21 Echocardiography in the Assessment of Atrial Septal Defects

Edmund A. Bermudez, MD, MPH

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INTRODUCTION

Historically, the evaluation of patients with suspected congenital heart disease relied heavily on the results of the physical exam and cardiac catheterization. In the current era, the initial assessment of congenital lesions can now be safely and easily accomplished with cardiac ultrasound, often without the need for further invasive testing. One of the most frequently encountered acyanotic congenital lesions in clinical practice is the atrial septal defect (ASD), of which echocardiography provides invaluable information in the initial assessment, follow-up, and management. The spatial resolution of echocardiography allows for accurate classification of ASDs and a comprehensive evaluation of associated cardiac pathology. A quantification of shunt flow and hemodynamics can be easily accomplished with Doppler, and provide an objective means of follow up for these patients. Finally, echocardiography can be used to guide percutaneous closure of ASDs, providing an important avenue for minimally invasive intervention. Thus, echocardiography is currently the basis for the initial diagnosis, follow-up, and when appropriate, management of ASDs.

ANATOMIC AND ECHOCARDIOGRAPHIC OVERVIEW

During embryological development, the right and left cardiac atria are formed with the growth of the atrial septum. This dividing wall between the atria originates with the growth of two separate septa: septum primum and septum secundum. The first to be formed is the septum primum, which grows from the superior aspect of the atria wall inferiorly, toward the enodcardial cushions between the atria and ventricles (Fig. 1A). In its normal development, the septum primum attaches to the endocardial cushions, with eventual resorption of the superior attachment. Concurrently, a second septum develops slightly to the right of the septum primum, with progressive growth from the superior aspect of the atrium toward the endocardial cushions, but does not reach or attach to this area. Therefore, when developed, the septum primum attaches inferiorly to the floor of the atrial cavity and the septum secundum superiorly to the roof the atria. Together, these two septa overlap to form the basis of the interatrial septum, between which the foramen ovale is formed (Fig. 1B). The foramen ovale is patent during development but normally closes

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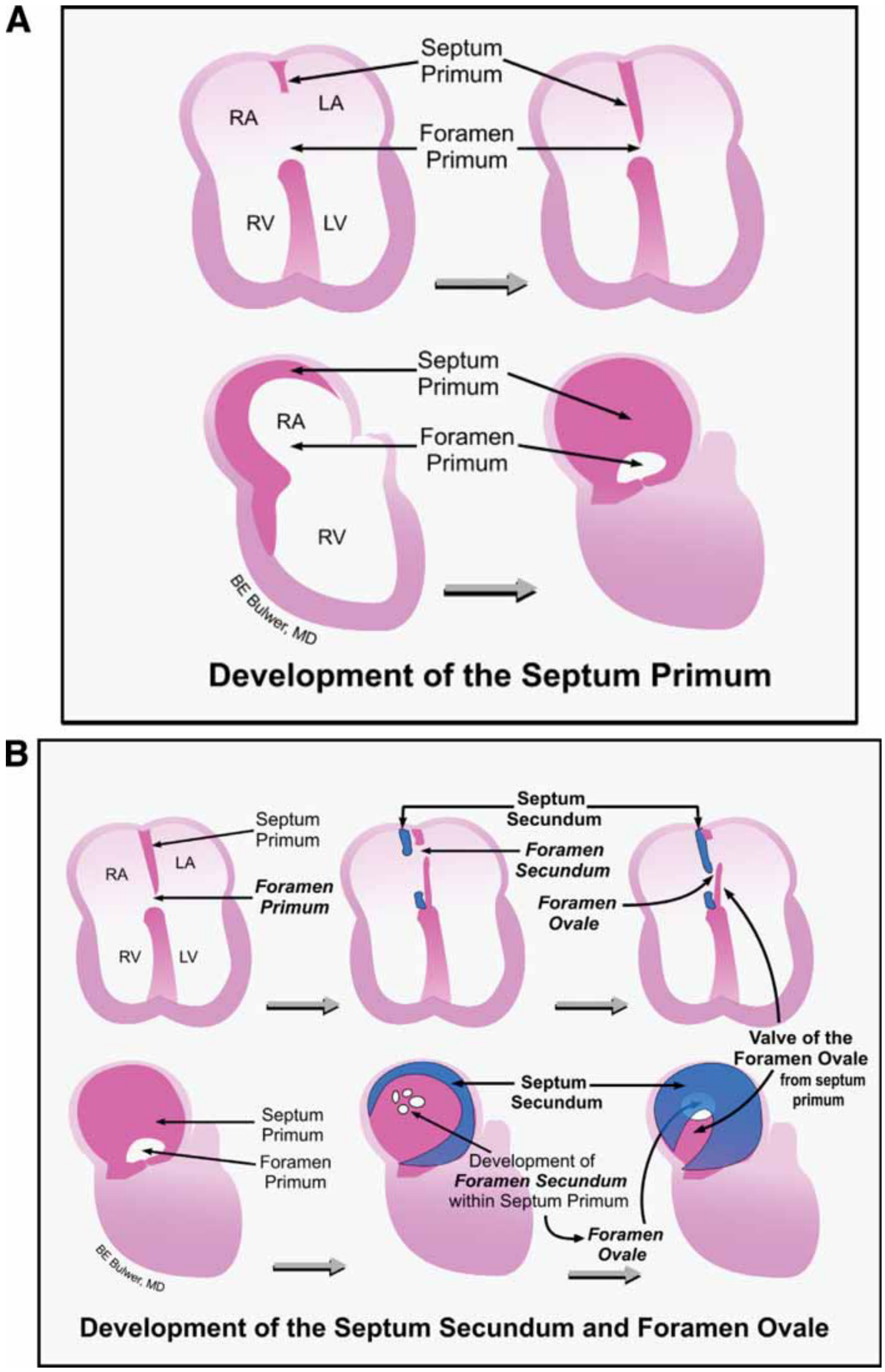

