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Chest Wall Deformities: An Overview on Classification and Surgical Options

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1. Introduction

Chest wall malformations (CWMs) represent a wide spectrum of anomalies, with a relatively high incidence and a significant impact on the life of patients. Besides a minority of cases with functional respiratory impairment and symptoms, the clinical importance of these anomalies derives primarily from the fact that the majority of children and their parents seek medical advice for psychosocial concerns, sometimes severe, usually due to poor cosmesis and aversion to sports and public exposure. Despite the relatively high incidence, of CWMs are often misdiagnosed or neglected by physicians, thus resulting in a significant delay or mistakes in the diagnostic work up or in the therapeutic management. In the last 12 years, however, since the introduction of the Nuss technique for pectus excavatum (PE) (Nuss et al., 1998), the interest of the scientific community about CWMs has dramatically increased, as well as the number of publications on this topic. A wide range of CWMs exist. Some malformations are very well defined and others are part of a wide spectrum of deformities. Confusion still exists in the literature about CWMs nomenclature and classification. A classification is of paramount importance because of the treatment implications. Other controversial issues are the treatment options: many different surgical techniques or other therapeutic alternatives have been proposed, especially in the last decade, so it can be difficult for a pediatrician or even a surgeon to advise correctly the patients about the possible correction techniques. In this chapter we will propose a simple classification, published few years ago by Acastello (Acastello, 2006), and modified by us, distinguishing CWMs into five types, according to the origin of the anomaly (Table 1). Following this classification, we will go through each of the most important CWMs, with the aim of reviewing and updating this topic, focusing particularly on treatment options.

2. Classification of CWMs

Acastello (Acastello, 2006) classified CWMs in 5 types, depending on the site of origin of the anomaly (cartilaginous, costal, chondro-costal, sternal, clavicle-scapular). We will follow hereafter his classification, with minor modifications regarding PC classification, as explained later in the text (table 1).

2.1 Type I: Cartilaginous anomalies

2.1.1 Pectus excavatum (PE)

PE is the most frequent thoracic malformation, with an incidence of 1/100 to 1/1000 live births, and accounting for around the 90% of all CWMs (Fokin et al., 2009; Lopushinsky & Fecteau, 2008). It is characterized by the presence of a variably deep sternal depression associated to a malformation of the lowest condrosternal joints. Usually it is congenital, but in some cases (around 15%) it appears later during development. In the latter there is a frequent association with malformations of the muscular connective tissue, such as Marfan and Ehlers-Danlos syndrome(Colombani, 2009; Fokin et al., 2009; Kelly, 2008; Kotzot & Schwabegger, 2009; Lopushinsky & Fecteau, 2008). The etiology of PE is not clear, and many hypotheses have been proposed (Kelly, 2008). The role of vitamins or other nutrients deficiencies is probably not influent at all, while a connective tissue disorder and genetic predisposition could play a role. PE shows a familial recurrence in up to 40% of the cases (Kelly, 2008), more rarely we can observe the presence in a PE family of other CWMs such as PC. A study on genetics of PE showed that the most frequent transmission pattern seems to be the autosomal dominant, but there are families with autosomal recessive and X-linked patterns (Creswick et al, 2006). The overgrowth of costal cartilages could be the pathogenetic mechanism leading to the development of PE (Fokin et al., 2009; Haje et al, 1999; Kelly, 2008). Collagen type II disorders have been demonstrated in the costal cartilages in PE (Feng et al, 2001), as well as overexpression or downregulations of some genes playing a role in the metabolism of cartilage and connective tissues, as collagen genes, matrix metalloproteinases, tumor necrosis factor-alpha, and filamin (Fokin et al., 2009).

Type I: cartilagineous	Pectus excavatum (PE)	
	Pectus carinatum (PC)	
Type II: costal	type 1	} agenesis, hypoplasia, sovrannumerary, bifid, fused, dysmorphic, rare (always complexes)
	True PC type 2	
	Simple (1 or 2 ribs)	} Jeune, Jarcho-Levin, Cerebrocostomandibular, others
	Complex (3 or more ribs)	
Type III: condro-costal	Syndromic (always complex)	
	Poland Syndrome	
Type IV: sternal	Sternal cleft (with or without ectopia cordis)	
	Currarino Silverman Syndrome	
Type V: clavicle-scapular	Clavicular	Simple or Syndromic
	Scapular	Simple or Syndromic
	Combined	

Table 1. Acastello classification of CWMS according to the site of the defect, modified by us. In the original classification, PC type Currarino Silverman Syndrome was called as superior or type 2 and included in cartilagineous anomalies. We have distinguished superior or type 2 PC into two different anomalies: the true PC type 2, very rare, and the Currarino Silverman syndrome, that we moved into sternal anomalies (see text).

