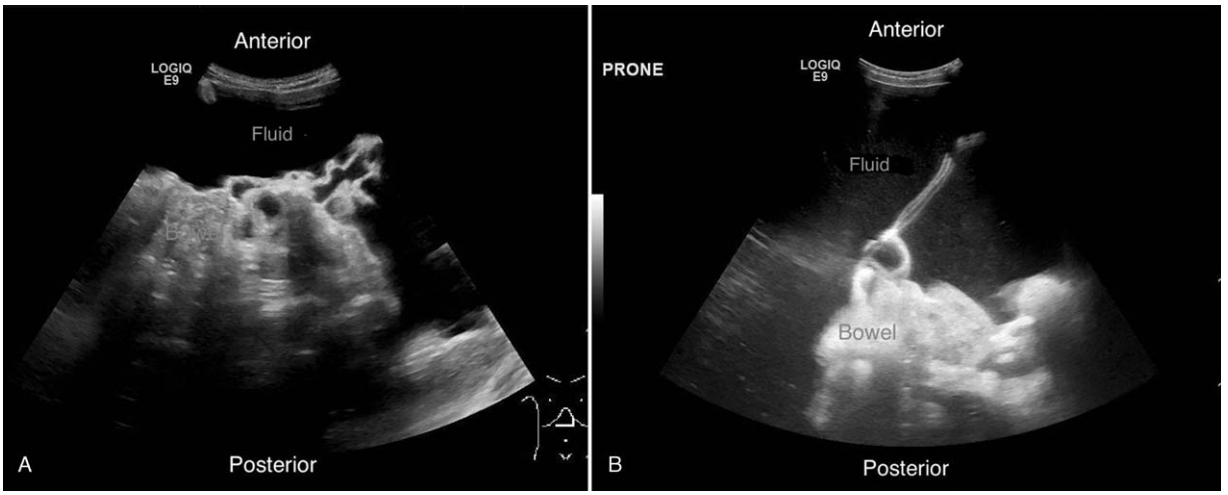

FIGURE 2. Ultrasound imaging showing bowel laying posterior to fluid in both supine (A) and prone (B) positioning.

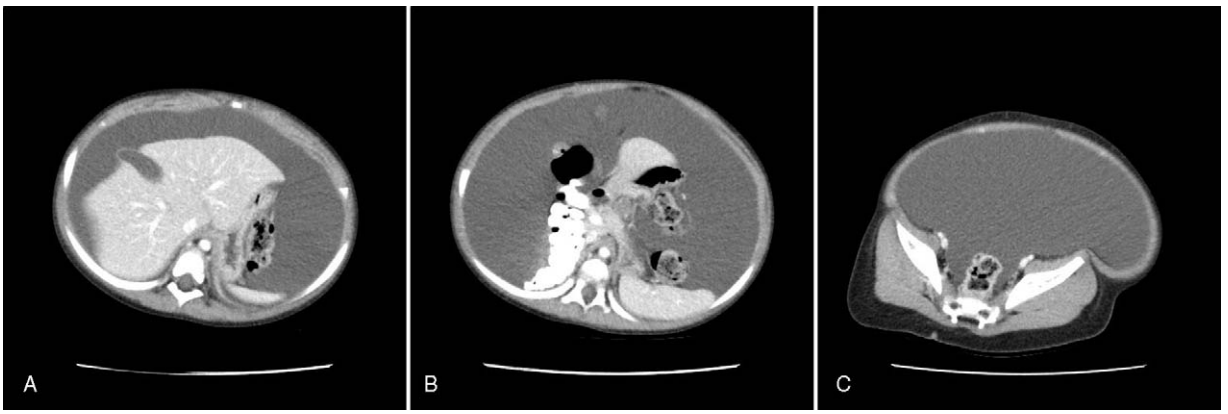

FIGURE 3. Computed tomography scan demonstrating findings in keeping with ascites at the level of the upper (A), mid (B) and lower (C) abdomen.

the diagnosis is often delayed as investigations are undertaken to rule out causes for ascites. In the clinical context of an otherwise healthy child with normal cardiac and liver function, omental lymphatic malformations should, however, be considered higher on the differential to avoid prolonged delays in treatment. As illustrated in our case, the use of supine and prone ultrasound imaging can help in distinguishing true ascites from a massive omental lymphatic malformation based on the relationship between the small bowel and fluid. Once the diagnosis is suspected, surgical resection is the definitive management.

