PART A Atrial and Ventricular Septal Defects

1 Pathology of Atrial and Ventricular Septal Defects

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Introduction

Congenital deficiencies of the atrial and ventricular septa are among the most common of congenital cardiac lesions. Ventricular septal defects occur in 24–35% while atrial septal defects occur in 4–11% of liveborn babies with congenital heart disease [1]. These defects can occur in isolation, in combination, or in association with many other defects. A ventricular septal defect is an integral part of tetralogy of Fallot, double-outlet ventricles, and common arterial trunk. It is also frequently encountered in association with complete and congenitally corrected transposition, pulmonary atresia, univentricular atrioventricular connections and coarctation, or interruption, of the aortic arch. When occurring in isolation, diagnosis may be delayed, sometimes well into adult life or later decades. In this chapter, we focus on these septal defects occurring as isolated lesions and discuss some of the complicating anomalies. For a better understanding of the anatomy of atrial and ventricular septal defects, it is pertinent to begin with a review of the normal cardiac septum.

Normal septal structures

Atrial septum

A cursory look from the right atrium gives the impression of an extensive septal structure. In particular, anterosuperior to the oval fossa, the seemingly vast expanse of “atrial septum” is the right atrial wall overlying the aortic root (Fig. 1.1a). Sectional cuts demonstrate the septum limited to the floor of the foramen ovale and the muscular rim immediately around it (Fig. 1.1b). The peripheral structures are the infolded right atrial wall anterosuperiorly, superiorly, posteriorly, and inferiorly, and the fibrofatty sandwich of the atrial and ventricular musculature anteroinferiorly [2]. The superior and posterior parts of the rim, often called the “septum secundum,” are mainly the infolded right atrial wall between the base of the superior caval vein and the insertion of the right pulmonary veins to the left atrium. This infolding from the epicardial aspect is known to surgeons as “Waterston’s groove,” through which the left atrium can be accessed without entering the right atrium. Posteroinferiorly, the rim is continuous with the wall of the inferior caval vein. The true septal component is formed by the floor of the foramen ovale (“septum primum”), which functions like a flap valve by closing against the muscular rim in postnatal life when pressure in the left atrium exceeds that in the right atrium (Fig. 1.1c). In the normal heart, the valve is adequate to overlap the muscular rim so that there is no potential for interatrial shunts. The valve is completely adherent to the rim but there is an adhesion gap, or probe patency, in approximately one-fourth of the population, and this provides the potential for right-to-left shunting through the foramen ovale [3–5]. The rim is an infolding of the muscular wall of the right atrium, and the flap valve is a thin sheet of fibromuscular tissue that is usually 0.5–1.5 mm thick. Fatty tissues of the interatrial groove fill the epicardial side of the fold of the muscular rim. The extent of fatty tissue varies, and when it appears excessive in the normal heart it can give the erroneous impression of lipomatous septal hypertrophy. In young adults, the upper limit of normal fat deposit is defined as 1.5 cm in the transverse dimension on echocardiography [6].

Ventricular septum

In the majority of hearts, the right ventricle is in its anticipated location relative to the left ventricle. When the heart is seen from the front, there is considerable overlapping of the ventricular chambers. The anteriorly situated right ventricle curves over the left ventricle such that the right ventricular outflow tract passes cephalad and a little leftward, crossing over the rightward-directed left ventricular outflow tract (Fig. 1.2a and b). The ventricular septum looks very different when viewed from the right and left ventricular aspects. Significantly, the inlet part on the right side is covered over by the septal leaflet of the tricuspid valve, whereas the corresponding part on the left side borders the aortic outflow tract and is devoid of septal attachments to the

mitral valve (Fig. 1.2c and d). This is because the acute angulation between inflow and outflow tracts in the left ventricle places the outflow tract between the septum and the “anterior” leaflet of the mitral valve. The septal attachment of the mitral valve is confined to the hinge line (also known as the annulus) of its leaflets, and this is seen only in the posteroinferior parts of the left ventricle close to the cardiac crux. The simulated echocardiographic four-chamber section displays the difference in levels of attachments of the hinge lines of the mitral and tricuspid valves at the septum (Fig. 1.1b). This offset arrangement between the two valves results in a part of the muscular ventricular septum being situated between the right atrium and the left ventricle. Although previously termed the muscular atrioventricular septum, its composition is a sandwich of right atrial wall on one side, crest of the muscular ventricular septum on the other, with intervening fibrofatty tissue from the inferior atrioventricular groove, which ingresses from the epicardium at the crux of the heart. Adjourning the “sandwich” anterosuperiorly is the central fibrous body together with the membranous component of the cardiac septum. The central fibrous body contains the penetrating bundle of His. Its continuation, the atrioventricular conduction bundle, is sandwiched between the crest of the muscular septum and the membranous septum (Fig. 1.3a). This feature is particularly relevant when considering holes in the vicinity of the membranous septum (Fig. 1.3b). The hinge line of the tricuspid valve crosses the membranous septum, effectively dividing it into atrioventricular and interventricular components (Fig. 1.3a). Viewed from the left ventricular aspect, the membranous septum is adjacent to the aortic valve. It adjoins the interleaflet fibrous triangle that lies between the right and the noncoronary leaflets. Thus, the landmark for the course of the atrioventricular conduction bundle is the septal area between the
right and noncoronary aortic sinuses. From there, the atrioventricular bundle branches into the right and left bundle branches (Fig. 1.3a). The cord-like right bundle branch passes through the muscular part of the septum to emerge subendocardially close to the insertion of the medial papillary muscle of the tricuspid valve. On the left side of the septum, the left bundle branch descends in the subendocardium to branch into three main radiating and interconnecting fascicles. The branching bundle and the proximal portion of the left bundle branch are, therefore, closely related to the septal aspect of the outflow tract immediately beneath the aortic valve (Fig. 1.3b).

Anterosuperior to the membranous septum is the pulmonary outflow tract exiting from the right ventricle. The musculature anterior to the membranous septum is the supraventricular crest, comprising the ventriculoinfundibular fold and its insertion into the septomarginal trabeculation at the septum (Fig. 1.2c). The septomarginal trabeculation is a characteristic muscle band looking like a tree trunk flattened against the ventricular septum in the right ventricle. It branches into two limbs that cradle the ventriculoinfundibular fold. One limb points anterosuperiorly to blend into the musculature of the subpulmonary infundibulum. The other limb points posteroinferiorly, and it is from this limb that the medial papillary muscle (also known as the conal muscle or muscle of Lancisi) arises to support the anteroseptal commissure of the tricuspid valve. The distal part of the septomarginal trabeculation extends into the moderator band that crosses the right ventricular cavity. The apical portion of the right ventricle bears coarse trabeculations that can obscure the presence of muscular septal defects.

**Figure 1.2** (a) This endocast of a normal heart viewed from the front shows the right ventricular outflow tract (solid arrow) crossing over the left ventricular outflow tract (broken arrow). (Note that the pulmonary valve is at a higher level than the aortic valve.) (b) The endocast viewed from the left side shows the relationship of the left ventricular outflow tract (broken arrow) to that of the right. The right ventricle is opened to show the septum and the three portions of the ventricle. (c) The limbs (arrows) of the septomarginal trabeculation (SMT) cradle the ventriculoinfundibular fold (VIF). (d) Dissection into the left ventricular outflow tract shows the proximity of the inlet and outlet portions. The asterisk marks the membranous septum. Ao, aorta; LA, left atrium; LV, left ventricle; PT, pulmonary trunk; RV, right ventricle.
In the outflow portion of the right ventricle, the conical sleeve of subpulmonary infundibulum is continuous with the ventriculoinfundibular fold. The infundibulum elevates the pulmonary valve away from the septum. Thus, in the normal heart, it is not possible to define a discrete muscular structure equivalent to an outlet septum separating aortic and pulmonary outflow tracts. The outlet septum is expressed in malformations of the outflow tract such as in hearts with Fallot’s tetralogy, in which it is an exclusively right ventricular structure.

