Percutaneous approaches to closure of coronary artery fistulas

Coronary artery fistula (CAF) is the most common hemodynamically significant congenital coronary anomaly. Blood is shunted from the coronary artery to a cardiac chamber, vein or intrathoracic vessel, bypassing the myocardial capillary network. The majority of the fistulas are small, and are detected incidentally and do not require intervention. The natural history of a CAF is variable and even spontaneous closure may be detected in rare cases. Treatment of CAF is indicated for symptomatic patients and for those asymptomatic patients with significant shunt or large fistulas that create risk for future complications, such as infective endocarditis, pulmonary hypertension or heart failure. Traditionally, surgery has been the main therapeutic method for the closure of CAF with proven efficacy and safety. However, since the first report of the use of percutaneous intervention in CAFs, many investigators have reported successful results using several techniques. Recent developments in the percutaneous closure methods have made this approach an effective and safe alternative to surgery, with the advantage of it being a less-invasive procedure requiring a shorter hospital stay with decreased costs and fewer complications.

KEYWORDS: coils ▪ coronary artery fistula ▪ detachable balloons ▪ occluder devices ▪ percutaneous closure ▪ surgical repair

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Coronary artery fistula (CAF) is an uncommon entity, which is associated with several potential complications, such as heart failure, myocardial ischemia, infective endocarditis and even rupture. Over the past few years a surgical approach has been the standard therapeutic option. However, recently, progress has been made in percutaneous interventional methods, with proven efficacy and safety.

In this article, we review the percutaneous methods used for the closure of a CAF. The comparison of several techniques is made to highlight the current treatment options in this rare abnormality.

Definition

Coronary artery fistula is defined as an abnormal communication – bypassing a capillary system – between a normal coronary artery and another cardiovascular structure, including cardiac chambers, veins or intrathoracic vessels. Previously, persistence of embryonal vascular sinusoids in the myocardium was the proposed mechanism for the formation of a CAF; however, the exact course of its embryogenesis is not yet clear [1]. According to the Hackensellner's involution–persistence theory, persistence of pulmonary buds with accompanying involution of aortic buds results in the formation of coronary arteries from pulmonary buds [2]. CAFs may originate from any of the three major coronary arteries, including the left main trunk. The most frequent site of origin is the right coronary artery, followed by the left anterior descending artery. Over 90% of CAFs drain into the low-pressure venous system or right heart chambers [3].

Epidemiology, classification & pathophysiology

■ Epidemiology

Coronary artery fistulas are rare abnormalities. The reported incidence of CAFs in the literature varies depending on the diagnostic method or ethnic group of the study population. As most CAFs are small or asymptomatic, true incidence is not known. Angiographic series reveal an incidence of 0.08–0.22% [4–7].

■ Classification

Coronary artery fistulas are divided into two groups: solitary CAFs and coronary artery left ventricular multiple microfistulas. Most of the solitary CAFs are congenital and not gender-specific [8]. In addition to congenital CAFs, acquired forms can be developed due to chest trauma, Takayasu arteritis, endomyocardial biopsy, cardiac surgery, pacing-lead erosion and complications during percutaneous coronary angioplasty procedures [9–14]. Although the size of the CAF is an important determinant of intervention, there is no consensus regarding the absolute size of a CAF for categorization. Latson *et al*. deliberately classified the CAF as: small fistulas (not larger than twice the diameter of the coronary artery at any point and do not cause coronary artery dilatation); medium-sized fistulas (larger than twice the diameter of the coronary artery but less than three-times as big); and large fistulas (larger than three-times the expected proximal normal coronary artery diameter) [15]. In the Dutch Registry, Said *et al.* defined small fistulas as having a vessel diameter of less than 2 mm, medium-sized fistulas between 2 and 8 mm and large fistulas over 8 mm [16].

■ Pathophysiology

'The coronary steal phenomenon' is the most widely accepted hypothesis for the ischemic symptoms of CAF. Myocardial segments distal to the fistula remain ischemic due to the diversion of coronary flow to the lower-pressure areas throughout the fistula and compensatory dilatation occurs in the proximal segment. High-flow fistula may also cause volume overload and increase pulmonary flow due to left-to-right shunt, resulting in heart failure or pulmonary hypertension. Usually, small CAFs have a benign natural history and even spontaneous regression has been reported [4,17]. However, large CAFs may progressively enlarge and the affected coronary artery and chamber may be dilated over time due to increased blood flow [18].