PE patients have often a typical aspect: they are slim and tall, with some degree of joint laxity, rounded shoulders with a kyphotic habit and a “pot belly” (Colombani, 2009). The association with scoliosis or kyphoscoliosis, reported in about 15%-50% of cases (Frick, 2000; Waters et al, 1989), sometimes represents a matter of concern for the patients and families, however spine deformities almost never represent a serious clinical problem and usually do not require any treatment (Waters et al, 1989). Cardiac anomalies are only seldom associated with PE, but mitral valve prolapse is frequent (Kotzot & Schwabegger, 2009). In our experience we observe PE in some neonates with diaphragmatic hernia or children with respiratory obstruction (mostly for hypertrophic tonsils). These particular types of PE are the only ones that can ameliorate significantly or disappear during infancy (Kelly, 2008). In all other cases PE is usually mild at birth but over the years it can become severe, progressing especially during pre - adolescent and adolescent age. When the deformity is very pronounced, patients may manifest dyspnea at exertion, lack of stamina, palpitations and thoracic pain or discomfort (Acastello, 2006; Colombani, 2009; Fokin et al., 2009; Kelly, 2008; Lopushinsky & Fecteau, 2008; Nuss et al., 1998; Williams & Crabbe, 2003). In cases of severe malformations there can be physiological repercussions. Many studies have tried to elucidate the implications of PE on the respiratory and cardiac function (Colombani, 2009; Kelly, 2007, 2008; Williams & Crabbe, 2003). Sternal depression causes a leftward displacement of the heart. In some patients, it is possible to find compression of the right ventricle or atrium with different degrees of dysfunction on the echocardiogram. Inferior cava vein can also be compressed. Lung functional tests can report some degree of dysfunction, more on stress conditions than on rest. Usually the most common pattern of PE patients is a restrictive one, but also obstructive or mixed patterns are not uncommon, while asthma induced by the exercise is rare. The aesthetic consequences greatly affect the self esteem and self image of most of the patients. They are usually extremely shy, withdrawn and not practicing any activity that may imply exposing their chests. Usually they do not go to the swimming pool or beach and they tend to isolate themselves. The thoracic deformity predominates over other physical alterations.

2.1.1.1 Diagnostic assessment and classification

As PE includes a large spectrum of anomalies with different degrees of gravity, it is important to assess the severity in order to select the best treatment. Different indexes have been proposed for this purpose. While we are allowed to calculate some of them during simple patient evaluation, measuring the depth of the excavation by a caliper (Colombani, 2009) or pulvimeter (Fokin et al., 2009), Computerized Tomography (CT) scan is necessary to calculate Haller index (Haller et al., 1987), the most widely accepted one, based on the division between lateral and antero-posterior thoracic diameters. It is recognized that Haller index higher than 3 or 3.25 indicates surgical correction. Another important feature to be considered is if PE is symmetric or asymmetric. The latter, usually more depressed on the right side due to a variable degree of sternal rotation, can be an important factor influencing the final result. In females with asymmetric PE the sternum is usually rotated towards the right side and the right breast is apparently hypoplastic, mimicking a PS and possibly creating some diagnostic difficulties for physicians without large experience in CWMs.

The shape of PE, extremely variable from case to case, is crucial in determining the type of surgical approach and the prognosis of the correction. It is possible to classify morphologically PE as follows (Fokin et al., 2009; Kelly, 2008; Nuss, 2008):

- Grand Canyon (figure 1): It is a severe and deep PE. There is a deep long canal in the sternum. These cases can be corrected with the retrosternal bar, but the correction is

extremely difficult, especially when thorax is largely ossified and sternum is extremely rotated (figure 1). A higher complication rate after correction is reported, compared with the other types. In these cases, modified open procedures can be a valid option for correction.

- Punch or cup shape (figure 1): PE is extremely localized, usually on the inferior part of the sternum. It is more often symmetric. This type of PE can be very difficult to correct at any age and sometimes the outcome is partial.
- Saucer type: It can be symmetric or asymmetric. It is an extensive depression, the thorax is usually quite flat and the deep area is along the complete anterior chest.
- Transversal PE (figure 1): The depression is transversal and below the sternum.
- Eccentric PE: The sternal depression is eccentric to midline. It is the highest degree of asymmetric PE.
- PE with flaring chest (figure 1): The main feature of this type of PE (but sometime this is an isolated malformation, without associated PE) is the flaring chest at the level of the last ribs.
- PE-PC: it is a combined malformation with a sunken chest and cartilage protrusion beside the sternum edge.
- Superior PE: this is a very rare PE, localized in the upper part of sternum and cartilage ribs. Lower sternum is normal.

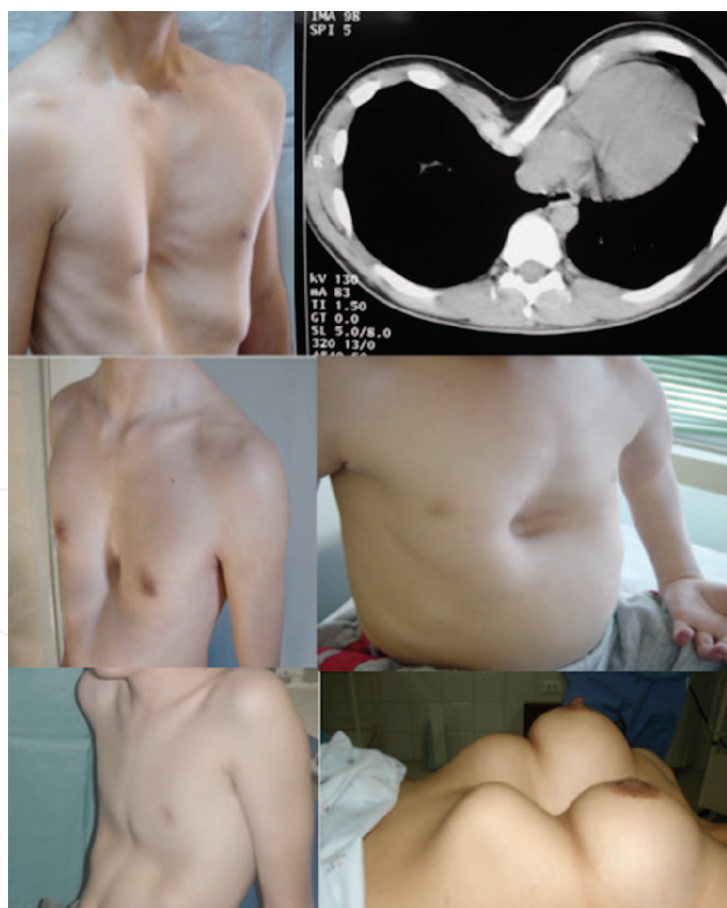


Fig. 1. *First row: Grand Canyon shape PE (left), Computerized Tomography of the thorax showing Grand Canyon PE with sternal rotation (right); Second row: Punch-shaped PE; Third row: Transversal PE (left), PE with flaring chest (right)*

2.1.1.2 Treatment options

Patients are selected for surgical correction if they demonstrate two or more of the following criteria (Kelly, 2008):

- symptoms;
- history of progression of the deformity;
- paradoxical movement of the chest wall with deep inspiration;
- a chest CT scan with Haller index greater than 3.25;
- cardiac compression;
- cava vein or pulmonary compression identified;
- abnormal pulmonary function studies showing significant restrictive disease;
- cardiac pathology secondary to the compression of the heart;
- history of failed previous repair;
- significant body image disturbance.

