GI Grand Rounds

"A Lifetime of Abdominal Pain"

12/9/2004

Tim Edwards

PMH
Aug 25, 1992

- 4 year old male presents to a pediatric gastroenterologist for primary complaint of anorexia, intermittent abdominal pain which occasionally awakens him at night
- These problems have been present for >1 year
- Negative UGI study
- EGD with mild gastritis
- Rx with tagamet 40mg/kg/day and caffeine free diet

May 19,1994

- Seen for c/o abdominal pain with vomiting at bedtime
- Been doing well off all medications for 1 year
- Weight 44 lbs; height 43 inches
- Exam within normal limits
- GES T1/2 prolonged at 135 minutes
- Rx with Cisapride 5mg 20minutes QAC+HS for gastroparesis.

October 19, 1994

- Seen in f/u for gastritis and GERD
- Doing well on Tagament and Propulsid
- No abdominal pain
- Recently started with loose stools
- Height 46 in, weight 47.5 lbs
- Exam within normal limits
- Propulsid stopped with recurrence of severe abdominal pain within several days

September 21, 1998

- F/U of GERD and gastroparesis
- Prilosec 20mg/day and Propulsid 10mg BID-TID
- C/O crampy abdominal pain
- No vomiting
- 2-3 loose stools per day without blood or mucus
- No weight loss
- Height 55 inches; weight 90 lbs

February 25, 2003

- Seen for recurrent abdominal pain associated with vomiting for three weeks
- Off medications for 2 years
- Daily epigastric/substernal pain. Pain usually postprandial.
- Emesis is nonbilious, previously ingested food
- Lost 6 lbs
- Weight 121 lbs; Height 65 in
- Exam within normal limits
- Placed on bland diet and Nexium for recurrent GERD

August 18, 2003

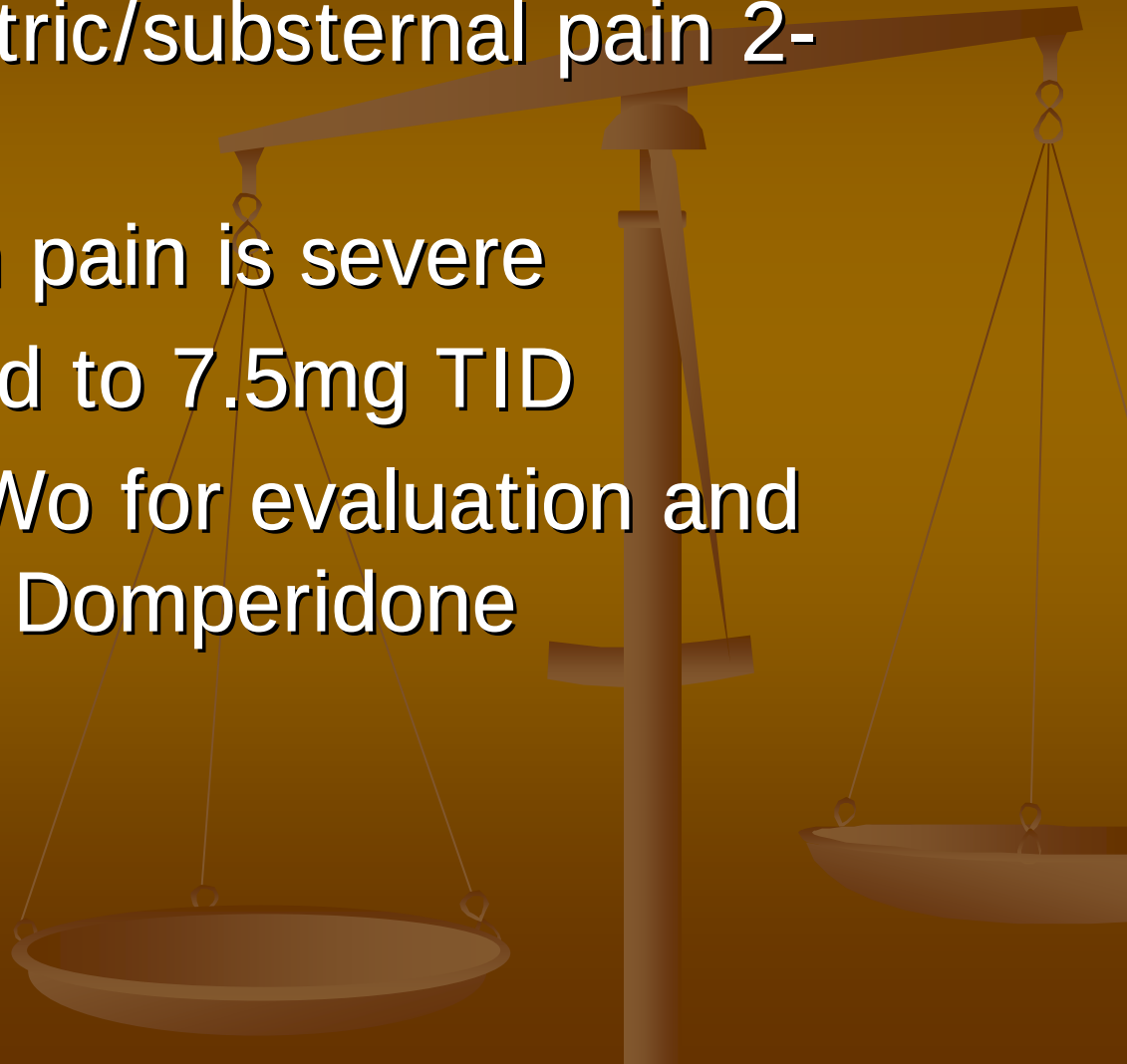
- Recent EGD within normal limits
- Biopsies normal, no celiac disease
- No improvement with Zelnorm 3mg BID
- Once per week with severe crampy abdominal pain relieved with nonbilious vomiting
- Weight 112 lb; height 66 inches
- Exam within normal limits
- 4H GES with T1/2 161 minutes

January 27, 2004

- F/U GERD and gastroparesis
- Recurrent abdominal pain has returned
- Prominent regurgitation
- Frequent nausea
- No diarrhea
- Decreased appetite; lost 4 lbs since October
- Increased Nexium 40mg BID; Increased Reglan to 5 mg TID

February 8, 2004

- Still with epigastric/substernal pain 2-3X/wk
- Will vomit when pain is severe
- Reglan increased to 7.5mg TID
- Referred to Dr Wo for evaluation and potential use of Domperidone



April 20, 2004

- Doing well on Nexium QAM and reglan BID
- No N/V/abdominal pain
- Repeated UGI with SBFT and CT abd/pelvis were without evidence of obstruction, stricture, or IBD
- Blood tests were without suggestion of secondary causes of gastroparesis
- Trial of Domperidone 10mg TID

