Pectus Excavatum Repair and Rehabilitation

Surgical Indications and Considerations

Anatomical Considerations: Pectus deformities refer to abnormal growth and alignment of the costal cartilages (ribs) and their attachment to the sternum.[1] There are two primary deformities of which this Practice Guideline will focus on the more common of the two, the pectus excavatum.

1. Pectus excavatum (PE): an inward displacement of the cartilage causing a posterior (inward) deformity at the lower portion of the sternum, sometimes referred to as a "cavus" or "funnel" chest.
2. Pectus carinatum (PC): an outward displacement of the cartilage causing an anterior protrusion of the sternum, sometimes referred to as a "pigeon breast" deformity.

Pathogenesis: Pectus excavatum deformities can be congenital, acquired or both. PE occurs congenitally if the cartilage is overgrown, deformed or weak allowing the sternum to be pulled inward due to the negative pressure created during inhalation in the thorax.[2-4] PE can also be acquired due to a dynamic muscle imbalance of the primary respiratory muscles: diaphragm, intercostals and abdominals. If the intercostal muscles are overpowered by the diaphragm, the sternum will be pulled inward resulting in a PE.[5-7] This is a fairly common secondary development for infants and children with increased respiratory workloads secondary to pulmonary disorders and/or neuromuscular disorders.[8, 9] Children with a genetic predisposition to PE who also have an imbalance in their primary respiratory muscles will see an exacerbation of their sternal deformity.

Epidemiology: Pectus deformities are the most common congenital chestwall deformities. The rate of occurrence varies among published reports from 1/300 live births to 1/1,000. [2, 10-13] PE is far more common than PC deformities. PE is generally reported as occurring in approximately 85% of the pectus deformities.[2, 3]. Pectus excavatum is more prevalent in males than females, with reports of incidences varying from 3 to10 times more often. [3, 14, 15]

Diagnosis: The diagnosis of a PE is made visually by observing the shape of the anterior chestwall. Common tests to determine the extent of the PE limitation include physical, physiologic and psychological tests:

1. Physical limitations:
    a. Radiographs or CT of chest and spine on coronal and sagittal planes to determine PE severity and other possible musculoskeletal restrictions such as scoliosis and kyphosis [16]
    b. Haller Index rating of CT scan to rate the severity of the PE [16, 17]

2. Pulmonary limitations:
    a. Pulmonary Function Tests to determine the amount of lung restriction and lung growth retardation [14, 18]
    b. Exercise Stress Tests to determine the extent of exercise intolerance due to the compression of the lungs and heart [17]
3. Cardiac limitations:
    a. ECG to determine any electric abnormalities [19, 20]
    b. Echocardiogram to determine the extent of the cardiac compression and to rule out other vessel abnormalities associated with connective tissue disorders that commonly present with PE [19, 20]
4. Quality of Life limitations:
    a. Survey outcomes to determine the effect of the PE on the child's self image and willingness to participate in peer related activities [21, 22]

Non-operative versus Operative Management: Typically, there are 2 options presented to the patient: have surgery or do nothing. The literature notes musculoskeletal problems frequently occur with or because of the PE, yet only 2 articles suggest physical therapy for the patients with PE and no article suggested PT as a regular screening test for PE. [23, 24] Currently, there is no literature to suggest that PE can be influenced by physical therapy, however I will share my non-surgical experience with this population later in the guideline.

Surgical Procedure: Two different surgical procedures are commonly performed to reduce the PE deformity: the modified Ravitch [25]or the Nuss procedure. [26]

- The modified Ravitch procedure is based on the technique described by Dr. Ravitch in 1949. It is also called the "Open Repair" technique and is championed by Dr. Eric Fonkalsrud at UCLA who has extensive experience performing this technique and reporting on outcomes for the past several decades. [2] The open repair surgically corrects and or resects the damaged cartilage (usually up to 4 rib cartilages). The sternum is then elevated to the proper position and a strut is inserted to maintain the alignment for around 6 months. The patient is hospitalized for 2- 5 days for the initial surgery. The strut removal is done as an outpatient. [2] The Ravitch procedure repairs only the damaged cartilage on the anterior chest wall. It does not affect the relationship of the entire rib with the thoracic spine.

