



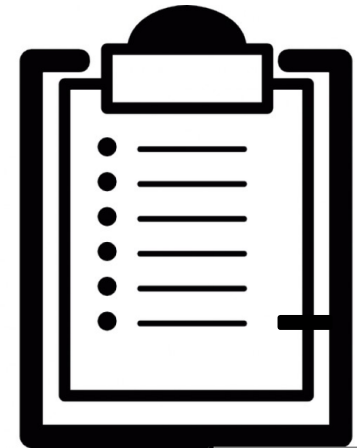
# **Chest Wall Deformities: Old Diagnosis, New Evidence**

May 6, 2021

*We will get started shortly. Your lines are muted upon entry. This event will be recorded.*

# Welcome!

- Today's event is being recorded
- All participants are muted on entry
- Ask questions using the Q&A Box
- Please fill out satisfaction survey



# About Oregon Health Forum



- Advancing health policy solutions through meaningful community dialogue
- Affiliate of The Lund Report news source
- Nonprofit supported by donations, sponsorships
- [OregonHealthForum.org/make-a-donation](https://OregonHealthForum.org/make-a-donation)

# Check out our upcoming event!



*Free Admission*

## **Racism as A Public Health Crisis: From Declaration to Action**

May 20 • 3pm • Zoom Livestream



**ADVANCING HEALTH POLICY  
THROUGH COMMUNITY DIALOGUE**

# Thank you to our annual sponsors!



**The Heatherington Foundation**  
for Innovation and Education in Health Care

**Thank you to our event co-host!**



**PeaceHealth  
Sacred Heart Medical Center**

**Continuing Medical Education**

# Chest Wall Deformities: Old Diagnosis, New Evidence

May 6 | 6-3:30pm

## Moderator:

**Farzana Molvi, MD**  
*Practicing Pediatrician*

## Panelists:

**Kimberly Ruscher, MD, MPH**  
*Pediatric Surgery, PeaceHealth*

**Garret Zallen, MD**  
*Pediatric Surgery, PeaceHealth*  
*Director of Chest Wall Deformities Program,*  
*Shriners Hospital for Children*

**Sudeshna Banerjee, MD**  
*Cardiology and Interventional Cardiology, PeaceHealth*



**Farzana Molvi, MD**  
Practicing Pediatrician



**Kimberly Ruscher, MD, MPH**  
Pediatric Surgery, PeaceHealth



**Garret Zallen, MD**  
Pediatric Surgery, PeaceHealth  
Director of Chest Wall Deformities Program,  
Shriners Hospital for Children



**Sudeshna Banerjee, MD**  
Cardiology and Interventional Cardiology,  
PeaceHealth





# Chest Wall Deformities

## Old diagnosis, new evidence

---



PeaceHealth

*The Spirit of Health*



## No disclosures

- Sudeshna Banerjee, MD, Cardiology and Interventional Cardiology, PeaceHealth
- Kimberly Ruscher, MD MPH, Pediatric Surgery, PeaceHealth
- Garret Zallen, MD, PeaceHealth Pediatric Surgery and Director of Shriners Hospital for Children Chest Wall Deformities Program



## Objectives / Outline

- Describe chest wall deformities, associated syndromes
- Indicated work up and screening for associated connective tissue disorders
- Cardiology perspective on physiology and work up
- Treatments for pectus
- Considerations in adults
- Q&A



# Why talk about chest wall deformities?

---

Kim Ruscher, MD MPH FACS

## Chest Wall Deformities: An Old Problem

*The chest becomes sharp pointed and not broad, becomes affected with difficulty breathing and hoarseness: for the cavities which inspire and expel do not obtain proper capacity.*

– Hippocrates

# Why talk about chest wall deformities?



- Common!
  - A generation of patients who were not treated
- Physiologic impact
- Psychosocial impact
- Correctable



## Case: 54 year old female

- Presents with increasing SOB with exertion
  - Lifetime history misshapen chest
  - Has seen PCP, Pulmonology, Cardiology







## Quotes from patients

- ‘I was told I had a cosmetic problem.’
- ‘My doctor told me there were no treatments for this.’
- ‘I was sent to an orthopedist and a thoracic surgeon before anyone sent me to the right person.’
- ‘I was told to get breast implants to hide the problem.’



# Defining her problem

- Role of the surgeon
  - Make a diagnosis
  - Deciding who need surgery
  - Which surgery is right



# Work up of Chest Wall Deformities

---

Garret Zallen, MD FACS



# Common Chest Wall Deformities

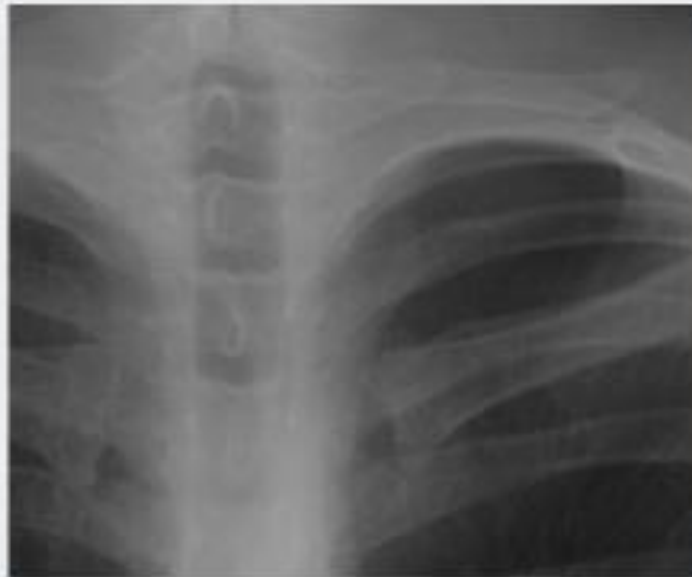
- Pectus Excavatum
- Pectus Carinatum
- Poland Syndrome
- Other Uncategorized deformities-
  - Rib flaring
  - Cervical ribs
  - Slipped Rib

