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Stenting Of Branch Pulmonary Artery In An Adult Patient With Repaired Tetrology Of Fallot With Right Pulmonary Artery Origin Stenosis: A Case Report

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Abstract
Now a days due to improved paediatric cardiac care we see patients with repaired or unrepaired congenital heart diseases in adult age more often. Pulmonary artery (PA) stenosis is a common association with Tetrology of Fallot (TOF). It is common to see some degree of residual pulmonary artery stenosis post intra cardiac repair. These stenosis can, sometimes, progress and present in adulthood. Management of patients with peripheral or branch PA stenosis have primarily been catheterization with surgical management of these cases reserved for limited indications. Here we report a case of Right PA ostial stenosis in an adult patient with history of surgical correction of TOF in childhood. We also describe the procedural aspects in the correction of this defect. We here discuss the indications, contraindications and complications of transcatheter and surgical approach. Lack of practice guidelines and recommendations with respect to patient selection, hardware selection, follow up, antiplatelet regimen and operative hemodynamics makes this procedure difficult to learn, monitor and follow.

Keywords
Pulmonary artery stenosis; Pulmonary artery stenting; Peripheral pulmonary stenosis.

Introduction
The incidence of congenital heart disease in general population is around 0.8 per 1000 live births. With advanced paediatric cardiac care more and more children with congenital heart disease grow to enter the adult age group. It is not uncommon to see patients coming with significant cardiac symptoms having history of repaired or unrepaired congenital cardiac defects. Management of these groups of patients require special skills and competence. Peripheral pulmonary artery stenosis or branch pulmonary artery stenosis can present as an isolated lesion or in association with other cardiac defects. Up to 20% of patients with Tetrology of Fallot (TOF) have some degree of pulmonary artery stenosis. According to the location, four types of pulmonary artery stenosis can be defined i.e. Stenosis of the pulmonary trunk, Stenosis of the pulmonary artery bifurcation with extension into right or left branch pulmonary artery (PA), Multiple peripheral PA stenosis, Stenosis of both the pulmonary trunk and peripheral arteries.[1] Management of patients with peripheral or branch PA stenosis have primarily been

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catheterization followed by balloon angioplasty or stenting with surgical management of these cases reserved for limited indications. Here we report a case of Right PA ostial stenosis in an adult patient with history of surgical correction of TOF in childhood.

Case Summary

Our patient was a 21 year old female who had history of surgical repair of TOF at the age of 7 years with patch closure of ventricular septal defect and patch repair of right ventricular outlet obstruction. She came with chief complaints of exertional breathlessness and fatigue, New York Heart Association (NYHA) class II, for last 2-3 year which have been insidious in onset and gradually progressive. Her clinical examination was not suggestive of any significant abnormality. Her electrocardiograph (ECG) (Figure 1) showed sinus rhythm with incomplete right bundle branch block with secondary ST-T abnormalities. Her chest radiograph showed mild cardiomegaly with sternal sutures in place. On echocardiogram she was found to have significant right pulmonary artery (RPA) stenosis with peak/mean gradient of 65/42 mmHg across the RPA ostium. However the degree and length of stenotic segment could not be elucidated on echocardiogram so patient was advised to undergo Computed Tomography (CT) of Heart. On CT Cardiac examination (Figure 2) she was found to have partial thrombosis of BT shunt between left subclavian artery and left pulmonary artery, double superior vena cava with left sided draining into coronary sinus, and right pulmonary artery ostial stenosis for a length of 2.3 cm with calibre of 5 mm; main pulmonary trunk measured 28 mm and left pulmonary artery measured 16 mm. After proper counselling and consent patient was posted for pulmonary artery stenting.

Procedure Details:

One day prior to the procedure patient was given loading with clopidogrel 600 mg and aspirin 300 mg. Patient was posted for RPA stenting under local anaesthesia with mild IV sedation. Right femoral artery and venous access was obtained with 7F and 9F introducer sheath respectively. 70 IU/Kg Heparin was given intravenously. Right ventriculogram using NIH catheter (Figure 3) was diagnostic for long RPA stenosis measuring 32.44 mm in length with diameter at the point of maximum stenosis of 6 mm whereas the dimensions of main pulmonary artery and right pulmonary artery post stenosis was 25.8 mm and 13.4 mm respectively. There was also evidence of mild pulmonic regurgitation. Patient's hemodynamic data was obtained (Table 1).6F 3.5 JR diagnostic catheter over double length 0.035 inch angled tip terumo wire was used to cross across the lesion for pressure measurement. Another 6F right femoral venous access was obtained and NIH catheter was passed into right ventricle up to pulmonic valve to mark its position so that the stent is deployed well above pulmonic valve. The terumo wire was exchanged for 0.035 inch 260 cm Amplatz ultra stiff wire. With wire position across the lesion the JR catheter was removed. Amplatz wire was then loaded with Bard Venova self expanding venous stent system with 14 X 60 mm stent dimension. Extra length of stent was taken to keep a margin of 5 mm on both ends of the lesion. Venous stent system was placed across the lesion under fluoroscopy and gently released into right pulmonary artery to main pulmonary artery (MPA) (Figure 4). Post stenting right ventriculogram (Figure 5) was suggestive of significant increase in minimum RPA diameter to 11 mm from 6 mm previously. Post procedure hemodynamic data was obtained (Table 1). There was significant decrease in right ventricular and pulmonary artery pressure post stenting, without any increase in pulmonary regurgitation. Post procedure 2d Echo done showed stent in good position in RPA and MPA well above the pulmonic valve. Procedure was uneventful. Patient tolerated the procedure well and was discharged on dual antiplatelets (Aspirin 75 mg OD and Clopidogrel 75 mg OD).

