ORIGINAL ARTICLE

UDC: 617.541:616.712/.713-007-053.2-071.3
https://doi.org/10.2298/VSP190430071K

How to assess chest wall deformity in children with *pectus excavatum* – evaluation of the agreement among methods

Kako izvršiti procenu deformiteta grudnog koša kod dece sa *pectus*-om *excavatum*-om – procena podudarnosti metoda

Marko Kostić*, Aleksandar Sretenović†, Milan Savić*, Marko Popović*, Sanja Kostić‡, Davor Stamenović§

Clinical Centre of Serbia, *Clinic for Thoracic Surgery, Belgrade, Serbia; †University Children's Hospital, Belgrade, Serbia; ‡University Hospital "Bežanijska Kosa", Belgrade, Serbia; §Clinic for Thoracic Surgery "ViDia Kliniken", Karlsruhe , Germany

Abstract

Background/Aim. *Pectus excavatum* (PE) is the most common deformity of the frontal aspect of the chest wall in children. A particular dilemma arises about the degree of deformity that should be subjected to surgical treatment. The aim of this study was to compare several morphological methods of evaluating the degree of deformity and determine the matching among them, as well as to determine the connection between the functional and morphological abnormalities of echocardiography. **Methods.** The study included 35 patients with PE, aged between 7 and 15 years. A noninvasive evaluation of chest deformity was carried out in all patients by photographic method (surrogate of clinical examination), native X-ray imaging, and computed tomography (CT), as well as by echocardiographic examination. **Results.** In our group of patients, males were more common (67.5%), as well as children with the Haller index (HI) > 3.5 [represented in most children (86.7%)]. A significant correlation of the index of the affected sternum segment (ASt) and the total length of the sternum (ASt/LSt) determined by the photographic method with that determined by the CT scan of the chest was established (p = 0.001). In addition, the correlation between HI, determined by the X-ray method and CT images was presented (p = 0.012). In contrast, despite the high frequency of echocardiographic abnormalities (69%), those were not mutually correlated with the degree of pronounced morphological deformities of the chest wall. **Conclusion.** A detailed clinical examination and photographic evaluation method, combined with the X-ray method, can determine the severity of deformity with a high degree of agreement with the CT chest findings. In this way, it is possible for children with PE, who are not candidates for surgical treatment, to be spared from repeated CT scans that are carried out in order to monitor the development of chest deformities with growth. Echocardiographic evaluation remains an integral part of the assessment of children with PE.

Key words:
child; echocardiography; funnel chest; methods; radiography; severity of illness index; tomography, x-ray computed.

Apstrakt

Uvod/Cilj. *Pectus excavatum* (PE) je najčešći deformitet frontalnog aspekta zida grudnog koša kod dece. Posebnu dilemu predstavlja stepen deformiteta koji treba da bude podvrgnut hirurškom lečenju. Cilj rada bio je da se uporedi više morfoloških metoda procene stepena deformiteta i utvrdi podudarnost između njih, kao i povezanost između funkcionalnih i morfoloških abnormalnosti na ehokardiografiji. **Metode.** Ispitivanjem je obuhvaćeno 35 bolesnika sa PE, uzrasta od 7 do 15 godina. Kod svih bolesnika je sprovedena neinvazivna procena deformiteta grudnog koša fotografskom metodom (surogat kliničkog pregleda), nativnom radiografijom (RTG) i kompjuterizovanom tomografijom (CT), kao i ehokardiografskim pregledom. **Rezultati.** U ispitanom uzorku češće je bio zastupljen muški pol (67,5%), kao i deca sa Halerovim indeksom (HI) > 3,5 (86,7% dece). U cilju procene podudarnosti metoda, utvrđena je značajna korelacija indeksa aficiranog segmenta sternuma (ASt) i ukupne dužine sternum LSt (ASt/LSt) određivanog na fotografiji sa onim koji je određivan na CT snimku grudnog koša (p = 0.001). Pored toga, pokazana je i korelacija između HI određenog putem RTG i CT snimka (p = 0.012). Suprotno navedenom, uprkos visokoj