Clinical presentation

Although CAFs are uncommon and usually incidentally detected abnormalities, they may cause significant morbidity and mortality in any age group. The adult population is mostly asymptomatic unless the fistula causes significant hemodynamic shunt (pulmonary flow/systemic flow ≥2) or complications causing myocardial ischemia, heart failure, infective endocarditis or rupture of dilated fistulas. The symptoms and complications of medium- to large-sized CAFs tend to be seen in the adult age group [19]. They may present as angina pectoris, atypical chest pain, dyspnea, dizziness, syncope, fatigue, palpitations or even sudden cardiac death [8]. Patent ductus arteriosus, pulmonary arteriovenous fistula, ruptured sinus of Valsalva aneurysm, aortopulmonary window, internal mammary artery to pulmonary artery fistula, anomalous origin of left coronary artery or right coronary artery from the pulmonary artery and systemic arteriovenous fistula should be considered in differential diagnosis.

■ Diagnostic evaluation

In children, the course and drainage sites of anomalous coronary arteries can be demonstrated by transthoracic echocardiography owing to an optimal acoustic window. In adults, transesophageal echocardiography is more useful, particularly in proximal lesions [20]. Although cardiac catheterization and coronary angiography is the gold-standard method to obtain the best hemodynamic data, shunt calculation and visualization of coronary anatomy, emerging imaging modalities such as multidetector computed tomography and MRI can accurately delineate the anatomy of coronary vessels and the origin, course and drainage site of the fistula [21,22].

Indications for intervention

Several factors including patient age, size, morphology and anatomy of a fistula and associated cardiac disorders should be considered for the best management of CAFs [23–27]. Traditionally, asymptomatic patients with small shunts are managed conservatively owing to the benign course and the possibility of spontaneous closure. In symptomatic patients presenting with myocardial ischemia, β-blockers or calcium channel blockers may relieve the symptoms by decreasing myocardial oxygen demand [28]. Symptomatic patients with left-to-right shunt and complications including myocardial ischemia, congestive heart failure, pulmonary hypertension, dysrhytmias and infective endocarditis should undergo interventional procedure. Percutaneous closure can be performed in the case of CAF associated with infective endocarditis. There is still controversy regarding intervention for large shunts in asymptomatic patients because of the lack of data about the natural history of unintervened CAF. Despite the case reports of spontaneous closure or nonprogression, several studies recommend closure of large CAFs owing to the risk of future complications [29–32]. According to the current American College of Cardiology (ACC)/American Heart Association (AHA) guidelines for adults with congenital heart disease, a large CAF should be closed regardless of symptoms and small-to-moderate CAFs should be closed in the presence of documented myocardial ischemia, arrhythmia, otherwise unexplained ventricular systolic or diastolic dysfunction, enlargement or endarteritis [33]. Therefore, all of the major CAFs should be treated as soon as the diagnosis is made.

Principles of treatment

■ Surgery versus interventional closure

In a series examining surgical closure, good short- and long-term results were reported. In a surgical review of CAF in 17 patients consisting

Table 1. Experiences in percutaneous closure of coronary artery fistulas.

