Current Topics in Pediatric Surgery for the Primary Care Provider

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Speakers Disclosure Statement

Dr. Gates discloses that he has no relevant financial relationships with commercial interests.

Dr. Gates does not anticipate discussing unlabeled uses of any commercial products or any investigational products.
Introduction

- Vascular Malformations
- Common Skin Lesions
- The Coccygeal Dimple
- Breast masses in adolescents
- The latest in Inguinal Hernia Repair
- Intussusception
- The Spectrum of Malrotation
- Pyloric Stenosis
- Pectus Deformities
Vascular Malformations
Vascular Malformations

• Venous malformations
  – Hemangiomas
    • Rapid growth phase between birth and 3 months
    • Gradually involute and usually resolve by age 5-7
    • Large or more than one
      – Liver ultrasound to evaluate for other lesions
  • Indication for surgery
    – Arteriovenous shunting
    – Platelet consumption (Kassebach-Merritt)
    – Bleeding and ulceration
• Medical therapies
Vascular Malformations

- Venous Malformations
Vascular Malformations

- Lymphatic Malformations
  - Usually a collection of abnormal lymphatic channels with a vascular component
  - Microcystic and macrocystic
  - Role of ultrasound and MRI
    - Characterization
    - Deep tissue extent
Vascular Malformations

• Lymphatic Malformations
  – Complications
    • Infection and cellulitis
    • Anatomic dysfunctions
      – Tracheal occlusion
      – GI tract
        » Protein loss
        » Pancreatitis
        » Obstruction
        » Chylous ascites
      – Chest
        » Chylothorax
        » Venous Occlusion
Vascular Malformations

• Lymphatic Malformations
  – Treatment
    • Surgery
    • Sclerotherapy – Ethanol, Picibanil
    • Laser – Integrated with other therapies and spaced over months
    • Radiofrequency ablation
    • Chemotherapy - Rapamycin
Vasuclar Malformations

- Lymphatic malformations
Common Skin Lesions

• Pilomatrixoma
  – Calcifying epithelioma of Malherb
  – Benign hair follicle tumor
  – Diagnosis: Pearly, mobile, subcutaneous lesion
  – Associations
    • Gardner’s syndrome
    • Rubenstein-Tybi syndrome – myotonic dystrophy
      – Genetic counseling
• Facial Dermoid Cysts
  – 50% of childhood orbital neoplasms
  – Stratified squamous epithelium mixed with multiple cell types
  – Form during embryogenesis as the fetal suture lines close trapping an epithelial rest
    • Most common in the fronto-zygomatic suture
Common Skin Lesions

• Lymph Nodes
  – Differentiate between acute, subacute, chronic
  – Determine possible infectious causes
    • Chronic otitis, pharyngitis
    • Cat scratch disease
    • Mycobacterium
Common Skin Lesions

• Lymph Nodes
  – Signs suggesting malignancy
    • Size > 2.5 cm
    • Firm/fixed
    • Supraclavicular or axillary
    • Non-tender
    • Progressive
    • “B”Symptoms
  – Role of ultrasound
Coccygeal Dimples
Coccygeal Dimples

• Incidence – 4.8% of children
  – Problematic lesions
    • Spinal dysraphism – 1/2500
    • Spinal lipoma – 1/4000
    • Dermal sinus – 1/2500
Coccygeal Dimples

• Indications of High Risk
  – Larger than 0.5 cm
  – Located more than 2.5 cm above the anal verge
  – Overlying cutaneous lesions
    • True hypertrichosis
    • Skin tags
    • Telangectasia
    • Subcutaneous mass
    • Aplasia cutis
    • Abnormal pigmentation
  – Asymmetric gluteal crease
Coccygeal Dimples

• Further evaluation
  – Avoid probing
  – Ultrasound imaging
    • Prior to 4 months of age
    • Operator sensitive
  – MRI
    • Requires sedation
  – Neurosurgical consultation
Coccygeal Dimples
Breast Masses in Adolescents
Breast Masses in Adolescents

- Infections
  - Treatment

- Asymmetric breast bud development

- Gynecomastia
  - Usually unilateral
  - Usually resolves as puberty progresses
  - Don’t forget the testicular exam!
Breast Masses in Adolescents

• Tumors
  – Incidence: 3.2% teenage girls
  – 95% will be a benign fibroadenoma
    • 10% regress spontaneously
    • < 0.02% are malignant
    • Juvenile fibroadenoma or Giant fibroadenoma
  – Other tumors
    • Phyllodes
    • PASH
    • Intraductal papilloma
    • Lymphangiomas
    • Malignancy
Breast Masses in Adolescents
Inguinal Hernias
Inguinal Hernias

- Congenital hernia
Inguinal Hernias
Inguinal Hernia
Inguinal Hernia

• Evaluation
  – Hydration status
  – Abdominal x-rays
Inguinal Hernia
Inguinal Hernia

• Evaluation
  – Hydration status
  – Abdominal x-rays

• Reduction
  – Dark, quiet room
  – Hydration
  – Consider conscious sedation
FIG. 64-2. Manual reduction of an inguinal hernia. Two fingers form an inverted V to allow reduction through the external inguinal ring.
Inguinal Hernia

