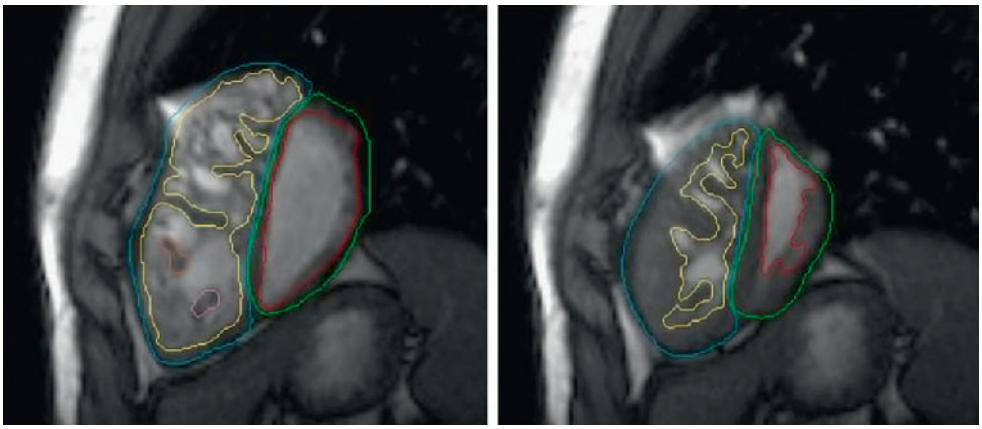

FIGURE 6-2 Short axis magnetic resonance images of the right and left ventricles with epicardial and endocardial tracings of both ventricular cavities. There are a predefined number of slices through the heart with a constant thickness. The volumes of the left and right ventricles in each slice are calculated and summed together in end diastole and end systole to determine the total right and left ventricular volumes (i.e., Simpon's method).

pulmonary valve annulus, or both. Reintervention is necessary in approximately 10% of adults with repaired TOF after 20 years of follow-up. With longer follow-up, the incidence of reintervention continues to increase. The most common indication for reintervention is pulmonary valve replacement for severe pulmonary valve regurgitation.

Prognosis

In the developed world, the unoperated adult with TOF has become a rarity because most patients undergo palliation (i.e., stenting) or repair in childhood. Survival of the unoperated patient to the seventh decade has been described but is rare. Only 11% of unrepaired patients are alive at 20 years of age and only 3% at 40 years.

Late survival after repair of TOF is excellent. Survival rates at 32 and 35 years are 86% and 85%, respectively, compared with 95% for age- and sex-matched controls. Importantly, most patients live an unrestricted life. However, many patients over time develop late symptoms related to numerous, long-term complications after TOF repair. Late complications include endocarditis, aortic regurgitation with or without aortic root dilation (typically due to damage of the aortic valve during VSD closure or to an intrinsic aortic root abnormality), LV dysfunction (from inadequate myocardial protection during previous repair or chronic LV volume overload due to long-standing palliative arterial shunts), residual pulmonary obstruction, residual pulmonary valve regurgitation, RV dysfunction (due to pulmonary regurgitation or pulmonary stenosis), atrial arrhythmias (typically atrial flutter), ventricular arrhythmias, and heart block.

Transposition of the Great Arteries

Definition and Epidemiology

Transposition of the great arteries (TGA) represents 3.8% of all congenital heart disease. In complete TGA, the aorta arises from the right ventricle and the pulmonary artery from the left ventricle. As a result, the systemic venous flow (i.e., blood with low oxygen content) is returned to the right ventricle and is then pumped to the body through the aorta without passing through the lungs for gas exchange. The pulmonary venous flow (i.e., oxygenated blood) returning to the left ventricle is then pumped back to the lungs. As a result, the systemic and pulmonary circulations run in parallel. Oxygenation and survival depend on mixing between the systemic and pulmonary circulations at the atrial, ventricular, or PDA level. In 50% of cases, there are other anomalies: VSD (30%), pulmonary stenosis (5% to 10%), aortic stenosis, and coarctation of the aorta (≤5%).

The first definitive operations for TGA (i.e., atrial switch procedures) were described by Senning in 1959 and Mustard in 1964. In these procedures, the systemic and pulmonary venous returns are rerouted in the atrium by constructing baffles. The systemic venous return from the superior and inferior vena cavae is directed through the mitral valve and into the left ventricle, which is connected to the pulmonary artery. The pulmonary venous return is then directed through the tricuspid valve into the right ventricle, which is connected to the aorta. These procedures leave the left ventricle as the pulmonary ventricle and the right ventricle as the systemic ventricle.

Over the past 10 to 20 years, the arterial switch procedure has gained popularity. During the procedure, the great arteries are transected and reanastomosed to the correct ventricle (i.e., left ventricle to the aorta and right ventricle to the pulmonary artery) along with coronary artery transfer. Operative survival after the arterial switch procedure is very good, with a surgical mortality rate of 2% to 5%.

Pathology

Most infants who do not have surgical intervention die in the first few months of life. For adults born with complete TGA who have had an atrial switch procedure, the right ventricle continues to be the systemic ventricle, and the left ventricle is the subpulmonic ventricle. Long-term follow-up series have demonstrated that the right ventricle can function as the systemic ventricle for 30 to 40 years, but with longer follow-up, systemic ventricular dysfunction continues to increase. At the 35-year follow-up, approximately 61% of patients have developed moderate or severe RV dysfunction.