Correspondence to: Marko Kostić, Clinical Centre of Serbia, Clinic for Thoracic Surgery, Koste Todorovića 26, 11 000 Belgrade, Serbia. E-mail: kostmarko@gmail.com

učestalosti ehokardiografskih abnormalnosti (69%) one nisu bile u korelaciji sa stepenom izraženosti morfoloških deformiteta zida grudnog koša. **Zaključak.** Detaljnim kliničkim pregledom i fotografskom metodom procene, zajedno sa RTG metodom, može se utvrditi težina deformiteta sa visokom podudarnošću u odnosu na CT grudnog koša. Na ovaj način, moguće je decu sa PE koja nisu kandidati za hirurško lečenje poštedeti od ponavljanja CT pregleda grudnog koša koji se sprovode u cilju praćenja razvoja deformiteta tokom rasta deteta. Ehokardiografska procena ostaje neophodni i sastavni deo procene stanja dece sa PE.

Ključne reči:
deca; ehokardiografija; pektus ekskavatum; metode; radiografija; bolest, indeks težine; tomografija, kompjuterizovana, rendgenska.

Introduction

Pectus excavatum (PE) is the most common deformity of the frontal aspect of the chest wall [1]. It is characteristic of the expressed depression of the sternum, as well as the lower costal cartilage, which disrupts the human figure and consequently leads to a person's withdrawal and development of complexes. With its complexity, PE deformity leaves its pressure and effects on the heart. It could also reduce the volume of the chest, which, as a consequence, has an impact on the respiratory system. Thus, the most common application for treatment of the deformity is surgical, which is the most acceptable solution for its correction [2].

The key problem in solving PE is in defining the morphology and severity of the deformity, after which it is necessary to determine precise indications for treatment and assess the risk.

The aim of this study was to describe the dysmorphia and anatomical deformity of PE, comparing the different diagnostic methods that are currently being applied.

Methods

Patients

Thirty-five patients with anatomical deformity of PE, referred to our Clinic in the period between March 2008 and March 2016, were included in the study regarding the evaluation aimed at the optimal approach to treat this deformity.

The study protocol was approved by the Ethics Committee of the University Children's Hospital – Tiršova, Belgrade, Serbia, and written informed consent was obtained prior to study engagement from each patient's legal representative (in all cases, they were parents or closest relatives).

The overall evaluation of this category of patients included history, physical examination, photographs, but also particularly important procedures of thoracic imaging – native chest X-ray, computed tomography (CT) chest scan, and echocardiographic examination [3].

Photographs were made by a digital camera at a distance of 1 m in order to indicate the median or sternal line of the chest, the vertical length of the affected sternum (ASt), the total vertical length of the sternum (LSt), and the vertical deformation length (LDEF) (Figure 1).

CT chest scans were conducted by Siemens Somatom Emotion 16-slice, with a mediastinal and bone window (on 5 mm display) with reconstructions and 3D display.

