Pectus Deformities 101: When and How they Matter

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Disclosures

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Pectus Excavatum

Sternal depression typically beginning over the midportion of the manubrium and progressing to the xiphoid process.
Pectus Excavatum

- 80-90% anterior chest wall disorders
- 1 in every 300 to 1000 live births
- Approx. 4x more common M:F
Pectus Excavatum

- Usually sporadic

- May be associated with:
  - Marfan syndrome
  - Ehlers Danlos syndrome
  - Osteogenesis Imperfecta
  - Spinal Muscular Atrophy
  - Noonan syndrome
Pectus Excavatum: Etiology

- Abnormal rib growth
- Abnormal cartilage development
- Response to pulmonary conditions
Pectus Excavatum: Natural History

1/3 of cases present in infancy

Some spontaneous improvement (rare); After 6 no improvement

During adolescent growth spurt: 1/3 worsen & 2/3 stable
Pectus Excavatum: Clinical Significance

Severity of chest wall defect

Cardiopulmonary morbidity

Psychosocial Impact/ Cosmetic Concerns
Pectus Excavatum: Symptoms

- Exercise Intolerance: 82%
- Chest Pain: 68%
- Poor Endurance: 67%
- Shortness of Breath: 42%

Pectus Excavatum: Evaluation

Sternal tilt
Pectus Excavatum: Evaluation

Pectus Severity Index (Haller Index) = lateral diameter of the chest sternum to spine distance

Normal PSI: <2.5

Pectus Excavatum PSI: >2.5

Surgical Pectus Excavatum PSI: >3.25
Pectus Excavatum: Evaluation

Pectus Severity Index (Haller Index) = lateral diameter of the chest sternum to spine distance

Mean PSI:
*3.6 isolated cardiac dysfunction
*4.4 isolated pulmonary dysfunction
*4.9 combined dysfunction

Pectus Excavatum: Evaluation

Mild PE:
* Cardiopulmonary function typically normal
* Cosmetic concerns indication for referral
Pectus Excavatum: Evaluation

Moderate to Severe PE:
* CT to evaluate the severity of PE
* Pulmonary Function testing
* Cardiac evaluation/Echocardiogram
  - Functional systolic murmur 18%
  - Bundle Branch Block 16%
* +/- Exercise Testing

Kelly et al. Semin Pediatr Surg 2008
Fonkalsrud et al. World J Surg 2009
10-39% of patients with PE have associated scoliosis

- Tauchi et al. *Eur Spine J* 2018
  - 20 patients with both Pectus Excavatum & scoliosis who had surgery for scoliosis
    - 8 AIS, 10 syndromic, and 2 NMS
    - 11/20 patients had worsening of Haller index
      - Preop 4.8 -> Postop 5.3
      - 9/11 had syndromic/NMS
Pectus Excavatum & Scoliosis

10-39% of patients with PE have associated scoliosis
Multiple reports in the literature of severe hypotension in the prone position during spine surgery in P.E. patients

* Alexianu et. al. *Anesth Analg* 2004
* Bafus et. al. *J Spinal Disord Tech* 2008
* Galas et. al. *Congenital Heart Dis* 2009
No pressure on the central portion of the chest

Consider intraoperative TEE
Pectus Excavatum: Treatment

Open Repair (Ravitch):
cartilaginous repair of ribs
Pectus Excavatum: Treatment

Minimally Invasive Repair (Nuss):
insertion of bar

Late childhood or early adolescence
At 6 months after bar insertion the TLC, FRC, VC, and FEV(1), increased and prior to bar removal the FRC increased.

At 6 months after Nuss bar removal, none of the lung function variables showed any significant change compared to the preoperative values.
Pectus Excavatum: Treatment

Chen et al. J Cardiothoracic surg 2012

* Meta Analysis of 23 studies

* Similar improvement in pulmonary function at 1 yr after Nuss or Ravitch

* Long term follow up favored the Nuss procedure
Pectus Carinatum

Protrusion of the anterior chest wall

* Latin for “chest with a keel”

* Chondrogladiolar prominence: “chicken breast”
  most common
  middle and lower sternum arch forward
  costal cartilage is concave/depressed
Pectus Carinatum

- 1 in every 1500 live births
- Approx. 4x more common M:F
- Familial incidence
  (25% of patients report affected family members)
Pectus Carinatum: Etiology

- Abnormal rib/sternum growth
- Abnormal anterior cartilage growth
- Biomechanical abnormalities in costal cartilage
Pectus Carinatum

• >90% diagnosed after age 11

• Can worsen dramatically during adolescent growth spurt

• Spontaneous improvement does not occur

• Isolated pectus carinatum: most have no associated morbidity
11 yo w/Morquio syndrome

Take care if doing an occipitocervical fusion to avoid longer fusions so it will not prevent airway access.

If fusing longer consider extending neck so patient could have trach if needed.
Pectus Carinatum: Bracing

Improvement in approx 90%

Flexible deformity-> less pressure, correct more quickly but need longer wear hours (recommend 20 hours/day)

Stiffer deformity->higher pressure, correct more slowly but shorter wear hours (recommend 8 to 12 hours)

Lee et al. J Pediatr Surg 2013
Pectus Carinatum: Surgery

Rare

Reserved for severe cases where bracing was ineffective or patient unable to tolerate

Essentially same techniques as PE reconstruction
Take Home Points

• Consider referral if Haller Index >3, symptoms of exercise intolerance, dyspnea or bothered by cosmetic deformity

• If patient has severe PE and is having scoliosis surgery position prone so there is no pressure on sternum- be prepared for hypotension- consider intraoperative TEE

• Pectus carinatum is predominantly cosmetic and often responds well to bracing

• Avoid long cervical fusion with severe pectus carinatum, or put in some extension to allow airway access