Case Studies in Abdominal Pain

Lawrence S. Friedman, MD

Newton-Wellesley Hospital
Massachusetts General Hospital
Harvard Medical School
Tufts University School of Medicine

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NEWTON-WELLESLEY HOSPITAL

MASSACHUSETTS GENERAL HOSPITAL

Patient 1

21 y.o. M, 2nd opinion for abdominal pain and vomiting

2 years earlier: Episodes of burning mid-abdominal pain

1 year earlier: Acute burning mid-abdominal pain and vomiting; hypertension; no nausea

7-lb. weight loss

Abrupt cessation after 10 days

Patient 1
Workup

Normal laboratory studies

Normal head CT (except small pineal cyst)

Normal abdominal and pelvic CT (??Meckel's inflammation)

Normal renal artery ultrasound

Normal MRCP

Normal colonoscopy

Normal laparoscopy

Patient 1

Fine after discharge

Similar episode 6 months later:

- Hospitalized for dehydration
- 20-lb. weight loss
- TPN

Patient 1
2nd Workup

Normal abdominal and pelvic CT

EGD: mild gastric erythema

Normal head CT

Normal UGI and SBFT

Normal colonoscopy

Normal video capsule endoscopy

Normal gallbladder ejection fraction

Negative tests for celiac disease, porphyria

Abrupt cessation after 10 days

Diagnosis?

Further tests?

Patient 1

Further History

FH: Migraine headaches

Paternal grandmother
Paternal aunt
? Father

Occasional marijuana use

Cyclic Vomiting Syndrome

Stereotypical episodes of vomiting with regard to onset (acute) and duration (<1 week)

Three or more discrete episodes in the prior year

Absence of nausea and vomiting between episodes

Supportive: history or family history of migraine headaches

Rome III, 2006

Cyclic Vomiting Syndrome
Treatment

Prevention:

- Discontinue marijuana and other triggers
- Amitriptyline
- Anti-seizure medications
  - Zonisamide
  - Levetiracetam

Treatment:

- Sumatriptan
- Benzodiazepines
- Antiemetics
- Analgesics

Hejazi RA, et al. Aliment Pharmacol Ther 2011; 34:263-73.
Nass J, et al. Clin Gastroenterol Hepatol 2010; 8: 245-7.

Patient 1
Follow-up

Discontinued marijuana

Amitriptyline, increased to 50 mg hs

Sumatriptan prescription (not taken)

No attacks in over 4 years

Patient 2

79 y.o. M, 2nd opinion for abdominal pain and nausea

9 months earlier: mild intermittent nausea

7 months earlier: chronic nausea

diffuse abdominal discomfort 1 to 2 hours after eating

10-lb. weight loss

No vomiting, bloating, distention, change in bowel habits

Patient 2
Prior Evaluation

Routine labs normal

Amylase 176/110 U/L

Lipase 24 U/L

ESR normal

Celiac serology negative

***H. pylori* antibody negative**

Patient 2
Prior Evaluation (cont'd)

Abdominal CT (x2):	**diverticulosis** **calcifications in aorta and bilateral ileac vessels** **kidney cysts** **enlarged prostate**
Abdominal ultrasound:	**unremarkable**
Prior colonoscopy:	**diverticulosis** **6-mm adenoma**

Patient 2
Upper Endoscopy

Normal

Histology: Gastric body and fundic mucosa with intestinal metaplasia, pseudopyloric metaplasia, and linear neuroendocrine cell hyperplasia

Antrum with reactive gastropathy and prominence of G cells

Interpretation: Autoimmune gastritis

Vitamin B12 level: 537

Patient 2
Prior Management

Prednisone 10 mg PO twice daily

Symptoms worsened

Discontinued by patient after 4-5 days

Trials of omeprazole twice daily and ranitidine prn: ?helped

Trial of rifaximin (for small intestinal bacterial overgrowth): no benefit

Placed on ondansetron 4 mg PO twice daily

Patient 2

Past history:	**HBP, laryngeal nodule, depression**
Medications:	**Irbesartan** **Lorazepam** **Ondansetron**
ETOH:	**1-2 glasses of wine with dinner, discontinued 6 months earlier**

Patient 2
Physical Examination

Chronically ill-appearing, sallow

P 78 reg, BP 124/80 mm Hg

Bilateral Dupuytren's contractures

Chest, heart, abdomen unremarkable: no abdominal mass, no succussion splash, no bruits

Patient 2
Additional Testing

Gastric emptying scan:	normal
Serum gastrin (on omeprazole):	336 pg/mL
Parietal cell antibodies:	positive (68.4 units)
Intrinsic factor antibodies:	negative
Celiac serology:	negative
Serum cortisol:	13.9 mcg/dL
Review of gastric histology:	automimmune gastritis

Diagnosis?

