

## Case Report

# Surgical treatment of a bilateral aneurysmatic aortic/coronary-to-pulmonary fistula associated mitral and tricuspid valve regurgitation: a rare case report

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**Abstract:** Coronary artery fistula is a rare abnormality which may cause severe complications such as congestive heart failure, infective endocarditis and even cardiac tamponade. We report a case of successful surgical treatment of a 63-year-old man with bilateral coronary artery to pulmonary fistulas, complicated by coronary aneurysm, severe mitral regurgitation, tricuspid regurgitation, atrial fibrillation, pulmonary arterial hypertension. We have documented a rare case of CAF arising from bilateral coronary connecting to the pulmonary and concomitant severe MR, TR, AF and PAH. We incised the main pulmonary artery, and a terminal fistula with 4-mm diameter was identified. We closed this fistula with pledgetted mattress sutures using two pairs of 5-0 prolene. Then the thin wall of the aneurysm was partially excised, no addition connection was found, we sutured and reinforced it with 7-0 prolene and opened pulmonary artery was repaired. Through a trans-septal approach, the mitral valve was repaired by using a size 32 Edwards C-E Physio Annuloplasty Ring and the tricuspid was repaired with a size 32 Edwards MC3 Tricuspid Annuloplasty Ring. During operation, no hemodynamic instability was observed. The postoperative course was uneventful and patient was discharged in an excellent condition on postoperative day 8. At six-month follow-up, he was asymptomatic, and the repeat echocardiogram showed a normally sized and functioning heart with normal LVEF (72%), without any residual shunt and valvular regurgitation. In conclusion, due to the rarity of such a condition, the careful assessments and appropriate therapeutic strategies should be tailored to the patients.

**Keywords:** Bilateral coronary artery fistulas, coronary aneurysm, pulmonary fistulas

## Introduction

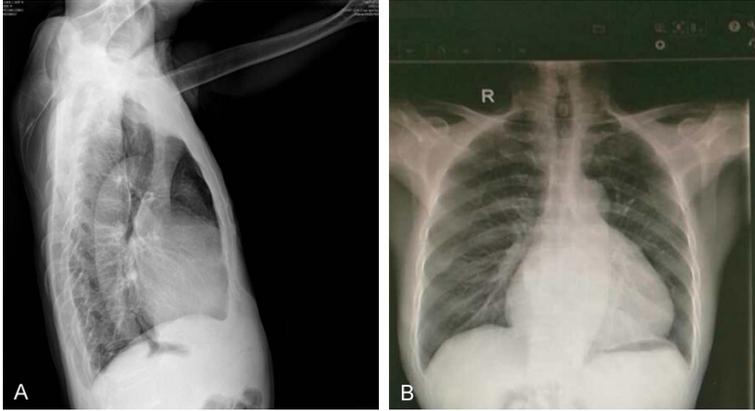
Coronary artery fistula (CAF) is a rare cardiac anomaly with a reported incidence of 0.1% to 0.2% in the adult population referred for cardiac catheterization. The right coronary artery (RCA) is the most common site of origin (approximately 50%), followed by the left anterior descending artery (LAD, approximately 40%) [1, 2]. Most cases are congenital, but previous reports have indicated that CAF can also be acquired secondary to trauma or from invasive cardiac procedures. Fistulous connections of the coronary artery into a cardiac chamber or major vessel often causes a longstanding left to right shunt, which can produce significant volume overload, with progressive dilatation of both mitral and tricuspid annuli, with consequent severe mitral regurgitation (MR) and tri-

cuspid regurgitation (TR). To the best of our knowledge, bilateral coronary fistulas concomitant with coronary aneurysm, MR, TR, atrial fibrillation (AF), pulmonary arterial hypertension [3, 4], as shown in the current case, is the first report in the literature.

