Management of Congenital Chest Wall Deformities

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ABSTRACT

Congenital chest wall deformities are considered to be anomalies in chest wall growth. These can be categorized as either rib cage overgrowth or deformities related to inadequate growth (aplasia or dysplasia). Rib cage overgrowth leads to depression of the sternum (pectus excavatum) or protuberance of the sternum (pectus carinatum) and accounts for greater than 90% of congenital chest wall deformities. The remaining deformities are a result of inadequate growth. Evolution in the management of congenital chest wall deformities has made significant progress over the past 25 years. This article will review chest wall deformities and the current management strategies of these interesting anomalies.

KEYWORDS: Congenital, chest wall, deformity, pectus

A brief review of the complexities of thoracic cavity development will assist in understanding the pathophysiology of the deformities presented. Chest wall deformities occur when there is anomalous skeletal development and/or formation of the thoracic cavity.¹ The formation of the thoracic cavity occurs after the intra-embryonic cavity has formed (around the 4th week of gestation). There is rapid growth of the lung buds with concurrent formation of the diaphragm. Anatomically, a thick plate of mesodermal tissue, known as the septum transversum, occupies the space between the thoracic and abdominal cavities. Lateral to the septum transversum and foregut are the pericardioperitoneal canals. At the beginning of the 5th week of gestation, the lung buds rapidly grow caudally and laterally at the pericardioperitoneal canals inducing development of the thoracic wall and pleuropericardial membranes. During the 7th week of gestation, the pleuroperitoneal folds extend medially

and ventrally fusing with the mesentery of the esophagus and the septum transversum, thus dividing the body cavities by forming the diaphragm.² Coinciding with the development of the thoracic cavity, the sternum also develops during the 6th week of gestation. From a management standpoint, the complex development of the sternum can be simplified to involve three stages: formation, chondrification, and ossification. Formation begins with the condensed mesenchyme where paired parallel mesenchymal bands migrate from the lateral plates and fuse in the midline by the 10th week of gestation. During second stage, chondrification, the sternum forms in a band-like fashion and begins immediately once the sternal plates fuse. The last phase is ossification and begins in the manubrium of the sternum approximately during the 6th month of gestation. Ossification occurs in isolated centers starting in the manubrium, subsequently in the middle body during the 7th month,

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and finally in the lower body during the first postnatal year. The xiphoid process will undergo ossification between the 5th and 18th years of life, and the multiple ossification centers do not coalesce until after puberty.

PECTUS EXCAVATUM

Pectus excavatum (PE), or funnel chest, is the most common congenital abnormality of the chest wall. It is characterized by variable depression of the sternum and lower costal cartilages. Pectus anomalies can be evident early in life and are three times more common in boys than in girls.

Pathogenesis

Various theories have failed to demonstrate a cause for PE.³ Its association with scoliosis and Marfan syndrome suggests a possible link with connective tissue disorders. Studies show a pattern of inheritance as high as 37%.

Clinical Presentation

Some patients with PE suffer from limitations in physical activity but for the most part are asymptomatic. The body habitus of these patients is typically asthenic, and the thoracic depression ranges from mild and superficial to a more severe deformity in which the sternum almost meets the spine. In the latter case, the functional limitations may be evident. The posterior angulation of the sternum usually begins below the second costal cartilage. This curve is accompanied by anomalies in the adjacent costal cartilages and occasionally the ribs in older patients. The defect is often asymmetric, and the degree of depression of the sternum can be evaluated by computed axial tomography (CT scan) as proposed by Haller.⁴ The *Haller index* can be calculated from the ratio between the transverse and anteroposterior chest diameters. In normal individuals, the average Haller index is ~2.5, whereas in significant pectus deformity it is 3.25 or greater. Two recent studies validated the use of chest radiographs to calculate the depth of depression and concluded that CT is not necessary to calculate the index or determine the need for surgery.⁵

PE is present in approximately two thirds of patients with Marfan syndrome and is associated with scoliosis and congenital heart defects in 15% and 1.5% of patients, respectively. Often, pulmonary function tests and echocardiography are obtained to assess the patient's cardiopulmonary status preoperatively. According to multiple studies, surgery does not significantly change the patient's baseline lung function but can improve ejection fraction if cardiac compression is evident.⁶ We do not recommend the routine use of these studies in asymptomatic patients, but insurance companies often request these prior to approval for surgical correction.

