

## Coronary Artery Fistulae

Gautam Reddy, MD; James E. Davies, MD; David R. Holmes, MD; Hartzell V. Schaff, MD; Satinder P. Singh, MD; Oluseun O. Alli, MD

### Case Presentation

Two patients presented to our cardiology clinic

Mr. H was 70 years old with a history of cardiac transplantation for ischemic cardiomyopathy. He presented with worsening dyspnea. Two years ago, he could run up 2 flights of stairs but had recently started to become short of breath while walking short distances. He also described pedal edema and progressive orthopnea. His medical history was significant for coronary artery disease. On examination, we noted an elderly male (6'0"; 85 kg). The heart rate was 90 bpm with a blood pressure of 156/90 mmHg. The SaO<sub>2</sub> was 98% on room air. The heart sounds were normal, but we auscultated a III/VI continuous machinery murmur over the left upper sternal border. His ECG showed sinus rhythm and left ventricular hypertrophy. The chest radiograph displayed mild cardiomegaly. We performed a transthoracic echocardiogram that was remarkable for concentric left ventricular hypertrophy with normal ejection fractions. Right heart catheterization revealed normal filling pressures and cardiac output. An endomyocardial biopsy showed no evidence of allograft rejection.

Ms. B was 75 years old and also presented with dyspnea. For the past 4 months, she had noted shortness of breath after walking ≈1 mi, whereas she was able to go much farther on her daily walks before the onset of symptoms. Her medical history was significant for hypertension, dyslipidemia, and migraines. There was no previous history of cardiac disease. On examination, we noted a frail elderly female (5'6"; 50 kg). Her heart rate was 62 bpm with a blood pressure of 132/90 mmHg. The SaO<sub>2</sub> was 99% on room air. The heart sounds were again normal but we auscultated 2 murmurs—one was a II/VI continuous murmur over the left upper sternal border and the other was a II/IV diastolic decrescendo murmur best heard in the aortic area. The ECG showed respiratory sinus arrhythmia. Her chest radiograph showed ascending aortic dilation. The transthoracic echocardiogram displayed aortic root dilation (5.5 cm) as well as a trileaflet aortic valve, moderate aortic regurgitation, and normal biventricular ejection fractions.

### Discussion

*Dr Reddy:* What Diagnostic Considerations Should We Entertain?

*Dr Holmes:* The presentation here is exertional dyspnea. The initial differential is broad and includes cardiac, pulmonary, hematologic, and metabolic abnormalities. However, the presence of the continuous murmur, assuming it is related to their symptoms, raises the possibility of a congenital or acquired intrathoracic arteriovenous communication.

A continuous murmur is produced by a shunt between 2 vascular systems with a marked difference in pressure throughout the cardiac cycle. These can include patent ductus arteriosus, ruptured sinus of Valsalva aneurysm, coronary arterial fistulae, pulmonary arterial fistulae, aortopulmonary windows, coarctation of the aorta, or branch pulmonary artery (PA) stenosis.

*Dr Reddy:* What diagnostic modalities would you use?

*Dr Alli:* We could perform coronary angiography or use noninvasive modalities, such as cardiac computed tomographic (CT) or cardiac magnetic resonance (MR) imaging. Given that endomyocardial biopsies are known to result in iatrogenic coronary fistulae, I would recommend coronary angiography for the first patient. For the second patient, the differential remains wide, so I would recommend CT or MR instead. These modalities with their wide field of view can better evaluate many of our differentials.

*Dr Reddy:* Mr. H undergoes invasive coronary angiography. This shows a large left anterior descending (LAD) coronary artery with abnormal termination into the right ventricle (Figure 1; Movies I and II in the Data Supplement). Cardiac CT coronary angiography is performed for Ms. B. This shows a dilated aortic root but there is also an abnormal tortuous vascular communication originating from the LAD and terminating into the PA (Figure 2; Movie III in the Data Supplement). What is your diagnosis at this point?

*Dr Alli:* These are coronary fistulae that are rare anomalies. The incidence ranges from 0.05% to 0.9% in several large selected series.<sup>1,2</sup> They are defined as abnormal communications between coronary arteries and vascular structures without an interposed capillary bed. They are classified into coronary cameral fistulae, where the fistula terminates in a cardiac chamber, and coronary arteriovenous fistulae, where the fistula terminates into a venous structure. These include the vena cavae, coronary sinus, bronchial veins, and PA. We use a comprehensive classification to describe these anomalies (Table). Mr. H therefore has a complex Sakakibara type B

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From the Division of Cardiovascular Diseases, The University of Alabama, Birmingham (G.R., J.E.D., S.P.S., O.O.A.); and Division of Cardiovascular Diseases, The Mayo Clinic, Rochester, MN (D.R.H., H.V.S.).