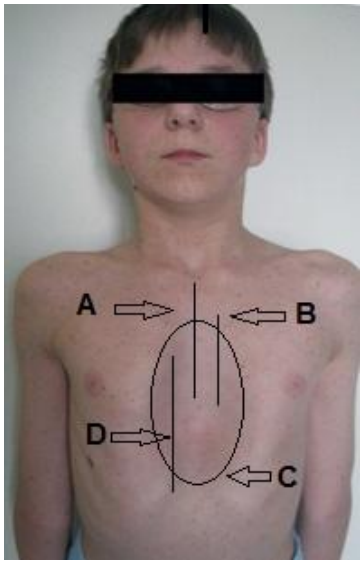
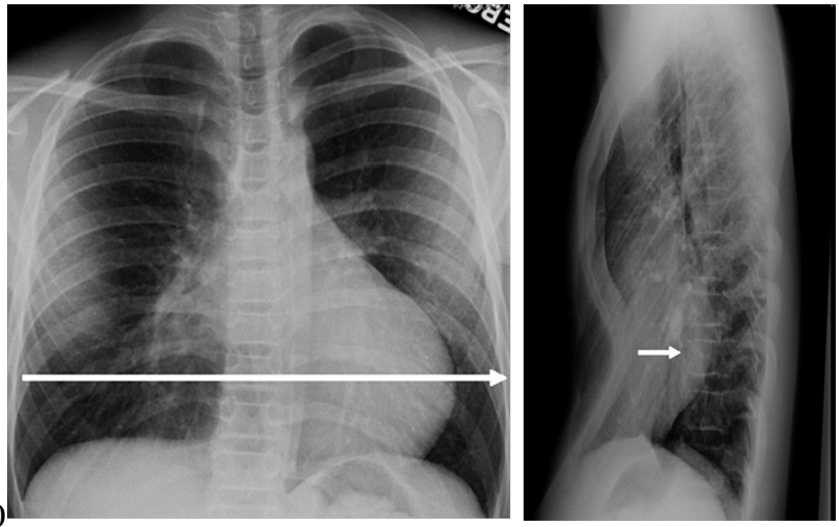


Fig. 1 – Photographic evaluation of chest wall deformity.
A - medial sternal line, length of sternum (LSt);
B - the length of the affected sternum (ASt)
C - a form of deformity; D - length of deformity (LDEF).

Echocardiographic examinations were performed by the Philips Sonos 7500. Standard echocardiographic techniques were applied, including two-dimensional echocardiography (2D mod), M-mode, and Color Doppler echocardiography. The echocardiographic analysis included the detection of any change in the morphology and function of the heart, particularly the valves, a type of syndrome prolapse, mitral valve dysplasia, mitral valve regurgitation, pulmonary artery dilatation truncus. The values of the longitudinal movements of the mitral annular (MAPSE) and the tricuspid valve (TAPSE) were also determined by the M-mode technique.

The description of dysmorphology in our patients was largely based on Cartoski et al. [4] (Nuss's group, Children's Hospital of King's Daughters, London, UK). Thus, the following indices were calculated: from photographs – ASt/LSt index = B/A × 100; symmetry index left = [L/(L+R) × 100]; symmetry index right = [R/(L+R) × 100]; ASt/LDEF index = B/D × 100; LSt/ LDEF index = A/D × 100). Then, from native X-ray (Figure 2) following indices were calculated: the Haller index (Hi) = T/A (right-side view); Hi = T/A (left-side view). And finally, from CT scans (Figure 3) following indices of sternal angle were calculated (at the level of the measured



A) **B)**

Fig. 2 – Native chest radiograph: illustrative case of a patient with *pectus excavatum*:
A) transversal diameter (arrow);
B) the shortest distance between the sternum and the front end of vertebrae (arrow).

