

Current Practice & Updates in Pediatric Chest Wall Deformities

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Objectives

At the conclusion of this activity, learners will be able to:

1. Identify the different classifications of chest wall deformities
2. Understand & explain updates and current management of the most common types of pediatric chest wall deformities
 - a) Pain control, surgical technique, activity restrictions
3. Apply this knowledge to your unique practice scenarios

Chest Wall Deformities



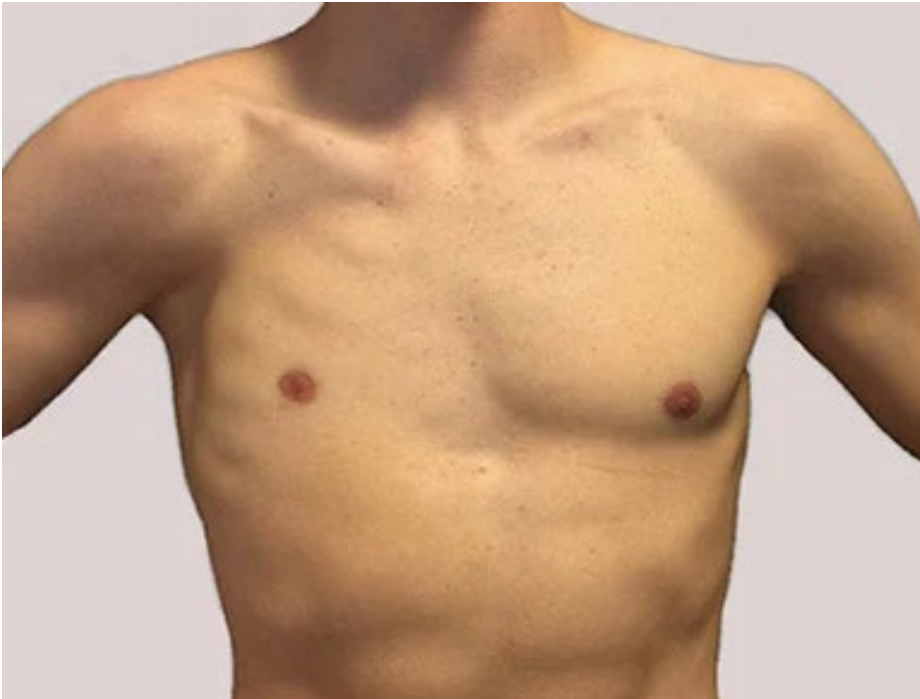
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Chest Wall Deformities



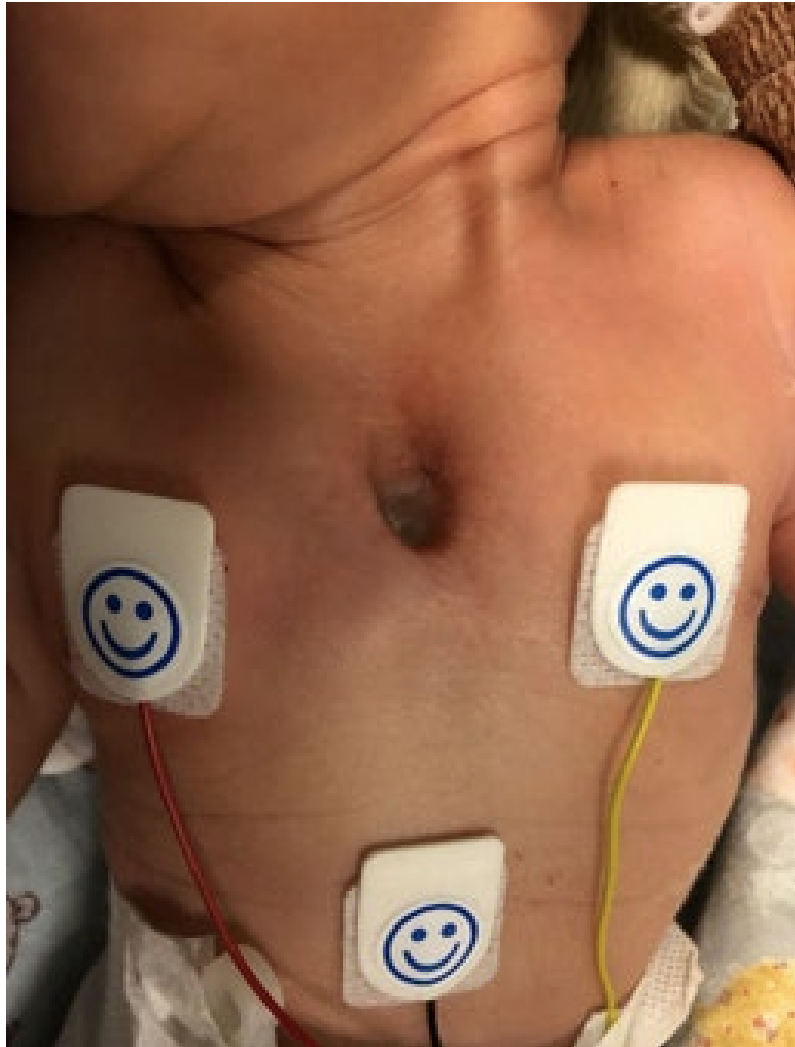
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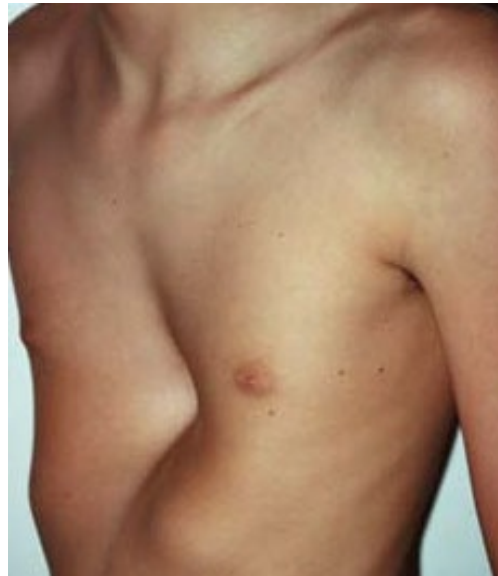
Chest Wall Deformities



