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Case Report

Surgical Management of a Large Right Coronary Artery to Left Ventricle Fistula in a Neonate: A Case Report

Yu Ri Lee, M.D.[1], Eun Seok Choi, M.D., Ph.D.[2]

[1]Department of Thoracic and Cardiovascular Surgery and [2]Division of Pediatric Cardiac Surgery, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Korea

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Corresponding author
Eun Seok Choi
Tel 82-2-3010-3587
Fax 82-2-3010-6966
E-mail gninihs@naver.com
ORCID
https://orcid.org/0000-0002-0618-4686

A fistula from the right coronary artery draining into the left ventricle is a rare form of coronary artery fistula. Here, we describe the case of a symptomatic neonate with a large fistula of this type. The neonate was successfully treated with surgical closure of the fistula.

Keywords: Coronary artery fistula, Left ventricle, Neonate, Case report

Case report

Coronary artery fistulas are rare congenital coronary artery anomalies, among which fistulas from the right coronary artery (RCA) draining to the left ventricle (LV) are extremely rare. Here, we describe the case of a symptomatic neonate with a large RCA fistula draining to the LV. The patient was successfully treated with surgical management. This study was approved by the Institutional Review Board of Asan Medical Center (IRB number: S2022-0493-0001; IRB approval date: April 20th, 2022), and the need for informed consent was waived because of the retrospective nature of the study.

The male patient whose coronary artery fistula was detected on prenatal ultrasonography was born at 39 gestational weeks. Postnatal echocardiography demonstrated an enlarged RCA with a proximal diameter of 6.1 mm, which was directly connected to the base of the LV (Fig. 1). An LV aneurysm was identified on the basal posteroinferior side into which the fistula drained. No paradoxical motion was observed on the aneurysm. A 12-lead electrocardiogram and cardiac enzyme marker test showed no abnormal findings. Heart computed tomography confirmed that the LV aneurysm was round and measured 13.3×12.5 mm, and the dilated, tortuous RCA fistula opened immediately right and superior to the neck of the LV aneurysm (Fig. 2). Congestive heart failure symptoms, including tachypnea, developed at 9 days after birth because of diastolic run-off through the fistula. Although inotropic infusion and high-flow nasal cannula therapy were initiated, symptoms persisted. At that point, surgical closure of the fistula was planned.

Surgery was performed at the age of 19 days, and the patient weighed 3.5 kg. After median sternotomy, a piece of the pericardium was harvested for closure of the fistula. The enlarged RCA was observed arising from the anterior side of the aorta and connecting to the base of the LV (Fig. 3). Conventional cardiopulmonary bypass was initiated with the ascending aorta and bicaval cannulation. After aortic cross-clamping, an antegrade infusion of del Nido cardioplegic solution was performed. During cardioplegic infusion, the fistula was compressed using a finger to prevent cardioplegic steal into the LV through the fistula. A longitudinal incision was made along the distal RCA near the fistula. No normal branch of the RCA was identified near the fistula opening. The LV-side opening of the fistula was then exposed and closed with an autologous pericardial patch using a 7-0 polypropylene continuous suture. The smooth surface of the pericardium faced the coronary artery. The incised RCA was closed with a 6-0 polypropylene

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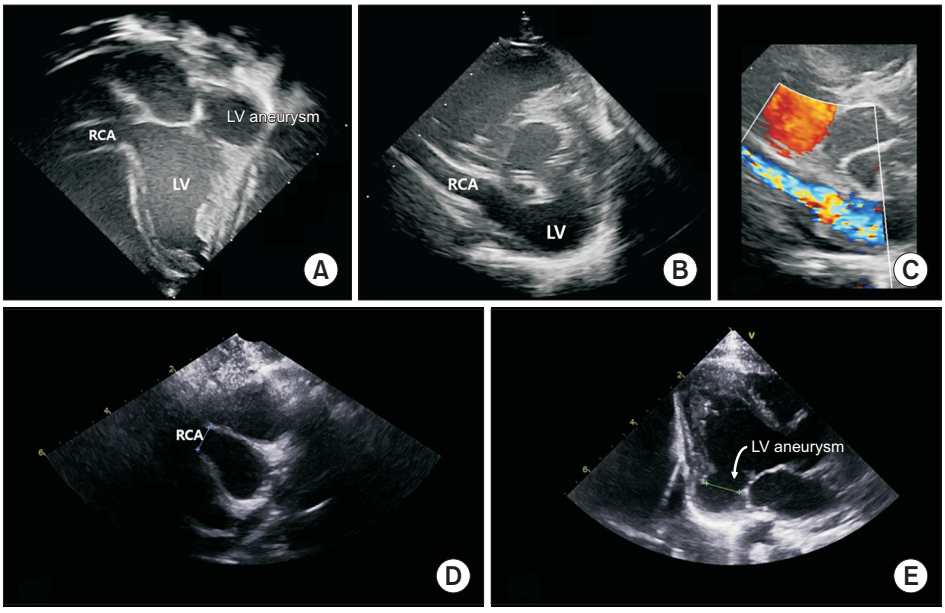

