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Pectus Excavatum: A review of current treatment possibilities

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Abstract

Anterior chest wall depression known as Pectus excavatum (PE) , is currently the most common congenital chest wall deformation, with incidence estimated at 1/400 births, five times more frequently in boys than girls (5:1 ratio in occurrence). ¹ Etiology of this deformity is formed by several hypothesis, including overgrowth of the ribs, developmental failure of the bony thorax or weakness and abnormal flexibility of the sternum. While theoretical concepts of its origin may differ, the common ground is the characteristics of said deformation: a posterior depression of the sternum and the lower costal cartilages. ² Surgical treatment options were described since 1949 with Ravitch procedure and since then a number of new ones has been developed.

For many years Ravitch and Nuss procedures were the main way to treat pectus excavatum, with a growing bias towards latter one. Since the Nuss procedure characterizes with less invasive approach and fewer overall complications ³ its popularity outgrew Ravitch procedure and currently is described as MIRPE – Minimally Invasive Pectus Excavatum Repair. ⁴ However afterward then many new techniques were developed, focusing on reducing overall procedure risks, improving patient safety and quality of life and including new technical possibilities.

The aim of this study was to gather and evaluate current literature to highlight new treatment options.

Keywords: MIRPE, Taulinoplasty, Magnetic mini-mover

1. Introduction

Epidemiology

Patients with musculoskeletal disorders are often submitted to primary care with chest wall deformities. The most common of them all is pectus excavatum (PE) characterized by posterior depression of the anterior chest wall resulting in a "funnel chest", with prevalence estimated at around 1/400 births. This malformation is more likely to occur in males than females with ratio of 5:1, however researchers have pointed that female incidence may be decreased because of the more developed breast tissue disguising the effect. ^{1,5}

1.1 Patient's characteristics

The defect usually becomes progressively more pronounced during pubertal growth spurt ⁶ and may sometimes be correlated with genetic disorders affecting accurate growth of the connective tissue, such as Marfan Syndrome, Ehlers-Danlos, Poland or MASS. ^{7,8} In the clinical environment children with PE usually display thin, tall postures with pot belly and forward-drifted shoulders that in some cases can lead to scoliosis. ⁹ Depression of the sternum can differ in some cases showing only cosmetical defect and in more severe instances displacing the heart with reduced lung volume. The inward deformation of the sternum can lead to compression on the right side of the heart and restrictive pulmonary deficits. The origin of pectus excavatum may be attributed to uneven overgrowth in the costochondral regions. Research indicates that individuals with asymmetric pectus excavatum exhibit shorter ribs on the more severely depressed side of the defect. ¹⁰

1.2 Evaluation

Diagnostic assessment of PE is usually executed using noncontrast computed tomography or magnetic resonance with the help of Haller Index (HI). This mathematical function describes relation between transverse and anteroposterior diameter of the chest wall, with its physiological score of 2.5 to 2.7. Severity of the deformation is then characterized by the increase of HI, with 3.25 often defined as the point at which the surgical treatment should be implemented. HI growth correlates with escalating heart displacement and pulmonary strain, reaching 30 in some of the most severe cases and sometimes showing substantial cardiopulmonary defects even at levels usually not qualifying for surgical treatment based

solely on the HI. Clinical evaluation is also greatly suitable in relation to HI, as in some cases lateral depression may show sternum surface being posterior to the anterior wall of the spine, which results in unmeasurable Haller Index results.¹¹⁻¹⁴ Additional diagnostic methods include electro- and echocardiography, cardiopulmonary exercise testing, blood chemistry analysis and sometimes pulmonary functions in order to evaluate severity of the patient's systemic strain caused by the chest malformation.¹⁵

2. Treatment

2.1 Noninvasive treatment

Important role in correct treatment of PE, often overlooked by physicians is the physical exercise, not to resolve problem but a mean to slow progression of mild and moderate malformations, correct patient's posture, prevent further complications and relapse after treatment.^{16(p315),17(p118),18(p270)}

There are two main treatment methods described in the literature that can be implemented instead of surgical treatment: vacuum bell and compression orthosis.

