The ECG shows RV hypertrophy. Chest x-ray shows a normal-sized, boot-shaped heart (*coeur en sabot*) with a prominent RV and a concavity in the region of the pulmonary conus. Pulmonary vascular markings are typically diminished, and the aortic arch and knob may be on the right side. Echocardiography demonstrates the malaligned VSD with the overriding aorta and the site and severity of PS, which may be subpulmonic (fixed or dynamic), at the pulmonary valve or in the main or branch pulmonary arteries. Classic contrast angiography may provide details regarding the RV outflow tract, pulmonary valve and annulus, and caliber of the main branches of the pulmonary artery, as well as about possible associated aortopulmonary collaterals. Coronary arteriography identifies the anatomy and course of the coronary arteries, which may be anomalous. Cardiac MRI and CT complement echocardiography and provide much of the information gathered by angiography as well as additional functional information. MRI is considered the clinical gold standard for quantification of RV volume and function as well as quantification of the pulmonary regurgitation severity.

TREATMENT TETRALOGY OF FALLOT

For a variety of reasons, only a few adults with tetralogy of Fallot have not had some form of previous surgical intervention. Reoperation in adults is most commonly for severe pulmonary regurgitation or pulmonary stenosis. Long-term concerns about ventricular function persist. Ventricular and atrial arrhythmias occur, respectively, in 15% and 25% of adults and may require medical treatment, electrophysiologic study and ablation, defibrillator placement, or transcatheter or surgical intervention, usually including pulmonary valve replacement. Transcatheter pulmonary valve replacement is widely used in patients meeting anatomic criteria. The aortic root has a medial tissue defect; it is commonly enlarged and associated with aortic regurgitation. Endocarditis remains a risk despite surgical repair.

COMPLETE TRANSPOSITION OF THE GREAT ARTERIES

This condition is commonly called *dextro-* or D-*transposition of the great arteries*. The aorta arises rightward anteriorly from the RV, and the pulmonary artery emerges leftward and posteriorly from the LV, which results in two separate parallel circulations; some communication between them must exist after birth to sustain life. Most patients have an interatrial communication, two-thirds have a patent ductus arteriosus, and about one-third have an associated VSD. Transposition is more common in males and accounts for ~10% of cyanotic heart disease. The course is determined by the degree of tissue hypoxemia, the ability of each ventricle to sustain an increased workload in the presence of reduced coronary arterial oxygenation, the nature of the associated cardiovascular anomalies, and the status of the pulmonary vascular bed. Patients who do not undergo surgical palliation generally do not survive to reach adulthood. The long-term outcomes in those that have undergone surgery are in large part determined by the type of surgery performed. By the third decade of life, ~30% of patients with "atrial switch" operations will have developed decreased RV function and progressive tricuspid regurgitation, which may lead to congestive heart failure. Pulmonary vascular obstruction develops by 1–2 years of age in patients with an associated large VSD or large patent ductus arteriosus in the absence of obstruction to LV outflow.

TREATMENT TRANSPOSITION OF THE GREAT ARTERIES

The balloon or blade catheter or surgical creation or enlargement of an interatrial communication in the neonate is the simplest procedure for providing increased intracardiac mixing of systemic and pulmonary venous blood. Systemic pulmonary artery anastomosis may be indicated in the patient with severe obstruction to LV outflow and diminished pulmonary blood flow. Intracardiac repair may be accomplished by rearranging the venous returns (intraatrial switch, i.e., Mustard or Senning operation) so that the systemic venous blood is directed to the mitral valve and, thence, to the LV and pulmonary artery, while the pulmonary venous blood is diverted through the tricuspid valve and RV to the aorta. The late survival after these repairs is good, but arrhythmias (e.g., atrial flutter) or conduction defects (e.g., sick sinus syndrome) occur in ~50% of such patients by 30 years after the intraatrial switch surgery. Progressive dysfunction of the systemic subaortic RV, tricuspid regurgitation, ventricular arrhythmias, cardiac arrest, and late sudden death are worrisome features. Preferably, this malformation is corrected in infancy by transposing both coronary arteries to the posterior artery and transecting, contraposing, and anastomosing the aorta and pulmonary arteries (arterial-switch operation). For patients with a VSD in whom it is necessary to bypass a severely obstructed LV outflow tract, corrective operation employs an intracardiac ventricular baffle and extracardiac prosthetic conduit to replace the pulmonary artery (Rastelli procedure).

SINGLE VENTRICLE

This is a family of complex lesions with both atrioventricular valves or a common atrioventricular valve opening to a single ventricular chamber. Associated anomalies include abnormal great artery positional relationships, pulmonic valvular or subvalvular stenosis, and subaortic stenosis. Survival to adulthood depends on a relatively normal pulmonary blood flow, yet normal pulmonary resistance and good ventricular function. Modifications of the Fontan approach are generally applied to carefully selected patients with creation of a pathway(s) from the systemic veins to the pulmonary arteries.

TRICUSPID ATRESIA

This malformation is characterized by atresia of the tricuspid valve; an interatrial communication; and, frequently, hypoplasia of the RV and pulmonary artery. The clinical picture is usually dominated by severe cyanosis due to obligatory admixture of systemic and pulmonary venous blood in the LV. The ECG characteristically shows RA enlargement, left-axis deviation, and LV hypertrophy.

Atrial septostomy and palliative operations to increase pulmonary blood flow, often by anastomosis of a systemic artery or vein to a pulmonary artery, may allow survival to the second or third decade. A Fontan atriopulmonary or total cavopulmonary connection may then allow functional correction in patients with normal or low pulmonary arterial resistance pressure and good LV function. There are a number of important long-term considerations with the Fontan circulation, including the development of arrhythmias, progressive liver dysfunction, thromboembolic complications, and potential long-term need for heart or heart and liver transplantation.

EBSTEIN'S ANOMALY

Characterized by a downward displacement of the tricuspid valve into the RV, due to anomalous attachment of the tricuspid leaflets, the Ebstein tricuspid valve tissue is dysplastic and results in tricuspid regurgitation. The abnormally situated tricuspid orifice produces an "atrialized" portion of the RV lying between the atrioventricular ring and the origin of the valve, which is continuous with the RA chamber. Often, the RV is hypoplastic. Although the clinical manifestations are variable, some patients come to initial attention because of either (1) progressive cyanosis from right-to-left atrial shunting, (2) symptoms due to tricuspid regurgitation and RV dysfunction, or (3) paroxysmal atrial tachyarrhythmias with or without atrioventricular bypass tracts (Wolff-Parkinson-White [WPW] syndrome). Diagnostic findings by two-dimensional echocardiography include the abnormal positional relation between the tricuspid and mitral valves with abnormally increased apical displacement of the septal tricuspid leaflet. Tricuspid regurgitation is quantitated by Doppler examination. Surgical approaches include prosthetic replacement of the tricuspid valve when the leaflets are tethered or repair of the native valve.