Congenital Heart Disease: Acyanotic Disorders—Shunts

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Larry T. Mahoney, MD; David J. Skorton, MD

Atrial Septal Defects

Atrial septal defects (ASDs) occur in three main locations: the region of the fossa ovalis (such defects are termed ostium secundum ASDs); the superior portion of the atrial septum near the junction with the superior vena cava (SVC) (sinus venosus ASDs); and the inferior portion of the atrial septum near the tricuspid valve annulus (ostium primum ASDs). The ostium primum ASDs are considered to be part of the spectrum of atrioventricular septal defects (AVSDs).

Ostium secundum ASDs are the most common variety, accounting for over half of ASDs. A frequent accompanying defect is mitral valve prolapse. Relatively less prevalent is the sinus venosus defect. Anomalous pulmonary venous return is a common associated abnormality. The proximity of the sinoatrial node to the ASD may lead to sinoatrial node dysfunction and atrial arrhythmias.

Pathophysiology

ASDs are associated with left-to-right shunts of varying degrees. The main determinants of the direction and magnitude of shunt flow are the size of the defect and the relative compliances of the left ventricle (LV) and right ventricle (RV).

Clinical Presentation

Most patients with ostium secundum or sinus venosus ASD are asymptomatic through young adulthood. As the patient reaches middle age, compliance of the LV may decrease, increasing the magnitude of left-to-right shunting. Long-standing atrial dilatation may lead to a variety of atrial arrhythmias, including premature atrial contractions, supraventricular tachycardia, and atrial fibrillation. A substantial number of middle-aged patients will report dyspnea, particularly with exertion, even if they do not have pulmonary hypertension. Approximately 10% of patients with ostium secundum ASDs will progress to pulmonary hypertension associated with pulmonary vascular obstructive disease (Eisenmenger syndrome). As the pulmonary pressure rises, the left-to-right shunt will diminish and eventually be replaced by a right-to-left shunt; cyanosis and pulmonary hypertension will develop.

The hallmark of the physical examination in ASD is the wide and fixed splitting of the second heart sound. A systolic murmur (from increased pulmonary flow) is common, and if a large left-to-right shunt is present, the additional flow across the tricuspid valve may lead to a diastolic rumble reminiscent of tricuspid stenosis.

Laboratory Tests

All patients with suspected ASD should have an electrocardiogram, a chest x-ray, and an echocardiogram.

Electrocardiography. The QRS axis usually is normal in ostium secundum ASD but may be slightly rightward, and an rSR' pattern is common in the right precordial leads. In sinus venosus
ASD, the axis may be normal or relatively horizontal (less than 30°). Ectopic atrial rhythms or other evidence of sinoatrial node dysfunction may be seen.

**Radiologic studies.** The chest x-ray reveals enlargement of the right atrium (RA), the RV, and the main pulmonary artery. The pulmonary vessels exhibit diffuse enlargement because of increased pulmonary blood flow. Magnetic resonance imaging, magnetic resonance angiography (MRA), or cardiac catheterization will identify anomalous pulmonary veins; these modalities should be considered when there is suspicion of this associated abnormality in patients with sinus venosus ASD.

The patient with secundum ASD who has pulmonary hypertension may benefit from right-sided heart catheterization to ascertain the level of pulmonary arterial pressure and resistance.

**Echocardiography.** Echocardiography can confirm the presence of an ASD, determine its size, permit calculation of shunt flow through it, and identify any associated anomalies.

**Management**

Large ASDs (defined as those with a pulmonary-to-systemic flow ratio [Qp:Qs] of over 1.5:1) should be closed to prevent the development of pulmonary hypertension and reduce the risk of paradoxical emboli. Direct surgical closure has been the method used, but devices are now available that permit catheterization-based closure of many defects. Postclosure management includes periodic assessment for the development of atrial arrhythmias. The need for endocarditis prophylaxis varies.

**Atrioventricular Septal Defects**

The septal leaflet of the tricuspid valve normally inserts into the septum slightly closer to the apex than does the septal leaflet of the mitral valve [see Figure 2 -- omitted]. Thus, the small portion of septal tissue superior to the tricuspid septal leaflet insertion separates the RA from the LV and so is called the atrioventricular septum. The term AVSD refers to a complex spectrum of disorders involving abnormalities of the atrioventricular septum and, frequently, the atrioventricular valves. Nomenclature for this spectrum of disorders has varied; synonymous terms include atrioventricular canal defect and endocardial cushion defect.

