

Response to reviewers' comments:

Professor Jing Yu,

Manuscript No: 27421

Title: Pediatric living donor liver transplantation for congenital hepatic fibrosis using a mother's graft with von Meyenburg complex: A case report

# Reviewer 03474922

This case report is interesting in the context of pediatric liver transplantation given the organ donor shortage. It gives a solution to the medical team and should not be a limitation to consider donation.

Thank you very much for your favorable comments and valuable advices.

Abstract: Give a good understanding of the case. The part concerning the donor might be a bit long for the abstract.

We changed the part concerning the part of the donor to be shortened.

Background: Good introduction, well structured and easy to read. But the sentence: "due to the lack..." is in contradiction with the sentence above "therefore it may be...". The authors try to persuade the reader that it could be a valid option for a transplant and this part might put a doubt in his mind. Even though one should stay cautious given the lack of data so far.

Thank you very much for your valuable advices. We cut the sentence 'Therefore, it may be considered appropriate to use a liver graft for liver transplantation (LT) in case of von Meyenburg complex' because it was redundant. In addition, we added the sentence ', and considered the possibility of a liver with von Meyenburg complex for a further expansion of living donor pool.'

Case Presentation: The story is well written. However, some more details about the following aspect of the story would be interesting: - Were there any other symptoms apart from the hematemesis that would have led to the liver biopsy? - Why the donor didn't get a biopsy in order to establish the diagnosis with certainty before transplantation? What was the rationale? - Is the CT at POD 28 a standard of care ?

if not, what had lead the team to do it? - What about the follow up ? For how long has she been transplanted? How often does she have medical visits? - What about immunosuppression?

She was also accompanied with melena, but it had not been serious compared to the hematemesis. We considered it was not enough worth describing.

As you say, we considered that the liver biopsy for donor should be performed. However, the condition of the recipient had got worse suddenly, therefore we did not have enough time to do. In addition, the diagnosability of MRI for von Meyenburg complex was reported high (reference 12, 13), therefore we decide to perform the LDLT without histological evaluation.

We routinely performed the CT examination at POD 28. We added the point in the sentence.

We check the routine blood test (including standard liver function test and renal function test) and echo check per 3 months for first 5 years and per 6months after that. We also examine the CT scan at 1 months, 6 months, and 1 year after LT with the use of CT volumetry. From 1 year to 5 years after LT, the volumes were examined annually; thereafter they were examined every 2 years. (Transplantation proc.2016)

We will perform the same follow up for the recipient. In addition, we will add the tumor marker (CEA, CA19-9) check, when the blood examination will be performed.

Immunosuppression regimen was tacrolimus and methylpredonisolone. We added the information.

Discussion: Also easy to read. End of third paragraph: " to our experience..." please add references when you write about previous report. Again could you please be more specific when you write about oncologic surveillance. What would be your recommendation? Same comment regarding renal function. Figure 3: an arrow would help to better see what lesion we are suppose to see.

Thank you very much for your favorable comments. We added the reference number again at the point.

As we mentioned above, we check the routine blood test (including standard liver

function test and renal function test) and echo check per 3 months for first 5 years and per 6 months after that. In addition, we perform the CT scan per year. We will perform the same follow up for the recipient. We will add the tumor marker (CEA, CA19-9) check, when the blood examination will be performed. Although it is unknown whether they are valid or cost effective, we will perform special detailed follow up for such a rare case.

We added an arrow to in Figure 3a and 3c.

# Reviewer 03017199

Comments: This manuscript describes Pediatric living donor liver transplantation for congenital hepatic fibrosis using a mother's graft with von Meyenburg complex: A case report. Despite the work is potentially interesting and the phagocytic activation is of great clinical relevance, some points need attention...

Thank you very much for your favorable comments.

Specific comments:

1. Markers of liver function should be provided.

We added Table 1 which demonstrated the preoperative liver function of the recipients. We also added the liver function of the donor in the case presentation.

2. The authors should more discuss what is innovative or profitable of this method in this case.

As we demonstrated in the abstract, there exist only two case reports of LT with a donor having von Meyenburg complex, and this is the first report of LDLT. To our experience and according to previous reports, graft liver with von Meyenburg complex shows no problem regarding graft function in the early post-LT phase. However, long time prognosis of the liver graft with von Meyenburg complex is unknown, especially concerns about liver function, risk of carcinogenesis, and renal function. We emphasized these problems again, and indicate our follow up plan for the patient.