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Review

Treatment Strategies for Coronary Artery Fistulas: A Contemporary Review of Anatomy, Diagnosis, and Management

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Abstract

Coronary artery fistulas (CAFs) are rare but clinically important congenital or acquired anomalies characterized by abnormal vascular connections between one or more coronary arteries and cardiac chambers or great vessels. Although these fistulas, which bypass the myocardial capillary bed, are often asymptomatic and detected incidentally, large or aneurysmal lesions can lead to serious complications such as myocardial ischemia, arrhythmias, heart failure, and infective endocarditis. The proliferation of advanced imaging technologies—especially electrocardiogram (ECG)-synchronized coronary CT angiography and cardiac magnetic resonance (MR)—has allowed for a more detailed assessment of anatomical features, drainage patterns, and hemodynamic effects of CAFs, enabling a comprehensive diagnostic approach that goes beyond conventional echocardiographic methodology. The presence of symptoms, shunt ratio, anatomical complexity, and the risk of complications are the primary factors guiding therapeutic decisions. Transcatheter closure has become the preferred option in suitable cases due to its minimally invasive nature, while surgical ligation is still indicated for complex fistulas. In addition, pharmacologic therapy—including beta-blockers, calcium channel blockers, and antiplatelet agents—may be employed as a temporizing measure in select low-risk or pediatric patients. Risk stratification models incorporating anatomical and hemodynamic parameters are increasingly used to support treatment decision-making, particularly in asymptomatic cases. Long-term and regular follow-up is critical due to the risks of residual flow, coronary dilatation, and thromboembolic events in the postprocedural period. This review provides a comprehensive overview of current diagnostic and therapeutic approaches to CAFs and discusses clinical decision criteria to support individualized treatment planning and long-term follow-up strategies in a structured manner. There is still a need for multicenter, prospective studies on the timing of treatment and the selection of treatment modalities, especially in pediatric and asymptomatic patient groups.

Keywords

coronary artery fistula; coronary vessel anomalies; hemodynamics; risk assessment; diagnostic imaging

Introduction

Coronary artery fistulas (CAFs) are rare vascular anomalies in which one or more coronary arteries form an abnormal connection with the cardiac chambers or great vessels, bypassing the myocardial capillary network [1]. CAF was first described by Krause in 1865 [1]. The first successful surgical correction was performed by Björk in 1947 [2]. Most cases are of congenital origin; however, acquired forms that develop after trauma, myocardial infarction, surgical procedures, or percutaneous interventions have also been described [1,2]. From an embryologic perspective, CAFs are believed to result from the persistence of myocardial sinusoidal channels that normally regress during development. In healthy cardiac morphogenesis, these sinusoids give rise to Thebesian veins; when regression fails, anomalous conduits may persist between coronary arteries and cardiac chambers or vascular structures [3,4]. The congenital form of CAF is frequently associated with other developmental cardiac anomalies, including atrial septal defects, ventricular septal defects, and tetralogy of Fallot, and reflect shared embryologic pathways [5]. The incidence of CAFs varies between 0.002% and 0.3%, depending on the population studied and the imaging modality used [6–8]. With the development of advanced imaging techniques such as multislice computed tomography (MSCT), coronary computed tomography angiography (CCTA), and cardiac magnetic resonance (MR), the rate of detection of undiagnosed cases has significantly increased [1,8,9]. While many CAFs are asymptomatic and detected incidentally, some cases may present with severe clinical manifestations such as dyspnea, angina, arrhythmias, myocardial ischemia, heart failure, and even endocarditis [1,10,11]. In pediatric populations, spontaneous closure has been reported in a minority of small, narrow-necked fistulas, particularly during infancy [12]. The pathophysiologic effects of the fistula vary according to the shunt volume,

anatomic origin, drainage point, and associated structural cardiac anomalies [13]. The most common artery of origin is the right coronary artery (RCA), and the most common drainage site is the pulmonary artery [14,15]. Even small fistulas that were previously considered benign may lead to complications such as volume overload, aneurysm formation, thrombosis, and rupture when left untreated. However, recent cohort studies and expert consensus indicate that such adverse outcomes are uncommon in small, asymptomatic CAFs, most of which follow a benign course under conservative monitoring [16,17]. Various classification systems have been proposed to assess the clinical significance of a fistula based on the vessel of origin, its communicating structure, the number of fistulas, and the hemodynamic effect [18]. These include angiographic classifications such as the Sakakibara and Liberthson types, which remain useful in guiding interventional strategies. These classifications guide both the diagnostic workup and the appropriate treatment, which largely depend on the anatomical complexity of the fistula, clinical symptoms, and the risk of complications. However, controversies remain regarding the timing and indications for intervention in asymptomatic or borderline cases [1,19]. Asymptomatic and small-sized CAFs are usually managed conservatively, while transcatheter coil embolization or surgical ligation is preferred for symptomatic or hemodynamically significant cases [17,20].

This review aims to provide a comprehensive overview of the anatomy, pathophysiology, clinical manifestations, imaging-based diagnosis, and current treatment strategies for CAFs. This includes a structured clinical approach incorporating anatomical, clinical, and hemodynamic predictors—reflected in a risk stratification model—to assist in therapeutic decision-making.

Anatomy and Classification

CAFs are rare vascular anomalies characterized by abnormal connections between a coronary artery and a heart cavity or great vessel. These fistulas bypass the myocardial capillary network, resulting in blood flow directly to low-pressure structures. The vast majority of cases are congenital; however, they can also occur following trauma, cardiac surgery, myocardial infarction, or procedures such as percutaneous coronary interventions and endomyocardial biopsy [1,2]. The incidence of these anomalies in patients undergoing invasive coronary angiography (ICA) is approximately 0.2–0.3% [15,21,22]. However, with the widespread use of advanced imaging techniques such as coronary computed tomography angiography (CCTA), these rates have increased, since CAFs can be detected in more detail [13,23]. In some reports, CCTA have prevalence rates approaching 0.9%, particularly in asymptomatic individuals undergoing screening [14,24]. The most common vessel from which CAFs originate is the RCA (approximately 50–60%), followed by the left anterior descending artery (LAD) (25–30%) and the left circumflex artery (LCX) (10–15%) [15,22,25]. It may also originate from the left main coronary artery, diagonal branches, or conus artery. The most common drainage site for a CAF is the pulmonary artery (30–63%), followed by the right atrium, right ventricle, coronary sinus, superior vena cava, and, more rarely, the left heart chambers (left atrium or left ventricle) [1,13,26]. Approximately 10% of fistulas have multiple drainage points or are plexiform, which can complicate its diagnosis and treatment planning [5,14,27,28].

Rare variants such as bilateral CAFs or fistulas draining into pulmonary veins or the coronary sinus require special diagnostic modalities due to their atypical hemodynamics and higher procedural complexity [29,30].

With the introduction of coronary computed tomography angiography, the morphologic classification of CAFs has significantly improved. Five main morphologic types have been described in the literature:

1. Linear (a simple vascular access);
2. Spiral or tortuous (tortuous structure);
3. Aneurysmal (enlargement of the vessel wall; risk of rupture or thrombosis);
4. Web-like (plexiform, thin and intertwined vasculature);
5. Mixed types [1,13,31,32].

