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Jin-Lei Wang
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Dear Editor,

Thank you very much for giving us the opportunity to revise our manuscript. We sincerely thank you and the Reviewers for your positive and constructive comments on our manuscript entitled 'Surgical treatment of atlantoaxial dysplasia and scoliosis in spondyloepiphyseal dysplasia congenita: A case report' (Manuscript NO.: 87238, Case Report).

We have carefully considered your thoughtful comments and have made revisions according to the editor and reviewer's suggestions. All revisions are highlighted with yellow color in the revised manuscript. We believe that your comments are of vital importance and have significantly improved our manuscript. Hereby we re-submit the revised manuscript as well as a detailed point by point response to the comments.

We would like to express our great appreciation to you and the Reviewers for your patience and support during this process.

Thank you and best regards.

Yours sincerely,
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Response to Reviewer

Dear professor, thank you very much for reviewing our manuscript and giving us your constructive comments and suggestions. Here are our responses to your comment point-by-point:

1."…It is a well written case report concisely explaining about the topic."

- **Response:** We are deeply grateful for your positive comments and invaluable suggestions, which have significantly contributed to the improvement of our manuscript. Following your constructive feedback, we have undertaken a thorough revision of the content and conducted further language refinement to uphold the manuscript's academic excellence. Your patience and advice have been immensely beneficial, and we are sincerely thankful for your continued support.

2."…Authors may consider including previous x-ray and compare the previous Cobb angle to the current Cobb angle status of the patient."

- **Response:** Thanks for your constructive suggestion. We contacted the patient and their family and added the spine X-ray from their first hospital visit to Figure 1A, following your invaluable advice. But it is a pity that the imaging film before the patient came to our hospital can no longer be found.
- Upon comparing the spine X-ray from the initial visit with the one obtained three months post-atlantoaxial surgery, we observed an increase in both the proximal thoracic curve (from 32° to 36°) and the main thoracic curve (from 62° to 65°). This trend is described in the discussion section. Additionally, the progressive aggravation of scoliosis played a substantial role in our decision to prioritize scoliosis correction surgery ahead of hip arthroplasty, a consideration elaborated upon in the discussion section. We have highlighted the relevant portions in yellow color in the revised manuscript.

3."…Authors should include bending/traction x-ray films. These would have been done to evaluate UIV and LIV."

- **Response:** Thanks for your comment. We have included Bending X-ray films (Figure 1D-E) in the manuscript.
- The selection of the upper and lower instrumented vertebrae (UIV and LIV) was a crucial aspect of the pre-operative planning. The Bending X-ray results confirm that both the proximal thoracic curve and the main thoracic curve are structural curves, necessitating the inclusion of both curves in the surgical plan[1]. Considering the patient's balanced preoperative shoulder alignment, we have chosen the UIV at the T3 level. However, in situations where there is a notable left shoulder elevation before surgery, UIV at T2/T1 may be considered.
- As for the LIV, the center sacral vertical line (CSVL) passed between the pedicles of L4 in the bending X-ray. However, CSVL did not intersect with the pedicle of L3, so we opted to select L4 as the LIV. For these patients with syndromic scoliosis, due to the influence of the primary disease, we usually choose a relatively 'conservative' range of surgery compared to adolescent idiopathic scoliosis. The selection of UIV and LIV in this case is also described and marked yellow color in the discussion section.

4."…Authors may explain the controversy of operating scoliosis or hip dysplasia first."

- **Response:** Thanks for your invaluable suggestion. During the initial visit, the patient complained of progressive scoliosis for 5 years. Additionally, the patient presented with concurrent issues of atlantoaxial dysplasia and hip dysplasia. Even though the patient has severe bilateral hip joint damage, the presence of neurological damage due to atlantoaxial dysplasia makes it crucial to prioritize saving neurological function. Additionally, it was observed that the Cobb angle of scoliosis was still increasing three months after atlantoaxial decompression and fixation surgery. To avoid the potential need for more extensive surgery if scoliosis worsens, the decision was made to undergo scoliosis correction surgery before hip replacement surgery.
- Furthermore, based on both X-ray and three-dimensional CT data, the patient exhibits relatively good pelvic balance after the atlantoaxial surgery. Consequently, the priority of hip surgery for enhancing pelvic balance is relatively low, contributing to our decision to address scoliosis as the initial treatment. We added this part to the discussion section and marked it as yellow.

5."…Since the case report includes cervical myelopathy as presenting complaints, the author may include surgical steps, follow-ups and details about C1-C2 fixation."

- **Response:** Thanks. In accordance with your constructive comments, we have added details and follow-ups on C1-2 surgery to the revised manuscript (in 'TREATMENT' section and 'OUTCOME AND FOLLOW-UP' section).
- In atlantoaxial decompression and fixation surgery, skull traction was implemented under anesthesia during the surgical procedure, and the atlantoaxial reduction was observed to be satisfactory. Consequently, one-stage posterior atlantoaxial reduction and fixation procedure was employed. During the procedure, lateral mass screws were placed on both sides of the atlas and pedicle screws were placed on both sides of the axis and fixed with connecting plates, and autogenous iliac cancellous bone were used for bone graft fusion.
- Three months after the atlantoaxial decompression and fixation surgery, the compression of the cervical spinal cord was significantly relieved, and the instability of the atlantoaxial joint was improved (Figure 4). The patient's muscle strength of bilateral proximal lower limbs was significantly improved to grade V. The Hoffman signs and Babinski signs were negative.
- At the postoperative 2-year follow-up, the patient reported no symptoms of discomfort, such as limitation of neck movement or weakness of the lower extremities, the atlantoaxial joint reduction remained effective and spinal correction retained its stability (Figure 6E-H).

- We would like to express our gratitude once again for your valuable suggestions.

References

1. Lenke LG, Betz RR, Harms J, Bridwell KH, Clements DH, Lowe TG et al. Adolescent idiopathic scoliosis: a new classification to determine extent of spinal arthrodesis. J Bone Joint Surg Am 2001; 83: 1169-1181 [PMID: 11507125 DOI: 10.2106/00004623-200108000-00006]