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Special Anesthesia Concerns for Patients with Prader-Willi Syndrome: The Winthrop University Hospital Center Experience

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Prader-Willi syndrome (PWS) is a complex syndrome whose features include muscular hypotonia, central nervous system abnormalities, behavior problems, obesity, hypogonadism and skeletal abnormalities. Decreased pulmonary reserve secondary to chest wall deformity (i.e., scoliosis), hypotonia and obesity may complicate ventilatory management during and following anesthesia. These individuals have in addition other characteristics such as an abnormal physiologic response to hypercapnia and hypoxia, a narrowed oropharyngeal space, hypoplastic dental enamel, thick secretions, prolonged and exaggerated response to sedatives, and increased risk of gastric aspiration that can lead to potential difficulties in airway management during the perioperative period. Familiarities with these issues can help to facilitate the anesthesia experience and prevent the number and severity of postoperative complications. We report the common complications observed in children with PWS that underwent surgical procedures.

We retrospectively reviewed the records of 13 patients (9 male) who underwent a total of 19 surgical procedures with general anesthesia. Age ranged from one to 15 years of age with a mean of 5.1 years of age. Specific surgeries consisted of adenotonsillectomy (T&A) (n=7), orchiopexy (n=8), Broviac central catheter placement (n=3), and one testicular biopsy. All cases were performed under general inhalational anesthesia. In 13 cases the airway was secured with endotracheal intubation and in 6 cases with laryngeal mask airway (LMA). Mean anesthesia time was 97 minutes (range = 35–285 minutes). There were a total of 6 adverse events (31.6%); four respiratory, one prolonged emergence from anesthesia, and one post-operative ileus. All complications occurred within 24 hours of surgery. The following chart gives specific details:

Age (years)	Sex	Procedure	Airway	Complication
3	Male	T&A	ETT	Prolonged Emergence
7	Male	T&A	ETT	Atelectasis
2	Male	T&A	ETT	Pulmonary Edema
1	Male	Orchiopexy	LMA	Apnea
1	Male	Orchiopexy	LMA	Pulmonary Edema
1	Male	Orchiopexy	LMA	Ileus

Young age and type of airway management were associated with perioperative difficulties. All patients with postoperative complications were male. Half of the patients who received ventilation via LMA had a complication post-operatively compared with only one quarter of those ventilated through an endotracheal tube. The configuration of the oropharyngeal space with narrow high arched palate makes placement and correct fit of a laryngeal mask more difficult. We did not have problems with dental injuries during intubation as might be suspected with hypoplastic dental enamel. Duration and type of anesthesia did not correlate with complication rate. All patients recovered to baseline and were discharged from the hospital without further incident.

Children with Prader-Willi syndrome may be at increased risk for post-operative complications, most of which occur within 24 hours after surgery. The data presented here stress the importance of preoperative assessment, airway management, and postoperative monitoring. Staff working perioperatively with these patients should be knowledgeable about anatomical and pathophysiological features unique to the syndrome. Close monitoring in the intensive care unit for at least 24 hours post-operatively may be warranted.