2.1.1 Vacuum bell (VB) Therapy

This method can be implemented in pediatric patients with mild and moderate deformities. Procedure consists of a bell-shaped suction device with pump regularly fixed every day, for extending periods of time during the therapy to anterior chest wall. Negative pressure created by removing air inside the device lifts the sternum reducing malformation. Upon excluding cardiac anomalies and other contraindications, patients can commence the daily application. The recommended regimen involves users applying the device twice daily for 30 minutes each, spanning the initial 4–6 weeks. As time progresses, the daily application duration of the VB varies widely among patients. Researchers state that the duration and frequency of daily application hinge on each patient's individual choice and motivation. Scheduled follow-up visits for patients occur at 3 to 6 monthly intervals, involving clinical examination, measurement of the depth of PE, and photo documentation. Electronic VB model enables the assessment of the correlation between applied negative pressure and the elevation of the anterior chest wall. The focus of clinical examination is on gauging the improvement in the depth of PE and monitoring for potential side effects, including persistent hematoma and/or

skin irritation. Future studies should assess whether initiating VB therapy before puberty proves more beneficial compared to starting during puberty or at a later stage. Even though this method on its own produces desired results in 20-30% of patients, combining VB with surgical approach like MIRPE produces excellent results in estimated 90% of patients. ¹⁹⁻²¹

2.1.2 Braces with exercise

Second method described by Brazilian researchers in 2021 is the use of wearable compression braces with some physical exercises. Patients qualified for this procedure shown mild, moderate, and severe states of malformation with mean age of 12.8 years. Promising results of 83% of patients showing good results after 24 months of treatment when their deformation was described as “flexible” and 62% with “poor flexibility” or “rigid”. However, patients using braces irregularly even with regular physical exercises displayed good results only in 38% of the cases. While complications were generally not significant, approximately 5% of the patients manifested skin rashes, superficial skin injuries, and transient hyperpigmentation in pressure areas. In 15% of cases, patients reported discomfort or transient pain in areas supported by braces, which was alleviated by partially releasing the pressure. Importantly, these complications did not lead to the discontinuation of device use. Among three patients, mild overcorrection occurred, and this was effectively managed by adjusting the device use time or introducing a second brace to address iatrogenic pectus carinatum. Notably, those who completed follow-up showed no instances of relapse. Encouraging results create a good alternative option to invasive surgery. ^{20,22,23}

3 Surgical approach

3.1 Surgery admission

Recommendations for invasive surgical approach aside of the cosmetic discomfort experienced by patient, include diagnostic imaging showing cardiopulmonary compression with HI of more than 3.25, correction score of 28% and more, evidence of cardiopulmonary disability and recurrence of malformation after previous surgery. Age recommendations include patients in their mid-adolescent years, which allows them to complete growth and lowers risk of recurring deformation. ^{15,24,25}

3.2 History of surgical treatment

Historically the first widely accepted way to surgically treat PE was described in 1949 by Ravitch ²⁶ and modified six years later by Rehbein and Wernicke ²⁷ – this open intervention procedure consists of deformed cartilages and xiphoid resection, sternal osteotomy with anterior fixation by the use of metal bars, thus correcting the depressed sternum and stabilizing chest wall for optimal recovery. Whole procedure could take up to 2 or 5 hours, leaving relatively sizable scar and in most cases requiring considerable number of wound drains. Initial hospitalization usually lasts 6 to 8 days with additional 2 or 3 days a year later, in order to remove metal bars used for stabilization. ⁹

