

# An overview of pectus deformities and rehabilitation approaches

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## ABSTRACT

Pectus deformities, particularly pectus excavatum and pectus carinatum, are among the most common congenital chest wall abnormalities in children. These deformities may have varying degrees of physical, psychological, and functional effects. This review provides a concise overview of the current state of knowledge regarding the history, epidemiology, etiopathogenesis, clinical presentation, classification, and rehabilitation of these deformities, focusing on the pediatric population.

**Keywords:** Deformity, exercise, pectus carinatum, pectus excavatum, rehabilitation.

Pectus, characterized by an irregularly shaped deformity of the anterior chest wall, is a common disorder and may present as an abnormal protrusion or depression of the sternum.<sup>[1]</sup> The prevalence of these deformities is estimated to be higher in males, at around one in 300 to 400 live births.<sup>[2]</sup> This condition has been observed to occur during childhood or early adolescence, when accelerated growth spurts lead to bone and cartilage development. Many additional problems, such as cardiopulmonary dysfunction, cosmetic problems, and psychological distress, occur in patients due to pectus deformities. The most common forms of pectus deformity are pectus excavatum (PE), also known as “shoemaker’s chest,” and pectus carinatum (PC), also known as “pigeon chest.”<sup>[1]</sup> However, there are also more complex “mixed” pectus deformities and other congenital or acquired chest wall abnormalities that occur together with these deformities. The historical process of pectus deformities dates back to ancient times, as evidenced by their depiction in medical texts and works of art. The first person to collect literature on PE was the physician Schenck von Grafenberg in the early 16<sup>th</sup> century.<sup>[3]</sup> The term “pectus excavatum” was first coined in the 19<sup>th</sup> century, while “pectus carinatum” was described later.<sup>[3]</sup> The earliest

information on chest wall deformities is found in Greek and Roman medical records.<sup>[3]</sup> However, these findings appeared to lack anatomical or pathological detail. Technological advances in radiology and thoracic surgery enabled better diagnosis and treatment.<sup>[4]</sup> Many studies have focused on the etiology and treatment of pectus deformities, including genetic, biomechanical, and psychosocial aspects and new developments in surgical techniques.

The etiology of PE and PC remains incompletely understood. The underlying pathology is thought to involve a disturbance in cartilage development. The prevailing contemporary hypothesis suggests that anterior or posterior sternal displacement occurs due to overgrowth of the lower costal cartilage and ribs, with the diaphragm playing a critical role in facilitating this displacement.<sup>[5]</sup> Although there is a lack of conclusive evidence supporting genetic transmission, a familial association has been reported.<sup>[6]</sup> Chest wall deformities can present in various forms, including symmetrical, asymmetrical, or combined configurations.

## CLASSIFICATION OF PECTUS DEFORMITIES

Pectus deformities are classified based on the direction and severity of sternal displacement.

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### 1. Pectus excavatum (Shoemaker's chest)

Pectus excavatum is the most prevalent anterior chest wall deformity, accounting for 90% of all congenital chest wall deformities. Pectus excavatum is characterized by an inward collapse of the sternum and costochondral cartilage structures (Figure 1). The prevalence of the condition is higher in males than in females, with a ratio of approximately five-to-one.<sup>[7,8]</sup> Pectus excavatum is often identifiable at birth; however, milder cases may remain undetected until the adolescent growth spurt, when the deformity becomes more pronounced. Additional anatomic characteristics associated with PE include sloped ribs, a protuberant abdomen (commonly referred to as a “pot belly”), and rounded shoulders. These features tend to become more evident as the child matures and undergoes further skeletal development.<sup>[8,9]</sup> The precise etiology remains to be elucidated; however, genetic factors, aberrant growth of costal cartilages, and connective tissue disorders (e.g., Marfan syndrome and Loeys-Dietz syndrome) have been identified as contributing factors. Pectus excavatum is a disorder arising from abnormal embryonic development. The underlying mechanism involves the improper fusion of the ribs with the sternum, a process that initiates around the first month of gestation, continues throughout

