Anesthetic Management of a Child with Progeria

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Introduction: Hutchinson-Gilford Progeria Syndrome (HGPS) is a rare disease of accelerated aging in children ending in premature death at an average of thirteen years secondary to atherosclerotic heart disease(1). There are only a handful of case reports in the literature discussing the anesthetic management of children with this syndrome (2-7). We present a child with HPGS requiring general anesthesia for mandibular plating to emphasize important considerations when caring for these children.

Case Presentation: A 4 year old, 8.7 kg male with atypical HGPS and severe micognathia developed progressive respiratory difficulty and severe obstructive sleep apnea. Imaging revealed a rare finding of an unstable, non-fused mandibular symphysis with total collapse in both the transverse and anteroposterior dimensions. He presented for dissection and mobilization of his mandibular symphysis, mandibular plating, bone morphogenic protein grafting, and glossopexy under general anesthesia. Prior anesthetic history required sedated nasal fiberoptic bronchoscopy.

The patient was admitted preoperatively to start fluid replacement with dextrose to prevent hypoglycemia and a catabolic state. Sedation was started with IV midazolam, ketamine and a dexmedetomidine infusion. A left nasoopharyngeal airway was placed for insufflation of oxygen while a right nasal flexible fiberoptic approach was undertaken. This was technically very difficult given his extremely hypoplastic mandible and minimal anterior mandibular displacement resulting in significantly decreased oro- and hypopharyngeal space. With each breath his tongue would fall back and obstruct the posterior pharynx despite a constant maximal jaw thrust and tongue pull. After securing the airway with a 3.5 cuffed ETT, particular attention was made with positioning and for adequate temperature regulation. The patient remained hemodynamically stable throughout with no EKG changes of note. Anesthesia was maintained with sevoflurane, fentanyl, and rocuronium. The intraoperative course was uneventful and the patient was extubated awake after appropriate reversal of neuromuscular blockade.

Discussion: Although HGPS is a rare condition, these patients often require surgery at some time during their short lives. Children with HPGS present many challenges to the anesthesiologist such as a potential difficult airway, advanced cardiovascular disease, risk for perioperative hypothermia and hypoglycemia, and significant osteopenia predisposing them to pathological fractures and pressure sores with surgical positioning. Despite the appearance of advanced age, emotionally and developmentally these patients are still children and interaction with them at the appropriate chronologic developmental stage is essential.

REFERENCES