Fig. 1. (A) The first step in the formation of the interatrial septum is the growth of the primum septum. **(B)** Partial resorption of the primum septum leaves fenestrations that coalesce in the formation of a second interatrial connection—the ostium secundum. This occurs concurrently with the descent of a second septum—the septum secundum—to the right of the septum primum. Both septae fuse except in the region called the foramen ovale which permits oxygenated blood to bypass the fetal lungs and hence to the fetal systemic circulation.

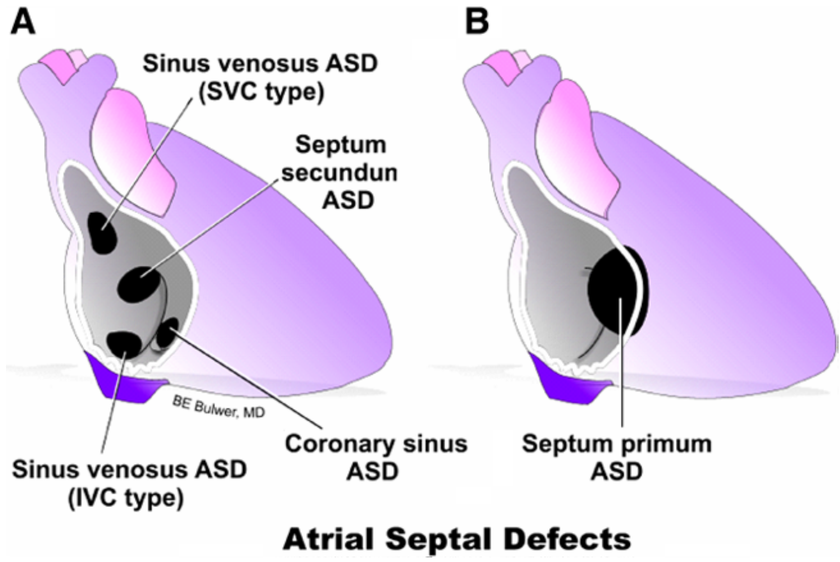

Fig. 2. The anatomic distribution of atrial septal defects. Four major types are described as shown.

shortly after birth when left-sided pressures exceed right and seal the two septa together.

ASDs form when the interplay of septal formation is deranged. Several forms of interatrial communication can therefore result. Four major ASDs can be classified and correspond to the anatomic region where the defect arises: secundum defect, primum defect, sinus venosus defect, and coronary sinus defect (Fig. 2). Occasionally, when the septum primum and secundum do not fuse after birth, a persistent orifice may be present, the so-called patent foramen ovale (PFO). This foramen can be physiologically patent with interatrial shunting in the direction of higher to lower pressure. Each of these congenital lesions of the atrial septum can be easily characterized by echocardiographic measures.

Transthoracic echocardiography is useful in the diagnosis and assessment of ASDs. Three main transthoracic views can be used to assess for ASDs in the majority of cases: (1) parasternal short axis at the level of the aortic valve, (2) apical four-chamber view, and (3) subcostal four-chamber view. In the apical four-chamber view, the atrial septum is a relatively deep structure and susceptible to echo signal dropout. Echolucency of this region should therefore be interpreted with caution without confirmation in other views or with color Doppler. Of these views, the subcostal four-chamber view may be most suited for the two-dimensional assessment of ASDs, as the ultrasound beam is most perpendicular to the septa in these views.

Table 1
ASD Types and Associated Cardiac Anomalies

Type	*Associated anomalies*
Septum secundum	Atrial septal aneurysm
Septum primum	Cleft mitral valve
Sinus venosus	Anomalous pulmonary venous drainage
Coronary sinus	Persistent left superior vena cava

Associated anomalies are not mutually exclusive.

Typically, secundum and primum septal defects can be seen with transthoracic imaging. However, multiplane transesophageal echocardiography may be needed to visualize sinus venosus or coronary sinus defects with certainty. Multiplane transesophageal imaging is highly accurate in visualizing the anatomical relationships and associated anomalies sometimes seen with ASDs (Table 1).

The use of color and spectral Doppler can enhance the detection of these defects. Color flow Doppler can confirm the presence and direction of interatrial shunting across visualized aspects of the atrial septum (Fig. 3; please *see* companion DVD for corresponding video). The color flow signal may also aid in measuring the size and number of defects. For example, should several color flow jets be seen across the area of the septum, a fenestrated defect is implied. Thus, color should be used in different views in order to assess for these flow abnormalities.