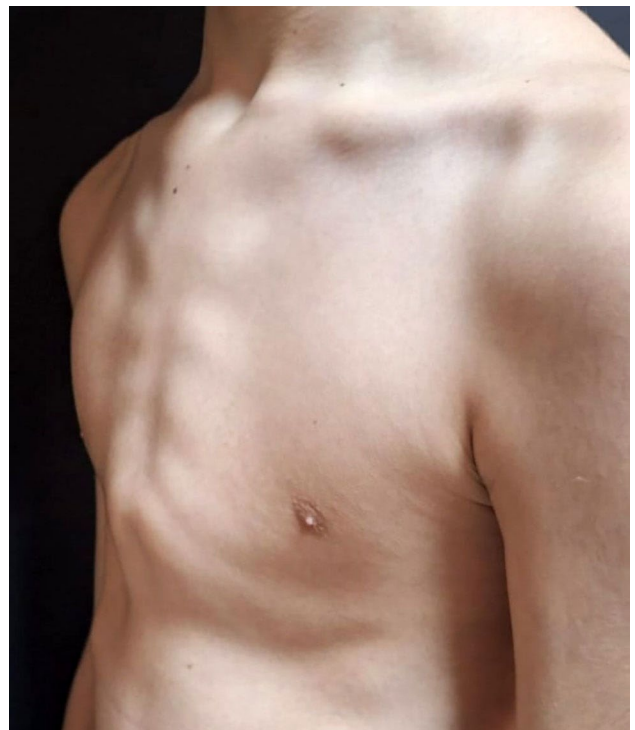
fetal development, and is ultimately completed by the final ossification of the chest wall during adolescence. Although the condition demonstrates a familial inheritance pattern, with approximately 40% of patients having a positive family history, no specific genetic aberration has been definitively identified as the direct cause of this deformity.<sup>[10]</sup> However, this finding is somewhat contradicted by the constant sex ratio of five-to-one in favor of males. This inconsistency may be attributed to the influence of multiple pectus disease-associated alleles.

### 2. Pectus carinatum (Pigeon chest)

Pectus carinatum is characterized by a protrusion of the sternum or rib cage (Figure 2).<sup>[11]</sup> When the sternal manubrium is prominently affected, the deformity is commonly termed “pigeon breast.” In contrast, the more frequent chondrogladiolar prominence, involving protrusion of the sternal body, is often described as “chicken breast.”<sup>[12]</sup> In contrast to PE, the diagnosis of PC is not usually made in infancy; rather, it is identified during the teenage years, a period characterized by accelerated growth. The condition can be observed as early as the 10<sup>th</sup> year of life, reaching its zenith at the age of 16 for females and 18 for males.<sup>[13]</sup> Pectus carinatum is often an asymptomatic condition. When symptoms



**Figure 1.** Pectus excavatum deformity.



**Figure 2.** Pectus carinatum deformity.

do occur, they primarily present as tenderness at the site of the protrusion. In symptomatic and more severe cases, additional symptoms due to decreased chest wall compliance include tachypnea, dyspnea on exertion, and decreased exercise tolerance.<sup>[14]</sup> The exact etiology of PC is unknown. However, it has been proven that a significant proportion of patients (25 to 33%) with a family history of chest wall deformities show a strong genetic predisposition.<sup>[13,14]</sup> In the diagnosis of PC, clinical examination is supported by detailed imaging techniques such as lateral chest radiographs or computed tomography (CT) scans. The radiographically calculated Haller index is used to determine the severity of the deformity. The Haller index is calculated from the ratio of the transverse diameter of the chest wall to the maximum anteroposterior diameter.<sup>[14]</sup>

### 3. Other rare types

#### *Pectus arcuatum*

This is characterized by a combination of excavatum and carinatum deformities, resulting in a “wave-like” appearance.<sup>[13]</sup>

#### *Poland syndrome*

A rare condition characterized by unilateral chest wall hypoplasia and associated hand abnormalities.<sup>[15]</sup>

#### *Jeune syndrome*

A severe form of chest wall deformity associated with skeletal dysplasia and respiratory compromise.<sup>[16]</sup>

#### *Rib flare*

In both PE and PC, as well as in cases of mixed deformities, the lower ribs may exhibit flaring, characterized by an upward pull of the ribs and the costal margin, either on one side (typically the left) or on both sides, resulting in protrusion. Frequently, rib flaring is perceived as the most prominent deformity associated with PE or PC deformities that are otherwise minor in nature. Conversely, rib flaring can manifest in isolation, devoid of any discernible additional abnormalities.<sup>[17]</sup>

#### *Pectus posture*

Pectus excavatum and PC have been associated with other skeletal problems and a “pectus syndrome,” defined as a cluster of symptoms and signs that occur together and characterize a condition.<sup>[17]</sup> This syndrome is often described by patients as

