Case Reports

A Rare Cause of Pulmonary Hypertension:

Congenital Bilateral Atresia of the Superior Pulmonary Arteries and Bilateral Stenosis of the Inferior Pulmonary Arteries

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Bilateral absence (atresia) of the superior pulmonary arteries, combined with bilateral stenosis of the inferior pulmonary arteries, has not to our knowledge been reported before now. We report such a case in a 48-year-old woman, together with the medical and percutaneous catheter interventions used to treat her condition. ***(Tex Heart Inst J 2014;41(1):73-5)***

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The absence of a main branch of the pulmonary artery (PA) was first described by Fraentzel in 1868.[1] The patient who survives to adulthood with this condition usually presents with an abnormal chest radiograph, but with no concomitant cardiovascular abnormalities, with few symptoms, and with any of a variety of erroneous diagnoses.[2] When pulmonary hypertension is present in the absence of a PA branch, the patient's condition can be improved by revascularization of the side with the absent artery. In stenotic PAs, intravascular stents have been used to good effect. We report a case of combined bilateral atresia and stenosis, a condition that to our knowledge has not previously been reported. Further, we describe the medical and percutaneous catheter interventions used to treat the patient, and their effectiveness.

Case Report

In August 2012, a 48-year-old woman was admitted to our hospital with dyspnea and fatigue. Since childhood, she had experienced dyspnea upon moderate exercise, but she had never received a diagnosis. She now described dyspnea of progressive severity over the past 4 months. On physical examination, there was mild pretibial edema, mild hepatomegaly, and a pulmonary systolic murmur. Her pulse rate was 67 beats/min, and her arterial pressure was 120/80 mmHg. Laboratory test results were all within normal limits, except for an increased D-dimer value (650 mg/dL). An electrocardiogram showed right atrial and ventricular hypertrophy. Transthoracic echocardiography revealed extensive dilation of the right atrium and the right ventricle, together with abnormally high PA pressure on systole (105 mmHg, calculated by observing tricuspid regurgitation) (Fig. 1). Echocardiograms did not show any other pathologic finding. Computed tomography showed no pulmonary emboli or thrombus. Wegener granulomatosis and Takayasu arteritis were excluded by consultation with our rheumatology staff. Because of the increased pulmonary pressure, we performed right-sided heart catheterization, upon which a vasoreactivity test was negative. The PA pressure was 98/75/65 mmHg, and the pulmonary capillary wedge pressure was 11 mmHg. During the right-sided heart catheterization, we performed oximetric tests and pulmonary angiography. The oximetric tests showed no intracardiac shunts; the pulmonary angiograms revealed bilateral superior PA atresia and bilateral inferior PA stenosis (Fig. 1). After this procedure, we excluded Noonan and LEOPARD syndromes, because there were no features characteristic of these diseases. After consulting our departments of thoracic surgery, cardiovascular surgery, and radiology, we decided in favor of PA stenting.

Our patient, however, rejected interventional therapy. She subsequently underwent outpatient therapy for 6 months: diuretic agents, aspirin, endothelin receptor antagonists, and bosentan. At the 6-month follow-up appointment, we found that functional

capacity was diminished and that PA pressure was elevated. On that occasion, we detected by echocardiography a PA pressure of 115 mmHg and severe tricuspid regurgitation.

Our patient thereupon agreed to the interventional procedure, and we used a fractional flow reserve guidewire to pass the stenosis. The distal–proximal gradient was 20 mmHg on the right side and 13 mmHg on the left side. Self-expanding stents were placed to remedy the bilateral inferior PA stenosis (Fig. 2). After the procedure, the distal–proximal gradients lessened. The patient was discharged from the hospital on the fol-

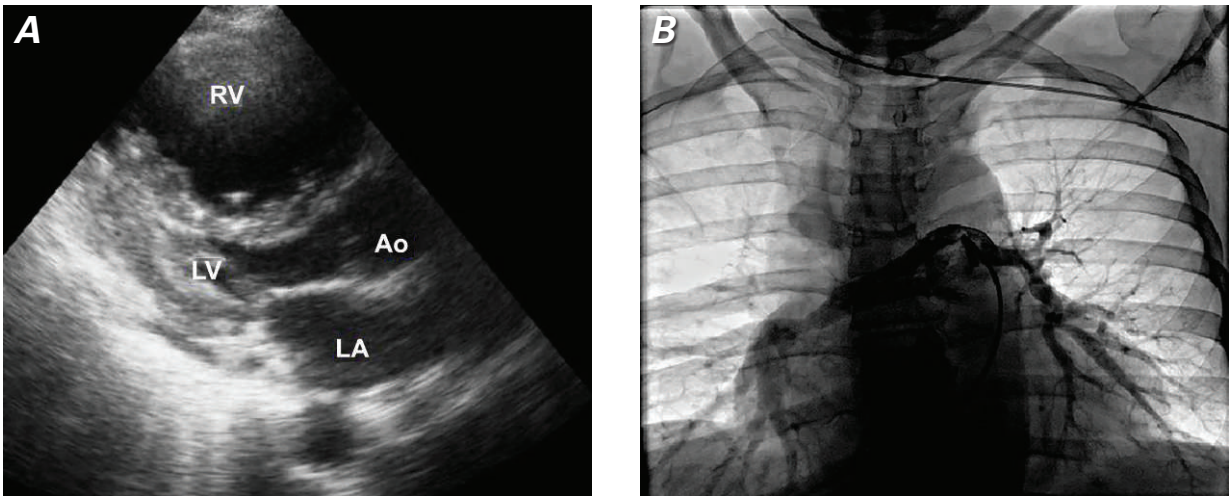

Fig. 1 ***A****) Echocardiogram shows dilated right ventricle.* ***B****) Pulmonary angiogram shows absence of the bilateral superior pulmonary arteries and bilateral inferior pulmonary stenosis.*