The ideal age for correction is a matter of debate (Lopushinsky & Fecteau, 2008; Nuss, 2008). Both open and Nuss procedures are feasible with good results in adult age (Aronson et al., 2007), however it is widely accepted that surgical correction has to be preferably performed in young patients before complete ossification of the thorax, that makes it harder and can jeopardize the final result. Fixing PE in the first years of life is probably unnecessary, and it could carry the risk of relapse (Nuss, 2008) or post-operative severe complications as acquired Jeune syndrome, according to the different techniques (Haller, 1996). A good age for correction with Nuss technique is usually considered from the age of 9 to 15 years of life (Nuss, 2008).

The first description of a PE repair was in 1911 from Meyer (Meyer, 1922). For many years the corrective procedures followed the principle introduced by Sauerbruch (Sauerbruch, 1931) and consisted mainly in resection of costal cartilages and mobilization of the sternum, sometimes with fracture of its anterior body. During the 20's, the resection was accompanied by external sternal traction which lasted for weeks. In 1939 Brown (Brown, 1939) recommended the resection of the ligament between sternum and diaphragm. Later, techniques were modified and standardized. A particular place has to be assigned to the pioneer of the modern era, Ravitch, who described his procedure in 1949 (Ravitch, 1949), consisting in the resection of all deformed costal cartilages without external traction. The modification proposed by Welch (Welch, 1958) preserved the perichondrium in order to facilitate rib regeneration. Another key point was the fixation of the sternum, in order to reduce flail chest and recurrence. It was performed initially with a bone graft (Dorner et al., 1950), later with a steel bar passed posterior to the sternum (Adkins & Blades, 1961). Many means of sternal fixation have been proposed during the following years, some of them absorbable. A totally new concept was introduced by Wada in 1970 (Wada et al., 1970) who described sternal turnover, in which the sternum was completely detached and removed, rotated by 180 degrees, then sutured back to the ribs. Another approach, attempted in mild cases, and proposed for the first time in 1972 by Standford (Stanford et al., 1972), was performed by filling the concavity of PE with some prosthetic material, as Silastic® or other subsequent modifications, as using omental flap (Grappolini, 2008). A revolutionary new technique was proposed by Nuss in 1997, and published after one year (Nuss et al., 1998), consisting of implanting a retrosternal metallic bar which is bent and rotated in 180° in order to obtain an immediate correction of the deformity. This bar is inserted through small lateral incisions and neither costal resections nor sternotomy are required. In Nuss procedure both the approach and the principle were completely new. In fact, although the result of the

correction is obtained immediately during operation, the efficacy of Nuss procedure in long term is based on the principle of thoracic cage remodeling under the action of the force determined by the retrosternal bar. For this reason the bar should remain for at least 3 years and then is removed through an outpatient procedure. This bar is fixed to the chest wall muscles and is stabilized via a lateral device that avoids slippage. The technique is simple but requires extreme care and experience. Initially the retrosternal tunnel for the bar was completed blindly, later thoracoscopy was introduced. Thoracoscopy is absolutely necessary, since the bar must pass very close to noble structures such as the heart which is sometimes very attached to the sternum. Other, less adopted, conservative procedures have been described, based on a suction device (Vacuum Bell) (Schier et al., 2005) or magnetic forces (Harrison et al., 2007), and proposed as attempts to correct PE without any surgical maneuver, but results still need to be proved.

Results in all series (Acastello, 2006; Kelly et al., 2007; Lopushinsky & Fecteau, 2008; Nuss, 2008) are usually good in more than 80-90% of cases, depending on the gravity, type of PE and age of correction. The largest experience of 1215 patients is reported by Nuss and colleagues (Kelly et al., 2010), who report a 95.8% surgeon's satisfaction rate, 93% patient's satisfaction rate and a 92% parent's satisfaction rate. It remains to be defined which technique between Nuss procedure and open resections can guarantee better results, however nowadays Nuss procedure is far more used because it is less invasive and does not leave anterior scars. The complications observed both in open and mini-invasive procedures are wound infections, hematomas, bar shifts, pneumothorax, transient Horner syndrome, bleeding from thoracic vessels, overcorrection or mild correction (Acastello, 2006; Haller et al., 1987; Kelly et al., 2010; Lopushinsky & Fecteau, 2008; Nuss, 2008; Park et al., 2004). Complications of an extensive open procedure, particularly at early age, are floating sternum (Prabhakaran et al., 2001) and acquired Jeune syndrome (Haller et al., 1996), while in Nuss procedure pericarditis and allergy to nickel (component of the metal bar) have been reported occasionally (Nuss, 2008). Very few heart lesions and deaths were reported, mainly in cases of procedure done without thoracoscopy (Moss et al., 2001; Nuss, 2008). In case of Nuss bar infection, this can be managed successfully conservatively (Van Renterghem et al., 2005). Recurrence is reported in a range between 2% and 5% (Kelly et al., 2010; Lopushinsky & Fecteau, 2008). In our experience, in case of suboptimal result, one or more lipofilling treatments can improve significantly the final outcome but there are no published series yet. In females with breast asymmetry due to the sternal rotation, Nuss or open procedures alone can correct the breast aspect, but in case some degree of asymmetry persists, breast augmentation can be required, (Rapuzzi et al., 2010).

