Abdominal pain in children: Case Reviews

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Objectives:
- Review characteristics of chronic abdominal pain in children
- Distinguish functional abdominal pain from organic causes of abdominal pain in children
- Using a series of cases, discuss the presentation, evaluation and management of several different causes of abdominal pain in children including:
  - Acid peptic disease
  - Acute intermittent porphyria
  - Choledochal cyst
  - Cholelithiasis/cholangitis
  - Functional abdominal pain
  - Pancreatitis
  - UPJ obstruction

Case 1
- A 13 y.o. girl is brought to your office for evaluation of abdominal pain that has been present over the past 4 months. Her pain is poorly localized to the periumbilical area without localization or radiation. She rates her pain as a 6/10. Her pain is worse in the morning and also may be present in the afternoon and before bed. There are no specific triggers such as diet or activity that precipitate her pain. She does not have a history of change in her appetite, nausea or vomiting, change in her stool pattern or weight loss. She is otherwise healthy.
- Her parents are most concerned because she has missed 1-2 days of school per week for the past 3 months.
- Her physical examination is normal.
How to proceed?

- Laboratory evaluation
  - CBC
  - CMP
  - WSR
  - Total IgA/tissue-transglutaminase IgA
  - Stool guaiac
  - UA
  - Stool for ova and parasites
  - Lactose breath test
  - Fructose breath test
- All are normal

Question 1

The most likely diagnosis as the cause for her chronic abdominal pain is:

A. Crohn’s disease
B. Celiac disease
C. Pancreatitis
D. Functional abdominal pain
E. Familial Mediterranean Fever

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Chronic Abdominal Pain in Children

- One of the most common complaints in children and adolescents
- 13% middle school children
- 17% high school children
- Functional vs. organic
- 80-90% of children diagnosed with functional abdominal pain

“Red flags” in the evaluation of children with chronic abdominal pain

- History
  - Persistent RUQ or RLQ pain
  - Dysphagia
  - Persistent vomiting
  - Gastrointestinal blood loss
  - Nocturnal diarrhea
  - Pain that awakens child from sleep
  - Arthritis
  - Unexplained fever
- Physical examination
  - Involuntary weight loss
  - Deceleration in linear growth
  - Hepatomegaly or splenomegaly
  - Perianal disease
  - Delayed puberty
- FH of celiac disease, IBD or PUD

Important facts: Functional Abdominal Pain in Children

- Rome III criteria for Functional Gastrointestinal Disorders, 2009
- Definition: Functional abdominal pain
  - Symptom of chronic or recurrent abdominal pain in children where there is no identifiable structural, inflammatory, infectious, neoplastic or metabolic cause.
  - Symptoms that occur at least once a week for a duration of at least 2 months.
- Classification of functional abdominal pain disorders
  - Functional dyspepsia
  - Irritable bowel syndrome
  - Childhood functional abdominal pain
  - Pain may be localized to the epigastric, periumbilical or lower abdomen
- No “red flags”
- Physical examination and limited laboratory evaluation is normal
Functional Abdominal Pain in Children

- Uncommon under the age of 5 years
- Females slightly more than male
- Children with functional abdominal pain experience real pain, not feigned
- Pathogenesis-Children with functional bowel disorders, may have an abnormal bowel reactivity to physiologic stimuli (meal, gut distension, hormonal changes), noxious stressful stimuli (inflammatory processes), or psychological stressful stimuli (parental separation, anxiety) leading to the development of visceral hyperalgesia
- Functional abdominal pain is a "positive diagnosis" not a failure to identify the true cause for a child’s abdominal pain

Question 1b:
The most appropriate initial treatment for a child with functional abdominal pain is:

A. Education, reassurance and limiting pain-induced disability
B. Cognitive-behavioral therapy
C. Self-hypnosis
D. Tricyclic antidepressants or selective serotonin reuptake inhibitors
E. Acid suppression/antispasmodics /laxatives
Case 2

- A 13 y/o boy was brought to your office for evaluation of epigastric abdominal pain that had been present for the past 2 months. His mother, an adult gastroenterologist, had treated him with a PPI with partial improvement in his symptoms. He had associated nausea and he had lost 2.5 kg over the previous two months. His examination was remarkable for moderate epigastric discomfort and otherwise normal.
  Labs: hgb 11.5 g/dL, hct 35.2 %; WBC, plt count, CMP and WSR were normal.

