

DIU cardiologie interventionnelle
année universitaire 2023-2024

Anomalies Coronaires congénitales (ANOCOR) classification

Pierre Aubry

Hôpital Bichat, Paris 75018

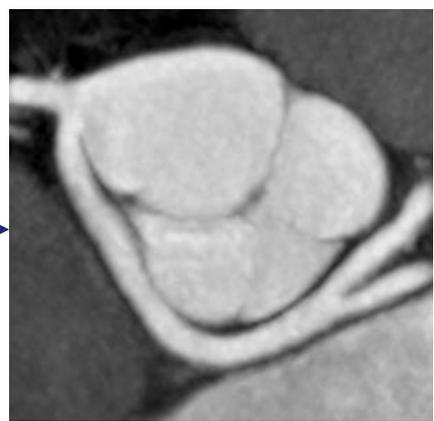
Centre Hospitalier, Gonesse 95500



ANOMALIES CORONAIRES CONGENITALES

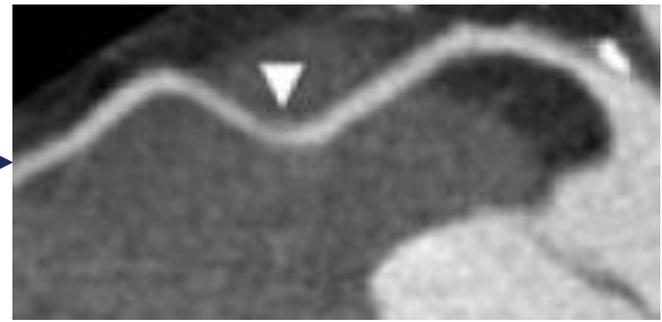


Connexion proximale



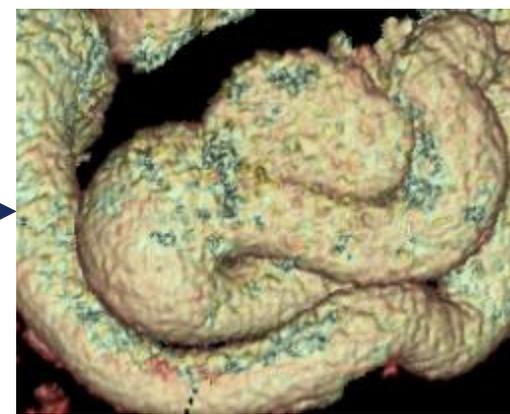
ANOCOR

Trajet



PM

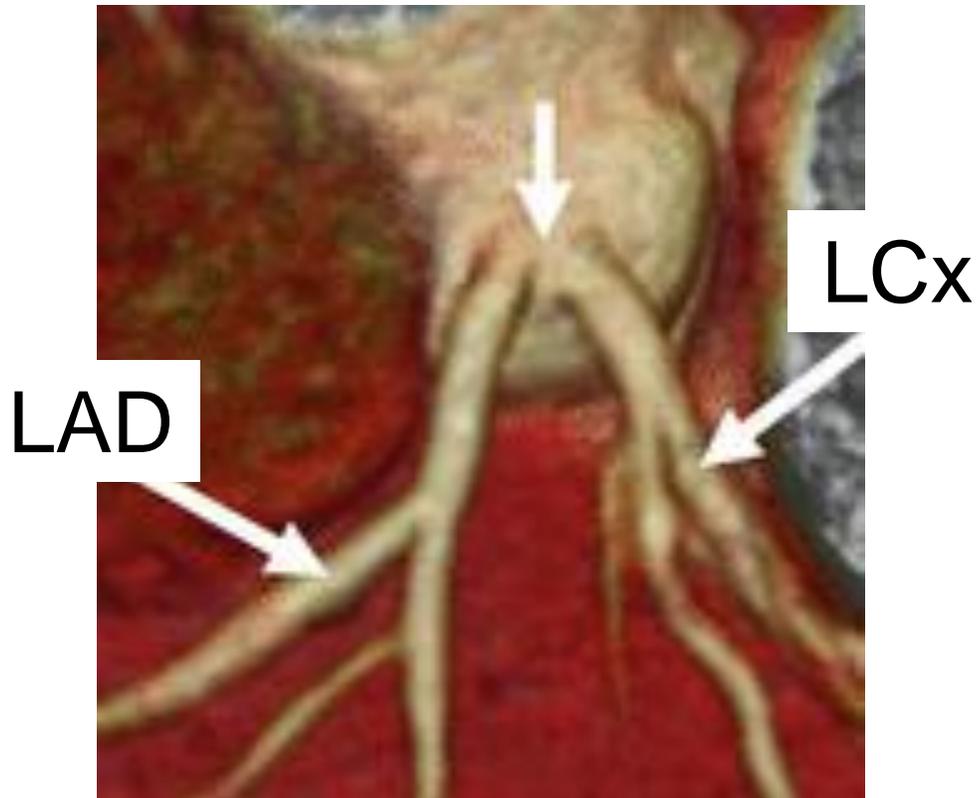
Connexion distale



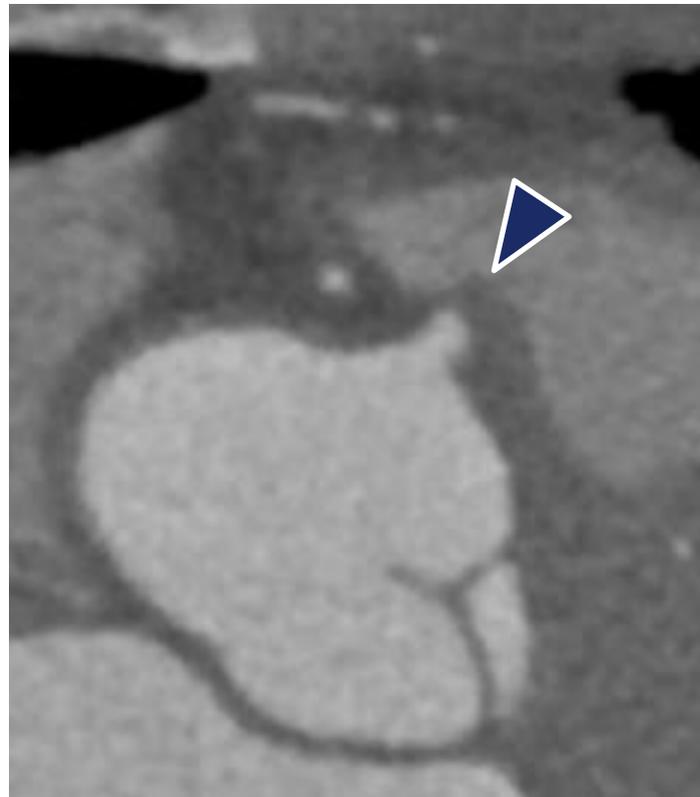
Fistule

- **Variantes anatomiques**
- Anomalies sur le trajet
- Connexions anormales distales
- Connexions anormales proximales

Variantes anatomiques coronaires



Absence de tronc commun

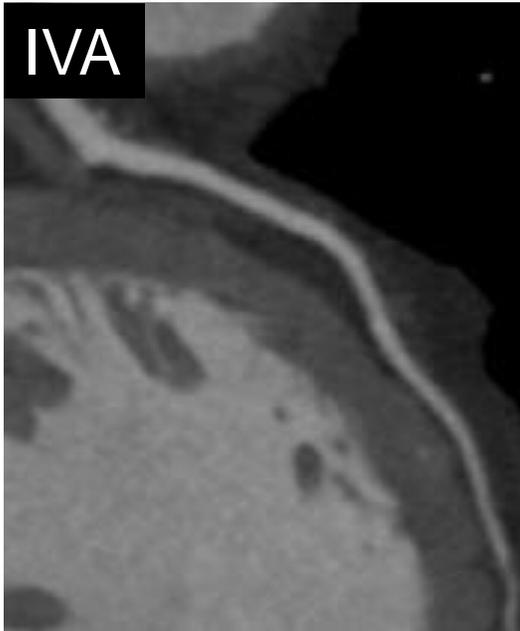


Rotation aortique anti-horaire (anatomique)



- Variantes anatomiques
- **Anomalies sur le trajet**
- Connexions anormales distales
- Connexions anormales proximales