- **Evaluation**
  - Hydration status
  - Abdominal x-rays

- **Reduction**
  - Dark, quiet room
  - Hydration
  - Conscious sedation

- **Timing of Repair**
  - Allow time for edema to resolve
  - Within 1-2 weeks
  - Parent education
Inguinal Hernia
Intussusception
Intussusception
Intussusception

- Abdominal Pain
- Vomiting
- Bloody Stools
  - sign of intestinal ischemia and mucosal sloughing.
Intussusception
Intussusception
Intussusception Management

• OPERATIVE
  – Manual reduction with appendectomy (50%)
  – Resection (45%)
    • Necrotic bowel
    • Pathologic lead point
  – Spontaneous reduction (5%)
Intussusception Management

- Radiologic Reduction
  - Reduced (>90%)
  - Not Reduced (<10%)
  - Perforated (<1%)
- Mortality (<1%)

- Admit
- Reduced (60%)
- Repeat Enema
- Not Reduced (40%)
- Operation (10%)
- Recur (5%)
Intussusception

- **Lead points**
  - more common in *older* children
  - nearly always found in adults with intussusception
Intussusception

**Lead Points:**

- Meckel’s diverticulum (most common)
- polyps
- duplications
- lymphomas
- submucosal hemorrhage with Henoch Schoenlein purpura
- hemangiomas
- lymphosarcomas
- inspissated meconium in the terminal ileum (Cystic Fibrosis)
Intussusception
Post-operative Causes

- Unique to pediatric population
  - Prolonged surgery with extensive bowel retraction
  - Wilm’s tumor
  - Nissen fundoplication
  - Retroperitoneal surgery
- Often confused with post-operative ileus
- Symptoms develop 1-2 weeks after surgery
  - Distension, nausea/vomiting, cramping
    - Suspect if marked increased volume of gastric drainage
  - Ultrasound for diagnosis
- Surgical exploration required
Reduction of Intussusception
Malrotation
Malrotation

- Wide variations of severity
- Abnormality of rotation of the midgut around the superior mesenteric artery with subsequent abnormal fixation of the midgut in the peritoneal cavity.
Gasless abdomen = volvulus
Rotational Anatomy
6 – 9 weeks
Rotational Anatomy
11 – 12 weeks
Final Rotational Anatomy
Ladd’s Procedure

- Derotation
- Division of Ladd’s Bands
- Widen the mesenteric pedicle
- Appendectomy
- Place Colon on left, small bowel on right
• Late Presentation
  – Chronic, intermittent abdominal pain
  – Incidental finding for other abdominal surgery
Pyloric Stenosis
Pyloric Stenosis

• Incidence – 4/1000 live births
  – 5:1 boys to girls

• Presentation
  – Non-bilious projectile vomiting
  – Visible peristaltic waves in LUQ
  – Hypochloremic metabolic alkalosis
  – 75% will have a palpable “olive”
Pyloric Stenosis

• Ultrasound
  – Operator dependent
  – Pyloric dimensions
    • Thickness >3 mm
    • Length >14 mm

• Upper GI
  – Elongated pyloric channel
  – Antral shouldering
  – May be confused with pyloric spasm
Pyloric Stenosis

- Repair
  - Open
    - RUQ or umbilical incision
  - Laparoscopic
    - Minimal scar
    - Faster recovery
    - Faster return to full feeds
    - Less post-operative emesis
    - Decreased pain
Pyloric Stenosis
Chest Wall Deformities

- Pectus Excavatum
- Pectus Carinatum
- Poland Syndrome
- Failure of Sternal Fusion
- Asphyxiating Thoracic Dystrophy
  - Cerebrocostomandibular Syndrome
  - Jarcho-Levin Syndrome
  - Jeume Syndrome
Chest Wall Deformities

- Pectus Excavatum
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Chest Wall Deformities

Pectus Excavatum

Pectus Carinatum
Pectus Excavatum

- Incidence = 1/1000
  - M>F (4:1)
- Multiple patterns of inheritance
- Variations
  - Punched in
  - Saucer shaped
  - Eccentric
  - Horns of Steer
  - Costal flare
Variations
Punched In
Variations
Saucer Shaped
Variations
Eccentric
Variations
Horns of Steer
Variations
Costal Flare
Pre-operative Symptoms

Data from Kings Daughters Children's Hospital
Cardiac Effects

• Most common symptoms:
  – Dyspnea
  – Exertional chest pain
  – Palpitations

• Cardiac Compression  85%
• Cardiac Displacement  73%
• Mitral Valve Prolapse  13%
• Arrhythmias/Murmurs  27%
Cardiac Compression

- Long term effects
  - Decreased cardiac output
  - Similar to RV dysplasia
    - Arrhythmias, V-tach
    - Sudden death as young adult
  - Trabecular hypertrophy
- 95% fully resolved after surgery
  (Coln 2006)
Mitral Valve Prolapse

• 1% Incidence in pediatric population
• 43 - 65% incidence in patients with pectus excavatum
  – 43 to 93% resolution after pectus repair
  (Shambarger 1987, Coln 2006)
Pulmonary Effects

- Left lung compression
- Increased work of breathing
  - restrictive abnormality
- PFT mild → severe restrictive, obstructive airway
- Atelectasis
- Exertional dyspnea
- Increased incidence of pneumonia, asthma, bronchiectasis
Pulmonary Effects

- After placement and subsequent removal of the pectus bar, patients have significant improvement in pulmonary function.
- Children over age 11 have a greater degree of improvement.

