Advanced Pediatric Emergency Medicine Assembly

March 23-26 2015
New York, NY

Non-Traumatic Surgical Emergencies:
Avoid Missing These Life Threatening Conditions in Children

The speaker will present a case-based approach to the care of the pediatric patient with surgical emergencies to include the latest updates in appendectomy, evaluation of a child for intussusception, and recognizing midgut volvulus with malrotation. The speaker will also review latest diagnostic and treatments for nontraumatic surgical emergencies in children.

OBJECTIVES

- Review the current literature regarding appendectomy—delayed approach, the perforated appendix, pre-op antibiotics.
- Discuss the surgical emergencies of the intestines including malrotation and midgut volvulus, pyloric stenosis, intussusception, and appendicitis

3/23/2015
2:15 PM-2:45 PM
Grand Ballroom
MO-11

DISCLOSURES:
(+) No significant financial relationships to disclose
Case #1: Will

• 13 yo boy presented with a four day history of abdominal discomfort and constipation. Parents were treating symptomatically. 36 hours before arriving in the ER, he took a warm bath and “felt better” for the next 8 hours. He then developed vomiting (12 episodes in 24 hours, yellow in color) and the abdominal pain recurred with increased intensity.

• The PCP phoned in a prescription for Zofran
Case #1

- **Physical Examination**
  - HR 117  BP 115/69
  - General: ill appearing, lips dry, eyes slightly sunken
  - Abdomen: diffusely tender with rebound and guarding.
    Moderately distended. Absent bowel sounds

- **Ultrasound:** Dilated appendix with an appendicolith
  and free fluid within the abdomen consistent with appendicitis and concern for possible rupture

APPENDICITIS
Appendicitis: background

• Most common pediatric surgical emergency
  - Lifetime risk 8.6% boys: 6.7% girls
  - Incidence peaks in the second decade of life

• Symptoms: dull periumbilical pain that migrates to the RLQ
  - 44% of children present with “atypical” findings
  - Ruptured?
    • children < 4y, 80%
    • children 10-17, 10%-20%

• Laboratory studies: elevated WBC(>12,000) increases the odds of appendicitis (OR=6.5)

Pepper, V et al Surg Clinics NA 2012

Appendicitis: Differential

• Inflammatory
  - Mesenteric adenitis, IBD, intussusception, omental infarction

• Infectious: viral, bacterial, or parasitic

• Vascular: HSP

• Congenital:
  - Meckel’s diverticulum, duplication cysts

• Genitourinary conditions:
  - Pyelonephritis, ovarian pathology, PID, infected urachal remnants
Management: Pain control

• It is appropriate to control pain with short-acting intravenous analgesics while the child is in the ED
  - .05-.1 mg/kg of morphine
  - 1 mcg/kg fentanyl

• Administration of analgesics does not effect the diagnostic accuracy of the physical examination nor alter the clinical outcome

Appendicitis: what is helpful?

• In children with abdominal pain
  - Fever, rebound tenderness, mid-abdominal pain that migrates to the RLQ each individually increase the likelihood of appendicitis.
    • The absence of those signs decreases the likelihood of the disease
  - WBC < 10,000 decreases the likelihood of appendicitis as does ANC < 6750
  - These signs and symptoms are most useful in combination particularly in identifying who doesn’t require additional evaluation

Bundy et al JAMA 2007
Pediatric Appendicitis Scores

<table>
<thead>
<tr>
<th>PAS</th>
<th>Alvarado score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Migration of pain</td>
<td>Migration of pain</td>
</tr>
<tr>
<td>Anorexia</td>
<td>Anorexia</td>
</tr>
<tr>
<td>Nausea/vomiting</td>
<td>Nausea/vomiting</td>
</tr>
<tr>
<td>RLQ tenderness</td>
<td>RLQ tenderness</td>
</tr>
<tr>
<td>Cough/hop/ percussion tenderness RLQ</td>
<td>Rebound</td>
</tr>
<tr>
<td>Increase in temperature</td>
<td>Increase in temperature</td>
</tr>
<tr>
<td>WBC&gt;10,000</td>
<td>WBC&gt;10,000</td>
</tr>
<tr>
<td>PMNs&gt;75%</td>
<td>PMNs&gt;75%</td>
</tr>
<tr>
<td>Total</td>
<td>Total</td>
</tr>
<tr>
<td>1-3 negative; &gt; =8 positive</td>
<td>1-4 negative; 9, 10 positive</td>
</tr>
</tbody>
</table>

