Reviewer’s report

Title: A Dutch Guideline for the Treatment of Scoliosis in Neuromuscular Disorders

Version: 2 Date: 27 July 2008

Reviewer: ATHANASIOS TSIRIKOS

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Paper Title: “A Dutch Guideline for the Treatment of Scoliosis in Neuromuscular Disorders”

Reviewer’s Comments

I would like to thank you very much for the opportunity to review this article. I have been very interested to read this paper, which addresses a topic that is of particular importance to spinal deformity surgeons and applies to our everyday practice. Even though there have been significant advancements on the treatment of patients with neuromuscular conditions who develop spinal deformities over the past few decades, there is still no general consensus on several issues surrounding the approach to these patients and controversy still exists around the indications for surgical management. Therefore, a paper which attempts to address these issues and aims to formulate an agreed guideline across the multidisciplinary group involved in the management of such patients should be of considerable interest.

Overall, I feel that the paper is well reasoned and balanced across the authors’ own views but also any published data in the literature. The authors have covered in sufficient detail previous reports on the different aspects of their topic and I did not feel that there was any important reference missing. The writing format of the text is satisfactory and I have only a few corrections to make. I would, therefore, recommend that this paper should be accepted for publication in Scoliosis after minor but essential revisions, which I am covering in details below.

Comments:

1. The title of the paper is slightly misleading in the sense that when most authors refer to neuromuscular scoliosis in general, they include disorders affecting the central nervous system, such as cerebral palsy, which is by definition the most common neuromuscular condition causing permanent disability in children and adolescents, as well as myelomeningocele (spina bifida). I can appreciate the resemblance in approach for patients with DMD and SMA where the generalised muscle weakness and the similar pattern of clinical presentation allows for a more direct comparison, as well as easier extracted conclusions which can apply to guidelines compared to, for example, patients with CP where the pathology and pattern of presentation are different. Therefore,
I do not find it unreasonable to confine the study to only patients with DMD and SMA and extrapolate these conclusions also to patients with more rare forms of congenital myopathies. However, I would recommend that, in this case, the title of the paper should be changed to represent accurately the groups of patients included in the study and this could be: “A Dutch Guideline for the Treatment of Scoliosis in patients with Duchenne Muscular Dystrophy and Spinal Muscular Atrophy”, or alternatively “A Dutch Guideline for the Treatment of Scoliosis in patients with Myopathic Conditions”.

2. On Page 7, 3rd paragraph, under Recommendation, the authors suggest that “the vital capacity is a possible indicator of the progression of scoliosis”. In my view, they should make a comment to clarify that decline in vital capacity is, however, expected to occur as part of the natural history of the condition in patients with DMD and/or SMA and this is not necessarily related to the development of scoliosis. This statement is additionally supported by the fact that surgical stabilisation of the spine and correction of the scoliosis does not influence further decline in respiratory function which is expected to occur as part of the natural process of the disease.

3. On Page 8, 1st paragraph, under Evidence, last sentence, in my view the authors should also add a statement to say that a spinal brace, especially if this is rigid, can potentially restrict chest movements and affect the patient’s respiratory function with detrimental consequences. In my opinion, for the few of these patients where trunk support is required and surgical correction cannot be performed, posture improvement could be easier and safer achieved by seating adaptations built on the patient’s wheelchair rather than the use of a spinal orthosis. If the authors agree, it might be helpful to include this statement in their recommendations.

4. On Page 9, under Steroid Treatment, I would expect the authors to refer to the associated problem of osteopenia that these patients inherently have but which is also induced by the corticosteroid management. I would suggest that the authors could include their recommendations for bone quality assessment through a bone mineral density scan, as well as their suggestions for treatment of a diagnosed osteopenia and osteoporosis possibly with bisphosphonates, especially in view of the need for scoliosis surgery and the importance of reducing the risk of pseudarthrosis.

