It’s Tuesday!

January 13, 2015
A 16-year-old girl comes to your office for a follow-up visit from the emergency department, where she went for the acute onset of knee pain and swelling. The emergency department physician has ordered an antinuclear antibody test, which was positive at 1:320. Further history reveals that she has had intermittent joint pains for several weeks and dark-colored urine. Findings on her physical examination are normal, except for an effusion in her right knee. You decide that further evaluation for systemic lupus erythematosus is warranted.

Of the following, the MOST specific test in helping you make the diagnosis for SLE is

A. Anti-cardiolipin antibody measurement
B. Anti-double stranded DNA antibody measurement
C. Anti-Ro antibody measurement
D. Complement measurement
E. Veneral disease research laboratory test
What do you want to know?
Let’s review!

- Refers to oral expulsion of gastric contents with contraction of abdominal and chest wall muscles

- NORMAL - Slight yellow tinge
  - Small amounts of bile into stomach

- ABNORMAL -
  - **Green** or **bright yellow** → “Bilious”
  - Often associated with intestinal obstruction
Why is there no “standard” workup?
Why is he vomiting?

<table>
<thead>
<tr>
<th>Neonate (&lt;1 month)</th>
<th>Infant (&gt;1-12 months)</th>
<th>Toddler (&gt;1-4 years)</th>
<th>Child (4-12 years)</th>
<th>Teenager (13-19 years)</th>
</tr>
</thead>
<tbody>
<tr>
<td>GER or GERD</td>
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<td>Gastroenteritis</td>
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<tr>
<td>Feeding intolerance</td>
<td>Acute otitis media</td>
<td>Urinary tract infection</td>
<td>Pharyngitis</td>
<td>Peptic ulcer disease</td>
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<tr>
<td>Pyloric stenosis</td>
<td>Protein intolerance</td>
<td>Pharyngitis</td>
<td>Post infectious gastroparesis</td>
<td>Cyclic vomiting</td>
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<tr>
<td>Meconium ileus</td>
<td>Gastroenteritis</td>
<td>GERD</td>
<td>Eosinophilic esophagitis</td>
<td>Eosinophilic esophagitis</td>
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<tr>
<td>Congenital atresia or webs</td>
<td>Pyloric stenosis</td>
<td>Eosinophilic esophagitis</td>
<td>Appendicitis</td>
<td>Pregnancy</td>
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<tr>
<td>Malrotation with midgut volvulus</td>
<td>Intussusception</td>
<td>Celiac disease</td>
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<td>Poisoning/toxic ingestion</td>
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<tr>
<td>Necrotizing enterocolitis</td>
<td>UTI</td>
<td>Intracranial lesion</td>
<td>Pancreatitis</td>
<td>Migraine</td>
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<tr>
<td>Metabolic disorders</td>
<td>Malrotation with midgut volvulus</td>
<td>Malrotation</td>
<td>IBD</td>
<td>Diabetic ketoacidosis</td>
</tr>
<tr>
<td>Hirschsprung disease</td>
<td>Intracranial lesion</td>
<td>Poisoning/toxic ingestion</td>
<td>Trauma (duodenal hematoma)</td>
<td>Ruminating syndrome</td>
</tr>
<tr>
<td>Protein intolerance</td>
<td>Metabolic disorders</td>
<td>Adrenal insufficiency</td>
<td>Poisoning/toxic ingestion</td>
<td>Drug abuse</td>
</tr>
<tr>
<td>Infection (UTI or meningitis)</td>
<td>Child abuse</td>
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<td>Appendicitis</td>
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<tr>
<td></td>
<td>Munchausen syndrome by proxy</td>
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<td>Gallstone</td>
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<td></td>
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<td>Pancreatitis</td>
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<td>Bulimia</td>
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<tr>
<td></td>
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<td>IBD</td>
</tr>
</tbody>
</table>
Even more detailed differential

