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LETTER TO THE EDITOR

Prenatal Diagnosis Of Congenital High Airway Obstruction Syndrome

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DEAR SIR

A healthy 33-year-old primigravida was referred to our hospital at 35 weeks of gestation due to fetal anomalies discovered on routine ultrasound (US). Repeat scan showed fetal weight 1400 g; large echogenic lungs, fetal hydrops, polyhydramnios and centrally placed heart (Fig.1a). Congenital high airway obstruction syndrome (CHAOS) was suspected and a magnetic resonance imaging was performed which showed volumetric increase of the lungs, right hand agenesis, prominent forehead, polyhydramnios, diaphragmatic inversion, fetal hydrops, as well as tracheal alterations suggestive of agenesis, although the site could not be characterized (Fig.1b).

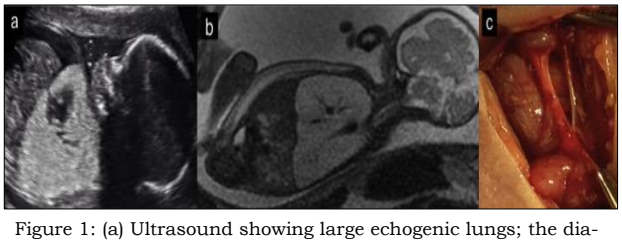

Figure 1: (a) Ultrasound showing large echogenic lungs; the diaphragm is inverted and there is disproportionately gross ascites. (b) Coronal high-resolution T2-weighted MRI at 35 weeks' gestation showing markedly distension of both lungs and diaphragmatic inversion. (c) Intra-operatory aspect of the trachea (EXIT): one centimeter below the thyroid cartilage, it consisted of an atretic strand.

The mother was submitted to an emergency caesarian section at 36 weeks of gestation due to premature labor. Ex-utero intrapartum treatment (EXIT) was performed: after the orotracheal intubation failed, a tracheostomy was attempted, but one centimeter below the thyroid cartilage, the trachea consisted of an atretic strand (Fig.1c). A decision to stop resuscitation was made. The baby was a girl with birth weight of 1455 g. In addition to the airway malformation, she had low implantation of the ears and agenesis of the right hand. The family declined a post-mortem examination.

CHAOS is a severe condition with high mortality rate, and survival beyond neonatal period is rare in cases of tracheal agenesis (TA). The incidence is 1:50.000 newborns, with 2:1 male to female ratio. [1] Once diagnosed, a detailed malformation scan is warranted to exclude conditions that may affect the prognosis, especially in countries that allow pregnancy termination. [2,3] Fetal surgery and EXIT are options to manage CHAOS, but prenatal diagnosis is essential to planning and only 2.3% of cases of TA are diagnosed prenatally. [1] For patients who are not candidates for intrauterine intervention, EXIT remains the option with the best chance of survival. [3] The objective is to evaluate and secure an airway below the level of obstruction, while utero-placental gas exchange is preserved. [4]

In cases of TA, the long-term goal is tracheal reconstruction. If tracheoesophageal fistula (TEF) is present, intubation is recommended (ventilation may be accomplished through the TEF after esophageal intubation). Surgical management, then, consists of esophageal ligation distal to the TEF, proximal esophagostomy (to drain saliva) and feeding gastrostomy until reconstruction can be planned. [1] If the intubation fails, a tracheostomy should be attempted, but success depends on finding tracheal tissue that can be intubated. [5] A systematic review by Smith et al. found that the mortality rate is 92.6% at 1 year, and none of the 7 cases without TEF survived more than 24 hours. [1] The decision

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to operate on a patient with CHAOS must be individualized, taking into account the remaining airway length, the presence of TEF and other malformations. Despite diagnostic and surgical improvements, the prognosis remains very poor and, in cases without a TEF, the condition is invariably fatal.

Consent: Authors have submitted signed consent form from legal guardian of the patient and available with editorial office.

Authors' contribution: All the authors equally contributed in concept, design, drafting of manuscript, and approved final version of the manuscript.

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