A Review of Current Treatment Trends in Pediatric Chest Wall Abnormalities

Abstract

Pediatric chest wall abnormalities have physiologic and psychological implications on pediatric patients. Current trends are for minimally invasive repair of these abnormalities to improve exercise function and cardiopulmonary reserve. This article provides an overview of current diagnosis, treatment and new nonsurgical management options for treatment of the most common pediatric chest wall abnormalities. Nonsurgical treatments as well as ancillary reconstructive surgical techniques provide a better cosmetic and functional result with improved tolerance to these procedures.

Keywords: Pediatric chest wall malformations; Pectus excavatum; Pectus carinatum; Nuss procedure; Jeune syndrome; Poland Syndrome

Introduction

Pediatric chest wall abnormalities are defined as any structural abnormality affecting the sternum, ribs, musculature, or thoracic spine, which alters the normal structure or limits the function of the thorax [1]. Of these deformities, pectus excavatum and carinatum are the most common sporadic malformations with both physiologic and psychological implications in patients affected. In some cases, pectus malformations can be associated with genetic syndromes and familial inheritance with the syndromic manifestations exhibiting cardiac and renal anomalies in association with the chest wall deformity [2]. Jeune’s Asphyxiating Thoracodystrophy Syndrome, deﬁant sternum, and musculoskeletal deformities after repair of large congenital diaphragmatic hernia are other chest wall abnormalities that can be more severe in their course and difficult to manage. These can be life threatening due to the inability of the thoracic cage to expand, result in exposure of the heart, or are associated with other cardiac malformations [3]. Poland Syndrome is a malformation of the ribs and chest wall musculature, associated with hypoplasia or aplasia of the breast or nipple and ipsilateral limb abnormalities, with correction and repair being more of a cosmetic issue than a functional problem, as there is no underlying pulmonary defect [3,4].

Due to the diversity and varying severity of chest wall abnormalities in pediatric patients, there is a spectrum of different management techniques to improve the cardiopulmonary reserve, exercise tolerance and function, as well as psychologic and self-esteem issues commonly associated with these conditions in children. In this review we examine some of the more common and severe chest wall abnormalities seen in the pediatric surgical practice and review the current trends in the minimally invasive management of these defects.

Chest wall Abnormalities

Sternum

Pectus excavatum is the inward displacement of sternum and costal cartilages, resulting in compression and leftward displacement of heart [3]. There is a depression of the anterior chest wall as a result of dorsal deviation of the sternum and the third to seventh rib or costal cartilage [2]. Incidence of excavatum is 0.1 to 0.8 per 100 persons, with a birth prevalence of 1/400, and a male preponderance [2,3]. Most commonly sporadic, though it may be associated with 32 different syndromes including Noonan Syndrome, Marfan syndrome, and Turner Syndrome [1,2]. Evaluation of these patients requires routine echocardiography to evaluate for cardiac compression, as well as static and dynamic pulmonary function testing. Additionally, genetic evaluation with thorough family history and anamnesis with work up is required in those with associated aortic root dilatation to evaluate for Marfan’s or other connective tissue syndromes [2]. In pectus excavatum, rotation and severity of the depression are the most important preoperative measurements as they are most likely to cause symptoms. Evaluation of the Haller Index (The ratio of the transverse diameter of the thorax to the shortest anteroposterior diameter between the sternum and vertebra) estimates the severity of the deformity (Figure 1a & 1b). A normal Haller Index is around 2.5 and values above 3.25 are considered to be severe.

Pectus carinatum is the second most common deformity of the anterior chest wall, defined by the outward displacement of the sternum or abnormal protrusion of the ribs [3]. As with excavatum, pectus carinatum has a male preponderance, however in preoperative evaluation of carinatum, only static and exercise pulmonary function testing are required [2]. Carinatum deformities may be further subdivided into keel chest, lateral...
pectus carinatum, and Pouter Pigeon Breast, with keel type deformity being the classical pectus carinatum, lateral pectus being a less common deformity represented by the outpouching of the lateral ribs and Pouter pigeon breast being the most severe [3]. Pouter Pigeon breast is represented by the protrusion of the manubriosternal junction and adjacent ribs with premature ossification of the sternum resulting in a z-shape deformity. This causes reduced compliance of the chest wall with impairment of respiratory movements. It is the only deformity on the pectus carinatum spectrum associated with cardiac abnormalities, most commonly ventricular septal defect [3]. Pouter pigeon breast is also associated with Noonan and Turner syndromes [2].