- The Nuss procedure was introduced by Dr. Donald Nuss at the Children's Hospital of the King's Daughters in Norfolk, VA in 1998. [26] His technique is also called "minimally invasive repair of pectus excavatum (MIRPE)". The Nuss procedure uses a Lorenz pectus bar, which is a thin long metal sheet that resembles the shape of old fashioned bicycle handle bars. The bar is inserted laterally through a small incision, fed under the anterior chest wall, and finishes on the opposite lateral wall. The bar is then "flipped" to mechanically reverse the PE deformity by lifting the anterior chestwall. The bar remains in place for 2 – 4 years to support normal growth of the ribs and cartilage around its support. [27] Although the Nuss procedure is less invasive from a surgical perspective, once the bar is flipped, the shape of the entire anterior chest wall and the relationship of the ribcage to the thoracic spine is changed. Because of this, more joints are affected and

the patients generally report more pain than with the Ravitch procedure.

Both procedures have good outcomes, but each has its own followers. [13] Fonkalsrud reported the outcomes of the modified Ravitch and Nuss procedures that were performed at 2 large hospitals during 1996 – 2000 and is presented below. [28] Recent research shows that with continual modifications of both procedures improving the overall outcomes, decisions about which surgical technique to use should depend on the patient and the surgeon's particular situations. [28, 29]

TABLE

Patient Parameters Compared

Parameter	Nuss Repair	Modified Ravitch Repair
Number of patients	68	139
Average age (yr)	12 (5-19)	17.3 (3-53)
Mean pectus severity index	4.2 (3.2-9.5)	4.9 (3.1-9.8)
Previous pectus repair	0	9
Operating time (min)	75 (45-130)	212 (110-260
Blood loss (mL)	90 (10-120)	90 (15-400)
Length of hospitalization (d)	6.5 (5.8)	2.9 (2-5)
Epidural used	66	0
Pneumothorax	7	3
Transient pericarditis	0	3
Intravenous analgesics (average d)	5 (3-7)	1.7 (1-3)
Patients placed in ICU	2	0
Bar displacement (flipped)	6	0
Reoperations	7	0
Rehospitalizations for pain	2	0
Return to school/work (average d)	18 (14-26)	12 (8-18)
Number sternal bars removed electively	18	107
Bar removal operating time (average min)	25 (17-40)	19 (15-31)
Time to bar removal (avg mo)	24 (23-26)	6 (5.5-6.5)

NOTE: Values in parentheses are ranges.

The overall the morbidity and mortality rates related to either procedure are very low. [10, 30-36] Reported complications of both procedures include:

- failure of the strut or bar to hold the corrected deformity
- cardiac complications
- pulmonary complications

Pre-operative Rehabilitation: Generally, surgeons do not require any pre-operative preparation other than to complete medical tests. There is no report of pre-op physical therapy (PT) evaluations in any of the literature that I used for this assignment. I also directly contacted the offices of the 2 primary PE surgeons: Dr. Fonkalsrud and Dr. Nuss. Both program coordinators indicated that they do not use PT for regular pre-op or post-op screening or intervention. Tina Gustin, program coordinator at Dr. Nuss' office, indicated that the PTs are involved in general bed mobilization education following surgery. She stated that the patients are given a simple pre-op exercise form-sheet from the doctor and encouraged to "improve their posture and strengthen their back muscles" before and after surgery on their own. Gale Tiemann, Dr. Fonkalsrud's assistant, said they do not routinely use physical therapy.

POSTOPERATIVE REHABILITATION

Procedure 1: Nuss or minimally invasive repair of pectus excavatum (MIRPE): There were no detailed post-op protocols lists published in the articles or on the Nuss website, so Dr. Nuss' office was contacted directly for this information.