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Clinical Remission and Normalization of Laboratory Studies in a Patient With Ulcerative Colitis and Primary Sclerosing Cholangitis Using Dietary Therapy

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Primary sclerosing cholangitis (PSC) is a chronic cholestatic disease that is frequently progressive, leading to liver cirrhosis, portal hypertension, and eventually to end-stage liver disease (1). PSC is characterized by inflammation and fibrosis of the intrahepatic and extrahepatic biliary ducts (2). The majority of PSC cases are associated with underlying inflammatory bowel disease (IBD) (1). The etiology of PSC is complex with multiple mechanisms potentially being involved including an immunologically mediated bile duct injury triggered in genetically susceptible patients by toxic or infectious agents that may gain access through

the diseased colon (3,4). Recently a resurgence of interest has developed around diet as a treatment for IBD. Much of this interest has been spawned by the understanding of the link between diet, the microbiome, and its relationship to Crohn disease and ulcerative colitis (UC) (5). Case reports and observational studies have demonstrated the resolution of active UC with instigation of the Specific Carbohydrate Diet (SCD) as primary therapy (5–7). The association between PSC and UC is well known, and although the SCD has been used to treat UC in many patients, the effects of SCD therapy on concurrent liver disease have not been documented. Herein we present a patient with UC and PSC who responded clinically, biochemically, and endoscopically to nutritional therapy as primary therapy for her UC and PSC.

CASE HISTORY

The patient was a 13-year-old girl presenting with abdominal pain, weight loss, and bloody diarrhea. One month before diagnosis, she was treated with a course of azithromycin for pneumonia. Two weeks later, she developed crampy abdominal pain and bloody diarrhea, occurring 3 to 4 times per day. She developed fevers, increased fatigue, nausea, and vomiting. Initial evaluation at an outside emergency department (ED) revealed normal complete blood count and urinalysis. *Clostridium difficile* antigen/toxin as well as stool cultures were negative. Erythrocyte sedimentation rate (ESR) was elevated at 54 mm/hour. The patient was transferred to Seattle Children's Hospital. On examination, she was afebrile with normal vitals. She had diffuse abdominal tenderness on examination, but no peritoneal signs. Given a history of bloody diarrhea with negative stool studies, she underwent endoscopic evaluation, which revealed erythema, edema, and friability in the stomach, and duodenal bulb. Colonoscopy revealed moderate-to-severe erythema, edema in the entire colon, and ulceration in the proximal colon. The terminal ileum was also erythematous and edematous. Biopsies revealed moderate-to-severe chronic and mild-to-moderate active colitis with chronic active ileitis (Fig. 2). Given endoscopic findings in the ileum, an initial diagnosis of Crohn disease was given and patient was initiated on exclusive enteral nutritional (EEN) therapy. Patient was reclassified later as UC after review of biopsies, imaging, serology, and conferencing with proceduralist. The patient responded well with gradual resolution of abdominal pain and decreased stooling to 2 to 3 semi-formed nonbloody stools per day. C-reactive protein decreased from a maximum of 4.2 to 1.6 mg/dL before discharge from hospital. At follow-up after discharge, CRP completely normalized (Fig. 1).

Initial laboratory studies at Seattle Children's also revealed elevation of alkaline phosphatase (1378 IU/L; normal 130–560 IU/L) and GGT (851 IU/L; normal 5–55 IU/L) (Fig. 1). Magnetic Resonance Cholangiopancreatography (MRCP) during hospitalization showed common bile duct dilatation with regions demonstrating a beaded appearance as well as an abrupt transition at the distal common bile duct suggestive of possible stricture. There was evidence of mild dilatation and beading appearance of the intrahepatic bile duct as well. Further evaluation revealed an elevated immunoglobulin G titer with a normal IgG subclass, antimitochondrial antibody and anti-liver kidney microsomal antibody negativity, and serum carbohydrate antigen (SCA) 19-9 elevation of 906 U/mL (normal <55 U/mL). She was ASCA IgA- and IgG negative and ANCA positive (Table 1). She underwent percutaneous liver biopsy as well as endoscopic retrograde cholangiopancreatography with sphincterotomy and stricture dilatation. Biopsy results were consistent with biliary ductal obstruction and PSC (Fig. 2).

She was maintained on EEN for 10 weeks and then transitioned to the SCD. She remained asymptomatic and clinically well for over a year. She has 2 bowel movements per day, formed, nonbloody, nonmucousy. She has had good weight gain, good energy, no oral ulcerations, no joint pains, and no rashes. She has had complete normalization of her labs with normal ESR, C-reactive protein, haemoglobin, and albumin as well as her liver enzymes including GGT and alkaline phosphatase (Fig. 1). Initial calprotectin done 6 weeks after initial diagnosis was 1127 mg/kg (normal <163 mg/kg). This normalized at 14 (108 mg/kg) and 34 weeks (78 mg/kg). In addition, repeat SCA 19-9 as well as immunoglobulins normalized (Table 1). Repeat endoscopy/colonoscopy 1 year after diagnosis, revealed normal appearing mucosa in the stomach, duodenum, terminal ileum, and colon. Biopsies revealed mild chronic colitis with focal activity (Fig. 2). MRE results showed focal regions of minimal intrahepatic biliary duct prominence within hepatic segment IV.