Author (year)	Age (median)	Patients treated	Devices used (n)	Follow-up	Morbidity (n)	Mortality	Residual fistula (%)	Ref.
Qureshi *et al.* (2001)	5 months–70 years	40	Coils (35) Detachable balloon (3) Combination of detachable balloon and coils (1)	1 month–10 years	Embolization of the occlusion device (6)	1	1 (3%)	[66]
Okubo *et al.* (2001)	8 months–14 years	13	Coils (10) Rashkind double-umbrella device (1) ADO (2)	1–31 months (mean: 14.6 months)	Migration of coils (4) Transient arrhythmias or changes in the resting ECG (4)	0	4 (30.7%)	[67]
Armsby *et al.* (2002)	12 years (5–71 weeks)	41	Coils (29) Balloons (8) Umbrellas (2) Covered stent (1) Coils and balloons (2)	1 day–4 years (mean: 1 year)	Coil embolization (4) Transient ST–T changes (4) Acute myocardial infarction (1) Transient dysrhythmia (1)	1 (due to embolization of coil into left coronary artery)	3 (7%)	[37]
Behera *et al.* (2006)	21 days–56 years (median age: 4.3 years)	6	ADO (6)	3–14 months (median: 8.5 months	Transient palpitations (1) Atrial arrhythmia (1)	0	1 (16%)	[26]
Liang *et al.* (2006)	2.1–12 years	4	Gianturco coils (4)	26–83 months (mean: 49 months)	–	0	0	[24]
Collins *et al.* (2007)	26–78 years	11	Coils (12) Drug-eluting stent (1)	Mean: 2.6 ± 2.1 years	Coronary dissection (1) Coil embolization (2)	0	0	[25]
Abdelmoneim *et al.* (2007)	42–78 years	6	Coils (6)	Mean: 2.5 years	Transient chest pain (2)	0	1 (17%)	[68]
Kabbani *et al.* (2008)		5	Microcoils (2) Hydrocoils (3)	3 months–7 years	–	0	0	[46]
Zhu *et al.* (2009)	5–56 years	20	ADO (10) AVP (4) Detachable coil (4) Cook coil (1) AMPLATZER® ventricular septal occluder (1)	3 months–10 years (mean: 4.7 years)	Transient ST–T changes (4)	0	1 (5%)	[60]
Bruckheimer *et al.* (2010)	0.5–52.2 years	9	ADO (6) AVP (3)	0.2–5.8 years (median: 1.7 years)	Transient ST–T changes (1)	0	0	[69]

ADO: AMPLATZER Duct Occluder®; AVP: AMPLATZER® Vascular Plug; ECG: Electrocardiography.

Figure 1. Devices used in percutaneous coronary artery fistula closure.

of infants and children, early surgical management of CAF was reported to be a safe and effective treatment resulting in a 100% survival and 100% closure rate [34]. In a study consisting of 41 patients with CAF, no operative mortality was reported after surgery and 96.9% of the patients were asymptomatic at a mean follow-up duration of 9.1 years [35]. In another surgical closure study by Wang *et al*. that included 52 patients, no residual shunt was reported before hospital discharge with good follow-up results for a mean period of 3.14 ± 1.84 years [36]. Although contemporary studies report no death after surgical closure, the surgical approach is not free of mortality and morbidity [19,23,36]. Accompanying congenital anomalies were more common in the surgical literature compared with percutaneous closure studies.

Despite the effectiveness of surgical closure, a similar success rate for transcatheter closure made the emerging percutaneous techniques an important alternative to surgery. In addition to the proven safety and efficacy, percutaneous techniques do not have many of the disadvantages of the surgical approach, which include sternotomy, cardiopulmonary bypass and longer hospital stay [37,38]. Although surgery seems to be associated with higher procedural success, it should be stressed that most of the available data were from single-center experiences in small patient groups. Furthermore, recurrence or residual shunts may also occur in approximately 10% of the patients who undergo surgery [35]. In a study including the report of short-term findings in 33 patients after the percutaneous closure of CAF by Armsby *et al*., complete occlusion was accomplished in 27 patients (82%) without procedural death or long-term morbidity in a median follow-up of 2.8 years [37]. These results showed similar early effectiveness, morbidity and mortality when compared with the surgical literature (Table 1).

■ Technical details & devices

Since the first percutaneous intervention by Reidy *et al*. [39], various devices have been introduced for the transcatheter closure of CAFs, including Gianturco coils, stainless-steel coils, detachable

Table 2. Comparison of devices used in percutaneous coronary artery fistula closure.

	Amplatzer® Vascular Plug	AMPLATZER Duct Occluder	Detachable balloon	Coils	Covered stent	Chemicals
Selected fistula size	Medium–large	Medium–large	Medium–large	Small–medium	Small–medium	Small–medium
Retraction before deployment	Yes	Yes	Yes	Yes/no	No	No
Clinical experience	High	High	Low	High	Moderate	Low
Delivery approach	Antegrade or retrograde	Antegrade	Retrograde	Antegrade or retrograde	Retrograde	Retrograde

balloons, platinum microcoils, double-umbrella devices, covered stents, AMPLATZER® Duct Occluder (ADO), AMPLATZER® Vascular Plug (AVP), polyvinyl alcohol foam and several chemicals including cyanoacrylate. The comparison of percutaneous methods used for the closure of CAF is illustrated in Figure 1 & Table 2. The most commonly used methods will be reviewed in the following sections.

