

To appreciate the physiology of heart defects, it is necessary to understand the role of pressure gradients, flow, and resistance within the circulation. As blood is pumped through the heart, it (1) flows from an area of high pressure to one of low pressure and (2) takes the path of least resistance. In general, the higher the pressure gradient, the faster the rate of flow; and the higher the resistance, the slower the rate of flow.

Normally, the pressure on the right side of the heart is lower than that on the left side, and the resistance in the pulmonary circulation is less than that in the systemic circulation. Vessels entering or exiting these chambers have corresponding pressures. Therefore, if an abnormal connection exists between the heart chambers (e.g., a septal defect), blood will necessarily flow from an area of higher pressure (left side) to one of lower pressure (right side). Such a flow of blood is termed a **left-to-right shunt**. Anomalies resulting in cyanosis may result from a change in pressure so that the blood is shunted from the right to the left side of the heart (**right-to-left shunt**) because of either increased pulmonary vascular resistance or obstruction to blood flow through the pulmonic valve and artery. Cyanosis may also result from a defect that allows mixing of oxygenated and deoxygenated blood within the heart chambers or great arteries, such as occurs in truncus arteriosus.

## Classification of Defects

There are typically two classification systems used to categorize congenital heart defects. Traditionally, cyanosis, a physical characteristic, has been used as the distinguishing feature, dividing anomalies into **acyanotic defects** and **cyanotic defects**. In clinical practice, this system is problematic because children with acyanotic defects may develop cyanosis. Also, more often, those with cyanotic defects may appear pink and have more clinical signs of HF.

A more useful classification system is based on hemodynamic characteristics (blood flow patterns within the heart). These blood flow patterns are (1) **increased pulmonary blood flow**; (2) **decreased pulmonary blood flow**; (3) **obstruction to blood flow** out of the heart; and (4) **mixed blood flow**, in which saturated and desaturated blood mix within the heart or great arteries. As a comparison, [Fig. 23-3](#) outlines both classification systems. With the