Anesthesia for a patient with Congenital Central Hypoventilation Syndrome
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Introduction
Congenital Central Hypoventilation Syndrome (CCHS) is a rare syndrome comprised of three features: cyanosis, apnea, and hypotonia. It is also known as Ondine’s Curse or primary alveolar hypoventilation. It was first described in 1962 by Severinghaus and Mitchell who observed hypoventilation during sleep in three patients following surgery on the cervical spinal cord or brainstem. Most children born with this syndrome do not survive infancy and currently there are only about 200 people around the world who carry this diagnosis. There are several causes of CCHS ranging from an inborn error to spinal cord injuries. In most cases there is no response to hypoxia or hypercarbia when the patient is asleep. The following is the case of a young adult with this syndrome who relies on a diaphragmatic implanted pacer to breath when she is asleep. She presented for removal of hardware from her face following a Leforte I maxillary advancement for her protuberant jaw.

Case report
A 23yo, 49kg female with CCHS due to PHOX2B polyalanine expansion mutation, chronic respiratory failure during sleep, attention deficit hyperactivity disorder and mid face hypoplasia presented to the operating room for removal of hardware from the right maxilla s/p Leforte I surgery. The patient was born with respiratory failure and required a tracheostomy as a neonate. The patient continued to have hypventilation during sleep and was diagnosed with CCHS. She required mechanical ventilation during sleep through her tracheostomy. In 2004 the patient had a diaphragmatic pacer placed laparoscopically which was activated a couple months after the surgery. Soon after the placement of the diaphragmatic pacer the tracheostomy site was closed. Since the patient now had her CCHS under control with the diaphragmatic pacer she proceeded with her midface advancement in 2008. She returned to the operating room for screw removal secondary to increasing pain at the hardware site. The patient stated that she only used her diaphragmatic pacer when she was asleep as it would be painful to use it when she was awake. The anesthetic plan was to for an IV induction with midazolam, fentanyl and propofol. Ventilation was adequate and the patient was subsequently paralyzed with vecuronium, easily intubated and placed on the ventilator. The procedure lasted for twenty minutes and the patient was reversed and extubated without incident. Following surgery the patient complained of increasing pain at the surgical site and was given several doses of morphine.

Discussion
While performing an anesthetic on a patient with CCHS several questions come to mind. Is it safe to paralyze this patient? Can residual muscle relaxant create difficulty in pacing? How does a magnet affect this patient’s pacer? What narcotic is best? This patient was paced only when she was asleep but can she be paced awake and if so should she be sedated? How awake does the patient need to be to have a normal response to hypercarbia and hypoxia. Since this condition is extremely rare there are only a few case reports in the literature that address these questions. In most cases the patients were not given muscle relaxant for fear that ventilatory support would be compromised. In these cases the patient did not have a diaphragmatic pacer in place. Since this patient did have a pacer it was assumed to be safe to use muscle relaxant.

References: