A Primer on Scoliosis Surgery for Prader-Willi Syndrome

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- Individuals with PWS often have decreased bone density. With growth hormone, the bone strength is usually satisfactory. Occasionally, the surgeon may need to add fixation points. This also presents a risk for Posterior Junctional Kyphosis (PJK) and Distal Junctional Kyphosis (DJK).

- Individuals with PWS have a different center of balance. They often have increased cervical-thoracic kyphosis, and walk with their head jutted forward. Avoid the temptation to improve their kyphosis. Most of the revision cases referred to me have failed due to overcorrection of the kyphosis, leading to a compensatory kyphosis above the construct, leading to PJK, and screw pull-out. I try to make my highest instrumented vertebrae T3 or lower, so that they have plenty of room to adjust kyphosis. In cases of spinal fusion for kyphosis, my goal is often to only correct the kyphosis to about 60°.

- Patients should have a sleep study, to rule out central or obstructive sleep apnea, and a pulmonary consult. They may need CPAP or BiPAP post-operatively.

- Patients with PWS can have both central and obstructive apnea and typically have an altered response to anesthesia. This can cause problems with extubation immediately post operatively. Frequently they may need to remain intubated in the ICU while they recover from anesthesia, anywhere from a few hours to overnight. Planning for that can help expedite transfer out of the operating room.

- Skin picking is a huge problem. Patients will tend to pick their incisions leading to infection. Placement of an incisional VAC (Vacuum Assisted Closure) will help with any seepage, but more importantly can act as an early alarm system, should they try to scratch their incisions in early post-op. I will usually outfit them with a light brace to protect the incision for about 2-3 months.

- Patients with PWS can frequently have gastrointestinal problems:

a. They are always very, very constipated. You may want to do a 1-2 week slow bowel clean up pre-op, so that there's less in place to cause post-op constipation. Start constipation medications early post-operatively.

b. They will be hungry immediately after surgery, but their actual ileus (caused by inactivity of the bowel) usually last longer than usual. These patients are susceptible to

gastroparesis (paralysis of stomach motility), causing them to have severe distension, to the point they can even have gastric perforation. Advance diet really, really slowly, relative to bowel sounds. Limited water is OK for first few days. Once solids started, get an abdominal x-ray to make sure there is no huge gas distension. Erythromycin can act as a gastric motility aid (better than Reglan for kids with PWS).

c. Have your dietician talk to family before admission to find out what the current diet plan is (usually 800 – 1000 calories per day).

d. Make sure nursing unit knows that there's a food seeking issue, and do not underestimate the craftiness of these children in finding food. This latter point is especially important if others are bringing food into the room, or the patient has a roommate.

- Patients with PWS frequently have impulse control or emotional/temper issues. It helps to have someone from psych or social work talk to family and school, and have a plan way ahead of time. The children respond well to well displayed schedules, so that they know what to expect, especially in relationship to meals. Plan ahead to have a family member with the patient essentially 24 hours per day. Soft restraints are often needed, to prevent pulling of intravenous lines and drains, as well as skin picking of the operative site.