Cardiac Ischemia in Pediatric Patients

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KEYWORDS

• Congenital heart disease • Kawasaki disease
• Coronary artery anomaly • Coronary artery ostial stenosis

Children and teenagers are frequently brought to primary care physicians with complaints of chest pain. In the minds of the patients, their parents, and their physicians, thoughts of a cardiac event with an unpleasant outcome are often conjured up in such situations. If a child experiences chest pain while he or she is on the school grounds, the teachers and school officials may become alarmed, demanding an immediate medical consultation. There is prevailing fear of sudden death or cardiac disability among lay people as well as medical professionals. This fear of chest pain in a child is triggered by our vivid experience with an adult who suffered acute coronary syndrome or news reports of a high-profile case of sudden cardiac death in a young athlete. Atherosclerotic heart disease as a basis for myocardial ischemia in children is very rare. The great majority of chest pain experienced by children and teenagers are noncardiac in origin. Cardiac ischemia in children is usually not an isolated disease in an otherwise normally formed coronary artery, but is part of more complex congenital or acquired diseases. Myocardial infarction in a child is seldom manifested as classic pressure-like angina pectoris, but may take nonspecific symptoms such as unusual irritability, nausea and vomiting, abdominal pain, shocked state, syncope, seizure, or sudden unexpected cardiac arrest. Some patients may develop silent nonfatal infarction.[1] Although cardiac ischemia is not a frequent occurrence, it must be recognized as a serious, life-threatening event. Pediatricians must be aware of these conditions, and stand ready to take prompt and appropriate actions to avoid irreversible consequences. This article lists and characterizes major causes of cardiac ischemia in children, describes signs and symptoms of each, and provides therapeutic considerations.

DEFINITION AND BACKGROUND

The word ischemia is derived from two Greek roots: *ischō*, to keep back, plus *haima*, blood. Myocardial ischemia implies inadequate perfusion of the myocardium usually as a result of coronary artery obstruction anywhere along the course of the epicardial

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Pediatr Clin N Am 57 (2010) 1261–1280
doi:10.1016/j.pcl.2010.09.007 **pediatric.theclinics.com**
0031-3955/10/$ – see front matter

artery from the ostium in the aorta to the minute intramyocardial branches. Obstruction may be due to intrinsic narrowing of the vessel lumen due to thickening of the wall, presence of thrombus within its lumen, extrinsic compression of the vessel from a nearby structure, kinking or stretching of the artery itself, or abnormal vasoreactivity or spasm.

Typically, in a normal subject, there are two coronary arteries arising from the right- and left-facing sinuses of Valsalva. The right coronary artery (RCA) typically courses along the right atrioventricular groove adjacent to the tricuspid valve ring and reaches the posterior crux of the heart. It gives off branches sequentially to the sinus node, right atrium, right ventricle (RV), atrioventricular node, and posteroinferior wall of the left ventricle (LV) in a majority of patients (so-called right dominant pattern). The main trunk of left coronary artery (LCA) tunnels under the main pulmonary artery and, as it resurfaces, bifurcates into the left anterior descending artery (LAD) and the left circumflex artery (LCX). The LAD courses on the anterior surface of the LV along the attachment of the ventricular septum to the free wall, supplying blood to the LV anterior wall and about two-thirds of the ventricular septum. The LCX courses along the left atrioventricular groove just outside the mitral valve ring, and gives off a large branch to the lateral wall of the LV. In a minority of patients the LCX crosses the posterior crux of the heart and extends into the posterior descending artery (so-called left dominant coronary pattern).

Embryologically, primordial coronary vessels are formed by endothelial precursor cells migrating from the liver and form networks of channels along the differentiating epicardium of the heart tube. These primitive vessels penetrate into the myocardium. These ingrowing vessels merge, acquire smooth muscle coats, and transform themselves into arteries.[2] The main right and left arterial channels eventually connect to the aorta. In normal subjects, there are no well-developed connections (collateral arteries) linking the RCAs and LCAs. Intercoronary collaterals may develop rapidly, especially in young children, when one of the major arteries is blocked by disease process.

Coronary arteries provide oxygen and fuel (in the form of glucose and free fatty acid) to actively contracting myocardial cells. Because increased tension within the ventricular walls during systole impedes blood flow, most of the coronary blood flow occurs during diastole. Thus, the aortic diastolic pressure is an important determinant of coronary perfusion. Any pathologic condition that lowers the diastolic pressure, such as aortic insufficiency or presence of an abnormal run-off from the aorta (eg, patent ductus arteriosus or arterovenous fistula) tends to have a negative impact on coronary perfusion.

CLASSIFICATION

Coronary artery diseases which form the basis of myocardial ischemia in children may be classified in terms of cause. Major classes include (1) congenital anomalies of the coronary arteries, (2) coronary artery complications associated with congenital heart disease, (3) coronary artery sequelae of Kawasaki disease, and (4) myocardial ischemia associated with hypertrophic cardiomyopathy (**Box 1**).

CONGENITAL ANOMALIES OF THE CORONARY ARTERIES

Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA) or Bland-White-Garland Syndrome

This particular anomaly is most likely come to the attention of a primary care physician in an infant between a few weeks to 12 months of age (**Fig. 1**). There are a few patients with this anomaly who remain symptom-free and survive until adulthood. The