

Transcatheter management of combined patent ductus arteriosus and left pulmonary artery stenosis in congenital rubella syndrome: A series of three patients and an insight into case selection for intervention

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ABSTRACT

Transcatheter closure of patent ductus arteriosus (PDA) is now the standard of care with some exceptions. Best treatment for peripheral pulmonary artery (PA) stenosis in small children is still debatable. Surgical augmentation, balloon dilatation, and stenting are the available options with each having its own risks and limitations. Here, we are reporting complete transcatheter management of three cases of congenital rubella syndrome who had PDA and left branch PA stenosis by device closure and stent placement, with successful outcome in two cases and complication, leading to surgery in one. We also share our understanding of case selection for a better outcome.

Keywords: Branch pulmonary artery stenosis, congenital rubella syndrome, patent ductus arteriosus, stenting, transcatheter intervention

INTRODUCTION

Combined occurrence of patent ductus arteriosus (PDA) and branch pulmonary artery (PA) stenosis is frequently found in association with congenital rubella syndrome (CRS). This makes transcatheter management a challenge in these children, especially when they are small. While PDA ligation or device closure are standard treatment options, management of severe branch PA stenosis in children remains controversial. Both percutaneous balloon dilation and direct surgical augmentation of the stenosed artery are known to have unpredictable results.

We report three cases of CRS with PDA and proximal left PA (LPA) stenosis that were managed primarily in the cardiac catheterization laboratory with successful

outcomes in two of them. One patient developed an immediate complication that was managed by emergency open-heart surgery. In retrospect, we propose that transcatheter management of this combined lesion is feasible and related complications could be avoided by careful case selection.

CASE REPORTS

Case 1

A 2.5-year-old male child, diagnosed as a case of CRS with a history of cataract surgery, was referred for cardiac evaluation. Clinically, the patient had features of failure

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to thrive (weight 8.5 kg-below 3rd centile). He had a Grade 3/6 ejection systolic murmur that was heard all over the precordium. Systemic examination otherwise was unremarkable. The chest X-ray (CXR) showed mild cardiomegaly and differential pulmonary vascularity with less vascularity of the left lung field. A 12-lead electrocardiogram was within the normal limits.

Transthoracic echocardiogram (TTE) revealed a 2 mm PDA (Left to right shunt) and stenosis of LPA origin. The LPA measured 4 mm at origin and 7 mm distally with a pressure gradient of 40 mmHg across the stenosis. Right PA (RPA) measured 9 mm throughout its length.

Written informed consent was obtained for the procedure. The procedure was performed under general anesthesia. The right femoral artery and vein along with the left femoral venous access were established. Angiograms were performed after full heparinization (100 U/Kg). We used left anterior oblique (LAO) view with cranial and caudal angulations (LAO-cranial 70°-80°/20° and LAO Caudal 45°/25°) [Figure 1] to define the LPA stenosis. The duct was profiled in the usual left lateral view.

At first, PDA was crossed with a (4F) right Judkins catheter (Cordis de Mexico S.A de C.V) and 0.025" × 260 cm straight tip GLIDEWIRE (Terumo medical corporation, Somerset, New Jersey) from the aortic end and the wire was snared through the venous side. After that LPA was crossed with a 0.025" × 260 cm angled tip GLIDEWIRE and (5F) multipurpose catheter. The GLIDEWIRE was exchanged with Amplatzer super stiff wire (Cook medical incorporation, Bloomington, Indiana) and (5F) venous sheath was exchanged with 7F amplatzer PDA delivery system (AGA Medical Corporation Golden Valley MN, USA). An 8 mm × 20 mm × 135 cm Formula™ 535 vascular balloon – Expandable stent was passed over the Amplatzer wire and positioned across the stenosis segment of LPA and was inflated after confirming its position on angiography. After securing the stent in LPA,



Figure 1: Pulmonary artery angiogram in left anterior oblique-caudal view showing left pulmonary artery origin stenosis

PDA was closed with a 4 mm × 4 mm Amplatzer duct occluder (ADO) II (AGA Medical Corporation Golden Valley MN, USA) from the aortic end [Figure 2].

Postprocedure, echocardiogram and angiogram confirmed complete ductal closure and optimum stent flow. The patient was extubated on table and maintained on heparin infusion (10U/Kg/h) overnight which was discontinued the following day after the first dose of aspirin. He was discharged home on oral aspirin (5 mg/kg) after 48 h. Both the device and stent were found to be in position at 12-month follow-up without any complications. Importantly, he achieved good weight gain on 1-year follow-up.

Case 2

A 2-year-old boy, known case of CRS was referred for cardiac evaluation. He weighed 8 kg (<3rd percentile for age) and had a continuous murmur at the second left intercostal space. Echocardiogram revealed a 4 mm PDA and severe LPA origin stenosis. The LPA measured 3 mm at origin and 6 mm distally, with a pressure gradient of 55 mmHg across the stenosed segment. RPA measured 8 mm throughout its length.

