Aortopulmonary artery fistula: A rare complication of balloon dilatation of the pulmonary artery

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ABSTRACT

Acquired aortopulmonary fistula (APF) in the setting of repaired congenital heart disease is extremely rare but potentially fatal, so timely diagnosis and treatment are critical. We present a case of an 8-year-old female with a history of complex Taussig-Bing anomaly, who underwent an arterial switch procedure with LeCompte maneuver and ventricular septal defect closure early in life. The patient developed neopulmonary stenosis and branch pulmonary artery (PA) stenosis, for which she underwent patch augmentation and balloon dilatation of the left PA. The patient presented with a fistula between the ascending aorta and the left branch PA, confirmed by echocardiography and cardiac catheterization. She underwent repair of the APF with a homograft patch reconstruction of the ascending aorta.

Keywords: Aortopulmonary fistula, balloon angioplasty, LeCompte maneuver, transthoracic echocardiogram

CLINICAL SUMMARY

The patient is an 8-year-old female with a history of complex Taussig-Bing anomaly (double-outlet right ventricle with a subpulmonary ventricular septal defect [VSD] and aortic hypoplasia), who underwent arterial switch procedure with LeCompte maneuver and VSD closure early in life. Subsequently, the patient developed neopulmonary stenosis (native aortic valve) and branch pulmonary artery (PA) stenosis, for which she underwent patch augmentation at the age of 3 years. Balloon dilatation of the left PA (LPA) was performed at the age of 5 years with a 9-mm balloon (resulting in increase in left lung flow from 8% to 14%) and again at 6 years with a 12-mm balloon (resulting in increase in left lung flow from 14% to 23%). During the follow-up visit, the patient was noted to have a new continuous murmur. Echocardiogram demonstrated the presence of continuous flow from the ascending aorta into the left branch PA [Figures 1, 2 and Video 1]. The aortopulmonary fistula (APF) was therefore felt to be secondary to the balloon angioplasty rather than surgical left pulmonary angioplasty. The patient was electively scheduled for cardiac catheterization. During catheterization, saturation was 80% in the superior vena cava, 81% in the right ventricle, 86% in the right PA, and 99% in both the LPA and descending aorta. The calculated Qp/Qs was 1.4:1 with Rp/Rs of 0.08. A fistula was confirmed between the ascending aorta and the left pulmonary artery, which was the source of the murmur [Figure 3 and Video 2]. The fistula was not suitable for device closure because it was located within the residual stenosis of the ascending aorta and the residual stenosis of the LPA. Covered stent implantation into the LPA was considered, but the fistula was located in a curved segment of the proximal LPA and the covered
implantation into the LPA. Surgical revision was therefore scheduled. She underwent repair of the APF with a homograft patch reconstruction of the ascending aorta. She tolerated the procedure well and after recovery was discharged home.

**Differential diagnosis**

Aortic pseudoaneurysm, aortic dissection, anomalous coronary artery from PA.

**DISCUSSION**

Although congenital APFs are a well-known association with congenital heart disease, acquired APFs are rare. The estimated prevalence of APF is about 3.7%.[1] The etiologies of acquired APF include rupture of aortic aneurysms, trauma, aortic dissection, and infective endocarditis with prosthetic aortic valve replacement.[2,3] APF caused by rupture of an infected thoracic aortic false aneurysm after endovascular stent placement has rarely been reported.[4]

Hemodynamic consequences of APF are related to the development of left-to-right shunting, including acute pulmonary edema and right heart failure. The common symptoms of APF (in older patients) include chest pain, hemoptysis, and dyspnea or other respiratory symptoms.

Transthoracic echocardiogram is commonly the first diagnostic modality, utilizing color flow Doppler interrogation to assess continuous (systolic and diastolic) high-velocity pulmonary flow. High oxygen saturation in the PA blood and an oxygen step-up between the right atrium and the PA, as determined during cardiac catheterization, are confirmatory. Computed tomography (CT) and cardiac magnetic resonance imaging are useful to evaluate the aorta, detect valvular vegetations, and diagnose extracardiac complications.

APF should be suspected in a patient who has an aortic aneurysm and shows signs of congestive cardiac failure. Early diagnosis using transthoracic echocardiography or CT and prompt surgical intervention are crucial to a successful outcome. Although coil embolization can be performed successfully in some cases,[5] when a Lecompte maneuver has been performed, this is not possible as there is often insufficient length to the fistulous connection. Endovascular stent-graft repair of the APF can be performed in older and larger patients;[6] however, this would not be possible in a small patient. In this particular case, the patient underwent patch repair of the fistulous connection with separation of the aorta from the LPA.

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**Conflicts of interest**

There are no conflicts of interest.
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