Current status of surgical treatment for pectus excavatum deformity.

Christophoros N. Foroulis, MD, FETCS, Kyriakos Anastasiadis, MD, FETCS, Nikolaos Charokopos, MD, George Missias, MD, Christos Papakonstantinou, MD

Aristotle University of Thessaloniki Medical School, AHEPA University Hospital, Department of Thoracic and Cardiovascular Surgery

ABSTRACT: Pectus excavatum is the commonest anterior chest wall deformity which is the result of overgrowth and elongation of costal cartilages that push the normal sternum in. The deformity has serious pshychologic impact and limits the exercise tolerance of the patient, because of displacement of the heart within the left hemithorax.
Repair of pectus excavatum is performed by the classical Ravich technique or the minimally invasive Nuss repair. The Ravich operation consists of resection of the deformed cartilages, bending the sternum in its normal position and maintenance of the sternum in the correct position by using a metal bar or a strong synthetic mesh. The Nuss procedure involves the thoracoscopic placement of a pre-formed curved metal bar behind the sternum which rises the sternum to the desired position. The selection of the technique depends on the age of the patient and the degree and symmetry of the deformity.
The Nuss technique is suitable for children between 5 and 15 years old who have flexible chest wall and symmetric deformities, while the Ravich procedure is indicated in young adults and asymmetric forms. Indeed, the Nuss technique still undergoes modifications to become safer and simpler and further it needs expertise and special equipment.

Key Words: Anterior chest wall deformities, Pectus excavatum, Ravitch repair, Nuss procedure, Minimally invasive pectus Excavatum repair.

INTRODUCTION

Chest wall deformities include a spectrum of malformations, such as pectus excavatum, pectus carinatum, Polland's syndrome, sternal clefts with or without ectopia cordis and pentalogy of Cantrell. Pectus excavatum accounts for 90% of all anterior chest wall deformities and it is by far the commonest deformity. The reported incidence of pectus excavatum varies from 1:400 to 1:200 live births and the male to female ratio is 3-4:1[1,2]. The deformity is uncommon in blacks[1,2,3]. Forty percent of patients with pectus excavatum have another family member with a chest wall deformity[3].

The etiology of pectus excavatum and pectus carinatum is unknown. Abnormalities of the connective tissue properties of the costal cartilages which result in overgrowth and elongation of the costal cartilages are considered to be the main responsible factor, while the ribs and sternum are normal[2-4]. According to the mechanical model that appears in Figure 1, if the elongated costal cartilages push the sternum in a pectus excavatum deformity is created while if they push the sternum out the result is a pectus carinatum deformity[2].

IMPACT OF PECTUS EXCAVATUM DEFORMITY ON DAILY LIFE AND ASSOCIATED CONDITIONS

Pectus excavatum deformity has serious psychologic impact and seriously affects the personality and social life of patients. Persons have poor self image, are very sensitive and tend to avoid sport activities and especially swimming, in order to hide the deformity from

Corresponding author: Christophoros N. Foroulis, AHEPA University Hospital, Department of Thoracic and Cardiovascular Surgery, 1 Stilponos Kiriakidi Street, 54636 Thessaloniki, Greece, Tel.: +30 2310 994705, Fax: +30 2310 994871, e-mail: foroulis@med.auth.gr

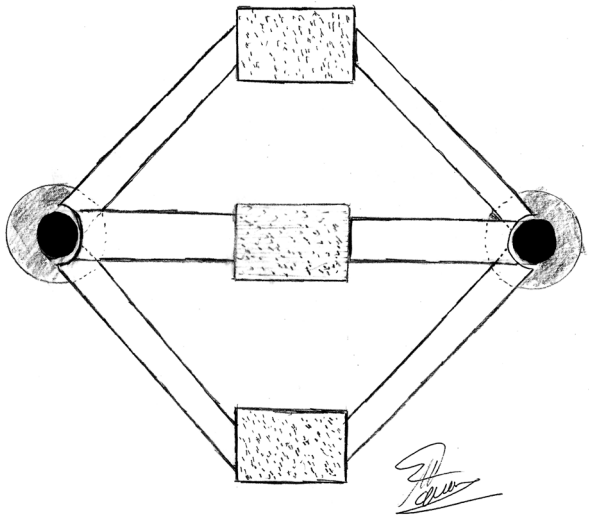

Figure 1. The mechanical model that represents pectus deformities.

