Coronary Artery Fistula from A to Z
: Assessed by ECG-gated Coronary CT Angiography

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What is coronary artery fistula (CAF)?

- Unusual congenital or acquired coronary artery abnormalities that can be defined as direct vascular connections from a coronary artery to a cardiac chamber, great vessel or other structure, without intervening capillary network.
Etiologies of coronary artery fistula (CAF)

**Embryological development of CAF**

- In the early embryologic period, persistence of sinusoidal connections between the lumens of the primitive tubular heart that supply myocardial blood flow.
- Normally, the intramyocardial sinusoids become narrowed and persist only as thebesian vessels in the adult.
- If obliteration of the intramyocardial trabecular sinusoids fails, a fistulous communication persists between the coronary arteries and a cardiac chamber.

**Congenital**
- Embryonic (m/c)
  - Multiple; systemic hemangioma

**Acquired**
- Disease related
  - Acute myocardial infarction
  - Hypertrophic cardiomyopathy
  - Dilated cardiomyopathy
  - Tumor
- Iatrogenic
  - Balloon angioplasty
  - Coronary Artery Bypass
  - Grafting
  - Permanent pacemaker
  - Endomyocardial biopsy
- Traumatic
  - Penetrating and
  - nonpenetrating trauma

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Pathophysiology of coronary artery fistula (CAF)

- Single origin is the most common from, ranging from 74-90% of the cases.
- Origin of CAF can be any of the three major coronary arteries, including left main artery: right coronary artery (RCA) or its branches is the most common site with 55% of the incidence, followed by left anterior descending (LAD) and left circumflex arteries (LCX).
- Usually drains into low-pressure structures, and right-sided chamber, pulmonary artery, coronary sinus or SVC, therefore Left-to-right shunt exists in over 90% of cases, and rarely drain into left-sided chamber.

Clinical manifestations of coronary artery fistula (CAF)

- In 1963 Haller and Little described the clinical presentations of CAFs as a triad of cardiac murmur, atrial or ventricular left to right shunting, and a large tortuous coronary artery.
- Most patients are asymptomatic, therefore CAFs are usually discovered incidentally.
- However, according to the size and localization of the CAF and severity of the left to right shunt, various symptoms from mild dyspnea, fatigue to congestive heart failure and myocardial infarction are occurred.

<table>
<thead>
<tr>
<th>Pathophysiology of CAF</th>
<th>Clinical features</th>
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<tbody>
<tr>
<td>Myocardial steal phenomenon</td>
<td>angina, infarction, and arrhythmia</td>
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<tr>
<td>Left-to-right shunt</td>
<td>pulmonary hypertension, congestive heart failure (volume overloading both ventricles)</td>
</tr>
<tr>
<td>Left-to-left shunt</td>
<td>volume overloading the left ventricle only</td>
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<tr>
<td>Other complications</td>
<td>endocarditis, rupture or thrombosis of the fistula or associated arterial aneurysm or embolic events</td>
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## Advantage and limitations of various imaging tools for CAF

<table>
<thead>
<tr>
<th>Modalities</th>
<th>Advantages</th>
<th>Disadvantages</th>
</tr>
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<tbody>
<tr>
<td><strong>CT</strong></td>
<td>- Non-invasive</td>
<td>- High radiation dose</td>
</tr>
<tr>
<td></td>
<td>- Short acquisition time</td>
<td>- Contrast media</td>
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<tr>
<td></td>
<td>- Excellent high spatial resolution</td>
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<td></td>
<td>- Accurate cross-sectional imaging in demonstrating routes of coronary artery</td>
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<td></td>
<td>- Multiple planes and three-dimensional views for cardiac and coronary anatomy.</td>
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<tr>
<td><strong>Angiography</strong></td>
<td>- Evaluation of proximal part of the CAF, size or number of fistulas</td>
<td>- Invasive</td>
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<tr>
<td></td>
<td></td>
<td>- Limited in evaluating the relation of CAF to other structures, distal part d/t dilution of the contrast medium</td>
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<tr>
<td><strong>Echocardiography</strong></td>
<td>- Lack of exposure to ionizing radiation</td>
<td>- Limited field of view</td>
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<tr>
<td></td>
<td>- Microbubbles as contrast medium can increase the identification of CAFs and their course</td>
<td>- Limited efficacy in visualization of complex anatomy, distal portals of CAFs, occluded vessels, and collateral vessels</td>
</tr>
<tr>
<td><strong>MR angiography</strong></td>
<td>- Non-invasive</td>
<td>- High cost</td>
</tr>
<tr>
<td></td>
<td>- Lack of exposure to ionizing radiation</td>
<td>- Long acquisition time</td>
</tr>
<tr>
<td></td>
<td>- High temporal resolution</td>
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## Comparison of Imaging tools for evaluation of CAF