Cleft sternum are rare abnormalities that result from varying degrees of failed fusion of the two sternal halves. Classification of sternal defts is based on the length and character of the defect. There are several types, with the most common being a broad V shaped superior defect, with the most severe form resulting in total cleft sternum, associated with diastasis rectus and inferior cleft sternum which is part of Cantrell’s pentalogy [3]. Cleft sternum, or complete nonfusion of the sternal halves results in an exposed heart and requires early surgical correction. In this case, the sternal defect paradoxically deepens upon inspiration and bulges with expiration, with the pulsation of the heart discernable through the skin [3,5].

Ribs
Jeune Asphyxiating Thoracodystrophy is a rare autosomal recessive disorder effecting about 1:130,000 live births (Figure 2). It can be diagnosed by prenatal ultrasound demonstrating polyhydramnios, small thorax, short limbs and unidentifiable fetal respiratory movements. It is identified clinically by a severely narrowed thorax with the chest being narrowed in both transverse and vertical axes. In this form, the rigidity of the chest does not allow chest wall expansion for adequate alveolar ventilation, leading to ventilator dependence and respiratory failure [6]. Additionally, there is an acquired form of Jeune Syndrome that is known as acquired restrictive thoracic dystrophy that is a severe iatrogenic deformity of the thoracic wall following an open pectus excavatum repair in the preadolescent period. Acquired thoraco dystrophy has been postulated to be due to the overzealous resection of the deformed costal cartilages at the time of primary repair and devascularization of the growth ossification center of the sternum with cartilage regeneration, scarring and bone replacement leading to frozen thorax [7]. In both the congenital and acquired forms, surgical correction is necessary to allow for the chest wall tp expand during respirations.

Figure 1: A. Preoperative Chest CT of Pectus Excavatum with HI of 3.75. B. Postoperative CT demonstrating repair at level of heart. C. Postoperative AP and Lateral of Nuss bar.
Muscle

Poland Syndrome is a unilateral malformation of the ribs, chest wall musculature, overlying breast and nipple and ipsilateral upper extremity. Cause is unknown but most commonly thought to be a disruption of the subclavian artery blood supply with hypoplasia of the internal thoracic artery or reduction of blood flow at crucial periods that leads to absence of the pectorals major muscle, with hypoplasia of branches of the brachial artery causing the associated hand abnormalities. An alternate hypothesis suggests that the deformity is a result of disruption of the lateral plate mesoderm between days 16 and 28 of gestation [3,4]. Poland syndrome is associated with some malignancies, and treatment is based on degree of the thoracic deformity and defect present. Often cardiopulmonary impairment is limited, surgical correction improves the structural integrity of the rib cage and improvement in the chest appearance can be obtained by various musculocutaneous flaps [3]. Surgical repair is usually considered in the more severe variants with hypoplasia of ribs and cartilages, II through IV or II through V, potentially causing respiratory depression [4].

Musculoskeletal deformity after repair of large CDH

Patients that require repair of large congenital diaphragmatic hernias, usually require a patch or autologous tissue transfer. In these patients there is an increased incidence of chest wall deformities and scoliosis postoperatively as they grow. The rate of pectus deformity ranges from 14% to 80%. Children who have diaphragmatic defects that can be closed primarily are significantly less likely to develop chest wall deformities than those who have flap or patch repairs, however there is no significant difference in the rates between either flap or patch repair of the larger hernia [8].

Non Operative and Operative Treatment Options

There are both nonoperative and operative management options for the treatment of various chest wall abnormalities. The timing of these repairs differs depending on the defect present. Sternal clefts associated with ectopia cordis need to be addressed early to provide coverage for the heart using a muscle flap, which can be then more appropriately addressed later for full reconstruction. Cleft sternum should be primarily repaired at about 3 months of age, before the thoracic cage becomes rigid [6]. Pectus defects are typically repaired in adolescence around 14 years of age, as are those defects that are acquired after repair of large congenital diaphragmatic hernia or in acquired Jeune syndrome.

Non operative

Non operative management techniques involve using a force provided to the chest wall defect, over prolonged periods of time to exact change in formation of the pliable chest wall during growth. These are usually well tolerated treatment modalities but success is limited by patient compliance and requires close patient follow up.

Orthotic bracing is the first line therapy for pectus carinatum [5]. It is typically well tolerated and provides good aesthetic
results, however is limited by patient compliance and skin issues including folliculitis and sensitivity to the components of the brace. It involves using a brace to provide force in an anteroposterior direction to compress the chest and results in progressive remodeling of the chest [6]. Non-surgical treatment avoids operative intervention and its complications, is less costly and is scarless. A custom brace is created, using the patient’s measurements, and is worn for 23 hours per day during the correction phase which lasts about 4-8 months. The maintenance phase is an additional 4-8 months and involves wearing the brace for 8 hours per day, usually at night. Upon completion of bracing, patients noted significant improvement in the appearance of their chest with improved social interactions and self-esteem. Failure was related to noncompliance [9]. Bracing is found to be an effective first line treatment with surgical repair being an option if bracing fails.

Vacuum Bell or suction cup method, is a noninvasive technique to treat pectus excavatum. It uses the concept of applying a vacuum deepest aspect of the pectus defect on the chest wall [5]. A preliminary study on the efficacy of cup suction demonstrates its ability to provide aesthetically pleasing results. It uses gradual correction through application of a minimal force applied over a long period of time. The vacuum is generally used 3x a day for 45-60 minutes each for the first week, then overnight and then over the course of the day, with serial measurements being performed at follow up. Once complete correction is obtained, when the deepest point is <5 mm, the patient is kept wearing the device in a retainer mode [10].

The magnetic mini mover procedure is less invasive and similar to the suction method, in it it requires bracing and follow up in the treatment of pectus excavatum. This procedure uses a magnet implanted in the retrosternal space, and then a second magnet placed in an external brace, allowing the attraction of the magnets to cause a slow remodeling of the chest wall over several months [5].

Operative

Surgical correction of pectus excavatum is offered to those patients who are attempting to improve their exercise tolerance with surgical correction being recommended if the patient has compression abnormalities on CT or MRI showing a Haller index of 3.25 or greater, pulmonary abnormalities notably the demonstration of restrictive lung disease by pulmonary function test evaluation, cardiac compression, mitral valve prolapse or conduction abnormalities, and performance abnormalities including exercise intolerance or lack of endurance. Surgical correction is also recommended for recurrence after previous open or minimally invasive repair [5].

Pectus Deformities

Ravitch Type or open procedure which involves a transverse incision in the anterior chest, below the nipple line in the future intramammary fold followed by the elevation of flaps to expose the body of the sternum and affected costal cartilages, with dissection carried to the sternomamural joint. Pectoralis muscles are then dissected off and elevated to the level of the costochondral joints. Cartilages of the 3rd to 6th ribs are exposed, grasped, and divided from their insertion. Laterally a 1 cm rim of cartilage is left in continuation of the adjacent rib for further growth plate development. Sternal angulation is corrected by performing a transverse osteotomy and removing a wedge of bone at the anterior surface of the sternum. This is then stabilized with a strut in the retrosternal plane [6]. Recurrence after primary open repair ranges between 2-37% [11]. Outcomes are excellent, disadvantages mainly a centrally located large scar. Complication rates are around 15%, most commonly wound infection, pneumothorax and recurrence [6]. For pectus carinatum, the open surgical procedure consists of making a transverse sternal osteotomy with costal cartilage resection, in similar fashion to the classical Ravitch procedure [6].

Nuss Procedure is a minimally invasive technique that may be used safely in initial or recurrent pectus excavatum. After failed Nuss or failed Ravitch procedure without any anatomic issues, a modified Nuss is still preferred, with recurrence of deformity after Nuss of about 1.4% [11]. This involves using a videothoracoscope placed into right chest for visualization, and creation of a subcutaneous tunnel in the anterior chest wall at the level of maximum depression, by introducing a large clamp through small incisions in the midaxillary line bilaterally. Once the lateral ridge of the defect is reached, a Lorenz introducer is advanced from the right side to the left in the retrosternal plan and the sternum is elevated. Retrosternal dissection is performed under direct visualization to prevent injury to pericardium and pleura. Umbilical tapes are tied to the introducer tip and passed from side to side while removing the introducer. Finally, a pectus bar shaped to correct the chest anomaly is passed from side to side along the plane, guided by the umbilical tapes. Bar is placed with concavity facing forward and flipped when correctly positioned. Unilateral or bilateral stabilizing bars are placed at either end of the Nuss bar and attached to the chest wall with heavy sutures (Figures 1C). There is a significant postoperative analgesia requirement with a prolonged duration of pain. Epidural analgesia and Morphine/Fentanyl patient controlled analgesia have both found good acceptance amongst the patient population. Epidural catheter placement may warrant intensive care unit stay in some hospitals. Most patients can be transitioned to oral analgesics around 5-7 days after discharge. Complications following pectus correction are discussed elsewhere. Modification for pectus carinatum in a minimally invasive fashion similarly involves using a C-shaped bar inserted subcutaneous through small lateral incisions, placed in front of sternum at the point of maximum protrusion and fixed to ribs on either side [6].

