

Guidelines for Postoperative Monitoring of Pediatric Patients with Prader-Willi Syndrome

Winthrop University Hospital Cares for over 300 patients with Prader Willi syndrome. The Prader Willi Center at Winthrop is a national referral resource for these patients. Many of them have chosen to perform their emergency and elective surgeries at our institution.

Background Information:

Patients with Prader-Willi syndrome are known to have increased morbidity after surgery due to:

- Abnormal physiologic 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 PWS

Recommendations:

- I. Infants and children with Prader Willi syndrome who undergo deep sedation and general anesthesia should be recovered overnight in a monitored unit, either the Pediatric Recovery Room or Pediatric Intensive Care Unit.
- II. Continuous monitoring of Pulse oximetry for 24 hours postoperative is strongly recommended with attention to airway and breathing
- III. A conservative approach to pain management and use of narcotic agents is recommended.
- IV. Full assessment of return of GI motility prior to initiation of intake by mouth because of the predisposition to ileus after surgery.
- V. Direct supervision (1:1) to prevent foraging postoperatively.
- VI. Monitor for picking of wounds.

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