Reviewer's report

Title: Life-threatening hypersplenism due to idiopathic portal hypertension in early childhood: Case report

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Reviewer: jean de Ville de goyet

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Case report type paper; the authors present a patient with a remarkable clinical presentation, being massive splenomegaly and secondary ipersplenism causing a lifethreatining clinical condition, mostly ventilation failure by abdominal distension and diafragmatic compression. They hypothetised idiopathic portal hypertension as primary disease, with splenectomy being the radical and complete treatment.

Major Compulsory Revisions:
They are few contradictions or inconsistencies:
- With the primary diagnosis in this child being idiopathic portal hypertension, it is a requisite to demonstrate that there was an increase in portal pressure: there is in fact no evidence at all in the paper that portal pressure was high. A grade 1 gastropathy is - per se and in the critical condition of the child - not indicative of portal hypertension, when all other facts do not support the latter hypothesis.
- The authors refers to the size of portal vein (that is within the normal range for age!) and hyperdynamic state as a sign of portal hypertension when in fact they write also "absence of formally abnormal portal vein flow on Doppler Ultrasound"
- Splenomegaly per se is not a sign of portal hypertension, as is not Hepatomegaly.
- In the worse condition of portal hypertesion, splenomegaly develops moderately with time and never causes such rapidly progressing organomegaly in children; The latter rapid development suggest rather a malformative growth. Vascular malformations and angiomas typically grow rapidly during early childhood and some can be associated with major trombopenia and coagulation disorders causing hemorrhagic diathesis. Even the results of immunochemistry would be consistent within the latter context.
- Overall, the authors cannot prove that portal hypertension was present and this is major weakness in discussing the case as presented; they may have faced a condition with a primary disorder of the spleen caused secondary ipersplenism and abdominal compression, and of course a resolution of the symptoms after splenectomy.

Minor Essential Revisions
The terminology "Banti syndrome", a term mostly used in the past to describe
any condition with splenomegaly of uncertain origin, has been quasi abandoned in literature and nearly disappeared from papers a decade ago. It is nowadays accepted that in most cases portal hypertension had a specific, but unrecognised, cause.

Discretionary Revisions

**Level of interest:** An article of insufficient interest to warrant publication in a scientific/medical journal

**Quality of written English:** Acceptable

**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'