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Total anomalous pulmonary venous connection and the nutmeg lung pattern in a fetus: prognostic indicator for counseling and outcome?

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Background: The term nutmeg lung has been used to describe the heterogeneous appearance of the lung parenchyma of primary or secondary congenital lymphangiectasia. We describe the nutmeg lung appearance seen in a fetal magnetic resonance imaging (MRI) scan in a fetus with pulmonary venous hypoplasia, ventricular septal defect and hypoplastic aortic arch with an adverse outcome, with special focus on the prognostic role of MRI in counseling and preparation for delivery.

Methods: The prenatal ultrasound and MRI findings of a fetus with complex congenital heart disease and congenital lymphangiectasia were reviewed and correlated with postnatal imaging findings and clinical outcome.

Results: A male fetus was referred for a cardiac evaluation at 35 and 4/7 weeks gestational age (GA) due to bilateral effusions and hypoechoic lung parenchyma. The echocardiogram revealed total anomalous pulmonary venous connection (TAPVC) with possible pulmonary venous hypoplasia, ventricular size discrepancy with right ventricular dominance and a mildly hypoplastic aortic arch. Significant bilateral pleural effusions and a small pericardial effusion, as well as abnormal echogenicity of the lung parenchyma were also identified. Subsequent fetal MRI consisting mainly of T2 weighted sequences, revealed abnormal lung parenchyma with pulmonary lymphangiectasis with grossly dilated lymphatic channels in the subpleural, interlobar, perivascular, and peribronchial areas as well as bilateral pleural effusions. Induction of labor was performed at 38 weeks (GA) with anticipation for urgent need for catheterization and possible ECMO (extracorporeal membrane oxygenation). Immediately after birth hemodynamic and respiratory instability developed and the pulmonary venous anatomy could not be imaged echocardiographically. Due to severe cyanosis and hemodynamic instability the newborn was placed on ECMO; subsequent catheterization revealed (TAPVC) with diffuse pulmonary venous hypoplasia, absence of a decompressing vein and coarctation of the aorta. The cardiac defects were deemed inoperable by cardiology and cardiac surgery and the parents chose palliative care; the newborn expired at 27 hours of life.

Conclusions: Presence of a nutmeg lung in utero may result from secondary pulmonary lymphangiectasia due to significant congenital heart defects such as TAPVC and hypoplasia. Its identification is essential for prenatal counseling and delivery planning, frequently involving EXIT to ECMO and catheterization laboratory.