

When a PFO is clinically suspected and a shunt across the region of the fossa ovalis cannot be detected, then a saline contrast bubble study is indicated. Please refer to Chapter 13 Paradoxical Embolism across a PFO.

Ventricular Septal Defects

A ventricular septal defect (VSD) is a congenital lesion where there is a communication between the left and right ventricles. VSDs account for approximately 20-25% of all congenital heart lesions.

With a VSD blood is usually shunted left-to-right as the pressure within the left ventricle (LV) is normally much higher than the pressure in the RV. Due to this left-to-right shunting, there is an increased volume of blood flow to the lungs, which in turn leads to an increase in the pulmonary venous return into the left heart. Therefore, in the presence of a significant left-to-right shunt across a VSD, there is dilatation of the left heart chambers. Dilatation of the right heart chambers may also occur when there is associated pulmonary hypertension. In order to appreciate the aetiology for each type of VSD it is important to first understand the embryological development of the IVS.

Embryological Development of the IVS

As for the formation of the IAS, the formation of the IVS is also quite complex. In particular, there are four distinct segments of the definitive IVS and each segment develops at different stages (Fig. 15.18). This complex staged formation of the IVS ensures that there is proper alignment of the AV valves with the respective ventricles and that the great arteries arise from their proper ventricles. As a result, in the fully developed heart, there are four major components of the IVS (Fig. 15.19). The

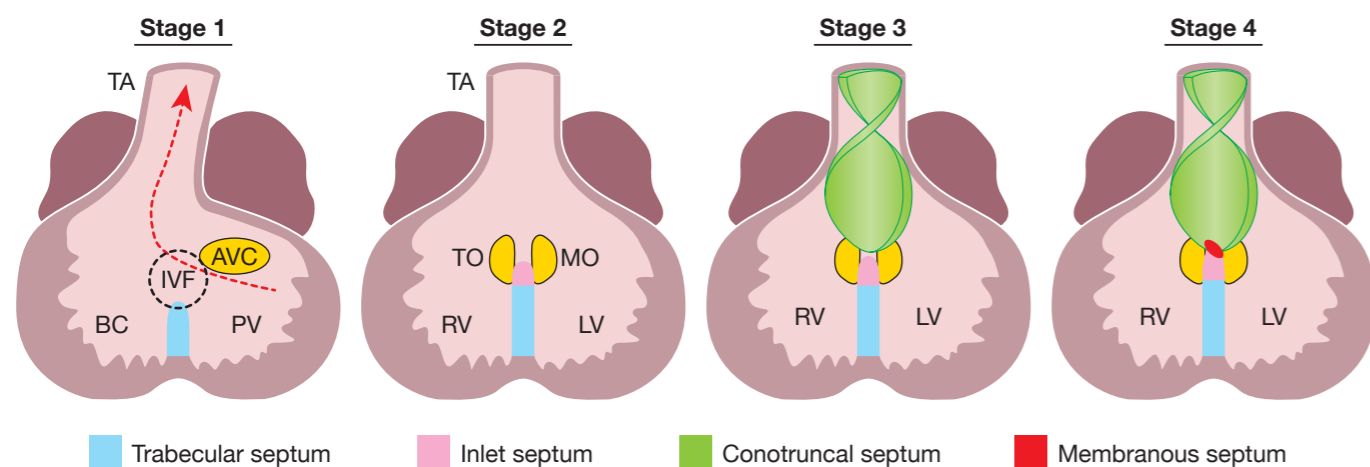


Figure 15.18 These schematics illustrate the formation of the interventricular septum (IVS). Note that the bulbus cordis (BC) becomes the definitive right ventricle (RV) and the primitive ventricle (PV) becomes the definitive left ventricle (LV). **Stage 1:** The trabecular IVS is the first to form, it grows from the apex towards the base of the heart. However, before this septum reaches the atrioventricular canal (AVC), it stops. This is because at this stage of development the truncus arteriosus (TA) only communicates with the bulbus cordis so blood flow from the primitive ventricle travels to the truncus arteriosus via the interventricular foramen (IVF). If fusion of the trabecular septum with the AVC occurred too soon the primitive ventricle would be shut off from the truncus arteriosus. **Stage 2:** As the truncus arteriosus moves to a central position and the AVC also shifts centrally to then divide into the tricuspid and mitral orifices (TO and MO), trabeculations from the inlet region form the inlet IVS which grows into the ventricular cavity perpendicular to the trabecular IVS. The fusion of these two septa forms the bulk of the muscular IVS. **Stage 3:** The conotruncal septum grows in a spiral direction and divides the truncus arteriosus into the aorta and pulmonary artery and the conus cordis into the right and left ventricular outflow tracts; thus, the outlet end of this septum forms the outlet muscular septum. This septum fuses with the endocardial cushions, the trabecular IVS and the inlet IVS. **Stage 4:** When there is complete division of the truncus arteriosus into the aorta and pulmonary trunk, the membranous IVS forms. Formation of the membranous IVS results in the complete closure of the interventricular foramen.

