



Management of acute cholangitis in a 3-year-old with hereditary spherocytosis: challenges of percutaneous intervention following failed ERCP

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Abstract

Introduction. Hereditary spherocytosis (HS) is a congenital haemolytic anaemia caused by a defect in the red blood cell membrane proteins, leading to chronic haemolysis in the spleen. The excessive production of bilirubin during haemolysis significantly increases the risk of developing pigment gallstones. In paediatric patients, these stones can lead to complications such as cholecystitis, acute cholangitis, or obstructive jaundice. Standard management involves symptom control, with major surgeries like cholecystectomy and splenectomy typically delayed until the child reaches five years of age to minimize infection risks.

Case report. A 3-year-old female patient was admitted with HS and diagnosed with biliary obstruction. Due to the patient's small paediatric anatomy, specifically the narrow major duodenal papilla (typically 2-5 mm in this age group), a standard Endoscopic Retrograde Cholangiopancreatography (ERCP) was technically unfeasible. From a methodological standpoint, a percutaneous transhepatic biliary drainage was selected and justified as the safest minimally invasive alternative for urgent biliary decompression. The patient's condition was complicated by the development of pancreatitis and later acute cholangitis caused by the bacterium *Enterobacter ludwigii*. After stabilization with specific antibiotic therapy (ceftazidime and metronidazole), the patient successfully underwent a laparoscopic cholecystectomy. The cholangiostoma was eventually removed, and the planned splenectomy was postponed until the patient reaches five years of age to minimize the risk of Overwhelming Post-Splenectomy Infection (OPSI).

Conclusion. This case highlights that HS is a major risk factor for early-onset gallstones and acute cholangitis in children. When ERCP is unfeasible due to age and anatomical constraints, percutaneous transhepatic drainage serves as a safe and effective alternative. Successful management relies on targeted antibiotic therapy, timely surgical intervention and multidisciplinary collaboration.

Keywords: hereditary spherocytosis, haemolysis, cholelithiasis, acute cholangitis, percutaneous transhepatic cholangiostoma, laparoscopic cholecystectomy, paediatric splenectomy.

1. Introduction

Hereditary spherocytosis is a congenital haemolytic anaemia caused by a defect in the red blood cell membrane protein: these erythrocytes become sphere-shaped and are destroyed in the spleen (1). This condition, due to the excessive production of bilirubin during haemolysis, significantly increases patients' predisposition to both gallbladder and bile duct stone disease (cholelithiasis). Gallstones in patients with hereditary spherocytosis, which are mostly composed of bilirubin, can cause complications such as cholecystitis, cholangitis, and obstructive jaundice (1). The incidence of biliary stone disease in children has increased significantly in recent decades, paralleling trends in obesity and metabolic syndrome. Studies have estimated a prevalence between 1.9 % and 4 % (2). Retrospective cholecystography studies with patients aged 1.4 to 22 years with hereditary spherocytosis (HS) have reported gallstones in 37-43% of cases (3).

Treatment for cholelithiasis in patients with hereditary spherocytosis is tailored to the severity of symptoms and the extent of haemolysis. Splenectomy is frequently the primary treatment method, as it significantly reduces haemolysis and bilirubin production, thereby indirectly lowering the risk of gallstone formation. However, splenectomy increases the risk of infections and, in certain cases, can lead to severe sepsis. It is more commonly known as Overwhelming Post-Splenectomy Infection (OPSI). This complication despite intensive treatment carries high mortality rate of up to 50%. To decrease this risk, strategies include partial splenectomy and delaying the surgery until five years of age (4,5).

Recent research indicates that even after a splenectomy, persistent haemolysis may still contribute to the formation of gallstones. Therefore, it

is essential to monitor patient's post-splenectomy (1). Prophylactic cholecystectomy is considered a preventive method for patients with significant hyperbilirubinemia, even in the absence of gallstone disease, especially in those with hereditary spherocytosis (6).

This article presents the case of a 3-year-old girl with hereditary spherocytosis who was treated for biliary duct obstruction. The case illustrates the complications of this rare genetic disease, specifically acute cholangitis and pancreatitis, and highlights the unique diagnostic and treatment challenges. These challenges required the collaboration of a multidisciplinary team (paediatric surgeons, radiologists, gastroenterologists, and onco-haematologists) to plan and monitor the patient's follow-up care

