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Brief Report

Partially anomalous pulmonary venous connection treated by interventional catheterization

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Abstract A young man had anomalous connection of the veins draining the upper lobe of the left lung to both a left-sided vertical vein and the left atrium. The ratio of pulmonary to systemic flows was 1.7:1, and he was symptomatic with evidence of volume overload of the right heart. He was successfully treated by percutaneous placement of an Amplatzer ductal occlusion device into the vertical vein.

Keywords: Laevoatrial cardinal vein; congenital heart disease; Amplatzer duct occluder

ISOLATED PARTIALLY ANOMALOUS PULMONARY venous connection has been found in from 0.4 to 0.7% of cases in adult autopsy series,[1,2] and more recently in 0.2% of computed tomographic studies.[3] Dual drainage to both the left atrium and to an abnormally connecting vein, however, is uncommon.[4] Until now, to the best of our knowledge, if patients are symptomatic, or have evidence of significant left-to-right shunting, treatment has always involved surgery.[5]

We report here a patient with anomalous connection of pulmonary venous return from the upper lobe of the left lung to both the left atrium and to a left-sided vertical vein. The vertical vein was closed successfully with an Amplatzer ductal occlusion device.

Case report

A previously asymptomatic boy, aged five, was found to have a cardiac murmur during a viral illness. The bruit was continuous in the left infraclavicular region, and was not obliterated by pressure on the neck. In 1979, he was studied by cardiac catheterization, and found to have partially anomalous pulmonary venous connection. Veins from the left upper lobe drained into an ascending vertical vein which flowed via the brachiocephalic vein to the superior caval vein. There was also a residual connection to the left atrium (Fig. 1). The veins from the left lower lobe, and those from the right lung, drained normally to the left atrium. Pressures in the right heart were normal, and,

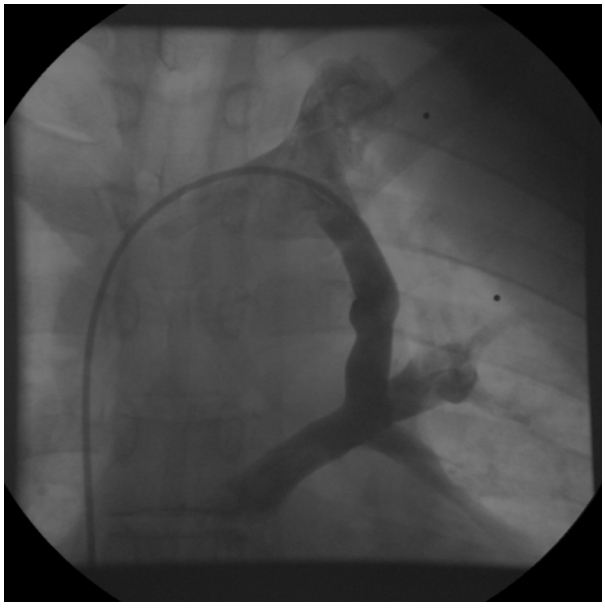

Figure 1.
A catheter has been passed via the superior caval vein and brachiocephalic vein into the left vertical vein. The pulmonary veins from the left upper lobe drain both into the vertical vein and the left atrium.

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on oximetry, the ratio of pulmonary to systemic flows was calculated at 1.3 : 1. Conservative management was recommended.

He next presented for medical care at the age of 28 years. He had a single episode of chest pain, and a history of decreased exercise tolerance. Ventricular ectopic beats, bigeminy, and short runs of ventricular tachycardia were noted on monitoring in a coronary care unit. The pain resolved, and there was no evidence of myocardial damage. An echocardiogram showed dilation of the right atrium and right ventricle.

Further cardiac catheterization confirmed the previous anatomic findings, but now the ratio of pulmonary to systemic flows was 1.7 : 1. An attempt was made to occlude the vertical vein using a 12 mm Cook coil. Unfortunately, the coil embolized to the right pulmonary artery, from where it was retrieved with Cook vascular retrieval forceps.

