

Surgical Treatment of Spinal Deformity in Prader-Willi Syndrome

Harold van Bosse, MD, Stuart Hershman, MD, David Feldman, MD, Alice Chu, MD

Introduction: In children with Prader-Willi Syndrome (PWS), scoliosis or kyphoscoliosis of greater than 10° occurs in 40-90% of patients. Fifteen percent of all patients require bracing or surgical treatment. Bracing is often ineffective because of the underlying hypotonia and difficulty with brace fitting and compliance. Previous series of children with PWS undergoing spinal fusion have reported frequent serious and occasionally fatal complications, related to anaesthesia, hardware fixation, pulmonary compromise, post-operative infections and fixation failure due to osteopenia. The purpose of our study was to review the peri-operative period of a series of children with PWS treated surgically for their spinal deformities, with emphasis on pre-operative assessment, intra-operative and immediate post-operative management, and complications.

Materials and Methods: Between November 2002 and August 2007, a total of five patients underwent spinal surgery. Three (ages 12-18 years) had a primary posterior spinal fusion, one (26 years old) had a revision of a failed posterior fusion, and one (6 years old) had implantation of non-fusion spinal instrumentation, with subsequent lengthenings.

Results: All patients had an extensive pulmonary work-up pre-operatively, including pulmonary function tests and sleep studies. One underwent treatment for sleep apnea (tonsillectomy). For all patients, an average of 14 levels were instrumented. Intraoperative blood loss for the 4 fusion procedures was 1000ml on average, with an average reinfusion 340ml of cell saver. One patient required a single unit of packed cells. All patients were managed post-operatively in a pediatric intensive care unit for an average of 1.4 days (range 1-2), many with BiPAP or CPAP. Hospital length of stay averaged 6 days (range 5-7). All patients were fully ambulatory at the time of discharge. The first patient in this series (the 6 year old) had dehiscence of the wound and a subsequent deep infection. Otherwise, there were no other major intra-operative or post-operative complications. Average follow-up from the index procedure is 22 months. All patients were found to have cervicothoracic kyphosis pre-operatively, and variably post-operatively.

Conclusion: In our series of five patients with PWS treated for spinal deformity by spinal fusion, the single serious complication encountered was a post-operative infection. A thorough pulmonary evaluation is required pre-operatively. Surgical treatment should emphasize segmental fixation, decreased operative time, and avoid an anterior approach. The cervicothoracic kyphosis needs to be recognized, and instrumentation levels chosen appropriately. Post-operatively the patients may need to stay intubated initially, should be managed in a pediatric intensive care unit, and may require BiPAP or CPAP. They should also be inhibited from skin picking the post-operative wound. With such considerations, the surgery can be performed safely with satisfactory results. A better understanding of the underlying syndrome, use of segmental instrumentation and avoidance of anterior procedures, has made surgical treatment safer.