ATRIAL SEPTAL DEFECT (ASD) WITH PAH: NATURAL HISTORY & MANAGEMENT

Prepared by **Dr. Sidhartha Mullapudi**, DrNB (Cardiology), MD (General Medicine)
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I NATURAL HISTORY OF OSTIUM SECUNDUM (OS) ASD

- **Core Principle:** The right ventricle (RV) in an ASD patient relies on the interatrial communication when pulmonary vascular resistance (PVR) rises. If severe irreversible Pulmonary Arterial Hypertension (PAH) develops and PVR becomes fixed, closing the defect traps suprasystemic pressure in the right heart, precipitating acute RV failure and low cardiac output. Clinical decision-making hinges on invasive hemodynamic assessment via Right Heart Catheterization (RHC).
- **Childhood and Adolescence:** Ostium secundum ASD is the most common congenital heart defect presenting in adulthood; patients are typically asymptomatic during the first two decades of life because the compliant, thin-walled RV readily accommodates the left-to-right (L-to-R) volume shunt.
- **Third to Fourth Decade (The Hemodynamic Turning Point):** As left ventricular diastolic compliance decreases with aging, systemic hypertension, or coronary artery disease, left atrial pressure rises relative to right atrial pressure, augmenting the L-to-R shunt across the ASD.
- **Clinical Syndrome:**
  - **Right Heart Volume Overload:** Progressive right atrial (RA) and RV dilatation lead to exertional dyspnea, fatigue, and functional tricuspid regurgitation.
  - **Atrial Tachyarrhythmias:** Chronic RA stretch and interstitial fibrosis trigger Atrial Flutter and Atrial Fibrillation in 15 to 20 percent of adults over 40 years of age (rising to over 40 to 50 percent past age 60).
- **Late Stage (PAH and Eisenmenger Syndrome):** Chronic high pulmonary blood flow induces endothelial shear stress and medial/intimal remodeling of the pulmonary arterioles. Moderate-to-severe PAH develops in approximately 5 to 10 percent of unoperated ASDs (less frequent and occurring later in life than in post-tricuspid shunts like VSD or PDA). When PVR approaches or exceeds systemic vascular resistance (SVR), the shunt reverses to right-to-left (R-to-L), producing central cyanosis and clubbing (Eisenmenger Syndrome).

II ASSESSMENT FOR DEVICE CLOSURE (ANATOMICAL AND HEMODYNAMIC WORKUP)

A Anatomical Assessment (TEE or ICE)

- **Rim Requirements:** Transcatheter closure of a secundum ASD requires sufficient surrounding septal tissue rims of at least 5 mm (superior/SVC, inferior/IVC, atrioventricular/mitral valve, right upper pulmonary vein, and posterior/coronary sinus rims) to anchor the occluder and avoid impinging on adjacent structures.
- **The Aortic (Anterosuperior) Rim:** Frequently deficient (less than 5 mm in up to 40 to 50 percent of cases), which is not an absolute contraindication to device closure if other rims are adequate and device oversizing is avoided.
- **The Inferior (IVC) Rim:** Deficiency of the inferoposterior (IVC) rim (less than 5 mm) is a major contraindication to percutaneous device closure because the device cannot anchor inferiorly, carrying a high risk of device embolization or inferior vena cava obstruction.
- **The AV Valve Rim:** Must be at least 5 mm to prevent device encroachment on the anterior mitral leaflet or septal tricuspid leaflet.

B Hemodynamic Assessment (Right Heart Catheterization)

- **Indication for RHC:** Invasive RHC is mandatory in any ASD patient with non-invasive echocardiographic signs of elevated pulmonary artery pressure (tricuspid regurgitation velocity greater than 2.8 m/s or estimated PASP greater than 40 mmHg).
- **ESC 2020 / ACC/AHA 2018 Hemodynamic Thresholds:**
  - **PVR less than 3 Wood Units (WU):** Safe for standard percutaneous or surgical closure when RV volume overload and Qp:Qs of 1.5 or greater are present (Class I, Level B).
  - **PVR 3 to less than 5 WU (The Borderline Zone):** Closure should be considered (Class IIa, Level C) when a significant net L-to-R shunt (Qp:Qs greater than 1.5) is present without exercise-induced desaturation.
  - **PVR 5 WU or greater:** Immediate complete closure is contraindicated due to advanced pulmonary vascular disease.