“poor posture” and is common, affecting one in three patients with pectus deformities. This condition is frequently disregarded by nonspecialist pectus healthcare professionals. The reasons why patients with pectus deformities develop this posture are unclear but are likely multifactorial. These include the effect on thoracic mobility, weakness of the shoulder and upper back muscles, and poor core strength.<sup>[18]</sup>

## IMAGING TECHNIQUES

Accurate and precise diagnosis of pectus deformities requires a comprehensive approach that includes clinical evaluation first, followed by imaging studies and functional evaluations. Only physical examination is sufficient for diagnosis in most cases.<sup>[19]</sup> However, depending on the accompanying clinical findings, further diagnostic tests may be necessary. Imaging modalities commonly used for diagnosis include chest radiographs, CT, and magnetic resonance imaging. Two-dimensional images of the chest wall are examined with posteroanterior and lateral chest radiographs, and the degree of sternal displacement can be demonstrated. While useful for initial screening, they provide limited anatomical detail. Computed tomography is widely used and accepted as the gold standard method in the evaluation of pectus deformities. This advanced imaging technique generates detailed three-dimensional reconstructions of the chest wall, allowing for precise calculation of the Haller index (a ratio used to quantify the severity of PE). It also facilitates the evaluation of cardiac or pulmonary compression due to the deformity. Magnetic resonance imaging serves as a viable alternative to CT by eliminating radiation exposure, particularly in pediatric patients. Magnetic resonance imaging provides high-resolution images of bone and soft tissue, providing detailed information.<sup>[20,21]</sup> In pectus deformity, in the context of functional evaluation, pulmonary function tests and cardiac evaluations are mandatory, specifically in patients with respiratory and cardiac complaints. Pulmonary function tests should be performed to understand the potential impact of pectus deformity on lung function and respiratory mechanics.<sup>[22]</sup> For detecting arrhythmia or conduction abnormality, an electrocardiogram should be done, and afterward, an echocardiogram is recommended to evaluate cardiac compression and overall cardiac function.<sup>[23]</sup> Cardiopulmonary exercise testing should be performed, which provides important information about functional limitations

and physiologic adaptations associated with pectus deformity during exertion and evaluates the patient's functional capacity.<sup>[24]</sup>

## REHABILITATION IN PECTUS DEFORMITIES

Rehabilitation in pectus deformities is gaining increasing importance. Rehabilitation is recommended for patients with mild to moderate deformities who do not require surgery and after chest deformity surgery. Rehabilitation principles are applied with a biopsychosocial approach. Rehabilitation of pectus deformities should be planned individually, taking into account the severity of the condition, the type of treatment chosen, and the general health status of the patient. The rehabilitation approach includes efforts to correct the deformity with orthosis, correct posture, increase lung capacity, provide cardiopulmonary fitness, regulate physical activity, provide self-confidence with psychological support, and eliminate anxiety.

### *Vacuum bell therapy in pectus excavatum*

Vacuum bell therapy (VBT) is applied to draw the sternum and soft tissue forward (Figure 3). The duration of treatment varies according to the patient's age, the flexibility of the chest wall, and the severity



**Figure 3.** Vacuum bell therapy in children with pectus excavatum.

of the deformity. Early treatment is crucial. There are quite variable protocols regarding treatment. It is initially started for 30 min twice a day and gradually increased over the next four to six weeks to 2 h per day. It is recommended to apply up to 8 to 12 h per day in the following period and to continue for at least one year. The duration of treatment can continue for 18 to 36 months depending on various factors.<sup>[25]</sup> According to a 15-year longitudinal study, treatment failure in PE was significantly associated with deeper and more severe sternal depressions, as well as increased chest wall flexibility. Consistent overnight use of VBT markedly improved outcomes. In this study, the success rate in overall VBT was stated as 41%. Females are more likely to initiate treatment earlier due to earlier onset of puberty and concerns about physical appearance. However, 39.3% of female patients were found to discontinue treatment prematurely following breast development. This may be attributed either to the fact that breast development can mask the appearance of PE, reducing the perceived need for further treatment, or to poor fit of the female model VBT after breast development begins.<sup>[26]</sup>

