

Pulmonary Stenosis & RVOT Obstruction

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Right Sided Obstructive Lesions

- Tetralogy of Fallot
- Tetralogy of Fallot with Pulmonary Atresia and Major Aorto-Pulmonary Collaterals
- Pulmonary Atresia Intact Ventricular Septum
- Valvar and Supravalvar Pulmonary Stenosis
- Infundibular Stenosis, Double Chambered Right Ventricle



Valvar Pulmonary Stenosis

- Incidence: 8-10% of all cases of congenital heart disease
 - One of the most common lesions
- Manifests at any age
 - Neonatal critical PS is a small fraction of these patients.



Morphology

- Six Subgroups:
 - Domed
 - Tricuspid
 - Bicuspid
 - Unicommissural
 - Dysplastic (syndromic association)
 - Hypoplastic annulus
- Thickened leaflets with commissural fusion
- Frequently post-stenotic dilatation present.



Pathophysiology

- Infundibular hypertrophy from the increased pressure load results in a fixed and dynamic obstruction to right ventricular outflow.
- Higher RV pressures in the setting of a poorly compliant right ventricle leads to tricuspid regurgitation, high CVP, increased right to left shunt through PFO.



Neonatal Critical Pulmonary Stenosis

- Usually trileaflet valve with commissural fusion.
- Increased right to left shunt during fetal life leads to hypoplastic tricuspid valve and RV in half of pts.
 - Severe hypoplasia is uncommon
- RV hypertrophy is common.
- Coronary fistulas and RV coronary dependency are uncommon.



Pathophysiology

- Pulmonary valve obstruction and RV hypertrophy result in a non-compliant RV and blood shunts R-> L at the atrial level.
- Tricuspid regurgitation is often present.
- Pulmonary blood flow may be dependent on the ductus arteriosus.



Presentation & Diagnosis

- With ductal closure, cyanosis worsens and the child can become critically ill
- Ductal patency can provide clinical stability, but patients often become tachypneic from the pulmonary overcirculation
- CXR shows cardiomegaly and an enlarged right atrium.
- Echocardiogram is the diagnostic study of choice.



Initial Management

- IV access
- Resuscitation of metabolic acidosis
- PGE1
- Mechanical ventilation
- Inotropic support



Neonatal PS: Intervention

- Balloon angioplasty has now supplanted surgical valvotomy as the 1st line procedure of choice.
- Child can then be weaned from PGE1
- Mild to moderate cyanosis may persist for some time as the RV compliance improves.



Valvar PS in older infants and children

- Often asymptomatic. Diagnosis picked up on echo performed for a murmur.
- Symptoms may be elicited on exercise.
- Can develop over time as RV hypertrophy worsens.



Grading Scale

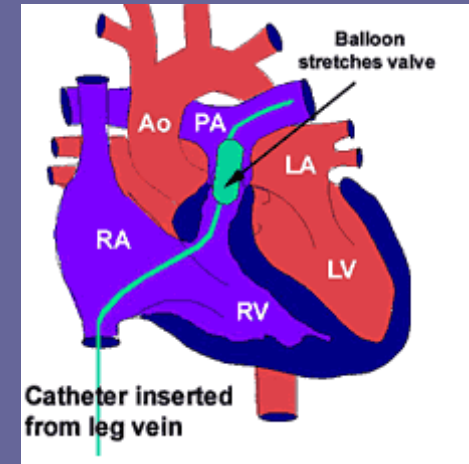
- Normal:
 - PV Gradient $< 10\text{mmHg}$
 - Resting RV pressure $\leq 30\text{mmHg}$
- Mild
 - PV Gradient $< 36\text{mmHg}$, peak velocity $< 3\text{m/s}$
 - RV to LV pressure ratio ≤ 0.5
- Moderate
 - PV Gradient > 36 but < 64 mmHg, peak velocity 3-4m/s
 - RV to LV Pressure ratio between 0.5 and 1
- Severe
 - PV Gradient $> 64\text{mmHg}$, peak velocity $>4\text{m/s}$
 - RV to LV pressure ratio > 1

