

Step by Step Balloon Pulmonary Valvuloplasty

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Questions to be answered....

- How to diagnosis....?
- How to manage.....?
- How to dilate.....?
- How to follow?

Introduction

- Congenital pulmonary valve stenosis is a common congenital heart disease.
- Isolated pulmonary valve stenosis comprises 7-12% of all congenital heart disease, and accounts for 80% to 90% of all lesions that cause RVOT obstruction.
- Approximately 20% of patients with valvular PS have a dysplastic valve, and if part of Noonan syndrome, these patients have an autosomal dominant trait with variable penetrance that has been mapped to chromosome 12
- While surgical pulmonary valvotomy has been available as a treatment since 1956, it requires a median sternotomy, use of cardiopulmonary bypass, and post-surgical ICU admission with multi-day hospitalization.
- Recognizing the potential advantages of a less invasive approach, the first report of the static balloon dilation of the pulmonary valve was by Kan et al. in 1982.

Causes of pulmonary valve disease

- Congenital heart diseases mostly associated with dysfunction of pulmonary valve either as a primary component or post repair.
- Acquired pulmonary valve disease in the adult population is unusual and mostly relates to uncommon conditions such as carcinoid disease.
- Even though rheumatic heart disease is common, pulmonary valve is the least affected one(rare).
- Infective endocarditis is on upraise as there are increased cases of IV drug abuse and CKD patients on dialysis.
- Surgical revision of RVOT is commonly performed in this population.

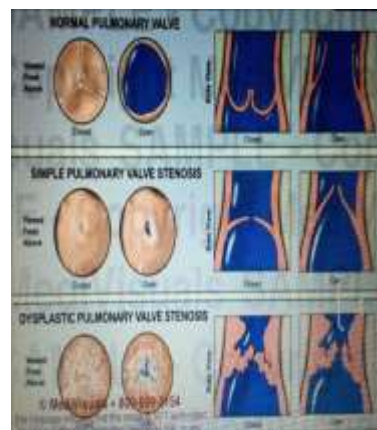
Patho-physiology

- Whether caused by commissural fusion or by a dysplastic valve, PVS can result in a significant increase in RV after-load and RV hypertension.
- This in turn can cause RV hypertrophy, including hypertrophy of the infundibular outflow tract, resulting in dynamic RV outflow obstruction.
- Right ventricular hypertension can exert stresses on the tricuspid valve leading to thickening of the leaflets and its chordal attachments with potential tricuspid regurgitation.
- If left untreated, persistent RV hypertension can lead to RV dilation with both systolic and diastolic dysfunction and right-heart failure.

Pulmonary valve morphology

The classic form of pulmonary valve stenosis characterized by a narrow central opening but a preserved, mobile valve mechanism. It is described as a “dome” shaped valve with raphes present, but the leaflets do not completely separate, sometimes fused

Dysplastic pulmonary valves are tri-leaflet but markedly thickened cusps with disorganized myxomatous tissue without significant fusion and frequently associated with hypoplastic valve annulus



Pre-procedural evaluation>>>How to diagnosis....?

* Clinical Examination

* ECG

* 2D ECHO:

- Grading of the pulmonary valve stenosis by echocardiography.
- Peak gradient in the RVOT – subvalvular, valvular, supra-valvular.
- Calculation of the pulmonary annulus.
- Morphology of the valve.
- Distal pulmonary arteries size.

* Cardiac catheterization

- Usually not required for diagnosis.
- Usually done before the procedure of balloon dilation.
- Pullback gradient from distal pulmonary artery to MPA, Pulmonary valve, sub-valvular ,RV.
- RV angiography in AP cranial and lateral.

Indications

- Severe or Moderate symptomatic Pulmonary Valve Stenosis, bi- or tricuspid valve.
- As a palliative procedure before surgical correction in TOF ???!!!.
Now contra-indicated
- No evidence for mild PS will benefit from PBPV.
- Can be done in neonates, infants, adolescents, adults. Age is not a predictor of response.
- **Not suitable**
 - Dysplastic valves and pulmonary hypoplasia.
 - Primary fibromuscular subvalvular narrowing
 - Primary fibromuscular supra-valvular narrowing.
 - DCRV

The AHA 2008 has refined the recommendations for when Balloon Valvuloplasty

severity	RV/LV pressure	Gradient across PV (Peak)
Mild	$< \frac{1}{2}$	<35 to 40 mmHg
Moderate	$\frac{1}{2} < RVP < \frac{3}{4}$	>40 mmHg
severe	$\geq \frac{3}{4}$	>60 to 70 mmHg

The AHA 2010 has refined the recommendations for when Balloon Valvuloplasty>>>How to manage.....?