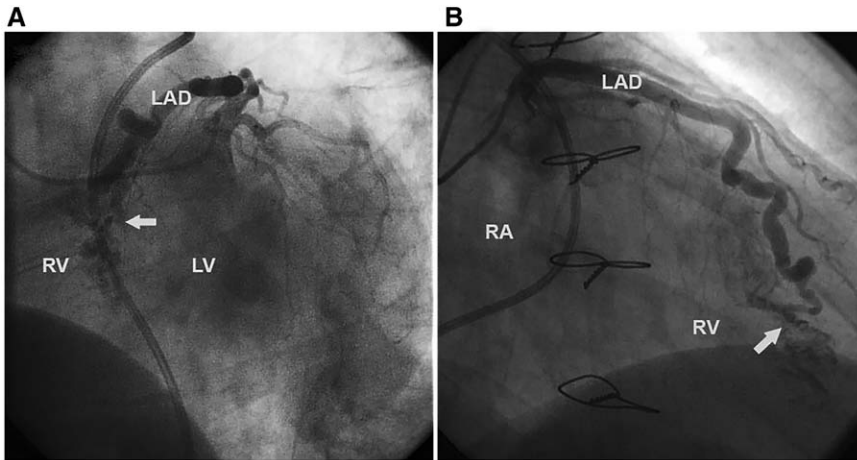
The Data Supplement is available at <http://circinterventions.ahajournals.org/lookup/suppl/doi:10.1161/CIRCINTERVENTIONS.115.003062/-/DC1>. Correspondence to Oluseun O. Alli, MD, Division of Cardiovascular Diseases, University of Alabama at Birmingham Cardiovascular Medicine, 510 20th St, S FOT 920 Birmingham, AL 35294. E-mail [oalli@uab.edu](mailto:oalli@uab.edu)

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**Figure 1.** Invasive coronary angiography showing complex Sakakibara type B fistula (arrow) from the terminal left anterior descending (LAD) into the right ventricle (RV). The proximal native vessel is dilated and tortuous. **A**, Left anterior oblique projection differentiates between right- and left-sided chambers as site of termination (arrow). This fistula terminates into a right-sided cardiac chamber. **B**, Right anterior oblique projection differentiates between the right atrium (RA) and the RV as the site of termination (arrow).

coronary cameral fistula from the LAD to the right ventricle, whereas Ms. B has a simple Sakakibara type A coronary arteriovenous fistula from the LAD to the PA.<sup>3</sup>

*Dr Reddy:* What is the best way to delineate the origin and termination of a coronary arteriovenous fistula during coronary angiography?

*Dr Holmes:* We begin with 2 projections: a straight right anterior oblique and a straight left anterior oblique. The former delineates whether the fistula is terminating into the atria or the ventricles. The latter will place the septum end on and differentiate between fistulae terminating in the right and left sides of the heart. Fistulae terminating into venous vessels or the coronary sinus are harder to localize. We can insert a balloon-tipped catheter into the PA and a pigtail catheter into the aortic root to provide localizing landmarks. Further angiographic projections are used to lay out the fistula's origin and course. It is not always easy, especially when the fistula is large, tortuous and overlaps its own origin and termination. In such cases, we may rely on CT or MR imaging with 3-dimensional (3D) coronary reconstruction to completely delineate the fistula.

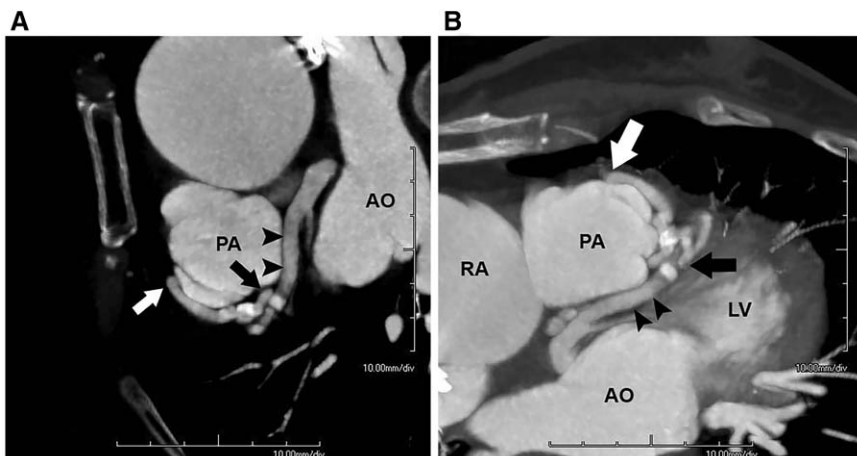
*Dr Reddy:* We know that Mr. H had several normal coronary angiograms before this presentation, and that Ms. B has no previous history of coronary or cardiac disease. Could you comment on the etiologies of the coronary fistulae in these cases?

*Dr Alli:* Coronary fistulae can be either congenital or acquired. The majority, and I estimate this to be >90% of all

fistulae, are congenital and sporadic. Acquired fistulae are rare. They can result from a variety of procedures, including coronary stenting, structural interventions, pacemaker placement, radiofrequency ablation, coronary bypass grafting, valve surgeries, epicardial procedures, endomyocardial, or transbronchial biopsies. They have also been described secondary to infective endocarditis, coronary vasculitis, and penetrating chest trauma.<sup>4</sup> Mr. H probably has an endomyocardial biopsy-related coronary fistula, whereas Ms. B probably has a congenital coronary fistula.

*Dr Reddy:* What symptoms do coronary fistulae produce?

