CHEST WALL DEFORMITIES

Celia Flores, PA-C
Department of Pediatric General Surgery
PRE-TEST QUESTION 1:

Chest wall deformities are always due to a genetic disorder.

• True
• False
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?

- Yes
- 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
Pectus carinatum is primarily a cosmetic concern.

- True
- False
PRE-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
OUTLINE

• Pectus Excavatum
• Pectus Carinatum
GENERAL OVERVIEW

Structural abnormalities of the chest wall
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

EPIDEMIOLOGY AND PATHOPHYSIOLOGY

• 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)
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:

- 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
- 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
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 – 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
PECTUS PROGRAM

TREATMENT – SURGICAL INTERVENTION

Nuss procedure **First Line**

- “Minimally invasive” thoracoscopic technique
- Curved steel bar inserted under the sternum to remodel the cartilage
- Length of treatment ~ 2-3 years

https://www.youtube.com/watch?v=R8SrRzJqbJ8
COMPLICATIONS

• Postoperative pain
• Bar displacement
• Cardiac complications

• Pneumothorax
• Recurrence
• Allergic reaction

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

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.
- Cryosurgery may reduce the amount and duration of pain after the procedure. Results are different in each patient.

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

IMAGING
You will receive a chest X-ray immediately following surgery and on the day you leave the hospital. The imaging results will be reviewed with you on 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.

PEDIATRIC SURGERY CLINIC NUMBERS
Texas Children’s Hospital
Texas Medical Center 832-822-3135
West Campus 832-227-2250
The Woodlands 936-267-7333
texasci.com/pediatric-surgery

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 to show to the airport security personnel.

Can I have an MRI done with the bar in place?
The company that produces the bar has discovered information summarized in the footnote regarding magnetic resonance imaging (MRI) 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, antibiotic 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 the weight they have lost, the bar becomes less obvious.


MRI Information The Nuss Bar Implant Part No. D1-3171 is MR-conditional according to the terminology specified in the American Society for Testing and Materials (ASTM) International, Designation F2735-06, Standard Practice for Marking Medical Devices and Other Items for Safety in the Magnetic Resonance Environment. ASTM International, 100 Barr Harbor Drive, West Conshohocken, Pennsylvania 19428. Non-conditional testing demonstrated that the Nuss Bar Implant, Part No. D1-3171 is MR-Conditional. A patient with this implant can be scanned safely immediately after placement under certain conditions. Magnetic field interactions degrade magnetic field of 3 Telsa and above. Maximum gradient magnetic field of 720 Gauss/cm or more. Please see the link listed above for MR-related heating specifications. MRI imaging may be compromised if the area of interest is in the same area or relatively close to the position of the nuss bar implant. Therefore, optimization of MRI imaging parameters to compensate for the presence of this device may be necessary. Please see the link listed above for specific on the actual information.

Pediatric Surgery Clinic Numbers
Texas Medical Center 832-822-3135
West Campus 832-227-2250
The Woodlands 936-267-7333
texasci.com/pediatric-surgery
PECTUS CARINATUM
PECTUS CARINATUM

“Pigeon chest”
Protrusion of the sternum and costal cartilages

Types:

• Chondrogladiolar prominence (most common)
• Chondromanubrial prominence
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 is primarily cosmetic
PECTUS CARINATUM – 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
  - Abnormal PSI < 2.5
- +/- genetics workup
- +/- Cardiology
- +/- Orthopedics (scoliosis)
TREATMENT – SURGICAL INTERVENTION

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**

Manually corrected by moderate compression (bracing)- done by custom fitting a circumferential external brace that puts sustained force on the most prominent portion of the sternum or cartilages in order to cause regression or flattening of the chest wall.
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.
Girls: 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 sleep on your stomach.
- 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 832-822-3133 for any questions you have about your Dynamic Compression Brace or treatment plan.
Please watch this short YouTube video to learn about the Chest Wall Deformity Clinic at Texas Children’s Hospital.

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

https://www.youtube.com/watch?v=DebGyEskDFQ
REFERRAL TO CHEST WALL DEFORMITY CLINIC

• EPIC Referral to Pedi Surgery Clinic “Chest wall deformity”
• Clinics available at Katy, Woodlands and Main Campus
POST-TEST QUESTION 1:

Chest wall deformities are always due to a genetic disorder.

- True
- False
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?

- Ultrasound of Chest Wall
- Chest MRI
- **Ch**est CT
- Bone density scan
- 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.

- Surgery
- Bracing
- Chest exercises
- 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?

- Yes
- 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
Pectus carinatum is primarily a cosmetic concern.

- True
- False
POST-TEST QUESTION 7:

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

- 1-5 years old
- 6-11 years old
- **12-18 years old**
- 18 – 25 years old

Explanation: For the majority of patients, who present during the pubertal growth spurt, the surgeon can offer a trial of bracing or surgical repair, depending on the patient's characteristics and preferences. This is when the chest wall is most compliant. This timing takes advantage of the ease of remolding an immature chest wall while minimizing the time that the defect can recur with future growth.
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?