These morphologic features not only define the anatomy but also guide risk assessment and interventional planning, especially in the case of complex or aneurysmal fistulas. From an embryologic perspective, congenital CAFs are thought to result from the failure of embryonic sinusoidal connections between the developing coronary arteries and the cardiac chambers to regress [1,4,5,33]. Normally, these vascular channels disappear during fetal development, but sometimes they become permanent and predispose to abnormal vascular connections. These channels typically regress to form Thebesian veins. When they persist, fistulous communications—especially into right heart chambers or the pulmonary artery—may occur [3,34]. In contrast, acquired CAFs may lack the tortuous configuration of congenital types and often display a more linear morphology. These fistulas typically arise after myocardial trauma, cardiac surgery, or catheter-based interventions [3,5,35,36]. Classification systems in the literature are generally based on anatomical, hemodynamic, and morphologic factors. Anatomically, fistulas are divided into two main groups: coronary–cameral (draining into cardiac cavities) and coronary–vascular (opening into large vessels such as the pulmonary artery or coronary sinus) [1,5,37]. In addition, the Sakakibara classification—based on the origin of the fistula relative to the coronary artery (proximal vs. distal)—and the Liberthson system—combining clinical and angiographic criteria—are widely used in research and clinical practice [1,18,33,38]. Hemodynami-

cally, CAFs may be small and asymptomatic fistulas, or large and clinically significant fistulas with volume overload and the risk of ischemia or heart failure [13,17,39,40]. Age-related progression is also relevant. Pediatric cases often remain asymptomatic with small-caliber tracts, while adult patients may exhibit progressive dilation, tortuosity, or aneurysmal changes due to chronic left-to-right shunting [13,16,20,41]. Currently, one of the most useful classifications is based on imaging. Using ECG-synchronized CCTA, the origin, course, and end site of the fistula can be identified with high accuracy, which greatly contributes to the diagnosis and treatment of CAFs [1,8,38,42].

A detailed evaluation of the originating vessel, drainage point, and morphologic structure of CAFs is critical for accurate diagnosis, risk analysis, and treatment planning. More frequent use of advanced imaging modalities enables earlier and clearer identification of these anomalies, thus paving the way for individualized treatment strategies based on anatomical and clinical findings.

Pathophysiology

The physiopathologic effects of CAFs are largely dependent on the size and anatomical location of the fistula and the direction and volume of the shunt. Patient-specific variables such as age, systemic vascular resistance, and comorbidities (e.g., pulmonary hypertension or diabetes mellitus) may significantly influence shunt physiology and the development of symptoms [16,43,44]. These anomalies create a direct connection between a high-pressure coronary artery and a low-pressure cardiac cavity or great vessel. This creates a continuous left-right shunt, allowing blood to pass directly to the draining structures without passing through the myocardial capillary network. The two most important clinical consequences of this abnormal flow are increased volume overload and myocardial ischemia due to the coronary steal phenomenon [1,16,24,45]. Volume overload occurs when a significant proportion of the blood leaves the coronary circulation and is diverted to low-pressure structures (e.g., the right atrium, right ventricle, or pulmonary artery). This causes dilatation and functional strain on the relevant chamber, depending on the drainage site of the fistula. Over time, this overload can lead to cardiac disorders such as right heart failure, pulmonary hypertension, and ventricular dysfunction, especially in the presence of large or multiple fistulas [13,46–49]. A Qp/Qs ratio greater than 1.5 is generally considered hemodynamically significant and may warrant closure even in the absence of overt symptoms [50]. Chronic turbulent flow through the fistulous tract can generate shear stress on the vessel wall, contributing to aneurysmal dilation, which in turn increases the risks of thrombosis, rupture, and embolic complications [10,44,50]. Such abnormal hemodynamics may also disrupt endothelial integrity by facilitating bacterial adhesion and increasing susceptibility to infective endocarditis [51]. Factors such as changes in vascular resistance with age and anatomical remodeling may cause even small fistulas to become more clinically significant [45,50]. The coronary steal phenomenon is one of the main mechanisms responsible for the development of myocardial ischemia in CAFs. In this condition, oxygenated blood is diverted to the unloading structure via the low-resistance fistula pathway, so that the myocardial territory responsible for the coronary artery from which the fistula originates does not receive adequate perfusion. This is particularly pronounced during increased demand (such as exercise). The steal phenomenon is more common in fistulas originating from the left coronary system or in the presence of concomitant aneurysmal dilatation [52–54]. This has been demonstrated by fractional flow reserve (FFR) measurements and perfusion imaging methodology, which have documented reversible ischemia in areas distal to the fistula [55]. Other pathologic effects of CAFs include aneurysmal dilatation of coronary segments due to increased flow. These dilatations carry significant risks for thrombosis, rupture, and embolic events. In addition, the presence of continuous turbulent flow at the fistula site may facilitate the development of infective endocarditis and predispose to arrhythmias due to chamber enlargement or myocardial scar formation [1,5]. Progressive myocardial remodeling in response to chronic volume and pressure overload may further increase the risk of arrhythmias, especially in patients with longstanding or untreated fistulas [5,22,29]. Although many small-sized CAFs remain asymptomatic and are often detected incidentally, the natural course of these lesions is unpredictable. Spontaneous closure is rare—occurring in less than 1% of cases—and is generally limited to low-flow, narrow-necked fistulas in infants or children [16,56]. Therefore, close monitoring or preventive closure strategies are recommended, especially in cases that are at risk for complications [5]. In neonates and infants, large fistulas may present early in life with symptoms such as heart failure, rapid breathing, feeding difficulties, and failure to thrive. In contrast, in adults, symptoms such as exercise-induced dyspnea, chest pain, palpitations, and even myocardial infarction may occur, especially in untreated lesions or lesions that grow over time [13,45]. The decision to intervene should be based not only on the presence of symptoms but also on the hemodynamic burden, signs of ischemia, and anatomical risk factors such as aneurysmal enlargement or complex morphology [1,13,17]. A comprehensive risk stratification framework—integrating shunt volume (Qp/Qs), fistula morphology, drainage location, and patient-specific modifiers—is essential to guide clinical decision-making and tailor treatment plans [1,10,57].

Table 1. Age-stratified clinical and anatomical classification of CAFs based on hemodynamic significance, morphological pattern, and drainage site.

Age group and clinical profile	Predominant clinical features	Estimated Qp/Qs	Anatomical morphology	Drainage site	Associated risk modifiers	Recommended management
Neonate – 1 year (Infant)	Tachypnea, poor feeding, hepatomegaly, failure to thrive	>1.5	Plexiform or saccular, proximal origin (RCA/LAD)	Pulmonary artery	Congestive signs, hepatomegaly, failure to thrive	Urgent closure (surgical or catheter-based)
1–10 years (Child)	Murmur, often asymptomatic, exertional dyspnea	<1.5	Linear, small-caliber tract from RCA/conus	Right atrium/right ventricle	None or incidental murmur	Echocardiographic follow-up, Qp/Qs monitoring
11–18 years (Adolescent)	Exertional dyspnea, palpitations, chest discomfort	1.2–1.5	Mild tortuosity, LAD or diagonal branches	Coronary sinus/right heart chambers	New-onset arrhythmias, chamber enlargement	Selective intervention if symptomatic or arrhythmic
19–60 years (Adult)	Angina, palpitations, exertional dyspnea, silent ischemia	>1.5	Aneurysmal or tortuous, LAD/LCX origin, acquired	Pulmonary artery/coronary sinus/RA	Ischemia, coronary steal, hypertension	Invasive closure guided by FFR/CCTA/CMR
>60 years (Geriatric)	Fatigue, dyspnea, atrial fibrillation, silent ischemia	1.0–1.5	Progressive saccular change, fragile walls, bilateral	Dual drainage (RA + coronary sinus)	DM, HTN, frailty, valvular disease	Multidisciplinary geriatric-tailored decision
Asymptomatic + Aneurysm >10 mm	Clinically silent but anatomically high-risk	>1.5	Saccular aneurysm, proximal RCA or LMCA origin	Pulmonary artery or coronary sinus	Rupture risk, thrombosis, endocarditis	Prophylactic closure per guidelines

