Objectives After completing this article, readers should be able to:

1. Review the anatomy of ventricular septal defect (VSD) and atrial septal defect (ASD).
2. Describe the clinical variables in children who have VSD and ASD.
3. Delineate the evaluation of VSD and ASD.
4. Describe the rationale for medical or surgical management of VSD and ASD.

Introduction
Congenital heart defects affect slightly less than 1% of liveborn infants. Two defects, ventricular septal defect (VSD) and atrial septal defect (ASD), account for about 30% of congenital heart disease: VSD for 20% and ASD for 10%. These defects may occur as isolated lesions or as a component of complex defects. This article focuses on the isolated defects. Most children who have either VSD or ASD are diagnosed rapidly and will have normal life expectancy.

Ventricular Septal Defects
VSDs are defined by location and size. Each characteristic determines, in part, the pathophysiology and need for therapy.

Definition
VSDs are described by the anatomic location of the defect as seen from the right ventricle. The most common types are perimembranous, outlet, inlet, and muscular. Perimembranous VSDs (70% of all VSDs) are located in the upper fibrous region of the septum beneath the septal leaflet of the tricuspid valve and usually represent a single defect. Outlet VSDs (5% of all VSDs) are found between the left and right ventricular outflow tracts just above the parietal and septal bands and just below the pulmonary valve. They usually are single defects. Inlet VSDs (5% of all VSDs) are seen in the posterior portion of the ventricular septum beneath or behind the tricuspid valve and typically are single defects. Muscular VSDs (20% of all VSDs) are located in the muscular wall between the left and right ventricles in any portion of the septum, although the most common locations are at the apex, at the anterior portion of the septum, or in the central portion. These defects may be multiple and are difficult to see and judge in size from the right ventricular side (because of the coarse trabeculations).

Most children who have VSDs have only one of the previously noted types, but small muscular defects may coexist with defects in other regions of the ventricular septum. The most commonly associated cardiac defects are patent ductus arteriosus and coarctation of the aorta. Males and females are equally affected by VSDs.

Typically, VSDs are referred to as tiny (very small), small, medium, or large. The defects can be measured by echocardiography or angiography. The common reference measurement is the diameter of the aortic valve annulus. Those that are about the same size as the annulus are considered large, those half the diameter of the aortic root are considered moderate, and those less than half the aortic root size are considered small. For comparison, the aortic annulus of the newborn normally measures 10 mm and that of an adult is 20 mm.
Epidemiology

The etiology of VSD is not known, but a combination of genetic and other factors has suggested a multifactorial cause. Isolated VSD generally is not associated with genetic syndromes, although many children who have chromosomal abnormalities or recognized syndromes may have VSD because it is the most frequent congenital heart defect. Some, but not all, cases of inlet VSD are diagnosed in children who have trisomy 21. Failure to form the membranous septum, failure of conotruncal septation, and failure to fuse the muscular wall may lead to VSD.

Clinical Aspects

The clinical presentation of VSD varies with the size and location of the defect. Recent echocardiographic investigations have documented tiny muscular VSDs in up to 50 per 1,000 asymptomatic newborns. Most small VSDs are identified when the pediatrician hears a systolic murmur after the first day or two of life, typically at the 2-week examination. The murmur, which is usually loud (grade 3) and high-pitched, is heard in a baby who otherwise appears very well. Some children who have moderate-to-large VSDs do not have the typical loud, harsh murmur heard until they are 6 weeks old; by that time, the child may be exhibiting signs of pulmonary overcirculation with tachypnea, poor feeding, and poor weight gain. Recurrent pulmonary infections also may be associated with VSD.

