

Very late trans-catheter recruitment of congenitally “absent” pulmonary artery

Sir,

Unilateral absence of pulmonary artery (PA) is a rare congenital anomaly caused by discontinuity and complete duct-dependency of one main PA, thereby resulting in its complete disappearance at physiologic neonatal duct closure.^[1] The ideal treatment of this malformation should be the recruitment of the discontinuous PA.^[2,3] However, this option is widely deemed unsuitable after the neonatal period due to the belief that the definitive fibrous transformation of the arterial duct (AD) results in irreversible loss of the dependent PA.

A 7-year-old child was referred due to easy fatigability and repeat events of acute dyspnea and dry cough arising during effort at high altitude. History, cardiac and respiratory examination, as well as EKG, were unremarkable. Oxygen saturation was 99%. Chest X-ray showed normal cardiac silhouette with mild hypoplasia and hypoperfusion of the left lung. Transthoracic echocardiography did not show any cardiac anomaly but a moderate dilation of the right PA and the absence of the left PA. The aortic arch was right-sided with the mirror-image pattern of the epi-aortic vessels and evidence of a tiny, blind-bottomed ductal stump arising from the left innominate artery [Figure 1a]. At computed tomography scan, this picture was confirmed and several tiny aortic-pulmonary collaterals, arising from the thoracoabdominal aorta as sole feeding vessels to the left lung, were imaged. However, despite the patient's age, interventional cardiac catheterization aiming to recruit the “absent” PA was planned, based on the hypothesis that the feeding AD was not irreversibly closed. At the hemodynamic evaluation, mild PA hypertension (mean PA pressure 24 mmHg, RV/Ao pressure ratio 0.4) was recorded. At aortic angiography, the ductal stump [Figure 1b] was successfully recanalized with a hydrophilic soft-tip coronary guide-wire, submitted to serial dilations with coronary balloons of increased diameter and finally stabilized by two telescopic implantated coronary bare-metal stents dilated to 3.75 mm (Coroflex Blue Neo, B. Braun Melsungen AG, Germany). This approach resulted in effective recruitment of a severely hypoplastic left PA (4.2 mm, z-score - 5.4) [Figure 1c and Video 1]. Postprocedure hospital course was uneventful, and the patient was discharged under anti-platelet (acetyl-salicylic acid 5 mg/kg) and mild diuretic therapy (frusemide 0.5 mg/kg).

Unilateral absence of PA has been reported in complex congenital heart malformations, including ventricular septal defects, tetralogy of Fallot, ventricular septal defect-pulmonary atresia, and heterotaxia syndrome or, less commonly, in otherwise normal heart.^[1,2] In this latter setting, it is usually asymptomatic, misdiagnosed as congenital “absence” of the PA, or even undiagnosed until adulthood. However, failure to diagnose and treat may result in hypoplasia of the associated PA, compensatory development of aortopulmonary collaterals, increased susceptibility to pulmonary infections, pulmonary hemorrhage, as well as thoracic asymmetry leading to scoliosis. In the small percentage of symptomatic patients, this malformation may be incidentally diagnosed with a consequence of respiratory symptoms in pediatric age or effort intolerance and hemoptysis in adulthood.

To date, AD stenting is considered a valuable and cost-effective alternative to surgical palliation, being

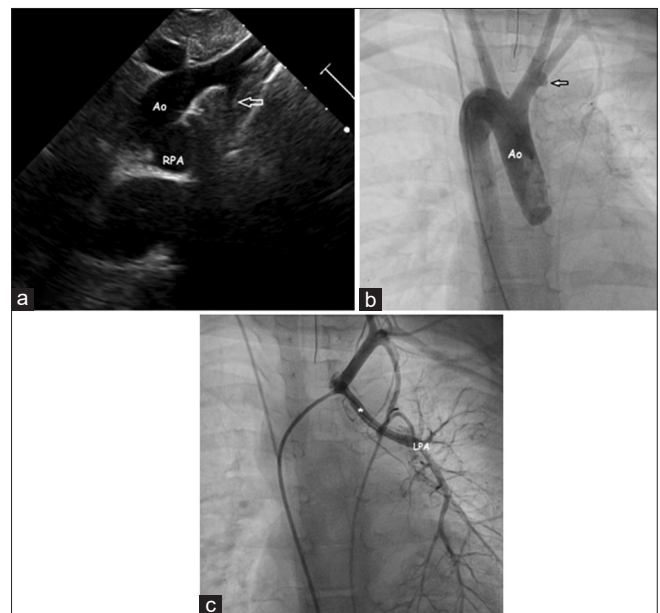


Figure 1: (a) Echocardiographic imaging of the aortic arch in supra-sternal view showing a blind-bottomed ductal stump arising from the base of the left innominate artery (arrow). (b) Aortic angiography in PA views showing a right aortic arch with the mirror-image pattern of the epi-aortic vessels and a duct stump (arrow) arising from the left innominate artery. (c) After recanalization and stenting of the AD (asterisk), a hypoplastic LPA is clearly imaged. Ao: Ascending aorta, LPA: Left pulmonary artery, AD: Arterial duct, PA: Pulmonary artery

at lower risk and promoting effective growth of the duct-dependent PA^[3,4] in view of its later surgical reimplantation. Since AD closure is “virtual” for a long period before the fibrous transformation, transcatheter duct recanalization may be successfully attempted even during early or late infancy by implantation of highly flexible coronary stents.^[3,5] Conversely, treatment of this condition in late childhood or in adults is widely deemed unworkable, owing to the belief that fibrous transformation of the AD may result in definitive loss of the dependent PA. However, as reported in this letter, the recanalization of a long-standing closed duct may be attempted as a cost-effective approach even very late after infancy, so promoting the potential growth of the dependent PA and hence improving the long-term prognosis of these patients. In our patient, the recruitment of the “absent,” although hypoplastic, PA might hopefully induce mid-term catch-up vessel growth in view of surgical unifocalization.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity.

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Conflicts of interest

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