Author’s response to reviews

Title: Patients with Glycogen Storage Diseases Undergoing Anesthesia: A Case Series

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Author’s response to reviews:

Dear Dr. Riha,

We wish to thank you and your reviewer for the thoughtful critique of our manuscript "Patients with Glycogen Storage Diseases Undergoing Anesthesia: A Case Series" (BANE-D-17-00182). We found these comments helpful in improving the strength of this manuscript. We have a point by point response to each point and have revised the manuscript accordingly. Please let us know if any further changes are requested.

We look forward to your decision.

Sincerely yours,

Toby N Weingarten, MD

MAJOR COMMENTS

1. In Methods section I would suggest to add a definition for metabolic acidosis (similar to definition of hypoglycemia) which was used by the authors during review of their cases.

RESPONSE: Metabolic acidosis was defined as pH imbalance primarily secondary to a reduction in bicarbonate (HCO3−) of 22 mmol/L or lower. This definition has been added in the text under Method section line 11, page 5.
MINOR COMMENTS

1. Page 10/last paragraph: I would suggest ...disturbances despite intraoperative administration of LR...

RESPONSE: This has been changed in the text under Conclusions section line 22, page 10.

TECHNICAL COMMENTS

1. Page 9/end of first paragraph: the reference to the statement of MHAUS should be numbered and moved to the list of References including the date when the link was checked.

RESPONSE: This reference has been numbered 24 and it has been moved to References section line 11, page 11.

Zdenek Turek, M.D., Ph.D. (Reviewer 2): Authors presents retrospective analysis of perioperative course in patients suffering from Glycogen Storage Disease (GSD) as a typical topic in "Anesthesiology and rare coexisting disease"

Despite understandable and reasonable retrospective character of such presented data, the level of importance for current anesthesia regular practice seems to be very high.

Reviewer's Comments

A) According to case mix - no patient in evaluated cohort underwent regional anesthesia (peripheral nerve blockage, spinal or epidural anesthesia) - were these pts excluded or were not found in retrospective period? - please, this could be mentioned in study limitations.

RESPONSE: This is a very good point. In our study we included all patients with a diagnosis of GSD who underwent surgery at our institution under anesthesia care (general, MAC or regional). However in our cohort study none of the patients underwent regional anesthesia. This has been added under Limitations section line 8, page 10

B) According to available knowledge, type I GSD is associated with platelet dysfunction, which is in part correlated to the extent of the dyslipidemia (1,2,3).


- Are there any signs of unusually increased perioperative blood loss in retrospectively observed cohort of patients?

RESPONSE: This is a very good point. The current thinking is that the observed platelet dysfunction is secondary to an acquired vWF deficiency. This has been added to the Discussion regarding GSD-1 (Page 8, line 7) and a reference provided (22). Suggested therapy is also added to this area as well as a reference (23). A reference to this effect has been provided. However, in this cohort, despite the majority of our type I GSD underwent major procedures like liver transplant; none of them experienced unexpected increased blood loss.

C) Hypoglycemia during liver transplant - due to GSD type I or as a consequence of surgical procedure itself? - please, to be mentioned in the discussion

RESPONSE: Thank you for this comment. All patients with type I GSD had a history of hypoglycemia and all were consequently treated with intraoperative dextrose solution. In this setting I think it would be hard to report how much the liver transplant could have contributed to the hypoglycemia. Of note in only one patient (patient #4) the intraoperative glucose immediately before stating the dextrose solution was 34. This patient underwent liver transplant. A comment in regards has been added under Discussion section lines 28, 35; page 7

Technical details

- side 7, line 41 (LR) - Ringer's lactate? This abbreviation is not defined above.

- Table 2, pt. # 12 - abbreviation is not defined below, (Alfentanil?)

RESPONSE: Thank you for identifying these errors. LR definition has been added in Discussion section line 41 page 7. Alfentanil (AL) abbreviation has been added to table N.2