

# Guidelines on Scoliosis Monitoring and Treatment for Children with Prader-Willi Syndrome

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## BACKGROUND

Children with Prader Willi Syndrome have an incidence of developing scoliosis at rates between 40 -90%. Approximately 15% of children with Prader Willi Syndrome will develop severe or significant curves, requiring bracing or surgery. The earlier the curve is detected, the better the possibilities for treating the curve with casting or bracing.

There are two peak ages for scoliosis presentation in children with PWS. Under the age of 4 years, most of the curves are C-shaped, and are most likely related to the hypotonia. The second peak, centered around 10 years of age, typically is the more common idiopathic S-shaped curve. Fifteen percent of curves diagnosed before 4 years of age subsequently required surgical treatment, 41% of curves diagnosed after 4 years of age required surgical correction, as per a PWSA-USA survey of membership. Spinal deformities in children with PWS are often diagnosed late. This delay appears to be due to unique characteristics of spinal deformities in children with PWS, rather than the presence of obesity. Fewer children with PWS now develop obesity, and often the curves are diagnosed prior to the onset of obesity. What seems to be the more important factor is that spinal deformities in children with PWS have less vertebral rotation than seen in other children with scoliosis curves of a similar size. Vertebral rotation causes the asymmetry of the chest wall seen during forward bending, usually the first sign of scoliosis. Therefore, the child with PWS may have a moderate curve radiographically, but only mild findings clinically. For that reason, there should be a much lower threshold for working up clinical findings in children with PWS, compared to otherwise unaffected children.

## CONDITIONING

Children with PWS should be encouraged to be as

active as possible, particularly those activities that build core musculature, strengthen the abdominals and the back muscles. In addition to sports and recreational activities, focused physical therapy and hippotherapy, emphasizing core muscle strengthening, may help improve a hypotonic curve in a young patient with a flexible deformity. If kyphosis is noted to develop, a physical therapy regimen should be included with specific exercises to strengthen the back extension muscles. A number of parents have also found pilates to be beneficial.

## MONITORING

Scoliosis in infants with PWS is unlikely to develop prior to the influence of gravity across the spine. Therefore, monitoring of spine should begin when the child first sits independently, usually around the first birthday. Yearly screening radiographs of the seated or standing spine should be used in addition to a clinic examination. If a deviation greater than 10° from straight is noted, radiographs should be obtained more frequently, depending on the age of the child, size of the curve, and apparent risk towards progression.

## CASTING

If scoliosis is detected in the infant, spinal casting has been shown to be effected before 3 years of age. Curves over 20° should undergo casting with the Mehta technique, with a goal of decreasing the curves to as close to 0° as possible. Even curves over 90° can obtain some correction with casting, and should be attempted in this age range. In general, a child's third birthday is often seen as the upper limit to starting casting, but in practice casting has been initiated even up to the fourth birthday. The casts are changed every 2 months in children under the 2 years of age, and every 3 months in children over 2 years of age. Casts are continued until the curve has been successfully reduced, or if correction plateaus

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over successive cast. The child is then braced to maintain the size of the curve.

## **BRACING**

Brace treatment should be considered for curves over 20° in children under 10 years of age, and for curve over 25° in children 10 years or older. The brace, a thoracic-lumbar-sacral orthosis or TLSO, should be worn 22 hours per day, allowing an extra hour or two out-of-brace for physical activities. When the brace is prescribed, initial radiographs in brace should be obtained to verify moderate correction, as compared to the out-of-brace radiograph. Smaller spine curves in children with Prader-Willi syndrome are often flexible, although it will not be possible to obtain a 50% correction of all curves. Follow-up out-of-brace radiographs are obtained every 4 to 6 months, having the child remain out of brace over night, up until the time of the radiograph.

## **SURGICAL INTERVENTION IN THE GROWING CHILD**

In growing children (usually up to 10 years of age) with progressive scoliosis that cannot be maintained under the 50°, a surgical intervention is needed. Performing a definitive spinal fusion at this age may improve the child's deformity, but will restrict spine and chest growth. At maturity, the patient will be adult sized, but with a child sized chest. Therefore, implantation of an expandable device is likely required. (In practice, once I have identified a curve that will require surgery, I try to postpone intervention until the curve cannot be maintained below 50° in an in-brace radiograph.) The goal of this type of surgery is to decrease the curve's size initially, then prevent it from progressing while allowing for spinal growth. There are two kinds of expandable implants, one is the "growth rods" or "nonfusion spinal instrumentation" (NFSI), the other being the "vertical expandable prosthetic titanium rib" (VEPTR) device. The VEPTR device has proven to be problematic in children with PWS, primarily due to the low bone mineral density leading to frequent failure from rib fractures. For that reason, we

recommend the use of a NFSI. For PWS, the construct that has worked well is a two segment fusion cranially, and a two segment fusion caudally, each with bilateral pedicle screws, for four pedicle screws anchor above and below. The segments are spanned with dual rods. A typical construct would be from T3 to L4, fusing T3 and T4 to act as the upper anchor, and L3 and L4 as the lower anchor (see figure). A characteristic of PWS is a cervical-thoracic junction kyphosis. Therefore the proximal extent of the fixation should be as low as possible (T3 or T4), and a moderate amount of existing thoracic kyphosis can be accepted. Over-correction of thoracic kyphosis appears to drive the cervical-thoracic junction kyphosis towards progression. The construct is lengthened every 6 months, to keep up with spinal growth. Near skeletal maturity (as determined by bone age), the construct will need to be converted to a definitive fusion.