Note: Qp/Qs, Pulmonary-to-systemic flow ratio; DM, Diabetes mellitus; HT, Hypertension; FFR, Fractional Flow Reserve; CCTA, Coronary Computed Tomography Angiography; CMR, Cardiac Magnetic Resonance; RCA, Right Coronary Artery; LAD, Left Anterior Descending artery; LCX, Left Circumflex artery; LMCA, Left Main Coronary Artery; RA, Right Atrium. Risk factors and treatment decisions should always be individualized based on patient-specific clinical, anatomical, and hemodynamic characteristics.

Clinical Manifestations

The clinical manifestations of CAFs vary widely and depend primarily on the size and morphology of the fistula, the pressure gradient between the feeding artery and the drainage site, and the presence of associated structural cardiac anomalies. Small and hemodynamically insignificant fistulas may remain asymptomatic for years and are often detected incidentally during ECG or ICA performed for unrelated indications [5,10,13,17,43]. They may also be discovered during evaluation for unexplained murmurs, subtle ECG changes, or nonspecific symptoms such as fatigue or palpitations [58]. A recent multicenter study reported that over 60% of asymptomatic CAFs were diagnosed incidentally during assessment for non-cardiac complaints [11].

Spontaneous closure, although rare (1–2%), has been observed in infants with narrow-necked, low-flow fistulas and is believed to result from thrombosis or endothelial proliferation [16]. When symptoms develop, they typically evolve gradually, reflecting progressive volume overload, rising Qp/Qs ratios, and age-related decline in vascular compliance [1]. Symptom burden increases over time, especially in untreated or aneurysmal lesions [13]. In neonates and infants, large CAFs may cause signs of congestive heart failure, including tachypnea, feeding difficulties, hepatomegaly, and failure to thrive. Although uncommon, these cases warrant prompt recognition and urgent intervention [19]. In older children and adults, the predominant symptoms include exertional dyspnea, chest discomfort, fatigue, and palpitations. In pediatric patients, dyspnea is linked to right-sided drainage, while palpitations are more common in adolescents and adults with left-sided or multichamber fistulas [5]. A continuous, machinery-like murmur, particularly along the left sternal border, is a hallmark finding across all age groups and represents uninterrupted shunting from the high-pressure coronary artery to a low-pressure receiving structure [15]. Auscultation remains an important diagnostic modality, especially in pediatric and geriatric populations, and often initiates further evaluation [59].

The coronary steal phenomenon is central to the ischemic symptoms of CAFs. In this mechanism, blood is diverted from the coronary arterial circulation to a low-resistance drainage site, thereby reducing perfusion to the myocardium, particularly during exertion. This can lead to exertional angina, ECG changes, and abnormal stress perfusion imaging even in the absence of atherosclerotic coronary artery disease [52]. Stress modalities such as myocardial perfusion Single-photon emission computed tomography, stress echocardiography, and cardiac MRI are highly sensitive for detecting perfusion deficits in CAF patients, including those who are asymptomatic [60]. Arrhythmias are frequently encountered in adults and may include supraventricular tachycardias, atrial fibrillation, and premature ventricular contractions. These are often secondary to volume overload and structural remodeling of the atrial or ventricular chambers [5]. Nearly one-third of adult CAF patients develop clinically significant arrhythmias, which may precede or accompany structural complications [5,22]. Large fistulas draining into right-sided cardiac structures or the pulmonary artery can increase right ventricular preload and pulmonary circulation pressures, potentially leading to right or biventricular failure over time [29,53].

Severe complications, though infrequent, include infective endocarditis, coronary aneurysm rupture, thromboembolism, and sudden cardiac death. These are more likely in untreated, massive, or aneurysmal fistulas [17]. In certain cases, these complications may be the initial presentation of a previously silent lesion. Therefore, early recognition and comprehensive risk stratification are essential [1,13]. CAF-related endocarditis, particularly when associated with valvular disease or delayed diagnosis, carries a high mortality [35]. Risk scoring systems have been proposed that incorporate age, Qp/Qs ratio (>1.5), aneurysm size, fistula origin, and drainage site to guide treatment decisions and predict prognosis [26,38,49].

An age-stratified clinical and anatomical classification integrating hemodynamic burden, morphologic pattern, and drainage characteristics has recently been proposed to guide individualized management decisions, as shown in Table 1 (Ref. [13,31,34,37,39,42,44,46,60]).

The severity of symptoms does not always correlate with the size of the fistula. Some large CAFs may remain asymptomatic, whereas smaller yet tortuous and high-flow fistulas may result in significant ischemia or arrhythmias. Morphologic predictors of adverse outcomes—including proximal origin, aneurysmal dilation, and extreme tortuosity—have been proposed as criteria for prophylactic closure, even in asymptomatic individuals [52]. Multimodal imaging with transthoracic and transesophageal echocardiography (TTE), coronary CT angiography, cardiac MRI, and ICA remains crucial for diagnosing CAFs and quantifying their hemodynamic significance [27,28]. In older adults, CAFs may present with vague symptoms such as exertional fatigue or dyspnea, which are frequently attributed to aging or comorbidities. A persistent murmur or unexpected left-to-right shunt on imaging should prompt suspicion for a CAF [22,23]. In this age group, comorbid conditions including hypertension, diabetes mellitus, and valvular disease may obscure the clinical picture, emphasizing the need for a tailored evaluation [10]. Current expert recommendations endorse a combination of geriatric assessment tools and individualized imaging strategies for optimal care in elderly patients [32]. The possibility of coronary steal should also be considered in the differential diagnosis of chest pain in patients without known coronary artery disease [19]. Even in asymptomatic cases, intervention may be warranted if the fistula is large,

Table 2. Comparative overview of diagnostic imaging modalities for CAFs adapted from recent studies [25–30].

