

defects. Syndromes associated with heart defects include DiGeorge syndrome, a syndrome characterized by deletion of part of chromosome 22q11 (interrupted aortic arch, truncus arteriosus, tetralogy of Fallot, and posterior malaligned VSDs); Noonan syndrome (pulmonic valve anomalies and cardiomyopathy); Williams syndrome (aortic and pulmonic stenosis); and Holt-Oram syndrome (upper limb anomalies and atrial septal defect [ASD]). Extracardiac defects (such as tracheoesophageal fistula, renal abnormalities, and diaphragmatic hernia) are seen in association with heart anomalies.

Circulatory Changes at Birth

Blood carrying oxygen and nutritive materials from the placenta enters the fetal system through the umbilicus via the large umbilical vein. The blood then travels to the liver, where it divides. Part of the blood enters the portal and hepatic circulation of the liver, and the remainder travels directly to the inferior vena cava (IVC) by way of the ductus venosus. Oxygenated blood enters the heart by way of the IVC. Because of the higher pressure of blood entering the right atrium, it is directed posteriorly in a straight pathway across the right atrium and through the **foramen ovale** to the left atrium. In this way, the better-oxygenated blood enters the left atrium and ventricle to be pumped through the aorta to the head and upper extremities. Blood from the head and upper extremities entering the right atrium from the superior vena cava is directed downward through the tricuspid valve into the right ventricle. From there it is pumped through the pulmonary artery, where the major portion is shunted to the descending aorta via the **ductus arteriosus**. Only a small amount flows to and from the nonfunctioning fetal lungs ([Fig. 23-2, A](#)).