Trajet coronaire intramyocardique



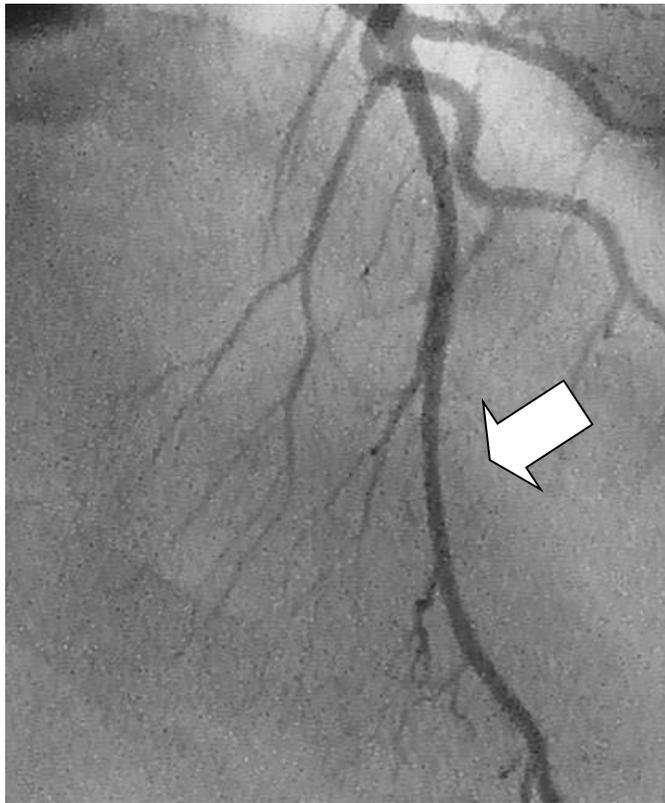
Trajet normal



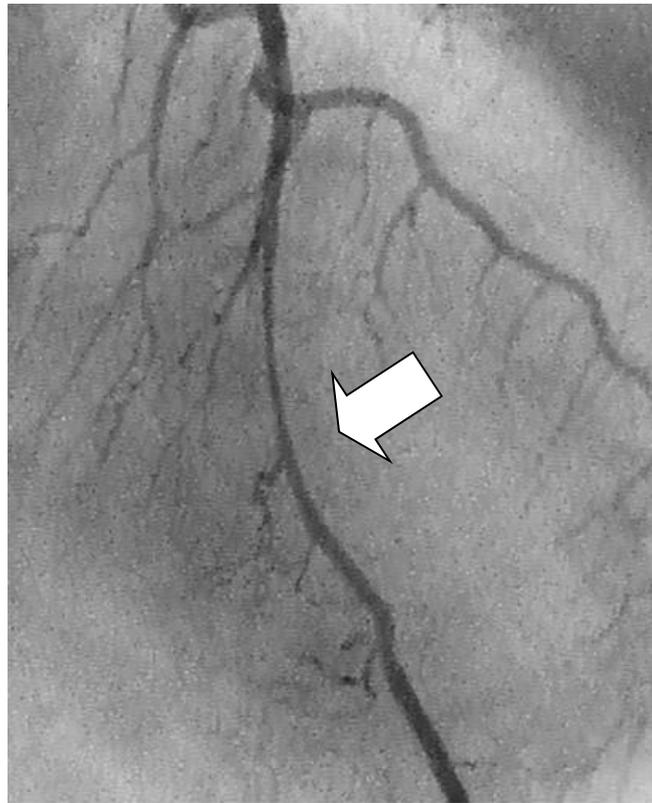
Pont musculaire

- coronarographie 5%
- scanner coronaire 20%

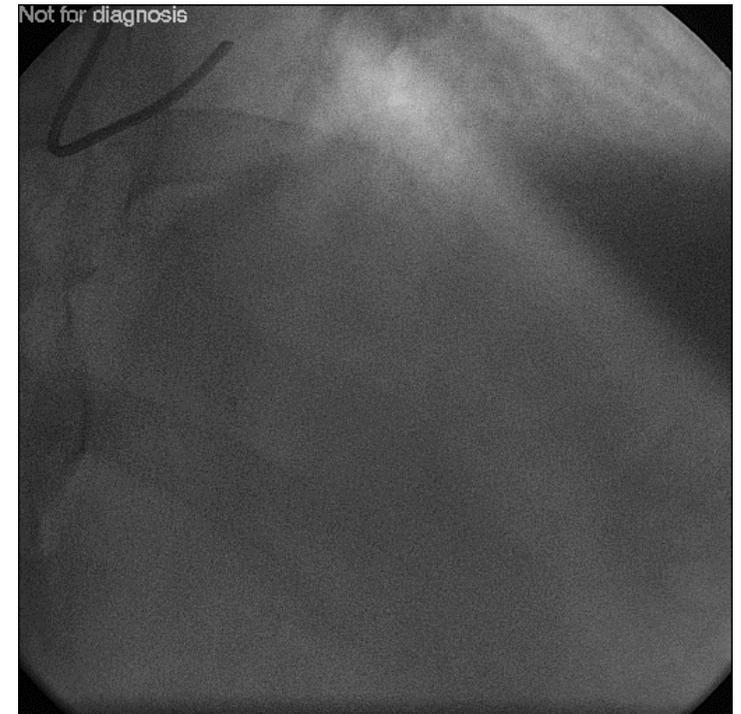
Trajet coronaire intramyocardique



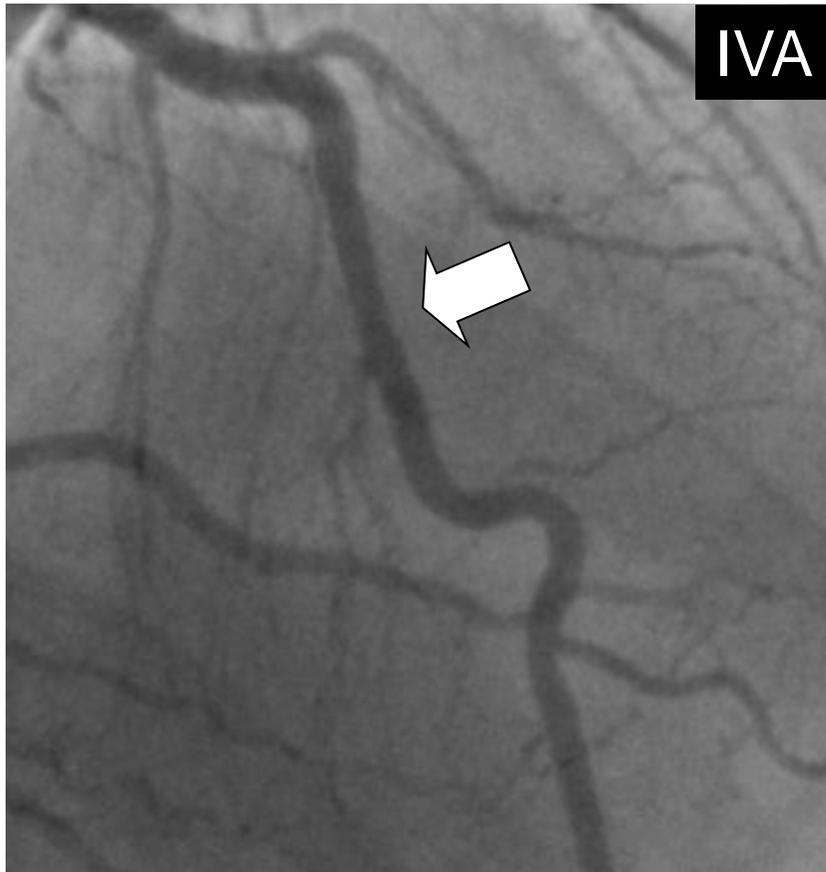
diastole



systole



Trajet coronaire intramyocardique

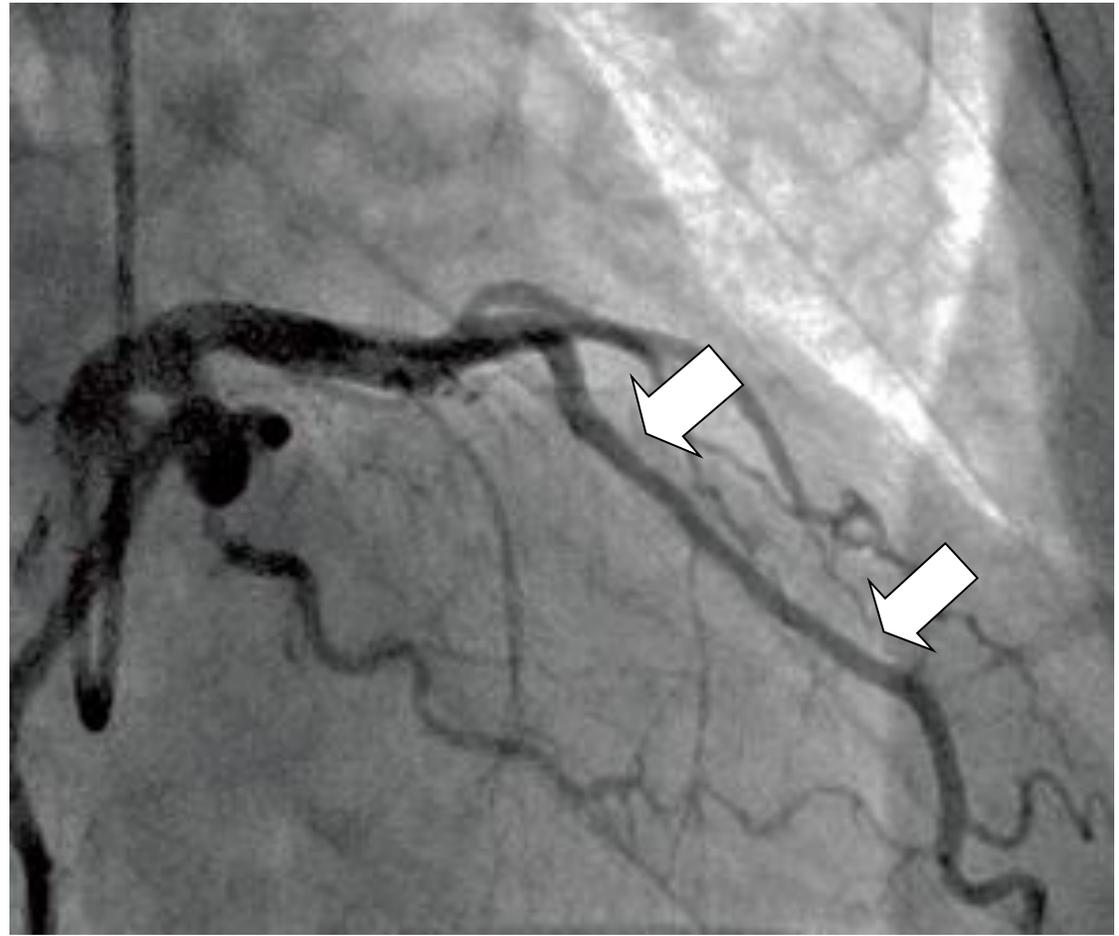
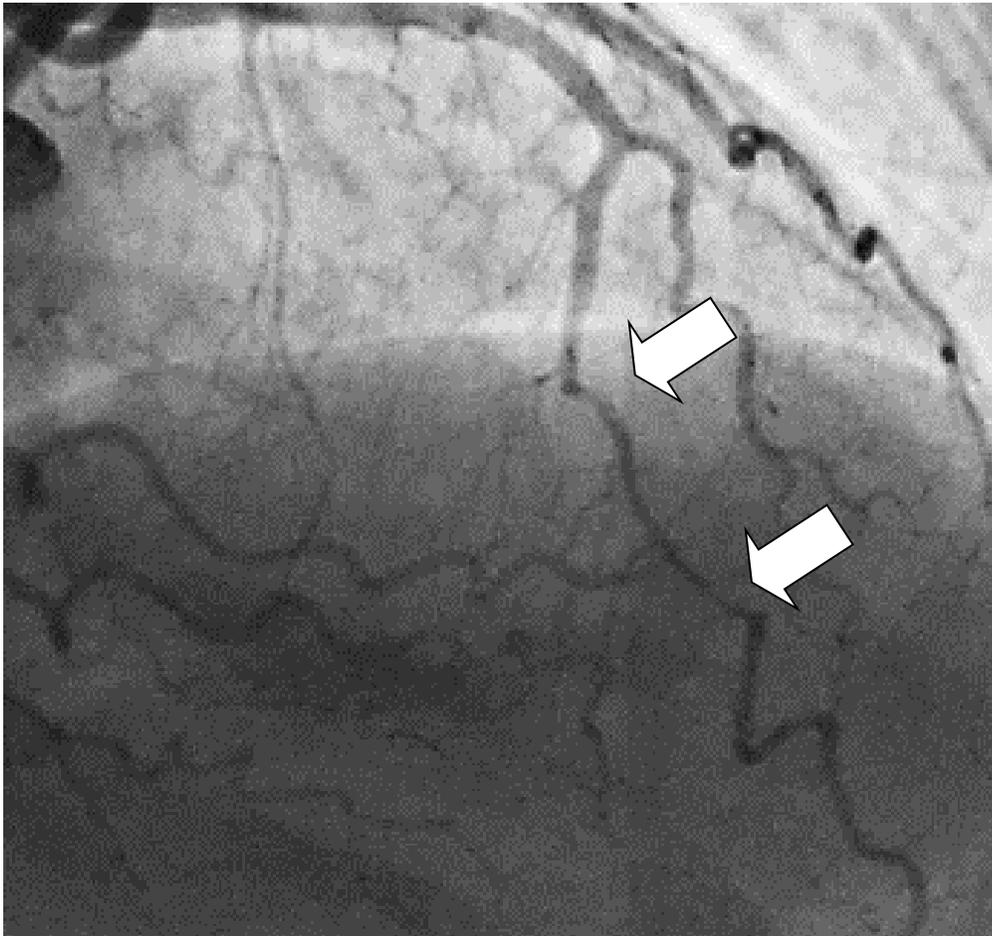


Systole avant nitré



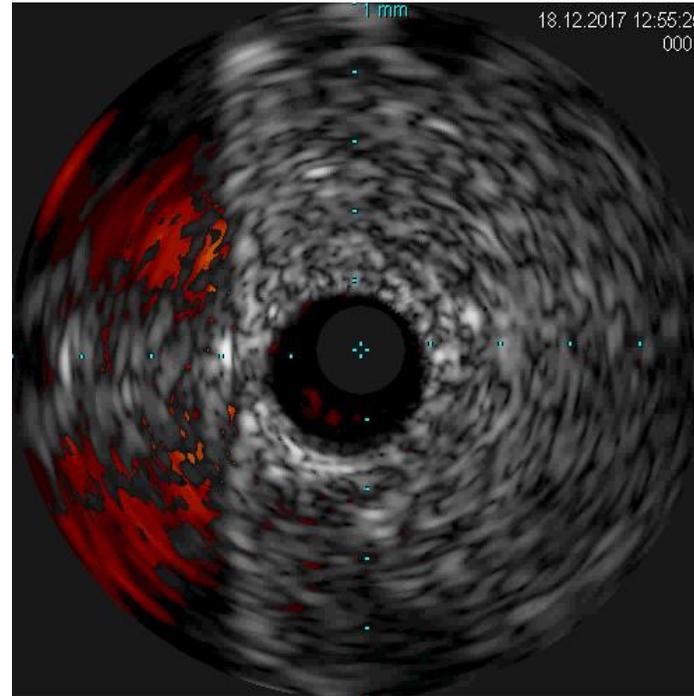
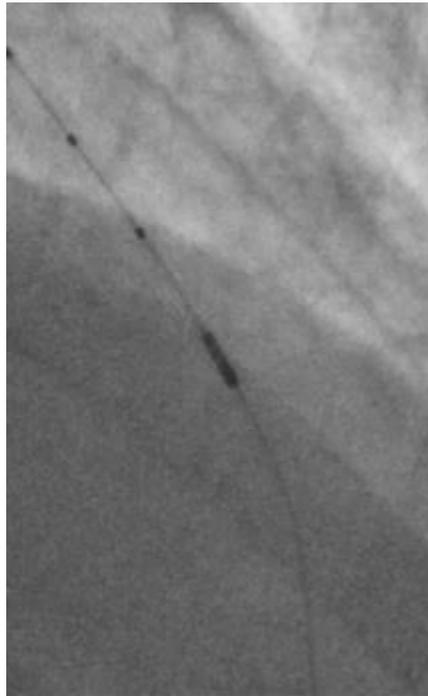
Systole après nitré

Trajet coronaire intramyocardique ?

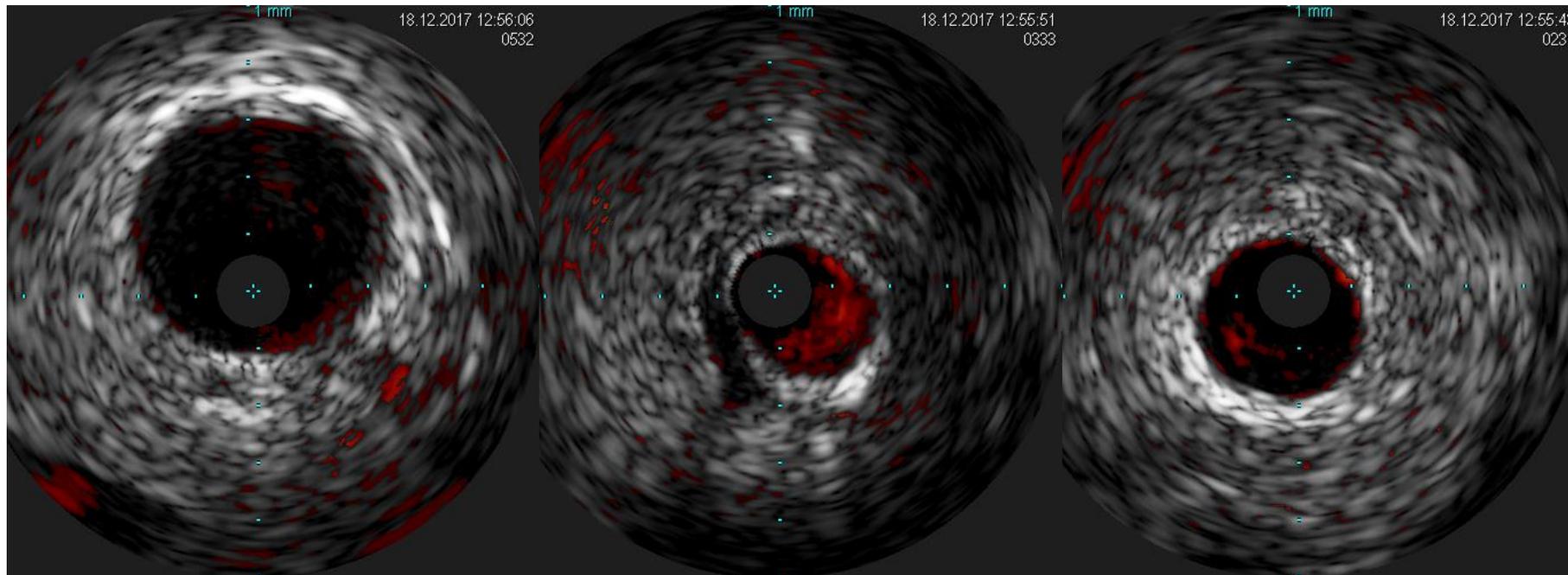


Trajet coronaire intramyocardique ?

