DOI 10.18019/1028-4427-2017-23-2-241-247

Clinical types of chest wall deformities (literature review)

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The article presents an overview of embryonic and postnatal development of the chest wall describing current classification for a variety of chest wall and rib malformations. The clear identification of specific anomalies and understanding of antenatal and postnatal development of the chest wall contribute to optimizing the choice of time and technique to treat the cohort of patients.
Keywords: chest wall, embryogenesis, chest wall, ribs, deformity

Congenital chest wall deformities represent a wide spectrum of sternocostal anomalies, with a relatively high incidence. Although significant pulmonary dysfunction is rarely seen in the patients the majority of children and their parents seek medical advice for psychosocial concerns due to poor cosmesis and aversion to sports and public exposure. Chest wall deformities 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.

Some deformities may occur in isolation or as part of a multiple malformation syndrome.

Embryogenesis of the human chest wall

The anlage of ribs and vertebrae is a closely related process of intrauterine development. Ventromedial surface of each somite gives rise to a group of mesenchymal cells that are known as the sclerotome bilaterally migrating to midline and accumulating around the chord. The sclerotome develops into the vertebrae. Clusters of cells form the anlage of the vertebral body. With the vertebral core developed clusters of mesenchymal cells spread dorsally and laterally and form the anlage of neural arches and ribs. Blastema gives rise to cartilage with buds being at the vertebral body and then chondrofication centers develop in neutral and costal processes. The chondrofication centers rapidly increase in volume and fuse with each other until cartilage is formed. Cartilaginous vertebra is primarily an integral component with no signs of partition that is seen later with the cartilage replaced by the bone. Cartilaginous ribs are separated from vertebrae at early ossification, and vertebrae remain unpartitioned (**Fig. 1**). The median ossification center gives rise to vertebral body. The centers located in the neutral processes spread dorsally forming the plate and neural arch. Spinous process develops due to the centers spreading dorsally from the meeting point. Transverse processes with articulating costal tubercles are formed by lateral location of ossification centers developing in neutral processes. The centers fuse with each other ventrally. The rib is formed due to spreading ossification from its center. Secondary epiphyseal centers develop in the costal head and tubercle postnatally. They remain separated from the rest of the rib by cartilaginous plates throughout growth. No fusion of the secondary epiphyseal centers with the rest part of the rib occurs until growth to the skeleton is complete [1].

The above is related to thoracic vertebra formation with well-defined relations between the rib and the vertebra. A costal element is present in each of the vertebrae, but it is reduced and changed in different vertebral sections (**Fig. 2**).

Cervical rib on the seventh cervical vertebra, an extra rib arising from the first lumbar vertebra, cleft rib, sternum bifidum at the costochondral junction are most common skeletal anomalies due to specificity of embryogenesis (**Fig. 3**) [1].

Komolkin I.A., Agranovich O.E. Clinical types of chest wall deformities (literature **review)**. *Genij Ortopedii*. 2017. T. 23. No 2. pp. 241-247. DOI 10.18019/1028-4427-2017-23-2-241-247

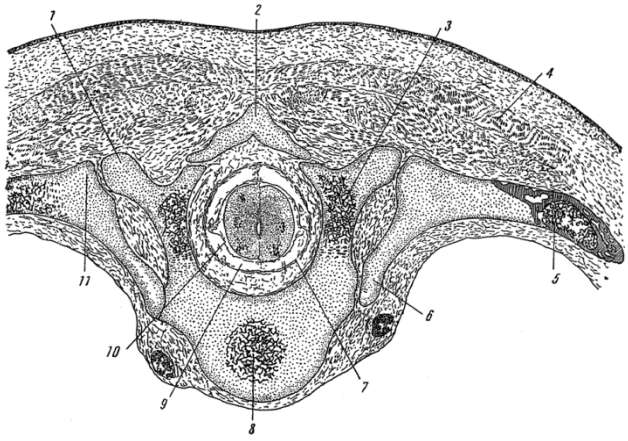

Fig. 1 Cross section of a human embryo at 12-week gestation (patietosacral distance of 72 mm) showing developing vertebra and ribs (cited according to B.M. Petten, 1959): 1 – transverse process; 2 – spinous process of adjacent vertebra; 3 – lateral ossification center; 4 – spinal muscles; 5 – costal ossification center; 6 – costal head; 7 – dura mater; 8 – median ossification center of the vertebral body; 9 – subarachnoid space; 10 – pia mater; 11 – costal tuberosity

