Recanalization of an occluded left pulmonary artery: A case report and review of the literature

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ABSTRACT

We report an 8-year-old male child with tetralogy of Fallot (TOF), who developed left pulmonary artery (LPA) atresia, following surgical repair of TOF and left pulmonary arterioplasty at the age of 6 years. He underwent successful radiofrequency recanalization and stenting of the LPA. The LPA exhibited satisfactory growth for 3 months, following recanalization and stenting.

Keywords: Branch pulmonary artery atresia, pulmonary artery stent, radiofrequency

INTRODUCTION

Pulmonary artery stenosis is estimated to occur in 2–3% of congenital heart disease, especially in tetralogy of Fallot (TOF) with pulmonary atresia or stenosis. The stenosis commonly occurs at the insertion of the patent ductus arteriosus (PDA) into the pulmonary artery, where it is sometimes referred to as pulmonary coarctation. Pathophysiological effects of pulmonary artery stenosis can be secondary to reduced ipsilateral pulmonary flow giving rise to dyspnea, poor lung growth, or rarely, increased right ventricular pressure.

CASE REPORT

We report an 8-year-old male child who was referred to our hospital with cardiac failure. He had TOF repair and left pulmonary arterioplasty 2 years ago at the referring hospital. Echocardiogram revealed a large residual muscular outlet ventricular septal defect (VSD), likely following VSD patch dehiscence. There was good flow to the right pulmonary artery, but flow to the left pulmonary artery (LPA) was not visualized. Cardiac catheterization was performed to delineate the LPA.

The presence of VSD was confirmed via angiogram with a Qp:Qs of 1.9:1. The right ventricular pressure was high at 80 mmHg in systole with an end-diastolic pressure of 15 mmHg while the right pulmonary artery pressure was 49/10 (mean 27) mmHg. Angiogram in the main pulmonary artery showed dilated right pulmonary artery while the LPA was completely occluded. No stenosis was noted at the right pulmonary artery. Pulmonary vein reverse wedge angiogram via transseptal puncture demonstrated patent but hypoplastic distal LPA.

A 5 Fr multipurpose catheter was used to probe the blind-ending channel of the proximal LPA from the pulmonary trunk using a 0.014” BMW coronary guidewire (Abbott Vascular, CA, USA) in an attempt at recanalizing the LPA but was unsuccessful. Through the 5 Fr multipurpose catheter at the blind-ending channel, radio-frequency (RF) energy (5 W for 1–2 s) was applied using a Cereblate PA 120 wire (Osypka AG, Rheinfelden, Germany) powered by a Baylis generator (Baylis Medical, QC, Canada). The RF wire was then replaced with the 0.014”
DISCUSSION

Branch pulmonary artery stenosis may occur at the insertion of the PDA in TOF with pulmonary stenosis or atresia. Its cause has been postulated to be due to presence of ductal tissue in the pulmonary arterial wall, which constricts as part of the PDA closure process. In series involving transcatheter treatment of branch pulmonary artery stenosis, TOF stood out to be the most common cardiac lesion to be affected. Pulmonary artery balloon angioplasty and/or stenting (rehabilitation) is one of the most common procedures performed in cardiac catheterization laboratories treating congenital heart disease.

Recanalization of occluded branch pulmonary arteries should ideally be performed as soon as possible as after the vessel is occluded; there will be organization and tissue ingrowth into the occluded segment. It is generally much easier to recanalize vessels before this tissue organization proceeds to a significant extent. The occlusion can be crossed by simple guidewire manipulation. In cases of chronic occlusion, however, direct penetration of the tissue is necessary. This type of direct penetration can be accomplished using sharp instruments such as a transseptal needle or the stiff end of a high tip load guidewire. However, there is inherent risk of failure to apply sufficient force in the required direction due to the tortuosity of the course or too much force applied causing penetration of vessel wall. In this situation, radiofrequency (RF) energy can be used to “clear a pathway” to allow for passage of guidewire with much less applied force. The flexibility of the RF wire allows better positioning at the targeted spot.

