An isolated congenital anomalous origin of the coronary arteries is a rare disorder and is implicated in approximately 0.1 % of cases of congenital heart disease. An anomalous origin of the left coronary artery (LCA) arising from the pulmonary artery (PA) (the Bland-Ga...
rate in infancy. In rare cases, if the patient is able to survive the first year of life, the chances of survival until adulthood are good [3, 5]. Its frequency among adults submitted to coronary artery angiography for suspected ischemic heart disease is low. Among 1750 adults undergoing coronary angiography, 2 had the LCA originating from the PA [5].

The proximal coronary arteries develop from the peritruncal area and extend into the great vessels [3, 6]. The pulmonary buds involute and the aorta buds mature rapidly in coronary arteries. The anomalous coronary anatomy described here is probably due to abnormally persistent involution of these angioblastic buds. During the neonatal period, blood flows from the PA to the anomalous LCA in an anterograde fashion as a result of high pulmonary vascular resistance. At approximately 2 months a drop occurs in perfusion pressure in the PA and in the aberrant coronary artery, accompanied by a decline in oxygen saturation, which leads to retrograde flow through the anomalous LCA [7]. This may provoke myocardial infarction and deterioration of left ventricular function. Survival during the initial period of life depends on the development of collateral circulation between the right and the left coronary artery. However, this collateral circulation introduces a second pathophysiological mechanism, namely a “coronary steal” phenomena which can occur following a left-to-right shunt from the RCA to the PA via the aberrant LCA.

If clinical manifestations are well known in infants and include left heart failure or mitral valve regurgitation, symptoms are far more varied in adults; they include angina pectoris, dyspnea, syncope and sudden death. Patients may be asymptomatic and this anomaly usually becomes medically apparent when a precordial murmur is detected. Congestive heart failure is rare, but sudden death is common. Of 14 adults with this anomaly reviewed by George and Knowlan, 10 died suddenly, and of the 10, sudden death was the initial clinical manifestation of the coronary anomaly in eight [8].

Definitive diagnosis is generally made by angiography: while contrast is being injected into the RCA, the LCA is opacified by collaterals and the connection between the LCA and the PA can be visualized. It is impossible to visualize the LCA originating from the PA during the injection of contrast medium into the PA because of the reverse flow in the anomalous LCA. A se-