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>CTA</th>
<th>Angiography</th>
<th>Echocardiography</th>
<th>MRA</th>
</tr>
</thead>
<tbody>
<tr>
<td>Spatial resolution</td>
<td>++</td>
<td>+++</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Temporal resolution</td>
<td>++</td>
<td>+++</td>
<td>++</td>
<td>+</td>
</tr>
<tr>
<td>Ionizing radiation</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Imaging duration</td>
<td>Shorter</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Iodinated contrast medium</td>
<td>++</td>
<td>++</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Evaluation of origin</td>
<td>++</td>
<td>++</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Hemodynamic assessment and flow</td>
<td>-/+</td>
<td>++</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>multiple fistulas and complexity</td>
<td>++</td>
<td>++</td>
<td>-/+</td>
<td>+</td>
</tr>
<tr>
<td>Drainage route and distal entry</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>-/+</td>
</tr>
<tr>
<td>Confirmation of thrombosis of fistula</td>
<td>+</td>
<td>+</td>
<td>-/+</td>
<td>-/+</td>
</tr>
<tr>
<td>extracardiac structures</td>
<td>++</td>
<td>-</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td>Need for B blocker</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>-/+</td>
</tr>
<tr>
<td>Therapeutic</td>
<td>-</td>
<td>+</td>
<td>-</td>
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Modified from AJR 2014; 203:W244–W252
## Role of Coronary CT angiography (CCTA) for CAF

### Detection and Diagnosis for the CAF

- Location of the Origin
- Location of the drainage site
- Number of origin site, drainage site and fistulous tracts
- Vessel size of the fistulous tract
- Complexity

### Presence of the associated other congenital heart abnormalities

- Tetralogy of Fallot
- Atrial septal defect, ventricular septal defect
- Patent ductus arteriosus
- Pulmonary atresia with intact ventricular septum
### Protocols of CCTA for the assessment of CAF

<table>
<thead>
<tr>
<th>Type of scanner</th>
<th>320 × 0.5 mm MDCT</th>
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<tr>
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<td>32 × 2 × 0.6 mm MDCT</td>
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**Scan parameters**

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Value</th>
</tr>
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<tbody>
<tr>
<td>Slice collimation (mm)</td>
<td>0.5</td>
</tr>
<tr>
<td>Slice thickness (mm)</td>
<td>0.5</td>
</tr>
<tr>
<td>Peak kilovoltage (kVp)</td>
<td>80–120</td>
</tr>
<tr>
<td>Tube current (mA)</td>
<td>Varies with body mass index</td>
</tr>
</tbody>
</table>

**Scan range**

From tracheal bifurcation to the inferior margin of the heart

**Gating method**

- Retrospective ECG gating and ECG-dependent tube current modulation
- Prospective gating due to less radiation exposure

**Contrast medium**

Nonionic iodinated contrast medium: 60–90 mL at 5–6 mL/s

**Scan triggering method**

Bolus-tracking technique for better opacification of fistula

**Scan delay**

determined on the basis of the peak enhancement in the ascending aorta
Coronary-Pulmonary Fistula

Coronary-Cameral Fistula

Coronary-Bronchial Fistula

Coronary-Venous Fistula

Coronary-Extracardiac connections
Coronary-Pulmonary Fistula

- Fistula with **pulmonary artery**
- 15% of coronary fistulas drain into the pulmonary arterial circulation
- 0.13% of all coronary angiograms
- Remnants of the **splanchnic vascular system** and normally regress with the formation of the pulmonary arterial system later in fetal life
- However, they may persist when the central pulmonary arterial system fails to develop adequately
- The **left to right shunt ratio is usually small** in coronary-pulmonary fistula
- Shunt murmur loudest near the **second intercostal space to the left** of sternum on physical examination.
A. Coronary-Pulmonary Fistula

Fig. 1. A 55-year-old man with dyspnea.
A & B. Serial axial CT (A) and Volume rendered CT image(B) shows dilated and tortuous branch (arrowheads) which is connected between proximal LAD (yellow arrow) and main pulmonary trunk (red arrow).
C. Invasive coronary Angiography also confirmed the dilated vessel (arrowhead) which is originated from proximal LAD (yellow arrow) and drained into main pulmonary trunk (red arrow).
Fig. 2. A 62-year-old man with incidental findings on health screening. A & B. Serial axial CT images (A) and volume rendered image (B) show dilated and tortuous vessels (arrowhead) from proximal LCX (yellow arrow), connected to right pulmonary artery (red arrow). B. Coronary Angiography also confirmed the coronary-pulmonary fistula (arrowhead) originating from proximal LCX (yellow arrow) draining into right pulmonary artery (red arrow).
B. Coronary-Cameral Fistula

