Transcatheter Coil Embolization of a Huge Right Coronary Artery to Right Ventricle Fistula

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Coronary artery fistula is a rare congenital heart disease. Treatment including medical control or intervention is only indicated for symptomatic children. It has been reported that transcatheter coil embolization is appropriate for symptomatic patients with a single fistula, safe accessibility to the feeding coronary artery, and the absence of large branch vessels. In this case report, we present a 4-year-old boy who was found by cardiac catheterization to have a huge isolated right coronary artery-to-right ventricle fistula. The patient initially presented at one year of age with an asymptomatic continuous grade II/VI murmur over the left-lower sternal border. Echocardiography demonstrated a continuous turbulent flow in the right ventricle and a dilated aneurysmal right coronary artery, suggesting the presence of a right coronary artery fistula. As the patient was asymptomatic, he was followed closely without definitive treatment. Over the next three years, he gradually developed mild exertional dyspnea and decreased exercise tolerance. Serial electrocardiograms revealed biventricular hypertrophy, and echocardiography revealed progressive right coronary arterial dilatation. Cardiac catheterization eventually confirmed the diagnosis, and the fistula was occluded by percutaneous transcatheter coil embolization. The boy remained well after the procedure with resolution of his symptoms. In conclusion, coil embolization is a safe alternative for the treatment of coronary artery fistula.

Key Words: Coronary artery fistula • Percutaneous transcatheter coil embolization

INTRODUCTION

Coronary artery fistula is a rare anomaly found in approximately 0.13% to 0.2% of patients undergoing diagnostic coronary angiography.¹,²,³ It reportedly accounts for only 0.2% to 0.4% of all congenital cardiac defects but for about 50% of pediatric coronary anomalies.²,⁵,⁸ Krause first described the anomaly in 1865. It is often congenital but may be acquired as a result of trauma or invasive cardiac procedures.¹,² The incidence of associated congenital cardiac anomalies was 13% in the adult and 21% in the pediatric population.² It is defined as any abnormal communication through which coronary artery blood is shunted into a cardiac chamber, great vessel, or other vascular structure without passing through the myocardial capillary bed. Fistulas are classified based on their point of termination as (1) a coronary-cameral fistula ending in any of the cardiac chambers or (2) a coronary arteriovenous fistula ending in either a systemic or pulmonary vein. We report a case of a huge right coronary artery-to-right ventricle fistula treated with transcatheter coil embolization.

CASE REPORT

A 4-year-old boy had first been seen at our hospital 3 years previously because of a continuous cardiac murmur. He was asymptomatic, hemodynamically stable, and had normal activity, feeding, and development. On
physical examination, he was not cyanotic, and there was no evidence of other congenital anomalies. Ausculation revealed a grade II/VI continuous murmur over the left-lower sternal border. There was no hepatomegaly and the peripheral pulses were normal.

Transthoracic 2-dimensional and color-Doppler echocardiography initially showed a dilated aneurysmal right coronary artery (RCA) and an abnormal continuous turbulent flow in the right ventricle (RV). On the parasternal short axis view, a dilated RCA orifice (5.8 mm in diameter) near the aortic root was seen with an abnormal Doppler flow signal during diastole. The apical 4-chamber Doppler image demonstrated a large continuous turbulent shunt within the RV directed toward the interventricular septum. The diagnosis of an isolated congenital RCA-to-RV coronary fistula was suspected after the initial examination and echocardiography. As he was asymptomatic, the patient was then regularly followed up in our hospital without any medication or intervention.

He was well until age 4, when he started to have mild exercise intolerance. Chest radiography revealed slight cardiomegaly with lung markings that were mildly increased compared to earlier films. The electrocardiogram revealed sinus rhythm with biventricular hypertrophy (Figure 1A). On echocardiography, the diameter of the RCA was increased from the previous 5.8 mm to 9.1 mm (Figure 2A) and the right intraventricular continuous

Figure 1. (1A) Twelve-lead electrocardiography before coil occlusion, revealing sinus rhythm with biventricular hypertrophy. (1B) Four months after coil occlusion, the electrocardiogram reveals persistent biventricular hypertrophy (the voltage calibration is full-standard in the limb leads and half-standard in the precordial leads). There is normal sinus rhythm without ischemic changes or conduction disturbance.
flow had increased in amount (Figure 2B). In addition, the aortic root was mildly dilated, there was trivial aortic regurgitation, and both ventricles were hypertrophic.

