

Scoliosis and Prader-Willi Syndrome: Results of the Latest PWSA (USA) Survey

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Introduction: Spinal deformities are estimated to occur in 40-90% of children with Prader-Willi syndrome (PWS). Approximately 15% of all children with PWS will require treatment, either bracing or possibly surgery. To understand the factors influencing these spinal deformities, the membership of the PWSA-USA were queried by a mailed survey.

Materials and Methods: A general survey was mailed to the membership families of the PWSA-USA in 2007. Of the 1603 responses, 639 indicated they had a child with a spinal deformity. A more specified questionnaire was sent to these families, resulting in 210 responses, of which 200 stated that their child had been diagnosed with a spinal deformity. This report summarizes the results of these surveys.

Results: There were 113 patients eighteen years or younger at the time of the survey (date of birth at or after January 1, 1989). In the 0 to 5 years old range, 16% of children had a spinal curve. In the older child, 6 to 18 years, the prevalence was 35%, both for children with uniparental disomy and chromosomal deletion patterns of PWS. Age at curve detection for this group was 5 years old.

There were 87 patients older than 18 years old, 80 indicating they primarily had scoliosis, 7 primarily had kyphosis, for an overall prevalence of 46%. The average age of curve detection was 11 years old. Thirty-four patients required bracing, of which 19 progressed to surgery. Of the 15 who were braced and did not progress to surgery, 8 were on growth hormone (GH), 2 started prior to the diagnosis of a spinal deformity, the other 6 after diagnosis of the curve. Of the 53 patients who did not use a brace, 11 had surgery. Twelve of the 42 patients who were not braced and did not have surgery used GH, 6 started prior to the diagnosis of a spinal deformity, the other 6 after diagnosis. Compliance data was available for 31 of the 34 braced patients – 15 were fully compliant, of which 11 needed surgery; 7 were mostly compliant, of which 2 needed surgery; 7 were sometimes compliant, of which 3 needed surgery, and 2 were never compliant, neither

needing surgery. Six patients used a Milwaukee brace, 19 used a Boston/Wilmington style brace.

Thirty patients of the older-than-18 year old group needed surgery, at an average of 13 years and 10 months. Curve magnitude ranged from 30° to 90°, with many around 70°. Six patients required a second procedure, and one patient required 5 procedures altogether. All had posterior spinal fusions, except one who apparently also had an anterior procedure.

Conclusion: Spinal deformities are very common in children with PWS. Of patients older than 18 years of age 46% were diagnosed with a curve, and of those 52% required treatment (17% bracing, 13% surgery, 22% bracing and surgery). From the data available, we are unable to determine what affect GH has on the development or progression of spinal deformities in children with PWS. We are also unable to determine the efficacy of bracing. Until more data is available, the recommendations should be that patients with PWS be screened yearly for scoliosis with radiographs. Once diagnosed with scoliosis, they should be followed every 3-4 months radiographically. At this time, we still recommend bracing for curves over 20° to 25°, although this may change upon further study. The data at this time does not support discontinuing GH in the face of scoliosis.