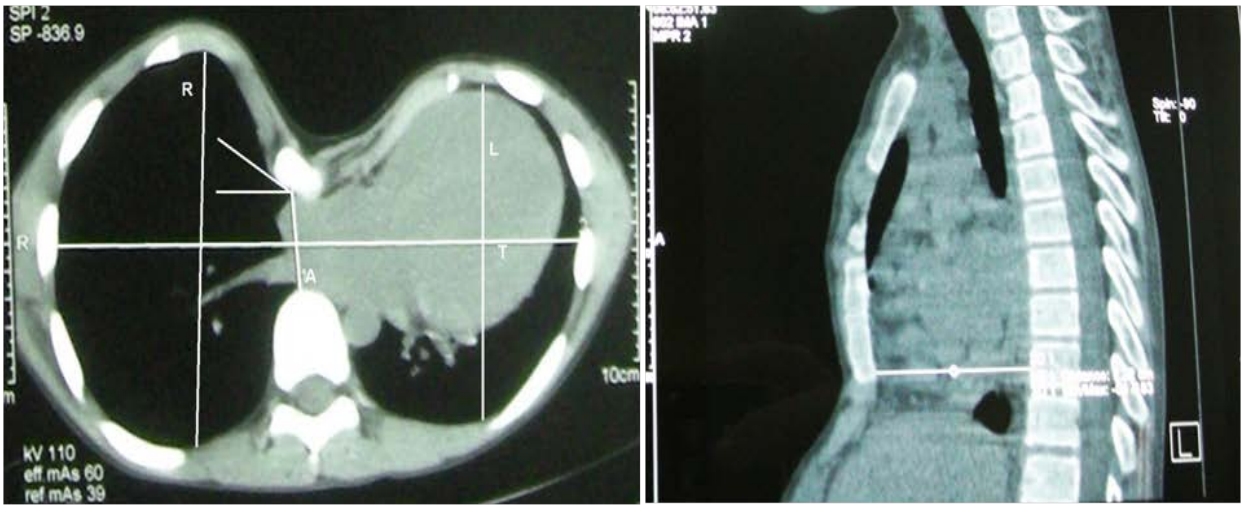


Fig. 3 – Computed tomography (CT) chest scan of a child with *pectus excavatus* deformity:
A - the shortest distance between the sternum and the front end of vertebra; T – transversal diameter Sternal angle; R – the largest distance of the right hemi-thorax in the anterior-posterior direction; L – the largest distance of the left hemi-thorax in the anterior-posterior direction; LSt - vertical length of sternum; ASt – length of affected sternum.

Hi): Hi = T/A; asymmetry index left = (L/R x 100); asymmetry index right = (R/L x 100); chest shape index (right-side view) = T/R x 100; chest shape index (left-side view) = T/L x 100; ASt/ LSt index.

The severity of pectus deformity was determined in accordance with the HI, according to which deformity levels are divided into: mild < 3.20; moderate: between 3.21 and 3.50; and severe: > 3.51. Normal HI considered values between 2.5–2.7 [5].

Concerning the degree of sternal torsion severity, which is usually graded as large, i.e., pronounced (> 30°) or small (<30°), we decided, based on the clinical judgment, to expand this division introducing the medium degree of this deviation (transitional form), in order to provide additional precision. Therefore, our division in this study implied three levels of sternal torsion severity: small (< 20°), medium (20–29°), as a transitional form, and large (> 30°).

Statistical analysis

Descriptive statistics were generated for all variables. The correlation of the findings with the use of different evaluation techniques was evaluated by the Pearson's correlation coefficient. The Student's *t*-test and Mann-Whitney *U*-test were used to test the differences when appropriate; significance level was set at the $p < 0.05$ level. The SPSS for Windows (release 16.0; SPSS, Chicago, IL, USA) was used to perform the statistical analysis.

Results

During the study, we evaluated 35 children with anatomical deformity of PE, at the age of 12.5 ± 2.4 years (range 7–15). According to age, in order to evaluate the growth effect on the development of PE deformities, patients

were further divided into age groups of 7–9 (19.4%), 10–12 (22.6%), and 13–15 (58.1%) years. In our sample, the male group was almost twice as big as the female group (65.7% males vs. 34.3% females).

Clinical and diagnostic characteristics of PE chest deformities are shown in Table 1.

Table 1

Clinical and diagnostic characteristics of *pectus excavatum* chest deformity

Parameter	Mean ± SD
Photographic method	
ASt/LSt index	47.11 ± 7.68
symmetry index R	48.34 ± 5.14
symmetry index L	51.62 ± 5.11
ASt/LDEF index	60.71 ± 9.56
LSt/LDEF index	131.44 ± 26.75
Radiographic method	
HI R	4.55 ± 1.30
HI L	4.47 ± 1.08
Computed tomography	
sternal angle	22.17 ± 11.74
HI	4.93 ± 1.69
asymmetry index R	101.43 ± 10.44
asymmetry index L	99.55 ± 9.99
chest shape index (right-side view)	176.31 ± 23.58
chest shape index (left-side view)	177.36 ± 17.56
ASt/LSt index	46.04 ± 12.50

ASt – affected sternum; LSt – length of sternum; L – left; R – right; LDEF – length of deformity; HI – Haller Index; SD – standard deviation.

