Chest-deformities: a proposal for a classification

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Background: **In this article we assess the significance of classifying chest-deformities based on morphological findings in type-related treatment and its results.**

Data sources: **Recent publications on chest-deformities in children and youth were retrieved from PubMed and Medline and from our clinical and intraoperative findings.**

Results: **Chest-deformities are diagnosed by thorax-measurements using a flexible meter projected on a graph-paper by MR/CT investigations and color coded videorasterstereography. In addition an ultrasound guided mediastinal analysis is performed on the heart, the great vessels and mediastinal organs. These investigations could determine meticulously the morphology of the sternum, the sterno-costal segments and the costal arch, enabling to find different chest wall deformities, i.e., 11 different types. The clinical and surgical significance of such a classification can be shown by comparing postoperative results of non-classified chest-deformities with those of classified. Preoperatively non-classified chest-deformities often have postoperative asymmetric shapes, partial local recurrences, costal arch eversions and a platythorax. Such a classification can be used to analyze and predict so-called "secondary associated alterations" of the vertebral column or mediastinal organs.**

Conclusions: **Determining the specific type of a thorax deformity could be considered a type-related physiotherapy as conservative treatment or vacuum treatment and if surgery is indicated a type-related surgical correction can be performed. A type-related and adapted surgical correction can prevent subsequent mitral valve prolapse, recurrent infections, vertebral disturbances caused by kypho-scoliosis and increasing psychological irritation. Typing chest-deformities are an additional and essential help for the surgeon to perform specific surgical procedures: detorsion of the sternum, correction of the sterno-costal region, the costal arch bow and the kind of chest wall immobilization by metal struts. It can also compare the postoperative results more accurately in similar types of chest-deformities.**

World J Pediatr 2011;7(2):118-123

Key words: **chest-deformities;**
classification;
funnel chest

Review article

Introduction

Chest-deformities have an incidence of approximately 1:1000.[1] Genetic studies have proved a congenital origin.[2] Chest-deformities must not always be present at birth. They can develop gradually in the first 5 years of life.[3] Histological and electron microscopic investigations have shown an increased growth of cartilage at the sterno-costal junction.[4] Chondrocyts in this region have a hypoplastic structure causing a diminished stability of the sterno-costal region. Chest-deformities are seen in 4.3% of patients associated with 29 different syndromes such as Marfan syndrome or Jeune syndrome.

Since August 2010, 2926 patients have been operated on at the Pediatric Surgical University Hospitals in Münster and Erlangen in a period of 35 years (1975-2010). These patients accounted for 28% of all admitted patients with chest-deformities.

This review aims to find out whether chest-deformities differ in morphology and whether a derived classification is of value for surgery, physiotherapy and comparison of the results.

Diagnostic methods

Before the start of specific diagnostic procedures,[5,6] we speak to the parents about the history, the onset of symptoms, psychological alterations of the children and check the heart, lung and vertebral column.[7,8] In addition an ultrasound examination of the heart provides some information about the shape and location of the heart, the mitral and aortic valve and the configuration of the aortic arch (dilatation of the aorta).

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doi: 10.1007/s12519-011-0263-y

With a flexible meter, modelled to the anterior and posterior part of the thorax, the shape of chest deformity can be reproduced on a graph-paper. This non-radiation method can determine the depth and protusion of the deformity, the external sterno-vertebral distance as well as torsion of the sternum or eversion of the costal arch (Figs. 1, 2).[9]

1. MR/CT investigations are performed in a transverse and vertical direction. They can diagnose malformations of the sternum including the mediastinal shape of the sternum, the sterno-costal junction, compression and dislocation of the heart and the mediastinal organs. In addition, curvatures of the spine related to the chest deformity can be determined (Fig. 3).[10,11]

2. We have used color-coded videorasterstereography for 8 years to illustrate the shape of the anterior wall of the thorax color coded. With this method pre- and postoperative results are compared accurately (Fig. 4).

Types of chest-deformities

The above mentioned diagnostic methods are used to investigate chest-deformities accurately.[12]

In a 35-year period, we operated on 2926 children and adolescent patients between the age of 4 and 18 years with chest-deformities. Preoperative findings were found to be correlated with intraoperative findings concerning the different shapes of chest-deformities. The most reliable diagnostic methods were measurements of the chest using a flexible meter and MRI. Based on these measurements, we found 11 types of chest-deformities (Fig. 5).

