

A Mystery Block, Or Two

Ellen Clark PGY-1, Phillip Blatt PGY-2 & Vishnu Manoranjan MD

Oregon Health & Science University, Portland, OR

Admission Diagnosis

Necrotizing Pancreatitis complicated by WOPN

Initial Presentation

18 year-old female with history of obesity, cholelithiasis, and marijuana use disorder admitted for 1 mo at an OSH for acute necrotizing pancreatitis of unclear etiology

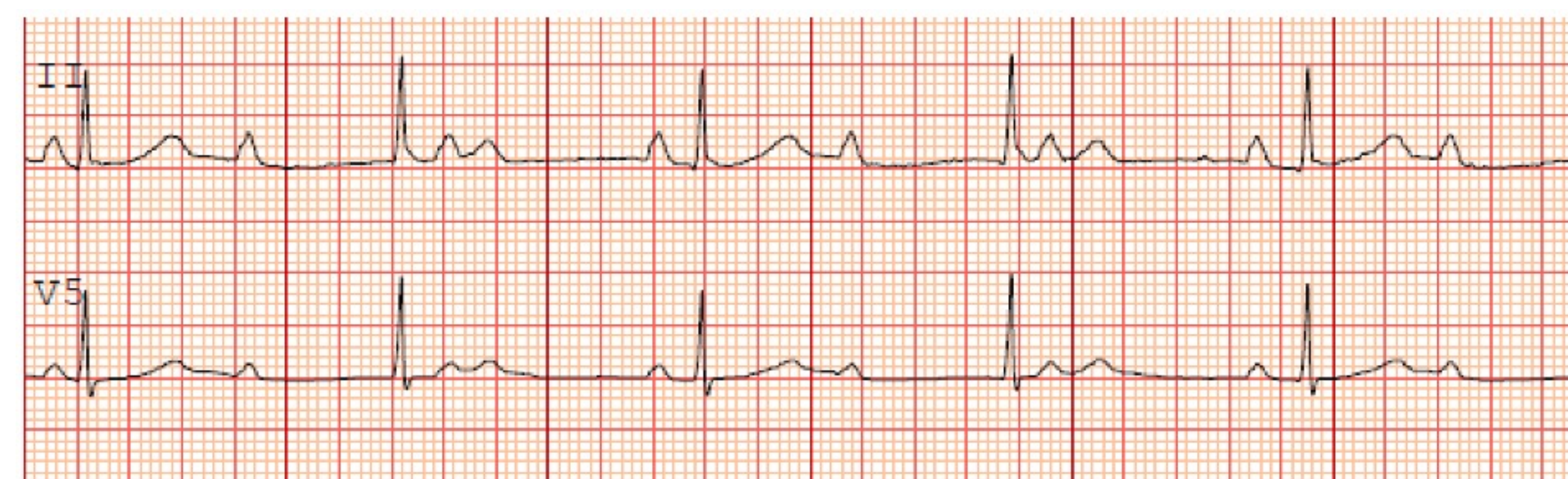
- Developed a large peri-pancreatic fluid collection with near occlusion of the portal venous confluence, and signs of partial GOO
- Also found to have asymptomatic complete heart block, no prior ECGs
- Transferred to OHSU for surgical intervention
- No recent trauma, travel, rash or tick bites
- No current or prior alcohol use
- Family history of acute pancreatitis, no cardiac arrhythmias/blocks