Clinical pathways for appendicitis

- Diagnostic accuracy of clinical pathway using PAS and ultrasound evaluated

- Patients determined to be at low (PAS 1-3) medium (PAS 4-7) or high risk (PAS 8-10)
  - Low risk patients discharged with phone f/u
  - High risk patients immediate surgical evaluation
  - Intermediate risk received an ultrasound

Saucier A et al Ped 2013
Clinical pathway for appendicitis

- 196 children enrolled: 44 low risk; 119 medium risk; 33 high risk
  - 65 had appendicitis
  - Ultrasound performed on 128 patients: 48 were positive
  - CT performed on 1 child
  - Negative appendectomy rate 3/68 (4.4%)

- Diagnostic accuracy 94%

Saucier, 2013

Utility of pediatric appendicitis scores

- Review of literature to determine accuracy of the Alvarado and PAS Scores

- Conclusions:
  - For children with pretest probability of acute appendicitis of <60%, an Alvarado score below 4 rules out the diagnosis
  - Observation without imaging is appropriate management
  - PAS did not identify clinically useful low or high risk groups

Accuracy of diagnostic imaging for appendicitis

- 1810 children with concern for appendicitis analyzed
  - 49% boys
  - 68% assessed by CT/46% assessed by ultrasound (13% had both ultrasound and CT)
  - Sensitivity of ultrasound increased with increasing pain duration: this association not seen with CT
  - Proportion of equivocal CT readings decreased with increasing pain duration


Evaluation of appendicitis by POCUS

- Investigation of accuracy of POCUS in diagnosing appendicitis
  - 13 PEM ultrasonographers were given focused training
  - After the training, 264 diagnostic studies including 85 with pathology verified appendicitis were performed
  - Bedside ultrasonography had sensitivity of 84%, specificity of 93%

- Conclusion: emergency physicians can learn to use ultrasound to accurately evaluate children with suspected appendicitis

Sivitz, AB et al Annals Emerg Med 2014
Ultrasound: appendicitis

CT scan: appendicitis
Antibiotics for appendicitis

• Historically: amp, gent and clinda

• Both piperacillin/tazobactam (Zosyn) and cefoxitin (Mefoxin) have been shown to be at least as efficacious

• Total length of therapy determined by presence of perforation

Emergency vs urgent appendectomy

• 365 children treated for appendicitis over a 28 month period at 2 children’s hospitals

  - Outcomes were compared between patients who underwent emergent (within 8 hours of presentation) vs urgent (more than 8 hours) appendectomy

  - No significant differences in postoperative outcomes including readmissions, wound infection, postoperative abscesses nor in LOS

• Conclusion: “Performance of an urgent appendectomy a time convenient to the surgeon should be considered within the standard of care”

  Taylor et al. J Ped Surg 2005
Effects of delay on rate of perforation

• 202 children undergoing appendectomy: 197 had appendicitis

• If symptoms present <18 hours, incidence of perforation 10%

• If symptoms present 36 hours, incidence of perforation 44%

• Most of the prolonged delay was attributed to prehospital rather than in-hospital factors.


Is appendicitis an emergency?

• Risk of perforation increases with delay in recognition and surgery for acute appendicitis

  - Review of database of 683,590 children and adults with discharge diagnosis of appendicitis

    • 80% surgery on day of admission; 18% on days 2-4 and later operations accounted for 1%

    • Perforation rate 28% on day of admission, 33% on hospital day 2 and 78% by hospital day 8

Papandria, D et al J Surg Research 2013
What do the surgeons think?

