

Surgical Management of a Double Coronary Cameral Fistula Associated with Mitral and Tricuspid Regurgitation, Patent Foramen Ovale, and Chronic Atrial Fibrillation

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ABSTRACT

Coronary cameral fistula, a rare acyanotic congenital or acquired anomaly that involves a large communication between one of the coronary arteries and a cardiac chamber, is the most important hemodynamically significant coronary artery abnormality. This case report is the first description of the combination of double coronary artery cameral fistulas associated with pulmonary hypertension and congestive heart failure and complicated by mitral and tricuspid regurgitation, patent foramen ovale, and chronic atrial fibrillation.

INTRODUCTION

Coronary cameral fistula, a rare acyanotic congenital or acquired anomaly that involves a large communication between one of the coronary arteries and a cardiac chamber, is the most important hemodynamically significant coronary artery abnormality. This case report is the first description of the combination of double coronary artery cameral fistulas associated with pulmonary hypertension and congestive heart failure and complicated by mitral and tricuspid regurgitation, patent foramen ovale, and chronic atrial fibrillation.

CASE REPORT

A 56-year-old man was referred with a 6-month history of mild shortness of breath (New York Heart Association class II). He had been found to have atrial fibrillation as an incidental finding 18 months previously. A physical examination revealed cardiomegaly with a continuous murmur in the pulmonary area and a pansystolic murmur at the apex.

A transthoracic echocardiography evaluation showed enlargement of all 4 cardiac chambers with severe mitral and tricuspid regurgitation due to annular dilatation. The pulmonary artery systolic pressure was 62 mm Hg.

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The most striking findings were an aneurysmal proximal right coronary artery with large vessels on the external surface of the heart appearing to take a tortuous course in the left atrioventricular groove (Figures 1 and 2). A transesophageal echocardiographic examination confirmed a grossly dilated right coronary artery draining into the coronary sinus (Figure 3). A computed tomography arteriogram clarified the diagnosis but also demonstrated involvement of the circumflex artery, which was not only mildly dilated and tortuous but also drained into the right coronary artery fistula distal to the take off of the posterior descending coronary artery and proximal to the second last tortuous fistulous loop on the inferior lateral wall in the atrioventricular groove (Figure 4).

Because of the patient's significant symptoms and to prevent progressive biventricular dysfunction, we decided to proceed with surgical intervention. The procedure was performed with cardiopulmonary bypass. The cardiopulmonary bypass time was 215 minutes, and the aortic cross-clamp time was 132 minutes (patient temperature, 32°C).

The first fistula from the right coronary artery to the coronary sinus was clearly seen at sternotomy (Figure 5). These large fistulous vessels were approximately 3 cm in diameter and were carefully dissected from each other. A double ligature and pledgeted suture were used to occlude the right coronary artery fistulous connection distal to the take off of the posterior descending coronary artery and what also appeared to be a small inferior wall coronary branch just distal to the inferior cardiac veins in the inferior interventricular groove. This branch was proximal to the site where the circumflex fistula entered. We confirmed that this vein crossed superficial to the fistula and then proceeded to the distal coronary sinus. When we attempted to go around the fistula, we opened the distal coronary sinus, which we then repaired. The fistula connection distal to the identified site of entry of the circumflex fistula was then similarly double-ligated with a pledgeted suture.

The regurgitation of the mitral valve, which was secondary to annular dilatation, was repaired by implantation of a Carpentier-Edwards ETLogicx 32-mm annuloplasty ring (model 4100; Edwards Lifesciences, Irvine, CA, USA). Similarly, the tricuspid regurgitation, which was also secondary to annular dilatation, was repaired with

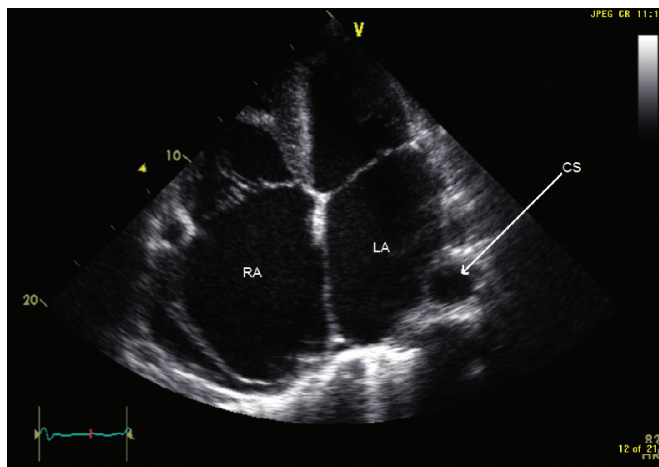


Figure 1. Transthoracic echocardiographic apical 4-chamber view showing dilatation of all cardiac chambers with a markedly dilated coronary sinus (CS). RA indicates right atrium; LA, left atrium.

a Carpentier-Edwards MC3 34-mm annuloplasty ring (model 4900; Edwards Lifesciences). The patent foramen ovale was closed with a running suture.

