

Clinical Presentation and Diagnostic Approach to Isolated Congenital Coronary Anomalies.

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Introduction:

Isolated congenital coronary anomalies (ICCA) are rare malformations affecting the pediatric population and can be isolated or associated with other heart diseases. Its clinical presentation varies widely from sudden cardiac death to heart murmur or absence of symptoms, being a casual diagnosis. The aim of this study is to analyse the initial approach to ICCA. The clinical spectrum is assayed according to each anomaly and the capacity of electrocardiographic and echocardiographic assessment to establish the diagnosis.

Methods:

Retrospective monocentric study including the patients under 18 years registered with the diagnosis of ICCA from 1996 to 2017.

Results:

ICCA were diagnosed in 27 patients. Anomalous origin of left coronary artery from pulmonary artery (ALCAPA) was the commonest anomaly (41%) followed by right coronaries from left sinus (22%) and fistulas to right ventricle (19%).

The clinical onset with life-threatening symptoms (cardiac failure or cardiac arrest) determines an affection on left coronary artery, the 10 patients admitted due to heart failure being diagnosed of ALCAPA later. Anomalies affecting the right artery are usually clinically silent or appear when studying a heart murmur or chest pain with normal EKG. An association between the presence of a murmur and right ventricle fistulas is described.

At the time of diagnosis, 37% of the patients had electrocardiographic signs of myocardial infarction, appearing in 73% of patients with subsequent ALCAPA.

Echocardiography determined the diagnosis globally in 85% of the cases and 73% of the patients with ALCAPA (left coronary reversal flow being the main echocardiographic marker). Regarding ALCAPA, the diagnostic value of mitral insufficiency, left ventricle dilatation is presented, as well as a decrease in the ejection fraction at diagnosis. In 52% cases, another imaging study was performed to accurate the diagnosis (22% CT scan and 37% coronariography).

Conclusions:

A great clinical variety is described, with higher clinical severity in the cases affecting the left coronary artery. The use of echocardiography alone to diagnose coronary anomalies is reliable in most cases. Even so, there are situations, especially in the presence of severe symptomatology, in which performing other techniques to reach the diagnosis is required.