Reviewer's report

Title: Abernethy malformation: a case report

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Reviewer: stephanie Franchi-Abella

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Major compulsory revisions:

1. Abstract: Introduction : the authors write that Abernethy malformation is a liver disease this is not true it is a vascular malformation with no liver disease. Liver modifications (small volume, nodules, hepatic test disturbance) are secondary to portal privation.

2. Abstract: Conclusion and discussion: We agree with the authors that long term follow-up is necessary but closure of the shunt when possible should be discussed to cure and even prevent complications.

3. Discussion: I do not agree when the authors write that “the patient is usually symptom free” “in the early course of the disease”: As reported in the literature, neonatal cholestasis, hypoglycaemia or cardiac failure can occur and spontaneously resolve. Hepatic encephalopathy is not so rare in children. Infraclinic forms are more frequent.

4. Discussion: In our experience in a tertiary reference center for liver disease in children, and as reported by other authors, portal veins are present in the parenchyma even if extremely hypoplastic and not visible with usual imaging modalities. Liver biopsy is not indicated to evaluate the possibility to close the shunt. Balloon-occlusion test of the fistula is useful to evaluate the portal branches and evaluate portal pressure after occlusion in order to choose a one or two step closure procedure.

5. Discussion: It is now reported by several teams that porto-systemic shunts usually called Abernethy 1 malformations can be closed safely but sometimes require a two-step procedure to let the portal branches enlarge slowly and avoid acute severe portal hypertension. Surgical or radiological closure should be the first treatment even preventively. Liver transplantation should be reserved to exceptionally complex anatomy where closure of the shunt is not possible. Liver transplantation is a complex procedure that leads to multiple and sometimes severe complications. It should not be recommended as the first therapeutic option. Closure of the porto-systemic shunt leads to restoration of a “normal” hepatic hemodynamic and most complications resolve after closure with no significant complication reported until now. This is the first therapeutic option.

6. Conclusions: US is the method of choice to screen liver disorders. We disagree with the authors when they write that porto-systemic shunt is a challenging diagnosis: the first step is to recognise on US, CT or MRI that the portal anatomy is not usual and the second step is to find the abnormal
communication between the portal and the systemic veins. This only requires knowledge of the anatomy and systematic analysis when performing the exams.

7. Bibliography is not up-to-date and does not give references concerning closure of shunts ((eg: S Franchi-Abella, JPGN 2010 )

Minor essentials revisions

1. Figure 1 Change the legend. The figure shows the direct communication between the portal trunk and IVC. It does not give information about arterial supply. Change the legend

2. Figure 2: give information about the technic: contrast injection and MIP reconstruction. figure 2b does not give more information than figure 2a. It should be deleted.

3. Figure 5 it is difficult on one figure that does not shows the arteries to be sure that intrahepatic portal branches are not seen. This figure is not necessary.

Level of interest: An article whose findings are important to those with closely related research interests

Quality of written English: Needs some language corrections before being published

Statistical review: No, the manuscript does not need to be seen by a statistician.

Declaration of competing interests:

'I declare that I have no competing interests'