A MIDDLE AGED WOMAN WITH ABDOMINAL PAIN A. Das, L. Lu. Baylor College of Medicine, Houston, TX. (Tracking ID # 173286)

LEARNING OBJECTIVES: 1) Consider atypical presentation of appendicitis in patients presenting with abdominal pain of unclear etiology. 2) Review the sensitivity and specificity of the radiographic studies in diagnosing appendicitis/appendiceal rupture.

CASE: A 50 year-old female presented with a 4 day history of intermittent left lower quadrant pain (LLQ). The pain was dull, non-radiating, variable in intensity, and worsened by lying on the left side or walking. She reported some decreased appetite with an episode of bilious vomiting, but was able to tolerate oral intake. She denied weight loss, fever, chills, diarrhea, constipation, dysuria, hematuria, past history of abdominal surgeries, or prior history of similar abdominal pain. Vital signs showed temp 100.3°F, BP 108/46, pulse 65, and respiration 12. Physical exam revealed normal bowel sound with tenderness to palpation in the lower abdomen, left more than right and mild rebound tenderness. Rectal exam was nontender with guaiac negative stool. Laboratory results including CBC, basic metabolic panel, liver function tests, amylase, lipase, and urinalysis were normal. Abdominal/pelvic CT revealed proximal small bowel distention without evidence of diverticulitis or appendicitis. A pelvic ultrasound followed by a trans-vaginal ultrasound showed left adnexal cystic lesions consistent with benign adnexal cyst. On the second day of admission, she developed small bowel obstruction with an acute abdomen and underwent an exploratory laparotomy revealing pelvic abscess with a perforated appendiceal tip. After appendectomy and pelvic abscess drainage, she recovered uneventfully with complete resolution of her abdominal pain.

DISCUSSION: Appendicitis is a common cause of an acute abdomen. The diagnosis is usually based on well-established clinical symptoms and physician experience. The typical clinical presentation of appendicitis is the gradual onset of vague periumbilical abdominal pain shifting to the right lower quadrant over approximately 24 hours associated with nausea, vomiting, and anorexia. Epigastric pain, non-specific abdominal pain, LLQ pain or pain in other locations may be initial presentation of appendicitis due to different evolving stage or abnormal appendiceal position. Approximately one third of patients with appendicitis present with localized pain outside of the right lower quadrant. However, LLQ pain as the manifestation of appendicitis is relatively rare and can be misleading. Rare causes of left sided appendicitis include situs inversus totalis, midgut malrotation, and right sided appendicitis with abnormal length projecting into left lower quadrant. The first two causes of left sided appendicitis are very rare, and only a few cases have been reported in the literature. For patients presenting with atypical symptoms of appendicitis, clinical observation with serial abdominal examination should be done. Computer tomography is often used to assist physicians in making diagnosis, but its sensitivity is 0.94 (95% CI 0.91-0.95) and specificity is 0.95 (95% CI 0.93-0.96). The sensitivity and specificity of ultrasonography are 86% and 81%, respectively. As illustrated in our case, the patient’s CT did not reveal appendicitis nor appendiceal rupture. There have been numerous citations for malpractice law suits mainly in the emergency department for missed appendicitis. Thus, high suspicion for atypical presentation of appendicitis and careful observation are indicated in patients with abdominal pain of unclear etiology.
ALL THAT FLATTENS VILLI IS NOT CELIAC SPRUE: CHRONIC DIARRHEA AND MALNUTRITION SECONDARY TO A PERIPHERAL T-CELL LYMPHOMA

P.M. Mckie1; A.S. Oxentenko1; K.M. Swetz1. 1Mayo Foundation for Medical Education and Research, Rochester, MN. (Tracking ID # 173034)

LEARNING OBJECTIVES: 1) Understand the evaluation and typical presentation of celiac sprue. 2) Recognize cases of refractory sprue and construct an appropriate differential diagnosis of alternative or secondary causes.

