

## VERTICAL PDA STENTING IN PULMONARY ATRESIA WITH INTACT INTERVENTRICULAR SEPTUM

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Date Received: December 29, 2013

Date Revised: January 26, 2014

Date Accepted: March 03, 2014

### **Contribution**

All the authors contributed significantly to the research that resulted in the submitted manuscript.

**All authors declare no conflict of interest.**

### **ABSTRACT**

Pulmonary atresia with intact interventricular septum (PA-IVS) is a rare congenital lesion. PA-IVS involves complete blockage of the pulmonary valve located on the right side of the heart. This blockage thus prevents the flow of blood to the lungs. Because of this lack of blood flowing through the right side of the heart, the structures on that side, such as the pulmonary valve and the tricuspid valve, may be abnormally small. The genetic cause of PA-IVS is unknown. We report a 6-month-old infant with congenital heart disease including; pulmonary atresia, intact ventricular septum, right ventricular hypoplasia, and vertical persistent ductus arteriosus (PDA) arising from left subclavian artery with stenosis at pulmonary end. PDA stenting was performed with successful hemodynamic after procedure.

**Key Words:** Pulmonary Arteria, Intact Ventricular Septum, PDA Stenting

### **INTRODUCTION**

Pulmonary atresia with intact ventricular septum (PAIVS) is a disease having morphologic variability, it affects not only the pulmonary valve but also the RV cavity, the tricuspid valve and coronary arteries. PAIVS involves complete blockage of the pulmonary valve located on the right side of the heart. This blockage thus prevents the flow of blood to the lungs. Because of this lack of blood flowing through the right side of the heart, the structures on that side, such as the pulmonary valve and the tricuspid valve, may be abnormally small. its genetic cause is unknown. It is a rare congenital cardiac defect which occurs in about 46 cases per every 100,000 live births.<sup>1</sup> its Surgical management depends on variables such as right ventricular (RV) size, tricuspid valve diameter and coronary anatomy. Fistulous connections between the small right ventricle and the coronary arteries may exist, which results in a dual source of coronary perfusion. In that case usually there is stenoses or complete interruption of the coronary arteries so the myocardial perfusion depends on the right ventricle. In contrast, there is usually uniformity in the anatomy of central pulmonary arteries (PAs), almost always normal sized, and in the source of pulmonary blood flow, generally a left sided arterial duct. Patients in PAIVS depends on ASD or PDA.<sup>2,3</sup> Where PDA is the only source for pulmonary circulation, then it should be patent enough to allow good pulmonary circulation. But as the patient get older, the PDA

get stenosed, so patient either needs surgical total correction or palliative procedures i.e Blalock Taussig (BT) shunt or PDA stenting.

We are here reporting a case PAIVS with least common type of PDA, comprising less than 5% of PDA type, which has the peculiar appearance of a BT shunt.<sup>4</sup> This long tubular ductus arises from the left subclavian artery (LSCA) (or from the right subclavian artery in a right aortic arch) and joins with the pulmonary artery perpendicularly. There is usually a tight constriction at its insertion site of pulmonary artery. Similarly was the constriction in our patient which was successfully stented.

## CASE REPORT

A 6 months old baby presented with easy fatiguability, cyanotic spells palpitation and shortness of breath. Echo shows large ventricular septal defect (VSD), Tetralogy of Fallot (TOF) with pulmonary atresia, moderate size PDA. CT scan shows large vertical PDA (Figure 1). Saturation was 54% at room air, so it was decided to go for PDA stenting with total correction of TOF in future. Right Femoral Artery was cannulated with 5 F sheath and Right Femoral Vein was cannulated with 6 F sheath. MPA 2 catheter used to cross SVC and angiogram taken showing right sided SVC and left innominate vein. RV angiogram showing overriding of aorta with severe pulmonary atresia with no forward flow and good size RV. Pressures were taken as, RV= 70/10 mmHg, Ao pressure 79/36(52)mmHg and LV=80/7mmHg. Measurements were taken which were showing Descending aorta = 10mm, MPA = 14.2mm, LPA = 9.75mm, RPA=8.25mm and MC goon ratio=1.8. Catheter passed through descending aorta to subclavian artery as shown in Figure 2 & 3. Injection given showing PDA of length 35mm arising from subclavian artery with severe pulmonary end

**Figure 1: CT Angiogram Showing Large Vertical PDA**



**Figure 2: Angiogram Showing PDA with Constriction at its Distal End where it Joins with Pulmonary Artery**



stenosis of PDA as shown in Figure 2. BMW wire used to cross PDA as shown in Figure 3. Direct stenting with Driver Stent of 4x15mm at 18 atm done as shown in Figure 5. Patient saturation improved from 62% to 90%. There was also small collateral from subclavian to LPA. Optimal stenting was done. Patient was given low molecular weight heparin and was stated on antiplatelets. No complication occurred.