Discussion

Many patients with TOF have significant residual branch PA stenosis after surgical repair.[2] And in some cases it may increase in severity as the age progresses. These patients have reduced exercise tolerance compared with other patients with TOF.[2] The common indication for treatment of such patients include- Right ventricular hypertension (right ventricular pressure > 2/3 systemic pressure), marked asymmetry of pulmonary blood flow (ipsilateral lung perfusion <35% than predicted in unilateral stenosis), or Segmental PA hypertension (mean distal pressure > 25 mmHg).[3]

The treatment options for these groups of patients include surgical correction, balloon angioplasty and stenting. Surgical correction has high peri-operative complication and mortality rates in such patients primarily owing to the fibrosis consequent to previous surgical intervention. Since the results of surgery for peripheral pulmonary stenosis have been quite unsatisfactory, transcatheter intervention remains the first line therapy for these patients.[4] In a study done by Bush et al, where he defined successful balloon angioplasty as 50% or more increase in predilatation diameter, whereas restenosis was said to occur if there was 50% or more loss in initial diameter gain, he found angiographic success rate of 64% and restenosis rate of 35% in one year follow up.[5] Balloon angioplasty is associated

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significant complications such as vessel rupture, dissection, aneurysm formation or even death and is often followed by recurrence. In the early literature the success rate of stenting was found to be 72.5%.[6] However, the more recent data suggests that the success rate for stenting is around 85% where success was defined as Post-stent PA vessel diameter/pre-stent distal PA diameter more than 0.7 and/or >50% decrease in gradient across the stenosis and/or >25% decrease in sub-pulmonic ventricular pressure.[7] The complication(s) seen with pulmonary artery stenting are bleeding event, device embolization), airway event requiring intubation, event requiring ECMO (extra corporeal membrane oxygenation, cardiac arrest, emergency cardiac surgery, other surgery due to cath complication, air embolus, tamponade and embolic stroke. The overall complication rate is around 14% with 9% being classified as major complications requiring intervention to avoid significant morbidity or mortality.[7] The intra-procedural mortality with pulmonary artery stenting has been found to be 1.7%.[7] Also the restenosis is very rare after successful stenting with rates as low as than 5% in one year follow up.[8] In another study transcatheter pulmonary artery intervention were associated with 22% risk of procedure-related adverse event, with major complications occurring in up to 10%.[9] The risk of complications were higher if there was hemodynamic vulnerability, emergency surgery, low patient weight, use of cutting balloons, balloon dilatation at higher pressures, as well as operator experience of <10 years or in a centre where such procedures are not routinely performed.

In cases with peripheral pulmonary artery stenosis balloon pulmonary angioplasty (BPA), specially in paediatric age group, still remains the acceptable first line option with stenting preferred for central or proximal branch pulmonary artery stenosis (proximal to branching points), stenosis due to kinking or tension (post-Lecompte manoeuvre), compliant obstructions that fail balloon dilation, external compression (i.e., left PA stenosis from compression by oversized neo-aortic reconstruction post-Norwood Stager I), Stenosis due to obstructive intimal flaps, restenosis post-BPA.[3] Given the current higher success rate of stent implantation, older children with proximal PA stenosis are managed preferably with stenting.

Surgical correction have been reserved for proximal peripheral pulmonary stenosis in association with heart defect requiring repair, supra-valvular pulmonary stenosis where stent placement is difficult due to adjacent pulmonic valve, stenosis at the PA bifurcation, Stenosis after PA band application, stenosis at Blalock-Thomas-Taussig shunt insertion site, or after failure of transcatheter techniques.[3]

At present there are no practice guidelines or consensus recommendation about antiplatelet regimen to be followed after pulmonary artery stenting. We used dual antiplatelet therapy in our patient with combination of aspirin and clopidogrel. Limited stent designs are available in market for pulmonary artery stenting including balloon expandable stents and self expanding stents system. We used Venova self expanding venous stent system. There had been no randomised head to head trials between different stent designs to suggest superiority of one over another.