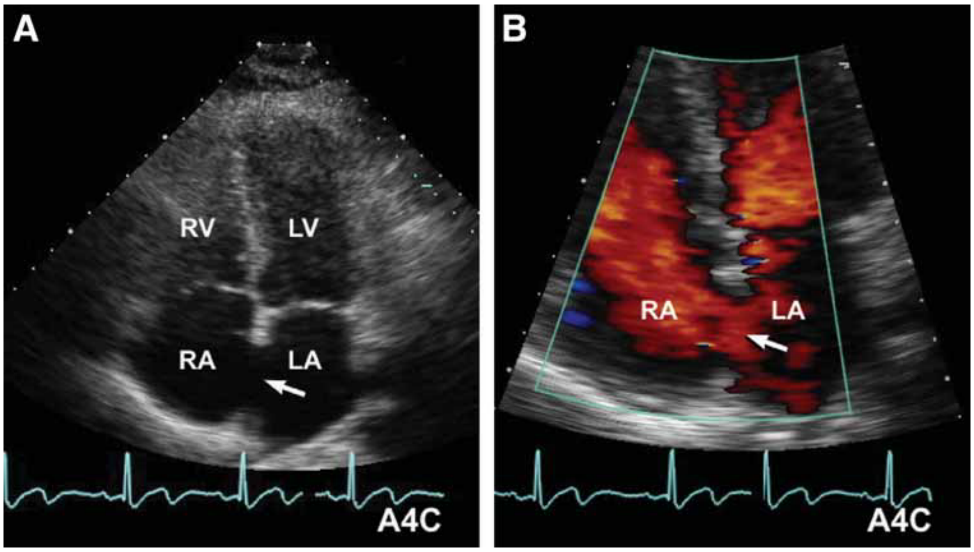

Fig. 3. Atrial four-chamber view showing defect or possible drop out in the interatrial septum (arrow, **A**). Color flow Doppler interrogation showed a typical pattern of predominant left-to-right shunting as seen with secundum atrial septal defects **(B)**. (Please *see* companion DVD for corresponding video.)

Should the result of a color and spectral Doppler exam be equivocal, an intravenous agitated contrast saline injection can be used to detect interatrial shunting. This effect is dependent on the fact that the bubbles created from the agitated saline are filtered in the pulmonary vasculature and do not cross to the left heart. Thus, contrast should only be seen in the right heart chambers in the absence of intracardiac and intrapulmonary shunting. In the typical case of an uncomplicated ASD, left atrial pressures are higher than right atrial pressures and direct predominant left-to-right shunting with occasional early crossing of contrast bubbles to the left heart from right-to-left shunting. Contrast is injected from an upper extremity and enters the right atrium via the superior vena cava (SVC) and tends to be directed more toward the tricuspid valve than the atrial septum, in contrast to blood from the inferior vena cava (IVC), which is usually directed more toward the atrial septum via the eustachian valve. An adequate contrast injection is attained when the bubble contrast is seen to appose the area of the septum. Valsalva maneuver is often used to assess for interatrial shunting with contrast by accentuating right-to-left shunting. The presence of contrast bubbles in the left heart early after injection signifies an interatrial shunt, whereas the appearance of late contrast bubbles (>3–5 cardiac cycles) in the left heart suggests an intrapulmonary shunt (Fig. 4; please *see* companion DVD for corresponding video).

Typically, ASDs confer a volume load on the right heart. Therefore, a pattern of right ventricular overload is often seen. The right ventricle is often dilated with preserved systolic function. The right atrium is often enlarged. Because of the volume load, ventricular septal motion may be abnormal and display paradoxical motion. The pulmonary pressures will vary depending on the hemodynamic impact of the lesion. In the extreme and rare case, Eisenmenger's physiology may result, with pulmonic pressures equal to or higher than systemic blood pressure.

Quantification of shunt flow can be accomplished with a comparison of pulmonary blood flow to systemic blood flow. The Qp/Qs ratio has been used to measure the hemodynamic impact ASDs. This result is found by calculating flow across the pulmonary valve and dividing this by flow across the aortic valve (Figs. 5 and 6). Flow is calculated by multiplying the cross-sectional area of the outflow tract with the time-velocity integral (TVI). Therefore,

$$Q_P / Q_S = \frac{\pi\,(\text{radius of RVOT})^2 \times \text{TVI}_{\text{RVOT}}}{\pi\,(\text{radius of LVOT})^2 \times \text{TVI}_{\text{LVOT}}}$$

Accurate Qp/Qs calculations necessitate precise measurements, particularly pertaining to the size of the valvular annuli, and are valid in the absence of significant regurgitant or stenotic lesions of the semilunar valves.

