prostaglandin E_1 infusion is needed to maintain ductal patency and ensure adequate systemic blood flow.

Surgical treatment: A multiple-stage approach is used. The first stage is a Norwood procedure, which involves an anastomosis of the main pulmonary artery to the aorta to create a new aorta, shunting to provide pulmonary blood flow (usually with a modified Blalock-Taussig shunt), and creation of a large ASD. Postoperative complications include imbalance of systemic and pulmonary blood flow, bleeding, low cardiac output, and persistent heart failure. A new modification of the first stage repair is the use of a right ventricle–to–pulmonary artery homograft conduit instead of a shunt to supply pulmonary blood flow (Sano procedure). The second stage is often a bidirectional Glenn shunt procedure (see Fig. 23-10) or a hemi-Fontan operation. Both involve anastomosing the SVC to the right pulmonary artery so that SVC flow bypasses the right atrium and flows directly to the lungs. The procedure is usually done at 3 to 6 months of age to relieve cyanosis and reduce the volume load on the right ventricle. The final repair is a modified Fontan procedure (see Tricuspid Atresia, Box 23-3).

Transplantation: Heart transplantation in the newborn period is another option for these infants. Problems include the shortage of newborn organ donors, risk of rejection, long-term problems with chronic immunosuppression, and infection (see Heart Transplantation, later in the chapter).

Prognosis: For the first-stage repair, survival rates vary widely in different centers. Much progress has been made, and some experienced centers are reporting mortality rates of about 10% (Tweddell, Hoffman, Mussatto, et al, 2002). Long-term problems with repair include worsening ventricular function, tricuspid regurgitation, recurrent aortic arch narrowing, dysrhythmias, and developmental delays. There is a risk of mortality between surgical procedures. The mortality for the later two operations is less than 5%.

ASD, Atrial septal defect; *HF,* heart failure; *IV,* intravenous; *IVC,* inferior vena cava; PAH, pulmonary artery hypertension; *PDA,*