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Coronary artery fistulas: a systematic review and pooled analysis of anatomy, risk stratification and contemporary management

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ABSTRACT

Objective Coronary artery fistulas (CAFs) are rare congenital or acquired anomalies characterised by abnormal coronary communications, with variable natural history and no consensus on optimal management.

Methods A comprehensive state-of-the-art review and pooled analysis of published literature was conducted, integrating clinical, anatomical, imaging, management and outcome data from 1966 patients with 1986 CAFs. Comparative descriptive analyses between complicated and uncomplicated cases were performed to identify recurring features associated with adverse clinical outcomes.

Results Specific anatomical and clinical patterns were more frequently observed in patients with complications, including fistulas originating from the right coronary artery or left main coronary artery, drainage into venous or left-sided structures, large-calibre fistulas and reduced ventricular function. Based on these recurring patterns, two pragmatic phenotypes of CAFs (higher-risk and lower-risk) were identified. Given the descriptive nature of the available evidence and the absence of formal multivariable modelling, these findings should be considered hypothesis-generating rather than indicative of independent predictors.

Conclusions CAFs are heterogeneous vascular anomalies with variable clinical presentation and substantial potential for complications. An integrated assessment of anatomical configuration, fistula size, drainage pattern and ventricular function may support structured clinical evaluation and individualised management. We propose a practical anatomy- and risk-informed framework to guide multidisciplinary decision-making and follow-up strategies.

WHAT IS ALREADY KNOWN ON THIS TOPIC

⇒ Coronary artery fistulas (CAFs) are rare coronary anomalies with highly variable clinical presentation and natural history. Current management is largely individualised and no standardised risk stratification framework exists to guide treatment decisions, particularly in asymptomatic patients.

WHAT THIS STUDY ADDS

⇒ In a pooled analysis of 1966 patients, we identify anatomical and functional features associated with adverse outcomes and describe pragmatic higher-risk and lower-risk CAF phenotypes. We propose an anatomy-based and risk-based management framework integrating multimodality imaging and therapeutic selection.

HOW THIS STUDY MIGHT AFFECT RESEARCH, PRACTICE OR POLICY

⇒ This framework may support more structured Heart Team decision-making and follow-up strategies for CAFs. Prospective registries and guideline development are warranted to validate risk-based management pathways.

INTRODUCTION

Coronary artery fistulas (CAFs) are rare congenital or acquired communications between a coronary artery and a cardiac chamber, great vessel or vascular structure, bypassing the myocardial capillary bed.[1] Most are congenital, arising from failed involution of embryonic sinusoids; acquired forms occur after trauma, surgery or percutaneous coronary intervention.[2]

Epidemiological data suggest a prevalence of 0.002%–0.3% in the general population, with higher detection rates in angiographic cohorts due to advances in cardiac imaging.[1–3] Although many CAFs remain asymptomatic, a substantial proportion lead to clinically significant sequelae, including myocardial ischaemia, volume overload, arrhythmias, endocarditis, aneurysmal degeneration and, rarely, rupture.[1–4]

Complications are more frequent in lesions with complex anatomy, large shunt magnitude (Qp:Qs) or drainage into left-sided chambers or the coronary sinus (CS).[5 6] Anatomically, CAFs exhibit marked heterogeneity: the right coronary artery (RCA) is most commonly involved (38%), followed by the left anterior descending (LAD) and circumflex (LCx) arteries. Drainage most often occurs into the pulmonary artery (PA), right atrium (RA) or right ventricle (RV).[1–3] Multivessel and bilateral fistulas are less common but may evolve into aneurysms over time.[7]

Despite decades of clinical observation, optimal management remains controversial. Options include conservative follow-up, transcatheter closure with embolic devices and surgical ligation—each with variable success rates and risks depending on anatomy and patient characteristics.[2 8] Critically, no formal guidelines exist to guide decision-making, particularly in asymptomatic or incidentally detected cases.[9]

This review synthesises evidence from published case reports and series to characterise the natural history, anatomical features associated with risk and outcomes of CAFs. Given the rarity of the condition and the predominance of observational data, formal multivariable modelling is limited by heterogeneity. Nevertheless, pooled analysis reveals consistent patterns in presentation and complications. We propose a practical, anatomy- and risk-informed framework for patient stratification and management. Unless otherwise specified, data reflect the combined cohort of congenital and acquired CAFs, with distinctions made where relevant. The aim of this systematic review and pooled analysis was to characterise the anatomy, clinical presentation, natural history and management of CAFs, and to identify clinical and anatomical features associated with adverse outcomes in order to propose an anatomy- and risk-based management framework. These observations should be interpreted as descriptive and hypothesis-generating, given the heterogeneity of the available data.

METHODS

A systematic review with pooled analysis of published case reports, case series and observational studies was conducted in accordance with PRISMA 2020 guidelines. PubMed, Embase and Scopus were searched up to April 2025 using MeSH terms and keywords including “coronary artery fistula,” “CAF,” “fistulous connection” and “abnormal coronary drainage.” Eligible studies included case reports, case series and observational studies with clinical, anatomical or therapeutic data. Narrative reviews, abstracts without full text and duplicate publications were excluded. A study-level summary table was generated by grouping individual patient records according to first author and year of publication (online supplemental table S1).

