Atrial Septal Defect, Ostium Primum

Elizabeth Laverriere | Todd J. Kilbaugh

Risk

- Ostium primum ASD is a variant of AV canal defect. Classified as an ASD, it is actually an endocardial cushion defect.
- Less common than secundum ASDs, this defect comprises 0.5-1% of all congenital heart defects, and 15-20% of ASDs.
- Approximately 50% of pts with primum ASD are female.
- Ostium primum is most commonly associated with the genetic defect trisomy 21 (Down syndrome). It is also associated with Holt-Oram and Noonan syndrome.

Perioperative Risks

- Periop mortality rate: 1.5-6%, lower mortality if repair is done before onset of pulm Htn.
- Late in clinical course of unrepaired, clinically significant ASDs; CHF is common with a L-to-R shunt.
- Increased risk of atrial dysrhythmias, heart block, and air embolus with surgical repair.
- Significant risk of mortality if Eisenmenger syndrome (eventual reversal of the shunt into a cyanotic R-to-L shunt) has occurred. Surgical repair is often not recommended at this point.

Worry About

- AV valves, which are usually abnormal
- Abnormal conduction axis
- CHF and Eisenmenger syndrome

Overview

- Failure of inferior atrial septum to close at the level of the tricuspid and mitral valves.
- Symptoms present earlier, are more severe than in secundum ASD, and include dyspnea, fatigue, atrial arrhythmias, recurrent respiratory infections, and failure to thrive.
- L-to-R shunt increases pulmonary blood flow and right-sided volume overload.
- Progression of clinical course: CHF (more common than in secundum pts) and shunt reversal (R-to-L flow across ASD).
- Frequently associated with mitral regurgitation with a cleft in the anterior mitral leaflet (can cause left-sided volume overload) and/or tricuspid regurgitation.
- Diagnosis by ECHO; characteristic appearance: Absence of the lower atrial septum.
- Cardiac catheterization may be required to assess PVR and pulm Htn in large shunts.

Etiology

- Failure of septum primum to fuse with endocardial cushion to close ostium primum

Usual Treatment

- Asymptomatic pts require no medications.
- Diuretics are used for CHF.
- ACE inhibitors may be used for afterload reaction in the presence of mitral regurgitation.
- Antiarrhythmics are occasionally needed for atrial dysrhythmias.
- Percutaneous closure is not possible because the rim of the inferior atrial tissue is inadequate to prevent the device from impinging on the valves.
- Surgery is the definitive management, usually between 2-5 y, but it may be earlier if there is mitral regurgitation, CHF, or failure to thrive. The incision is median sternotomy, submammary, lateral thoracotomy, or transxiphoid. The defect is closed under direct visualization using CPB. Endocarditis prophylaxis is indicated for 6 mo after repair. Persistent AV valve abnormalities may require long-term prophylaxis.
- Consult a geneticist if syndromic.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Difficult intubation	Down syndrome	Down syndrome facies	
CV	Atrial dysrhythmias, right-sided heart failure L-to-R shunting, hypertrophic RA and RV Mitral regurgitation	Palpitations SOB, frequent fatigue Cyanosis if shunt reversal	Irregular rate and rhythm Right heart enlargement Normal S_1, wide, fixed splitting of S_2, and soft systolic ejection murmur; diastolic rumble	ECG (tall P wave, RBBB + RVH) CXR—cardiomegaly ECHO Cardiac catheterization Dye dilution study
RESP	Increased pulm blood flow Increased PVR	SOB, frequent URIs	Rales; wheezing	CXR—increased pulm vascular markings
GI	Feeding difficulty Hepatic dysfunction if severe CHF	FTT Jaundice	Hepatomegaly	LFTs; PT/ INR
CNS	Embolic stroke from chronic AFIB	Various neurologic changes		Head CT, cardiac ECHO
RENAL	Renal dysfunction if severe CHF			Cr and BUN

Key References: Rivenes SM: Ostium primum atrial septal defects. *eMedicine* <http://emedicine.medscape.com/article/890880-overview>, 2015 (Accessed 10.03.16.); Wasnick JD, Hillel Z, Kramer D, et al: Anesthesia for patients with congenital heart disease. In Wasnick JD, Hillel Z, Kramer D, et al editors: *Cardiac anesthesia and transesophageal echocardiography.* New York, 2011, McGraw-Hill.

Perioperative Implications

Preoperative Medications

- Oral or IV midazolam before procedure
- Antibiotic prophylaxis

Monitoring

- Routine monitors, arterial line, CVP; intraop TEE to assess anatomy before CPB, AV valve regurgitation, and function, ventricular function and to check for air and residual shunt after CPB; central and peripheral temp monitoring

Induction

- IV or inhalational: IV induction theoretically slowed by L-to-R shunt because of increased pulmonary blood flow; inhalational induction speed is increased; may place an epidural with loss of resistance to saline technique to avoid air embolism; must be placed 1 h before heparinization

Maintenance

- Balanced anesthetic: Combination of opioids, inhalational agent, and muscle relaxants.
- Reduced fraction of oxygen to maintain PVR and decrease L-to-R shunt.
- Avoid nitrous oxide to minimize size of air bubbles.
- Watch for shunt reversal with hypoxemia, hypercarbia, and hypothermia.

Extubation

- If intraop course is uneventful, then pt may be extubated at the end of the procedure.
- Control BP with milrinone, nitroprusside, or nitroglycerin.
- Keep mechanically ventilated if the repair has been complex or arrhythmias are present.

Adjuvants

- Watch for supraventricular dysrhythmias and AV conduction defects; must have pacing wires during surgical repair.

Postoperative Period

- Adequate analgesia for sternotomy or thoracotomy pain; pacemaker available for transient heart block

Anticipated Problems/Concerns

- Air emboli with vascular access.
- Dysrhythmias: SA node or AV node dysfunction.
- Depressed systolic function; Inotropic support, diuretics, and afterload reduction may be helpful.
- Third-degree AV block with repair of low-lying defects.
- Residual pulmonary Htn, which can lead to tricuspid regurgitation and right ventricular failure.
- Residual mitral valve insufficiency may remain or worsen.
- Endocarditis, especially with a residual cleft mitral valve.
- Pts with left ventricular outflow tract obstruction are at higher risk for reoperation.