Second method of surgical treatment for PE and to this day a gold standard in managing this deformation is the Nuss ²⁸ method first described in 1998 as the minimally invasive repair of pectus excavatum (MIRPE). This procedure involves insertion of a metal, U-shaped bar through a small incision on the side of chest, guided by a thoracoscopic control, rotated 180° after setting it in the right position, fixing the sides with stabilizers to lateral muscles of the chest wall. This operation lasts usually less than an hour, does not require ICU care and leaves patient with 2 or 3 considerably small scars. Initial hospitalization takes up 4-5 days with additional 2-3 days after 3-year period, in order to remove the bar. Wide implementation of this treatment option was found to be less invasive, shorter and preferred by the patients. ⁹

3.3 Comparison of Ravitch and Nuss method

These two methods to this day are the most popular choices for surgical treatment of PE, although MIRPE is more often chosen because of its less invasive character. However, as reviewed in many studies, the application of Ravitch method was favorable in the adult patient population, where overall complications and reoperations rates were lower. Also, when considering mixed populations, both pediatric and adult alike, Ravitch method showed lower rates of bar displacement. The last considerable difference between these two methods is the ability to change the anatomy of ribs, which comes helpful in cases of chest trauma to stabilize broken ribs, or in some cases when patients present rib malformations along the pectus excavatum i.e., as a result of vitamin D deficiency. ^{3,29,30}

3.4 Modifications to Ravitch and Nuss method

Since the 20th century both Ravitch's and Nuss's methods have been modified by several physicians. The most popular alteration to the Ravitch's approach are the Leonard procedure³¹ and the Robicsek method³². Leonard procedure utilizes sheathed wire placed behind the sternum fixed to an external brace. A curvilinear incision is made over the sternum, and following the mobilization of the pectoral muscles, the lower costal cartilage is excised, leaving the perichondrium in place. Subsequently, a wedge osteotomy is carried out. Instead of employing a bar, a wire is positioned behind the sternum and drawn up through separate stab incisions, securing it to an external brace. Patients are required to wear the brace for approximately 3 months.³³ Robicsek method relies on using a mesh hammock to secure the sternum. Bilateral resection of the defective costal cartilages and a transverse wedge sternotomy are performed. Following the mobilization of the sternum into the correct position, stabilization is achieved from behind using a Marlex mesh, which is securely affixed to the remaining cartilage.³⁴

When considering derivatives of MIRPE currently there are three main methods available. First one is MOVARPE technique (minor open videoendoscopically assisted repair of pectus excavatum) which consist of implantation of similar metal bar with accessory sternum osteotomy and multiple chondrotomies, that allows correction of particularly complex malformations with solid effect and high efficiency. The location and number of distorted ribs are individually chosen based on the shape and severity of the deformity. Employing the split muscle technique, rib cartilages are incised or partially resected to relax the chondrocostal arches. In cases with convex rib arches, a wedge resection is performed, while in concave ribs, a simple incision is adequate to correct the deformed cartilage curvature. In symmetric cases, the posterior side of the sternum is left uncut, but in asymmetric cases, complete transection is necessary to correct malrotation, alleviating the elevation of the deformed central thoracic wall unit. Using a bone hook, the sternum, and the anterior thoracic wall—now relaxed due to the multiple incisions—are elevated, resulting in the intentional greenstick fracture of the posterior sternum compacta. The operation then proceeds as a conventional MIRPE technique with video-assisted thoracoscopy, typically involving the implantation of a single pectus bar. Light activities are permitted after 3 weeks, and unrestricted sports activities are allowed 3 months post-surgery.³⁵

Second is the so-called Erlangen technique characterized by mobilizing sternum with retrosternal dissection through anterior incision, minimal cartilage resection and implantation of metal bar transternally amidst constant tension control provided by the use of tensiometer attached to the sternum. This way operators can precisely measure forces needed to elevate the chest. When the sternum is severely and rigidly depressed, coupled with a pronounced curvature, an osteotomy of its ventral cortex is carried out near the apex of the curve. Following the successful mobilization of the sternum, the Erlangen metal plate is transsternally positioned. The divided costal cartilages are individually reshaped, and the congruent sections are reconnected using robust absorbable sutures. In some instances, these costal cartilages may be shortened to attain congruence. After the placement of wound drains, the chest wall is closed. The patient is promptly transferred to a standard ward and is discharged from the hospital within 6 to 10 days, with plate removal scheduled after one year.