Results

Typing chest-deformities enables to get further information about necessary operative steps, accurate findings of secondary alterations of the vertebral

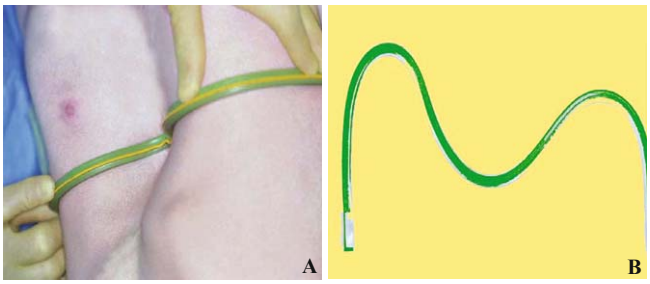


Fig. 1. With a flexible meter which is placed on the anterior side of the thorax (**A**), the shape of the deformity can be reproduced on a graph-paper (**B**). The external sterno-vertebral distance can be determined if the flexible meter is also placed on the posterior side of the thorax. The distance is compared with normal values. The size of protusion or the depth of the thorax can be expressed as a percentage of the normal findings age related.

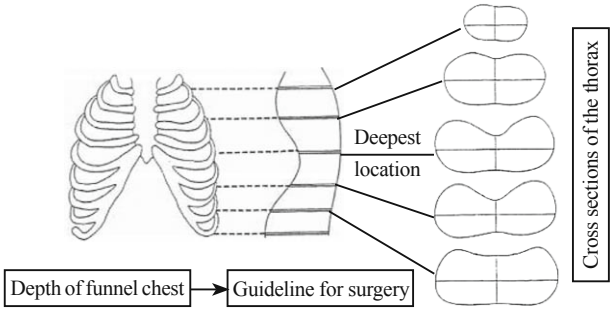


Fig. 2. Determination of chest deformities using a flexible meter.

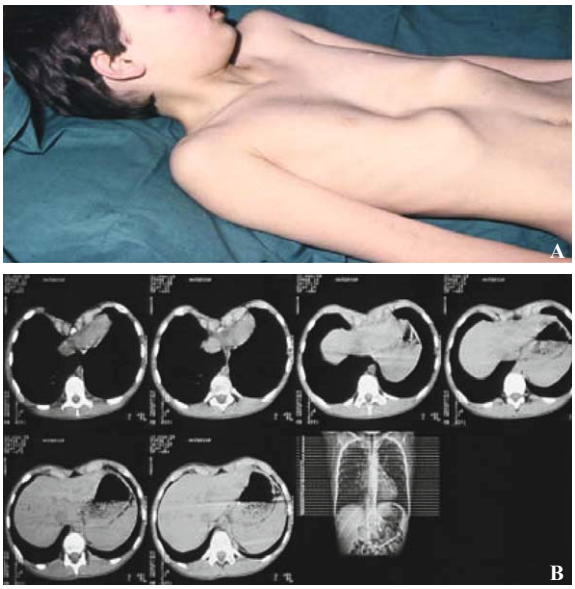


Fig. 3. Significance of thorax MR/CT-measurements: this is an MR-investigation in a 12 years old boy with a funnel chest (**A**). The depth was 45% of the normal sterno-vertebral distance. The funnel chest is symmetrically shaped. The sternum is not twisted. The heart is shifted to the left side and compressed by the sternum (**B**).