EAE/ASE Echocardiographic assessment of valve stenosis



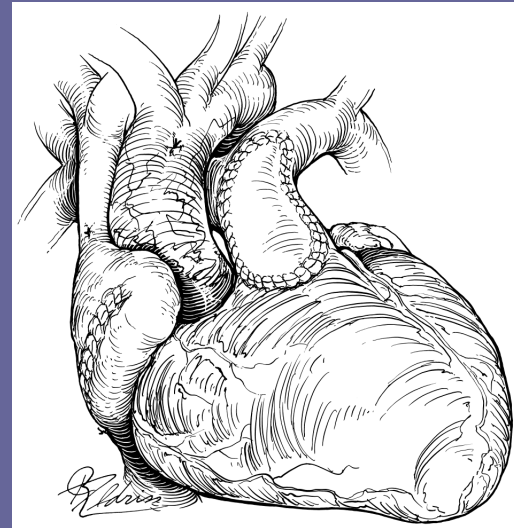
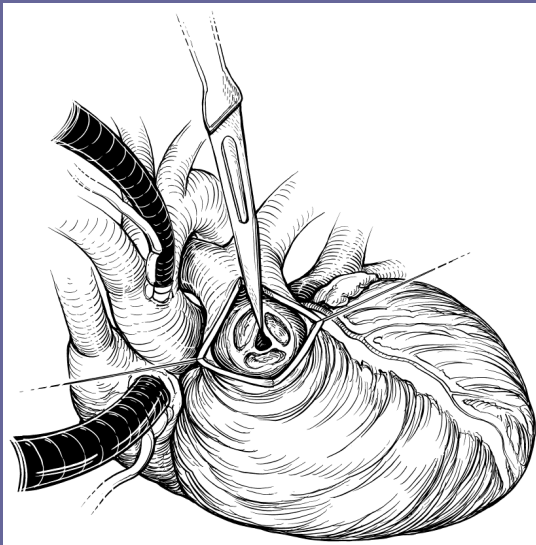
Management

- Balloon valvuloplasty is 1st line treatment
- Indications:
 - Symptoms
 - Mean PV gradient $> 50\text{mmHg}$
 - RV/LV pressure ratio > 0.66
 - Progressive RV hypertrophy
- Difficulties:
 - Small annulus size
 - Dysplastic valve type
 - Supravalvar PS
- Results:
 - Low mortality, $< 2\%$
 - Freedom from re-intervention: 75%