# The Easy Ones

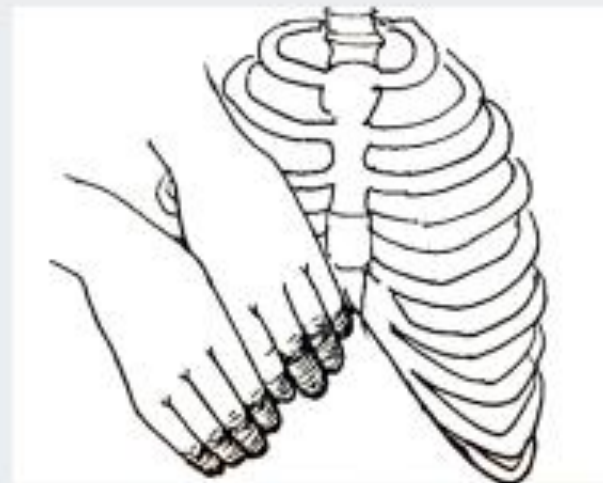
Rib Flare



Cervical Rib

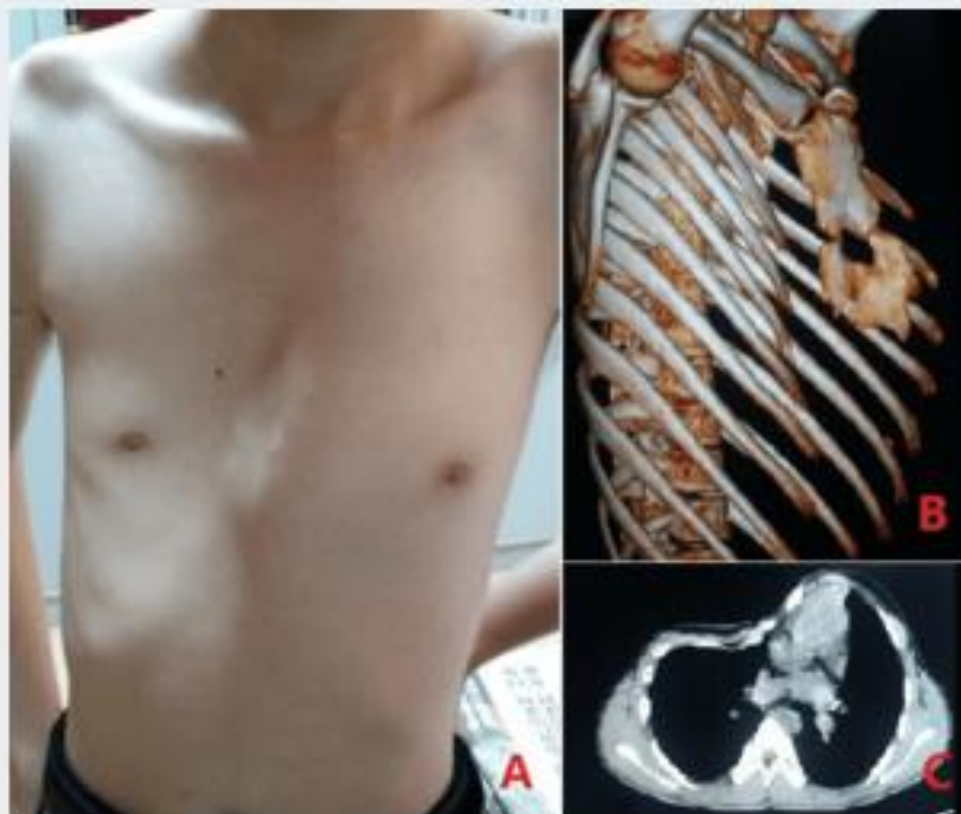


Slipped Rib



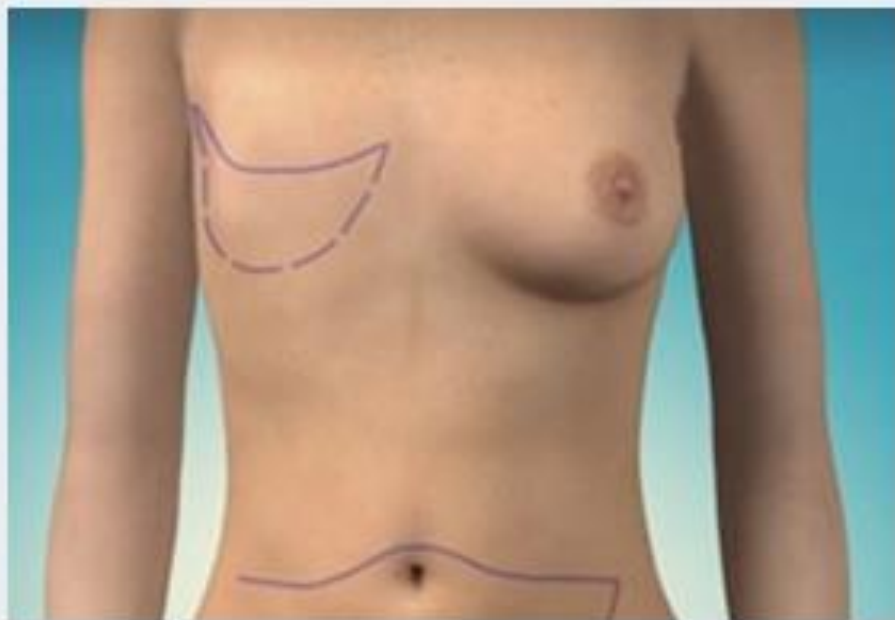
# Poland Syndrome

- Congenital absence of elements of the chest wall
  - occasionally the upper extremity
- Not progressive, but can change during childhood and puberty
- No real physiological consequences except in extreme cases
- May require bracing if sternum and ribs rotate to unaffected side



# Poland Syndrome

- Females can have significant breast asymmetry
- Reconstruction only possible after breast development is complete
- Prosthetics can be very helpful until reconstruction