• Fistulas with the cardiac chambers
• 0.08% to 0.3% of unselected patients undergoing diagnostic coronary angiography
• 90% with the right-sided chambers, rest drain to the left side of the heart, or to both
• Coronary-cameral fistulas to left heart chambers are uncommon; when present, they result in a left-to-left shunt and can manifest with signs and symptoms that resemble those of aortic valve insufficiency.
• Arterioluminal fistula; direct and focal communication with the cardiac chamber concerned, or arteriosinusoidal, where arterial blood communicates with the cardiac chambers via a sinusoidal network.
B. Coronary-Cameral Fistula

Fig 3. A 42-year-old male with cardiac murmur
A. Volume rendered CT images show diffusely dilatated RCA (arrows), which drains into LV chamber (arrowheads).
B. Serial axial CT images demonstrate dilated RCA along the AV groove (arrows) which drains into LV chamber (arrowheads).
C. Curved multiplanar reformatted image depicts fistulous communication of dilated RCA (arrow) with LV chamber (arrowheads).
Fig. 4. Incidental findings in 55-year-old male.

A-C. Volume rendered CT images show diffusely dilated OM branch (arrows), which drains into LV chamber (arrowhead).

D. Curved multiplanar reformatted image depicts fistulous communication of dilated LCX /OM branch (arrow) with LV chamber (arrowheads).
Fig. 5. 63-year-old male with atrial fibrillation and dyspnea.
A-C. Volume rendered CT images show calcified, dilated RCA (arrow), coursing along the LV draining into RV with aneurysmal change distally (arrowhead).
D. Short axis CT images reveal an origin of dilated RCA (arrow) and RV drainage site (arrowhead).
C. Coronary-Bronchial Fistula

- Less than 1% according to CT findings
- Most commonly from the LCX draining into the bronchial artery
- Can be symptomatic with hemoptysis (probably due to accompanying bronchiectasis)
- Typically found in patients with pulmonary artery occlusive disease or chronic lung disease
- 12 reported cases in 2003, found that the most common associations were bronchiectasis (67%) followed by CAD (33%). (Jim et al.)
- PTE procedure can be complicated by MI due to distal thrombus embolisation into the coronary artery branch -> MI has been reported as a serious complication of bronchial artery embolization.
- Prophylactic coronary-to-bronchial artery embolization will become a new treatment indication, which has to be determined individually, depending on the severity of the underlying disease and symptoms.
C. Coronary-Bronchial Fistula

Fig. 6. A 58 year old man with intermittent cough.

A. Volume rendered CT image shows SA nodal branch of proximal LCX (yellow arrow) running along LA, which continues with bronchial arteries (red arrow).

B-D. Serial axial CT images show tortuous SA nodal branch of LCX (yellow arrows) along the retroaortic course forming a fistulous communication with bronchial arteries (red arrows).

E. Axial CT image in lung window shows cystic bronchiectasis in LLL.

(Ao = Aorta; PA= Pulmonary artery; LV = left ventricle)
D. Coronary-Venous (Cardiac vein) Fistula

- Fistulous track with **coronary sinus**.
- particularly at **high risk of long-term morbidities** after closure

**Fig. 7.** A 57 year old female with chest pain.

A-D. Volume rendered CT images show tortuous first diagonal branch (D1) and second diagonal branch (D2) of LAD, which is connected to dilated middle cardiac vein (MCV) draining into coronary sinus (CS).

(D1 = 1\textsuperscript{st} branches of diagonal branch; D2 = 2\textsuperscript{nd} branches of diagonal branch; MCV = middle cardiac vein; CS = coronary sinus)
E. Coronary-Extracardiac Connections

- 10.7% to 16% of all coronary artery fistulas
- Best obliterated by operation, on CPB to completely close all fistulous tracts.

Fig. 10. A 48 year old male with mild dyspnea.
A. CT volume rendered image demonstrates tortuous diagonal branches of LAD (D1), which connected (arrowheads) with dilated tortuous internal mammary artery (IMA) and inferior phrenic artery (IPA) from thoracic aorta. It is also continuous with the left marginal vein (LMV), which drains into great cardiac vein (GCV) (arrows).
B. CT 2 chamber image depict inferior phrenic artery (IPA) from thoracic aorta extending into diagonal branch of LAD (D1).
• Small fistulas -> no treatment.
• Large fistula with significant shunt -> Treatment

1) **Conservative medical management**
   • Antiplatelet agents and antibiotics for prevention of the thrombosis and endocarditis.
   • Spontaneous closure of the fistula secondary to spontaneous thrombosis; 1%–2% of cases

2) **Percutaneous therapeutic embolization**

3) **Surgical ligation**

→ Results from the transcatheter and surgical literature show that both approaches have similar early effectiveness, morbidity, and mortality
Transcatheter Embolization

- Materials: Coils, umbrella devices, detachable balloons, vascular plugs, and covered stents
- Delineating the Distal vessel entry site is important!
  - Wide vessel entry -> higher likelihood of coronary steal and migration into the drainage site of coils used for treatment
  - Narrow distal entry and multiple tiny vascular lakes -> incomplete occlusion of the shunt if a coil is deployed distal to a branch point.

