Another associated defect may be a VSD. The presence of a VSD increases the risk of HF because it permits blood to flow from the right to the left ventricle, into the pulmonary artery, and finally to the lungs. However, it also produces high pulmonary blood flow under high pressure, which can result in high pulmonary vascular resistance.

Clinical manifestations: These depend on the type and size of the associated defects. Newborns with minimum communication are severely cyanotic and have depressed function at birth. Those with large septal defects or a PDA may be less cyanotic but have symptoms of HF. Heart sounds vary according to the type of defect present. Cardiomegaly is usually evident a few weeks after birth.

Therapeutic management (to provide intracardiac mixing): The administration of IV prostaglandin E_1 may be initiated to keep the ductus arteriosus open to temporarily increase blood mixing and provide an oxygen saturation of 75% or to maintain cardiac output. During cardiac catheterization or under echocardiographic guidance, a balloon atrial septostomy (Rashkind procedure) may also be performed to increase mixing by opening the atrial septum.

Surgical treatment: An arterial switch procedure is the procedure of choice performed in the first weeks of life. It involves transecting the great arteries and anastomosing the main pulmonary artery to the proximal aorta (just above the aortic valve) and anastomosing the ascending aorta to the proximal pulmonary artery. The coronary arteries are switched from the proximal aorta to the proximal pulmonary artery to create a new aorta. Reimplantation of the coronary arteries is critical to the infant's survival, and they must be reattached without torsion or kinking to provide the heart with its supply of oxygen. The advantage of the arterial switch procedure is the reestablishment of normal circulation, with the left ventricle acting as the systemic pump. Potential complications of the arterial switch include narrowing at the great artery anastomoses and coronary artery insufficiency.