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Commentary: Neonatal pneumonectomy for unilateral pulmonary venous atresia; simple lessons in unusual times

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In this issue of the *Journal*, Chiu and Maeda,¹ from Stanford University, report on a neonate who carried the fetal diagnosis of arch hypoplasia with an abnormality of the left-sided pulmonary veins. Postnatally, the diagnosis of arch hypoplasia was confirmed, and the baby was found to have unilateral pulmonary vein atresia. As the workup revealed a lesion not amenable to surgical repair, a left pneumonectomy was performed with repair of the arch at day of life 6 with a good outcome. The pathology of the resected specimen revealed changes consistent with pulmonary lymphangiectasia secondary to pulmonary vein atresia.¹

Pulmonary vein atresia is a rare diagnosis, and most reports in the literature are case reports or small case series.^{2,3} Many of these children are diagnosed later in life. They suffer predominantly from recurrent pulmonary infections, exercise intolerance, and occasional hemoptysis.³ However, diagnosis and management of this rare entity in the newborn period has been reported before, and pneumonectomy appears to be the most successful surgical strategy applied in newborns and older children alike.⁴

With the continuous evolution of fetal diagnosis and the widespread adoption of advanced fetal imaging, the early detection of fetal cardiovascular abnormalities is helping identify patients and pathologies that otherwise would go undetected for years. In this case report, fetal diagnosis

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CENTRAL MESSAGE

Fetal diagnosis can result in finding rare congenital lesions, and surgical teams need to be prepared to perform unusual operative procedures to ensure an optimal outcome.

was accurate for the arch pathology but also could point to a suspicious pulmonary venous abnormality. This led a highly effective congenital team to the timely diagnosis of a rare and unusual condition. After that, a courageous team of skilled surgeons proceeded with the effective conduct of an unusual neonatal operation that led to a good, durable outcome.

What can one learn from all this? The obvious lesson is that a logical, well thought-out plan based on teamwork and on the study of past experiences of others can lead to success even in the most unusual and rare complex situations—a simple lesson that appears at times so complicated in the confusing and difficult times in which we all live.

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