According to the HI value, one patient (3.3%) had a mild degree of chest deformity (< 3.2), 3 patients (10%) had a moderate degree (3.2–3.5), and 26 patients (86.7%) had a severe degree of deformity (> 3.5).

In the present sample, we determined a significantly higher incidence of patients, 86.7% of subjects (p = 0.004), with a severe degree of chest wall deformity (according to the HI), which is the expected distribution for tertiary health centers.

Further analysis within the group of patients with the most severe degree of chest deformity (HI > 3.5) showed that most patients belong to the age group from 13–15 years (61.5%).

The angle of sternal torsion was changed in a similar way as the age-related increase of the HI, thus a high degree of rotation was observed in all subjects with the HI > 3.5 in the age group from 13–15 years.

Comparing various methods of evaluating PE deformities, it was found that the ASt/LsT index, determined by the photographic method, significantly correlated with the same index (r = 0.608, p = 0.001) when determined with CT.

Furthermore, considering the HI as the most widely accepted indicator of chest deformity, a consistent correlation was shown between the index determined by the X-ray method (right profile) and the one obtained by measuring by the CT scan (r = 0.528, p = 0.012) (Figure 4).

Similar correlation was established between the X-ray-determined HI (right profile) and ASt/LSt index measured by CT scan (r = 0.536, p = 0.012).

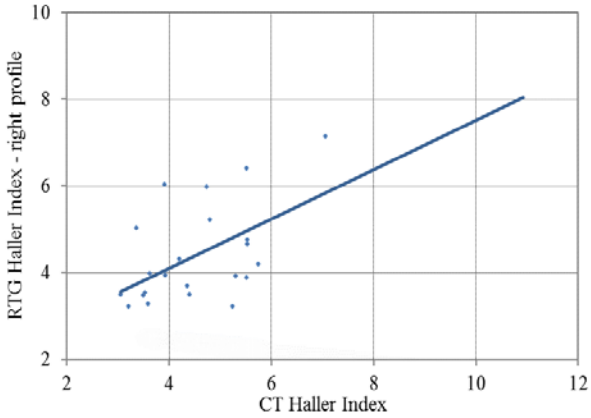


Fig. 4 – Line graph shows correlation between the Haller index measured by computed tomography (CT) scan and native radiography of the chest (right profile) in children with *pectus excavatus* deformity.

Since the asymmetry of chest cavity forms is a very common finding in children with PE deformity, defining the correlation is of particular importance. Therefore, if the irregularity of the chest cavity is considered, the right-side chest shape index (CT parameter) showed a high correlation with the asymmetry indices for the right (r = -0.618, p = 0.001) and left half (r = 0.696, p = 0.001) of the chest, CT measured as well (Figure 5).

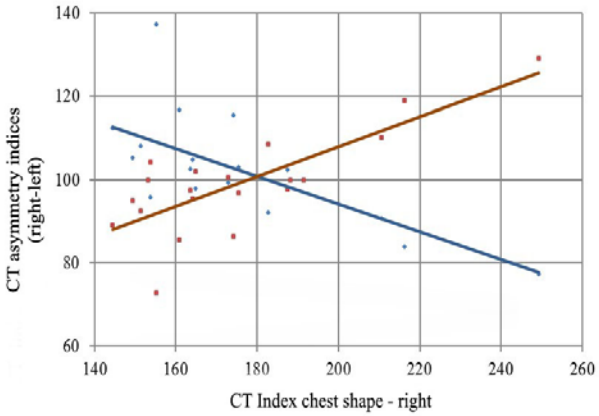


Fig. 5 – Line graphs show correlation of asymmetry indices and chest shape index (right profile blue, left profile red) in children with *pectus excavatum* deformity [both measured by computed tomography (CT) scan].

