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NUBPL: a mitochondrial Complex I deficiency disorder

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Reviewer 1

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*Only major points from review and responses included.

Reviewer 1

The authors reviewed here the current knowledge on NUBPL, a protein involved in the biosynthesis of Fe-S clusters. The review is well-written and informative about the syndromes and phenotypes associated with mutations in NUBPL, the diagnosis, and the clinical management.

My only suggestion is to include a scheme recapitulating the biosynthetic pathway of Fe-S clusters to help the readers to put NUBPL into context.

Authors

We thank the reviewer for this analysis of our work and we hereby include a figure to illustrate mitochondrial Fe-S cluster biogenesis (new Fig. 1). Therefore, the other figure numbers were changed accordingly. We have amended the manuscript with in depth description of the mitochondrial iron-sulfur cluster biogenesis to complement the new figure.