- Inpatient hospitalization is around 5 - 8 days. Patients are instructed to sleep supine, avoid "heavy lifting", and avoid twisting their spine. PT is utilized for general bed and ambulation mobilization. These limitations are in place for 4 weeks.
- For the first 6 weeks, walking is the only allowed "exercise".
- After 6 weeks, the patients can engage in swimming and other low risk activities that will not cause a sudden jerking motion of the chest or cause a blow to the chest or spine.
- After 8 weeks, the patients can begin slowly begin a weight lifting program.
- At 12 weeks, the patients can re-engage with all physical activities except those with a high risk for full body contact such as football, wrestling, contact martial arts, etc.
- After 2-3 years when the chest bar is removed, the patient can re-engage in any sporting activity.

Goals: To maintain the position of the chest bar during the 2-3 year post-op period to allow the bones, joints and cartilage to reshape themselves along the improved chest alignment until the results are permanently maintained by the patient's own musculoskeletal system.

Interventions: PT is rarely utilized beyond initial inpatient mobilization period.

Procedure 2: Modified Ravitch or open repair technique: No published post-op protocols were available for the modified Ravitch either, so Dr. Fonkalsrud's office was contacted directly for this information.

- Inpatient hospitalization is around 2 – 5 days. PT is utilized for general bed and ambulation mobilization.
- For 3 – 4 months, the patients are instructed to avoid lifting over 10 pounds. They are also told to avoid lifting their arms above 90 degrees suddenly or to twist their bodies suddenly. No sports or no gym classes are allowed during this time
- After the strut bar is removed at 6 months, the patients can re-engage in full activities including contact sports.

Goals: Maintain the position of the chest bar during the 6-month post-op period until the cartilage and sternal junction is healed. Once it is healed, the results should be permanently maintained by the patient's own musculoskeletal system.

Intervention: PT is rarely utilized beyond initial inpatient mobilization period.

Procedure 3: Alternative Interventions to surgery: I work with a pediatric pulmonologist (Dr. Steen Boas) and a pediatric thoracic surgeon (Dr. Marleta Reynolds who uses a modified

Ravitch procedure) as a team to evaluate the best intervention strategy for children with PE. The goal of the pre-op evaluation is:

- to determine the physiologic restrictions (cardiac, pulmonary, connective tissue, etc.) if any, on the child as a result of the PE
- to assess the child's potential for physical rehabilitation as a means of reducing the PE and related postural abnormalities
- to determine the need for surgery
- to determine the best sequence of intervention strategies

PT Goals: Through the use of specific intervention strategies including those listed below, teach the patients how to use their own bodies to minimize the deformity and related physical and physiologic restrictions to the best of their own ability and to help determine whether surgery is necessary to attain a better result with all aspects considered.

- Musculoskeletal techniques to improve mobility of all affected joints, most commonly the chest, shoulders, spine, neck, and pelvis
- Neuromuscular re-education from a developmental perspective to teach these patients how to correct muscle imbalances that have been present their entire lives. In particular, retraining focuses on imbalance as they relate to breathing, postural control and trunk stabilization.
- Cardiopulmonary interventions to teach more efficient breathing patterns and coordination with movement. Implement related pulmonary programs as necessary such as airway clearance (patients often have true pulmonary disorders such as asthma and broncho-pulmonary dysplasia).
- Integumentary techniques to reorganize the underlying connective tissue structures that limit full erect posture and normal UE positioning.

Intervention: In our multidiscipline assessment, PT is regularly utilized to screen the patients for chest wall mobility, postural abnormalities, respiratory and trunk muscle imbalance, development delays of effective movement strategies, integumentary restrictions, swallowing/aspiration problems, etc. If a patient is deemed a PT candidate, surgery is put off for 6 – 12 months to see if physical rehabilitation can reduce the cosmetic and physiologic restrictions adequately enough to decide that surgery is no longer necessary. The outcomes of our team interventions are currently being collected and will hopefully be analyzed and published at a later date.

References

1. Medline Plus E. Pectus Excavatum at Medline plus. *Medline plus Health Information.* *http://www.nlm.nih.gov/medlineplus/ency/imagepages/2927.htm* [Internet website]. 10/6/03. Available at: http://www.nlm.nih.gov/medlineplus/ency/imagepages/2927.htm. Accessed 10/10/03.

