CHEST WALL DEFORMITIES

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OUTLINE

• Pectus Excavatum
• Pectus Carinatum
  -Rib Flaring
• Poland syndrome
GENERAL OVERVIEW

(a) Skeleton of the thoracic cage, anterior view

- Jugular notch
- Clavicular notch
- Manubrium
- Sternal angle
- Body
- Xiphisternal Joint
- Xiphoid process
- Sternum
- True ribs (1–7)
- False ribs (8–12)
- Intercostal spaces
- Costal cartilage
- Costal margin
- Floating ribs (11, 12)

https://www.slideshare.net/DrMohammadMahmoud/bones-and-joints-of-the-thorax
PECTUS EXCAVATUM
PECTUS EXCAVATUM

• “Funnel chest”

• Deformity of the chest wall characterized by a sternal depression typically beginning over the mid-portion of the manubrium and progressing inward through the xiphoid process
PECTUS PROGRAM

• Pectus Excavatum accounts for 90% of anterior chest wall disorders
• 1:400-1000 live births
• 3-5x more prevalent in males than females
• Usually sporadic but may be associated with connective tissue disorders (Marfan, Ehlers-Danlos, osteogenesis imperfecta), neuromuscular disease (spinal muscular atrophy) or some genetic conditions (Noonan or Turner syndrome and MEN2)

EPIDEMIOLOGY AND PATHOPHYSIOLOGY
PECTUS EXCAVATUM

Symptoms:

• Exercise intolerance  82%
• Chest pain  68%
• Poor endurance  67%
• Shortness of breath  42%
PECTUS EXCAVATUM

Natural History:

• 1/3 of cases present in infancy
• Spontaneous regression has been reported but is rare
• After 12yr of age, the PE worsens in 1/3 of patients during the adolescent growth spurt and remains the same in 2/3
• As PE worsens, simple symmetrical lesions may progress to more complex asymmetric deformities
PECTUS EXCAVATUM- DIAGNOSTICS

Diagnostics:

- CT scan: Pectus severity index (PSI) = Hallar index: describes the depth of the pectus defect by comparing the ratio of the lateral diameter of the chest to the distance between the sternum and spine
  - Normal PSI = 2.5
  - Surgery if PSI > 3.25

https://radiopaedia.org/articles/haller-index
PECTUS EXCAVATUM- DIAGNOSTICS

• Pulmonary function tests to assess restrictive lung disease
• Exercise testing to assess for cardiopulmonary limitation
• Echocardiogram if significant displacement of the heart or cardiopulmonary limitation
• +/- genetics workup
• Dermatology referral for skin patch test
PECTUS EXCAVATUM

Clinical significance depends on three factors:

1. Severity of the chest wall defect (Haller Index or Chest Severity Index)

2. Cardiopulmonary morbidity (chest pain, shortness of breath, exercise intolerance)

3. Psychosocial impact due to patient’s appearance
TREATMENT

• Non Surgical
  • Vacuum bell

• Surgical
  • Modified Ravitch Procedure
  • **Nuss Procedure** **first line**
TREATMENT

- Vacuum bell therapy: non-invasive therapy
- Not well studied
- Ideal for younger patients with mild
  - Must have flexible chest and;
  - Depth of PE <1.5cm and;
  - Child <12yr age

TREATMENT – SURGICAL INTERVENTION

Must meet two or more criteria:

• Haller index >3.25
• Cardiac compression or conduction abnormalities
• PFT showing restrictive disease
• Psychosocial concerns
TREATMENT – SURGICAL INTERVENTION

Modified Ravitch Procedure

• “Open procedure”

• Resection of the subperichondrial cartilage, a sternal osteotomy underneath the angle of the PE and temporary internal fixation to support the sternum

https://thoracickey.com/congenital-chest-wall-anomalies/
TREATMENT – SURGICAL INTERVENTION

Nuss procedure **First Line**

- “Minimally invasive” thoracoscopic technique
- Curved steel bar inserted under the sternum to remodel the cartilage
TREATMENT – SURGICAL INTERVENTION

Nuss bar Removal

• Done after 2-3 years
• Outpatient procedure
COMPLICATIONS- NUSS PROCEDURE

• Postoperative pain
• Bar displacement
• Cardiac complications
• Pneumothorax
• Recurrence
• Allergic reaction

https://www.semanticscholar.org/paper/Experience-in-100-cases-with-the-Nuss-procedure-a-Uemura-Nakagawa/3f6bd857fb126c4aded256ff112829be826f30a24
OUTCOMES