Imaging modality	Advantages	Limitations	Clinical role	Contraindications	Relative Cost	Sensitivity/Specificity
TTE	Non-invasive, widely available, hemodynamic estimation	Limited acoustic window, especially in adults	First-line screening for large fistulas	None significant	Low	~90%/~85%
TEE	Higher spatial resolution, better posterior visualization	Semi-invasive, requires sedation	When TTE is inconclusive; detailed surgical planning	Esophageal disease, patient intolerance	Moderate	~95%/~90%
CCTA	High-res 3D anatomy, fast scan, excellent for complex CAFs	Radiation, contrast-induced nephropathy	Anatomical mapping, aneurysm detection	Renal insufficiency, iodine allergy	Moderate–High	~97%/~90%
Cardiac MRI	No radiation, flow quantification, viability analysis	Limited access, longer acquisition time	Follow-up imaging, shunt quantification	Metal implants, severe renal dysfunction	High	~88%/~89%
ICA (Coronary Angiography)	Real-time flow assessment, therapeutic procedures	Invasive, may miss small or tortuous fistulas	Gold standard for procedural planning	Contrast allergy, high bleeding risk	Very High	~95%/~98%
IVUS	High-resolution wall and lumen visualization	Catheter-based, limited field of view	Adjunct to ICA when anatomy unclear	Same as ICA	Very High	Not precisely quantified
Hybrid Imaging (PET/CT/MRI)	Combines anatomy and perfusion; ischemic evaluation	Expensive, not widely available	Complex or symptomatic cases with uncertain flow	Same as MRI/CT depending on modality	Very High	Variable (protocol dependent)

Note: TTE, transthoracic echocardiography; TEE, transesophageal echocardiography; CCTA, coronary computed tomography angiography; MRI, magnetic resonance imaging; ICA, invasive coronary angiography; IVUS, intravascular ultrasound; PET, positron emission tomography; CT, computed tomography; CAF, coronary artery fistula.

drains into right-sided structures, or shows aneurysmal transformation—especially in young or physically active individuals [50].

In conclusion, CAFs encompass a broad clinical spectrum, ranging from incidental findings to life-threatening emergencies. Therefore, timely diagnosis, individualized imaging evaluation, and comprehensive risk stratification are essential for effective management.

Diagnostic Imaging and Evaluation

Overview of Imaging Strategies

Several different and often complementary imaging modalities can be used to identify and anatomically delineate CAFs. CAFs, as well as the vessels from which they originate, vary in size, and the sites of opening and flow dynamics differ. Hence, the choice of an imaging technique is critical to ensuring accurate diagnosis and effective treatment planning. The summary of the principal imaging modalities used in the diagnosis of CAF is presented in Table 2 (Ref. [25–30]) and offers a framework for clinicians to systematically compare the advantages, limitations, clinical roles, contraindications, relative costs, and sensitivity/specificity of the available modalities and to help choose the most appropriate modality for each patient.

Transthoracic Echocardiography (TTE)

Since it is both non-invasive and widely available, TTE is among the most frequently chosen modalities, in particular, for pediatric and asymptomatic patients. It can reveal dilated coronary arteries or abnormal flow patterns indicating the presence of a fistula. Color Doppler-guided TTE can recognise large fistulas and estimate their haemodynamic significance. Characteristic Doppler findings are observed when continuous, turbulent flow is directed into the great vessels of cardiac chambers [24,61]. However, due to the acoustic window limitations on TTE, this modality cannot always assess the full extent of fistulas in adults or those with complex anatomies [62,63].

Transesophageal Echocardiography (TEE)

Given its higher spatial resolution, TEE is particularly appropriate to cases where fistulas are associated with posterior cardiac structures or cannot be visualised on transthoracic imaging. Example are cases when the information generated by TTE is inadequate or cannot supply the sufficient anatomical mapping detail required before surgery [64]. This modality requires sedation and is of limited use in patients with esophageal disease or those who cannot tolerate the insertion of a probe [65].

Multislice Computed Tomography Angiography (CT Angiography)

A relatively recent and non-invasive modality, CT angiography generates high-resolution three-dimensional (3D) visualisation of the coronary arteries as well as the origin and end sites of the fistula tract. Used in tandem with an ECG, it can detect aneurysmal changes and complex branching patterns [42]. An imaging review of the use of CT angiography in pediatric patients found it outperformed ECG in estimating the full extent and morphology of CAFs [8]. Furthermore, the images generated by dynamic four-dimensional (4D) CT gives a more detailed picture of flow characteristics and vascular wall dynamics than other modalities, especially in pediatric patients and for multi-orifice CAFs. Its principal limitations are radiation exposure and contrast-induced nephropathy. When CAFs are hemodynamically significant (Qp/Qs ratio >1.5 or aneurysms >10 mm), CCTA can provide a better initial anatomical assessment than TTE, since it has higher spatial resolution and can delineate complex anatomy. When ischemia is suspected, cardiac MRI is advised to evaluate myocardial function. When interventional closure is planned, ICA is advised [42].

Cardiac Magnetic Resonance Imaging (MRI)

Cardiac MRI can complement other modalities to assess the functional and hemodynamic effects of CAFs by providing a quantitative analysis of shunt flow and assessing the volumes of the heart chambers and myocardial viability. These data are vital in cases of significant left-right shunting and to determine the impact of the coronary steal phenomenon [24,66]. Since ionizing radiation is not used, MRI is also recommended for patients requiring long-term follow-up. MRI enhances the measurement of Qp/Qs using phase-contrast flow imaging. In addition, myocardial fibrosis or ischaemic burden are identified through late gadolinium enhancement (LGE). However, it is contraindicated for patients with metal implants or severe renal dysfunction [67,68].

Invasive Coronary Angiography (ICA)

ICA is regarded as the ‘gold standard’ in CAF diagnosis, especially when percutaneous treatment is planned. ICA is the most suitable modality to determine the origin, extension, and drainage point of the fistula through the assessment of real-time blood flow. Furthermore, it allows catheter-based closure procedures to be performed in a single session [15,22,69]. A disadvantage is that, if the right angles are not used or contrast flow cannot be properly monitored, it may miss small or tortuous fistulae [13]. In such cases, it can be complemented by high-resolution intravascular ultrasonography (IVUS) to determine the origin of the fistula tract, wall structure, or aneurysmatic segments

[70]. Both modalities are invasive and can pose risks such as bleeding, vascular injury, and contrast-related complications. In borderline cases, clinicians can apply fractional flow reserve (FFR) to estimate the functional significance of the coronary steal to help determine whether interventions are appropriate [71].

Hybrid and Functional Imaging

Hybrid imaging modalities using both CT angiography and positron emission tomography (PET or MRI) have recently proven valuable in generating anatomical and functional information. In symptomatic patients, hybrid modalities can enhance diagnostic reliability because they allow anatomical findings to be combined with perfusion data [72,73]. The simultaneous structural and metabolic assessment enabled by hybrid PET/CT and PET/MRI is useful in evaluating inflammatory complications, such as endocarditis, and myocardial viability. Their disadvantages are high cost and restricted availability. Nevertheless, hybrid modalities are still recommended in complex or high-risk cases [37].