In addition, we used a bipolar and monopolar radiofrequency device (Cardioblate; Medtronic, Minneapolis, MN, USA) to carry out a modified biatrial Maze III procedure with excision of the left atrial appendage.

After termination of bypass, we observed that the intervening segment was filled and slightly distended, but there was no run off into the coronary sinus.

The patient was extubated 13.5 hours later, and his postoperative recovery was uneventful. He was discharged in sinus rhythm on the 10th postoperative day. This gentleman remains well at follow-up, and a repeat transthoracic echocardiography evaluation showed marked reductions in the sizes of all 4 chambers with no significant mitral or tricuspid regurgitation (Figure 6).

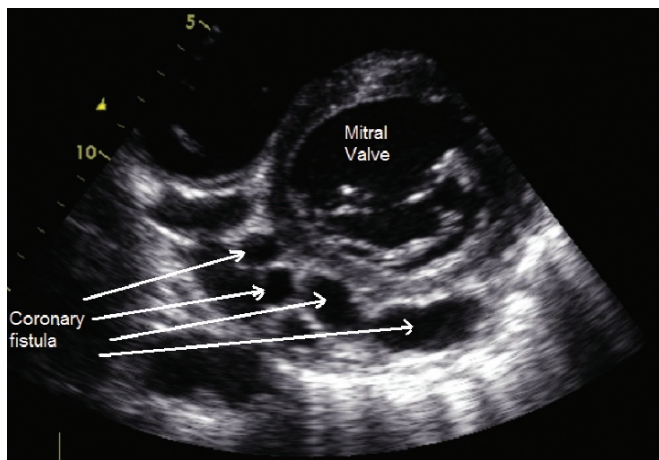


Figure 2. Transthoracic echocardiographic parasternal short-axis view showing abnormal vessels on the posterior surface of the left ventricle.

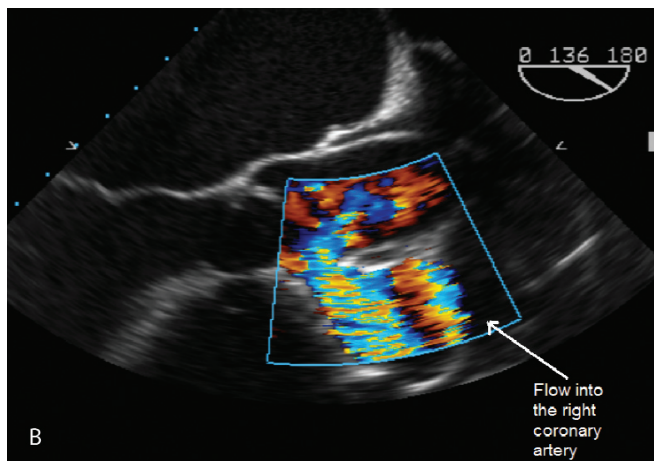
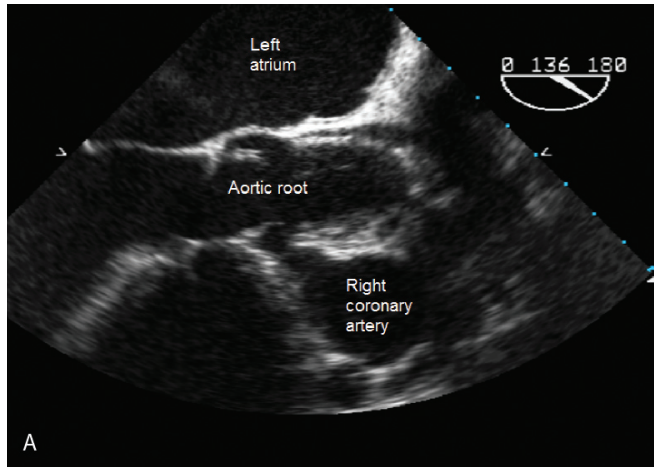


Figure 3. Tracheoesophageal long-axis projection showing a grossly dilated proximal right coronary artery.