Careful physical examination, including documentation of the pulses, is the most important tool for the primary pediatrician. If the murmur is high-pitched and well localized to the lower left sternal border and the child has a normally active precordium, the pathophysiologic effects of the VSD are minimal and the defect probably is small to moderate. If the murmur is medium-pitched, with an active precordium in a tachypneic patient, the VSD is likely to be moderate to large. A large VSD usually results in pulmonary overcirculation, the symptoms of which often are referred to as heart failure (in fact, the heart muscle works well, but pulmonary blood flow is excessive). There are no absolute or firm recommendations as to necessary laboratory tests. A chest radiograph is useful to confirm the presence or absence of cardiomegaly and increased pulmonary blood flow and can eliminate a pulmonary process as the cause of the tachypnea. Care must be taken in interpretation if the radiologist does not often read children’s radiographs. An electrocardiogram also can be obtained in the office, although test results are so nonspecific that they may appear normal in the face of significant cardiac findings and “abnormal” in a healthy child who has a murmur. Again, caution is advised in relying on computer-generated interpretation or interpretation by an adult cardiologist. The electrocardiogram may demonstrate left ventricular hypertrophy in the presence of a large left-to-right shunt, and the presence of significant right ventricular hypertrophy suggests the need for further evaluation.

Experienced clinicians are fairly adept at recognizing the small, hemodynamically insignificant VSD. An infant who has such a lesion may need no evaluation beyond that of an experienced practitioner listening to the child and confirming that the growth and respiratory pattern are normal. Echocardiography may be useful to confirm the diagnosis (Fig. 1). The most cost-effective method of evaluation is to refer the child to a pediatric cardiologist rather than to order an echocardiogram first (as is the common practice in internal medicine). Children who have significant murmurs and symptoms related to the

Figure 1. A. Echocardiographic image from subcostal view demonstrating VSD just under the aortic valve. B. Same image with color Doppler demonstrating flow into the right ventricle across the VSD (orange color) and flow out the left ventricular outflow tract (blue color).
cardiovascular system always should be referred. Cardiac
catheterization is employed for the most complex lesions
and for evaluation of the VSD in which pulmonary
hypertension is suspected.

Management and Prognosis

Management of the VSD depends on the size and patho-
physiology of the lesion. Patients who have tiny and small
defects that are associated with no cardiovascular symp-
toms should receive well-child care and antibiotic pro-
phylaxis when appropriate for prevention of infectious
endocarditis. Infants and children who have moderate
and large VSDs that produce respiratory symptoms and
impair growth should receive more aggressive therapy,
which may include diuretics, digitalis, afterload reduc-
tion, and increased caloric density of feedings. The use of
digitalis is controversial because the ventricular
function usually is normal. Digitalis does have a weak
diuretic effect and may blunt the sympathetic re-
response seen in this condition. Some VSDs, usually
those that are peri-
membranous or muscular,
become smaller with time,
and the child can be fol-
lowed and managed with-
out surgery. Most inlet or outlet VSDs require surgical
repair because spontaneous closure is rare. Growth fail-
ure despite maximal medical management is an indica-
tion for surgical closure. A child weighing as little as 2 kg
can be placed on cardiopulmonary bypass support and
undergo successful closure of the defect. If there is
associated aortic insufficiency, even a small VSD may
require surgical closure.

We recommend that large VSDs be closed surgically
(primary with suture alone or with a patch) in the first
year of life and that moderate-size defects be closed in the
second year of life. The occasional defect complicated by
irreversible pulmonary hypertension and vascular dam-
age to the lungs is not considered operable. There is a
growing role for catheterization-placed devices to close
certain VSDs in children in whom surgery may not be
advisable. In some instances, large muscular VSDs can be
closed with a device. Injury to chordal attachments of the
atrioventricular valves, injury to the conduction system,
and device failure limit the use of these procedures.

The operative mortality is less than 5% for uncompli-
cated VSD. Children who have VSDs that either close
spontaneously or with surgery generally do well, with an
average life expectancy. Lifetime risks are infectious en-
docarditis, aortic insufficiency, and arrhythmia. There is a
slightly increased risk for having another child who has a
heart defect in a family in which one child has a VSD.

In general, the prognosis for a child who has a VSD is
very good. Most VSDs are hemodynamically insignifi-
cant, close spontaneously, and are not associated with
any other conditions. The VSDs that are moderate or
large and are associated with growth failure or recurrent
pulmonary infections can be corrected surgically, with a
low mortality rate, few postoperative sequelae, and good
results. Antibiotic prophylaxis is recommended for all
VSDs preoperatively and for children who have a persis-

Atrial Septal Defects

ASDs may be located at various sites in the septum and
range in size from small to large. They are two to three
times more common in girls than in boys and usually
occur in otherwise healthy children. Rarely, ASD may be
associated with an inheritable disorder that has skeletal or
other defects, such as the Holt-Oram syndrome.