Treatment and Management

The treatment of CAFs depends on multiple factors, including the patient's age, clinical findings, size of the CAF, anatomical complexity, hemodynamic impact, and the risk of future complications. In many cases, CAFs are incidently detected and are asymptomatic. When CAFs are larger or symptomatic, potential complications include myocardial ischemia, arrhythmias, heart failure, aneurysms, and endocarditis, and therefore definitive treatment is indicated [17,40]. In a very few cases, CAFS close spontaneously, particularly in neonates or young children with narrow-necked fistulas, small shunt volumes, and low intraluminal pressure, potentially due to progressive endothelial proliferation or thrombotic occlusion [37]. However, these cases account for $<2\%$ of the total; the majority require structured monitoring or active closure. Treatment is recommended in several types of cases: where the CAF is symptomatic, with hemodynamically significant shunts (Qp/Qs >1.5); in the presence of ischemia due to the coronary steal phenomenon; and where there is progressive chamber dilatation or risk of complications (e.g., thrombosis, rupture). When CAFs are small and asymptomatic, a conservative approach and long-term follow-up may be preferable; however, as small and asymptomatic CAFs can grow or become symptomatic, early closure can be advisable to prevent future complications [37,40]. There is disagreement in the literature over the optimal timing for intervention. Some studies recommend early surgical or transcatheter closure in all anatomically suitable patients, whatever their symptoms, since the risks of myocardial ischaemia, bacterial endocarditis, and coronary aneurysm rupture can increase as patients age. Other studies recommend that the approach should be selected based on the patient's age, fistula size, and shunt volume. Jaffe *et al*. [74] advise long-term monitoring for small asymptomatic CAFs with low-flow shunts, whereas other studies recommend that simple lesions should be closed in children by the age of three. In contrast, it is generally agreed that closure is not suitable for patients with Eisenmenger physiology or severe pulmonary hypertension [38,44]. This controversy has led to the emergence of five distinct perspectives regarding the timing of treatment in patients with CAFs [36,40]. Some authors advocate early surgical or transcatheter closure in all diagnosed cases, even if asymptomatic, due to the progressive risk of myocardial ischemia, bacterial endocarditis, and the formation of coronary aneurysms with advancing age [17,37]. Others propose deferring closure until around age three for simple fistulas in asymptomatic children, reserving intervention for those with large shunt volumes, aneurysm formation, or associated cardiac defects [29,40]. Jaffe *et al*. [74] and subsequent studies suggest that small, low-flow CAFs may be monitored conservatively due to their potential for spontaneous closure over time [34–36]. A fourth view recommends withholding all intervention in stable, asymptomatic patients without additional cardiac anomalies [5,6]. Finally, recent systematic reviews increasingly support early closure—especially in pediatric patients—based on the age-associated rise in adverse outcomes, such as aneurysmal rupture or infective endocarditis [36]. These divergent perspectives emphasize the need for individualized decision-making that integrates anatomical features, hemodynamic relevance, patient age, and the risk for long-term complications.

The two principal approaches to treating CAF are transcatheter (percutaneous) closure and surgical ligation. The choice of the approach should be tailored to the anatomical features of the patient and the experience of the facility where the intervention is performed.

Medical Management

Medical management is usually selected when CAFS are small-calibre, asymptomatic or mildly symptomatic, and have hemodynamically insignificant shunt ratios (Qp/Qs <1.2) [41]. When medical therapy is combined with routine image monitoring, observational studies have found positive mid-term outcomes [1]. Various pharmacological treatments are available and are designed to control symptoms and reduce thrombotic risk. Such treatments include beta-blockers (e.g., metoprolol 25–50 mg/day) and calcium channel blockers (e.g., diltiazem 60–120 mg/day), which may be helpful in reducing myocardial oxygen consumption, thus mitigating ischemic symptoms [42]. In cases of mild aneurysmal dilation, low-dose aspirin (75–100 mg/day) has been shown to prevent thrombus forma-

tion [43]. However, pharmacological treatment does not cure the anatomical defect and is most appropriate for patients who require management while awaiting definitive closure or high-risk cases where invasive intervention is deemed unsuitable [44,56]. When imaging reveals large aneurysms, progressive dilation, or mural thrombus, options include anticoagulation or dual antiplatelet therapy (DAPT) [43,44]. Special consideration should be given to pediatric patients, particularly those whose anatomies are still evolving. Medical treatment is frequently used as an interim measure in this population. It is vital that doses are based on weight, and multidisciplinary assessment is required. As noted above, spontaneous closure has been seen in a minority of pediatric cases, especially those with fistulas with narrow necks [51].

Regular clinical follow-up with ECG or CCTA (e.g., every 6 to 12 months) is required to monitor anatomical progression and prevent complications in patients with CAF. The long-term effectiveness of medical therapy and its role in risk stratification and clinical decision-making remain unclear, and future prospective studies are necessary [36,51].

Transcatheter (Percutaneous) Closure

Where cases are anatomically suitable—in particular, single and wellcircumscribed fistulae with a narrow drainage area—transcatheter coil embolisation or device occlusion is usually selected because of its minimal invasiveness and short recovery time. Several devices can be used for this approach based on the size and location of the CAF, including platinum coils, Amplatzer vascular plugs, ductus occluders, and covered stents [69]. The first successful transcatheter occlusion of a coronary artery fistula was achieved by Jaffe *et al*. [74] in 1983 using a detachable balloon, marking a paradigm shift towards nonsurgical management. In the following decades, improvements in occlusion technologies—including microcoils, lowprofile Amplatzer Vascular Plugs II/IV, ductus arteriosus occluders, covered stents, and 3D imageguided navigation—have enabled closure of increasingly complex CAF anatomies [75–80]. Device selection should be tailored to the characteristics of the fistula such as diameter, tortuosity, neck morphology, and proximity to major coronary branches. Vascular plugs offer highflow shunt occlusion, coils remain suitable for small and tortuous tracts, and duct occluders excel in aneurysmal or larger fistulas [76,77]. Percutaneous closure is most effective in cases with single fistulas, minimal tortuosity, welldefined distal landing zones, absence of thrombus or infection, and sufficient distance from major coronary branches. Conversely, this approach is contraindicated in patients with severe pulmonary hypertension, large thrombotic burden, active infective endocarditis, or complex multitract anatomies that impede endovascular access—as detailed in the current consensus and recent devicebased guidelines [75,76,80]. Recent functional studies using quantitative flow ratio (QFR) and fractional flow reserve (FFR) have shown significant improvement in donorvessel perfusion postocclusion, particularly in mediumsized CAFs, reinforcing the clinical benefit of transcatheter closure in ischemic or hemodynamically significant fistulas [76–78].

In summary, interventional closure offers high procedural success (85–95%) with short recovery and lower morbidity, and should be strongly considered in anatomically appropriate, symptomatic or hemodynamically significant CAFs. A multidisciplinary approach integrating imaging, anatomy, and riskstratification is essential to obtain successful outcomes.

Surgical Closure

Many researchers and practitioners still regard surgical ligation as the ‘gold standard’, especially in cases of complex or tortuous anatomy and where percutaneous methods are not appropriate. Surgical closure is preferred for multiple fistula tracts, large aneurysms, fistulas that progress into the myocardium, and where there are technical difficulties in gaining transcatheter access [31,45]. Surgical success is increased by the use of intraoperative angiography or interventions performed in hybrid operating rooms [45,46].

Clinical Relevance and Risk Stratification of CAFs

Historical Framework and Existing Classification Systems

Several classification and risk stratification frameworks have been developed to guide therapeutic decisionmaking in CAFs. The criteria proposed by Konno and Endo in 1973 [81] remain foundational, highlighting indications such as significant left-to-right shunting (Qp/Qs >1.3), myocardial ischemia, heart failure, a history of endocarditis, and the presence of coronary aneurysms. Anatomical classification by Sakakibara differentiates lesions by origin and drainage patterns, noting that distally originating fistulas draining into right-sided chambers or the pulmonary artery typically produce higher shunt volumes and are associated with increased symptomatology [82]. Moreover, complex or multiple fistulous tracts are associated with elevated procedural risk and technical challenges [80]. Rigatelli and Rigatelli’s 2005 [83] clinical impact classification further refines this framework by stratifying anomalies into four hemodynamic risk categories—benign, relevant, severe, and critical—based on their physiologic burden rather than solely anatomical features. Contemporary guidelines from the American Heart Association (AHA) and the European Society of Cardiology (ESC) support intervention for all large or hemodynamically significant CAFs and for smaller

Table 3. Proposed risk scoring system for CAF severity.