Further tests?

Patient 2
Workup

Doppler studies of mesenteric vessels:

70% stenosis of celiac trunk

Patient 2

Now what?

Patient 2

Placement of celiac artery stent

Resolution of discomfort, improvement in nausea (some GERD symptoms)

Worsening depression

Vitamin B12 supplementation

Chronic Mesenteric Ischemia

- <5% of cases of intestinal ischemia
- Almost always caused by mesenteric atherosclerosis
- Despite collateral pathways, single-vessel disease (especially of celiac artery) does occur

Hansen KJ, et al. J Vasc Surg 2004; 40:45-52.
Moawad J, et al. Surg Clin North Am 1997; 77:357-69.

Chronic Mesenteric Ischemia
Clinical Features

Abdominal cramping or discomfort usually within 30 minutes after eating, increasing and resolving over 1-3 hours

Progressive; pain can become continuous

Fear of eating ("sitophobia")

Weight loss

Possibly nausea, bloating, episodic diarrhea, malabsorption, constipation, GI bleeding

Chronic Mesenteric Ischemia
Diagnosis

Abdominal plain films and CT often normal; vascular calcifications may be seen

Duplex US, MRA, angiography: reveal anatomic limitations to splanchnic blood flow but do not establish presence or absence of intestinal ischemia

Often at least 2 of 3 major splanchnic vessels are severely stenotic or occluded

Moneta GL. Semin Vasc Surg 2001; 14:186-92.
Gentile AT, et al. Am J Surg 1995; 169:476-9.

Chronic Mesenteric Ischemia
Treatment

Surgical revascularization is traditional approach

Percutaneous transluminal mesenteric angioplasty alone or with stent insertion is now preferred

Symptom relief in up to 99%, but relapse rate as high as 28%

Schermerhorn ML, et al. J Vasc Surg 2009; 50:341-8.
Pecoraro F, et al. Ann Vasc Surg 2013; 27:113-22.
Mensink PB, et al. Gut 2011; 60:722-37.
Silva JA, et al. J Am Coll Cardiol 2006; 47:944-50.

Patient 3

53 y.o. M with right-sided chest and right subcostal pain for several months

Age 19:	**Fundoplication for refractory esophagitis**
	Eventual resumption of medical therapy (PPI) to control symptoms
Age 38:	**History of binge drinking** **Epigastric pain and diarrhea** **Steatorrhea** **EUS: chronic pancreatitis** **Pancreatic enzymes** **Symptoms resolved**

Patient 3
Past Medical History

Atrial fibrillation: Maze procedures twice

Homozygous factor V Leiden mutation

Protein C deficiency

Venous thromboembolism and pulmonary emboli

IVC filter

Anticoagulation

L4-5 and L5-S1 diskectomy

Depression

Patient 3

Presents with right-sided chest and right subcostal pain for several months

- **Stabbing**
- **Right lower chest/right upper quadrant**
- **Worse with movement, not inspiration**
- **No dyspnea**
- **Limits ability to climb stairs**
- **Recent URI treated with levofloxacin**
- **Weight loss of 15 lbs. attributed to pain**

Patient 3
Family History

Heart disease (father MI, age 46)

Pancreatic cancer (maternal grandmother)

Colon cancer (maternal grandfather)

Abdominal aortic aneurysm (maternal uncle)

Patient 3
Medications

Lanzoprazole

Creon

Warfarin

Atorvastatin

Celexa

Multivitamin

Patient 3
Physical Examination

RUQ discomfort on “twisting” body

Afebrile, normal pulse and BP

Normal heart and lungs

Abdomen obese, 2-cm area of distinct tenderness in RUQ, no guarding or rebound, no mass or organomegaly, normal bowel sounds

No rash

Normal neurologic examination

Patient 3

Studies

CBC:	normal
CMP:	normal
Amylase:	96 U/L
Lipase:	140 U/L
INR:	2
CXR:	sternal wires, otherwise normal
CT chest and abdomen:	2 small fat hernias, no pulmonary emboli, no aortic dissection, normal pancreas, no gallstones, normal liver
EKG:	RBBB and left anterior hemiblock (unchanged)

Diagnosis?

