



## Different techniques for biliary diversion in progressive familial intrahepatic cholestasis



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### ABSTRACT

**Background:** Progressive familial intrahepatic cholestasis (PFIC) is a cholestatic liver disease of childhood. Pruritus resulting from increased bile salts in serum might not respond to medical treatment, and internal or external biliary drainage methods have been described. In this study, we aimed to evaluate different internal drainage techniques in patients with PFIC.

**Patients and methods:** Between 2009 and 2014, seven children (4 male, 3 female, 3 months–5 years old), (median 2 years of age) with PFIC were evaluated. The patients were reviewed according to age, gender, complaints, surgical technique, laboratory findings and outcome. In each two patients, cholecystoileocolonic anastomosis, cholecystojejunocolonic anastomosis and cholecystocolostomy were performed. Cholecysto-appendico-colonic anastomosis was the technique used in one patient.

**Results:** Jaundice and excessive pruritus were the main complaints. One of the patients with cholecystoileocolonic anastomosis died of comorbid pathologies (cirrhosis, adhesive obstruction and severe sepsis). Temporary rectal bleeding was observed in all the patients postoperatively. Regardless of the surgical technique, pruritus was dramatically decreased in all the patients in the postoperative period.

**Conclusion:** Regardless of the technique, internal biliary diversion methods are beneficial for the relief of pruritus in PFIC patients. Selection of the surgical method might vary depending on the surgeon's preference and the surgical anatomy of the gastrointestinal system of the patient.

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Progressive familial intrahepatic cholestasis (PFIC) is an autosomal recessive liver disease characterized by intermittent attacks of cholestasis, which could start at any age and progress to hepatic failure [1]. Pruritus is a frequent symptom in cholestasis. It develops as a result of the increase of many substances in serum induced by failed bile excretion. Pruritus might not respond to medical treatment [2]. Surgical biliary diversion methods with internal or external drainage might be useful for resolving pruritus. Biliary drainage has been reported to delay the progressive course of the disease in many patients [3–5]. Biliary diversion might save time and even be therapeutic in patients for whom liver transplantation is necessary. In this study, we evaluated four internal drainage methods and their results in seven patients with PFIC.

### 1. Patients and methods

Between 2009 and 2014, seven PFIC patients (4 male, 3 female) from 3 months to 5 years of age (median 2 years) were evaluated. The patients were reviewed in terms of age, gender, complaints, the surgical technique and laboratory findings. All the patients received medical treatment consisting of ursodeoxycholic acid, a second-generation

antihistamine and cholestyramine. In cases with grade 4 pruritus (severe pruritus with epidermal bleeding), medical treatment was determined to have failed, and these patients underwent surgery. The patients were operated on by three different surgeons, who had similar experience performing biliary diversion surgery.

Of the three surgeons who operated on the seven children included in this report, the first surgeon performed one cholecystojejunocolonic (CCJC), one cholecysto-appendico-colonic (CCAC) and one cholecystoileocolonic (CCIC) anastomosis. Another surgeon performed one CCJC, one cholecystocolostomy (CCCT) and one CCIC anastomosis. The last surgeon performed one CCCT. Pruritus was assessed from 0 to 4, according to the Whitington and Whitington scale [6]. ALT (alanine aminotransferase enzyme), AST (aspartate transaminase enzyme), GGT (gamma glutamyl transferase enzyme), ALP (alkaline phosphatase) and direct-indirect bilirubin levels were evaluated preoperatively and postoperatively. A CCIC anastomosis was performed in two patients. For CCIC anastomosis, a 10-cm long isoperistaltic ileal segment was divided from the ileum and placed behind the colon transmesocolically; the proximal end was anastomosed to the gallbladder, whereas the distal end was connected to the transverse colon as an end-to-side anastomosis (Fig. 1). A CCAC anastomosis was performed in one patient, and a CCJC anastomosis was performed in two patients. For the CCJC, a 10 cm long isoperistaltic jejunal segment was divided distal to 40 cm