2.1.2 Pectus carinatum (PC)

PC is the second most frequent malformation. Its incidence is estimated to be 5 times less frequent than PE (Colombani, 2009; Fokin et al., 2009), with a strong male predominance. In some areas of the world, however, PC is almost equally or more frequent than PE (Acastello, 2006; Martinez-Ferro et al., 2008; Peña et al., 1981). The deformity is a protrusion of the sternum and chondrocostal joints (figure 2).

The etiology is unknown, but the pathogenetic mechanism could be the same than for PE, consisting in an overgrowth of the ribs (Haje et al., 1999). The same anomalies in the costal ribs than in PE have been reported in PC patients (Fokin et al., 2009). Familial cases are not uncommon (Fokin et al., 2009; Martinez-Ferro et al., 2008) and in some families it is possible to observe both PC and PE cases (Martinez-Ferro et al., 2008). Connective tissue disorders, Noonan syndrome and cardiac anomalies are seldom associated with PC (Kotzot &

Schwabegger, 2009). PC usually appears later in life than PE, mainly during pre-puberty or puberty, but in some cases it is possible to observe infants or children with this anomaly. PC has the tendency to increase rapidly during the growth spurt. The same symptoms than in PE can be observed, but more frequently some degree of thoracic pain than respiratory complaints (Colombani, 2009). Usually the cardiac and pulmonary function are less implicated than in PE (Fokin et al., 2009), but psychological effects of PC can be severe and they are the fundamental indication to the surgical correction.

2.1.2.1 Diagnostic assessment and classification

PC is classified according to the localization and symmetry into the following types (Colombani, 2009; Williams & Crabbe, 2003):

- Type 1, inferior or Chondrogladiolar (figure 2): It is the most frequent type. The sternal protrusion is located in the inferior or mid sternum. The last ribs can be slightly or severely depressed on lateral aspects. It is more often symmetric.
- Type 2, Superior or Chondromanubrial. In some reports it is called also Currarino-Silverman syndrome (Currarino & Silverman, 1958) or Pouter Pigeon Breast, but there is confusion in the literature regarding superior PC. Actually in our experience we have observed two different anomalies of superior PC that we have to differentiate. The most frequent is a sternal malformation characterized by a premature fusion and ossification of manubrio-sternal joint and the sternal segments, resulting in a high symmetric carinatum chest deformity with a short thick sternum with a depression in the lower third. (figure 2). This anomaly is sometimes described in the literature as Currarino Silverman syndrome or Pouter Pigeon Breast (Currarino & Silverman, 1958). The aspect is of a superior PC with an inferior PE. The sternum on a lateral view is S-shaped. Although this anomaly is included into the cartilaginous anomalies and called type II PC in Acastello classification (Acastello, 2006), we classified it as part of sternal anomalies because of the sternal origin of the anomaly (see table 1).



Fig. 2. Type I PC (left image); Type II PC or Currarino-Silverman syndrome (middle image). Lateral Computerized Tomography reconstruction shows S shape of the sternum (right image)

- The second anomaly we have observed in few cases is a superior PC without the typical features of Currarino-Silverman syndrome (figure 3). The sternum has a normal length and is not depressed in the lower third. This anomaly is probably due, similarly to inferior PC, to a cartilage anomaly. We propose to use the term superior PC only for the latter and to include this anomaly in the first category (cartilaginous anomalies) of CWMs classification. This entity is extremely rare in our experience and it has not been described hereto, to the best of our knowledge. The term of superior PC for the description of Currarino Silverman syndrome can be confusing and should be avoided.

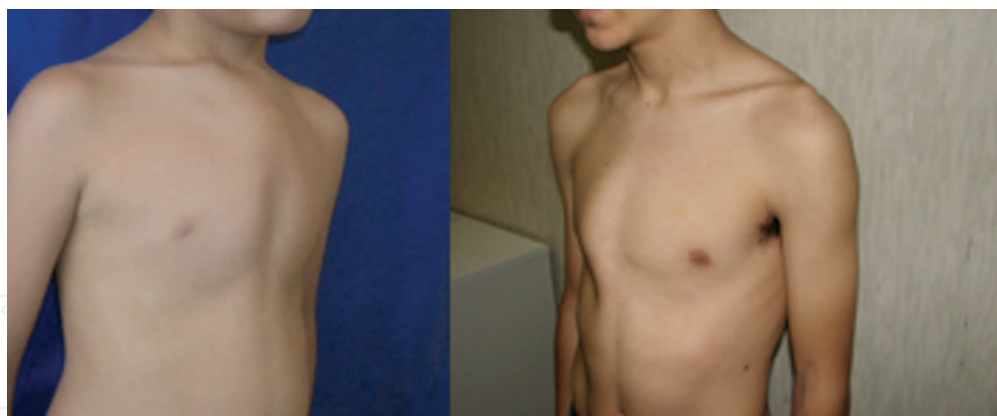


Fig. 3. Superior PC without features of Currarino-Silverman syndrome (left); Unilateral PC (right)

Other types of PC described are:

- Lateral or unilateral PC (Fokin et al., 2009): asymmetric by nature, it consists in a protrusion of some costal cartilages near chondro-sternal joint on one side (figure 3). The sternum can be rotated towards the opposite side.
- Reactive PC (Swanson & Colombani, 2008): this type of PC is a complication of a PE correction, in which in the first months or years after Nuss or open procedure the sternum progressively displaces ventrally. It is more frequent in patients with connective tissue disorders.

