[NM-299] Anesthetic Management in an Infant with Walker-Warburg Syndrome

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Introduction: Walker-Warburg Syndrome (WWS), also known as HARD Syndrome, is a rare form of an autosomal recessive muscular dystrophy presenting with weakness and associated with other congenital anomalies such as smooth-appearing brain, ocular anomalies, genitourinary anomalies, cleft lip and/or palate, ear shape/placement anomalies, and other varying abnormal facies. Life expectancy is less than three years. There are only few reports of anesthesia for infants with WWS and difficult airway management has not been reported for these patients.
Case Report: Our patient was a 3 month old, 6.5 kg male. He had a prenatal diagnosis of noncommunicating hydrocephalus,suffered from hypothyroidism, GERD and decreased gastric motility, bilateral auditory neuropathy, lissencephaly and ocular anomalies. Genetic testing confirmed WWS. He presented for laparoscopic assisted gastrostomy tube placement. He had been intubated at birth and was still intubated when presenting for prior surgery to drain his hydrocephalus, but the report of his initial intubation at an outside hospital was not available. His physical exam was notable for macrocephaly, external ventricular drain, he had truncal hypotonia with hypertonic extremities. His mandible and mouth appeared small.
The patient was preoxygenated and induced via a PICC line with midazolam and remifentanil. Rocuronium was used for muscle relaxation and he was easily ventilated by mask. With a shoulder roll in place, direct laryngoscopy with a Miller 1 and Macintosh 1 blade yielded a grade 3 view to two operators. A Glidescope with a Mac 1 blade yielded a view, but it was impossible to maneuver the tube into the glottic opening. Attempts to combine direct and videolaryngoscopy with an optical stylet failed. Eventually, a supraglottic airway (Air-Q) was placed and used for fiberoptic intubation with a 3.0 cuffed endotracheal tube.
Discussion: Anesthetic experience for patients with WWS is limited and actual reports are scarce. Recommendations are typically limited around the concerns regarding the muscular dystrophy. Succinylcholine should be avoided as with any myopathy. There may be concerns regarding the use of propofol or volatile anesthetics causing rhabdomyolysis. Our patient presented with craniofacial and oropharyngeal features resulting in difficult intubation. Airway management was successful following an approach of escalating airway management equipment while providing safety through intermittent mask ventilation.
Conclusion: Patients with WWS may be at risk of difficult intubation and this should be part of preoperative anesthetic considerations.
