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Percutaneous transhepatic cholecysto-cholangiography (PTCC): An alternative to intraoperative cholangiography in high risk infants suspect for biliary atresia

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1. Introduction

Persistent conjugated hyperbilirubinemia must be evaluated promptly to exclude the diagnosis of biliary atresia (BA), otherwise there is progression to cirrhosis and liver failure. Success of a portoenterostomy (PE) is time dependent, and therefore BA must be excluded as promptly as possible. There is clear evidence that improved outcomes are achieved when definitive operation is performed earlier [1,2]. Timely diagnosis is therefore imperative.

There is a dearth of literature and clinical practice to support a multifactorial workup in these infants, including infectious, metabolic and genetic laboratory evaluation, multiple imaging modalities including ultrasonography and HIDA scan; and invasive procedure like liver biopsies [3,4]. If the comprehensive work up suggests biliary atresia, then the gold standard for diagnosis of biliary atresia is intraoperative cholangiography (IOC) via laparotomy. Neonates that are critically ill or have significant co-morbidities may be high-risk surgical candidates, and thus minimizing operative time or avoiding an operation may be beneficial. An additional modality for excluding BA is percutaneous transhepatic cholecysto-cholangiography (PTCC). We present three infants undergoing PTCC by interventional radiology (IR) revealing patency of the biliary system without the need for IOC.

2. Case report

PTCC is typically performed with general mask anesthesia or general endotracheal anesthesia, if already intubated, with intravenous gram negative antibiotic coverage initiated before the procedure. Ultrasound is used to identify and cannulate the gallbladder via a percutaneous transhepatic approach with a small needle (21–25 ga). Usage of transhepatic access allows for a tamponade effect on the gallbladder puncture site. After sonographic confirmation of needle placement, iodinated contrast is gently injected under fluoroscopy to evaluate the biliary tree. Biliary atresia is excluded with opacification of both the intrahepatic biliary structures and extrahepatic flow into the duodenum.

In our case series, Patient 1 presented for evaluation outside of the immediate neonatal period. He presented after prolonged phototherapy and anesthesia because it does not require general anesthesia.

with an open approach under general anesthesia. Recent studies have demonstrated efficacy with a laparoscopic approach compared to an open approach, albeit this is still under general anesthesia. PTCC may be a safer alternative for patients who are at a high risk for open surgery and anesthesia because it does not require general anesthesia.
patients 2 and 3 were felt to be high risk for surgery. After our success with Patient 1, this procedure was attempted in higher risk patients 2 and 3; both of whom were seen in consultation in the neonatal intensive care unit.

All patient data is presented in Table 1. All three patients were felt to require cholangiography in order to exclude biliary atresia.

### Table 1

<table>
<thead>
<tr>
<th>Patient Data</th>
<th>Case 1</th>
<th>Case 2</th>
<th>Case 3</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Demographics</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Age at presentation/consultation</td>
<td>7 weeks</td>
<td>6 weeks</td>
<td>8 weeks</td>
</tr>
<tr>
<td>Gestational age (weeks)</td>
<td>38</td>
<td>31</td>
<td>32</td>
</tr>
<tr>
<td>Gender</td>
<td>Male</td>
<td>Male</td>
<td>Male</td>
</tr>
<tr>
<td>Comorbidities</td>
<td>None</td>
<td>Coarctation of the aorta, renal insufficiency, respiratory insufficiency, 2q37 deletion</td>
<td>Dehydrated hereditary stomacytosis, fetal chylous ascites, intracranial hemorrhage with hydrocephalus, restrictive lung disease</td>
</tr>
<tr>
<td><strong>Duration of conjugated hyperbilirubinemia</strong></td>
<td>5 weeks</td>
<td>4 weeks</td>
<td>8 weeks</td>
</tr>
<tr>
<td><strong>Baseline Laboratory Values</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total Bilirubin mg/dL</td>
<td>11.9</td>
<td>6</td>
<td>3.6</td>
</tr>
<tr>
<td>Conjugated Bilirubin mg/dL</td>
<td>9.1</td>
<td>5.1</td>
<td>2.9</td>
</tr>
<tr>
<td>Alkaline phosphatase u/L</td>
<td>938</td>
<td>281</td>
<td>366</td>
</tr>
<tr>
<td>γ-Glutamyl transpeptidase u/L</td>
<td>271</td>
<td>86</td>
<td>95</td>
</tr>
<tr>
<td>Aminotransferase (AST/ALT)</td>
<td>675/393</td>
<td>257/162</td>
<td>104/56</td>
</tr>
<tr>
<td><strong>Pre-procedural Diagnostics</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ultrasound Abdomen</td>
<td>Normal gallbladder</td>
<td>Normal gallbladder</td>
<td>Complex loculated ascites, normal gallbladder</td>
</tr>
<tr>
<td>HIDA</td>
<td>Not obtained</td>
<td>No evidence of biliary excretion over 24 h</td>
<td>Not obtained</td>
</tr>
<tr>
<td>Liver biopsy</td>
<td>Cholestatic liver disease(^\text{a,b,c,d,e})</td>
<td>Cholestatic liver disease(^\text{a,b,c,d,e})</td>
<td>Cholestatic liver disease(^\text{a,b,d})</td>
</tr>
</tbody>
</table>

\(^\text{a}\) Periportal ductal proliferation.
\(^\text{b}\) Fibrosis (portal, peri-portal, bridging).
\(^\text{c}\) Incomplete nodule formation.
\(^\text{d}\) Bile plugging.
\(^\text{e}\) Giant cell transformation of hepatocytes.

