Reviewer's report

Title: VACTERL Spectrum with Portal Hypertension - a case report

Version: 3 Date: 14 May 2009

Reviewer: Wolfgang Kreisel

Which of the following following best describes what type of case report this is?: New associations or variations in disease processes

Has the case been reported coherently?: Yes

Is the case report authentic?: Yes

Is the case report ethical?: Yes

Is there any missing information that you think must be added before publication?: Yes

Is this case worth reporting?: Yes

Is the case report persuasive?: Yes

Does the case report have explanatory value?: Yes

Does the case report have diagnostic value?: No

Will the case report make a difference to clinical practice?: No

Is the anonymity of the patient protected?: No

Comments to authors:

The authors have submitted a revised and improved version of their manuscript. However, I am not so happy with the result. In the manuscript one can find some incorrect details, several typing errors, and the arguments that the findings in the present paper are not the result of an extrahepatic portal vein thrombosis with partial reopening of the portal vein, but rather a congenital defect, are not completely convincing.

Abstract:

First phrase: The occurrence of VACTERL spectrum and extrahepatic portal hypertension has not been reported in the literature.

This shorter phrase seems to be better (in my opinion) later: embryonic (not embroyonic)
Introduction*

Portal hypertension consists ......This is incorrect. Esophageal varices etc. are late consequences of portal hypertension and cannot be found in every case of portal hypertension Better: Late consequences of portal hypertension may be: esophageal varices, gastric varices, splenomegaly, ascites, and "caput medusae".

Second last paragraph: .... and Doppler sonography (not: the dopplers) What does it mean: The portal vein was thready (...) with collateral vessels involving both left and right portal veins. Are these collaterals going parallel to left and right main portal vein branch?

Interpretation of: There was no intrahepatic duct dilatation.

*Discussion*

Second paragraph.

respiratory (respiratory), patients (patient) In the paragraph starting with "Our patient with portal hypertension...

The authors state that their data support the assumption the patient has rather a congenital defect of the portal vein than a portal vein obstruction. I think the arguments are not conclusive, apart of the fact that one phrase is found in duplicate. Have Ando et al. showed that the feature of extrahepatic portal vein obstruction in young patients is so different from that found in the present patient? If yes, the conclusion of the authors is correct. But they should comment on this! The authors want to differentiate between extrahepatic portal vein obstruction and portal vein thrombosis. This should be explained in more details.

Paragraph starting with: "The frequency in PVT.... Do the authors want to say that in their patient they have looked for presence of these coagulation defects cited here, and have found none of them?

*Conclusion*

Typing errors: patient (pateint), embryonic (embroyonic)

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