Author's response to reviews

Title: Budd-Chiari Syndrome: Long term success via hepatic decompression using transjugular intrahepatic porto-systemic shunt

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Author's response to reviews: see over
Dear Editor Dr Zauner,

thank you very much for the comments of the reviewer which were extremely helpful to further improve our manuscript.

In the following we are giving a point-by-point response to the comments of the reviewer Martin Rösslle.

Abstract

1. **Introduction** paragraph, line 8-9:
   We changed the statement as suggested by the reviewer: “The aim of this study was to report our single centre long term results of patients with BCS receiving one of three treatment options i.e., medication only, TIPS or OLT on an individually based decision of our local expert group.”

2. **Results** paragraph, 2. sentence:
   We added “plasmatic”, as suggested by the reviewer.

3. **Results** paragraph, 3. sentence:
   We changed the sentence as suggested: “… and one patient died from liver failure before any specific treatment could be initiated.”

4. **Results** paragraph, 4. and last sentences:
   We changed the sentences as suggested: “Eleven of 13 TIPS patients required 2.5 ± 2.4 revisions (range: 0-8). One patient died from his underlying hematologic disease. The residual 12 patients still have stable liver function not requiring OLT. All 4 patients who... Survival in the TIPS group was 92.3% and in the OLT group 75% during a median follow-up of 4 and 11.5 years, respectively.”

5. **Conclusions** paragraph:
   We dropped the paragraph and inserted instead: “Our results confirm the role of TIPS in the management of patients with acute, subacute and chronic BCS. The limited number of patients with OLT does not allow to draw a meaningful conclusion. However, the underlying disease may generate major complications, a reason why OLT should be limited to patients who cannot be managed by TIPS.”
Introduction

6. We dropped the following two sentences: “Recommendations and guidelines for the management of BCS have been derived from retrospective studies. So far, only one prospective study on BCS has been published.” Therefore we added: “The present treatment recommendations of BCS are not based on randomized studies but on a small number of retrospective studies and one prospective study. The aim of this single centre retrospective study was to further enlarge the body of patients evaluated with the intention to allow steady optimization of present treatment strategies” at the end of the Introduction section (see 9.).

7. 2. paragraph:
We replaced “condition of the liver” by “severity of liver disease”.

8. 2. paragraph, last sentence:
We completed the references and rearranged them.

9. We changed the text as suggested (see 6.).

Patients and Methods

10. paragraph 4:
We clearly mentioned the reasons why the 4 patients in the OLT group did not receive TIPS treatment: “Patients who underwent OLT as initial therapy were not candidates for TIPS. Two patients were transplanted before TIPS had been introduced as a treatment option of BCS, namely in 1988 and 1992. The other patients had cirrhosis with signs of chronic liver failure in terms of hepatic encephalopathy and high bilirubin levels so that liver transplantation was considered essential in these patients.”

11. Second page, first paragraph:
We deleted the paragraph “significant narrowing of the lumen on venography” as suggested.
Results

12. First paragraph:
We deleted lines 3-8 and wrote instead: “Myeloproliferative disorders (especially PV) were the cause of BCS in 6 patients, 7 patients had plasmatic coagulation abnormalities, 1 patient took oestrogen medication as a possible underlying hypercoagulable condition and in 6 patients the aetiology of BCS remained unclear.”

13. Second paragraph:
We agree with the reviewer. Patients with pain alone are rather uncommon in the literature. But we checked our data and could confirm our findings.
14. We joined the first to the second paragraph and introduced the subheading “Patients Characteristics”.
15. Third paragraph:
We introduced the subheading “Treatment and Outcome”.
Discussion

We restructured the Discussion as suggested.

16. First and second paragraph:
We dropped the sentences 1-4 and begun with: “In this study, we assessed the clinical course and long term outcome of patients with BCS referred to our university hospital analysing the data of all patients admitted with a primary diagnosis of BCS between 1988 and 2008.”

17. We completely retyped the third paragraph following the reviewers suggestions.

18. We inserted more references supporting this statement.

19. We dropped the two sentences as suggested.

20. 3. paragraph:
We dropped: “None of our patients in the TIPS group had to undergo OLT, subsequently.” and added the rest of the paragraph to the third paragraph (see 17.).

21. 4. paragraph:
We dropped the statement: “Four patients were considered non-eligible for TIPS placement and had to undergo OLT as initial therapy.” and wrote instead: “Four patients underwent OLT as initial therapy. Two were transplanted before TIPS had been introduced as a treatment option of BCS and two had cirrhosis with signs of chronic liver failure.”

22. We added a figure legend to the text: “Figure 1. Probability of survival of all BCS patients. On the x-axis the time of follow-up in years is given. On the y-axis the probability of survival is shown.”

Because of the large time span of 20 years on the x-axis and the early events within a few days our software (SPSS) is not able to give a correct projection so that the graph does not start at 1.0.

We did not present the survival curves of the subgroups because the low number of cases in the subgroups do not allow a valid Kaplan Meier estimate.

Yours sincerely, Alexandra Zahn and Peter Sauer.