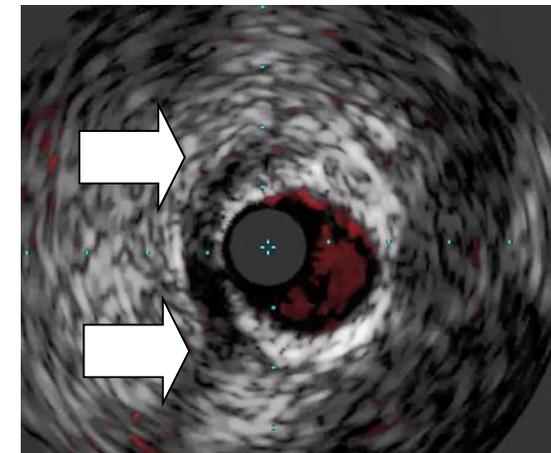
Imagerie endocoronaire



Trajet coronaire intramyocardique



Half-moon sign



IVA I

IVA II (pont musculaire)

IVA III

Trajet coronaire intramyocardique

situation non rare

douleur thoracique suspecte
(événement cardiaque fréquent)
+
trajet coronaire intramyocardique
(particularité anatomique non rare)

lien de causalité ?

trajet intramyocardique incident

trajet intramyocardique pathologique

Trajets coronaires intramyocardiques

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THE PRESENT AND FUTURE**JACC STATE-OF-THE-ART REVIEW**

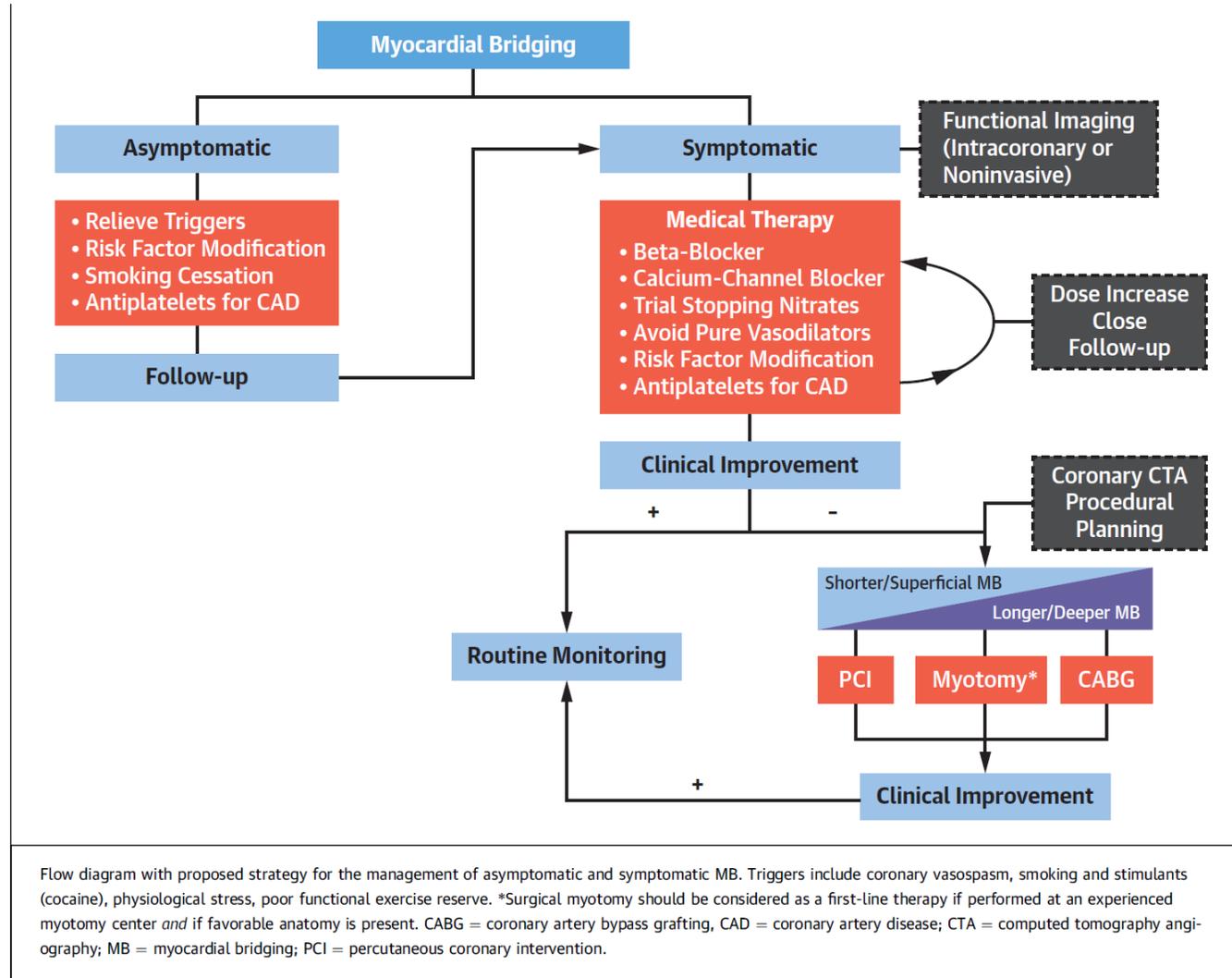
Myocardial Bridging: Diagnosis, Functional Assessment, and Management

JACC State-of-the-Art Review

David Sternheim, MD,^{a,*} David A. Power, MD,^{a,*} Rajeev Samtani, MD,^a Anapoorna Kini, MD,^a
Valentin Fuster, MD, PhD,^{a,b} Samin Sharma, MD^a



Trajet coronaire intramyocardique



Trajets coronaires intramyocardiques

Description dans un compte-rendu

- trajet intramyocardique : certain/vraisemblable
- compression systolique : oui (degré) / non

- Variantes anatomiques
- Anomalies sur le trajet
- **Connexions anormales distales**
- Connexions anormales proximales

Connexions anormales distales ou fistules coronaires

Prévalence angiographique $\approx 0.1\%$

Classification

- Vaisseau donneur
- Origine sur vaisseau donneur
- Site de connexion distale
- Nombre
- Taille de la fistule
- Retentissement

Fistules coronaires - sites de connexion distale

ANOMALIES DE
TERMINAISON

=

FISTULES
CORONAIRES

CORONARO -
CAMÉRALES

DRAINAGE : OD VD OG VG
CONGÉNITALES +++

CORONARO -
VEINEUSES

DRAINAGE : VCS VCI SC
CONGÉNITALES +++

CORONARO -
PULMONAIRES

DRAINAGE : TRONC PULMONAIRE
CONGÉNITALES SOUVENT

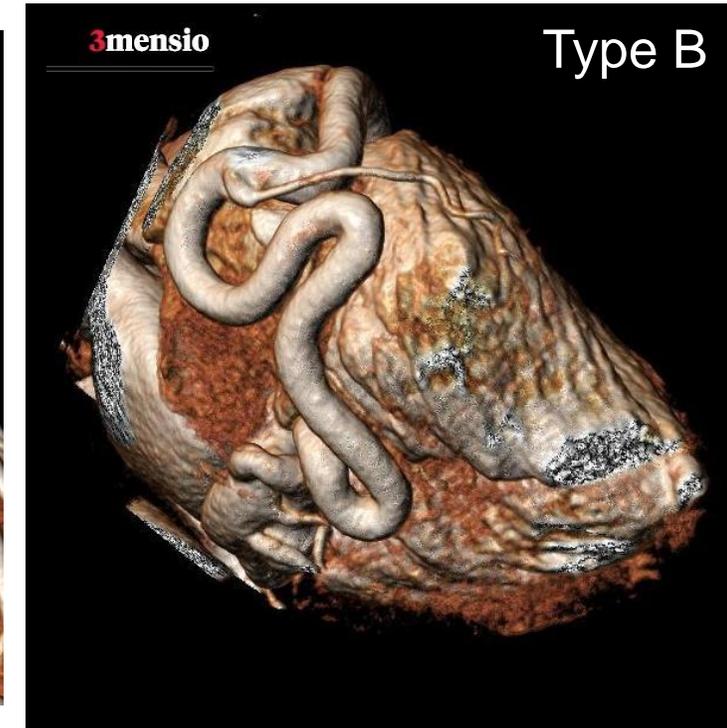
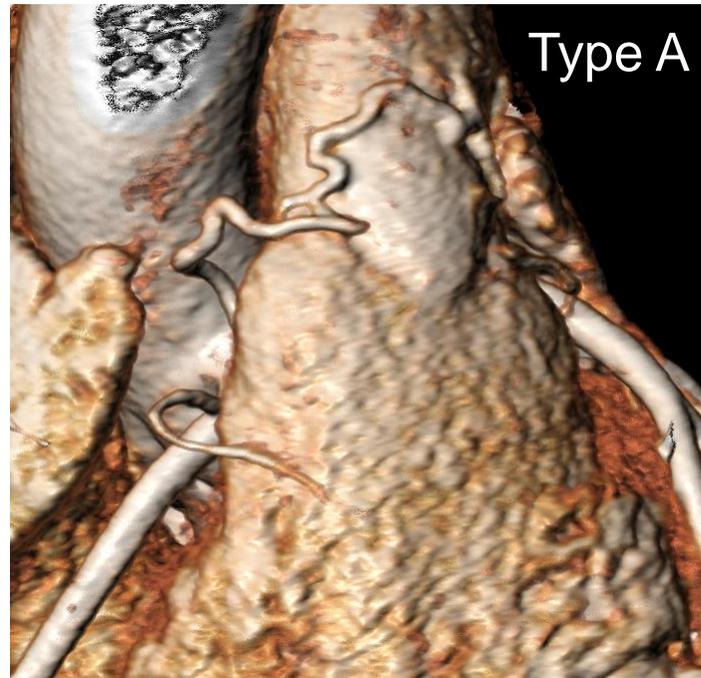
CORONARO -
BRONCHIQUES

ANASTOMOSES AVEC
RESEAU ARTERIEL BRONCHIQUE
ACQUISES SOUVENT

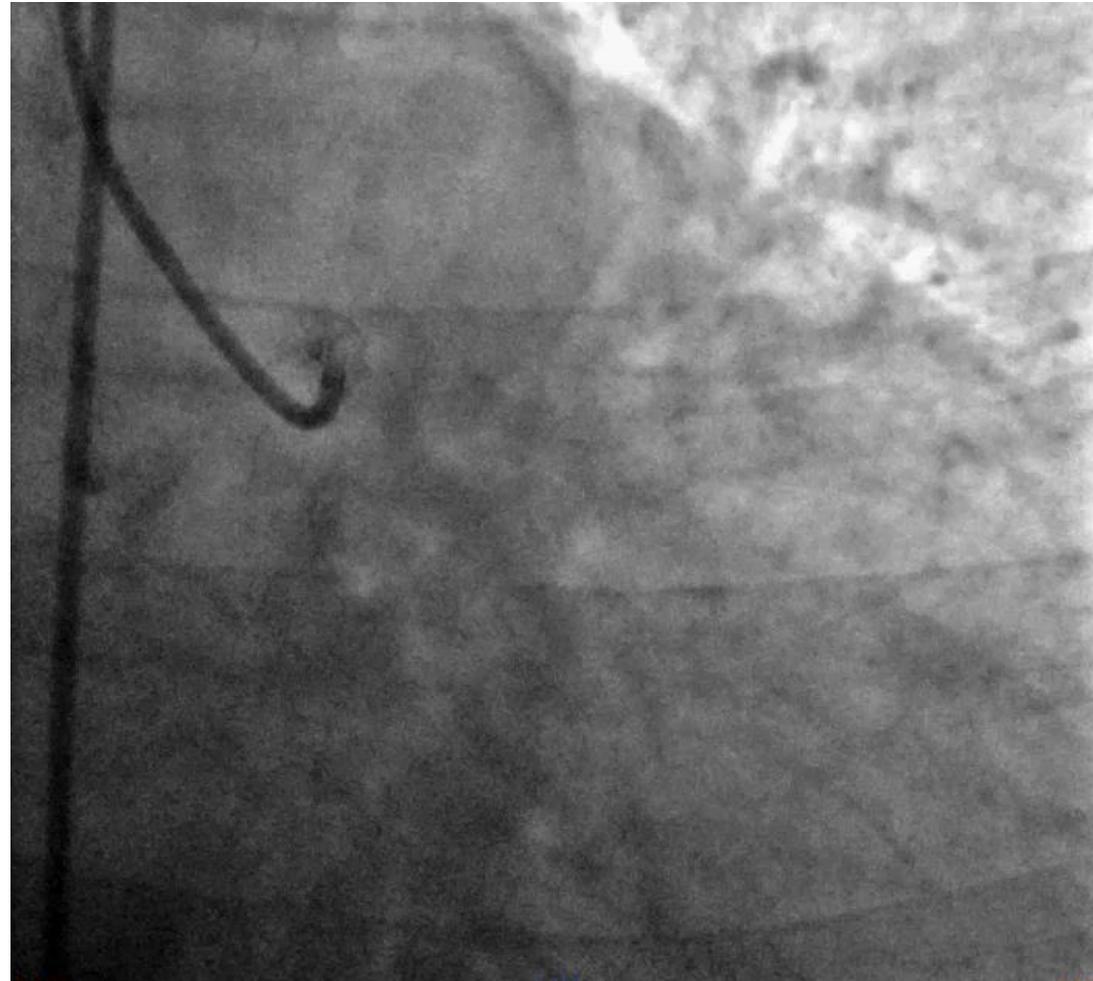
Fistules coronaires

Classification

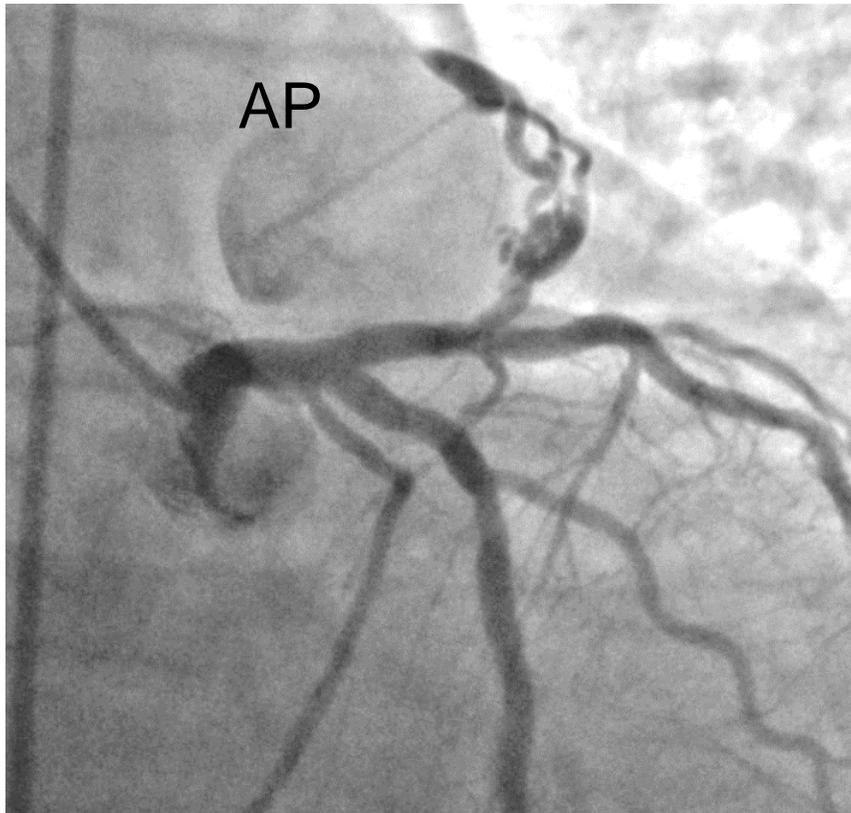
- Vaisseau coronaire donneur
- Origine sur vaisseau donneur
 - type A : tiers proximal
 - type B : tiers moyen ou distal