For convenience in describing the location of holes in the ventricular septum, the right ventricle can be considered as having three portions, although there are no anatomic lines that would allow division of the muscular septum into three parts (Fig. 1.2c). Thus, the inlet portion of the right ventricle is taken to be the portion receiving the tricuspid valve while the outlet portion is the part leading to the semilunar valves, and is mainly musculature proximal to the subpulmonary infundibulum. The remaining portion, the trabecular portion, is the most extensive. Owing to the configuration of the left ventricular inflow and outflow tracts, with the aortic outlet wedged between the septum and the mitral valve, much of the inlet portion of the right ventricle overlaps the outlet portion of the left ventricle (Fig. 1.2d).

**Atrial septal defects**

Although generally categorized as atrial septal defects (ASDs), some of the defects commonly referred to as ASDs are interatrial communications rather than deficiencies of the atrial septum. This is because the extent of the atrial septum is rather limited when a septum is defined as a partition that separates two adjacent chambers, and passage through the septum will not exit the heart (see Atrial septum). Strictly speaking, the septum that separates the two atrial chambers is the valve that is circumscribed by the muscular rim of the foramen ovale (Fig. 1.1a). Defects within this area, usually termed “secundum defects,” are true atrial septal defects (Fig. 1.4a). By contrast, sinus venosus defects, coronary sinus defects, and “ostium primum” defects are outside the confines of the true atrial septum although, unequivocally, they permit interatrial shunting (Fig. 1.4a) [3].

**Patent foramen ovale**

As discussed above, the patent foramen ovale (PFO) is very common. It exists because of an incomplete circumferential adhesion of the septum primum (the flap valve of the foramen) to the septum secundum (the rim of the fossa). The gap, the last
Figure 1.4 Locations of various interatrial communications in the right atrium. (a) The oval fossa defect (1) is the true atrial septal defect. The superior (2) and inferior (3) sinus venosus defects are related to the entrances of the corresponding caval veins. The coronary sinus defect (4) is at the site of the coronary sinus orifice. The atrial component of the atrioventricular septal defect (broken line) is indicated by the number 5. The orange shapes represent the sinus and atrioventricular nodes. (b) The patent foramen ovale (arrow) lies at the anterocephalad margin of the oval fossa. (c) Cut through the atria viewed from behind shows the infolded right atrial wall (asterisk) forming the right margin of the tunnel-like patent foramen ovale (arrow). (Note the aortic root emerging immediately anteriorly.) (d) Long-axis cut through the left heart shows the proximity of the exit (arrow) of the patent foramen ovale to the anterior wall of the left atrium. This part of the atrial wall can be exceedingly thin and it borders the transverse pericardial sinus (triangle) and the noncoronary aortic sinus (N). (e) Right atrial view shows an oval-shaped defect in the atrial septum owing to deficiency of the flap valve. (f) The valve of the oval fossa is lacking and only a few strands remain in the fossa. Ao, aorta; cs, coronary sinus; ICV, inferior caval vein; LV, left ventricle; MV, mitral valve; PT, pulmonary trunk; R, right coronary aortic sinus; SCV, superior caval vein; TV, tricuspid valve.

part of the valve to become adherent, is located at the anterocephalad margin of the rim (if viewing the right atrium in a simulated right anterior oblique projection; Fig. 1.4b). The adhesion gap leaves a slit-like tunnel that allows a probe to be passed obliquely from the right atrium into the left atrium in approximately 25% of cadaver hearts. The length of the tunnel depends on the extent of overlap between the flap valve and the rim [7,8]. In the left atrium the exit site of the probe is at the crescentic margin of the flap valve, and this is closely related to the anterior wall of the left atrium (Fig. 1.4c). This part of the wall can be exceedingly thin, and perforations can lead to the transverse pericardial sinus and the aortic root (Fig. 1.4c and d).

