Reviewer’s report

Title: Rare Causes of Scoliosis and Spine Deformity: Experience and Particular Features

Version: 1 Date: 13 April 2007

Reviewer: ATHANASIOS TSIRIKOS

Reviewer’s report:

Manuscript title: “Rare causes of scoliosis and spine deformity: experience and particular features”.
Reviewer’s recommendation: Accept for publication following major compulsory revisions
Reviewer’s comments

A). General comments.
This is a case series of patients with spinal deformities due to the presence of uncommon underlying syndromic conditions. Syndromic or miscellaneous scoliosis constitutes the forth type of structural scoliosis (with idiopathic, congenital and neuromuscular being the other 3 types), as described previously in the literature and in accordance with the classification introduced by the SRS.
Even though there are previous reports, where patients with individual syndromic disorders have been investigated in relation to the development of a spinal deformity and the specific treatment considerations (for example patients with Marfan’s syndrome, osteogenesis imperfecta, Rett’s syndrome), there is limited published data on the assessment and management of a mixed group of patients with such conditions originating from a single institution. This can be considered a strong point of the current report.
By definition this group consists of patients with very diverse underlying diagnosis and clinical features, which have significant implications in the development and the management of their spinal deformity. Therefore, no uniform conclusions can be extracted from the investigation of a mixed group of patients with such conditions other than the fact that a multidisciplinary approach is required during the detailed preoperative assessment but also the intensive perioperative management of these patients, in order to minimise the associated medical and surgical risks related to the surgical correction of their spinal deformity and achieve an optimum outcome. The individual characteristics of the underlying syndromic condition, which commonly affect the type, the natural history, as well as the severity of progression of the spinal deformity, have to be taken into consideration and need to be addressed meticulously to reduce the risk of intraoperative or postoperative complications, which can be occasionally life threatening.
It is the reviewer’s opinion that this is a message that needs to be given more emphatically both in the text and in the abstract, as it represents the only conclusion that the paper can offer in the existing literature, which already discusses all these conditions separately in previously reported case series or case reports.

B). Specific comments.
1. In the first paragraph of the abstract (background), scoliosis (not spine deformity) should be divided into idiopathic, congenital, neuromuscular and syndromic or miscellaneous. The reviewer would also suggest that the type of scoliosis or spinal deformity reported in the present study would be much better described if the term syndromic or miscellaneous was being used in the text to characterize the whole group of patients.
2. In all case presentations where the patients underwent surgical treatment, the postoperative outcome should be discussed in more details (for example: what was the % of deformity correction, postoperative scoliosis angle and sagittal balance of the spine, possible specific intraoperative technical complications if they occurred). The length of clinical and radiographic follow-up should also be described, as well as the final outcome regarding spinal fusion, residual or recurrent spinal deformity, prevalence of pseudarthrosis, instrumentation related complications, late wound infections, survivorship of the patients.
3. The reviewer considers that it would add to the description and better understanding of the individual cases in the text, if preoperative and postoperative (AP) and lateral radiographs of all reported patients were included. If the authors have also obtained preoperative and postoperative clinical photographs of these patients, I would welcome their appearance in the paper. Subsequent to the figure changes, the figure legends should also be adjusted.
4. In the description of case 6 (page 9), the authors would have to specify what “prophylactic chemoprophylaxis” was utilised during surgery.
5. In the description of case 9 (page 10), I would not agree with the authors’ statement that correction of the lumbar hyperlordosis would result in further anterior spinal decompensation. As the authors describe, in this group of patients the presence of a significant anterior pelvic tilt due to severe weakness of the hip extensors results in the development of an increased compensatory lumbar lordosis, as an attempt of the spine to balance. Patients with FSH muscular dystrophy commonly lose gradually their ambulatory function.
and become confined on their wheelchair with poor sitting balance due to a fixed lumbar hyperlordosis. In these patients, surgical correction of the spinal and pelvic deformity in the form of a combined anterior and posterior spinal fusion with anterior lumbar multilevel closing wedge osteotomies and fusion of the spine to the sacrum with pelvic fixation can improve significantly their posture, as well as their sitting balance. I would, however, agree that in the patient described as case 9 and based on the clinical information provided, surgical treatment would not improve her level of function and, therefore, the decision taken was correct.

6. In the description of case 10 (page 11), it would have been useful if the authors could explain what was the medication treatment for the patient with the dystonia once it was decided that surgical treatment was not appropriate, especially since this was a dopa non-responsive dystonia.

7. At the beginning of the discussion (page 11), an introductory section to discuss spinal deformities related to syndromic disorders would have been useful before the individual conditions are presented separately.

8. In page 12, 1st paragraph the sentence to read: “Congenital spine deformities have also been reported” needs to be referenced.

9. In page 15, paragraph 1, line 3 “anterior” should be changed to “posterior”.

10. In page 16, paragraph 2 “functional spine deformity” is better described as “non-structural or postural spinal deformity”.

11. The figures are labelled incorrectly and have to be changed: Figure 3 is figure 3A, figure 4 is figure 3B, figure 5 is figure 4A, figure 6 is figure 4B, figure 7 is figure 4C, figure 8 is figure 5, figure 9 is figure 6, figure 10 is figure 7A, figure 11 is figure 7B.

Thank you very much for giving me the opportunity to review this paper for Scoliosis.

Yours sincerely,

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What next?: Accept after minor essential revisions

Level of interest: An article of limited interest

Quality of written English: Needs some language corrections before being published