[NM-295] Laryngeal clefts- Associated syndromes and Management

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Laryngeal clefts are congenital anomalies of the posterior larynx forming a communication between the trachea and esophagus. The incidence of laryngeal clefts is rare, occurring 1 in 10,000 to 20,000 births. We present two patients with laryngeal clefts, one with a mild and the other with a severe form.

Patient A was a 4 week old 3.8 kg baby girl born full term with cleft lip, cleft palate, atrial septal defect and ventricular septal defect who presented with coughing during feeds. Imaging showed contrast traveling from the esophagus into the anterior trachea. Direct laryngoscopy and bronchoscopy with spontaneous ventilation revealed an H-type tracheoesophageal fistula and a grade 1 laryngeal cleft. The patient underwent a tracheoesophageal fistula ligation and conservative management was recommended for the laryngeal cleft. Opitz G was considered as a possible diagnosis for the patient’s syndrome.

Patient B was a 19 day old 1.1kg born at 29 weeks who presented with respiratory distress. The patient was intubated at birth and had recurrent desaturations and bradycardia when extubated. Direct laryngoscopy and bronchoscopy were performed at bedside instead of the operating room due to desaturation with movement of the endotracheal tube. The patient was found to have a grade 3 laryngeal cleft which required surgical repair with ECMO or cardiopulmonary bypass. Due to the patient's size, prematurity, and guarded prognosis, the family decided to withdraw care.

Laryngeal clefts account for 0.2 to 1.5% of congenital airway anomalies and can be associated with syndromes such as Opitz G and Pallister Hall. Approximately 20% of laryngeal clefts can present with other airway abnormalities such as tracheoesophageal fistulas. Prognosis is dependent on the grade of the cleft, associated comorbidities, and pulmonary status. As a result, an interdisciplinary team is important for optimal management.

References

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