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Case Report

# Ileal Bypass for Pruritus Relief in a 3-Year-Old Boy with Advanced Progressive Familial Intrahepatic Cholestasis: How Effective Is It?

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## Abstract

Progressive familial intrahepatic cholestasis (PFIC) is a group of liver disorders that manifest in early childhood with cholestasis and pruritus resulting progressively in liver failure. We present a case of a 3-year-old boy with advanced PFIC from refractory pruritus. In order to offer an effective treatment of pruritus, our patient underwent ileal bypass and after a 2-month period free of symptoms, unexpectedly relapsed after a Rota viral infection. Finally, the child underwent orthotopic liver transplantation. Patients with advanced PFIC do not seem to benefit from nontransplant invasive interventions regarding the relief of pruritus.

## Keywords

children, cholestasis, ileal bypass, pruritus

## INTRODUCTION

PFIC is a group of liver disorders inherited in an autosomal dominant pattern which become manifest in early childhood with cholestasis, pruritus, increased serum bile acids, and usually normal gamma-glutamyl transpeptidase, resulting progressively in liver fibrosis, cirrhosis and failure.[1] The current management of PFIC includes medical treatment with ursodeoxycholic acid and multiple anti-pruritic drugs and endoscopic or surgical interventions, such as NBD, partial external biliary diversion (PEBD) and ileal bypass (IB).[2-7] In cases when medical and surgical treatment fails, the patients undergo liver transplantation.

## CASE REPORT

*A 15-month-old previously healthy boy presented with jaundice, pruritus, passage of pale-colored feces, and dark urine. The physical examination revealed yellowish pigmentation of the skin and sclera of eye, multiple scratches on his face and trunk and hepatomegaly. Blood biochemistry tests revealed direct hyperbilirubinemia with serum bilirubin 4.53 mg/dl, aspartate transaminase 114 IU/L, alanine aminotransferase 123 IU/L, alkaline phosphatase 521 U/L, while gamma-glutamyl transpeptidase was normal. The abdominal ultrasound examination showed no pathological findings.*

*Furthermore, he underwent extensive laboratory metabolic, serological, and immunological tests, which were within normal limits. Moreover, alfa 1 antitrypsin levels and phenotype were normal. The concentration of bile acids was very high (326.5 μmol/L). The boy underwent a liver elastography, which revealed severe fibrosis (F2-3 METAVIR), and liver biopsy, which demonstrated intrahepatic cholestasis and minimal portal and peri-centrivenular fibrosis. Liver molecular genetics test detected MYO5B variants associated with autosomal microvillus inclusion disease and cholestasis. Finally, the biopsy of small intestine did not reveal any abnormal findings.*

*Subsequent to appropriate medical evaluation of the patient's clinical presentation and the results of the laboratory, radiological, pathological, and genetic tests the child was considered to have progressive familial intrahepatic cholestasis (PFIC).*

*The initial treatment included administration of ursodeoxycholic acid, phenobarbital, medium-chain triglyceride-enriched formula, and fat-soluble vitamins. During a follow-up period of 1 year, the patient recovered gradually from the jaundice, liver function tests became normal and the concentration of bile acids fell significantly (23.86 μmol/L). On the other hand, pruritus did not improve and the additional prescription of rifampicin, ondasentron, naltrexone, and prednilozone was ineffective. As a result, the patient suffered from severe itching that interfered with daytime and nighttime activities with abrasions.*

*As the medical therapy was unsuccessful regarding the pruritus relief, endoscopic nasobiliary drainage (NBD) was attempted and a 6-F nasobiliary drain was placed in the common bile duct after sphincterotomy. Unfortunately, after 18 hours the catheter was pulled out accidentally and this method was rejected because of low compliance of the parents with the medical instructions about the care of catheter. After 2 months, the child underwent ileal bypass (IB). More specifically, 45 cm of the terminal ileum were excluded and an ileo-colic anastomosis in a distance of 5 cm from the ileocecal valve was created. There were no post-operative complications and the pruritus improved rapidly.*

*The child was free of symptoms, but after 2 months, when he was re-admitted to the hospital because of a Rota viral infection, pruritus relapsed and the serum bile concentration began to rise again. After two years, the boy underwent orthotopic liver transplantation; his symptoms resolved, and he is now being closely followed up by the team at the transplantation center.*

## DISCUSSION

For patients with persistent pruritus despite medical therapy, invasive treatment options have been suggested, aiming to the reduction of the hepatic and systemic concentration of bile acids. NBD has been proven effective, but in our case it failed, as the child was too young and the parents were not careful regarding the care of catheter.[4,8] PEBD is another treatment option, which improves pruritus by interrupting the enterohepatic circulation and reducing the bile acid pool, but it has carries the disadvantage of a permanent stoma and its care.[5,6,9,10] In our case, we rejected this method to avoid causing damage in the liver anatomic region because of the high possibility of future transplantation.

In order to offer an effective treatment of pruritus, our patient underwent IB, as it is safe, with low post-operative complication rate, encouraging results and inhibits the intestinal re-absorption of bile acids, without the creation of external biliary fistula.[7,10] After the procedure, our patient was asymptomatic, but then unexpectedly relapsed after a Rota viral infection. As result, pruritus recurred, indicating IB procedure failed to eliminate bile acids, probably because of the gradual adaptation of the remaining ileum or the advanced disease of the child, but it is remarkable the sudden and unexplained onset of pruritus after the treatment of gastroenteritis.

Patients with mild PFIC seem to benefit from nontransplant invasive interventions regarding the relief of pruritus.[10-12] In our case, the child with advanced PFIC did not respond to surgical treatment with IB and was transplanted. Perhaps, earlier in the course of the disease, a bypass of a different length of intestine could lead to a better outcome and prevent or delay the liver transplantation.

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## Competing interests

The authors have declared that no competing interests exist.

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# Шунтирование подвздошной кишки для облегчения зуда у 3-летнего мальчика с прогрессирующим семейным внутрипечёночным холестазом: насколько оно эффективно?

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## Резюме

Прогрессирующий семейный внутрипечёночный холестаз (ПСВХ) представляет собой группу заболеваний печени, которые проявляются в раннем детстве холестазом и зудом, приводящими к прогрессирующей печёночной недостаточности. Мы представляем случай 3-летнего мальчика с прогрессирующим ПСВХ из-за рефрактерного зуда. Чтобы предложить эффективное лечение зуда, нашему пациенту было проведено шунтирование подвздошной кишки, и после 2-месячного бессимптомного периода у него неожиданно развился рецидив ротавирусной инфекции. Наконец, ребёнку была выполнена ортотопическая трансплантация печени. Пациенты с прогрессирующим ПСВХ, по-видимому, не получают пользы от нетрансплантационных инвазивных вмешательств в отношении облегчения зуда.

## Ключевые сова

дети, холестаз, илеошунт, зуд