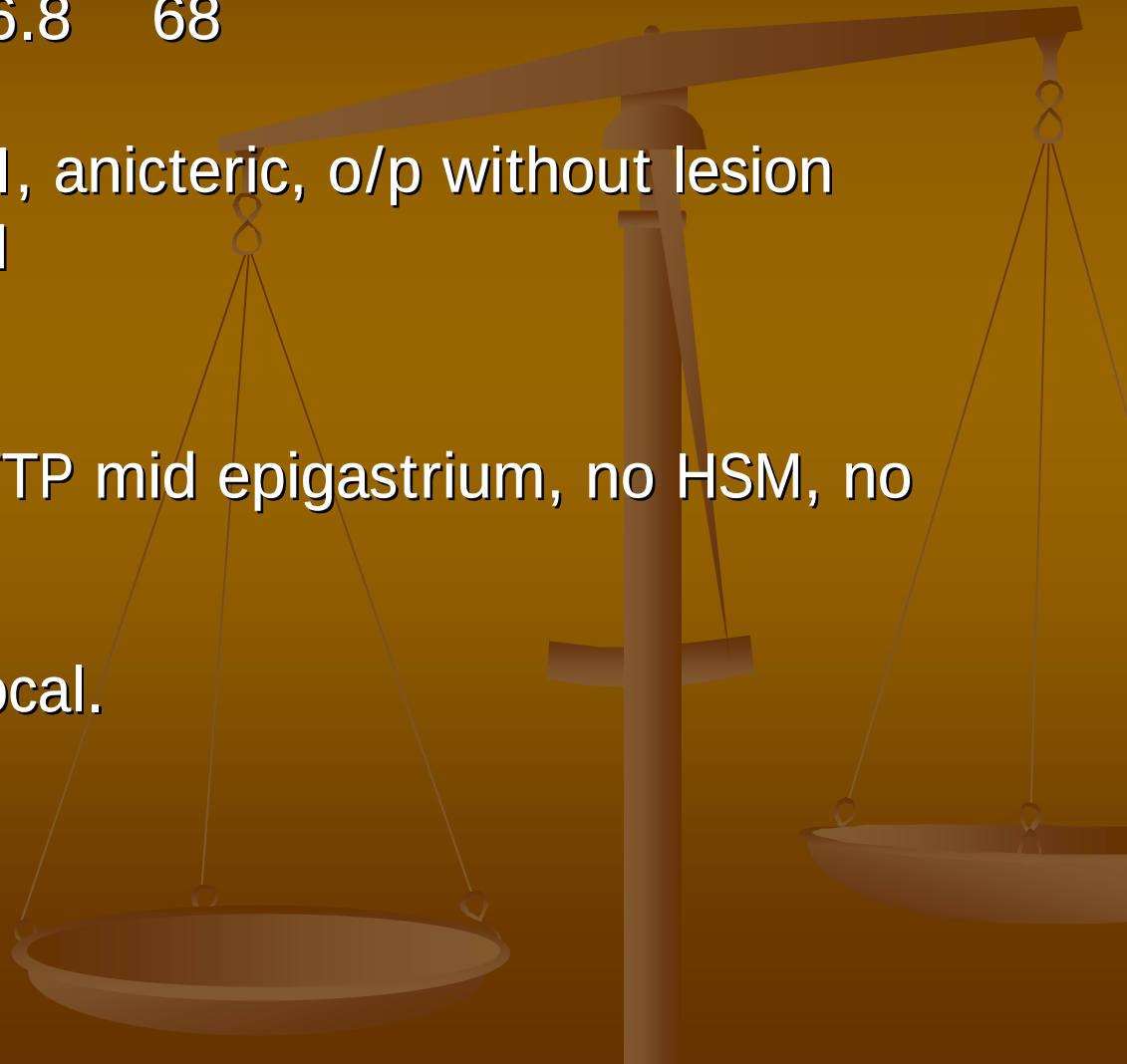
July 30, 2004

- Admitted to U of L Hospital from Dr Wo's clinic for 2 days of sharp, constant, nonradiating, epigastric pain which was relieved with vomiting
- No PO intake for 2 days
- Increase in typical GERD pain
- No diarrhea

- PMH: As outlined previously. O/W negative.
- PSHx: None
- FmHx: Noncontributory
- Social Hx: Does well in school, no ETOH, drugs
- All: NKDA
- Meds: Nexium 40mg PO BID, Domperidone 10 mg PO BID
- ROS: 11 lb weight loss previous 3 weeks

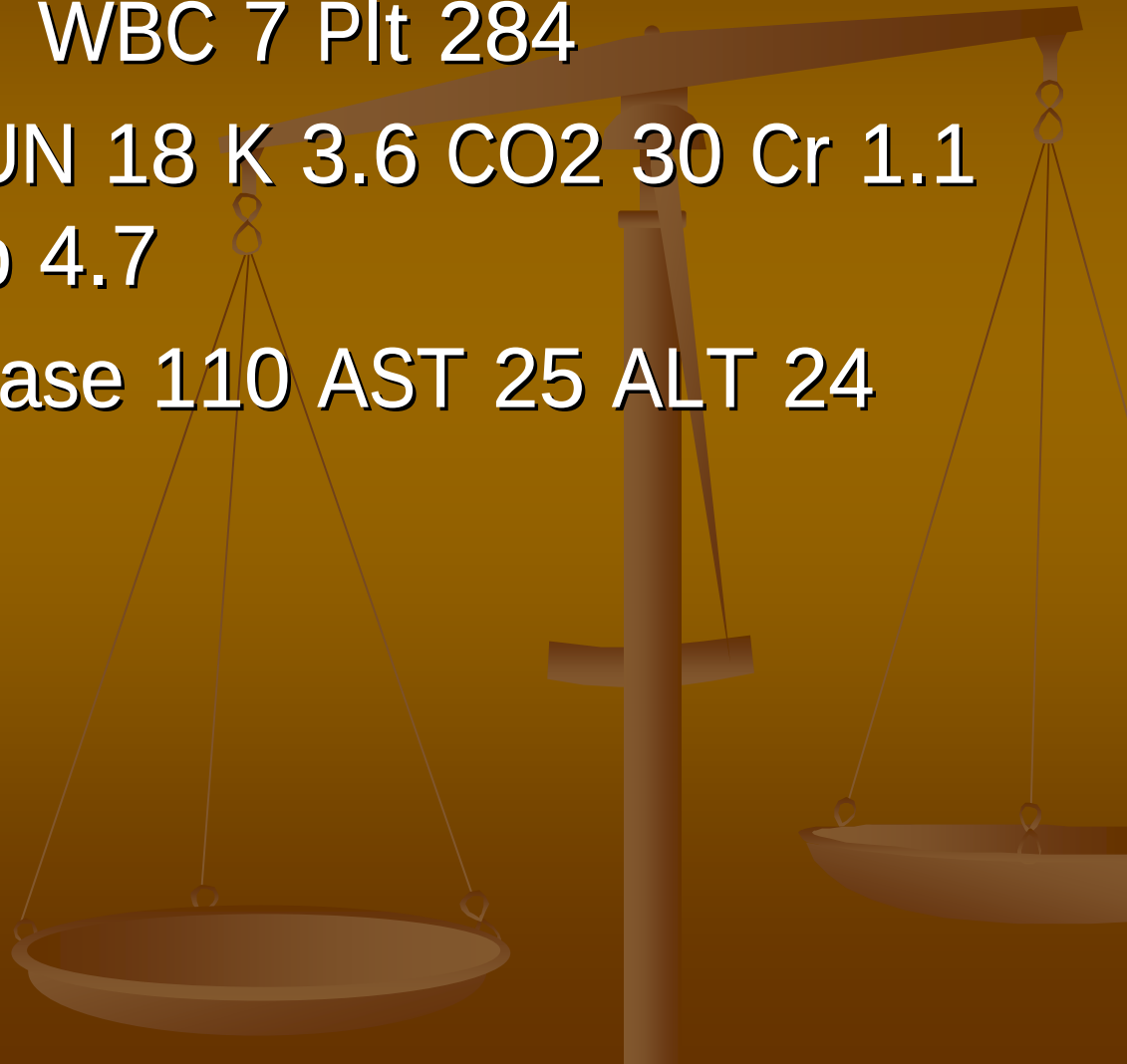
Physical Exam

- VS: 112/79 12 96.8 68
- Gen: NAD
- HEENT: NC/AT, EOMI, anicteric, o/p without lesion
- Neck: No TM, no LAN
- CV: RRR
- Lungs: CTA B
- Abd: S/ND/minimal TTP mid epigastrium, no HSM, no masses
- Ext: No c/c/e
- Neuro: AAOX3, nonfocal.