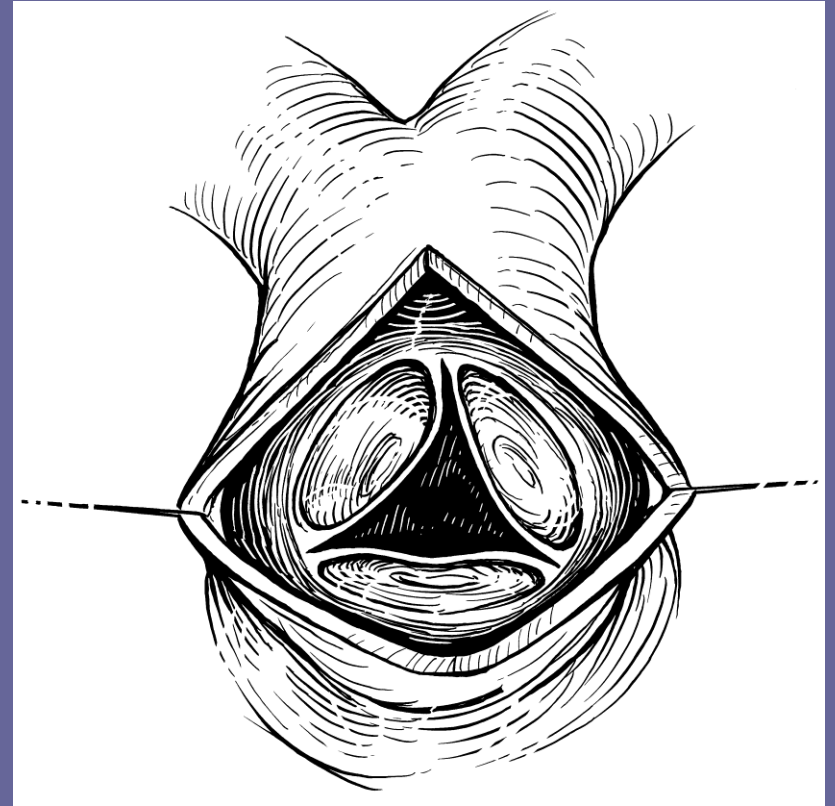
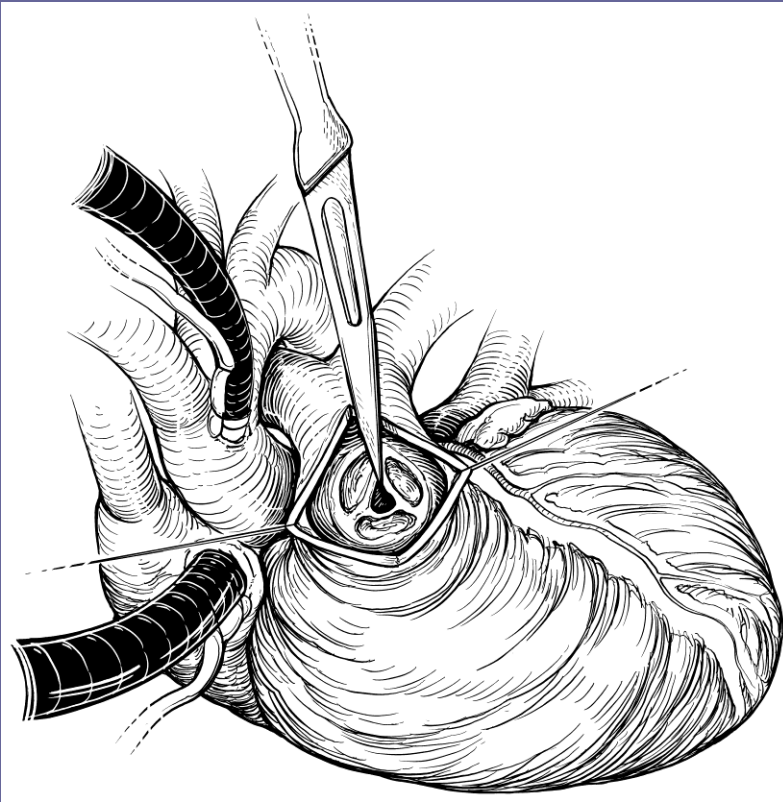