Two reviewers independently screened titles and abstracts, extracted data on demographics, presentation, anatomy, imaging, management and outcomes. The final pooled cohort comprised 1966 patients with 1986 CAFs from published case reports and series (online supplemental figure 1). This review was registered in PROSPERO (CRD420251144984). The protocol is publicly available and no amendments were made after registration.

Descriptive statistics summarised demographic, anatomical and clinical characteristics. Comparative analyses between complicated and uncomplicated CAFs used proportional and mean differences. Associations between features and complications or treatment selection were evaluated descriptively, without formal inferential statistical testing. Anatomical comparisons were limited to cases with complete information on both origin and drainage. Formal multivariable modelling was not feasible due to heterogeneity and incomplete reporting; findings should therefore be interpreted as exploratory and hypothesis-generating rather than indicative of independent associations.

RESULTS

Study population and clinical characteristics

A total of 1966 patients with 1986 CAFs were included in the pooled analysis, derived from published case reports and case series (online supplemental table S1).

Congenital CAFs accounted for 90.7% of cases, while acquired forms represented 8.3% (table 1). Among acquired aetiologies, iatrogenic causes were reported in 4.4% of patients, including complications of coronary artery bypass grafting, valve surgery, percutaneous coronary intervention and endomyocardial biopsy, followed by trauma (2.0%), neoplastic (0.4%) and infectious causes (0.3%) (table 1).

In the pooled cohort, 65.3% of CAFs were diagnosed incidentally (table 1), while 18.5% presented acutely and 16.3% with chronic symptoms (table 1). Among symptomatic patients, the most frequently reported symptoms were chest pain (33%), dyspnoea (26.6%) and palpitations (5.8%), followed by murmurs (6.7%), arrhythmias (2.6%), fatigue (1.7%) and syncope (1.5%) (table 1).

Concomitant cardiovascular conditions included coronary artery disease (9%), valvular heart disease (5.5%) and structural or aortic abnormalities (5.7%) (table 1).

Anatomical characteristics

A total of 1986 CAFs were analysed (table 2). The most common site of origin was the RCA (38.2%), followed by the LAD (36.2%) and the LCx (16.2%). Left main (LM) origin was observed in 7.8% of cases, while other origins accounted for 1% (table 2).

Regarding termination, the most frequent drainage sites were the PA (34%), RA (28.3%) and left ventricle (LV) (11.9%). Additional drainage sites included the CS (11%), left atrium (LA) (3.6%), RV (2.3%), superior vena cava (2.6%), inferior vena cava (0.1%) and extracardiac locations (2.7%), while 3.5% were not specified (table 2).

Table 1 Baseline demographic, etiological and clinical and anatomical characteristics of the study population

Baseline characteristic, n (%)	Overall population (n=1966 by patients)
Age, years (mean±SD)	48.68±21.12
Paediatric age	213 (10.8)
Adult age	1551 (78.9)
Male gender	910 (46.3)
Cigarette smoking history	86 (4.4)
COPD	33 (1.7)
Asthma	10 (0.5)
Diabetes mellitus type II	66 (3.4)
History of CAD	98 (5)
Previous PCI	28 (1.4)
Previous cardiac surgery	115 (5.9)
Congenital aetiology	1783 (90.7)
Acquired aetiology	164 (8.3)
Idiopathic	26 (1.3)
Iatrogen	87 (4.4)
Trauma	39 (2)
Neoplastic	8 (0.4)
Infectious	5 (0.3)
Clinical presentation	
Incidental	1284 (65.3)
Acute	363 (18.5)
Chronic	321 (16.3)
Symptoms or signs	
Murmur	132 (6.7)
Chest pain	648 (33)
Dyspnoea	523 (26.6)
Palpitation	113 (5.8)
Syncope	30 (1.5)
Fatigue	33 (1.7)
Arrhythmia	51 (2.6)
Concomitant pathology	
Valvular heart disease	109 (5.5)
CAD	177 (9)
Pre-existing arrhythmia	31 (1.6)
Extracardiac condition	43 (2.2)
Aortic syndrome or other cardiac structural disease	111 (5.7)

Percentages may not sum to 100% due to missing data.
CAD, coronary artery disease; COPD, Chronic Obstructive Coronary Disease; PCI, percutaneous coronary intervention.

Table 2 Anatomical characteristics of coronary artery fistulas

Origin of fistulas, n (%)	Overall (n=1986 by fistulas)
Right coronary artery	759 (38.2)
Left anterior descending coronary artery	719 (36.2)
Left circumflex coronary artery	321 (16.2)
Left main coronary artery	155 (7.8)
Other	19 (1)
Termination of fistulas, n (%)	Overall (n=1986 by fistulas)
Pulmonary artery	675 (34)
Right atrium	563 (28.3)
Left ventricle	236 (11.9)
Coronary sinus	219 (11)
Left atrium	71 (3.6)
Right ventricle	45 (2.3)
Superior vena cava	52 (2.6)
Inferior vena cava	2 (0.1)
Extracardiac	53 (2.7)
Other/not specified	70 (3.5)
Multiplicity, n (%)	Overall (n=1966 by patients)
Single fistula	1581 (80.4)
Multiple fistulas	385 (19.6)

A single fistula was present in 80.4% of patients, whereas 19.6% had multiple fistulas (table 2).