Risk Factor	Criteria	Points
Shunt Volume (Qp/Qs ratio)	Qp/Qs <1.5:1 (small shunt)	0 points
	Qp/Qs 1.5–2.0:1 (moderate shunt)	1 point
	Qp/Qs >2.0:1 (large shunt)	2 points
Aneurysm of Fistula/Coronary	None or <10 mm (no significant aneurysm)	0 points
	10–30 mm (moderate aneurysm)	1 point
	>30 mm (giant aneurysm)	2 points
Fistula Complexity (number and morphology)	Single fistula tract, non-tortuous	0 points
	Multiple fistulous connections or very tortuous course	1 point
Symptoms (ischemia, heart failure, etc)	No symptoms (incidental finding)	0 points
	Symptomatic (angina, dyspnea, etc.)	1 point
Complications (endocarditis, HF, MI)	No major complication history	0 points
	History of endocarditis, fistula-related MI, or HF decompensation †	1 point
Anatomical Origin of Fistula	Proximal third of coronary (proximal origin)	0 points
	Distal two-thirds of coronary (distal origin)	1 point
Drainage Site	Drains to left heart (e.g., LV/LA)	0 points
	Drains to right heart or PA (low-pressure)	1 point

† If the patient has experienced a major complication—such as infective endocarditis, fistula-related myocardial infarction, or heart failure hospitalization—add 1 additional point to the total score, beyond the point assigned for symptoms alone.

Note: Qp/Qs, pulmonary-to-systemic flow ratio; HF, heart failure; MI, myocardial infarction; CHF, congestive heart failure; LV, left ventricle; LA, left atrium; PA, pulmonary artery; CAF, coronary artery fistula; TTE, transthoracic echocardiography; TEE, transesophageal echocardiography; CCTA, coronary computed tomography angiography; MRI, magnetic resonance imaging; ICA, invasive coronary angiography; IVUS, intravascular ultrasound; PET, positron emission tomography; CT, computed tomography.

lesions associated with myocardial ischemia, arrhythmias, ventricular dysfunction, or endarteritis [84,85]. A Qp/Qs threshold of ≥1.5 is commonly employed to define clinical significance [86].

In this context, the CAF-RS (Coronary Artery Fistula Risk Stratification) system offers a composite risk framework incorporating anatomical (fistula size, drainage site, aneurysm presence, structural complexity) and clinical (symptomatology, shunt severity) parameters. This tool is designed to enhance individualized risk assessment, facilitate evidence-based treatment planning, and reduce unnecessary interventions in contemporary clinical practice. These variables are integrated into a composite scoring tool as presented in Table 3 (Ref. [13,31,37,45,81–83,85–88]).

Rationale for a New Risk Scoring System

While existing classifications provide qualitative guidance, there is no standardized quantitative risk score for CAF severity. We propose a novel CAF Risk Score that incorporates key factors identified in the literature as predictors of adverse outcomes. The goal is to create a scoring matrix that stratifies patients into Low, Intermediate, or High risk categories, to assist in clinical decision-making and timing of intervention. The score is based on five domains, each grounded in published evidence or guideline criteria:

Shunt Size/Volume

Estimated by the shunt flow ratio (Qp/Qs). A larger left-to-right shunt correlates with volume overload and risk of heart failure. We use Qp/Qs thresholds of ~1.5:1 and 2:1, reflecting moderate and large shunts respectively. These cut-offs align with congenital heart disease guidelines (often using Qp/Qs ≥1.5 as significant) and surgical series (Konno's >30% shunt ≈ Qp/Qs >1.3) [81]. Higher Qp/Qs also tends to predict pulmonary hypertension if prolonged [63].

Aneurysm Size of the Fistula or Feeding Artery

Many CAFs cause aneurysmal dilation of the coronary artery due to chronic high flow. Aneurysm size is a surrogate for both shunt chronicity and wall stress. Literature indicates that aneurysms ≥30 mm (3 cm) carry high risk of rupture and are generally regarded as "giant" coronary aneurysms [31]. Even smaller aneurysms are concerning: cases of rupture have been reported with aneurysms ~10 mm [32], and any aneurysm formation was deemed an indication for surgery in Konno's criteria [81]. Therefore, this score grades aneurysm dimension as <10 mm (small), 10–30 mm (moderate), or >30 mm (large)—reflecting escalating risk of rupture or thrombosis in the fistulous segment.

Fistula Morphology and Complexity

A single, short fistulous tract is usually lower risk, whereas multiple fistula connections or a tortuous, plexi-

form course indicates a complex lesion that tends to shunt more blood and is harder to close percutaneously [13]. Complex fistulas often lead to diffuse arterial dilation and multiple exit channels (sometimes termed "artery of serpentine" on angiography). They have higher rates of complications like aneurysm formation and incomplete closure [37]. Complexity is thus incorporated as a binary factor (single vs. multiple/tortuous).

Clinical Presentation—Symptoms and Complications

The presence of symptoms attributable to the fistula (e.g., exercise angina, dyspnea, fatigue, arrhythmias, or signs of heart failure) is a pivotal indicator of clinical significance [44]. Any symptom suggests the fistula is causing physiologic burden (often via myocardial ischemia from steal or volume overload). In addition, certain serious complications can occur even in asymptomatic patients, notably infective endocarditis (due to turbulent flow), or rarely, fistula thrombosis with myocardial infarction [45]. A history of endocarditis is an unequivocal high-risk marker (warranting closure per Konno's criteria) [88], and heart failure or ventricular dysfunction attributable to the shunt also indicates advanced physiological impact [51,60]. This domain therefore awards points for being symptomatic and extra points for any major complication (endocarditis, fistula-related MI, heart failure hospitalization, etc.) [45,51,60].

Anatomic Location—Origin and Drainage

As noted, fistula origin and drainage significantly influence hemodynamic impact. A fistula originating from the proximal third of a coronary artery may divert flow early, but often the distal artery still has normal caliber (implying less total runoff) [36]. In contrast, a distal origin often means the entire artery has enlarged to channel blood, suggesting a larger runoff and more severe steal [31]. We assign higher risk to distal-origin fistulae [13,31,37]. Likewise, the drainage site is critical: low-pressure drainage (into the right atrium, RV, pulmonary artery or venous system) causes continuous runoff and volume load, whereas a fistula draining into the left ventricle (high-pressure system) typically has minimal net shunt [46]. Therefore, fistulas draining to the right heart/pulmonary circuit are scored higher risk than those draining to the left heart (Notably, the vast majority of congenital CAFs drain to the right side [13], so this factor mainly flags the uncommon left-side drainage as lower risk).

Each of these five domains is assigned a point value in our proposed scoring table. The scoring is weighted to reflect the relative importance: for example, a large Qp/Qs or a giant aneurysm might contribute more points than a single symptom. The total cumulative score then places the patient into a risk category that correlates with recommended management, as detailed after the table.

Scoring interpretation: Patients are stratified by total score into risk categories: Low Risk = 0–2 points, Intermediate Risk = 3–4 points, and High Risk = ≥5 points (maximum possible score is 8). These categories reflect an increasing likelihood of current or future adverse events and can guide management as follows.