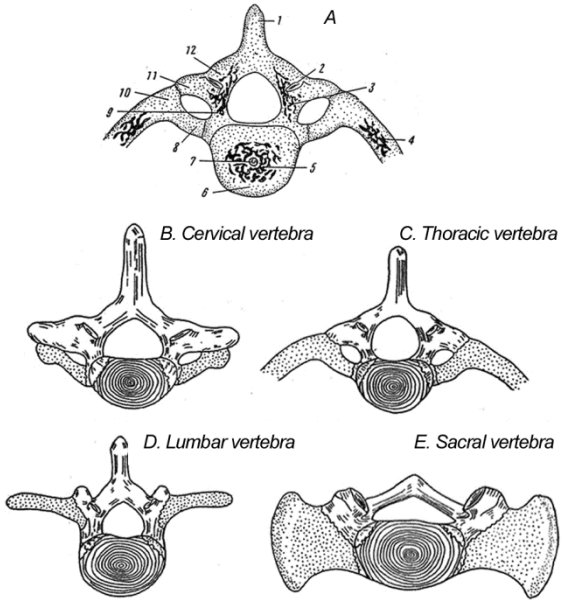

Fig. 2 Components and ossification centers of growing vertebra (cited according to B.M. Petten, 1959): A – various ossification centers located in thoracic vertebra and related ribs; B-E – typical components of different vertebrae. A.: 1 – spinous process; 2 – articular process; 3 – lateral ossification center; 4 – costal ossification center; 5 – median ossification center; 6 – vertebral body; 7 – chord; 8 – costal head; 9 – base of neural arch; 10 – costal tubercle; 11 – transverse process; 12 – plate

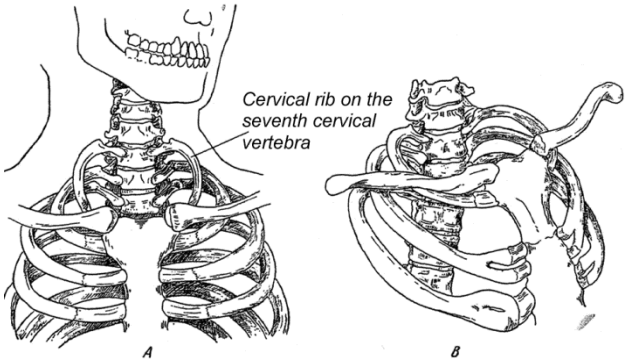

Fig. 3 Costal abnormalities (cited according to B.M. Petten, 1959): A – cervical rib (after Koring); B – bifurcated ribs (after Arey) (cited according to B.M. Petten, 1959)

The sternum forms from a pair of vertical mesenchymal bands referred to as sternal bars. They fuse in the midline to form the cartilaginous models of manubrium. The process begins at the cranial end and costal cartilage grows from vertebra to sternum. At 9 weeks sternum is placed at midline, with ribs reaching the sternum. By the time sternum and adjoining ribs acquire cartilaginous structure with starting costal ossification. The sternum undergo secondary transverse partition into several cartilages, each having ossification center mostly located in the center with occasional two left and right entities. They finally fuse at puberty. Such anomalies as sternal cleft , sternal foramen and forked xiphoid process are associated with the paired element of sternal anlage (**Fig. 4**) [1].