Treatment

Surgical repair of PE is indicated when the deformity is severe and when the defect has a significant impact on the patient's self-esteem. We describe the two most frequently performed surgical repairs and briefly discuss other therapeutic techniques.

TRANSVERSE STERNAL OSTEOTOMY WITH SUBPERICHONDRIAL COSTAL CARTILAGE RESECTION (MODIFIED RAVITCH PROCEDURE)

This technique was described by Ravitch more than 60 years ago. In its original version, apart from dividing the intercostal vascular bundles, the resection of the affected costal cartilages included the corresponding perichondrium.⁷ It was later modified to perform a subperichondrial dissection, thus preserving the intercostal bundles and costal growth plates.

Description A transverse incision is made in the anterior chest below the nipple line. This incision should correspond with the future inframammary fold. Skin flaps are then elevated to expose the body of the sternum and affected costal cartilages. The dissection is carried to the level of sterno-manubrial joint. After optimal exposure, the pectoralis muscles are dissected off the cartilages and elevated bilaterally to the level of the costochondral joints. The perichondrium corresponding with the affected costal cartilages is opened sharply (Fig. 1). The cartilages of the 3rd, 4th, and 5th ribs are exposed, grasped, and divided sharply from their insertion into the sternum. Laterally, a 1-cm rim of cartilage is left in continuation to the adjacent rib for further growth plate development. The next step is the correction of the sternal angulation, which is achieved by performing a transverse osteotomy and by removing a wedge of bone at the anterior surface of the sternum. This is then reapproximated with heavy sutures, and some surgeons stabilize the sternum with a strut placed in a retrosternal plane. A closed drainage system is left in place, the pectoral muscle flaps are reapproximated in the midline, and the skin is closed in layers. Perioperative antibiotics are used, and adequate narcotic analgesia is provided. If the sternal strut is used, it can be removed at 6 months after the procedure.

Outcomes The results of the Ravitch procedure are excellent. Its major disadvantages compared with minimally invasive techniques are the presence of a centrally located relatively large scar and a longer operative time. Hecker et al showed a complication rate of up to 15%, the most common being wound infection, pneumothorax, and recurrence.⁸

MINIMALLY INVASIVE REPAIR OF PE (NUSS PROCEDURE)

The minimally invasive repair is now the preferred method of PE repair in children. It was introduced by

Figure 1 Modified Ravitch procedure for pectus excavatum. (a) Inframammary skin incision. (b) Subperichondrial dissection and exposure of costal cartilage. (c) Resection of 3rd, 4th, and 5th costal cartilages. (d) Appearance after repair.

Donald Nuss in 1988 and has undergone some modifications.

Description Under general anesthesia, the video thoracoscope is introduced in the right chest. A subcutaneous tunnel is created across the anterior chest wall at the level of maximum depression. This is accomplished 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 plane and the sternum is elevated. Retrosternal dissection is performed under direct thoracoscopic vision to avoid injury to the pericardium and pleura. Double 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. The bar is inserted with its concavity facing forward and is flipped over when correctly positioned (Fig. 2). This maneuver pushes up the depressed sternum against the downward force of the chest. To prevent bar displacement, unilateral or bilateral stabilizing bars are placed at either end of the Nuss bar and attached to the chest wall with heavy sutures. Reinforcement wires placed around the ribs have

also been proposed to prevent bar migration. Despite the less invasive approach, the analgesic requirement is significant, and the duration of postoperative chest pain may last longer than that with the Ravitch open procedure. The bar is usually removed after 2 or 3 years.

Outcomes Randomized studies have demonstrated the superiority of the Nuss over the Ravitch procedure. Its advantages are primarily cosmetic. Reported complications include wound infection, pneumothorax, bar displacement, allergy to the bar, and overcorrection leading to pectus carinatum.⁹ Bleeding from vessel erosion is rare. There is also the occasional verbal report of cardiac arrhythmia or cardiac injury with bar placement, and these potentially lethal complications are of significant concern. The results of the Nuss technique have not been as favorable in the adult population. In these patients, the chest is less flexible making them more susceptible to postoperative complications and pain.