SECUNDUM ASD

Secundum ASDs are one of the most frequently encountered congenital cardiac defects in clinical practice

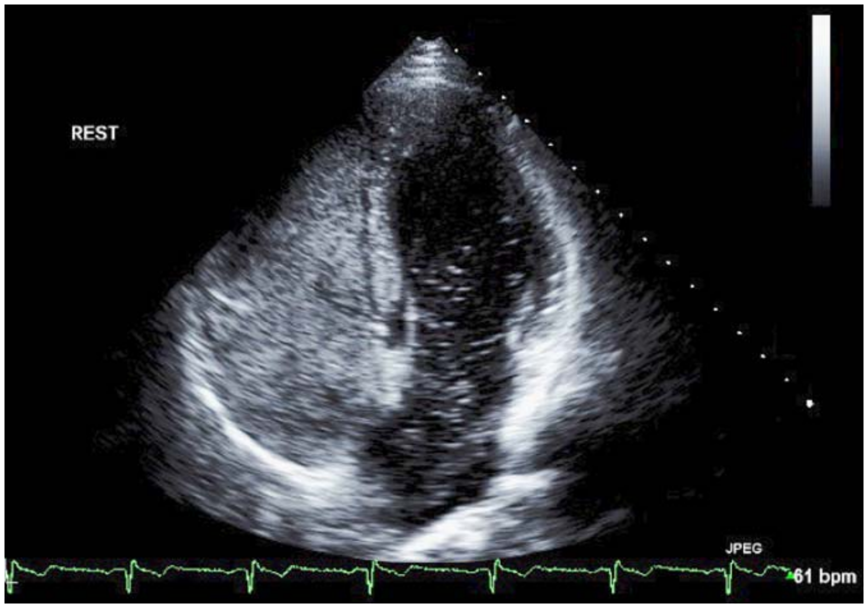

Fig. 4. Agitated saline contrast echocardiography in this 28-yr-old female who complained of migraine headaches shows good opacification of right heart chambers. Clear evidence of right-to-left shunting (appearance of saline bubbles into left heart chambers) occurred early during rest. She was diagnosed with a secundum atrial septal defect. (Please *see* companion DVD for corresponding video.)

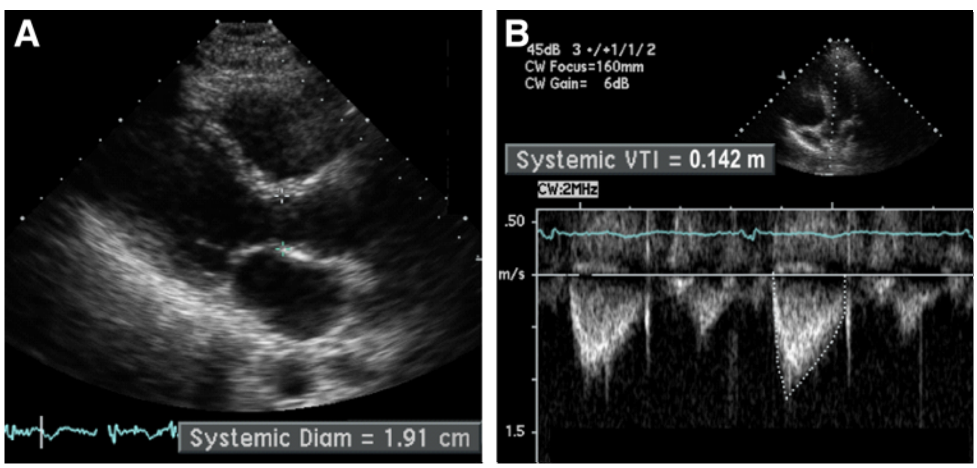

Fig. 5. Quantification of shunt flow (Qp:Qs) requires measurement (*see* Anatomic and Echocardiographic Overview section). Systemic flow calculation requires measurement of left ventricular outflow tract diameter **(A)** and the velocity time integral (VTI) by PW Doppler **(B)**.

along with bicuspid aortic valves. The lesion results from the incomplete development of the septum secundum, leaving a central patency between the right and left atria (Fig. 7; please *see* companion DVD for corresponding video). Occasionally, there may be more than one opening in the septum so that a fenestrated septum results. This is often clearly delineated on transthoracic imaging with the use of color flow Doppler around the area of the central portion of the septum (Fig. 8). Shunt flow can be seen with the use of pulse wave Doppler across the defect. Usually some transient right-to-left shunting can be seen with these defects, typically corresponding with ventricular contraction. Occasionally, with the use of intravenous agitated saline in the right heart, an echolucent silhouette of blood flow may be seen in the right atrium when predominant left-to-right shunting is present, the so-called "negative contrast effect." However, because saline contrast is often injected from an upper extremity, this "negative contrast effect" should be interpreted with caution as this can also be produced by contrast-free caval blood flow emanating