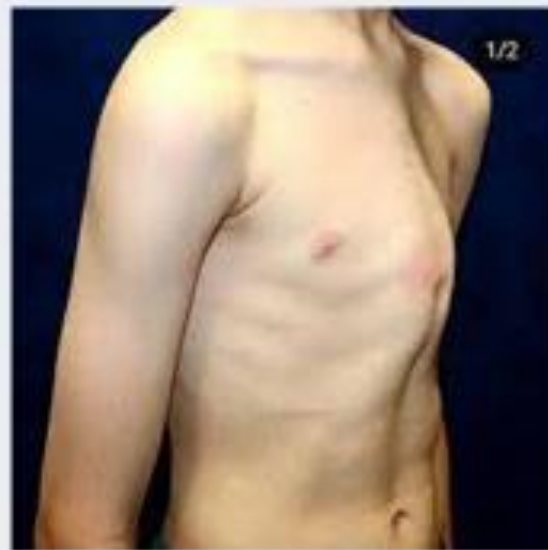




# Poland Syndrome

- **Work-up:** usually none, as this is a developmental issue and not associated with other genetic syndromes
- **Strength and function issues:** Rare, even with significant muscular hypoplasia
- **Counseling:** Setting expectations is critical, especially regarding breast development in adolescence

# Pectus Carinatum



# Pectus Carinatum

**Treatment:** non-surgical bracing

- **Highly effective:** but requires cooperation from the patient and often a significant amount of encouragement
- There may be an upper age limit to bracing

**Ravitch Procedure:** Very severe cases may require surgical correction

**Physiological consequences:** none associated with Pectus Carinatum

**Associated syndromes:** similar to Pectus Excavatum



# Pectus Excavatum

- **Frequency:** Most common chest wall deformity, 1:300-500
  - Boys > girls: or is it??
- **Presents:** at birth, or often during adolescent growth spurt
- **Physiological consequences:** Can be significant
- Non-surgical options are available, but very limited effectiveness

**Surgical correction is gold standard**





## Associated Syndromes: Pectus Excavatum and Carinatum

- Most common: Ehlers-Danlos, Marfan and Loeys-Dietz
  - Connective tissue disorders
- Depending on the sub-type may involve dilation of blood vessels
- Some may have a specific appearance-
  - Marfan (Marfanoid Habitus)
  - Loeys-Dietz (Hypertelorism)



# Associated Syndromes: Pectus Excavatum and Carinatum

- Most patients **do not** have any other associated diagnosis
- Conversely if you have Ehlers-Danlos, Marfan or Loeys-Dietz you have a high likelihood of having a chest wall issue
- Given that these are relatively rare:  
**Who and how do you screen?**
- Red Flags:
  - More severe chest wall deformities
  - Unusual body habitus
  - Family history of these disorders
  - Other joint issues or unexplained joint pains (knees or ankles)
  - Hypermobility

## Associated Syndromes: Pectus Excavatum and Carinatum

- Rule of thumb



# Work up for associated syndromes

- **Echocardiogram:** looking for aortic root and or aortic valve dilation
- **Eye exam:** specifically looking for subluxation of the lens
  - If both are negative then can essentially rule out Marfan, except in young patients
- Echo: can also help screen for the subtypes of Ehlers-Danlos that are life threatening as well as Loeys-Dietz
- **Ehlers-Danlos:** Difficult to diagnose, especially the Hypermobility syndrome sub-type (most common)
  - Currently no specific treatment
- **Genetics** consult can be very helpful



# Pectus excavatum: A Cardiology Perspective

---

Sudeshna Banerjee, MD, FACC  
Interventional Cardiology, Oregon Heart and Vascular Institute



PeaceHealth

*The Spirit of Health*

Not always as harmless as it seems...

- 59-year-old
  - Palpitations
  - Fatigue
  - Postural dyspnea
- Multiple examinations
  - PVCs
  - AV nodal tachycardia
- 6 years later: HF unknown cause
- 8 years later: orthopneic
- 10 years later: surgery

*BMJ Case Rep.* 2009; 2009: bcr10.2009.2329  
Published online 2009 Dec 14. doi: [10.1136/bcr.10.2009.2329](https://doi.org/10.1136/bcr.10.2009.2329)  
Myth exploded



# Symptoms

Exertional dyspnea

Chest discomfort

Palpitations

Exercise limitations

Less Common

- Gastric Fullness
- Dysphagia
- Reflux
- Anxiety

# Indications for Surgery

- Symptoms
- Cardiac compression on imaging
  - Up to 80% in one series
- Cardiopulmonary indications\*\*\*



# Mechanisms of Impairment

## Mechanical Compression of the Right Heart Chambers

- Limitation of diastolic filling and stroke volume
- PE Surgery results in increase RV size with corresponding increase in RV Stroke Volume (SV) and Cardiac Output (CO)
  - RV Cardiac Output: 38% increase overall, 65% increase in patients older than 30

## Poor Respiratory Mechanics

- Decreased chest wall motion and increased abdominal contribution
- Post surgery: Improved anaerobic oxygen uptake, aerobic oxygen capacity and oxygen uptake per heartbeat