To best assess the gravity of PC and the degree of asymmetry, some radiological indexes have been proposed (Egan et al., 2000; Stephenson & Du Bois, 2008), measurable on CT scan, but in clinical practice they are less used than Haller index for PE. CT scan remains the gold standard radiologic evaluation for PC.

2.1.2.2 Treatment options

The standard correction has been performed through costal excision surgery, as for PE. Ravitch in 1952 (Ravitch, 1952) was one of the first who described the surgical technique for PC. While Lester in 1953 (Lester, 1953) proposed the resection of the lower third portion of the sternum, Howard (Howard, 1958) introduced the principle of sternal osteotomy, usually required to correct the defect. Osteotomy on the anterior sternal plate can be performed transversally or, in case of asymmetric PC, in an oblique fashion. Recently, some modifications to the Ravitch and Welch procedure were proposed (Del Frari & Schwabegger, 2011; Fonkalsrud & Anselmo, 2004), attempting to reduce the invasiveness of this approach, by reducing the extent of muscle and cartilage resection. The best treatment for PC type 2 and Currarino Silverman syndrome remains the open procedure (Brichon & Wihlm, 2010), while the following alternatives, minimally invasive or conservative techniques, have been recently proposed for type 1 PC, with good results:

- The *orthotic brace system*, proposed already in 1992 (Haje & Bowen, 1992) but popularized only recently by different groups approximately at the same time (Banever et al., 2006; Frey et al., 2006; Kravarusic et al., 2006; Rapuzzi et al., 2010, Swanson & Colombani, 2008), is based on the principle of reshaping the thorax during puberty due to thoracic malleability (as in Nuss procedure for PE) by applying a dynamic compression on it. Martinez-Ferro (Martinez-Ferro et al., 2008) added to this system the possibility to measure the pressure necessary to the correction and to regulate it

(dynamic compression system, DCS). He observed good results in a large proportion of patients, if the brace is used for most of the time during day and night. A significant proportion of non compliant patients (13.8%) who abandoned the treatment, and some minor complications (hematomas, ulcerations, back pain) in 12.5%, were reported (Martinez-Ferro et al., 2008). Moreover, this approach cannot correct a rigid ossified thorax, so it can be applied only to adolescent patients.

- Intrathoracic compression procedure (Abramson's procedure) (Abramson, 2005): the concept is the same as the orthotic brace, but the system is placed surgically. Through two lateral incisions, a metallic bar is inserted in presternal space under the pectoralis muscles and fixed to lateral stabilizer in order to push back the sternum. It is like a reverse Nuss procedure and it is based again on the same principle of the thoracic malleability. As the previous approach, it has an age limit. It has the advantage of obtaining immediately the result without the need of wearing an external brace. In the Abramson experience, at 5 years the results were good; the bar was removed usually after two years or more (Abramson et al., 2009).
- Thoracoscopic cartilage resection (Kim & Idowu, 2009): described recently, it is performed by resecting under thoracoscopic view uni- or bilaterally, according to the type of defect, the anomalous costal cartilages, without damaging the internal thoracic vessels. It can be associated in severe cases with an intrathoracic compression procedure according to Abramson technique in order to stabilize better the sternum.
- Thoracoscopic complete cartilage resection with perichondrium preservation (CCRPP) (Varela & Torre, 2011): reported by our group, this procedure differs itself from the previous because cartilages are prepared both laterally and medially to the internal thoracic vessels, up to the chondrosternal joints. Internal thoracic vessels are coagulated and cartilages completely excised, leaving the anterior perichondrium intact.
- Mini-invasive submuscular dissection (Schaarschmidt et al., 2006): the pectoralis muscle dissection is performed by subpectoral CO2 insufflation, the resection of the ribs, the sternal osteotomy and the insertion of trans-sternal steel struts are performed through a vertical pre-sternal incision under endoscopic view. Recently, the same Authors reported some technical variations (Schaarschmidt et al., 2011), abandoning the pre-sternal incision and performing a more extense submuscular dissection and two lateral incisions between the anterior and middle axillary lines. These should allow the creation of a submuscular and presternal tunnel in order to implant a Nuss metal bar presternally. Specific eight-hole stabilizers are though required.
- Minimal access treatment of PC (Hock, 2009): the bar is inserted as in Abramson procedure through two lateral incisions above the sternum, but it passes on both sides into the thoracic cavities; thoracoscopy was not used.

Reactive PC after Nuss procedure can be simply corrected with the withdrawal of the bar; in case of failure or in other cases an open procedure is advised; alternatively a mini-invasive technique can be attempted.

2.2 Type II: Costal anomalies

2.2.1 Dysmorphic cartilaginous type II CWMs (not syndromic)

This group is a spectrum of costal anomalies. Cartilages are malformed and the consequence can be a unilateral or bilateral depression in the thoracic wall. The treatment consists in a cartilage excision.

A rare malformation belonging to this group of malformation is the so called “intrathoracic rib” (figure 4), classified into different types (Kamano et al., 2006):

- *type Ia* is a supernumerary rib articulated with a vertebral body, *type Ib* is a bifid rib taking origin close to the vertebral body;
- *type II* a bifid rib arising more laterally;
- *type III* is a not bifid rib depressed into the thoracic cavity.

Flaring Chest consists into an hypertrophy or fusion of the cartilages in the lower costal margin. Open resection of all these malformed cartilages is an option treatment.

Cartilage rib asymmetries are quite frequently seen, they appear like isolated protrusion in the cartilage ribs. In the majority of cases the ribs are fused (figures 4).

