Do I Need a Pediatric Surgeon?

Common Reasons for a referral

Bryan Dicken, MSc, FRCSC, FAAP
Stollery Children's Hospital
University of Alberta

Faculty Disclosure

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Objectives

* At the completion of this discussion, participants will be able to:
  1. Identify the child at risk of Hirchsprung's disease, and initiate a work-up and referral
  2. Identify the child at risk of Biliary Atresia, and the appropriate investigations and referral
  3. Recognize the spectrum of chest wall anomalies, and initiate appropriate investigations and referral

Constipation vs. Hirchsprung's

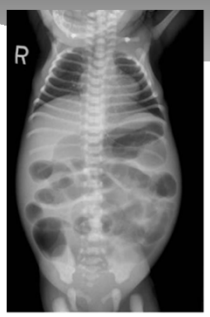
- Case:
  - 2 day old infant
    - Failure to pass meconium with 24 hrs of life
    - Abdominal distension
    - Feeding intolerance (frequent "spitting up")
    - Now with bilious emesis
  - PMHx
    - Unremarkable
    - No family history, uneventful spontaneous vaginal delivery, all the "parts" are present (ie: normal anus)

Constipation vs. Hirchsprung's

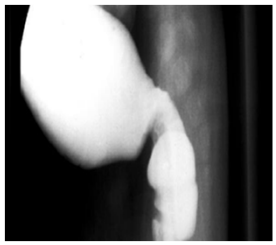
- What now?

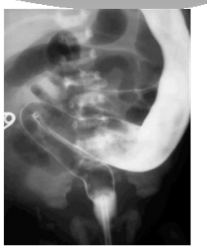
Constipation vs. Hirchsprung's

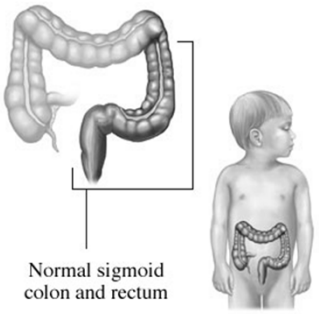
Now What?

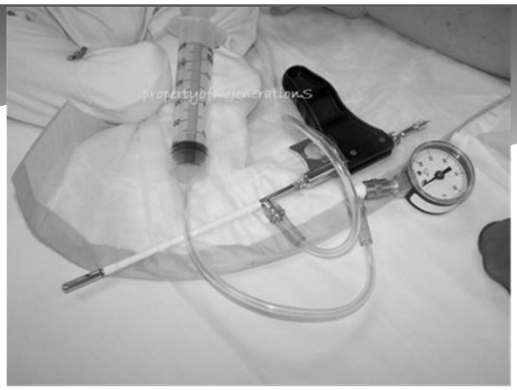


Hirchsprung's?

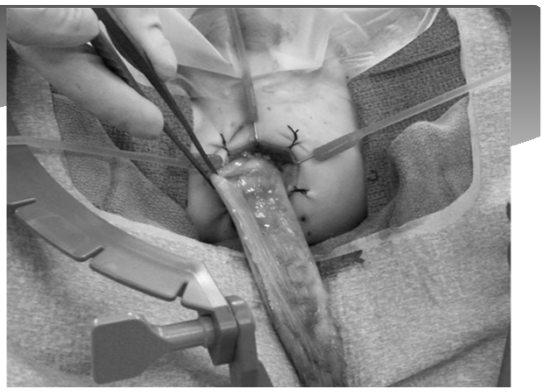








Suction Rectal Biopsy



That is not what we are talking about!

- More commonly, 2 – 5 yrs old
  - Presents with abnormal stooling as "long as the mother can remember"
  - Frequently "seems to be in pain" when trying to stool, the "nothing" comes out for 2 weeks
  - Associated with abdominal distension
  - Normal growth and development
  - Have tried everything under the sun to initiate stooling
- What now?