Name that type...

Chest Wall Deformities

- Represent abnormalities of the thoracic cavity
- Classified as primary or secondary
- Symptoms vary & depend largely upon the etiology of deformity
 - Cosmetic disturbances → dyspnea → cardiac arrhythmias



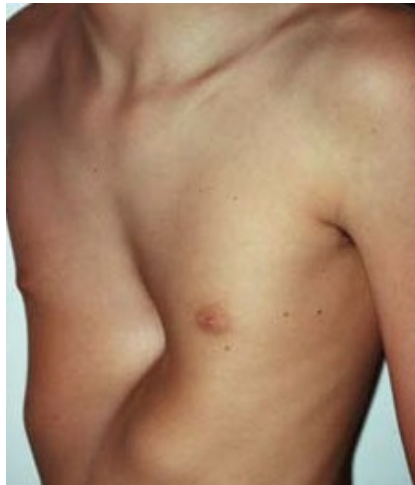
Types of Chest Wall Deformities

1. **Pectus Excavatum*****
2. **Pectus Carinatum*****
3. Poland Syndrome
4. Sternal Cleft
5. Other (Jeune Syndrome)



Pectus Excavatum

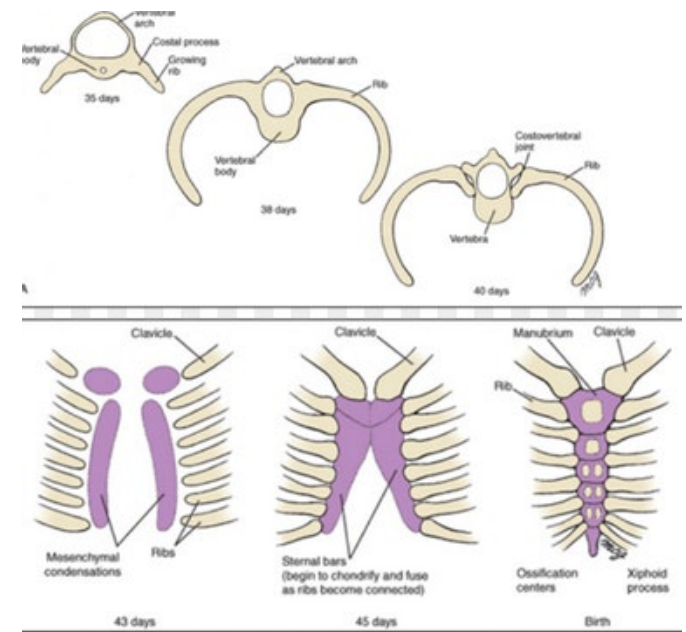
- **Most common chest wall deformity**
 - Represents 90% of all congenital chest wall deformities
 - 1:300-400 white males
 - 4:1 M- F ratio
- **Genetics**
 - 90% sporadic
 - 10% associated w/ syndrome (Marfan's, Ehler-Danlos, Noonan)



Pectus Excavatum

- **Embryology**

- **Day 35 GA:** Trunk develops from mesoderm w/ ribs
- **Day 45 GA:** thoracic cage has costal cartilages that connect ribs 1-7 to sternum
- **Week 6 GA:** endochondral ossification of ribs
- **Week 7-9 GA:** ribs contact sternal bars and fuse in midline



