Gomez-Lopez-Hernandez syndrome, also known as cerebello-trigeminal dermal dysplasia, is a rare neurocutaneous syndrome classically characterized by the triad of rhombencephalosynapsis, trigeminal anesthesia, and bilateral parietal alopecia. Associated clinical features include a characteristic facial appearance (mid-face hypoplasia, hypertelorism, and low-set, posteriorly rotated ears), brachycephaly, strabismus, ataxia, developmental delay, short stature, and corneal opacities. Given the associated congenital anomalies, anesthetic care may be required for various surgical interventions. We report a 7-month-old with Gomez-Lopez-Hernandez syndrome scheduled for laparoscopic gastrostomy with tube placement and frenulotomy under general anesthesia. The potential perioperative implications of such patients are reviewed and options for anesthetic care discussed.

Introduction

Gomez-Lopez-Hernandez syndrome (GLH) syndrome, also known as cerebello-trigeminal-dermal dysplasia was reported by Gómez in 1979 and subsequently by López-Hernández in 1982. GLH syndrome is one of the more uncommon neurocutaneous syndromes, with only 30 cases reported in the literature since its initial description in 1979. The classical triad of clinical findings includes rhombencephalosynapsis, trigeminal anesthesia, and bilateral parietal or parieto-occipital alopecia. Rhombencephalosynapsis is the most characteristic malformation seen in GLH syndrome. It is characterized by posterior fusion of the cerebellar hemispheres and by agenesis or hypogenesis of the vermis, fusion of the dentate nuclei, and apposition or fusion of the cerebellar peduncles. Additional clinical features of GLH syndrome may include craniofacial anomalies such as brachycephaly or brachyturricephaly, a distinct facial appearance (midface hypoplasia, hypertelorism, and low-set, posteriorly rotated ears), strabismus, ataxia, developmental delay, short stature, and corneal opacities secondary to trigeminal anesthesia. As no chromosomal abnormalities have been determined using karyotyping or subtelomere screening, it is postulated that de-novo chromosomal arrangements or spontaneous dominant mutations are responsible for the clinical findings. Given the associated congenital anomalies, anesthetic care may be required for various surgical interventions. We report a 7-month-old male with Gomez-Lopez-Hernandez syndrome scheduled for laparoscopic gastrostomy with tube placement and frenulotomy requiring general anesthesia. The potential perioperative implications of such patients are reviewed and potential anesthetic regimens discussed.
Case Report