MAGNETIC MINI MOVER PROCEDURE

This is a novel technique in development by Harrison et al in which a magnet is implanted in the retrosternal space of patients with PE. A second magnet is placed in

Figure 2 Nuss repair of pectus excavatum. (a) Severe pectus excavatum in a teenage girl. (b) Placement of pectus bar in the retrosternal plane. (c) Bar in place and ready to be flipped. (d) Final appearance.

an external brace. Both magnets attract each other causing a slow remodeling of the chest over several months.¹⁰

THE VACUUM BELL

Another device of interest, proposed by Haecker et al, is a suction cup applied to the chest wall to create a vacuum that sucks the deformity outward. When applied to the chest, the sternal lift is evident. The vacuum bell would need to be applied daily for adequate duration to ensure reformation of the chest anomaly. The device may also be used temporarily during the Nuss procedure to enlarge the retrosternal precardiac space when placing the bar in a severe deformity.¹¹

PECTUS CARINATUM

Pectus carinatum (PC) is the second most common congenital abnormality of the chest wall. It occurs in ~1 of every 2500 live births and is more common in males (4:1). The defect is usually identified in the early years of puberty. The body of the sternum and adjacent costal cartilages protrude, creating a defect similar to a pigeon's breast.¹ Although most defects are symmetrical, they can also be asymmetric and combined with PE. According to a classification based on the salient part of

the sternum, two variants can be identified: chondrogliadiolar and chondromanubrial. Other congenital anomalies associated with this defect are the sternal fusion defects, cardiac defects, and chondrocostal hypoplasia. Up to 26% of family members of children with PC may have the deformity.

Pathogenesis

No specific cause is identified, but there is an association with other skeletal abnormalities such as scoliosis suggesting that connective tissue disease in some cases may play a role in its pathogenesis.

Clinical Presentation

Most patients with this defect are asymptomatic. Patients with severe anomalies may complain of pain when lying in the prone position.

Treatment

NONOPERATIVE

Recent evidence suggests that nonoperative management with a brace that provides anteroposterior compression

results in progressive remodeling of the chest. The brace must be worn 14 hours a day for at least 2 years. Surgical correction is often reserved for those who fail conservative management or in noncompliant patients.¹²

OPERATIVE

The surgical treatment of PC is somewhat similar to that of PE. The Ravitch and Nuss techniques have been modified appropriately for the repair of a protruding defect rather than a depression.

Transverse Sternal Osteotomy with Costal Cartilage Resection

This technique is equivalent to the modified Ravitch procedure. A transverse incision on the anterior chest wall is made, and the pectoralis major muscle is mobilized to expose the affected costal cartilages. The perichondrium of the cartilages is opened, and the cartilages are excised sharply from the insertion to the sternum. Again, the costal growth plate is spared by leaving a rim of cartilage attached to the adjacent rib. Often, the sternal body and/or gladiolus have complex curves causing asymmetric defects. Multiple transverse osteotomies may be necessary to achieve the desired result. This includes the removal of a wedge of bone for realignment or sometimes interposition of cartilage (Fig. 3). After repair, a strut may be left in place for stability and can be removed after 6 months.

Minimally Invasive Repair of PC Similar to the Nuss procedure, a C-shaped bar is inserted subcutaneously through small lateral incisions. The bar is located at the point of maximum protrusion and, unlike the technique for PE, the bar is placed in front of the sternum and fixed in a compressing position to the ribs on either side.¹³

Outcomes Both surgical techniques provide equivalent surgical outcomes. The minimally invasive technique has an aesthetic advantage.