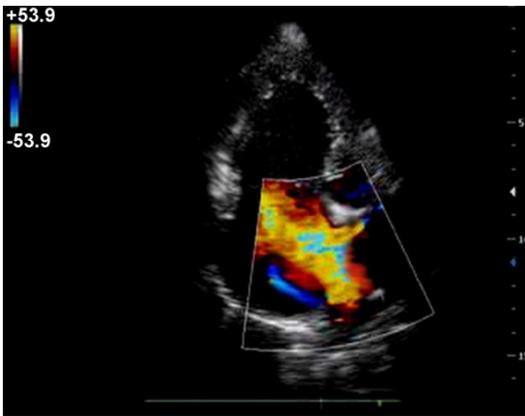
## Case report

A 63-year-old man was admitted to our hospital with progressive dyspnea on exertion for more than two years. His medical history involved Billroth's subtotal gastrectomy because of duodenal ulcer at the age of 33. There was no family history of congenital heart disease and no history of trauma. The vital signs were normal. On auscultation, his heart rate was irregular, a grade 4/6 systolic murmur could be heard at the apex of heart, as well as a grade 3/6 con-

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**Figure 1.** Chest X-ray radiography (XR) showed the increased cardiothoracic ratio. A. The lateral chest XR image. B. The frontal chest XR image.



**Figure 2.** Transthoracic echocardiography (TTE) illustrated the anterior mitral leaflet.

tinuous murmur over the second left sternal border. Electrocardiogram showed atrial fibrillation with low voltage QRS. Chest X-ray radiography (XR) showed an increased cardiothoracic ratio of 0.7 with mild pulmonary congestion (**Figure 1**). Transthoracic echocardiography (TTE) (**Figure 2**) demonstrated the anterior mitral leaflet was prolapsed resulting in severe regurgitation, moderate tricuspid regurgitation was also present. Left ventricular function was normal (LVEF, 61%), with a giant left atrium (43 × 60 × 94 mm) and enlarged right atrium (47 mm). The mean pulmonary arterial pressure was 28 mmHg. Coronary angiography confirmed a fistula arising from the dilated right aortic sinus of Valsalva (**Figure 3A**). The fistula had a tortuous trace, formed an aneurysm of 4 mm and merged with a second fistula arising from the proximal LAD (**Figure 3B**) and drained into the main pulmonary artery. The rest of the

coronary arteries were not dilated and no significant atherosclerotic stenosis or thrombosis were found. All laboratory values were unremarkable.

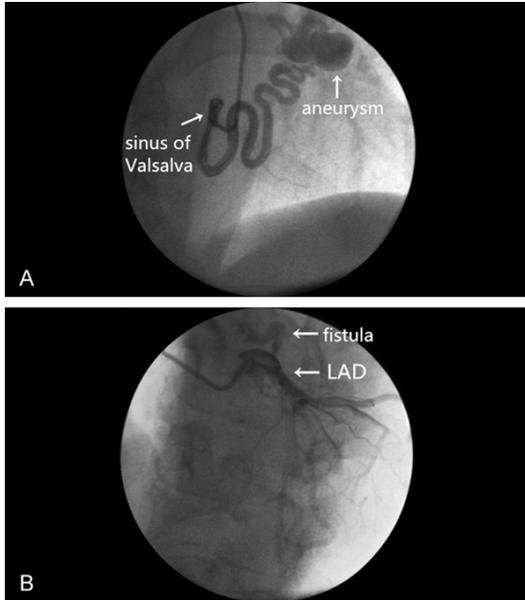
Through a median sternotomy, cardiopulmonary bypass (CPB) was established with aortic, superior and inferior vena cannulation. Cardiac arrest was achieved by using antegrade blood cardioplegia every 20 min. We incised the main pulmonary artery, and a terminal fistula with 4-mm diameter was identified. We closed this fistula with pledgetted mattress sutures using two pairs of 5-0 prolene. Then the thin wall of the aneurysm was partially excised, no additional connection was found, we sutured and reinforced it with 7-0 prolene and opened pulmonary artery was repaired. Through a trans-septal approach, the mitral valve was repaired by using a size 32 Edwards C-E Physio Annuloplasty Ring and the tricuspid was repaired with a size 32 Edwards MC3 Tricuspid Annuloplasty Ring. We didn't do the MAZE procedure because the patient refused. The cross-clamp time and cardiopulmonary bypass time were 99 min and 147 min, respectively. During the operation, no hemodynamic instability was observed. The postoperative course was uneventful and the patient was discharged in an excellent condition on postoperative day 8. At six-month follow-up, he was asymptomatic, and the repeat echocardiogram (**Figure 4**) showed a normally sized and functioning heart with normal LVEF (72%), without any residual shunt and valvular regurgitation.