Institutional Review Board approval is not required by Nationwide Children’s Hospital for presentation of single case reports. The patient was a 7-month old, 5.9 kg infant presenting for laparoscopic gastrostomy with tube insertion and frenulotomy due to feeding difficulties and failure to thrive. The patient was diagnosed with Gomez-Lopez-Hernandez syndrome during the neonatal period based on findings from brain magnetic resonance (MR) imaging and the characteristic physical features. He was delivered as twin B at 37 weeks gestation via Cesarean section. Routine ultrasound examination at 20 weeks gestation had shown the abnormal brain structure. Pregnancy was complicated by a bicornuate uterus and a velamentous insertion of the umbilical cord. Birth weight was 2640 grams with Apgars of 9 and 9. The patient was born to a 27 year old mother and a 32 year old father. The medical history of the mother included dyslexia and attention deficit disorder. She was also found to have Factor V Leiden deficiency and treated with enoxaparin during pregnancy. She took prenatal vitamins and there was no known exposure to tobacco, alcohol, drugs, or other teratogens. The patient’s 2 year old brother was being evaluated for possible autism. Continuous positive airway pressure (CPAP) was required for approximately 12 hours after delivery due to respiratory distress. Post-delivery karyotype and microarray tests were normal. MR imaging of the brain revealed rhombencephalosynapsis (RES). He had abnormal movements including head shaking/rolling, motor developmental delay, and bilateral parietal alopecia. Associated co-morbid conditions included feeding problems, gastroesophageal reflux, diarrhea, and failure to thrive. The patient’s past surgical history included two lingual frenulotomies. Medications at the time of his current surgery included ranitidine, albuterol, lansoprazole, hyoscyamine, and multivitamins. There were no known allergies. His pre-operative vital signs included a resting respiratory rate of 36 breaths/minute, an oxygen saturation of 97% by pulse oximetry on room air, a heart rate of 121 beats/min, and a blood pressure (BP) of 97/51 mmHg. Preoperative physical examination revealed an abnormal head shape and nystagmus. There were no clinical signs of respiratory and cardiovascular dysfunction. Airway examination revealed congenital ankyloglossia and a Mallampati Class II view. Preoperative laboratory evaluation including electrolytes, renal function, and blood glucose were normal. The patient was admitted on the day of surgery. He was held nil per os for solids for 6 hours and for clear liquids for 2 hours prior to surgery. The patient was transported to the operating room and routine American Society of Anesthesiologists’ monitors were placed. Nitrous oxide (50%) in oxygen was administered via the face mask for one minute. Anesthesia was then induced with sevoflurane. No anticholinergic agent was administered prior to anesthetic care. The baseline heart rate was 120 beats/min with a blood pressure (BP) of 105/75 mmHg. The concentration of sevoflurane was increased by 2% every 3-4 breaths to a maximum inhaled concentration of 8%. Because of the patient’s non-reassuring airway examination, no neuromuscular blocking agents were administered. Spontaneous ventilation was maintained throughout induction and endotracheal intubation. Light cricoid pressure was applied, which brought the vocal cords into view (Cormack-Lehane score of 1). Endotracheal intubation was easily achieved using a Miller 1 blade and a cuffed 3.5 endotracheal tube. The patient tolerated anesthetic induction and endotracheal intubation without adverse effects. Mechanical ventilation was initiated for the case. Maintenance anesthesia consisted of sevoflurane (end-tidal concentration 2-3.5%) in 50% oxygen and air, dexmedetomidine (0.3 µg/kg), and fentanyl (2.5 µg/kg). HR decreased from 152 to 128 beats/min and mild hypotension occurred after the administration of fentanyl, which did not require therapy. The blood pressure stabilized between 70-80/30-40 mmHg during the case. The surgical procedure lasted approximately 1.5 hours. There were no intraoperative surgical complications. A total of 120 mL of lactated Ringer’s solution was administered during the procedure. Wound infiltration was administered by the surgeon for postoperative analgesia using 3 mL of 0.25% bupivacaine with 1:200,000 epinephrine. The patient was transported to the Post-Anesthesia Care Unit (PACU) and his trachea was extubated uneventfully in the PACU. He was hemodynamically stable, spontaneously breathing, and discharged from the PACU without any complications. The remainder of the postoperative course was uneventful.
Discussion

Patients with known genetic syndromes may pose a variety of challenges to the anesthesia provider. As with all anesthetic care, appropriate preoperative preparation begins with a thorough history and physical examination. Of primary concern to the anesthesia provider in patients with diagnosed or potential genetic syndromes is the potential for difficulties with airway management and endotracheal intubation related to craniofacial abnormalities including micrognathia, midface hypoplasia, and other associated dysmorphic features. The appropriate equipment for dealing with the difficult airway including an indirect videolaryngoscope should be readily available prior to anesthetic induction or airway management. As was accomplished in our patient, general anesthesia can be induced by the incremental inhalation of sevoflurane with the maintenance of spontaneous ventilation until the airway is secured or adequate bag-valve-mask ventilation is demonstrated. In many instances, endotracheal intubation can be accomplished without the use of neuromuscular blocking agents with sevoflurane supplemented with intravenous propofol or an opioid (remifentanil) if needed. Despite the dysmorphic features noted in our patient, airway management was not complicated and direct laryngoscopy with minimal cricoid pressure provided a grade I view of the glottic structures.

