

Atrioventricular Septal Defects (AVSD)

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Anatomy and Physiology

AVSDs, or canal defects, are characterized by abnormal endocardial cushion development, resulting in deficiency of the atrioventricular septum. There is a spectrum of size of the atrial and ventricular components of the defect which may range from very small to very large and altered formation of the atrioventricular valves (1,2). In the *complete form* of this malformation there is an inferior interatrial communication or ostium primum defect, an interventricular communication at the superior aspect of the inlet or posterior muscular septum and a common atrioventricular valve. In the *partial form*, an ostium primum ASD is accompanied by a cleft or commissure in the left-sided atrioventricular valve and two functionally distinct atrioventricular valvular orifices are generally identified. The prevalence of these defects is frequent among patients with Down syndrome.

Complete AVSDs are typically associated with non-restrictive intracardiac shunting, excessive pulmonary blood flow, and excessive systemic pressures in the right ventricle and pulmonary artery. Without intervention this may result in early pulmonary vascular changes and the development of fixed pulmonary vascular obstructive disease. The severity of atrioventricular valve regurgitation also influences the clinical presentation. Partial AVSDs are less likely to be associated with pulmonary overcirculation severe enough to cause significant heart failure symptoms.

Long-Term Outcome. Most adults with the complete form of this defect have undergone complete repair in childhood. In some patients, initial palliation may have consisted of pulmonary artery banding to restrict pulmonary blood flow, followed by subsequent definite repair. Over the last several decades, the surgical approach has evolved from a two-stage intervention to a single surgical strategy of primary repair in infancy (3). The long-term outlook after repair of AVSDs is generally good. In a few patients, uncorrected defects have resulted in Eisenmenger's physiology, rendering them inoperable candidates. This is associated with significant late morbidity and early death (4,5,6). Although definitive repair is usually accomplished during childhood, various publications have documented the results of surgical intervention in adults with partial forms of defects. Patients older than 40 yr of age may undergo reparative surgery with low operative risk (7); however, they may require long-term surveillance because late mitral valve dysfunction may occur. Among 50 patients who underwent surgery for partial AVSDs (mean age, 36.6 yr; 39 of them being intervened for the first time for a substantial shunt), a low operative risk was reported and excellent long-term results were achieved (8). Complications after repair of an AVSD include residual intracardiac shunting, left atrioventricular valve stenosis or regurgitation, and subaortic obstruction.

Transesophageal echocardiography (TEE)

In patients with AVSDs, TEE is useful in confirming the anatomy and defining the type and extension of the septal defects (9). Two- and three-dimensional TEE imaging has been shown to

be of benefit preoperatively, not only during the initial repair but also when reinterventions have been necessary (10,11). The deficiency in the atrial and ventricular septa and the large common atrioventricular valve can be readily identified in the mid esophageal four-chamber view (Fig. 1) The ventricular component of the defect is best defined during systole when the atrioventricular valves are closed. If there are dense chordal attachments to the crest the ventricular component may be delineated with color flow imaging. Charecterization of the “bridging leaflets,” which span the common orifice, assists in the classification of these defects into types A, B, or C as proposed by Rastelli et al. (1) according to the anterosuperior bridging leaflet morphology. (Fig. 2). The posteroinferior leaflet almost invariably has chordal attachments to the crest of the septum. Other information of interest that is well outlined by TEE includes atrioventricular valve competency, associated ventricular outflow obstruction, and noninvasive assessment of pulmonary artery pressures. Additional muscular VSD’S can be found in 10% of the patients. Other lesions less frequently encountered include tetralogy of Fallot (3.5%), valvar pulmonar stenosis, double outlet right ventricle, truncus arteriosus and ventriculoarterial discordence. One of the most frequent associated lesions is left ventricular outflow obstruction, including subaortic stenosis either due to chordal attachments to the crest of the septum or due to a subaortic fibrous ridge (4%), valvar stenosis and aortic subaortic coarctation. Presence of a patent ductus arteriosus is common, particularly in Down’s Syndrome and can create an additional risk of Pulmmary hypertension. In the postoperative patient, TEE can assist in the determination of residual defects, status of the atrioventricular valves, and evaluation of ventricular function. Tee is also helpful in the situation of assessing ventricular size, in unbalanced AVSD’s where one ventricle is smaller an alternative surgical approach is pulmonary artery banding which allows the diminutive ventricle to grow.

TRANSESOPHAGEAL ECHOCARDIOGRAPHY (TEE) IN THE EVALUATION OF AVSD

Tee Planes & Information Provided	Postsurgical Evaluation
ME 4-CH and 2-CH--morphology of AVV, bridging leaflets and their attachments in the complete form of the defect, severity of AVV regurgitation. Size/location of intracardiac defects.	Residual shunts, AVV regurgitation, LVOT obstruction, ventricular function.

