Repeat percutaneous recanalizations of a discontinuous pulmonary artery: A very "lucky" vessel

Giovanbattista Calabri[1], Alberto Clemente[2], Giuseppe Santoro[3]

[1]Pediatric Cardiology, "Meyer" Pediatric Hospital, University of Florence, Florence, Italy, [2]"Ospedale del Cuore", Tuscany-CNR Foundation "G. Monasterio", Massa, Italy, [3]Pediatric Cardiology and GUCH Unit, Heart Hospital "G. Pasquinucci", Tuscany-National Council of Research Foundation "G. Monasterio", Massa, Italy

ABSTRACT

Pulmonary artery (PA) discontinuity with ductal origin of a major pulmonary branch is a rare congenital anomaly that can be diagnosed as an isolated lesion or in association with major cardiac malformations. Arterial duct (AD) closure results in complete disappearance of the dependent PA, thus leading to the misdiagnosis of "congenital PA absence." Neonatal AD transcatheter recanalization is considered a cost-effective approach in view of later, lower-risk surgical recruitment of the disconnected PA. However, repeat percutaneous recanalizations of a completely occluded PA, the first one as native duct-dependent lesion and the second one several months after its surgical reimplantation, have so far never been reported in the literature. This paper reports on a neonate who serendipitously received at a few weeks of age the diagnosis of "congenital" absence of the right PA. She was successfully submitted to transcatheter AD recanalization and then surgical recruitment of the dependent PA about 8 months later. However, complete occlusion of the reconnected PA was diagnosed some few months after the surgical repair. This vessel was once again recruited by percutaneous approach and it is still patent and in catch-up growth after 6 months from the second recanalization procedure.

Keywords: Interventional cardiac catheterization, pulmonary artery discontinuity, stent

INTRODUCTION

Pulmonary artery (PA) discontinuity with arterial duct (AD) origin of a major pulmonary branch is a rare congenital anomaly. It has been reported in complex congenital heart malformations, including ventricular septal defects, tetralogy of Fallot, ventricular septal defect-pulmonary atresia, and heterotaxia syndrome,[1-3] though it could be also infrequently found as isolated malformation.[4,5] In this anatomic arrangement, pulmonary flow depends on AD patency, thereby resulting in complete disappearance of the PA at the physiologic neonatal duct closure. This outcome is usually asymptomatic, and in the case of isolated PA discontinuity, it may lead to the misdiagnosis of congenital "absence" of the PA. Transcatheter AD stenting has been proposed as an appealing and cost-effective alternative to surgical palliation of this malformation,[4,6-10] in view of lower-risk surgical repair beyond early infancy. Although transcatheter PA recruitment has been performed also late after duct closure,[8] repeat recanalization procedures of the same PA overtime have not been so far reported in literature. We report on a 20-month-old infant who was submitted to two recanalization procedures, the first one on a native, isolated discontinuous PA in the late neonatal period and the second one on the same vessel about 1 year after surgical unifocalization.

Access this article online

Quick Response Code:

Website: www.annalspc.com

DOI: 10.4103/apc.APC_142_19

How to cite this article: Calabri G, Clemente A, Santoro G. Repeat percutaneous recanalizations of a discontinuous pulmonary artery: A very "lucky" vessel. Ann Pediatr Card 2020;13:163-6.

Address for correspondence: Dr. Giuseppe Santoro, Via Camillo Sorgente, 31, 84125 Salerno, Italy. E-mail: santoropino050562@gmail.com

Submitted: 23-Sep-2019 **Revised:** 15-Oct-2019 **Accepted:** 18-Dec-2019 **Published:** 20-Mar-2020