• In a series of 1170 patients, patient-reported satisfaction was excellent in 93% and good in 6% after Nuss procedure*

• Marked improvement in chest shape as well as self-image

• Subtle improvements in lung function at rest after surgical correction

• Subjective improvement in exercise tolerance

INTRAOPERATIVE INTERCOSTAL NERVE CRYOABLATION (INC)

- Performed by disrupting peripheral nerve structures and temporarily blocking conduction
- Historically has had many applications ranging from trigeminal neuralgia and rib fractures to back and joint pain and peripheral neuritis
NUSS PROCEDURE WITH INC AT TCH

• Decreased length of stay
• Decreased opioid requirements
• Decreased urinary retention
• Not associated with increased odds of slipped bars or allodynia
**PEDIATRIC SURGERY**

**What to Expect After a Nuss Procedure**

**PAIN**
- Pain after surgery is common. Your pain will be managed by a team of anesthesiologists specialized in pain control.
- You will be placed on a regimen of pain relievers, muscle relaxers and neuromodulators that will be adjusted as needed.
- Cryotherapy may reduce the amount and duration of pain after the procedure. Results are different in each patient.

**SWELLING/BURSING**
- Swelling and bruising around the incision and where the bar was placed may occur and last for several weeks after surgery.
- You can take non-steroidal anti-inflammatory medication (NSAIDs) such as ibuprofen as needed to assist with swelling.

**IMAGING**
- You will have a chest X-ray immediately following surgery and on the day you leave the hospital. The imaging results will be reviewed with you on your day of discharge or at your first follow-up visit.

**ACTIVITY RESTRICTIONS WHILE IN THE HOSPITAL**
- Activity restrictions are necessary to make sure the Nuss bar stays in place and becomes stable.
- Weight loss is common for patients after surgery.

**Activity in the Hours Following Surgery**
- You should be able to walk to the bathroom and around the room with assistance.

**Activity in the Days After Surgery**
- You should be able to walk to the bathroom and around the hospital floor with assistance as needed. Walking will help improve blood flow, speed wound healing and help to maintain normal breathing function.

**FATIGUE**
- It is common to feel tired or fatigued for a few days after surgery.
- It is recommended to add extra protein in your diet during this period.

**GOING HOME**
- Length of stay varies per patient but patients with a Nuss bar usually stay in the hospital for approximately 1-2 days.
- In order to go home, you need to reach the following milestones:
  - 12 hours without pain medicine
  - 12 hours without fever
  - Eating a regular diet

**ACTIVITY RESTRICTIONS AFTER GOING HOME**
- No sports or physical education for up to 6 weeks. You may only participate in activities like walking that are:
  - Not strenuous for you
  - Non-contact
  - Non-impact

**PLAN FOR FOLLOW-UP APPOINTMENTS**
- You will receive a phone call 3-5 days after discharge to evaluate pain control.
- The first follow-up visit will be scheduled 2 weeks after leaving the hospital.
- At 6 weeks, you will visit the clinic to receive an exercise program to increase strength and range of motion and a release to participate in sports.
- At the 6-week follow-up visit, you can schedule 3-month, 1-year and 2-year follow-up visits.
- At the 2-year follow-up visit, we will discuss scheduling the bar removal procedure at the 3-year point.

**NUSS PROCEDURE FAQ**

**Am I able to travel by airplane if so after how long?**
- In most cases, you can travel by airplane one week after surgery, unless otherwise instructed.

**Do I need a letter for airport security?**
- During your hospital stay, you may be provided with a letter/card to use for airport security.

**Can I have an MRI done with the bar in place?**
- The company who produces the bar has disclosed information summarized in the brochure regarding magnetic resonance imaging if you are being scanned, make sure to inform the radiologist about your implant and defer decision-making to them.

**Are antibiotics required prior to dental procedures due to the implanted bar?**
- No, prophylastic antibiotics are not necessary.

**Is a clicking/popping noise or sensation normal?**
- Yes, some patients report a clicking or popping noise after surgery; this can be normal.

**When should I be concerned and seek medical attention?**
- If you notice a sudden change in the appearance of your chest and/or experience severe, unexplained chest pain, fever and/or shortness of breath, please go to the emergency department.

