7 Heart Defects and Endocarditis

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Congenital Heart Disease in Adults

Congenital cardiovascular anomalies are present in 0.8–1% of all newborns. The number of different anomalies is so large that their complete description would be beyond the scope of this book. This chapter therefore focuses on the capabilities of modern cardiac imaging techniques in the most common types of congenital heart disease in adults—atrial septal defects, patent foramen ovale, and ventricular septal defects.

Atrial Septal Defect

Anatomy and Pathophysiology

Atrial septal defects (ASDs) account for ~10% of all congenital heart diseases and for 22–40% of congenital heart disease in adults. They are associated with varying degrees of left-to-right shunting of arterialized blood into the pulmonary circulation, depending on the size of the septal defect and the relative pressures. In extreme cases the shunt flow may be several times the volume flow of that in the systemic circulation. Atrial septal defects are primarily characterized by a volume overload on the right heart, which eventually lead to cardiac failure. With a left-to-right shunt above the level of the tricuspid valve, the great dilatory capacity of the pulmonary vessels can forestall a pressure rise in the pulmonary artery and right ventricle. Comparable to the ventricular septal defect however, a functional and/or organic rise in vascular resistance will develop over time in the pulmonary circulation, causing the pressure in the right ventricle to rise. When the pulmonary vascular resistance exceeds that of the systemic circulation in the presence of a ventricular or atrial septal defect, a "shunt reversal" occurs, leading to marked cyanosis. This phenomenon is termed the **Eisenmenger reaction**.

Three etiological types of atrial septal defect are distinguished:

- An **ostium secundum atrial septal defect (ASD II)** is located in the central portion of the atrial septum in the region of the fossa ovalis. It is the most common type, accounting for 60–70% of atrial septal defects.
- An **ostium primum atrial septal defect (ASD I)** is usually a large defect that comprises 15–25% of atrial septal defects. It results from a failure of fusion of the septum primum with the endocardial cushion between the atrioventricular valves. As a result, it is commonly associated with mitral valve defects and occasionally with tricuspid valve defects.
- A **sinus venosus atrial septal defect** is present in 5–15% of cases. It occurs in the posterosuperior portion of the atrial septum between the termination of the superior vena cava and the fossa ovalis. It is frequently associated with anomalous termination of the right pulmonary veins in the right atrium.

Clinical Features

Symptoms

- Generally asymptomatic until the third decade; 70% of patients are symptomatic by the fifth decade
- Dyspnea, rapid fatigability (manifestations of heart failure).
- Palpitations (in patients with atrial arrhythmias)
- Proneness to respiratory infections
- Approximately 15% of patients show clinical symptoms of associated mitral insufficiency

Complications

- Atrial fibrillation or flutter
- Mitral insufficiency

Imaging

Echocardiography

2D Echocardiography

- Transthoracic scanning enables direct visualization of the septal defect (excepting sinus venosus defects) (**Fig. 7.1**).
- Transesophageal scanning permits direct visualization of a sinus venosus defect (**Fig. 7.2**).
- Enlargement of the right atrium and right ventricle
- Detection of associated congenital anomalies

3D Echocardiography

- Enables direct visualization of the ASD in the frontal view to assess the location, size, and shape of the defect.
- Can detect an AV canal in patients with ASD I.
- Allows direct visualization of a cleft in the anterior mitral valve leaflet in ASD I.

Color Doppler Echocardiography

- Detection of shunt flow by demonstrating a flow jet through the atrial septum into the right atrium
- Frequently primary detection of a sinus venosus defect is based on an abnormal flow signal near the termination of the superior vena cava (**Fig. 7.2b**).
- Detection of an AV canal in patients with ASD I
- Detection of associated mitral insufficiency in ASD I

Contrast Echocardiography

- Detection of a left-to-right shunt based on the washout phenomenon in the contrast-filled right atrium (**Fig. 7.1b**).

CW Doppler Echocardiography

- Pulmonary hypertension can be diagnosed by assessing the systolic pulmonary artery pressure based on the regurgitant signal from the tricuspid valve.

