For research article: An assessment of dietary intake, feeding practices, growth and swallowing function in young children with late onset Pompe disease: a framework for developing nutrition guidelines. Pendyal et al.

Response to Reviewer 3 Comments

1. Summary

Reviewer comment: The work presented by Pendyal et al. is very well written and meticulously organized. The authors provide a comprehensive data presentation, covering all major aspects of the study in deep detail. The same is true for the discussion section in the manuscript, where the authors reflect to every single detail of the presented data in order to define the excellent framework for recommended nutritional guidelines at the end of the discussion.

> As the authors point out, various studies need to be performed in individuals with this disease to derive important information for helping parents to apply the most appropriate diet to their children and improve life quality and expectancy. From a metabolic standpoint I could envisage a need for the follow up of the process of gluconeogenesis in situations of increased protein/amino acid intake. Most of the aminoacids are either glucogenic or partially glucogenic, with the exception of lysine and leucine, which are purely ketogenic. Thus, the substitution of carbohydrate by protein and the age at which that needs to occur requires appropriate evaluation of this gluconeogenic pathway. The same would be true for the glycogenesis pathway, to determine how impactful the excess protein intake would be on glycogen stores. With a prevalence of 1 in 20,000 this disease affects many individuals worldwide that would certainly benefit from dietetic guidelines supported by metabolic research.

Response: Thank you very much for taking the time to review this manuscript. We are glad that you find our paper to be well written and comprehensive. We appreciate your critical review of our paper and your feedback. We agree that overall glycogen metabolism and gluconeogenesis have implications on disease and should be taken into consideration when prescribing diets to children with LOPD. We are grateful for the time you took to review our manuscript.

2. Questions for General Evaluation	**Reviewer's Evaluation**	**Response and Revisions**
Does the introduction provide sufficient background and include all relevant references?	Yes	Thanks for the positive general evaluation
Are all the cited references relevant to the research?	No comment by author	

Is the research design appropriate?	Yes
Are the methods adequately described?	Yes
Are the results clearly presented?	Yes
Are the conclusions supported by the results?	Yes

3. Point-by-point response to Comments and Suggestions for Authors

Comments 1: Page 3, line 135: "14 74%)" should be 14 (74%);

Response 1: Thank you for pointing this mistake out. We have made this correction in the FOIS-P score of children to 14 (74%). We want to kindly notify you that this information now appears on page 3, paragraph 4 and in line 143 [vs 135] due to some edits made in the previous section.

Comments 2: Page 7, line 288: "muascle"; correct to muscle

Response 2: We apologize for this spelling error. We have, corrected this error by correctly spelling the word "muscle". The correction now appears on page 7, paragraph 3, line 294 of the revised manuscript.

Comments 3: Page 10, line 397: "between those were received ERT"; this segment in the sentence needs correction.

Response 3: Thank you pointing out this mistake in the sentence structure. This sentence which acknowledges the limitations of the study to not being to compare results between children who were on ERT and those who were not on ERT, now appears on page 10, paragraph 1 and line2 410-412 of the revised manuscript.