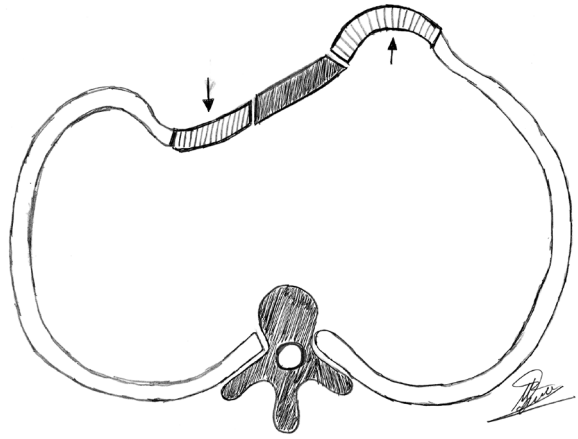

Figure 2. Combined pectus excavatum and carinatum deformity (mixed form). Note the rotation of the sternum toward the depressed side.

their peers[1-4]. In most cases the adverse psychological impact of the deformity in patient's social life is the "driving force" behind the strong request for repair pectus excavatum[4].

The main symptoms in severe pectus excavatum deformities are chest pain, shortness of breath and exercise intolerance[1-4]. Restrictive ventilatory pattern and ventilation/perfusion mismatch may be observed[2-4]. However, most of the studies in the past failed to demonstrate consistent improvement of pulmonary function after the repair of the deformity[5]. A reduction in stroke volume and cardiac output is detected in some studies because of the displacement of the heart within the left hemithorax and because of the compression of the right ventricular outflow tract by the depressed sternum which can also produce a functional systolic murmur[1-4].

Pectus deformities are frequently associated with scoliosis (20%-65%), mitral valve prolapse (15%-60%), Marfan syndrome (2%), asthma and chronic bronchitis[1-4]. The incidence of congenital heart disease among patients with pectus deformity is not different from that observed in the general population[4].

CLASSIFICATION OF PECTUS EXCAVATUM DEFORMITY

Pectus deformity is classified in 3 forms, according to Chin EF:[6]

(1) Local, narrow and symmetric form, where the sternal depression is mainly situated in the lower part of the sternum
(2) Broad and symmetric form, where the sternal depression is extended from the manubrium down to the xiphoid process
(3) Asymmetric form

From a practical point of view, pectus deformity is classified in symmetric and asymmetric forms. Asymmetric forms and mixed forms which represent a combination of carinatum and excavatum deformity are complex forms that need expertise for their optimal repair. In asymmetric and mixed forms the sternum is rotated toward the depressed side that is usually the right side (Figure 2)[2,3].

Pectus deformity becomes evident since the age of two years and worsens during the periods of rapid growth which for boys is between the age of 12 and 15 years[2,4]. The severity of the pectus excavatum deformity is objectively estimated by anthropometric indexes, such as the Haller index measured on chest

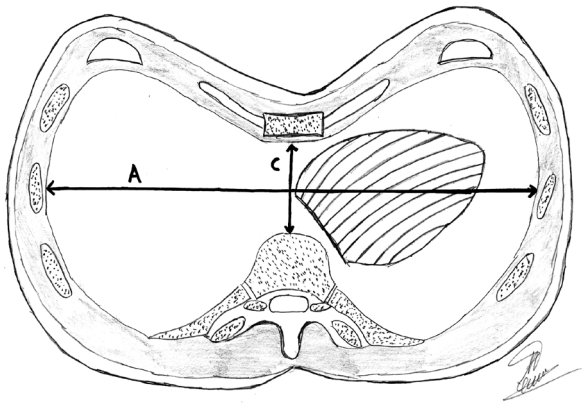

Figure 3. The way to calculate the Haller index from chest CT scan images. Haller index = A/C.

CT scan and the Lower Vertebral Index measured on lateral plain chest radiography. The Haller index is the commonly used anthropometric index for the estimation of the severity of a pectus excavatum deformity. (Figure 3)[7-9].