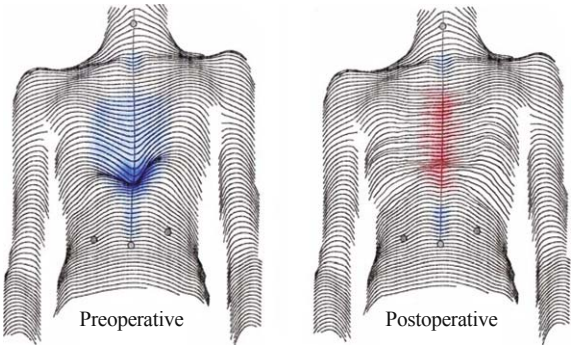
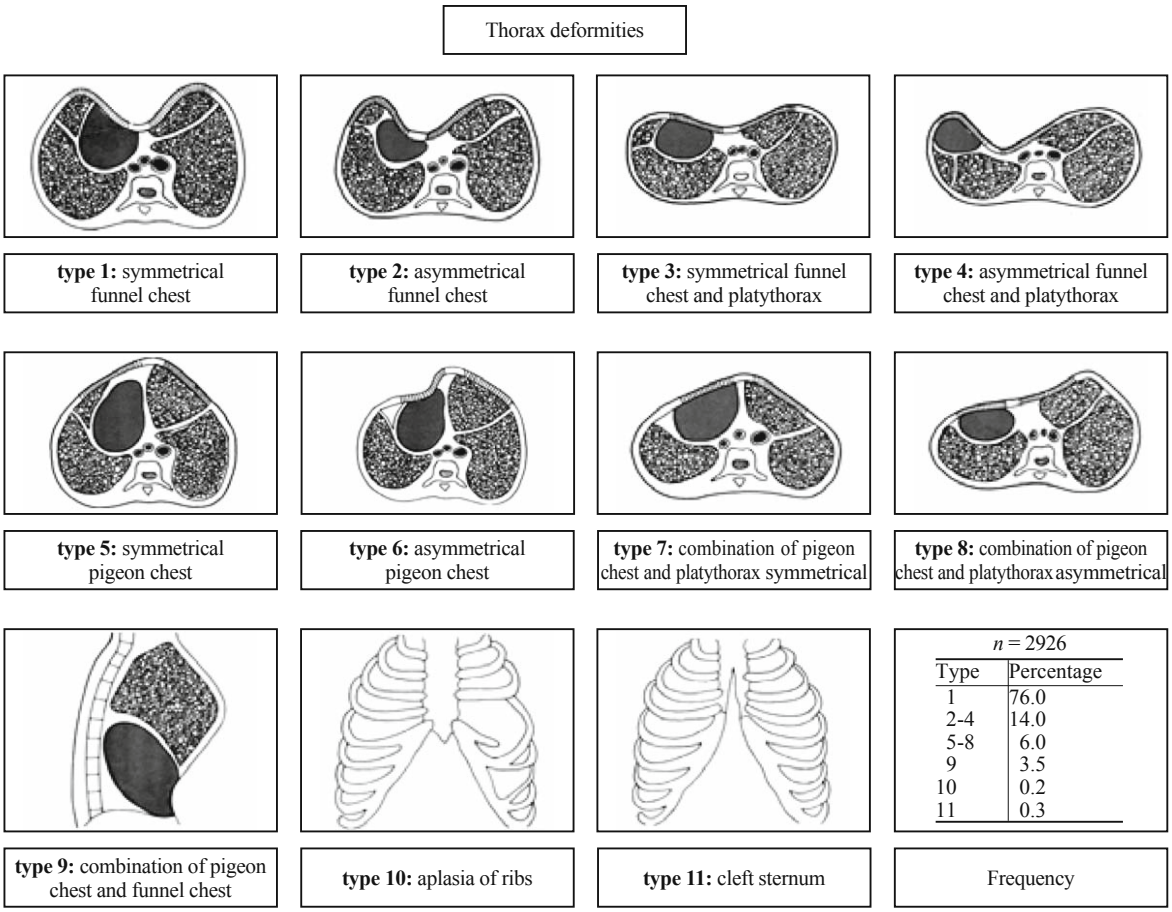


Fig. 4. The color-coded videorasterstereography enables an easy documentation of depths or protusions of the sternum, the ribs and shows hypoplasia of muscle layers. Pre- and postoperative findings can be compared with each other, and the outcome of physiotherapy can be ascertained easily by improvements of the muscle layers. It is also possible to documentate symmetric and asymmetric contures of the thorax. Depressions are in blue, protusion are in red color.



n = 2926

Type	Percentage
1	76.0
2-4	14.0
5-8	6.0
9	3.5
10	0.2
11	0.3

Fig. 5. Survey of different types of chest-deformities.