To date, there has been only one previous publication regarding anesthetic care for patients with GLH syndrome, which involved the provision of general anesthesia for a 2-year-old girl during MR imaging of the brain. Given the dysmorphic facial features, the authors were prepared for a potentially difficult airway prior to anesthetic induction as a GlideScope® and equipment for fiberoptic endotracheal intubation were available. Similar to our case, anesthesia was induced with the inhalation of sevoflurane and the maintenance of spontaneous ventilation. During anesthetic induction, upper airway obstruction occurred with persisted despite placement of an oral airway. A laryngeal mask airway (LMA) was placed which resulted in the resolution of upper airway obstruction. The LMA was left in place for the imaging procedure. Spontaneous anesthesia was maintained and anesthesia provided by a propofol infusion at 7.2 mg/kg/hour (120 µg/kg/min). No perioperative issues were noted. In their review, the authors identified several potential features which may impact perioperative care including potential airway concerns, psychiatric and behavioral problems (hyperactivity, depression, and self-injurious behavior), trigeminal anesthesia with recurrent corneal and facial scarring, parietal scalp alopecia, muscular hypotonia which may result in upper airway obstruction, as well as ventriculomegaly and the potential for hydrocephalus.

The cognitive performance of patients with GLH syndrome may range from moderate-severe impairment to normal cognitive function. Baseline mental and intellectual impairment should be assessed to help differentiate pre-existing problems from postoperative complications. Documentation of neurologic deficits is suggested prior to anesthetic care. Symptoms such as hyperactivity, depression, self-injurious behavior and bipolar disorder may require premedication or sedation. Careful postoperative observation and protection should be employed to avoid self-injury in behavioral disorder cases.

Given the potential for trigeminal anesthesia, one of the major concerns regarding perioperative care is protection from damage to the insensate areas including the cornea. Although trigeminal anesthesia is considered one of the classic triads of GLH syndrome, it is not invariably present. As cognitive performance permits, the presence of trigeminal anesthesia should be documented. Corneal ulcerations, clouding, keratitis, or facial scarring related to trigeminal anesthesia may be present preoperatively and should be carefully documented during the preoperative examination. Corneal abrasions may result from direct trauma to the unprotected eye during anesthetic induction and airway management or more commonly during the postoperative period especially in patients with altered cognitive function and hyperactivity issues. Loss of pain perception and inhibition of protective corneal reflexes may further increase the risk of corneal injury. As such, careful attention to corneal protection is suggested during perioperative care.

Scalp alopecia in GLH syndrome was originally described as bilateral parietal or occipital non-scarring alopecia, but it may present in many other distributions. As alopecia may be hidden by surrounding scalp hair,
it is important to document the areas of alopecia to ensure differentiation from a possible pressure-related complication due to prolonged positioning in the operation room setting.

Given the invariable involvement of the central nervous system, hypotonia is invariably present in patients with GLH syndrome. Involvement of the upper airway with dyscoordination of the pharyngeal musculature may result in upper airway obstruction during anesthetic induction. This is also relevant during the perioperative period when the effects of residual anesthetic agents and neuromuscular blocking agents may exacerbate poor baseline function and result in perioperative respiratory insufficiency. Muscular hypotonia can also impact on postoperative respiratory function at the level of upper airway and the thoracic musculature and diaphragm. As was chosen for our patient, the use of short acting anesthetic agents should be considered. In many cases, neuromuscular blocking agents are not required even for endotracheal intubation. These patients may also require prolonged postoperative care in a monitored setting following major surgical procedures. Although no such problems were noted in our patient, the previous report of anesthetic care noted upper airway obstruction during anesthetic induction that was not relieved by placement of an oral airway, but was resolved with insertion of an LMA.

Hydrocephalus may coexist with developmental anomalies of central nervous system. Ventriculomegaly with the development of hydrocephalus has been reported in GLH syndrome as well as rhombencephalosynapsis. Clinical signs and symptoms of increased intracranial pressure should be identified during the preoperative assessment.

In summary, patients who have GLH syndrome may present for diagnostic imaging or various surgical procedures requiring general anesthesia. The current report is only the second in the literature to provide guidance regarding the anesthetic care of such patients. Potential perioperative implications include a potentially difficult airway given the associated dysmorphic features of the head and face, psychiatric and behavioral problems (hyperactivity, depression, and self-injurious behavior), trigeminal anesthesia with recurrent corneal and facial scarring, parietal scalp alopecia, muscular hypotonia, as well as ventriculomegaly and the potential for hydrocephalus.
References


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* Train-of-four
† Post-tetanic count
‡ Second twitch


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