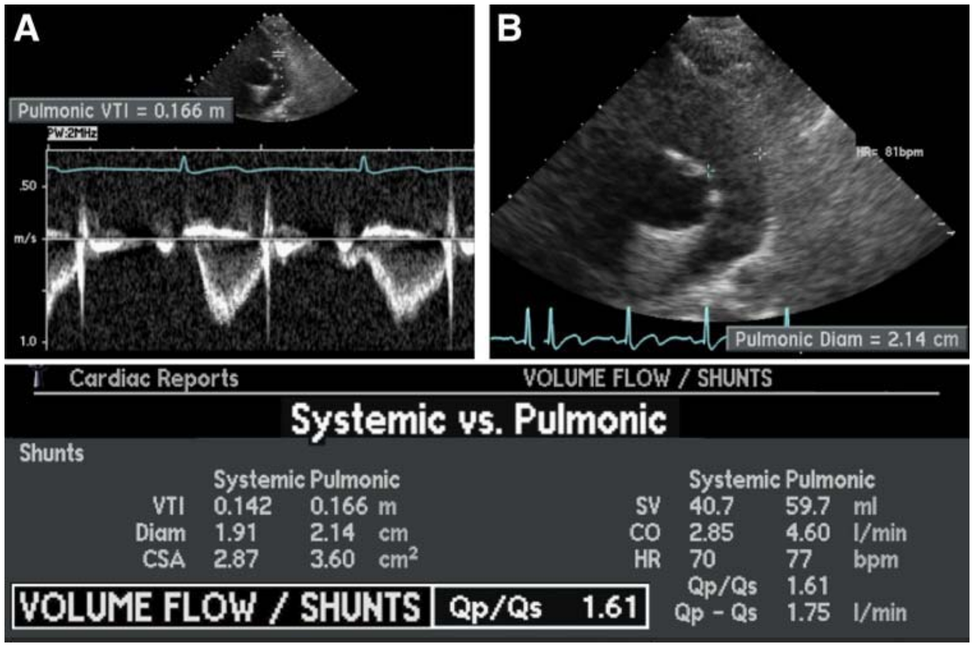

Fig. 6. Pulmonary flow is similarly calculated following measurement of the right ventricular outflow tract at the level of the pulmonary valve **(B)**.

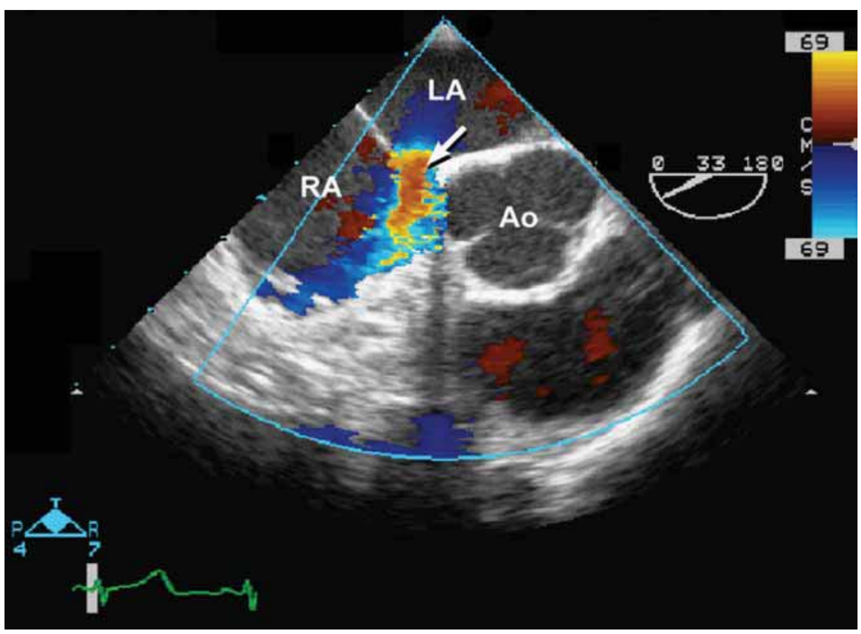

Fig. 7. Midesophageal view of the interatrial septum with color Doppler applied shows left-to-right shunt via a secundum atrial septal defect. (Please *see* companion DVD for corresponding video.)

from the IVC flushing against the area of the fossa ovalis.

Secundum ASDs may be associated with other cardiac anomalies. Occasionally, atrial septal aneurysms may be seen with secundum ASDs. In these cases, the atrial septum is hypermobile and bows into either atrial chamber, varying with respiratory variation. When rheumatic involvement of the mitral valve is concomitantly present with an ASD, Lutembacher's syndrome is said to be present.

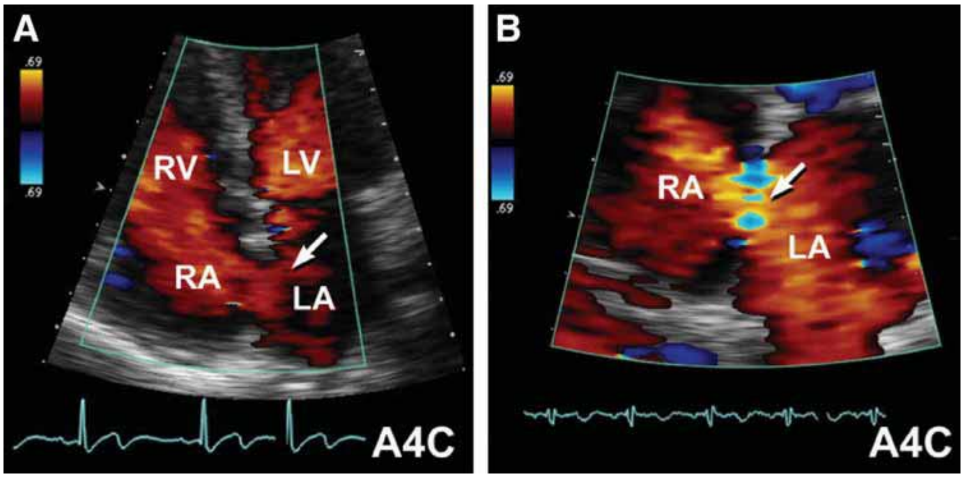

Fig. 8. Transthoracic apical four-chamber view (A4C) with color flow Doppler interrogation of the interatrial septum shows left-to-right shunt via a secundum defect.

