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PECTUS EXCAVATUM: OVERVIEW AND CASE STUDY

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INTRODUCTION

This case study presents diagnosis and treatment of a patient with pectus excavatum (PE). PE is characterized by depression of the lower sternum and is the most common congenital abnormality of the anterior chest wall. PE ranges in severity from a minor depression to a deep concave hollow capable of displacing the heart and intrathoracic structures. The etiology of PE, though not well understood, resembles other multifactorial inheritance pathologies. PE could be mild and asymptomatic but could also progress in severity, causing physical symptoms and psychosocial problems, especially during adolescence.
Evaluation of PE involves chest radiography or computed tomography scan plus or minus more advanced studies depending on the severity of the ailment. Current treatment of PE involves modified and less invasive surgical procedures with better outcomes and lesser complications. Adequate education, evaluation, follow-up, and referrals are important for nurse practitioners to achieve optimal health outcomes for PE patients.

OVERVIEW

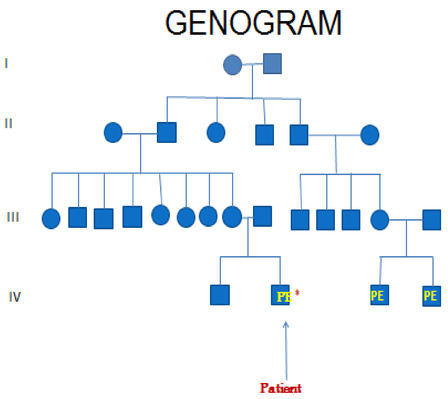
Pectus excavatum (PE), also called "funnel chest" is a common anterior chest wall disorder (1)
The etiology of PE is not well understood, but hypotheses include:
Multifactorial inheritance (1)
Abnormally short central diaphragmatic tendon (5)
Abnormal development of the coastal cartilage (5)
Genetic disorder resulting to overgrowth of the coastal cartilage and ribs (2,3)
Abnormal intra-uterine pressure on sternum and coastal cartilages (2,5)

Epidemiology
Occurs 1:400 births (1,2,3)
Makes up ~ 90% anterior chest wall abnormalities (2)
3- 5 times more in males than females (1,2)
Less prevalent in Blacks and Hispanics (1)
1/3 of PE occurs in first year of life
Rarely resolves spontaneously (2)
~1/3 of cases progresses in severity especially in adolescence during growth spurt (2)
Severity may remain the same throughout life (2)

Clinical Presentation
Psychosocial problems related to appearance, more common in females than males (1,2,6)
SOB, easy fatigability, exertional intolerance, chest pain, especially in adolescents (1,2,6)
CT scan may show moderate to severe cardiopulmonary displacement/implications (1,2)
PFT may reveal respiratory restrictions (2)
Stress test may reveal cardiopulmonary limitations
ECG may reveal conduction abnormalities
ECHO may reveal right ventricular outflow obstruction.

CASE PRESENTATION

Three-month old Hispanic male brought to primary care clinic with complaint of depression of anterior chest wall by his mother. No accompanying symptoms such as respiratory distress reported.
Prenatal Hx: G2P1T1L1. GBS+, normal prenatal ultrasound
Birth Hx: L=54.6cm; wt. 9 lbs 7oz (4.281 kg); HC=36.2cm; APGAR – 1min/9; SVD; 40 6/7 weeks; breast/bottle-fed
Neonatal Hx: APGAR 9/5min
Newborn Screen: Result WNL
Feeding: Similac 6 oz q4hr.
Developmental Hx: WNL
Immunization. Up to date
Family Hx: MGM, PGF DM type 2; two second cousins have PE
Social Hx: Parents married, Hispanic, 1brother
Allergies: NKDA.
Medication: none
ROS: unremarkable except sternal depression



EVALUATION

Physical exam of patient only revealed a mild depression of lower sternal area.
Physical exam is useful for revealing any coexisting disorder such as scoliosis; symptoms such as tachypnea, heart murmur (suggestive of coexisting disorders e.g. Marfan syndrome), tachycardia (in severely displaced heart)
Chest radiograph was not done because it is limited in assessing severity of PE but some studies suggest it adequately measures PE severity, less expensive, has lower radiation exposure risk, and can reveal early kyphoscoliosis (2)
CT scan was done - revealed a PE index of 3.2 and compression of the right side of the heart anteriorly.

