent growth spurt.

which presents with asymmetrical or mixed carinatum/excavatum deformities and often has a long trench (usually asymmetrical).⁵

Pectus excavatum is present at birth and is often recognized during childhood; however, a marked increase in severity of the depression often occurs during the adolescent growth spurt as the skeletal system is fully maturing.¹ When the body reaches adult height, the deformity usually stabilizes; however, in some individuals, the deformity can worsen, particularly if the patient develops other complications (for example, scoliosis).³ Many patients do not undergo corrective surgery in childhood, leading them to experience

evaluation.⁶ Anatomic abnormalities including decreased thoracic volume and cardiac compression are thought to explain these effects.¹

Patients often have normal pulmonary function test results; however, it is thought that the increased work of breathing due to a partially restricted chest wall accounts for the decrease in exercise tolerance and dyspnea.^{1,3} Furthermore, cardiac compression impedes the heart's ability to properly expand and contract, leading to a reduction in stroke volume and cardiac output.³ As a result, patients often experience fatigue and compensatory tachycardia.¹ Symptoms and presentation vary among patients; therefore, the primary care provider should be knowledgeable of the classic presentation.

■ Effect on body image

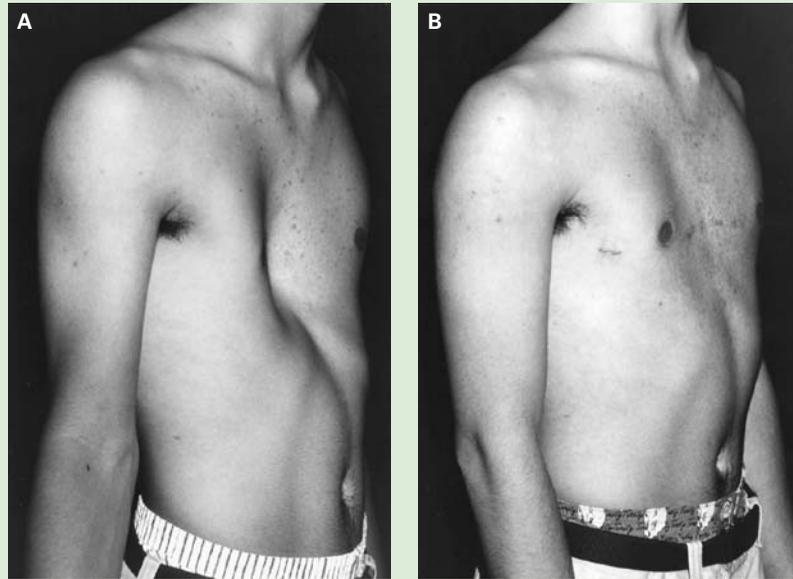
As with many other deformities, the impact on body image can be significant. Patients will often present not with physical symptoms, but rather those arising from a psychological cause. Children with pectus excavatum often have increased self-awareness, embarrassment, and shame, resulting in a lowered self-esteem, feelings of inferiority, depression, shyness, and social anxiety.^{3,7} The consequences of these emotions manifest in avoidance to participate in sports and other activities, which may impact their overall physical and social development.³ In many cases, the degree of deformity does not warrant surgical repair due to physical limitations, and thus, repair is considered cosmetic and not often covered by insurance.⁷ A study by Habelt et al. investigated the psychological distress felt by children with pectus excavatum and whether this could be a justified indication for treatment of the disorder.⁷ The authors concluded that psychological indication for treatment was justified.⁷

■ Imaging

In patients who present with concerning symptoms and physical characteristics, primary care providers may decide to order preliminary studies to further evaluate the chest wall; therefore, it is important that primary care providers have a general understanding of the various tests that may be requested by a surgeon for further evaluation. A stress test, pulmonary function test (PFT), lab studies (for example, complete blood cell count and comprehensive metabolic

Pectus excavatum preoperative and postsurgical repair

Photo A shows a 14-year-old boy with pectus excavatum before surgery. Photo B shows the boy postoperatively after his surgical repair of pectus excavatum using a retrosternal strut.



