Difficult Airway Associated with CHARGE Syndrome

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INTRODUCTION
Airway management for a patient with CHARGE syndrome can be difficult and planning and preparation of resources is essential.

CHARGE syndrome is an autosomal dominant genetic disorder involving the mutated gene CHD7 on chromosome 8 (1). The acronym “CHARGE” denotes the nonrandom association of coloboma, heart anomalies, choanal atresia, retardation of growth and development, and genital and ear anomalies (1). These often present in various combinations and to varying degrees. We present a clinical case that involved difficulty airway secondary facial anomalies during general anesthesia for tracheostomy.

CASE REPORT
The case involved a one day old Caucasian male born at an outside hospital via spontaneous vaginal delivery at 37 3/7 weeks by exam to a 22 y/o G3P2 female with APGAR scores of 8 at both 1 and 5 minutes. The patient received routine care in the delivery room except for the need of blow by oxygen secondary to poor color. He was noted to have complete situs inversus, dextrocardia, and severe oropharyngeal anomalies including left malformed nare, left coloboma, synthalia, left facial cyst, and left choanal stenosis. Because of poor venous blood gas (pH 6.9, PaCO2 99, PaO2 20, base excess –15), intubation was attempted but was unsuccessful. A nasal trumpet was successfully placed with an improved blood gas after bag mask ventilation. The patient remained stable on room air. He was then transported to our hospital via life flight for higher level of care. The patient was brought into the operative room the same day for tracheostomy due to the need for a definitive airway. Due to his severe facial anomalies, awake fiberoptic was initially used to visualize the vocal cords through the nasal trumpet. After the patient was given midazolam 0.25mg for agitation, the nasal trumpet was removed and replaced with 3.0 uncuffed endotracheal tube. Nasal fiberoptic was again utilized to visualize the vocal cords and place the endotracheal tube. Induction was followed with propofol and volatile agent. The patient tolerated the procedure well and the tracheostomy was uneventful.

DISCUSSION
One of the differential diagnoses for complete situs inversus and oropharyngeal anomalies is CHARGE syndrome. Up to 56% of the patients with CHARGE association have upper airway abnormalities apart from choanal atresia and cleft lip and palate (2). Up to 50% of the patients need tracheotomy for associated airway abnormalities and for salivatory retention, swallowing disorders and chronic aspiration (3). Airway difficulty often exists due to choanal atresia and cleft lip or palate. Choanal atresia is a congenital unilateral or bilateral bony or membranous obstruction of the nasopharynx which can contribute to respiratory distress or asphyxia. Both complete and partial nasopharyngeal obstruction exacerbates airway obstruction with relaxation of upper airway tone after sedation or induction of general anesthesia. Moreover, micrognathia increases the difficulty of direct laryngoscopy and predisposes the patient to upper airway obstruction. In such a patient, mandibular hypoplasia creates a limited resting spot of the tongue causing a relative macroglossia and increased incidence of obstruction and desaturation. These airway difficulties tend to be more pronounced as the patient ages. Induction and airway management should weigh the potential complications of airway difficulty with the potential increased risk of aspiration.

UPDATE
Since patient’s initial airway surgery, patient had aortic coarctation repair and G-button placement at 1 month of age. He also had left maxillary mass removal, excision left cheek appendage, reconstruction of left maxillary deformity and left facial cleft repair at 3 months old. His most recent surgeries included release of synagithia and placement of mandigular distractor. He is currently doing well at home while on humidified blow-by during the day and trach collar at night.

REFERENCES