To avoid shunt-induced high-output heart failure or other complications, we decided to close the suspected RCA-to-RV fistula. On cardiac catheterization, there was step-up of the oxygen saturation in the RV (oxygen saturation: superior vena cava: 74%; inferior vena cava: 75%; right atrium: 74%; RV: 84%; main pulmonary artery: 83%; left ventricle: 97%; aorta: 97%), with a calculated pulmonary-to-systemic flow ratio of 1.6. Pressures in the right heart were normal. Aortic root angiography demonstrated a huge aneurysmal RCA with a fistula (2.3 mm in the narrowest dimension) draining into the RV (Figure 3A). The contour of the RCA was smooth, and no major distal branches near the termination were seen.

Figure 2. Echocardiography before coil occlusion. (2A) Two-dimensional echocardiography, short-axis view, demonstrating a large lumen and orifice of the right coronary artery (9.1 mm) (large arrow) compared with the orifice of the left coronary artery (2.75 mm) (small arrow). (2B) Color-Doppler echocardiography, apical 4-chamber view, showing continuous turbulent flow (arrow) in the right ventricle.

Figure 3. Angiography before coil occlusion. (3A) Aortic root angiography, anterior-posterior view, revealing an aneurysmal right coronary artery with a fistula (large arrow) shunting into the right ventricle. The left coronary artery (small arrow) is far smaller than the right coronary artery. Selective left coronary artery angiography, anterior-posterior view (3B) and lateral view (3C), revealing left coronary artery dominance. The left coronary artery (small arrow in 3B) has a widened myocardial distribution. The left circumflex artery gives rise to the posterior descending artery (small arrow in 3C).
On selective left coronary artery angiography (Figure 3B and 3C), the artery appeared to be normal in size. The left circumflex artery supplied the posterior descending artery, contributing to left coronary dominance. Based on these findings, an isolated RCA-to-RV coronary artery fistula was diagnosed, and transcatheter coil embolization was considered appropriate.

To evaluate the safety of occlusion, we initially inflated the catheter balloon to occlude the distal RCA near the site of the fistula for 20 minutes while monitoring the hemodynamic status and electrocardiogram. The vital signs remained stable and there was no evidence of myocardial ischemia during the observation period. A coil was selected that would have a diameter nearly 1.5 times that of the fistula and a length great enough to complete at least 5 entire circles. Based on the size of the fistula, the coil diameter needed to be at least 10.8 mm and the length at least 5.5 cm. We therefore inserted a 38-11-7 coil (Occluding Spring Coil, Cook, Bloomington, IN, U.S.A.), having an 11-mm diameter and 7-cm length, which succeeded in occluding the fistula. Only a trivial residual shunt was demonstrated by selective RCA angiography after the procedure (Figure 4). The aortic pressure increased from 98/58 mmHg prior to occlusion to 118/80 mmHg afterwards, indicating cessation of the RCA-to-RV shunt.

The patient tolerated the procedure well, and his murmur disappeared. The cardiac enzymes were within normal limits on the day of the procedure (CK/CKMB: 138/9.2 IU/L, Troponin-I: 0.17 ng/mL), and on the next two days (CK/CKMB: 79/6.6 IU/L and 79/8.3 IU/L). Four months later, an electrocardiogram revealed sinus rhythm and persistent biventricular hypertrophy (Figure 1B). No immediate or late arrhythmias, conduction disturbance, or myocardial ischemic changes appeared on the electrocardiogram after the procedure. A follow-up echocardiogram revealed a normal bilateral coronary flow pattern, persistently dilated RCA without thrombosis, normal ventricular kinetic motion bilaterally, and adequate valvular function despite trivial aortic regurgitation. The patient did well over the subsequent four months, without complaints of chest discomfort or exertional dyspnea. He received no medication after occlusion of the fistula. However, given the long-term risk of thromboembolic events, we will continue to monitor the child.