Management - Surgical

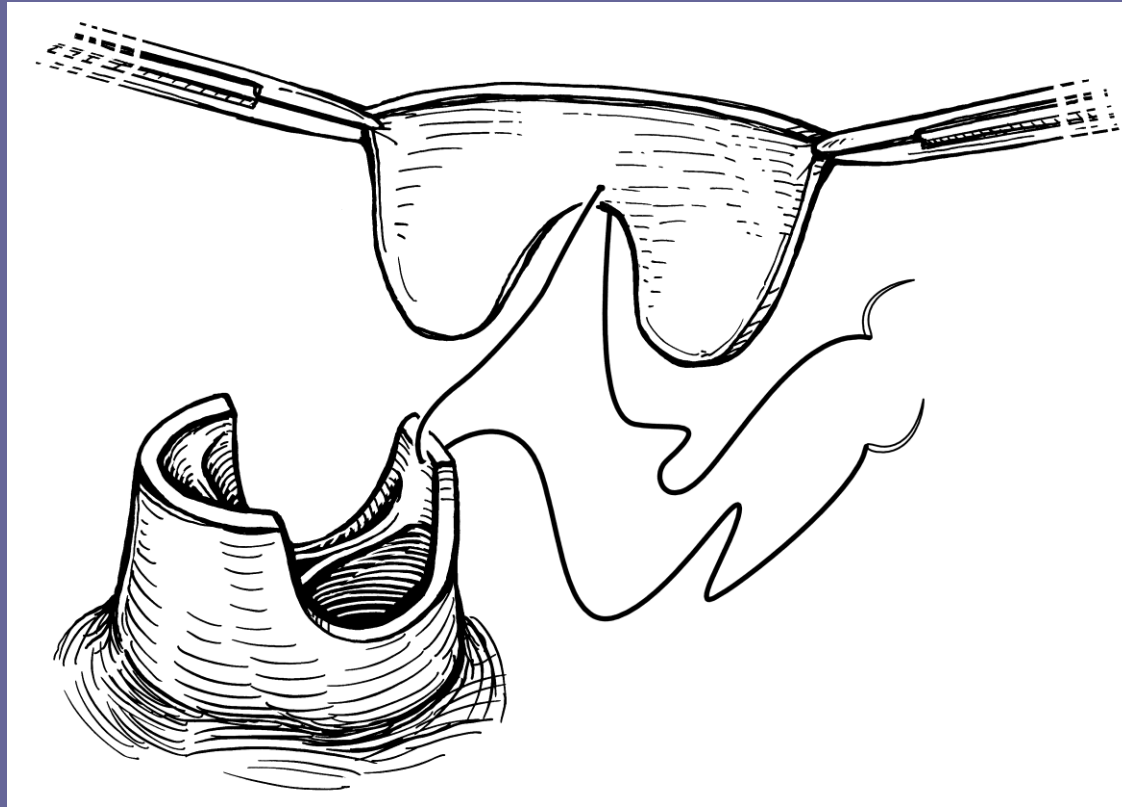
- Indications:
 - Failure of primary balloon valvotomy
 - Dysplastic valve anatomy, Hypoplastic annulus
 - Supravalvar stenosis