DISCUSSION

The first description of a coronary artery fistula was in 1865 by Krause [1865]. The incidence of this disorder is 0.1% to 0.4%, with the vast majority of cases being congenital [Yamanaka 1990; Kale 2006]. Acquired fistulas due to acute myocardial infarction, blunt and penetrating trauma, and iatrogenic fistulas due to mitral valve surgery, endomyocardial biopsy, pacemaker insertion, radiofrequency ablation, and angioplasty have all been described. Only 0.4% of all congenital cardiac malformations are due to coronary artery fistulas [Davis 1994], but they represent the most common congenital anomaly of the coronary arteries [Levin 1978].

Coronary artery fistulas are isolated in 55% to 80% of cases or are associated with other congenital anomalies (such as tetralogy of Fallot, patent ductus arteriosus, ventricular septal defect, atrial septal defect, or pulmonary atresia-intact ventricular septum) in approximately 20% to 45% of cases. The single fistulas are more common (74%-90% of cases), multiple fistulas are present in 10.7% to 16% of cases, and fistulas originating from both coronaries are present in 4% to 18% of cases [Dodge-Khatami 2000].

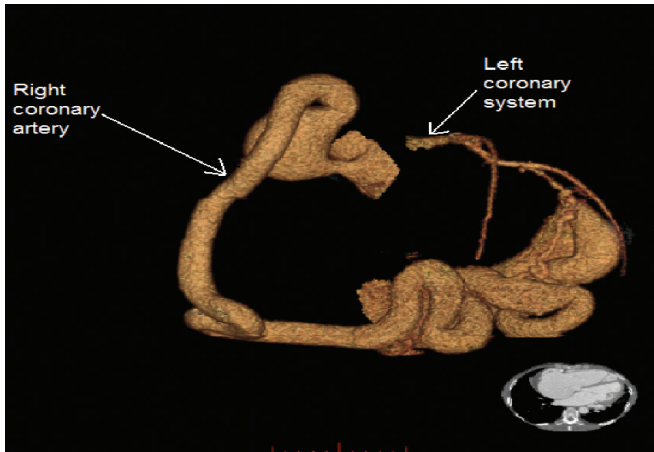


Figure 4. Computed tomography coronary arteriogram showing the course of the hugely dilated right coronary artery, plus the left coronary artery.

In heart transplant recipients, the prevalence of coronary artery fistulas is 7.7% to 8%, which is significantly higher than in the healthy population [Sandhu 1989; Somers 1991].

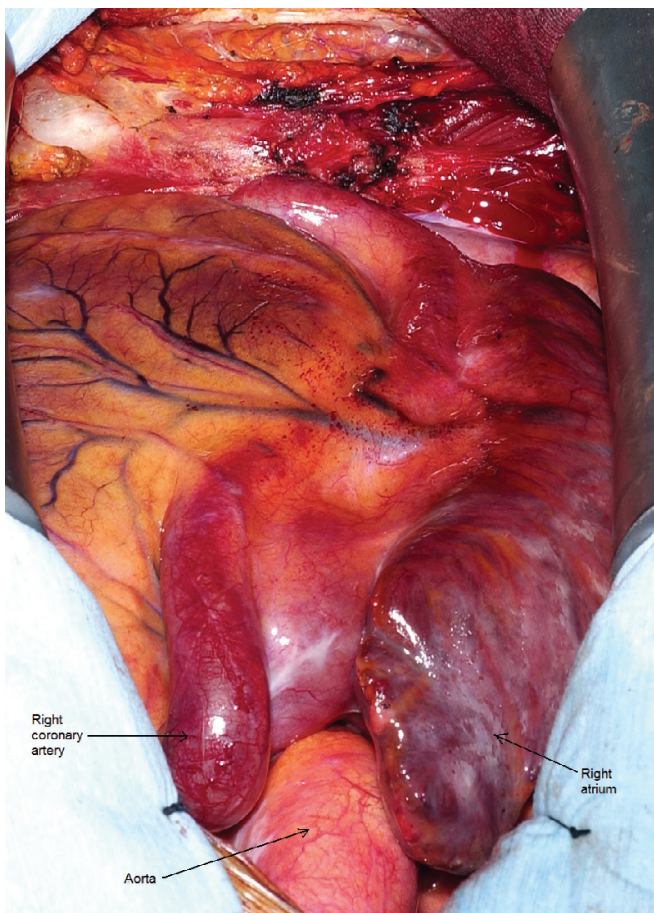


Figure 5. The course of the dilated right coronary artery is shown prior to cannulation of the heart.