Pectus Excavatum

- Embryology

- 1st ossification of sternum begins at 6 months → complete by 12 years
- Perichondrium produces layers of cells in radial direction
 - Forms the sternocostal joints

- Pathophysiology

- Underlying cause of PE is unknown

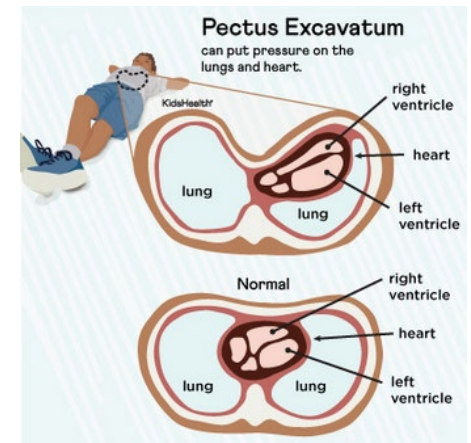
Pectus Excavatum

- **Presentation**

- Do all patients with pectus excavatum have symptoms?
- Appearance of the chest wall that causes the family to seek medical attention

- **Symptoms**

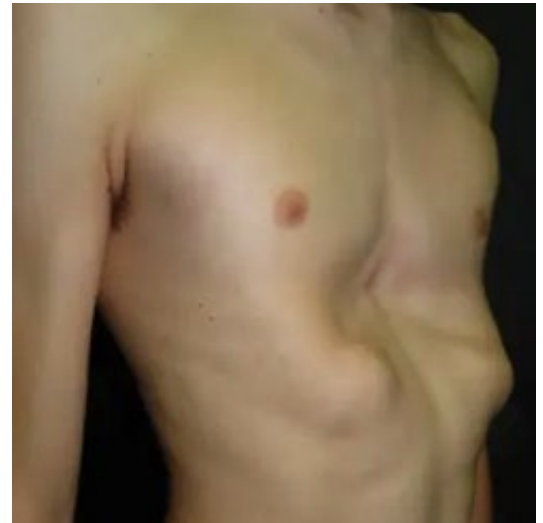
- Psychosocial
- Shortness of breath, limitation of exercise, chest wall pain



**STIGMA
SOCIAL
ANXIETY
LOW SELF
ESTEEM**

Pectus Excavatum

- Challenges
 - Asymmetry of the defect
 - Rib flaring
- Timing of repair
 - Defect may be present at birth
 - Often worsens with growth
 - Surgical results best in 12-16 year old's



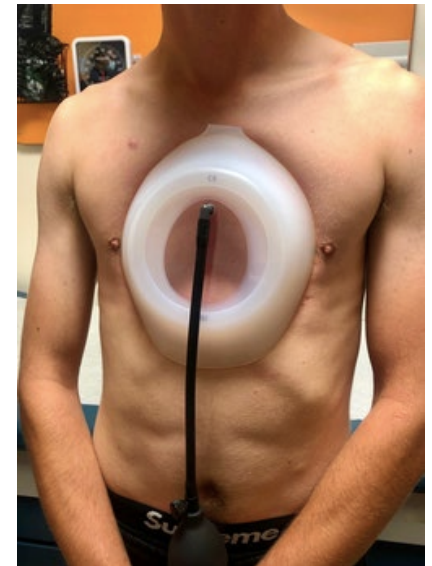
Pectus Excavatum: Treatment

- Surgery

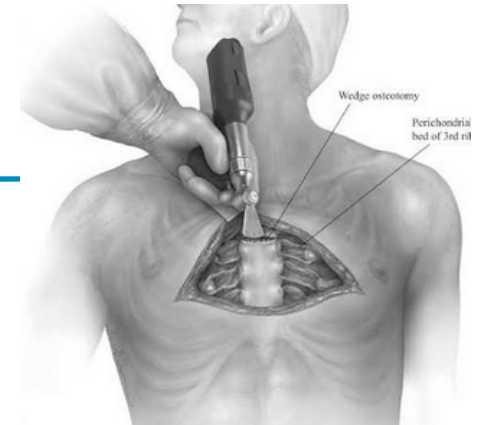
- Relatively few options available for patients who do not want surgery

- Vacuum bell

- Pulls the chest forward
- Requires long-term repeated uses for very modest correction
- Success relies heavily on patient motivation