Morphologically, there are two forms of PFO [7]. The first is the valve-competent form in which, under normal circumstances, the valve is large enough to overlap the muscular rim, much like a door closing against a door frame. Although forming a perfect seal, some of these valves are aneurysmal in appearance and bow into the right and left atrial chambers with the respiratory phases. The second form is the
Defects within the oval fossa

Usually these so-called “secundum” defects are located at the site of the embryonic “ostium secundum” rather than a deficiency of the “septum secundum” since the septum secundum is largely the infolded right atrial wall. Deficiencies, perforations, or complete absence of the valve guarding the foramen ovale (the embryonic “septum primum”) are the most common types of interatrial communications with a spectrum of sizes. The simplest form is one resulting from the valve being too small to overlap the muscular rim and so leaving an oval-shaped aperture between the rim and the edge of the valve (Fig. 1.4e). This form is most amenable to transcatheter repair providing there are adequate muscular borders without impinging upon the orifices of the pulmonary veins, the atrioventricular valves, the caval veins, or the coronary sinus [9–11]. Even so, the location of the valve is variable [9]. In some cases, it may be more anteriorly situated or more posteriorly situated. The valve itself may be perforated with single or multiple fenestrations. Sometimes, it appears like a net or is represented by a filigreed remnant (Fig. 1.4f).

When the valve is completely absent, or nearly completely absent, the defect is the hole surrounded by the muscular rim of the fossa. If the anterior rim is deficient, it is worth bearing in mind the proximity of the anterior margin to the transverse sinus and the aortic root. The right coronary and noncoronary aortic sinuses are in the immediate neighborhood. A deficient posterior rim reduces the distance to the orifices of the right pulmonary veins and also increases the proximity to the epicardium due to effacement of the infolding. Occasionally, the defect may extend toward the inferior caval vein or toward the atrioventricular junction. In the case of the latter situation, the distance of the defect from the annular attachment of the mitral valve may become reduced, increasing the risk of damaging the mitral valve during device closure. Although defects in the oval fossa do not alter the basic disposition of the sinus and atrioventricular nodes of the conduction system, these very large defects will reduce the distances between the margin of the defects and the atrioventricular node or the orifice of the coronary sinus.

In cases associated with persistent left superior caval vein draining into the coronary sinus, the coronary sinus is usually enlarged. In these cases, the muscular margin between the coronary sinus orifice and the defect needs to be evaluated carefully. Cases of successful device closure without obstructing coronary venous return have been reported [11].

Sinus venosus defects

These defects are usually located in the mouth of the superior caval vein and described as superior sinus venosus defects. The inferior sinus venosus defects are related to the inferior caval vein and are far less common. The key feature of sinus venosus defects is that they exist outside the confines of the true atrial septum (see Atrial septum). This is not to say that they cannot become confluent, or coexist, with deficiency of the oval fossa.

In the case of a superior sinus venosus defect, the mouth of the superior caval vein typically overrides the atrial septum above the superior rim of the oval fossa (Fig. 1.4a) [12]. Anomalous insertion of the right pulmonary veins into the wall of the superior caval vein is usual in this situation. The defect, therefore, has a well-defined inferior border, the superior rim of the oval fossa, which encloses epicardial fat. Roofing the defect is the overriding caval vein. Owing to the lack of a superior rim for anchorage, currently available devices for closing atrial septal defects are unsuitable. Surgical repair of this defect should take account of potential obstruction to the superior caval pathway following restoration of pulmonary venous return to the left atrium. Also at risk is the sinus node and its arterial supply should there be the need to widen the cavoatrial junction [13].

Sinus venosus defects related to the mouth of the inferior caval vein have similar features to those of superior sinus venosus defects. In the inferior position, the defect’s roof is delineated by the posteroinferior rim of the oval fossa and the orifice of the inferior caval vein opens to both left and right atria (Fig. 1.4a). The lower right pulmonary vein can attach anomalously to the wall of the inferior caval vein. This type of defect is remote from the anticipated locations of the sinus and atrioventricular nodes but it lacks an inferior rim for device anchorage.