DISCUSSION

PSC is generally a progressive disease, which ultimately leads to severe complications including cholestasis and hepatic failure. Hepatic transplant is the only curative therapy. Approximately 50% of symptomatic patients do not survive beyond 15 years from diagnosis unless transplanted. Currently, there is no effective medical therapy that alters PSC disease progression, although ursodeoxycholic acid and vancomycin have been associated with biochemical improvements in PSC (8).

The intestinal microbiota of patients with PSC is characterized by decreased microbial diversity, and a significant overrepresentation of *Enterococcus*, *Fusobacterium*, and *Lactobacillus* genera. This dysbiosis is present in patients with PSC with and without concomitant IBD and is distinct from IBD without PSC (9). As the relationship between PSC, IBD, and the intestinal microbiome is becoming better elucidated, therapeutic options aimed at manipulating the intestinal microbiome will likely play an important role in treatment. A prime example is vancomycin, which has been shown to have a beneficial effect in PSC (10). Although dietary therapy has not been rigorously studied in PSC, a disease without known therapy to prevent progression, we suggest that diet may play a critical role. It is known that immunosuppressive therapy can help control symptoms, but the effect on the intestinal microbiome from immunosuppression differs from the changes resulting from dietary therapies. Whereas immunosuppression may help control IBD-associated enteral inflammation, it may not address underlying triggers of pathogenesis such as the microbiota, dietary exposures, and the resultant metabolites produced. This case is the first to report clinical remission and laboratory normalization of PSC with dietary therapy. Though IBD activity may be a confounder, we suggest that both control of IBD activity

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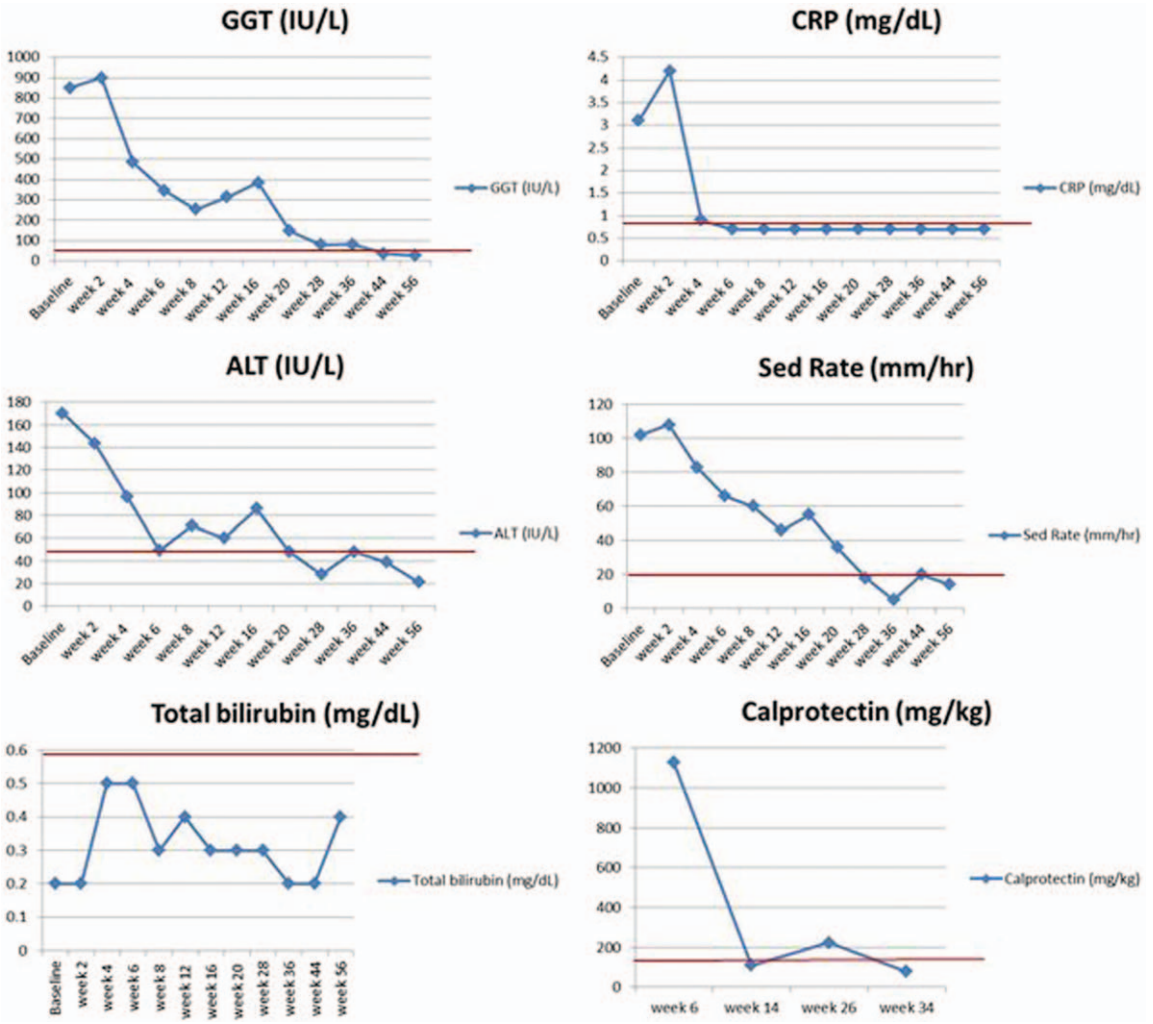

