Author’s response to reviews

Title: Surgical Correction of Hyperlordosis in Facioscapulohumeral Muscular Dystrophy. A Case Report

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Author’s response to reviews:

Dear Prof. Allaix, Prof. Biber and Prof. Samadian,

Thank you very much for the comprehensive review of our manuscript entitled: "Surgical Correction of Hyperlordosis in Facioscapulohumeral Muscular Dystrophy. A Case Report" (BSUR-D-17-00076).

We sincerely appreciate the reviewers’ comments and feel encouraged by their professional questions and concerns. Below we provide a point-by-point response to the comments and highlight the revised parts of the manuscript in bold.

Reviewer #1 (Professor Biber):

(1) Question: Lee reported a similar patient, who however was ambulatory. In my view, this is a big difference to your patient! Maybe the operation is more beneficial in wheelchair patients. This is an issue to discuss. The passage "They thought the excessive extend of back muscles maintained upright posture and gave chance to stand and walk. Thus, the corrective for hyperlordosis wasn't recommended due to the possible damage to compensatory mechanism by hyperextended back extensor muscles." is hardly understandable for me. What are "hyperextended back extensor muscles" - we are talking about hyperlordosis. Extensors tend to be shortened, not extended ... Please re-phrase this passage and make your thoughts more explicit.
Response: Thank you for insightful review. The previous expression of our idea in passage may be a little ambiguous. So, the passage was re-phrased as follows: “They thought the relatively strong back extensor muscles made the spine hyperextended to maintain upright posture, and gave chance to stand and walk. Thus, the corrective for hyperlordosis wasn’t recommended due to the possible damage to compensatory mechanism by back extensor muscles.” (Discussion, line 8, page 4)

(2) Question: The major topic of your paper is the speculation about the indication of the method (suitable for which patients? only the wheelchair-bound?)

Response: Our conclusion in manuscript, “Corrective surgery for spinal deformity, such as hyperlordosis and scoliosis, could do some help in posture and improve the quality of life especially in wheelchair-dependent patients”, is deduced based on our one case with compared to another one case reported by Lee et al. The indication of corrective surgery for hyperlordosis in FSHD patients may not be easily concluded based on only two cases. However, the experience from our case can give us inspiration that the wheelchair-dependent FSHD patients may benefit from the corrective surgery for hyperlordosis. And further investigation with more patients is needed to clarify this indication. We hope that this explanation could make this issue more clear.

(3) Question: Further suggestions for minor revisions:

Response: It is very kind of you to point the improper expressions in this manuscript and give so detailed suggestions to me. Thank you for your careful review. Here are modified details following your advices.

Abstract, l4: "can't" -> cannot. Then start a new sentence: "While surgical treatment usually is neither reported nor recommended in previous studies, we report the first ...

Revision: Hyperlordosis is common in facioscapulohumeral muscular dystrophy (FSHD), which cannot be controlled by bracing. While the surgical treatment usually is neither reported nor recommended in previous studies, we report the first corrective surgery for hyperlordosis in one wheelchair-dependent FSHD patient. (Abstract, line 4, page 2)

Abstract, l17, and Conclusion, l32: "under controversial" -> "under controversy" OR "controversial". Then start a new sentence: "We report the first successful case of operative treatment by corrective spine surgery in these rare and demanding patient collective..."

Revision: The correction for hyperlordosis in FSHD is controversial. We report the first successful case of operative treatment by corrective spine surgery in these rare and demanding patient collective. (Abstract, line 13, page 2) (Conclusion, line 24, page 4)

Background, l1, and Conclusion, l29: If you refer to FSHD as the "third" form, you have to comment on the other forms. Otherwise just say "... is a rare form of ..."
Facioscapulohumeral muscular dystrophy (FSHD) is one of the most common forms of muscular dystrophy with prevalence of 1/20000 to 12/100000, (Background, line 2, page 3) (Conclusion, line 22, page 4)

Although FSHD progresses slowly, about 20%-36.9% patients need the wheelchair during the rest of their life. (Background, line 7, page 3)

Hyperlordosis is common in FSHD, which results in weakness of pelvic extensor and paraspinal muscles. (Background, line 9, page 3) (Discussion, line 2, page 4)

However, corrective surgery is not reported and even not recommended in patients from previous studies. (Background, line 11, page 3)

We report the first corrective surgery for hyperlordosis in one wheelchair-dependent FSHD patient, who shows improvement in posture, (Background, line 13, page 3)

Indicating that correction for hyperlordosis can help to improve posture and quality of life for wheelchair-dependent FSHD patients. (Background, line 14-15, page 3)

She had been wearing the brace for more than 4 years (20 hours/day) (Case presentation, line 20, page 3)

Case presentation, l30: bilateral inability to completely close the eyes, ...

Discussion, l6: "isn't" -> is not

But corrective surgery is not introduced, (Discussion, line 5, page 4)
Discussion, l15: "more powerless" -> weaker

Revision: Unlike the patient reported by Lee, the back extensor muscles of this girl are weaker due to the deterioration of FSHD and fail in maintaining compensatory mechanism. (Discussion, line 12, page 4)

Discussion, l22: What is "ritual life"?

Revision: which has obvious influence on the daily life (Discussion, line 16, page 4)

Discussion, l22: "So, the correction is necessary because the rigid internal system could hold the upright sitting posture in consideration of none powerful paraspinal muscles available" please re-phrase: what is the "internal system"? can it be "in consideration"? "none powerful" is "powerless" or "weak" or "insufficient"

Revision: So, the correction is necessary because the rigid internal fixation system could hold the upright sitting posture, while the paraspinal muscles were weak. (Discussion, line 16-18, page 4)

Reviewer #2 (Professor Samadian):

Question: The authors report a good example of hyperlordosis treatment in a known disease. They corrected the hyperlordosis and explained the result of surgery. As a case report its good for publication, but for decision making and case selection of this type of corrective surgery we need more case and long term follow up of the patients.

Response: Thank you for your positive review on our case. As your opinion said, the decision making and case selection of corrective surgery for FSHD patient needs more cases and long term follow-up. We totally agreed with it and it will be our target to investigate further for providing more information available for reference.

Many thanks to you, Prof. Allaix, and the two reviewers, Prof. Biber and Prof. Samadian, for the time and efforts spent on this paper. I sincerely hope this revised manuscript will be accepted for publication.