MANAGEMENT OF VALVULAR PULMONARY STENOSIS (PS)

Prepared by **Dr. Sidhartha Mullapudi**, DrNB (Cardiology), MD (General Medicine)
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I INTRODUCTION, MORPHOLOGY AND HEMODYNAMIC GRADING (ESC 2020 ACHD)

- **Core Principle:** Valvular Pulmonary Stenosis (PS) accounts for 80 to 90 percent of all right ventricular outflow tract (RVOT) obstruction and is almost always congenital. The primary therapeutic goal is to relieve **Right Ventricular (RV) pressure overload** before chronic hypertrophy leads to subendocardial ischemia, myocardial fibrosis, RV systolic/diastolic failure, or tricuspid regurgitation.
- **Two Distinct Anatomical Morphologies (Crucial for Treatment Selection):**
  - **Classical Dome-Shaped Valvular PS (85 to 90 percent):** Thin, pliable trileaflet or bicuspid valve with **commissural fusion**, a mobile systolic "doming" configuration, a normal pulmonary annulus, and prominent **post-stenotic dilatation of the main and left pulmonary arteries** (due to the high-velocity jet preferentially directed into the left PA). Responds excellently to **Balloon Pulmonary Valvuloplasty (BPV)**.
  - **Dysplastic Pulmonary Valve (10 to 15 percent — Classic in Noonan Syndrome):** Markedly thickened, myxomatous, immobile cartilaginous-like cusps **without commissural fusion**, frequently accompanied by a **hypoplastic pulmonary annulus** and minimal post-stenotic PA dilatation. Responds poorly to balloon dilation and typically requires **surgical valvotomy/annular patch**.
  - **Doppler Echocardiographic Severity Grading (ESC 2020 ACHD / ACC-AHA 2018 Guidelines):** Graded by Continuous-Wave (CW) Doppler **Peak Jet Velocity (Vmax)** and **Peak Instantaneous Gradient (4v2)** across the pulmonary valve (assuming normal cardiac output):

Severity Grade	Peak Jet Velocity (Vmax)	Peak Instantaneous Doppler Gradient	Mean Doppler Gradient
Mild PS	Less than 3.0 m/s	**Less than 36 mmHg**	Less than 20 mmHg
Moderate PS	3.0 to 4.0 m/s	**36 to 64 mmHg**	20 to 40 mmHg
Severe PS	**Greater than 4.0 m/s**	**Greater than 64 mmHg**	**Greater than 40 mmHg**

II CLINICAL INDICATIONS FOR INTERVENTION (ESC 2020 ACHD / ACC-AHA 2018 GUIDELINES)

- **Class I Indications (Recommended):**
  - **Severe PS (Peak Doppler Gradient greater than 64 mmHg / Vmax greater than 4.0 m/s):** Intervention (**Balloon Pulmonary Valvuloplasty [BPV]**, provided the valve is not anatomically unsuitable/dysplastic) is **recommended in all patients with severe PS regardless of symptoms**.
  - **Symptomatic Moderate PS (Peak Gradient 36 to 64 mmHg):** Intervention is recommended in symptomatic patients (exertional dyspnea, fatigue, angina, syncope, or cyanosis from right-to-left atrial shunting across a PFO/ASD) with moderate PS when alternative causes of symptoms have been excluded.
  - **Critical Neonatal Valvular PS:** Emergency **Class I** indication for BPV in cyanotic neonates with duct-dependent pulmonary blood flow and right-to-left atrial shunting.
  - **Class IIa / IIb Indications (Should or May Be Considered):**
  - **Asymptomatic Moderate PS (Peak Gradient 36 to 64 mmHg):** Intervention may be considered in the presence of **progressive RV systolic dysfunction, severe RV hypertrophy, progressive Tricuspid Regurgitation**, right-to-left atrial shunting, arrhythmias, or **prior to planned pregnancy** (when peak gradient exceeds 50 mmHg).
  - **Class III (Not Indicated):**
  - **Mild PS (Peak Gradient less than 36 mmHg):** Benign natural history without progression in adulthood; requires **no intervention** and no restriction from competitive sports or pregnancy.