CONCERNS ON SURGICAL REPAIR OF PECTUS DEFORMITY

The decision to proceed with surgical repair is based on the answer to the following questions:

Which is the severity of the deformity and which is the degree of physiologic and psychological embarrassment?

Which is the optimal age for repair if needed?

Which is the appropriate technique of repair in each individual? The 3 available options today are the classic Ravitch repair, the minimally invasive pectus excavatum repair (MIPER or Nuss procedure) and the prosthetic reconstruction of the deformity.

The optimal age for repair still remains debatable. Early pectus repair, between 2 and 5 years, is proposed by some experienced pediatric surgeons in the past and the main argument supporting early repair is to avoid the expected adverse psychological impact of the deformity during clildhood and adolescence[2]. However, increase concern exists today about long term recurrence rate and the future impairment of growth of the chest wall if the Ravitch technique will be used for repair in this early age[1,3]. Surgery during adolescence and especially around the age of 15 years is considered today to be the optimal age for repair. The chest wall has less remaining growth after the age of 15 and the recurrence rate of the deformity is considered to be very low[1,2]. Repair after the age of 18 is also an acceptable option with good results[1]. The repair that is performed between 18 and 22 years has minimal risk for recurrence and is therefore considered to be a permanent repair. Indeed, the recently widely accepted, attractive Minimally Invasive Pectus Excavatum Repair (MIPER/Nuss procedure) should ideally be offered in children between 5 and 12 years old who have "flexible" chest[1,2,10,11].

All patients should undergo a complete medical evaluation to determine the potential effects of pectus excavatum on their heart and lung function or to discover any other associated cardiopulmonary abnormality. Preoperative work-up should routinely include spirometry, arterial blood gas or pulse oximetry, chest radiography and chest CT scan, electrocardiogram and Doppler echocardiography[1,2].

The two principles of the repair are to correct the sternal depression and to maintain the sternum in the corrected position[1-3]. Repair is usually performed by either the classical **Ravich repair** or the minimally invasive **Nuss repair**.

The basic principles of the **Ravich repair** are the bilateral resection of the deformed costal cartilages that is followed by a transverse osteotomy at the beginning of the sternal concavity which permits to bend the sternum anteriorly using a posterior support (metal bar or a strong synthetic mesh)[2,3,12]. Our team prefers the e-PTFE mesh as posterior sternal support because the patient will avoid the reoperation after 2 or 3 years to remove the bar and because this material is strong and resistant to infection[13]. Any associated sternal tilting in asymmetric or mixed forms can be corrected by a reversed "Z" wire. (Figure 4) Both pectoralis major muscles are mobilized at the beginning of the Ravich procedure to expose the sternum and the deformed costal cartilages. The pectoralis major muscles are re-approximated at the end of the procedure in the midline, offering that way a very good cosmetic result[1,2,4,12].

The observed complications of the Ravich repair for pectus deformities are atelectasis, pneumothorax (6%), seromas (5%), displacement of the supporting metal bar, mild pectus recurrences (<5%) as the result

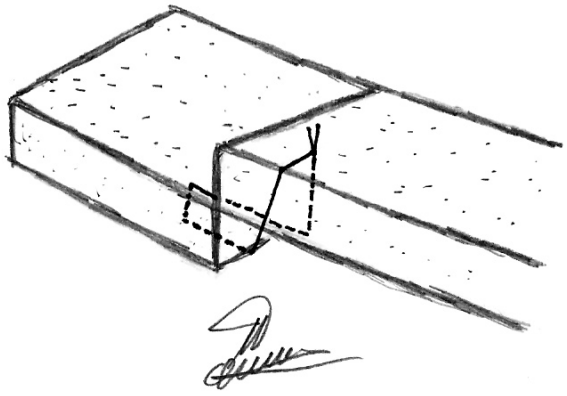

Figure 4. Correction of the tilted sternum by a reversed "Z" wire.