Definition

Secundum ASDs (75% of all ASDs) are located in the
central portion of the septum (fossa ovalis). Tissue may
be totally deficient or strands of tissue may traverse the
opening (fenestrated ASD). This ASD may close and may
be associated with mitral valve prolapse. Primum ASDs
(15% of all ASDs) are seen in the lower portion of the

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septum just above the atrioventricular valves. They usually are associated with a cleft in the anterior leaflet of the mitral valve and generally are considered a part of the atrioventricular septal defects group. Mitral regurgitation is common, and the defect size generally does not decrease with time. Sinus venosus ASDs (10% of all ASDs) are found in the upper portion of the septum at the junction with the superior vena cava, usually are associated with partial anomalous pulmonary venous return, and typically do not decrease in size with time. Persistent patent foramen ovale (PFO) is located in the middle portion of the atrial septum, may close with time, and remains probe-patent in 20% to 25% of affected people throughout life. Recently, there has been concern that they may represent a risk factor for stroke.

Commonly, the murmur of an ASD may not be identified until the prekindergarten physical examination. A grade 3 systolic ejection murmur may be heard at the upper left sternal border with a widely split second heart sound in a child who otherwise has been well. Often, an ASD may be identified when an echocardiogram is performed for another reason (evaluation of a murmur or an ill neonate) (Fig. 2). Although classical teaching has suggested that spontaneous closure occurs less frequently in ASD than in VSD, recent studies using echocardiography have demonstrated that spontaneous closure occurs frequently in ASD.

Epidemiology
The etiology of ASD has been attributed to multifactorial causes, with errors occurring in the programmed complex embryology of the formation of atrial septa. Secundum ASDs, which account for most ASDs, are formed by excessive resorption of the septum primum or deficiency of the septum secundum. Primum ASDs are formed when growth of the endocardial cushions is deficient. Sinus venosus ASDs are formed by incomplete septation of the upper portion of the atrial division and are uncommon. The foramen ovale is functionally closed in most people by 2 to 3 months of age, although 20% to 25% remain probe-patent throughout life.

Clinical Aspects
Small ASDs or PFOs produce no discernible symptoms. A baby who has an ASD or PFO may have mild cyanosis with prolonged crying. Murmurs may be minimal or absent in a child who has a small ASD. The size of the defect in the atrial septum and the compliance of the right ventricle determine the pathophysiology of the defect. Moderate or large ASDs are associated with significant left-to-right shunt and an increase in pulmonary blood flow. This increased flow to the right atrium, right ventricle, and pulmonary artery produces a murmur at the upper left sternal border. When the left-to-right shunt is large enough, a diastolic flow rumble may be audible at the lower left sternal border, representing increased flow across a normal tricuspid valve. Right ventricular compliance determines the amount of flow across the defect, most of which occurs during diastole. In general, the left atrial pressure is higher than the right, favoring left-to-right shunting. After the newborn period, the right ventricle becomes thinner and more compliant than the left ventricle, thus determining the lower right atrial pressure (a reflection of the right ventricular end-diastolic pressure) and left-to-right flow. As the volume of pulmonary blood flow increases with time, pulmonary valve closure is delayed, and the second heart sound is typically widely split and fixed. The child who has a loud systolic murmur and wide splitting of the
second heart sound should be evaluated by a pediatric cardiologist.

Somatic growth in children who have all but large ASDs generally is normal. The chest radiograph usually demonstrates mild-to-moderate cardiomegaly with increased pulmonary vascular markings and a prominent main pulmonary arterial segment. If the shunt is large, the older child or adult may complain of shortness of breath and exercise limitation. Results of electrocardiography usually are normal in children who have PFO or small ASDs. Right ventricular hypertrophy is seen with moderate or large ASD. The pattern of right ventricular hypertrophy of volume overload may be indistinguishable from incomplete right bundle branch block or terminal conduction delay.

