An eleven years old boy was diagnosed as a case of Tetralogy of Fallot (TOF) with absent left pulmonary artery (LPA) since two years of age. He had history of total corrective surgery in 2007 from India. He developed severe right pulmonary artery origin stenosis, severe tricuspid and pulmonary valve regurgitation after surgery and redo surgery was performed in 2009. During redo surgery an orthotropic conduit was placed between right ventricular (RV) out flow tract and pulmonary artery. He developed significant conduit dysfunction in the form of stenosis and regurgitation in 2011 and planned for Melody® trans-catheter pulmonary valve replacement. Finally in 25th December 2012, first ever case of pulmonary valve replacement in South Asia was performed in catheterization laboratory of CMH Dhaka with technical support from a Saudi cardiac team. Patient was discharged three days after the procedure with a fully functioning Melody® valve without any stenosis or regurgitation.

So PPVI has the potential to become the standard procedure in the treatment of dysfunctional conduit. We report the first case of PPVI in South Asia where a Melody® with 20 mm ensemble was used in 11 years old boy successfully.

**Case report:**

J, an eleven years old boy was diagnosed as Tetralogy of Fallot (TOF) with absent left pulmonary artery at the age of two years. He had his first surgery on 27th July 2007 in Narayana Hridayalaya, Bengalore, India which was a trans-atrial repair.

During his follow up, some residual and new problems were observed in echocardiography, chest x-ray and electrocardiography. Patient was symptomatic also in the form of exertional dyspnea.

So cardiac catheterization was repeated on 3rd September 2008 and severe right pulmonary artery (RPA) origin stenosis, severe tricuspid regurgitation (TR) and severe pulmonary regurgitation (PR) was noticed.

He was referred again to same centre of India for Redo surgery.

This time an orthotropic conduit (23 mm aortic homograft) was placed between RV out flow tract (RVOT) to pulmonary artery (PA) on 22nd January 2009.

He was doing well in first one year of follow up.

Later he again developed easy fatigability. His echocardiography showed severe conduit stenosis (15th December 2011) of PPG 65 mm Hg with calcified area.
of 10 mm, free PR and dilated RA RV, RVOT with RVOT diameter of 18 mm. It is a common complication of conduit placement.

He was planed for PPVI as parents were refusing further surgery. Communication for procurement of valve from Medtronic, USA started.

It took about a year to arrange things to have a launching program in Combined Military Hospital (CMH) Dhaka for the first ever case of PPVI not only in Bangladesh but also in South Asia.

J was taken into cath Lab on 25th December 2012 for the procedure.

Procedure:

Equipment required.
- Short introducer sheath 18F
- Pigtail catheter 7F
- Multi track catheter 7F
- High pressure balloon Neumed 22 mm X 4 cm
- Inflation device
- BIB balloon catheter 22 mm X 4 cm
- Dilator 18F
- CP stent 39 mm and 42 mm
- Melody valve
- Ensemble delivery system (20 mm)

Steps:

Right femoral vein and right femoral artery accessed. Patient was heparinized as per standard protocol. Pressure run was recorded in Aorta, LV, RV and pulmonary artery.

Aortogram was done to assess coronary artery arrangement.

RVOT angiogram was done to assess morphology of RVOT and conduit, and severity of stenosis. Pulmonary angiogram was done to look for severity of regurgitation.

A 14F Mullin sheath was guided over a landerquest wire into PA and dilator was removed. Balloon dilatation of conduit was done with 22 mm high pressure balloon.

A CP stent of 39 mm length was placed inside conduit to cover the calcified area.

Another 42 mm CP stent was again implanted to cover the whole length of conduit and BIB balloon of 20 mm was used for expansion of stent.

The Melody valve was washed in 3 saline baths for 5 minutes each to clear the gluteraldehyde preservative.

The Ensemble of 20 mm was flushed and the balloons were de aired.

The venous access was dilated with dilators upto 22 mm.

The valve was crimped on the balloon catheter of the delivery system carefully to orientate it in the direction of blood flow and finally covered with the outer sheath.