- *Class I*

1. Clinically significant pulmonary valve obstruction in the presence of RV dysfunction were considered definitive indications for balloon pulmonary valvuloplasty. (level of evidence A)
2. Balloon valvotomy is recommended for asymptomatic patients with a domed pulmonary valve and a peak instantaneous Doppler gradient greater than 60 mm Hg or a mean Doppler gradient greater than 40 mm Hg (in association with less than moderate pulmonic valve regurgitation). (*Level of Evidence: B*)
3. Balloon valvotomy is recommended for symptomatic patients with a domed pulmonary valve and a peak instantaneous Doppler gradient greater than 50 mm Hg or a mean Doppler gradient greater than 30 mm Hg (in association with less than moderate pulmonic regurgitation). (*Level of Evidence: C*)

- *Class IIb*

1. Balloon valvotomy may be reasonable in asymptomatic patients with a dysplastic pulmonary valve and a peak instantaneous gradient by Doppler greater than 60 mm Hg or a mean Doppler gradient greater than 40 mm Hg. (Level of Evidence: C)
2. Balloon valvotomy may be reasonable in selected symptomatic patients with a dysplastic pulmonary valve and peak instantaneous gradient by Doppler greater than 50 mm Hg or a mean Doppler gradient greater than 30 mm Hg. (Level of Evidence: C)

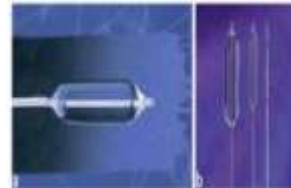
- *Class III*

1. Balloon valvotomy is not recommended for asymptomatic patients with a peak instantaneous gradient by Doppler less than 50 mm Hg in the presence of normal cardiac output. (Level of Evidence: C)
2. Balloon valvotomy is not recommended for symptomatic patients with PS and severe pulmonary regurgitation. (Level of Evidence: C)
3. Balloon valvotomy is not recommended for symptomatic patients with a peak instantaneous gradient by Doppler less than 30 mm Hg. (Level of Evidence: C)

AHA 2010.....Indication for intervention in RVOT obstruction

Indications	Class ^a	Level ^b
RVOTO at any level should be repaired regardless of symptoms when Doppler peak gradient is ≥ 64 mmHg (peak velocity ≥ 4 m/s), provided that RV function is normal and no valve substitute is required	I	C
In valvular PS, balloon valvotomy should be the intervention of choice	I	C
In asymptomatic patients in whom balloon valvotomy is ineffective and surgical valve replacement is the only option, surgery should be performed in the presence of a systolic RVP ≥ 80 mmHg (TR velocity ≥ 4.3 m/s)	I	C
Intervention in patients with gradient ≤ 64 mmHg should be considered in the presence of: <ul style="list-style-type: none"> • symptoms related to PS or, • decreased RV function or, • double-chambered RV (which is usually progressive) or, • important arrhythmias or, • right-to-left shunting via an ASD or VSD. 	IIa	C
Peripheral PS, regardless of symptoms, should be considered for repair if $\geq 50\%$ diameter narrowing and RV systolic pressure ≥ 50 mmHg and/or lung perfusion abnormalities are present.	IIa	C

Selection of Balloon Size



Effective balloon size:

- The target balloon diameter should be 120-140% of the pulmonary valve annulus diameter.
- Annulus ratios of less than 120 may result in poor long-term outcome and restenosis.
- Balloon size with $>150\%$ the diameter of the normal pulmonary valve annulus will result in rupture & damage of RVOT.
- Double balloons if used – total circumference of the two balloons should be equal to 120-140% of the circumference of the valve annulus.

Ring JC et al, Am J Cardiol 1985;55(1):210-4.

Balloon length:

- There is limited data regarding the length of balloons used for pulmonary valvuloplasty.
- In general, shorter balloons make it difficult to maintain position across the pulmonary valve annulus during inflation.
- On the other hand, longer balloons carry a risk of injury to more remote structures.
- This may include disruption of tricuspid valve architecture with resulting tricuspid insufficiency, trauma to conduction system with risk of heart block, or injury of the pulmonary artery.
- It has been recommended to use 20 mm length for neonates and infants, 30 mm for children and 40 mm for adolescents and adults.

