Author's response to reviews

Title: Treatment of scoliosis in patients affected with Prader-Willi syndrome using various different techniques

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Author's response to reviews: see over
Cover letter giving a point-by-point response to the concerns.

Referee no. 1 Shimizu Katsuji

Thanks for you comments.

Referee no. 2 Seung-Woo Suh

Regarding the little number of patients, the study refers to, the whole available scientific literature report a number of patients lower than 50 (patients surgically treated for scoliosis. Generally speaking the literature dealing with scoliosis surgery in PWS consists of only few case reports. The cases reported by Accadbled et al. were operated on in 3 various different centres (Spine (Phila Pa 1976). 2008 Feb 15;33(4):394-401; Complications of scoliosis surgery in Prader-Willi syndrome; Accadbled F, Odent T, Moine A, Chau E, Glorion C, Diene G, de Gauzy JS.), whereas the six reviewed by the current authors all underwent surgery in the same centre. Since this disease is classified as rare and the authors have dealt with the surgical treatment of scoliosis associated with this pathology, the number of cases is quite above the average (some scientific works refer only to one case, see Spine J. 2007 May-Jun;7(3):345-8., Scoliosis associated with Prader-Willi syndrome, Yamada K, Miyamoto K, Hosoe H, Mizutani M, Shimizu K.).

The phenotypic features of the Prader Willi Syndrome is quite different from that of neuromuscular scoliosis; risks are strictly related to hypotonia and obesity of patients who presents with compulsive forms of behaviour entailing difficulties in their postoperative management. The characteristics of each patients were detailed because PW scoliosis cannot be classified as idiopathic scoliosis (Lenke, King, etc.). Bracing cannot control this type of deformity; however, surgery is associated with a higher risk of complications. In our opinion, it is not fair to consider these deformities as Duchenne scoliosis and muscular dystrophy, since in spite of the elements in common – such as hypotonia and osteoporosis – PW patients have expectations and quality of life which are different from the other subjects. In fact, their Body Mass Index is often high and, although hypotonic, they are not wheelchair-bound and can walk and often perform compulsive movements. Therefore, they need immediate stability of the spine which can be obtained through the surgical technique described. Such a technique is associated with high rate of complications, related to the specific features of this syndrome. The purpose of our article was to help the surgeon to recognize and consider these features.
Some minor language reviews were performed and highlighted in the text.

Referee no. 3 Toru Maruyama

In Table I, the authors have added preoperative and postoperative Cobb angle of primary scoliosis, in addition to correction rate.

Regarding the choice of the instrumentation, the referee is right when he says that the first case (hybrid instrumentation) was highly successful; however, the worst case of the present series was the second, where the same type of instrumentations was applied. In any case, conclusions have been modified according to the suggestion and feeling that the final choice should always be made considering all the of the peculiar features of the patients.

Case no. 4: no lengthening was performed, as the growing rod technique would require, since it was not possible to reach an agreement with the patients’ parents. However, if the referee consults the authors’ website, he can see a more recent case affected with PWS, who is being treated following the technique standard procedure, i.e. periodical lengthening until final fusion. This patient, being too recent, is not included in the current series. The authors do know this technique and agree with the referee’s suggestion.

Page 13, line 14: Two reinterventions were performed: one on account of mobilization (case no. 6), and the other to achieve final fusion after applying the growing rod technique (case no. 4). Text has been amended.

English revisions have been performed.

The authors would like to thank this referee for his helpful comments; being an Institute with scientific purposes, scientific research is part of our daily activity.