

Postoperative Monitoring of Patients with Prader-Willi Syndrome

Treatment Recommendations

The following recommendations have been reviewed by members of the Clinical Advisory Board of the Prader-Willi Syndrome Association (USA) with special acknowledgment to Dr.'s Moris Angulo, Mary Cataletto and Mary Lyn Quintos-Alagheband of Winthrop University Hospital, Mineola, NY (which cares for over 300 patients with the syndrome) for their contribution to this document. The unique health needs of individuals with this disability must be carefully researched and evaluated by all health care professionals. For questions or consultations, please feel free to contact the PWSA (USA) office at 1-800-926-4797 from 10:00 am to 5:00 pm EST weekdays.

BACKGROUND INFORMATION:

Patients with PWS are known to have increased morbidity after surgery due to:

- Abnormal physiological response to hypercapnia and hypoxia
- Hypotonia
- Narrow oropharyngeal space
- High incidence of central, obstructive and mixed apnea
- Thick secretions
- Obesity
- Increased incidence of scoliosis with decreased pulmonary function
- Prolonged exaggerated response to sedatives
- Increased risk for aspiration
- Decreased pain sensation is common in persons with PWS

Patients with PWS may experience greater challenges with compliance to pre- and postoperative treatment procedures due to:

- Extreme food seeking behavior and hyperphagia due to hypothalamic dysfunction.
- High incidence of gastroparesis and slow motility of the intestinal tract.
- Extreme skin picking which may interfere with wound healing.
- Altered temperature regulation – fever may be absent in the presence of infection. There does not seem to be a higher incidence of malignant hyperthermia.

RECOMMENDATIONS:

- Patients with PWS who undergo deep sedation and general anesthesia should be recovered overnight in a monitored unit.
 - Infants and children may require intensive care monitoring
- Continuous monitoring of pulse-oximetry for 24 hours postoperative with attention to airway and breathing.
- A conservative approach to pain management and use of narcotic agents.
- Full assessment of return GI motility prior to initiation of intake by mouth because of the predisposition to ileus after surgery.
- Scheduling procedure as early in the day as possible to prevent prolonged time period where food seeking could take place.
- Direct supervision (1:1) to prevent foraging postoperatively.
- Monitor for picking at wounds and/or incisions. May require additional dressings and other barriers to prevent access to wound.
- Close observation of wound for signs of infection.
- Utilization of respiratory therapy interventions to prevent atelectasis and/or postoperative lung infection.
- Due to the hypotonia and obesity the individuals with PWS are at risk for deep venous thrombi (DVT) and pulmonary embolism. Patients should be under the guideline for DVT prophylaxis.