CASE REPORT

A 2.1 kg neonate, born at 34 weeks of gestation, was referred to our institution at 3 weeks of age due to easy fatigability at feeding and failure to thrive. Cardiovascular examination and electrocardiogram (EKG) were unremarkable. A chest X-ray showed mild hypoperfusion of the right lung. At echocardiography, moderate dilatation of the left PA without any evidence of the right PA was imaged, prompting to suspect an anomalous origin of the "absent" PA from an already-closed AD. Thus, interventional cardiac catheterization was planned despite an anticipated long-standing and stabilized closure of the AD. At hemodynamic evaluation, PA pressure was at half-systemic level, but it was considered as a physiologic finding of neonatal age. At aortic angiography, the left aortic arch with normal origin of the epiaortic vessels and a tiny duct stump sited at the base of the right innominate artery were imaged [Figure 1a]. In addition, the right lung was faintly and homogeneously perfused by a large, trifurcated major aortopulmonary collateral which could have been misdiagnosed as an anomalous PA arising from the descending thoracic aorta [Figure 1b]. The attempt of AD recanalization was performed using a hydrophilic, soft tip coronary guidewire (ChoICE Floppy, Boston Scientific Co., Heredia, Costa Rica) followed by the implantation of a chromium-cobalt 3.5-mm large coronary stent (Vision, Guidant Corporation, Santa Clara, CA, USA) [Figure 2a]. Finally, the aortopulmonary collateral was embolized using an Amplatzer Vascular Plug device Type 4 (Abbott-St. Jude Medical, Plymouth, MN, USA) to avoid any flow competition and overcirculation in the right pulmonary vascular bed [Figure 2b]. Based on a significant growth of the right PA imaged by serial echocardiographic evaluations, surgical recruitment of the duct-dependent PA was performed at the age of 8 months. After the surgery, the patient was lost at follow-up and reappeared at clinical evaluation about 1 year later. At that time, although the infant was completely asymptomatic, the echocardiographic examination failed to image the surgically reconnected right PA, despite a normal shape, volume, parietal thickness, and global function of the right ventricle. Computed tomography (CT) scan confirmed the presence of a complete fibrous occlusion of the PA at the site of reimplantation, so resulting in blood flow exclusion of the right lung. Thus, a second interventional catheterization was planned to try a further recruitment of the lost PA. At hemodynamic evaluation, normal PA pressure was recorded despite the total lack of perfusion of the right lung. The most suitable catheter shape was chosen on the basis of CT scan three-dimensional (3D) reconstruction to maximize the thrust force of whatsoever drilling tool we would have used. Thus, a right coronary Amplatz AR1-shaped 4 Fr catheter was leaned against the supposed origin of the right PA [Figure 3a] and through it two high load tip, chronic occlusion coronary guidewires (Conquest Pro 12, Asahi Intecc, Aichi, Japan) were simultaneously and repeatedly pushed, so progressively drilling the fibrous occlusion [Figure 3b]. Again, serial balloon dilatations and finally, implantation of 8-mm large peripheral stent (Rx Herculink Elite, Abbott, Santa Clara, CA, USA) completely recruited the vessel [Figure 3c]. The patient was discharged home under anticoagulant therapy after few days. At 3 months from the procedure, the baby was in good clinical conditions and showed a normal tolerance at effort. Thus, control cardiac catheterization, magnetic resonance imaging, or radionuclide scan were not deemed cost-benefit options in the mid-term follow-up evaluation. However, patency and perfusion of the repeatedly recruited lung were evaluated by CT scan that confirmed the normal patency and perfusion of the right PA, the diameter of which increased by 110%, from 2.4 mm (*Z*-score – 7.8) to 5 mm (*Z*-score – 2.8) [Figure 3d].

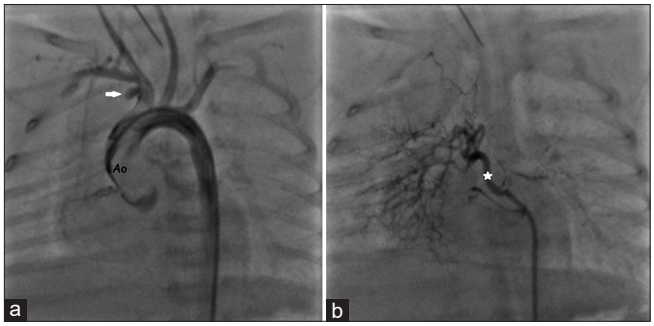

Figure 1: (a) Aortic angiography in pulmonary artery view showing a left aortic arch with a normal pattern of the epiaortic vessels and a duct stump (white arrow) arising from the base of the innominate artery. (b) Descending aorta injection imaging an anomalous aortopulmonary vessel (white star) arising from the middle thoracic aorta and distributing to the entire right lung, so mimicking the right pulmonary artery peripheral distribution. AO: Ascending aorta