Constipation vs. HD

- The devil is in the details, specifically the history
  - 50-90% of HD present during the neonatal period with abdominal distension, bilious vomiting, and feeding intolerance
  - Delayed meconium (>24 hrs) occurs in 90%.
    - Problem, often parents cannot remember, or perceptions are strongly influenced by recent events

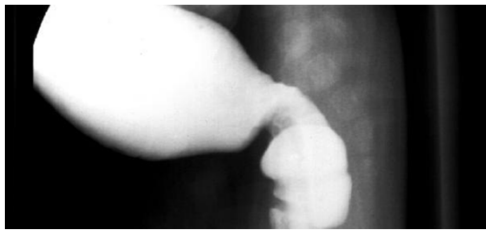
HD?

- Features suggestive of HD:
  - Failure to thrive
  - abdominal distension
  - need for enemas/stimulation for evacuation.
- Features NOT suggestive of HD:
  - Delayed onset (ie: develop around potty training)
  - Incontinence (overflow incontinence)
  - Toilet pluggers

HD?

- Investigations:
  - If child is well, and has no history of suggestive features, then bowel management routine and reassurance is appropriate
  - If child has suggestive features
    - Start with bowel management routine
    - Arrange contrast enema
      - Look for reversed rectum:sigmoid ratio, transition zone, or retained contrast on 24-hr post-evacuation film

HD?



What now?

HD?

- Refer to pediatric surgeon for OPEN rectal biopsy
  - Bedside suction rectal biopsy only effective in children < 12 months old.
  - Older children require full-thickness biopsy for confirmation.
- Treatment
  - In older children, often requires diverting ostomy with delayed pull-through due to over-distended colon (depends on contrast study)

Jaundice

- Jaundice is COMMON, and that is the problem!
  - Majority of cases hyperbilirubinemia (HB) is unconjugated and resolves without intervention
  - Importance of identifying the rare infant whose Bilirubin persists beyond 2WEEKS cannot be overstated
    - These neonates should be presumed to have biliary obstruction from choledochal cysts or biliary atresia until proven otherwise

Jaundice

- Biliary Atresia
  - 1:10,000
  - Female predominance
  - No genetic etiology, but higher in Asia than in both Europe and North America
  - Etiology unknown and likely multifactorial
  - 20% are associated with other congenital malformations

Jaundice

- Case:
  - 10 day female infant presents for routine check-up
  - Parents concerned about "yellow color"
    - Neonatal bilirubin:
      - Day 1 of life (98 umol/L [N < 120])
      - Day 2 of life (181 umol/L)
      - Day 3 of life (232 umol/L)
      - Conjugated bili (116 umol/L [N< 5 umol/L) 12 weeks of age
      - Cbili 137 umol/L at 16 weeks of age

Jaundice

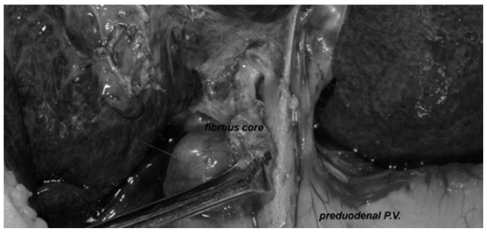
Jaundice

- Combination of progressive jaundice, acholic stools, dark urine and hepatomegaly should raise suspicion
  - Jaundice refers to elevated DIRECT bilirubin, which is not reflected in neonatal assessment
  - 50% of cases stools are initially pigmented
  - Outcomes strongly linked to timing of surgical intervention
    - Surgery in infants less than 12 weeks of age had significantly better outcomes than those beyond 12 weeks

Jaundice

- Back to case:
  - Patient referred to Stollery for assessment
    - Ultrasound – assess for GB, liver texture, exclude alternate obstructive etiologies
    - Labs – assess residual function, and exclude perinatal infections
    - Liver biopsy – accurate nonsurgical test to differentiate from other cholestatic causes
    - HIDA scan – less valuable in cases with extensive jaundice and compromised function
  - Referred for surgical consultation

