Successful surgical treatment of a gigantic congenital coronary artery fistula immediately after birth

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Abstract

A foetus was prenatally diagnosed with a gigantic (12 mm) coronary artery fistula (CAF) from the left anterior descending (LAD) coronary artery to right ventricular apex at 38 weeks of gestation. LAD was dilated to 10 mm with partial aneurysmal changes. Because of concern for sudden ischaemic cardiogenic shock soon after birth, the child was electively delivered by caesarean section, with surgical fistula closure subsequently performed 1 h after birth. We also highly suspected the presence of a clinically significant accessory diagonal branch just around the fistula, thus direct fistula closure from outside the heart without cardiopulmonary bypass was abandoned and cardiopulmonary bypass was initiated. The terminal end of LAD was carefully opened, and the fistula was directly closed with four pairs of 6-0 polypropylene mattress sutures under cardioplegic arrest, while the opened terminal end of LAD was also repaired with plegetted 6-0 polypropylene mattress and over-and-over sutures. After 4 days of post-surgical extracorporeal life support for over-systemic pulmonary hypertension, the patient recovered without complications. Although postoperative echocardiography 5 months after the operation showed normal cardiac function without ventricular asynergy, the dilated and aneurysmal LAD remained unchanged.

Keywords: Coronary artery fistula • Newborn • Coronary artery aneurysm

INTRODUCTION

With recent improvements in foetal cardiac Doppler colour flow mapping, coronary artery fistula (CAF) is often diagnosed during the prenatal period, and some newborns require surgical or catheter-based intervention in neonatal period [1–4]. The optimal timing and the method of treatment depend on the size and location of communication between the coronary artery and the cardiac chamber. However, surgical intervention for cardiopulmonary collapse immediately after birth is required in some cases. We report a case of a gigantic CAF from left anterior descending (LAD) coronary artery to right ventricular apex, for which we successfully performed surgical CAF closure under cardiopulmonary bypass within 1 h after birth.

CASE

Foetal echocardiography at 38 weeks of gestation revealed a dilated and partial aneurysmal LAD (8–10 mm), communicating with the right ventricular apex through a 12-mm wide orifice (Fig. 1A). Sudden cardiopulmonary collapse soon after birth caused by severe coronary steal condition along with a physiological decrease in pulmonary arterial pressure was highly anticipated, thus elective caesarean (C)-section procedure and subsequent surgical repair were planned.

The child was delivered by elective C-section at 40 weeks of gestation with a birth weight of 3564 g, and then the surgical repair was started 50 min after birth. Using a median sternotomy, the dilated, aneurysmal and tortuous LAD was identified, and the distal portion was found to be in communication with the right ventricle (RV) apex (Fig. 2A and B). We attempted to compress the terminal end of LAD to assess whether the direct closure of CAF from the outside was possible. However, ST waves on the II and V5 leads elevated and blood pressure decreased, which indicated that a clinically significant accessory diagonal branch was arising from nearby fistula. Hence, we initiated cardiopulmonary bypass. Antegrade cardioplegia was then infused after aortic cross-clamping by pressing the fistula with a finger.

The aneurysmal terminal end of LAD was incised open under cardioplegic arrest and a 12-mm wide fistula was identified. We closed the orifice of fistula with four pairs of 6-0 polypropylene mattress sutures (Fig. 2C), then repaired the opened LAD using 6-0 polypropylene mattress sutures with fresh autopericardial strips, which were reinforced with 5-0 polypropylene continuous sutures.

After weaning from cardiopulmonary bypass, systolic pulmonary arterial pressure was 48 mmHg and systemic systolic blood pressure was 37 mmHg. Thus, extracorporeal life support was initiated to treat the over-systemic pulmonary hypertension and then successfully removed 4 days later. The postoperative course was uneventful and the patient was discharged at 2 months of age without any symptoms. Five months after surgery, echocardiography showed that both ventricles were normally sized and functioning, without any ventricular wall motion asynergy and a
tiny residual shunt from the LAD artery to the RV. On the other hand, cardiac computed tomography revealed that the dilated and aneurysmal LAD had not regressed (Fig. 1B). Presently, Coumadin administration is being continued with a target prothrombin time–international normalized ratio of 1.5, according to the protocol for the treatment of coronary artery aneurysm associated with Kawasaki disease.

COMMENT

A CAF is known to range from asymptomatic with spontaneous closure to a life-threatening condition. Although rare, it should not be overlooked that some patients in this anatomical group develop sudden heart and/or respiratory failure soon after birth because of a severe form of the lesion. In such cases, only quick closure of the fistula allows the survival of the affected newborn [5].

A result of the presence of such a fistula is that physiological pulmonary hypertension is severe and surgical intervention might be delayed. In our experience, the coronary artery connecting CAF can have some stenotic portions that regulate blood flow shunted from the coronary artery to cardiac chambers. However, the present patient had a non-obstructed rather than an excessively dilated LAD coronary artery. In fact, the diameter of LAD was much larger than that of ascending aorta.

![Figure 1:](image1)

**Figure 1:** (A) Foetus echocardiography showing the dilated and partially aneurysmal LAD coronary artery, communicating with the right ventricle at the apex through a 10-mm wide fistula (white arrows). (B) Three-dimensional computed tomography image obtained at 5 months after the operation showing that the dilated and aneurysmal LAD coronary artery has not regressed.

![Figure 2:](image2)

**Figure 2:** (A) Intraoperative macroscopic image and (B) related illustration showing the dilated and aneurysmal LAD coronary artery (white arrows). (C) An incision was made on the aneurysmal terminal end of LAD to explore the 12-mm wide CAF. The CAF was then closed with four pairs of 6-0 polypropylene mattress sutures. RV, right ventricle; LV, left ventricle; Ao, aorta; PA, pulmonary artery.
previously published reports have noted such a CAF, thus we were concerned that an unexpected systemic and/or coronary steal condition might develop. As a result, we decided to perform surgical intervention immediately after finding the ECG change. We believe that undertaking the urgent operation soon after birth was justified and, along with assistance from the collaborating medical team, greatly contributed to the rapid successful treatment of the present patient. The fate of the dilated and aneurysmal LAD remains unclear. Because of the risk of rupture or thrombus formation, long-term follow-up examinations and continual anticoagulation therapy are mandatory [5].

In summary, we treated a newborn with a gigantic CAF from LAD to RV with successful surgical closure within 1 h after birth. Cardiopulmonary bypass and cardioplegic arrest were required so as to not injure the branching diagonal coronary artery originating from the nearby fistula. Postoperative echocardiography findings revealed normal cardiac function without ventricular asynergy, while computed tomography showed that dilation and aneurysmal change of LAD was not regressed.

**Conflict of interest:** none declared.

**REFERENCES**