**DISCUSSION**

Our patient had a primary or isolated coronary artery fistula. In such cases, the lesion is the only cardiac abnormality or may be associated with a patent ductus arteriosus or atrial septal defect. Secondary or complicated fistulas, on the other hand, are associated with major cardiac anomalies. Coronary artery fistulas arise most commonly from the RCA (55%), the rest arising from the left coronary artery (35%), both coronary arteries (5%), or an anomalous single coronary artery (3%).17 The vast majority terminate in the right side of the heart (> 90%), primarily the RV (42.5%), or the right atrium (34%), pulmonary trunk (15%), coronary sinus (7%), and, rarely, in the left atrium (5%) or left ventricle (3.5%).2,5,17

The pathophysiology of coronary artery fistula is myocardial stealing or reduction in myocardial blood flow distal to the site of termination. The mechanism is related to the pressure gradient and runoff from the coronary vasculature to a low-pressure receiving cavity. The natural history of coronary artery fistula is highly variable.2,4,5,15 Some patients remain asymptomatic throughout life and survive into the ninth decade. In contrast, an emergency such as that of a 4-day-old infant with severe cardiorespiratory failure requiring extracorporeal mem-
brane oxygenation has also been reported. The incidence of symptomatic cases ranges from 30% to 55% and varies by age, with only 19% of patients under 20 years old reporting symptoms compared with 63% over 20.\textsuperscript{4,5,8} Reported pediatric fistula-related complications include congestive heart failure (12%), myocardial infarction (4%), bacterial endocarditis (3%), and death (6%). Other rare life-threatening complications included giant aneurysmal dilatation of the fistula, dissection, and rupture with cardiac tamponade, myocardial infarction, and sustained ventricular tachycardia. In pediatric patients, the most frequent presenting finding is congestive heart failure.\textsuperscript{2,5}

In the absence of symptoms, a continuous murmur may be the only sign that a cardiac anomaly is present.\textsuperscript{2,5} In asymptomatic pediatric patients with a small single lesion, spontaneous fistulous closure may occur in 23% of the cases.\textsuperscript{2,5} The reported incidence of spontaneous closure in all age groups is 1%, and in those under 20 years of age, 2%.\textsuperscript{2,5} Another review stated that spontaneous closure is more common in the pediatric (8.5%) than in the adult population (3%).\textsuperscript{2} Even though spontaneous closure is thus quite rare, there is no urgency to close coronary artery fistulas immediately in asymptomatic patients.\textsuperscript{2,5}

The symptoms are mainly dependent on the severity of the shunt, with small shunts being symptomatically silent. Such patients can be monitored periodically to make sure the condition is not progressing. In a large fistula, the intracoronary diastolic perfusion pressure may drop profoundly, leading to myocardial ischemia, or pulmonary overperfusion may ensue, eventually causing heart failure. The coronary vessel will attempt to compensate by progressive enlargement of the ostia and feeding artery. Our patient was initially asymptomatic, so it was reasonable to follow him at that point. However, over the next 3 years, the diameter of the proximal RCA almost doubled, from 5.8 mm to 9.1 mm. His biventricular hypertrophy reflected the long-term left-to-right shunt. Fortunately, the volume overload was compensated for by the hypertrophic heart and did not result in myocardial ischemia. The patient’s unusual left coronary dominance may also have played a role in protecting the myocardium from ischemia. However, the child ultimately developed heart failure symptoms so that intervention was indicated.

The goal of treatment for symptomatic patients is abolition of the fistula in a way that doesn’t interfere with the native coronary artery supply. Factors to consider in deciding between surgery and coil occlusion include the size of the fistula, its location and drainage pattern, and the possibility of associated cardiac lesions. Prior to the availability of occlusive devices, surgery was the only choice. While the reported surgical morbidity and mortality (3%) and incidence of recurrence (4% of patients) are low,\textsuperscript{9} such procedures still require a median sternotomy and usually cardiopulmonary bypass. A few recent reports have described a number of successful surgical repairs without circulatory bypass.\textsuperscript{16} However, less invasive procedures are still preferable.

