Undiagnosed late onset central hypoventilation syndrome: Perioperative challenges and Anesthetic Implications

Author(s): Mohamed Mahmoud, MD, Yuvan Bryan, MD, Senthilkumar Sadhasivam, MD

Affiliation(s): Department of Anesthesiology, Cincinnati Children’s Hospital Medical Center, Cincinnati, USA.

Introduction: The differential diagnosis of delayed emergence in Post Anesthesia Care Unit (PACU) is most commonly due to residual anesthetic agents, sensitivity to narcotics and sedatives and residual neuromuscular weakness. Rarely slow awakening after anesthesia may be due to metabolic or neurologic disorders. We present a case of delayed emergence due to an undiagnosed late onset central hypoventilation syndrome (LO-CHS) in a toddler that resulted not only in delayed awakening but also unanticipated respiratory complications following a tonsillectomy that required re-intubation, ventilatory support and eventually a tracheostomy.

Case report: 2-yr-old, 14.6 kg girl was admitted to the hospital for history emesis with two episodes of oxygen desaturation in the emergency department (ED). An airway exam showed enlarged tonsils and adenoids and an X-ray exam of chest was normal. She was scheduled for an elective tonsillectomy, adenoidectomy, microlaryngoscopy and bronchoscopy to evaluate the airway for possible explanation for this episodes of oxygen desaturations. Intravenous induction was done with propofol 45 mg, vocal cords were sprayed with 2 ml of lidocaine 4%. Microlaryngoscopy and bronchoscopy were performed under insufflation with sevoflurane 6%. No gross anatomical airway abnormality was found by the surgeon. Trachea was intubated with an internal diameter 4 mm, non-cuffed endotracheal tube with a peritubal leak at 15 cm H₂O. The patient received the following drugs during the procedure; morphine 2-mg, dexamethasone 4-mg and ondansetron HCL 2-mg. No muscle relaxant was used during the procedure. Despite discontinuing sevoflurane, there were no efforts of spontaneous breathing at the end of the procedure. Patient was transferred to PACU with endotracheal tube in situ. Spontaneous breathing did not resume even after 1 hour of assisted ventilation in the PACU. A peripheral venous blood gas analysis showed normal electrolytes and a respiratory acidosis with a PvCO₂ of 120 mmHg. The patient was hyperventilated. Due to suspicion of hypersensitivity to narcotic, 20 mcg of naloxone was titrated every five minutes. Patient resumed spontaneous breathing, followed commands, so the trachea was extubated in PACU. For a close observation of respiration, the patient was transported to the Pediatric Intensive Care Unit. Due to persistent apnea requiring intermittent naloxone, naloxone infusion was started immediately in the Pediatric Intensive Care Unit but she was reintubated next day for persistent hypercarbia (PaCO₂ 130 mmHg) despite spontaneous breathing with continuous naloxone infusion. On the third post operative day, the patient’s mother mentioned that the child’s father had a tracheostomy for central hypoventilation syndrome (CHS) which presented during his infancy. Pulmonologists and sleep specialists were consulted to evaluate this patient and they suggested that given
the family history, the current perioperative scenario was suggestive of a rare delayed onset central hypoventilation syndrome. To confirm the diagnosis and to rule out other causes further exams were done. An ultrasound exam of diaphragm ruled out a paralyzed diaphragm as a cause for hypoventilation. A sleep study done when the patient remained intubated showed evidences of central hypoventilation and an increased upper airway resistance that clinically manifested with snoring, paradoxical respiration and intermittent airway obstructive events. On the seventh post operative day, she was extubated and placed on BIPAP during sleep only. Tracheostomy was done three weeks later with sevoflurane and local anesthesia only and she was discharged later on home ventilatory support at night.

Discussion: Delayed emergence is a relatively frequent occurrence in the recovery room. It is usually related to drugs administered in the course of the procedure, and their timing of administration. Less commonly, it is caused by metabolic or electrolyte disturbance, hypotension, hypoxia, or intracerebral pathology. All except intracerebral pathology were excluded in this case. Under normal circumstances, the sedative effects of morphine, propofol, and sevoflurane would not hamper a rapid emergence from general anesthesia at the end of the procedure. Since the child started to breath spontaneously after titrating naloxone, we felt oversensitivity to morphine was the most likely the reason but the patient was reintubated next day despite being on naloxone infusion overnight making narcotic oversensitivity unlikely the reason. Without knowing the family history of CHS which was mentioned later to intensive care unit team, it was very difficult for anesthesiologist to figure out the reason for the delayed wake up and consistent drowsiness until the diagnosis of delayed central hypoventilation syndrome was made several days later. Congenital central hypoventilation syndrome (CCHS) or ondine’s curse and late onset central hypoventilation syndrome (LO-CHS) are diagnosed in the absence of neuromuscular, pulmonary or cardiac disease or an identifiable brain stem lesion. CCHS presents at birth whereas LO-CHS had not been reported well previously in children after infancy. Typically children with late onset central hypoventilation have normal growth, development and cognitive function until 1.5-4 years of age. Hypercarbic respiratory failure is often precipitated by adenotonsillar hypertrophy, anesthesia or pharmacologic respiratory depressants in these patients. The prevalence of CCHS is unknown, but registries suggest that there are, at minimum, 180 living children world wide with the condition (1). LO-CHS has been described in at least 11 cases in the literature (2). They are characterized by generally adequate ventilation while the patient is awake but hypoventilation with shallow rates while sleep. These children do not perceive the challenges of hypoxemia and hypercarbia; they are likely to exercise farther and longer than their friends without sensing their physiologic compromise, resulting in fatal hypoxemia/hypercarbia. Several ventilatory support options are available for children with LO-CHS. The older child with entirely normal airway may benefit from diaphragmatic pacing by phrenic nerve stimulation while awake and BIPAP mask ventilation while asleep. Tracheostomy and home mechanical ventilation is required for younger children. In conclusion surgery in a patient with undiagnosed delayed central hypoventilation syndrome is an anesthetic challenge because of its respiratory consequences and its rarity. LO-CHS can mimic many diseases including myasthenia gravis, altered airway or intrathoracic anatomy, diaphragm dysfunction or brain stem
abnormality. A proper history from family and clinical suspicion of LO-CHS will help to identify these patients and help anesthesiologists to warn patients/parents potential serious respiratory consequences following relatively minor surgeries.

References: