

Format for ANSWERING REVIEWERS



December 11, 2013

Dear Editor,

Please find enclosed the edited manuscript in Word format (file name: 6448-review.doc).

Title: Clinical and Radiographic Features of Hutchinson-Gilford Progeria Syndrome: Case Report.

Author: Daniel Berretta Alves, Juliana Melo Silva, Tatiany Oliveira Menezes, Rosely Santos Cavaleiro, Fabrício Mesquita Tuji, Marcio Ajudarte Lopes, Alexandre Augusto Zaia, Ricardo Della Coletta

Name of Journal: World Journal of Clinical Cases

ESPS Manuscript NO: 6448

The manuscript has been improved according to the suggestions of reviewers:

1 Format has been updated

2 Revision has been made according to the suggestions of the reviewer

(1) Abstract: "The disease is caused by heterozygous mutations in lamin A (LMNA) and ZMPSTE24." This should read: The disease is caused by mutation in the lamin A/C gene (LMNA). "and ZMPSTE24" should be dropped, as mutations in this gene do NOT cause HGPS.

(2) Introduction: "Mutations in the lamin A (LMNA) gene are the main cause of HGPS. However, mutations in the ZMPSTE24 gene, which encodes a metalloproteinase that is specifically involved in the post-translational proteolytic processing of prelamin A to mature LMNA, has also been described in HGPS patients.¹⁴" This should read: Mutations in the lamin A/C (LMNA) gene are the cause of HGPS. THE REFERENCES AFTER THIS SENTENCE SHOULD BE: Eriksson et al. Nature 2003;423:293-8 (current reference 20) and De Sandre-Giovannoli et al. Science. 2003;300:2055. Mutations in ZMPSTE24 do NOT cause HGPS; it is caused by mutations in LMNA, see OMIM: <http://omim.org/entry/176670>

(3) Discussion: "the diagnosis of HGPS is based on the recognition of common clinical features and the detection of mutations in the LMNA gene and eventually in the ZMPSTE2 gene." Remove ...and eventually in the ZMPSTE24 gene.

(4) The authors should discuss the findings in this case with those in Chen et al. Genet Couns 2012;23:1-7 and cite that paper. 5. Reference 20: "Erlksson" should be Eriksson.

(5) Please cite at the abstract the age and sex of the child evaluated (11-year-old boy).

(6) Would suggest you to change the last phrase, to be more attractive for readers, because as you comment the strong point of your manuscript are the dentomaxillofacial features observed in this case, and this must be emphasized. I suggest something like: "Some common and uncommon HGPS-associated features, as alterations in the facial sinuses and hypoplasia of the condyles, were recognized in our patient, what may contribute to expand the phenotypic spectrum of this syndrome from a dentomaxillofacial perspective."

(7) When describing the syndrome, I would suggest you to cite the main age of the deaths and their common reasons (further to highlight the importance of the diagnosis and early intervention to avoid/delay damages).

(8) I would also suggest you to include something regarding the importance of recognizing dentomaxillofacial features, as well as regarding the contribution of your paper to the literature (the importance of a dentistry to know about and look for cited alterations in patients with this syndrome, especially because of the multidisciplinary attention that these patients deserve, as well as for the early diagnosis)

(9) I suggest you to change the sentence "This case highlights some common and uncommon dentomaxillofacial features associated with HGPS."

(10) At the discussion, you say: "The permanent teeth in the current report were macroscopically normal in shape and color", maybe you can include photos of the occlusal view to better illustrate the dental condition and the maxillomandibular relation (if you have one). **Patient's trismo doesn't permit to take the photo.**

(11) Although the references are good, I suggest you to add some other recent published papers (I've found some interesting ones: Zhang H, et al; Pereira S, et al; Chen CP, et al.

(12) Figure 2 and 3 are with a poor quality (maybe a consequence of the pdf version?). Please check.

3 References and typesetting were corrected

Thank you again for publishing our manuscript in the World Journal of Clinical Cases.

Sincerely yours,

A handwritten signature in blue ink, reading "Daniel Berretta M. Alves".

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