*Dr Holmes:* Coronary fistulae can present with protean manifestations. Systemic hemodynamic effects result from shunting of blood through the fistula and the resultant high output state. Fistulae producing left to right shunts, such as those terminating in the right-sided chambers or pulmonary arteries, can lead to pulmonary hypertension or biventricular volume overload depending on the fistula size and flow pattern. Large fistulae that produce left to left shunts, such as coronary to left atrium or left ventricular fistulae, produce left ventricular volume overload. The clinical presentation is usually that of heart failure. Coronary fistulae can also produce disturbances in coronary hemodynamics. The low pressure sink of the fistula results in preferential flow through it and can produce a coronary steal phenomenon in the distal native coronary artery. The response of the coronary artery is to undergo



**Figure 2.** Computed tomographic coronary angiography showing a large simple Sakakibara type A fistula (arrow) from the left anterior descending (arrowheads) into the pulmonary artery (PA). **A**, Transverse section. **B**, Coronal section. The site of origin (black arrow) and site of termination (white arrow) are clearly delineated. AO indicates aorta; LV, left ventricle; and RA, right atrium.

**Table. Classification of Coronary Artery Fistulas**

Vessel of origin
Left coronary artery
Left anterior descending
Ramus intermedius
Left circumflex coronary
Right coronary artery
Anomalous coronaries
Segment of origin
Sakakibara type A: originating from the proximal third of the native vessel
Sakakibara type B: beyond proximal third or as continuation of native vessel
Site of termination
Coronary arteriovenous fistulas
Vena cavae
Coronary sinus
Pulmonary artery
Pulmonary veins
Bronchial vessels
Other (azygos, costal, brachiocephalic veins etc)
Coronary cameral fistulas
Left atrium
Right atrium
Left ventricle
Right ventricle
Mode of termination
Macrofistulas: discrete fistulous vessel visible on invasive angiography
Microfistulas: visible contrast blush without discrete fistulous vessel
Fistula morphology
Simple: fistula with single origin, single vessel, and single termination
Complex: other fistula morphologies including plexiform variants
Fistula number
Single
Multiple
Angiographic sizing (applicable to simple macrofistulas only)
Large: >2× distal reference vessel diameter
Medium: 1–2× distal reference vessel diameter
Small: <1× distal reference vessel diameter
Physiological abnormalities
Vascular shunting
Left to right physiology (pulmonary hypertension)
Left to left physiology (left ventricular overload)
Coronary steal phenomenon
Acute intrinsic changes
Thrombosis (with possible distal coronary embolization)
Dissection
Vasospasm
Rupture (with possible hemopericardium and tamponade)
Chronic intrinsic changes
Aneurysm formation
Vascular calcification
Mediastinal compression

dilation proximal to the fistula, but this may not compensate for the steal. These patients present with chronic angina.

Intrinsic pathophysiologic changes in the fistula are uncommon. Acute changes can include thrombosis, embolism, and dissection. Extension of these conditions into the native vessel can produce acute coronary syndromes. Intrapericardial rupture of the fistula usually leads to hemopericardium and tamponade. Superinfection produces infective endarteritis. Chronic changes include dilation and ectasia, which can rarely produce symptoms by compression of mediastinal structures.

Arrhythmic symptoms such as palpitations or syncope may also occur. Atrial fibrillation is perhaps the most common arrhythmia although almost any supraventricular or ventricular arrhythmia can occur, especially once the atria and ventricles start to dilate. Presentation with sudden cardiac death before the onset of symptoms is rare, but the exact incidence is unclear.

*Dr Reddy:* Our patients do not seem to have significant comorbidities that could otherwise explain their symptoms of exertional dyspnea. The creation of Mr. H's fistula seems to correlate temporally with his symptoms. However, as Ms. B's fistula is thought to be congenital, how do we explain her late presentation?

*Dr Alli:* The progressive enlargement of coronary fistulae can lead to insidious onset of symptoms, and many patients initially underestimate the severity of their functional impairment. In general, the size of fistula dictates the severity of symptoms. I have seen neonates with large fistulae presenting at birth, and have incidentally discovered small fistulae in asymptomatic octogenarians. Overall, the clinical course is highly variable and rather unpredictable, but the majority of fistulae enlarge over time and eventually become symptomatic.

*Dr Reddy:* Our patients have symptoms attributable to their coronary fistulae, that is, progressive exertional dyspnea. They have no other comorbidities that could be treated or optimized to improve their symptoms. Before we consider fistula closure, is there a role for medical management to slow progression or ameliorate symptoms?

*Dr Alli:* There is no medical therapy proven to slow the progression of coronary fistulae. In rare cases where the risks of intervention absolutely outweigh the benefits, therapy should be directed at the underlying pathophysiology. Because shunt-induced volume overload results in neurohormonal activation mechanisms similar to that in systolic heart failure, consider treatment with diuretics,  $\beta$ -adrenergic blockers and angiotensin-converting enzyme inhibitors. In patients with coronary steal, therapy should be directed at decreasing myocardial oxygen consumption. However, vasodilators such as nitrates might dilate the fistula, which can worsen steal symptoms. Fistula-related arrhythmias can be treated with standard anti-arrhythmic therapy but may not respond. Overall, the evidence base for medical therapy in this setting is extremely limited.