Biliary Atresia



Biliary Atresia

- Outcomes
  - If Kasai done within 12 weeks
    - 30% long-term bile flow
    - 30% initial flow, then fail
    - 30% fail immediately
      - Delayed diagnosis, infection
      - Liver fails within months
      - 94% mortality if untreated

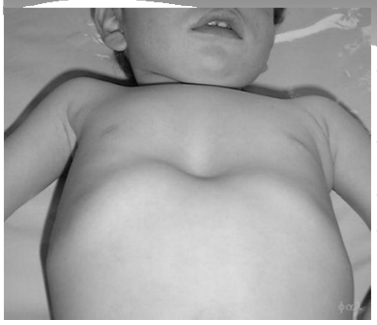
Biliary Atresia

- Bottom:
  - BA is rare, but consequences of missed diagnosis very high
  - Jaundice/Hyperbili beyond 10 days of life should be assessed with fractionated bilirubin, and followed for trend
    - If in doubt refer/consult early

Pectus Excavatum

- Most common congenital chest wall deformity
- 1 in 300 live births; 3:1 male predominance
- Results from unbalanced posterior growth of costal cartilages that are often fused, deformed or rotated
- 90% identified during infancy, becoming more pronounced at puberty

Pectus Excavatum



- Body of sternum exhibits posterior curvature
- Right side often slightly more curved in
- Familial in 1/3 of cases
- **Marfan syndrome** predisposition

Pectus Excavatum

- General:
  - Worsening of appearance and symptoms reported during periods of rapid bone growth
  - Teenagers
  - 30% familial
  - Etiology is unknown
    - Intrinsic abnormality of coastal cartilage suggested
      - Marfan Syndrome (5 – 8%)
      - Ehlers – Danlos Syndrome (3%)
      - Scoliosis (15 – 21%)

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Pectus Excavatum

- Presentation:
  - Highly variable!
    - Chest and Back Pain
    - Poor exercise tolerance
    - Palpitations
    - Cardiac Murmur – Mitral valve prolapse
    - Body-image and psychological/interpersonal-related complaints

Objective Testing

- Cardiovascular Testing:
  - Echo indicated pre-operatively
    - MVP present in 17 – 65% (versus 1% population)
      - Resolves in ~ 50% of patients post-op
    - Right atrial and ventricular compression
    - Dysrhythmia in 16%
    - Cardiac work capacity related to severity of pectus defect
      - Stroke volume from rest to exercise was 18.5%
        - expect 51% increase in normal subjects
        - Post-op, cardiac index increased by 38% due to improved stroke volume

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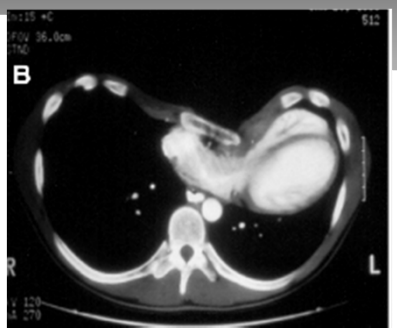
Cardiac Compression



Selection Criteria

- If 2 or more criteria apply:
  1. CT chest shows cardiac/pulmonary compression
  2. CT index > 3.25
  3. Echo shows compression, displacement, MVP, murmur or conduction abnormality
  4. PFT shows restrictive and/or obstructive disease
  5. Previous repair has failed