AVSD : Long term outcome

Partial or intermediate forms

- Presentation of unrepaired partial defect (ostium primum ASD and cleft mitral valve) in adulthood not uncommon; most are symptomatic by 40 years of age
- in the short term, the results of surgical repair similar to those after closure of secundum ASDs
- mitral valve regurgitation, sub AS and atrioventricular block may develop or progress occasionally mitral stenosis results, with surgical revision required in 5% to 10% of patients

Complete forms

- □ most presently corrected in infancy □
- If prior palliation with pulmonary artery banding, may have resulted in inadequate protection of pulmonary vascular bed □
- uncorrected defect in the adult often associated with Eisenmenger's syndrome
- first degree atrioventricular block is common and complete atrioventricular block may occur

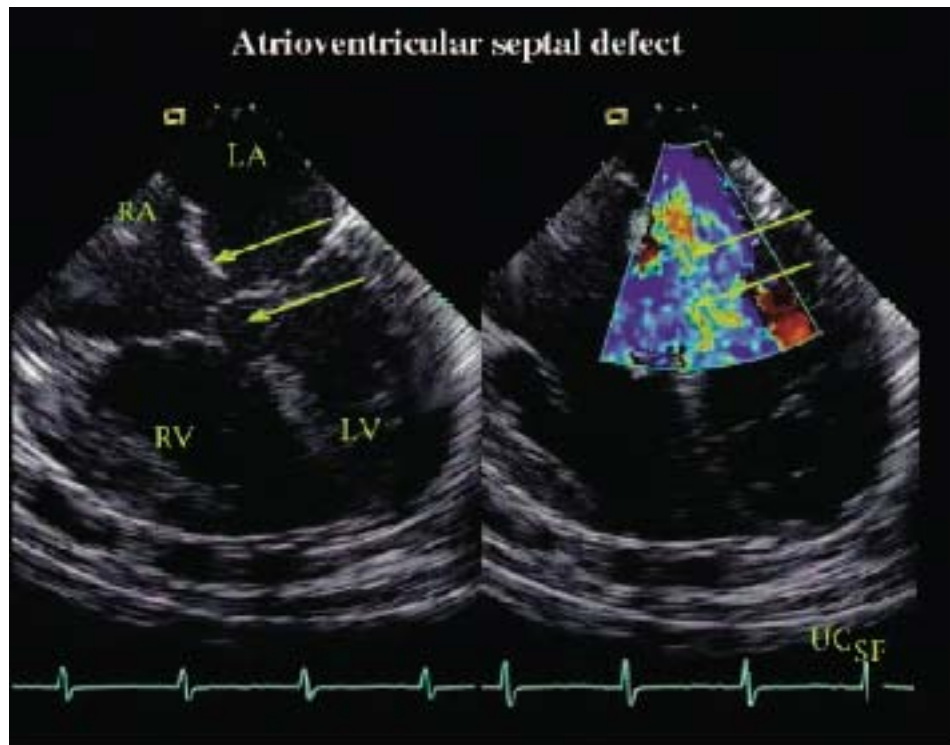


Figure 1. Complete Atrioventricular Septal Defect. Left: Mid-esophageal four-chamber view demonstrating a complete atrioventricular septal defect. The malformations characteristic of this lesion are shown, namely a primum atrial septal defect at the inferior aspect of the interatrial septum (upper arrow) and the posteriorly located, inlet-type ventricular septal defect (indicated by the lower arrow). Bridging of the common atrioventricular valve over the ventricular septum is seen. LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle. Right: Color flow Doppler showing extensive left-to-right atrial and ventricular level shunting.

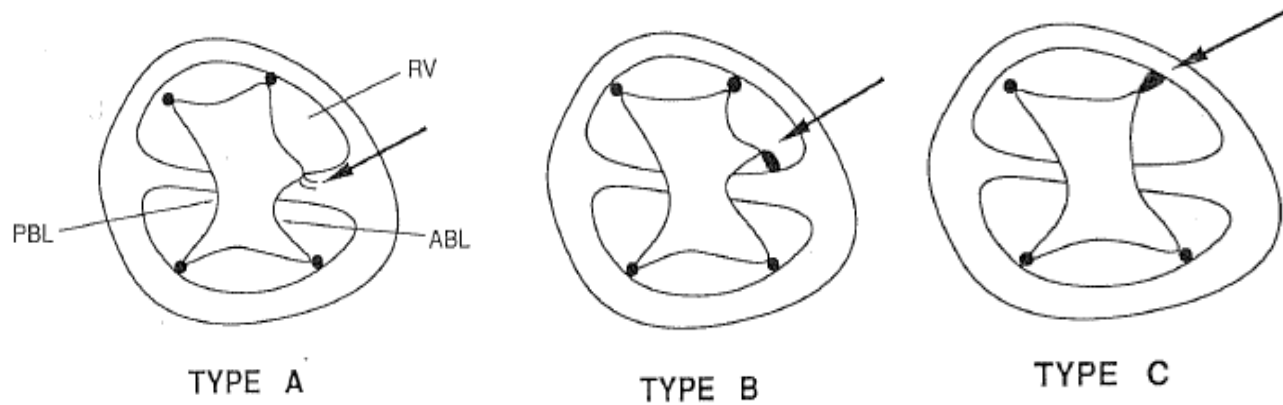


Figure 2. Schematic demonstration of the three different types of atrioventricular septal defects according to Rastelli et al. (1). **Top.** The anterior bridging leaflet (ABL) is attached to the crest of the ventricular septum (arrow), characteristic of a type A defect. **Middle.** The anterior bridging leaflet is attached to a papillary muscle on the right side of the ventricular septum (arrow), characteristic of the type B defect. **Bottom.** The anterior bridging leaflet is unattached to the septum but is attached to a large anterior papillary muscle in the right ventricle (arrow), characteristic of a type C defect. (From Higgins C, Silverman NH, Kersting Sommerhoff B, Schmidt KG, *Congenital heart disease: Echocardiography and magnetic resonance imaging*. New York: Raven Press, 1990)

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