Essential Point
In the presence of clinical suspicion of a left-to-right shunt at the atrial level, it is possible to overlook a high-sited sinus venosus type of ASD if the atrial septum appears normal. Often a sinus venosus defect can be detected only by multiplanar transesophageal echocardiography in color Doppler mode.

Magnetic Resonance Imaging and Computed Tomography

The standard planes for detecting an ASD in MRI are horizontal long-axis slices and short-axis slices through the heart. Spoiled gradient-echo sequences are preferred. They are of advantage over SE and balanced SSFP sequences in that they can also detect smaller defects by the associated zones of turbulent flow, for example.

Determination of the maximum size of the ASD is essential in selecting patients for interventional repairs, an unsuitable procedure for large defects. Both MRI and multislice CT (MSCT) can document the enlargement of the right atrium and right ventricle (**Fig. 7.3a**). In addition to demonstrating morphologi-

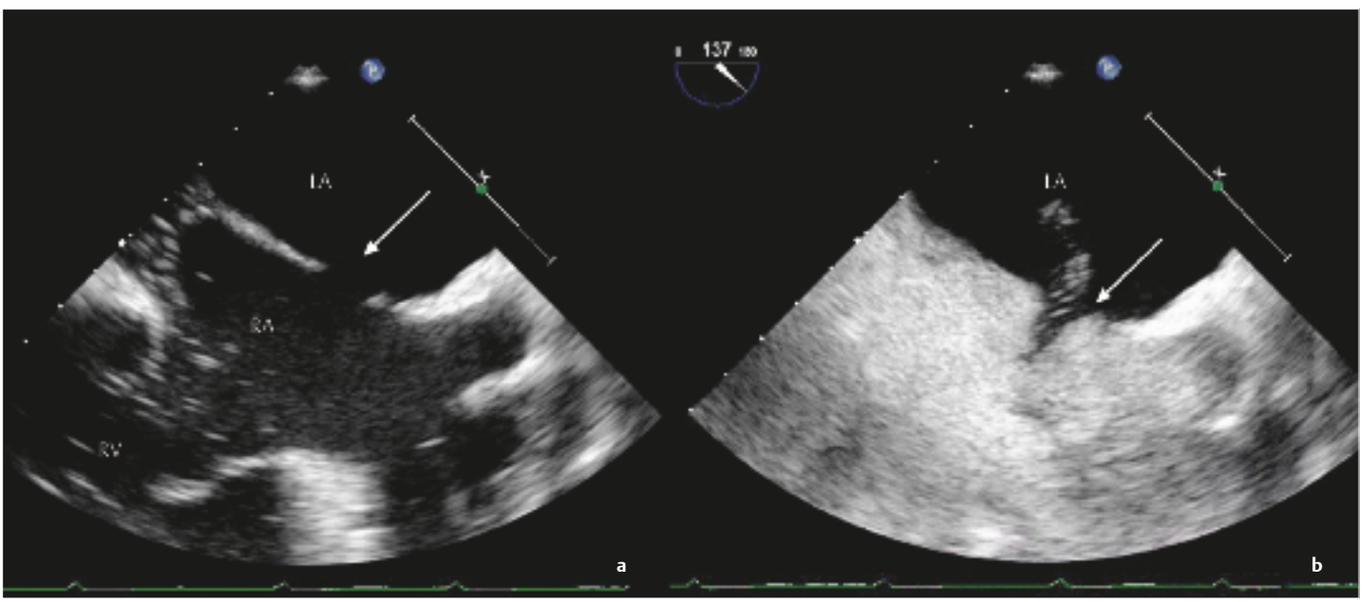


Fig. 7.1 a, b Transesophageal echocardiography of ASD II (arrow).
a Prior to contrast wash-in.
b Typical appearance of the washout phenomenon (arrow) in a left-to-right shunt at the atrial level. Various bubbles can be seen entering the left atrium after passing through the shunt.