• Survey of 1052 members of the American Pediatric Surgical Association
  - 46% response rate
  - 4% considered nonperforated appendicitis to be an emergency
  - 14% would consider coming in from home to perform an overnight appendectomy
  - 92% believe that waiting until daytime does not result in increase in complications
    • Surgeon fatigue and OR availability most common factors underlying delay

Dunlop, JC et al Acad Ped 2012

Case #1: Hospital course

• Given Zosyn, IVF
• Underwent laparoscopic appendectomy
• Post-operative pain was difficult to control and he was unable to tolerate oral feeds
• Started on TPN
• Underwent IR drainage of intrabdominal abscesses on HD 12
• Discharged on hospital day 15
Case #2: Noel

- 6 mo boy was well until four days prior to presentation when he developed vomiting. He was seen by the PCP who prescribed Zofran. The emesis continued. He also had crying spells during which he pulled his legs up to his abdomen. Mother also reported poor oral intake and decreased urine output. He did not pass stool for four days until the day of presentation when a “dark streak” was noted.

Case #2

- Physical examination
  - BP 101/65  HR 157  TEMP 98.3
  - General: WD/WN infant alert
  - Abdomen: NT/ND normal bowel sounds. Becomes tense with crying

- Ultrasound: Ileocolic intussusception
Intussusception: background

• Most common cause of small bowel obstruction in children (56/100,000/year)
  - Occurs most often around the ileocecal junction

• Demographics:
  - Peak incidence between 5-10 mos.: 90% are less than 2 years.
  - Males: female 2:1

• Classic presentation: abdominal pain, sausage shaped abdominal mass and currant jelly stool
  - Seen < 15% of patients

• Divided into two groups:
  - Idiopathic (90%): no clear lead point or disease trigger
  - Secondary to a lead point: anatomic, tumor, vascular, infectious
Intussusception risk after Rotavirus vaccine

- Data analyzed from infants 5-39 weeks of age who were enrolled in 3 health plans and received either monovalent (Rotovirax) of pentavalent (Rotateq) rotavirus vaccine between 2004-2011

- RV5 associated with an increase in intussusception of 1.5/100,000 recipients of the first dose
  - No risk seen after doses 2 and 3
  - Represents one tenth the risk of tetravalent rotavirus vaccine (RotaShield)

Yih, WK et al NEJM 2014

Classic presentation: intussusception

- Review of 219 patients with intussusception
  - 60% male
  - 60% under 1 year of age (median 7 mos)
  - 27% between 13 and 35 mos (median 23 mos)
  - 15% were older than 3 years (median 5 years)

- Presentation:
  - < 1yr more likely to present with emesis, irritability and guaiac positive or bloody stools
  - >1yr abdominal pain was most common complaint

- Mandeville, K et al PEC 2012
Predictors of intussusception

- 115 children <5yrs of age on whom contrast enemas were performed for suspicion of intussusception
  - 59% had the diagnosis
- Independent predictors of intussusception included
  - Male gender
  - Rectal bleeding
  - Highly suggestive abdominal radiograph
  - History of emesis

Kuppermann et al Arch Ped Adol Med 2000

Intussusception: diagnosis

- Imaging choices:
  - Radiographs: low specificity and low sensitivity (45%)
  - Ultrasound: sensitivity and specificity approach 100%
    - Also can assess the presence of a pathological lead point and identify alternative diagnoses.
    - Predict the reducibility of an intussusception as trapped peritoneal fluid between two loops of bowel and lack of color Doppler are associated with less reduction

Applegate, KE Ped Radiol 2009
Abdominal plain films: intussusception

Ultrasound: intussusception
Comparison of 2-view AXR with US

• Retrospective cohort of 286 children 3 mo – 3 yr who presented to ED with symptoms suggestive of intussusception and had both 2-view abdominal XR and ultrasound performed