Further tests?

Patient 3

Neurologic consultation:

- **Anterior cutaneous nerve entrapment syndrome (ACNES)**
- **R/o thoracic radiculopathy**
- **Unlikely: biliary disease**
  pancreatitis
  ischemic bowel disease

Anterior Cutaneous Nerve Entrapment Syndrome (ACNES)

- Entrapment of a cutaneous branch of a sensory nerve derived from a neurovascular bundle emanating from spinal levels T7 to T12
- Related to intra- or extra-abdominal lesion, edema, or fibrosis (scar)
- Pain is discrete, localized
- "Hover sign": guarding from the examiner's hand
- "Carnett's sign": increased tenderness with tensing of abdominal muscles

Hershfield N, et al. J Clin Gastroenterol 1992; 14:199-202.
Lindsetmo RO, et al. Am J Surg 2009; 198:129-34.
Costanza C, et al. Clin Gastroenterol Hepatol 2004; 2:395-9.

Anterior Cutaneous Nerve Entrapment Syndrome

Treatment

Avoidance of certain movements

Non-narcotic analgesics

Physical therapy

Injection therapy

? Laparoscopy, lysis of adhesions, subcutaneous nerve resection

Kuan L, et al. Taiwan J Obstet Gynecol 2006; 45:239-43.
Nazareno J, et al. Can J Gastroenterol 2005; 19:561-5.
Paajanen H. Surg Endosc 2006; 20:1835-8.

Patient 3
Follow-up

Gabapentin

Thoracic MRI: no radiculopathy

Nerve block (trigger point injection)

Resolution

CPC

A 59-year-old man with fatigue, abdominal pain, anemia, and abnormal liver function

Friedman LS, et al. N Engl J Med 2014; 370:1542-50.

Presentation

Epigastric distress, ankle edema for 3 days

Personal distress

Difficulty sleeping

Dysgeusia

Nausea

T 37.3° BP 110/68 mm Hg ankle edema

Hct 30.6

Omeprazole and sucralfate

Next Days

Pain in both legs, dysgeusia

Loose stools

Increased abdominal pain, pain in right knee and both shoulders, fatigue

T 37.2° C P 89/min BP 131/89 mm Hg

Tender right knee

ED Visit

Diffuse abdominal pain, worse with eating, radiating to left side of chest, neck, shoulder, back

Constipation for several days, nausea, shortness of breath

BP 187/90 mm Hg / 129/80 mm Hg

Key Test Results

Hct	31.6%, basophilic stippling
ESR	3 mm/hr
Na	138 → 126 mmol/L
T. Bili	1.8 mg/dL (D. bili 0.4 mg/dL)
ALT	349 U/L
AST	179 U/L
Fe	166 mcg/dL
TIBC	211 mcg/dL
Ferritin	274 ng/mL
Abd x-ray	Dilated colon
CT	Large amount of stool in colon

Summary
59-year-old Man with Acute Abdominal Pain

- Stress
- Difficulty sleeping
- Fatigue
- Nausea
- Dysgeusia
- Constipation
- Edema, varicosities
- Muscle and joint pains
- Tachycardia
- Labile blood pressure
- Aminotransferase elevations
- Increased iron saturation
- Acute anemia
- Basophilic stippling
- Colonic pseudo-obstruction
- Hyponatremia

Diagnosis?

Diagnostic test?

Acute Porphyria

Abdominal pain
Nausea
Stress, restlessness
Constipation, pseudo-obstruction
Pain in extremities
Tachycardia, episodic hypertension
Syndrome of inappropriate ADH
(Red-colored urine)

Not: dysgeusia, basophilic stippling

Puy H, et al. Lancet 2010; 375:924-37.