The sternal ossification occurs late. The first ossification point develops in manubrium at 5 to 6 months of gestation, then in the sternal body at 5 to 9 months of fetal growth and in the xiphoid process at 6 years. The fusion of sternal body fragments is normally complete by 25 years of age; xiphoid process and the body fuse at the 25 to 28 years, and later (and not always) the body and manubrium at the age of 25 to 30 years (**Fig. 5**) [1, 2].

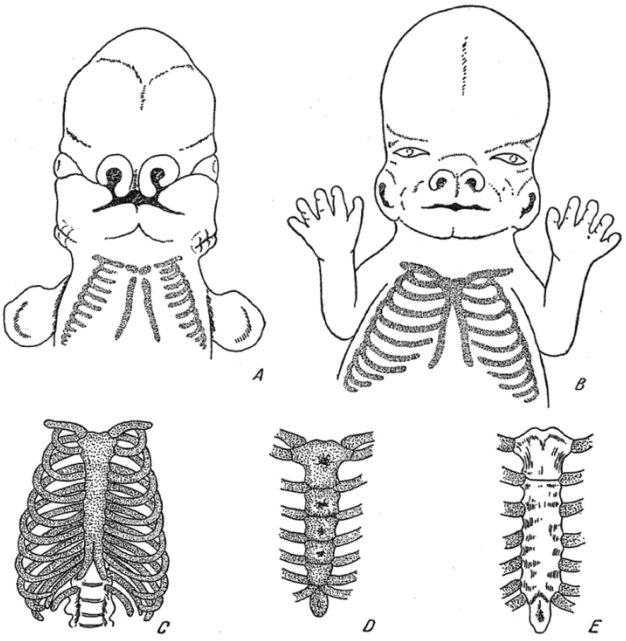

Fig. 4 Sternal development (cited according to B.M. Petten, 1959): A – at 6 weeks; B – at 8 weeks; C – at 9 weeks; D – at 5.5 months; E – sternum of an adult

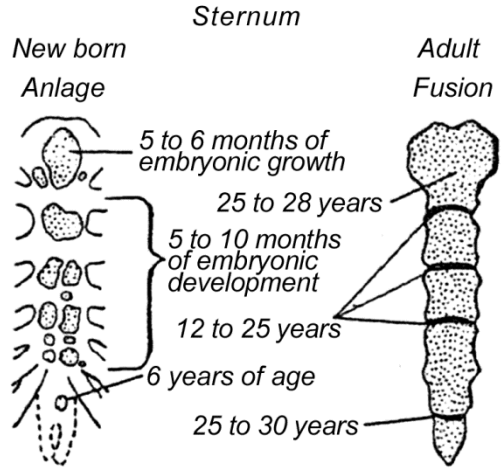

Fig. 5 Sternal ossification (cited according to B.M. Petten, 1959)

Classification of chest wall malformations

Classification of chest wall deformities offered by Acastello and modified by M. Torre et al. (2012) identified 5 major types [3].

Type I. Cartilaginous abnormality:

- Pectus excavatum
- Pectus carinatum (type 1 and type 2).

Type II. Costal deformity:

- Simple (1 or 2 ribs) associated with agenesis, hypoplasia, sovrannumerary, bifid, fused, dysmorphic, rare (always complex);
- complex (3 or more ribs). associated with agenesis, hypoplasia, sovrannumerary, bifid, fused, dysmorphic, rare (always complex);
- syndromic (always complex): Jeune, Jarcho-Levin, cerebrocostalmandibular, others

Type III. Chondro-costal deformity:

- Poland syndrome.

Type IV. Sternal deformity:

- Sternal cleft (with or without ectopia cordis);
- Currarino Silverman syndrome.

Type V. Clavicle-scapular malformation:

- Simple or syndromic clavicular;
- Simple or syndromic scapular;
- Combined abnormality.