Natural history and complications

Overall, 41.5% of patients presented with at least one complication (table 3). The most frequently reported complications were coronary aneurysm (23.2%) and volume overload (20.4%) (table 3). Acute coronary syndromes occurred in 2.6% of cases, fistula rupture in 1.3% and arrhythmias in 0.7% (table 3).

Median fistula diameter was 8.4 mm (IQR 5.0–15.0) in patients without complications and 10.0 mm (IQR 6.0–29.5) in those with complications (table 3). Larger diameters were observed in aneurysmal fistulas (13.0 mm (IQR 8.0–30.0)) and in cases of rupture (30.0 mm (IQR 15.0–37.5)) (table 3).

Median Qp:Qs was 1.40 (IQR 1.29–1.60) in uncomplicated cases and 1.55 (IQR 1.30–2.00) in patients with complications (table 3).

Complication rates increased across fistula diameter categories, from lesions <5 mm to those >20 mm. A progressive increase in complication rates was observed with increasing fistula diameter and shunt magnitude. Complication rates rose from approximately 50% in lesions with Qp:Qs<1.2 to over 70% in those with Qp:Qs>2.0 (figure 1).

Diagnostic imaging

The most frequently reported diagnostic modality was invasive coronary angiography, performed in 52.7% of patients (table 4). Transthoracic echocardiography (TTE) was used in 24.7% of cases and coronary CT angiography (CCTA) in 10.8% (table 4).

Other imaging modalities included nuclear imaging (1.6%), cardiac MRI (0.7%), and intraoperative diagnosis (0.05%) (table 4).

Table 3 Clinical course and complications of coronary artery fistulas

Complication	n, (%)	Diameter, mm (median (IQR))	Qp:Qs (median (IQR))
Any complication	817 (41.5)	10.0 (6.0–29.5)	1.55 (1.30–2.00)
Coronary aneurysm	456 (23.2)	13.0 (8.0–30.0)	1.60 (1.30–2.00)
Volume overload	401 (20.4)	10.1 (5.8–25.0)	1.65 (1.40–2.21)
Acute coronary syndrome	51 (2.6)	7.0 (4.0–30.0)	1.29 (1.06–1.53)
Arrhythmia	14 (0.7)	–	–
Rupture	26 (1.3)	30.0 (15.0–37.5)	2.10 (1.90–2.30)
No complication	1148 (58.5)	8.4 (5.0–15.0)	1.40 (1.29–1.60)

Management strategies

Overall, 36.6% of patients were managed conservatively, while 63.4% underwent an interventional procedure (table 4). Among treated patients, surgical repair was performed in 869 cases (69.7%), and transcatheter closure alone in 365 cases (29.3%) (table 4).

Among percutaneous procedures, device use included coils in 51.8% of cases, vascular plugs in 28.9%, covered stents in 6.7% and balloon occlusion in 3.6% (table 4). Residual shunt after transcatheter closure was reported in 48.5% of cases (table 4). Among patients undergoing transcatheter closure, device selection varied according to anatomical and clinical characteristics. Coils were more frequently used in smaller or distal fistulas and in cases with PA termination, whereas vascular plugs were more commonly employed in larger, high-flow fistulas, LM origin or paediatric patients (figure 2).

Surgical techniques included on-pump direct closure in 68.9% of cases, off-pump approaches in 2.2% and hybrid procedures in 0.9% (table 4). Concomitant procedures included coronary artery bypass grafting in 7.7% of patients and valve surgery in 3.4% (table 4).

DISCUSSION

Principal findings

This comprehensive pooled analysis of 1966 patients with 1986 CAFs provides an integrated overview of their anatomical characteristics, clinical presentation, natural