5. On Page 12, 1st paragraph, under Recommendation, I would suggest that the authors could include their recommendations on the postoperative management of these patients in regard to their associated respiratory compromise. For example, in my practice, this would include often elective endotracheal ventilation for 24-72 hours depending on the severity of preoperative respiratory dysfunction, frequent use of non-invasive ventilation following extubation and cough assist machines in patients with an ineffective cough, intensive respiratory physical therapy, as well as early mobilisation of the patients on a reclining wheelchair with the aim to reduce the risk of pulmonary infection. I believe that this is the only part of the paper where such recommendations could be presented since guidelines on respiratory management are not included further down in the text.
6. On Page 12, 3rd paragraph 3, under Discussion, I would welcome the authors’ guidelines on whether severe ventricular insufficiency in patients who are already on cardiac medication would be considered in their suggestions as a contraindication for scoliosis surgery. Do the authors have a specific cut-off for cardiac impairment beyond which surgery would be contraindicated? Unfortunately, the current literature does not provide an answer to this question and I think that it would be interesting to present the working group’s views.

7. On Page 13, 2nd paragraph, under Recommendation, do the authors have any suggestions on the need for nutritional supplementation perioperatively in patients with DMD and SMA who are usually malnourished which would decrease primarily the risk for infection? In addition, in patients with severe associated hip deformity and a scoliosis with pelvic tilt, which pathology would they recommend to correct first: the scoliosis including correcting the pelvic obliquity and fusing to the sacrum or the hip displacement? It would again be very interesting to present the working group’s consensus.

8. On Page 13, 3rd paragraph, under Evidence, I would expect the authors to mention also the need for hypotensive anaesthesia in order to reduce blood loss intraoperatively, as well as to discuss the use of blood preservation techniques and present their experience and recommendation. In general, the collective experience of such a multidisciplinary group of clinicians and investigators would be of exceptional value to the audience who are treating patients with such conditions. Therefore, wherever possible I would encourage the authors to include their group opinion, especially in regard to aspects of management where the literature does not provide valid information.

9. On Page 14, 1st paragraph, under Evidence, I would be interested to read the authors’ experience on initial anterior release and fusion as a supplementary procedure to increase flexibility of the curve, for example in patients with fixed severe lumbar scoliosis and rigid pelvic obliquity, and before a posterior instrumented arthrodesis is performed. Do the authors have clear indications and recommendations for anterior surgery in this group of patients?

10. On the same paragraph, I think that the authors should mention that fusion to L5 in this group of patients who have limited arm function, which allows mobility of the lumbosacral junction can be performed in patients with minimal pelvic obliquity and, in addition to the advantages already mentioned in the text, this could preserve at least to some extent the ability for independent postoperative feedings.

11. On the same section of the text, 2nd paragraph, where the authors discuss application of growing rods for these deformities, I would suggest that they mention the significant risks associated with the repeat lengthenings that are usually performed every 6 months (serial anaesthesics, instrumentation problems, wound infections) for such a medically compromised group of patients.

12. On Page 15, 1st paragraph, under Recommendation, I would welcome the authors’ comment on whether they feel there is any role for the use of BMPs, especially in revision cases with a documented non-union.

13. On Page 16, last paragraph, how soon after scoliosis surgery and how often
thereafter would the authors recommend monitoring of the patients’ cardiorespiratory function?

14. On Page 19, 5th paragraph, under Discussion/conclusion, I would suggest that the authors include a statement to say that postoperative bracing could be detrimental to the respiratory recovery, especially at an early stage following surgery when the pulmonary function is significantly reduced.

Corrections on writing format:
1. Abstract, 1st paragraph, “…and is perceived as unaesthetic”, what do the authors mean? Please correct.
2. Introduction, 1st paragraph, “Usually contractures…present”, does not read well. This could be changed to: “Usually joint contractures, as well as nutritional disorders are present”.
3. Page 3, line 12, change “live” to “life”.
4. Page 3, line 15, “and revalidation” should be omitted.
5. Page 5, line 21, “tracheostomy” should be corrected.
6. Page 5, line 38, “heart” should be corrected.
7. Page 11, last paragraph, “lung function” should be corrected. This mistake is repeated in the following 2 paragraphs.

Thank you again for the opportunity to review this manuscript.

Kind regards,

Yours sincerely,

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**Level of interest:** An article of importance in its field

**Quality of written English:** Acceptable

**Statistical review:** No, the manuscript does not need to be seen by a statistician.