Successful angioplasty results in our patient is suggested by post-stent PA vessel diameter (11 mm)/pre-stent distal PA diameter (13.4 mm) more than 0.7, >50% decrease in gradient across the stenosis (44 mmHg pre-stent to 12 mmHg post stenting) and >25% decrease in sub-pulmonic ventricular pressure (from 60 mmHg pre stent to 44 mmHg post stenting).

Conclusion

There is limited literature of pulmonary artery stenting in adult population. Lack of practice guidelines and recommendations with respect to patient selection, hardware selection, follow up, antiplatelet regimen and operative hemodynamics makes this procedure difficult to learn, monitor and follow. Also there are no multicentre prospective trials to assess long term mortality and morbidity benefits associated with the procedure.

Table 1: Pre and Post Procedure Hemodynamic Data

	Pre Procedure (mm/mmHg)		Post Procedure (mm/mmHg)	
	Dimension	Pressure	Dimension	Pressure
Aorta		148/72		146/72
Right Ventricle		60 systolic		44
Main Pulmonary Artery	25.8	60/10		34/15
At RPA lesion	6		11	
Distal Right Pulmonary Artery	13.4	18/7		22/14
Gradient Across RPA Lesion		42		12

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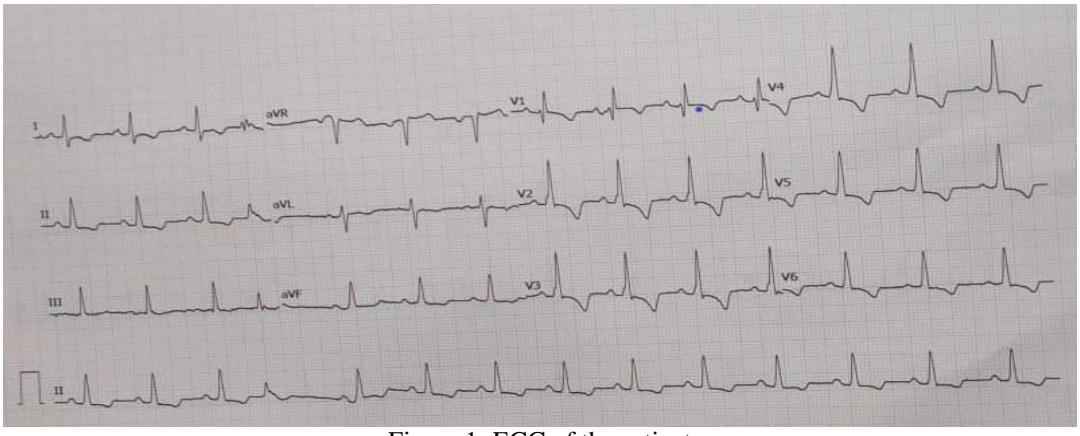

