Reviewer's report

Title: Phosphodiesterase 5 Inhibitors Lower both Portal and Pulmonary Pressure in Portopulmonary Hypertension. A case report.

Version: 3 Date: 5 May 2007

Reviewer: Florian S Fuchs

I am familiar with the literature and believe that this case meets one of the 7 criteria for evaluation in the journal: New associations or variations in disease processes

Has the case been reported coherently?: Yes

Is the case report authentic?: Yes

Is this case worth reporting?: Yes

Is the case report persuasive?: No

Does the case report have explanatory value?: Yes

Does the case report have diagnostic value?: Yes

Will the case report make a difference to clinical practice?: Yes

Comments to authors:

General
Authors report about treatment of portopulmonary hypertension and about the diagnostic approach for pharmacological testing the effects of PDE5 inhibitors on portal hypertension. Portopulmonary hypertension is an important kind of pulmonary arterial hypertension and good therapeutical possibilities for this patients are needed.

Revisions necessary for publication
- In the chapter "Backgrund" the authors say that PDE5 inhibitor are the standard therapy of IPAH. So one might think, PDE5 inhibitors are the only standard therapy of IPAH.
- The authors describe a patient with alcoholic liver cirrhosis. A prothrombin time of 68% and an elevated bilirubin of 3,3 mg/dl leads to Child Class B (minimum of 8 points). Ascites was absent, did the patient suffer from encephalopathy, did he have a reduced albumine?
- The patient had no severe reduction of his performance status (NYHA II, 6MWT 522 m, normal arterial blood gas analyses before and after 6MWT, high values in CPET). Was it neccersary to initiate a therapy of the pulmonary hypertension? Why was ist done with sildenaifl two but not three times daily?
- Did the authors exclude any other cause for the described episodes of syncope? Left heart status in echocardiography?
- Why was tadalafil used for reactivity testing in right heart catheterization? It would have been interesting to compare PDE5 inhibitor and a standard testing substance (NO, adenosin, prostacyclines). Mean PAP was reduces less than 10 mmHg an PVR was reduced about exactly 25%, so there was no distinct effect of tadalafil on pulmonary haemodynamics.
- The PWP is missing, which seems to be relevant for exclude a left ventricular cause for pulmonary hypertension. Over all any other kinds of pulmonary hypertension were not excluded properly.
- The authors should state, why they used two different PDE5 inhibitors for testing the pulmonary haemodynamics?
- One could not concluse that PDE5 inhibitor are no risk factor for bleeding from esophageal varices as the authors do.
- In figure 1 cardiac index but not cardiac output is shown.
- In table 1 systemic blood pressure was high during first pulmonary haemodynamic evaluation. Did the patient receive any medication for this or any other baseline medications?

What next?: Revise and resubmit
Quality of written English: Acceptable