Note: The ventricular septum is a complex curved structure; however, for simplicity the formation of the IVS is illustrated in a two-dimensional plane.

muscular IVS forms the bulk of the IVS and includes the inlet, trabecular, and outlet (infundibular) components. The inlet septum separates the tricuspid and mitral valves and extends from the septal tricuspid leaflet to the distal cordal attachments of the tricuspid valve. The outlet (infundibular) septum separates the right and left ventricular outflow tracts and extends up to the pulmonary valve. The trabecular IVS is the largest part of the muscular IVS, extending from the inlet septum out to the apex and up to the outlet septum. The membranous septum accounts for a small section of the IVS; it is located at the base of the heart between the inlet and outlet components of the muscular septum and below the right and non-coronary cusps of the aortic valve. The septum between the insertion of the septal tricuspid leaflet and the anterior mitral leaflet represents the atrioventricular septum, because it lies between the RA and the LV.

Types of VSD

There are four types of VSD (Fig. 15.20) and these defects are named based on their anatomic location within the IVS. The perimembranous VSD is located beneath the aortic valve bordering the septal tricuspid leaflet and inferior to the crista supraventricularis. This defect is the most common type of VSD accounting for approximately 80% of all VSDs. These defects are also referred to as paramembranous, membranous or infracristal VSDs.

Muscular VSDs are the most common defects in infancy but as many of these defects spontaneously close; overall, they account for only about 20% of all VSDs. Muscular VSDs are located anywhere along the trabecular septum and they are bordered only by muscle. Muscular VSDs can be classified as anterior (anterior to the septal band), mid-muscular (posterior to the septal band), apical (inferior to the moderator band), and posterior (beneath the septal tricuspid leaflet). Muscular VSDs may also be multiple.

Inlet VSDs result from deficiency of the inlet septum; these defects are located posterior and inferior to the membranous IVS beneath the tricuspid and mitral valves. An inlet VSD, also referred to as an AV canal type defect, accounts for approximately 5% of all VSDs.

A supracristal VSD occurs due to deficiency in the IVS above and anterior to the crista supraventricularis; these defects are located anterior to the membranous IVS and beneath the semilunar valves. Supracristal VSDs are also referred to as outlet, infundibular, conal, doubly committed or subarterial defects; they account for approximately 5% of all VSDs. VSDs usually occur as isolated lesions. However, VSDs are often components of other more complex congenital cardiac anomalies such as tetralogy of Fallot (see below). Also VSDs often coexist with patent ductus arteriosus (PDA) or

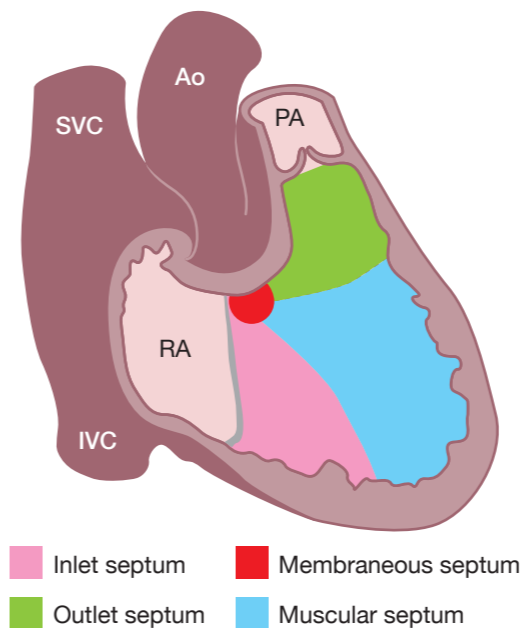


Figure 15.19 Four components of the interventricular septum are shown from a right ventricular aspect. See text for details. Ao = aorta; IVC = inferior vena cava; PA = pulmonary artery; RA = right atrium; SVC = superior vena cava.