### Notes

- Patient 1 is a 31 week gestation male with no significant co-morbidities. He presented for evaluation with a five week history of hyperbilirubinemia and persistent jaundice requiring prolonged phototherapy outside of the immediate neonatal period. His total bilirubin was 11.9 mg/dL, direct bilirubin of 9.1 mg/dL, and GGT of 271u/L (normal 10–78). An abdominal ultrasound was obtained which was unable to identify a common bile duct but was able to visualize the gallbladder. Liver biopsy revealed cholestatic liver disease with periportal ductal proliferation, fibrosis, incomplete nodule formation, bile plugging, and giant cell transformation of the hepatocyte.

- Patient 2 is a 31 week gestation male with 2q37 chromosomal deletion, significant coarctation of the aorta, and respiratory insufficiency. The patient required aggressive cardiac and ventilator support in the first several months of life. He presented with four weeks of hyperbilirubinemia with an accompanied elevated GGT. His total bilirubin was 6.0 mg/dL and direct bilirubin of 5.1 mg/dL, despite Actigall use. He underwent an ultrasound which revealed a normal gallbladder. HIDA scan showed hepatic uptake without clear biliary excretion over a twenty-four period. Liver biopsy revealed cholestatic liver disease with periportal ductal proliferation, fibrosis, incomplete nodule formation, bile plugging, and giant cell transformation of the hepatocyte.

- Patient 3 is a 32 week gestation male found to have severe ascites of unknown etiology. The patient required bi-weekly paracentesis and developed intracranial hemorrhage with resultant hydrocephalus and restrictive lung disease. He presented with an 8 week history of persistent hyperbilirubinemia with a total bilirubin on 3.6 mg/dL and a direct bilirubin of 2.9 mg/dL with a slowly elevating GGT. An ultrasound revealed a normal gallbladder but had difficulty visualizing the common bile duct. Liver biopsy revealed cholestatic liver disease with periportal ductal proliferation, fibrosis, and bile plugging.

All three patients underwent PTCC (Figs. 1–3). The gallbladder was accessed with ultrasound guidance via a percutaneous transhepatic route and contrast was injected with opacification of the gallbladder,
cystic duct, main hepatic bile ducts, and flow of contrast to the duodenum. These findings ruled out biliary atresia in all cases. All three patients tolerated this procedure without any complications. Further evaluations were done to delineate the underlying cause for their hyperbilirubinemia. Patient 1 was found to have idiopathic neonatal hepatitis with resolution of hyperbilirubinemia. Patient 2 was felt to have multifactorial neonatal hepatitis which resolved. Patient 3 was found to have a genetic condition and had eventual resolution of hyperbilirubinemia.

3. Discussion

Persistent direct hyperbilirubinemia beyond the neonatal period requires a comprehensive work up to exclude BA. Once the comprehensive laboratory and infectious work up is completed, further investigation with radiographic imaging is essential. This includes US and at times a HIDA, albeit a HIDA scan may not be obtained in all cases or centers. If there is still concern for BA, a liver biopsy is obtained to help provide insight to the persistent cholestasis. There are cardinal features on a liver biopsy which implies BA. These include: expanded portal tracts with bile duct proliferation, portal fibrosis, absence of sinusoidal fibrosis, portal tract edema and inflammation, canalicul and bile duct plugs [10,11].

If at this point in the work-up BA is still considered possible, an intraoperative cholangiogram (IOC) is warranted to further delineate biliary anatomy. There are several approaches to IOC including open, laparoscopic, and percutaneous. At our institution, in addition to local anesthesia, we use mask general anesthesia unless they are already intubated and then they receive general endotracheal anesthesia. If patency of the entire biliary system is identified, then a surgical procedure can be avoided. If patency cannot be identified, then operative exploration with a PE is warranted. PTCC may also shorten operative time, as an IOC may not have to be performed. An IOC may be required even after a PTCC when the porta hepatitis is not consistent with the PTCC findings.

All three of the infants described in this series had concern for BA on their comprehensive workup. PTCC allowed the two poor surgical candidates to avoid a surgical procedure to rule out BA. PTCC is a feasible alternative to surgery to exclude the diagnosis of biliary atresia even in patients with significant comorbidities. While the gold standard is intra-operative cholangiography, PTCC may be an option for those patients with severe comorbidities.

Informed consent

Informed consent was waived by our IRB due to the fact that the data collected for this study was for a case report and was de-identified.

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Authorship

All authors attest that they meet the current ICMJE criteria for authorship.

Conflict of interest

The authors have no conflict of interest to disclose.

Appendix A. Supplementary data

Supplementary data related to this article can be found at https://doi.org/10.1016/j.epsc.2018.05.003.

References


Joseph Sujka: Data collection, data analysis and interpretation, drafting article, critical revision of article, approval of article, statistics.

Katrina Weaver: Concept and design, data collection, data analysis and interpretation, drafting article, critical revision of article, approval of article.

Ashwini S. Poola: Data collection, drafting article, critical revision of article, approval of article.

Douglas Rivard: Concept and design, data analysis and interpretation, approval of article, statistics.

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