[NM-274] Rhizomelic Chondrodysplasia and Its Respiratory Challenges

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Purpose: Rhizomelic Chondrodysplasia Punctata (RCDP) is a rare congenital syndrome that is part of a subgroup of the larger classification, Chondrodysplasia Calcificans Punctata (CCP). RCDP is manifested clinically by shortening of long bones, cataracts, malformation of facial features, failure to thrive, and severe mental retardation. These patients often have extremely low birth weight, body length, and head circumference Diagnosis of this disease is made clinically through calcified stippling of the hyaline cartilage. Several case reports of RCSP have been written with anesthetic concerns focused mainly on tracheal stenosis. We present other respiratory challenges encountered on a 4-week-old patient with a diagnosis of RCDP, who presented for congenital cataract extraction.
Clinical features: A 4 week old, 2.7kg patient, with a past medical history of RCSP and Gastrostomy tube (G tube) dependency, presented to our institution for a cataract removal. A pre-operative clinic visit was not arranged prior to her presentation for surgery. Her past surgical history was significant for G tube placement, performed at another institution. There were no perioperative complications reported to the mother after the G tube procedure. Anesthesia induction was achieved with oxygen and sevoflurane via face mask, followed by establishment of intravenous (IV) access. Rocuronuim 3mg IV was then administered to facilitate the intubation process. Upon direct laryngoscopy, this patient was found to have a grade 3 view with deep cricoid pressure application. In between laryngoscopy attempts, it was necessary to maintain the patient on 100% fractional inspired oxygen level as she experienced significant desaturations, while also in the midst of challenging mask ventilation. After successful advancement of a 3.0 uncuffed endotracheal tube (ETT), breathe sounds were absent in the left lung fields, until the ETT was withdrawn to a depth of 7cm at the lips. This raised a clinical suspicion of a shortened trachea. Following the operation the patient was successfully extubated, and monitored overnight at in inpatient unit.
Discussion: It is essential that the anesthesiologist understands the respiratory challenges affiliated with RCSP patients prior to inducing anesthesia. While tracheal stenosis and respiratory tract cartilage changes have already been reported in the literature, our encounter with challenging mask ventilation, difficult layrngoscopic view and clinically relevant tracheal shortening, has yet to be described. Airway preparation for the care of an RCDP patient should include having a variety of difficult airway equipment as well as an experienced airway practitioner readily available. These patients stand a risk for rapid desaturations, difficult mask ventilation, difficult layrngoscopic attempts, and endobrochial intubations.
References:
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