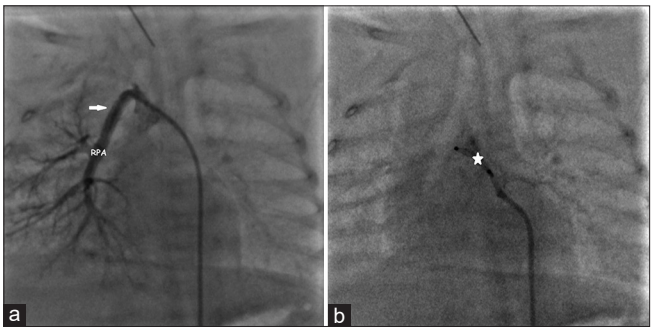

Figure 2: (a) After recanalization and stenting of the arterial duct (white arrow), the "true" right pulmonary artery is clearly imaged. (b) Finally, the anomalous aortopulmonary collateral is embolized with a vascular plug (white star). RPA: Right pulmonary artery

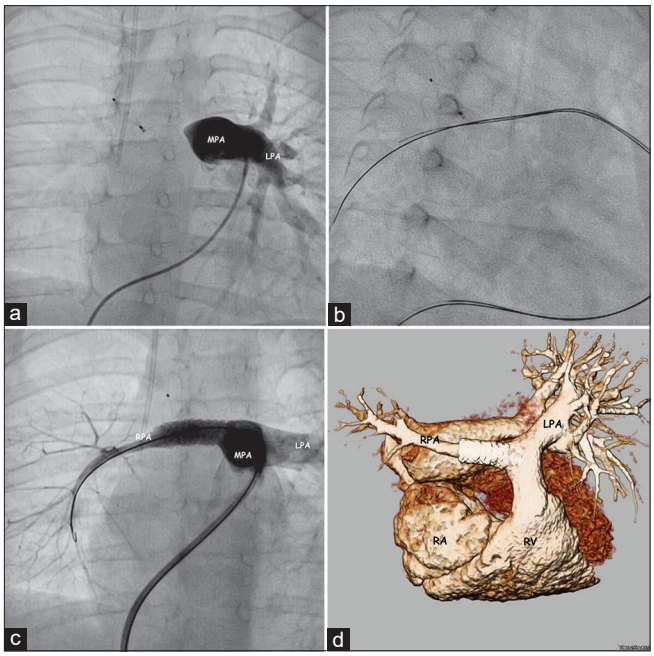

Figure 3: (a) Pulmonary angiography failing to show the right pulmonary artery. (b) The tough fibrous tissue occluding the vessel is progressively drilled using two high load tip, chronic occlusion coronary guidewires simultaneously pushed. (c) Stent implantation results in complete recanalization of the right pulmonary vascular bed. (d) Three-dimensional computed tomography scan imaging of the pulmonary artery over a mid-term follow-up. LPA: Left pulmonary artery, MPA: Main pulmonary artery, RPA: Right pulmonary artery

DISCUSSION

Discontinuity with complete duct dependency of a PA is a rare congenital malformation, accounting for 1:200,000 live births. It is frequently associated with complex congenital heart disease or, less commonly, found in otherwise normal heart.[1-5] This malformation occurs when there is involution of the ipsilateral proximal sixth aortic arch, which gives origin to proximal PA, with persistent connection between distal PA and AD, originating from the distal sixth aortic arch. In this anatomic arrangement, the affected PA receives blood flow through an AD while the contralateral PA is normally connected to the main PA. At the time of postnatal duct closure, the affected PA ceases to receive blood flow, but this does not result in cyanosis or symptoms because the remainder of the heart is normally formed and can, therefore, escape detection in the neonatal period. Thus, this entity is often described as unilateral "absence" of a PA. Failure to treat this condition may result in hypoplasia of the associated PA, compensatory development of aortopulmonary collaterals to the affected lung, increased susceptibility to pulmonary infections, pulmonary hemorrhage, as well as thoracic asymmetry caused by asymmetric lung volumes, leading to scoliosis.

