A Primer on Scoliosis Surgery for Prader-Willi Syndrome

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1. **Bone mineral density:** Individuals with PWS often have decreased bone density. With growth hormone, the bone strength is usually satisfactory, but still less robust than typically developing peers. Worrisome indications are a history of fracture, or if the bowel gas pattern on a spine xray actually obscures the vertebra it overlays. The surgeon will probably prefer to add fixation points. This also presents a risk for Posterior Junctional Kyphosis (PJK) and Distal Junctional Kyphosis (DJK).

2. **Sagittal alignment:** The big issue in planning spine surgery for patients with PWS has to do with managing their kyphosis. Individuals with PWS have a different center of balance, prefering a head forward position when walking or standing, with their head anterior to the usual body axis. There is usually an associated thoracic hyperkyphosis or cervical-thoracic kyphosis. In general, patients with PWS have kyphoscoliosis rather than lordoscoloisis. The revision rate for PWS scoliosis cases is very high, mostly due to proximal or distal junctional kyphosis (PJK or DJK) and screw pull-out. This appears related to attempts to correct their kyphosis to what is considered "normal"; the patient subsequently develops a compensatory kyphosis above or below the construct, to reestablish their preferred posture. In the immediate post-operative period, the underlying generalized hypotonia probably is culpable as well. Avoid the temptation to improve their kyphosis to what is typical for a child with an idiopathic spine deformity. Attempt to build at least 50° of kyphosis into the fixation, and when a spinal fusion is primarily indicated for kyphosis, only correct the kyphosis to about 60° during surgical instrumentation. Try to leave a noninstrumented region of spine above the construct to allow the patient to sub-cervical motion segments to gradually adjust their kyphosis. I try to end at T4 whenever possible, and only instrument above T3 in the rarest of circumstances, regardless of the curve. PJK will destroy the surgical results early, whereas DJK is more gradual, taking years to become problematic, both compounded by the underlying poor bone mineral density. In some cases, I have added a chin support to the post-operative TLSO to remind the patient to keep their head up - the support can be removed from the brace for sleeping.

3. **Pulmonology and respiratory:** persons with PWS often have respiratory findings as well as disordered sleep, in part due to sleep apnea, both obstructive and central.
    a. **Sleep apnea:** Patients should be seen by their pulmonology and possibly have a sleep study, to rule out central or obstructive sleep apnea. Occasionally a ENT consultation is needed for consideration of adnoidectomy and/or tonsillectomy pre-operatively.
    b. **CPAP or BiPAP:** Due to obstructive and/or central sleep apnea, evaluations with BiPAP/CPAP titrations may be necessary. The patients must have repeat studies showing normalization with their settings anywhere from 3-6 weeks post initial study, prior to surgery. This may slow down the surgical clearnace, but there is nothing worse than having to initially introduce BiPAP/CPAP immediately postoperatively

for these behaviorally challenged patients. Despite concerns, these patients adapt well to the masks, even our very young patients undergoing spine casting for scoliosis.

c. **Skin picking:** Skin picking may seem, by description, to be just a social inconvenience, but it can be a huge problem. Patients will tend to pick their surgical incisions leading to dehiscence and infection, which can ultimately lead to hardware removal. Placement of a post-operative 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 period. Rarely restraints were needed immediately post-op, and IV sites and monitors are always a target. I will then usually outfit them with a light brace to protect the incision for about 2-3 months. They need the greatest supervision during bathing, when in my experience the roving hands take but seconds to wreak destruction.

4. **Gastrointestinal issues:** Patients with PWS frequently have gastrointestinal concerns. They have decreased gastric motility, and radiographically can have evidence of moderate to extreme constipation, even if they have a bowel movement daily. Lastly, patients with PWS have an insatiable hunger and a constant obsession with food; it is common for their first concern to be, on awakening in the operating room, when they will be able to eat. Ironically, they have a more stubborn ileus compared to typical patients.

   a. **Constipation:** Patients with PWS are always very, very constipated. Pre-operatively, a 1-2 week slow bowel clean out is helpful, so that there is less fecal material in place to cause post-op constipation. Start constipation medications early post-operatively; lactulose works well and is well tolerated by children with PWS.