Fistule IVA-AP



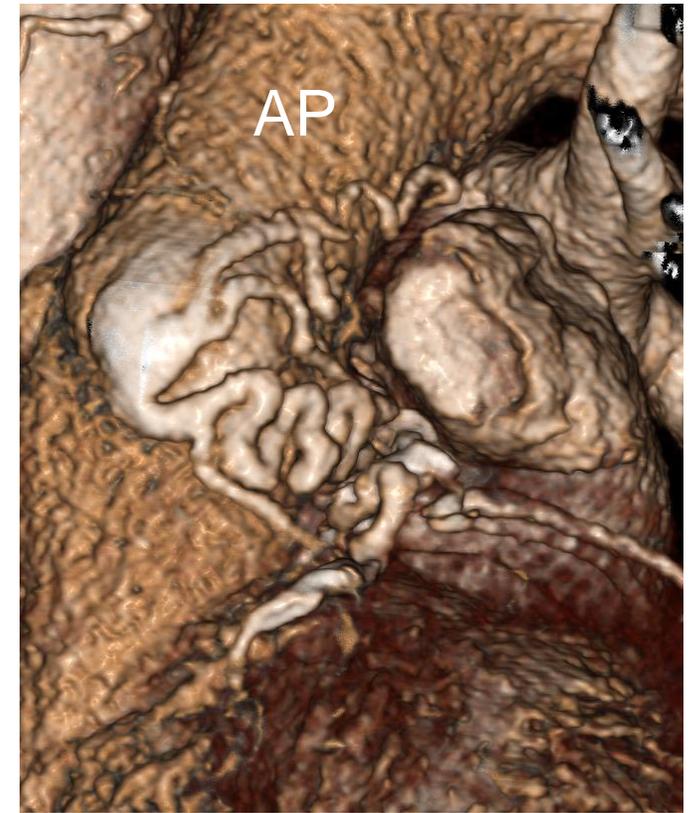
Fistules coronaropulmonaires

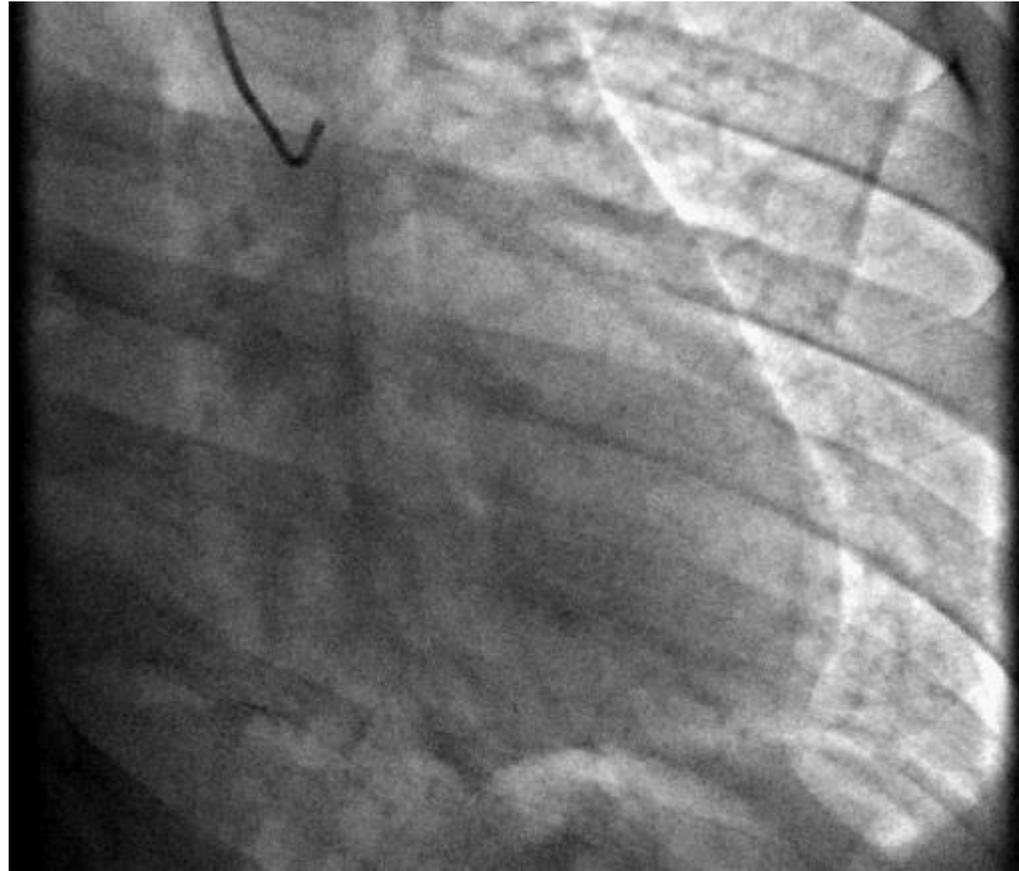


Fistule IVA-AP



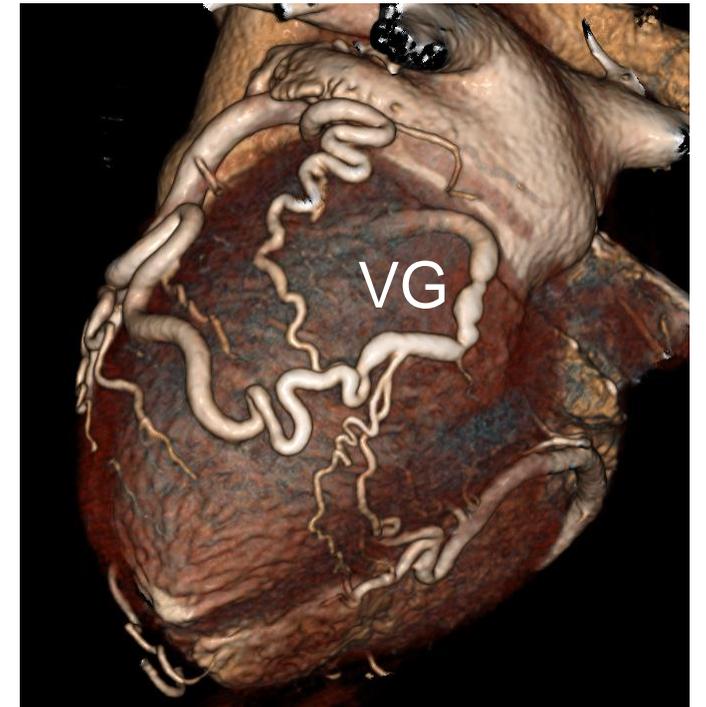
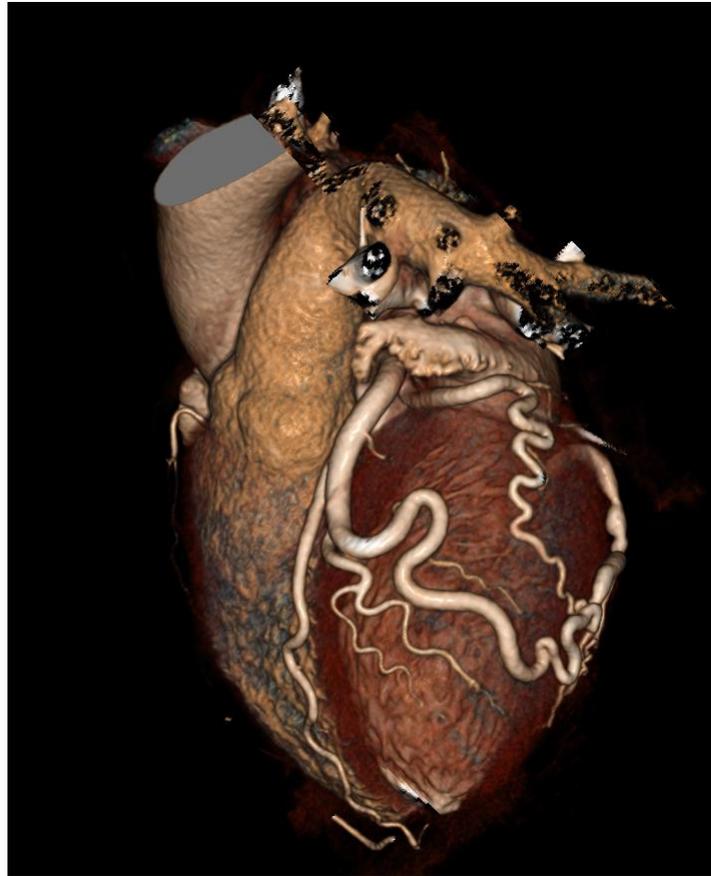
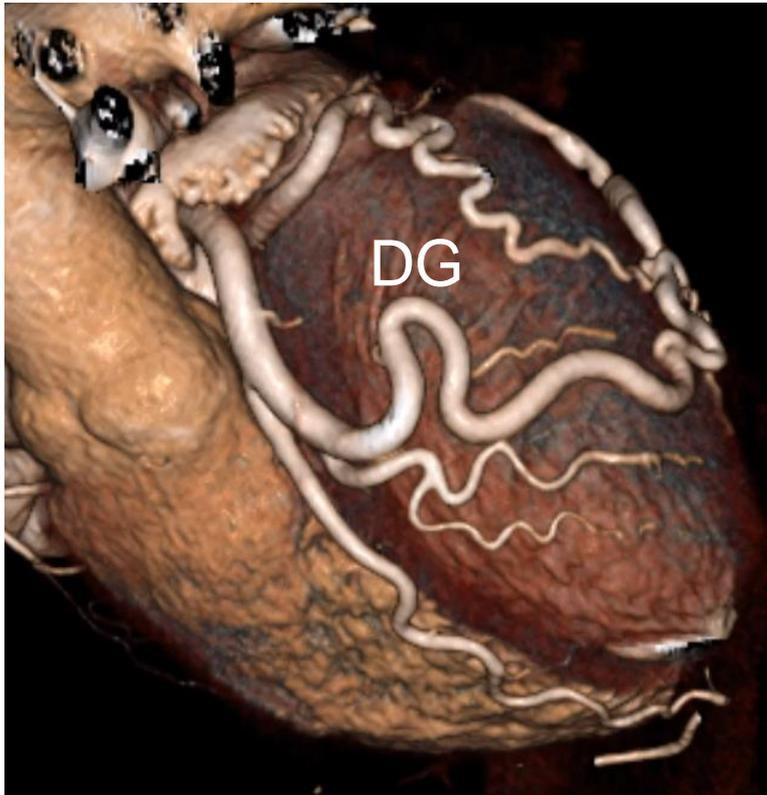
Fistules IVA-AP





Fistule CX-sinus coronaire (type B)

Fistules coronarogastrocardiaques gauches (type B)



Fistules multiples

Fistules coronaires

JACC: CARDIOVASCULAR INTERVENTIONS
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VOL. 14, NO. 13, 2021

STATE-OF-THE-ART REVIEW

CORONARY

Coronary Artery Fistulas

Indications, Techniques, Outcomes, and Complications of Transcatheter Fistula Closure



Mohammed Al-Hijji, MD,^{a,b} Abdallah El Sabbagh, MD,^c Stephanie El Hajj, MD,^a Mohamad AlKhouli, MD,^a
Bassim El Sabawi, MD,^a Allison Cabalka, MD,^a William R. Miranda, MD,^a David R. Holmes, MD,^a
Charanjit S. Rihal, MD^a

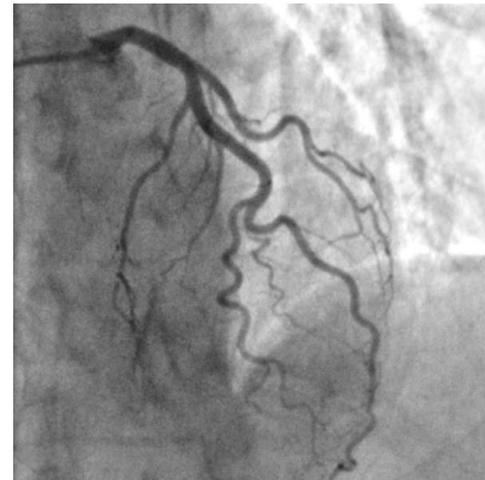
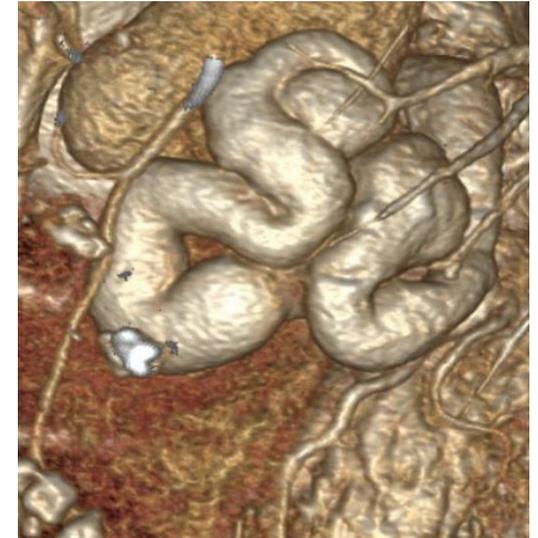
Classification

- Taille du vaisseau donneur (type A)
 - fistule large :
>2 fois taille vaisseau donneur en aval connexion
 - fistule moyenne :
1-2 fois taille vaisseau donneur en aval connexion
 - fistule petite :
<1 fois taille vaisseau donneur en aval connexion