column and intrathoracic organs, and precise recommendations for postoperative physiotherapy for preventing recurrences (Table 1).[13-18] To evaluate the significance of such classification in chest-deformities, we compared postoperative results in patients of the same age, gender and surgical techniques used with those without use of this classification. Thus complications and extraordinary courses related to the surgical technique could be excluded (Table 2).[19]

Preoperative chest wall typing in patients with chest-deformities is helpful to differentiate surgical planning: detorsion, splitting and elevation of the sternum, immobilization of the anterior chest wall by one or additional metal struts, placed above, transverse or behind the sternum, single or multiple chondrotomies, correction of costal arch protrusions, and reimplantations of excized cartilage/bone covered by bio-absorbable patches.

Symmetrical funnel chests types 1-3 are found in 58% of costal arch protrusions. Therefore it is recommended to correct this costal arch protrusion by additional chondrotomies after the sternum is elevated in a normal anatomical position.

Asymmetrical funnel chests types 2-4 are found in 56% of costal arch protrusions of different dimensions or sternal torsion. They need a wedge osteotomy at the level of the third intercostal space and detorsion of the sternum, depending on the degree of the torsion. This operation can be done by either open surgery or minimal invasive surgery via a thoracoscopic procedure. In addition, the different levels of the costal arch protrusions should be corrected in a straight way, otherwise a pseudo funnel chest may occur.

In case of funnel chests with a platythorax (types 3 and 4) it is recommended not only to lift up the sternum in a normal position but also to elevate the sterno-costal junction and small costal segments.

In case of combination of pigeon chest and funnel chest (type 9), several surgical steps must be planned to avoid recurrences: elevation or subsiding of parts of the chest wall by multiple chondrotomies and multiple sternotomies.

For the surgeon, a preoperative classification of chest-deformities can help to plan the procedure more accurately in order to get better results. In other words, classifying chest-deformities enables to identify

Table 1. Significance of preoperative typing chest-deformities in planning the surgical procedure and in regard to postoperative results

Type	Description	Malformation sternum	Malformation sterno-costal junction	Malformation costal arch	Platy-thorax	Special points
Type 1 (45%)	Symmetrical funnel chest otherwise normal thorax	Sternal depression	Sterno-costal depression	Bilateral costal arch protrusions	No	Location of deepest sternal depression either in upper or middle or lower part
Type 2 (15%)	Asymmetrical funnel chest otherwise normal thorax	Sternal depression and sternal rotation	Sterno-costal depression and different steep walls of funnel	Bilateral costal arch prortusions at different levels	No	Location of deepest sternal depression either in upper or middle or lower part
Type 3 (22%)	Symmetrical funnel chest and platythorax	Sternal depression and flat thorax	Sterno-costal depression and flat thorax	Minimal costal arch protrusions bilateral	Yes	No
Type 4 (8%)	Asymmetrical funnel chest and platythorax	Sternal depression and sternal rotation and flat thorax	Sterno-costal depression and different steep walls of funnel, flat thorax	Minimal costal arch protrusion unilateral	Yes	No
Type 5 (2%)	Symmetrical pigeon chest, otherwise normal thorax	Sternal protrusion	Sterno-costal protrusion	No	No	Malformation starts at manubrium – sternal junction
Type 6 (1%)	Asymmetrical pigeon chest otherwise normal thorax	Sternal protrusion and sternal rotation	Sterno-costal protrusion and different steep walls of protrusion	No	No	Malformation starts at manubrium – sternal junction
Type 7 (2%)	Symmetrical pigeon chest and platythorax	Sternal protrusion and flat thorax	Sterno-costal protrusion and flat thorax	No	Yes	No
Type 8 (1%)	Asymmetrical pigeon chest and platythorax	Sternal protrusion and sternal rotation and flat thorax	Sterno-costal protrusion, different steep walls of protrusion, flat thorax	No	Yes	No
Type 9 (2%)	Combination of funnel chest and pigeon chest	Sternal depression and sternal protrusion	Sterno-costal protrusion and sterno-costal depression	No	No	Sternal malformation at different parts of the sternum
Type 10 (1%)	Aplasia of the ribs	No	No	No	No	Special surgical technique
Type 11 (1%)	Manubrium - sternal cleft	Sternal cleft	No	No	No	Special surgical technique

