Cardiology in the Young

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Anomalous origin of a single coronary artery from the pulmonary artery associated with patent ductus arteriosus

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Brief Report

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Abstract

We report an unusual case of a 12-month-old boy diagnosed with anomalous origin of a single coronary artery from the pulmonary artery associated with patent ductus arteriosus. The patient survival was attributed to left-to-right shunt (patent ductus arteriosus) allowing for appropriate myocardial perfusion. Successful surgical correction involved patent ductus arteriosus closure, mitral annuloplasty and reimplantation of the coronary artery into the aortic root.

Anomalous origin of a single coronary artery from the pulmonary artery is a rare and haemodynamically significant congenital heart malformation, which can lead to myocardial ischaemia or infarction and sudden cardiac death.[1] Its actual incidence is not known, but it is rarer than anomalous left coronary artery from the pulmonary artery (1/300,000 births[2]) and anomalous right coronary artery from the pulmonary artery (1/50,000 births[3]).

Published case reports of patients with anomalous origin of a single coronary artery from the pulmonary artery suggest cardiac ischaemia is usually present prior to diagnosis, which is often established on autopsy.[4] Main clinical signs are similar to those of anomalous left coronary artery from the pulmonary artery and include dilated cardiomyopathy, ischaemic mitral valve regurgitation, or hyperechogenic papillary muscles.[5] Diagnosis is complex owing in part to the low incidence of the condition but can be suspected on the electrocardiogram and subsequently confirmed by echocardiography or CT scan. Surgical treatment involves the reimplantation of the single coronary artery into the aortic root.[5]

Anomalous origin of a single coronary artery from the pulmonary artery is typically associated with other cardiovascular defects,[6–8] the number and severity of which determine the clinical course. It can also occur as an isolated disease leading to early death during the first year.[1]

We describe here a particular case of isolated anomalous origin of a single coronary artery from the pulmonary artery with unusually preserved left ventricular function and prolonged survival attributed to a patent ductus arteriosus.

Case report

A 12-month-old-boy (7.7 kg and 77 cm) was referred to our hospital from his home country with a diagnosis of anomalous left coronary artery from the pulmonary artery (based on transthoracic echocardiography and cardiac catheterisation). The patient presented with growth retardation, dyspnea, and asthenia. Physical exam disclosed regular heart sounds and a left parasternal systolic ejection murmur. The electrocardiogram revealed sinus rhythm and Q-waves in lateral and lower leads. The echocardiogram showed a well-preserved left ventricular function (left ventricular ejection fraction: 67%), a severely dilated left ventricle (left ventricular end-diastolic diameter: 41 + 6.9 mm, mean ± SD), as well as patent ductus arteriosus, 3.5 mm in diameter, with left-to-right shunting. However, typical features of anomalous left coronary artery from the pulmonary artery such as severe ischaemic mitral valve regurgitation, hyperechogenic papillary muscles or left ventricular dysfunction were not observed. Unexpectedly, the right coronary artery was not found at the usual location and the left coronary artery was seen originating from the postero-inferior sinus of Valsalva of the pulmonary artery (Fig 1). It splits from a short common trunk into three vessels, one of which oriented towards the right atrioventricular groove. These findings raised the suspicion of a single coronary artery arising from the pulmonary artery. Haemodynamical tolerance and survival could be explained by the patent ductus arteriosus which maintained a higher pressure of oxygenated blood in the pulmonary artery and hence a perfusion of the single coronary artery.

Given the patient's age and the cardiopathy severity, a cardiac CT scan under general anaesthesia was deemed dangerous. Diagnosis was confirmed intraoperatively (Fig 2), when it became

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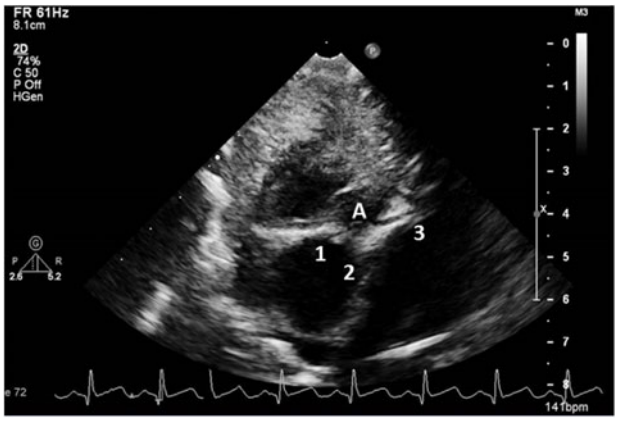

Figure 1. Parasternal short axis view of three coronary vessels originating from the main pulmonary artery. A: Main pulmonary artery; 1: right coronary artery; 2: circumflex artery; 3: left anterior descending artery.