CASE: A previously healthy 69-year-old male presented with a 1-year history of persistent diarrhea. Small bowel biopsies elsewhere revealed villous atrophy suggestive of celiac sprue, although serologic markers were negative. His diarrhea did not improve with a gluten-free diet, and he subsequently developed significant malnutrition associated with weight loss and peripheral edema. On presentation to our institution, he was cachectic and debilitated. He had a diffuse xerotic, hyperkeratic rash over his entire body and scalp. Additionally, he had prominent lymphadenopathy, anasarca, and loss of pubic hair. Laboratories revealed anemia, prerenal azotemia, hypoalbuminemia, and multiple vitamin deficiencies. Tissue transglutaminase (IgA and IgG) and endomysial antibodies were negative. Dermatologic evaluation suggested acquired ichthyosis secondary to underlying malignancy. CT enterography revealed bulky adenopathy in the mesentery, retroperitoneum, and inguinal regions. Upper endoscopy revealed scalloped duodenal folds, and repeat small bowel biopsies showed marked villous atrophy similar to prior biopsies. Pathology showed lamina propria lymphocytosis with CD3 positive T cells, without intraepithelial lymphocytosis. Cytogenetic analysis revealed clonal T-cell receptor gene rearrangement, consistent with a low-grade peripheral T-cell lymphoma. An inguinal lymph node biopsy showed an anaplastic T-cell lymphoma of the same T-cell phenotype, suggesting aggressive transformation. The lamina propria (versus intraepithelial) lymphocytosis, low-grade (versus high-grade) morphology and the immunophenotyping were all consistent with a peripheral T-cell lymphoma rather than an enteropathy-associated lymphoma. Hematology consult was initiated but the patient expired of multiorgan system failure prior to treatment.

DISCUSSION: While small bowel villous flattening and intraepithelial lymphocytosis are the histologic hallmarks of celiac disease, clinical history, serologic markers and a response to a gluten-free diet help to substantiate the diagnosis. Herein, we present the case of a patient with chronic diarrhea and malnutrition, with villous atrophy and “lymphocytosis” suggestive of celiac disease, who had an unexpected clinical course. Gastrointestinal involvement by a peripheral lymphoma accounted for this patient’s chronic diarrhea and malnutrition. Instead of the intraepithelial polyclonal lymphocytosis seen in celiac disease, this patient exhibited a monoclonal CD3 positive T-cell expansion of the lamina propria consistent with lymphoma. The presence of lymphadenopathy and acquired ichthyosis were additional features suggestive of an underlying malignancy. The low-grade (versus high-grade) T-cell morphology and the immunophenotyping were all consistent with a peripheral T-cell lymphoma rather than an enteropathy-associated lymphoma. Serologically-negative celiac disease not responding to a gluten-free diet should raise the suspicion for other pathologic processes that can cause small bowel villous flattening and lymphocytosis, namely lymphoma.
THE GUT STOPS HERE—AN INTERESTING CASE OF CHRONIC INTESTINAL PSEUDO-
OBSTRUCTION D.D. ÖLveczky1, J. Potter2. 1Beth Israel Deaconess Medical Center, Cambridge, MA; 2Harvard
University, Boston, MA.  (Tracking ID # 172716)

LEARNING OBJECTIVES: 1. Discuss the differential diagnosis of chronic intestinal pseudo obstruction (CIP).  
2. Recognize pseudo-obstruction as a presenting symptom of primary amyloidosis

CASE: An 82-year-old woman presented with 2 months of anorexia, dysphagia, constipation and weight loss, and 
one episode of non-bloody, non-bilious emesis. Imaging revealed a partial small bowel obstruction. PMH was 
notable only for a salpingotomy for an ectopic pregnancy 53 years prior. She was treated conservatively and failed 
multiple feeding trials; subsequent exploratory laparotomy revealed dilated proximal small bowel and thickening of 
the distal small bowel without a clear obstruction or transition point. The biopsy was negative for inflammatory 
bowel disease, lymphoma or amyloid but showed increased numbers of plasma cells, consistent with a reactive 
process. She remained TPN-dependent with worsening anasarca and proteinuria. Radiology studies demonstrated 
persistent pseudo-obstruction. Six weeks later she developed hematemesis: EGD demonstrated distal esophageal 
erosions and a nodular, firm, friable pyloric mucosa. Congo red staining of samples from the pylorus revealed 
amyloid. Serum and urine immunofixation electrophoresis revealed free Bence Jones Kappa protein consistent with 
primary amyloidosis.