2. Fonkalsrud EW. Pectus Excavatum, Pectus Carinatum and Pectus [internet website]. 2004. Available at: http://www.surgery.medsch.ucla.edu/asp/Clinical.asp?Clinical_Service=Pectus%20Excavatum,%20Pectus%20Carinatum%20and%20Pectus. Accessed 7-26-04, 2004.

3. Malek MH, Fonkalsrud EW. Cardiorespiratory outcome after corrective surgery for pectus excavatum: a case study. *Medicine & Science in Sports & Exercise.* 2004;36(2):183-190.

4. Waters P, Welch K, Micheli LJ, Shamberger R, Hall JE. Scoliosis in children with pectus excavatum and pectus carinatum. *Journal of Pediatric Orthopedics.* 1989;9(5):551-556.

5. Bach JR, Bianchi C. Prevention of pectus excavatum for children with spinal muscular atrophy type 1. *American Journal of Physical Medicine & Rehabilitation.* 2003;82(10):815-819.

6. Lissoni A, Aliverti A, Tzeng AC, Bach JR. Kinematic analysis of patients with spinal muscular atrophy during spontaneous breathing and mechanical ventilation. *American Journal of Physical Medicine & Rehabilitation.* 1998;77(3):188-192.

7. Massery MP. Chest development as a component of normal motor development: implications for pediatric physical therapists. *Pediatric Physical Therapy.* 1991;3(1):3-8.

8. Lund DP, Mitchell J, Kharasch V, Quigley S, Kuehn M, Wilson JM. Congenital diaphragmatic hernia: the hidden morbidity. *Journal of Pediatric Surgery.* 1994;29(2):258-262; discussion 262-254.

9. Vanamo K, Peltonen J, Rintala R, Lindahl H, Jaaskelainen J, Louhimo I. Chest wall and spinal deformities in adults with congenital diaphragmatic defects. *Journal of Pediatric Surgery.* 1996;31(6):851-854.

10. Fonkalsrud EW, Dunn JC, Atkinson JB. Repair of pectus excavatum deformities: 30 years of experience with 375 patients. *Annals of Surgery.* 2000;231(3):443-448.

11. Molik KA, Engum SA, Rescorla FJ, West KW, Scherer LR, Grosfeld JL. Pectus excavatum repair: experience with standard and minimal invasive techniques. *Journal of Pediatric Surgery.* 2001;36(2):324-328.

12. Cincinnati Children's Hospital M. Pectus Excavatum [internet website]. 2004. Available at: http://www.cincinnatichildrens.org/health/info/chest/diagnose/pectus-excavatum.htm?&MSHiC=28591&L=10&W=pectus+&Pre=%3CFONT+STYLE%3D%22color%3A+%23000000%3B+background%2Dcolor%3A+%23FFFF00%22%3E&Post=%3C%2FFONT%3E. Accessed 7-26-04, 2004.

13. Williams AM, Crabbe DC. Pectus deformities of the anterior chest wall. *Paediatric Respiratory Reviews.* 2003;4(3):237-242.

14. Xiao-Ping J, Ting-Ze H, Wen-Ying L, et al. Pulmonary function for pectus excavatum at long-term follow-up. *Journal of Pediatric Surgery.* 1999;34(12):1787-1790.

15. Haller JA, Jr., Scherer LR, Turner CS, Colombani PM. Evolving management of pectus excavatum based on a single institutional experience of 664 patients. *Annals of Surgery.* 1989;209(5):578-582; discussion 582-573.

16. Haller JA, Jr., Kramer SS, Lietman SA. Use of CT scans in selection of patients for pectus excavatum surgery: a preliminary report. *Journal of Pediatric Surgery.* 1987;22(10):904-906.

17. Croitoru DP, Kelly RE, Jr., Goretsky MJ, Lawson ML, Swoveland B, Nuss D. Experience and modification update for the minimally invasive Nuss technique for pectus excavatum repair in 303 patients. *Journal of Pediatric Surgery.* 2002;37(3):437-445.