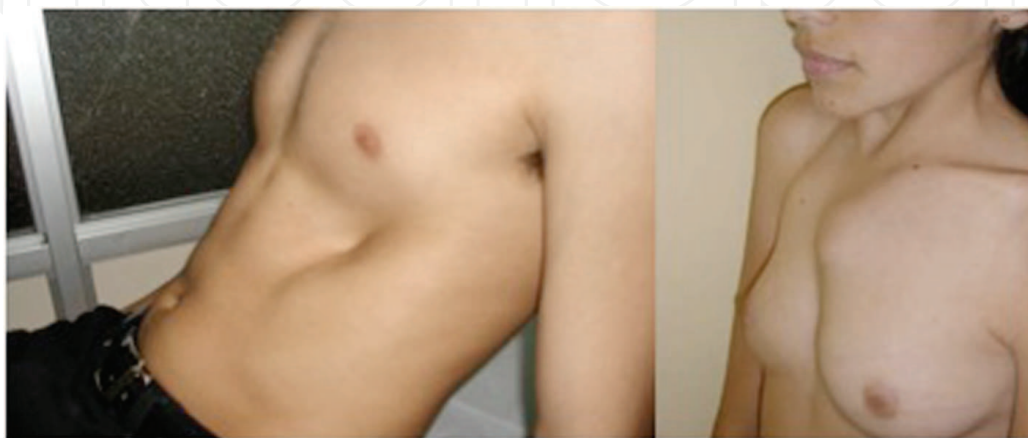


Fig. 4. *Left*: Dysmorphic anomaly of the ribs (type II of Acastello classification); *Right*: Dysmorphic and fused ribs

2.2.2 Syndromic type II anomalies

2.2.2.1 Jeune syndrome

Jeune Syndrome or asphyxiating thoracic dystrophy is an autosomal recessive disorder, originally described by Jeune in 1954 (Jeune et al., 1954) in a pair of siblings. The frequency of the condition is estimated 1/ 100.000 to 1/30.000 live births. Jeune syndrome is characterized by many bone abnormalities, the most pronounced being a long, narrow thorax with a reduced thoracic capacity causing the lungs to not have enough room to expand and grow. Both antero-posterior and lateral thoracic diameters are reduced, so respiratory distress may be severe. Prognosis is poor in patients who have respiratory symptoms during the first months of life, resulting in death during infancy.

All patients have small chests with short, wide and horizontal ribs (figure 5). There is variability in the severity of clinical and radiographic features, and two variants of Jeune syndrome exist:

- Severe variant: It represents the 70% of cases, and it usually is lethal during the infancy. The thorax is extremely small, conversely the abdomen seems prominent; respiratory failure is the rule.
- Mild variant: In 30% of cases ribs are less affected, respiratory symptoms are manageable and survival is prolonged. Renal or liver dysfunctions are present in some cases, and they can lead to death patients affected by this type of malformation.

Surgical repair techniques have typically involved median sternotomy (with graft interposition), resulting in poor outcomes (Philips & van Aalst, 2008). Lateral thoracic

expansion, realized by rib incisions and suture in a staggered fashion (Davis et al., 1995), or more recently vertical expandable prosthetic titanium rib (VEPTR) (Waldhausen et al., 2007) are techniques proposed more recently that seem to offer some good results. The mild type of Jeune syndrome may not require any treatment.

2.2.2.2 Cerebrocostomandibular syndrome

Cerebrocostomandibular Syndrome is a rare entity. There is no clinical experience in the world. We have diagnosed one case in the last ten years, the main feature is a lack of development of the rib cage. There are only costal vestiges. There is flail chest and mechanical ventilation is required since birth. In some cases the thoracic cage agenesis is unilateral (figure 5). This defective costal development is also associated with features of the Pierre-Robin anomalad. Cerebral maldevelopment or malfunction is also common (Drossou-Agakidou et al., 1991).

2.2.2.3 Costal agenesis

Costal agenesis is limited to some ribs and is not syndromic. They are rare conditions. Lung herniation occurs. It may require thoracoplasty using the same technique used for PS.

2.2.3 Rare type II CWMs

There is a series of CWMs rarely observed, not included in a standard classification. As they differ each one from the other the treatment must be personalized.

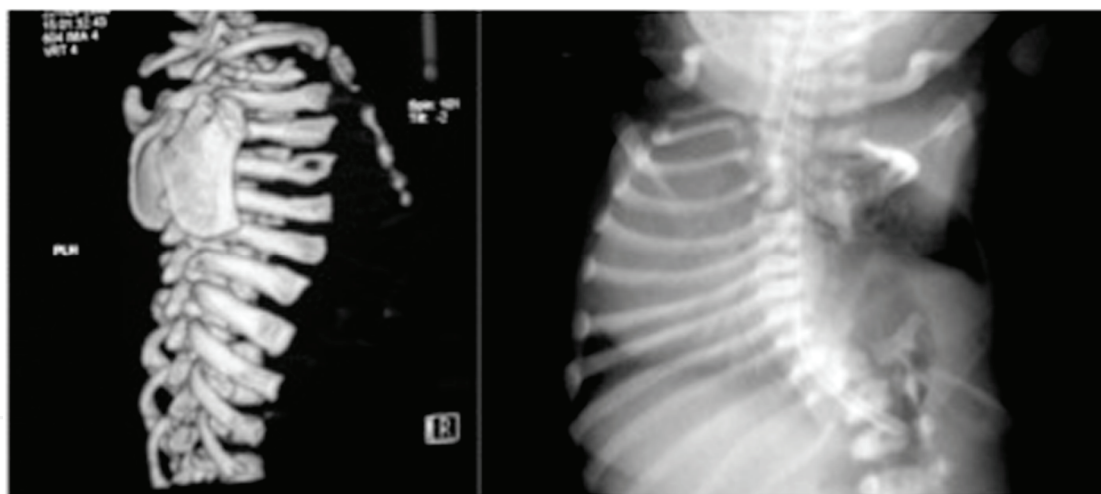


Fig. 5. *Left*: Jeune syndrome (Asphyxiating thoracic dystrophy), type severe. Computerized Tomography reconstruction of the rib cage shows the typical ribs of Jeune syndrome; *Right*: Cerebrocostomandibular syndrome. Unilateral agenesis of thoracic cage shown at thoracic X-Ray