The right asymmetry indices were in a negative correlation, which means that the higher index value corresponded to the lower values of the right side of the chest shape index and *vice versa*. Contrary to that, the left asymmetry index was in a high positive correlation, which means that the higher index value corresponded to the higher values of the right side of the chest shape index.

Echocardiographic examinations in our sample of patients showed a high incidence of abnormalities (69%). The most common abnormalities were elongation, dysplasia, and mitral valve prolapse, with very frequent findings of

Table 2

Echocardiographic findings in children with *pectus excavatum* chest deformity

Variables	Children n (%)
Elongation of the mitral valve	14 (56.0)
Dysplasia of the mitral valve	6 (24.0)
Prolapse of the mitral valve	6 (25.0)
Combined prolapse and dysplasia of the mitral valve	8 (33.3)
Pressure	3 (12.5)
Regurgitation	11 (45.8)
Combined prolapse and dysplasia of the mitral valve with regurgitation	10 (41.7)

combined abnormalities (Table 2). Contrary to that, the least common disorder in our group of subjects was the pressure on the right ventricle or the atrial septum (12.5%).

Comparison of morphological abnormalities evaluated by different photographic or radiological parameters with functional findings defined by echosonography revealed the existence of only one relevant connection; both indices of asymmetry (photographically determined), right-sided ($r = -0.412$; $p = 0.045$) and left-sided ($r = 0.420$; $p = 0.040$), showed an association with the findings of a mitral valve prolapse in PE patients.

Discussion

The key findings of this study indicate that there is a significant correlation in the application of clinical assessment by the photographic method with radiological methods (X-ray and CT scan) of the chest dysmorphism in children with PE.

The distribution of the male sex in our sample was almost twice greater than the female sex (65.7% vs. 34.3%), which is common in this type of chest wall deformity.

In our study, the male sex was prevalent, but not with a statistically significantly higher incidence of PE. However, studies of other authors show that the incidence of this disease is higher (3– 5 times) in males [6–9], and according to some authors, even 80% of patients are male [10]. Therefore, the comparability of data is limited due to the specificity of our sample.

The chronological course of the disease shows that the symptoms are less pronounced in early childhood, but they increase with age [11]. The prenatal diagnosis of PE was seldom reported [12]. PE is more frequently present at birth, but in large series, only one-third of patients have deformities clearly expressed in early childhood [6, 13]. In most cases, greater deformation is observed in puberty, when otherwise there is a rapid increase.

In our patient sample, the correlation between the degree of PE deformity, expressed by the HI, concerning the subject's age did not show statistical significance, even though the HI in the majority of patients (16/18), in the oldest age group (13–15 years), was higher than 3.5. In contrast, the angle of rotation of the sternum showed a positive correlation with age in our sample of patients. All the remaining CT parameters applied in this study did not show a significant linear connection either with age or with the HI value, resulting in a dilemma about the significance of these indices in the preoperative assessment of patients.

However, as we already stated, the high degree of agreement of the HI measured on CT scan images and photographic analysis of radiographic images from the profile are even more important for the concept of this research.

In terms of the cut-off HI values from which the indication for operative treatment is derived, Kilda et al. [14] state that changes in the HI values, comparing the values before and after the surgery, were not observed if the preoperative value was 3.12. From this observation, they suggest that surgical interventions are conducted in children whose degree of deformity measured by the HI exceeds 3.1.