Coil embolization

Currently, coil embolization is the most commonly preferred method for percutaneous closure of CAF either by retrograde or anterograde approaches (Figure 2). In case of multiple feeders, coil embolization should not be preferred because closure of CAF is difficult with a single intervention. Distally located fistulas, tortuous anatomy, an adjacent vessel at risk, the need for concomitant coronary bypass and larger fistulas do not favor coil embolization [34]. The potential complications are transient arrhythmias, coil embolization into the great vessels or recoil into the major coronary artery leading to acute myocardial infarction and, occasionally, sudden death. The coil needs to be up to 30% larger than the area of target vessel to avoid migration and embolization of the coil. After the implantation of the first coil in the correct position, different sizes of coils can be deployed until complete occlusion is achieved [40].

Coils are made of metallic wires with variable size, shape and rigidity, which are either pushable or detachable. Coils are mainly divided into two groups: standard steel coils (Gianturco coils) and platinum microcoils [41,42]. Generally, coils are preferred in the occlusion of small fistulas; however, pushable coils such as Gianturco coils have been used in large and aneurysmal fistulas [43]. Platinum microcoils are available with or without fibers embedded into the device. Electrolytically detachable coils have also been used to occlude CAFs and can easily be retrieved from the delivery catheter until a precise location is found. These coils have the advantage of electrothrombosis to help the closure of the fistula [44]. Interlocking detachable coils have the advantage of retraction and repositioning before delivery, as do the electrolytically detachable coils. Until the optimal position is achieved, these coils can be easily manipulated, in contrast to the traditional platinium or steel coils, which cannot be retrieved or retracted before deployment. Furthermore, these coils contain Dacron® fibers to promote thrombogenicity. Flexibility and delivery in small catheters (<3 Fr) made those coils an attractive option, especially in tortuous fistula anatomy [45]. Hydrocoils are made of hybrid material and have the potential to expand up to nine-times their volume after contact with blood owing to an outer layer of hydrophilic, acrylic polymer gel, and are used successfully to occlude large CAFs [46].