Existing guidelines recommend intervening on all large fistulae irrespective of symptoms.<sup>5</sup> This reflects their propensity to become symptomatic. For small to medium fistulae, the recommendation is to intervene if they are symptomatic. However, the guidelines do not provide a guide to sizing. We size all simple fistulae (single origin, 1 vessel, and single termination) during coronary angiography by using the native vessel as a reference. For complex fistulae, we do not use

size as a criterion for intervention. A minority of fistulae may remain stable and asymptomatic and can be followed clinically. Meticulous history and physicals should be the mainstay of interval evaluation in these cases. We use treadmill testing or cardiopulmonary exercise testing if we suspect occult symptoms. The onset of new cardiac symptoms in the presence of a known coronary fistula generally mandates angiography.

*Dr Reddy:* A discussion was conducted with the patients' where the diagnosis, prognosis, and treatment options, including medical therapy, transcatheter intervention, and surgical closure were described. Please tell us about your thought process before this discussion.

*Dr Holmes:* More information was needed to decide what route of closure was viable in each case and to estimate the risk benefit ratio of an intervention. We begin with the history and physical, laboratory investigations, electrocardiography, and chest radiograph. Echocardiography is used to evaluate the physiological effects of the fistula and to screen for coexisting cardiac conditions. This is followed by CT or MR coronary imaging with 3D reconstruction, which provides exquisitely detailed images that are invaluable for procedural planning. This is also useful when other cardiac anomalies or extracardiac diagnoses are suspected. We require that patients have invasive coronary angiography before closure because finer details such as blood flow patterns, device landing zones, or surgical ligation sites may not be fully evident on CT or MR images. This allows complete procedural planning.

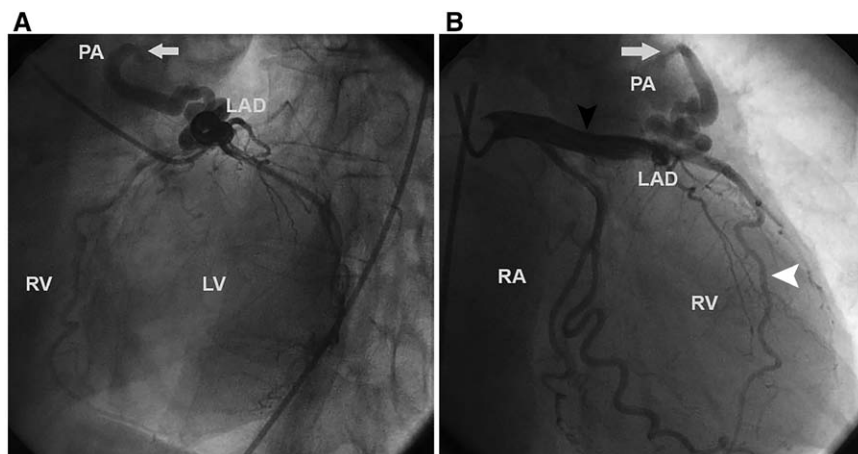
Depending on the individual's comorbidities, we may perform additional studies such as pulmonary function testing, aortic angiography, and right heart catheterization. It is important to note that the invasive shunt fraction is unreliable for fistulae that terminate in the PA because proper sampling of true mixed venous blood is rarely possible in this case. Also, there are no defined criteria for shunt fraction, invasive or non-invasive, that correlates with symptoms because of coronary fistulae. Therefore, we do not recommend the measurement of shunt fraction to guide closure. There is no evidence supporting the use of intracoronary pressure or flow measurements to guide fistula intervention, although these may still be used to guide angioplasty of concomitant coronary artery disease. Overall because this is an elective setting, it is important to exclude differential diagnoses and optimize comorbidities.

*Dr Reddy:* Nowadays it is rare to find a patient who does not have comorbidities, and we are frequently confronted with decision making in the setting of complex cardiopulmonary disease. How would you evaluate a patient with symptoms of cardiac disease who also had multiple cardiac comorbidities in addition to a coronary fistula?

*Dr Holmes:* Coronary fistulae can produce nonspecific symptoms, making this a difficult question to answer. When there is coexistent myocardial, valvular, pericardial, or atherosclerotic disease, it may be impossible to quantify the fistula's contribution to the patient's symptom burden. The question of obvious or occult pulmonary, hematologic and metabolic diseases, or deconditioning may also arise. I would recommend cardiopulmonary exercise stress testing in such cases. We also obtain other tests based on the nature of the comorbidities. One must then weigh the contributions from the comorbidities and optimize each one medically. If symptoms persist, I would intervene on one abnormality at a time and then reevaluate. As to which abnormality to intervene on first, this is where clinical judgment must be used. It is important to look at the characteristics of the fistula, ask whether it is compatible with the presenting symptoms, and consider the risk/benefit ratio of intervention. In the end, especially if the fistula is large and symptoms are persistent, I would empirically close the fistula.

