

Introduction:

- CCHS is a rare neuropathological condition characterized by failure of the autonomic control of breathing during sleep
- Profound concerns for the anesthesiologist in the perioperative period due to absent ventilatory response to hypoxemia and hypercarbia
- Aim to evaluate our anesthetic practice and perioperative outcome among these patients at Children's Hospital Los Angeles

Methods:

- IRB approved retrospective study
- Included all CCHS patients who underwent procedures under GA from 2000-2016
- Reviewed charts for demographics, PHOX2B genotype, ventilatory support, comorbidities, anesthetic and perioperative course
- Endpoints were time to extubation or return to home ventilator settings, and hospital stay

Results:

- 19 patients (42% males, mean age of 6.9 years) underwent a total of 74 procedures
- PHOX2B mutation was seen in 18 patients
- 14 patients were ventilator dependent during sleep
- 7 used PPV via tracheostomy
- 7 had diaphragmatic pacers
- LMA in 1 and ETT intubation in 21 cases
- Mean time to extubation was 1.52 days
- Mean time to discharge from hospital was 13.3 days
- 22 cases received Midazolam, 2 requiring assistance
- Long-acting NMBs used in 24 cases (17 reversed)
- Intraoperative opiates were given in 32 cases
IV morphine (2), IV fentanyl (29), epidural morphine (1)

Table 1. Peri-Operative Anesthetic Complications in CCHS patients (%)

Complications	% Occurrence
Cyanosis	1.35
Hypotension	1.35
Readmission following seizure	1.35
Tachycardia	2.7
Bradycardia	1.35
Hypotension	4.05
Severe respiratory acidosis	1.35
Diaphragm pacer dysfunction	1.35
Flushing	2.7
Fever	4.05
Low urine output	2.7
Metabolic acidosis	1.35
Atelectasis	1.35

Figure 1. Surgery Performed (%)

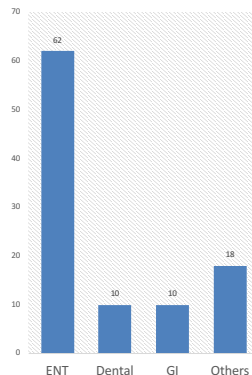
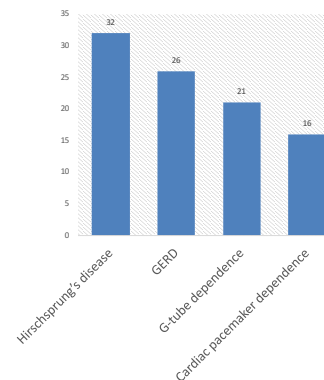


Figure 2. Common Comorbidities (%)



Discussion:

- Literature on optimal anesthetic care is lacking, existing mostly as case reports
- As a referral center for CCHS, we present our anesthetic experience in these children
- Anesthetic management is challenging with goals including avoidance of hypoxia, hypercarbia, acidosis, hypo or hypervolemia, and hypothermia while providing adequate anesthesia and pain control
- Long acting opioids have profound respiratory depression and should be avoided
- Regional techniques are excellent for post-op pain control, however, it is often difficult to use this as the primary anesthetic in pediatric population
- Long acting NMBs should be appropriately reversed
- Premedication with benzodiazepines should be avoided due to the increased risk for apnea in an inappropriate setting
- Parental presence on induction should be considered

Conclusion:

- Imperative for the anesthesiologist to have a good understanding of the condition and its associated comorbidities as well as secondary complications to achieve the best outcome

References:

- Niazi AU, Mocon A, Varadi RG, Chan VW, Okrainec A. *Ondine's curse: anesthesia for laparoscopic implantation of a diaphragm pacing stimulation system.* Can J Anesth 2011; 58:1034-1038.
- Visser WA, Fanyar Z, Luiten EJT. *Thoracic paravertebral block for awake breast surgery in a patient with congenital central hypoventilation syndrome (Ondine's Curse).* J Clin Anesth 2013; 25: 604-605.