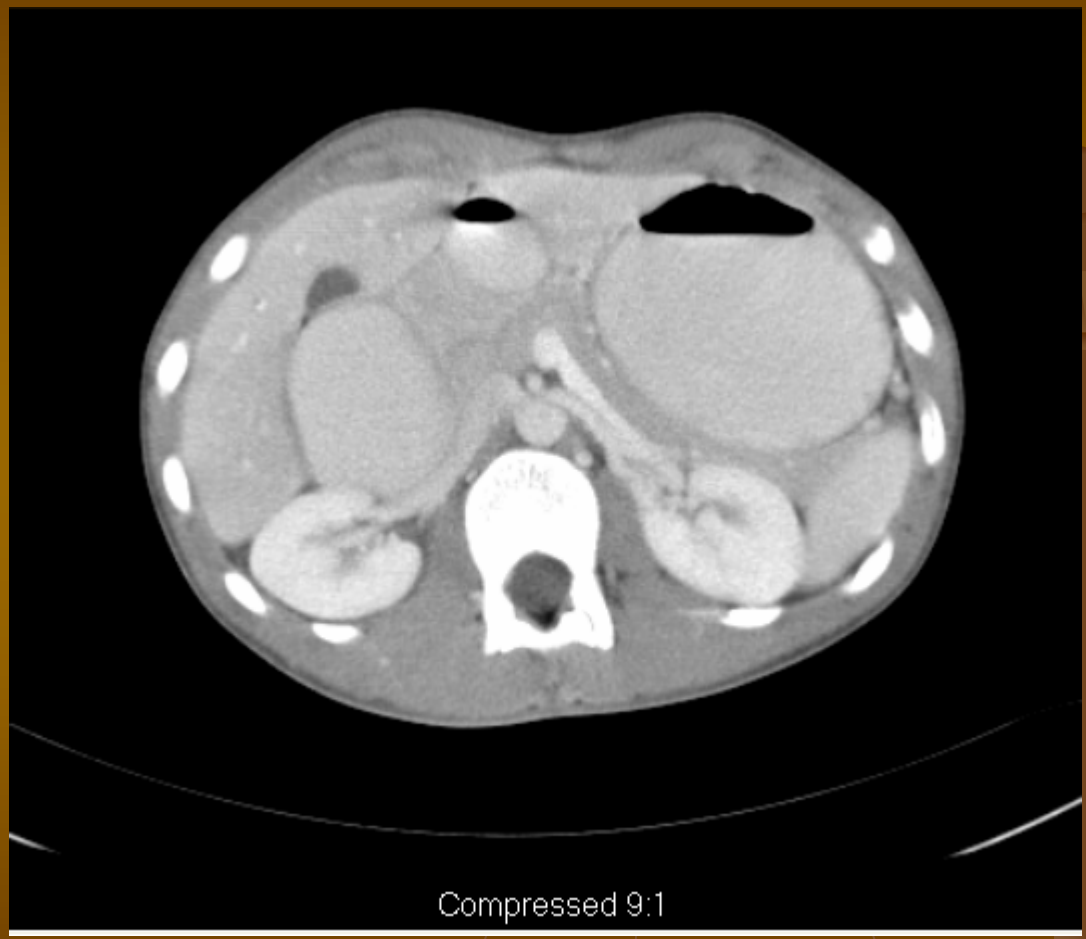
Laboratory

- Hgb/Hct 16/46 WBC 7 Plt 284
- Na 137 Cl 94 BUN 18 K 3.6 CO2 30 Cr 1.1 Ca 9 Tp 8.1 Alb 4.7
- Amylase 53 Lipase 110 AST 25 ALT 24 Tbili 0.08



Imaging

- 7/30/2004 CT Abdomen
- Marked dilatation of the stomach and proximal duodenum with a transition point near the third portion of the duodenum.
- May be secondary to focal dysmotility versus obstruction secondary to the mesentary and its vascular structures simulating a SMA syndrome.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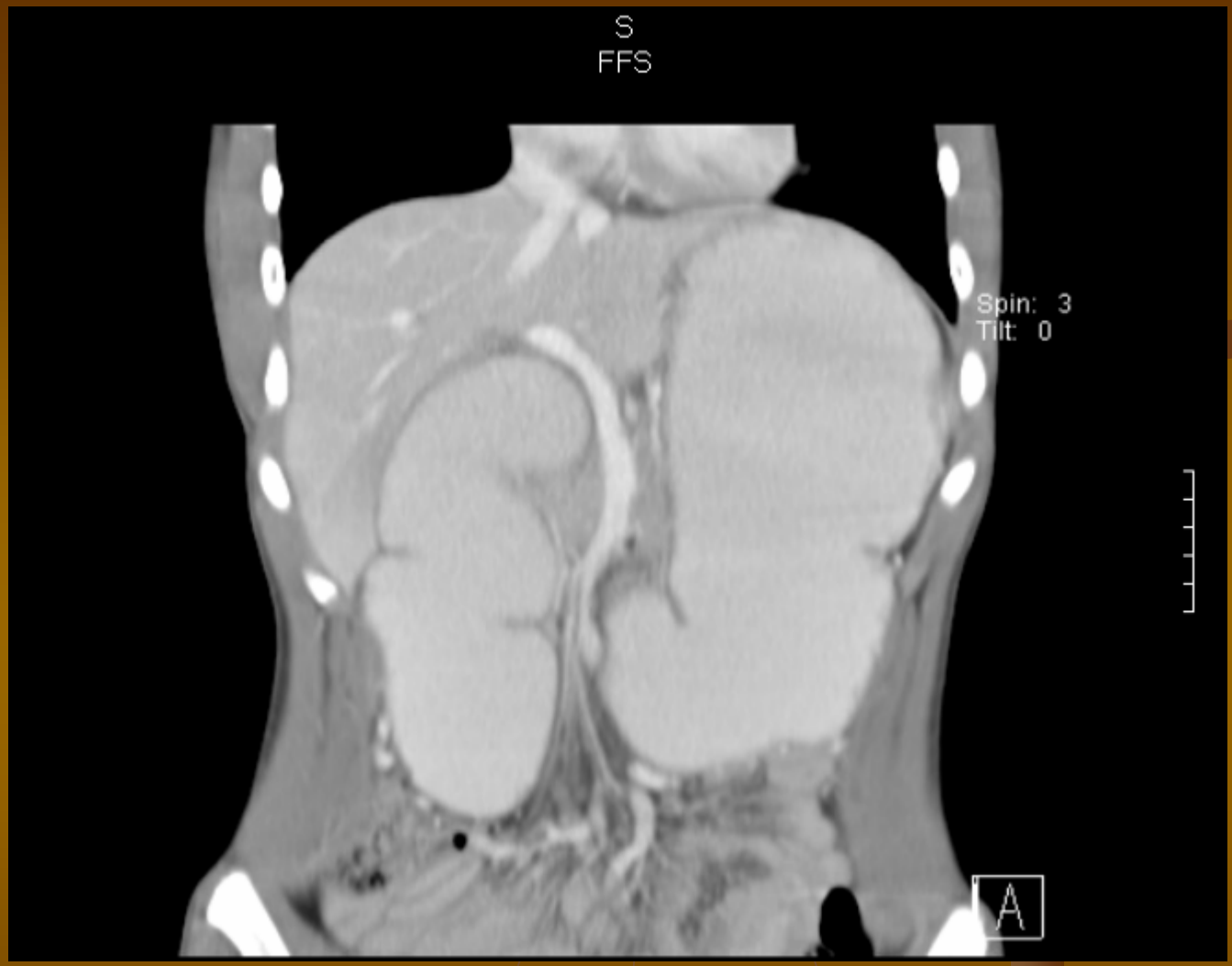
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FFS
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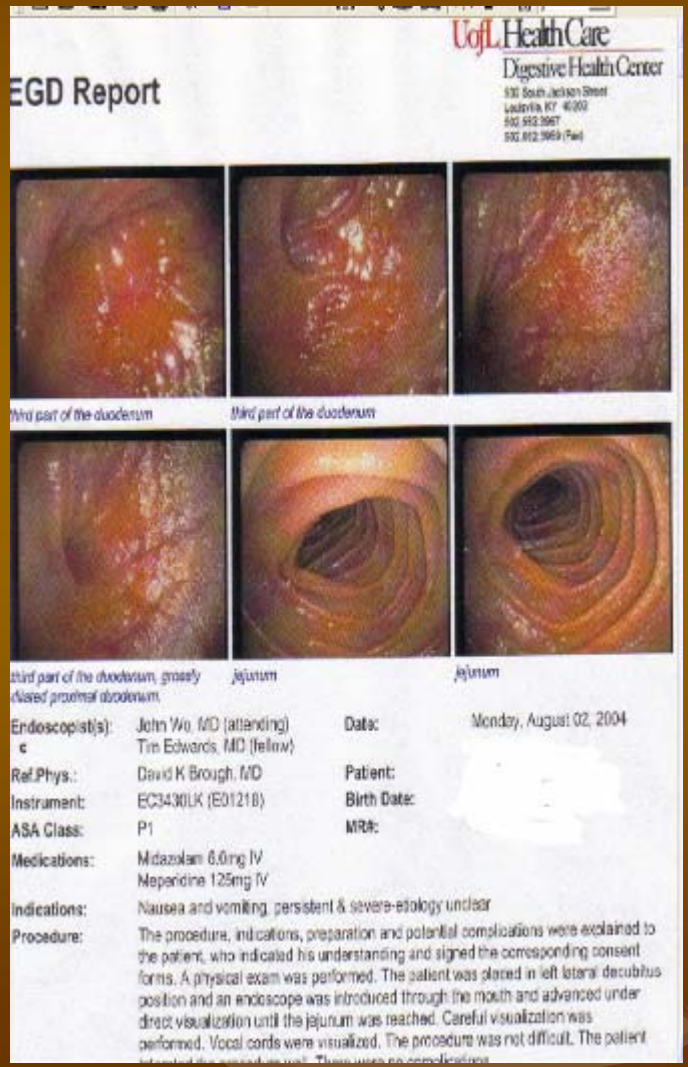
Upper GI limited