CONGENITAL DIAPHRAGMATIC HERNIA

Congenital diaphragmatic hernia (CDH) is a rare disorder that occurs in ~1 in 5000 live births. It is characterized by a defect in the diaphragm that leads to herniation of abdominal contents into the affected hemithorax. Compression caused by herniated viscera in the chest leads to pulmonary hypoplasia. CDH can be classified according to its location: anteromedial (Morgagni) when the defect is near the sternum and posterolateral (Bochdalek) when the defect is located at the junction between the costal and vertebral parts of the diaphragm (spino-costalis hiatus).

CDH is more common on the left side, and its origin is thought to be persistence of the pleuroperitoneal canal.¹⁴

Figure 3 Open repair of pectus carinatum. (a) Preoperative appearance. (b) Subperichondrial cartilage resection and correction of sternal angulation with cartilage interposition. (c) Postoperative appearance.

Clinical Presentation

CDH children often have severe physiologic impairment as a result of pulmonary hypoplasia and pulmonary hypertension. Although deceptively stable, the child's condition often deteriorates after 24 hours when homeostasis mechanisms are exhausted and ventilation requirements become critical and life threatening. Medical therapy for pulmonary hypertension is frequently required.

Treatment

The surgical treatment of CDH is a challenge. It is necessary to stabilize the patient first before correcting the defect.¹⁵ The timing of repair depends primarily on the successful recovery from the initial insult. Signs of improvement are minimum ventilation requirement, the resolution or stabilization of pulmonary hypertension, and stable global clinical status. Despite advances in respiratory therapy and protection against barotrauma with permissive hypercapnia, some patients require support with extracorporeal membrane oxygenation (ECMO). The criteria used to determine the need for ECMO are derived from clinical and physiologic parameters and remain somewhat controversial. When ECMO is used, most surgeons wait to repair the CDH after decannulation.¹⁶ The surgical steps to correct CDH consist of gentle reduction of the herniated

viscera, excision of the sac (when present), and repair of the diaphragmatic defect (Fig. 4). When the hernia defect is small, primary repair can be performed. With large defects, a tension-free repair should be done. The options include synthetic or bioprosthetic patch, reinforcement with prerenal fascia, and flaps with intercostal or abdominal muscle. Patch repair has gained popularity, especially when combined with minimally invasive techniques. Several studies compare laparoscopy or thoracoscopy with conventional surgery.¹⁷ The validity of these studies is limited as they include older children with delayed presentation and physiologically fit babies with absent or minimal pulmonary hypertension. Minimally invasive surgery should be used cautiously in the fragile neonate.

Outcomes

Recurrence after surgical correction of CDH is high, and dislodgement of the mesh and recurrence of the hernia is reported in as many as 46% of repairs.¹⁸ Although this seems high, it is even greater in the group with patch repair due in part to the associated defects of the abdominal wall musculature.¹⁹ Repair of recurrent CDH represents a challenge due to postoperative adhesions between the herniated viscera and the diaphragm or lung. The disruption can be approximated again if there is adequate diaphragm or repaired with mesh. The

Figure 4 Congenital diaphragmatic hernia. (a) Chest radiograph depicting a left-sided CDH (note the bowel gas pattern in the left hemithorax). (b) Right-sided CDH defect after reduction of herniated liver. (c) Pledgetted repair of left CDH defect.

authors repair hernias with or without mesh using pledgetted nonabsorbable mattress suture with satisfactory results. Techniques including intercostal and abdominal flaps are successfully used in patients with recurrent CDH.

POLAND'S SYNDROME

Poland's syndrome is a rare congenital anomaly in which there are varying degrees of hypoplasia or aplasia of the thoracic wall components.²⁰ It occurs in ~1 in 30,000 live births and is rarely bilateral. The only consistent anomaly in this syndrome is the underdevelopment or absence of the pectoralis muscles. Associated anomalies of the rib cage are present in up to 60% of cases, including hypoplasia or aplasia of the ribs and their cartilages. Other deformities include amastia, athelia, brachydactyly or syndactyly, and occasionally hypoplasia of the subclavian vessels.

Pathogenesis

The exact cause of the syndrome is not identified. Familial predisposition is described.

Clinical Presentation

The asymmetry is evident on the affected side. Hypoplasia of the pectoral muscles rarely causes functional limitation in the ipsilateral extremity, perhaps due to compensation by other muscle groups. Costochondral defects and absence of ribs can result in disfiguring anomalies. In addition, women may have amastia, athelia, or ectopia of the nipple.