**Is it normal to notice the bar sticking out on the side?**
- Patients tend to lose weight the first few weeks following surgery and during this time the bar may become more noticeable. Over time, as patients gain weight, the bar will become less obvious.

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**PEDIATRIC SURGERY CLINIC NUMBERS**

<table>
<thead>
<tr>
<th>Location</th>
<th>Phone Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Texas Medical Center</td>
<td>832-822-3135</td>
</tr>
<tr>
<td>West Campus</td>
<td>832-227-2250</td>
</tr>
<tr>
<td>The Woodlands</td>
<td>936-267-7333</td>
</tr>
</tbody>
</table>

[txchildrens.org/pediatric-surgery]
PECTUS CARINATUM
PECTUS CARINATUM

• “Pigeon chest”
• Protrusion of the sternum and costal cartilages

• Types:
  • Chondrogladiolar prominence (most common)
  • Chondromanubrial prominence

https://www.uptodate.com/contents/pectuscarinatum?search=pectus%20carinatum&source=search_result&selectedTitle=1~18&usage_type=default&display_rank=1
EPIDEMIOLOGY AND PATHOPHYSIOLOGY

• 1:1500 live births
• 4x more prevalent in males than females
• Strong familial incidence (up to 25% of affected patients reporting chest wall abnormalities in family members)
• More common in Caucasian and Hispanic populations
• Usually sporadic but may be associated with musculoskeletal abnormalities (scoliosis in 15-30% of patients), connective tissue disorders (Marfan, Ehlers-Danlos, osteogenesis imperfecta) or some genetic conditions (Noonan or Poland)
PECTUS CARINATUM

Natural History:

• 90% of patients diagnosed after 11th birthday
• Worsens dramatically during adolescent growth spurt
• <10% have a severe defect by 3yr of age
• Spontaneous improvement does not occur
PECTUS CARINATUM

• Most patients are asymptomatic
• Correction has been primarily cosmetic but is known to have significant psychosocial impact
• Signs and Symptoms:
  • Chest pain or discomfort, physical exercise intolerance, scoliosis, impaired shoulders and kyphotic position

PECTUS CARINATUM – DIAGNOSTICS

- CT scan: Pectus severity index (PSI) = Hallar index
  - Normal PSI= 2.5
  - Abnormal PSI < 2.5
- +/- genetics workup
- +/- Cardiology
- +/- Orthopedics (scoliosis)
PECTUS CARINATUM TREATMENT

- Observation
- Bracing
- Minimally invasive techniques
  - Reverse Nuss or Abramson procedures
- Ravitch Procedure
Ravitch Procedure

• “Open procedure”

• Performed through a horizontal chest incision across the mid chest. In this repair, the abnormal costal cartilages are removed, preserving the lining that covers the outside of the cartilage, allowing the sternum to be pushed downward in a more normal position
TREATMENT – NON-SURGICAL INTERVENTION

Bracing **First line**

https://spinaltech.com/pectus-carinatum-orthosis
https://www.ratchetingbuckles.com/applications/medical/orthopedic-prosthetic/pectus-brace/
TREATMENT - DYNAMIC COMPRESSION BRACING

- Custom fitted
- Low profile
- Light weight (Aluminum)
- Each segment is replaceable or adjustable
DYNAMIC COMPRESSION BRACING AT TCH

• Eligibility criteria:
  • >10 years of age
  • Pectus carinatum located below the armpit line

• What treatment looks like:
  • Initial visit with surgeon
  • Monthly Adjustment clinics with APP until chest is corrected
  • 3 month wean period followed by yearly follow ups
OUTCOMES

• 75-95% resolution/improvement in patients who comply with protocol

• Effectiveness of bracing = patient compliance/severity

Pectus Carinatum
Dynamic Compression Brace Information

TYPE OF BRACE
You have been fitted with a Dynamic Compression Brace.

RECOMMENDATIONS
Try to wear the brace during the day.
Wear the brace every day.
Grip: Wear a bra without an underwire. The wire can cause skin irritation under the brace.
Avoid weightlifting using your upper body muscles during the length of your treatment.

TEST YOUR SKIN
To make sure that the Dynamic Compression Brace isn’t causing skin breakdown, you need to perform a Red-to-White Finger Test once every day.