- August 3, 2004
- There is a filling defect seen in the second or third portion of the duodenum which may be c/w intrinsic vs extrinsic defect, but intrinsic defect is favored. The etiology may be ectopic pancreatic tissue, large adenomatous polyp, small bowel tumor or other multiple extrinsic causes such as SMA syndrome.
- Non-obstructive bowel pattern

EGD



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EGD Report

third part of the duodenum | third part of the duodenum

third part of the duodenum, grossly dilated proximal duodenum | jejunum | jejunum

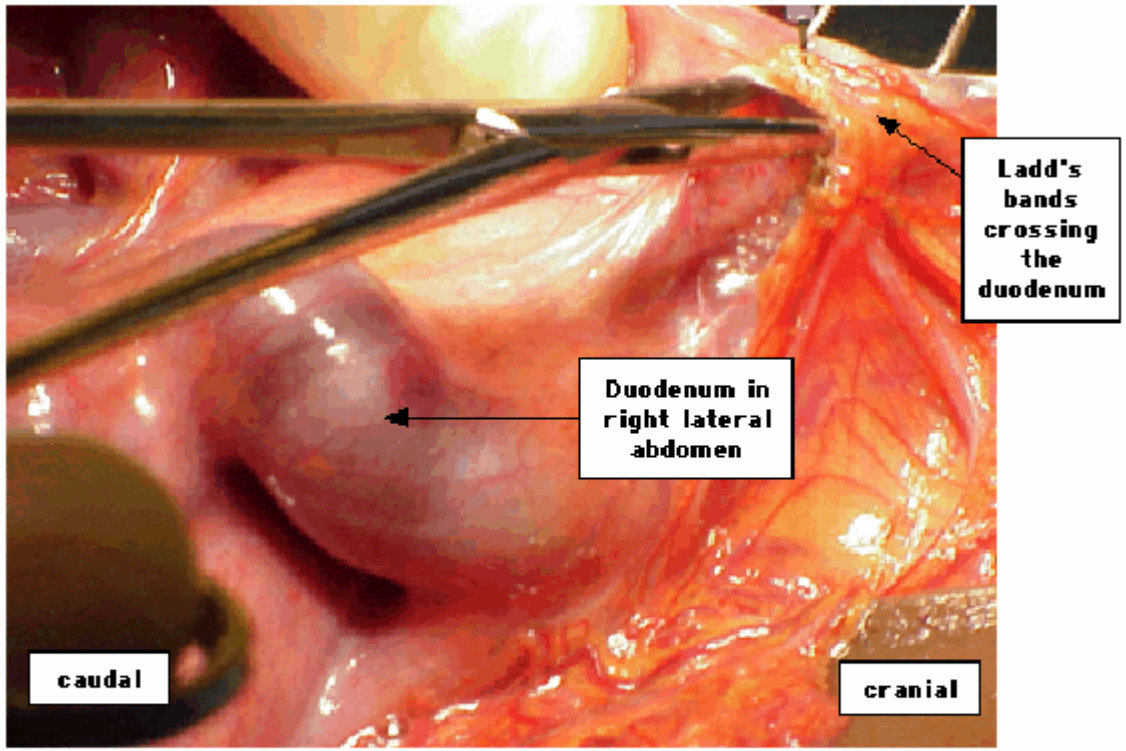
Endoscopist(s): c	John Wo, MD (attending) Tim Edwards, MD (fellow)	Date:	Monday, August 02, 2004
Ref.Phys.:	David K Brough, MD	Patient:	
Instrument:	EC3430LK (E01218)	Birth Date:	
ASA Class:	P1	MR#:	
Medications:	Midazolam 6.0mg IV Meperidine 125mg IV		
Indications:	Nausea and vomiting, persistent & severe-etiology unclear		
Procedure:	The procedure, indications, preparation and potential complications were explained to the patient, who indicated his understanding and signed the corresponding consent forms. A physical exam was performed. The patient was placed in left lateral decubitus position and an endoscope was introduced through the mouth and advanced under direct visualization until the jejunum was reached. Careful visualization was performed. Vocal cords were visualized. The procedure was not difficult. The patient		

Definitive Therapy

- Given endoscopic and radiologic evidence of extrinsic lesion resulting in obstruction a surgical consultation was obtained.
- On 8/4/2004 the patient was taken to the OR for exploratory laparotomy for diagnosis of source of duodenal obstruction

Operative Findings

- Liver, gallbladder, spleen, and stomach appeared normal
- Unable to locate the ligament of Trietz
- The duodenum was not fixed in usual retroperitoneal position
- Thick fibrous band of tissue crossed the 4th portion of the duodenum as an obstruction point
- Cecum and ascending colon were mobile and not attached to the lateral abdominal wall
- This was c/w intestinal malrotation



Malrotation The cecum is located in the mid-upper abdomen and is fixated to the right lateral abdominal wall by bands of peritoneum, known as Ladd's bands. Ladd's bands cross the duodenum causing extrinsic obstruction. Courtesy of Mary L Brandt, MD.