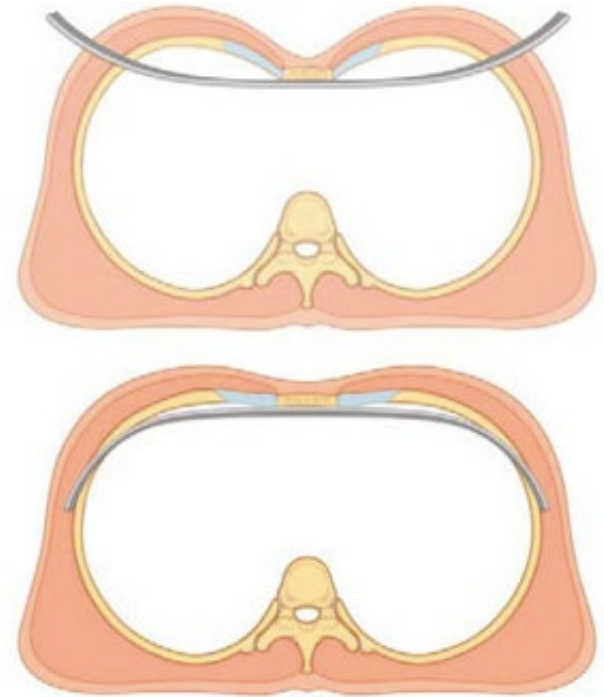
Pectus Excavatum



- **Historical surgical technique**
 - **Open Ravitch procedure**
 - Transverse incision at infra-mammary line
 - Myocutaneous flaps raised
 - Dissection of pectoralis muscles off of deformed portion of chest
 - Perichondrium is incised bilaterally at all involved levels
 - Periosteal elevated used to resect cartilage
 - Perichondrial sheaths preserved for ingrowth of new cartilage
 - Plus/minus wedge osteotomy of sternum and sternal strut

Pectus Excavatum

- **Current surgical standard of care**
 - **Minimally invasive (thoracoscopic) Nuss Procedure**
 - Place temporary metal bar under sternum
 - Bar acts as a brace
 - Allows sternum to reform in more correct position
 - Bar stays in for ~3 years
 - Requires surgery for bar removal

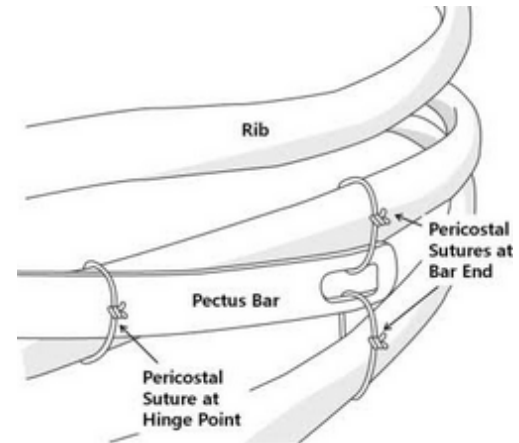
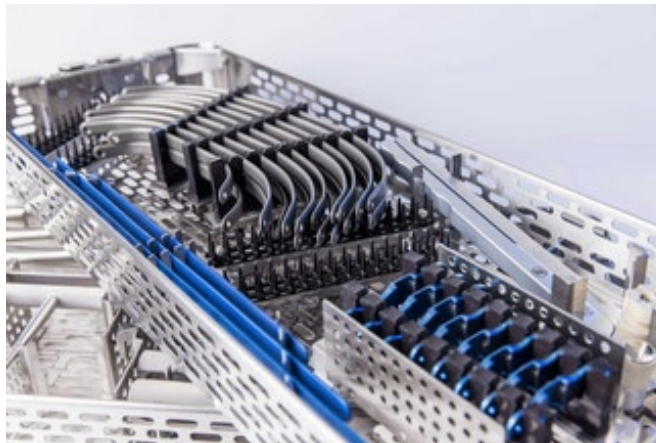