Risk Categories and Management Implications

Low Risk (0–2 Points)

Typically small, asymptomatic fistulas with no aneurysm and trivial shunt. These patients can usually be managed conservatively with periodic monitoring. For example, a patient with an incidentally found small CAF (Qp/Qs 1.2, no aneurysm) would score 0 and be classified as Low Risk. Such a fistula has a low probability of causing harm in the near term [81]. Management would involve endocarditis prophylaxis as recommended [42], and serial follow-up (e.g., echocardiography every 3–5 years to monitor for enlargement or chamber dilation) [39]. No intervention is necessary unless the score worsens over time.

Intermediate Risk (3–4 Points)

These fistulas have some concerning features (moderate shunt or aneurysm, or mild symptoms, etc.) but not all high-risk criteria. For instance, a moderately large fistula (Qp/Qs ~1.7) with a 12 mm aneurysm, causing occasional chest discomfort, would score 1 (shunt) + 1 (aneurysm) + 0 (complexity) + 1 (symptoms) + 0 (no complication) = 3 points (Intermediate). Management in this category is individualized: many will warrant closure, especially if the patient is symptomatic or if any risk factor is progressive [17]. If asymptomatic, a case-by-case approach is taken, weighing patient age and surgical risk. In adults with intermediate features, guidelines tend to favor closure if there is any suggestion of myocardial ischemia or if the shunt is hemodynamically significant (Qp/Qs ≥1.5) [38]. In children, an intermediate-risk fistula often meets criteria for elective closure to prevent long-term damage; for example, closure is advised if Qp/Qs ≥1.5 or if right ventricular enlargement is present even in the absence of symptoms [65]. Thus, most intermediate-risk patients, especially younger ones, will be referred for closure, but the timing and modality (catheter vs. surgery) will depend on anatomy and patient-specific factors.

High Risk (≥5 Points)

Patients in this range have large shunts and/or advanced complications (e.g., Qp/Qs ≥2.0, giant aneurysm, symptoms with ischemia, etc.). This category correlates with the classic indications for intervention regardless of age [58]. For example, a symptomatic patient with a large tortuous fistula (Qp/Qs 2.5, 5 cm coronary aneurysm,

angina) might score 2 (shunt) + 2 (aneurysm) + 1 (complexity) + 1 (symptoms) + 0 (no prior endocarditis) + 1 (distal origin) + 1 (right-side drainage) = 8 points. High-risk CAFs should be promptly closed given their significant hazard for heart failure, rupture, or ischemic events [25]. Indeed, current guideline advice is that all large fistulas be closed even if asymptomatic , and any with complications absolutely require closure [63,68]. The scoring simply helps to identify these cases. Typically, high-risk patients will undergo intervention soon after discovery and medical stabilization [68].

Application in Pediatric and Adult Populations

The proposed CAF Risk Score is intended to be applicable across age groups, but there are special considerations in pediatric patients versus adults.

Pediatric Considerations

Children with significant CAFs often have more cardiovascular reserve and may remain asymptomatic despite substantial shunts. However, prolonged shunting can cause irreversible changes (e.g., pulmonary vascular disease or ventricular dysfunction) if not corrected. Therefore, in pediatrics there is a tendency to intervene at a lower threshold. For instance, even an asymptomatic infant with a large shunt (Qp/Qs $\geq$1.5) and right heart dilation would be scored as Intermediate or High risk and typically referred for closure in infancy [45]. Additionally, spontaneous closure of CAF in childhood, while documented, is extremely rare—thus observation is usually only for truly small, low-risk fistulas. The scoring system's thresholds (particularly Qp/Qs and symptoms) should be interpreted in context of a child's potential for compensating; an Intermediate-risk score in a child may prompt earlier intervention than the same score in a middle-aged adult, in order to preempt complications. One study from a multicenter CAF registry noted that asymptomatic neonates/infants with hemodynamically significant fistulas (Qp/Qs $\geq$1.5 or cardiac enlargement) were routinely treated early, with excellent outcomes [37].

Adult Considerations

In adults, the decision to close a fistula also weighs co-morbidities and procedural risk. A small, asymptomatic CAF in an elderly patient (Low risk by score) might be managed conservatively if their surgical risk is high. On the other hand, most adults over 20 with a sizable fistula become symptomatic with time [34], so an Intermediate or High score in an adult is usually met with an intervention recommendation. Another age-related factor is the development of concomitant atherosclerotic coronary disease; older patients might experience ischemia from a combination of coronary stenoses plus the steal from a fistula [20]. Our scoring system indirectly accounts for this by focusing on symptoms and ischemia. An older patient with borderline anatomy might be considered for transcatheter closure if their anatomy is favorable, to avoid the higher risk of surgery in advanced age [28].

In summary, pediatric patients often undergo closure at slightly lower scores (preventatively), whereas adults, especially if asymptomatic, might be observed until a certain score threshold or symptom triggers the need for action. Nonetheless, a High-risk score in any age typically mandates treatment, while Low-risk can often be watched.

Guiding Treatment Strategy With the Score

Importantly, the CAF Risk Score is not only a trigger for whether to treat but can also inform how to treat (transcatheter vs. surgical approach).

Low-risk Patients (Score 0–2)

Low-risk patients (score 0–2) are generally not immediate candidates for invasive treatment. They should be managed with medical therapy as needed e.g., beta blockers for mild symptoms, prophylactic antibiotics to prevent endarteritis and surveillance. If intervention is contemplated (for example, an otherwise low-risk young patient who is undergoing another cardiac surgery), the score highlights which factors are present. Usually, though, observation is chosen [84].

Intermediate-Risk Patients (Score 3–4)

Here, careful anatomical review is necessary to decide between percutaneous closure and surgery. Key considerations from the score include:

- If the fistula is anatomically simple (single, proximal origin, non-tortuous) and the patient has no contraindications, a transcatheter closure is often preferred [76,77]. A proximal, narrow-necked fistula is ideal for device or coil embolization, which avoids sternotomy and cardiopulmonary bypass. Our score would likely be lower in the "complexity" sub-score for such cases, reinforcing that less invasive closure is feasible. In fact, per AHA guidelines, transcatheter treatment is recommended for suitable symptomatic patients who have a narrow, single CAF, especially if proximal and without other cardiac anomalies [85].
- If the fistula is complex, distal, large-caliber, or has multiple channels (thus scoring points in those domains), surgical closure becomes more appropriate [31,45]. A higher complexity score signals that a catheter approach might be challenging (risk of incomplete closure, device migration, or inability to cannulate all channels) [80]. For example, a tortuous multi-hole fistula (score +1 complexity) with a distal insertion (score +1) might best be ligated surgically to ensure all connections are closed. Surgery is also favored if there is a large or diffuse aneurysmal seg-

ment requiring resection or if concomitant cardiac conditions need correction [88].

• The presence of certain complications in the score (e.g., endocarditis) usually mandates surgery for definitive treatment (excision of infected tissue), whereas purely hemodynamic issues without complication might be resolved via catheter [51].

Thus, a patient in the Intermediate range will be evaluated by a multidisciplinary team. Those with lower complexity scores and proximal anatomy will be triaged to transcatheter closure, often using coils, vascular plugs, or occluder devices [78,79]. Those with higher complexity or unfavorable anatomy will go to surgery for direct ligation or patch closure, often performed under cardiopulmonary bypass if the fistulous connections are intramural [49]. Our scoring system's individual components (especially the Complexity, Origin, and Drainage factors) thus guide the operator in choosing the safest and most effective therapy.