2.3 Type III: Chondrocostal anomalies

2.3.1 Poland Syndrome (PS)

Occurring in approximately 1/30,000 live births (Freire-Maia et al., 1973), PS is characterized by the absence or hypoplasia of the pectoralis major muscle, frequently combined with other ipsilateral abnormalities of the chest wall, breast and upper limb (Kelly, 2008). The defect is essentially unilateral and in two thirds of cases right-sided. There is a male preponderance with a ratio of about 2/1 with females. Very rare bilateral cases have been described (Baban

et al., 2009; Karnak et al., 1998). The etiology is unknown, but the most accredited hypothesis is the interruption of the vascular supply in subclavian and vertebral artery during embryonic life (Bavinck & Weaver, 1986), leading to different malformations in the corresponding districts. According to this, PS could be actually interpreted as a sequence.

Alternately, paradominant inheritance (Happle, 1999) or the presence of a lethal gene survival by mosaicism (van Steensel, 2004) have been proposed to explain the origin of this anomaly. PS is usually sporadic, but the occurrence of familial cases has raised the hypothesis of a possible transmission with an autosomal dominant pattern; however there is still no evidence of that. Association of PS with other anomalies, as Moebius (Parker et al., 1981), Klippel Feil syndromes and Sprengel anomaly (Bavinck & Weaver, 1986), has been reported.

PS phenotype is extremely variable (Alexander et al., 2002; Shamberger et al., 1989). The thoracic defect is usually evident at birth, but it can be undiagnosed until the child gets older. The pectoral muscle deficiency causes an asymmetric aspect but if there are costal anomalies associated the defect is more evident. In case of rib agenesis, particularly if multiple (the most affected ribs are the third and the fourth), lung herniation and paradoxical respiratory movements are always present. Ribs can also be smaller or anomalous. Anomalies like a PE or PC or both can occur, but in less than 10% of cases they require surgery. Breast region and nipple are frequently involved. A mild degree of breast hypoplasia to a complete absence of mammary gland are constant features. Associated cardiac and renal anomalies, as well as scoliosis, have been reported, but they are uncommon (Alexander et al., 2002). Dextroposition is reported frequently, always associated with left PS, and it seems to be caused by mechanical factors during embryonic life in patients with multiple left rib agenesis (Torre et al., 2010). Patients with PS are asymptomatic, and there are usually no limitations due to the muscle defects. Upper limb is frequently involved, from the classical symbrachydactily to split hand or other defects (Al-Qattan, 2001; Shamberger et al., 1989).

Thoracoplasty finds its main indication in cosmetic reason (Ravitch, 1966). Only rarely it is necessary a thoracoplasty in the infancy. There is no evidence of the utility of thoracoplasty for protection against thoracic traumatic injuries in children with rib agenesis. In case of surgical correction in pediatric age, some options are available, from costal transposition described again by Ravitch in 1966 (Ravitch, 1966), to the repair with absorbable or not absorbable prostheses (Moir & Johnson, 2008; Urschel, 2009). According to some Authors (Acastello, 2006), costal transposition and the consequent stabilization of the thorax could prevent the progression of thoracic deformity, but there is no consensus about this concept. Most Authors (Moir & Johnson, 2008; Urschel, 2009) prefer to wait until puberty and further, in order to correct in one or more times the thoracic flail chest and the pectoral defect. At this age, the most frequent issue in PS is breast and pectoral reconstruction in female patients. Correction with prostheses alone or in association with other surgical procedures (latissimus dorsi or rectal abdominal muscle transposition, lipofilling, or omental flap or other techniques) has been advocated (Urschel, 2009), but the surgical approach has to be tailored on the single case. In males the same techniques can be applied, but the indication to the surgical procedure has to be evaluated case by case, because the esthetical defect is less important. Martinez-Ferro described latissimus dorsi transposition flap using a minimally-invasive approach (Martinez-Ferro et al., 2007). Usually teams including

pediatric or thoracic surgeons together with plastic surgeons can treat PS patients with the highest chance to get the best results.

2.4 Type IV: Sternal anomalies

2.4.1 Sternal cleft

A defect in the sternum's fusion process causes the sternal cleft, a rare idiopathic CWM. Acastello et al. found that sternal cleft (SC) accounted for 0.15% of all CWMs (Acastello et al., 2003). The Hoxb gene might be involved in the development of SC (Forzano et al., 2005). Known from many centuries, these malformations have been classified in many different ways. To our knowledge the clearest classification has been proposed by Shamberger and Welch (Shamberger & Welch, 1990) and includes 4 types:

- Thoracic ectopia cordis: the heart is ectopic and not covered by skin. Usually the heart, in an anterior and cephalic ectopia, has intrinsic anomalies. The sternal defect can be superior, inferior, central (rare) or total. Abdominal wall defects as omphalocele can be associated. Thoracic cavity is hypoplastic, and for this reason the surgical correction is usually not able to save the life to these patients. Isolated survival after surgery has been reported (Dobell et al., 1982).
- Cervical ectopia cordis: much rarer than previous type, the heart is more cranial, sometimes with the apex fused with the mouth. Associated craniofacial anomalies are frequent. Prognosis is always negative.
- Thoraco-abdominal ectopia cordis: the heart is covered by a thin membranous or cutaneous layer. An inferior sternal defect is present. The heart, located into the thorax or into the abdomen, is not rotated as in previous types but intrinsic anomalies are common (Major, 1953). This kind of anomaly is generally found as part of a field defect known as the pentalogy of Cantrell (Cantrell, 1958). Prognosis after surgical repair can be good.
- Sternal cleft is the most common of this group of CWMs, and consists in a congenital malformation of the anterior thoracic wall, arising in a deficiency in the midline embryonic fusion of the sternal valves. The incidence is unknown, and it is more frequent in females (Acastello, 2006).