Pectus Excavatum

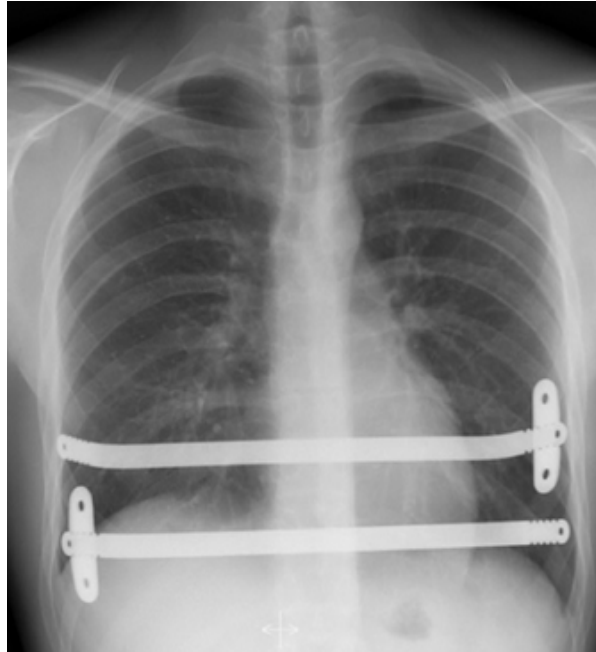
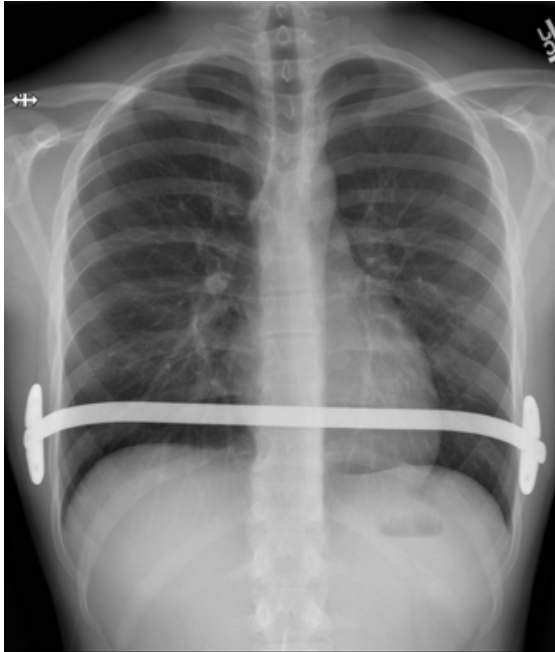
- Updates in surgical technique
 - Use of preformed titanium bars rather than steel
 - Less allergic risk
 - Easier removal
 - Methods for fixation of bars
 - Pericostal sutures
 - Stabilizer (1 or 2)
 - Number and orientation of bars