Ao = aorta; LA = left atrium; LV = left ventricle; RV = right ventricle

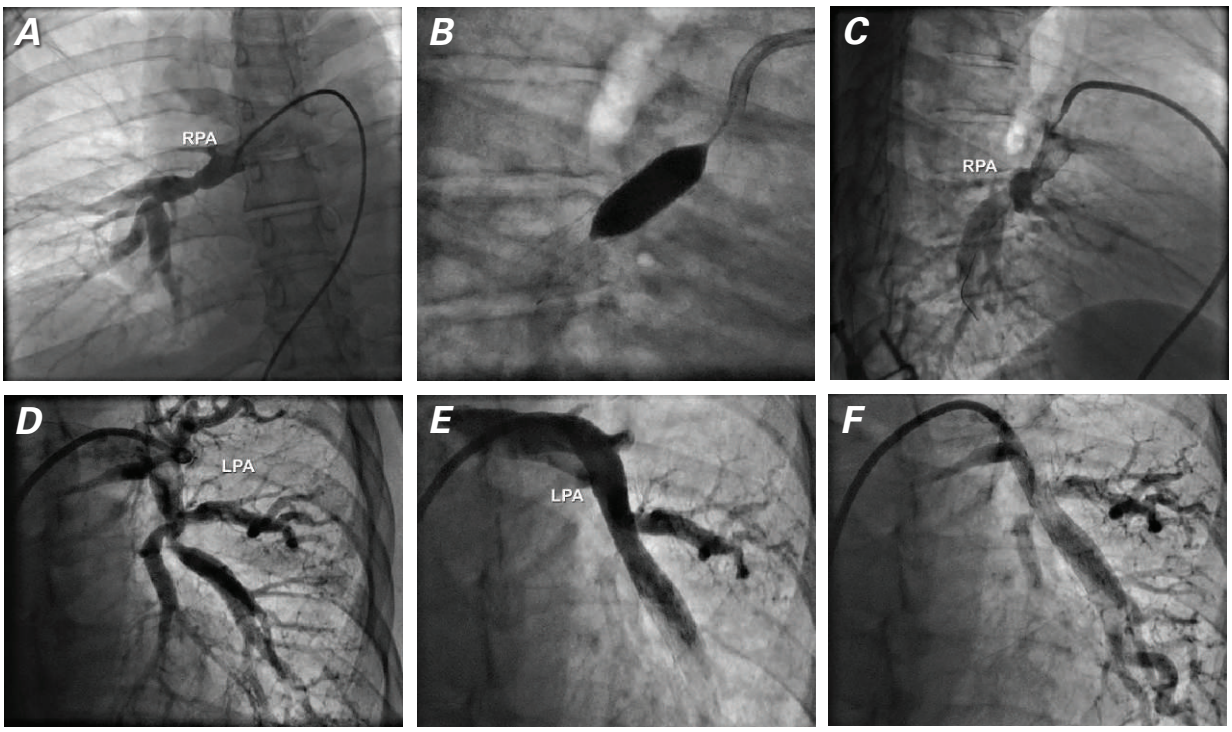

Fig. 2 *Pulmonary angiograms show* ***A****) stenosis of right inferior pulmonary artery,* ***B****) the right pulmonary artery during postdilation after stent implantation,* ***C****) the right pulmonary artery after stent implantation,* ***D****) the left pulmonary artery before stent implantation,* ***E****) the left pulmonary artery after stent implantation, and* ***F****) the left pulmonary artery after stent implantation, revealing no major stenosis.*

LPA = left pulmonary artery; RPA = right pulmonary artery

lowing day, but we continued the bosentan treatment. At the 3-month follow-up evaluation, echocardiography revealed a systolic PA pressure of 45 mmHg and a mean PA pressure of 22 mmHg. The patient's functional capacity had improved from New York Heart Association functional class III to I–II. At the 6-month follow-up after stenting, repeat echocardiography revealed the same pressure gradients, together with improvement of right ventricular size and function.

Discussion

Absence of the pulmonary arteries is a very rare condition, which according to the current literature occurs in 1 of 200,000 individuals.[3] The embryologic explanation for the absence of PAs is involution of the proximal 6th aortic arch and persistence of the connection of the intrapulmonary PA to the distal 6th aortic arch.[4] Absence of a left PA is more frequent in patients with tetralogy of Fallot or truncus arteriosus, whereas absence of a right PA is most often an isolated finding.[5] Other congenital heart defects that have been associated with the absence of PAs are coarctation of the aorta, either isolated or in combination with a ventricular septal defect; subvalvular aortic stenosis; transposition of the great arteries, either isolated or in combination with ventricular septal defect or pulmonary stenosis; Taussig-Bing malformation and coarctation; congenitally corrected transposition and pulmonary stenosis; and scimitar syndrome.[4] Although the absence of any PA is rare, the bilateral absence of PAs is, to the best of our knowledge, a condition heretofore undescribed in the literature.

Recurrent pulmonary infections, decreased exercise tolerance, chest pain, and mild dyspnea during exertion are the usual symptoms of absent PA. Our patient was admitted with dyspnea and fatigue. Although the infections are usually mild, they can have devastating effects, such as necrotizing bronchopneumonia that can lead to neonatal pneumonectomy. Less frequently found are hemoptysis and signs of pulmonary hypertension.[6]

The diagnosis of absence of a PA is difficult, especially when chest radiographic abnormalities are first noted in adulthood. The diagnosis is based on the patient's history, physical examination, and laboratory results, coupled with a high degree of suspicion. Without evidence of air-trapping on an expiratory radiograph, the posteroanterior and lateral chest radiographs show less hyperlucency than does normal hyperlucent lung. Also, magnetic resonance imaging[7] and digital subtraction angiography are useful in the definitive diagnosis of absent PA. In our patient, we discovered the pathologic condition incidentally, while we performed pulmonary angiography for pulmonary embolism after echocardiography revealed such suspicious pathologic findings as right ventricular dilation and elevated PA pressure. During this procedure, pulmonary angiography also revealed bilateral stenosis of the inferior PAs.

When pulmonary hypertension is present in a patient with absence of PA, his or her condition can be improved by revascularization of the side with the absent artery. In most cases, there is an identifiable artery at the hilum that can be used for revascularization.[8] Surgical repair of branch PA stenosis is difficult and often unsuccessful.[9] Since 1989, intravascular stents have been used successfully in treating vascular stenosis—predominantly PA stenosis. For branch PA stenosis, balloon dilation with intravascular stent placement provides a greater increase in vascular diameter and a greater reduction in pressure gradient than does balloon dilation alone.[10] For our patient, we performed balloon dilation, followed by stent implantation.

Conclusion

To the best of our knowledge, we have described the first procedure in which stents—in the bilateral absence of superior PAs—were implanted in stenotic bilateral inferior PAs. After stent implantations, the patient's PA pressure dropped and her dyspnea, fatigue, and functional capacity improved substantially. We continued bosentan treatment after hospital discharge. When a patient presents with dyspnea of unknown origin, physicians should keep in mind the possibility of PA atresia or stenosis.

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