**Pathophysiology**

The spectrum of AVSDs ranges from a simple ostium primum ASD to a complete AVSD, which allows free communication among all four cardiac chambers. Variations of the anatomy of the anterior leaflet of the mitral valve and the septal leaflet of the tricuspid valve include a cleft or other abnormality in either or both of these leaflets; accessory chordae that attach in anomalous locations and alter function of the valve leaflets; or a common atrioventricular valve leaflet that bridges the septal defect. Physiologic consequences vary according to the extent of the anomaly; for example, the addition of a cleft mitral valve anterior leaflet adds varying degrees of mitral regurgitation (MR). Larger defects that also involve the ventricular septum, as well as complete AVSDs, can be associated with torrential left-to-right shunts or an admixture of venous and arterial blood.

Patients with an unrepaired complete AVSD are at risk for developing pulmonary hypertension. Eisenmenger syndrome is particularly common in AVSD patients who also have Down syndrome (trisomy 21).

**Clinical Presentation**

Patients with isolated ostium primum ASDs may be asymptomatic until adulthood and then may present with fatigue, dyspnea, or symptoms related to atrial arrhythmias. Severe regurgitation of either atrioventricular valve can produce symptoms of heart failure or arrhythmias. Symptoms related to pulmonary hypertension occur in those patients who develop Eisenmenger syndrome.

Patients with only an ostium primum ASD will have clinical findings similar to those of patients with an ostium secundum ASD. The presence of a cleft in either atrioventricular valve will be associated with a pansystolic murmur. Finally, an additional pansystolic murmur can be found in patients with a complete AVSD.

**Laboratory Tests**

**Electrocardiography.** Left axis deviation is present in the majority of patients. The combination of physical findings of ASD along with left axis deviation on the ECG suggests the presence of an AVSD. RV conduction delay may be present as well.

**Radiologic studies.** The chest x-ray shows cardiomegaly and pulmonary vascular engorgement because of the left-to-right shunt.

**Echocardiography.** Echocardiography defines the specific anatomy and functional importance of
the defects. Preoperative echocardiographic assessment includes estimation of the severity of atrophicventricular valve regurgitation, the Qp:Qs ratio, and pulmonary arterial pressures. Postoperatively, echocardiography is used to identify and assess the significance of residual atrophicventricular valve regurgitation or residual shunt.

**Management**

The rare patient who presents in adulthood with complete AVSD should be evaluated for pulmonary hypertension. If pulmonary pressures are normal or if pulmonary hypertension is not prohibitive (i.e., pulmonary vascular resistance is less than 50% of systemic vascular resistance), then surgical closure of the defect and repair of the atrophicventricular valve anomalies should be undertaken. Postoperatively, patients are assessed for the adequacy of atrophicventricular valve repair and are monitored for evidence of residual shunt. In patients with residual MR, management focuses on the need for and timing of reoperation, which may involve either repair or replacement of the mitral valve. The patient should also be followed for the development of atrial arrhythmias.

**Ventricular Septal Defects**

VSDs are among the most common congenital cardiac disorders seen at birth but are less frequently seen as isolated lesions in adulthood. This is because most VSDs in infants either (1) are large and nonrestrictive (i.e., they permit equilibration of pressures between the ventricles) and therefore lead to heart failure, necessitating early surgical closure, or (2) are small and close spontaneously.

Classification systems for VSD vary but usually are referenced to the embryologic divisions of the ventricular septum into inlet, outlet, muscular, and membranous portions [see Figure 3 -- omitted]. The most common defects are perimembranous defects. Inlet VSDs, located more posteriorly, may be part of the spectrum of AVSDs (see above). Single or multiple defects may occur in the muscular septum (muscular VSD). Finally, outlet VSDs include subpulmonary defects, which may allow prolapse of an aortic cusp, leading to associated aortic regurgitation (AR).

**Pathophysiology**

Nonrestrictive VSDs permit equilibration of ventricular pressures between the RV and LV, whereas small defects produce a large pressure gradient across the defect, so right heart pressures remain normal. The magnitude of shunt flow across moderate or large VSDs depends on the relative resistances of the systemic versus the pulmonary vascular bed. Rarely, clinicians may encounter adult patients who have large, nonrestrictive defects in the absence of other lesions. Moderate pulmonic stenosis at either the valve or the subvalvular level may create increased resistance to right ventricular outflow sufficient to reduce the left-to-right shunt; consequently, patients with VSD and mild to moderate pulmonic stenosis may reach adulthood without experiencing symptoms. Adults with long-standing VSD and large shunts may develop Eisenmenger syndrome.

