Author’s response to reviews

Title: Recurrent femoral shaft fractures in a child with gnathodiaphyseal dysplasia: a case report

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Author’s response to reviews:

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Sebastian Farr, M.D.
Editor-in-Chief
BMC Musculoskeletal Disorders

Dear Dr. Farr:

We sincerely appreciate the opportunity to submit the revised version of our manuscript titled: “Recurrent femoral shaft fractures in a child with gnathodiaphyseal dysplasia: a case report” (BMSD-D-18-01696) to BMC Musculoskeletal Disorders.
We would like to thank the reviewers for their valuable comments on our manuscript. We have revised the manuscript in accordance with the reviewers’ comments and have provided point-by-point responses.

We hope our revision will meet the quality requirements for publication in BMC Musculoskeletal Disorders.

Thank you again for your support. We look forward to your reply.

Sincerely,

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Response to Reviewers

Manuscript Number: BMSD-D-18-01696

Title: Recurrent femoral shaft fractures in a child with gnathodiaphyseal dysplasia: a case report

Response to Dr. Lars Helbig (Reviewer 1)
We sincerely appreciate your review of our manuscript and for the comments providing insight into scope for improvements. We have revised the manuscript thoroughly and have added supplementary information as suggested by your comments.

1. the authors should show all X-rays in two planes

Response: We have added the lateral plane images for all figures, except figures 5 and 6, for which lateral radiographs were not available.

2. the two fractures at half-pin insertion sites are not indicated. All fractures should be indicated by arrows.

Response: We added a new figure that shows the two fractures at the half-pin insertion sites. Furthermore, we have added arrows indicating the fractures.

3. In my opinion the supracondylare fracture was not addressed with the Ender nail. Could the authors explain their procedure in more detail?

Response: We totally agree the reviewer’s opinion that the Ender nail was not indicated for the supracondylar femoral fracture. The supracondylar fracture occurred 9 weeks before the revision surgery with the Ender nail. At the time of the Ender nail insertion, the supracondylar fracture site was totally asymptomatic. We considered that this fracture was healing, and that additional internal fixation was unnecessary, although the risk for refracture at that site was high. We therefore prioritized the diaphyseal fracture, which was the indication of the Ender nail.

We added a more detailed explanation about the supracondylar fracture and indication for the Ender nail as follows: “After this operation, the patient sustained a refracture (33-D/4.1 in AO-PCCF) at the same fracture site, followed by a supracondylar fracture (33-M/3.1 in AO-PCCF) at a distant site of the femur (Figure 5) and two consecutive fractures at the half-pin insertion sites (Figure 6). The supracondylar fracture occurred without any triggering activity before beginning weight-bearing exercise. The supracondylar fracture was successfully treated conservatively, but she sustained two more consecutive diaphyseal fractures (33-D/4.1 and 33-D/4.2 in AO-PCCF)
at the half-pin insertion sites (Figure 6). She eventually underwent a revision surgery for the diaphyseal fractures with an Ender nail (Ender nail®, MIZUHO Co., Ltd., Tokyo, Japan).” (Case presentation section, line 107-115, page 6).

We would like to thank you again for your valuable comments on our manuscript. We hope that the revised manuscript is now suitable for publication.

Response to Dr. Sebastian Farr (Reviewer 2):

We wish to express our sincere appreciation for your valuable comments regarding our paper and for providing suggestions that helped us improve the manuscript significantly.

1. Page 4-line 34: what do you mean by "sporadic"? non-hereditary?

Response: We changed the word “sporadic” to “non-hereditary” as suggested (Case presentation section, line 84, page 5).

2. Page 4-line 45-46: did you consider bisphosphonate therapy for this entity for bone improvement? are there any recommendations in the literature?

Response: We appreciate the reviewer’s comment on this point. Although we tried to start bisphosphonate therapy, the patient’s mother did not provide consent.

We have added the following text to the Discussion and Conclusion (Page 7, line 1-12) as:

“To prevent recurrent fractures, pharmacological therapy for bone fragility may be considered. Among OI patients, bisphosphonate therapy is currently the most common medical treatment and has been reported to decrease the incidence of long-bone fractures [27]. On the other hand, little is known about medications for patients with GDD. Ghada et al. reported a case of GDD treated with a bisphosphonate [1]. They demonstrated that the bisphosphonate therapy did improve BMD of the spine. However, no apparent benefit was observed in terms of frequency or severity
of fractures. We proposed bisphosphonate therapy for our patient but could not obtain the caregivers’ consent. They were mainly concerned about the potential adverse effects, particularly bisphosphonate-related osteonecrosis of the jaw, as the patient had GDD-associated lesions in the jawbone, which had required multiple surgical interventions. Therefore, further studies are needed to clarify the role of pharmacological therapy for GDD patients.”