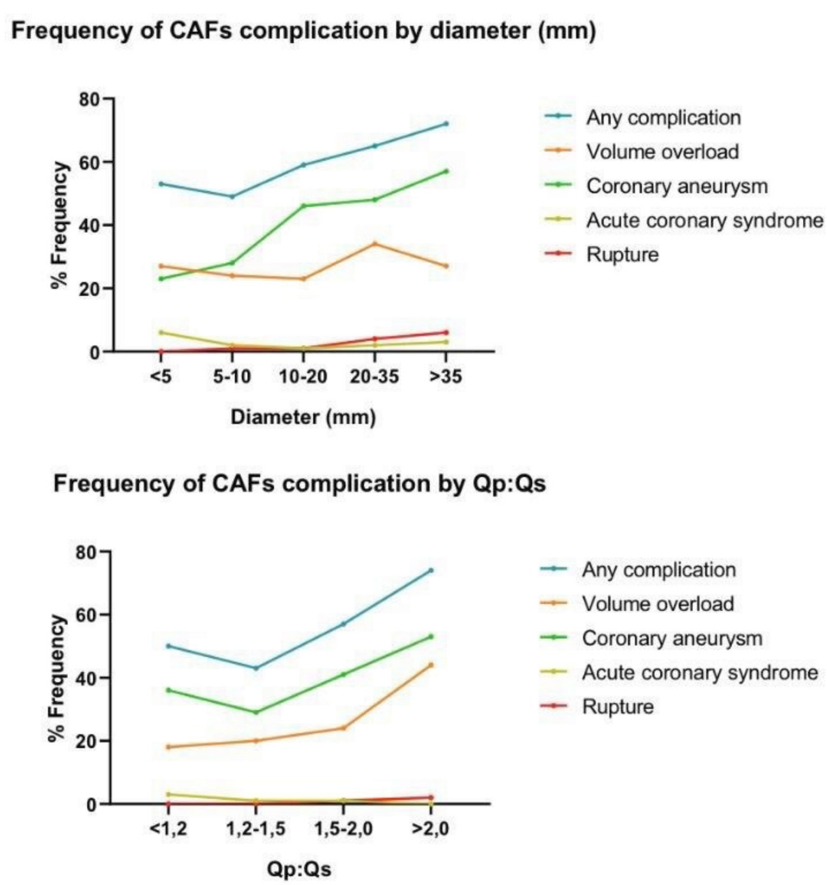

Figure 1 Frequency of specific complications by fistula diameter and shunt (Qp:Qs) classes. CAFs, coronary artery fistula.

Table 4 Diagnostic pathways and therapeutic management of coronary artery fistulas

Category	Modality/strategy	n (%)	Clinical role/remarks
Diagnostic imaging	Transthoracic echocardiography	485 (24.7)	Common first-line test in patients with murmur or vague symptoms; useful for chamber size, ventricular function and shunt suspicion. Limited anatomical definition in adults.
	Coronary angiography	1036 (52.7)	Most frequently used diagnostic modality; gold standard for defining coronary origin and planning intervention, but limited for distal anatomy and shunt quantification.
	Coronary CT angiography	212 (10.8)	Increasingly used as first-line or early second-line imaging; excellent spatial resolution for defining origin, course, and termination; key for procedural planning.
	Cardiac MRI	14 (0.7)	Limited availability; provides functional assessment, ventricular volumes and Qp:Qs quantification.
	Nuclear imaging/other	31 (1.6)	Occasionally used to assess ischaemia; low specificity for anatomical diagnosis.
	Intraoperative diagnosis	1 (0.05)	Rare incidental finding during cardiac surgery.
Therapeutic strategy	Conservative management	720 (36.6)	Preferred in asymptomatic patients with small-calibre fistulas, no significant shunt and no chamber enlargement; requires structured imaging follow-up.
	Any closure*	1246 (63.4)	Indicates patients who underwent a definitive interventional strategy.
	Surgical closure (overall)	869 (44.2)	Favoured for large, tortuous, aneurysmal, multifocal fistulas, venous or left-sided drainage, or when concomitant cardiac surgery is required.
	On-pump direct closure	599 (68.9†)	Standard surgical approach allowing definitive control of origin and termination.
	Off-pump epicardial closure	19 (2.2)†	Selected proximal and superficial fistulas; risk of incomplete closure in deep or intramyocardial tracts.
	Hybrid/other	8 (0.9)†	Selected complex anatomies or incomplete preoperative mapping.
	Transcatheter closure (any)	388 (19.7)	Preferred in anatomically suitable fistulas with single origin and favourable landing zones.
	Only PCI	365 (94.1)‡	Definitive percutaneous treatment.
	PCI followed by surgery	23 (5.9)‡	Due to residual shunt or technical failure; already included in the surgical closure group.
Percutaneous devices	Coils	201 (51.8)	Preferred for small, tortuous, or distal fistulas.
	Amplatzer/vascular plug	112 (28.9)	Favoured for large-calibre or high-flow fistulas.
	Covered stent	26 (6.7)	Used mainly for venous terminations or complex anatomy.
	Balloon occlusion	14 (3.6)	Rarely used; selected cases.
	Other	2 (0.1)	—
	Residual shunt after PCI§	188 (48.5)	Detected at follow-up imaging.
Concomitant procedures	CABG	152 (7.7)	Often performed in patients with coexisting coronary artery disease.
	Valve surgery	66 (3.4)	Combined procedures in selected patients.
	CABG+valve surgery	5 (0.25)	—
	Other	4 (0.2)	—

*‘Any closure’ represents the overall number of patients who underwent a definitive intervention. This total includes: (1) all surgical closures (n=869, which already incorporate the 23 patients who underwent PCI followed by surgery); (2) patients treated with PCI alone (n=365) and (3) patients in whom the type of closure was not reported (n=12). ‘Only PCI’ therefore contributes entirely to the ‘any closure’ total, whereas ‘PCI followed by surgery’ should not be double-counted.
†Percentages calculated relative to the surgical closure group.
‡Percentages calculated relative to the transcatheter closure group.
§Residual shunt after PCI includes mild or trivial residual flow.
CABG, coronary artery bypass grafting; PCI, percutaneous coronary intervention.