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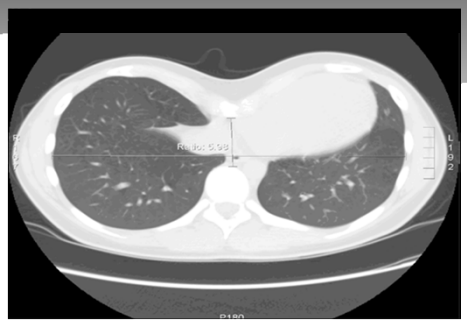
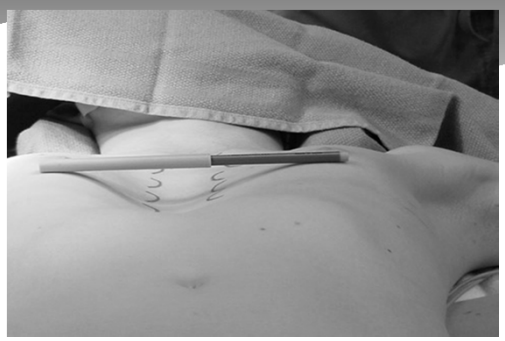
Pectus Excavatum



Haller Index = 5.98

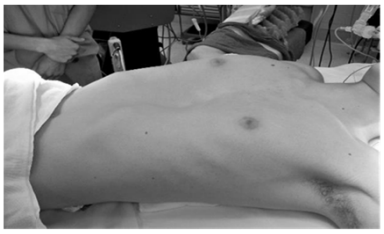
Pectus Excavatum



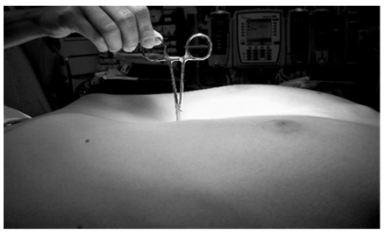
Pectus Excavatum

- Symptoms – chest pain, SOB
- 2% have CHD
  - Echocardiogram + PFTs
- Surgical correction:
  - Optimal timing in late childhood and adolescence
    - More pliability of costal cartilages (to prevent thoracic dystrophy)
  - Relation to cardiopulmonary impairment debatable
  - Improvements in exercise tolerance, SOB, fatigue

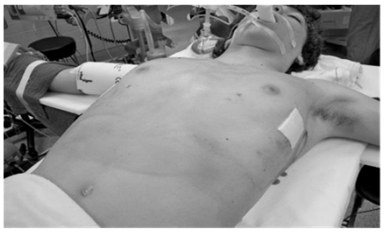
Pectus Excavatum



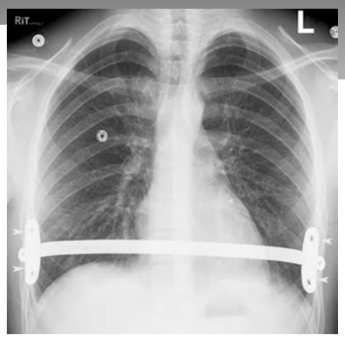
Pectus Excavatum



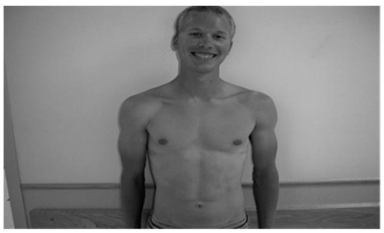
Nuss Bar

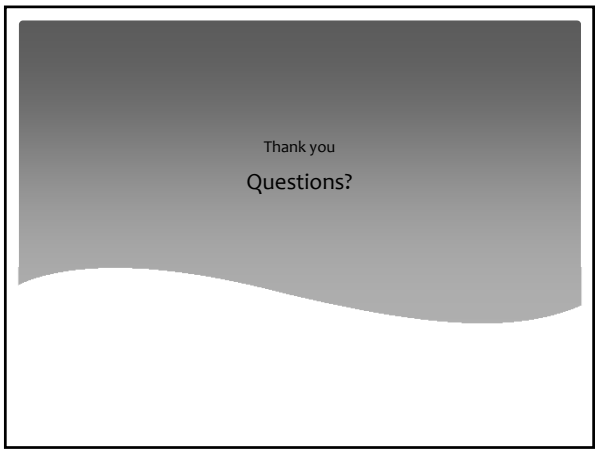


Nuss Bar



Post-op Nuss Procedure




Thank you
Questions?