BMW guidewire over which a 2 mm × 10 mm coronary balloon was threaded and inflated at the atretic segment of LPA [Figure 1c]. The BMW wire was then replaced with a 0.035” Amplatz Super Stiff Guidewire to introduce an 8 Fr Mullin’s sheath. A 6 mm × 20 mm cutting balloon (Boston Scientific, MA, USA) was then used to allow for further balloon dilatation [Figure 1d and e]. A 10 mm × 17 mm Genesis XD (Cordis Corporations, FL, USA) was used to stent the atretic segment, and the stent was further dilated using an 8 mm × 40 mm Powerflex PTA balloon (Cordis Corporations, FL, USA). The patient was discharged 3 days after the procedure with aspirin at 5 mg/kg and readmitted 4 weeks later for revision of the VSD patch. The aspirin was planned for long term to reduce the risk of in-stent stenosis.

Repeat cardiac catheterization 3 months later showed that the LPA stent was patent and the LPA had increased in size with improved left lung arborization [Figure 2a]. A localized segment of the left lower lobe artery was stenosed (2.4 mm) as compared to segments proximal and distal to it (4.6 and 5 mm, respectively) [Figure 2b]. It was redilated using an 8 mm × 20 mm peripheral Flextome cutting balloon (Boston Scientific, MA, USA) over a V-18 ControlWire (Boston Scientific, MA, USA) with satisfactory results [Figure 2c]. In addition, a Formula 418 Expandable stent (Cook Medical, IN, USA) was implanted to the stenotic segment distal to the first stent. Post-re-intervention, the LPA and distal pulmonary arterial tree appear healthy and are of satisfactory size for future growth [Figure 2d]. A repeat cardiac catheterization 4 months after the initial intervention showed reduction of the right ventricular pressure of 41 end-diastolic pressure 8 mmHg (simultaneous aortic pressure: 91/50 mmHg, mean: 66 mmHg).
while the energy delivery is controlled. This method of recanalization showed successes in recanalizing occluded vessels.[9,10] The major disadvantage of this approach, however, is that there is destruction of tissue, rather than penetration, which may increase the chance of bleeding if the penetrating implement exits the vascular lumen.[11]

As such, it is imperative that the pulmonary vein wedge angiography is performed to determine the patency of the vessel distal to the occluded site and to ascertain that the RF wire is within the vessel lumen, following recanalization before advancing larger caliber catheter or balloon.

It is generally accepted that younger children have high potential for growth, following rehabilitation.[12] The alveolar and pulmonary arterial growth is less as the child grows. However, our patient has demonstrated that satisfactory growth may still be achievable in older children. This success suggests that rehabilitation should be attempted in a growing child irrespective of age as long as there is still growth potential.

The real challenge in such branch pulmonary artery rehabilitation lies in the feasibility of multiple redilatation of small stents to trail somatic growth. It is imperative that stents implanted permanently in a branch pulmonary artery of a growing child should be redilatable to an adult size pulmonary artery diameter later. The Genesis XD and formula stents are among those that may be used. In addition, in severe cases of stenosis – cutting balloons, which are lined with 3–4 rows of atherotomes, may be used. They have the advantage of scoring the fibrotic tissues longitudinally without applying extra vessel stress to allow for more radial expansion during balloon dilatation. This allows for placement of larger stents as in our patient.

Looking forward, bioabsorbable stents now used in coronary stenting may be a promising proposition.[13] The stent dissolves over a period thus giving rise to opportunity for future restenting using a larger stent. However, its use beyond the coronary arteries has had very limited application. They may lack the radial strength for densely fibrotic lesions, for example, in lesions similar to our case. Moreover, current available sizes are only suitable for coronary vessels.

Recanalization and rehabilitation of chronically occluded pulmonary artery with RF energy, cutting balloons, and stents is feasible and beneficial even in older children.

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Conflicts of interest
There are no conflicts of interest.

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