

## Postoperative Monitoring of Patients with Prader-Willi Syndrome

### *Treatment Recommendations*

#### **Background Information:**

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

- 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.

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*Reviewed September 2022*