Coronary sinus defects

Defects termed “coronary sinus defects” cover a spectrum ranging from a hole at the site of the orifice of the coronary sinus and absence of the coronary sinus itself, to a single or multiple fenestrations along the course of the coronary sinus, allowing it to communicate directly with the left atrium. Absence of the wall of the coronary sinus together with the adjoining portion of the left atrial wall results in the deficiency described as unroofing of the coronary sinus [14]. The defect usually leaves the persistent left superior caval vein connecting directly to the left atrium. When existing as a hole at the orifice of the coronary sinus, it may be amenable to device closure [15]. However, closing a large defect at the site of the orifice of the coronary sinus may jeopardize the atrioventricular node because the triangle of Koch becomes foreshortened.

“Ostium primum” defects

This type of defect, although producing an interatrial shunt, is not a true atrial septal defect (Fig. 1.4a). Hearts with
in isolation as most require little if any attention. A major determinant of outcome is the size of the defect. The majority become proportionally smaller with time. Spontaneous closure of the defect occurs in up to half of cases recognized in childhood [17,18], and it has been suggested may also occur in adult life. Generally, those who are asymptomatic are likely to have small defects.

This review of the morphology is restricted to isolated ventricular septal defects. For reasons already discussed, the ventricular component of atrioventricular septal defects will not be included. Also excluded are septal defects following myocardial infarction.

**Description of ventricular septal defects**

Over the decades, there have been many classifications of ventricular septal defects. In more recent decades, however, three main categories of ventricular septal defects are recognized: *perimembranous* (or membranous/infracristal), *muscular* (or trabecular), and *doubly committed and juxtaarterial* (or infundibular/supracristal/subpulmonary) (Fig. 1.5). These descriptions are applicable to defects existing in isolation as well as in association with other malformations. The distinction between perimembranous and muscular septal defects highlights the relationship of the defect’s margins to the atrioventricular conduction system [19].

The location of any hole in the septum between ventricles can be described relative to the three portions of the normal right ventricle, i.e., inlet, apical trabecular, and outlet, with the approach by the surgeon usually from the right side of the heart. Furthermore, the size of the defect and any associated misalignment of septal structures need to be considered in any treatment strategy.

**Muscular defects**

The muscular defect is characterized by having completely muscular borders. Reportedly, it accounts for 5% of all ventricular septal defects, but its true incidence may be considerably higher as small muscular defects tend to close spontaneously. They can be described as being located in the inlet, outlet, or apical trabecular portions of the right ventricle. Muscular defects, especially those in the apical portion, may be multiple, giving the septum a Swiss cheese appearance, but these are rare. The thick right ventricular trabeculations overlying the septum may make it difficult to visualize or approach these defects from the right side. Some defects appear very small on the right ventricular side but actually form a large confluent defect when examined from the left side (Fig. 1.6a and b).

Muscular defects located in the inlet portion may be partially hidden by the septal leaflet of the tricuspid valve. A rim of muscle separates the border of the defect from the hinge of the tricuspid valve, distinguishing it from a perimembranous defect located in the inlet portion (Fig. 1.5). By virtue of its location, the atrioventricular conduction bundle is related to

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**Figure 1.5** Three types of ventricular septal defect as seen from the right ventricle. The atrioventricular conduction bundle (red line) is shown skirting close to the posteroinferior margin of the perimembranous defect (1). By contrast, the bundle is related to the superior margin of a muscular inlet defect (2). Muscular defects can be located anywhere in the muscular septum and are represented by the yellow ovals. The doubly committed and juxtaarterial defect (3) are intimately related to the arterial valves.

**Ventricular septal defects**

This is one of the most common congenital heart malformations. The incidence is much higher when the defect exists...
the superior margin of the muscular defect. This is in distinct contrast to perimembranous defects (see below). When a muscular inlet defect co-exists with a perimembranous defect, the atroventricular conduction bundle runs in the muscle bar separating the two defects (Fig. 1.5).