III MANAGEMENT OF ADULTS WITH ASD AND PAH (ESC 2020 / ESC-ERS 2022 STANDARDS)

A The "Treat-and-Repair" Strategy

- **Target Patient:** Patients presenting with baseline PVR of 5 WU or greater who still maintain a net left-to-right shunt or reversible pulmonary vascular reactivity.
- **Stepwise Protocol (Class IIb, Level C in ESC 2020):**
  - **Upfront Targeted PAH Therapy:** Initiate PAH-specific pharmacotherapy—typically Endothelin Receptor Antagonists (Bosentan, Macitentan, or Ambrisentan) combined with PDE-5 inhibitors (Sildenafil or Tadalafil).
  - **Repeat Invasive Hemodynamic Evaluation:** Repeat RHC after 3 to 6 months of medical therapy.
  - **Fenestrated Closure Criterion:** If PVR declines to less than 5 WU in the presence of a significant net L-to-R shunt (Qp:Qs greater than 1.5), fenestrated ASD closure may be considered.
- **Role of the Fenestrated Device:** A custom or factory fenestrated ASD occluder (containing a calibrated 4 to 8 mm central communication) preserves a controlled decompression "pop-off valve" from the right atrium to the left atrium, preventing acute right ventricular failure if pulmonary pressures rise post-procedure; targeted PAH medical therapy must be continued indefinitely after fenestrated closure.

B Management of Eisenmenger Syndrome

- **Target Patient:** Patients with fixed PVR of 5 WU or greater that fails to fall below 5 WU on PAH therapy, bidirectional or net right-to-left shunting (Qp:Qs less than 1.5), or resting/exercise arterial desaturation.
- **Strict Contraindication (Class III — Harm):** ASD closure (percutaneous or surgical) is strictly contraindicated, as eliminating the right-to-left decompression shunt precipitates right ventricular failure and reduces systemic cardiac output.
- **Comprehensive Medical Management:**
  - **PAH-Targeted Therapy:** Oral Bosentan is Class I recommended (validated by the BREATHE-5 trial) to improve exercise capacity and hemodynamics in symptomatic Eisenmenger patients, with PDE-5 inhibitors, macitentan, or prostanoids used in combination therapy.
  - **General Precautions:** Avoid pregnancy (maternal mortality historically 30 to 50 percent and still prohibitive), volume depletion/dehydration, systemic vasodilators, and unacclimatized high-altitude exposure.
  - **Hematologic Care:** Routine phlebotomy is contraindicated (Class III) because it induces iron deficiency, generating rigid microcytic erythrocytes that paradoxically worsen blood viscosity and stroke risk. Isovolumic phlebotomy is reserved strictly for moderate-to-severe hyperviscosity symptoms with hematocrit exceeding 65 percent in an iron-replete, hydrated patient.
- **End-Stage Disease:** Heart-lung transplantation or bilateral lung transplantation with concomitant ASD repair.

IV SUMMARY TABLE FOR EXAM

Hemodynamic Profile on RHC	ESC 2020 Closure Recommendation	Management Strategy
PVR less than 3 WU (Qp:Qs 1.5 or greater)	**Class I** (Recommended)	Standard Percutaneous Device Closure (or surgical repair if anatomy unsuitable)
PVR 3 to less than 5 WU (Qp:Qs greater than 1.5)	**Class IIa** (Should be considered)	Standard or Fenestrated Closure in an expert ACHD/PH center
PVR 5 WU or greater (Reversible to less than 5 WU on PAH therapy with Qp:Qs greater than 1.5)	**Class IIb** (May be considered)	Treat-and-Repair: Upfront PAH therapy, repeat RHC, Fenestrated Closure + lifelong PAH drugs
Persistent PVR 5 WU or greater / Eisenmenger (Qp:Qs less than 1.5 or R-to-L shunt)	**Class III** (Contraindicated / Harm)	Defect closure prohibited; lifelong PAH therapy (e.g., Bosentan); Heart-Lung transplant evaluation

KEY REFERENCES

- **Baumgartner H, De Backer J, Babu-Narayan SV, et al.:** 2020 ESC Guidelines for the management of adult congenital heart disease, *European Heart Journal*, 2020.
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- **Stout KK, Daniels CJ, Aboulhosn JA, et al.:** 2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease, *Circulation*, 2019.