Bars and Fixation Methods

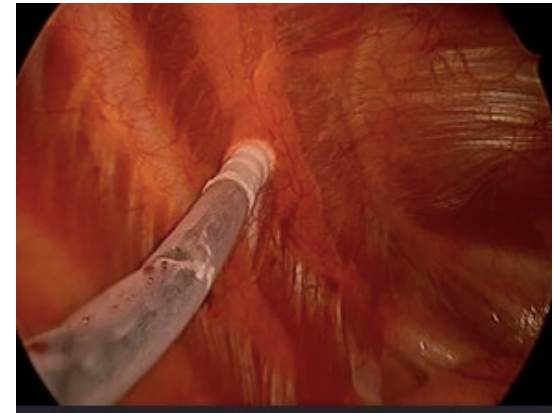


Bar Number & Orientation

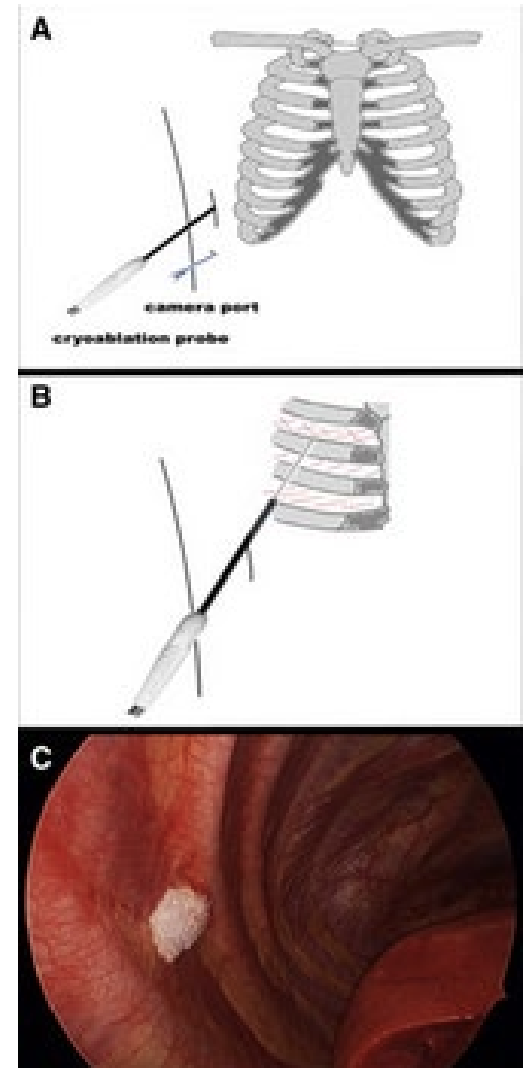
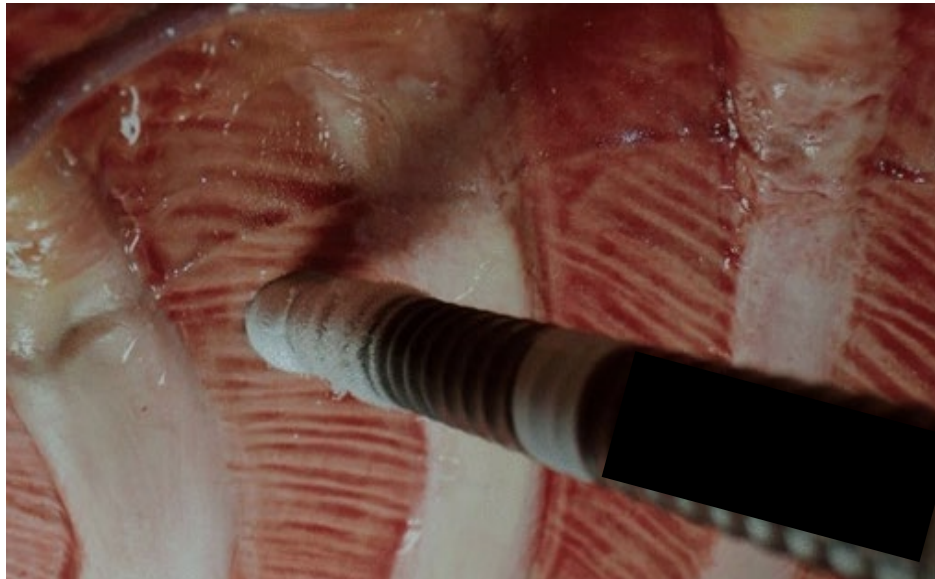
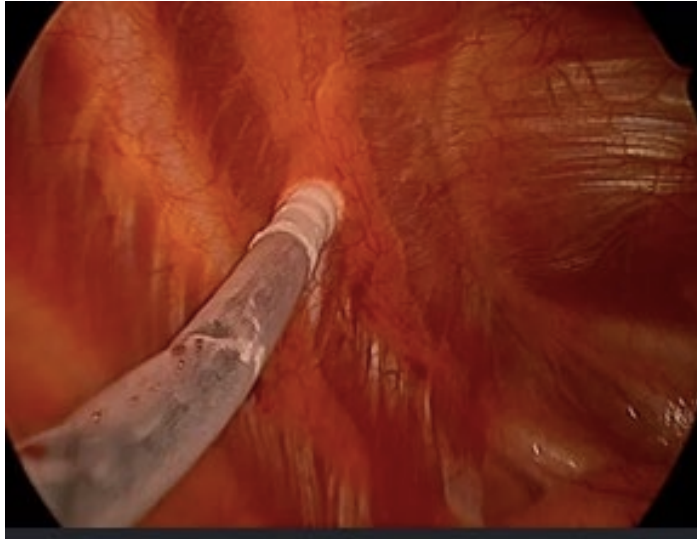


Pectus Excavatum

- Historical pain control strategy
 - Thoracic epidural
 - Patient controlled anesthesia (PCA)
 - Elastomeric pain infusion catheters
 - Opioids
- Updates in pain control optimization
 - Multi-modal approach
 - Intercostal nerve cryoablation

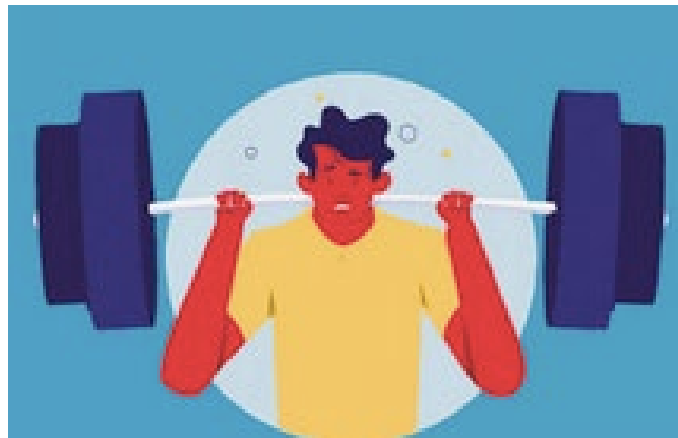


Intercostal Nerve Cryoablation



Pectus Excavatum: Follow-up

- Surgery Clinic
 - 4 weeks post-op
 - 12 weeks post-op w/ CXR
- Activity restrictions
 - No heavy weight lifting & no physical contact to chest x 3 months
 - Non-contact sports & isolated extremity weight lifting is allowed at time of discharge
 - NO ACTIVITY RESTRICTIONS after 3 month visit if no issues



Pectus Carinatum

- Abnormal costal cartilages cause outward protrusion of chest wall → "pigeon chest"
- Represents only 5-10% of chest wall deformities
 - 1: 1,500 children
 - 4:1 M – F ratio
- Genetics
 - Suggested genetic predisposition (1:4 patients have family history of chest wall deformity)