Muscular defects high in the outlet portion are very rare. Seemingly safe from the pulmonary valve, they may be sited close to the aortic valve on the left ventricular side. When the muscular rim in the superior border is narrow, such defects may be difficult to distinguish from doubly committed and juxtaarterial defects (see below).

The majority of muscular defects are not round [20]. They range from being slit-like to the more usual oval shape or D shape (Fig. 1.6a–c). Furthermore, the defects tend to “burrow” through the septum such that their opening on the right side of the septum is offset from that on the left side. In some
cases the opening on one side is larger than on the other side and in others the hole is crossed by muscle bars on one side (Fig. 1.6a and b).

**Perimembranous defects**

This is the most common type of ventricular septal defect. Being in the environs of the membranous septum, it is related to several important structures. As these defects usually involve a more extensive area than that occupied by the normal membranous septum, they are best described as perimembranous. The key feature of perimembranous defects in all hearts with concordant atrioventricular connections is either a remnant of the membranous septum or fibrous continuity between the atrioventricular valves at the posteroinferior border (Fig. 1.6d and e). It is at this part of the border that the atrioventricular conduction bundle emerges from the central fibrous body to become subendocardial and, in most cases, the branching portion of the bundle appears on the left side of the septal crest.

Perimembranous defects vary in shapes and sizes and can extend toward the inlet, outlet, or apical trabecular portions. Large defects have been described as confluent. In our pathological series [20], the majority of perimembranous defects were oval or round in shape when the septum was viewed en face from the right side. When located in the inlet, they are partially, or even entirely, covered over by the septal leaflet of the tricuspid valve (Fig. 1.6d and e). Cords tethering the leaflet usually cross the defect on the right ventricular aspect but in some cases are inserted to the septal crest or to the left side of the septum. In this location, the defect tends to be oval shaped with its long axis parallel to the valvar hinge line. The tricuspid and mitral valves lose their “offset” relationships and their hinge lines form the “roof” of the defect in long-axis echocardiographic sections. Owing to there being a defect, the atrioventricular conduction bundle is displaced more posteroinferiorly than normal, and it is still related to the area of fibrous valvar continuity in the posteroinferior margin of the defect. The medial papillary muscle is located in its antero-superior border. Perimembranous inlet defects have been described as atrioventricular canal type defects [21] but differ in not having the hallmark of a common atrioventricular junction that characterizes atrioventricular septal defects [16].

Perimembranous defects that excavate toward the outlet portion are close to the semilunar valves. Although variable in shape, they tend to be more circular than the other forms of perimembranous defects. In hearts with normal arterial relationships, the aortic valve minimally overrides the septal crest. Aortic, mitral, and tricuspid valvar continuity forms the fibrous posteroinferior border (Fig. 1.3b). Again, it is this fibrous border that harbors the atrioventricular conduction bundle. The medial papillary muscle supporting the tricuspid valve is close to this quadrant of the hole. The distance of the rim of the defect from the conduction bundle depends on the size of the remnant of the membranous septum.

Some perimembranous defects excavate toward the trabecular portion. These tend to be oval or triangular with their long axis directed toward the cardiac apex. In these, the medial papillary muscle is located at the apical quadrant of the defect. Again, the atrioventricular conduction bundle runs in the posteroinferior border. On the left ventricular side of the septum there may be a rim of muscle between the defect and the aortic valve.

Tissue tags adjacent to perimembranous defects can be accessory tissues from the tricuspid valve or the membranous septum. Also described as ventricular septal aneurysms, these are involved in spontaneous closure or diminution in size of the defects [22]. True membranous septal defects are very rare and they are small. Even rarer are defects (described as Gerbode defects) that arise owing to the absence of the atrioventricular component of the membranous septum, resulting in shunting from the left ventricle to the right atrium. Shunts at this level are more often a result of perimembranous defects that are associated with a deficiency in the septal leaflet of the tricuspid valve.