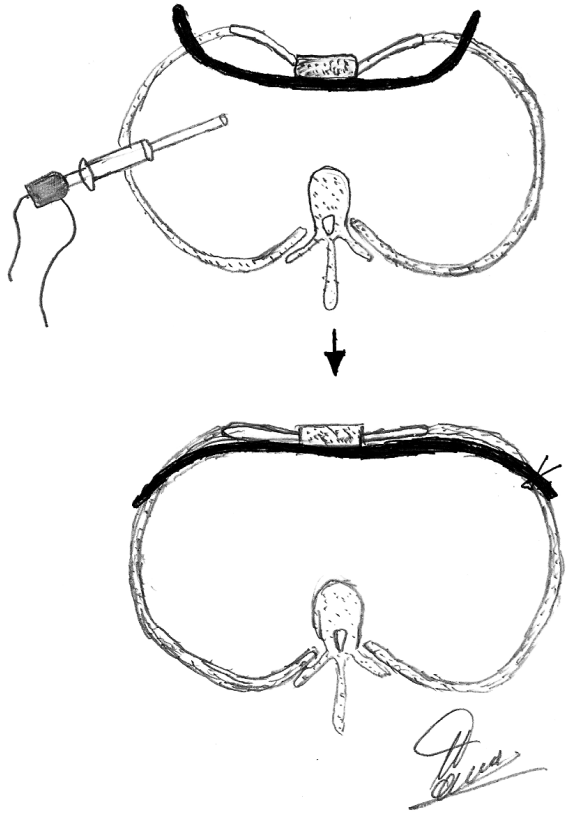

Figure 5. The minimally invasive Nuss procedure.

of inadequate initial repair or of rapid growth of the chest during adolescence and restriction of the chest wall (acquired Jeune's syndrome) that is mainly observed after a too early (between 2 and 4 years old) and too extensive repair of the deformity[1,2,4,14]. The cosmetic result of the Ravich repair is very good in most instances while the repair significantly improves cardiovascular function, but nor pulmonary function[5].

The **minimally invasive endoscopic Nuss procedure** is currently the state-of-the-art thoracoscopic surgical repair for pectus excavatum and it is the best option for most of the patients[10]. The Nuss procedure avoids cartilage resection and transverse sternal osteotomy. A curved preformed stabilizing metal bar is placed under the sternum just below the point of the maximal depression. The metal bar is advanced below the sternum with the convex facing the sternum until reaching the contralateral chest wall under thoracoscopic guidance. The bar is advanced through a tunnel created at the beginning of the operation by an introducer (long clamp). The bar is then forcefully turned over and its convex will face now the mediastinum, rising that way the sternum to the desired position. (Figure 5) The whole procedure is accomplished trough 2 small incisions in the lateral chest wall on both sides under thoracoscopic guidance[1,4,10,15]. Recently the titanium bar represents an excellent material for posterior sternal support in Nuss procedure which is resistant to infection and corrosion within a biological environment and which has also extraordinary tissue biocompatibility. Additional advantages of the titanium bars are that they do not respond to metal detectors and permits the performance of MRI examinations[16]. Unfortunately this material is at the moment unavailable in the Greek market.The advantages of the Nuss procedure are the minimal invasiveness of the procedure, the reduced operating time and the minimal blood loss[1,4,10,15].

A quite good correction of pectus excavatum deformity can be achieved with the Nuss procedure in symmetric forms of the deformity. Indeed, the Nuss technique has certain limitations such as any associated inferior costal flaring that precludes a good cosmetic correction, the asymmetry (> 15^{o}) of the deformity or mixed forms of the deformity, the thickening of the costal cartilages and the rigid chest together with a

strong musculature in young adults and extreme sternal depression, where the depressed sternum reaches the spine, that makes the Nuss procedure technically difficult and dangerous. In addition the Nuss procedure needs expertise in thoracoscopic surgery and special equipment to achieve the optimal result.

The main reported complications of the Nuss procedure are bar displacement requiring revision (4%-8%), infectious complications (2%) and pleural effusions (2%). Rare complications are reactive pericarditis (0.5%), cardiac injury during passage of the introducer behind the sternum (< 0.5%), damage to the internal thoracic artery or erosion of the artery and pseudoaneurysm formation by the metal bar (< 0.5%) and sternal erosion by the pressure applied by the bar (< 0.5%)[10]. Overcorrection of the deformity and reactive postoperative pectus carinatum deformity are also reported and represent failure of surgery to achieve the optimal repair or the later further overgrowth of costal cartilages, especially in Marfan patients[17]. Of importance is also the fact that as the experience ongoing, the Nuss procedure undergoes continuously modifications in order to become safer and to ameliorate the rate of existing complications.