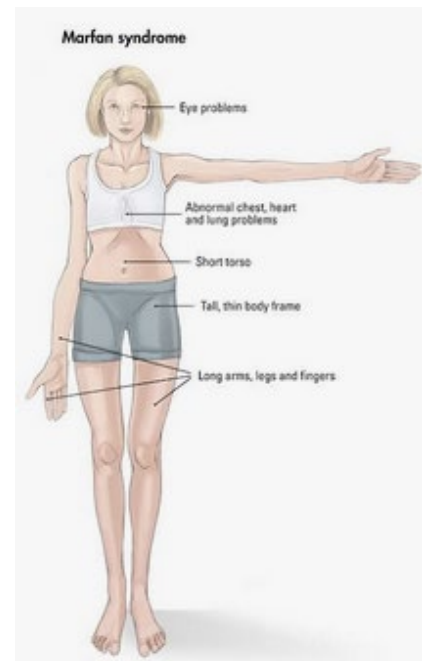
Pectus Carinatum

- **Associations**

- Marfan syndrome, Noonan syndrome
- Poland syndrome , neurofibromatosis, vertebral anomalies
- 10-20% also have scoliosis

- **Pathophysiology**

- Unclear etiology
- Abnormal development of the diaphragm & excessive growth of ribs/costal cartilages have been implicated



Pectus Carinatum

- Presentation

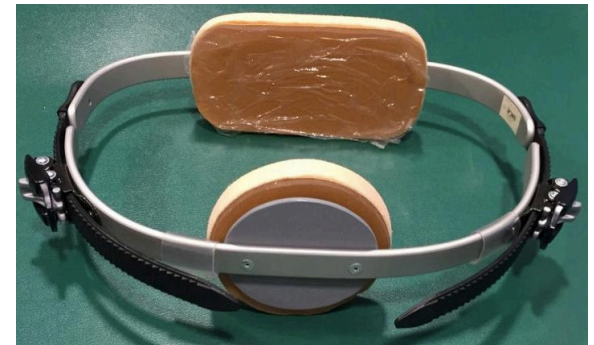
- Can be present at birth → often subtle & not recognized until puberty
- Largely considered a purely aesthetic anomaly*
- Lacks same degree of cardiopulmonary compromise as PE

- Symptoms

- Mild shortness of breath, exercise intolerance
- Asthma
- Tenderness in the area of sternal protrusion when lying prone
- Significant psychological impact**

Pectus Carinatum: Treatment

- Historical management
 - Observation
 - Surgical correction (Ravitch procedure)
- Current management
 - Observation
 - Children <10-12 yo w/ mild defects
 - Bracing***
 - Since 2006, widely adopted and preferred 1st line treatment
 - Surgery
 - Ravitch procedure



Pectus Carinatum: Treatment

- **Bracing**
 - Interest likely initiated by successful results of Nuss procedure
 - Concept of continuous tension on a compliant adolescent chest wall
 - Can induce permanent changes to chest wall topography
 - Health care system factors
 - Insurance coverage by third-party payers influences treatment decisions



Pectus Carinatum: Treatment

- Bracing
 - Centers work w/ orthotics company to design braces
 - Bracing can be dynamic in nature
 - Use a pressure measuring device
 - Calculates pressure needed to reshape chest wall
 - Brace therapy divided into compression and maintenance phases



Pectus Carinatum: Treatment

- **Bracing**

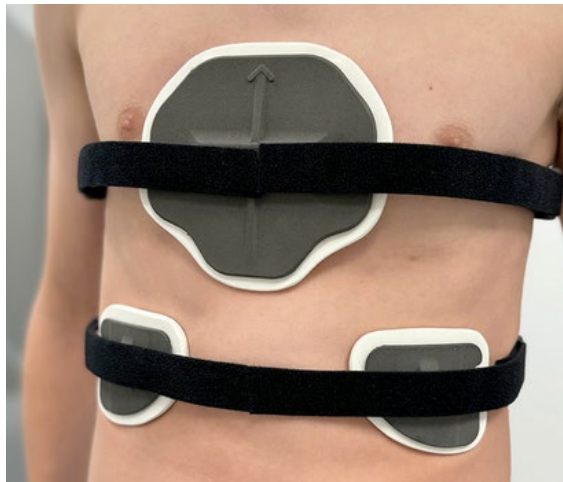
- Current guidelines lacking regarding duration and intervals of compression

- **Duration/Interval Examples**

- Compression phase: **23 hours/day x 3-4 weeks**
- Maintenance phase: **8 hours/day** bracing x **6 months**

OR

- Compression phase: **12-20hours/day x 2-weeks**
- Maintenance phase: **8 hours/day x 6 months**



Pectus Carinatum: Treatment

- **Bracing Results**
 - Overall reported success rate varies widely: 40-100%
 - Bracing DOES NOT preclude patients from operative repair
- **Limitations**
 - Self-motivation and compliance with treatment
 - Compliance affected by wide range of factors
 - Studies show if initial improvement is seen in first 3 months → more likely to complete therapy

