Reviewer's report

Title: Deficiency of ADAMTS-13 in Pediatric Patients with Severe Sepsis and the impact on outcome.

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Reviewer: MOREL Nathalie

Reviewer's report:

This article is interesting. In fact, the asked question is important. However, authors must modify some points, both to make the article more robust and to be easily readable. Furthermore, it is maybe necessary to use these results with caution, because, time to obtain results of ADAMTS 13 is maybe different and in some centers long; the time is maybe sometimes too long in comparison to possible quickly poor evolution.

Please, could you answer to the following questions or correct mistakes, before a decision on publication can be reached (major compulsory revisions).

1/On the content:
- In materials and methods:
  o Is it possible to briefly recall severe sepsis criteria?
  o Are you sure that this type of study is descriptive, cross-sectional? Outcome data necessites a longitudinal study. Otherwise, retrospective character must be indicated.
  o ADAMTS 13 analysis: What type of test was used to describe decrease of ADAMTS-13 levels? Activity? Antigen?
  o Precise what is assayed in outcome: mortality? Could you give average time of follow up in your study, and, time of follow up for the last patient included in the study.
- In results:
  o At second line: sum of the two percentages (56,3% and 43,8%) is not equal to 100: could you verify the rounding of percentages.
  o “Clinical characteristics were compared and were not statistically significant”. You can’t use “statistically” because statistical tests were not performed for all characteristics compared. In fact, statistical tests are not necessary in this case but you can’t write “statistically”: Clinical characteristics were compared and don’t seem to be different between two groups.
  o Precise time of follow up in this part too; otherwise it’s difficult to understand proportion of alive patients, dead patients, and patients lost to follow-up.
- In discussion:
  o “our results revealed that pediatric patients with severe sepsis seem to have a
higher incidence of ADAMTS13 deficiency. (in fact, you didn’t give a statistical test to affirm that incidence is really higher.” Furthermore, who is the control group used?

- “Septic patients with ADAMTS13 deficiency were more thrombocytopenic”: results allowing to affirm that, and statistical tests must be detailed in results paragraph.

- For figures:

  - Table 1: Give the signification of used abbreviations: PT, APTT. You speak about which type of cultures? (blood cultures?). Give the p-value for negative cultures, if you gave it for positive cultures.

  - Figure 1: Length of hospitalisation in place of hospitalisation’s duration. Please give units of two axes. Please, could you give title of Y axe without abbreviation.

2/On the form:

- Some precisions could be done to make easier reading and comprehension:

  - In summary and in full text: “comparison was done with regard to some clinical and biological characteristics and outcome between two groups”

  - First paragraph of discussion: could you reformulate the first sentence, because I think we could not class defect in ADAMTS-13 because of acquired antibodies in primary causes.

  - Second sentence of discussion: precise that specific treatment must be performed in emergency.

- Some mistakes must be corrected:

  - Please, could you unify abbreviation use for “von Willebrand Factor”: vWF or VWF but not both.

  - In introduction: “decreased levels of ADMATS-13 are particularly seen in TTP”.

  - In introduction: “systemic lupus erythematosus”.

  - In materials and methods: “kept at -80°C temperature until analyses”.

  - At the fourth paragraph of results: “difference was not statistically significant”.

  - In discussion: “may have change the results (and not changed).”

- Proofreading by an anglophone seems essential.

  - In fact, style is perfectible and some sentences are difficult to understand (for example, “thus the burden…until end of introduction”; Prefer “preventive strategies are needed… in place to “there is a need for preventive strategies”; “the outcome these of patients”; “dead” in place to “expired”; “ADAMTS13 supplementation and use of synthetic granulocyte…”).

  - Presentation of references must be unify and concordant to instructions to authors of the review: particularly, caution to the presentation “;” or “.” between names of authors and title of article. Caution to presence or not of month of publication.
**Level of interest:** An article whose findings are important to those with closely related research interests

**Quality of written English:** Needs some language corrections before being published

**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' below.