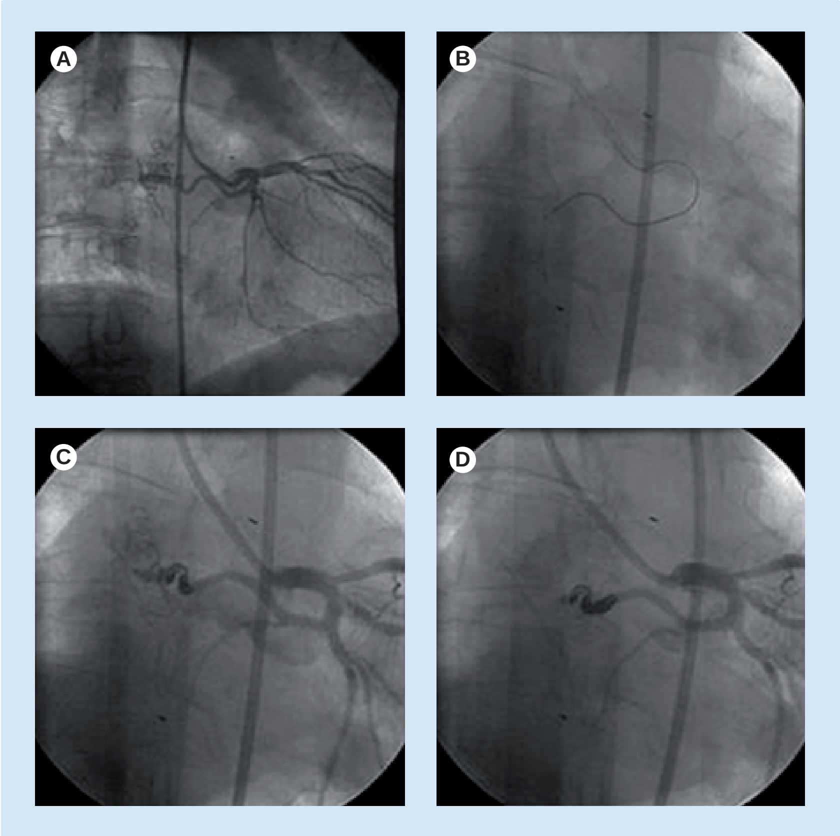

Figure 2. Coil embolization of coronary artery fistulas. (A) Coronary artery fistula from circumflex artery to pulmonary artery causing ischemia shown by myocardial perfusion scintigraphy. **(B)** Coronary artery fistula was crossed with a 0.014-inch wire. **(C)** Multiple detachable coils were deployed into the fistula. **(D)** Control angiography revealed no residual flow through the fistula.

Detachable balloons

Detachable balloons are no longer preferred in the closure of CAFs since the advent of newer methods. They require large introducer catheters and are inflated with contrast medium or silicone material (Figure 3). They are floated within the arterial vessel with the direction of blood flow and immediate occlusion is achieved when the balloon is inflated and detached. Detachable balloons are a reliable and safe technique for occluding large CAF, with several advantages over other methods, which are:

- The effects of occlusion on myocardium can be assessed before the final release of the balloon;

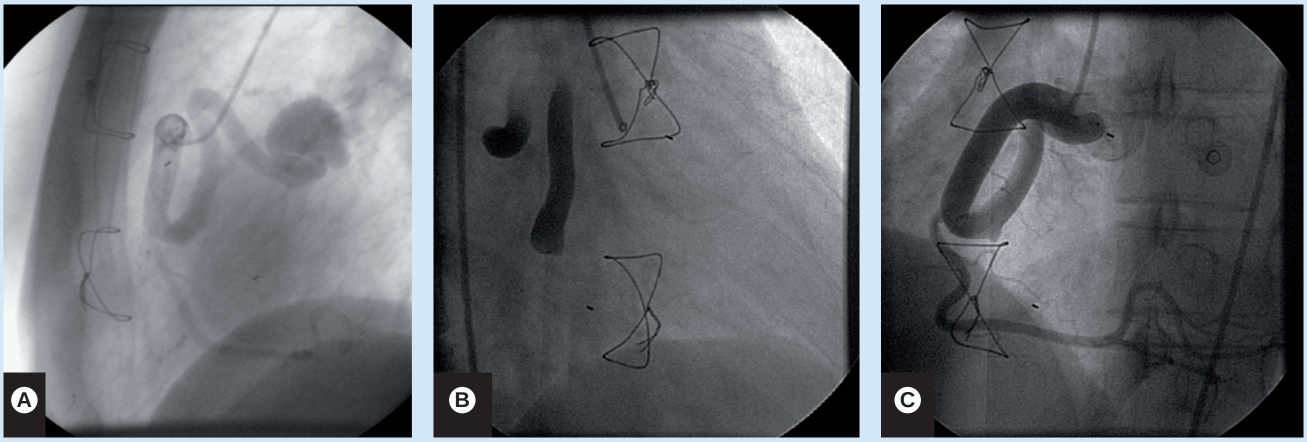

Figure 3. Closure of coronary artery fistulas with detachable balloon. (A) Large coronary artery fistula originating from the right coronary artery and draining into the right atrium. **(B)** The view of a detachable balloon deployed into the fistula from the right anterior oblique projection. **(C)** No residual flow was observed after deployment.

- Detachable balloons can be easily inflated and constructed to fit the size of the fistula;
- Balloons can be easily controlled;
- They can be directed to the lesion site easily from the coronary artery because of the same direction of blood flow.

Immediate and reversible occlusion can be achieved with these balloons during detachment. After the withdrawal of the microcatheter, the balloons are closed by a valve. Detachable balloon is a reliable and safe way to occlude medium- to large-sized CAFs. However, owing to infrequent use, available data are mainly limited to occasional case reports [47,48].

