

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

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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:

1. 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.
2. Continuous monitoring of Pulse oximetry for 24 hours postoperative is strongly recommended with attention to airway and breathing
3. A conservative approach to pain management and use of narcotic agents is recommended.
4. Full assessment of return of GI motility prior to initiation of intake by mouth because of the predisposition to ileus after surgery.
5. Direct supervision (1:1) to prevent foraging postoperatively.
6. Monitor for picking of wounds.

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