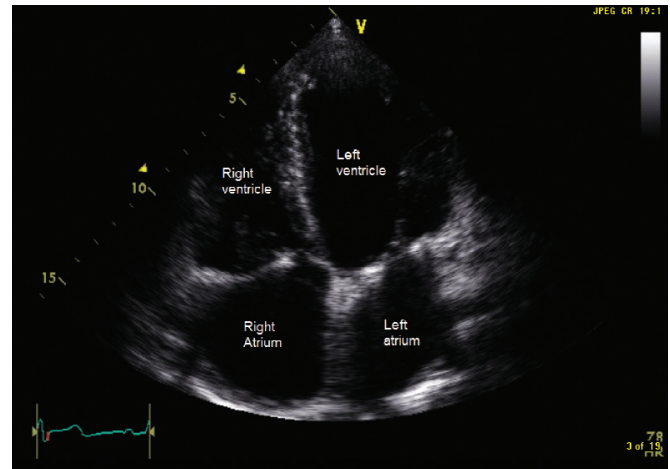


Figure 6. Transthoracic echocardiographic apical 4-chamber view taken 4 months postoperatively showing a marked reduction in the size of all 4 chambers. The ventricular end-diastolic dimensions in particular have been reduced from 6.8 cm to 4.9 cm. There was no significant mitral or tricuspid regurgitation.

In a large review of 187 patients with coronary artery fistulas, 81% of the patients younger than 20 years were asymptomatic. In contrast, patients older than 20 years were symptomatic in 63% of the cases; these patients presented with congestive heart failure, bacterial endocarditis, myocardial infarction, fistula rupture, arrhythmias, and sudden death [Liberthson 1979]. One of the most important clinical findings is the presence of a continuous heart murmur [McLellan 1989; Lau 1995]. The severity of the symptoms depends on the magnitude of the left-to-right shunt and the extent of the “coronary steal phenomenon.” It is widely accepted that symptomatic patients should be treated.

The mainstay of surgical therapy is ligation of the fistula, a procedure that was first described by Biörck and Crafoord in 1947 and currently remains the most effective treatment [Biörck 1947].

The angiographic classification by Sakakibara et al in 1966 [Sakakibara 1966] suggests 2 types of fistulas: type A (proximal type), in which the proximal coronary segment is dilated to the origin of the fistula and the distal end is normal, and type B (distal type), in which the coronary artery is dilated over the entire length and terminates as a fistula in the right side of the heart (end artery type), with the regular branching of the coronary proximal to the fistula. The surgical relevance of this classification is that type A is treated by epicardial ligation of the fistula distal to the fistula origin. Normal branch flow is maintained, and no cardiopulmonary bypass is required. Type B requires ligation of the precapillary end by intracameral purse-string sutures at the site of the termination with cardiopulmonary bypass [Sakakibara 1966; Dodge-Khatami 2000].

The operative mortality rate approaches 0% to 2%, depending on the complexity of the fistula [Urrutia-S 1983; Mavroudis 1997]. Surgical complications include myocardial infarction, residual fistula or recurrence, cerebrovascular accident, mediastinitis, complete heart block, and arrhythmias [Reul 2002].

Alternative therapeutic options include coil embolization, an Amplatzer vascular plug, and deployment of a polytetrafluoroethylene-coated stent [Mavroudis 1997; Mestre Barceló 2004; Balaguru 2006; Said 2006].

The surgical approach depends on the type of fistula, the coexisting anomalies, and the patient's comorbidities. The management of asymptomatic patients is still the subject of debate. Sherwood et al reported a spontaneous closure rate of 23% (7/31) in patients with small-sized fistulas and found no adverse outcomes in children and adolescents with incidental asymptomatic fistulas that were managed conservatively [Sherwood 1999]. Therefore, when deciding on whether to proceed with operative intervention specifically in children, it is important to consider the possibility of spontaneous closure [Farooki 1993].