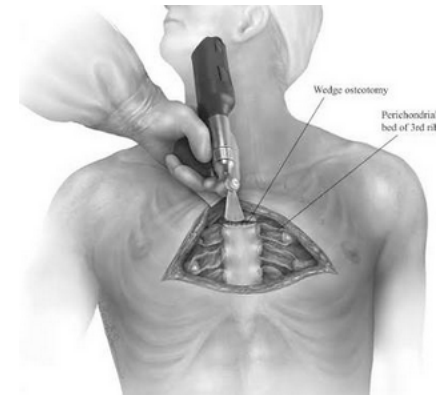
Pectus Carinatum: Treatment

- Indications for surgical management
 - Patients who have failed bracing
 - Patients w/ moderate to severe PC (Haller index < 2.0)
 - Patients with symptoms of tachypnea, reduced endurance, or pain with exertion
 - Patients w/ asymmetric pectus deformities
 - Female patients w/ large breasts (may have difficulty w/ bracing)
 - Patients > 20 years of age

Pectus Carinatum: Surgery

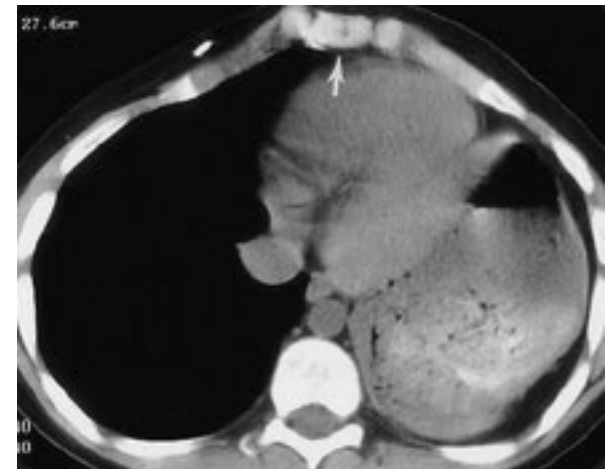
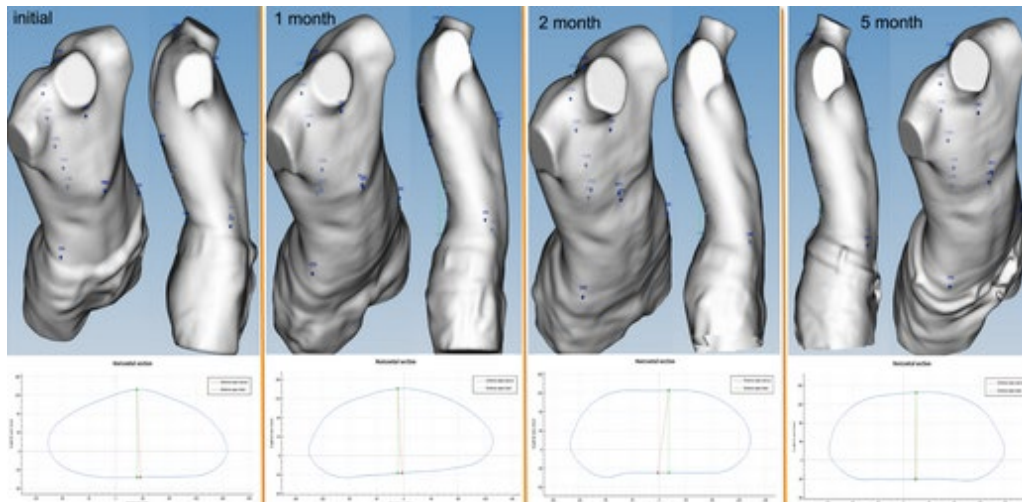
- **Ravitch procedure**

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Pectus Carinatum: Follow Up

- No clear follow-up guidelines for surgeons
 - During bracing
 - After surgical repair
- Novel monitoring techniques
 - White light scanning



Pectus Carinatum: Follow Up

- White light scanning (WLS)
 - Safe, fast
 - Radiation-free optical imaging method
 - Captures millimeter-scale surface contours
 - Map volumetric changes & compare serial scans over time without repeated XR exposure



Summary Review

- **Pectus excavatum and carinatum comprise ~95% of pediatric chest wall deformities**
- **1st line therapy for PE:**
 - **Minimally invasive thoracoscopic repair (Nuss procedure)**
- **1st line therapy for PC:**
 - **Bracing**

Summary Review

- **Updates in management for PE**
 - Titanium preformed Nuss bars
 - Cryoablation for pain control
 - Limited activity restrictions
- **Updates in management for PC**
 - Bracing as 1st line therapy
 - Use of white light scanning for monitoring

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