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Prenatal Diagnosis of Left Pulmonary Arteryto-Pulmonary Vein Fistula

and Its Successful Surgical Repair in a Neonate

Pulmonary arteriovenous fistula is a rare disease. To the best of our knowledge, prenatal diagnosis of a fistula between the left pulmonary artery and the left pulmonary vein has not been described in the medical literature. We report a case of the prenatal diagnosis of a left pulmonary artery-to-pulmonary vein fistula, followed by successful neonatal surgical repair. **(Tex Heart Inst J 2015;42(2):169-71)**

irect communication between the branches of the pulmonary artery (PA) and pulmonary veins (PVs), without an intervening pulmonary bed, is a rare anomaly, the prevalence of which is 2 to 3 per 100,000 live births.¹ Central cyanosis, exertional dyspnea, and decreased arterial oxygen saturation usually accompany the lesion in neonates. We report a case of prenatal diagnosis and surgical repair of a left PA-to-PV fistula in a neonate.

Case Report

In June 2012, a 29-year-old pregnant woman (gravida III, para 0) was referred to the Ukrainian Children's Cardiac Center for fetal echocardiography at 21 weeks' gestation, for investigation of a suspected fetal cardiac malformation.

Transabdominal fetal echocardiography was performed. Color-flow Doppler mode revealed bidirectional shunting at the level of the foramen ovale, together with anomalous systolic and diastolic flow from the left PA into the left atrium (Fig. 1). Distal narrowing of the fistula was identified. Peak velocity was 273 cm/s (Fig. 2). Serial follow-up fetal echocardiograms performed at regular intervals throughout the pregnancy showed no features of fetal heart failure or hydrops fetalis.

Spontaneous labor occurred at 38 weeks' gestation with delivery of an 8.2-lb girl (Apgar scores of 6 and 7 at 1 and 5 min, respectively). The patient's blood oxygen saturation was initially 86% on room air. Transthoracic echocardiograms confirmed the presence of abnormal flow from the dilated left PA to the left atrium. Computed tomograms enabled us to definitively identify the fistula, which originated from the left PA, proximal to the takeoff of the lobar arterial branches (Fig. 3).

Because of unresponsive hypoxemia (arterial oxygen saturation, 60%), the infant underwent surgical repair at 2 days of life. Median sternotomy was performed, and cardiopulmonary bypass (CPB) was initiated via aortic and right atrial cannulation. The left PA was dissected well into the hilum, where its trifurcation was seen. To identify which vessel was fistulous, we passed a silastic loop around each of the left lobar branches, temporarily weaned the patient from CPB, and individually tightened the vascular loops—evaluating the patient's arterial oxygen saturation on each occasion. We then double-ligated the fistulous vessel; oxygen saturation reached the normal range after the neonate's separation from CPB.

The postoperative course was uneventful, with extubation at 6 hours. Transthoracic echocardiograms showed no residual left PA-to-PV shunt. The infant was discharged from the hospital on the 14th postoperative day.

At the 2-month, 6-month, and 1-year clinical follow-up examinations, the child was in good health and had normal echocardiographic results. Both of the mother's previous pregnancies had, for unknown reasons, resulted in miscarriage. Her partner and she underwent comprehensive genetic consultation, including their own testing for hereditary hemorrhagic teleangiectasia—with negative results.

Discussion

Direct communication between the branch PAs and PVs without an intervening pulmonary bed has been variously called a pulmonary arteriovenous fistula (PAVF), or a pulmonary arteriovenous malformation. A PAVF is a rare anomaly, with a prevalence of 2 to 3 in 100,000 live births and a male-to-female ratio of 1:1.5 to 1:1.8.¹

Central cyanosis, exertional dyspnea, and decreased oxygen saturation typically accompany the lesion in neonates. Sequelae are cerebral and systemic emboli, cerebral abscesses, and fatal rupture of the fistula. Untreat-



Fig. 1 Fetal echocardiogram (4-chamber view in color-Doppler mode) shows bidirectional shunting at the foramen ovale and pathologic flow into the left atrium (asterisk). See the supplemental motion image for anomalous systolic and diastolic blood flow into the left atrium.

A = anterior; L = left; P = posterior; R = right

Supplemental motion image is available for Figure 1.



Fig. 2 Fetal echocardiogram (pulsed-wave Doppler mode) shows high-velocity flow and distal constriction (arrow) of the fistula.







Fig. 3 After the patient's birth, this computed tomogram with 3-dimensional volume-rendering shows a fistula between the left pulmonary artery and left upper pulmonary vein **A**) in posterior view (fistula indicated by asterisk), **B**) in schematic representation, and **C**) in posterior-inferior view.

LA = left atrium; LPA = left pulmonary artery; LUPV = left upper pulmonary vein; MPA = main pulmonary artery; RPA = right pulmonary artery

Supplemental motion image is available for Figure 3C.

ed, PAVF can lead to heart failure. To prevent sequelae, particularly systemic and cerebral emboli, early surgical intervention should be performed. Indications for intervention are severe cyanosis or marked polycythemia.

More than 80% of PAVFs are congenital. Approximately 70% of cases are associated with hereditary hemorrhagic telangiectasia or Osler-Rendu-Weber syndrome.² Our patient (the child) had only one of the 4 diagnostic criteria of hereditary hemorrhagic telangiectasia (in accordance with Shovlin and colleagues³), which suggests that this pathologic origin is unlikely.⁴

The apparent first diagnosis of PAVF in a child was made by Thomas Churton, and was recorded thereafter by James Braithwaite as "multiple aneurysm of the pulmonary artery," in a report of a British medical society in 1897.⁴ Since then, more than 500 cases of PAVF have been described in the medical literature,² predominantly in adolescents and adults. There have been rare descriptions of the prenatal diagnosis of fistulas between right PAs and PVs.⁵⁻⁹ To the best of our knowledge, prenatal diagnosis of a fistula between the left PA and PV has not been previously reported.

We believe that clear imaging of the PVs and Doppler echocardiographic visualization of the origin of pulmonary venous flow can lead to correct prenatal diagnosis in such cases. Thereafter, accurate perinatal management and appropriate surgical timing can achieve excellent long-term results.

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