PRIMUM ASDs

Primum ASDs result from the incomplete development of the septum primum with the endocardial cushions in the region of the atrioventricular (AV) valves. Also called partial AV canal defects, this lesion results in an atrial communication that is seen inferiorly, in the lower portion of the atrial septum that normally is fused with the AV valvular apparatus. The AV valves are therefore affected, typically involving the mitral valve, where a cleft may be seen in the anterior leaflet. Mitral regurgitation may also be seen. Interventricular shunts are not generally seen with this abnormality.

The features of this defect can usually been seen with transthoracic imaging. In the four-chamber view (apical or subcostal), the AV valves may appear to lie on the same plane, forming a "T," referring to the alignment of the AV valves with the ventricular septum. An echolucent space can be seen above the AV valves, usually with a nonrestrictive shunt seen by color flow Doppler. The "goose neck" deformity seen angiographically on left ventriculography can occasionally be seen echocardiographically as an abnormal appearing left ventricular outflow tract.

SINUS VENOSUS SEPTAL DEFECT

The sinus venosus ASD is an interatrial communication that arises near the junction of the vena cava with the right atrium (Fig. 9; please *see* companion DVD for corresponding video). Therefore, this defect can present in two distinct morphologies. More commonly, the defect is located superiorly at the site of the SVC drainage to the right atrium but can be present inferiorly at the junction of the IVC and right atrium. In either case, these defects may be difficult to define anatomically via transthoracic imaging alone. However, transesophageal echocardiography can easily define the defect and any associated pathology. Imaging the atrial septum in the superior-inferior dimension (bicaval transesophageal view) often may reveal the defect.

Sinus venosus defects can be associated with other cardiac pathology. Commonly, the right upper pulmonary vein is anomalous when the venosus defect is seen at the junction of the SVC and right atrium. The anomalous vessel can connect to the SVC in the more typical case. However, the anomalous connections can be diverse in either morphology of sinus venosus defect, necessitating a meticulous definition of pulmonary venous drainage in these cases as a further cause of left-to-right shunting.

CORONARY SINUS ASD

This rare type of ASD presents an interatrial communication via an unroofed coronary sinus. Therefore, the defect is seen at the site of origin of the coronary sinus. A persistent left SVC is associated with this defect and can often be seen to drain into the coronary sinus or left atrium.

This defect may not be visualized on routine transthoracic echocardiography. However, the defect may be suggested by the constellation of right heart overload, a dilated coronary sinus, and the presence of a persistent left SVC. Multiplane transesophageal echocardiography

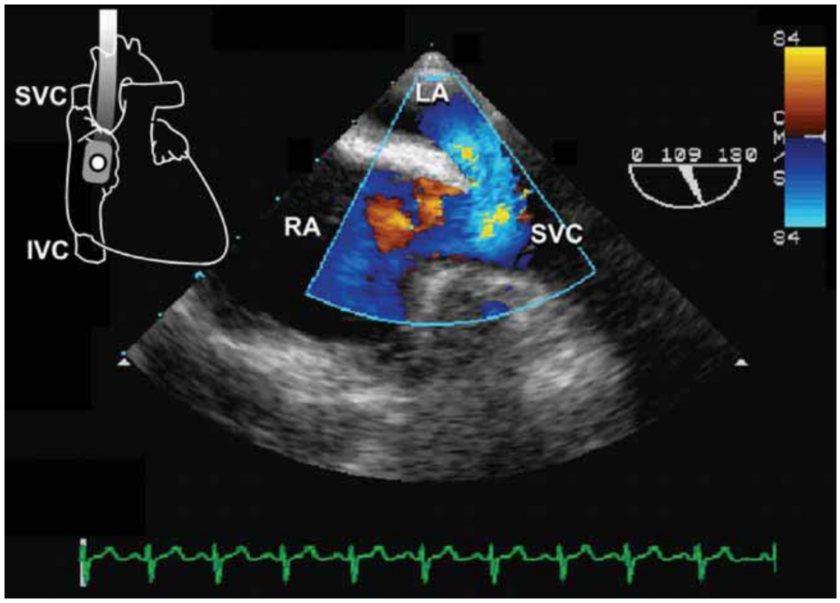

Fig. 9. Mid esophageal bicaval view showing sinus venous defect (superior vena cava [SVC]-type). Note the direction of flow across the defect—from left atrium to right atrium. (Please *see* companion DVD for corresponding video.)