Patient preparation>>>How to dilate....?

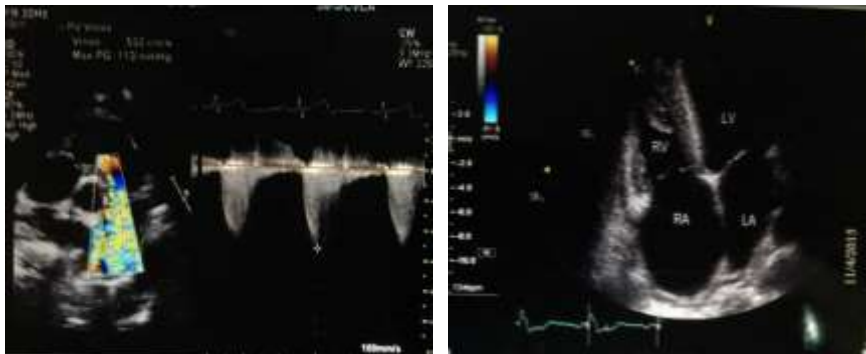
- Fasting from the night before procedure
- Local anaesthesia is usually enough.
- General anaesthesia for children.
- Venous access – unilateral or bilateral (8F)
 - Jugular venous access
 - Hepatic – Axillary vein
- Catheter is used.
- Wires: Stiff – Soft tip
- Inj. Heparin (50-100 IU/ kgm) at the start of procedure.
- One dose of broad spectrum antibiotic at the start of procedure.

Relationship Between Peak Instantaneous Doppler Echocardiographic Pressure Gradients and Peak-to-Peak Cardiac Catheterization Gradients

- There are recent data that suggest the peak-to-peak gradient by cardiac catheterization correlates best with the **mean** Doppler (and not peak instantaneous Doppler) gradient in this situation and that the peak instantaneous gradient systematically overestimates the peak-to-peak cardiac catheterization gradient by slightly more than 20 mm Hg.
- Correlation of the echocardiography-Doppler gradient with other clinical findings is important.

Echocardiography

Female girl, 5Y old, C/O dyspnea, palpitation with effort, auscultation loud systolic murmur. Echo: Severe valvular PS with peak RVOT PG: 113mmHg, post stenotic dilation of MPA, no PR, RVH.



Outcomes

Procedural success is high in children and adults with congenital valvular pulmonary stenosis.

Residual gradient of <35 mmHg in most of the patients.

Complications occur very rare in the form of:

- Femoral vein injury, or thrombosis.
- Pulmonary tear, TV PM rupture—children, cardiac perforation, tamponade.
- Arrhythmias (RBBB). High Grade AV nodal block. Complete HB
- Transient RVOT obstruction which referred to as a “suicidal right ventricle”
- Significant blood loss
- Pulmonary incompetence.
- Seizures, cerebro-vascular accidents
- Recurrence of stenosis with need for intervention
- Pulmonary insufficiency, pulmonary edema.
- Cardiac arrest

How to follow?

- There is little progression in PS severity when the gradient is less than 30 mm Hg; such patients can be followed up at least every 5 years with a clinical examination and Doppler echocardiogram.
- Those with more significant stenosis should be followed up on a yearly basis.
- If a dynamic outflow tract obstruction exists, therapy with drugs that slow the heart rate and improve diastolic filling time (beta blockers), and those that might potentially reduce the systolic gradient and improve lusitropy (ie, calcium channel blockers and disopyramide) may also be used clinically.
- Elevated right-sided heart pressures, edema, and ascites can be treated with thiazides, loop diuretics, and aldosterone antagonists as appropriate.

Conclusion

- Percutaneous pulmonary valvuloplasty is the treatment of choice for pulmonary valve stenosis in neonates, children and adults.
- As the technique was refined and catheter and balloon technology have advanced, the results of balloon pulmonary valvuloplasty have improved and the approach has become the standard of care for treating pulmonary valve stenosis.

- Complications are rare and can be avoided by using the proper balloon diameter and length, short inflation time and with meticulous attention to the technique
- The key for successful procedure is adequate assessment of the valve size, obtaining stable wire position, and choosing an appropriate balloon diameter.
- Good results are manifested as immediate reduction in gradient, reduction in RV: systemic pressure ratio, increase in the width of the jet and better motion of the pulmonary valve leaflets.

**Thanks For Your
Attention**