The Porphyrias
Classification

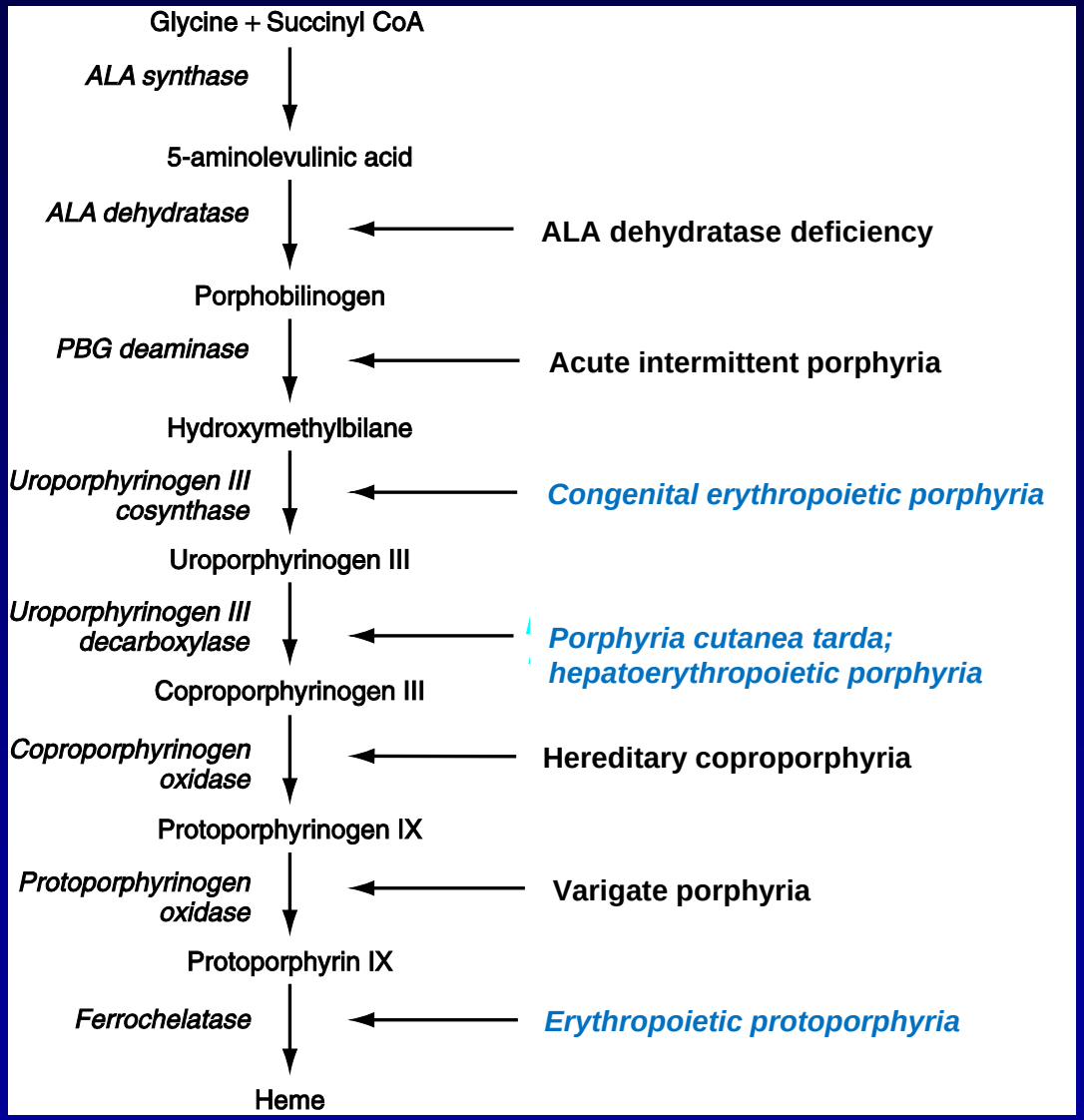
Acute (Neurovisceral)

Cutaneous

Acute (Neurovisceral) Porphyrias

	Enzymatic Defect	Mode of Inheritance	Usual Age of Onset	Major Site of Expression	Major Biochemical Findings
Acute intermittent porphyria	PBG deaminase	Autosomal dominant	Adulthood	Liver	Urine: ALA < PBG
Plumboporphyria	ALA dehydratase	Autosomal recessive	Childhood	Liver	Urine: ALA
Hereditary coproporphyria	Coproporphyrinogen oxidase	Autosomal dominant	Adulthood	Liver	Urine: ALA > PBG, coproporphyrin Stool: coproporphyrin
Variegate porphyria	Protoporphyrinogen oxidase	Autosomal dominant	Adulthood	Liver	Urine: ALA > PBG, coproporphyrin Stool: coproporphyrin, protoporphyrinogen