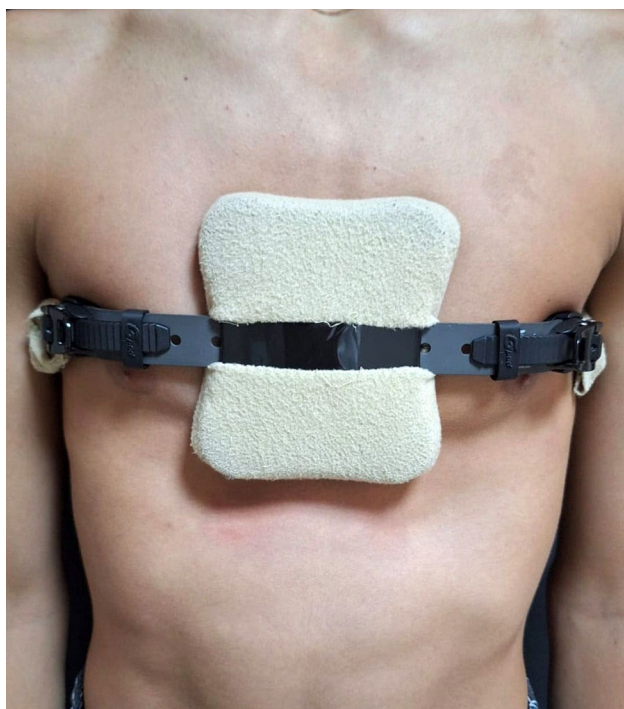
Vacuum bell therapy is associated with common but mild and transient complications. Common adverse effects include discomfort or moderate chest pain, petechiae, skin discoloration (9.7%), seroma (6.5 to 29%), and dorsalgia (6.5 to 50%), all of which resolved spontaneously. Adolescents and adults were more prone to transient subcutaneous hematoma and paresthesia than children under 10 years of age. Rare complications included skin thickening, costal flare, asymmetric PC, and rib fractures.<sup>[25]</sup>

### *Orthosis for pectus carinatum*

The orthosis approach began in the 1970s. Vidal et al.<sup>[27]</sup> applied cast vests and exercises to 52 patients after treatment with casts. In 2008, Martinez-Ferro et al.<sup>[28]</sup> introduced a dynamic compression system that became the prototype of dynamic orthoses. This orthosis aims to eliminate the carinatum deformity in patients with elastic sternocostal complexes.<sup>[29]</sup> Recently, there is increasing interest in the use of orthoses in PC.

Orthotics are an effective conservative treatment for patients with prominent PC deformity (Figure 4). Orthotics carry fewer risks and are less expensive than surgery.<sup>[30]</sup> The goal is to reshape the chest wall and to maintain the sternum in a less prominent position after the orthotics is





**Figure 4.** Orthosis in children with pectus carinatum.

removed. The orthotics are placed at the highest point of the chest to compress the sternum and reposition it in a more normal alignment. The success of this method depends on the severity of the condition and the patient's age, with younger patients generally having better outcomes due to more malleable bones. Older patients, whose bones are denser, may experience less effective results. According to most authors, the orthosis should be worn for at least 20 h per day.<sup>[31]</sup> The Calgary orthosis protocol is the most popular.<sup>[32]</sup> During the correction phase, it is recommended to wear the orthosis for 23 h per day, gradually increasing the duration. One hour is left for hygiene procedures and a series of exercises. During the maintenance phase, it is recommended to wear the orthosis for 8 h per day (mostly at night). Some authors recommend using the orthosis for 8 to 16 h per day during the correction phase.<sup>[31]</sup> Treatment usually takes 12 to 24 months, influenced by the severity of the deformity and the initial corrective pressure.<sup>[30]</sup>

The poor outcome rate of orthotic treatment for PC reaches 38%, mainly due to complications such as soft tissue damage, pain, ineffective correction, relapse, tachycardia, vasovagal episodes during orthosis application, and finger paresthesia.<sup>[31]</sup>

### **Posture**

The term “pectus posture” describes a distinctive posture in patients with pectus deformities, usually with their shoulders shifting forward and thoracic kyphosis (excessive bending of the upper back) developing.<sup>[17,18,33]</sup> This posture may be a compensation that patients develop consciously or spontaneously to hide the deformity in the chest wall. It is still unclear whether this posture is due to aesthetic concerns only or whether it is a postural disorder that develops directly due to the pectus deformity. In the study by Alaca et al.,<sup>[34]</sup> it was determined that patients with PE had worse posture according to the New York Posture Assessment scores. In addition, the rate of patients having at least one posture disorder was determined as 95% in PE and 92% in PC patients. The rate of thoracic kyphosis in PC patients was higher than in PE patients. These deformities affect not only the cervical and thoracic regions but also the entire spine. The study conducted by Mete et al.<sup>[17]</sup> revealed that adolescents with PE and PC experience reduced spinal mobility, alignment issues, and impaired spinal position sense. The findings emphasize the importance of assessing the spine during physical exams of adolescents with chest wall deformities.