Furthermore, Potts [15] believes that the surgical attitude for surgical treatment of PE is somewhere between the two extremes, from having to operate on the vast majority of children with PE to not having to operate on any child at all. Patients selected for surgical treatment should have at least two or more defined criteria, which include the HI. Yoshida et al. [16] suggest that the progression of asymmetry on the right side occurs in children aged 10–12 years and that after 13 years, half of the children with PE have serious asymmetries. Based on these results, they decide upon the optimal period for performing the surgical treatment. However, several authors suggest that the time for surgery is problematic in younger children. They advocate that deformation correction should be carried out at a later stage of teenage growth, allowing the patient to complete growth and reduce the possibility of recurrence or damage [17–21]. Young children with severe cardiopulmonary symptoms may also be candidates for the implementation of an operative procedure. However, a corrective surgery at premature age may result in an inappropriate growth of the chest wall and other complications, including recurrence [22–24].

Many patients with PE have noticed decreased physical abilities, as well as minor chest pains, which is one of the indications for these major thoracic surgeries, for ensuring normal cardiac function [17, 25].

Cardiological examination of patients with PE is important because of the presence of a significant percentage of patients with right ventricular compression as well as mitral valve prolapse. In different studies, mitral valve prolapse was present in 17%–65% of patients, in contrast to the normal population where it was present in only 1% of patients [13, 26, 27]. Therefore, prolapse

of the mitral valve in PE cases could be a direct consequence of compression. Cardiac conduction disorders, such as first-degree atrioventricular block, right bundle branch block, and Wolff-Parkinson-White syndrome, were present in up to 16% of patients [28].

The majority of children in our study had some form of cardiac abnormality with the highest incidence of elongation, dysplasia, and prolapse of the mitral valve. In an attempt to determine the relationship between indicators of morphological abnormalities of the chest wall and echocardiographically defined disturbances, the only correlation was found between the index of symmetry obtained by the photographic method and the frequency of mitral valve prolapse.

Conclusion

According to our experience and the results of this study, it can be suggested that the optimal algorithm evaluation, that takes into account the rationality and reliability of diagnostic approaches, includes detailed clinical examination. The examination is further documented by standardized photographs of the chest, followed by either unavoidable native X-ray, while the CT scan is reserved for children for which the possibility of operative treatment is considered with a high level of suspicion. In our opinion, due to the high incidence of abnormal findings and because of the noninvasive examination, the echocardiographic examination should be an integral and indispensable part of the evaluation of this category of patients.