During the operation, no hemodynamic instability was observed. The postoperative course was uneventful and the patient was discharged in an excellent condition on postoperative day 8. At six-month follow-up, he was asymptomatic, and the repeat echocardiogram (**Figure 4**) showed a normally sized and functioning heart with normal LVEF (72%), without any residual shunt and valvular regurgitation.

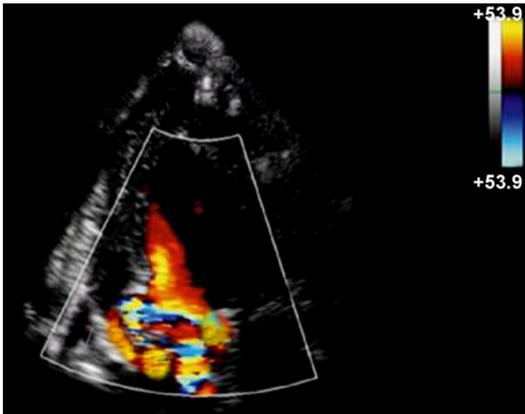
### Discussion

CAFs are not common coronary artery abnormalities. Most of CAFs are unilateral, but bilateral or multiple fistulas are very rare entities. Among the 54 patients reported by Canga Y etc, bilateral fistula was mentioned in 3 and bilateral aneurysms were found in only 1 of all patients [4]. The longstanding shunt may cause coronary arteries and heart chambers enlarged progressively and increase the volume overload, subsequently resulting in MR, TR

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**Figure 3.** Bilateral fistulas from dilated right aortic sinus of Valsalva (A) and left anterior descending artery (B), formed an aneurysm, draining into the pulmonary artery.



**Figure 4.** The echocardiogram at six-month follow-up illustrated a normally sized and functioning heart.

and PAH. In addition, myocardial ischemia due to a coronary steal phenomenon may also occur, which can worsen MR. Eventually, congestive heart failure is inevitable. In extremity, CAF may rupture, leading to cardiac tamponade and sudden death [5].

Coronary angiography is still the gold standard method for the diagnosis of CAF. The precise features of CAFs, including runoff, shunt size, and the route of origin and drainage can be showed. However, some non-invasive methods

such as TTE, transesophageal echocardiography (TEE), and MDCT can also play important role in the diagnosis of CAFs. Echocardiography can provide clues such as coronary dilation, chamber enlargement, and color Doppler supplies evidence with abnormal blood flow. MDCT even provides more detailed than selective coronary angiography in some aspects but with minimal trauma.

The management of CAF is still controversial [6]. Many CAFs are small and have no clinical manifestation, they do not need treatment immediately. However, when the patient is symptomatic or serious complications are concomitant, CAFs are recommended to be closed as early as possible. The main treatments include surgical closure and transcatheter occlusion. Surgical intervention is safe, and is strongly recommended in cases with complications and other cardiac abnormalities. Surgical correction of the CAF in this case is indicated to prevent progressive congestive heart failure, significant aneurysmal formation with potential rupture or embolization. Transcatheter embolisation is regarded as a new way and is widely accepted by many people since it is less invasive. However, when misused, it can result in serious complications [7, 8]. When there are additional complex heart diseases needed to deal with at the same time, like this case, transcatheter embolisation is not suitable.

In summary, we have documented a rare case of CAF arising from bilateral coronary connecting to the pulmonary and concomitant severe MR, TR, AF and PAH, who was successfully treated by incising the aneurysm, closing the outflow orifice into the PA, and repairing the mitral and tricuspid valves. Due to the rarity of such a condition, we recommend that careful assessments and appropriate therapeutic strategies should be tailored to the patients.

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### Disclosure of conflict of interest

None.

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