After proper parental counseling and consent, the patient was taken up for intervention. The procedure was performed under conscious sedation. Vascular access and angiograms were performed as described before. The PDA was closed by an 8 mm × 6 mm Amplatzer™ Duct Occluder I device (AGA Medical Corporation Golden Valley MN, USA) from the venous side. An 8 mm × 20 mm × 135 cm formula™ 535 vascular balloon – Expandable stent was deployed and inflated across the stenosed LPA while still holding the PDA device with the delivery cable [Figure 3]. The device was released after the stent was completely inflated. Postprocedure, angiogram demonstrated optimum placement of both the device and stent with no local complications. Heparin infusion was continued overnight, and the patient was discharged home on low-dose aspirin (5 mg/kg). This patient has also remained well at 1-year follow-up.

Case 3

An 18-month-old baby girl, diagnosed case of CRS, was referred for failure to thrive. She weighed 6.2 kg (<3rd centile) and had right eye cataract. She had resting tachypnea and a Grade 3/6 systolic murmur over pulmonary area and back. CXR showed cardiomegaly with differential pulmonary blood flow (plethoric right lung). TTE showed 6 mm PDA, with the left-to-right shunt and LPA origin stenosis. LPA measured 4.5 mm at origin and 8 mm distally, with a pressure gradient of 50 mmHg. RPA was 8 mm.

Transcatheter PDA device closure and LPA stenting were planned under general anesthesia, and consent

was obtained for the same. The PDA measured 6 mm on angiogram. A 10 mm × 8 mm ADO I was deployed across the duct which easily slipped through. Further, a 10 mm muscular VSD device was (Lifetech Scientific Corporation, Shenzhen, PRC) used to close the duct. Keeping the device attached to its cable, an 8 mm × 20 mm × 135 cm formula™ 535 vascular balloon – Expandable stent was inflated across the stenosed segment of LPA [Figure 4].

The PDA device was released after the LPA stent was securely placed. Postprocedure, angiogram showed stable device and stent position [Figure 5]. Echocardiography showed optimum device position and mild flow acceleration across the main PA; however, the stented LPA had good flow in it. After a few hours of being shifted to the intensive care unit, the patient developed hemodynamic compromise. Repeat echocardiogram showed PA disc of the PDA device was causing near total occlusion of the main pulmonary artery (MPA) flow, resulting in acute right ventricular (RV) outflow

obstruction and RV dysfunction sets in. Within the next few hours, prominent left ventricular dysfunction was also evident.

In view of rapid clinical deterioration and emergent biventricular dysfunction, the child was urgently taken to the operation theater. Both the device and stent were removed, and the patient underwent ductal ligation with LPA-plasty. The patient was extubated within 24 h of surgery and recovered uneventfully. She was discharged home on the 5th postoperative day.

DISCUSSION

CRS is an infectious embryo-fetopathy which is estimated to affect 100,000 infants every year, mainly in the developing countries.^[1] Acquired during the first 8–10 weeks of gestation, CRS is diagnosed based on the presence of classic triad of ophthalmologic, cardiac, and auditory defects. The most common cardiac abnormality in CRS is the combination of PDA and branch PA stenosis,



Figure 2: Retrograde closure of patent ductus arteriosus by amplatzer duct occluder II device after deployment of left pulmonary artery stent

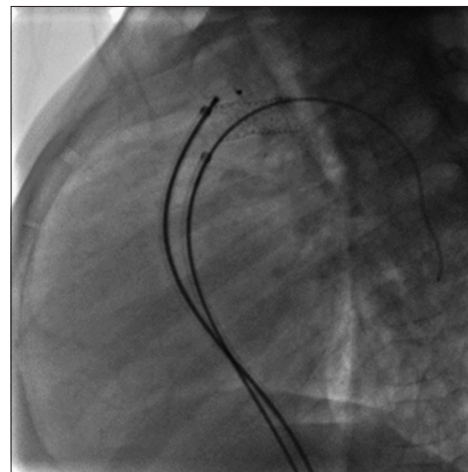


Figure 3: Deployment of stent across left pulmonary artery while the patent ductus arteriosus device was kept on hold



Figure 4: Deployment of stent across left pulmonary artery, while 10 mm muscular VSD device across ductus was kept on hold