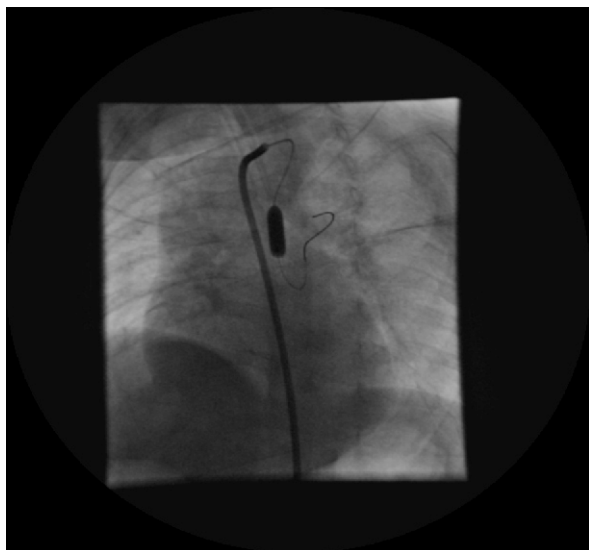
## DISCUSSION

Ductal stenting is preferable now in some centers as there is waiting list for this critical surgery and sometimes the

**Figure 3: Angiogram Shows Wire Crossed Through PDA From Aorta**

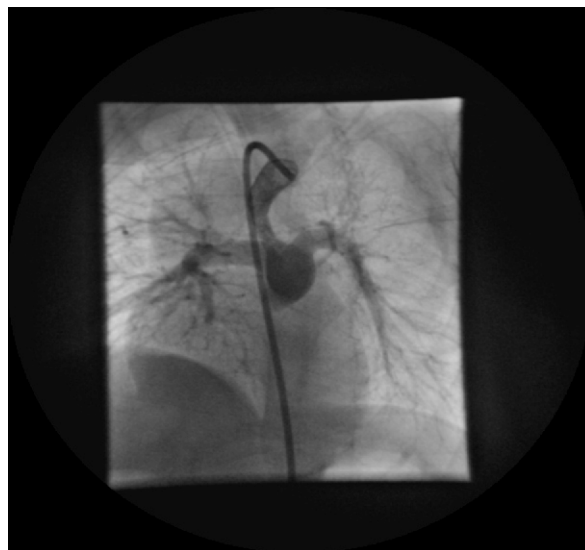


**Figure 4: Angiogram Showing PDA Stenting**



pulmonary arteries are too small for Blalock Taussig shunt. Successful PDA stenting depends on the morphology of the duct. Ductal stenting is an alternative to conventional shunt surgery in duct dependent congenital heart disease.<sup>2</sup> With new generation coronary stents which have excellent profile, flexibility and trackability, ductal stenting may be achieved safely and with considerably less difficulty than previously described.<sup>3</sup> With advances in interventional techniques and cardiac surgery, the management of PAIVS continues to evolve. Ductal stenting is an alternative to conventional shunt surgery in duct dependent pulmonary circulation.<sup>2-4</sup> Advances in surgical technique and postoperative care in the past two decades have led to improved survival and long term outcome in patients with complex congenital heart disease.<sup>4,5</sup> Nowadays corrective cardiac surgery is increasingly performed at an earlier age so obviating the need for palliative procedures in lesion like tetralogy of Fallot (TOF). However, palliative Blalock Taussig (BT) shunt is commonly performed today in relatively complex duct dependant lesions. Some conditions like Hearts with single ventricle physiology and pulmonary atresia also require staged palliation or can only be corrected at a later age.<sup>3</sup> Thus, in the modern era, BT shunt as a palliation is an operation that is almost exclusively performed in the neonatal period or early infancy. Ductus arteriosus stenting, is an attractive as a less invasive alternative for first stage palliation.<sup>2,5</sup> The focus of this case report is the practical aspect of ductal stenting in the catheterization laboratory of PDA from subclavian artery. This type of ductal stenting can be approached by the retrograde trans-arterial method as approached in our case. In this technique the tip of a 4F long sheath is placed near the origin of the LSCA, then a 4F JR catheter is used to engage the proximal part of the LSCA, and the ductus is crossed

**Figure 5: Post PDA Stenting Angiogram**



with a an extra support choice PT wire to anchor deeply in a branch pulmonary artery.<sup>4,5</sup> As described above, and angiograms are taken for measurements of stent length to be use after guidewire positioning. It is best to place a short stent in the distal part of the ductus to cover just enough of the ductal constriction and a second stent for the proximal part.<sup>4,5</sup> Acute stent thromboses is very serious and life threatening complication.<sup>5</sup> In our case which was the least common of PDA location but successful stenting of the PDA improves the saturation of patient.

## CONCLUSION

Ductal stenting is a less invasive alternative to BT shunt as first stage palliation in neonates with duct dependant cyanotic Congenital Heart Disease. Case selection and choice of technique is crucial to the success of the procedure and outcome. This is largely based on the morphological characteristics of the ductus, its shape, size, length, origin from the aorta, tortuosity and its insertion onto the pulmonary artery.

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