Furthermore, we added reference [27] accordingly.

3. Page 4-line 51: please provide a detailed AO classification for your fractures

Response

In accordance with the reviewer’s comment, we have added AO pediatric comprehensive classification (AO32-D/4.1, 32-D/4.2 and 33-M/2.1) to the case presentation section. Furthermore, we have added reference [5] accordingly.

4. Page 5-line 7-11: were all these fractures one after another or at once, e.g. on one day? please clarify

Response

We appreciate the Reviewer’s comment on this point. We revised this part to clarify the timeline of the fracture as: “After this operation, the patient sustained a refracture (33-D/4.1 in AO-PCCF) at the same fracture site, followed by a supracondylar fracture (33-M/3.1 in AO-PCCF) at a distant site of the femur (Figure 5) and two consecutive fractures at the half-pin insertion sites (Figure 6). The supracondylar fracture occurred without any triggering activity before beginning weight-bearing exercise. The supracondylar fracture was successfully treated conservatively, but she sustained two more consecutive diaphyseal fractures (33-D/4.1 and 33-D/4.2 in AO-PCCF) at the half-pin insertion sites (Figure 6).” (Case presentation section, line 6-12, page 4)

5. Page 6-line 14: did you use normal or hydroxylapatite-coated pins?

Response

We used non-hydroxylapatite-coated normal pins. We have added the word “non-hydroxylapatite-coated” in the case presentation section (Page 5, line 57).
6. Page 7-line 1ff: I am not sure whether the authors use the wording "FIN" in the correct meaning. FINs or ESINs should be used for femur fractures in children. However, for weak bone diseases such as OI or also GDD, rigid nails such as rushpins are usually recommended for increased stability. The authors should elaborate some more on this topic. I am not sure whether flexible nails can provide enough stability for this entity. Moreover, elongating nails such as Fassier-Duval for OI are not really flexible - they are rigid nails.

Response

We appreciate the reviewer’s comment. We totally agree the reviewer’s opinion that a flexible intramedullary nail provides less rigidity and is not an ideal fixation method for patients with brittle bone diseases. In this case, no solid nails that could cover adequate lengths of the femur were available in our country; therefore, we chose the Ender nail. Moreover, there might be confusing statements about the implication of FIN and rigid nails.

To make this point clear, we revised the whole paragraph as follows: “The IM device is also used as a first-line treatment of long-bone fractures among patients with osteogenesis imperfecta (OI), which is a systemic osseous fragility disease similar to GDD [22]. For patients with OI, more rigid IM devices are preferred to obtain stability. Two types of IM devises are used for fractures in OI: elongating and non-elongating rods. Elongating rods have self-extending designs to follow bone growth. Recently developed elongating rods, such as Sheffield and Fassier–Duval medullary rods, have become the standard treatment in patients with OI [22]. El-Adl et al. reported that elongating rods are better than non-elongating rods, such as Kirschner wires or rush pins, with regards to mobility status, longevity, and incidence of complications requiring reoperations [23]. In a previous report on a case of GDD, although the authors did not provide the description in detail, they mentioned that the patient was treated using elongating rods (Fassier–Duval medullary rods) [24]. Given that recurrent refractures in our patient were a result of treatment with EF, we recommend IM devices as the first choice for the treatment of femoral shaft fractures in GDD patients. In this case, we utilized the Ender nail as an IM device because elongating rods are not commercially available in our country and because other non-elongating IM devices such as rush pins and Kirchner wires only have limited size variations, which were not able to cover adequate lengths of the femur in our patient. It remains unclear whether fixation with a single Ender nail provides enough stability for patients with GDD. Considering the favorable results in OI, rigid and elongating rods might be a better option than non-elongating IM devices.” (Discussion and Conclusion section, line 174-193, page 8-9)

Thank you again for your valuable comments on our paper. We hope that the revised manuscript is now suitable for publication.