Type I. Cartilaginous abnormality

Pectus excavatum is the most frequent thoracic malformation, with an incidence of 1/100 to 1/1000 live births and accounting for 90 % of all chest wall deformities [4, 5]. The anomaly is characterized by the presence of a variably deep sternal depression associated with deformity of the lower chondrosternal joints. In 15 % of the cases the abnormality is seen during development. In later occurrence there is a frequent association with malformations of the muscular connective tissue (Marfan syndrome, Ehlers-Danlos syndrome) [4–8]. The etiology of pectus excavatum is not clear, and many hypotheses have been proposed. ДThe role of vitamins or other nutrient deficiencies is probably not influent at all, while a connective tissue disorder and genetic predisposition can play an important role for pectus excavatum. According to Kelly (2008), the condition shows a familiar recurrence in up to 40 % of the cases, other deformities such as pectus carinatum, can be rarely seen in a family with pectus excavatum [7]. The most frequent transmission pattern can be the autosomal dominant, but there are families with autosomal recessive and X-linked patterns [9]. According to several authors, the overgrowth of costal cartilages can be the pathogenic mechanism leading to the development of pectus excavatum [4, 7, 9]. Collagen type II disorders have been shown in the costal cartilages in pectus excavatum, as well as overexpression or downregulation of some genes playing a role in metabolism of cartilage and connective tissue (collagen genes, matrix metalloproteinsases, tumor necrosis factor alpha, and filamin) [4, 11].

Morphological grouping of the deformities are as follows [4, 12]:

1. Grand-Canyon is a severe and deep pectus excavatum with a deep long canal in the sternum. The deformity correction is extremely difficult, especially when thorax is largely ossified and sternum is extremely rotated. A higher complication rate is reported as compared with other corrections.

2. Punch or cup shape is a local deformity with the inferior portion of sternum being mostly involved. This type of pectus excavatum can be very difficult to correct at any age, and sometimes the outcome is partial.

3. Saucer type can be symmetric or asymmetric with an extensive depression along the complete anterior chest.

4. Transversal type is characterized by transversal depression below the sternum.

5. Eccentric pectus excavatum is characterized by the sternal depression being eccentric to midline. It is the highest degree of asymmetric deformity.

6. Pectus excavatum with flaring chest is the flaring chest at the level of the last ribs. This is an isolated malformation.

7. Pectus excavatum-pectus carinatum is a combined malformation with a sunken chest and cartilage protrusion beside the sternum edge.

8. Superior type is a very rare pectus excavatum localizing in the upper part of sternum and cartilage ribs. Lower sternum is normal.

Pectus carinatum is the second most frequent deformity. The incidence is estimated to be 5 times less than pectus excavatum with a strong male predominance [4, 6]. However, pectus carinatum is almost equally or more frequent than pectus excavatum in some countries. The deformity is a protrusion of the sternum and chondrocostal joints. The etiology is unknown but the pathogenic mechanism can be the same than that for pectus excavatum consisting in an overgrowth of the ribs [10]. Familial cases are not uncommon, and both types of the deformities can be observed in some families [13]. Connective tissue disorders, Noonan syndrome and cardiac anomalies are seldom associated with pectus carinatum [8]. Pectus carinatum is normally seen later in life as compared to pectus excavatum, mainly at pre-puberty or puberty, and in some cases the deformity can be observed at early childhood. Pectus carinatum has a tendency to progress rapidly during the growth spurt. Several symptoms are similar for both pectus excavatum and pectus carinatum, however, the latter is more associated with pain than respiratory disorders [6]. Cardiac and pulmonary dysfunctions are less evident for pectus carinatum as compared to pectus excavatum, but psychological effects of pectus carinatum can be severe and they can be the major indication for surgical correction [4].

Pectus carinatum is classifies according to localization and symmetry as follows [6]:

Type 1, inferior or chondrogladiolar is the most frequent type. The sternal protrusion is located in the inferior or middle third of sternum. The last ribs can be slightly or severely depressed on lateral aspects. It is often symmetric.