Table 2. Different postoperative results if preoperative chest-deformities have been classified

Results (1 y after operation)	Classifying chest-deformities regarding postoperative results	
	Not classified (n=43)	Classified (n=43)
Partial recurrence	7	2
Total recurrence	3	0
Costal arch protrusion	8	1
Asymmetrical surface	7	1
Parasternal protrusion	7	1
Sternal depression	6	0
Protrusion of ribs	5	0

morphological details of the chest wall accurately. This may be a useful indicator for surgical procedures to avoid local recurrences.[20]

In addition, such a classification proves to be useful for a differentiated physiotherapy. Patients with a funnel chest (types 1-4) in 87% have a poor posture with round-shaped shoulders and a curved abdominal wall due to costal arch protrusion in contrast to patients with a pigeon chest (types 5-8). Reasons for this are hypoplastic and weak muscles of the back, the shoulders and the abdominal wall. After reconstruction of the chest an intensive training of these muscles is necessary in order to avoid postoperative recurrences. It is important to speak with the parents and the patients before operation to plan postoperative physiotherapeutic treatments as part of the whole treatment.[21]

Furthermore, such a classification can be of diagnostic help regarding alterations of the vertebral column, the heart and the lung. Symmetrical chest-deformities (types 1, 3, 5, 7 and 9) can lead to kyphosis, whereas asymmetrical chest deformities (types 2, 4, 6 and 8) can lead to scoliosis of the vertebral column causing backache in youth and early adult life and later slip discs if no additional physiotherapeutic treatment is prescribed. These alterations of the vertebral column are described as secondary pathology of chest-deformities. If these secondary alterations are present it is recommended to train, after chest wall reconstruction, muscle layers of the back by a special physiotherapeutic therapy. Symmetrical funnel chests (type 1) can cause a compression of the heart, reduced heart beat volume, and functional disorders of the heart or mitral valve prolapse. They can also lead to a compression

of the trachea or of lung segments, documented by perfusionscintigraphy.[22]

Discussion

There are diseases in pediatric surgery, which have been investigated meticulously and in which a classification has been proposed for diagnostic or therapeutic purposes. Such diseases include esophageal atresia, anorectal anomalies, biliary atresia, diaphragmatic hernia and others. Chest wall deformities may be a disease in which different morphological types can be distinguished following morphological alterations of the sternum, the sterno-costal segments and the ribs. This classification describes accurately the morphological alterations of chest-deformities and provides information on possible secondary alterations of the heart, lung and vertebral column as well as recommendations for physiotherapy improving unilateral or bilateral deficiencies of muscle layers of the abdominal wall or back.[23]

Chest-deformities necessitate a preoperative plan for surgical reconstruction: location of chondrotomies, costal arch corrections, partial sternotomy or determination of the number, location and shape of metal struts. In our cases we compared preoperative morphological findings with intraoperative findings. Preoperative typing of chest-deformities helps to predict the prognosis of postoperative results: symmetrical funnel chests (type 1) have the best postoperative results and prognosis: a partial recurrence rate of 2.4% and a total recurrence rate of 0.8%, whereas asymmetric funnel chests with a plathythorax (type 4) have a partial recurrence rate of 4.2% and a total recurrence rate of 1.7%.

Further experience should be accumulated by a type-related conservative treatment of chest-deformities using physiotherapy.[24] Complications and extraordinary courses are internet-based and collected by the IDBEC (the International Database for Complications and Extraordinary Courses).[25-29]

Conclusions

Typing chest-deformities is advantageous for both patients and surgeons. Distinguishing between different types of chest-deformities enables the surgeons to plan the surgical procedure more accurately for detorsion of the sternum in asymmetric chest-deformities (types 2 and 4), correction of associated costal arch protusions for types 1 and 2, and physiotherapeutic treatment of muscle hypoplasia (type 4). Such a classification might be informative for surgeons to compare the results of different types of funnel chests with each other.

Funding: None.
Ethical approval: Not needed.
Competing interest: No benefits in any regard have been or will be received from any commercial party related directly or indirectly to the topic of this article.
Contributors: All authors contributed to the content and approved the final version.

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Received June 29, 2009
Accepted after revision September 27, 2010