<table>
<thead>
<tr>
<th>Gastrointestinal disorders</th>
<th>Metabolic disorders</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Esophagus: gastroesophageal reflux, eosinophilic esophagitis, achalasia, esophageal atresia, stricture, web, ring, foreign body</td>
<td>• Carbohydrate: galactosemia, hereditary fructose intolerance, pyruvate carboxylase deficiency</td>
</tr>
<tr>
<td>• Stomach: gastroenteritis, gastritis, peptic ulcer disease, pyloric stenosis, gastroparesis, bezoar</td>
<td>• Organic acid: phenylketonuria, urea cycle defect, maple syrup urine, tyrosinemia type 1</td>
</tr>
<tr>
<td>• Small intestine: malrotation, atresia, duplication, intussusception, volvulus, ileus, pseudoobstruction, necrotizing enterocolitis, celiac disease, Crohn's disease, duodenal hematoma, superior mesenteric artery syndrome, milk protein enteropathy</td>
<td>• Fatty acid oxidation defect: carnitine deficiency, MCAD, LCAD</td>
</tr>
<tr>
<td>• Colon: Hirschsprung disease, ulcerative colitis, appendicitis, constipation, hernia</td>
<td>• Lysosomal storage: mucopolysaccharidoses, Niemann–Pick disease, Wolman disease</td>
</tr>
<tr>
<td>• Liver: hepatitis, acute liver failure, hepatic abscess</td>
<td>• Peroxisomal disorders: Zellweger disease, infantile Refsum disease, acyl-CoA oxidase deficiency, adrenal leukodystrophy</td>
</tr>
<tr>
<td>• Gallbladder: cholecystitis, choledolithiasis, choledocho lithiasis, gallbladder dyskinesia, choledochal cyst</td>
<td>Endocrine disorders</td>
</tr>
<tr>
<td>• Pancreas: pancreatitis, annular pancreas, pancreatic divisum</td>
<td>• Diabetic ketoacidosis, adrenal insufficiency/adrenal crisis, hyperparathyroidism, pregnancy</td>
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<tr>
<td>• Peritoneum: peritonitis, peritoneal adhesion</td>
<td>Neurologic disorders</td>
</tr>
<tr>
<td>Renal disorders</td>
<td>• Hydrocephalus, tumor, intracranial bleeding (subdural hematoma), meningoen cephalitis, abscess, seizure, migraine, pseudotumor cerebri, motion sickness, ventriculoperitoneal shunt failure or infection</td>
</tr>
<tr>
<td>• Renal tubular acidosis, ureteropelvic junction obstruction, nephrolithiasis, renal insufficiency, uremia, hydronephrosis</td>
<td>Toxins and medications:</td>
</tr>
<tr>
<td>Infectious disorders</td>
<td>• Aspirin, iron, lead, digoxin, alcohol, marijuana, chemotherapeutic agent</td>
</tr>
<tr>
<td>• Urinary tract infection, meningitis, pharyngitis, sinusitis, otitis media, pneumonia, sepsis</td>
<td>Other</td>
</tr>
<tr>
<td></td>
<td>• Eating disorders (anorexia, bulimia), cyclic vomiting syndrome, rumination, overfeeding, psychogenic</td>
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</tbody>
</table>

LCAD=long chain acyl-CoA dehydrogenase; MCAD=medium chain acyl-CoA dehydrogenase.
So, what next?
Our patient...
Pyloric Stenosis
Epidemiology

- Occurs in 3-6/1000 infants
- Male:Female? **4:1**
- Most commonly: **firstborn, white infants**
  - 1/3 are first-born!
- 10-20% of infants of mother with history of pyloric stenosis
Etiology

- Underlying defect is redundant pyloric mucosa-> edema, obstruction, secondary muscle hypertrophy
- Hereditary and environmental?
- Use of **erythromycin** during first 2 weeks of life
  - Also: maternal use of macrolide during late pregnancy/breastfeeding
- Increased incidence with B and O blood types
- Abnormal muscle innervation
- Maternal stress during 3\textsuperscript{rd} trimester
Clinical Manifestations

• Age: 2-5 weeks of age
• Initial symptom: Nonbiliious emesis
• Weight loss and dehydration
• Are they hungry?
• Palpable small “olive-shaped mass” in mid-epigastric area (usually after emesis)
• Visible gastric peristaltic wave after feeding
• +/- Jaundice (Ictopyloric Syndrome = Hyperbili + PS in up to 8%)
  – Jaundice due to increased enterphepatic circ and decreased glucuronyl transferase activity
Diagnosis

• Clinical dx possible in 60-80%

• What is the major lab finding seen?
  
  *Hypochloremic hypokalemia*
  
  – Due to loss of gastric HCl
  – Remember, can still be acidotic if severe dehydration present

• What would you see on abdominal x-ray?
  
  – Dilated stomach bubble
Further Imaging

• Test of choice? **Abdominal US (90% sensitivity)**
  – By the numbers:
    • Thickness 3-4 mm
    • Pyloric length of 15-19mm
    • Pyloric diameter of 10-14mm

• Upper GI (has risk for aspiration)
  – May show elongated pyloric channel (string sign)
  – Bulge of pylorus into antrum (shoulder sign)
  – Parallel streaks of barium in the channel (double tract sign)
  – Narrowed ending of pyloric canal (beak sign)
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Management

• Correct dehydration
• Correct electrolytes
  – Bicarb should be less than 30 to prevent **Postoperative apnea**
• After about 24 hours….head to OR
  – Pyloromyotomy (lap or incision)
  – Split the pylorus without cutting through mucosa
Finishing up the patient…

Have a fantastic day!

Noon Conference: Dr Beatty
“The Business of Medicine”