## Impaired inspiratory muscle generation of negative pulmonary pressure

- Leads to decreased venous return → decreased CO



# Comprehensive evaluation

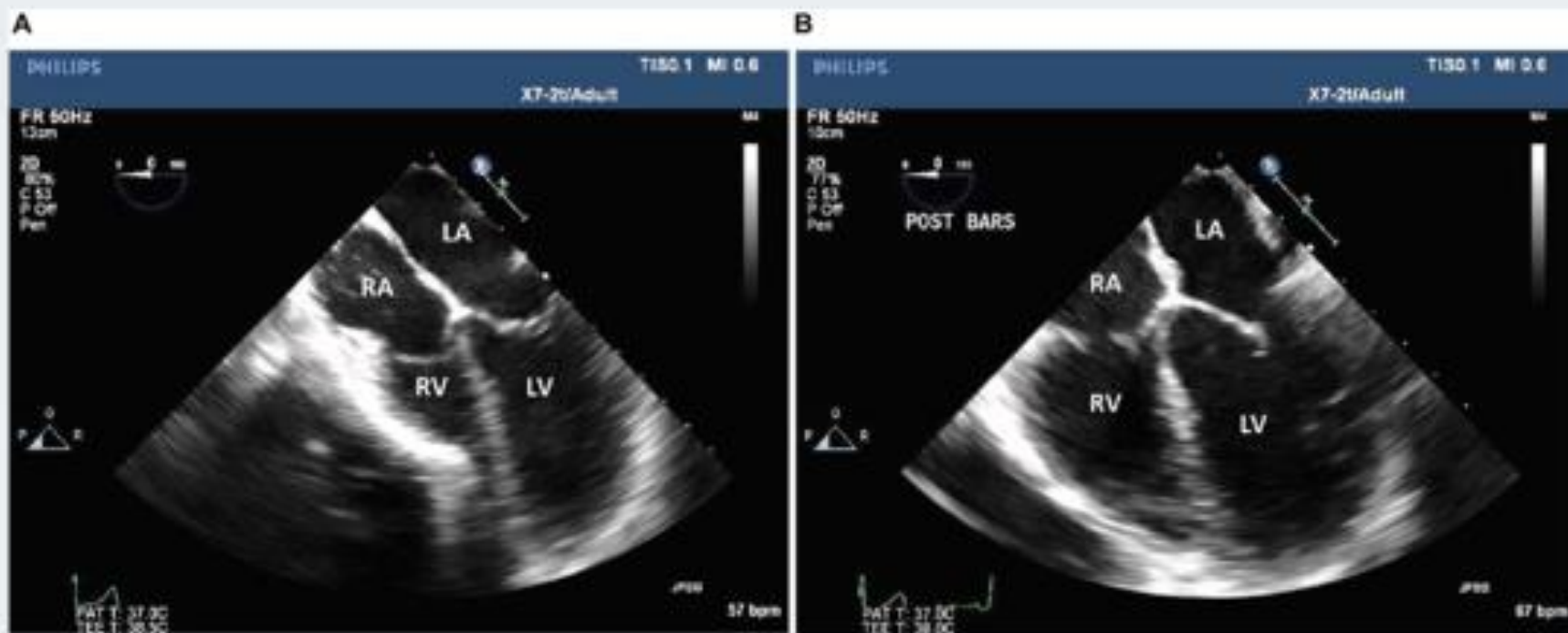
- Physical examination
  - Thoracic abnormalities (narrow chest wall, kyphosis)
  - Respiratory symptoms (tachypnea, reduced aerobic capacity, laryngomalacia)
  - Cardiac examination (tachycardia, systolic murmurs, ECG)
- Pulmonary Function
- Exercise testing (CPET)
  - Correlates with level of the severity of the chest wall deformity

# Chest CT and Xray

- Assess the degree of deformity
- Quantified by “Pectus Index”
  - Also: PSI, Haller Index
- Does not correlate with SV
- Does not correlate with subjective improvement post-operatively
- Describes cardiac displacement, degree and shape of abnormality
- Exposes patients to ionizing radiation



Fig. 2. Computed tomography with contrast showing bone projection

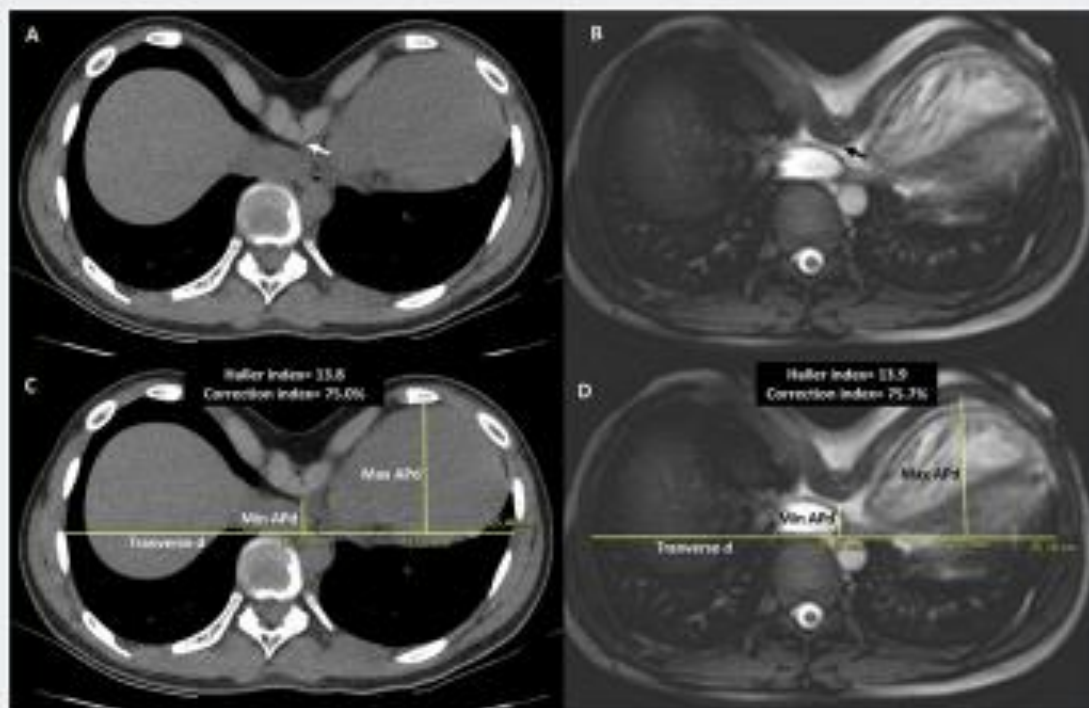


## Echocardiography

- Has demonstrated right ventricular outflow obstruction and reduced right ventricular systolic function in patients with severe PE
  - These measures improve after surgical treatment
- A study using intraoperative transesophageal echocardiography demonstrated relief of right heart chamber compression and improved cardiac output immediately after surgical repair

## Cardiac MR

- Powerful tool
- Chest and Cardiac anatomy
- Cardiac function
- Pulmonary Vascular anatomy and perfusion
- Blood flow
- Blood flow circulation times
  
- Decrease in RV short axis diameter, increase in long axis diameter and reduced RVEF
- No differences in LV parameters





# Final Thoughts

- PE is associated with significant cardiopulmonary effects
- Symptoms can increase with age
- Comprehensive evaluation with multiple modalities exist and are improving
- Surgery is high risk but successful with good outcomes in the hands of experts
- There are improvements in the objective physiologic indices with surgery as well as symptomatic relief



Thank you!