Causes of epigastric abdominal pain

- Functional dyspepsia
- GERD/Esophagitis
- Gastritis
- Peptic ulcer disease
- Pancreatitis

Question 2:

What test would be most helpful in establishing a diagnosis?

A. Abdominal ultrasound
B. CT scan
C. Upper endoscopy with biopsy
D. H. pylori antibody titer
E. Amylase and lipase
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H. Pylori gastritis/peptic ulcer disease

Peptic ulcer disease: Facts to know
- Duodenal > gastric
- Boys > girls
- Primary ulcers: duodenum
- Secondary ulcer: gastric
- Symptoms - variable pain pattern
  - + / - epigastric
  - + / - temporal relationship to meals
  - + / - nighttime pain
  - 40% vomiting
Peptic ulcer disease: Facts to know

- Causes of peptic ulcer disease:
  - H. pylori - most common cause of ulcers in children
  - NSAID/aspirin/alcohol
  - Stress: Shock, trauma, sepsis, head injury, burns
  - Excessive acid production:
    - Zollinger-Ellison syndrome - gastrin secreting tumors
    - Systemic mastocytosis
  - Other conditions:
    - Eosinophilic gastroenteritis
    - Menetrier's disease, hypertrophic gastritis
    - Autoimmune (atrophic) gastritis
    - Gastroduodenal Crohn's Disease

Helicobacter Pylori

- S shaped gm- bacillus
- 1983 - associated with chronic gastritis
- Colonizes the surface of the gastric mucosa
- Person-person transmission
- Associated symptoms and findings:
  - Acute / chronic nodular antral gastritis
  - Gastric ulcer and duodenal ulcer
  - Asymptomatic
  - Iron deficiency anemia
- Diagnosis:
  - "Gold standard" upper endoscopy/gastric mucosal biopsy
  - Rapid urease testing (CLO test)
  - 13C Urea breath test
  - H. pylori stool antigen testing
- Treatment - triple therapy
  - Proton pump inhibitor, amoxicillin and clarithromycin

Case 3

- A previously healthy 3 yo girl is seen in your office for evaluation of recent onset of right upper quadrant abdominal pain and scleral icterus. Her examination was remarkable for moderate jaundice and the presence of a right upper quadrant mass.

Laboratory studies were remarkable for:

- CBC: hgb 12.3 g/dL, hct 37.2 % WBC 13,700
- AST 60 U/L ALT 76 U/L bili 3.2 mg/dL
- Amylase 100, Lipase 75
Question 3:
What is the most likely diagnosis?
A. Intestinal duplication cyst  
B. Pyloric stenosis  
C. Hydronephrosis  
D. Choledochal cyst  
E. Constipation

Abdominal doppler ultrasound

Choledochal cysts: important facts
- Congenital cystic dilation of intra or extra hepatic biliary tree
- Types I-V
- 90% Type I - dilatation of the common bile duct
- 1 in 13,000 to 2,000,000
- Girls 4x > boys, more common in Asian population
- Present at any age, 50% diagnosed in infancy presenting with neonatal jaundice
- Clinical triad
  - Jaundice
  - Intermittent abdominal pain
  - Abdominal mass
- Complications: cholangitis and pancreatitis
- Diagnosis-ultrasound, MRCP
- Treatment: complete surgical resection - Potential for the development of cholangiocarcinoma
Case 4

A 16 yo boy is seen in the ED with acute onset of severe epigastric and LUQ abdominal pain radiating to his back associated with nausea and vomiting. He has otherwise been well with the exception of having a mild URI in the week prior to his visit. The day before, he was seen after he fell off of his bike while dirt biking. He is not on any medication. FH is negative for GERD, PUD, pancreatitis and gallstones.

On physical examination his vital signs are stable. He has evidence of moderate dehydration. He more comfortable sitting in the knee/chest position. He has moderate LUQ and epigastric discomfort.

Question 4

What laboratory test would be most helpful in confirming a diagnosis?

