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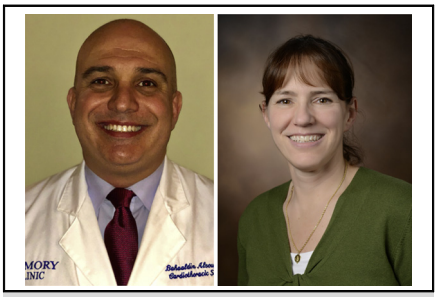
Commentary: If more is no good, then less is more

Bahaaldin Alsoufi, MD, and Deborah J. Kozik, DO



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CENTRAL MESSAGE

Lung development anomalies linked to pulmonary vein atresia can be a source of continuous morbidity and mortality. Resection of the affected lung might be better than its preservation in selected cases.

Chiu and colleagues[1] report an interesting and very rare presentation of a neonate with prenatal diagnosis of coarctation of the aorta, aortic arch hypoplasia, and left-sided pulmonary venous anomalies. After birth, he experienced respiratory distress and significant consolidation of the left lung. Workup that included echocardiography, computed tomography angiography, and pulmonary angiography eventually led to the diagnosis of left pulmonary vein atresia with a stenotic connection to a vertical vein that attached to the left innominate vein. The patient underwent sternotomy, aortic arch repair, and left pneumonectomy at age 6 days and was discharged home 32 days later.

Pulmonary vein atresia is a very rare congenital cardiac anomaly that is usually associated with diagnostic challenges leading to missed or delayed identification, in addition to management challenges due to the neonate's poor clinical condition (often requiring extracorporeal membrane oxygenation support) and associated lung disease that complicates early and late postoperative recovery.[2-5] Surgical repair of common pulmonary vein atresia has been reported in several cases, and although establishment of a patent anastomosis between the common pulmonary vein to the left atrium can be achieved (similar to surgical repair of total anomalous pulmonary venous connection), the results of surgery in common pulmonary vein atresia have varied, with morbidity and mortality largely related to associated lung disease.[2-6] There are strong associations between pulmonary vein atresia and spontaneous pneumothorax, hypoplasia of the pulmonary veins, pulmonary lymphangiectasia, and thickening of the pulmonary arteries with subsequent pulmonary hypertension. As a result, many of the patients reported in the literature required early extracorporeal membrane oxygenation, prolonged ventilation, and hospitalization for respiratory failure, in addition to showing evidence of chronic lung disease, effusions/chylothorax, and late death due to respiratory decompensation.[2-5]

In the case reported by Chiu and colleagues, the neonate had a unilateral disease. The pathophysiology was similar to unilateral pulmonary venous atresia, given that the connection to the vertical vein was stenotic. This is practically different from common pulmonary vein atresia in 2 areas: (1) there might not be a good confluence to allow safe attachment of the atretic unilateral vein to the left atrium, in contrast to common pulmonary vein atresia, where a better-formed confluence can be connected to the back wall of the left atrium; and (2) given the unilateral disease, resection of the affected lung can be an option, in contrast to common pulmonary vein atresia with bilateral lung development anomalies. Again, the significant associated lung disease can interfere with early recovery and cause significant morbidity due to lymphangiectasia and subsequent effusions, chylothorax, and respiratory compromise, in addition to the risk of future lung infections and the development of pulmonary hypertension. Despite the potential complications of pneumonectomy, this option might offer the advantage of eliminating the poorly developed lung

From the Department of Cardiovascular and Thoracic Surgery, University of Louisville School of Medicine, Norton Children's Hospital, Louisville, Ky.

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Address for reprints: Bahaaldin Alsoufi, MD, Department of Cardiovascular and Thoracic Surgery, University of Louisville School of Medicine, Norton Children's Hospital, 201 Abraham Flexner Way, Suite 1200, Louisville, KY 40202 (E-mail: balsoufi@hotmail.com).

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and related problems, and it can be well tolerated with compensation from the now-protected well-developed contralateral lung. In this case, "less is more," and the neonate might do better with 1 healthy lung than with 2 lungs where the diseased lung continues to create significant morbidity that could affect the function of the normal lung.

This is an extremely rare anomaly that many surgeons will not see in their career. However, in the event that one encounters a similar neonate or older child with unilateral pulmonary vein atresia, what should the proposed surgical treatment be? Clearly, it would depend on the morphology of the pericardial and hilar components of the affected pulmonary vein and whether or not there is a confluence that would allow safe attachment of the vein to the left atrium. In addition, it would also depend on the quality of the affected lung. However, the dilemma that remains is how can we assess the damage to the affected lung and the risk of preserving it versus resecting it? Is there a role for lung biopsy, and are there any pathological findings that could aid decision making? I suspect that this will continue to be a tough decision owing to the lack of data and experience with this very rare lesion, and that surgeons will have to rely mainly on their best possible judgment based on the available morphologic, hemodynamic, and clinical information available at the time of presentation.

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