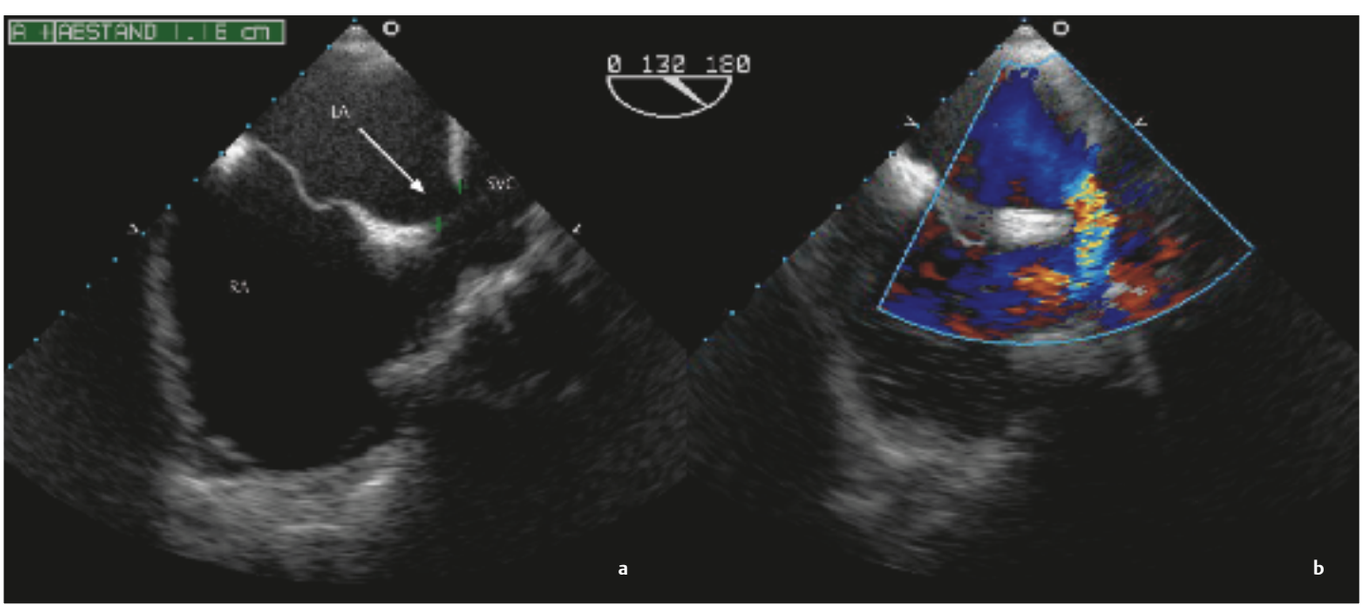


Fig. 7.2 a, b Sinus venosus atrial septal defect documented by transesophageal echocardiography.
a The size of the defect (arrow) is measured at the entry of the superior vena cava into the right atrium.
b Abnormal flow signal detected by color Doppler echocardiography.

cal features, MRI also enables an accurate assessment of the Q_p/Q_s ratio based on flow measurements in the ascending aorta and pulmonary trunk. This ratio is important in selecting patients for operative or interventional repair.[1] The Q_p/Q_s ratio can also be determined using cine MRI to identify right and left ventricular stroke volumes, provided the presence of an additional significant valvular defect can be excluded. MRI and CT additionally detect any anomalous pulmonary venous termination that may be present. Operative treatment is the only option available for correcting this anomaly[2] (**Fig. 7.3b**).

Patent Foramen Ovale

Anatomy and Pathophysiology

Patent foramen ovale (PFO) is a valvelike opening that persists postnatally between the septum primum and secundum in the region of the fossa ovalis owing to failed fusion of these septal elements. A right-to-left shunt at the atrial level may occur spontaneously or in response to a Valsalva maneuver. Autopsy results indicate that PFO has a prevalence of ~25% in the general population. Its clinical significance is that it places patients at risk for paradoxical emboli. This risk appears to be particularly high in patients with a hypermobile atrial septal aneurysm, which frequently coexists with PFO. Because the opening and volume of the shunt are generally small, hemodynamic complications do not occur.

Clinical Features

Symptoms

- The majority of patients with PFO are asymptomatic.