At a second catheterization, an Amplatzer duct occlusion device was successfully positioned in the vertical vein (Fig. 2). The vein was accessed with a multi-purpose catheter, and measured from 8 to 10 mm in diameter. A sizing balloon was used to assess the stretched diameter of the distensible vein, and an Amplatzer device of 12 by 10 by 16 mm was deployed via a long 9 French sheath, passed through the right atrium, superior caval vein, brachiocephalic vein, and into the persisting vertical vein. It was released when a satisfactory position was obtained, and there were no complications. The patient was subsequently treated with Aspirin for six months.

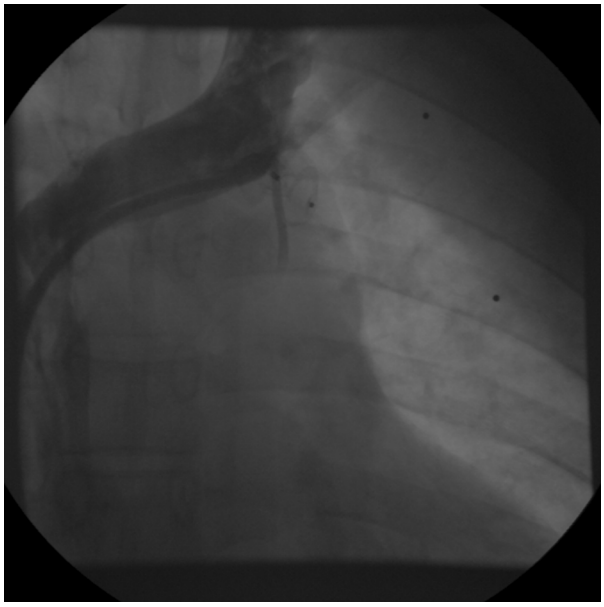

Figure 2.
Contrast injected into brachiocephalic vein shows successful occlusion of the vertical vein. The Amplatzer device is seated near the junction of the vertical and brachiocephalic veins.

Discussion

Our patient with partially anomalous pulmonary venous connection from the upper lobe of the left lung to both the left atrium and a left-sided vertical vein was asymptomatic during childhood. He had a relatively small shunt, the ratio of pulmonary to systemic flows being 1.3 : 1. No intervention was considered necessary at this stage. As a young adult, however, he developed symptoms, with echocardiographic evidence of right ventricular enlargement, and a shunt now measured at 1.7 : 1. These are indications for treatment. He was spared a thoracotomy by using an Amplatzer ductal occlusion device to obstruct the vertical vein, thus restoring all pulmonary venous return to the left atrium. This is only feasible when the anomalous pulmonary veins connect both to the left atrium and to the systemic veins.

We had initially attempted to occlude the vein using a coil, but the distensibility of the large vein, coupled with a significant shunt, led to embolization of the coil, even with purposeful oversizing. The ductal occlusion device has the advantage of being easily positioned, and can be recaptured and repositioned before final deployment.

Such interventional therapy has been described for a scimitar vein,[6] a persistent left superior caval vein which reopened subsequent to a total cavopulmonary anastomosis,[7] and a residual hepatic vein draining to the pulmonary venous atrium after a Fontan operation.[8] This device appears to be the treatment of choice in such unusual cases requiring venous occlusion. A device specifically designed for vascular occlusion will be released in the near future (by AGA Medical Corp.).

The anatomy described in our patient is known to occur in congenital lesions that obstruct the left heart, acting as a safety valve to divert pulmonary venous blood into the systemic venous circulation.[9] In hypoplastic left heart syndrome, the prevalence of such anomalous connections to a vertical vein, with normal connection to the left atrium, is said to be 2.5%.[10] These lesions can be dealt with at surgery, but catheter intervention is an alternative approach.

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