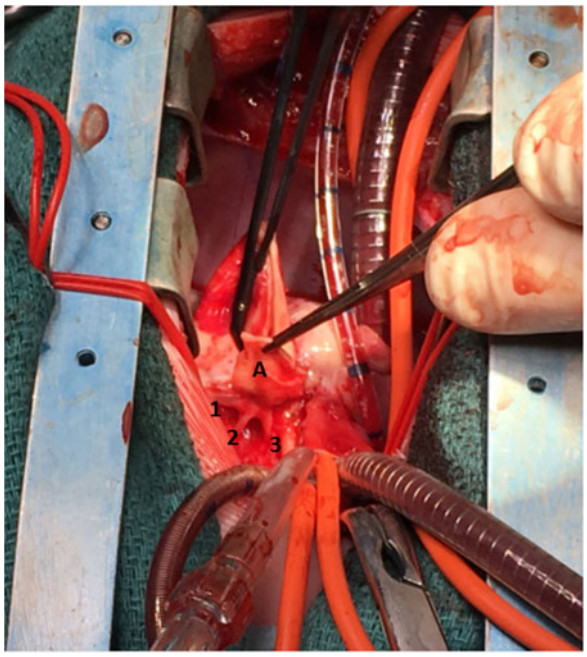

Figure 2. Surgical view after main pulmonary artery opening, splitting off into three coronary vessels. A: Main pulmonary artery; 1: right coronary artery; 2: circumflex artery; 3: left anterior descending artery.

apparent that the main pulmonary artery splits into the right coronary artery, circumflex artery, and left anterior descending artery. The patient underwent a surgical correction of the defect which consisted in ligation and division of the ductus arteriosus, reimplantation of the anomalous coronary artery into the posterior sinus of the aortic root, and in a mitral valve annuloplasty (with resorbable stitches). To obtain a safe myocardial protection, cardioplegia has been delivered in the aortic root and in the pulmonary trunk, after ligation of the ductus and occlusion of both pulmonary arteries. The post-operative course was uneventful; the left ventricle remained dilated although the left ventricular end-diastolic diameter decreased (37 + 5 mm), the left ventricular function was stable (left ventricular ejection fraction: 68%), and an intermittent mitral micro-leak was recorded. The patient was discharged 1 week after the procedure.

Discussion

Anomalous origin of a single coronary artery from the pulmonary artery is a rare disease, which is often diagnosed post-mortem by autopsy[4]. The diagnosis in living patients may be complicated by the presence of other cardiac congenital malformations. Echocardiography is helpful to determine the origin of the coronary arteries, but CT scan and cardiac catheterisation provide definitive diagnosis and an accurate appreciation of the anatomy of the coronary artery system, which is essential for successful surgical repair.

When anomalous origin of a single coronary artery from the pulmonary artery is present, blood delivery to the heart originates from the pulmonary artery. As pulmonary vascular resistance falls in the neonatal period, coronary perfusion decreases, resulting in ischaemia. Therefore, the condition is usually fatal unless adequate myocardial perfusion is achieved by an associated cardiac defect responsible for an increase in perfusion pressure and in pulmonary arterial oxygen saturation.[6] The most common defects are atrial septal defect, ventricular septum defect,[8] conotruncal defects (tetralogy of Fallot and truncus arterosius), and aorta coarctation.[7] In the case of our patient, the associated patent ductus arteriosus was the only left to right shunt sustaining the patient tolerance to a usually deadly condition. Other rare cases of long-term survival due to a defect allowing for near normal perfusion pressure have been reported, including a 39-year-old man with persistent truncus arteriosus[9] and a 7-year-old girl with a large ventricular septal defect.[10]

The surgical treatment of anomalous origin of a single coronary artery from the pulmonary artery has been associated with poor outcome and its success critically depends on a timely diagnosis, as well as appropriate pre- and perioperative management.[4] One of the anaesthetic challenges is to maintain coronary perfusion and myocardial contractility by keeping elevated pulmonary vascular resistances.[5] Correct delivery of cardioplegia to the myocardium is also crucial and is best achieved with an antegrade flow in both coronary arteries. Surgery was successful, neither complication nor post-cardiotomy syndrome was reported during the post-operative course and the patient could return to his home country.

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Conflicts of interest. None.

Ethical standards. The authors assert that all procedures contributing to this work comply with the ethical standards of the national guidelines and local regulations (Monegasque Law n°1.265 of 23 December 2002 on the protection of individuals with regard to the biomedical research) and with the Helsinki Declaration of 1975 as revised in 2008.

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