Prosthetic reconstruction is an alternative option for mild pectus excavatum deformities. (Figure 6) The implants that are used to fill the space between the depressed sternum and the skin are custom-made from information obtained on helical CT scan of the chest supported by special technology, the Computer Assisted Design and Computer Assisted Milling (CADCAM)[18,19]. Indeed, the use of subcutaneous implants (from silicone or silastic or polyethylene) to fill pectus excavatum deformity has no physiologic benefit for the patient and has also the danger of migration of the prosthetic material causing further cosmetic deformity[1].

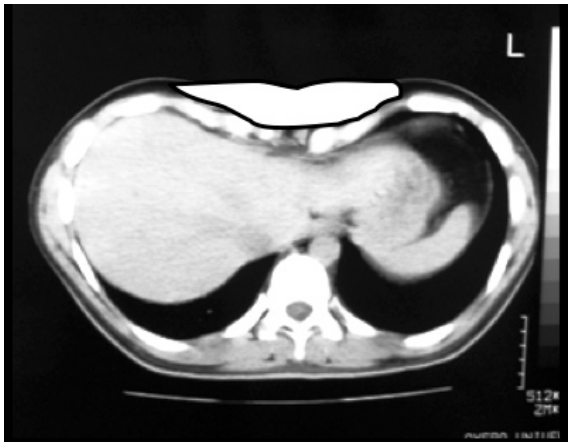

Figure 6. Prosthetic reconstruction (silicone implant) of pectus excavatum deformity.

SELECTION OF THE OPTIMAL APPROACH TO REPAIR A PECTUS EXCAVATUM DEFORMITY

A logical approach when we are going to decide on which is the appropriate technique to repair a pectus deformity in each case individually seems to be the following:

a) The Nuss operation is highly recommended in the paediatric population because of the flexibility of the chest wall, moreover the technique should be applied only by surgeons who have good skills in thoracoscopy and experience with the procedure.
b) In adolescents, although the complication rate is higher than in the paediatric group, the incidence of late complications such as bar displacement and persistent pain is acceptable.
c) In adults, however, the incidence of reoperation due to bar displacement, of chronic pain and of stabilizer-related wound infection is high and therefore, the Nuss procedure is not recommended for correction of pectus excavatum. In addition, in asymmetric and complex forms of the deformity the Nuss procedure is not recommended because the repair will be suboptimal.
d) The Ravich repair is applicable for patients of all ages and for all types of pectus deformity including complex forms and recurrent deformities.
e) Prosthetic reconstruction with subcutaneous implants is an acceptable option for mild, symmetric pectus deformities without physiologic embarrassment.

ILLUSTRATIVE CASE

A 21-year old man was presented with a mixed excavatum and carinatum deformity of the anterior chest wall. (Figure 7) The patient did not experience any exercise intolerance and the results of pulmonary function tests were normal. Doppler echocardiography did not reveal mitral valve prolapse or compression of the

Figure 7. Mixed pectus deformity - preoperative image (From AHEPA University Hospital, Department of Cardiothoracic Surgery).

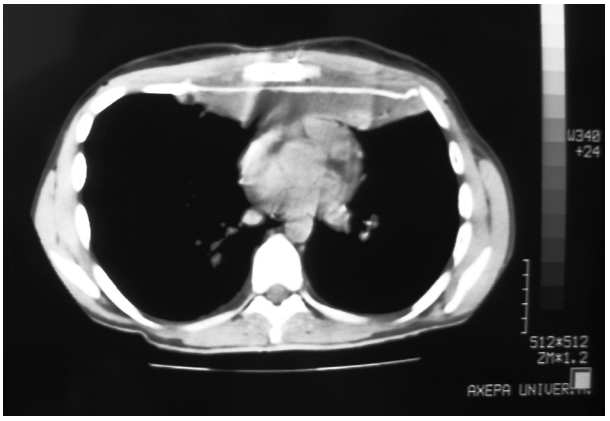

Figure 8a. Ravitch procedure: CT scan performed the 7th postoperative day - posterior sternal support with e-PTFE dual-mesh. (From AHEPA University Hospital, Department of Cardiothoracic Surgery).

right ventricular outflow tract. The Haller index measured in chest CT scan was 4.1. The deformity caused a serious psychologic embarassement and the patient asked for surgical correction of the deformity.