<table>
<thead>
<tr>
<th></th>
<th>2 view AXR</th>
<th>Ultrasound</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sensitivity</td>
<td>62%</td>
<td>98%</td>
</tr>
<tr>
<td>Specificity</td>
<td>86%</td>
<td>96%</td>
</tr>
<tr>
<td>Negative predictive value</td>
<td>89%</td>
<td>99%</td>
</tr>
<tr>
<td>False positive rate</td>
<td>13%</td>
<td>3%</td>
</tr>
<tr>
<td>False negative rate</td>
<td>37%</td>
<td>1.6%</td>
</tr>
</tbody>
</table>

Henderson et al. PIC 2013

Conclusion: ultrasound is superior screening method

Intussusception: treatment

• Consult surgery prior to enema reduction
  - To assess for peritoneal signs that preclude an enema
  - For post reduction management

• Reduction techniques:
  - Air enema: better technique: cleaner, safer, faster, less radiation than with liquid enema
    • Average reduction rate 74%
  - Hydrostatic (not barium) enema can also be used

• Main risk of reduction: perforation of the bowel (<1%)

• After enema reduction recurrence in 10%

Applegate, 2009
Air enema reduction: intussusception

Hydrostatic enema reduction: intussusception
Antibiotics before reduction?

• Patients from 2 centers were compared: one where antibiotics were administered and one where they were not
  - 118 children (70% male)
  - Median age 24 months
  - 56 (57%) patients received antibiotics

• Incidence of fever post-reduction was not statistically different between the groups

• Time to oral feeds was slightly shorter and total length of stay slightly longer for the antibiotic group

Al_Tokhais, T et al J Ped surg 2012

Intussusception: reduction under ultrasound

• Sonographic guidance with either air or water reduction techniques show reduction rates equivalent to those using fluoroscopy

• Risk of bowel perforation: 0.8%

Applegate 2009
After reduction by enema

- Retrospective chart review of 98 children ages 0-17 years with confirmed and uncomplicated enema reduction of intussusception
  - 10 episodes of recurrence in 7 children
    - Two had recurrence in <48 hours (3h and 5h)
    - Remainder had recurrence > 48 hours
    - No adverse events noted
  - Conclusion: ED observation for 6 hours is safe alternative to admission after reduction

Chien M et al J Emerg Med 2013

Role of surgery in intussusception

- On-site surgery at time of reduction?
  - Complications requiring immediate medical or surgical attention are rare
    - 1.6% of 433 cases (4 perforations, 2 unsuccessful reductions)

- Surgery is indicated when
  - Nonoperative reduction is incomplete
  - Persistent filling defect indicating a mass lesion is noted
  - Suspected or proven bowel perforation
  - Bowel necrosis

Nguyen H et al Ped Imaging 2014
Case #3: Sarai

• 4 week old former 36 week baby was well until the morning of presentation when she developed emesis (NB/NB). She was seen by her PCP who suspected pyloric stenosis and sent her in for a pyloric ultrasound.

• Physical examination
  - HR 169 TEMP 99.9 BP 106/56
  - General alert WDWN baby
  - Abdomen: distended without guarding or rebound
Volvulus: background

- Small bowel or midgut volvulus is most commonly due to malrotation
  - Presents with bilious emesis in the newborn (80%)
  - Presents commonly with abdominal pain and constipation in older infants and toddlers
    - May also see diarrhea and failure to thrive
    - Bilious emesis present in <50%
- Other causes of volvulus include Meckel's diverticulum, meconium ileus, mesenteric and duplication cysts and jejunal atresia or stenosis
- Diagnosis made by upper GI contrast study

Diagnosis: malrotation no volvulus

Diagnosis: malrotation with volvulus
Malrotation of the intestine in children

• Review of 102 children with malrotation: 64% male
  - 52 < 7 days
  - 13 from 8-30 days
  - 26 from 31-365 days
  - 11> 1 year

• Presenting symptoms: bilious emesis (47), intestinal obstruction (19), abdominal pain (11), bloody stools (7)

• 70 patients had congenital anomalies (50 single, 20 multiple)
  - GI anomalies most common