Intestinal Malrotation in the Adolescent

- Midgut malrotation is estimated to occur in approximately 1/500 live births
- The true incidence is unknown owing to those who remain asymptomatic and go undiagnosed
- Surgical series estimate 50-80% present in the first month of life
- 20% present within first year
- 10-20% present older than 1 year

Pathophysiology

- Intestinal malrotation can be simply defined as any deviation from the normal 270 degree counterclockwise rotation of the midgut during embryonic development

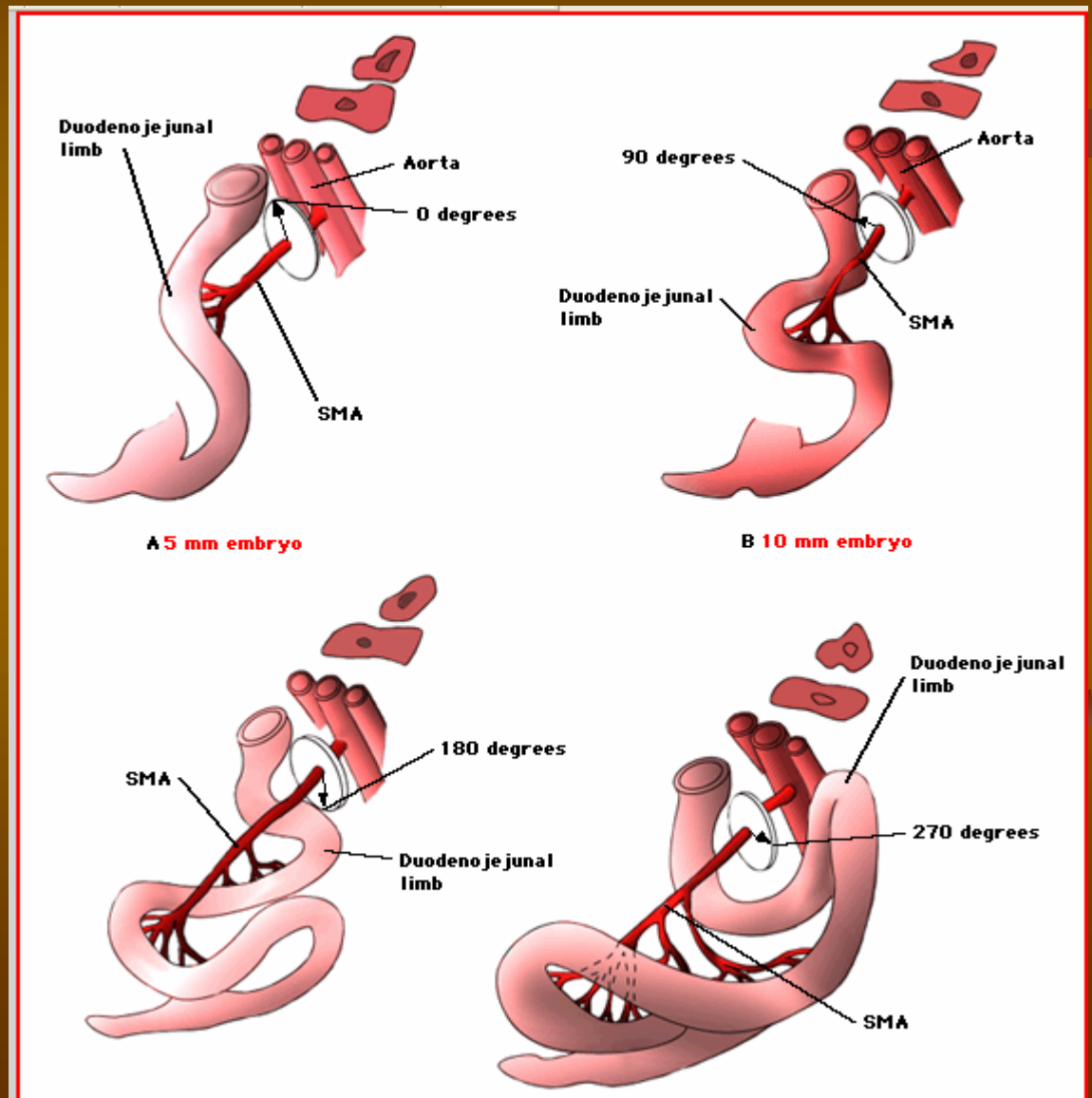
Normal Rotation

- In the first two months of development the growth of the intestines exceeds the capacity of the abdomen to contain them
- The bowel develops outside the abdomen in the yolk sac
- Normal counterclockwise rotation of the bowel is driven by the greater rate of proximal bowel growth as compared to distal bowel and the rapid growth of the fetal liver

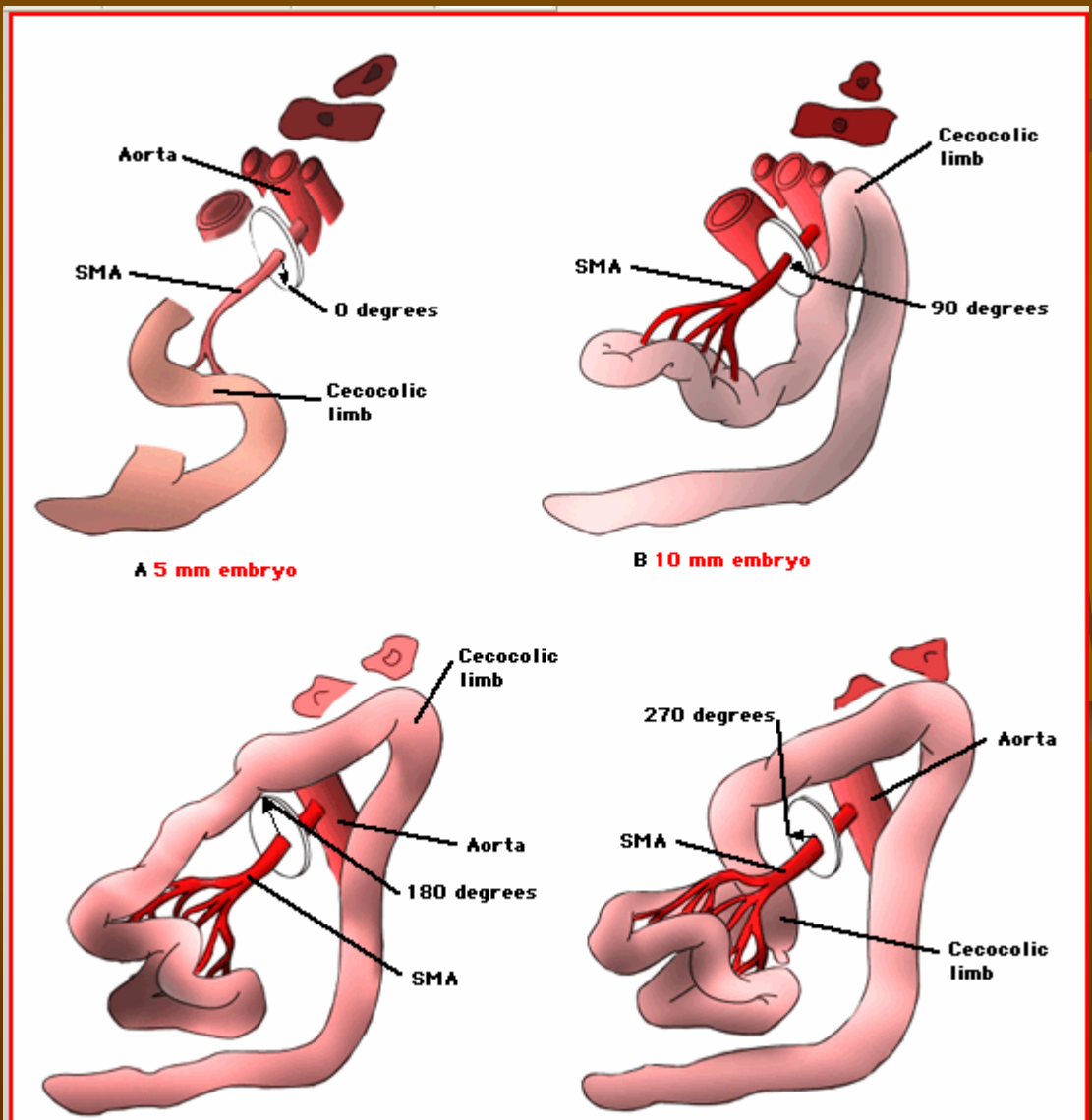
Return to the Abdomen

- First the duodenojejunal junction passes behind the SMA and becomes fixed to the upper left retroperitoneum. This forms the ligament of Trietz
- Second, the cecocolic junction passes from the left side of the abdomen, anterior to the SMA, assuming its position right of midline
- Overall, the bowel rotates 270 degrees counterclockwise from the original primary loop