**Doubly committed and juxtaarterial defects**

These defects account for 5–10% of ventricular septal defects and are more commonly found in the Orient and in Latin America. The feature that characterizes doubly committed and juxtaarterial defects is the lack of muscular separation between the arterial valves in the superior borders. These defects are roofed by the pulmonary and aortic valves. Only a fibrous raphe runs between the adjoining valvar leaflets. In some cases, the right coronary leaflet of the aortic valve prolapses into the defect. In many, the posteroinferior margin of doubly committed and juxtaarterial defects is muscular, owing to the fusion between the inferior limb of the septomarginal trabeculation and the ventriculoinfundibular fold. In others, the posteroinferior margin extends to the remnant of the membranous septum and becomes perimembranous. By the nature of their immediate proximity to the semilunar valves, these types of defects are unsuitable for closure using currently available devices. There is hardly any superior margin for safe anchorage without interfering with mobility of the semilunar valves or creating outflow obstruction.

**Atrioventricular septal defect**

As discussed above, the so-called primum ASD form is unlikely to be suitable for device closure. The form with a common valvar orifice (so-called complete form or atrioventricular canal defect) with both atrial and ventricular defects is also unsuitable. The bridging leaflets of the atrioventricular valve can be compromised. The rarest form that has only a ventricular component of the defect, when carefully selected, may be amenable to device closure. A 4-year-old patient underwent successful implantation of a device with good outcome (Dr. Michael Rigby, personal communication, 2007).
In this child, there was accessory valvar tissue at the margin of the defect.

**Malalignment of septal structures**

Descriptions of ventricular septal defects are not complete without considering whether the septal components are aligned. Malalignment between atrial and ventricular septa or between components of the muscular ventricular septum have important consequences on the structures in the vicinity of the septal defect. Perhaps the best-known situation of septal malalignment is anterocephalad deviation of the outlet septum in hearts with Fallot’s tetralogy. In Fallot, the outlet septum is entirely in the right ventricle and its malalignment produces overriding of the aortic valve as well as subpulmonary stenosis. In contrast, malalignment of the outlet septum into the left ventricular outflow is associated with obstructive lesions of the aortic arch.

Malalignment between atrial and ventricular septa is exemplified by cases with straddling and overriding of the tricuspid valve. Whether existing with isolated ventricular septal defects or with other intracardiac defects, the cardinal feature is that the muscular septum does not extend to the crux of the heart but inserts to the right of the crux. The malalignment results in an abnormally located atrioventricular conduction axis with the atrioventricular node situated in the posterolateral margin of the tricuspid orifice and the bundle penetrating at the point at which the ventricular septum meets the right atrioventricular junction.

**Conclusions**

A good understanding of the morphological substrates and variations of atrial and ventricular septal defects is particularly relevant in the era of interventional cardiology [10]. An appreciation of the limited extent of the atrial septum helps in distinguishing true atrial septal defects from all other forms of interatrial communications, and also in identifying the ideal site for trans-septal puncture. Interaltral communications that are suitable for transcatheter device closure must be sufficiently remote from the atrioventricular valves, coronary sinus, pulmonary and caval veins, and the aortic root. To minimize the risk of complications, some of these defects, either because of their very large size, or because of their close relationship to these vital structures, require very careful evaluation before considering transcatheter device closure.

Description of ventricular septal defects need not rely on developmental concepts. The categorization described above draws attention to the location of the defect and the proximity of the defect margins to crucial cardiac structures. When considering transcatheter device closure of perimembranous ventricular septal defects, the size of the defect relative to the heart size is important on account of the locations of the aortic valve and the atrioventricular conduction bundle in the immediate vicinity. Ventricular septal defects roofed by the arterial valves and those associated with malalignment of the septal components are unlikely to be suitable for percutaneous device closure without risking damage to adjacent valvar structures.

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**References**