*Dr Reddy:* Ms. B underwent coronary angiography (Figure 3; Movies IV and V in the Data Supplement). Both patients elected to pursue closure of their fistulae. A transcatheter approach was recommended for Mr. H, whereas surgical closure was recommended for Ms. B. Please tell us how the route of closure was selected in each case.

*Dr Schaff:* Coronary artery fistulae can be closed by surgical methods or transcatheter techniques. Because acute changes necessitating emergent intervention is rare, I will limit the discussion here to elective management. There is little data to compare these 2 routes, because closure volumes are too low to support a randomized trial and there is no multicenter registry either. The existing guidelines based largely on expert opinion, which do not indicate a preference for either route. We must emphasize that this is one area where clinical expertise is critical for good outcomes; indeed, the guidelines recommend that closure be performed at centers of excellence by structural interventionalists and cardiac surgeons with



**Figure 3.** Invasive coronary angiography. **A**, The left anterior oblique projection shows fistula coursing superiorly. **B**, The right anterior oblique projection shows the fistula terminating in the main pulmonary artery (PA). The proximal vessel is dilated and tortuous (black arrowhead). The distal vessel is underfilled with contrast because of high volume runoff into the fistula (white arrowhead). LAD indicates left anterior descending; LV, left ventricle; RA, right atrium; and RV, right ventricle.

specific expertise in this procedure. Regardless of the route of closure, we advise multidisciplinary consultation between the patient, a structural interventionalist and a cardiac surgeon, in a manner similar to the Heart Team approach.<sup>6</sup>

We default to the transcatheter route given its minimally invasive nature and excellent outcomes. However, there are several factors that need to be considered first. We use an algorithm that incorporates 2 critical questions (Figure 4). (1) The fistula amenable to transcatheter closure and (2) Surgical intervention mandated for coexisting conditions? If the answers are yes and no, respectively, we recommend transcatheter closure. When surgical intervention is necessary, and especially when a concomitant procedure is required, predicted surgical mortality should be taken into consideration. When this exceeds 10%, we consider hybrid interventions or transcatheter closure of the fistula plus another transcatheter procedure, such as transcatheter aortic valve replacement, transcatheter mitral valve repair, alcohol septal ablation, endovascular aneurysm repair, etc when indicated. Hostile chests or frail patients also negatively influence the decision to perform open surgical repair. In some cases, robotic approaches may reduce surgical risk.

Both patients here had symptoms attributable to their coronary fistulae, and closure was therefore indicated. Mr. H had a coronary artery fistula of the distal LAD into the right ventricle. This provided a stable landing zone. He did not have other cardiac conditions that would mandate surgery. We recommended transcatheter closure for him. Ms. B had a discrete fistula originating from the LAD, providing a discrete device landing zone, but her aortic root aneurysm was a concrete indication for surgical intervention. We recommended surgical closure for her. With careful selection and meticulous technique, success rates for closure approach 90% to 100%.

*Dr Reddy:* Mr. H underwent transcatheter closure of his fistula (Figure 5; Movies VI and VII in the Data Supplement). Please tell us how you planned and executed the closure.

*Dr Alli:* Once a fistula is selected for transcatheter closure, we refine the procedure. The body of the fistula must present a suitable landing zone for the selected device. The landing zone must be some distance away from the fistula ostium. If the device interferes with the native coronary, in situ thrombosis and distal embolization, dissection or spasm might occur. The landing zone must also be some distance from the fistula termination. If the device dislodges, embolization into the systemic circulation will result, potentially with catastrophic outcomes. The potential for device embolization is higher if the fistula does not narrow toward its termination. Additional characteristics, such as tortuosity, aneurysms, ramification, and marked calcification can challenge even skilled interventionalists.

Device selection is linked to landing zone morphology. Our standard approach for fistulas <5 mm in diameter is to use 0.018" hydrophilic vascular coils as the first line device. They are versatile to size and easy to deliver. The electronically detachable types are retrievable and repositionable before release, making them uniquely suited for the coronary setting in a try before you buy fashion. These are introduced through 2.8 French hydrophilic microcatheters placed in the landing zone over a 0.014" coronary guidewire. This system is advanced through a 6 French guiding catheter selected based on the patient's anatomy. For larger fistulae, we use a framing strategy where a large coil is used to create a framework on which we land smaller sealing coils. It is imperative that the coil be large enough to preclude distal embolization but small enough to allow complete deployment. We typically oversize the first coil to 1.5x of the landing zone diameter. When the