Fig. 1. (A–C) Preoperative transthoracic echocardiography. (A) The apical 4-chamber view shows a dilated right coronary artery (RCA) and left ventricle (LV) aneurysm. (B) The parasternal short-axis view shows the distal RCA draining to the base of the LV. (C) Color Doppler image visualizing the direction of blood flow from the distal RCA to the LV. (D, E) Postoperative transthoracic echocardiography at a 4-month follow-up. (D) In the parasternal short-axis view, the proximal RCA was measured as 6.28 mm. (E) In the apical 4-chamber view, LV aneurysm was measured as 11.3×8.1 mm (arrow).

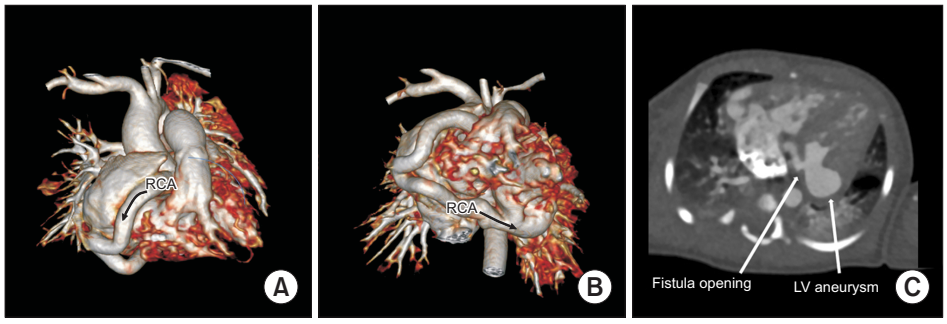

Fig. 2. Preoperative heart computed tomography. (A) A three-dimensional (3D) reconstruction shows the dilated right coronary artery (RCA) with a proximal diameter of 6.1 mm (arrow). (B) A 3D reconstruction shows the distal RCA (arrow) terminating into the base of the left ventricle (LV). (C) In the axial plane, the LV aneurysm measured 13.3×12.5 mm and the RCA fistula opened immediately right and superior to the neck of the LV aneurysm.

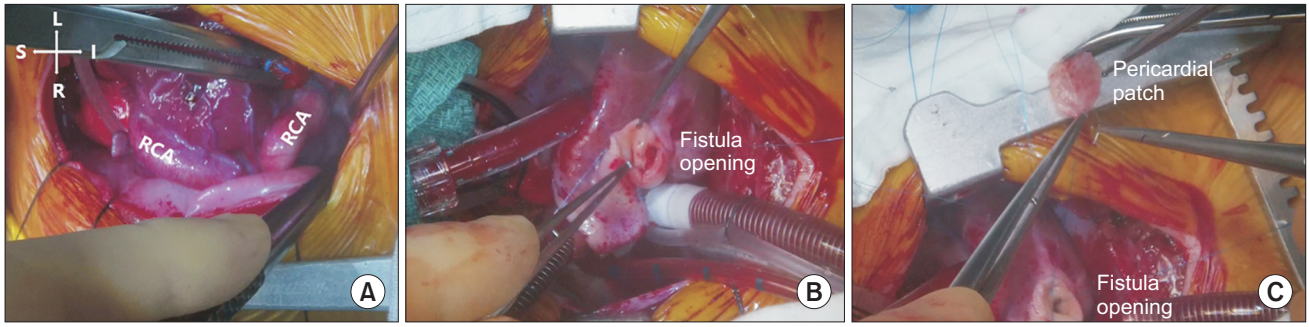

Fig. 3. Operative photographs. (A) The enlarged and tortous right coronary artery (RCA). (B) The opening of the fistula was exposed through the incision of the distal RCA. (C) Closure of the fistula opening with an autologous pericardial patch.

continuous suture. The opened RCA was de-aired before it was completely closed. The patient was successfully weaned from cardiopulmonary bypass, and intraoperative transesophageal echocardiography showed no leakage of the fistula. The cardiopulmonary bypass and aortic cross-clamping times were 116 minutes and 70 minutes, respectively.