Heme Synthesis Pathway



ALA, 5-aminolevulinic acid; PBC, porphobilinogen

Acute Porphyria
Triggers

Starvation, negative energy balance

Drugs*

Alcohol

Smoking

Infections

"Stress"

* Not pirbuterol, per the American Porphyria Foundation (www.porphyriafoundation.org)

Features Inconsistent with Acute Porphyria

Dysgeusia

Basophilic stippling

(First attack age 59)

Dysgeusia
Altered sense of taste

Chemotherapeutic agents
- Cyclophosphamide
- Cisplatin

Pesticides and other toxins
- Lead
- Zinc deficiency

Other drugs
- Albuterol, pirbuterol
- Histamine H1-receptor antagonist
- D-penicillamine
- Metronidazole
- Boceprevir

Xerostomia

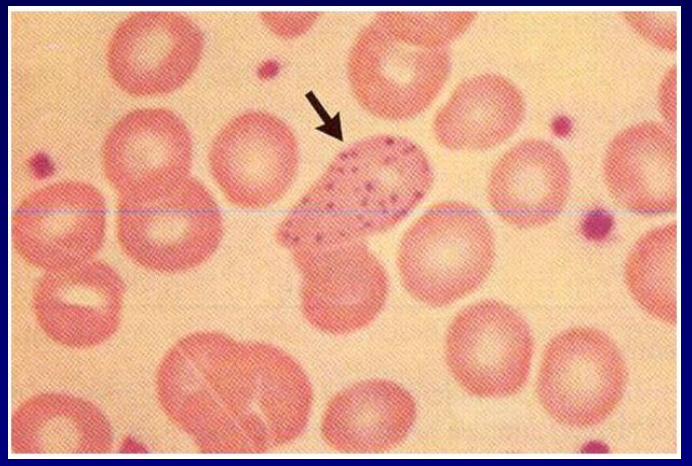
Basophilic Stippling

Sideroblastic anemia

Lead poisoning

Arsenic poisoning*

Thalassemia



Erythrocyte 5′-nucleotidase deficiency

Thrombotic thrombocytopenic purpura

*Associated with garlicky odor in breath, not true dysgeusia, and with severe diarrhea and pulmonary toxicity

Lead Poisoning

Features

- Abdominal pain ("lead colic")
- Nausea
- Dysgeusia
- Constipation, pseudo-obstruction
- Joint and muscle pains
- Hypertension
- Acute anemia
- Basophilic stippling
- SIADH
- Fanconi-type syndrome
- Neuropsychiatric effects

(Aminotransferase elevations may be seen)

Flora G, et al. Interdisc Toxicol 2012; 5:47-58.

Lead Poisoning can be Confused with Acute Porphyria

Lead inhibits ALA dehydratase

Overproduction of ALA in both

Plumboporphyria = deficiency of ALA dehydratase (children; one case in a 63-year-old man with polycythemia vera)

PBG also elevated in other porphyrias

(ALA and PBG are elevated during an attack)

Anderson KE, et al. Ann Intern Med 2005; 142:439-50.

Lead Poisoning
Signs

"Lead lines"

Bluish pigmentation at the gum-tooth line

Deposition of lead in bone

Blood Lead Levels

Elevated	≥10 mcg/dL
Acute lead poisoning	>100 mcg/dL
Inhibition of heme synthesis (ALA dehydratase)	~55 mcg/dL

↑ Free erythrocyte protoporphyrin

↑ Zinc protoporphyrin

Centers for Disease Control and Prevention. MMWR Morb Mortal Wkly Rep 2011; 60:841-5.
Ahamed M, et al. Sci Total Environ 2005; 346:48-55.

What was the source of lead exposure?