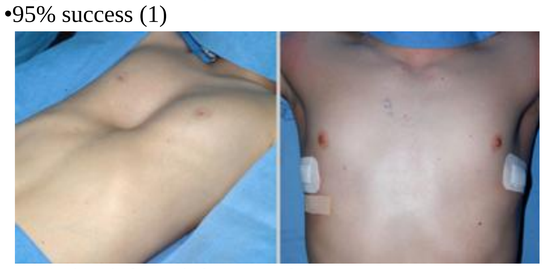
EVALUATION cont.

CT scan is useful for accurately measuring severity of PE; impact on heart, lung, great vessels.[2]
CT is used to calculate the pectus severity index (PSI) or Haller index which is measure of severity (ratio of lateral diameter: AP diameter of chest cavity).[1,2]
Haller index of 2.5 is normal; PE ranges from 3.2 to 12.7 with mean of 4.4. [1]
MRI is costly but could be used to measure severity with no exposure to radiation.[2]
PFT- often is normal but limited in measuring respiratory distress with exertion
Patient was referred to pediatric cardiologist where ECG and Echo were performed. ECG showed normal sinus rhythm at 153 beats per minute. Echo revealed normal intracardiac anatomy with slight compression of the right ventricle which was not hemodynamically significant.
ECG is useful for revealing dysrhythmias
Echo is useful in revealing right ventricular outflow obstruction.[2]
Exercise testing: may measure cardiopulmonary abnormalities better than spirometry.[2]

TREATMENT

Treatments using sternal magnets, sternal suction, prosthetic inserts, and physical therapy have been used in minor PE cases but there are no records of lasting results at discontinuation of these therapies (2)
*2 major surgical procedures currently used are:
Highly Modified Ravitch Repair Procedure
•Open procedure requiring resecting coastal cartilages, sternal osteotomy, bar- support of sternum posteriorly, and longer OR time (2,4)
•Rare complications: wound seromas, pleural effusion, pneumothoraces, bar migration, deformity recurrence if performed before growth spurt (1,2,4)
•Over 90% success rate

Nuss Procedure
•Minimally invasive procedure started in 1998; use of convex bar under sternum and coastal cartilages;which is left in place for 2-3 years; best result if performed in adolescence (16)
Complications:
•Hemothorax, pneumothorax, pleural effusions, pericarditis, pain, seromas, and wound infection (1)
Later complications:
Allergy to bar, pain, rash, overcorrection, and displacement of bar (1)
•95% success (1)

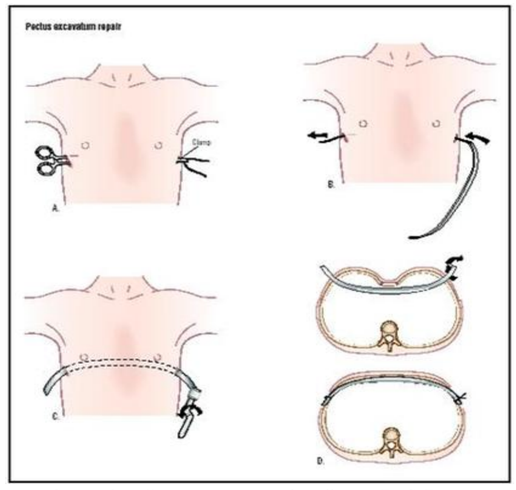


Boy's chest, before and after surgery for pectus excavatum (Nuss procedure) at Mayo Clinic. Source: http://www.mayoclinic.org/pectus-excavatum/enlargeimage5462.html

EDUCATION & FOLLOW-UP

Patient's parents were educated on the following:
- Nature of PE.
- Need to monitor for symptoms such as tachypnea, fast heartbeat, fatigue, cyanosis, etc. which may manifest as child gets older especially toward adolescence; development of any associated disorder.
- Importance of keeping clinic appointments.
- Surgery options; benefits, and complications
- Need for restrictions & avoiding trauma post-operatively

Patient will continue routine wellness check-up
Patient will follow up with cardiologist periodically to continue monitoring progression of the disorder.
Referral to other specialists /surgery will depend on : Index ≥ 3.25 (15) significant cardiopulmonary dysfunction; recurrence after surgery; psychosocial problem; progressing severity; worsening cardiorespiratory symptoms (1,2)



Source: http://www.surgeryencyclopedia.com/Pa-St/Pectus-Excavatum-Repair.html

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