A. AST
B. ALT
C. Amylase /lipase
D. Serum glucose
E. Urinalysis
Other causes of elevations of amylase

- Macroamylasemia
- Salivary gland trauma/infection (mumps)
- Intestinal obstruction/perforation
- Diabetic ketoacidosis
- Ovarian torsion
- Renal insufficiency
- Medications-opiates
- Diagnosis of pancreatitis- amylase and lipase 3X the upper limit of normal

Pancreatitis clinical presentation-

- Symptoms
  - Nonspecific
  - Sudden onset epigastric abdominal pain
  - Radiation to back / RUQ
  - Nausea / vomiting
- Physical findings
  - Epigastric/LUQ tenderness
  - Mild jaundice
  - Shock / hypotension
  - Ecchymoses of flank (Turner's sign)
  - Blue discoloration of the umbilicus (Cullen's sign)
- Laboratory findings
  - Amylase and lipase greater than 3x upper limit of normal
  - Ultrasound- pancreatic hypertrophy, decreased echogenicity, dilated pancreatic duct, peri-pancreatic fluid.
  - also to evaluate for the gallstones or biliary abnormalities

Causes of acute pancreatitis

- Biliary disorders
  - Gallstones
  - Cholelithiasis
- Trauma
- Pancreatic disorders
  - Cystic fibrosis
  - Diabetes
  - Pancreatic divisum
- Idiopathic
- Medications
  - Antibiotics
  - Anticonvulsants
  - Diuretics
  - Infection
- Metabolic
  - Hypercalcemia
  - Hyperlipidemia
- Vasculitis
- Duodenal ulcer
- Toxins
  - Alcohol
  - Organophosphates
  - Yellow scorpion bite
- Multisystem disease
  - Hemolytic uremic syndrome
  - Shock
  - Crohn's disease
  - Kawasaki disease
Chronic Pancreatitis - chronic recurrent abdominal pain

- Progressive pancreatic inflammatory disorder
- Recurring or persistent abdominal pain with the development of pancreatic endocrine or exocrine insufficiency

Causes of Chronic Pancreatitis

- Idiopathic
- Structural anomalies
  - Congenital ductal abnormalities
  - Choledochal cyst
  - Stricture
  - Pancreas divisum
- Hyperlipidemia - Type I, II and V
- Hypercalcemia
- Cystic fibrosis
- Inflammatory bowel disease
- Hereditary
  - PRSS1 gene mutation - encoding cationic trypsinogen
  - SPINK1 gene mutation - pancreatic trypsin inhibitor
  - CFTR mutations
- Autoimmune pancreatitis
- Organic acidemias
- Idiopathic fibrosing pancreatitis

Case 5

A 9 y.o. boy is seen with a history of chronic intermittent severe left sided abdominal pain for the past 3 months. His pain is localized to the left upper quadrant with radiation into his left flank. He was seen in the ED for evaluation of his abdominal pain, vomiting and dehydration. He has not had a history of fever, dysuria or polyuria.

- His examination is remarkable for left upper quadrant fullness

Question 5

The most likely diagnosis is:

- A. Cyclic vomiting
- B. Appendiceal colic
- C. Intestinal malrotation
- D. UPJ obstruction
- E. Diabetic ketoacidosis

CT scan of abdomen
Question 5

The most likely diagnosis is:
- A. Cyclic vomiting
- B. Appendiceal colic
- C. Intestinal malrotation
- D. UPJ obstruction
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CT scan of abdomen

UPJ obstruction: Facts
- Most common urinary obstructive lesion in children
- Congenital narrowing, most often partial obstruction, at the ureteropelvic junction
- Incidence: 1 in 500 live births
- Boys affected more than girls
- More frequent on the left side
- Symptoms:
  - Colicky abdominal pain, lateralizing abdominal, flank or back pain
  - Nausea/vomiting
  - Palpable renal mass
  - Hematuria
- Diagnosis
  - Ultrasound
  - Findings may only be present with acute episode of pain
- Treatment
  - Surgery

Case 6
- A 15 y.o. girl with sickle cell disease is seen in your office with a 6 month history of intermittent right upper quadrant pain and nausea. She has been admitted several times over the past 6 months with pain crisis, however this pain is distinctly different. Her pain is worse after meals and can occur at any time of the day. Her PMH is otherwise unremarkable. Her examination is normal, although she confirms that when her pain is present she localizes her pain (10/10) to the RUQ.
Question 6

