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Case Report

Endoscopic Retrograde Cholangiopancreatography with an Ultra-Slim Forward-Viewing Gastroscope in a 3.8 kg Infant: A Case-Report

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Abstract:

Background: Experience with pediatric endoscopic retrograde cholangiopancreatography (ERCP), particularly in infants, is limited. The lower incidence of biliopancreatic disorders in infants and the discontinuation of pediatric duodenoscopes raise concerns regarding ERCP's technical efficacy and safety in this vulnerable population, for whom conventional duodenoscopes are not recommended.

***Case*:** We report the case of a low-weight infant with short bowel syndrome under parenteral nutrition, who was referred for ERCP due to choledocholithiasis with progressive jaundice. We opted for an ultra-slim forward viewing gastroscope (5.4-mm distal end outer diameter and 2.2-mm working channel) with a needle knife sphincterotome (1.8-mm outer sheath diameter). This allowed sphincterotomy with adequate gallstone removal and complete normalization of cholestasis parameters without associated complications.

***Discussion and conclusion*:** The decreasing availability of pediatric duodenoscopes challenges the important role of ERCP as a diagnostic and therapeutic tool in infants, making their management a challenge. With this case we aimed to describe a previously unreported approach to a low-weight infant with gallstone disease requiring ERCP, which proved to be safe and effective.

Keywords: children, choledocholithiasis, ERCP, intestinal failure-associated liver disease, pediatric

Introduction

Endoscopic retrograde cholangiopancreatography (ERCP) has a well-recognized role in the treatment of biliopancreatic diseases in children[1]. However, experience with ERCP in children, particularly in infants, is limited[2,3]. The relatively lower incidence of biliary and pancreatic disorders in the pediatric population, improvements in magnetic resonance cholangiopancreatography in diagnosis and concerns regarding ERCP's safety and outcomes in children have challenged its widespread use in pediatrics. Additionally, pediatric duodenoscopes stopped being manufactured in 2014, so most of the duodenoscopes used are the ones designed for adults[4]. This lack of appropriately sized duodenoscopes and accessories makes ERCP technically more difficult to perform, particularly in small children. We report a case of a 3.8 kg infant who underwent ERCP with an ultra-slim forward-viewing gastroscope with favorable outcomes.

Case

A 6-month-old female infant weighing 3.8 kg was referred to our department for ERCP. She was born at 25 weeks of gestational age (extreme preterm), to a G1P0 33-year-old mother who had an unmonitored pregnancy. The mother was admitted in spontaneous labor and an emergent cesarian section was performed due to fetus in breech position. There was no time for fetal lung maturation. The baby was born with 636 g, with an Apgar score 4/5/5, and was immediately admitted to the neonatal intensive care unit.

The newborn presented with neonatal distress syndrome requiring invasive mechanical ventilation and hemodynamically instability requiring fluid replacement and vasoactive amines. Surfactant was administrated on the first and third days and empirical prophylactic antibiotic treatment with ampicillin and gentamicin was given since day one. She was also jaundiced (with a maximum value of total bilirubin of 7.59 mg/dL on the third day due its unconjugated fraction) for which she did phototherapy for 2 weeks, with improvement of total bilirubin values down to 2.10 mg/dL. Due to delayed sepsis with blood cultures on the 8th day positive for *Staphylococcus haemolyticus* and *Staphylococcus epidermidis*, she was treated according to antibiotic susceptibility testing with piperacillin-tazobactam, vancomycin and amikacin.

By the end of the first month of age, she developed septic shock with necrotizing enterocolitis complicated with perforation and pneumoperitoneum. She was transferred with 32 days to our center for surgical intervention. Laparotomy showed multiple perforations in the ileum and ascending colon, the first one 15-cm from the angle of Treitz. She underwent a large resection of the ileum, ileocecal valve, and ascending colon, being left with a jejunostomy which was later reconstructed as a jejuno-colic anastomosis. The infant was under parenteral nutrition (PN) since her

birth, which was maintained due to her short bowel syndrome (45-cm of remaining bowel).

At three months of age, she developed cholestasis, considered related to intestinal failure-associated liver disease (IFALD). Despite multiple PN adjustments, laboratory values were progressively worsening, reaching total/direct bilirubin values up to 15.39/10.70 mg/dL by six months of age (**Table 1**). On abdominal ultrasound, there was sludge in the gallbladder, intrahepatic and extrahepatic bile duct dilation and choledocholithiasis.

Table 1. Changes in laboratory results before and three months after treatment.

Laboratory parameters (units)	Normal range	Day before ERCP	3 months after ERCP
Total bilirubin (mg/dL)	0.20-1.20	15.39	0.51
Direct bilirubin (mg/dL)	0.05-0.30	10.70	0.26
Aspartate aminotransferase (IU/L)	20-67	138	34
Alanine aminotransferase (IU/L)	5-33	90	25
Gamma-glutamyl transferase (IU/L)	8-127	46	23
Alkaline phosphatase (IU/L)	134-518	691	349
International normalized ratio	0.96-1.04	1.68	1.36

ERCP: Endoscopic retrograde cholangiopancreatography

She underwent an ERCP under general anesthesia in the operating room by an experienced endoscopist trained in adult ERCP, who routinely performs more than 250 ERCPs per year. We used an ultra-slim forward viewing gastroscope with a 5.4-mm distal end outer diameter and 2.2-mm working channel (GIF-H190N, *Olympus* ®) with a needle knife sphincterotome (1.8-mm outer sheath diameter, *Endotec* ®).