Source: Shamberger RC. Chest wall deformities. In: *General Thoracic Surgery*. 7th ed. Philadelphia, PA: Wolters Kluwer/Lippincott Williams & Wilkins; 2009:612.

panel), chest X-rays, computed tomography (CT) scan of the chest, electrocardiogram, and echocardiogram may be included following the suspicion of pectus excavatum to confirm the diagnosis.⁸ For preliminary evaluation of chest wall deformities, the primary care provider may obtain chest radiographs (chest X-ray and noncontrast CT scan). These studies are useful in determining if a deformity is present in the bony or cartilaginous skeleton, and the three-dimensional view of the CT scan can reveal cardiac compression or displacement.¹ The severity of the deformity is often quantified using the Haller index. This index is derived from dividing the widest transverse diameter of the chest by the shortest anterior-posterior diameter on a simple CT scan.^{9,10} Greater than or equal to 3.25 is generally considered the acceptable threshold for surgical correction.¹⁰

Additionally, PFTs are used to elicit the physiologic effect of pectus excavatum; however, patients often have normal results. An echocardiogram may demonstrate a mitral valve prolapse and compression of the right atrium and right ventricle by the sternum. Following this preliminary workup, surgical referral is indicated if the patient is symptomatic, has experienced progression in the deformity, and has paradoxical movement of the chest wall with deep inspiration, a Haller index of greater than 3.0 to 3.25, cardiac compression or displacement, pulmonary compression,

abnormal PFTs showing significant restrictive disease, mitral valve prolapse, and/or significant body image disturbance. Surgeons typically require that two of the above criteria are met for patients to be considered for surgical intervention.¹

■ Surgical intervention

Currently, most surgeons wait for patients to reach adolescence or the early teenage years before performing a repair. By allowing the patient to reach skeletal maturity; repair during this time has a lower chance of recurrence. For patients who meet criteria for surgical intervention, there are two methods most often used: the highly modified Ravitch repair and the Nuss procedure. The modified Ravitch repair is an open procedure in which incisions are made allowing exposure of the lower four ribs.¹¹ The surgeon then resects the deformed costal cartilage, and a stainless steel strut is placed posterior to the sternum to support the new chest contour.¹¹ This strut will be removed 6 months following the repair in an outpatient procedure.¹¹ Proponents of the modified Ravitch claim lower cost, shorter hospitalization, and less postoperative pain.¹ (See *Pectus excavatum preoperative and postsurgical repair*.)

The Nuss procedure is a minimally invasive procedure that involves placement of convex stainless steel bars through small, lateral, bilateral thoracic incisions.¹¹ The bar(s) is inserted from a lateral incision under the sternum and then forcibly rotated to elevate the ribs and sternum.¹¹ The procedure is usually performed using laparoscopic visualization to prevent cardiac perforation.¹¹ The bar(s) are typically removed after 3 to 5 years after the anterior chest wall remodels.¹¹ This procedure is gaining popularity for cosmetic reasons, the smaller skin incisions, and shorter operative time.¹

■ Rationale for repair

Despite numerous published reports, there is no consensus in the literature as to whether surgical repair improves cardiovascular function.² One study found that exercise tolerance is not improved with either type of repair.⁴ In addition, Koumbourlis concluded that the overall effect on cardiopulmonary function following repair was modest and probably clinically insignificant.³ In contrast, a meta-analysis performed by Malek et al. found that average cardiovascular function improved significantly (by greater than one half a standard deviation) following surgical repair of pectus excavatum.²

These results, along with patient testimonials, have led to the argument that surgical repair is indicated in symptomatic patients regardless of physiologic testing.¹ Furthermore, these findings contradict arguments that surgical repair is primar-

ily cosmetic and yields minimal physiologic improvement.² One of the most compelling arguments in favor of repair using either technique is that most adult patients report relief of symptoms after repair.¹² Additionally, resolution of mitral valve prolapsed and release of chest wall entrapment is seen in a majority of patients when comparing pre- and post-operative diagnostic studies.¹²

■ Best patient outcomes

As reviewed during this article, pectus excavatum impacts both physical and psychological aspects of patient's lives. Certainly, appropriate and timely diagnosis of pectus excavatum supports best care and treatment. By being knowledgeable of the physical characteristics, typical symptoms, and impact on body image, primary care providers can accurately diagnose pectus excavatum and refer for surgical evaluation when appropriate. Surgical intervention has demonstrated improvement in symptoms and body image in many patients. A strong understanding of the disease process allows primary care providers to best care for their patients and achieves best patient outcomes. **NP**

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