Treatment

The surgical treatment of Poland's syndrome is largely cosmetic and is individualized according to the anomalies present. Surgery is rarely needed to improve the functional status of the patient. The concavity of the chest wall at the site of the defective pectoral muscle can be filled with muscle flaps or implants. One of the most popular flaps uses the latissimus dorsi muscle. In women with amastia, reconstruction should be performed when full development of the normal contralateral breast is completed.²¹ Techniques for repairing the costochondral anomalies in Poland's syndrome are adaptations to the Ravitch technique.²² When multiple ribs are hypoplastic or absent, contralateral costal grafts can be used. The periosteum of the donor rib is left behind to allow regeneration. The grafted ribs are then attached individually to the sternum with wires or heavy sutures. Wedge osteotomies are performed as needed to correct the abnormal curvature or rotations in the sternum.

Outcomes

With slight defects, the results are satisfactory. However, the treatment of severe or diffuse costochondral malformations remains a challenge.

STERNAL DEFECTS

Sternal defects are rare chest wall deformities that present with a broad spectrum of clinical severity from the potentially lethal ectopia cordis to the benign sternal cleft. Sternal defects are divided into four categories based on tissue coverage of the heart: (1) sternal cleft or bifid sternum, (2) thoracic ectopia cordis, (3) cervical ectopia cordis, and (4) thoracoabdominal ectopia cordis.²³

Sternal clefts/bifid sternums are malformations that occur as the sternal elements fail to fuse. The true incidence is unknown but may be around 0.15% of patients with chest wall deformities.²⁴ Sternal clefts are classified as complete or partial, with partial defects being either superior or inferior. Superior defects are the most common and are usually not associated with other anomalies. Sternal clefts may be "U"-shaped (proximal to the 4th cartilage) or "V"-shaped (reaching the xiphoid process). Inferior sternal clefts are usually associated with the pentalogy of Cantrell.

Clinical Presentation

Patients with sternal clefts may present as a spectrum from asymptomatic to recurrent respiratory infections and may have a dramatic appearance when the child cries or coughs. Surgery is advocated in even asymptomatic patients to protect the heart, allay parental anxiety, and decrease respiratory infections. Perioperative evaluation should include an echocardiogram, chest radiographs, and CT scan to completely evaluate the bony abnormality. If there are concerns for vascular anomalies, then a contrast CT scan is justified.

Management

Primary repair is performed at 3 months of age before the thoracic cage becomes rigid. Neonatal repair has also been advocated in the literature.¹⁸ A longitudinal incision is used that extends the length of the defect with the sternal bars lying below the subcutaneous tissue. Blunt mobilization of the posterior endothoracic fascia is performed to allow safe suture placement. The manner of approximation varies from primary suture closure, removing an inferior wedge of cartilage, or converting a partial sternal defect to a complete defect to allow suture approximation.¹ If at any time during repair the cardiac structures are at risk, primary repair should be abandoned for a more elaborate approach. The latter includes sliding or rotating chondrotomies or a pectoralis

major myoplasty. If sternal approximation is not possible with the chondrotomies, clavicular fracture lateral to the protuberant medial third has been advocated. When approximating the sternum primarily with one of the elaborate procedures, juxtaposition of the sternum with physiologic monitoring is recommended to verify the patient does not decompensate with the closure. If decompensation occurs, the remaining options include autologous tissue flaps, myocutaneous flaps, or prosthetic materials.²⁵ Autologous tissue repair is preferable to prosthetic materials because it usually allows sternal growth to occur in a normal fashion and is believed to offer a better cosmetic result.

ECTOPIA CORDIS AND PENTALOGY OF CANTRELL

Cantrell, Haller, and Ravitch in 1958 described a syndrome consisting of five embryologic defects: (1) ectopia cordis, (2) deficiency of the anterior diaphragm, (3) supraumbilical abdominal wall defect, (4) defect in lower

sternum, and (5) congenital intracardiac anomalies.²⁶ Other anomalies such as cleft palate and clubfeet were also described with the syndrome. The incidence of the syndrome is 5 per 1 million live births and is slightly more prevalent in males.