2. Case report

A 3-year-old patient with a congenital diagnosis of hereditary spherocytosis was first hospitalized in the paediatric surgery department for an initial six-week period. On the second day of hospitalization, an attempt was made to perform retrograde cholangiopancreatography (ERCP) to address the biliary obstruction. However, the procedure proved technically unfeasible due to the patient's age and the specific anatomical constraints inherent in a three-year-old child. The patient was subsequently started on a course of Unasyn. The following day, a transhepatic bile duct clearance was performed, which was followed by the development of pancreatitis as a complication. Ten days later, patient underwent percutaneous transhepatic cholangiography with the placement of a 6 Fr drain (Image 1). Following a cholangioscopy two weeks after the initial drainage placement drain was replaced with a larger 8.5 Fr cholangiostoma. Two days later a multidisciplinary

consultation, identified the management of the patient's pancreatitis as a clinical priority. The management plan specified that a minimally invasive intervention for bile duct clearance and a subsequent cholecystectomy would be performed only after the patient's condition stabilized. Furthermore, it was recommended that the splenectomy be deferred until the patient reaches approximately five years of age to decrease the risk of overwhelming post-splenectomy infection (OPSI). A follow-up cholangiography performed one day before discharge, revealed that all biliary stones and sludge had spontaneously migrated into the intestinal tract. The patient was subsequently discharged with the cholangiostoma in situ, and elective surgery was scheduled for early January.

However, twelve days after discharge due to a worsened clinical status, the patient was readmitted to the Department of Paediatric Surgery at the LSMU Kaunas Clinics, presenting with episodes of paroxysmal abdominal pain. According to the mother, the patient experienced two acute pain episodes requiring analgesic intervention. Objective clinical findings included a low-grade fever of up to 37.5°C and a visible intensification of jaundice in both the sclera and skin. Laboratory investigations confirmed a severe inflammatory response and significant cholestasis. C-reactive protein (CRP) was markedly elevated at 199.5 mg/l, and the white blood cell count (WBC) reached $26.43 \times 10^9/l$, indicating acute systemic infection. Despite the clinical jaundice, liver transaminases (ALT 32 IU/l, AST 45 IU/l) remained within normal ranges, while total bilirubin was severely increased to 126.48 $\mu\text{mol/l}$ (Table 1). The patient was hospitalized following a diagnosis of acute cholangitis. An emergency abdominal ultrasound revealed a completely contracted gallbladder containing sludge and several small calculi.

Additionally, the spleen was enlarged for her age, measuring approximately 10.0 cm. Bile cultures afterwards yielded *Enterobacter ludwigii*, prompting the initiation of intravenous ceftazidime and metronidazole. A follow-up ultrasound on the fifth day of the second hospitalization visualized a biliary stent extending into the duodenum. A laparoscopic cholecystectomy was successfully performed the next day. Postoperatively, a subhepatic drain and the cholangiostoma were left in place. One week after the surgery, a control cholangiography confirmed that the contrast medium flowed easily into the intestinal tract. Consequently, the cholangiostoma was removed, and the drainage tract within the hepatic parenchyma was sealed using Embocube particles. A final follow-up ultrasound showed that the biliary tree was not dilated. Eight days after the surgery, the patient was discharged in stable condition (Table 2). Her discharge recommendations included continued folic acid therapy until the scheduled splenectomy and adherence to a specialized post-cholecystectomy dietary plan.

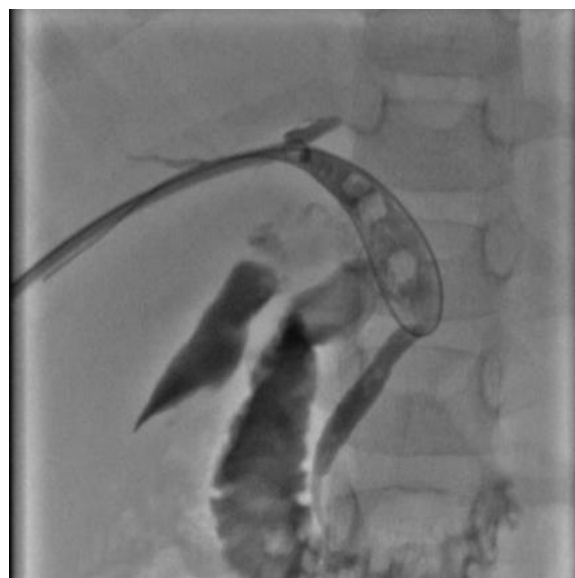


Image 1 Percutaneous Transhepatic Cholangiogram

Table 1 Laboratory Data Dynamics: Acute Phase vs. Discharge

Parameter	Reference Range	On Admission	At Discharge
C-reactive protein (CRP)	0 – 5 mg/l	199.5 mg/l	31.8 mg/l
White Blood Cell (WBC)	4.92 – 11.8 x 10 ⁹ /l	26.43 x 10 ⁹ /l	9.74 x 10 ⁹ /l
Total Bilirubin	1.7 – 15.4 µmol/l	126.48 µmol/l	75.86 µmol/l
Hemoglobin (Hb)	111 – 141 g/l	89 g/l	94 g/l
ALT (GPT)	7 – 45 IU/l	32 IU/l	140 IU/l
AST (GOT)	8 – 50 IU/l	45 IU/l	130 IU/l
MCV	72.1 – 87 fl	93.3 fl	87.5 fl