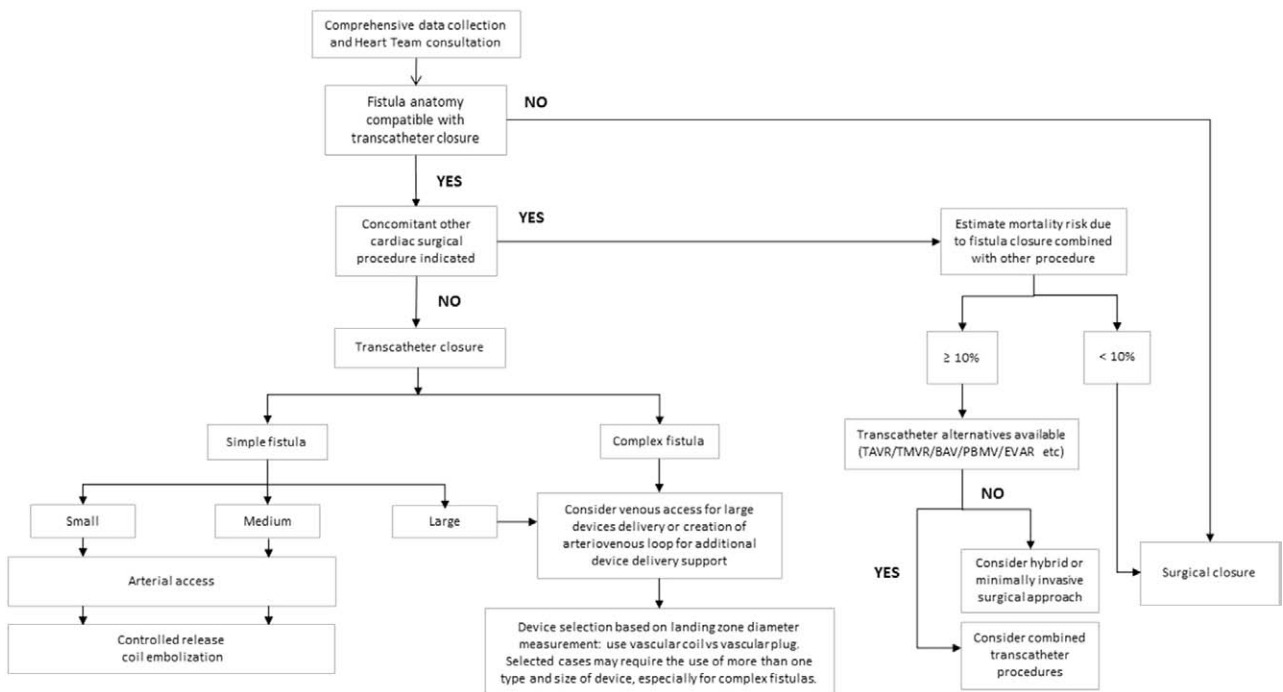
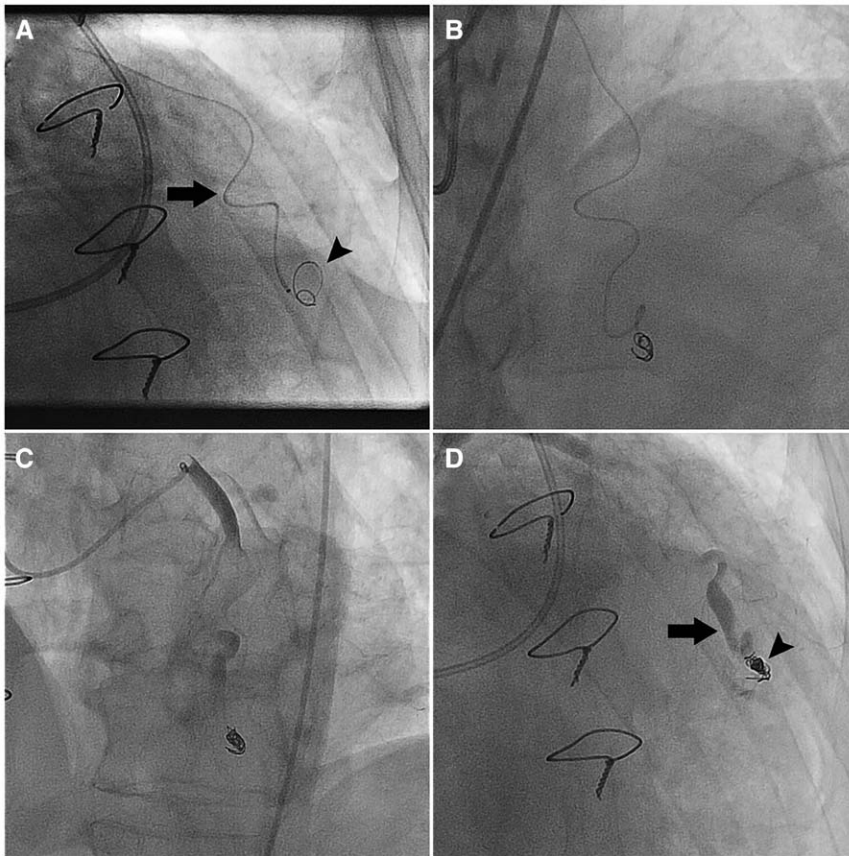


Figure 4. Algorithm for selection of coronary fistula closure method.



**Figure 5.** Transcatheter closure of left anterior descending (LAD) to right ventricle fistula. **A**, Insertion of microcatheter (arrow) over guidewire (arrowhead). **B**, Framing coil. **C**, Additional coils. **D**, Stasis of flow in the LAD (arrow) because of successful fistula occlusion (arrowhead).

fistula is  $>5$  mm in diameter, we may consider using devices, such as patent ductus occluders or vascular plugs.<sup>7</sup> We do not use embolization particles, detachable balloons, Grifka bags, or Rashkind umbrellas. When the landing zone is inadequate, we may deploy covered stents in the native coronary to occlude the ostium of the fistula. This mandates dual antiplatelet therapy and runs the risk of stent thrombosis. We use therapeutic heparin anticoagulation during all transcatheter closure procedures.