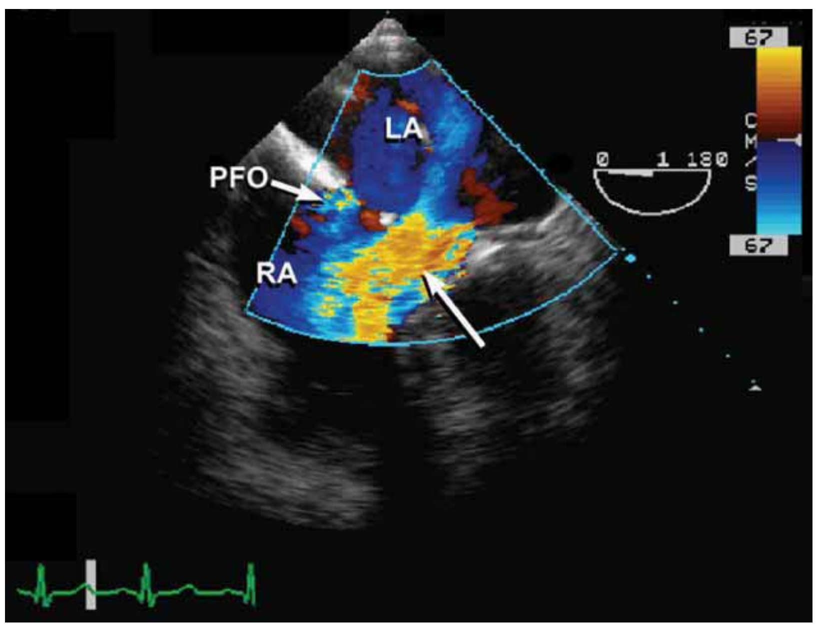

Fig. 10. Color flow Doppler demonstrates the presence of the coronary sinus ASD with left-to-right shunt. Note also the small shunt arising from a patent foramen ovale (PFO).

should be used to confirm the presence of this defect and associated pathology (Fig. 10). Intravenous injection of agitated saline contrast into the left arm can be used to evaluate the site of drainage of the left SVC (Fig. 11).

PATENT FORAMEN OVALE

This defect is not a true defect in embryological development of the either the septum primum or secundum. Rather, this lesion is the result of the failure of permanent fusion of the normally developed atrial septa.

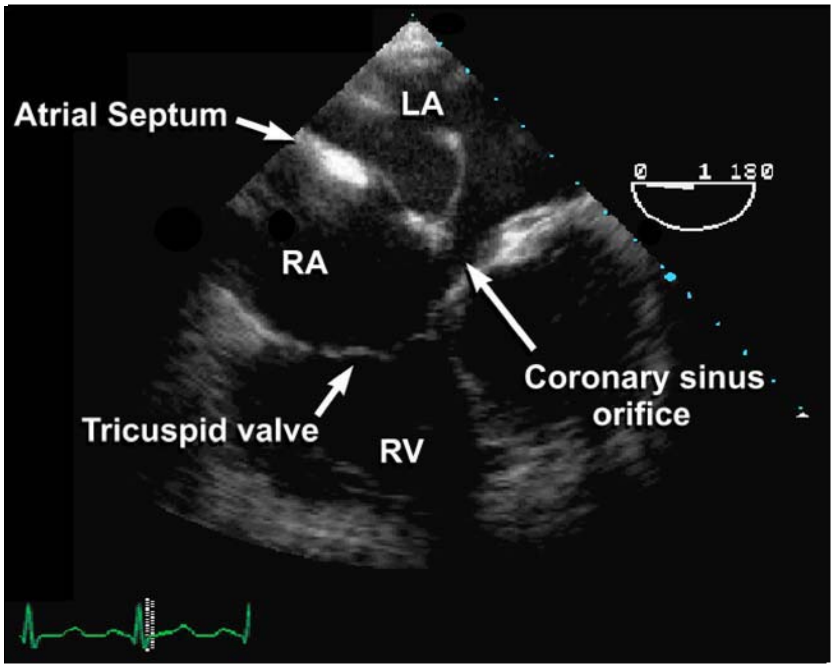

Fig. 11. Midesophageal view showing agitated saline contrast (bubble) study injection into the left arm. Bubbles were seen entering the coronary sinus, then the left atrium (LA), and then into the right atrium (RA).

Therefore, a dynamically patent orifice acting similar to a flap valve is present and may respond to interatrial pressure changes (Fig. 12; please *see* companion DVD for corresponding video; *see als0o* Chapter 17). Thus a right-to-left shunt may be present when right atrial pressures increase higher than left atrial pressures, such as with Valsalva, coughing, and so on. The defect may be functionally sealed when left atrial pressures exceed right, with only occasional right-to-left shunting.

The presence of this defect is easily evaluated with agitated saline contrast injection. Normally, the procedure is performed with and without Valsalva maneuver, as right-to-left shunting may not be seen under baseline conditions. The presence of contrast bubbles early after appearance in the right heart indicates the presence of right-to-left shunting. Color Doppler over the area of the fossa ovalis can often detect this defect. Transthoracic subcostal imaging or multiplane transesophageal echocardiography can usually illustrate the defect.

ECHOCARDIOGRAPHY IN THE MANAGEMENT OF ASDs

An accurate evaluation of the hemodynamic impact of the shunt is necessary following diagnosis. The degree of shunt flow is quantified by indexing pulmonary blood flow to systemic blood flow, the Qp/Qs ratio. Traditionally, when pulmonary blood flow exceeds systemic blood flow by 50% (a Qp/Qs ratio of 1.5:1.0), repair of the defect is recommended. However, in the case of ASDs with smaller shunts (ratios of <1.5:1.0), repair may still be appropriate owing to the risks for other problems including atrial arrhythmias, progressive pulmonary hypertension, congestive failure, or paradoxic emboli. PFOs have been repaired after cryptogenic stroke, especially in young patients.