Lead Poisoning

Sources

Work place exposures

Lead smelting and refinement

Coal combustion

Manufacture:

Batteries	Solder
Paints	Tin cans
Pigments	Lead glazing
Car radiators	Ammunition
Cable and wire	Cosmetics

Fischbein A, Hu H. In: Rom WM, Markowitz SB, eds. Environmental and Occupational Medicine. Philadelphia: Lippincott Williams & Wilkins, 2007:954-990.

Lead Poisoning
Sources

Home exposures

- Lead paint
- Home renovations
- Natural disasters
- Moonshine
- Lead glass (leaching)
- “Laced” marijuana

Morgan BW, et al. Ann Emerg Med 2003; 42:351-8.
Busse F, et al. N Engl J Med 2008; 358:1641-2.

Curious Features of Case

Patient's "distress because of personal issues"

No exposure or occupation history

Partner

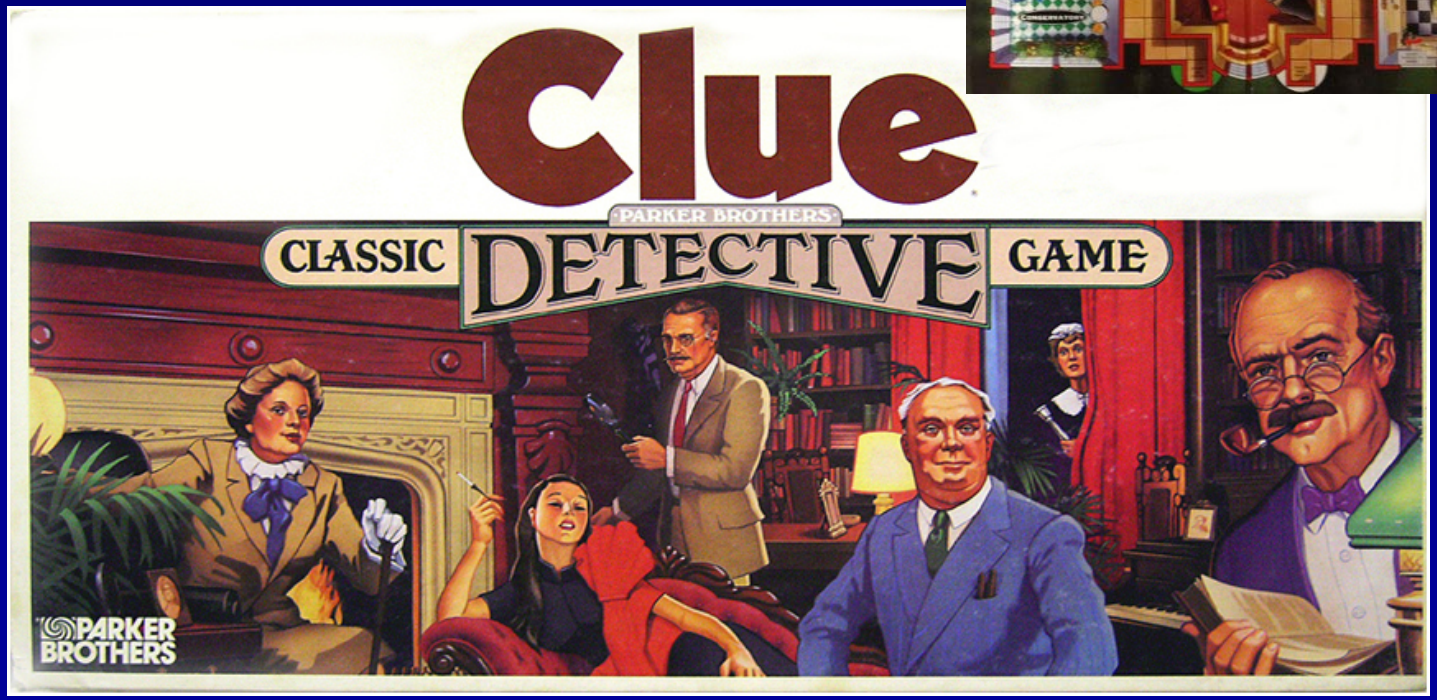
Phobic anxiety of lead poisoning?
or
A stressful relationship?

A Game of Clue???

The partner,

In the kitchen,

With lead!



Diagnosis

Lead poisoning

Lead level
91 mcg/dL

Zinc protoporphyrin
425 µmol/mole hgb