Duodenal atresia associated with pre duodenal portal vein Biliary atresia

A case report

Zubrina Solomon, A Theron, T. Govender
University of Witwatersrand
- Duodenal atresia incidence: 1 in 5000 to 10,000
- Males > females.
- Association with abnormalities of the biliary tree very rare.
- Pre-duodenal portal vein incidence: 1 in 10,000
- Syndromic forms of biliary atresia may include the presence of a pre-duodenal portal vein.

The case

- Female neonate
- Born 35 /40 with BW: 1810g
- RVD exposed, PCR negative and positive RPR.

**On examination:**
- Non bilious aspirates
- Distended epigastrium
- No stool on PR.

**AXR:** double bubble
Laparotomy

- D17 of life

**Findings:**
- Type 1: Duodenal atresia at the level of D2/D3.
- Contracted small gall bladder

**Operation:**
- Kimura DSD
- Liver biopsy

**Post operative clinical progression:**
- Progressed to full feeds
- No pigmented stools passed
# Liver function tests

<table>
<thead>
<tr>
<th></th>
<th>09/05/17 (D14 life)</th>
<th>22/05/17</th>
<th>13/06/17</th>
<th>30/06/17</th>
<th>23/07/17 (Post Kasai)</th>
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<tbody>
<tr>
<td><strong>Total Protein</strong></td>
<td>48</td>
<td>46</td>
<td></td>
<td>42</td>
<td>56</td>
</tr>
<tr>
<td><strong>Albumin</strong></td>
<td>27</td>
<td>25</td>
<td></td>
<td>25</td>
<td>29</td>
</tr>
<tr>
<td><strong>Total Bilirubin</strong></td>
<td>122</td>
<td>141</td>
<td>141</td>
<td>152</td>
<td></td>
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<tr>
<td><strong>Conjugated Bilirubin</strong></td>
<td>76</td>
<td>102</td>
<td></td>
<td>101</td>
<td>109</td>
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<tr>
<td><strong>Alanine transaminase</strong></td>
<td>53</td>
<td>11</td>
<td></td>
<td>421</td>
<td>86</td>
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<tr>
<td><strong>Aspartate transaminase</strong></td>
<td>157</td>
<td>91</td>
<td></td>
<td>344</td>
<td>273</td>
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<tr>
<td><strong>Alkaline phosphatase</strong></td>
<td>558</td>
<td>574</td>
<td></td>
<td>1039</td>
<td>353</td>
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<tr>
<td><strong>Gamma glutamyl transferase</strong></td>
<td>481</td>
<td>727</td>
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<td>529</td>
<td>355</td>
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<tr>
<td><strong>INR</strong></td>
<td></td>
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<td>0.99</td>
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</table>
Biopsy

Extensive bile duct proliferation, portal fibrosis associated with an inflammatory infiltrate with no bridging fibrosis.
D97 of life.

**Findings:**
- Type 2 biliary atresia, a pre-duodenal vein and a normal spleen.
- The left common hepatic artery on the left of the portal plate dividing into right and left only higher up with the right hepatic artery crossing over the portal plate.

**Operation:**
- Kasai Portoenterostomy incorporating the right hepatic artery into the portoenterostomy
The outcome

Post operative:
- Break down of the Roux en Y anastomosis

Clinical:
- No passage of pigmented stool
- Referred to GIT unit for workup for transplant
Discussion

- Intrinsic DA: failure of recanalization of the duodenum.
PDPV development
Biliary atresia connection

Possible blastogenetic abnormality that interferes with the connection and merging of duodenal, pancreatic and biliary canals at the middle line axis could lead to the obliteration and atresia of the extrahepatic bile duct, which may accompany the duodenal atresia.
The syndromic subtype of biliary atresia: developmental insult that occurs during the differentiation of the hepatic diverticulum from the foregut.

Aetiology is still uncertain

Association between BA and duodenal atresia is more frequently seen in the non syndromic type.

PDPV is usually discovered 1st

Surgical vigilance

Did we look for it?

Conclusion

- PDPV biliary atresia associated with DA is rare.
- The closely related embryological development of the duodenum and biliary tract may explain this phenomenon.
- The diagnosis of biliary atresia was suspected at the initial operation at 2 weeks of age at which point the patient was not jaundiced.
- PDPV: not cause for the duodenal obstruction.
- Focused and thorough Exploratory laparotomy must always be performed.