## **DEFINITIVE SPINAL FUSION**

In the older child, a definite fusion should be performed for curves exceeding 50° out of brace. A curve of this size has a 95% chance of progressing, even after skeletal maturity. Whereas in idiopathic scoliosis the lower age threshold for a definitive procedure is 10-12 years of age, children with PWS often have a later growth spurt, with delayed maturation, possibly related to the use of supplemental growth hormone. For that reason, it is advantageous to wait until 12 years of age for girls and 14 years for boys prior to fusion. Bone mineral density in children with PWS is frequently low, so multi-segmental pedicle screw constructs are recommended, maximizing the number of fixation points. The scoliosis is usually a kypho-scoliosis rather than the lordo-scoliosis seen in idiopathic scoliosis, but care should be taken not to over correct the kyphotic deformity, as this may secondarily exacerbate the cervical-thoracic junctional kyphosis. Also, as with the expandable implants, consideration should be given to keeping the upper level of the fusion no higher than T3 or T4, if possible, to prevent proximal junctional kyphosis from developing.

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## SURGICAL AND ANAESTHETIC CONSIDERATIONS

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In children requiring anesthesia, for spinal casting, hardware implantation, or even planned interval lengthens of a NFSI, the special characteristics of children with PWS should be well understood by the entire treatment team.

### **Respiratory**

The children have a number of respiratory issues, related to their hypotonia, as well as their obstructive and/or central sleep apnea. They are at high risk for postoperative pneumonia. The literature reflects a high rate of complications with anterior spinal procedures, and it is strongly recommended to avoid entering the chest. Preoperatively, a pulmonary function test is required, as is a sleep study in many cases. In cases of obstructive apnea, a tonsillectomy may be required preoperatively. Postoperatively, extubation may need to be delayed for a few hours, or even over night, until a patient has sufficiently awoken for a strong respiratory effort. Thereafter, the patient may require CPAP or BiPAP when sleeping, possibly for a few nights to a few weeks. Their hypotonia causes them to have a weak cough, and chest physical therapy should be aggressive.

### **Bone Mineral Density**

Children with PWS have decreased bone mineral, which may lead to loss of hardware fixation and/or pseudo arthrosis. Pre-operatively, it is important to have vitamin D and calcium levels optimized. Children with PWS who have been actively managed by their endocrinologist will likely have been on long term growth hormone and possibly sex steroid replacement. This treatment can optimize bone strength by puberty, but only if vitamin D and calcium levels have been sufficient. It is a good idea for patients to supplement their vitamin D and calcium intake, especially if surgery is being considered. Although radiographs notoriously underestimate bone mineral loss, a sure sign of an abnormality is if the vertebral density is nearly that of the soft tissue shadows, seen best at areas of overlapping bowel gas. Another warning sign is a diet low in dairy products (milk, yogurt, cheese). A DEXA scan may be helpful if 1) there is concern that the bone mineral density is critically low, and 2) there is enough time prior to surgery for the treatments to have an effect. In those critical cases, intravenous pamidronate

and vitamin D may make a meaningful difference.

Operative planning should maximize the number of spinal fixation points for performing the instrumentation for fusion. A multisegmental pedicle screw construct, with fixation at nearly every level has worked well.

### **Pain Tolerance**

Children with PWS have an increased pain tolerance, which may be helpful when attempting to mobilize them after the surgery. It could also be a reason why they awake slower from anesthesia.

### **Food Seeking and Gastrointestinal Issues**

The family and hospital staff needs to be vigilant for food seeking behavior, and its possible life threatening consequences. Preoperatively, the NPO interval may be violated, making induction of anesthesia dangerous. Postoperatively, the children should not be fed until they have normally active bowel sounds and flatus. Usually, they are hungry much earlier than other post-operative patients, but their ileus resolves a day or so later than expected. Also, the patient's preoperative calorie restrictions should be well known by the hospital's nutrition staff, and observed postoperatively. All treating staff should be aware that children with PWS do not vomit, nor lose appetite. Should either of those occur, they require an emergent nasogastric tube insertion, followed by an abdominal radiograph to evaluate for gastric dilatation, and possibly a CT scan to rule out free air. Gastroparesis leading to gastric necrosis is unfortunately a common cause of death in children with PWS.

### **Skin Picking**

A constant threat to the surgical incision is the habit of patients with PWS to skin pick, in essence scratching their wounds until they break down, leading to a dehiscence and a deep infection. This is probably the most common post surgical PWS complication seen. The skin picking may take the form of rubbing their back against a wall or furniture. In many cases, we have used a light post-operative brace to protect the skin for the first several weeks.

If the above points are carefully observed, surgery can be performed safely and uneventfully in children with PWS.