Such postural changes can lead to musculoskeletal problems over time. Poor posture can cause the spine not to be properly supported. Even after the chest wall deformity is corrected with surgical intervention, it is often difficult to correct the pectus posture. Even if the deformity is corrected, patients may still continue with the wrong posture, which can negatively affect the healing process. Poor posture can become a habit over a long period of time. Therefore, it is not enough to focus only on the deformity itself in the treatment process for pectus deformity. The patients' posture disorders should also be an equally important issue and should be included in the treatment. Whether or not surgery is performed, postural exercises and physical therapy can help patients achieve better posture and maintain spinal health.<sup>[17,35,36]</sup>

### **Exercise**

There is no clear consensus on the role of exercise training. However, it is suggested that it makes a significant contribution to the prevention or correction of deformities, the reduction of postoperative pulmonary complications, the promotion of aesthetic improvement, and the improvement of the patient's quality of life.<sup>[37,38]</sup>

The aim of the exercises is to correct posture, increase the flexibility of the spine and chest wall, lengthen tight or shortened structures, strengthen the muscles, improve breathing, and increase endurance.

The first step is to activate and stretch the structures surrounding the pectoralis major, upper back, and chest wall. It is recommended that the exercises be performed in conjunction with deep breathing exercises.<sup>[37,39]</sup> The main posture exercises include “Wall Angels” and “Chin Tucks.”<sup>[40]</sup>

Breathing exercises are one of the cornerstones of pulmonary rehabilitation in PE. Breathing exercises vary depending on factors such as the frequency of breaths per minute, the pattern of chest and abdominal movements, and the duration of respiratory phases. The breathing exercises include diaphragmatic, upper and middle/lower lateral costal breathing, and breathing exercises involving each lung lobe. Patients with asymmetric PE are allowed to breathe to the depressed side and are prevented from breathing to the opposite side. Respiratory muscle training also provides benefits by increasing both respiratory muscle strength and endurance. Its aim is to improve respiratory muscle function and relieve dyspnea. It should be added to the exercise program for patients with respiratory muscle weakness.<sup>[38,41]</sup> The exercise program includes muscle strengthening exercises. The aim is to strengthen the chest, back, and trunk muscles to support the structural integrity of the chest wall. Strengthening exercises for PE can improve the overall appearance and function of the chest. Strengthening exercises can promote better posture, reduce the appearance of the deformity, and potentially alleviate some of the physical discomfort associated with the condition.<sup>[37,38,42]</sup> Strengthening exercises for PC can help develop the muscles around the chest, which can help reduce the prominence of the sternum. This can make the deformity less noticeable.<sup>[37]</sup>

Aerobic exercise plays an important role in managing pectus deformities by improving overall cardiovascular health and lung function. Aerobic exercises, such as walking, running, swimming, or cycling, can help improve lung function and increase oxygen uptake, thereby alleviating some of the breathing difficulties associated with the condition. These activities promote better circulation and increase overall endurance, which can make physical activities feel less strenuous and improve

daily functioning. Although aerobic exercise does not correct the deformity itself, it is valuable for supporting overall health, managing symptoms, and promoting long-term physical well-being.<sup>[37,38]</sup>

### ***Physical activity and lifestyle changes***

Regular physical activity is essential to maintaining a healthy lifestyle. Lifestyle changes, such as weight control and avoiding prolonged inactivity, are also important components of long-term rehabilitation.

### ***Psychological counseling***

Psychological support plays an important role in improving the mental and emotional quality of life of people with pectus deformity. Rehabilitation programs should include mental health support, including counseling and cognitive behavioral therapy, to address the emotional impact and improve overall quality of life. Psychological counseling can help patients cope with these emotional challenges by providing strategies for managing anxiety and developing a positive body image. Support groups where patients can share their experiences and connect with others who are facing similar challenges can also be helpful in reducing feelings of isolation. Counseling can also help guide patients in evaluating treatment options and reduce stress.<sup>[43,44]</sup>

In conclusion, patients with pectus deformity may have varying degrees of clinical complaints, including posture disorders, spinal pathologies, and decreased functional capacity. Rehabilitation programs aim to improve functional outcomes, alleviate symptoms, and enhance the patient's quality of life. The use of orthosis or VBT to eliminate the deformity or reduce its progression has recently received increasing interest in children with pectus deformities. Rehabilitation methods, particularly exercises, increase the success of the treatment. Rehabilitation program for pectus deformities should be individualized, taking into account the severity of the condition, the type of treatment chosen, and the patient's overall health.

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