Duodenojejunal Rotation



Cecocolic Limb



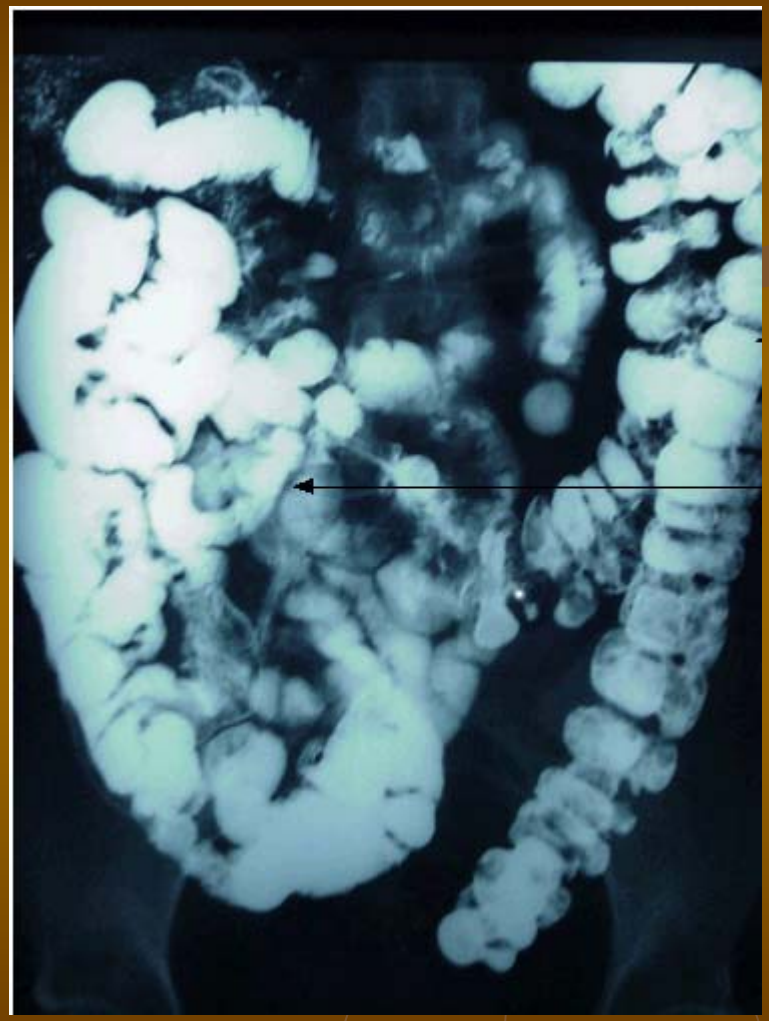
Normal Fixation

- At the completion of the rotation the intestines become fixed to the retroperitoneum by a broad based mesentery
- The mesentery extends from the ligament of Trietz to the ileocecal junction

Normal Mesenteric Fixation

Nonrotation

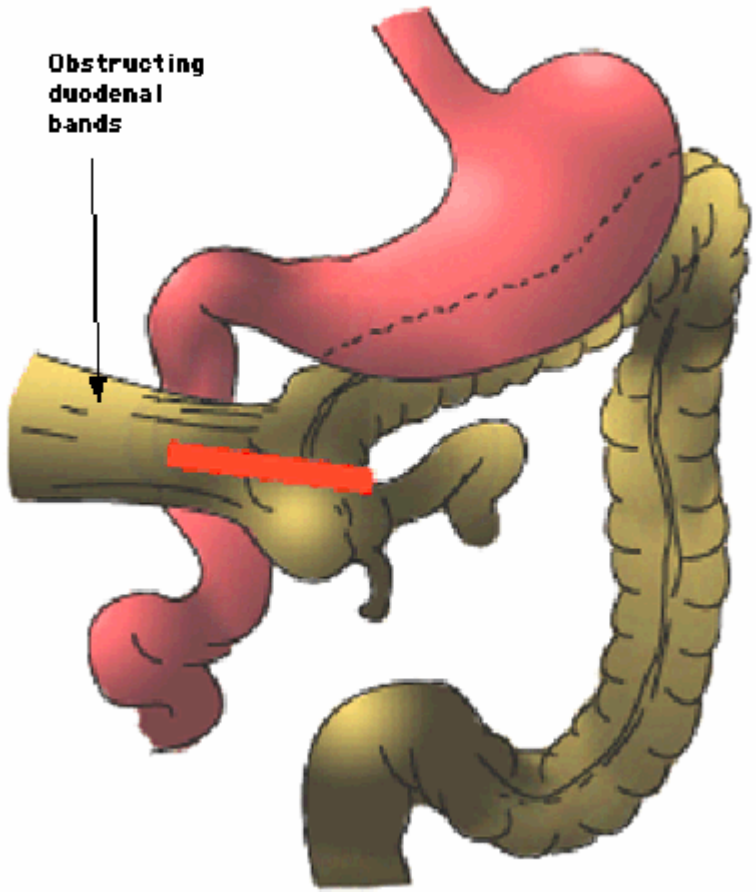
- Nonrotation occurs when the duodenojejunal and cecocolic limbs return the abdomen without any rotation
- The small bowel is located in the right abdomen
- The colon is located in the left abdomen



Malrotation

- Malrotation occurs with the duodenojejunal limb having no rotation. The cecocolic limb has partial rotation
- The cecum will be fixed to the right central abdominal wall by thick peritoneal bands. These bands may cause extrinsic compression of the duodenum
- This configuration results in a very narrow mesenteric attachment
- The narrow vascular pedicle predisposes to volvulus with subsequent ischemia and necrosis

Shortened Mesenteric Attachment



Clinical Presentation

- Majority of symptomatic malrotation is diagnosed within the first week of life
- The presentation of malrotation in adolescents and adults is highly variable
- Most will have intermittent abdominal pain
- The pain has an unusual nature in the it will be transient, vague, and not necessarily associated with any physical findings
- Often the pain is postprandial and may or may not be associated with vomiting
- Less common presentations include failure to thrive , malabsorption, diarrhea, motility disorders, and biliary obstruction

