Pectus Defects: An Update on Options and Timing of Treatment

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OBJECTIVES

- Describe types of different chest wall deformities and their incidence
- Discuss diagnosis, referral, and treatment of pectus defects
  - Pectus Excavatum
  - Pectus Carinatum
# Types of Chest Wall Deformities

<table>
<thead>
<tr>
<th>Condition</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pectus Excavatum</td>
<td>“sunken” or “funnel chest”</td>
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<tr>
<td>Pectus Carinatum</td>
<td>a chest wall protuberance</td>
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<tr>
<td>Poland’s Syndrome</td>
<td>varying degrees of dysplasia of the breast, the pectoralis muscles, ribs, and ipsilateral upper extremity hypoplasia</td>
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<tr>
<td>Jeune’s Syndrome</td>
<td>asphyxiating chondrodystrophy</td>
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<tr>
<td>Bifid Sternum</td>
<td>partial or complete failure of the midline fusion of the sternum</td>
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<tr>
<td>Pentalogy of Cantrell</td>
<td>defects involving the diaphragm, abdominal wall, lower sternum, pericardium, and heart</td>
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## Incidence of Chest Wall Deformities

<table>
<thead>
<tr>
<th>Condition</th>
<th>Incidence</th>
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<tbody>
<tr>
<td>Pectus Excavatum</td>
<td>88% (1 in 1000 births)</td>
</tr>
<tr>
<td>Pectus Carinatum</td>
<td>5%</td>
</tr>
<tr>
<td>Mixed Pectus Excavatum/Carinatum</td>
<td>6%</td>
</tr>
<tr>
<td>Poland’s Syndrome</td>
<td>0.8%</td>
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Pectus Excavatum

<table>
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<tr>
<th>PECTUS EXCAVATUM</th>
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<tr>
<td>• Most common congenital deformity of the chest</td>
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<tr>
<td>• Depression of the anterior chest wall of variable severity</td>
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<tr>
<td>• Abnormal growth of the costal cartilage causes the sternum to be pushed inward</td>
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</table>
PECTUS EXCAVATUM

- May have variations in depth and symmetry

- Progressive disorder
  - Deformity may be noted at birth and progresses with growth
  - During rapid pubertal growth, the defect may become more pronounced

PUNCHED-IN
WIDE-SHALLOW (SAUCER-SHAPED)

ASYMMETRIC
PECTUS WITH COSTAL FLARING

CLINICAL FEATURES

• Most young children are asymptomatic
  – Significant cardiac and pulmonary reserve
  – Chest wall is still very pliable

• As deformity becomes more severe and chest wall becomes more rigid → increasing subjective symptoms

• “Pectus posture” – thoracic kyphosis, forward-sloping shoulders, and a protuberant abdomen

• Psychosocial impact from body image
**CLINICAL FEATURES**

- Upon presentation during adolescents majority (95%) have subjective symptoms
  - Shortness of breath (67%)
  - Chest pain (66%)
  - Dyspnea on exertion (50%)

- Majority of echocardiograms (85%) and PFTs (72%) are normal

Yu YR et al. Preoperative resource utilization prior to minimally invasive repair of pectus excavatum.

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**SURGICAL EVALUATION**

- Consider surgical correction for children ≥ 14-years-old

- History and Physical
  - Identification of symptoms/limitations
  - Family history of pectus deformities
  - Marfan syndrome or other connective tissue disorders
  - History of metal allergy
  - Evaluate for mixed excavatum/carinatum defects
  - Evaluate for chest acne

- CT scan
  - Evaluation of cardiac compression
  - Haller Index (>3.2) vs. Correction Index (>10%)
SURGICAL EVALUATION

- Consider studies to evaluate cardiopulmonary symptoms
  - Echocardiogram
  - PFTs
- Begin posture and upper body strengthening exercise program
- Not all patients with pectus excavatum need surgery

HALLER INDEX – PECTUS PATIENT

<table>
<thead>
<tr>
<th>The Haller Index (HI)</th>
<th>the standard metric to evaluate the severity of pectus excavatum</th>
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</table>

Haller Index = 190.3 / 69.8 = 2.73
**HALLER INDEX – NORMAL CONTROL**

Haller Index = \( \frac{233.7}{62.8} = 3.72 \)

**THE CORRECTION INDEX**

**Correction Index (CI)** measures the depression of the sternum relative to the anterior chest

Correction Index = \( \frac{(90.3-69.8)}{90.3} \times 100 = 22.7 \)
Minimally Invasive Repair of Pectus Excavatum with Nuss Bar
MINIMALLY INVASIVE REPAIR PECTUS EXCAVATUM (NUSS BAR)
MINIMALLY INVASIVE REPAIR PECTUS EXCAVATUM (NUSS BAR)

MINIMALLY INVASIVE REPAIR PECTUS EXCAVATUM (NUSS BAR)
## POSTOPERATIVE COURSE

- Average hospital length of stay is 3 – 5 days
- Pain management is key
- No physical education, strenuous activity, or heavy lifting for 1 month
- May resume posture and upper body strengthening exercises after 3 months
- No contact sports for 6 months