# Surgical correction

---

Kim Ruscher



PeaceHealth

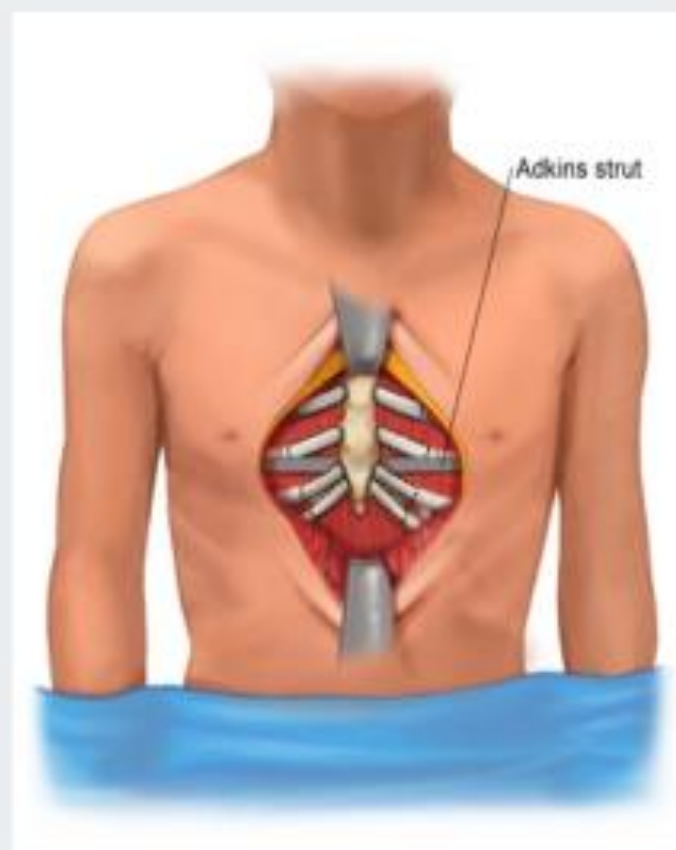
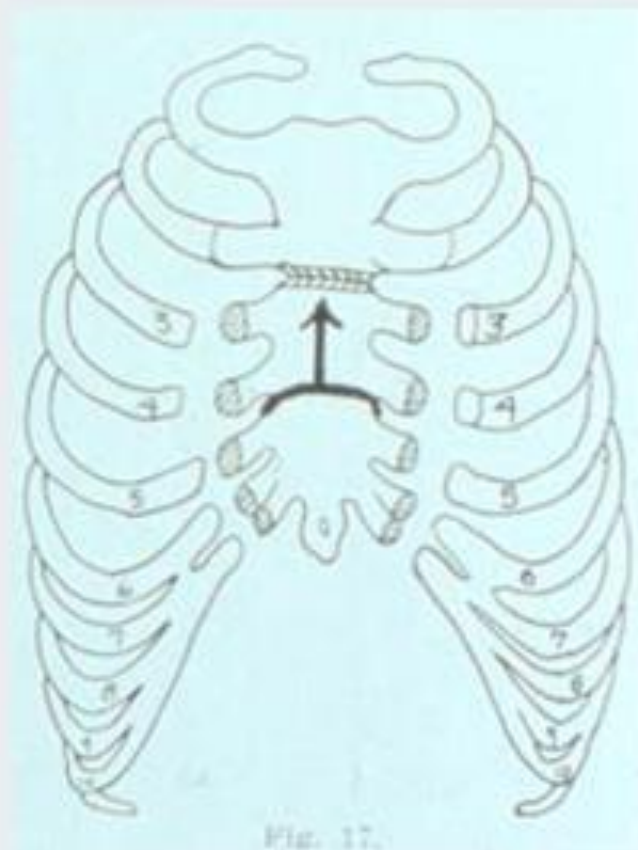
*The Spirit of Health*

# Pectus Excavatum: History



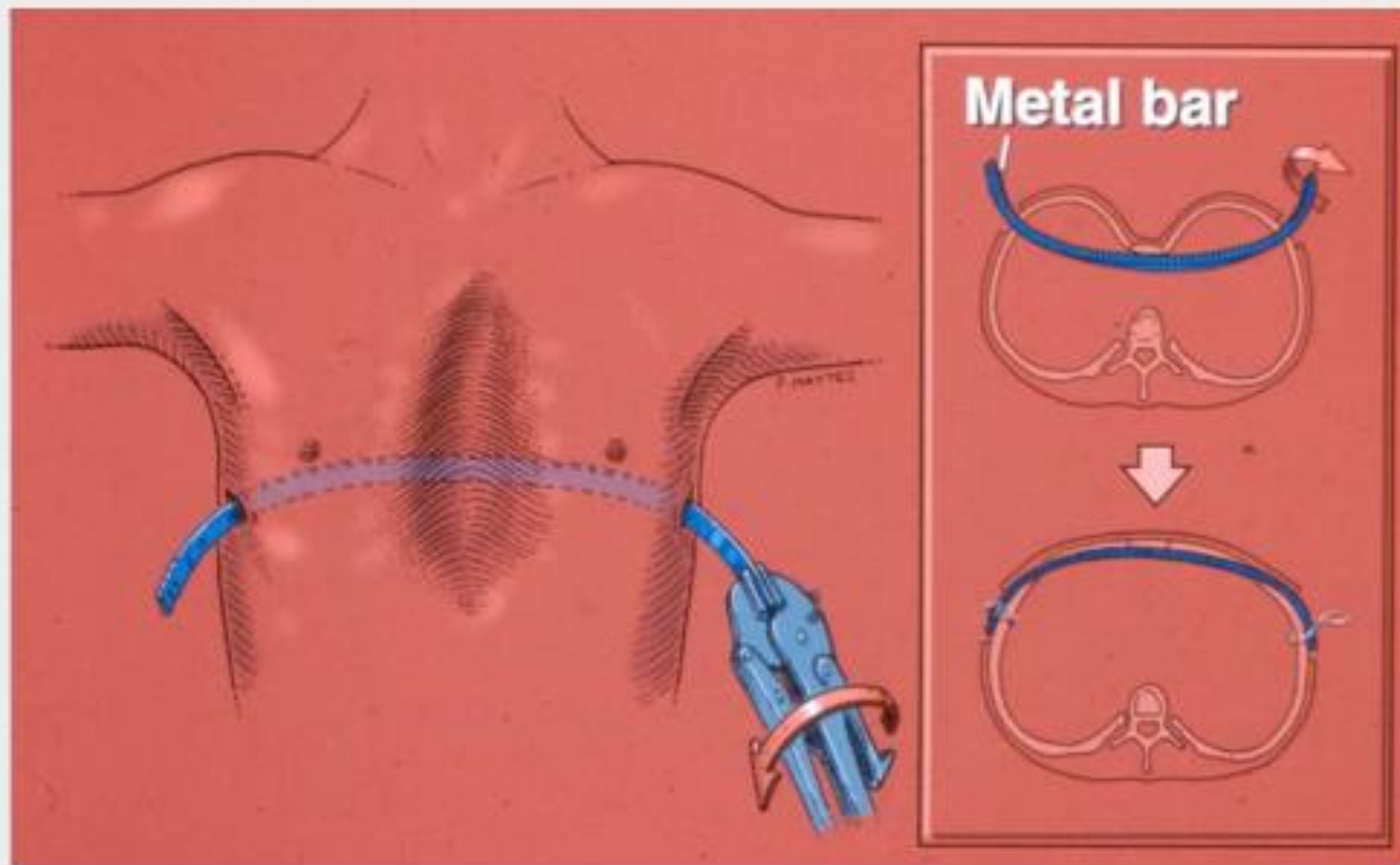
Nuss, Donald (2015, October). *Historical Perspective with Emphasis on Significant Modifications: a personal journey*. Presented at Advanced Pectus Course, Phoenix, AZ.

