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Barth syndrome: a genetic ailment with a lipid component and bioenergetic ramifications

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Manuscript submitted 2022-03-07, 2022-05-18 (revision)

Manuscript accepted 2022-05-20

<https://doi.org/10.26124/bec:2022-0005>

Reviewer 2

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Manuscript reviewed 2022-04-13

<https://doi.org/10.26124/bec:2022-0005.r2>

*Only major points from review and responses included.

Reviewer 2

Regarding the statement “Respirasomes are thought to increase substrate channeling and increase efficiency of electron transport”, the authors should consider noting that the actual biological role and importance of respirasomes is still debated (see and perhaps cite PMID 29937372, 28380371, 30382836). There seems to be an increasing consensus that the major benefit of supercomplexes is to minimize electron leak to oxygen (i.e., superoxide formation), which is consistent with mitochondrial oxidative stress being a primary component of BTHS pathophysiology and tafazzin/CL deficiency (PMID: 25247053, PMID: 33793303; PMID: 24813252). This is also worth noting, as the precise impact of altered cardiolipin composition and tafazzin deficiency on bioenergetics is still an area of active investigation, and appears to be more complex than overt ETS dysfunction (PMID: 24285538, PMID: 32665401).

Authors

We thank the reviewer for this very thorough comment. We have included the new suggested references in the body of the manuscript and we highlighted in yellow the well-articulated words of the reviewer to be a part of this review. This comment enhanced the quality of our manuscript significantly.