Associations with Malrotation

- Syndromes
  - Cornelia de Lange
  - Cantrell syndrome
  - Trisomies 13, 18, 21
  - Coffin-Siris syndrome
  - Heterotaxy
  - Marfan syndrome
  - Meckels syndrome
  - Mobile cecum syndrome

- Anatomic anomalies
  - Absence of kidney and ureter
  - Biliary atresia
  - Congenital diaphragmatic hernia
  - Small-bowel stenosis or web
  - Gastrochisis
  - Hirschsprung’s
  - Meckel’s diverticulum
  - Pyloric stenosis

Case #3

- In the ED the patient developed HR=200 and increased abdominal distension
  - NS fluid bolus (20 cc/kg) 3 times
  - Given Zosyn
  - Admitted to the PICU

- Went to the operating room where found to have a segmental volvulus with necrotic bowel. Had a resection of 30cm ileum.

- Discharged on HD 12 on oral feeds
Case #4: Adam

• 4 week old male infant was sent to the ED by his PCP with a two day history of vomiting. He was the product of a term, uncomplicated pregnancy and delivered vaginally. He went home on DOL #2. He has been bottle fed and has been taking 3-4 ounces every 3-4 hours until the vomiting began.

• Emesis is described as “clabbered milk”. He is hungry after vomiting. He had been given Pedialyte which he also vomits. Urine output has been normal.

Images
PYLORIC STENOSIS
Pyloric stenosis: background

• First described in 1627

• First successful surgical procedure 1908

• Most frequent surgical condition in the first few months of life
  - Most common surgical cause of vomiting (nonbilious, forceful)

• Incidence: 2-4/1,000 live births in Western population
  - Usually presents between 2-6 weeks of life
  - Males predominate
  - Family history of pyloric stenosis is a risk factor
  - Etiology: multifactorial

Pyloric stenosis: changing features

• Comparison of 2 groups of patients at the same hospital with confirmed diagnosis of pyloric stenosis
  - Group 1: 2004-2010
  - Group 2: 1984-1995

Taylor et al J Paed Child Health 2012
Pyloric stenosis: changing features

<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>% male</td>
<td>84.8%</td>
<td></td>
</tr>
<tr>
<td>Mean age at admission</td>
<td>5.4 weeks</td>
<td>5.9 weeks</td>
</tr>
<tr>
<td>Olive palpated</td>
<td>48%</td>
<td></td>
</tr>
<tr>
<td>Peristalsis visible</td>
<td>25%</td>
<td></td>
</tr>
<tr>
<td>Hematemesis</td>
<td>16%</td>
<td></td>
</tr>
<tr>
<td>Diagnosis on clinical examination alone</td>
<td>9%</td>
<td>74%</td>
</tr>
<tr>
<td>Ultrasound used</td>
<td>91%</td>
<td>16%</td>
</tr>
<tr>
<td>CL&lt;85</td>
<td>9%</td>
<td>26%</td>
</tr>
</tbody>
</table>

Taylor, 2012

Evaluation of pyloric stenosis in ED

- Evaluation of accuracy of PEM physician ultrasonography for infants with suspected pyloric stenosis
  - 67 patients: 10 with pyloric stenosis

- Trained PEM physicians identified all cases with 100% sensitivity and 100% specificity

Sivitz, A et al AEM 2013
Pyloric stenosis and bottle feeding

• Examination of the Danish National Birth Cohort
  - 70,148 infants followed for first 4 months of life
  • 65 had surgery for pyloric stenosis
  • Incidence in Denmark is 1-2/1000 live births
  - Information on feed practices obtained from 6 month phone interview.

• Bottle fed infants experienced a 4.6 fold higher risk of PS compared to breast fed infants

Krogh et al Ped 2012

Pyloric stenosis: breaking news

• Confirmation of two novel loci for infantile hypertrophic stenosis on chromosome 3 and 5

• Screened samples representing 796 case and 876 controls from patients of Danish origin and confirmed loci
  - Chromosome 3p25.1
  - Chromosome 5q35.2

Everett, KV and Chung EMK J Human Genetetics 2013