R E F E R E N C E S

1. *Shamberger RC.* Congenital chest wall deformities. Curr Probl Surg 1996; 33(6): 469–542.
2. *Abid I, Ewais MM, Marranca J, Jaroszewski DE.* Pectus Excavatum: A Review of Diagnosis and Current Treatment Options. J Am Osteopath Assoc 2017; 117(2): 106–13.
3. *Emil S.* Current Options for the Treatment of Pectus Carinatum: When to Brace and When to Operate? Eur J Pediatr Surg 2018; 28(4): 347–54.
4. *Cartoski MJ, Nuss D, Goretsky MJ, Proud VK, Croitoru DP, Gustin T,* et al. Classification of the dysmorphology of pectus excavatum. J Pediatr Surg 2006; 41(9): 1573–81.
5. *Daunt SW, Cohen JH, Miller SF.* Age-related normal ranges for the Haller index in children. Pediatr Radiol 2004; 34(4): 326–30.
6. *Blanco FC, Elliott ST, Sandler AD.* Management of congenital chest wall deformities. Semin Plast Surg 2011; 25(1): 107–16.
7. *Fokin AA, Steuerwald N, Ahrens WA, Allen KE.* Anatomical, histologic, and genetic characteristics of congenital chest wall deformities. Semin Thorac Cardiovasc Surg 2009; 21(1): 44–57.
8. *Park JM, Varma SK.* Pectus excavatum in children: diagnostic significance for mitral valve prolapse. Indian J Pediatr 1990; 57(2): 219–22.
9. *Sabiston JH Jr.* Congenital deformities of the chest wall. In: *Sabiston JH*, editor. Textbook of Surgery. Philadelphia: WB Sauders Co.; 1997. p. 1888–96.
10. *Nowak H.* Dieerblische Trichterbrust. Duetsche Med Wchnschr 1936; 62: 2003. (German)
11. *Jaroszewski DE, Fonkalsrud EW.* Repair of pectus chest deformities in 320 adult patients: 21 year experce. Ann Thorac Surg 2007; 84(2): 429-33.
12. *Salamanca A, Girona A, Padilla MC, Sabatel RM, Gonzales-Gomez F.* Prenatal diagnosis of pectus excavatum and its relation to Down's syndrome. Ultrasound Obstet Gynecol 1992; 2(6): 446–7.
13. *Shamberger RC, Welch KJ, Sanders SP.* Mitral valve prolapse asso-ciated with pectus excavatum. J Pediatr 1987; 111(3): 404–7.
14. *Kilda A, Basevicius A, Barauskas V, Lukosevicius S, Ragaisis D.* Radiological assessment of children with pectus excavatum. Ind J Pediatr 2007; 74 (2): 143–7.
15. *Potts WJ.* The Surgeon and the Child. Philadelphia, PA: W.B.Saunders, 1959.
16. *Yoshida A, Uemura S, Yamamoto M, Nouso H, Kuyama H, Muta Y.* Correlation of asymmetric chest wall deformity and growth in patients with pectus excavatum, J Pediatr Surg 2013; 48(4): 771–5.
17. *Kelly RE Jr.* Pectus excavatum: historical background, clinical picture, preoperative evaluation and criteria for operation. Semin Pediatr Surg 2008; 17(3): 182–93.
18. *Fonkalsrud EW.* Current management of pectus excavatum. World J Surg 2003; 27(5):502–8.
19. Colombani PM. Preoperative assessment of chest wall deformities. Semin Thorac Cardiovasc Surg 2009; 21(1): 58–63.
20. *Davis JT, Weinstein S.* Repair of the pectus deformity: results of the Ravitch approach in the current era. Ann Thorac Surg 2004; 78(2): 421–6.
21. *Nuss D, Kuhn M.* Our approach: minimally invasive surgical repair of pectus excavatum. Contemp Surg 2007; 63: 444–51.
22. *Haller JA Jr, Colombani PM, Humphries CT, Azizkhan RG, Loughlin GM.* Chest wall constriction after too extensive and too early operations for pectus excavatum. Ann Thorac Surg 1996; 61(6): 1618–24; discussion 1625.
23. *Kim DH, Hwang JJ, Lee MK, Paik HC.* Analysis of the Nuss procedure for pectus excavatum in different age groups. Ann Thorac Surg 2005; 80(3): 1073–7.
24. *Pilegaard HK, Licht PB.* Routine use of minimally invasive surgery for pectus excavatum in adults. Ann Thorac Surg 2008; 86(3): 952–6.
25. *Lawson ML, Cash TF, Akers R, Vasser E, Burke B, Tabangin M,* et al. A pilot study of the impact of surgical repair on disease-specific quality of life among patients with pectus excavatum. J Pediatr Surg 2003; 38(6): 916–8.
26. *Saint-Mezard G, Duret JC, Chanudet X, Larrue J, Bonnet J, Bricaud H.* Mitral valve prolapse and pectus excavatum. Fortuitous association or syndrome? Presse Med 1986; 15(9): 439. (French)
27. *Warth DC, King ME, Cohen JM, Tesoriero VL, Marcus E, Weyman AE.* Prevalence of mitral valve prolapse in normal children. J Am Coll Cardiol 1985; 5(5): 1173–7.
28. *Nuss D, Croitoru DP, Kelly RE.* Congenital chest wall deformities. In: *Ashcraft KW, Holcomp GW 3rd, Murphy JP*, editors. Pediatric Surgery. 4th ed. Philadelphia, PA: Elsevier Saunders, 2005. p. 245–63.

Received on April 30, 2019.
Accepted on May 29, 2019.
Online First June, 2019.