#### Chapter

# **Congenital cardiac malformations**

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#### Introduction

Relatively recent advances in pediatric cardiovascular surgery, catheter-based interventional therapies, intensive care and medical management have dramatically changed the landscape of the field of congenital heart disease (CHD). The complexity of the anatomy and physiology of patients surviving with CHD is increasing exponentially, and the majority will survive to adulthood. These changes are placing new demands on imaging to diagnose and plan medical management. There are a number of imaging modalities available to the clinician and radiologist when it comes to these evaluations. The primary modalities, and the ones highlighted in this chapter, include the chest X-ray (CXR), echocardiography, cardiac catheterization with X-ray angiography, cardiac magnetic resonance imaging (MRI) and cardiac CT angiography (CTA).

The following chapter is divided into (1) left-to-right shunt lesions: atrial septal defects (ASDs), ventricular septal defects (VSDs), patent ductus arteriosus (PDA), common atrioventricular canal defects (CAVCs) and partial anomalous pulmonary venous return (PAPVR); (2) obstructive lesions: coarctation of the aorta (CoA); (3) cyanotic lesions: tetralogy of Fallot (TOF), truncus arteriosus (TA), D-transposition of the great arteries (D-TGA), total anomalous pulmonary venous return (TAPVR) and the univentricular heart, and finally, (4) great vessel anomalies: aortic rings and pulmonary artery slings. For each defect, the anatomy, physiology, natural history and approach to repair are briefly discussed before presenting diagnostic imaging examples.

## Left-to-right shunt lesions

### Atrial septal defects

Isolated secundum atrial septal defect (ASD) occurs in 5–10% of all CHD, and about 30–50% of children with CHD have an ASD. ASDs occur twice as frequently in females. There are four types of atrial septal defect; these include secundum ASDs, sinus venosus defects, primum ASDs and coronary sinus defects (Figure 3.1).

Secundum ASDs are the most common, accounting for 50–70% of all ASDs. Secundum ASDs are the result of a defect in the septum primum; the tissue that covers the fossa ovalis (Figures 3.2, 3.3).



**Figure 3.1** Atrial septal defect – diagram. The location of the different types of atrial septal defect (ASD) are demonstrated. Secundum ASDs (ASD 2°) are the result of fenestrations in, or deficiency of, septum primum. Primum ASDs (ASD 1°) represent deficiencies in the endocardial cushion portion of the atrial septum. Sinus venosus ASDs occur at the junction of the superior vena cava with the right atrium, or less frequently, at the junction of the inferior vena cava with the right atrium. From the Multimedia Library of Congenital Heart Disease, Children's Hospital, Boston, MA, editor Robert Geggel, MD, www.childrenshospital. org/mml/cvp with permission.

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