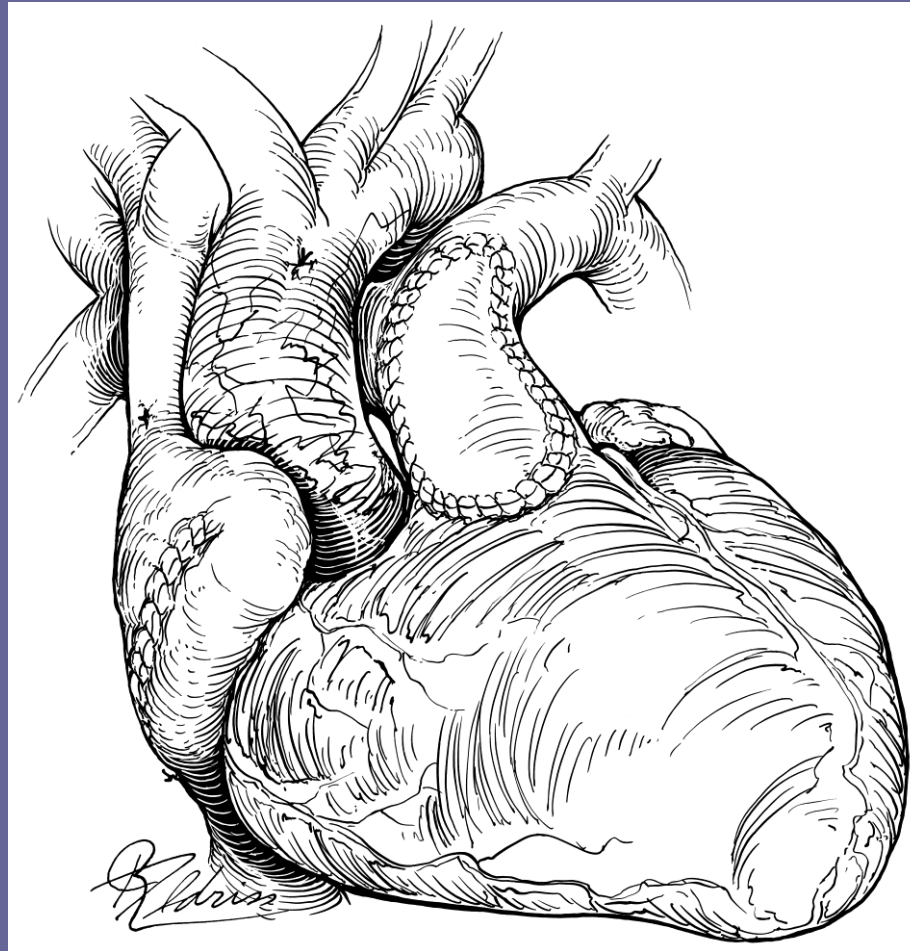
Valvotomy



Pantaloon Patch



Transannular Patch



Supravalvar Pulmonary Stenosis

- Associated with Noonan's syndrome
- Indications for treatment are the same as valvar PS
- Surgical patch arterioplasty is the primary treatment
 - Patch may extend out past pulmonary artery bifurcation
 - Peripheral artery stenosis is common and may require ballooning and stenting.



Infundibular Stenosis

- Isolated infundibular stenosis accounts for 5% of all cases of RVOT obstruction
- Distal infundibular stenosis:
 - Trans-atrial / Trans Pulmonary approach is attempted first with resection of obstructing muscle trabeculations.
 - Care is taken to avoid tricuspid papillary muscle injury.
 - If exposure is poor or RVOT is hypoplastic, a longitudinal ventriculotomy and patch is performed to widen the outflow tract
 - Pulmonary valve is usually normal and can be preserved.
- Double-Chambered Right Ventricle:
 - Proximal stenosis at the junction between main RV and outlet chamber is approached via transatrial route with muscle resection.
 - Usually associated with a VSD.



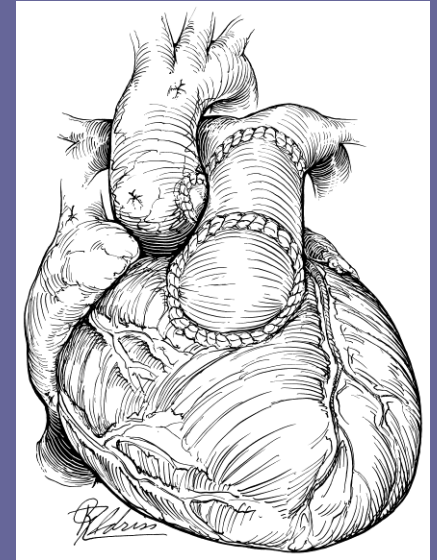
Operative Assessment

- TEE on all cases
- Pressure measuring line to assess RV and LV pressures.
 - RV/LV ratio greater than 0.66 will require further treatment (infundibular or trans-annular patch, conduit).
 - Esmolol can be given to test for dynamic residual gradient from hypertrophied RV
 - If ratio drops, can expect gradient to improve as RV remodels after relief of outflow tract obstruction



Valved Conduit RVOT Reconstruction

- If trans-annular incision required, it is occasionally preferable to have a competent pulmonary valve:
 - Pulmonary hypertension
 - Small RV
 - Poor RV function
 - Tricuspid regurgitation
 - Peripheral pulmonary stenosis
- Pulmonary or Aortic homografts are useful for infants and small children
- Porcine valved conduits or Contegra bovine jugular valved conduits can be used in larger children and adult primary and reoperative surgery



Results

- Age at implant and size of conduit are primary factors in freedom from reoperation.
- 5 Year Freedom from reoperation:
 - Homograft: 92%
 - Carpentier-Edwards Conduit: 87%
 - Contegra: 78%