Type 2, superior or chondromanubrial. It is also called Currarino-Silverman syndrome or Pouter Pigeon Breast [14]. There are two different anomalies of superior pectus carinatum that should be differentiated. 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 and depression in the lower third. This anomaly is described as Currarino-Silverman syndrome in literature. The malformation can also be characterized as superior type of pectus carinatum. The sternum is S-shaped on a lateral view.

Other types of pectus carinatum

Lateral or unilateral type is asymmetric by nature presented with protrusion of some costal cartilages near chondrosternal joint on one side. The sternum can be rotated towards the opposite side [4].

Reactive type is a complication of pectus excavatum correction with the sternum progressively displacing ventrally. The deformity is often seen in patients with connective tissue disorders [15].

Type II. Costal anomalies

Dysmorphic cartilaginous type II (non syndromic)

This group is a spectrum of costal anomalies resulting in unilateral or bilateral depression in the thoracic wall. The treatment includes cartilage excision.

A rare malformation of the group is called “intrathoracic rib” with several types in subclassified [16]:

Type I A is a supernumerary rib articulated with a vertebral body;

Type I B is a bifid rib taking an origin close to the vertebral body;

Type II is a bifid rib arising more laterally;

Type III is a not bifid rib depressed into the thoracic cavity.

Flaring chest is due to hypertrophy or fusion of cartilages in the lower costal margin. Open resection of all the malformed cartilages is indicated.

Cartilage rib asymmetries are quite frequent and appear as an isolated protrusion in cartilage ribs. Costal concrescence is observed in most of the cases.

Agenesis of ribs is a rare non-syndromic condition. Pneumocele can develop due to weak chest skeletal frame.

Syndromic type II anomalies

Jeune syndrome or asphyxiating thoracic dystrophy is a rare condition with the estimated frequency of 1/30 000 to 1/100 000 live births. It is characterized by multiple skeletal abnormalities with the most evident being a long, narrow thorax, short, wide, horizontal ribs and a reduced thoracic capacity resulting in respiratory deficiency. Prognosis is poor in patients who develop respiratory symptoms during the first months of life. The disease is an autosomal recessive disorder originally described by Jeune et al. in 1954 in a pair of siblings. There are two types of the disease with regards to clinical and radiological manifestations, severe and mild types.

Severe type represents 70 % of the cases normally resulting in lethal outcome during infancy due to severe respiratory deficiency. The thorax is extremely small.

Mild type is encountered in 30 % of the cases with the ribs less affected, respiratory symptoms being less evident and survival being prolonged. Renal or liver dysfunctions can lead to death in some cases [3].

Cerebrocostomandibular syndrome is a rather rare condition characterized by intrauterine lack of development of the rib cage and severe respiratory deficiency. There are only costal vestiges. Unilateral costal agenesis is seen sometimes. The anomaly is associated with Pierre-Robin syndrome. Cerebral maldevelopment or malfunction is also common [17].

Type III. Chondrocostal anomalies

Poland syndrome occurs in approximately in 1/30 000 live births and is characterized by the absence of hypoplasia of the pectoralis major muscle frequently combined with other ipsilateral anomalies of the chest wall, breast and upper limb [7, 18]. The defect is right-sided in 2/3 of the cases. There is a male preponderance with a ratio of about 2/1. Bilateral cases are very rare [19, 20]. The etiology is unknown but the most accredited hypothesis is disturbed intrauterine blood supply to subclavian and vertebral artery leading to different malformations in the corresponding areas [21]. Alternatively, paradominant inheritance or the presence of lethal gene survival by mosaicism has been suggested to explain the origin of the abnormality [22, 23]. The condition is normally sporadic but the occurrence of familial cases has raised the hypothesis of a possible transmission with autosomal dominant patter, however, there is still no evidence of that. Association of Poland syndrome with other anomalies as Moebius, Klippel Feil syndromes and Sprengel anomaly has been reported [21, 24]. Poland syndrome phenotype is extremely variable [25, 26]. The thoracic defect is usually evident at birth but it can be undiagnosed at early childhood. The pectoral muscles deficiency results in asymmetry, and the deformity is more evident in combination with costal agenesis. In case of costal agenesis, multiple in particular (the most affected ribs are the third and the fourth), lung herniation and paradoxical respiratory movements are present. Ribs can also be smaller or anomalous. Anomalies like pectus excavatum or pectus carinatum or both can occur and operative treatment is required in less than 10 % of the cases. Breasts are often involved, from mild hypoplasia to agenesis. Associated cardiac and renal anomalies, as well as spinal deformities have been occasionally reported [25]. Dextracardia is common and associated with left Poland syndrome and deemed to be caused by mechanical factors during embryonic life in patients with multiple left rib agenesis [27]. Upper limbs are frequently involved from classical symrachydactily to split hand or other defects [26, 28].

