Reviewer's report

Title: Natural course of scoliosis in proximal spinal muscular atrophy type II and IIIa. Descriptive clinical study with retrospective data collection of 126 patients

Version: 2 Date: 8 July 2013

Reviewer: Matthew E. R. Butchbach

Reviewer's report:

This manuscript provides a retrospective analysis of the natural course of scoliosis in type II and type IIIa SMA patients. It is indeed an interesting study but there are a couple of items which must be addressed.

Major Compulsory Revisions

1. In this manuscript, the data are presented in both tabular and graphical formats. As I see it, this presentation is redundant. Table 2 = Figures 1 and 2; Table 3 = Figure 3. Please present these data as either tables or graphs.

2. If the authors elect to present these data as graphs, then please use bar graphs instead of line graphs as line graphs imply repeated measures of the same set of patients (which I do not believe is the case here). Also, please be sure to include error bars (standard error).

3. There is no mention of statistical analysis in the methods sections. Please describe how you completed your statistical analysis.

4. In the methods, it is mentioned that the diagnosis of SMA was "confirmed either genetically or by muscle biopsy and neurophysiology." Why were all of the subjects’ diagnoses not confirmed genetically given that 1) there is a reliable genetic test for proximal SMA (SMN1 and SMN2) and 2) there are multiple types of SMAs with different genetic causes (i.e. SMN1 for 5q proximal SMA, DYNC1H1 for SMA-LED, AR and UBE1 for X-linked SMAs).

5. In those patients whose diagnoses were confirmed genetically, was there a correlation between SMN2 copy number and the severity of scoliosis/pelvic obliquity/relative vital capacity?

6. Are there relationships between sex and a) the type of scoliosis observed, b) maximum curvature, c) pelvic tilt and d) relative vital capacity?

Minor essential revision

1. In the figure and table legends, the authors describe their measures in patients "not operated on". The wording is difficult to read. Please reword.

Level of interest: An article of importance in its field

Quality of written English: Acceptable

Statistical review: Yes, and I have assessed the statistics in my report.
Declaration of competing interests:

I declare that I have no competing interests.