The valve over the ensemble was forwarded over the wire to the site of implantation.

The valve was uncovered at the optimum site. Check angiography was performed through side port to confirm the site of implantation. Dilatation of inner balloon was followed by dilatation of outer balloon to deploy valved stent.

The delivery system was pulled out carefully keeping the guide wire in position.

Repeat hemodynamic assessment was performed and angiography was done at pulmonary artery.

No PR or PS was noticed. Haemostasis was achieved by applying a tie with silk. Patient was shifted to cardiac ICU after extubation.

Echocardiography on next morning showed well functioning Melody valve with negligible residual
Tetralogy of Fallot, Transposition of the great arteries and Truncus arteriosus 6, 7, 8.

The surgical correction of above conditions often includes the creation of an artificial right ventricle (RV) to pulmonary artery (PA) connection by RV to PA conduit. After repair, over time, these conduits are prone to develop valve dysfunction leading to pulmonary regurgitation (PR), stenosis (PS), thrombosis, infections and calcifications 9, 10. Chronic PR after relief of RV outflow tract obstruction can lead to RV dilatation, biventricular dysfunction, heart failure symptoms, arrhythmia and sudden death 11. There is a very close relationship between the degree of PR and RV volumes which was confirmed by many studies 11-15. Tetralogy of Fallot is the most common from of cyanotic congenital heart disease with frequency of 06-08 per 1000 live birth 16. Total surgical repair was available for last 50 years with favorable outcome in most cases. But surgical repair of tetralogy of Fallot (TOF) may results in many anatomic and functional abnormalities. In the majority of patients in follow up period most common problems are pulmonary regurgitation and conduit stenosis. Though RV volume overload due to severe PR tolerated over years, there is now evidence that the compensatory mechanisms of the right ventricular myocardium ultimately fail and that if the load is not eliminated or reduced, the dysfunction might be irreversible 17. As a result early pulmonary valve replacement is recommended in many centers.

Pulmonary valve replacement can be performed electively with little risk and may improve symptoms of right ventricular failure and provide excellent midterm survival in above mentioned group of patients 18.

Our patient was a case of TOF with absent left pulmonary artery who had surgery on two occasions earlier, than pulmonary valve replacement was decided for conduit dysfunction.

Indications for PPVI are 19

- Severe pulmonary regurgitation.
- Residual RVOT obstruction>30 mm Hg.
- Electrocardiographic evidence of QRS prolongation >180 milliseconds.

Candidates for PPVI must also fulfill the anatomic requirements necessary for safe anchoring of the percutaneous valve: So conduit provide such properties
for PPVI. In contrast native or patched out flow after TOF repair tends to be dilated and remain dynamic and therefore do not provide a secure implantation site19.

In our case we have used Melody valve which was approved by Food and Drug Administration (FDA) in USA in January 2010.

Procedure related risks are stent fracture, embolization of valve, acute coronary compression etc. Coronary compression demands urgent surgical intervention.

Experience of PPVI in two centre over 100 patients showed excellent result regarding reduction of RVOT gradient and PR20.

Outcome of pulmonary valve replacement in 170 patients with chronic PR after relief of ROVT obstruction was also found acceptable in another study11.

Early result from the US clinical trial on Melody transcatheter pulmonary valve also recommends the procedure as safe and effective one21.

A multicentre survey was performed by Italian society of pediatric cardiology (SICP) to analyze the data of patients treated by Melody Medtronic valve22. This study result showed that the procedure is safe and successful. Major concerns are related to the occurrence of stent fracture and bacterial endocarditis.

Conclusion:
Percutaneous pulmonary valve implantation (PPVI) is a new treatment option in patient with RVOT conduit regurgitation and stenosis. Early results following PPVI have shown a significant reduction in right ventricular pressure and RVOT gradient. The most common complication of PPVI is stent fracture or PR in the context of endocarditis. We got excellent result in our first case. So PPVI has the potential to become the standard procedure in the treatment of dysfunctional conduit and will be easy to procure if price is considered for the developing countries.

References:
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