Jeune’s Syndrome

Operative techniques for the Repair of Jeune’s Syndrome, both acquired and congenital have a main goal to expand the thoracic cage in order to release the entrapped lung, unload the mechanical pressures on the heart, and improve diaphragmatic excursion. The most important step of the procedure is to release and elevate the sternum [7]. The goal is to expand thoracic volume, leading to improved lung expansion. This is done through a variety of methods, most commonly a combination of open techniques as described below. Vertical Expandable Titanium Rib (VEPTR) procedure involves using vertically oriented titanium struts attached to the ribs or transverse processes of the spine and...
become progressively lengthened in a series of procedures [6]. This technique is being performed by a few centers with excellent results. Median sternotomy with graft placement to expand the thoracic cavity, using either methyl metacrylate struts or bone grafts can be performed, with reasonable widening of the thoracic cavity [3,6,12]. Centers that do not perform VEPTER usually use the median sternotomy and bone graft expansion technique. Lateral Thoracic expansion technique divides the ribs and underlying tissues in a staggered fashion, these are fixed with titanium plates to permit chest expansion in a gradual manner [6].

Ancillary Reconstructive Surgery

Using customized silicone implants to provide cosmetic repair is a less invasive option than the Nuss or Ravitch procedures. They can be used as primary repair or rescue therapy for recurrence after bar removal. This method involves making a copy of the chest wall defect by taking an impression using either laser scanning, 3-d photography or conventionally. A prototype is then created and a wax mold generated and fit to the defect where it is modified, flashed and an implant is made and then implanted subcutaneously [13]. This method cannot be used in those who have cardiac or respiratory issues in addition to the cosmetic deformity in pectus, as the implant does not modify the chest wall.

Complications

Postoperative and post treatment complications exist in patients who undergo surgical repair of their pectus deformity. Surgical complications in the open Ravitch-type and Nuss procedure include failure of correction, infection, post-operative pain, and the potentially lethal cardiac perforation. Modifying the Nuss procedure, using techniques to lift the sternum and thoracoscopic techniques to visualize retrosternal passage of the bar, has decreased the risk of cardiac perforation, but the risk is still present. Infection after the Nuss procedure is rare but bar salvage is possible with conservative measures in a great majority of the cases. This may involve removing the metal stabilizer and intravenous antibiotics to achieve high serum antibiotic concentrations followed by a prolonged oral antibiotic maintenance course that may be necessary for up to three months [14]. Following surgical repair with implantable metal bars in the minimally invasive repair of pectus excavatum, metal allergy to nickel and chromium have resulted. Allergy symptoms can be misdiagnosed as infection early on and can present as rash, erythema, granuloma, pleural effusion, or pericarditis, as well as, fever, elevated ESR, but have negative cultures. Allergy is a delayed type IV hypersensitivity reaction. Metal (Nickel) allergy typically occurs in about 2.2% of patients treated by minimally invasive pectus repair. Treatment of allergy consists of bar removal or exchange with titanium bars. Prevention of allergy is also important, a thorough history of allergy consists of bar removal or exchange with titanium bars. Prevention of allergy is also important, a thorough history of allergy should be elicited and then dermal allergy testing should be performed with the use of titanium bars if allergy found [15]. With all surgical repairs, postoperative pain is not insignificant. Chronic pain should be managed in consultation with the pain service. In rare instances pain may be due to allergy or subclinical infection and must be excluded. Complications of bracing are minor, and include folliculitis, failure of bracing, recurrence, and poor compliance with treatment. The judicious increase in pressure with dynamic compression braces and adjustment of compression by serial pressure monitoring, decreases incidence of skin breakdown when compared to standard bracing techniques. However dynamic compression braces, which often need to be imported, are more expensive than standard braces.

Cardiac and Pulmonary Function in Pectus deformities

Pectus deformities can cause right ventricular malfunction. Postoperatively with sternal elevation, there is modification of the right ventricular structure and function leading to a statistically significant change in the pectus index and right ventricular volume index. The elevation of a pectus deformity can increase right ventricular systolic, diastolic and stroke volumes, although no correlation between the degree of elevation and these changes has been found. In one study, 57% of patients had exercise limitations and 38% had EKG changes of right bundle branch block and right axis deviation, six months after surgery, only 7.1% reported exercise limitation, there were no changes in EKG abnormalities after repair. Pectus index in patients repaired before surgery was 4.19 and after the operation the mean decreased to 2.47. Right ventricular end diastolic volume, right ventricular end systolic volume and the stroke volume index all increased significantly after surgery [16].