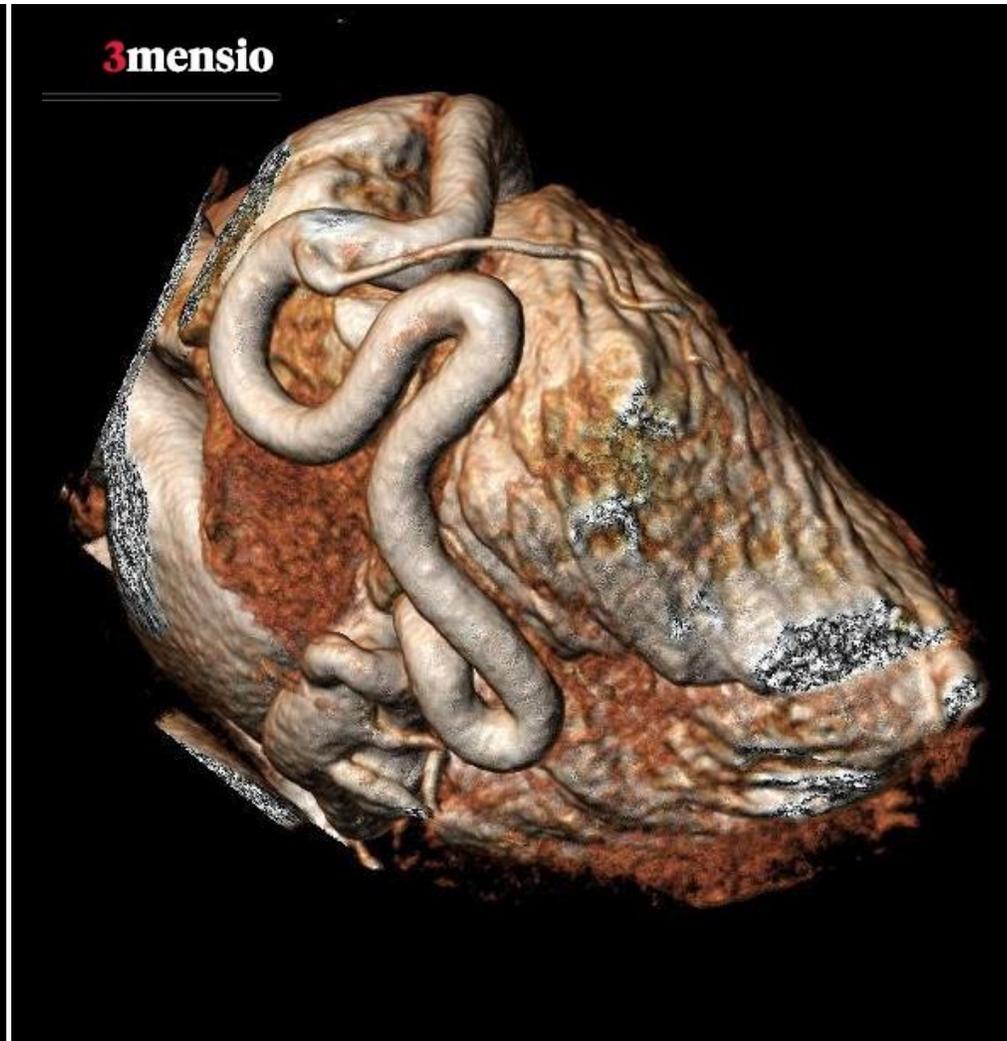
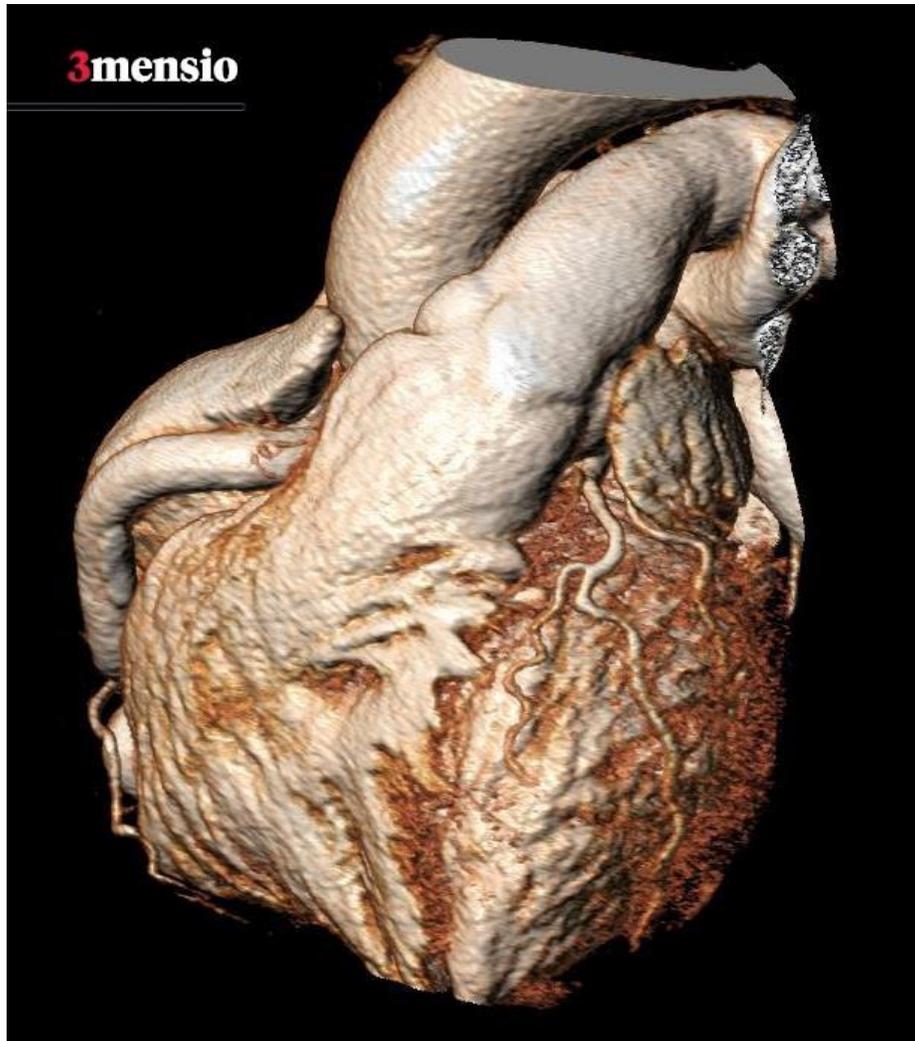
Reddy G et al. Circ Cardiovasc Int. 2015.

- Taille du vaisseau donneur (type B)
 - comparaison à artère controlatérale
 - mêmes critères que précédemment

Al-Hijji M et al. JACC cv int. 2021.



Fistule coronaire droite - sinus coronaire (type B)



Fistules coronaires

- Retentissement physiologique : pas toujours présent
 - shunt gauche-droit significatif
 - shunt gauche-gauche significatif
 - vol coronaire
- Retentissement anatomique : fréquent avec le temps
 - déformation anévrysmale
 - allongement vasculaire avec tortuosités
 - calcifications

- Insuffisance cardiaque droite
- VG dilaté
- Angor



4.17 Coronary anomalies

2020 ESC Guidelines for the management of
adult congenital heart disease

4.17.1.3 Coronary artery fistulae

A coronary artery fistula, whether congenital or acquired, is an abnormal connection between a coronary artery and cardiac chamber or vessel. Small fistulae have a good prognosis without treatment. Medium or large fistulae are associated with long-term complications (angina, myocardial infarction, arrhythmias, heart failure, and endocarditis). The presence of symptoms, complications, and a significant shunt are the main indications for percutaneous or surgical closure.

Fistules coronaires

JACC: CARDIOVASCULAR INTERVENTIONS

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STATE-OF-THE-ART REVIEW

CORONARY

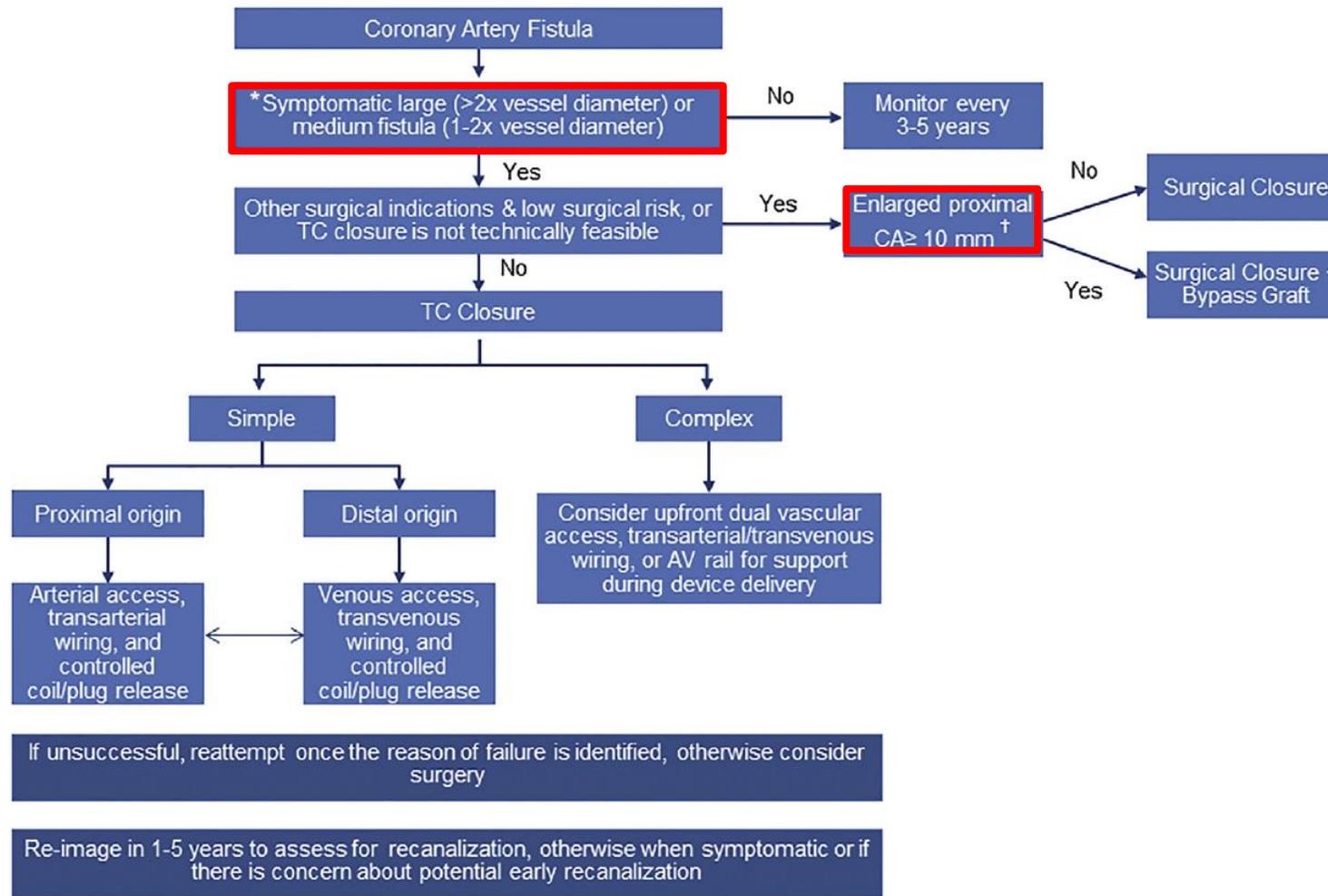
Coronary Artery Fistulas

Indications, Techniques, Outcomes, and Complications of Transcatheter Fistula Closure



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Bassim El Sabawi, MD,^a Allison Cabalka, MD,^a William R. Miranda, MD,^a David R. Holmes, MD,^a
Charanjit S. Rihal, MD^a

FIGURE 1 Algorithm of Coronary Artery Fistula Evaluation and Management



*Symptomatic fistulas are coronary artery (CA) fistulas that potentially lead to myocardial ischemia, vessel aneurysm and rupture, endocarditis, unexplained cardiac chamber enlargement/dysfunction, or arrhythmia. †An enlarged proximal CA with diameter ≥ 10 mm has a tendency to thrombose after fistula closure, resulting in myocardial infarction. A simple fistula has a single-vessel origin, simply defined pathways, and a clearly defined termination. A complex fistula is a large fistula with multiple origins and plexiform formation. AV = arteriovenous; TC = transcatheter.



- Procédure complexe
- Risque IDM (10%)

- Variantes anatomiques
- Anomalies sur le trajet
- Connexions anormales distales
- **Connexions anormales proximales**

- Embryologie et anatomie
- Classification anatomique
- Prévalence
- Imagerie
- Ischémie myocardique
- Mort subite

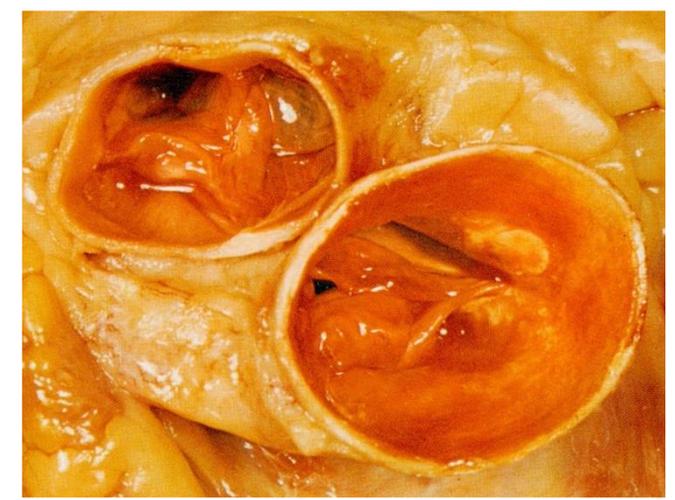
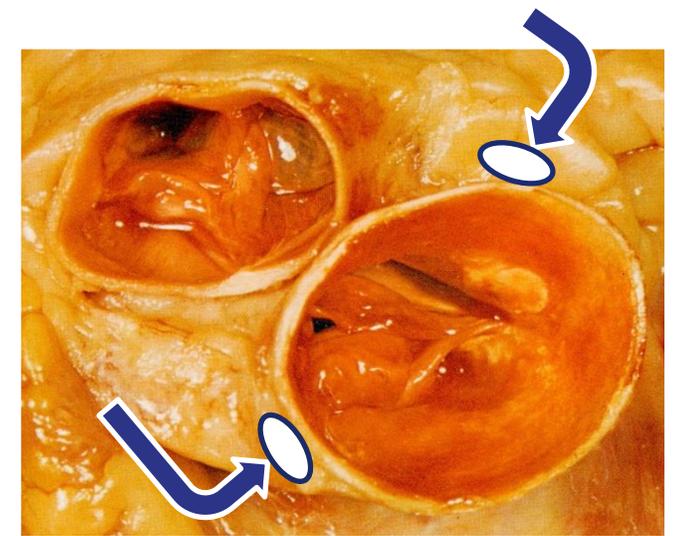
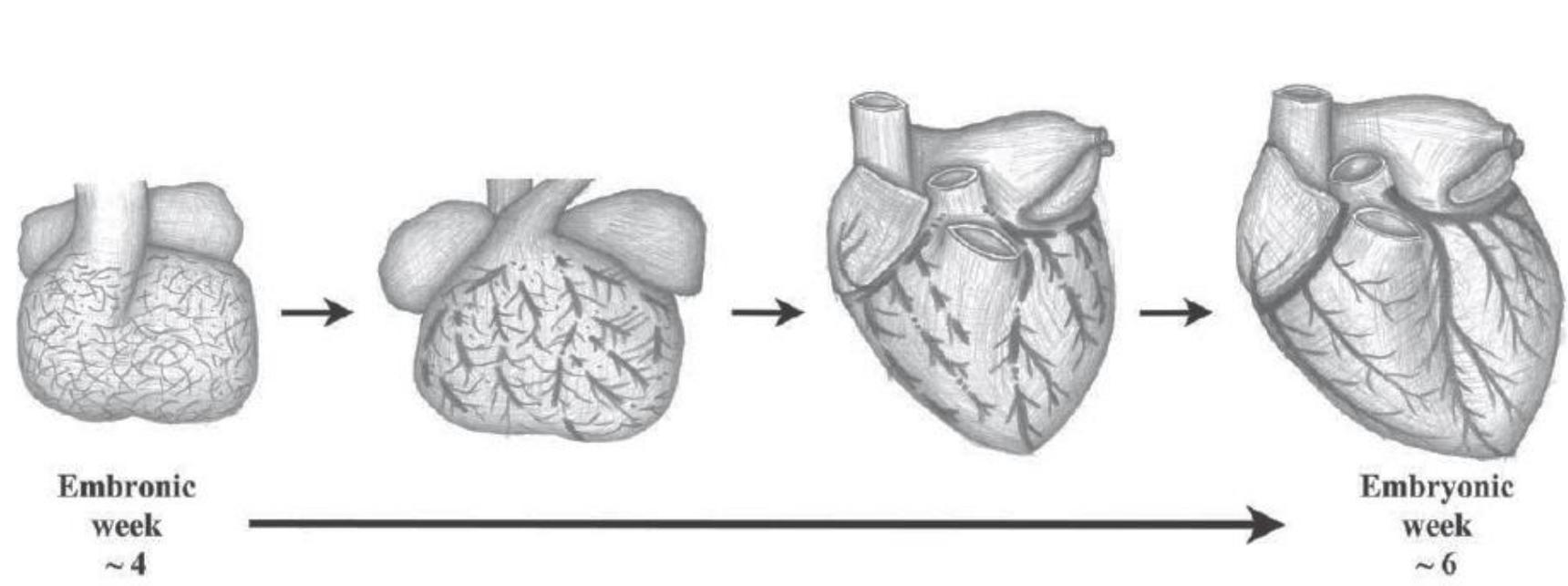
Origine anormale

