Transcatheter Occlusion of a Giant Coronary Artery Fistula in a Neonate

Shunsuke Yamada
Mieko Aoki-Okazaki
Manatomo Toyono

Corresponding Author: Manatomo Toyono, e-mail: manatomo@doc.med.akita-u.ac.jp

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Patient: Female, newborn
Final Diagnosis: Coronary arterial fistula
Symptoms: Respiratory distress
Medication: —
Clinical Procedure: —
Specialty: Cardiology • Pediatrics and Neonatology

Objective: Congenital defects/diseases
Background: Although large coronary artery fistulas are uncommon, they lead to substantial over-circulation in the pulmonary vascular beds and left heart system. Fistula occlusions are achieved via surgical or transcatheter technique; however, reports on successful outcomes of transcatheter treatment during the neonatal period are limited.

Case Report: A female infant was born at the gestational age of 37 weeks with a birth weight of 2615 grams via normal vaginal delivery. Cardiac auscultation revealed a loud continuous murmur emanating from the fourth right intercostal space. A right coronary artery-to-right ventricle fistula was confirmed using transthoracic echocardiography. The newborn developed respiratory distress 3 days after birth and was administered continuous positive airway pressure to assist breathing. On day 8, the ventilator was used through tracheal intubation due to gradual worsening of dyspnea. A 6-mm Amplatz Vascular Plug 4 (AGA Medical Corporation, Plymouth, MN) was chosen, as the minimum diameter of the coronary artery fistula was 5 mm. In view of the risk of myocardial ischemia with additional devices, the procedure was stopped despite persistent shunting. The newborn’s clinical condition significantly improved following the procedure and she was eventually weaned off ventilator support.

Conclusions: A self-expanding occlusion device was useful for relieving this life-threatening condition. Complete elimination of shunting is not always necessary, to avoid compromising myocardial circulation.

Keywords: Coronary Vessels • Fistula • Heart Failure • Heart Ventricles • Infant, Newborn • Therapeutics

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Background

Most coronary artery fistulas are small and asymptomatic. Large coronary artery fistulas uncommonly lead to substantial over-circulation in the pulmonary vascular beds and left heart system, potentially increasing the risk of myocardial ischemia, heart failure, and pulmonary hypertension [1]. The presence of congestive heart failure and/or distal coronary insufficiency warrants therapeutic interventions such as transcatheter occlusion or open surgical ligation on the coronary artery fistula [2,3]. Transcatheter occlusion of coronary artery fistulas is associated with good short-term procedural success rates and complication rates in selected patients [4]; however, reports on successful outcomes during the neonatal period are limited [2,5,6]. Here, we describe the transcatheter management of a neonate who developed congestive heart failure associated with a giant right coronary artery-to-right ventricle fistula.

Case Report

In a 28-year-old woman (gravida 1, para 1), with fetal cardiomegaly, was identified through routine scanning at the gestational age of 22 weeks. She was in good health without any maternal complications or high-risk factors. Further examination revealed an abnormal vessel originating from the ascending aorta and a dilated right ventricle in the fetus. The drainage site of the abnormal vessel was not identified; however, diastolic reversal in the aorta suggested a diagnosis of coronary artery-to-right ventricle fistula. The size of the abnormal vessel gradually increased during pregnancy. No interventions

Figure 1. Transthoracic echocardiograms. (A, B) Parasternal short-axis views. (C, D) Subxiphoid views. Arrows: Proximal right coronary artery markedly dilated. Ao – aorta. (Adobe Photoshop Elements 18.0).
were performed after the identification of the anomaly because normal fetal growth was periodically confirmed.

A female infant was born at the gestational age of 37 weeks with a birth weight of 2615 g via normal vaginal delivery. On physical examination, her respiratory rate was 65 per min, apical rate 140 per min, and blood pressure 50/27 mmHg (normal reference ranges: respiratory rate 30 to 50 per min, apical rate 120 to 160 per min, and blood pressure 70/40 mmHg or greater, respectively). Cardiac auscultation revealed a loud continuous murmur emanating from the 4th right intercostal space.