Complications

- Central or peripheral emboli, stroke, transient ischemic attacks, dizzy spells, migraines

Imaging

Echocardiography

M-Mode Echocardiography

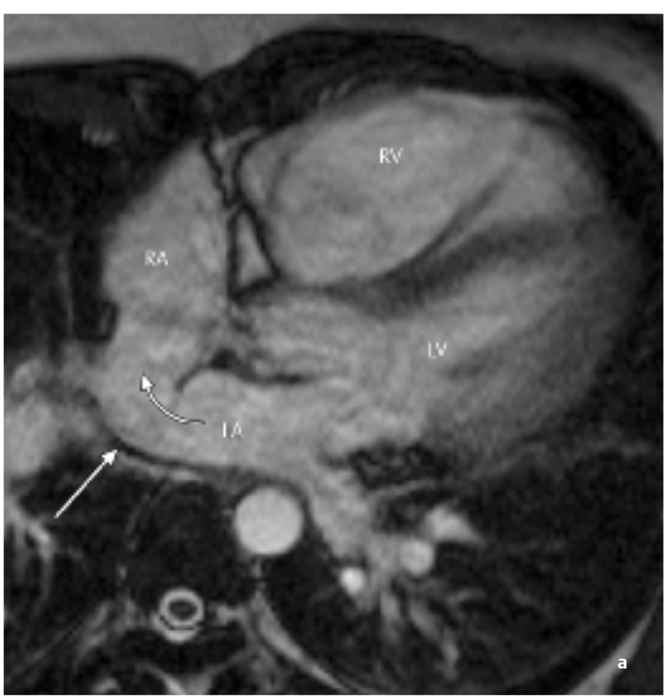
- M-mode can document hypermobile excursions associated with an atrial septal aneurysm (**Fig. 7.4a**).

2D Echocardiography

- Demonstrates the valvelike separation between the septum primum and secundum; but cannot reliably detect patent foramen ovale (2D visualization only by transesophageal scanning).
- Can detect an atrial septal aneurysm (frequently also detectable by transthoracic scanning.

Contrast Echocardiography

- Can reliably detect or exclude patent foramen ovale based on the right-to-left passage of contrast medium, either spontaneously or in response to a Valsalva maneuver (**Fig. 7.4b**).



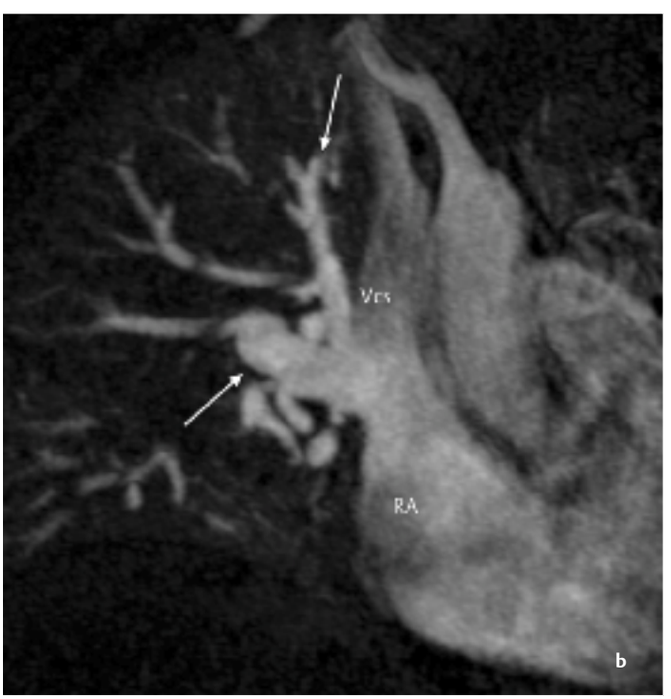


Fig. 7.3 a, b Large ASD II in a 32-year-old woman.
a Cine image in the four-chamber view demonstrates the large ASD (arrow).
b Contrast-enhanced 3D MRA further demonstrates partial anomalous pulmonary venous termination on the right side. The upper and middle lobe veins (arrows) open into the superior vena cava.