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REVIEW ARTICLE

Congenital coronary artery anomalies☆

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Abstract Congenital coronary artery anomalies are abnormalities in the origin, course or structure of these arteries, and their incidence varies from 0.2% to 5.6%. Although the majority of anomalies are asymptomatic, they are the second most common cause of sudden cardiac death in young athletes.

The aim of this study is to highlight the main anomalies with hemodynamic significance, particularly anomalous aortic origin of coronary arteries and anomalous left coronary artery from the pulmonary artery.

Anomalous aortic origins of coronary arteries account for 14–17% of all sudden cardiac deaths that unexpectedly occur in healthy children or young athletes during or immediately after physical exercise. The mechanism responsible for the compression/occlusion of the coronary artery originating from the opposite sinus is still unclear and several mechanisms have been proposed. The clinical presentation of these patients varies and physical examination is normal in most cases. Transthoracic echocardiography is the most commonly used test for diagnosis. The treatment, management and follow-up of these patients are the subject of debate.

The anomalous origin of the left coronary artery arising from the pulmonary artery is an even rarer condition and, without corrective surgery, most patients die within the first year of life. Echocardiography is also the method of choice to confirm this condition. The diagnosis of this anomaly in a seriously ill child is an indication for urgent surgery.

Due to the hemodynamic abnormalities caused by these abnormalities, early diagnosis and treatment are important.

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PALAVRAS-CHAVE
Anomalias das artérias coronárias;
Anomalias da origem das artérias coronárias na aorta;
Anomalias da origem das artérias coronárias na artéria pulmonar;
Morte súbita de origem cardíaca

Anomalias congénitas das artérias coronárias

Resumo As anomalias congénitas das artérias coronárias são alterações da sua origem, trajeto ou estrutura e a sua incidência varia entre 0,2 e 5,6%. Apesar de a maioria ser assintomática, são a segunda causa morte súbita de origem cardíaca em jovens atletas.

O objetivo deste trabalho é salientar as principais anomalias com repercussões hemodinâmicas, nomeadamente as anomalias da origem das artérias coronárias na aorta e as anomalias em que a artéria coronária esquerda tem origem na artéria pulmonar.

As anomalias da origem das artérias coronárias na aorta correspondem a 14-17% de todas as mortes súbitas de causa cardíaca, que ocorrem inesperadamente em crianças saudáveis ou jovens atletas durante ou imediatamente após o exercício físico. O mecanismo responsável pela compressão/oclusão da artéria coronária com origem no seio coronário oposto ainda não é claro e existem vários mecanismos propostos. A apresentação clínica desses doentes é bastante variável e o exame físico é na maioria das vezes normal. O ecocardiograma transtorácico é o exame mais usado para o diagnóstico. A abordagem, o tratamento e o seguimento desses doentes é ainda um tema controverso.

A origem anómala da artéria coronária esquerda na artéria pulmonar é ainda mais rara e a maioria dos doentes morre no primeiro ano de vida, se não for feita correção cirúrgica. O ecocardiograma é também o método de eleição para a sua confirmação. O diagnóstico dessa anomalia numa criança, gravemente doente, é indicação cirúrgica de urgência.

Pelas alterações hemodinâmicas que acarretam, o seu diagnóstico e tratamento precoce são importantes.

Introduction

Congenital coronary artery anomalies (CCAAs) were first described two millennia ago by Galen and Vesalius[1] and are abnormalities in the origin, structure, and course of these arteries.[2]

In fact, this topic has become increasingly well-known due to its association with sudden death,[3] even though most CCAAs are benign, with no hemodynamic or prognostic implications.[4]

Coronary artery disease is one of the leading causes of morbidity and mortality worldwide. Among these, CCAAs, although less prevalent, are a potential source of malignant arrhythmias, ischemia and myocardial dysfunction.[3] In young athletes, CCAAs are the second most common cause of sudden cardiac death (in 12% of deaths), and are generally triggered by vigorous physical exercise.[4,5] Sudden cardiac death can be defined as an unexpected death that is cardiac in origin. It generally occurs less than one hour after the onset of symptoms in an individual with no previously known fatal condition.[4] According to a study based on the Minneapolis Heart Institute Foundation's records, the two most common causes of sudden death were hypertrophic cardiomyopathy at 36% and CCAAs at 17%.[6]

Although most CCAAs are asymptomatic and involve no hemodynamic complications, some CCAAs are clinically significant.[7] The latter are classified into two subgroups: anomalous origin of coronary arteries from the opposite sinus (left coronary artery originating from the right coronary sinus and right coronary artery originating from the left coronary sinus) and anomalous left coronary artery from the pulmonary artery (ALCAPA).[3] These anomalies are associated with sudden death. However, there are other less common CCAAs that may also be related to sudden cardiac death. These include a single coronary artery with an interarterial course, atresia of the coronary ostium or artery fistulas.[4,6,8]

In the presence of CCAAs, sudden death is related to myocardial ischemia resulting in malignant ventricular arrhythmias. The physiological demands of physical activity depend on the type of exercise. The two types of exercise (isotonic and isometric exercise) involve the use of large muscle masses that increase venous return and left ventricular end-diastolic volume. Along with stimulating the sympathetic adrenergic system, said masses increase heart rate, blood pressure, cardiac output and myocardial contractility. The aim of these responses is to increase the oxygen supply to the myocardium with an increased blood flow, which in the context of these anomalies is compromised.[6,9,10]

These anomalies are more common in individuals with congenital heart disease (particularly tetralogy of Fallot, transposition of the great arteries and some forms of pulmonary atresia) than in individuals with a structurally normal heart. Symptoms usually occur earlier, as does the diagnosis. This affects the treatment and prognosis of the underlying disease.[7,11]

The real incidence of CCAAs in the general population remains unclear, ranging from 0.2% to 5.6%. This variation depends on the method (autopsy vs. angiography), the diagnostic criteria and the population studied.[4,12,13] There are