In sternal anomalies we have included also Currarino Silverman syndrome or Pouter Pigeon Breast, as already described above.

Sternal clefts are classified as being partial (figure 6) or complete (figure 6). The partial deformity can be superiorly or inferiorly located. The rarer inferior variety is often associated with a thoraco-abdominal ectopia cordis, while upper partial cleft (the most common variant) can be an isolated abnormality. The sternal clavicular joints are displaced laterally, but the clavicles have a normal length. There is a bulging of thoracic viscera in the midline across the defect, more evident during forced expiration. The complete form is much less frequent. There is a total lack of fusion; it produces an even bigger paradoxical movement than partial cleft and sometimes respiratory distress.

According to a recent review of the literature, SC is frequently associated with other defects (82%) (Torre et al., 2011). These must be carefully looked for before any surgical procedure, since they can lead to major complications. Some of them are evident on physical examination such as maxillofacial hemangiomas (Fokin, 2000), cleft lip or cleft palate, pectus excavatum, connectival nevi (Torre et al., 2011), supraumbilical raphe, or gastroschisis. Other defects must be ruled out, such as cardiac defects, aortic coarctation, eye abnormalities, posterior fossa anomalies, and hidden haemangiomas.



Fig. 6. Partial sternal cleft in a 4-year-old patient not operated at birth; Total sternal cleft in a newborn (right) also part of clinical conditions like PHACES syndrome (Metry et al., 2009), sternal malformation/vascular dysplasia, midline fusion defects, or Cantrell's pentalogy.

There is consensus that ideally correction of sternal clefts should take place during the neonatal period or in the first months of life (Acastello, 2006; Domini et al., 2000; Torre et al., 2008), to re-establish the bony protection of the mediastinum, prevent paradoxical visceral movement with respiration, eliminate the visible deformity and allow the normal growth of thoracic cage. The reason for preferring an early surgical approach is that primary closure is easier and there is no need of a big procedure, maybe necessary at older ages. In fact, after the first few years of life, primary closure requires sterno-clavicular disarticulation, sternal isolation, inferior sternal osteotomy and medialization of the neck muscles after separation of their sternoclavicular attachments laterally (Acastello et al., 2003). As it can bring the risk of a circulatory impairment due to cardiovascular compression, in some cases primary sternal suture can be impossible and prosthetic or autologous closure (De Campos et al., 1998) can be preferable because less invasive. Partial thymectomy can be useful to reduce the pressure on thoracic vessels (Torre et al., 2008). Many prosthetic materials have been described for sternal cleft repair (Domini et al., 2000). In our experience we have closed an upper cleft in one 8 year old female with an artificial bone tissue with an excellent outcome. Complications are not frequent, but PE can occur later in life in patients operated for sternal cleft. In case of prosthetic repair, there is an increased risk of infections and recurrence.

2.5 Type V: Clavicle-scapular anomalies

These anomalies usually are field of interest of orthopedic surgeons more than pediatric surgeons. We do not have experience of this type of CWMs.

2.6 Other anomalies

2.6.1 Post operative surgical deformities

This category includes cases in which thoracic deformity is due to the correction of a previous CWMs. We have experience of few cases of this kind of anomaly (figure 7). They were due to early multiple cartilage resections during an open correction of a PE and finally resulted, after many years, in a thoracic deformity that required an open revision procedure. As discussed above, the optimal age for pectus repair is controversial (Lopushinsky &

Fecteau, 2008; Nuss, 2008). Repair in early childhood is easier but cases of restrictive growth patterns of the chest wall have been reported. Jeune syndrome or acquired asphyxiating thoracic dystrophy is associated with open repair in children less than 4 years with extensive resection of five or more ribs (Haller et al., 1996) and damage to the cartilage growth centers (Robicsek et al., 2009). These children present with an extremely narrow chest. For these reasons most Authors postpone open surgical repair after 10 years of age (Lopushinsky & Fecteau, 2008; Nuss, 2008).



Fig. 7. Post surgical deformity in a 6-year-old boy operated of PE when he was one year old

3. Conclusions

CWMs are a large spectrum of anomalies. Etiology and genetic implication of CWMs are still largely unknown. Precise identification of the single malformation, its classification and an accurate diagnostic assessment, are the first fundamental steps in the modern approach. We have adopted the modified classification of Acastello, based on the origin of the anomaly. Identification of familial cases, possible associated syndromes and anomalies, clinical symptoms and psychological implications have to be considered. Among the therapeutic armamentarium, nowadays classical techniques and new approaches make us able to choose the more appropriate for the single patient, according to the surgeon's experience and preference but in particular tailoring the treatment on the individual clinical and psychological needs. A multidisciplinary approach is advisable in order to manage CWMs in all their complexity.

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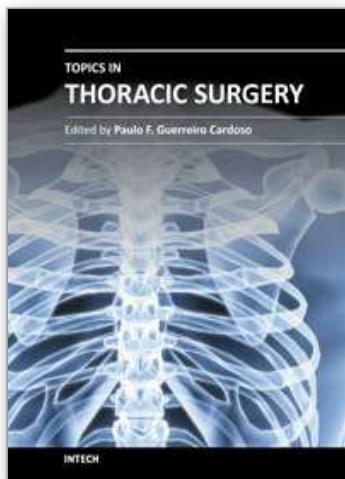
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