Push down on skin with 1 finger, remove the finger and observe as your skin goes from red to white in a few seconds. If the area does not turn white, do not wear the brace until the skin turns white when you repeat the test. This usually takes 1 to 2 days.

BREATHING EXERCISES
Hold your breath for 10 seconds after taking a very deep breath in and expanding your chest. Do 10 deep breaths at least 4 times every day with the brace on. This exercise helps improve chest wall elasticity.

THINGS NOT TO DO
• DO NOT use the Dynamic Compression Brace while exercising or doing physical activity.
• DO NOT make modifications to the brace on your own. You may only adjust shoulder straps.
• DO NOT dip on your own.
• DO NOT let your Dynamic Compression Brace have any contact with water.

TREATMENT PLAN
Wear your Dynamic Compression Brace for ________ hours every day.

Come to your follow-up visit on __________ at __________ am/pm.

TREATMENT PLAN
Call your doctor’s office at 832-822-3135 for any questions you have about your Dynamic Compression Brace or treatment plan.
RIB FLARING

• A common aspect of pectus deformities that is associated with the ribcage.

• Variation in shape of costal margin and it can change with age.
POLAND SYNDROME
EPIDEMIOLOGY AND PATHOPHYSIOLOGY

• Occurs in 1/10,000 to 1/100,000 live births
• Characterized by partial (28%) or complete (72%) absence of the pectoral muscles.
  • Most commonly unilateral (93%)
• Most of the time it appears sporadically
• More common in males than females
POLAND SYNDROME

• Associated anomalies may include:
  • Aplasia or hypoplasia of other chest wall muscles, breast tissue, or nipple
  • Absence of the costal cartilage of ribs 2-4 or 3-5
  • High riding scapula (Sprengel deformity)
  • Digital abnormalities
• No associated respiratory sx
• Diagnosed based on physical exam findings

https://musculoskeletalkey.com/61-modified-green-scapuloplasty-for-congenital-high-scapula-sprengel-deformity/
POLAND SYNDROME - TREATMENT

• Implant or injections to fill out the underdeveloped chest area
• Surgery on the ribs to help with breathing problems
Chest wall deformities are always due to a genetic disorder.

a. True
b. False
POST-TEST QUESTION 2:

A 14-year-old boy complains that he tires much more quickly during gym class than his peers. On physical examination, you note that he has a pronounced depression in the lower chest. What imaging study would you consider ordering to evaluate the severity of the chest wall defect?

a. Ultrasound of Chest Wall
b. Chest MRI
c. Chest CT
d. Bone density scan
e. Chest x-ray
A 13-year-old boy complains of a rapidly progressive protrusion of his lower chest. On physical examination, he has marked lower pectus carinatum. What would you recommend as first line therapy to this patient.

a. Surgery
b. Bracing
c. Chest exercises
d. Physical therapy
A 15-year-old girl presents to the clinic for eval of pectus excavatum. She reports subjective dyspnea on exertion and mild chest pain. You order a CT Chest which produces a Haller index of 3.5. Is this patient a candidate for a Nuss procedure based on this Haller index score?

a. Yes
b. No
Clinical significance of pectus excavatum depends on which of the following listed below.

a. Psychosocial impact
b. Severity of chest wall defect (Haller index >3.25)
c. Cardiac compression or conduction abnormalities
d. PFT showing restrictive disease
e. All of the above
POST-TEST QUESTIONS 6:

Pectus carinatum is primarily a cosmetic concern.

a. True
b. False
POST-TEST QUESTION 7:

The most appropriate age to start bracing a patient with pectus carinatum is

a. 1-5 years old
b. 6-11 years old
c. 12-18 years old
d. 18 – 25 years old
TEXAS CHILDREN’S HOSPITAL
CHEST WALL DEFORMITY CLINIC

- Now accepting new and return patients at all three campuses – Main, West, Woodlands and Austin

- https://www.youtube.com/watch?v=DebGyEskDFQ
Texas Children’s Hospital is affiliated with Baylor College of Medicine in the areas of pediatrics, pediatric surgery, and obstetrics and gynecology. Currently and throughout the 60-year partnership, Texas Children’s serves as Baylor’s primary pediatric training site, and more than 1,500 Baylor faculty are the division chiefs and staff physicians of Texas Children’s patient care centers.
COMMENTS/QUESTIONS?