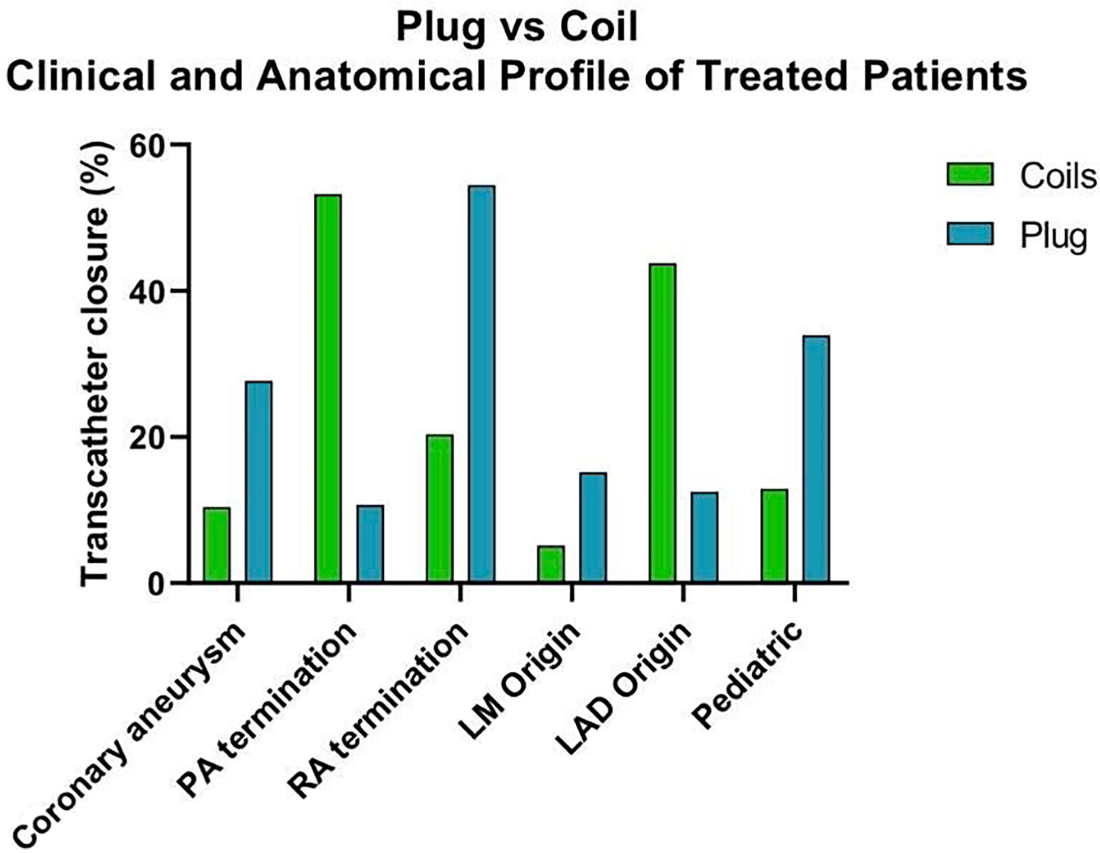

Figure 2 Clinical and anatomical characteristics associated with plug versus coil use in percutaneous closure of coronary artery fistulas. LAD, left anterior descending; LM, left main; PA, pulmonary artery; RA, right atrium.

history, and management strategies. Given the rarity of the condition and the predominance of case reports and small case series, this dataset represents one of the largest aggregated descriptions currently available in the literature.

In the present analysis, CAFs demonstrated marked heterogeneity across all domains, including origin, drainage site, number of tracts, clinical presentation and outcomes. Although a substantial proportion of cases were identified incidentally (65.3%), a considerable burden of disease was observed, with 41.5% of patients presenting with at least one complication. The most frequently reported adverse events were coronary aneurysm and volume overload, consistent with the haemodynamic consequences of chronic abnormal shunting and altered coronary flow patterns (table 3). Less frequent but clinically relevant complications included acute coronary syndromes, fistula rupture and arrhythmias.

Across the pooled cohort, specific anatomical and clinical features were more frequently observed in patients with complications. These included RCA and LM origin, drainage into venous or left-sided structures, larger fistula diameter and reduced ventricular function. These findings are consistent with previously described mechanisms such as coronary steal, progressive vessel dilation and volume overload, which may contribute to adverse clinical evolution.[1 10 11]

Importantly, the available evidence is largely derived from heterogeneous observational reports, with variable definitions, imaging modalities and follow-up durations. Therefore, the patterns observed in this analysis should be interpreted as descriptive rather than causal and they do not allow identification of independent predictors of adverse outcomes.