FIGURE 1. Laboratory evaluation at diagnosis and after initiation of dietary therapy.

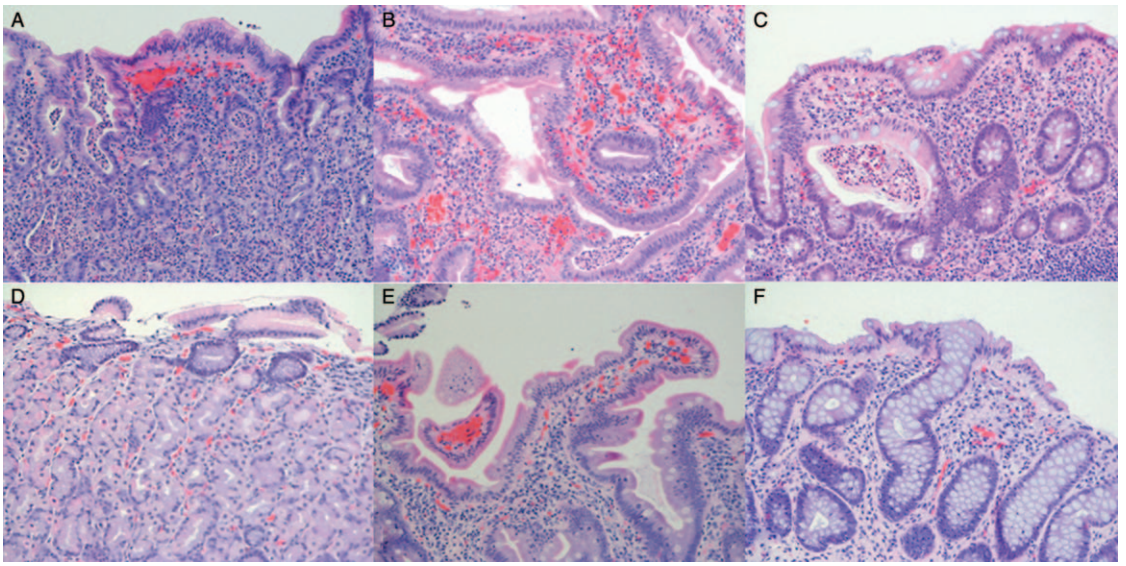

FIGURE 2. All images taken at 200× magnification. Biopsies taken on presentation: (A) Gastric biopsy with moderate active gastritis; (B) duodenal biopsy with chronic, active duodenitis; and (C) colonic biopsy with moderate chronic, active colitis. Biopsies taken after therapy: (D) gastric biopsy with no pathologic diagnosis; (E) duodenal biopsy with no pathologic diagnosis; and (F) colonic biopsy with mild chronic colitis.

TABLE 1. Laboratory evaluation of liver disease and at 6 months after diagnosis

	Labs at diagnosis	Follow-up labs (6 months later)
Prothrombin time (seconds)	13.8	
INR	1.1	
Immunoglobulin G level (mg/dL)	1810 (H)	1563
IgG total (mg/dL)	2110 (H)	1644
IgG subclass 1 (mg/dL)	1390 (H)	1151 (H)
IgG subclass 2 (mg/dL)	338	294
IgG subclass 3 (mg/dL)	151 (H)	111
IgG subclass 4 (mg/dL)	54.8	58.3
Saccharomyces cerevisiae Ab, IgA (U)	11.5	
Saccharomyces cerevisiae Ab, IgG (U)	<10.0	
Neutrophil specific antibodies	Positive	Negative
Antismooth muscle antibody	1:20	
Liver kidney microsomal antibody	Negative	
Antimitochondrial antibody	Negative	
CA 19–9 (U/mL)	906 (H)	13

and alterations in the intestinal microbiota may be within the pathway of effective therapy for PSC. Further prospective studies are merited and will help elucidate the role of diet in either primary or adjunctive therapy for PSC.

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