In the immediate postoperative period, the patient's vital signs were stable and no abnormal cardiac rhythm was detected. Postoperative echocardiography showed no leakage at the fistula closure site and a good LV ejection fraction of 69.9%. The patient was discharged on postoperative day 10 without complications. The patient continued to take aspirin and clopidogrel prescribed by his pediatric cardiologist after surgery. On postoperative echocardiography 4 months after repair, the proximal RCA was still dilated with a diameter of 6.28 mm. The LV aneurysm was measured as 11.3×8.1 mm; its contraction was still synchronous with the LV and no leakage of the previous fistula was found (Fig. 1D, E). The patient has now been followed up for 11 months and his condition is good.

Discussion

Coronary artery fistula is defined as an abnormal communication between the coronary arteries and cardiac chambers or vessels. It can present either as a part of a complex anomaly or occur in isolation. Its prevalence ranges from 0.2% to 0.9% [1,2]. In most cases, the exit chamber of the fistula is the right heart chamber, whereas fistulas terminating into the LV have been rarely reported [3,4].

Correction of the coronary artery fistula is generally indicated if the patient develops symptoms. Asymptomatic fistulas can also be corrected, since they may result in congestive heart failure, bacterial endocarditis, fistula rupture, or arrhythmia if left untreated [5-7]. Our patient showed early symptoms and signs of heart failure, including tachypnea and cardiomegaly. Although transcatheter closure of the fistula by angiography was an available treatment option [8], it would have been difficult to close the fistula safely through the transcatheter approach because the patient was a neonate and his fistula was extremely large.

Various surgical treatments have been reported for coronary artery fistulas: fistula ligation or division, tangential arterioplasty, transcoronary closure, intracardiac repair, and coronary artery bypass grafting. Fistula ligation is widely recognized as a safe treatment that can be performed without cardiopulmonary bypass [9]. In this case, fistula ligation could be considered a reasonable option because the fistula had a single origin, single vessel, and single termination. However, we avoided epicardial ligation of the fistula because the fistula was connected to the LV, which generated high pressure, resulting in an aneurysm at the residual fistula just distal to the ligation point. Intracardiac repair was another treatment option. However, direct incision of the LV was required for intracardiac approach and the incision had to be placed at the posteroinferior side of the LV, which was highly likely to lead to poor visualization of the surgical field. In this case, open patch closure of the fistula was considered to help maintain the original coronary arterial geometry and explore which coronary branches from the distal RCA should be preserved. For this reason, we chose a transcoronary approach—that is, patch closure of the opening of the fistula through a coronary incision under cardiopulmonary bypass—and it provided an excellent surgical field.

We did not dissect the distal RCA because we decided to make a coronary incision. We did not consider concomitant reduction-plasty of the RCA because the RCA was diffusely dilated, instead of having a fusiform aneurysm, and the enlarged RCA would have regressed in the neonatal period once the fistula was repaired. We decided not to perform coronary angiography after a discussion with the pediatric cardiology team because no significant coronary branch was found on either preoperative computed tomography or echocardiography. Moreover, because we planned to close the fistula at the entry site of the LV, it is not likely that any coronary branches of the distal RCA beyond the closing point would have been compromised.

An LV aneurysm accompanied a fistula in this case. Exclusion of the aneurysm was not performed concomitantly because the aneurysm contracted synchronously with the LV and its wall was not thin. Additionally, because the aneurysm was connected to the opening of the fistula, we assumed that it would not be dilated further after closure of the fistula. Postoperative echocardiography showed no significant change in the LV aneurysm in terms of size, wall motion, and thickness. Further follow-up is warranted.

In conclusion, we describe the case of a symptomatic neonate with a large fistula from the RCA to the LV accompanied by an LV aneurysm. The patient successfully underwent surgical closure of the fistula, and was followed up for 11 months without complications.

Article information

ORCID

Yu Ri Lee: https://orcid.org/0000-0002-2400-1556
Eun Seok Choi: https://orcid.org/0000-0002-0618-4686

Author contributions

Conceptualization: ESC. Data curation: YRL, ESC. Methodology: YRL, ESC. Project administration: YRL, ESC.

Visualization: YRL, ESC. Writing–original draft: YRL. Writing–review & editing: ESC. Final approval of the manuscript: all authors.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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