The patient underwent surgical correction of his mixed excavatum and carinatum deformity by the Ravitch technique. A dual-mesh e-PTFE mesh, 2mm thick was used for posterior sternal support after bilateral removal of the deformed 3rd to 10th costal cartilages and correction of the sternal tilting by two reverse "Z" wires just below the lower edge of manubrium.

The immediate postoperative (7th postoperative day) and the late postoperative result (3 months later) of the repair were very good. (Figures 8a and 8b) The patient currently follows an exercise program to strength and to enlarge the musculature of the chest wall for further improvement of his body image.

Figure 8b. The 3-month postoperative image of the patient: excellent final result of the Ravitch repair.

CONCLUSIONS

Surgical repair of pectus excavatum can significantly improve the body image, the associated with body image psychologic embarrassement of the patient and any limitation on physical activity experienced by the patient. Surgical repair for pectus excavatum significantly improves cardiovascular function, but not pulmonary function. Repair of severe forms of the deformity improves to a variable degree the exercise tolerance of the patient.

All the available techniques (Ravitch and Nuss procedures, prosthetic reconstruction) can be used. Indeed, the selection of the proper technique for each individual will be based on the morphology, the severity of the deformity and the age of the patient. The minimally invasive Nuss repair is very attractive however one's should always keep in mind the limitations and complications of the technique in order to apply an optimal, permanent and uncomplicated repair for pectus excavatum deformity.

Η σύγχρονη θέση της χειρουργικής θεραπείας για το σκαφοειδή θώρακα (pectus excavatum).

Χριστόφορος Ν. Φορούλης, Κυριάκος Αναστασιάδης, Νικόλαος Χαροκόπος, Γεώργιος Μίσσιας, Χρήστος Παπακωνστατίνου

Αριστοτέλειο Πανεπιστήμιο Θεσσαλονίκης, Ιατρική Σχολή, Πανεπιστημιακό Νοσοκομείο ΑΧΕΠΑ, Κλινική Χειρουργικής Θώρακα-Καρδιάς και Μεγάλων Αγγείων

ΠΕΡΙΛΗΨΗ: Ο σκαφοειδής θώρακας αποτελεί τη συχνότερη συγγενή δυσμορφία του προσθίου θωρακικού τοιχώματος και οφείλεται σε ανωμαλία και επιμήκυνση των στερνοπλευρικών χόνδρων, που εμβυθίζουν το φυσιολογικό στέρνο προς τη σπονδυλική στήλη. Η δυσμορφία έχει σημαντικές ψυχολογικές επιπτώσεις, ενώ σε σοβαρές μορφές υπάρχει περιορισμός της ικανότητας για άσκηση, λόγω μετατόπισης της καρδιάς εντός του αριστερού ημιθωρακίου και ελάττωσης του όγκου παλμού.

Η χειρουργική διόρθωση του σκαφοειδούς στέρνου γίνεται με την κλασσική τεχνική Ravitch, που συνίσταται στην εκτομή των μη φυσιολογικών στερνοπλευρικών χόνδρων, την ανάταξη του στέρνου και τη συγκράτησή του στην αναταγμένη θέση με οπίσθια υποστήριξη με μεταλλική ράβδο ή συνθετικό πλέγμα και τη νεότερη ενδοσκοπική τεχνική Nuss, που συνίσταται στη θωρακοσκοπική τοποθέτηση κατάλληλα διαμορφωμένης, κυρτής μεταλλικής ράβδου όπισθεν του στέρνου, που ανατάσσει με την πίεση που ασκεί το εμβυθισμένο στέρνο. Η τεχνική της διόρθωσης εξαρτάται από παραμέτρους όπως η ηλικία που γίνεται η διόρθωση, η βαρύτητα και συμμετρία της δυσμορφίας και η εμπειρία.