Anatomical and clinical features associated with complications

The distribution of anatomical and clinical features differed between the overall cohort and patients presenting with complications, highlighting recurring patterns that may be clinically relevant (table 5). Anatomical comparisons were restricted to cases with complete information on both origin and drainage (n=1723), whereas non-anatomical variables were analysed in the overall cohort with available data. In particular, fistulas originating from the RCA were more frequently represented among complicated cases, especially when associated with drainage into venous or left-sided structures. The RCA-to-CS configuration showed the largest excess among complicated CAFs, followed by RCA-to-LV and RCA-to-PA pathways, which were also more commonly observed in patients with adverse outcomes.

Fistulas involving the LCx, particularly those draining into the CS, were similarly more frequent among

Table 5 Anatomical distribution of coronary artery fistulas (CAF) and non-anatomical features in the total cohort and in patients with complications

Anatomical distribution	Total CAF (N=1723) n (%)	Complicated (N=735) n (%)	Difference (%)	Associated complications
RCA → coronary sinus	115 (6.7)	82 (11.2)	+4.5	Coronary aneurysm, arrhythmia
RCA → left ventricle	90 (5.2)	53 (7.2)	+2	Acute coronary syndrome, rupture, coronary aneurysm
LCX → coronary sinus	87 (5)	49 (6.7)	+1.7	Coronary aneurysm, volume overload, arrhythmia
LM → right atrium	53 (3.1)	28 (3.8)	+0.7	Volume overload, arrhythmia
RCA → pulmonary artery	161 (9.3)	74 (10.1)	+0.8	Volume overload, coronary aneurysm
LM → coronary sinus	7 (0.4)	6 (0.8)	+0.4	Coronary aneurysm, arrhythmia
RCA → right ventricle	23 (1.3)	12 (1.6)	+0.3	Volume overload, arrhythmia
LCX → left ventricle	21 (1.2)	12 (1.6)	+0.4	Acute coronary syndrome
LM → left atrium	9 (0.5)	6 (0.8)	+0.3	Acute coronary syndrome, coronary aneurysm
LM → left ventricle	8 (0.5)	5 (0.7)	+0.2	Acute coronary syndrome, rupture
LM → right ventricle	4 (0.2)	2 (0.3)	+0.1	Volume overload
Non-anatomical distribution	Total CAF (N=1966) n (%)	Complicated (N=817) n (%)	Difference (%)	Associated complications
Multiple fistulas	385 (19.6)	173 (21.2)	+1.6	Volume overload, aneurysm, arrhythmia
Adult age group	1551 (78.9)	732 (89.6)	+10.7	ACS, aneurysm, arrhythmia
Large diameter (>10 mm)*	124 (6.3)	78 (9.5)	+3.2	Aneurysm, rupture
Significant shunt (Qp/Qs >1.5)	116 (5.9)	74 (9.1)	+3.2	Volume overload
Reduced LVEF (<50%)	45 (2.3)	27 (3.3)	+1	Volume overload, ACS

Data are presented as n (%). Difference (%) represents the absolute percentage-point difference between complicated cases and the overall cohort. Anatomical distributions are reported for cases with complete information on both origin and drainage (n=1723). Non-anatomical variables are based on available data and may not include all patients due to missing values.
*Diameter thresholds based on reported measurements.
ACS, acute coronary syndrome; LCx, left circumflex; LM, left main; LVEF, left ventricular ejection fraction; RCA, right coronary artery.

complicated cases. These configurations are often characterised by tortuous anatomy and complex venous drainage patterns, which may contribute to progressive dilation and technical challenges during closure. LM coronary artery–origin fistulas, although less frequent, were proportionally more represented among complicated cases and frequently associated with complex anatomical features. LM-derived CAFs were associated with large vessel calibre, high-flow physiology and proximity to the coronary bifurcation, features that have been previously associated with aneurysmal transformation, ischaemic phenomena and increased procedural complexity.[1 5 10 11]

Fistula size also showed a consistent relationship with complications. Larger diameters were more frequently observed among complicated cases, particularly in aneurysmal and ruptured fistulas (table 3). This finding aligns with previous observations suggesting that progressive dilation may reflect chronic exposure to high-flow states and altered wall stress.[10 12 13] Similarly, adult age and reduced LV ejection fraction were more common among patients with complications (table 5), reflecting the cumulative effects of long-standing haemodynamic burden and potential myocardial impairment.

The proportion of patients with Qp:Qs>1.5 was slightly higher among complicated cases; however, this finding should be interpreted with caution due to inconsistent reporting and potential measurement variability (table 5). This finding may be influenced by several factors, including earlier intervention in patients with significant shunting, variability in measurement techniques and incomplete reporting across studies. While Qp:Qs remains an important descriptor of haemodynamic burden in individual patients, its role as a standalone marker within retrospective pooled datasets appears limited.[14 15]

Overall, these findings support a multifactorial model in which anatomical configuration, vessel size and patient-related factors collectively contribute to the observed heterogeneity in clinical outcomes.