**Clinical Presentation**

With the exception of those patients who contract infective endocarditis or those with Eisenmenger syndrome, adults with VSD are asymptomatic.

The classic physical finding of a restrictive VSD is a harsh, frequently palpable, pansystolic murmur heard best at the left lower sternal border. Patients who have large defects that allow equilibration of ventricular pressures may present with less impressive murmurs than patients with small defects; the reason is that with small defects, there is a large gradient between the LV and the RV, which results in severe turbulence across the defect. When aortic cusp prolapse occurs, the murmur of AR will be audible.

**Laboratory Tests**

**Electrocardiography.** The ECG may be normal or show evidence of left ventricular hypertrophy (LVH) and a pattern of so-called diastolic overload, featuring prominent Q waves in left precordial leads V5 and V6 and in leads I and aVL.

**Radiologic studies.** The chest x-ray may be normal or show left ventricular enlargement and pulmonary arterial engorgement. Patients who have evidence of pulmonary hypertension should undergo right heart catheterization to determine the degree of pulmonary hypertension and the level of pulmonary resistance.

**Echocardiography.** Echocardiography is the procedure of choice for identifying the location, size, and hemodynamic significance of a VSD; the interventricular gradient should be determined (to estimate RV pressure), and an assessment should be made of increased pulmonary blood flow.

**Management**

Patients with ventricular septal defects in which the Qp:Qs ratio is greater than 1.5:1 should be considered for surgical closure. Patients with pulmonary hypertension may undergo closure if
Pulmonary resistance is no more than about 50% of systemic resistance. Aortic cusp prolapse with resultant AR may diminish the shunt magnitude, but the presence of a prolapse constitutes an additional potential indication for closure.

Early VSD operative closures were performed through a right ventriculotomy, but now, many defects—particularly those in the perimembranous septum—are closed through a transatrial approach; such an approach leads to fewer problems with RV dysfunction and arrhythmias. Continual progress is being made in the deployment of transcatheter closure devices. Currently, however, surgical closure of VSD is still the most common approach. Postclosure management involves assessment for residual or recurrent VSD and atrial or ventricular arrhythmias, as well as assessment of RV function.

**Patent Ductus Arteriosus**

During fetal life, the ductus arteriosus connects the pulmonary artery to the aorta. Soon after birth, as a result of changes in circulating prostaglandin levels and arterial oxygen saturation, the ductus constricts; later, it closes permanently. Failure of the ductus to close leads to the condition termed patent ductus arteriosus (PDA).

**Pathophysiology**

The shunt from aorta to pulmonary artery increases pulmonary blood flow and return to the left heart. The size of the defect and the relative resistances of the pulmonary and systemic vascular beds determine the degree of shunting. Adults who have PDAs commonly present either with a small lesion without a large left-to-right shunt or with larger lesions and Eisenmenger syndrome.

**Clinical Presentation**

Except for patients with Eisenmenger syndrome, most adults with small to moderate PDAs will be asymptomatic, unless endarteritis supervenes.

The pathognomonic physical finding of PDA is the continuous murmur. A continuous murmur is one that is audible throughout systole and into diastole to any extent. The classic PDA murmur is machinelike and extends through systole and to variable degrees into diastole, peaking in intensity at the time of S₂. The runoff of blood into the pulmonary artery in diastole will produce a wide pulse pressure because of low aortic diastolic pressure.

**Laboratory Tests**

**Electrocardiography.** The ECG in patients with PDA may be normal or may show evidence of LVH.

**Radiologic studies.** If the shunt is small, the chest x-ray may be normal. Patients with larger shunts will have associated cardiomegaly and increased vascular markings. In adults, calcium may be noted within the wall of the ductus.

**Echocardiography.** Echocardiography will identify the PDA and permit quantification of the Qp:Qs ratio.

**Management**

With the advent of reliable means of transcatheter closure of PDAs, common practice is to recommend that most PDAs be closed. In rare cases, the ductus may need to be closed surgically if transcatheter closure is not successful. Postoperative management includes assessment for the need of a residual shunt; although this is uncommon, Patients who develop pulmonary hypertension are managed in the same way as those with Eisenmenger syndrome [see Eisenmenger Syndrome -- omitted, below].

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Larry T. Mahoney, MD, Professor of Pediatrics and Director of Pediatric Cardiology, University of Iowa College of Medicine

David J. Skorton, MD, University of Iowa College of Medicine
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