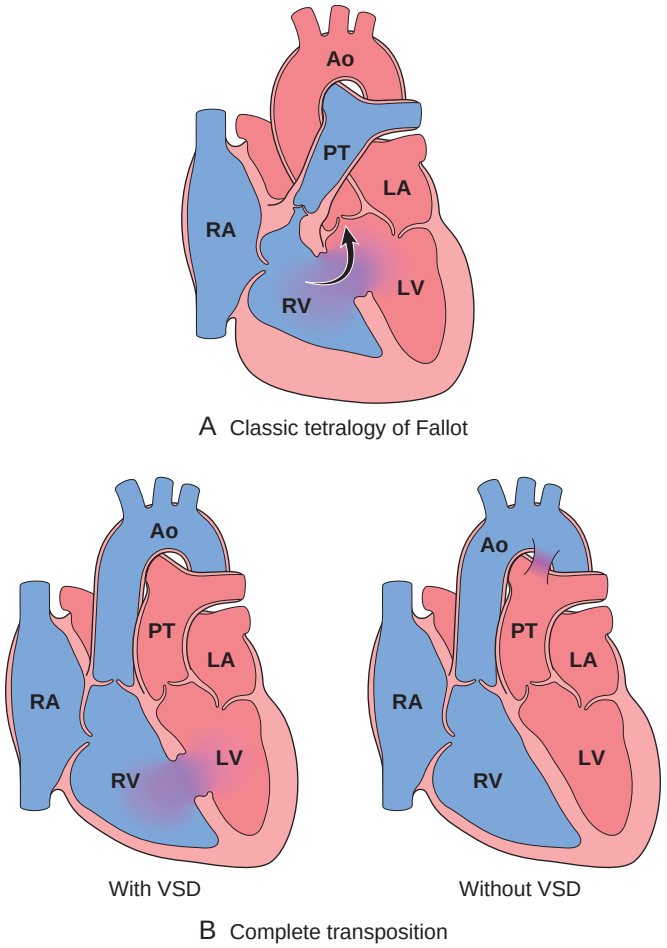

Figure 12-6 Common congenital right-to-left shunts *(cyanotic congenital heart disease).* **A,** Tetralogy of Fallot. The direction of shunting across the ventricular septal defect (VSD) depends on the degree of the subpulmonary stenosis; when severe, a right-to-left shunt results (*arrow*). **B,** Transposition of the great arteries with and without VSD. Ao, Aorta; LA, left atrium; LV, left ventricle; PT, pulmonary trunk; RA, right atrium; RV, right ventricle.

overriding the defect and both ventricular chambers. The obstruction to right ventricular outflow is most often due to narrowing of the infundibulum (subpulmonic stenosis) but can be accompanied by pulmonary valvular stenosis. Sometimes there is complete atresia of the pulmonary valve and variable portions of the pulmonary arteries, such that blood flow through a PDA, dilated bronchial arteries, or both, is necessary for survival. Aortic valve insufficiency or an ASD may also be present; a right aortic arch is present in about 25% of cases.

Clinical Features. Even untreated, patients with TOF can survive into adult life; 10% of untreated patients are alive at 20 years and 3% survive for 40 years. The clinical consequences depend primarily on the severity of the subpulmonary stenosis, since this determines the direction of blood flow. If the subpulmonary stenosis is mild, the abnormality resembles an isolated VSD, and the shunt may be left-to-right, without cyanosis (so-called "pink tetralogy"). *With more severe right ventricular outflow obstruction, right-sided pressures approach or exceed left-sided pressures, and right-to-left shunting develops, producing cyanosis (classic TOF).* Most infants with TOF are cyanotic from birth or soon thereafter. The more severe the subpulmonic stenosis, the more hypoplastic are the pulmonary arteries (i.e., smaller and thinner-walled), and the larger is the overriding aorta. As the child grows and the heart increases in size, the pulmonic orifice does not expand proportionally, making the obstruction progressively worse. The subpulmonary stenosis, however, protects the pulmonary vasculature from pressure overload, and right ventricular failure is rare because the right ventricle is decompressed by the shunting of blood into the left ventricle and aorta. Complete surgical repair is possible but becomes complicated for individuals with pulmonary atresia and dilated bronchial arteries.

Transposition of the Great Arteries

TGA produces ventriculoarterial discordance. Thus, the aorta lies anterior and arises from the right ventricle, while the pulmonary artery is relatively posterior and emanates from the left ventricle (Fig. 12-7; see also Fig. 12-6*B*). The atrium-to-ventricle connections are normal (concordant), with the right atrium joining the right ventricle and the left atrium emptying into the left ventricle. The embryologic defect in complete TGA stems from abnormal formation of the truncal and aortopulmonary septa. The result is separation of the systemic and pulmonary circulations, a condition incompatible with postnatal life unless a shunt exists for adequate mixing of blood.

The outlook for infants with TGA depends on the degree of blood "mixing," the magnitude of tissue hypoxia, and the ability of the right ventricle to maintain the systemic circulation. Patients with TGA and a VSD (approximately 35%) often have a stable shunt. However, dependence on a patent foramen ovale or ductus arteriosus for blood mixing (approximately 65%) is problematic. These systemic-to-pulmonary connections tend to close early and thus require intervention to create a new shunt within the first few days of life (e.g., balloon atrial septostomy). With

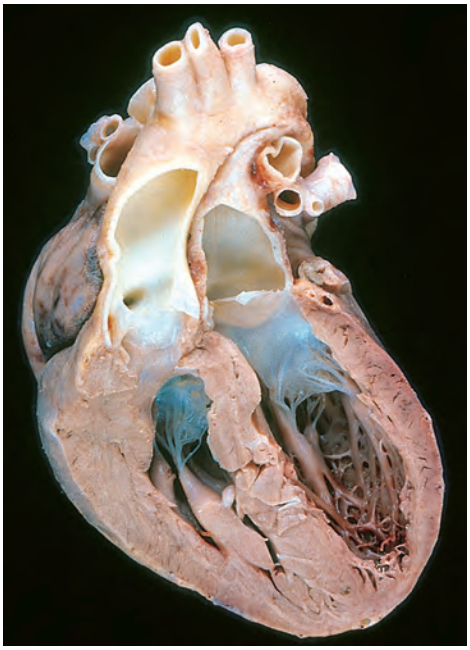

Figure 12-7 Transposition of the great arteries. (Courtesy William D. Edwards, MD, Mayo Clinic, Rochester, Minn.)