## PECTUS BAR REMOVAL

- After approximately 3 years
- Outpatient surgery
- Open lateral chest incisions only, and remove bar and stabilizers
- Minimal pain
- No activity restrictions
<table>
<thead>
<tr>
<th>SUMMARY POINTS – PECTUS EXCAVATUM</th>
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<tbody>
<tr>
<td>• Pectus excavatum defects may become more pronounced during pubertal growth</td>
</tr>
<tr>
<td>• Extensive preoperative workup (including echocardiogram and PFTs) is not necessary</td>
</tr>
<tr>
<td>• Surgical treatment is usually offered for children ≥ 14-years-old</td>
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<tr>
<td>• Nuss bar remains in place for 3 years</td>
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**Pectus Carinatum**
PECTUS CARINATUM

• Protrusion defect of the chest
• Characterized by an abnormal growth of the costal cartilages, resulting in protrusion of the sternum
• Suggested genetic causation with ~25% of patients with a family history of a chest wall defect

5% of all chest wall defects

4:1 male-to-female ratio

CHONDROGLADIOLAR (LOWER)
CHONDROMANUBRIAL (UPPER)

MIXED DEFECT
### Clinical Features

- Common symptoms include:
  - Dyspnea
  - Decreased exercise tolerance
  - Chest pain on exertion

- Psychosocial impact from body image

### Surgical Evaluation

- History and Physical
  - Identification of symptoms/limitations
  - Family history of pectus deformities
  - Evaluate for mixed excavatum/carinatum defects

- No radiographic evaluation or ancillary testing is necessary

- Consider treatment for children ≥ 12-years-old
TREATMENT OF PECTUS CARINATUM

First line treatment for the majority of pectus carinatum defects is:

**Bracing**

Surgery is reserved for bracing treatment failures

PECTUS CARINATUM BRACES OF THE PAST
INTRODUCING…

The Dynamic Compression Brace for Correction of Pectus Carinatum
**Dynamic Compression Brace**

- Brace to remodel costal cartilage into a more normal shape
- Uses the least amount of pressure needed to correct the carinatum
  - Decreases skin breakdown
  - Decreases discomfort
  - Increases compliance
- Avoid surgery!
PRESSURE OF INITIAL CORRECTION (PIC)

MEASURING FOR CUSTOM FIT BRACE
# TYPICAL TREATMENT PLAN

<table>
<thead>
<tr>
<th></th>
<th>Group 1</th>
<th>Group 2</th>
<th>Group 3</th>
<th>Group 4</th>
</tr>
</thead>
<tbody>
<tr>
<td>PC*</td>
<td>1-4</td>
<td>4-6</td>
<td>6-8</td>
<td>&gt;8</td>
</tr>
<tr>
<td>PT**</td>
<td>2,5</td>
<td>2</td>
<td>1,5</td>
<td>1</td>
</tr>
<tr>
<td>Use (hours/day)</td>
<td>24</td>
<td>12 to 24</td>
<td>6 to 12 (day or night)</td>
<td>3 to 6 (day or night)</td>
</tr>
<tr>
<td>Duration of Treatment</td>
<td>2-4 months</td>
<td>4-8 months</td>
<td>8-12 months</td>
<td>1-2 years</td>
</tr>
</tbody>
</table>

*PC=Pressure of Correction; **PT=Pressure of Treatment. Both pressures are measured in PSI. Values are estimates for guidance only; refer to a FMF Dynamic Compressor System trained physician or directly to PAMPAMED SRL for any doubts or questions.

# BRACE ADJUSTMENTS
PRESSURE ADJUSTMENTS

DYNAMIC COMPRESSION BRACE TREATMENT PLAN

- Recommend wearing brace 12 – 23 hours per day depending on pressure of correction

- Duration of treatment averages 8 months (varies based on initial pressure of correction)

- 1 – 2 month follow-up for brace adjustments throughout treatment course

- After complete correction maintain use in “retainer mode”
### Dynamic Compression Brace Skin Care

- Must monitor for skin breakdown throughout treatment course
- Redness of the skin is expected under the area of the compression plate
- Evaluate the skin for blanching
- Redness without blanching indicates potential for skin breakdown
- The brace should only be worn once the skin blanches again

### Dynamic Compression Brace Treatment Failure

- Wearing the brace as prescribed without treatment progression after 2 years
- Unable to tolerate the brace due to pain or discomfort
- Non-compliance
- Next step is surgery – Ravitch procedure
**SUMMARY POINTS – PECTUS CARINATUM**

- Pectus carinatum defects may become more pronounced during pubertal growth
- Imaging and ancillary testing is not necessary prior to treatment
- First line treatment is bracing and usually started around 12-years-old
- Average duration of bracing is approximately 8 months

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**TEXAS CHILDREN’S HOSPITAL PECTUS PROGRAM**

**Our Team**
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Allen L. Milewicz, MD
Jed G. Nuchtern, MD
Sohail R. Shah, MD, MSHA
Celia Flores, PA-C

[www.texaschildrens.org/pectus](http://www.texaschildrens.org/pectus)

For appointments call: 832-822-3135