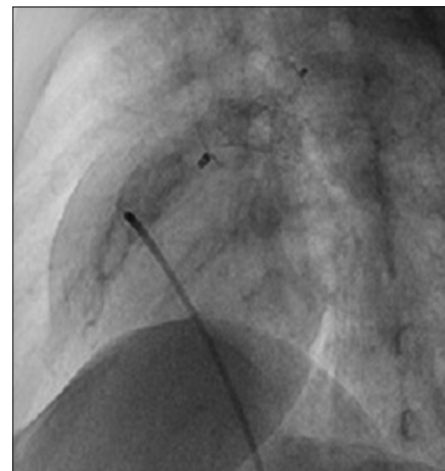


Figure 5: Post release angiogram showing stent and device *in situ*

followed by isolated branch PA stenosis and isolated PDA^[2] and other cardiac anomalies such as ventricular septal defect, atrial septal defect, tetralogy of Fallot, aortic and pulmonary valvar stenoses, coarctation of the aorta, transposition of great arteries, and tricuspid atresia. The fact that combined occurrence of PDA and branch PA stenosis which is an otherwise uncommon occurrence in the general population far exceeds any other cardiac defect in CRS, points toward predilection of Rubella virus for embryologic sixth arch structures.^[3]

Branch PA stenosis in CRS occurs in one of the three main forms: isolated or multiple stenosis of the pulmonary arteries or involvement of the bifurcation of the pulmonary trunk (coarctation of the PA or membrane above the pulmonary valve).^[4]

Branch PA stenosis may also occur with other congenital syndromes such as Williams syndrome, Alagille syndrome, and Noonan's syndrome or maybe acquired postoperatively. Children with branch PA stenosis are at a risk of developing ipsilateral PA hypoplasia^[5] and spontaneous disconnection from the main PA.^[6]

Intervention in branch PA stenosis is indicated in case of severe stenosis as evidenced by a pressure drop of more than 20 mmHg across the stenosed segment, elevated RV or proximal main PA pressure >50% to two-thirds of systemic pressure, or when there is differential pulmonary flow documented on the lung perfusion scan to be 35%: 65% or worse.

Branch PA stenosis is managed either surgically or by percutaneous intervention. Surgical techniques to manage these branch PA stenoses include pericardial or Gore-Tex patch augmentation, pericardial pedicle graft, and "lay open technique" (described by Kaneko *et al.*). However, technically demanding nature of surgical management combined with high rates (35%–50%) of restenosis at mid- and long term led to the introduction of balloon angioplasty in the early 1980s.^[7] An immediate success rate of 32%–72% is reported with standard balloon dilatation.^[8]

Acute failure of balloon angioplasty is attributed to high resistance to vessel tears, vascular recoil, and compression from the adjacent structures. Consequent to tissue recoil and scarring, restenosis is reported in up to 35%–50%, and complications such as PA aneurysm are seen on long-term follow-up.^[9] Because of the unpredictable and unsatisfactory long-term results following surgery and plain balloon angioplasty, PA stenting was introduced in 1991, which showed promising results in maintaining long-term vessel patency and scope for further dilatations to adult size.^[10] In addition, hybrid procedures with intraoperative placement of stents under direct vision are also another known treatment modality.^[11]

PA stenting is now widely accepted as the first-line therapy of branch PA stenoses with simultaneous management of additional lesions amenable to treatment by interventional therapy, in catheterization laboratory. The kind of stent to be implanted depends on the anatomic situation, the age and size of the patient, and the vascular access status. Usually, stents that may be re-dilated up to at least 18–20 mm to accommodate somatic growth in children are preferred. In very small children (below 10 kg), to date, palliative approach is considered with the use of smaller stents and a commitment for surgical removal or enlargement of these stents in future. However, with the advent of newer low profile, premounted, balloon-expandable stents, which can be expanded to up to two times the original size, definitive stenting of branch PA in small patients is made possible. The new generation hybrid open-cell design holds promise for significant postdilatation without marked shortening. The premounted system, with monorail or over the wire design, has good radial strength and shortens minimally even with over-dilatation up to 100% of its nominal diameter, which is a very important feature for any stent implantation, especially in small infants.^[12]

Device closure is the standard procedure for ductal closure, with very low complication rates. Accomplishing both ductal device and PA stent in the interventional laboratory remains an attractive and minimally invasive treatment option in children with CRS. Transcatheter intervention is a feasible option for coexisting PDA and branch PA stenosis in children with CRS, which not only saves them from the surgical risk but also, in appropriately selected patients can give very good long-term results.

We realized in retrospect that the rubella being an embryopathic virus may hamper the growth of the main PA segment in addition to affecting its branches. In the presence of a large PDA in a small child, especially in association with pulmonary arterial hypertension, such as in our third patient, any protrusion of PDA device into the MPA segment should be avoided as there is a potential for MPA obstruction.

CONCLUSIONS

The combination of PDA and branch PA stenosis can be successfully treated by transcatheter intervention. However, case selection is of critical importance in optimizing outcomes. To date, surgery is the preferred option for combined lesions with large ducts.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for their images and

other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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