Clinical Course

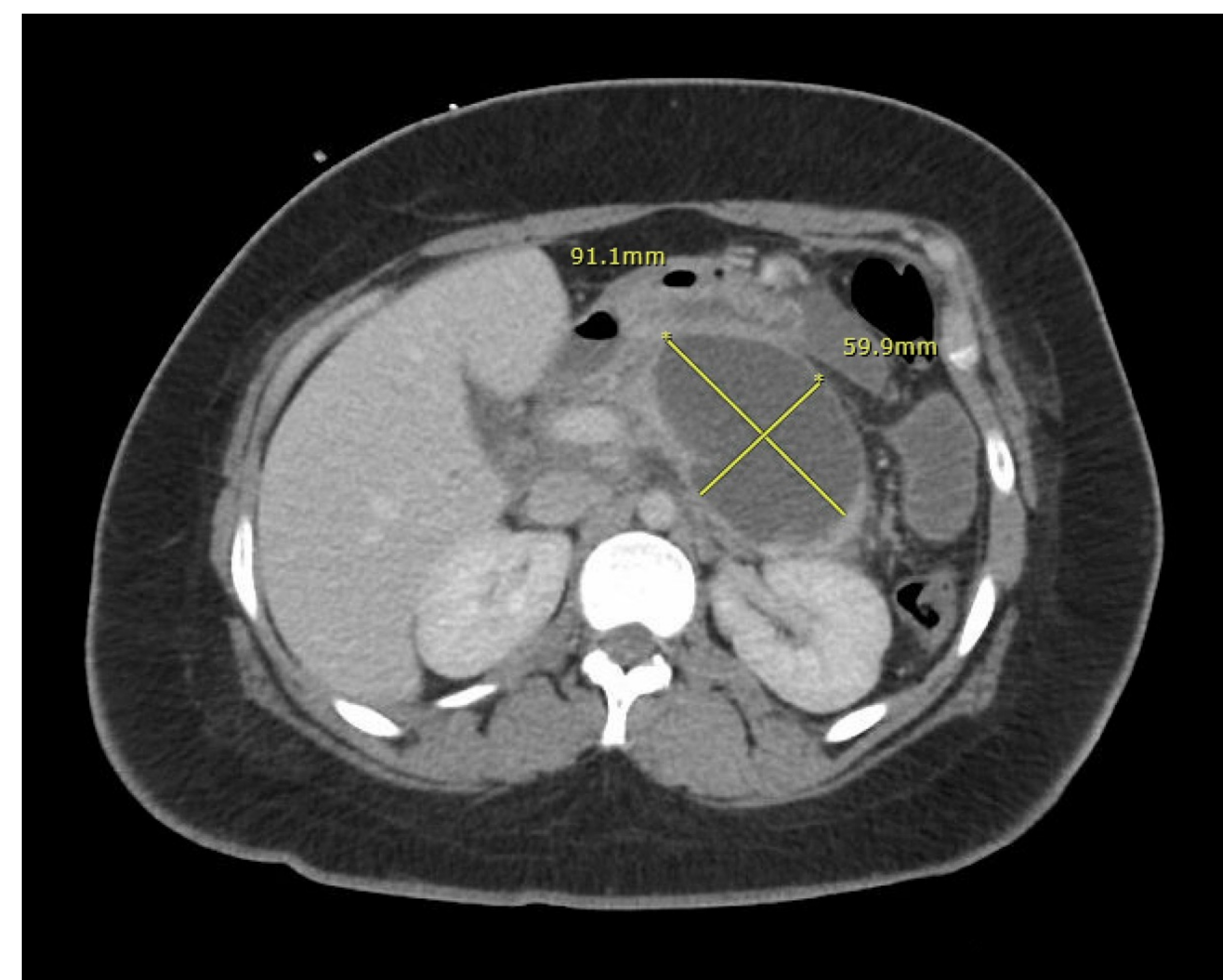
- **At outside hospital:**
 - No biliary duct obstruction: only mildly elevated GGT
 - Intermittent brady to 40s at OSH, increased to 100 with activity
 - cardiology at OSH felt block a/w acute pancreatitis, anticipated resolution with txt underlying disease
- **Overall improvement: normalizing of CRP, platelets and WBCs with conservative management**
- **Complete heart block required further eval prior to GI surgical intervention**
- **Her junctional rate augmented well with atropine, never required pacing, unremarkable work up**
- **Successful EUS cystgastrostomy:**
 - two large WOPN, s/p x2 pigtail stents
- **Holter on discharge**

Imaging & Labs

- **RUQ U/S** nonobstructive cholelithiasis without signs of cholestasis
- Repeat liver labs without signs of obstruction
- Normal TG levels, normal IgG4 subclass
- **MRCP** demonstrating decrease in size of peripancreatic fluid collection however still c/f worsening GOO
- **TTE and Cardiac MRI** unremarkable



ECG: Complete Heart Block: narrow QRS at rates >40s consistent with high junctional escape rhythm



CT A/P with IV Contrast on Hospital Transfer:

- Liver & biliary tree unremarkable
- Pancreas body and tail with WOPN 9.1cm x 6 cm
- New simple fluid collection within lesser sac 6 x 4cm
- Signs of disconnected pancreatic duct syndrome

Pancreatitis Evaluation

- Etiology unclear despite broad work up
 - Idiopathic / Genetic:** mat / pat grandparent with acute pancreatitis, (-) genetic panel
 - Biliary:** known cholelithiasis, although no signs of clear obstruction
- interval imaging with improvement, GI to follow up

Complete Heart Block Evaluation

- Etiology unclear, differential included:
 - Congenital:** no hx maternal CTD (maternal anti-ro/la)
 - Familial cardiac conduction disorder:** SCN5A, SCN1B, TRPM4 muts
 - Infectious:** Lyme (negative) or Chagas carditis (no recent travel history)
 - Inflammatory / genetic cardiomyopathies:** unremarkable cardiac MRI & (-) arrhythmia and cardiomyopathy genetic panel
- **On holt monitor, cardiology to follow up**

Key Points

- **High incidence of acute pancreatitis in US:** 2/3 cases: alcohol + gallstones; majority deemed "idiopathic" likely to have genetic risk, particularly in younger patients
- **Etiology of complete heart block in younger patient without a clear acquired or structural cause may be congenital or genetic (less common)**
- **Most asymptomatic patients with CHB will become symptomatic and require pacemaker (timing of insertion is not straightforward)**
- **This is a unique case with likely rare causes of 2 conditions known to have: high mortality rates & recurrence**

References

- Bai HX, Lowe ME, Husain SZ. What have we learned about acute pancreatitis in children? J Pediatr Gastroenterol Nutr. 2011 Mar;52(3):262-70.
- Forsmark CE, Baillie J; AGA Institute Clinical Practice and Economics Committee; AGA Institute Governing Board. AGA Institute technical review on acute pancreatitis. Gastroenterology. 2007 May;132(5):2022-44.
- Jaeggi ET, Hamilton RM, Silverman ED, Zamora SA, Hornberger LK. Outcome of children with fetal, neonatal or childhood diagnosis of isolated congenital atrioventricular block. A single institution's experience of 30 years. J Am Coll Cardiol. 2002 Jan 2;39(1):130-7.
- Schott JJ, Alshinawi C, Kyndt F, Probst V, Hoorntje TM, Hulsbeek M, Wilde AA, Escande D, Mannens MM, Le Marec H. Cardiac conduction defects associate with mutations in SCN5A. Nat Genet. 1999 Sep;23(1):20-1.