Prenatal findings suggesting a right coronary artery-to-right ventricle fistula were confirmed after delivery using transthoracic echocardiography (Figure 1A-1D). A chest X-ray on the first day indicated increased pulmonary vascularity within the bilateral lung fields; therefore, a continuous intravenous infusion of phosphodiesterase-3 inhibitors (olprinone, 0.3 µg/kg/min) was initiated. An electrocardiogram on the second day did not show ischemic change (Figure 2). The newborn developed respiratory distress 3 days after birth and was administered continuous positive airway pressure to assist breathing. On day 8, the ventilator was used through tracheal intubation due to gradual worsening of dyspnea. The plasma B-type natriuretic peptide level was 6753 pg/ml at the time. Enhanced computed tomography angiography exhibited a single stenotic site (neck) in the entire fistula.

A decision was made to percutaneously close the fistula; 22 days after birth, the procedure was performed under generalized anesthesia during which, unfractionated heparin was administered. As her bilateral femoral arteries were narrow, vascular access was via the common carotid artery using an introducer sheath to avoid prolonged attempts through the femoral artery; the approach was retrograde arterial loop. A soft-tipped stiff wire was advanced, followed by a 4-Fr catheter over the wire; selective coronary angiography was performed prior to the procedure to delineate the anatomy of the giant distal fistula (Figure 3A, 3B). Due to the tortuosity of the fistula requiring placement in a controlled fashion, and its high-flow communication, a self-expanding occlusion device was selected for closure. A 6-mm Amplatzer Vascular Plug 4 (AGA Medical

Figure 2. Electrocardiogram. There were no abnormal Q waves or ST-T changes in leads II, III, and aVF. (Adobe Photoshop Elements 18.0).
Corporation, Plymouth, MN) was chosen, as the minimum diameter of the coronary artery fistula was 5 mm; great care was taken not to compromise circulation to the myocardium. The correct location of the device was confirmed through angiography before its release from the delivery cable. After deployment of the device, to verify the correct position and a possible residual flow across the target structure (Figure 3C, 3D), a repeat angiography was immediately performed. In view of the risk of myocardial ischemia with additional devices, the procedure was stopped despite persistent shunting. The newborn’s clinical condition significantly improved following the procedure and she was eventually weaned off ventilator support. After a 2-week hospital stay, she was discharged home with a prescription for aspirin (5 mg/kg/day). There was no occurrence of post-procedural myocardial infarction, vascular complications, or stroke.

During the 16-month follow-up period, she remained both stable and asymptomatic, with no evidence of myocardial infarction; the size of her right ventricle had normalized, and the B-type natriuretic peptide plasma level was 10.5 pg/mL. Although post-procedural angiography revealed a small persistent shunting, the mean pulmonary arterial and pulmonary capillary wedge pressures were not elevated (13 and 9 mmHg, respectively).

Figure 3. Right coronary angiography. Pre-procedure: Frontal (A) and lateral projections (B); Proximal right coronary artery (white arrows) was markedly dilated. The “neck” of the fistula (red arrows) was defined proximal to it, entering the right ventricle (RV). Post-device deployment: Frontal (C) and lateral projections (D); A self-expandable device (yellow arrows) was deployed at the neck of the fistula. (Adobe Photoshop Elements 18.0).
Discussion

A coronary artery fistula is rarely large enough to exert a substantial effect on hemodynamics. For a fistula that is symptomatic, invasive treatment by transcatheter occlusion or surgical ligation is usually a reasonable choice, and both strategies show comparable results in long-term follow-up [7]. To date, reports on successful outcomes during the neonatal period have been limited (Table 1). According to the previous reports, the body weights of the patients who underwent transcatheter occlusion of coronary artery fistulas during the neonatal period ranged from 3.4 to 3.9 kg [2,5,6]; therefore, we would like to emphasize that our patient (2.6 kg) was the lightest. Even in a neonate with a body weight less than 3 kg, a fistula can be treated with transcatheter occlusion accessed via the common carotid artery. In addition, all neonatal cases were treated with self-expandable devices, leading to closure of the coronary artery fistulas.

