Being able to view the morphological details of the heart malformation in any patient with the diagnosis of atrioventricular septal defect (AVSD) is invaluable in surgical planning, especially in the current era of modified 1-patch versus 2-patch techniques. Real-time 3-dimensional transesophageal echocardiography (RT 3D TEE) is a recently developed technique that allows real-time high-quality images akin to viewing the heart specimen. We present the principal pathomorphological features of AVSD as visualized by RT 3D TEE.

The hallmark of AVSD is a common atrioventricular junction guarded by a common atrioventricular valve.1 In the normal heart, when seen from atrial perspective, the aortic valve is wedged between the mitral and tricuspid valve. In AVSD, because of the common AV junction, the aortic valve is in an “unwedged” position (Figure 1). The valve consists of 5 leaflets: the superior and inferior bridging leaflets, each of which overrides the ventricular septum, a left mural leaflet, a right anterior, and a right inferior leaflet (Figure 2). Whether the bridging leaflets are joined together distinguishes the complete form from the partial form. In partial AVSD (also termed “ostium primum ASD”), a tongue of leaflet tissue connects the bridging leaflets and the undersides of these leaflets are adherent to the crest of ventricular septum, dividing the common valve functionally in 2 separate valvar orifices and allows shunting at the atrial level only (Figure 3). By comparison, in the heart with complete AVSD, the common AV valve has a common valvar orifice. Although the bridging leaflets are tethered to the ventricular septum to varying extents, the superior bridging leaflet is often free-floating. Shunting exists at both interatrial and interventricular levels owing to the gap between the inferior margin of true atrial septum and the crest of ventricular septum (Figure 4). RT 3D TEE well demonstrates the relative sizes of the valvar orifices, allowing designation of balanced or unbalanced forms. The morphology of the leaflets of the AV valve and the so-called mitral cleft (Figure 5) is also well visualized. Left ventricular outflow tract obstruction is an important concern after surgical repair since the outflow tract is elongated and inherently narrow in this malformation. The scoop of the ventricular septum and the relationship of the superior bridging leaflet to the septal crest contribute to the characteristic “goose-neck” deformity of the left ventricle seen on angiography (Figure 6). The depth of the scoop at the left ventricular outflow tract is readily visible on RT 3D TEE, allowing the surgeon to see around the corner before carrying out surgical repair (Figure 7).

In summary, RT 3D TEE can allow demonstration of “living” anatomy of AVSD in detail, using novel perspectives that are invaluable for surgical planning.

Disclosures
None.

Reference

Key Words: atrioventricular septal defect / 3-dimensional transesophageal echocardiography

Figure 1. The “unwedged” position of the aorta. A, RT 3D TEE image from atrial perspective in a normal subject showing the aortic valve (Ao) wedged between the mitral (MV) and tricuspid (TV) valves. B, RT 3D TEE image from atrial perspective in a patient with AV septal defect. There is a common AV junction, and the aorta (Ao) is located anteriorly in an unwedged position. SB indicates superior bridging leaflet; IB, inferior bridging leaflet; LM, left mural leaflet; RA, right anterior leaflet; and RI, right inferior leaflet.
Figure 2. The 5 leaflets. A, RT 3D TEE image of the AV junction in a patient with AVSD and B, the corresponding view in an anatomic specimen. The common AV valve consists of 5 leaflets: the superior and inferior bridging leaflets, each of which overrides the ventricular septum, a left mural leaflet, a right anterior, and a right inferior leaflet. SB indicates superior bridging leaflet; IB, inferior bridging leaflet; LM, left mural leaflet; RA, right anterior leaflet; and RI, right inferior leaflet.

Figure 3. The common AV valve. RT 3D TEE shows the common valve seen “en face” from atrial perspective at systole (A) and diastole (B). In the systolic frame, the lines of apposition among leaflets are easily recognized. The diastolic frame shows the superior and inferior bridging leaflets joined together (arrow) in this heart with the “partial” form of AV septal defect. SB indicates superior bridging leaflet; IB, inferior bridging leaflet; LM, left mural leaflet; RA, right anterior leaflet; and RI, right inferior leaflet.

Figure 4. RT 3D TEE image of a “complete” AV septal defect. A, RT 3D TEE image of a “complete” AV septal defect; this perspective is obtained from a 4-chamber view slightly rotated rightward. B, the corresponding view in an anatomic specimen. The double dotted arrow marks the deficiency between the edge of the atrial septum (AS) and the superior crest of ventricular septum (VS) in a patient with “complete” AV septal defect. The bridging leaflets (BL) are not completely adherent to the crest of the VS. Note that the BL is floating, allowing the potential for both interatrial and interventricular shunts. LA indicates left atrium; LV, left ventricle; RA, right atrium; and RV, right ventricle. See also online-only Data Supplement Movie 1.
Figure 5. The cleft of the left AV valve. A, View from atrial perspective. B, View from a ventricular perspective. The common dictum “cleft of the mitral valve” is incorrect because the left AV valve is not similar to the bishop’s miter, being a trileaflet structure. The cleft is just the line of apposition between the superior (SB) and inferior (IB) bridging leaflets (arrows). Ao indicates aortic valve.

Figure 6. The left ventricular outflow tract. View at diastole (A) and at systole (B) displaying the inherently narrow outflow tract (O) of the left ventricle. SB indicates superior bridging leaflet; IB, inferior bridging leaflet. See also online-only Data Supplement Movie 2.

Figure 7. The scoop. View at systole (A) and a heart specimen (B) displaying the scoop of the ventricular septum (arrows) and the narrow outflow tract (O) viewed from the left ventricle. This area is hidden from the surgeon’s view during surgical repair. SB indicates superior bridging leaflet; IB, inferior bridging leaflet; LA, left atrium; and LV, left ventricle.
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Supplemental Material

Movie 1. RT 3D TEE movie of a "complete’ atrioventricular septal defect. The perspective is obtained from a four chamber view slightly rotated rightward. The movie shows both interatrial and interventricular defects.

Movie 2. RT 3D TEE movie of a ‘complete’ atrioventricular septal defect. The perspective is similar to a long axis view and shows the inherently narrow outflow tract.