   b. **Post-operative ileus:** The post-operative ileus lasts longer than usual for a similar procedure. Advance diet really, really slowly, relative to bowel sounds. Limited water is OK for first few days. Our protocol has been 2 ounces of clear liquids (including jello [think sugar-free] and popsicles) every 4 hours. Once that is tolerated, increase to 4 ounces every 4 hours. If that does well, obtain an upright abdominal view (or first upright spine) to evaluate stomach gas bubble and colon gas pattern, helping to determine if the ileus is resolving. If the signs are good, begin to gradually add a soft diet, and obtain daily abdominal x-rays until full diet is restored, making sure the gas pattern continues to improve. Erythromycin can aid gastric motility, it works better than Reglan for kids with PWS. Also consider methylnaltrexone, a μ-opioid–receptor antagonist, post-operatively, as it selectively blocks the narcotic effects on the gut.

   c. **Gastroparesis:** People with PWS are susceptible to gastroparesis (paralysis of stomach motility), where gastric emptying completely fails and the stomach becomes severely distended. Without treatment, the gas distension can stretch the stomach lining and muscular wall to the point of ischemic necrosis, and stomach rupture can follow. If a patient with PWS has increasing abdominal distension, abdominal pain of any sort, lack of appetite, or emesis, an emergency abdominal radiograph is necessary, and possibly an abdominal CT to evaluate for thinning of the stomach wall. Note that persons with PWS has a much higher pain tolerance, therefore mild abdominal discomfort can be an important symptom. Treatment is immediate NPO status and a nasogastric tube, IV hydration (the patient is likely dehydrated), and close observation.

d. **Food obsession:** They will be hungry immediately after surgery, and will look for opportunities to get food. This may include stealing food, eating non-food stuffs, tricking others into giving food, or foraging (ie. trashcans). Make sure the nursing unit knows that there is 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.

e. **Nutritional support:** People with PWS have caloric requirements much lower (~50%) than same sized typical patients and nearly all are on a strict calorie restricted diet, which should be observed before and after surgery. Have your dietician talk to family before admission to find out what the current diet plan is (usually 800 – 1000 calories per day).

5. **Anaesthesia and pain management:**

a. **General:** Despite earlier concerns, children with PWS have no known risk of malignant hyperthermia. They do exhibit slower arousal from anaesthesia, possibly due to their higher pain tolerance, and their apnea, both central and obstructive, can cause an altered response to anesthesia. This can cause problems with extubation immediately post operatively. On rare occasions 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.

b. **Reduced CO_2 :** Children with PWS have a reduced sensitivity to CO_2, so to be careful with supplemental O_2 so as not to drive down respiratory drive.

c. **Post-operative pain:** All our patients get dexmedetomidine and morphine (hydromorphone) either by patient controlled anaesthesia (PCA) pump, or, for the few who cannot use a PCA, just as a basal rate. If they are intubated then dexmedetomidine and fentanyl works great. We suggest staying away from Ketamine, including intra-operatively, due to the frequent behavior issues these patients have. The patients' high tolerance for pain usually means they require less narcotic than the average patient.

6. **Venous access:** patients with PWS are notoriously difficult for obtaining venous access, both due to the amount and quality of their subcutaneous fat, and their small veins. With the extended ileus most will experience, it is good to plan on an extended need for IV hydration. Therefore, we will usually have a central venous line (CVL) placed at the time of major surgeries. In patients whose skin picking is difficult to control, we suggest a tunneled CVL to prevent a catheter insertion site infection becoming a sepsis. Occasionally the pre-operative decision was made for a Broviac catheter, or, in very compliant patients, a peripherally inserted central catheter (PICC) line.

7. **Emotional well-being:** Patients with PWS frequently have impulse control or emotional/temper issues. It helps to have someone from psychology or social work talk to family and school pre-operatively, and have a plan much prior to admission. Children with PWS often have trouble with changes in routine, but having a well-displayed schedule can help limit anxiety, 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. Occasionally a 24 hour 1:1 companion from an agency is needed, both for staff and patient safety.

8. **Post-operative protocol summary:**
    a. Incisional negative pressure dressing for 5 days
    b. Post-operative brace to protect wound
    c. Bathing by post-operative day 5, with close supervision to prevent skin picking
    d. Avoid forward bending or picking up objects heavier than 10 pounds for 3 months
    e. Encourage walking for exercise daily on discharge (30-60 minutes per day)
    f. Allow swimming activities at 6 weeks if incision well healed

9. **Growth Hormone:** There is no data that links growth hormone supplementation to spine deformity initiation or progression in Prader-Willi syndrome. Growth hormone has many important benefits for these patients metabolically and physiologically. Therefore it is important not to cancel growth hormone supplementation. We tend to skip the dose on the day of surgery, then resume dosing on post-operative day 1.