(Goretsky 2005)
Psychosocial Effects

- 8 of 9 psychosocial indicators showed statistically significant improvement, including:
  - Body-image satisfaction
  - Feelings of sadness, frustration, restlessness, and isolation
  - Experience of social ridicule

(Lawson 2003)
Workup Prior to Surgical Referral

- Complete History and Physical Exam
- Radiographic studies
  - PA/Lateral chest x-ray
- Other radiographic and functional studies are patient specific and will be obtained AFTER the surgical consult
History

• When did the deformity manifest
  – Infancy vs. older child
  – Increase in depth often occurs during periods of rapid growth
  – Patients with pectus excavatum from infancy will have fewer symptoms due to adaptation

• Type of physical activity and change

• History of allergies
  – Jewelry, metal
Physical Exam

- Height / weight percentiles
- Cardiac exam - displacement of PMI
- Type of Pectus
- Symmetry / costal flaring
- Scoliosis / joint laxity / stria
Criteria for Referral

- Age over 8
- Cardiopulmonary symptoms
- Significant increase in deformity
Surgical Evaluation and Treatment Pathway

History and Physical Chest X-Ray

Mild

Physical Therapy

Follow-up in 6 months
Surgical Evaluation and Treatment Pathway

History and Physical
Chest X-Ray

Mild
Physical Therapy
Follow-up in 6 months

Severe
Physical Therapy

Pectus Bar
Surgical Evaluation and Treatment Pathway

History and Physical
Chest X-Ray

Mild
Physical Therapy
Follow-up in 6 months

Severe
Physical Therapy

CT Scan
Echocardiogram
Pulmonary Function Tests

Pectus Bar
So... what is a severe defect?
Radiologic Evaluation

Haller Index

\[ \frac{26 \text{ cm}}{2.5 \text{ cm}} = 10.4 \]
Severe Pectus Excavatum

• Haller index greater than 3.2
• Cardiac and pulmonary symptoms
  – Exercise intolerance
  – Chest pain with exertion
  – Cardiac murmur
Goals of Physical Therapy

• Strengthen chest wall musculature
• Improve chest wall flexibility
• Increase cardiovascular endurance
• Improve posture
• Deep breathing – chest expansion
Pectus Excavatum

• Effectiveness of Physical Therapy Program
  • Decreased post-operative length of stay
  • Earlier ambulation
  • Transferred supine to sit independently earlier
  • Improved post-operative pain scores compared to those children who received epidural catheters
Indications for Surgery

- Haller index > 3.2 in patients with longstanding pectus excavatum
- Decreased pulmonary function tests
  - Less than 80% of predicted
- Cardiac compression, mitral valve prolapse
- Symptomatic pectus excavatum
- Documented increase in deformity
- Psychosocial issues
Timing of Repair

- Optimal age range: 10 – 14 years
  - Before puberty, the chests are soft and malleable
  - Quick recovery with excellent results
- After puberty, the chest is stiffer routinely requiring 2 bars with a more difficult recovery
- HOWEVER patients > 20 years have been extremely happy with their results with an equal response
Overview of Procedure

Thoracoscopy to guide placement of the bar behind the sternum and anterior to the pericardium
Operative View
Results
Results
Post-operative Considerations

• Significant post-operative pain
  – Lessened by pre-operative physical therapy
  – Usually a 1-2 week convalescence
  – Minimal pain at 6 weeks
    • Plan operation for summer or winter break
• Restart physical therapy at 6 weeks
• Return to previous activities in 6 months
• Follow up at 6 month intervals
Long Term Results

972 Patients through 2010
   (20 year experience)
- Excellent result 830 (85.4%)
- Good result 114 (11.7%)
- Fair result 11 (1.1%)
- Poor result 6 (0.6%)
- Failed 11 (1.1%)

(Results from Kings Daughters Children’s Hospital)
Pectus Carinatum

- 10-20% of all pectus deformities
- Males > Female
- Four Variants
  - Upper
  - Lower
  - Mixed
  - Iatrogenic
    Any variant may be asymmetric
- Usually asymptomatic
Pectus Carinatum

• Pectus Brace
  – Fitted by sports medicine physical therapist
  – Combined with exercise program
  – Worn for 14-16 hours per day for 2 to 3 years
Results
Pectus Carinatum

- Pectus brace effectiveness
  - 75% Compliance
  - 85% Good or Excellent result
    (Stephenson 2008)

- Pectus brace complications
  - Pain or discomfort
  - Fit problems
  - Skin breakdown
  - Overcorrection
  - Shortness of breath
Pectus Carinatum Surgical Options

• Ravitch Procedure
  – Wedge resection of central costal cartilages

• Abramson procedure
  – External pectus bar
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QUESTIONS?