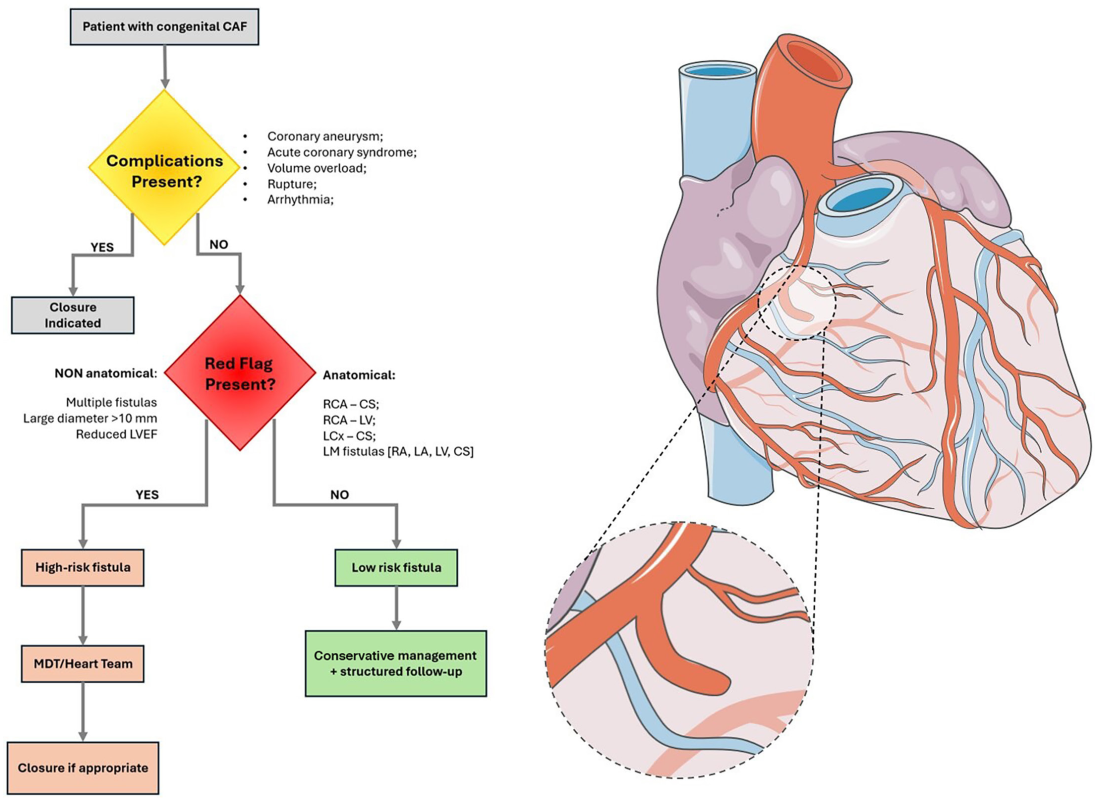

Figure 3 Proposed framework for risk stratification and management of coronary artery fistulas (CAF). CS, coronary sinus; LA, left atrium; LCx, left circumflex; LM, left main; LV, left ventricle; LVEF, LV ejection fraction; MDT, multidisciplinary team; RA, right atrium; RCA, right coronary artery.

Pragmatic risk framework and clinical implications

Based on the patterns observed in the pooled dataset, it is possible to describe a set of recurring anatomical and clinical features that tend to cluster in patients with complications (table 5). These include proximal coronary origin, particularly from the RCA or LM, drainage into venous or left-sided structures, large-calibre fistulas and reduced ventricular function. Rather than acting in isolation, these variables appear to interact in a multifactorial manner, contributing to the heterogeneity of clinical presentation and outcomes.

On this basis, a pragmatic distinction between higher- and lower-risk CAF phenotypes can be described. Higher-risk configurations are characterised by one or more of the following features: large vessel diameter, proximal origin (RCA or LM), complex or venous drainage (eg, CS), evidence of ventricular dysfunction or signs of haemodynamic impact. In contrast, lower-risk lesions are more commonly small, single, right-sided fistulas without associated chamber enlargement or functional impairment. This conceptual classification is consistent with prior reports emphasising the role of anatomical complexity and haemodynamic burden in determining clinical course and treatment selection.[1 5 10 11]

The proposed framework (figure 3) integrates these elements into a structured approach to evaluation and management. Initial assessment includes TTE for detection of chamber enlargement and functional abnormalities, followed by detailed anatomical characterisation using CCTA or ICA. Cardiac MRI may provide complementary information on shunt magnitude (Qp:Qs) and ventricular remodelling when required.[16–18]

This integrated imaging strategy allows comprehensive characterisation of fistula anatomy, drainage patterns, and physiological impact, forming the basis for subsequent decision-making. In patients with higher-risk features, evaluation within a multidisciplinary Heart Team is warranted to determine the indication for closure and the most appropriate therapeutic approach, taking into account anatomical complexity, procedural feasibility and patient-specific factors.[1 11 19] In contrast, lower-risk lesions may be managed conservatively with periodic imaging follow-up, particularly in the absence of symptoms or haemodynamic consequences.

Importantly, given the descriptive nature of the analysis and the absence of formal multivariable modelling, these features should not be interpreted as independent predictors of adverse outcomes. Rather, they should be considered clinically relevant patterns that may support structured assessment and individualised decision-making in the context of limited and heterogeneous evidence.