18. Haller JA, Jr., Loughlin GM. Cardiorespiratory function is significantly improved following corrective surgery for severe pectus excavatum. Proposed treatment guidelines. *Journal of Cardiovascular Surgery.* 2000;41(1):125-130.

19. Shamberger RC. Cardiopulmonary effects of anterior chest wall deformities. *Chest Surgery Clinics of North America.* 2000;10(2):245-252.

20. Shamberger RC, Welch KJ, Sanders SP. Mitral valve prolapse associated with pectus excavatum. *Journal of Pediatrics.* 1987;111(3):404-407.

21. Lawson ML, Cash TF, Akers R, et al. A pilot study of the impact of surgical repair on disease-specific quality of life among patients with pectus excavatum. *Journal of Pediatric Surgery.* 2003;38(6):916-918.

22. Roberts J, Hayashi A, Anderson JO, Martin JM, Maxwell LL. Quality of life of patients who have undergone the Nuss procedure for pectus excavatum: Preliminary findings. *Journal of Pediatric Surgery.* 2003;38(5):779-783.

23. Niedbala A, Adams M, Boswell WC, Considine JM. Acquired thoracic scoliosis following minimally invasive repair of pectus excavatum. *American Surgeon.* 2003;69(6):530-533.

24. Schoenmakers MA, Gulmans VA, Bax NM, Helders PJ. Physiotherapy as an adjuvant to the surgical treatment of anterior chest wall deformities: a necessity? A prospective descriptive study in 21 patients. *Journal of Pediatric Surgery.* 2000;35(10):1440-1443.

25. Fonkalsrud EW. Open repair of pectus excavatum with minimal cartilage resection. *Annals of Surgery.* August 2004 2004;240(2):231-235.

26. Nuss D, Kelly RE, Jr., Croitoru DP, Katz ME. A 10-year review of a minimally invasive technique for the correction of pectus excavatum. *Journal of Pediatric Surgery.* 1998;33(4):545-552.

27. Nuss D. The Nuss Procedure for Pectus Excavatum [internet website]. 2004. Available at: http://www.chkd.org/about_us/pectus_kidstuff.asp. Accessed 7-26-04, 2004.

28. Fonkalsrud EW, Beanes S, Hebra A, Adamson W, Tagge E. Comparison of minimally invasive and modified Ravitch pectus excavatum repair. *Journal of Pediatric Surgery.* 2002;37(3):413-417.

29. Jo WM, Choi YH, Sohn YS, Kim HJ, Hwang JJ, Cho SJ. Surgical treatment for pectus excavatum. *Journal of Korean Medical Science.* 2003;18(3):360-364.

30. Fonkalsrud EW, DeUgarte D, Choi E. Repair of pectus excavatum and carinatum deformities in 116 adults. *Annals of Surgery.* 2002;236(3):304-312; discussion 312-304.

31. Akcali Y, Ceyran H, Hasdiraz L. Chest wall deformities. *Acta Chirurgica Hungarica.* 1999;38(1):1-3.

32. Nuss D, Croitoru DP, Kelly RE, Jr., Goretsky MJ, Nuss KJ, Gustin TS. Review and discussion of the complications of minimally invasive pectus excavatum repair. *European Journal of Pediatric Surgery.* 2002;12(4):230-234.

33. Park HJ, Lee SY, Lee CS. Complications associated with the Nuss procedure: analysis of risk factors and suggested measures for prevention of complications. *Journal of Pediatric Surgery.* 2004;39(3):391-395; discussion 391-395.

34. Barakat MJ, Morgan JA. Haemopericardium causing cardiac tamponade: a late complication of pectus excavatum repair. *Heart (British Cardiac Society).* 2004;90(4):e22.

35. Ohno K, Morotomi Y, Ueda M, et al. Comparison of the Nuss procedure for pectus excavatum by age and uncommon complications. *Osaka City Medical Journal.* 2003;49(2):71-76.

36. Mansour KA, Thourani VH, Odessey EA, Durham MM, Miller JI, Jr., Miller DL. Thirty-year experience with repair of pectus deformities in adults. *Annals of Thoracic Surgery.* 2003;76(2):391-395; discussion 395.