High-risk Patients (Score ≥5)

These almost invariably require closure, and usually sooner rather than later. A total score ≥5 often implies a large shunt and/or symptoms, which are Class I indications for closure [85]. In terms of strategy, many high-risk fistulas will require surgery because they are large and complex. For instance, a fistula with a giant 4 cm aneurysm and multiple communications is high-risk and anatomically challenging—surgery allows resection of the aneurysm and oversewing of the origin, addressing all issues.

High-risk scores that come predominantly from shunt size and symptoms (e.g., a single large proximal fistula causing heart failure, but without multiple channels) might still be amenable to transcatheter closure if anatomy permits—but even then, these patients must be approached cautiously. Large fistulas can have high flow that risks embolization devices being swept into the pulmonary circulation [11], and often have accompanying proximal coronary dilation that might need surgical attention. The score highlights such issues (e.g., aneurysm size). Generally, a high total score correlates with the need for surgical expertise, either for primary surgery or as a backup if transcatheter occlusion fails. Notably, if a patient has a high score due in part to complication history (like endocarditis or rupture), surgery is almost always indicated to remove infected tissue or repair damage [51,88].

In all cases, the risk score is not a substitute for clinical judgment but a supplementary tool. It compiles evidence-based risk factors into a single framework to ensure none are overlooked. For example, a patient might be asymptomatic (0 points for symptoms) but have a huge aneurysm (+2), distal origin (+1), and moderate shunt (+1), summing to 4 points (Intermediate). Without the score, one might be misled by the lack of symptoms; the scoring highlights that anatomically this is a dangerous lesion (indeed, guidelines would advise closure for an aneurysm of that size to prevent rupture) [86].

Conversely, a very symptomatic patient with chest pain might alarm clinicians, but if the fistula is actually small (no aneurysm, trivial shunt), the score might be low (e.g., 1 or 2), prompting a careful search for other etiologies of symptoms before attributing everything to the fistula.

Limitations and Future Directions

This proposed scoring system is derived from retrospective literature data and expert opinion guidelines. It has not yet been prospectively validated. One challenge is quantifying some factors in practice—for instance, exact Qp/Qs calculation for a coronary fistula can be complex (it requires cardiac catheterization with the oxygen step-up method, which is not always performed if the decision to treat is made based on noninvasive imaging) [87]. In such cases, an estimate of shunt severity may be made by indirect measures such as chamber enlargement, Doppler flow volume, or ventricular dilation on echocardiography [26]. Similarly, aneurysm size should ideally be measured by angiography or CT; however, small differences (e.g., 28 mm vs. 32 mm) might technically change the score category, though in practice both measurements are concerning [44]. The thresholds chosen (10 mm and 30 mm) are somewhat arbitrary but are guided by case series and definitions of "giant" aneurysms in prior studies [42]. Refinement of these cut-offs may be possible as more data accumulate—for example, if a study demonstrates that rupture risk increases significantly beyond 20 mm, the scoring system could be adjusted accordingly [86].

Age is not explicitly a component of the numerical score, but as discussed, it significantly influences management decisions. In future iterations, one might incorporate an age-risk modifier—for instance, pediatric patients with a given score might be classified one level higher due to anticipated long-term morbidity [67]. For now, we prefer to keep age as a qualitative consideration separate from anatomical or clinical risk factors.

Finally, the score should ideally be validated in a prospective cohort of CAF patients to assess whether higher scores truly predict worse outcomes such as the need for intervention, development of complications, or adverse events [88]. An international CAF registry has already begun risk stratifying by some anatomical features (e.g., neonatal outcomes based on fistula size and complexity) [37]. As more outcomes data become available, the scoring system can be fine-tuned. We anticipate that such a structured tool will be useful in multi-disciplinary heart team discussions, ensuring a reproducible and evidence-based approach to what has historically been a case-by-case decision-making process.

Clinical Implications of the CAF-RS System

CAFs vary widely in presentation—from incidental and benign anomalies to urgent surgical pathologies. A literature-derived risk scoring system for CAF, as proposed here, offers a structured framework that integrates key clinical and anatomical variables—including shunt size, aneurysm presence, morphological complexity, symptomatic burden, and drainage characteristics—into a single composite metric. This approach synthesizes established classification models such as Konno's surgical criteria [81], Sakakibara's anatomical typology [82], and Rigatelli's clinical relevance stratification [83], in conjunction with current guideline recommendations from the American College of Cardiology/American Heart Association (ACC/AHA) and ESC [84,85]. The scoring table provides clear thresholds for when intervention should be considered and offers insight into whether a transcatheter or surgical approach may be more appropriate [84–86].

In clinical practice, such a scoring system may improve decision-making by stratifying patients into meaningful risk categories aligned with therapeutic pathways—in a manner analogous to existing models in valvular heart disease or congenital structural anomalies. Prospective validation of this system will be necessary to confirm its prognostic value and clinical utility. As outcome data accumulate, adjustments to weighting or thresholds (e.g., aneurysm diameter or Qp/Qs ratios) may further refine its performance. Ultimately, the goal is to ensure that patients with CAFs receive timely and appropriate intervention in high-risk scenarios, while unnecessary procedures are avoided in low-risk cases that can be safely monitored.

Conclusion

Although CAFs are rare among coronary anomalies, they are clinically significant, and care must be taken in making their diagnosis determining treatment strategies. Despite their generally asymptomatic nature (and, frequent incidental detection), large-diameter and aneurysmal fistulas, in particular, can cause serious complications including myocardial ischemia, arrhythmias, heart failure, and endocarditis. Consequently, it is vital to carefully assess and stratify risks. Several non-invasive imaging techniques have recently become available, including ECG-synchronised CCTA and cardiac MRI, which accurately delineate the fistula morphology, drainage patterns, and hemodynamic effects and can be used in conjunction with traditional ECG and ICA. Factors determining the need for intervention include clinical presentation, anatomical complexity, and long-term prognosis. Transcatheter closure has the advantages of being minimlly invasive, and result in effective mid-term results. It is therefore frequently selected where cases are anatomically suitable. However, in fistulas with complex or tortuous structures, surgical ligation remains the procedure of choice. Combined approaches may be used in selected patients. Long-term follow-up is vital in all cases, even for patients who have undergone closure, to monitor for residual flow, coronary dilatation, and thrombotic complications. Anatomical features and hemodynamics vary among CAFs; therefore, classification and planning must be based on anatomy and physiology. CAFS exist across a broad spectrum, from embryological origin to clinical manifestation, and thus, properly defined management strategies are required. Risk stratification models can be valuable in offering indications for intervention, particularly when cases are asymptomatic or borderline. Detailed imaging, multidisciplinary expertise, and approaches that are validated by up-to-date scientific data should be used to determine the best strategy for individual patient management. It is recommended that long-term multicentre studies be undertaken, since current databases are limited, in particular for pediatric patients. High-quality, prospective research, will also be valuable in better delineating the natural history of CAFs, optimising the time for closure, and for assessing long-term cardiovascular outcomes.

In conclusion, diagnostic and therapeutic management must be structured and based on anatomical considerations to mitigate potential complications and enhance long-term cardiovascular outcomes in patients with CAF.

Author Contributions

The author (MM) contributed to the conception, design, data acquisition, analysis, and interpretation of the work. The author was involved in drafting, revising, and critically reviewing the manuscript, and has approved the final version for submission and publication. The author agrees to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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Conflict of Interest

The author declares no conflict of interest.

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