Développement à partir de l'aorte

Connexion anormale

Développement vers l'aorte

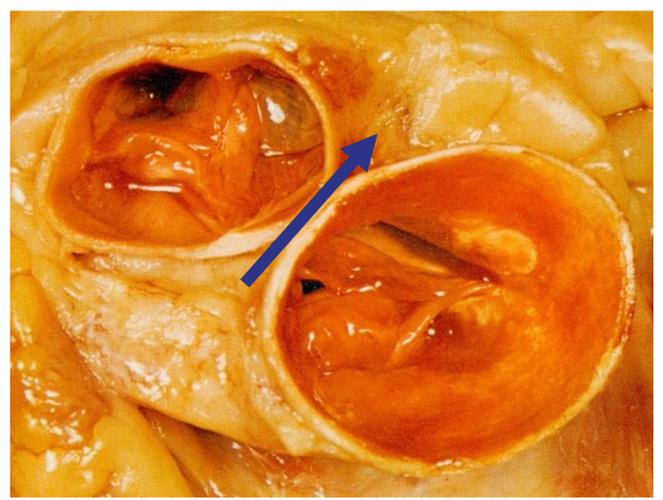
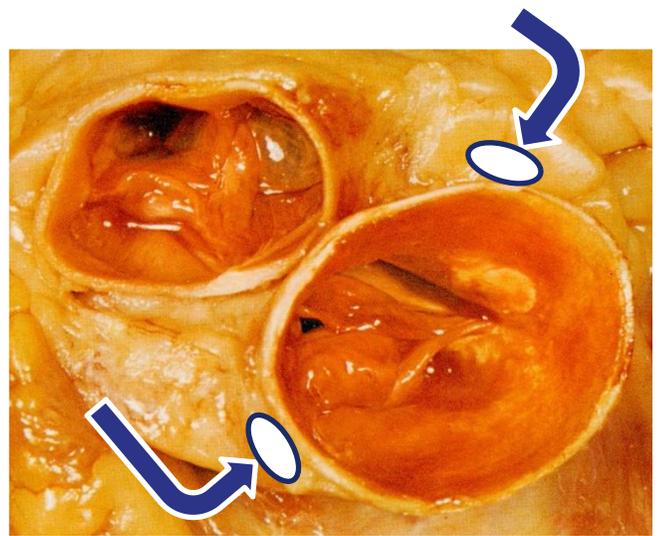
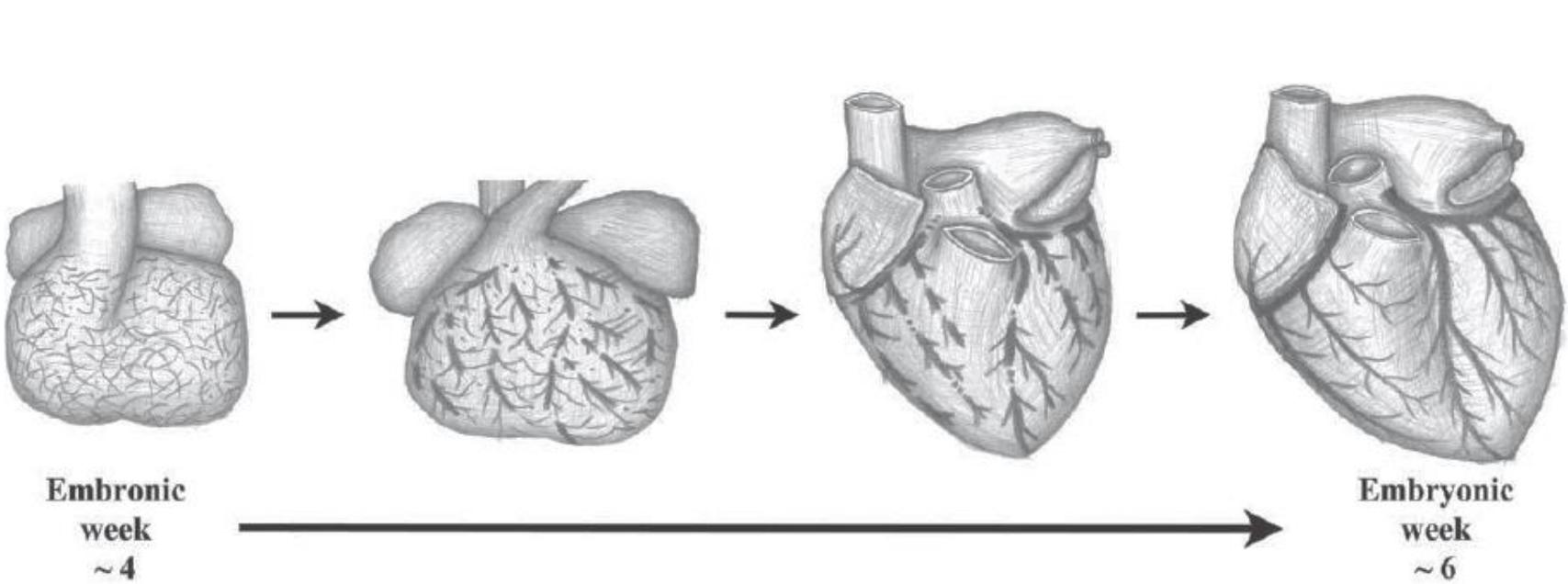
Embryologie et anatomie



Development of coronary vessels during embryogenesis.

Lluri G. *Clin Cardiol* 2014
Bogers AJ. *Anat Embryol* 1989

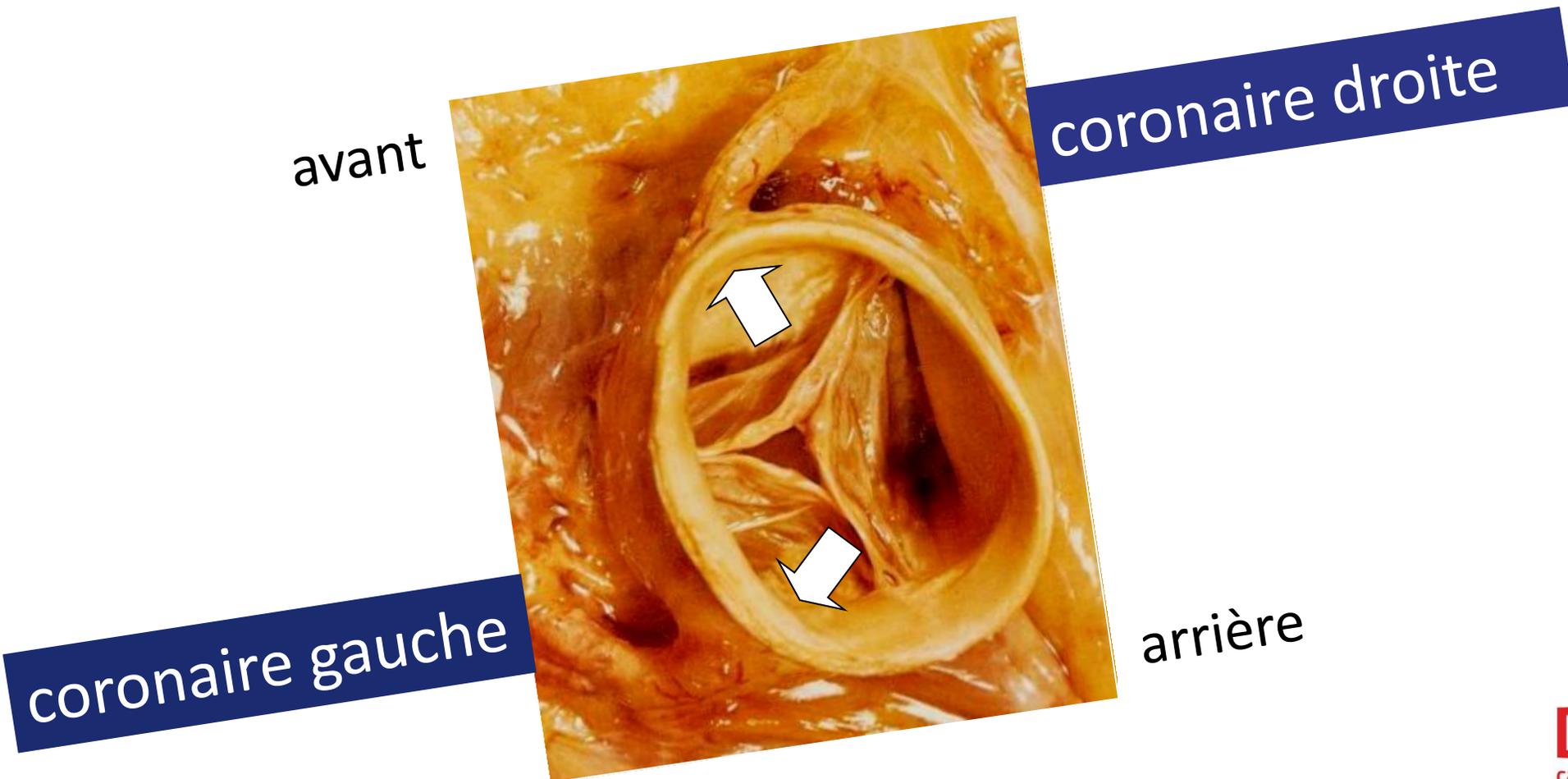
Embryologie et anatomie



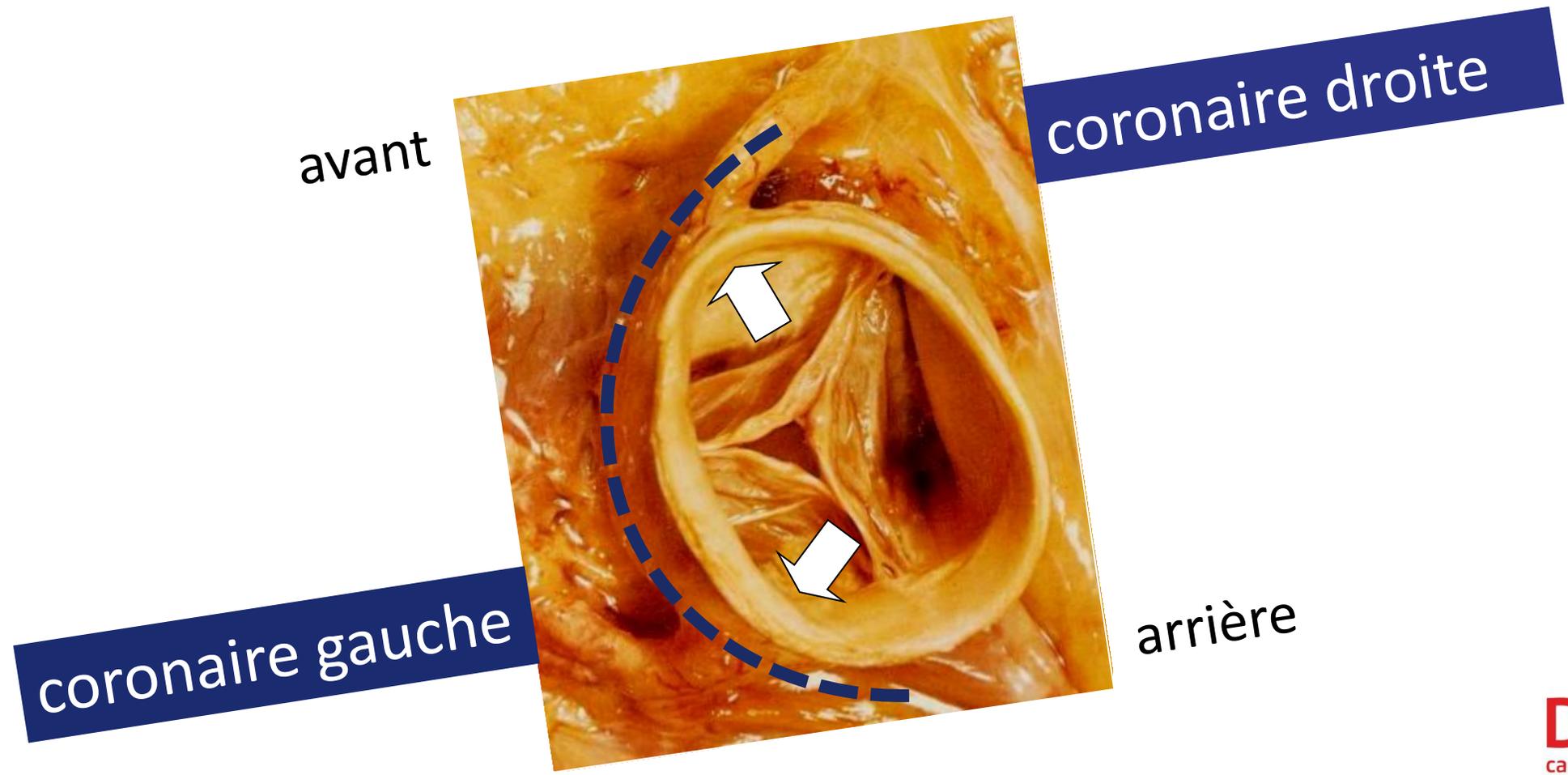
Development of coronary vessels during embryogenesis.