Percutaneous closure of secundum ASDs and PFOs have been performed using transcatheter devices in experienced centers. Recent clinical data suggests that percutaneous closure techniques are safe and effective, supporting a less invasive alternative to surgical closure. A variety of devices are available for transcatheter closure such as the Amplatzer and CardioSeal closure devices.

The deployment of transcatheter devices for ASD or PFOs are generally guided by echocardiographic techniques (Fig. 12; please *see* companion DVD for corresponding video). Transesophageal echocardiography or intracardiac echocardiography are important tools for the morphological characterization of the lesion. For example, an accurate description of the ASD size and rim measurements is invaluable information in guiding device deployment. During PFO

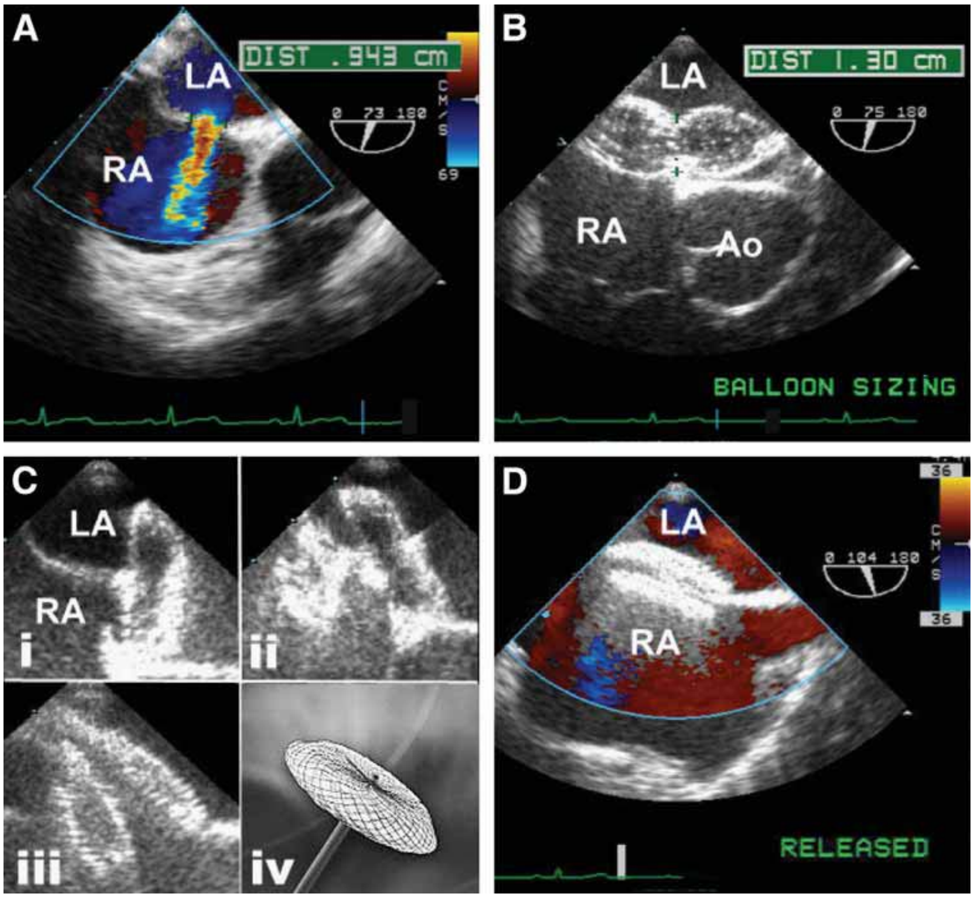

Fig. 12. Stages of percutaneous closure of a secundum atrial septal defect (ASD; **A**) using an Amplatzer device are shown on transesophageal echocardiography guidance. Balloon sizing of the defect **(B)** is followed by sequential deployment of the left atrial and right atrial arms of the device **(C)** (picture insert: AMPLATZER® Septal Occluder ASD). Following release of the device, interrogation of the closure was performed using Color Doppler **(D)** and agitated saline contrast. (Please *see* companion DVD for corresponding video.)

closure, transeptal puncture can be guided as well as an assessment of the PFO tunnel size and septal stiffness. Overall, echocardiography during device deployment is important for device sizing, positioning, and the assessment of residual shunting postdevice deployment (Fig. 12; please *see* companion DVD for corresponding video).

SUMMARY

In the current era, echocardiography provides a comprehensive assessment of ASDs. From initial diagnosis, classification, and assessment of associated lesions, echocardiographic techniques have largely supplanted cardiac catheterization. Further, as the placement of transcatheter devices for shunt closure is becoming an increasingly popular alternative to surgical closure, echocardiography is rapidly expanding as an interventional tool. As cardiac ultrasound expands to intracardiac and three-dimensional technologies, its future role in the assessment of simple and complex congenital cardiac lesions will certainly continue to evolve.

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