What is the most appropriate imaging modality to order?
- A. CT scan
- B. Hepatobiliary scan
- C. Abdominal ultrasound
- D. MRCP
- E. KUB

Ultrasound of the RUQ
Question 6b
What type of stones would be found in a patient with sickle cell disease?
- A. Cholesterol stone
- B. Pigmented stone
- C. Calcium oxalate
- D. Uric acid

Gallstones: important facts
- Symptoms
  - Non-specific abdominal pain
    - Colicky right upper quadrant abdominal pain
    - Nausea / vomiting
    - Jaundice
    - Fatty food intolerance
  - Stone formation occurs from the precipitation of insoluble constituents in bile, which are cholesterol, bile pigments and calcium salts
  - Cholesterol stones, pigmented stones or mixed.
- Gallstone distribution by age
  - Infants -10%, typically do not require surgical intervention
  - Children- 6mo to 10yr-21%
  - Adolescents- 68%, most often cholesterol stone
Gallstones: important facts

- Predisposing factors:
  - Hemolytic disease-sickle cell disease and hereditary spherocytosis
  - Obesity
  - Hyperlipidemia
  - Infection
  - Pregnancy
  - Cystic fibrosis
  - Total parenteral nutrition
  - Ileal disease or resection
  - Medications- Ceftriaxone, Furosemide, Oral contraceptives
- Diagnosis- ultrasound
- Complication: cholecystitis, pancreatitis

Cholecystitis

- Results from inflammation of the gallbladder, typically secondary to gallstone obstruction of the cystic duct
- Can be acute or chronic
- Presenting symptoms of RUQ pain, radiating to the back with associated nausea, +/- fever and jaundice
- RUQ tenderness- Murphy’s sign
- Differential diagnosis
  - Hepatitis, hepatic abscess, pancreatitis, gonococcal pericholangitis, PUD, pneumonia, pyelonephritis, and kidney stones
- Laboratory evaluation: CBC, CMP, amylase, lipase, UA
- Diagnosis
  - Ultrasound- findings of gallstones, gallbladder dilatation, thickened gallbladder wall
  - Hepatobiliary scan

Case 7

- A 15 yo is seen in the ED with severe abdominal pain that has become progressively worse over the previous 3 days. Her pain is poorly localized over her entire abdomen. She has associated nausea and in the past 24 hours has become more lethargic, confused and experienced diffuse muscle weakness.
- In the past she has had similar although less severe episodes of pain. She has been evaluated by a pediatric gastroenterologist in the past and she has had a “million dollar” workup, all done at times when she has been asymptomatic.
- As the resident in the ED you obtain a careful family history which includes a history of Crohn’s disease, ulcerative colitis, gallstones, and acute intermittent porphyria in a maternal grandmother.
Question 7
Which test would be most helpful in establishing a diagnosis?
- A. Urinalysis
- B. Urine amino acid screen
- C. Urine catecholamines
- D. Urine HCG
- E. Urine porphobilinogen

Acute intermittent porphyria: things to remember
- Autosomal dominant disorder-variable penetrance
- Deficiency of enzyme porphobilinogen deaminase
- Many people never develop symptoms/symptoms are variable and nonspecific
- Symptoms occur as acute attacks
- More common in women than in men
- Pain is neuropathic not inflammatory
Acute intermittent porphyria: things to remember

**Symptoms**
- Severe abdominal pain in 80-90% of patients
- Nausea / vomiting
- Muscle weakness
- Confusion / hallucination
- Seizures

**Precipitants**
- Low carbohydrate intake
- Medications - sulfonamides, barbiturates
- Hormonal changes

**Diagnosis**
- Urine porphyrin screen- porphobiligen(PBG) and delta-aminolevulinic acid (ALA) during acute attack
- Decreased RBC porphobilinogen deaminase
- DNA testing for PBGD mutations

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**Answer key**
- 1.D
- 1b.A
- 2.C
- 3.D
- 4.C
- 5.D
- 6.C
- 6b.B
- 7.E

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Good Luck !!!!