PFO is of concern to physicians, particularly neurologists or internists, caring for adults. Studies of patients who have had strokes documented a higher-than-expected incidence of PFO compared with controls. Probe-patent PFO is very common, and stroke is far less common, although it is very significant in terms of mortality, morbidity, and to the health care dollar. PFOs are identified in virtually all infants in whom echocardiography is performed. Routine documentation of closure has not been current practice. Closure of PFOs, either by surgery or a device placed during interventional catheterization, in patients who have had strokes is a matter of ongoing debate and evolving practice. In general, stroke patients who are younger than 60 years of age and have a large PFO and no other identifiable cause of embolic stroke are considered for device closure following a second event (either stroke or transient ischemic attack).

Management and Prognosis

Currently, surgical closure is recommended for all moderate and large ASDs. The defect may be closed with sutures (primary closure) or a patch. The risks of nonclosure include right heart enlargement and decreased right ventricular compliance, paradoxic embolism, arrhythmia, and a small chance of the development of pulmonary vascular obstructive disease (<10%). Most ASDs are closed electively in childhood, usually before the child enters school. Long-term follow-up studies suggest that functional outcome is superior to that of those who have the defect closed later in life. The surgical mortality is less than 1%, and the morbidity is low. The risk of atrial arrhythmias continues postoperatively. Transcatheter closure is becoming more common practice as devices gain United States Food and Drug Administration approval. The cost and need for general anesthesia are similar to that of traditional surgery, although the risks of cardiopulmonary bypass and the surgical scar are eliminated with device closure. ASDs can be corrected surgically with a limited thoracotomy, and an inframammary incision may be used for cosmetic reasons.

The prognosis for patients who have ASD is excellent. Endocarditis prophylaxis is recommended for 6 months after closure until the patch is covered by endothelium. Sports participation is allowed both pre- and postoperatively. There is a small recurrence risk for subsequent children in the family or with offspring of the child who has the ASD.

Suggested Reading

Hanson LK, Oshoj H. High prevalence of interatrial communications during the first three months of life. Pediatr Cardiol. 1977;18:83–85
Locke JE. Patent foramen ovale is indicted, but the case hasn’t gone to trial. Editorial. Circulation. 2000;101:838
**PIR Quiz**

Quiz also available online at www.pedsinreview.org.

5. Which of the following statements about the relative occurrence of ventricular septal defects (VSDs) is most accurate?

A. Nearly 75% of all VSDs are of the inlet form.
B. Nearly 75% of all VSDs are of the muscular form.
C. Nearly 75% of all VSDs are of the outlet form.
D. Nearly 75% of all VSDs are of the perimembranous form.
E. Perimembranous, muscular, outlet, and inlet VSDs occur with approximately equal frequency.

6. Which of the following statements about the relative gender incidence of VSDs is most accurate?

A. Almost all VSDs occur in female infants.
B. Almost all VSDs occur in male infants.
C. Nearly 75% of VSDs occur in female infants.
D. Nearly 75% of VSDs occur in male infants.
E. VSDs occur equally in male and female infants.

7. Small VSDs that ordinarily do not cause problems in development or general health usually are associated with a murmur that sounds:

A. Loud and high-pitched.
B. Loud and low-pitched.
C. Soft and high-pitched.
D. Soft and low-pitched.
E. Soft but varies significantly in pitch.

8. VSDs large enough to produce pulmonary overcirculation usually are indicated by which one of the following complexes of symptoms?

A. A harsh, gurgling heart murmur.
B. Generalized bloating and cyanosis.
C. Rapid breathing.
D. Rapid breathing, poor feeding, and poor weight gain.
E. Shallow breathing and facial bloating.

9. Approximately what portion of patent foramen ovales close by the age of 2 to 3 months?

A. Virtually 100%.
B. 90% to 95%.
C. 85% to 90%.
D. 75% to 80%.
E. 65% to 70%.

10. Which of the following statements regarding treatment of patients who have atrial septal defect (ASD) is most accurate?

A. Medical management is advocated first, with surgery reserved for unresponsive cases.
B. Medical management is effective for almost all cases.
C. Spontaneous closure is common; watchful observation is recommended into adolescence.
D. Surgical closure is recommended for all moderate and large ASDs.
E. Surgical closure is recommended for almost all ASDs.
Ventricular and Atrial Septal Defects
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