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:** 21 July 2013

**Reviewer:** Kathryn J. Swoboda

**Reviewer's report:**

The authors provide a retrospective review of 126 patients evaluated at their center with SMA types II and IIIa who had not yet been operated on for scoliosis, to provide a perspective on scoliosis progression, type and severity as well as pelvic obliquity in the two populations, and provide available data on respiratory function concomitant with these changes. While there are certain flaws associated with the retrospective nature of the data collection and the biased sample of patients who were presumably already noted to have scoliosis, this is an important study for the main reason that the authors have stated, that limited data has been published on patients with SMA and scoliosis.

A few suggested edits would improve the paper further (compulsory revisions):

Compulsory revisions
1) It is important to know over what period of time these data were examined, and what methods they used to ensure that all patients evaluated during that time period were included.
2) Similarly, it would be of value to know how many patients were excluded because they had already had surgery, if possible.
3) The mean age of sitting independently was surprisingly late for their type 2 cohort, indicating they likely had a biased cohort of patients, referred to their group for evaluation due to the fact that scoliosis had already developed. This is to be expected, but the authors should put this caveat to highlight weaknesses in methodology. In point of fact, this is valuable, since delayed sitting may be predictive of the severity of weakness indicating a higher risk for scoliosis.

Strongly suggested additions/revisions:
4) This study highlights the need for a comprehensive strategy to prospectively identify and follow all patients from an early age and monitor them for disease progression; give the expertise of these authors, some suggested guidelines as to when to refer such patients and how frequently to monitor them, particularly with regard to frequency of x-ray evaluation, would be of value.

**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.