Case report

Surgical repair of coronary sinus orifice atresia

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Abstract

We report a 10-month-old boy who underwent a bi-directional Glenn procedure and repair of coronary sinus orifice atresia. The left superior vena cava was the only vessel communicating with the coronary sinus in this case. The coronary sinus was allowed to communicate freely with the left atrium by creating a partially unroofed coronary sinus using a left superior vena cava flap.

Keywords: Coronary sinus; Surgery

1. Introduction

Coronary sinus orifice atresia is usually diagnosed at autopsy. This defect is an intrinsically benign anomaly because it is usually associated with unroofed coronary sinus or unobstructed retrograde drainage via left superior vena cava (SVC). However, if left SVC is the only coronary venous drainage vein, coronary venous hypertension may progress after Glenn or Fontan procedure without division of the left SVC. We performed a bi-directional Glenn procedure and simultaneous repair of this anomaly with division of the left SVC.

2. Case report

A 10-month-old boy was referred to our hospital due to severe cyanosis. Right modified Blalock-Taussig shunt had previously been performed for pulmonary atresia, atrioventricular discordance, ventricular septal defect, and hypoplastic left ventricle. Cardiac catheterization revealed a narrow left SVC. Left SVC venography demonstrated absence of the CS orifice, with retrograde flow into the innominate vein (Fig. 1). As the patient was a good candidate for cavopulmonary shunt, right-sided Glenn procedure and division of the left SVC with repair of the CS orifice atresia were planned.

Immediately after median sternotomy, an episode of paroxysmal supraventricular tachycardia occurred; heart rate was 220 and hemodynamic compromise was evident. However, cardioversion resulted in return to sinus rhythm.
After establishing cardiopulmonary bypass with bicaval cannulae, the right Blalock-Taussig shunt was ligated and divided. The right SVC was divided and a bi-directional Glenn procedure was performed. The left anterior aorta was retracted rightwards for adequate visualization, exposing the narrow left SVC, which continued to the slightly dilated CS on the LA. The stenotic segment of the left SVC was measured to be approximately 3.5 mm in diameter. Repair of CS orifice atresia was performed with cold crystalloid cardioplegic cardiac arrest (Fig. 2). The left anterior aorta was pulled rightwards to expose the left SVC and CS. The left SVC was ligated superiorly and divided. The proximal end of the left SVC joining the CS was then cut into the common wall of the CS and left atrium (LA), and this opening was enlarged inferiorly to extend into the CS and upwards to the LA wall. The roof of the CS was fenestrated inferiorly so that the CS communicates freely with the LA. The edges of the incision in the CS roof were oversewn with 7-0 suture material, taking relatively small bites to avoid a purse-string effect. The opening in the LA was closed with a left SVC flap. The suture line was finally completed with either a few interrupted stitches or with a 7-0 continuous suture. After the aortic cross-clamp was removed, sinus rhythm resumed with good cardiac contractions.

The patient recovered uneventfully and remains well at follow-up three months after surgery. Postoperative Doppler echocardiography showed undiminished flow at the new CS orifice and good ventricular wall motion.

3. Discussion

Atresia of the CS orifice is usually associated with an unroofed CS [1]. In this situation, cardiac venous blood returns to the left atrium without obstruction. CS orifice atresia associated with persistent left SVC in the absence of left atrial connection is a very rare cardiac anomaly [2]. The left SVC is the only vessel in communication with the coronary sinus in half the cases with this anomaly. CS orifice atresia with unobstructed retrograde drainage via left SVC is an intrinsically benign anomaly [1]. However, preoperative recognition and surgical intervention for this anomaly are necessary if ligation or division of the left SVC is indicated. Even if the left SVC remains, special care should be taken during median sternotomy, which may stretch and obstruct the narrow left SVC, and during direct cannulation of the left SVC. Interruption of coronary sinus drainage may lead to poor cardiac contractions [2,3] and can even have lethal consequences [4]. Moreover, cardiac dysfunction due to myocardial perfusion problems with resulting coronary venous hypertension may progress after Glenn or Fontan procedure is performed without division of the left SVC [5].

Surgical treatment for this anomaly has rarely been reported. Two options have been described: anastomosis of a connecting vein with the CS to the LA [3] and a creation of a new connection from the interior of the LA using a guide probe advanced through the open end of the left SVC [5]. Our method is not only a simple unroofing of the CS, but also creates a smooth, large opening between the CS and the LA using a left SVC flap and oversewing the edges of the CS roof incision. Additionally, it is appropriate for small infants in whom simple unroofing is hazardous on account of the diminutive LA and absence of the landmark of the atrioventricular node.

References