Upper endoscopy was unremarkable, except for a protruded duodenal papilla. Sphincterotomy led to the spontaneous extrusion of a 5-mm stone (**Figure 1**), followed by abundant drainage of biliary sludge. Fluoroscopy showed slightly dilated intrahepatic and extrahepatic bile ducts on opacification, without filling defects. The gallbladder was dilated, with multiple filling defects. By the end of the procedure, the bile ducts were clear, with adequate drainage. There were no procedure or anesthesia-related complications.

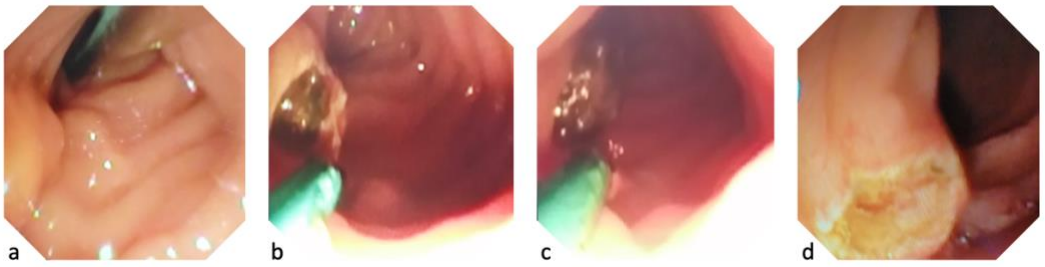

Figure 1. Upper endoscopy using an ultra-slim forward-viewing 5.5-mm gastroscope. **a.** Protruded duodenal papilla. **b.** Needle-knife sphincterotomy showing an impacted bile duct stone. **c.** Gallstone removal. **d.** Duodenal papilla after sphincterotomy.

In the following weeks, multiple PN adjustments were done, and she had a gradual decline in the cholestasis parameters, up to their complete normalization after three months (**Table 1**).

Discussion

Cholestasis is a significant complication in children on PN, with an overall incidence estimated in 29.9%[5]. Premature infants and children with intestinal failure or short bowel syndrome are susceptible to IFALD, characterized by progressive cholestasis, liver fibrosis, biliary cirrhosis, portal hypertension and cholelithiasis[6,7]. The pathogenesis of IFALD is complex and related to many factors, including prematurity, low birth weight, enzyme deficiencies, genetic alterations, anatomic features, and PN-related factors such as its composition[5,7]. PN was encountered as a risk factor in 10-17.6% children with gallstones[7], with greater risk if started at a younger age, without enteral feeding, and in cases of motility disorders with a stoma[8].

Gallbladder and biliary disease remain the most frequent indication for pediatric ERCPs, corresponding to 48% of the ERCPs performed in a US nationwide assay from 2005-2014[2], which included 11,060 patients under 20 years of age. However, only 5% of them were under the age of four.

In fact, there are few guidelines addressing the indications and optimal utilization of therapeutic ERCP in infants and neonates. The European Society of Gastrointestinal Endoscopy (ESGE) and European Society for Pediatric Gastroenterology, Hepatology and Nutrition (ESPGHAN) guidelines on endoscopy in children[1] recommend that therapeutic ERCP in pediatric patients (>1 year old) should be considered for common bile duct stones, bile leaks, benign/malignant biliary strictures, primary sclerosing cholangitis and parasitosis, and that diagnostic ERCP can be used in neonates and infants with cholestatic hepatobiliary disease in the diagnosis of biliary atresia. In children above 10 kg, therapeutic duodenoscopes are considered safe, but in neonates, infants and children weighing less than 10 kg, a

pediatric 7.5-mm duodenoscope with a 2-mm working channel is recommended, thus limiting the instruments and accessories that can be used with it.

In a retrospective study on 856 ERCPs performed in 626 pediatric patients between 1999-2018[3], 219 were under 1 year of age, 5% of whom with choledocholithiasis. In these patients, a pediatric duodenoscope was used and a 5-Fr biliary stent inserted without papilla sphincterotomy. They had conservative treatment with ursodeoxycholic acid up to 1-year of age, when the stent was removed and sphincterotomy done, with extraction of the choledocholithiasis.

Furthermore, since 2014, the only existing pediatric 7.5-mm duodenoscopes (PJF 7.5, PJF 160 and PJF 240 duodenoscopes by *Olympus* ®) stopped being supported for maintenance by the manufacturer and no new pediatric duodenoscopes have been produced, contributing to the disappearance of ERCP facilities in certain centers[4] and turning this procedure a real challenge in infants in whom adult duodenoscopes cannot be used, like in our case, making it necessary to consider different options in their management.

This decreasing availability of pediatric duodenoscopes and facilities is an unwanted development, as infant ERCP is an indispensable diagnostic and therapeutic tool[4], being at risk if the production of these duodenoscopes is not restored. Efforts by the worldwide endoscopic community to drain and centralize care in high-volume centers with experience in ERCP and assure continuing production and maintenance of pediatric duodenoscopes should be done. Additionally, sharing experience with the scientific community and conducting prospective multicentric studies on ERCP in infants would be desirable, to improve the quality of infant ERCP and offer these patients the best possible care.

Conclusion

In conclusion, therapeutic ERCP is expected to be increasingly needed at an early age, considering the greater number of children surviving with conditions like short bowel syndrome who require PN and develop choledocholithiasis. The lack of specific equipment for neonates and infants can challenge the safety and technical success of the procedure, creating the need to find and adapt solutions to this vulnerable population.

We aimed to describe a possible approach to a low-weight infant with gallstone disease requiring ERCP, allowing adequate drainage of the biliary tract and complete normalization of cholestatic parameters without associated complications. To the best of our knowledge, this is the first reported case in the literature of an ERCP with sphincterotomy using an ultra-slim forward-viewing scope in an infant of such weight.

Conflict of Interest

None declared.

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Parental Consent

The grandmother (child's guardian) provided informed consent regarding the publication of this article.

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