Preoperatively there is a modest reduction of vital capacity (VC) and total lung capacity (TLC) that improves after surgical correction. Preoperatively, measures of dynamic pulmonary function (forced vital capacity (FVC); Forced Expiratory volume (FEV1); and VC) are below expected normal levels for patients of similar weight and size. Additionally, preoperative exercise tolerance in patients with pectus is lower than age matched controls, numbers that further deteriorated in the immediate post operative phase.

In a meta-analysis performed reviewing pulmonary functional recovery after the Nuss and Ravitch procedures, it was demonstrated that forced expiratory volume over 1s (FEV1), forced vital capacity (FVC), vital capacity (VC), and total lung capacity (TLC) were similar 1 year after the procedures, however changes in pulmonary function years after surgery and bar removal favored the Nuss procedure. In the post op period, FEV1 changes favored the Ravitch procedure, and this advantage persisted through the 1 year time point, however 3 years after treatment and bar removal FEV1 changes favored the Nuss procedure. FVC recovery improvement was greater after the Nuss procedure than the Ravitch procedure and improved after removal of bars after 3 years. As with FVC, VC initially favored the Ravitch procedure in the immediate postoperative period, however both increased significantly to favor the Nuss procedure at 3 years post surgery. TLC though was unchanged at 1 year was reported to be improved at 3 years with the Nuss procedure [17]. There is improvement in perceived exercise tolerance postoperatively, although PFTs may worsen, likely due to the combined cardiac and pulmonary changes. The immediate post operative decrease in pulmonary function improves in patients studied 12 and 18 months after surgery to the patient’s pre operative baseline, with some patients even improving beyond initial PFTs [18].
Discussion

The management of patients with chest wall abnormalities is complex and requires a multidisciplinary approach. The spectrum of chest wall abnormalities varies in severity and in clinical impact on cardiac compression and respiratory mechanics. The most lethal of these abnormalities, cleft sternum with resulting ectopia cordis, need to be managed in the first days of life, with less severe sternal defects requiring correction within the first 3 months of life prior to the hardening of the thorax.

Pectus type deformities and Poland syndrome, require correction in the adolescent years. There are several nonsurgical strategies as well as primary surgical and reconstructive surgical techniques that can be used to correct these deformities for optimal cardiopulmonary as well as aesthetic results. There is extensive literature and studies to support the continuously improving safety of minimally invasive procedures in the repair of these deformities. Of them, the use of a video thoracoscope for direct visualization, using various devices to elevate the sternum externally including the vacuum bell device, sternal hook and T-fasteners, [6,12,19-21], makes the procedure safer with less risk of damaging the heart. Additionally, use of more investigational magnetic mini mover or the suction device can be used as a first line for less severe pectus excavatum deformities prior to definitive surgical correction in case of failure may become more broadly accepted, as providing adequate aesthetic results, and improvement in the self-esteem of the patients [6,9]. First line treatment for pectus carinatum continues to be orthotic bracing, with failures of treatment being selected for surgical management [9]. Ancillary surgical techniques using specialized implants have further improved the cosmetic correction of these defects without any significant increase in morbidity, though provide no improved functional result [13].

New studies have documented the efficacy of the minimal invasive repairs for even recurrent pectus after previous open or minimally invasive procedure [11]. Results of repeat Nuss procedure are good, without increase in the most common complications, of pain and pneumothorax. However, a patient is not a candidate for repeat minimally invasive repair if the thoracic deformity is extensive as in acquired Jeune’s syndrome. Repair of these patients requires either complete anterior chest wall reconstruction or a modified Ravitch procedure [7,11].

In initial treatment of pectus deformities, it is imperative that correction be done in the adolescent years, with the avoidance of over resection of the deformed costal cartilages [7] as the early repair of these defects and overly aggressive surgery have been related to the later occurrence of acquired Jeune syndrome, which is more difficult to treat than a primary pectus deformity and requires open surgical repair rather than minimally invasive repair.

Treatment of patients with congenital or acquired chest wall deformities is consistently improving in its results and safety profile [15]. Newer non operative techniques combined with the current minimally invasive techniques allow for improved results and patient satisfaction with improved patient safety.

References