Although right coronary artery-to-right ventricle shunting persisted after transcatheter therapy of the fistula in our case, post-procedural improvement was substantial. In a 21-year experience of transcatheter therapy of coronary artery fistula at the Mayo Clinic, 3 of 16 patients who had an acute persistent shunting of trivial grade showed an increase in residual flow during the follow-up period [4]. In a study of 15 years of transcatheter treatment at another institution, of the 9 patients who required re-intervention, the reason of re-intervention was persistent shunting only in 1, and those included myocardial ischemia, device embolization, and valve injury in the others [8]. Based on these experiences, it is quite possible that complete elimination of shunting is not always necessary. Closure of small-to-moderate coronary artery fistulas is recommended in the presence of symptoms of myocardial ischemia, threatening arrhythmia, unexplained ventricular dysfunction, or left atrial hypertension [9]. No data have been reported on asymptomatic coronary artery fistulas to establish the maximum magnitude of shunting through a fistula that is considered harmless.

Transcatheter treatment is associated with a high incidence of adverse events in children [10]. Known complications of transcatheter occlusion of coronary artery fistulas include residual shunt due to incomplete closure, myocardial ischemia due to closure of side branches of peripheral normal coronary artery, coil migration and embolization in the right ventricular system, transient ST changes on electrocardiograms, and atrioventricular block [7]. In addition, there is a significant risk of complication at the site of arterial puncture in infants weighing less than 4 kg [11]. The risk of complication is high in the first 6 months of life, and 0.23% of deaths are reported in neonates [12]. In contrast, in neonatal cardiac catheterization, low-birth-weight infants weighing less than 2.5 kg have significantly more minor complications than those weighing more than 2.5 kg, but there is no difference in the frequency of serious complications, according to 1 report [13]. The relationship between the number of years of experience of the operators and adverse events has been reported from the United States in a multicenter registry; the incidence of adverse events was significantly higher among operators with less than 5 years of experience [14].

In this case, the following conditions were met for transcatheter occlusion of the coronary artery fistula: (1) possible safe insertion of the catheter into the coronary artery fistula, (2) presence of stenosis at the planned embolization site of the fistula

Table 1. Characteristics of patients who underwent transcatheter occlusion of coronary artery fistulas during the neonatal period.

| Case | Gender | Body weight | Symptoms/findings | Day at closure | Origin | Termination | Size of fistula | Device | Outcome | Reference |
|------|--------|-------------|-------------------|----------------|--------|-------------|----------------|--------|---------|-----------|
| 1    | N/A    | N/A         | Congestive heart failure | 5              | Right coronary artery | Right coronary artery | N/A    | 4-mm Amplatzer Muscular Ventricular Septal Defect | Complete occlusion | [5]     |
| 2    | N/A    | N/A         | Congestive heart failure | 17             | Right coronary artery | Right atrium           | N/A    | 8/6 Amplatzer Ductal Occluder I | Complete occlusion | [5]     |
| 3    | Male   | 3.9 kg      | ST-T segment abnormalities | 7              | Left main coronary artery | Left ventricle | 4 mm | One 8-mm Amplatzer Vascular Plug 2, two 6.5-mm micro-vascular plugs | Complete occlusion | [6]     |
| 4    | Female | 2.6 kg      | Congestive heart failure | 22             | Right coronary artery | Right ventricle | 5 mm | 6-mm Amplatzer Vascular Plug II | Partial occlusion | Present case |
pathway, and (3) 1 opening to the right ventricle. Furthermore, by selecting the common carotid artery as the approach site, percutaneous treatment of the giant coronary artery fistula was successful in the neonatal period.

Coronary artery fistula is rarely reported in the fetal period [5,7,15-17], and in many of these the neonates developed early symptoms of congenital heart failure shortly after birth. In our case, congestive heart failure and cardiac enlargement were observed in the early neonatal period, as in these reports. This may have been related to a decrease in right ventricular systolic pressure due to decreased pulmonary vascular resistance after birth. In principle, early closure is the treatment of choice for large coronary artery fistula; fetal diagnosis will allow careful follow-up and treatment through the perinatal period.

Conclusions

Transcatheter occlusion of the giant coronary artery fistula using a self-expandable device was feasible even in a neonate whose body weight was 2.6 kg. Complete elimination of shunting was not necessary to relieve life-threatening congestive heart failure associated with the fistula. Treatment strategies for a such case need to be developed from multiple perspectives.

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Declaration of Figures Authenticity

All figures submitted have been created by the authors, who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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