Type IV. Sternal anomalies

Sternal cleft is a rare idiopathic anomaly developing due to impaired embryonic sternal fusion. The disease accounts for 0.15 % of all malformations of the chest wall [29]. The Hoxb gene

might be involved in sternal cleft [30]. Classification proposed by R. Shamberger, K. Welch (1990) identifies four types of the condition [31].

1. Thoracic ectopia cordis with ectopic heart not covered by skin. The heart is displaced anteriorly and upwards and anomalous. The sternal defects can be superior, inferior, central and total. Abdominal wall defects as omphalocele can be associated with the condition. Thoracic cavity is hypoplastic and surgical correction is a failure for the children. However, some cases of survival after the surgery have been reported [32].

2. Thoraco-abdominal actopia cordis: the heart is covered by a thin membranous or cutaneous layer with an inferior sternal defect present. The heart located in the thorax or abdomen is anomalous with absent rotation [33]. The anomaly is known as a symptom of Cantrell syndrome [34]. Prognosis for surgical repair is good for the patients.

3. Cervical ectopia cordis: the heart is more cranial, with the apex fused with the mouth in some cases and associated frequent craniofacial anomalies. The condition is rare with poor prognosis.

4. Sternal cleft is a congenital malformation of the anterior thoracic wall arising in a deficient fusion of sternal valves during embryogenesis [35–37].

Sternal anomalies are observed also with Currarino-Silverman syndrome.

Sternal clefts can be complete or partial. Partial deformity can be superiorly or inferiorly located. Inferior type is often associated with thoraco-abdominal actopia cordis, and superior partial cleft is mostly an isolated malformation. Sternal-clavicular joints are displaced laterally, and clavicles have normal length. There is bulging of thoracic viscera in the midline across the defect. Complete cleft is less common as compared to the partial type. Complete sternal cleft entails paradoxical movements and respiratory distress over the cleft. In 82 % of the cases the anomaly is associated with other malformations and a thorough workup is needed for the patients to avoid major complications [3]. Maxillofacial hemangiomas, cleft lip or cleft palate, pectus excavatum, gastroschisis, cardiac defects, eye abnormalities hidden hemangiomas can be evident on physical examination [3, 4].

Type V. Clavicle-scapular anomalies

are quite rare [3].

Other anomalies

Postoperative secondary chest wall deformities include malformations developed after previous surgical treatment with multiple cartilage resections in pectus excavatum. Extensive resection of 5 or more ribs in children less than 4 years lead to damage to the cartilage growth centers and secondary chest wall malformations with clinical manifestations being similar to those of Jeune syndrome [38, 39]. For the reasons, many authors would advocate operative treatment for children older than 10 years [5, 12].

CONCLUSIONS

Chest wall malformations represent a wide spectrum of anomalies. Etiology and impact of genetic factors are still vague. The clear identification of specific anomalies and understanding the development and growth of the chest wall are primary fundamental steps in current approach to the treatment of the condition that would contribute to optimizing the choice of tactics to treat patients with chest wall deformities.

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Received:15.11.2016

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