A Rare Case of a Type IV Laryngotracheal Cleft in Association with VACTERL

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A newborn of nonconsanguineous healthy parents, with a prenatal diagnosis of hydramnios, two interventricular communications, and persistence of the left superior vena cava, was born at 34 weeks and four days. After birth, he presented mild respiratory distress that required early continuous positive airway pressure. Attempts to initiate orogastric tube feeding on the first days of life were unsuccessful. The chest X-ray was normal. The echocardiogram confirmed the previous cardiac abnormalities. A computed tomography angiography showed an abnormal segmentation of vertebral corpses of T7, T9, T10, and T11, a partial fusion of several lower posterior costal arches on both sides and confirmed the cardiac abnormalities (Fig. 1). The upper digestive endoscopy showed an esophageal lumen that led to the stomach and one lumen that was not permeable (Fig. 2). The bronchoscopy identified a type IV laryngotracheal cleft. The renal ultrasound revealed renal pelvis dilatation of 5 mm in the left kidney. The comparative genomic hybridization array was normal. Given his myriad of clinical findings, with the absence of limb abnormalities, the patient was diagnosed with an incomplete VACTERL association.

VACTERL refers to patients with three or more congenital abnormalities on the vertebral, anorectal, cardiac, tracheal, esophageal, renal, and limb systems. Its incidence is estimated at approximately 1:10,000 to 1-4:40,000 live-born infants.[1] Laryngeal clefts are a rare congenital anomaly with an estimated incidence of 1:10,000-20,000 live births. They represent approximately 0.2%-0.5% of malformations involving the larynx.[2]

This particular case demonstrates a rare case of a type IV laryngotracheal cleft in association with VACTERL. To the best of our best knowledge, there are rather few cases describing this association, and even fewer with a successful surgical correction.[3,4]

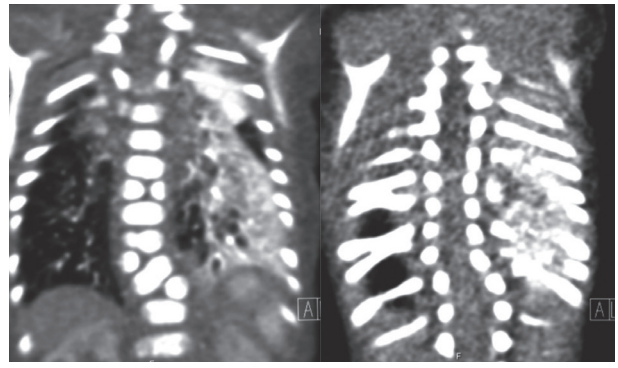

Figure 1. Thoracic tomography angiogram. The abnormality of the segmentation of vertebral corpses of T7, T9, T10, and T11 and the partial fusion of several lower posterior costal arches bilaterally.

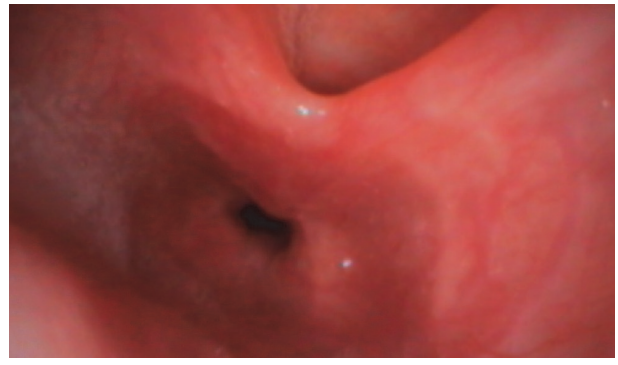

Figure 2. Upper digestive endoscopy. Absence of the posterior wall of the trachea with communication to the esophageal lumen, compatible with a type IV laryngotracheal cleft.

Keywords: Abnormalities, Multiple/diagnosis; Congenital Abnormalities/diagnosis; Infant, Newborn; Larynx/abnormalities; Trachea/abnormalities

WHAT THIS REPORT ADDS

- Familiarity with this entity is important because early treatment is imperative.
- Laryngotracheal clefts are rare, and when present, it is important to perform systematic work to evaluate the associated syndromes and anomalies.
- Delay in the management of laryngotracheal clefts and other anomalies can result in complications and a poor prognosis.

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