60. Congenital Heart Disease

The MR evaluation of congenital heart disease is a minimally invasive alternative to conventional cardiothoracic angiography, although sedation of uncooperative pediatric patients may be required. A common congenital disorder of the heart is a bicuspid aortic valve of which aortic stenosis is the most frequent complication (see Chapter 59). Further frequent findings in congenital heart disease (alone or in combination with other complex malformations) include atrial and ventricular septal defects (ASD / VSD), their significance depending upon the size of and flow through the defect. Absent other shunting, comparison of pulmonary artery and aortic outflow (with velocity-encoded MR) or left and right ventricular volumes (the right is normally larger with a VSD) allow quantification of shunt volume. The four chamber cine image of Fig. 60.1A demonstrates a VSD of the muscular septum—as opposed to the more common membranous defect—and resulting right ventricular dilatation and hypertrophy. A low SI jet into the left ventricle identifies the reversal of normal left to right flow across the defect, corresponding with an Eisenmenger-type physiology. Atrial septal defects are classified as secundum, arising from a defect in the fossa ovale, and less common primum types, arising from sinus venosus defects. Atrioventricular septal defects (AVSD) are characterized by an insufficient fusion of embryonic endocardial cushions thus resulting in a deficient separation of atria from ventricles at the level of the AV-valves in combination with defects of the lower atrial septum and the membranous ventricular septum.

Defects in the thin atrial septum are best visualized in four-chamber view cine images. Resulting dilatation of the right atrium and ventricle may occur, and septal configuration progressively loses concavity to the right (best seen in short axis cine views) as pressures therein increase. Quantitative measurements may be made with techniques similar to those above. A patent ductus arteriosus is best visualized on CE MRA and once detected warrants further scrutiny for other congenital disorders (such as Tetralogy of Fallot). Velocity-encoded MR must be obtained to evaluate flow across the ductus with other quantitative measurements performed as above. Aortic coarctation also frequently presents with other congenital abnormalities. A high grade coarctation is demonstrated on the thoracic CE MRA in Fig. 60.1B with prominent collateralization of the thoracic vasculature. ECG (and possibly respiratory) gated cine images of the heart and arch allow a detailed evaluation of cardiac function and aortic morphology to the level of the renal arteries. Velocity-encoded MR of the ascending and descending aorta allow assessment of the lesion's hemodynamic significance, while black blood images may aid detection of both focal areas of stenosis and lesions within the descending or abdominal aorta. Vascular rings are particularly well-seen

with contrast administration, allowing assessment of arterial impingement on the low SI trachea.

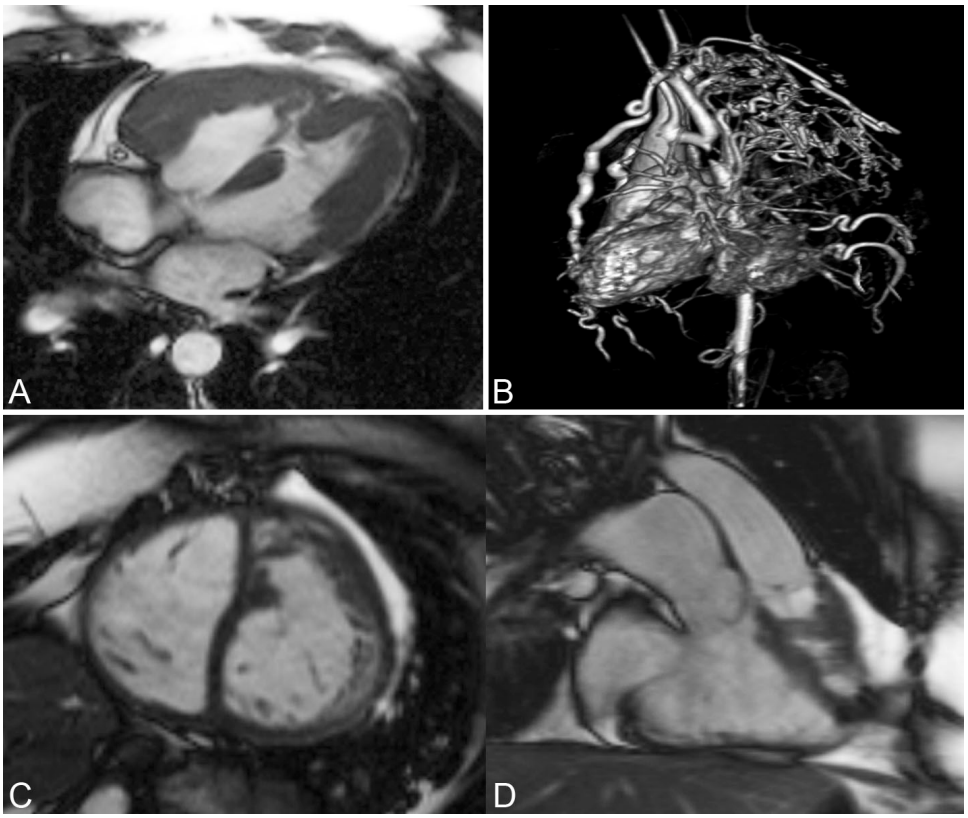

Fig. 60.1

The major cyanotic congenital disorders include transposition of the great arteries and Tetralogy of Fallot. In transposition, the left ventricle is connected to the pulmonary arteries and the right to the aorta. The dextro type of transposition relies on anatomical shunting to maintain oxygenation as deoxygenated blood is pumped through the aorta by the right ventricle and oxygenated blood through the pulmonary arteries via the left ventricle. Surgical interventions include atrial switches—whereby a baffle directs blood from the vena cava to left ventricle and pulmonary arteries, while pulmonary venous return is directed into the aorta via the right ventricle. Nowadays, arterial switching—whereby the pulmonary arteries and aorta are re-attached to their normal ventricles is preferred, resulting in a better long term outcome for patients. MR evaluation of postoperative atrial switches requires assessment of the baffle for leaks or stenosis and of the right ventricle for hypertrophy and dysfunction. Outflow tract obstructions, valvular regurgitation, and associated shunts may be quantitatively analyzed. With arterial switch procedures, additional attention should be paid to any pulmonary artery stenosis or aortic root dilatation. Levo transposition consists of atrioventricular and ventriculoarterial discordance,

whereby the systemic blood flow returns to the right-sided but morphologically left ventricle which empties into the pulmonary arteries, while pulmonary venous blood enters the left-sided but morphologically right ventricle which empties into the aorta. This appearance is demonstrated on the short-axis cine images of Fig. 60.1C. Here the identity of the posterior, although morphologically right ventricle is established by the moderator band. Additional cine images adapted to focus on the large vessels show the essentially parallel course of the ascending arteries (Fig. 60.1D). Definite surgical therapy herein requires a double switch procedure (atrial and arterial switch). Tetralogy of Fallot is the most common cyanotic congenital disorder, consisting of an overriding aorta, pulmonary stenosis, right ventricular hypertrophy, and a VSD. Preoperatively, cine MRI assesses right ventricular morphology and function, while CE MRA delineates aortopulmonary collaterals and determines ductus arteriosus patency. In operative planning, specific coronary artery imaging may be useful, particularly in detecting the relation of the arteries to the right ventricular outflow tract, a portion of which is often resected surgically. Postoperatively, the right ventricle should also be carefully examined for evidence of dysfunction, aneurysm, or recurrent stenoses in the RVOT or the pulmonary arteries.