AMPLATZER® Vascular Plug

The AMPLATZER® Vascular Plug is a self-expandable, cylindrical device made of nitinol mesh wires and is used in a variety of lesions including CAFs. As with other devices, the diameter of the device should be sized at 30–50% larger than the diameter of the reference vessel at the intended point of occlusion [27]. Moreover, unlike other AMPLATZER occlusion devices, the the AVP has no occlusive material. The main advantage of AVP is the high rate of occlusion without the need for delivery of multiple occlusion devices, as complete occlusion can be achieved with a single device.

AMPLATZER® Duct Occluder

The ADO is a self-expandable device used in the closure of patent ductus arteriosus, made of nitinol wire mesh. Polyester material within the device causes thrombosis and closure of the fistula. The device size should be 30% greater than the vessel diameter. The ADO should be placed at a location in the proximal portion of the fistula at a narrow site, so the device can be anchored to the area of interest. The reported advantages of ADOs in CAFs include single device usage, a high ratio of complete occlusion, antegrade placement, ease of placement and improved control during deployment of the device.

Covered stents

Covered stents are made of a membrane integrated into a coronary stent. These stents are mostly used to seal degenerated vein grafts and cover coronary artery perforations, aneurysms and fistulas [49,50]. Mostly, stents are covered with polytetrafluoroethylene material. Recently, covered stents have been introduced as an important alternative technique for closure of CAFs. Covered stents preserve the main vessel patency while occluding flow through the fistula. Covered stents are mainly preferred in cases of an accompanying coronary atherosclerotic lesion near to the origin of the CAF and may also be useful in plexus-like CAFs, in which the implantation of a single device is insufficient for the closure of the CAF [51,52]. The main limitations of covered stents are side branch occlusion, mismatch between proximal and distal vessel diameter (as seen in many patients with large CAFs) and a high risk of in-stent restenosis and stent thrombosis [53,54].

Chemicals

Embolization of fistulas with chemical materials is an alternative method for the percutaneous closure of CAFs. Mainly particulate materials such as gel foam and polyvinyl alcohol foam or liquid materials such as *N*-butyl cyanoacrylate have been used. Gel foam is composed of gelatin sponge, and polyvinyl alcohol foam is composed of synthetic plastic foam. Thrombus formation around those particles is the mainstay of the embolization procedures. *N*-butyl cyanoacrylate is a type of glue and this agent polymerizes immediately after contact with blood, which causes vessel occlusion. Successful results of percutaneous embolization of CAF with cyanoacrylate [55], polyvinyl alcohol foam [56] and gel foam [43] have been reported. In addition to isolated use in occluded CAFs, cyanoacrylate may be combined with coils in the case of residual flow after coil embolization.

■ Tips & tricks

Percutaneous closure of CAFs is mainly preferred in cases of proximal origin of fistula, the absence of multiple feeders, with a lack of adjacent coronary artery branches and for drainage to low-pressure areas [37,42]. Multiple drainage sites, extreme vessel tortuosity, aneurysm formation, acute angulation, close location of a side branch to a drainage site and distal location of the lesion not allowing delivery of a catheter are the main limitations for the percutaneous approach [34,37]. The selection of occlusion devices mostly depends on the age of the patient, associated cardiac diseases and the anatomic characteristics of the fistula. The therapeutic approach does not differ according to the type of CAF (either congenital or acquired).

Selective catheterization of the fistula and imaging the entire coronary anatomy is the most important step in percutaneous intervention. In cases of high-flow fistulas, balloon occlusion angiography can be used to identify all coronary and fistula anatomy and drainage sites. Selective cannulation of fistulas with small diameter-guiding catheters (e.g., 4 or 5 Fr) is a widely used method for proper positioning [25]. In cases of tortuous anatomy, cannulation of the coronary ostium with a large diameter-guiding catheter (e.g., 8 Fr) and afterwards, either high-torque floppy wires or a delivery catheter passing through this large guiding catheter can be used for positioning at the closure site of the fistula. In these cases, the 3-Fr Tracker (Target Therapeutics, CA, USA) or Micro Ferret catheter (Cook Medical, IN, USA) are commonly used to reach the target area and, afterwards, detachable microcoils can be deployed with the guidance of a 0.014-inch coronary guidewire. To prevent device migration, positioning the catheter proximal to the narrowest area of the fistula is essential for procedural success. Temporary balloon occlusion may be needed for the appropriate positioning of coils in high-flow CAF. Another strategy is to create an arteriovenous loop with a wire by crossing through the entire fistula from arterial site to venous site with a snare, so as to carry the devices in a large CAF. Coils are mostly preferred in cases of small-to-moderate CAF.