Η τεχνική Nuss θεωρείται ιδανική για τη διόρθωση της δυσμορφίας σε παιδιά ηλικίας 5-15 ετών που έχουν εύκαμπτο θωρακικό τοίχωμα και συμμετρική δυσμορφία, ενώ η τεχνική Ravitch ενδείκνυται σε νεαρούς ενήλικες και σε ασύμμετρες μορφές. Η τεχνική Nuss όμως χρειάζεται εμπειρία, έχει περιορισμούς και υπόκειται σε συνεχείς μικρο-τροποποιήσεις.

Λέξεις Κλειδιά: Δυσμορφίες του προσθίου θωρακικού τοιχώματος, Σκαφοειδές στέρνο, Επέμβαση Ravitch, Επέμβαση Nuss, Ελάχιστα επεμβατική αποκατάσταση του σκαφοειδούς στέρνου.

REFERENCES

1. Fonkalsrud EW. Current management of pectus excavatum. World J Surg 2003; 27: 502-508.
2. Robicsek F. Surgical treatment of pectus excavatum. Chest Surg Clin North Am 2000; 10: 277-296.
3. Huddleston CB. Chest wall deformities. In: Patterson GA, Cooper JD, Deslauriers J, Lerut AEMR, Luketich JD, Rice TW (eds), Pearson's Thoracic and Esophageal Surgery (3rd edition). Churchill Livingston, Philadelphia, 2008, 1236-1242.
4. Shamberger RC. Chest wall deformities. In: Shields TW, LoCicero J, Ponn RB (eds), General Thoracic Surgery (5th edition). Lippincott Williams and Willkins, Philadelphia, 2000, 535-561.
5. Malek MH, Berger DE, Malerich WD, Coburn JW, Beck TW, Housh TJ. Pulmonary function following surgical repair of pectus excavatum: a meta-analysis. Eur J Cardiothorac Surg 2006; 30: 637-643.
6. Chin EF. Surgery of funnel chest and congenital sternal prominence. Br J Surg 1957; 44: 360-376.
7. Rebeis Eb, Milanez JB, Fernandez A, Pinho Moreira LF, Jatene FB. Anthropometric index for pectus excavatum. Clinics 2007; 62: 599-606.
8. Brigato RR, Campos JRM, Jatene FB, Moreira LFP, Rebeis EB. Pectus excavatum: evaluation of the Nuss technique by objective methods. Interac Cardiovasc Thorac Surg 2008; 7: 1084-1088.
9. Chang J-H, Wan Y-L. Evaluation of pectus excavatum with repeated CT scans. Pediatr Radiol 1995; 25: 654-656.
10. Hebra A. Minimally invasive pectus surgery. Chest Surg Clin North Am 2000; 10: 329-339.
11. Kim HD, Hwang JJ, Lee MK, Lee DY, Paik HC. Analysis of the Nuss procedure for pectus excavatum in different age groups. Ann Thorac Surg 2005; 80: 1073-1077.
12. Ravich MM, Steichen FM. Atlas of General Thoracic Surgery. Philadelphia: W.B. Saunders Company, 1988: 10-27.
13. Kotoulas C Papoutsis D, Tsolakis K, Laoutidis G. Surgical repair of pectus excavatum in young adults using

the Dual Mesh 2-mm Gore-Tex®. Interact Cardiovasc Thorac Surg 2003; 2: 565-568.
14. Haller AJ,Jr, Colombani PM, Humphries TC, Azizkhan RG, Loughlin GM. Chest wall constriction after too extensive and too early operations for pectus excavatum. Ann Thorac Surg 1996; 61: 1618-1625.
15. Jacobs JP. Minimally invasive endoscopic pectus repair. http://www.ctsnet.org/sections/clinicalresources/thoracic/expert_tech-15.html
16. Osawa H, Mawatari T, Watanabe A, Abe T. New material for Nuss procedure. Ann Thorac Cardiovasc Surg 2004; 10: 301-3.
17. Swanson JW, Colombani PM. Reative pectus carinatum in patients treated for pectus excavatum. J Pediatr Surg 2008; 43: 1468-1473.
18. Marks MW, Iacobucci J. Reconstruction of congenital chest wall deformities using solid silicone onlay prosthesis. Surg Clin North Am 2000; 10: 341-355.
19. Hougaard G, Svensson H, Holmqvist KG. Casting the implant for reconstruction of pectus excavatum. Scand J Plast Reconstruct Surg Hand Surg1995; 29: 227-231.