After device selection, we plan vascular access. For simple fistulae, this can be done by either an arterial (retrograde) or venous (antegrade) approach. We prefer the venous route when larger sheath sizes (8 Fr+) are anticipated for the selected device. We use the transfemoral route for most arterial cases, although we have used the radial approach when femoral vessels were inadequate. If the fistula is complex or large and tortuous, device delivery can be difficult. In such cases, we plan for the creation of an arteriovenous loop across the fistula for additional support. Once access is achieved, we use a standard 0.014 coronary guidewire to access the fistula, either by the antegrade or by the retrograde route. If we are using coils, we insert the microcatheter over the wire and systematically deliver coils until cessation of flow is achieved. For other devices we land a delivery sheath in the fistula and then deploy the selected device. The ideal situation at the end of the procedure is no flow or minimal flow in the fistulous vessels.

*Dr Reddy:* Mr. H was noted to have extremely sluggish flow in the native LAD coronary artery with threatened native vessel thrombosis after delivery of vascular coils. Could we discuss the underlying pathophysiology here?

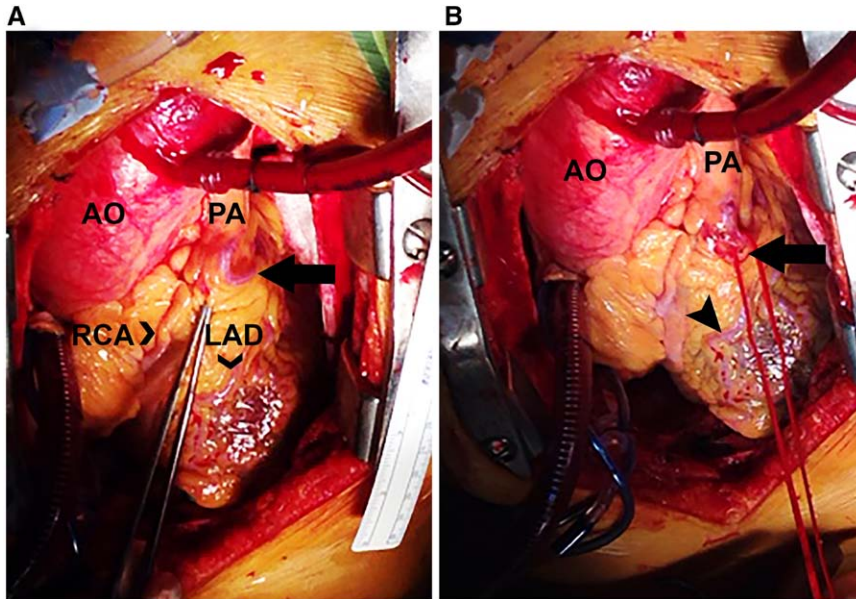
*Dr Holmes:* The Sakakibara fistula classification is important here. This depends on the site of origin of the fistulae. Type A fistulae originate from the proximal third of the native coronary. These are typically large complex coronary arteriovenous fistulae. Type B fistulae arise beyond the proximal third or represent terminal connections of the native coronary artery. These are typically small coronary cameral fistulae. The section of the native coronary proximal to the fistula is usually dilated. Mr. H had a type B fistula, where his dilated native LAD was terminating into the right ventricle. Because the high flow through the fistula was going through the entire length of the native coronary, closure of the fistula decreased the volume flow rate, and predisposed to stasis in the native coronary. This phenomenon carries a high risk of coronary thrombosis.<sup>8</sup>

*Dr Reddy:* How do you manage cases of slow flow in the short and long term?

*Dr Holmes:* Once we detect this phenomenon, we begin anticoagulation immediately, starting with unfractionated or low molecular weight heparin and then transitioning to warfarin for a goal international normalized ratio of 2.0 to 3.0. In the absence of bleeding, we recommend anticoagulation for 6 months. This allows enough time for the native coronary to remodel and for flow to normalize, at which point anticoagulation can be discontinued.

*Dr Reddy:* What are some other complications that may result from this procedure?

*Dr Holmes:* We have discussed the slow flow phenomenon related to type B fistulae. For type A fistulae, propagation of



**Figure 6.** Surgical closure of left anterior descending (LAD) to pulmonary artery (PA) fistula. The communicating vessel (arrow) courses toward the PA. The proximal coronary is dilated and the distal coronary is of normal caliber (arrowhead). Right coronary artery is also visible (RCA). **A**, Identification of fistula. **B**, Epicardial ligation. AO indicates aorta.

thrombus or device embolization down the native coronary can cause acute thrombosis and periprocedural myocardial infarction. This necessitates acute thrombus management or device retrieval. Other coronary complications include dissection and spasm. Coronary rupture can produce hemopericardium and tamponade. Transient ST segment changes and arrhythmias are frequent but rarely malignant. Death is extremely rare, even in high-risk individuals. Complications may also be related to vascular access and procedural sedation. In a series from our institute, overall complication rates were low.<sup>9</sup>

