Case report

Hypoplastic left heart syndrome with anomalous origin of left coronary artery from the right pulmonary artery: successful surgical treatment in a neonate

Matej Nosál*, Ikenna Chima Omeje, Rudolf Poruban

Department of Cardiac Surgery, Children's Heart Centre Slovak Republic, Limbova 1, 833 40 Bratislava, Slovak Republic

Received 17 March 2005; received in revised form 31 May 2005; accepted 13 June 2005

Abstract

Anomalous origin of the left coronary artery from the right pulmonary artery in association with hypoplastic left heart syndrome is a rare congenital anomaly. We describe a successful simultaneous surgery for both anomalies during the first stage palliation in a neonate.

Keywords: Congenital heart disease; Hypoplastic left heart syndrome; Coronary artery anomaly

1. Introduction

Coronary artery anomalies in hypoplastic left heart syndrome are a relatively rare finding. Changes in course and wall thickness of coronary arteries have been described in some variants of hypoplastic left heart syndrome [1]. In very rare cases, anomalous origin of coronary arteries may occur [2–5]. Anomalous origin of the left coronary artery from the right pulmonary artery in patients with hypoplastic left heart syndrome has been described in the literature, but to our knowledge, no successful simultaneous surgery for both anomalies within the first stage palliation has been reported in a neonate [2–5].

2. Case report

A neonate born in the 38th week of gestation, weight at birth 3000 g was referred to the Children’s Heart Centre at the age of 12 days with signs of multiple organ dysfunction, tachyypnoe and failure to thrive. Echocardiography confirmed a variant of hypoplastic left heart syndrome with a 6 mm mitral valve, hypoplastic left ventricle and a 4 mm aortic anulus with a bicuspid valve. There was a nonrestrictive shunting at the atrial level and mild tricuspid regurgitation. The coronary anatomy was considered to be normal (Fig. 1). After initial stabilisation, the patient was referred for first stage surgical palliation at the age of 18 days. During surgery, arterial and venous cannulae were placed into the ascending aorta and the right atrium, respectively, and the patient cooled to deep hypothermia. Upon initiation of cardiopulmonary bypass, there was a visible discouloration of the left ventricular myocardium. A revision of the origin of the coronary arteries revealed an anomalous origin of the left coronary artery from the proximal part of the right pulmonary artery, coursing across the roof of the left atrium towards the interventricular groove. We immediately instituted selective perfusion of the main pulmonary artery through an additional cannula with both pulmonary branches snugged, hence ensuring adequate perfusion of the left myocardium. During the cooling phase, cardioplegic solution was administered into the aorta and the main pulmonary trunk, which was subsequently transected. The left coronary artery was excised from the proximal part of the right pulmonary artery with a sufficient button of surrounding tissue and mobilised over a short section. Due to its course, the anomalous coronary artery was directly reimplanted into the posterior sinus of the main pulmonary trunk (Fig. 2). The pulmonary confluence was enlarged with a patch of native pericardium. Under circulatory arrest, the aortic arch was reconstructed by a modified technique using the native pulmonary trunk [6]. The interatrial septum was resected. On reinstitution of cardiopulmonary bypass, we observed normal perfusion of the left myocardium. A 3.5 mm modified aortopulmonary shunt was constructed on the right side. The patient was subsequently weaned from bypass on a low dose of inotropic agents and the chest was closed. The patient was discharged on the 14th postoperative day and underwent the second stage palliation—the bidirectional Glenn procedure at the age of 7 months.
3. Comment

Anomalous origin of the left coronary artery from the right pulmonary artery in hypoplastic left heart syndrome is mostly undetected preoperatively due to its low incidence, thus increasing the difficulty and complexity of the first stage palliation. Till date, only two cases of surgical treatment of this anomaly have been reported in the literature. One of these cases involved a successful coronary transfer in a 6-month-old patient with anomalous origin of the left coronary artery from the right pulmonary artery, which was diagnosed preoperatively [3]. The second case was in a neonate who died shortly after the operation [4]. In both cases, the coronary arteries were transferred into the native aorta. In our case, early perioperative manifestation of the anomalous origin of the left coronary artery was enhanced by direct cannulation of the ascending aorta, hence it was possible to apply selective perfusion and cardioplegia of the coronary artery prior to the coronary transfer. It also enabled direct coronary transfer into the pulmonary trunk during the cooling phase, without tension on the coronary artery and at the same time ensured a good surgical exposure. Although there is a low incidence of this anomaly, it is advisable to preoperatively verify the origin of coronary arteries in patients with hypoplastic left heart syndrome by echocardiography and in doubtful cases by angiography. At the same time it is expedient to check the coronary anatomy at the start of operation. It is also our opinion that implanting the coronary artery into the main pulmonary trunk under direct surgical view may facilitate the transfer and ensure a proper position of the coronary artery without tension.

References