Indications for Embolization

1) Proximal origin of fistula
2) Single narrow drainage site
3) Older patients high perioperative risk profile for open surgery
4) Absence of multiple fistulae
5) Absence of large branch vessels
6) Safe accessibility to the coronary artery supplying the fistula
Surgical Ligation

Types of surgery

- Epicardial identification and mobilization of the fistula, with ligation or division of the fistula.
- An arteriotomy can be performed in the dilated proximal coronary artery, close to the fistula site, to oversew the fistula from within the lumen.
- The fistulous connection(s) can be exposed from within the cardiac chambers or pulmonary artery and closed by direct suture or with autologous pericardial patch.

Indications for Surgical Ligation

1) Large high-flow fistula
2) Multiple complex communications
3) Tortuous and aneurysmal fistulous arteries
4) Associated clinically significant cardiac lesions
Potential role of CCTA for treatment of CAF

Evaluation of the CAF before the Treatment

- Location of the Origin & drainage site
- Course of the fistulous tract
- Number of origin site, drainage site and fistulous tracts
- Vessel size of the fistulous tract
- Complexity; angulation, totousity, associated connection with other structure

Embolization

Surgical ligation

Evaluation of the Complication after the Procedure for CAF

- Postoperative recanalization
- Persistent dilation of the coronary artery and ostium
- Thrombus formation, calcification
- Myocardial ischemia
Fig. 11-1. A 62 year old female with dyspnea before Coil embolization.
Axial CT images (A-B) and volume rendered image (C) and invasive angiography (D-E) show dilated and tortuous cornus branches of RCA (CB_R) and LAD (CB_L), which drain into pulmonary trunk (arrowheads).

Check lists before the procedure !!!
✓ Location of the origin: conus branch from RCA (CB_R), conus branch from LAD (CB_L)
✓ Location of the drainage site; main pulmonary trunk
✓ Course of fistulous track: Pre-pulmonary trunk (anterior to the RVOT)
✓ Number of fistulous track; at least 2
✓ Size of the vessel : CB_R (3mm), CB_L (4mm)
✓ Relation with other structures : no specific findings
Fig. 11-2. A 62 year old female with dyspnea after Coil embolization.
A. Invasive angiography shows no fistulous tract after successful embolization of coronary-pulmonary fistula with coils.
B-C Volume rendered (B) and axial CT images (C) show no connection between coronary artery and pulmonary trunk after successful occlusion of fistula after coil embolization,

Check lists for the post-procedural complication !!!
✓ Complete obliteration (+)
✓ recanalization (-)
✓ persistent dilation of the coronary artery (-)
✓ thrombus formation, calcification (-)
✓ myocardial ischemia (-)
**Summary**

**Advantages of CCTA for evaluating CAF**

- Non-invasive, Short acquisition time
- Excellent high spatial resolution
- Accurate cross-sectional imaging in demonstrating routes of coronary artery
- Multiple planes and three-dimensional views for cardiac and coronary anatomy

**Pre-procedural CAF check list**

- Location of the Origin & drainage site
- Course of the fistulous tract
- Number of origin site, drainage site and fistulous tracts
- Vessel size of the fistulous tract
- Complexity; angulation, totousity, associated connection with other structure

**Post-procedural CAF check list**

- Postoperative recanalization
- Persistent dilation of the coronary artery and ostium
- Thrombus formation, calcification
- Myocardial ischemia
Conclusion

- CAF is rare anomaly that could lead to variable symptoms from asymptomatic to serious ones including chest pain, stroke myocardial infarction depending on the degree of left to right shunt.

- ECG-gated Coronary CT angiography (CCTA) has high temporal and spatial resolution, allows three-dimensional view, which helps radiologists to assess the connections from origin to drainage site of various CAF non-invasively.

- It is also a promising tool for guiding the plan for noninvasive as well as invasive treatment and assessing the post-procedural complication.

- Radiologists should be fully aware of the critical role of the CCTA for evaluating CAF to help clinicians make clinical decision based on the reliable radiologic report.


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