Unilateral Absence of a Left Pulmonary Artery
Successful Therapeutic Response to a Combination of Bosentan and Warfarin

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Unilateral absence of a pulmonary artery is a rare anomaly, with only 108 case reports since the first observation by Frantzel in 1868. Absence of a left pulmonary artery occurs at an even more infrequent rate, accounting for one third of all unilateral absence of a pulmonary artery. A late presentation in life, as with the patient described below, is uncommon, and delayed diagnosis may be the result of atypical symptoms earlier in life that went unrecognized. Such an anomaly can present with a wide array of symptoms including chest pain, shortness of breath, hemoptysis, fatigue, and decreased functional capacity for physical exertion. Therapeutic options are many and include surgical resection of lung lobe/tissue and medical therapy with endothelin receptor antagonists, prostacyclin, and nitric oxide. Despite evidence surrounding the vast array of therapeutic options, there has been no published data describing combination therapy using bosentan and warfarin.

A 58-year-old white woman presented for an outpatient evaluation of dyspnea on exertion. The patient initially described progressive increase in shortness of breath over the previous 6 months; however, significant dyspnea with minimal activity had surfaced over the 4 weeks before presentation.

Initial examination revealed a normal first heart sound, whereas the second heart sound had a loud pulmonic com-
ponent. A II/VI holosystolic murmur, which increased with inspiration, was noted along the left sternal border at the second intercostal space.

An ECG revealed normal sinus rhythm with normal axis and evidence of right atrial enlargement. Antero-posterior and lateral chest roentgenogram revealed prominent right atrium and increased pulmonary vessel diameter on the right side. Echocardiography revealed a mildly hypertrophied right ventricle with normal function. Dobutamine stress echocardiography revealed normal left ventricular systolic function, grade I diastolic impairment, and right heart chambers that were normal at rest with poststress hypokinesis along the inferior wall of the left ventricle. Subsequent adenosine myocardial perfusion imaging revealed no stress-induced ischemia. An initial 6-minute walk test revealed a total distance traveled of 1568 feet.

Computed tomography (CT) of the thorax with contrast from 2 years before the evaluation was reviewed and revealed dilation of the right main pulmonary artery and its branches as well as no visualization of the left pulmonary artery.

Right and left heart catheterization, with complete hemodynamic and oximetric evaluation, was performed. Right heart catheterization revealed a right atrial pressure of 11 mm Hg, right ventricular pressure of 93/18 mm Hg, pulmonary artery pressure of 76/33 mm Hg, and a mean pulmonary artery pressure of 50 mm Hg, suggesting severe pulmonary hypertension. Pulmonary vascular resistance was calculated at 6.1 Woods units and cardiac index was calculated to be 3.08 L/min/m². Right heart catheterization did not reveal a shunt. Left heart catheterization revealed collateral blood flow from the left main coronary artery (Figure 1) and right coronary artery (Figure 2) supplying the lower and upper lobes of the left lung, respectively. Pulmonary angiogram demonstrated the absence of left pulmonary artery (Figure 3).

A 64-slice multidetector CT angiography was performed with a 10-second breath hold, β-blockers to optimize heart rate below 65 beats per minute, and nitroglycerin before
image acquisition. Retrospective gating was performed to reconstruct the images. CT angiography demonstrated complete unilateral absence of the left pulmonary trunk (Figure 4) as well as left upper and lower pulmonary veins (Figure 5). Additionally, severe dilation of the right pulmonary trunk was noted. Reconstruction of the cardiac CT displayed collateral blood flow from the left circumflex artery supplying the left lower lung lobe (Figure 6) and collaterals from the right coronary artery supplying the left upper lung lobe (Figure 7).

Initial medical treatment for this patient with unilateral absence of a pulmonary artery consisted of warfarin therapy with goal international normalized ratio of 2.0 to 3.0. Additionally, the patient was started on bosentan, an endothelin receptor antagonist approved for the treatment of pulmonary hypertension.

At 6-month follow-up, the patient’s clinical symptoms had improved from World Health Organization class III to class II with improvement in 6-minute walk distance from 1568 to 1754 feet. This is the first described case of unilateral absence of a pulmonary artery treated with warfarin and bosentan to display such improvements in symptoms.

Disclosures
None.

References
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