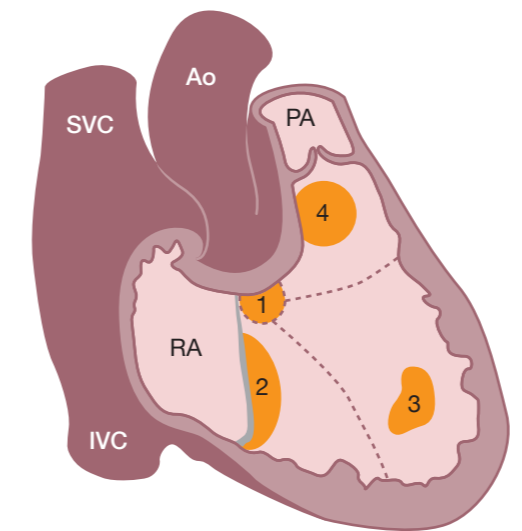


Figure 15.20 This schematic illustrates the four types of ventricular septal defect (VSD) from a right ventricular aspect: 1 = perimembranous VSD; 2 = inlet VSD; 3 = muscular VSD; 4 = supracristal or outlet VSD. Ao = aorta; IVC = inferior vena cava; PA = pulmonary artery; RA = right atrium; SVC = superior vena cava.

coarctation of the aorta. Furthermore, with a perimembranous or supracristal VSD, aortic regurgitation (AR) may occur due to distortion or incomplete support of the aortic valve cusps or annulus (see Fig. 7.43).

Role of Echocardiography in VSD

When assessing a patient with a VSD, it is important to:

- determine the type of defect
- estimate the anatomic size of the defect
- determine the shunt direction
- estimate the haemodynamic significance of the shunt
- estimate the RVSP.

VSDs Type and Shunt Direction

The VSD type is based on its anatomic location as well as the echocardiographic view in which the defect is seen. In particular, perimembranous and outlet VSDs can be differentiated from the parasternal short axis view (Fig. 15.21) while inlet VSDs and perimembranous VSDs can be differentiated from the apical views (Fig. 15.22).

The formation of aneurysmal tissue along the RV side of a perimembranous VSD is often seen (Fig. 15.23). On real-time imaging, aneurysmal tissue displays a characteristic 'windsock' appearance. The progressive development of aneurysmal tissue often leads to spontaneous closure of these defects.

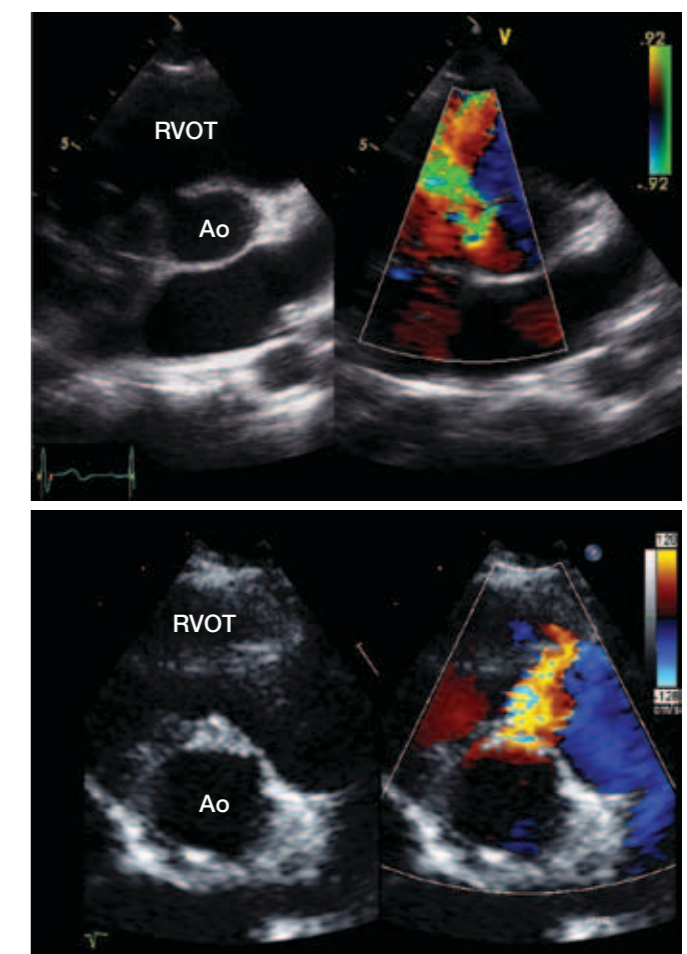


Figure 15.21 The parasternal short axis view can be used to differentiate between a perimembranous and a supracristal (outlet) VSD. A perimembranous VSD is located between 9 and 12 o'clock (top) while an outlet or supracristal VSD is located between 12 and 3 o'clock (bottom). Observe on the colour Doppler images flow through both VSDs is left-to-right; this appears as turbulent flow towards the transducer. Ao = aorta; RVOT = right ventricular outflow tract.