# Pectus Excavatum: Ravitch



Nuss, Donald (2015, October). *Historical Perspective with Emphasis on Significant Modifications: a personal journey*. Presented at Advanced Pectus Course, Phoenix, AZ.

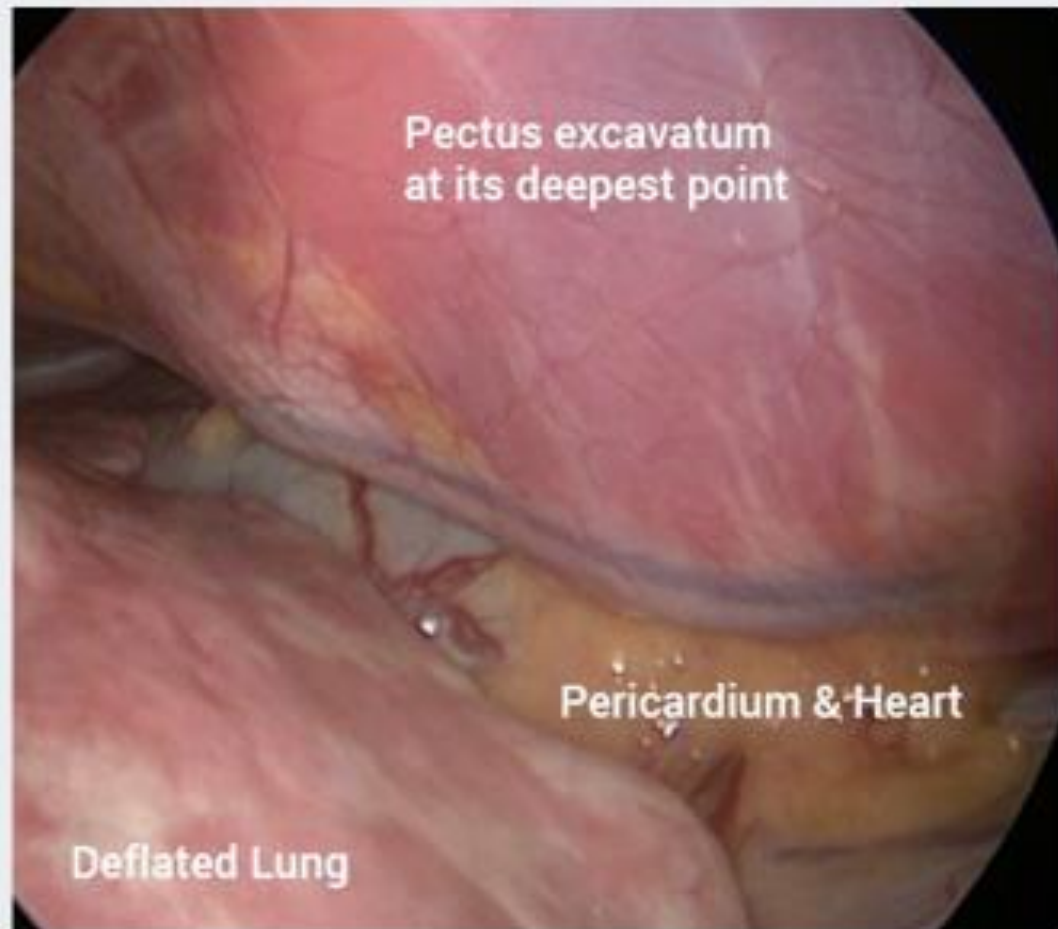
# Pectus Excavatum: Nuss procedure

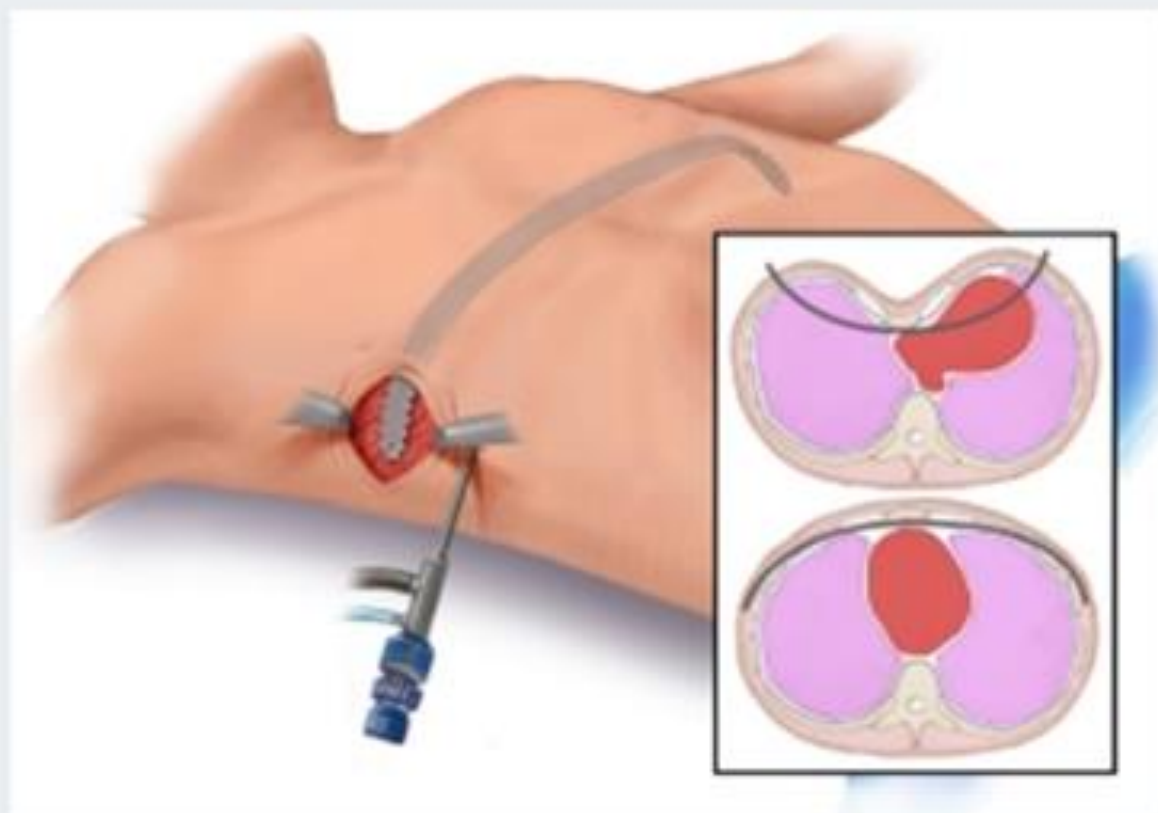


# Thoracoscopy

Visualization

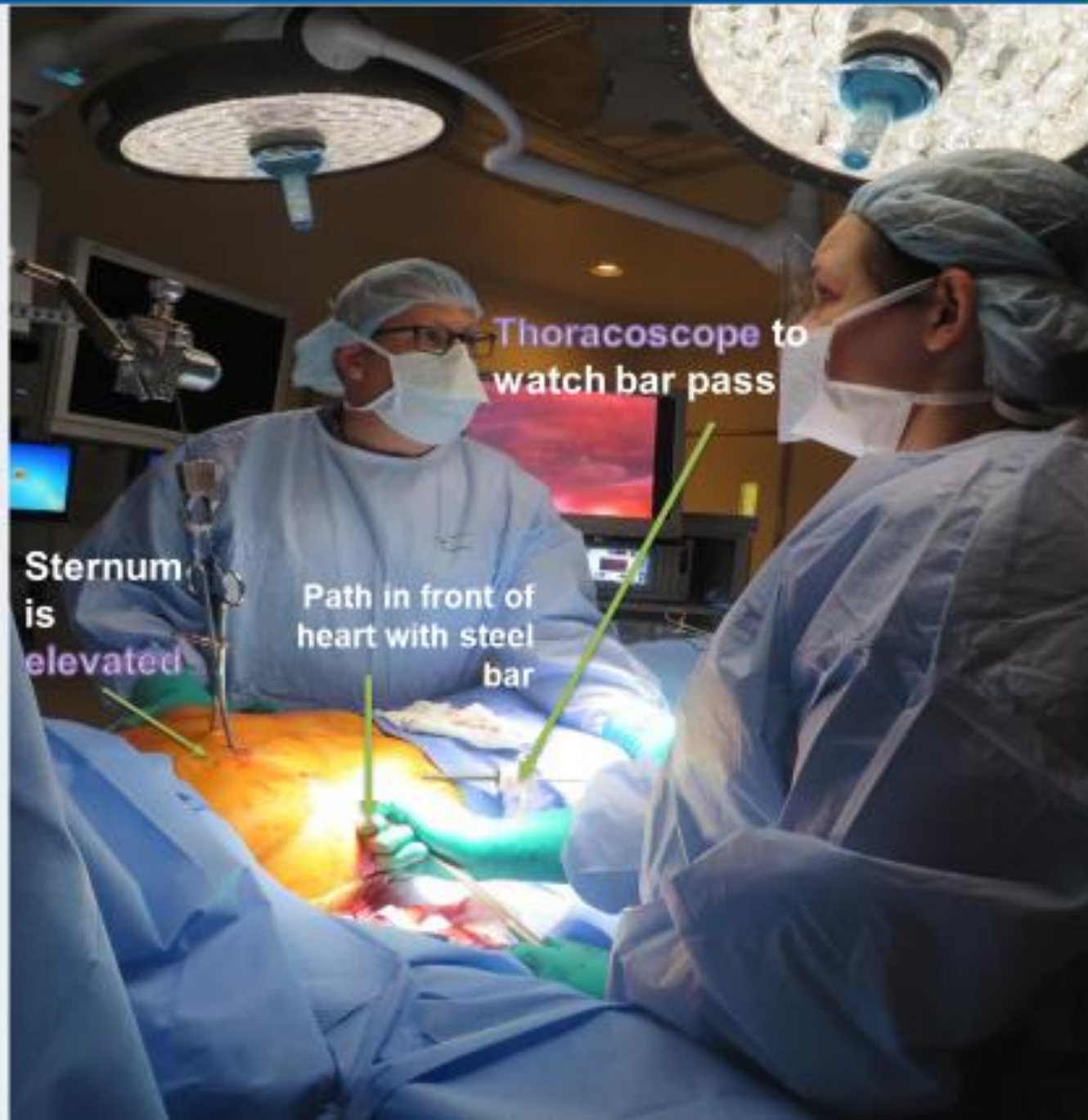
Decreased injury risk





## Minimally Invasive Repair Pectus Excavatum

Surgical standard of care



Thoracoscope to watch bar pass

Sternum is elevated

Path in front of heart with steel bar

PA  
SHIELDED





## Pectus Excavatum: pre/post





# ERAS: Enhanced recovery

## Multimodal pain relief

- Cryotherapy to intercostal nerves
- Rib blocks
- Oral and IV analgesics

Early ambulation and diet

Hospital stay 1-2 days



# Considerations in the adult with Pectus Excavatum

---

Garret Zallen



# The adult chest wall

- More rigid, resists correction
- Pectus Excavatum: Nuss bar may not hold up the chest wall
  - May require multiple bars, intercostal reinforcement and/or osteotomies or rib resections
- Pectus Carinatum: Adult ribs can remodel (COPD) but require more force to change shape.
  - This force may exceed the tolerance of the skin