Pathogenesis

The pathogenesis of Cantrell's pentalogy is unknown. It is thought to be lack of migration of the lateral mesodermal folds and incomplete development of the septum transversum in the embryo.

Clinical Presentation

More than half the cases of Cantrell's pentalogy present with the complete syndrome. Ectopia cordis is present in 63% of cases and is called cervical, thoracic, or thoracoabdominal depending on the area of cardiac evisceration.²⁷ The most common intracardiac anomalies associated with the syndrome include ventricular septal

Figure 5 Staged repair of ectopia cordis. (a) Thoracoabdominal ectopia cordis (note the thin layer of skin covering the eviscerated heart and bowel). (b) Placement of tissue expanders. (c) Appearance of expanded skin after 6 weeks. (D) Soft tissue coverage of defect.

defect (VSD) atrial septal defect (ASD) and tetralogy of Fallot. The diagnosis can be made in the first trimester of pregnancy by ultrasonography. Recently, magnetic resonance imaging (MRI) has been used to delineate the anatomic details of the pentalogy.

Treatment

The treatment requires a multidisciplinary approach. When the diagnosis is made early in pregnancy, the physician should counsel the parents about the high mortality associated with the syndrome. Termination of pregnancy may be an option. There are several methods for correcting the different components of the syndrome. The main objective is to cover the eviscerated heart with skin, muscle flap, or synthetic material, without returning the heart to the thoracic cavity (Fig. 5). This is because these patients usually have reduced thoracic capacity.²⁷ In a later surgery, the osseous defect should be corrected. With the advent of pediatric cardiovascular surgery, the mortality of patients with intracardiac defects has been reduced substantially. Patients with the worst prognosis are those who present with complex heart defects and those with cervical or thoracic ectopia cordis.

JEUNE'S SYNDROME/ASPHYXIATING THORACIC DYSTROPHY

Jeune's syndrome, or familial asphyxiating thoracic dystrophy (ATD), is a rare autosomal recessive disorder effecting 1 per 100,000 to 130,000 live births. It was first described by Jeune in 1955 in a pair of siblings with severely narrow thoraces. A genetic locus has been identified on chromosome 15q13 as well as in the IFT80 gene encoding intraflagellar proteins in a subset of patients.²⁸

Clinical Presentation

Prenatal diagnosis by ultrasound is described in the literature that may allow for in utero consultation with surgical subspecialists. Key features in prenatal diagnosis are polyhydramnios, abnormal small thorax, short limbs, and unidentifiable fetal respiratory movements.²⁹ Clinical manifestations of this syndrome may include dwarfism with short ribs, short limbs, fixed elevated clavicles, and a high incidence of C-1 stenosis.¹ The clinical spectrum runs from a mild form where the chest may support adequate respirations to a severe form where the chest is narrowed in both transverse and vertical axis, with horizontal ribs and irregular costochondral junctions. In the severe form, the rigidity of the chest does not allow chest wall expansion for adequate alveolar ventilation leading to ventilator dependence and respiratory failure.¹

Management

A variety of techniques are described to treat ATD with the main goal of expanding the thoracic volume leading to improved lung expansion. Multiple procedures are described that involve a median sternotomy with placement of graft material to expand the thoracic cavity. Graft material has included methyl methacrylate and bone grafts. Staged procedures are also described for the treatment of ATD. Davis and colleagues have described a lateral thoracic expansion technique where the ribs and underlying tissue are divided in a staggered fashion and fixed with titanium plates to permit chest expansion in a gradual manner.³⁰

Vertical expandable titanium rib (VEPTR) procedure has been described by Campbell and colleagues in patients with Jeune's syndrome as a technique to improve chest wall movement. Vertically orientated titanium struts are attached to the ribs and/or transverse processes of the spine and progressively lengthened in a series of procedures. A majority of patients treated with VEPTR have thoracoabdominal scoliosis and long-term follow-up, and functional outcomes in patients with Jeune's syndrome are needed.³¹

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