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Third one is Magnetic mini-mover procedure utilizing pulling forces generated by two magnets: one fixed to the sternum by a metal cradle and second placed in an external brace worn on the patient's chest for several months. The implant comprises a neodymium iron boron disc magnet (1 ½ cm diameter x 3/16 in. thick) and a ferromagnetic focusing plate encapsulated within a low-profile titanium shell. The back plate is linked to the anterior magnet through titanium cables that wrap around the sides of the sternum. The external brace is a custom orthotic crafted from polypropylene, molded to conform to each patient's anterior chest wall. It accommodates a second magnet, secured to the patient's chest wall by its attraction to the implanted magnet. The patient has the ability to adjust the force applied to the sternum using a screw mechanism, altering the distance between the brace and the implanted magnet. Promising results shown short operation time of around 30 minutes and initial hospitalization lasting one day. Although this method does utilize the use of metal bar, out of all minimally invasive surgical treatments mentioned is the least invasive.³⁷⁻⁴⁰

3.5 Newly developed procedures

Out of all described procedures there are two more methods disclosed in the literature with promising initial results: taulinoplasty and sternochondroplasty (SCP).

SCP is used to correct both pectus excavatum and carinatum. This procedure begins with resection of sternal cartilage and separating muscle bundles from sternum. After correcting of

the deformation with retrosternal tissue dissection, a metal bar is placed in order to support the sternum. Finally takes place the reconstruction of pectoral and abdominal muscles. Stabilizing bar is removed after 3-year period. Even though this method bares some similarity to the Nuss's procedure, initial results show that SCP provides shorter hospitalization, less overall complications, and longer post-operative epidural anesthesia. It's main downside to MIRPE is significantly longer surgery time averaging at 54.2 minutes in MIRPE and 229.5 minutes in SCP. ^{41,42}

Taulinoplasty is a method first described in 2016 and was designed to bypass the need for retrosternal involvement occurring in MIRPE. For this purpose, authors designed a special extrathoracic implantation and traction systems consisting of metal plate fixed with opening in the center and regulated screw with nut attached to the sternum. The screw is planted in the sternum at the lowest point of the defect and metal plate with its opening centering screw at its center. Then a traction system is mounted to the screw over external platform. After that traction system is tightened under thoracoscopic control lifting the sternum in result. Additional stabilization of the plate to the ribs can be done with few methods: screws, steel wires or Zip Fix ribbons. Initial results of this method can be described as encouraging in comparison with MIRPE showing shorter surgery time, hospitalization and analgesia time with similar efficiency and level of complications. ^{43,44} More time and result analysis is needed to correctly evaluate this methods efficiency.

4 Summary

Treatment evolution of pectus excavatum has been a subject of debate for many decades and has spurred many technically advanced ways to help all pediatric and adult patients struggling with this deformation. A development in both noninvasive and invasive techniques show dedication of researchers to maximally optimize the process with patients comfort in mind.

With the use of upcoming new materials, technology and involving new specialists to the cause the future can look promising for new, even more optimal ways to deal with PE as well as other pectoral malformations.

As we bring this overview to a close, it is evident that research on pectus excavatum is an unfolding narrative, continually evolving and adjusting to fresh insights. The importance of collaborative endeavors among diverse specialists persists as the key factor in discovering new possibilities for the treatment of PE. It is through these joint efforts that we will persist in

refining our comprehension, formulating efficient therapeutic approaches, and ultimately laying the groundwork for groundbreaking treatments to alleviate the impact of PE and other pectoral deformations on patients.

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