Lluri G. *Clin Cardiol* 2014
Bogers AJ. *Anat Embryol* 1989

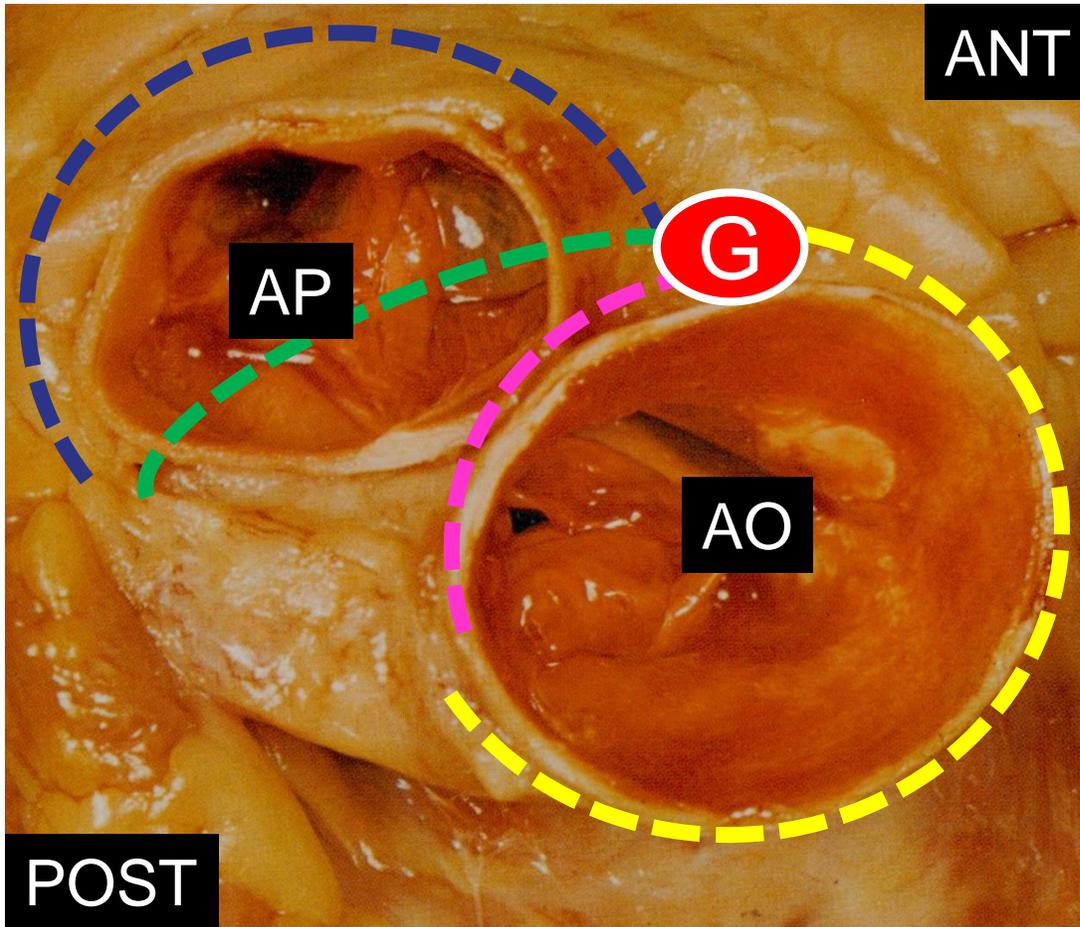
Connexions coronaires normales



Connexions coronaires anormales (---)

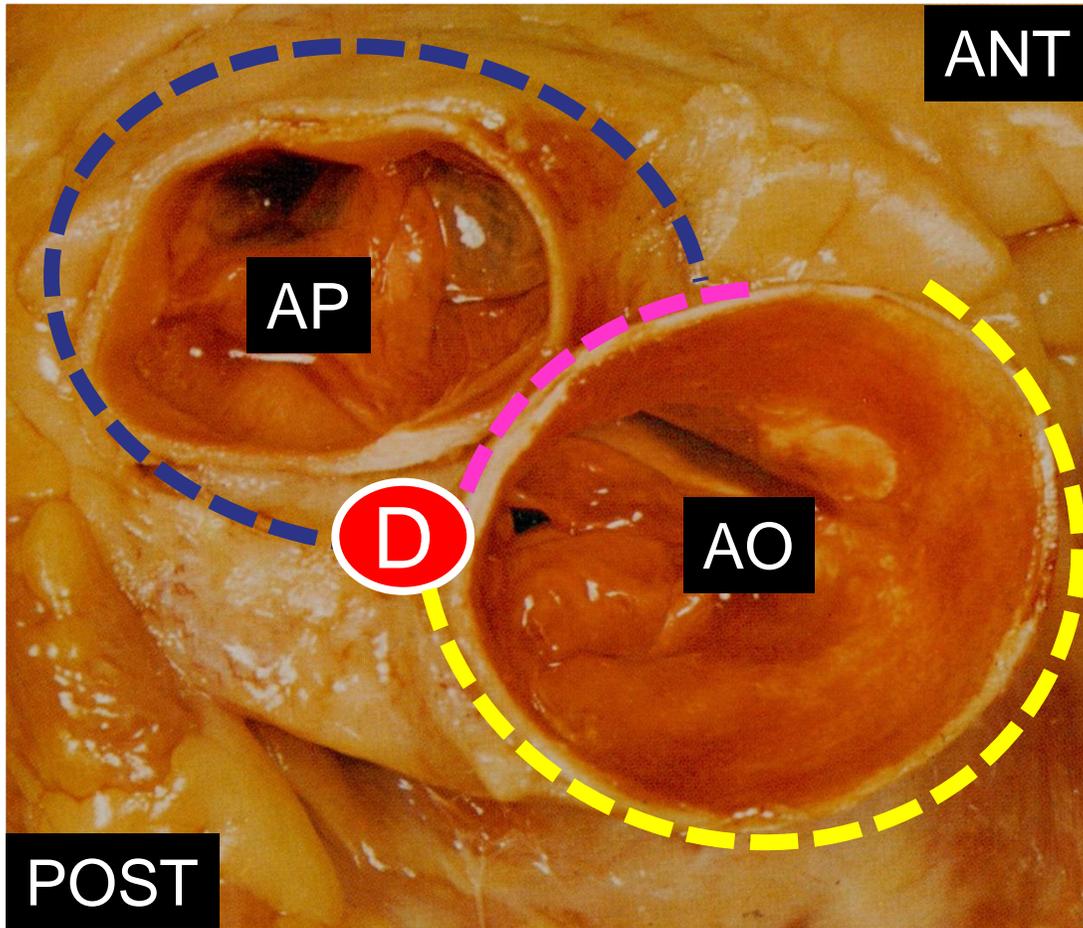


Trajets ectopiques pour la coronaire gauche



- prépulmonaire
- rétropulmonaire (intraseptal)
- interartériel (préaortique)
- rétroaortique

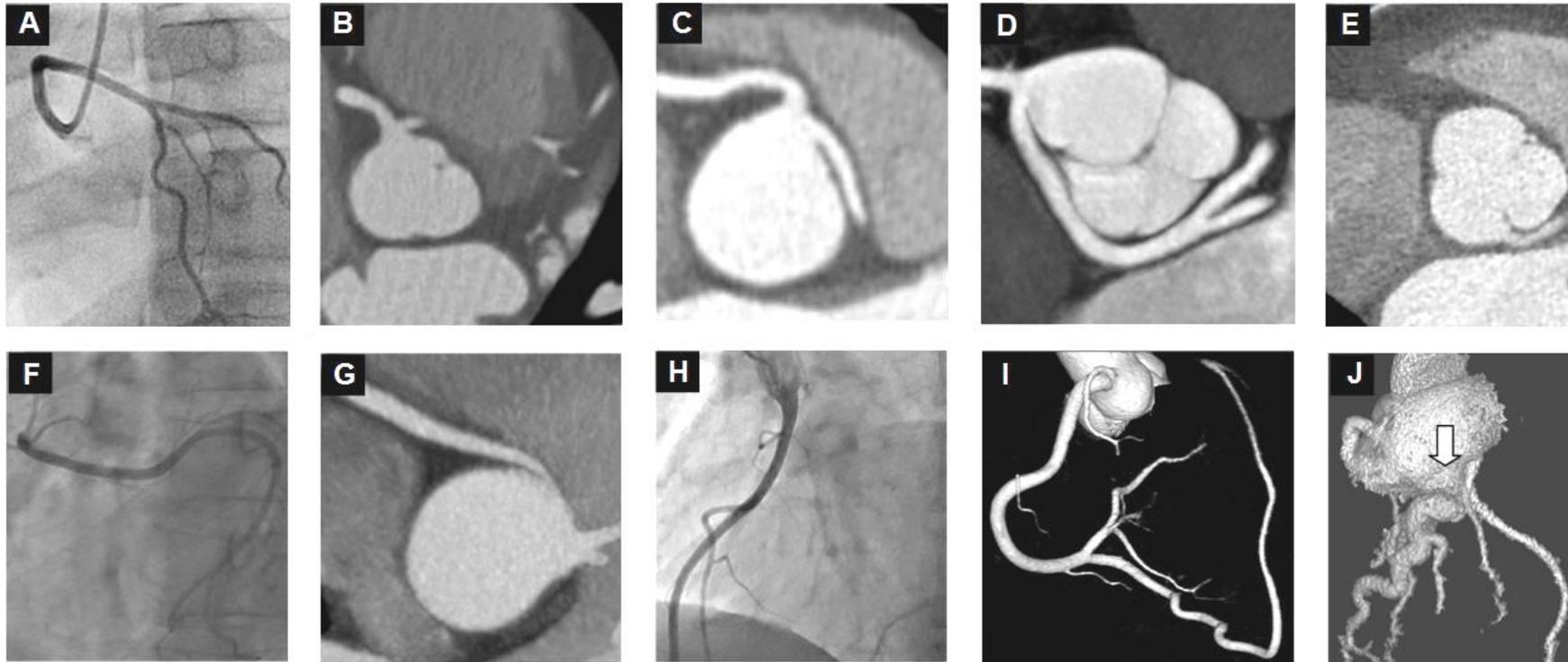
Trajets ectopiques pour la coronaire droite



- — — prépulmonaire
- — — interartériel (préaortique)
- — — rétroaortique

- Embryologie et anatomie
- **Classification anatomique**
- Prévalence
- Imagerie
- Ischémie myocardique
- Mort subite

Nombreuses formes anatomiques



Classification

- Type d'artère

Tronc
IVA
Circonflexe
Droite
Septale

- Trajet

Prépulmonaire
Rétropulmonaire
Interartériel
Rétroaortique
Normal

- Site de connexion

Artère controlatérale
Sinus controlatéral
Sinus non coronaire
Sinus approprié
Aorte thoracique
Artère pulmonaire

- Risques

Mort subite
Arrêt cardiaque
Arythmies V
Ischémie
Absents

Classification

- Liens de causalité :
 - entre anomalie coronaire et symptomatologie
 - entre anomalie coronaire et ischémie myocardique
 - entre anomalie coronaire et arythmie ventriculaire
 - entre anomalie coronaire et arrêt cardiaque

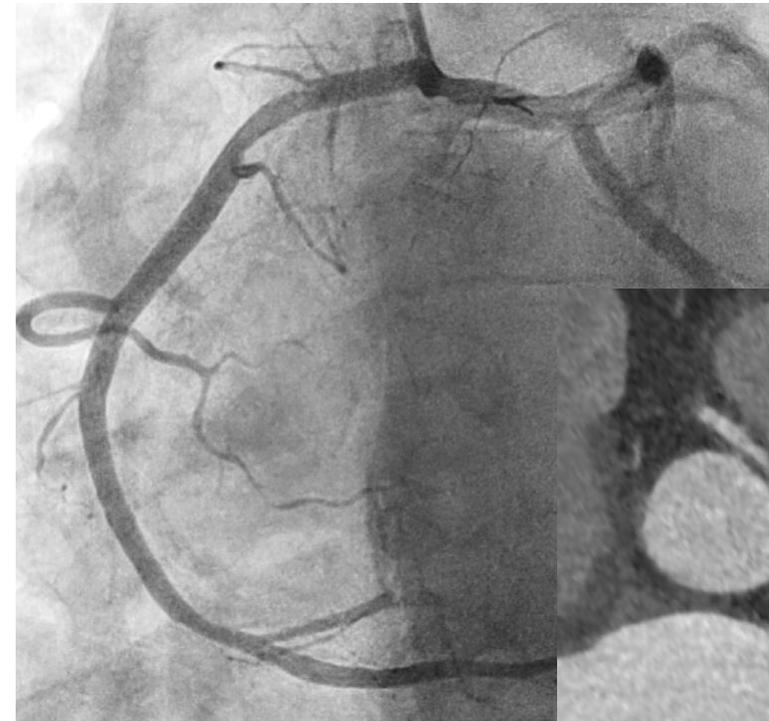
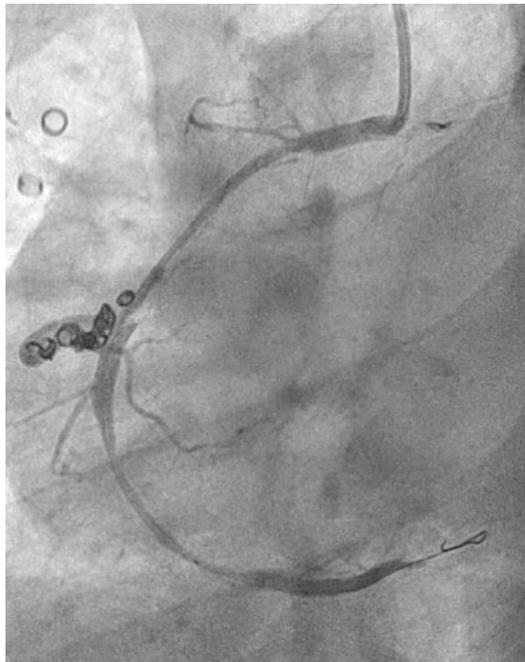
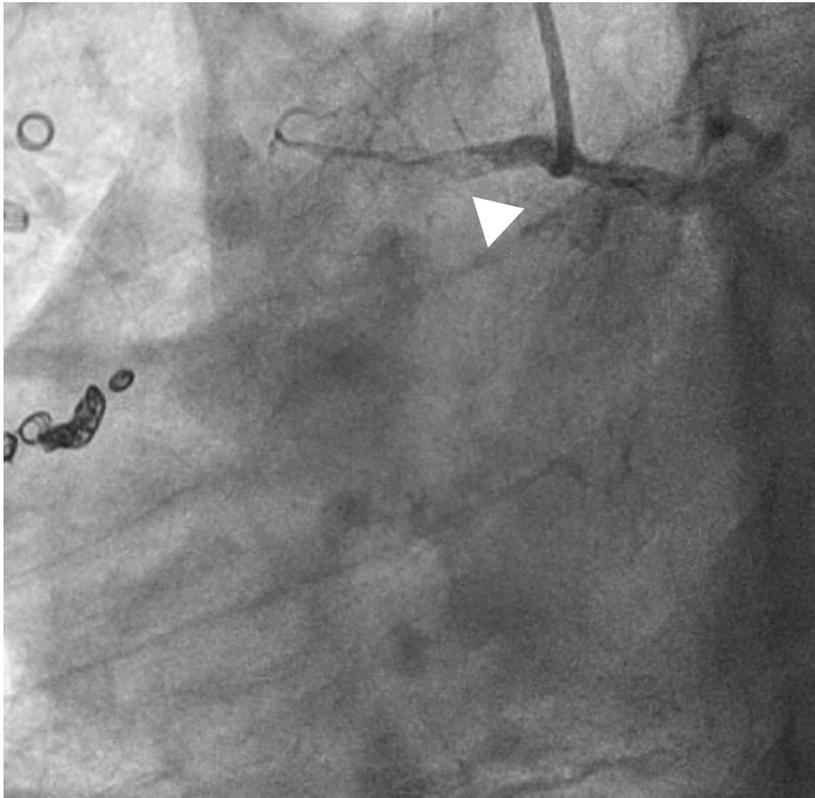
Absent

