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PEER-REVIEW REPORT

Name of journal: World Journal of Clinical Cases

Manuscript NO: 58935

Title: Neonatal isovaleric acidemia in China: A case report and literature review

Reviewer's code: 00503623

Position: Editorial Board

Academic degree: MD, PhD

Professional title: Professor

Reviewer's Country/Territory: United States

Author's Country/Territory: China

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Reviewer chosen by: Chen-Chen Gao

Reviewer accepted review: 2020-10-05 14:24

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Review time: 1 Hour

Scientific quality	[ ] Grade A: Excellent [Y] Grade B: Very good [ ] Grade C: Good [ ] Grade D: Fair [ ] Grade E: Do not publish
Language quality	[Y] Grade A: Priority publishing [ ] Grade B: Minor language polishing [ ] Grade C: A great deal of language polishing [ ] Grade D: Rejection
Conclusion	[Y] Accept (High priority) [ ] Accept (General priority) [ ] Minor revision [ ] Major revision [ ] Rejection
Re-review	[ ] Yes [Y] No
Peer-reviewer statements	Peer-Review: [Y] Anonymous [ ] Onymous Conflicts-of-Interest: [ ] Yes [Y] No

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SPECIFIC COMMENTS TO AUTHORS

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.