Clinical Presentation

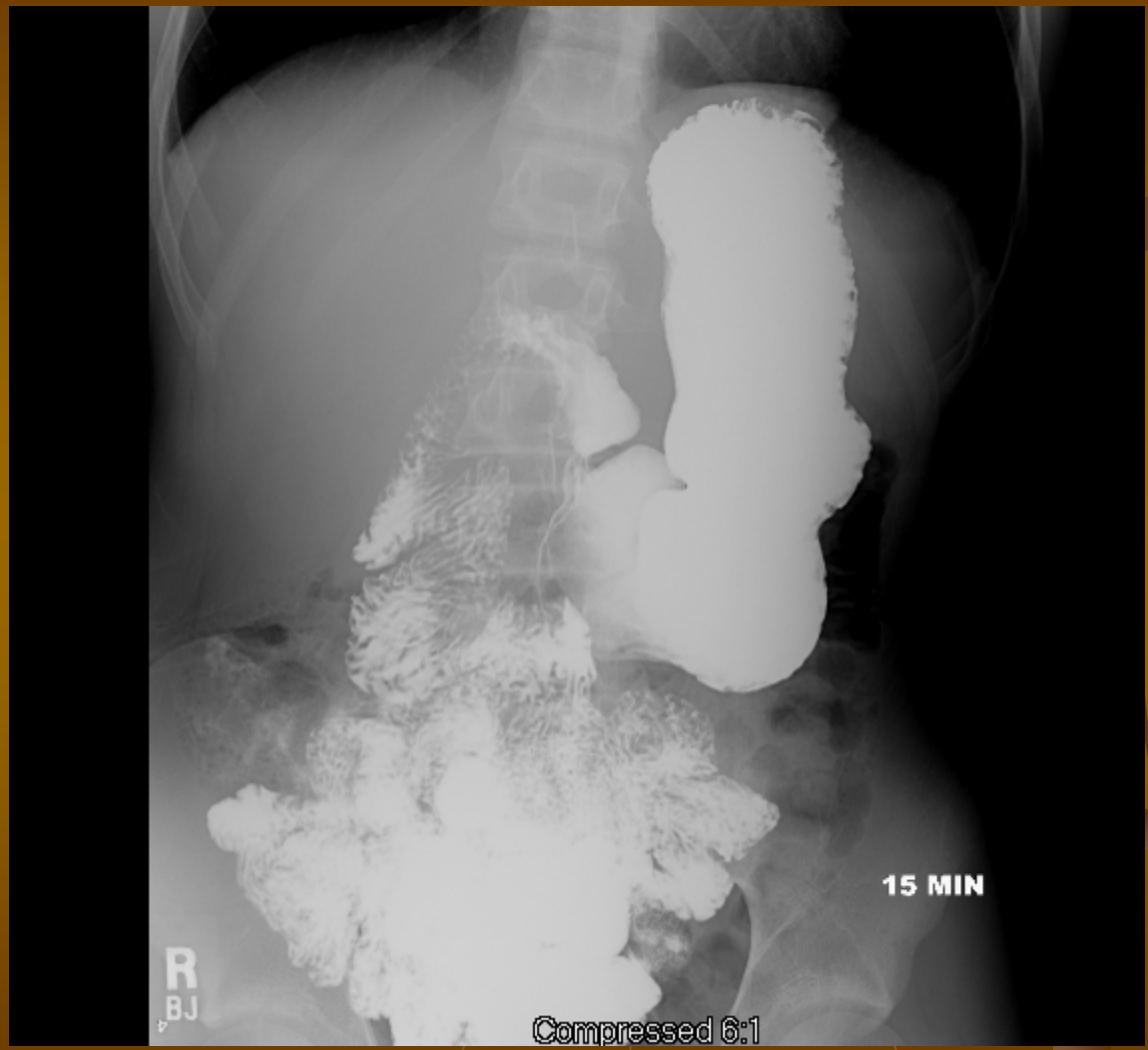
- Case studies report the time to diagnosis ranging from months to 17 years
- Common misdiagnoses include cyclic vomiting, food allergy, IBS, and motility disorders
- Often malrotation is first suspected in adolescents due to abnormal imaging studies or at laparotomy

Plain Films

- Conventional radiography is neither sensitive nor specific for malrotation
- Right sided jejunal markings and absence of stool filled colon in the right lower quadrant can be suggestive of malrotation
- Plain radiographs may be completely normal

Upper GI Series

- A limited Upper gastrointestinal barium series remains the most accurate tool for detection of malrotation
- Findings include failure of the duodenojejunal junction to cross the midline and lying below the level of the duodenal bulb and a clearly misplaced duodenum that has a corkscrew appearance
- 75% of cases have obvious signs of malrotation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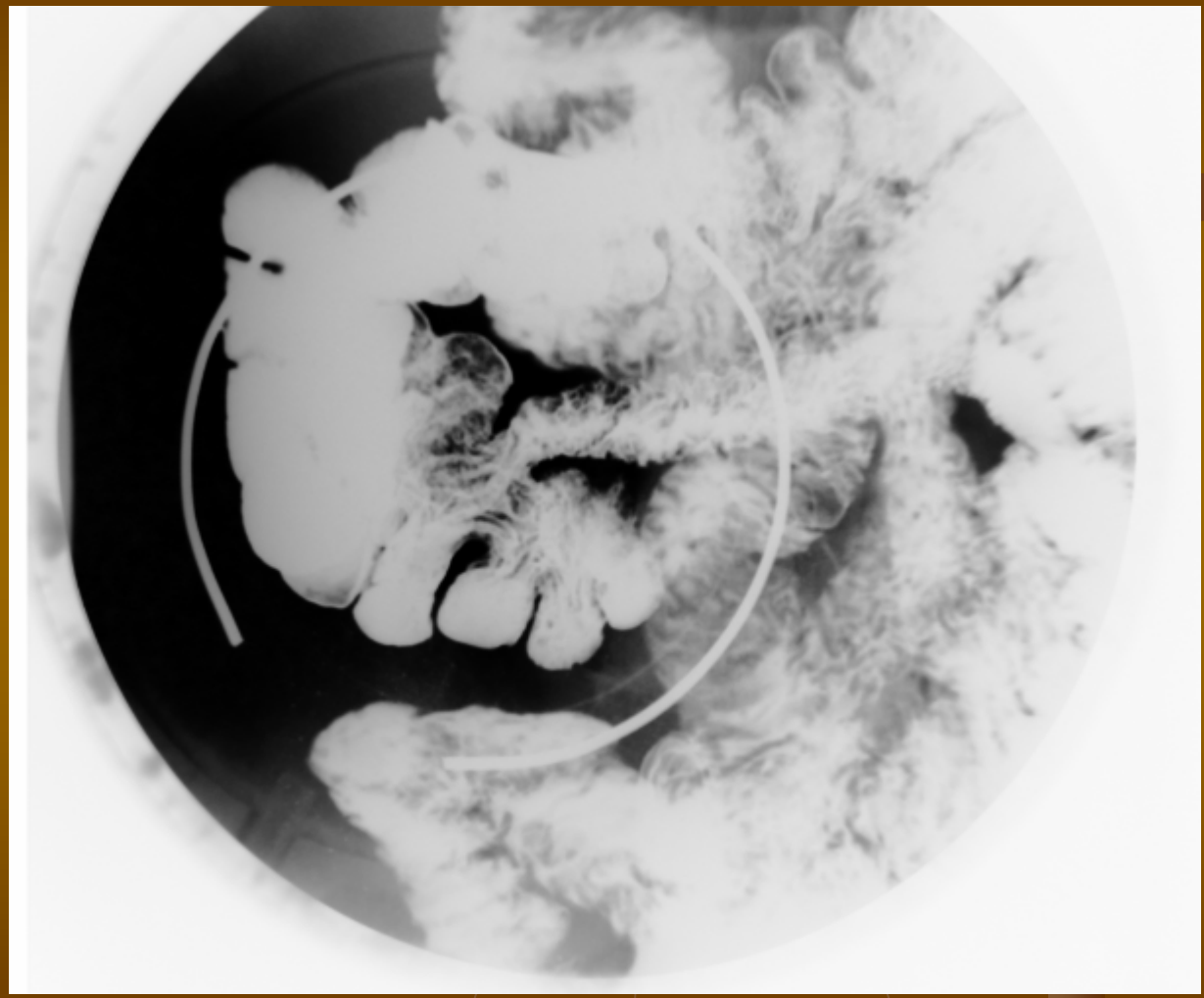
Barium Enema

- Contrast enema examination usually shows malposition of the right colon
- Contrast enema findings are nonspecific because the cecal location can be variable without malrotation
- 20% of patients with malrotation will have a cecum which assumes a normal position giving a false negative study