Percutaneous transcatheter coil embolization, first successfully performed in 1983, has therefore become the procedure of choice in many centers in order to avoid surgical risk.\textsuperscript{15} In addition to coils,\textsuperscript{9} use of a number of other devices has been reported, including detachable balloons, umbrellas, polyvinyl alcohol foam, and others.\textsuperscript{10-13} Coil occlusion is considered the best method because of improved control and delivery techniques.\textsuperscript{15} The risks of coil embolization are low, and there have been no reports of deaths in children.\textsuperscript{15} Only one fatality in an adult patient has been reported.

Although good outcomes can be achieved with coil embolization, only 36% of coronary artery fistulas are amenable to this method. Mavroudis and coauthors\textsuperscript{14} recommend criteria for selecting patients for coil occlusion, including the absence of multiple fistulae, a single narrow drainage site, absence of large branch vessels, and safe accessibility to the coronary artery supplying the fistula. In our patient, the smooth contour of the RCA, no RCA distal branch, and a single drainage site appeared amenable to coil occlusion. Additionally, the patient’s left coronary dominance may reduce the possible risk of myocardial infarction induced by coil embolization. This may explain why our patient has continued to do well without any sequelae.

Complications after coil occlusion are rare but may include transient T-wave inversions, transient arrhythmias, coil migration, coronary artery trauma or rupture, or total occlusion of a coronary artery. Persistent leakage after coil occlusion has been reported, but these leaks may be abolished by the placement of additional coils.\textsuperscript{15} Persistent coronary dilatation as late as 4 years following
fistula occlusion has been also reported. Low-dose aspirin therapy (3 to 5 mg/kg per day) is recommended until coronary normalization occurs. Warfarin may be added if the dilatation is severe (> 10 mm), particularly when coronary flow is sluggish. In our patient, no immediate or late complications occurred. However, because the proximal RCA remains dilated, regular monitoring of his condition is indicated.

In summary, transcatheter coil embolization of the coronary artery fistula is a safe and effective procedure in children. Because the long-term outcome after coil occlusion is unknown, patients require close follow-up and, in certain cases, anti-platelet therapy or low-dose anticoagulation.

ACKNOWLEDGMENTS

I wish to acknowledge my teacher, Dr. Ming-Ren Chen, and Dr. Mary-Jeanne Buttrey for assistance during the preparation of this manuscript.

REFERENCES

經皮心導管螺旋線圈栓塞術治療一個巨大的
右冠狀動脈至右心室動脈瘻管

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冠狀動脈瘻管是一種罕見的先天性心臟病，對於無症狀的病童可以先觀察，而對有症狀的
孩童則須施予適當的藥物或介入性治療。根據文獻，對於有症狀的單一病灶、心導管能安
全到達病灶、且無複雜的冠狀血管分枝的病患，適合以心導管彈簧線圈方式栓塞治療。我
們發表了一個四歲的男孩，經心導管檢查證實其患有一個巨大的單純性右冠狀動脈至右心
室動脈瘻管。病患於一歲時，於左下胸骨緣聽診發現 2 度的連續性心雜音至本院求診。心
臟超音波顯示右心室內出現一個連續性分流，與擴大的動脈瘤樣右冠狀動脈。由於無臨床
症狀，因此持續追蹤病患。三年後，男童逐漸表現出呼吸喘以及運動負荷減低的症狀，心
電圖出現雙側心室肥大，超音波追蹤顯示逐漸擴張的右冠狀動脈。最後經由心導管檢查確
定診斷，並以心導管栓塞方式治療，術後病人狀況良好，無併發症產生。結論，心導管彈
簧線圈栓塞術是一種安全的冠狀動脈瘻管替代性介入治療，並且，術後持續規則地追蹤其
可能出現的併發症是需要的。

關鍵詞：冠狀動脈瘻管、經皮心導管螺旋線圈栓塞術。