DISCUSSION: CIP is a rare syndrome that presents with symptoms and signs of bowel obstruction and 
radiographic evidence of intestinal dilation; however, no anatomic obstruction is found. Primarily due to an 
underlying neuromuscular disorder impacting small bowel motility, CIP can affect any part of the gastrointestinal 
tract. The primary process may be myopathic (eg. scleroderma), neuropathic (eg. diabetes mellitus, multiple 
sclerosis, Parkinson’s disease, amyloidosis, paraneoplastic), or both (eg. scleroderma, amyloidosis). Medications 
that affect the enteric nervous system, e.g. anti-cholinergic antidepressants, calcium channel blockers, and alpha-2 
adrenergic agonists, may also be implicated. Evaluation consists of: radiographic studies to exclude mechanical 
obstruction and alternative diagnoses e.g. Crohns disease; nutritional assessment; and confirmation of dysmotility 
using a transit test. Management options include: nutritional support; antibiotics for bacterial overgrowth; prokinetic 
agents; immunomodulator therapy; and surgery. Primary or immunoglobulin light chain-related (AL) amyloidosis is 
a rare plasma cell proliferative disorder in which fibrils of monoclonal light chains are deposited in various tissues. 
Presenting symptoms such as weight loss and fatigue are often non-specific. Other manifestations depend on the 
organs involved, most commonly heart, liver, kidneys, autonomic and peripheral nervous systems. Although 
amyloid deposits are frequently seen in the gastrointestinal tract, pseudo-obstruction occurs rarely. Diagnosis is 
made by tissue biopsy with Congo-red staining: if positive, serum and urine electrophoreses and immunofixation 
electrophoreses are performed. Treatment consists of oral melphalan and prednisone; response is usually limited. 
Young patients with high performance status may be offered high-dose, intravenous melphalan with autologous 
blood stem-cell support. Survival ranges from 6-21 months: multi-system involvement is a poor prognostic sign. 
Given the poor prognosis and frequent localized involvement, clinicians need to maintain a high index of suspicion 
to pursue the diagnosis even if initial biopsy of the affected organ is negative. Oncologist2006:11;824.
A BUG’S LIFE, FOR WHOM THE TOILET TOLLS: DISCOVERING THE UNEXPECTED CAUSE OF CHRONIC DIARRHEA IN A YOUNG, HEALTHY MAN. A. Casillas1; C. Lai1. 1University of California, San Francisco, San Francisco, CA. (Tracking ID # 172676)

LEARNING OBJECTIVES: 1) Learn the initial outpatient evaluation of chronic diarrhea, and recognize the importance of a detailed social history, including sexual practices, to detect risk factors for parasitic disease. 2) Identify the pathogenesis and clinical features of Entamoeba hystolytica/dispar.

CASE: A 28 yo man with self-diagnosed irritable bowel syndrome (IBS) presented with 3 weeks of rectal soreness and diarrhea. He recently had oral and anal intercourse with a male partner who tested positive for syphilis. Our patient was treated empirically for syphilis; RPR, HIV, and rectal swab for GC/Chlamydia returned negative. His symptoms did not resolve, and he returned to clinic 1 month later with persistent diarrhea ~8 stools per day with occasional blood and foul smell, epigastric pain and rectal pruritis. He denied fever or weight loss, diet changes, laxative use, sick contacts, or travel/camping. Exam revealed a thin man with normal vital signs, abdomen/rectum and GU region. Labs were normal: FOBT, UA, bacterial urethral and rectal swabs, TSH, CBC, lytes, and albumin. Stool cultures for bacteria, Giardia, and Cryptosporidium were negative, and O and P returned positive for Entamoeba hystolytica/dispar. He was treated with 10-days of metronidazole, and reported improved symptoms two weeks later. Because of his long-standing bowel symptoms, he underwent a colonoscopy, which ruled out inflammatory bowel disease (IBD). He was educated on hygienic anal and oral sex practices to prevent future fecal to oral acquisition and transmission of the parasite.

