

Answers to the reviewers:

Reviewer #1: This manuscript is a case report describing a rare recessive inherited organic acidemia caused by a genetic deficiency of isovaleric-CoA dehydrogenase, a mitochondrial matrix enzyme that catalyzes the oxidation of isovaleryl-CoA to 3-methylcrotonyl-CoA in the leucine degradation pathway. The disease, referred to as isovaleric acidemia or IVA, occurs mainly during neonatal period and exhibit a high rate of mortality. Based on GC/MS analysis of blood and urine the 12-day old patient exhibited very high concentration of isovaleryl glycine causing severe metabolic stress and eventually died. This report, in addition to the clinical characteristics of the patient, provide extensive review of the reported cases of IVA over the period of last 14 years. The report is well written, clearly presented and provides an intricate insides that may help clinicians to early diagnose and manage the IVA.

Answer: Many thanks to the reviewer for the comments on my article.

Science editor: 1 Scientific quality: The manuscript describes a case report of the neonatal isovaleric acidemia. The topic is within the scope of the WJCC. (1) Classification: Grade B; (2) Summary of the Peer-Review Report: The report is well written, clearly presented and provides an intricate inside that may help clinicians to early diagnose and manage the IVA. Based on GC/MS analysis of blood and urine the 12-day old patient exhibited very high concentration of isovaleryl glycine causing severe metabolic stress and eventually died. This report, in addition to the clinical characteristics of the patient, provide extensive review of the reported cases of IVA over the period of last 14 years; and (3) Format: There is 1 table and 1 figure. A total of 37 references are cited, including 3 references published in the last 3 years. There are no self-citations. 2 Language evaluation: Classification: Grade A. A language editing certificate issued by MedE was provided. 3 Academic norms and rules: The authors provided the Signed Informed Consent, and the CARE Checklist - 2016. The authors need to provide the signed Conflict-of-Interest Disclosure Form and Copyright License Agreement. No academic misconduct was found in the Bing search. The highest single-source similarity index in the CrossCheck report showed to be 6%. According to our policy, the overall similarity index should be less than 30%, and the single-source similarity should be less than 5%. Please rephrase these repeated sentences. 4 Supplementary comments: This is an unsolicited manuscript. The topic has not previously been published in the WJCC. The corresponding author has not published articles in the BPG. 5 Issues raised: I found the authors did not provide the original figures. Please provide the original figure documents. Please prepare and arrange the figures using PowerPoint to ensure that all graphs or arrows or text portions can be reprocessed by the editor. 6 Re-Review: Not required. 7 Recommendation: Conditionally accepted.

Answer: Thank you for the comments. We have rephrased some repeated sentences to insure the single-source similarity is less than 5%, the signed Conflict-of-Interest Disclosure Form, Copyright License Agreement and original figure is in the attachment.