In summary, surgical intervention in coronary artery cameral fistulas remains a safe and definitive treatment that prevents the development of complications. A high index of suspicion is often required to make the diagnosis. Surgical management can be curative in these challenging and rare cases.

REFERENCES

- Balaguru D, Joseph A, Kimmelstiel C. 2006. Occlusion of a large coronary-cameral fistula using the Amplatzer vascular plug in a 2-year old. *Catheter Cardiovasc Interv* 67:942-6.
- Biörck G, Crafoord C. 1947. Arteriovenous aneurysm on the pulmonary artery simulating patent ductus arteriosus Botalli. *Thorax* 2:65-90.
- Davis JT, Allen HD, Wheller JJ, et al. 1994. Coronary artery fistula in the pediatric age group: a 19-year institutional experience. *Ann Thorac Surg* 58:760-3.
- Dodge-Khatami A, Mavroudis C, Backer CL. 2000. Congenital Heart Surgery Nomenclature and Database Project: anomalies of the coronary arteries. *Ann Thorac Surg* 69(4 Suppl):S270-97.
- Farooki ZQ, Nowlen T, Hakimi M, Pinsky WW. 1993. Congenital coronary artery fistulae: a review of 18 cases with special emphasis on spontaneous closure. *Pediatr Cardiol* 14:208-13.
- Kale SB, Reddy V, Patil R. 2006. Iatrogenic coronary-cameral fistula following intracardiac repair of tetralogy of Fallot: case report. *Indian J Thorac Cardiovasc Surg* 22:25-27.
- Krause W. 1865. Ueber den Ursprung einer Accessorischen A. coronaria cordis aus der A. pulmonis. *Z Rationelle Med* 24:225.
- Lau G. 1995. Sudden death arising from a congenital coronary artery fistula. *Forensic Sci Int* 73:125-30.
- Levin DC, Fellows KE, Abrams HL. 1978. Hemodynamically significant primary anomalies of the coronary arteries: angiographic aspects. *Circulation* 58:25-34.
- Liberthson RR, Sagar K, Berkoben JP, Weintraub RM, Levine FH. 1979. Congenital coronary arteriovenous fistula: report of 13 patients, review of the literature and delineation of management. *Circulation* 59:849-54.
- Mavroudis C, Backer CL, Rocchini AP, Muster AJ, Gevitz M. 1997. Coronary artery fistulas in infants and children: a surgical review and discussion of coil embolization. *Ann Thorac Surg* 63:1235-42.
- McLellan BA, Pelikan PC. 1989. Myocardial infarction due to multiple coronary-ventricular fistulas. *Cathet Cardiovasc Diagn* 16:247-9.
- Mestre Barceló JL, Salido Tahoces L, del Río del Busto A, Camino López A, Moya Mur JL, Pey Illera J. 2004. Closure of an iatrogenic coronary artery fistula with a PTFE-coated stent [in Spanish]. *Rev Esp Cardiol* 57:699-701.
- Reul RM, Cooley DA, Hallman GL, Reul GJ. 2002. Surgical treatment of coronary artery anomalies: report of a 37 1/2-year experience at the Texas Heart Institute. *Tex Heart Inst J* 29:299-307.
- Said SA, van der Werf T. 2006. Dutch survey of coronary artery fistulas in adults: congenital solitary fistulas. *Int J Cardiol* 106:323-32.
- Sakakibara S, Yokoyama M, Takao A, Nogi M, Gomi H. 1966. Coronary arteriovenous fistula: nine operated cases. *Am Heart J* 72:307-14.
- Sandhu JS, Uretsky BF, Zerbe TR, et al. 1989. Coronary artery fistula in the heart transplant patient: a potential complication of endomyocardial biopsy. *Circulation* 79:350-6.
- Sherwood MC, Rockenmacher S, Colan SD, Geva T. 1999. Prognostic significance of clinically silent coronary artery fistulas. *Am J Cardiol* 83:407-11.
- Somers JM, Verney GI. 1991. Coronary cameral fistulae following heart transplantation. *Clin Radiol* 44:419-21.
- Urrutia-S CO, Falaschi G, Ott DA, Cooley DA. 1983. Surgical management of 56 patients with congenital coronary artery fistulas. *Ann Thorac Surg* 35:300-7.
- Yamanaka O, Hobbs RE. 1990. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. *Cathet Cardiovasc Diagn* 21:28-40.