# Adult physiology

- Maximal heart rate drops as we age
  - Direct decrease in cardiac output in severe Pectus Excavatum; it is rate dependent, not filling dependent- asymptomatic → symptomatic
- Nerve regeneration is slower
  - Prolonged effects of cryoablation
  - More issues with neuropathic pain
- Higher risk procedure
  - Especially if existing cardiac dysfunction (CAD or heart failure)

## Pectus and associated syndromes

- Timing of surgery needs to be coordinated around the need for repair of cardiac lesions
- Post operative management can be more difficult
- Higher post-operative complication rates

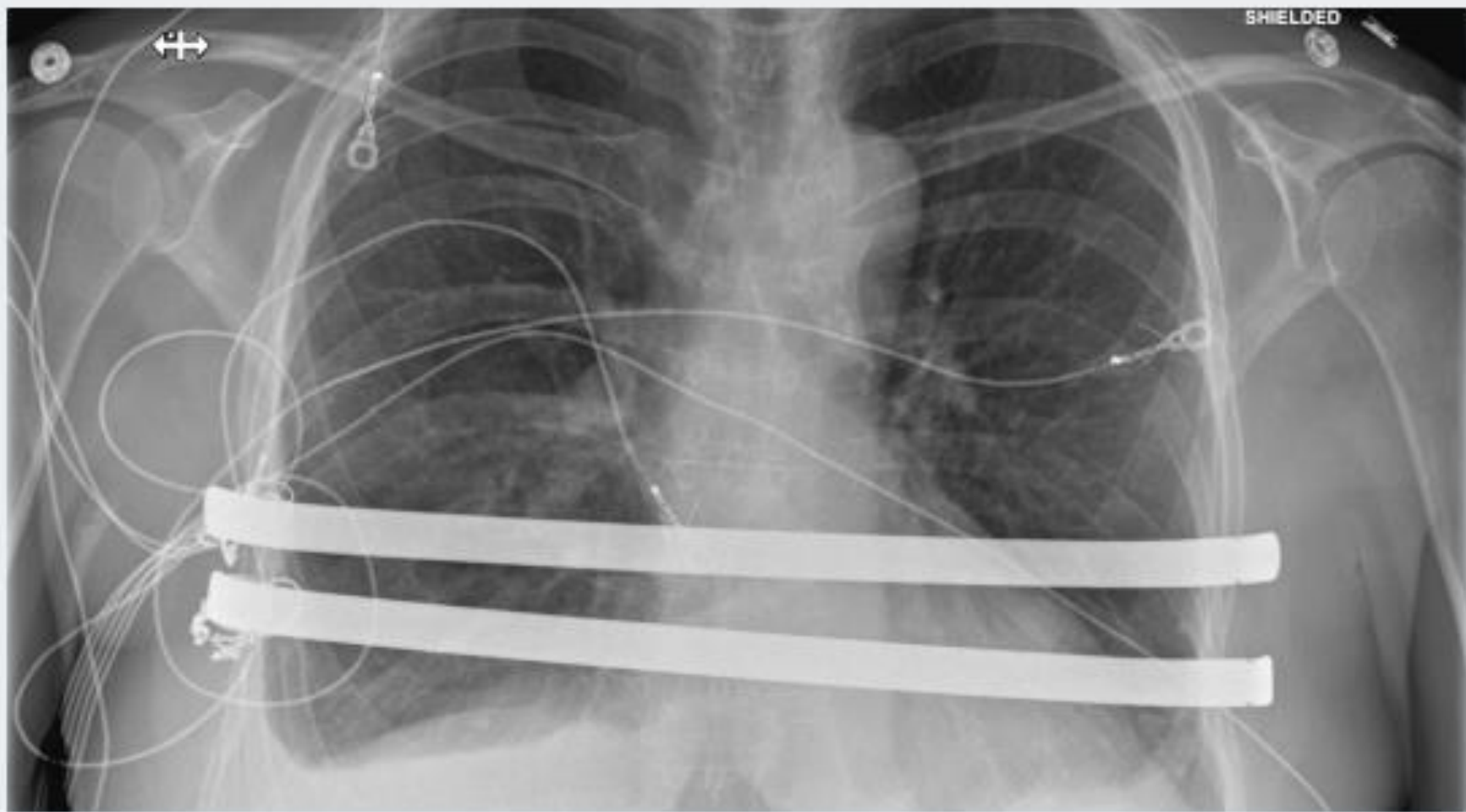




## Psycho-social considerations

- May have been mis-diagnosed and/or treated with inappropriate therapies
  - frustration, mistrust, depression and anger
- May have had a failed Ravitch procedure and reluctant to have additional procedures
- Were told that getting it repaired was unnecessary- stigma
- Gave up...as they didn't know anyone treated adults
- Safe to get pregnant with a Nuss Bar in place

# Our patient





# Summary: Chest Wall Deformities

- Common, correctable
- Often missed or underdiagnosed, especially in females
- Can have real physiological and emotional consequences
- Association with connective tissue disorders
- Safe, evidence-based treatment options
- Optimal time for repair is during or around adolescence
  - Even older adults can safely have correction
- Just covering the defect in Pectus Excavatum does nothing to solve the problem
- We are happy to see and evaluate children and adults or help with questions



## Real world – Eugene patient

*I was 12 when I was diagnosed with pectus excavatum. I began experiencing shortness of breath, a racing heart and pain—I had to stop doing all of the things that I enjoyed. I couldn't go hiking, I would get dizzy and I'd have to stop because I was afraid. I couldn't run, and I love to run. I looked so different that I felt really insecure.*

*This summer I hiked to the top of Spencer's Butte – I made it all the way to the top. It was amazing.*



PeaceHealth

*The Spirit of Health*

# Thank you to our annual sponsors!



**The Heatherington Foundation**  
for Innovation and Education in Health Care

**Thank you to our event co-host!**



**PeaceHealth  
Sacred Heart Medical Center**

**Continuing Medical Education**

# Audience Q&A

**Thank you for attending!**