Deployment of a coil that is too large for the vessel results in insufficient wrapping and the device remains elongated, therefore causing insufficient closure. Conversely, coils that are too small carry the risk of embolization. The size of the AVPs and ADOs should be 30% greater than the target vessel area, as with other devices, for appropriate deployment. These devices have the advantage of closure with a single device and can be used in large fistulas compared with coils. AMPLATZER devices are MRI compatible and may be selected in patients in whom medical status may require additional imaging. Currently, AVPs and ADOs are preferred in cases of large CAFs instead of detachable balloons. AVPs and ADOs also have the advantage of fistula occlusion with a single device. The AVP can be used in cases of multiple CAFs draining into a single area. The main advantage of covered stents is the exclusion of fistulous anatomy in device selection. Furthermore, covered stents can be used in patients with coexistent coronary atherosclerosis at the target occlusion area.

The main aim of catheter intervention in CAF is to occlude the fistula artery at a precise point, away from the native coronary artery. However, sometimes, if the embolization of CAF is too distal, it may cause migration of an occluding device or material into the relevant cardiac structures and pulmonary circulation. Closure device delivery should be distal enough within the fistula that the native coronary circulation is protected from device migration, clot propagation and side branch occlusion. Distal location of the fistula, presence of multiple feeders, tortuosity, side branch lesion, acute angulation, aneurysm formation and accompanying cardiac anomalies favor surgical intervention. However, percutaneous intervention can be performed in most of these cases with appropriate equipment. For example, in cases of high flow across the fistula, flow can be stopped with a balloon prior to intervention or superfloppy guidewires can be used in tortuous vessels. Despite early reports stating that a CAF with multiple drainage sites precluded percutaneous intervention, progress in experience and improvements in equipment and occlusion devices have enabled the use of transcatheter closure in CAFs with multiple drainage sites [37]. In cases where the fistula is located near to the origin of an adjacent vessel, percutaneous closure can be safely performed when the fistula is selectively and distally cannulated. The anatomical structure of fistulous vessel aneurysm, atherosclerosis and histologic abnormalities in the vessel wall may lead to complications [57]. Therefore, particularly in percutaneous interventions in large fistulas, gentle manipulation of catheters and devices is essential to prevent dissection or vessel rupture [37].

Multiple CAFs constitute 10% of all CAFs [58]. There are controversies about the optimal management of multiple CAFs, either percutaneous or surgical interventions. Mostly, multiple CAFs are referred for surgical closure; however, in some cases, percutaneous closure can be performed [27].

Mostly, retrograde approaches are used in percutaneous intervention to occlude CAFs. The main advantage of a retrograde approach is familiarity with this approach but, in some cases, an antegrade approach can be performed to occlude the fistula (e.g., short feeding vessel). The use of larger catheters, a straighter catheter course and avoiding femoral artery access are the potential advantages of the antegrade approach, despite the risk of device embolization due to lack of flow control.

A thorough analysis of past reports indicates that all symptomatic patients and asymptomatic patients with large CAFs should undergo closure of CAFs. Owing to the unusual nature of this abnormality, firm recommendations relevant to the optimal management are difficult to make. However, progress in technical and interventional tools extended the current concept of percutaneous intervention to CAFs. Furthermore, percutaneous interventions can be performed in cases with challenging coronary anatomy.

Complications, outcomes & follow-up

Early complications related to occlusion procedures are transient myocardial ischemia or dysrhythmias, myocardial infarction, distal coronary spasm, fistula dissection, device embolization and, very rarely, procedural death [37]. Residual flow is seen in approximately 10% of patients [59]. Zhu *et al.* reported that the trivial–mild residual incidence was 25% and recurrence was 5% among 24 patients after percutaneous closure in a mean follow-up of 4.7 ± 3.2 years [60]. Examining the use of a surgical approach, Rittenhouse *et al.* reported a recurrence incidence of 4% [61]. In fact, most of the recurrences were small and did not require additional intervention [62].