In this malformation, AD stenting has been proposed as a valuable and cost-effective alternative to surgical palliation, being at low risk and promoting a significant growth of the duct-dependent PA[8,10] in view of later, lower-risk surgical reimplantation of the disconnected PA. Since AD closure is "virtual" for a long period, due to the constriction of duct muscular layer before the fibrous transformation, transcatheter recanalization and recruitment of the discontinuous PA could be possible even beyond the perinatal period. It may be quite easily performed using very soft, hydrophilic coronary guidewires and completed by implantation of trackable, highly flexible chromium-cobalt coronary stents. However, significantly more challenging and risky is the recanalization of a long-standing occluded PA after surgical reimplantation since it is presumably due to organized, tough fibrous tissue inside a vessel which, in addition, has an unpredictable spatial orientation. In this setting, CT scan 3D reconstruction might be very useful to plan any recanalization procedure, being crucial in selecting the catheter shape as well as in guiding its spatial orientation at the time of perforation. The most commonly used drilling tools are radiofrequency catheters, intravascular needles, the stiff side of coronary guidewires, or chronic occlusion coronary guidewires with different tip loads. In our opinion, using multiple high load tip, chronic occlusion coronary guidewires simultaneously pushed could be the highest cost-benefit option, in that it maximizes the drilling power and, at the same time, leaves at low risk of complications this challenging procedure.

CONCLUSION

The knowledge of the anatomic and pathologic details of PA isolation in terms of spatial orientation of the occluded vessel and nature of the occlusion, makes the most of technical and technological advances of interventional catheterization. In our opinion, percutaneous vessel recanalization, either in native or postsurgical settings, should always be considered the first-choice approach, leaving the surgical recruitment as a last chance option.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

GS is Proctor, Abbott, Italy.

REFERENCES

1. Sotomora RF, Edwards JE. Anatomic identification of so-called absent pulmonary artery. Circulation 1978;57:624-33.
2. Trivedi KR, Karamlou T, Yoo SJ, Williams WG, Freedom RM, McCrindle BW. Outcomes in 45 children with ductal origin of the distal pulmonary artery. Ann Thorac Surg 2006;81:950-7.
3. Imanaka K, Shimizu S, Matsumoto J, Hashizume K, Tsuchiya K, Takemura T. Unilateral absence of pulmonary artery and ventricular septal defect in an infant. Ann Thorac Surg 1998;66:251-2.
4. Santoro G, Bigazzi MC, Palladino MT, Russo MG, Carrozza M, Calabrò R. Transcatheter palliation of tetralogy of Fallot with pulmonary artery discontinuity. Tex Heart Inst J 2005;32:102-4.
5. Welch K, Hanley F, Johnston T, Cailes C, Shah MJ. Isolated unilateral absence of right proximal pulmonary artery: Surgical repair and follow-up. Ann Thorac Surg 2005;79:1399-402.
6. Peirone A, Lee KJ, Yoo SJ, Musewe N, Smallhorn J, Benson L. Staged rehabilitation of ductal origin of the left pulmonary artery in an infant Fallot's tetralogy. Catheter Cardiovasc Interv 2003;59:392-5.
7. Kampmann C, Wippermann CF, Schmid FX. Transcatheter recanalisation and stenting of a closed ductus arteriosus in duct dependent lung perfusion. Heart 1998;80:206-7.
8. Butera G, Santoro G, Calabró R, Carminati M. Percutaneous treatment of ductal origin of the distal pulmonary artery in low-weight newborns. J Invasive Cardiol 2008;20:354, 356.
9. Kim GB, Ban JE, Bae EJ, Noh CI, Kim WH, Lee JR, *et al.* Rehabilitation of pulmonary artery in congenital unilateral absence of intrapericardial pulmonary artery. J Thorac Cardiovasc Surg 2011;141:171-8.
10. Santoro G, Capozzi G, Giordano M, Gaio G, Palladino MT, Iacono C, *et al.* Fate of duct-dependent, discontinuous pulmonary arteries after arterial duct stenting. Pediatr Cardiol 2017;38:1370-6.