Table 2 Chronological Timeline of the Case

Date	Event / Stage	Description
2022-09-21	Birth and Diagnosis	Patient diagnosed with hereditary spherocytosis.
2025-11-04	First Hospitalization	Admitted due to bile duct stone disease with obstruction.
2025-11-06	Failed Intervention	Attempted ERCP failed due to the patient's small/fine anatomy; initiated antibiotic treatment with Unasyn.
2025-11-07	Complication	Transhepatic bile duct clearance performed; followed by postoperative pancreatitis.
2025-11-17	Drainage	Percutaneous transhepatic cholangiography performed and a 6 Fr drain was inserted.
2025-12-03	Multidisciplinary Decision	8.5 Fr cholangiostomy inserted. Medical board decided to treat pancreatitis first and postpone surgery.
2025-12-18	Clinical Improvement	Follow-up cholangiography showed all stones and sludge had spontaneously flushed into the intestine.
2025-12-19	Discharge	Patient discharged with a functioning 8.5 Fr cholangiostomy; elective surgery scheduled for early January.
2026-01-01	Emergency Readmission	Onset of abdominal pain, fever (up to 37.5°C), and jaundice. Diagnosed with acute cholangitis.
2026-01-02	Infection Identification	Bile culture grew <i>Enterobacter ludwigii</i> ; targeted IV antibiotic therapy with Ceftazidime and Metronidazole initiated.
2026-01-07	Surgical Treatment	Laparoscopic cholecystectomy performed.
2026-01-14	Drain Removal	Follow-up cholangiography confirmed free flow to the intestine; cholangiostomy removed and the tract sealed with "Embocube" particles.
2026-01-15	Final Discharge	Patient discharged in stable condition; continued folic acid treatment and planned splenectomy at age five.

3. Discussion

This case corroborates documented findings in medical literature regarding the increased risk of early onset cholelithiasis and choledocholithiasis in patients with hereditary spherocytosis. Even in early childhood

due to chronic haemolysis, excessive bilirubin production leads to the formation of pigment stones, which can cause severe complications such as biliary obstruction and acute cholangitis. In this instance, the standard approach for managing bile duct

obstruction—Endoscopic Retrograde Cholangiopancreatography (ERCP)—was technically unfeasible due to the patient's paediatric anatomy. Gastrosopes that are specifically utilized for ERCP, specialized pediatric slim duodenoscopes ('Olympus PJF'-7.5, 7.5–8.5 mm) and ultra-slim gastrosopes ('ELUXEO EG-740N'), were incompatible with the narrow major duodenal papilla. Anatomical data indicates that in female patients of this age group, the major papilla typically measures only 2–5 mm in diameter (7). When endoscopic access fails, as a safe alternative, percutaneous transhepatic biliary (PTBD) drainage is performed. The PTBD procedure relies on using 22G Chiba needles and 5-8 Fr catheters. PTBD is rarely performed in children aged 3 years and younger outside specialized contexts like liver transplantation or neonatal cholestasis, with no population-level incidence data due to its infrequent use in high-risk cases only (8). However, PTBD provides effective biliary decompression with technical success (89–96%) and low complication rates (minor 10.8%, major 1.7%) in pediatric series (8). This case highlights the vital necessity of alternative safest minimally invasive interventions, such as percutaneous transhepatic cholangiography and drainage, as safe and effective management strategies.

Paediatric acute cholangitis due to *Enterobacter ludwigii* is a rare clinical finding. Reports of this specific opportunistic pathogen in children remain infrequent. In paediatric cholangitis, more typical organisms are usually Enterobacterales such as *E. coli*, *Klebsiella*, and *Enterobacter* spp (9). Therefore, this appears to be an unusual ethology in our patient necessitating targeted antibacterial therapy with ceftazidime and metronidazole. Additionally, biliary duct drainage via a cholangiostoma was required until

the primary focus of infection in the gallbladder could be definitively eliminated.

The acute conditions of cholangitis and pancreatitis demanded urgent collaboration from a multidisciplinary team—including paediatric surgeons, radiologists, gastroenterologists, and onco-haematologists—to achieve rapid clinical stabilization. This multidisciplinary approach was also essential for planning the subsequent cholecystectomy and scheduling the splenectomy to be postponed until the patient reaches five years of age.

Splenectomy was deferred until age 5 to reduce the risk of overwhelming post-splenectomy infection, since OPSI is more common in young children and many recommendations advise postponing elective splenectomy until after early childhood when feasible(10).

4. Conclusion

1. Hereditary spherocytosis is a significant risk factor for the development of choledocholithiasis and acute cholangitis in paediatric patients.

2. When Endoscopic Retrograde Cholangiopancreatography is unsuccessful due to the patient's age and specific anatomical constraints, percutaneous transhepatic drainage serves as a safe and effective alternative.

3. Successful management of acute cholangitis relies on targeted, specific antibiotic therapy and timely surgical intervention.

4. A multidisciplinary team approach is essential for managing complex cases of paediatric biliary obstruction.

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