DISCUSSION: In industrialized countries, the most common causes of chronic diarrhea (duration >4 weeks) in immunocompetent persons are IBS, IBD, and malabsorption syndromes. The American Gastroenterology Association recommends that initial evaluation include CBC, TSH, lytes, total protein, albumin, and FOBT (all normal in our patient); stool microbiology is also recommended. Although parasitic infections are not a common cause of diarrhea in the U.S., a study in the year 2000 found that 1/3 of 5,792 fecal samples sent for diarrhea work-up tested positive for intestinal parasites. Entamoeba hystolytica/dispar, the culprit for our patient´s diarrhea, causes symptoms in 10% of infected patients. Although Entamoeba hystolytica/dispar consists of two different parasites, they are reported as one, because they cannot be morphologically distinguished. Clinically, E. hystolytica is assumed the organism in all symptomatic disease, since E. dispar is non-pathogenic. In the U.S., all patients with Entamoeba-positive stool are treated with metronidazole. The amoeba is transmitted fecal-orally as cysts, and is predominantly seen in migrants from and travelers to endemic areas, institutionalized patients, and men who have sex with men (MSM). Amebiasis usually has a subacute onset (1-3 weeks) with mild diarrhea to severe dystentery; fever and weight loss are common. Untreated infections are characterized by years of diarrhea, pain, and weight loss-- and may be confused with IBD. To prevent fecal-oral spread, travelers should avoid unboiled water and uncooked produce. Pertinent to our patient, a recent study of MSM HIV-negative males demonstrated that cleansing of the anus and penis before and after sex was independently associated with a lower prevalence of parasitic infection. This case is a reminder that primary care physicians should take detailed social histories to ascertain patients´ risks for parasitic diseases.
LEARNING OBJECTIVES: 1. To recognize the emergence of Vancomycin and Metronidazole-resistant Clostridium difficile associated diarrhea (CDAD) 2. To explore newer therapeutic options like Rifaximin for refractory CDAD

CASE: A 57-year-old woman was referred to a gastroenterologist’s office with 6 months of chronic non-bloody diarrhea. She denied any nocturnal symptoms, fever, weight loss, abdominal pain, nausea, vomiting or change in appetite. Her symptoms dated back to a dental infection, treated with penicillin 6 months ago. She used OTC loperamide with partial relief. Her past medical history included osteoporosis, anxiety and depression controlled with medications. She denied smoking or alcohol use. Her physical examination revealed a benign abdomen. Her hematological and biochemical investigations were unremarkable including a normal TSH. Colonoscopy revealed normal colonic mucosa but biopsies revealed non-specific mild focal acute and chronic colitis. The stool aspirate was negative for ova, parasites and other bacteria but was positive for Clostridium difficile toxin (A/B). She was started on metronidazole, lactobacillus and counseled about contact precautions to prevent faeco-oral transmission and avoidance of antibiotics. With no improvement after 8 weeks, treatment with oral vancomycin (250 mg QID) was commenced. After four weeks of therapy, diarrhea resolved and Vancomycin was tapered over two weeks. But her diarrhea promptly recurred. The dose of Vancomycin was escalated back and cholestyramine was added; but yielded no response. With increasing severity of symptoms, an infectious disease consultation was sought. Repeated stool testing confirmed the persistence of C. difficile toxin and absence of other pathogens. Serum immunoglobulins were normal. Rifaximin (200 mg TID) was begun and within three days, her diarrhea abated with complete resolution after 20-day course of Rifaximin. The patient continues to be relapse free now for over 3 months.

DISCUSSION: CDAD in hospitalized patients in the US is associated with an incidence of 1%, an annual mortality rate of 2% and a 50% increase in hospital costs due to prolonged hospital stays. C. difficile is a ubiquitous, spore-forming, gram-positive, anaerobic bacterium that proliferates when gut flora is altered, commonly due to antibiotic use. It produces two toxins, A and B, which cause inflammation, fluid loss and diarrhea that may be complicated by dehydration, sepsis and toxic megacolon. Standard therapy consists of discontinuation of the inciting antibiotic, fluid resuscitation, avoidance of antimitility agents and 2-week course of oral metronidazole or vancomycin. Increasing incidence of severe and refractory CDAD may be multi-factorial due to resistance to metronidazole or vancomycin, the occurrence of strains that hyperproduce toxin, or generation of novel toxins. Rifaximin, from the Rifamycin family of drugs, approved by the FDA for the treatment of Travelers’ diarrhea, has shown promise as a treatment for CDAD both in vitro and in early clinical testing. The mean MIC of rifaximin has been found to be 10 fold lesser than metronidazole and 100 fold lesser that vancomycin. A recent pilot study in Italy with 20 patients demonstrated comparable efficacy of rifaximin to vancomycin. This case illustrates the efficacy of rifaximin in CDAD refractory to months of standard therapy. An ongoing multi center, double blind, randomized trial of rifaximin and vancomycin will hopefully provide more insights.