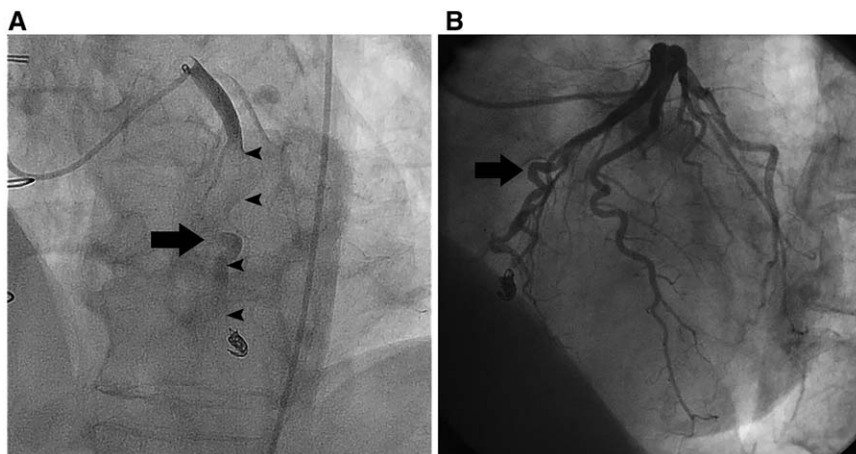
*Dr Reddy:* Ms. B underwent surgical closure of her fistula (Figure 6). Please tell us how you planned and executed the closure.

*Dr Davies:* First, we select the approach. Median sternotomy is preferred because it provides excellent visualization of the coronary vessels. When there is a single epicardial fistula on the lateral surface of the heart, closure may be accomplished via thoracotomy using open or robotic methods. The simplest method of closure is by epicardial interruption using ligatures or clips. This assumes that the fistula has enough structural integrity to sustain these materials. Large tortuous fistulae may have fragile walls, especially in case of

aneurysmal change. In such cases plication or excision may be required. Coronary artery bypass grafting is required when the native vessel is extensively malformed and likely to thrombose after closure. When the fistula is hard to localize epicardially, cardiopulmonary bypass and endocardial approaches are necessary. Endocardial closure is achieved by suture closure or patch placement. Hybrid approaches to locate drainage sites by placing a coronary wire through the fistula before surgery have been described.<sup>10</sup>

*Dr Reddy:* What complications can be anticipated after this procedure, other than those associated with general anesthesia, median sternotomy, and cardiopulmonary bypass?

*Dr Schaff:* Complications after surgical closure are higher than after transcatheter closure.<sup>11</sup> This may reflect selection bias because fistulae referred for surgical closure are usually not amenable to transcatheter closure or are associated with comorbid cardiac conditions requiring surgery. Specific complications include those seen with transcatheter closure, such as native coronary thrombosis or incomplete closure. If the fistula seen on echocardiography before the procedure, this may be used to confirm closure post procedure. We do not routinely perform coronary angiography to confirm closure



**Figure 7.** Flow profiles after transcatheter occlusion. **A**, Slow flow and prothrombotic state after closure (arrow). **B**, Remodeling of native vessel and flow improvement after 6 months (arrow).

unless the patient fails to improve or experiences complications after the procedure. It is also important to reevaluate the mitral and tricuspid valves during fistula closure. Especially when the ventricles are dilated, late valvular regurgitation occurs in a significant proportion of individuals and may require additional surgery after the index procedure.

*Dr Reddy:* Small residual fistulae are not uncommon after transcatheter or surgical closure. What is the outcome in these cases and how should we manage them?

*Dr Davies:* Small residual fistulae are rarely of clinical significance. In some cases, these residual may progressively enlarge over time and lead to symptoms, but this is rare in our experience. Another late problem may be the progressive enlargement of previously microscopic fistulae, but this should not be confused with recurrence of the original fistula. In either case, serial angiographic imaging of these entities is unnecessary. In the eventuality that residual fistulae produce clinical symptoms, they are generally amenable to transcatheter closure and rarely require repeat surgical intervention.

### Outcomes

*Dr Reddy:* Mr. H underwent transcatheter closure. He developed a slow flow state in the native vessel and was discharged home the next day on warfarin. Coronary angiography 6 months later showed normalization of flow, at which point anticoagulation was stopped (Figure 7; Movies VIII and IX in the Data Supplement). Ms. B underwent off-pump surgical closure via median sternotomy. Cardiopulmonary bypass was then initiated and a Bentall type composite aortic root replacement was performed. Her postoperative course was complicated by complete heart block for which a dual chamber permanent pacemaker was implanted. She was discharged home 5 days later. Both patients were seen in the outpatient clinic after 6 months. No late procedural complications were noted and their symptoms had completely resolved.

### Disclosures

None.

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KEY WORDS: cardiac surgery ■ congenital ■ coronary vessel anomalies ■ fistula ■ percutaneous coronary intervention



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Gautam Reddy, James E. Davies, David R. Holmes, Hartzell V. Schaff, Satinder P. Singh and  
Oluseun O. Alli

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