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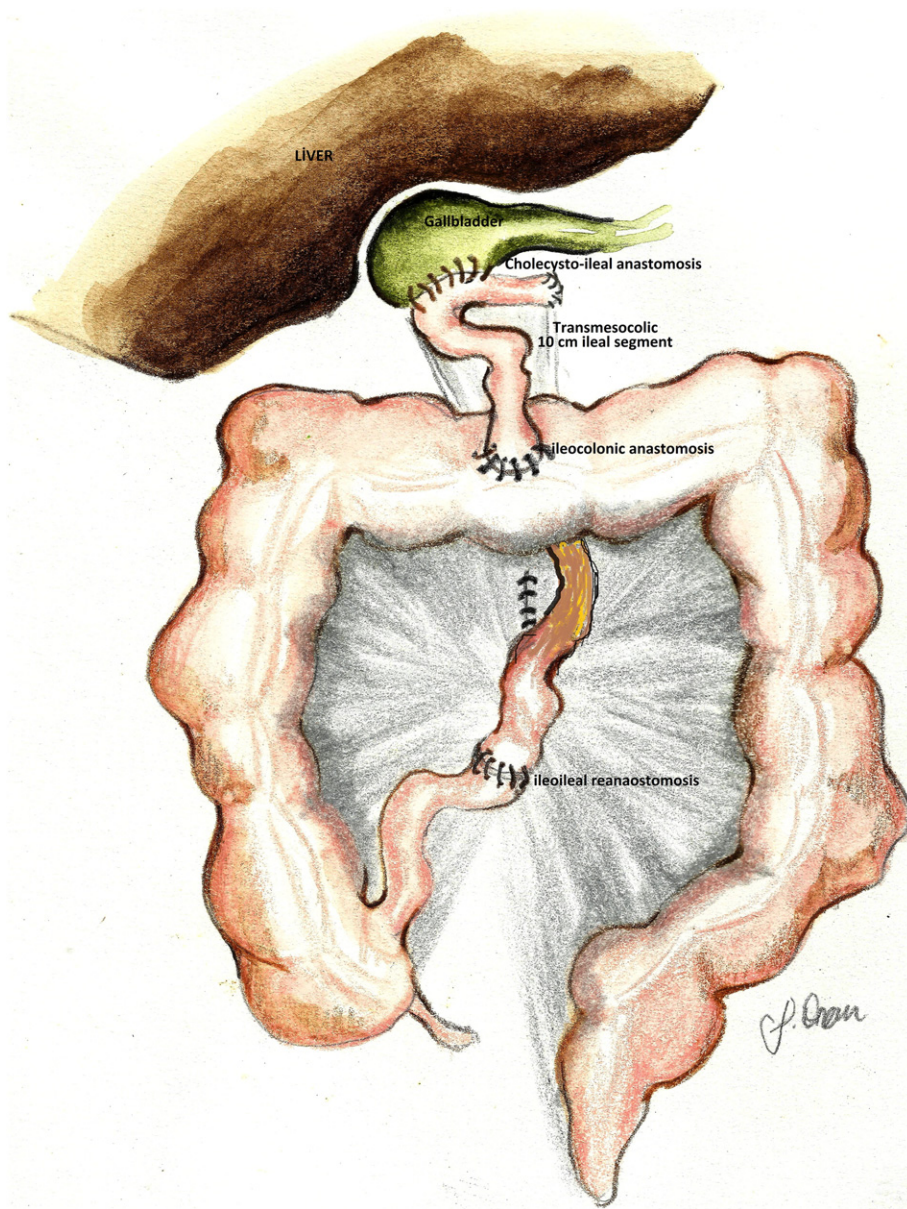


Fig. 1. Cholecystoileocolonic anastomosis.

from the ligament of Treitz. The proximal end was anastomosed to the gallbladder in an end-to-end fashion whereas the distal end was connected to the transverse colon as an end-to-side anastomosis. For the CCAC anastomosis, the appendix base was separated from the cecum, and the appendicular vessels were protected. A 1.5 cm-long incision was made on the gallbladder. The distal tip of the appendix was anastomosed to the gallbladder. The proximal tip of the appendix was anastomosed to the transverse colon in an end-to-side fashion. To protect the appendico-colonic anastomosis, a 1.5 cm long subserosal tunnel was created on the tenia of the colon (Fig. 2). In two patients, anastomosis was performed with the CCCT technique. The colon was separated from the splenic flexura, its mesentery was freed and the 10 cm-long colon segment was anastomosed to the gallbladder as a Roux-en-Y bypass. Colocolic anastomosis was performed on the antimesenteric side of the proximal colon in an end-to-side fashion, and thus the continuity of the colon was ensured (Fig. 3). The protocol for the postoperative follow-up consisted of evaluation of bilirubin and liver enzymes for the first month after surgery and then every three months.

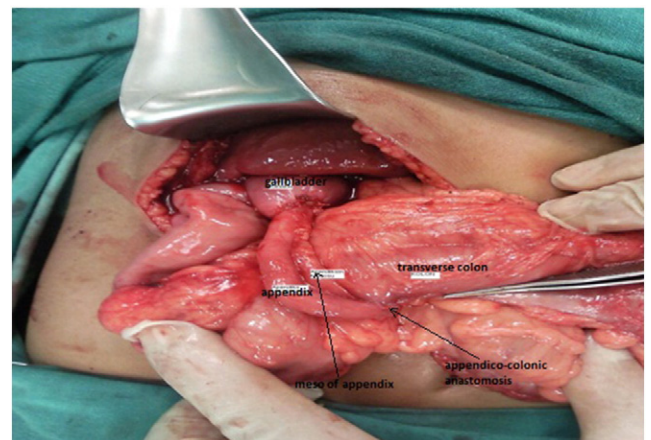


Fig. 2. Cholecysto-appendico-colonic anastomosis.

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