III PERCUTANEOUS BALLOON PULMONARY VALVULOPLASTY (BPV)

- **First-Line Gold Standard (Class I, Level B):** Since first described by **Kan et al. in 1982**, percutaneous **Balloon Pulmonary Valvuloplasty (BPV)** is the treatment of choice for isolated dome-shaped valvular PS across all age groups (neonates, children, and adults).
- **Procedural Technique and Balloon Sizing:**
  - **Vascular Access and Hemodynamics:** Femoral venous access; right heart catheterization measures baseline RV systolic pressure, PA pressure, and peak-to-peak catheter gradient, while a **lateral RV angiogram** delineates the hinge points of the pulmonary valve leaflets to precisely measure the **pulmonary valve annulus diameter**.
  - **Balloon-to-Annulus Ratio (Updated Contemporary Standard):** Historically sized at 1.2 to 1.4 times the annulus; contemporary consensus recommends a **Balloon-to-Annulus Ratio of 1.2 to 1.25 (not exceeding 1.3)** (or a double-balloon / Inoue balloon technique in adults with large annuli greater than 22 to 25 mm), which achieves identical commissural splitting while **significantly reducing the late incidence of severe Pulmonary Regurgitation**.
  - **Procedural Endpoint:** Rapid inflation with disappearance of the central "waist" on fluoroscopy, resulting in a **post-procedure peak-to-peak catheter gradient less than 30 mmHg** (or greater than 50 percent reduction).

IV SURGICAL TREATMENT (VALVOTOMY / RVOT RECONSTRUCTION)

- **Class I Indications for Surgery Over BPV (ESC 2020):** Surgery is recommended in severe PS when BPV has failed or is anatomically contraindicated due to:
- **Dysplastic Pulmonary Valve** (thick, immobile myxomatous leaflets without commissural fusion, classic in **Noonan Syndrome**).
- **Severe Pulmonary Annular Hypoplasia** or **Supravalvular Main PA Stenosis**.
- **Fixed Fibromuscular Subvalvular (Infundibular) Stenosis** or **Double-Chambered Right Ventricle (DCRV)**.
- **Concomitant Cardiac Lesions Requiring Surgery:** Severe **Pulmonary Regurgitation**, **severe Tricuspid Regurgitation** requiring annuloplasty, or large ASD unsuitable for device closure.

V COMPLICATIONS AND ACUTE POST-BPV "SUICIDE RIGHT VENTRICLE"

- **Dynamic Infundibular Obstruction ("Suicide Right Ventricle"):**
  - **Mechanism:** In patients with longstanding severe valvular PS, the RV infundibulum undergoes massive concentric muscular hypertrophy. Immediately after BPV relieves the fixed valvular afterload, the hypercontractile, hypertrophied infundibular walls collapse inward during systole, causing **acute severe dynamic subvalvular (infundibular) obstruction**, a secondary pressure gradient within the RV body, cavity obliteration, and **acute low cardiac output / systemic hypotension**.
  - **Definitive Management:** Treated immediately with **aggressive intravenous isotonic volume expansion (preload)** and **Intravenous Beta-Blockers (Esmolol or Propranolol)** (followed by oral Propranolol for 3 to 6 months until RV hypertrophy regresses). **Positive inotropes (Dobutamine, Dopamine, Adrenaline, Milrinone), vasodilators, and diuretics are strictly contraindicated**, as they exacerbate dynamic infundibular cavity obliteration.
  - **Late Pulmonary Regurgitation (PR):** Mild-to-moderate PR is common after BPV; however, 10 to 30 percent of patients (especially those treated historically with oversized balloons >1.3x annulus) develop **severe PR and progressive RV dilation** decades later, requiring **Transcatheter (TPVR) or Surgical Pulmonary Valve Replacement** using the same volumetric CMR criteria as repaired Tetralogy of Fallot.

VI PRACTICAL CONSIDERATIONS IN THE INDIAN CLINICAL CONTEXT

- **Syndromic Screening (Noonan vs. Williams vs. Congenital Rubella):** In Indian clinics, identifying syndromic features is a high-yield clinical skill: **Noonan Syndrome** (PTPN11 mutation; hypertelorism, ptosis, webbed neck, short stature, chest deformity, and **hypertrophic cardiomyopathy**) is classically associated with a **dysplastic pulmonary valve** (poor BPV response) and **left-axis deviation** on ECG; **Williams Syndrome** (7q11.23 deletion) is associated with **peripheral/supravalvular branch PA stenosis** and supravalvular AS; and **Congenital Rubella Syndrome** is associated with **branch PA stenosis + PDA**.
- **Valvular PS in Pregnancy:** Mild and moderate valvular PS are well tolerated during pregnancy (**mWHO Class I–II**). However, in women presenting in India with **severe PS (peak gradient greater than 64 mmHg)** prior to conception, **elective pre-pregnancy BPV** is recommended. If a pregnant woman develops refractory NYHA Class III/IV right heart failure from severe dome-shaped PS during pregnancy, **percutaneous BPV can be safely performed in the second trimester** (after organogenesis is complete) with pelvic lead shielding and Intracardiac/Transthoracic Echo guidance.

KEY REFERENCES

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