Figure 1: ECG of the patient

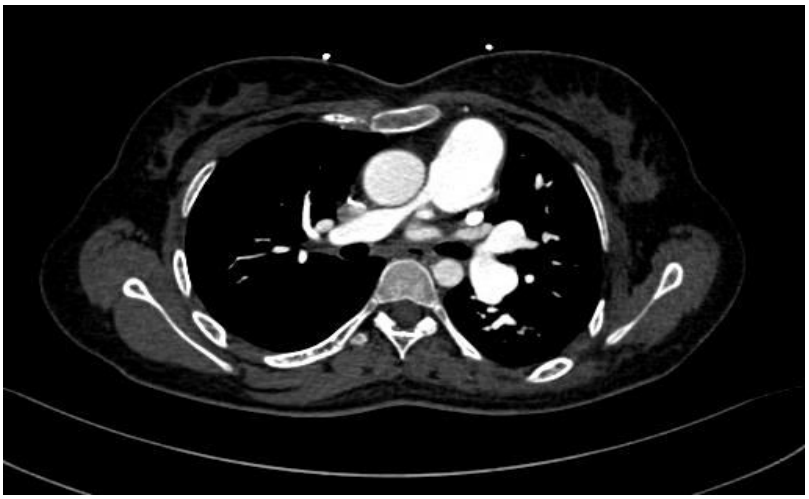

Figure 2: CT Cardiac showing significant RPA ostio-proximal stenosis

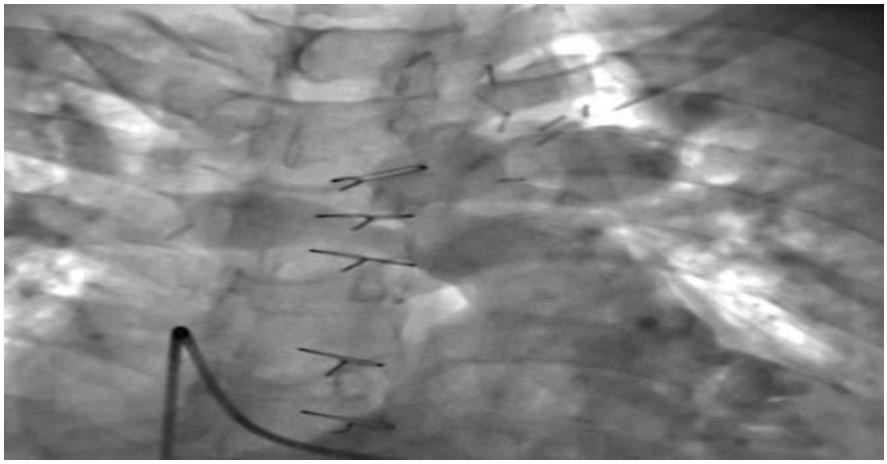

Figure 3: Right Ventriculogram depicting RPA stenosis.

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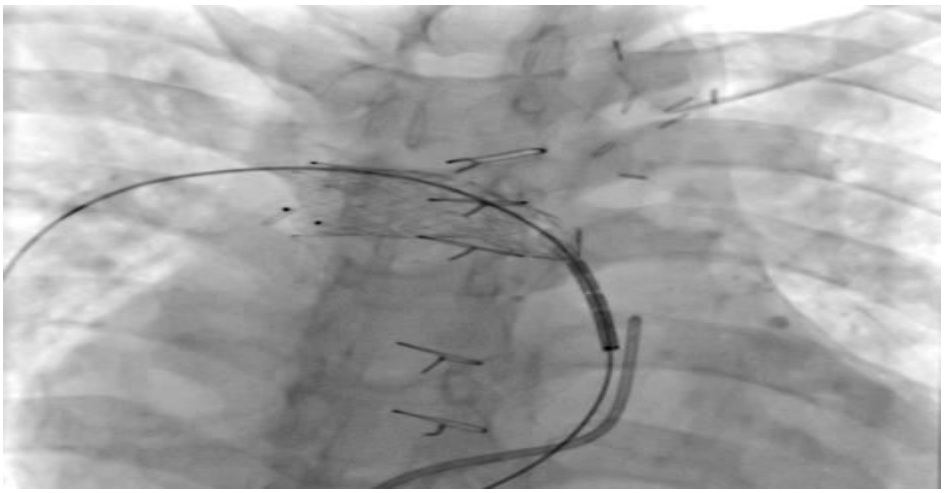

Figure 4: Self Expanding Stent being deployed under Fluoroscopy

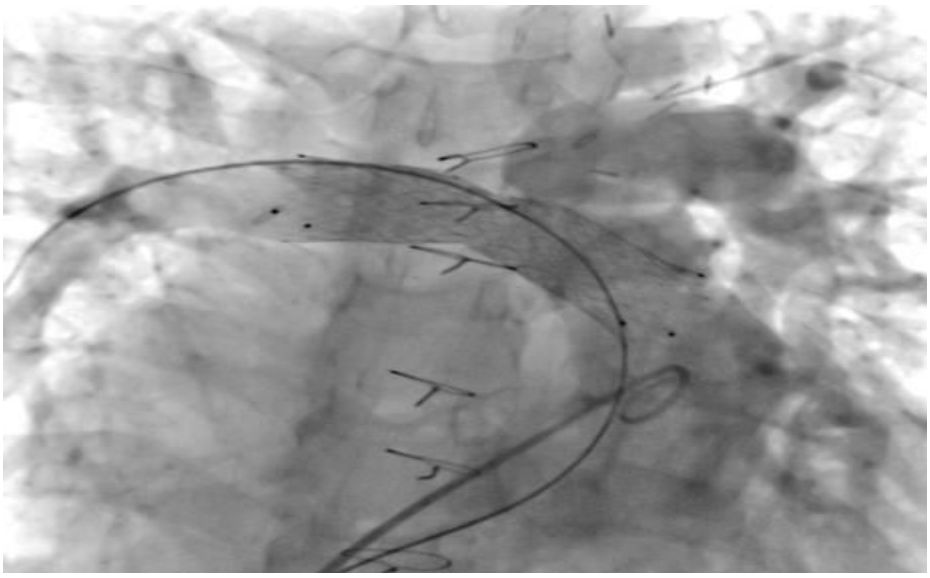

Figure 5: Post Stenting Right Ventriculogram showing significant increase in minimum RPA diameter.

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