Clinical management considerations

Management of CAFs remains heterogeneous and largely individualised, reflecting both the anatomical variability of the condition and the absence of standardised, evidence-based guidelines. In the present pooled analysis, a substantial proportion of patients underwent interventional treatment, although this distribution is likely influenced by publication bias, as cases managed conservatively are less frequently reported in the literature.

Therapeutic strategies include conservative management, transcatheter closure and surgical repair, with selection primarily driven by anatomical configuration, shunt magnitude, clinical presentation and the presence of complications. Rather than being mutually exclusive, these approaches should be viewed along a continuum of care, in which treatment decisions are tailored to the individual patient profile.

Transcatheter closure represents a less invasive option and is typically applied in anatomically favourable lesions, such as single, well-defined fistulas with accessible trajectories. Device selection—including coils, vascular plugs or covered stents—is guided by fistula size, flow characteristics and drainage pattern. Prior studies have demonstrated high procedural success rates, although residual shunting and recanalisation may occur, particularly in complex or high-flow lesions.[1 19–21]

Surgical repair remains a cornerstone in the management of CAFs, particularly in cases characterised by large, tortuous or multifocal fistulas, or when associated with other cardiac conditions requiring concomitant intervention. The ability to directly visualise and control both the origin and termination of the fistula allows for definitive closure, with reported low rates of recurrence and durable long-term outcomes.[13 22 23] However, surgical intervention is inherently more invasive and must be balanced against patient comorbidities and procedural risk.

Conservative management may be considered in selected patients, particularly those with small, asymptomatic fistulas and no evidence of haemodynamic impact. However, the natural history of untreated CAFs remains incompletely defined, and a proportion of patients may develop complications over time. For this reason, when a conservative approach is adopted, structured follow-up with periodic imaging is essential to monitor for changes in fistula size, shunt magnitude or ventricular remodelling.

Within this context, the proposed framework (figure 3) may help to organise clinical decision-making by integrating anatomical, functional and clinical variables. Rather than providing prescriptive recommendations, this approach supports a structured evaluation process in which the key question is not only how to treat a CAF but whether intervention is indicated and in which clinical context. In particular, multidisciplinary Heart Team discussion may be especially valuable in patients with higher-risk features or borderline indications for closure, allowing integration of imaging findings, procedural feasibility and patient-specific considerations.[1 11 19]

Overall, these considerations underscore the need for individualised management strategies and highlight the limitations of current evidence, emphasising the importance of prospective data collection and standardised reporting to better inform future recommendations.

LIMITATIONS

This study has several limitations. First, the analysis is subject to the limitations inherent to pooling data derived largely from retrospective case series and registries, in which patient selection, reporting accuracy and follow-up duration are heterogeneous. A significant publication bias must be acknowledged, as most reports describe patients undergoing intervention, whereas conservatively managed cases are less frequently published, potentially leading to an overestimation of procedural outcomes and complication rates.

In addition, anatomical and clinical variables were inconsistently reported across studies, limiting the ability to standardise classification and adjust for potential confounders. Thresholds for haemodynamic significance, including vessel diameter and Qp:Qs ratio, varied across reports and were not uniformly assessed using the same imaging modalities, introducing measurement variability.

Moreover, the relative contribution of individual features identified in table 5 should be interpreted with caution, as formal multivariable analyses were not consistently feasible across studies. As a result, these findings do not allow inference of independent associations and should be considered descriptive.

Finally, long-term outcomes beyond the immediate procedural period remain incompletely characterised, particularly with regard to the natural history of conservatively managed fistulas, the durability of transcatheter closure and the risk of late complications such as recanalisation or aneurysmal progression.

These limitations highlight the need for prospective multicentre registries, standardised reporting and uniform follow-up protocols to refine risk stratification approaches and better inform management strategies in this rare but clinically heterogeneous condition.

CONCLUSIONS

CAFs are heterogeneous vascular anomalies with a variable clinical course and a substantial burden of complications. In this pooled analysis, approximately 40% of patients presented with adverse events, most commonly coronary aneurysm and volume overload, highlighting that these lesions cannot be uniformly considered benign.

Specific anatomical and clinical features, including larger fistula diameter, proximal coronary origin (particularly RCA or LM), complex or venous drainage patterns and reduced ventricular function, were more frequently observed in patients with complications. In contrast, shunt magnitude (Qp:Qs) showed a less consistent relationship with adverse outcomes within this heterogeneous dataset.

On the basis of these observations, a pragmatic, anatomy- and risk-informed framework can be proposed to support clinical evaluation and management. Higher-risk configurations may warrant multidisciplinary assessment to determine the appropriateness and modality of intervention, whereas lower-risk lesions may be managed conservatively with structured follow-up.

Given the descriptive nature of the available evidence, these findings should be interpreted as hypothesis-generating rather than definitive. Further prospective studies, multicentre registries and standardised reporting are needed to validate risk-based approaches and to inform future guideline development in this complex and relatively uncommon condition.

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