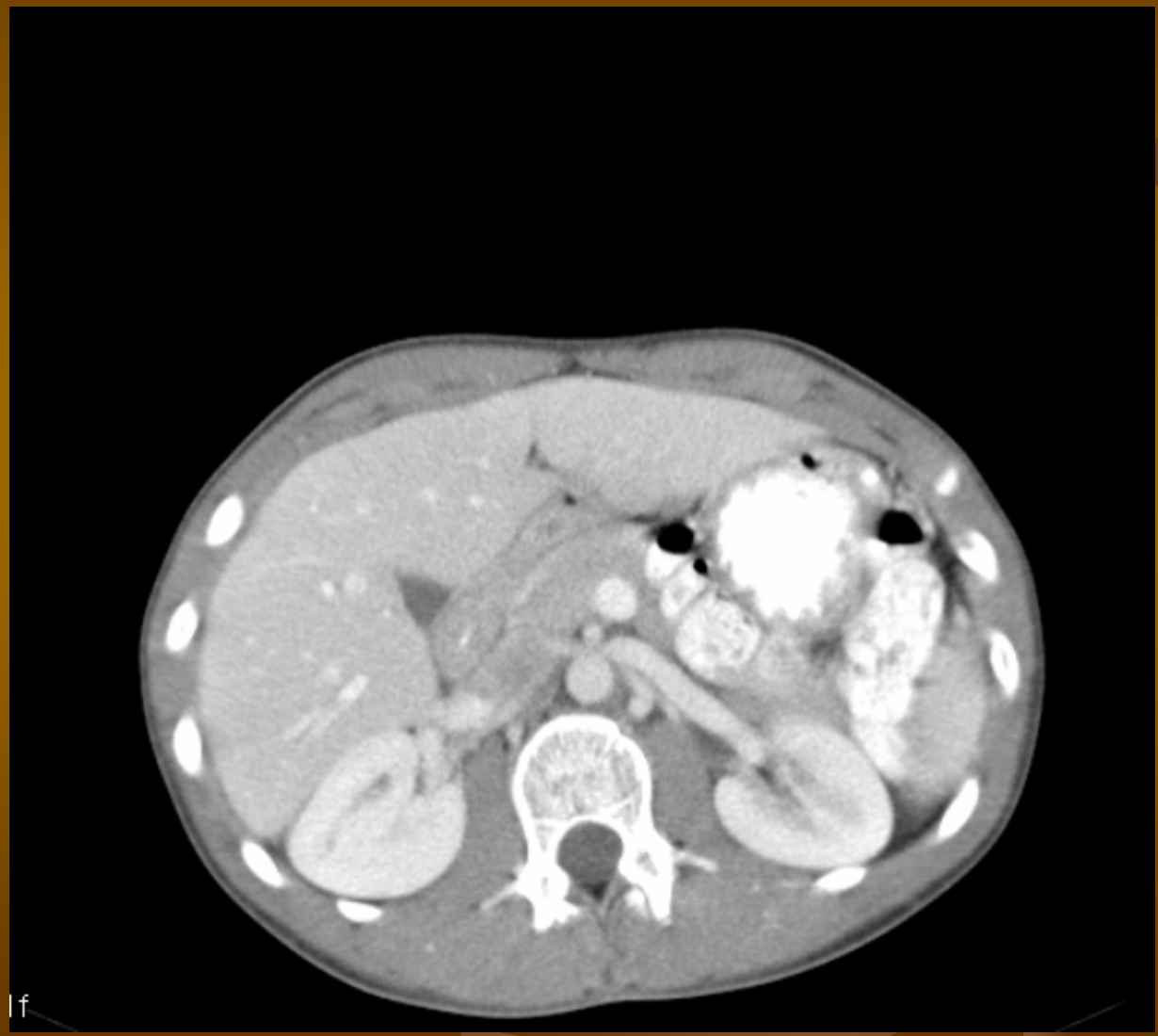


Ultrasound

- The role of ultrasound for diagnosing malrotation is not established
- A normal U/S does not rule out malrotation
- Findings which suggest malrotation are an abnormal relationship of the SMV and SMA; either anterior or to the left of the SMA
- The “whirlpool” sign of volvulus caused by the twisting of the vessels around the narrow mesenteric pedicle

CT Imaging

- Many cases of quiescent malrotation in adolescents and adults are detected by CT's obtained for other reasons
- CT can depict extra-intestinal findings not seen on conventional imaging
- Deviation of the normal SMV to SMA relationship (vertical or left-right inversion is suspicious for malrotation)
- The pancreas may show underdevelopment or absence of the uncinate process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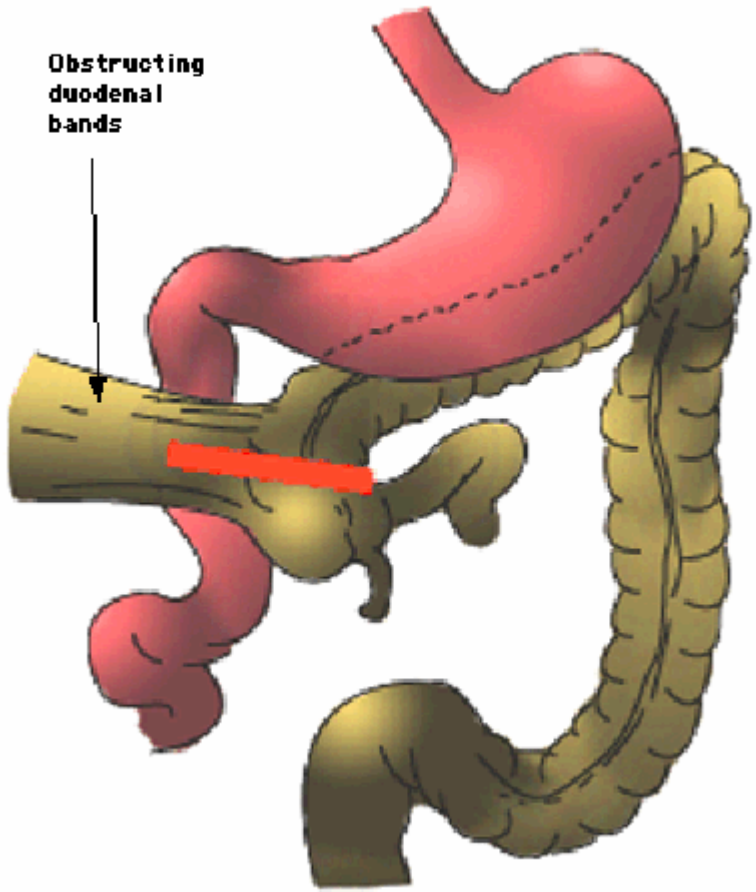
Treatment

- The treatment for malrotation, whether asymptomatic, related to acute duodenal obstruction, or incidentally found is surgical
- The “Ladd” procedure is used for treatment of duodenal obstruction secondary to malrotation
- He wrote his paper on this procedure in 1932

Shortened Mesenteric Attachment



Outcome

- Overall mortality rate is related to the presence or absence of volvulus and intestinal necrosis at the time of surgery
- Mortality approaches 0% in healthy patients without intestinal ischemia
- Recurrent volvulus can not be eliminated due to the inability to correct the underlying defect of malrotation. Estimated recurrence is 2-5%

Summary

- The clinical diagnosis of malrotation in adolescents and adults is rarely considered
- Adolescents/adults most often present with chronic abdominal pain with or without vomiting or chronic diarrhea
- Malrotation should be considered in any adolescent with intermittent abdominal pain, vomiting, diarrhea, or malabsorption
- Surgery is indicated in all cases of malrotation regardless of the discovery to reduce risk of volvulus and associated complications