Coronary artery dilatation may persist after intervention, either with percutaneous closure or surgical ligation, in intermediate to long-term follow-up [63]. Rupture of CAFs may occur in patients, independent of preceding dilation. Therefore, close follow-up is warranted, especially for thin-walled ectatic arteries. Although there is not a consensus regarding the antithrombotic or anticoagulant therapy for dilated coronary arteries after closure, some authors advocate antiplatelet therapy for dilated coronary arteries and anticoagulation with warfarin in severe coronary artery dilatation (>10 mm), especially in patients with sluggish coronary flow [63].

A small CAF has the most favorable course. Sherwood *et al.* reported that a small asymptomatic CAF had a benign natural history and patients were free of complications at a mean follow-up of 9.3 years [64]. Cheung and coauthors concluded that the benefits of surgery are uncertain for asymptomatic patients with mild shunting and regular follow-up has been recommended; surgical correction should be planned when symptoms develop or shunt increases [35]. In cases of asymptomatic small fistulas, routine clinical evaluation combined with imaging including both ventricular dimensions and fistula morphology is reasonable because of the risk of increase in size with time.

The follow-up algorithm is limited for CAF after intervention. Long-term follow-up is essential owing to the possibility of postprocedural recanalization or residual flow, persistent dilation of the coronary artery, late thrombosis and myocardial ischemia. Therefore, even though most patients become asymptomatic after intervention, they should not be dismissed from follow-up. Clinical evaluation with chest x-ray, electrocardiogram and echocardiography should be used in the follow-up. When myocardial ischemia is suspected, myocardial perfusion scintigraphy should be performed. In our institution, follow-up visits were performed at 1, 3, 6 and 12 months during the first year and annually thereafter. In the majority of studies, color-Doppler study was integrated into echocardiographic analysis of both intervened and unintervened CAFs. The changes in affected chamber dimensions should be checked at each visit. Coronary angiography should be performed in symptomatic patients if the clinical condition is attributed to fistula recanalization or other complications. However, the necessity for coronary angiography in long-term follow-up in asymptomatic patients is not clear. Asymptomatic patients should be followed-up with noninvasive tests. Newer imaging modalities such as multidetector computed tomography and cardiac MRI may produce valuable data regarding the patency or residual flow of the fistula in those cases [65]. Although reported long-term outcomes after percutaneous closure are promising, routine follow-up with either invasive or noninvasive tests should be the standard of care in both intervened and unintervened cases.

Future perspective

Currently, percutaneous management of CAFs provides a high degree of procedural success with a very low risk of serious complications. Device selection and delivery technique should be based on the anatomic and morphologic characteristics of the fistula. Although surgical ligation has previously been the standard treatment for CAF, specialized techniques, equipment and newer devices have made the percutaneous approach a safe and effective first-line treatment modality in most patients with suitable anatomy, with good follow-up results. In our opinion, with the advent of new devices and equipment, outcomes of the percutaneous approach will be improved, and most of the CAFs are amenable to percutaneous closure. More studies evaluating the efficacy and safety of percutaneous approaches with long-term follow-up results will provide valuable data in the near future.

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Executive summary

Indications for intervention

- Symptomatic patients with left-to-right shunt and complications including myocardial ischemia, congestive heart failure, pulmonary hypertension, dysrhythmia and infective endocarditis should undergo percutaneous intervention.

Principles of treatment

- Percutaneous closure of a coronary artery fistula is recommended at the proximal origin of the fistula, in the absence of multiple feeders and if there is a lack of adjacent coronary arterial branches. The selection of occlusion device mostly depends on the cost of the device, the operator's experience, the age of the patient, associated cardiac diseases and the anatomic characteristics of the fistula.

Tips & tricks

- Positioning the catheters as proximal as possible to the narrowest area of the fistula is integral to procedural success to prevent device migration. The selected device should be slightly larger than the target vessel diameter to allow proper positioning of the device.

Complications, outcomes & follow-up

- Long-term follow-up is essential owing to the possibility of postprocedural recanalization or residual flow, persistent dilation of the coronary artery, late thrombosis and myocardial ischemia after percutaneous intervention.

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