Possible/Probable

Certain

Classification

Homme de 35 ans – Maladie Rendu-Osler – MVA cérébrales/pulmonaires
SCA ST+ inférieur lors activité sportive



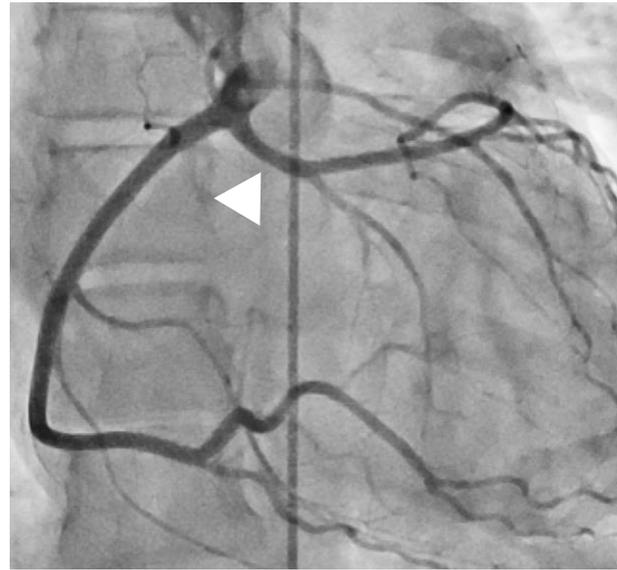
- Embryologie et anatomie
- Classification anatomique
- **Prévalence**
- Imagerie
- Ischémie myocardique
- Mort subite

Prévalence



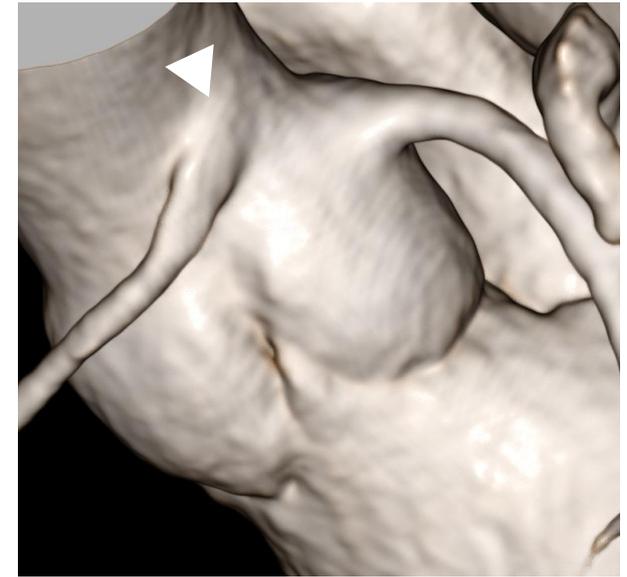
Echocardiographie

0.2%



Coronarographie

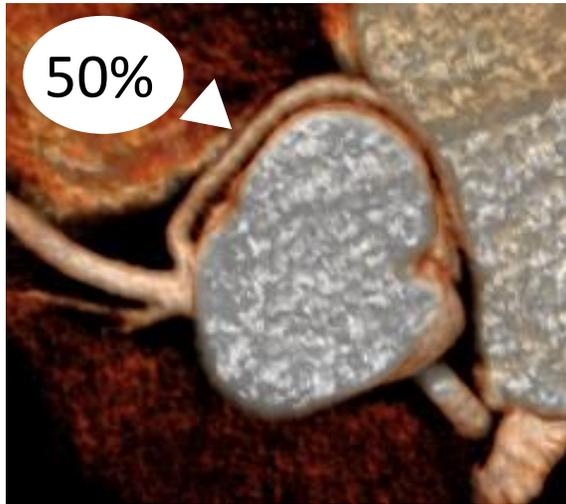
0.8%



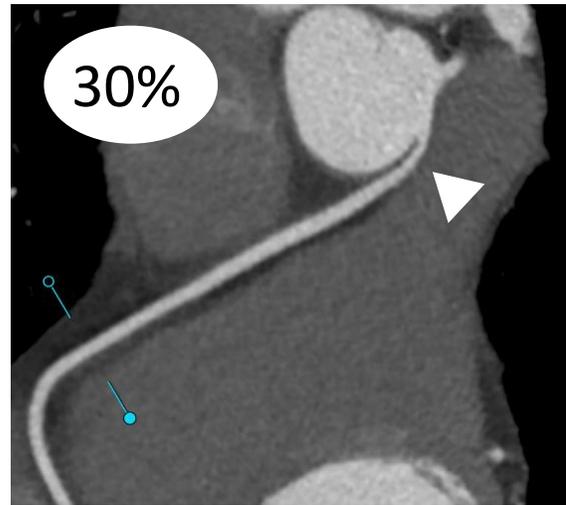
Scanner

1.2%

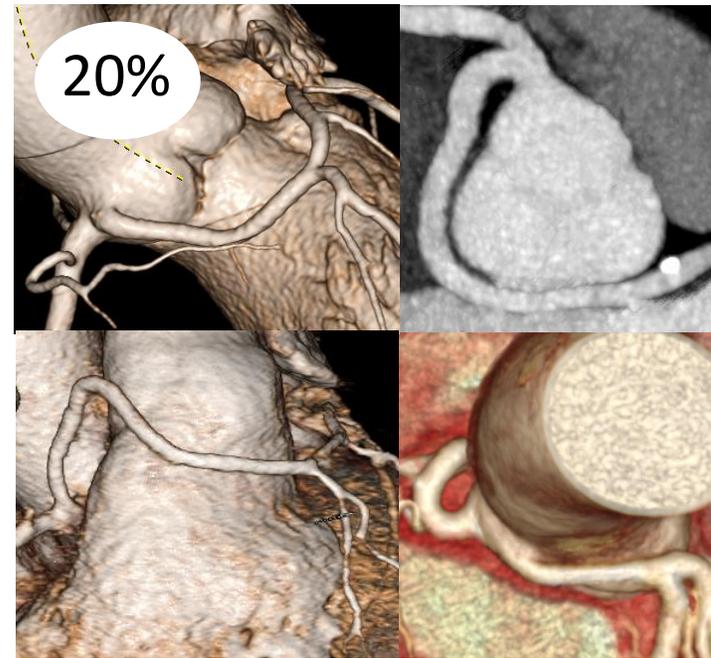
Prévalence selon l'artère coronaire



Circonflexe
rétroaortique (99%)



Droite
interartérielle (95%)



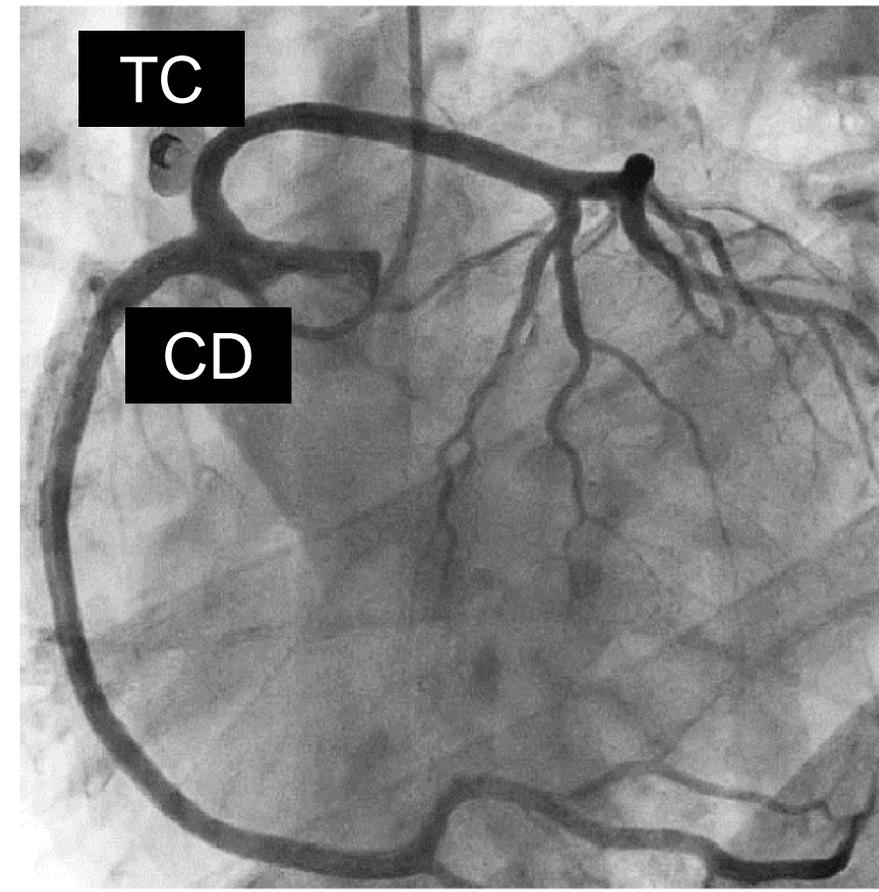
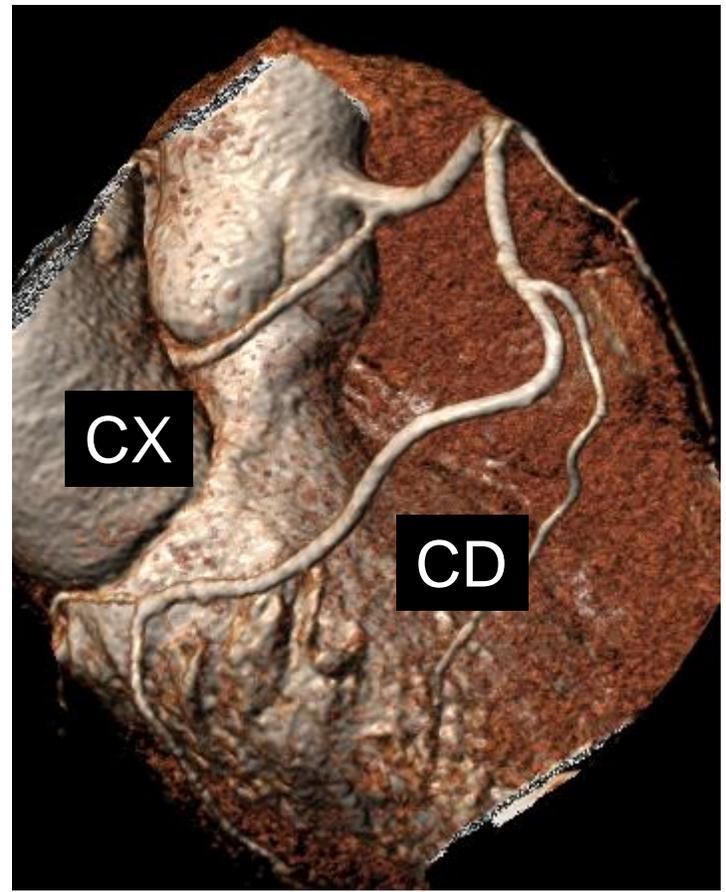
- TC/IVA
rétropulmonaire
(45%)
- prépulmonaire
(30%)
- rétroaortique
(15%)
- interartériel
(10%)

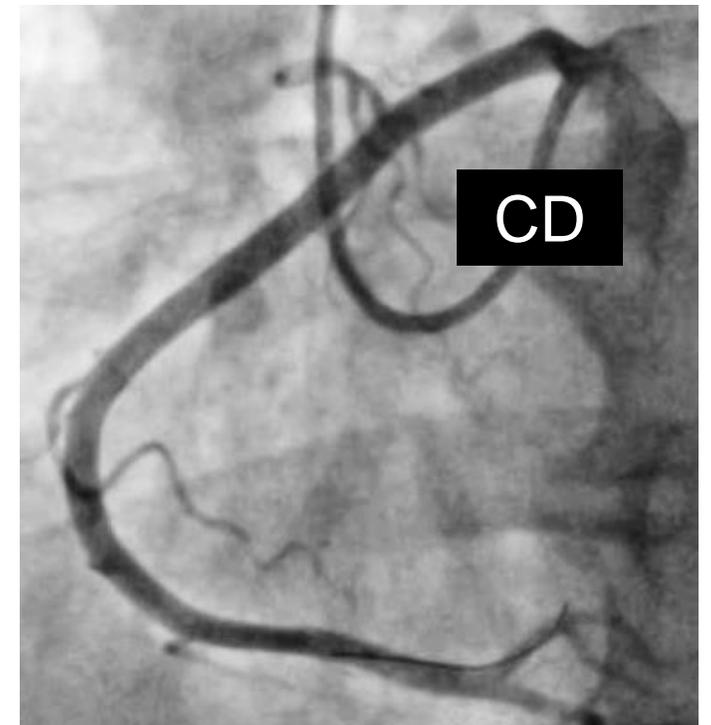
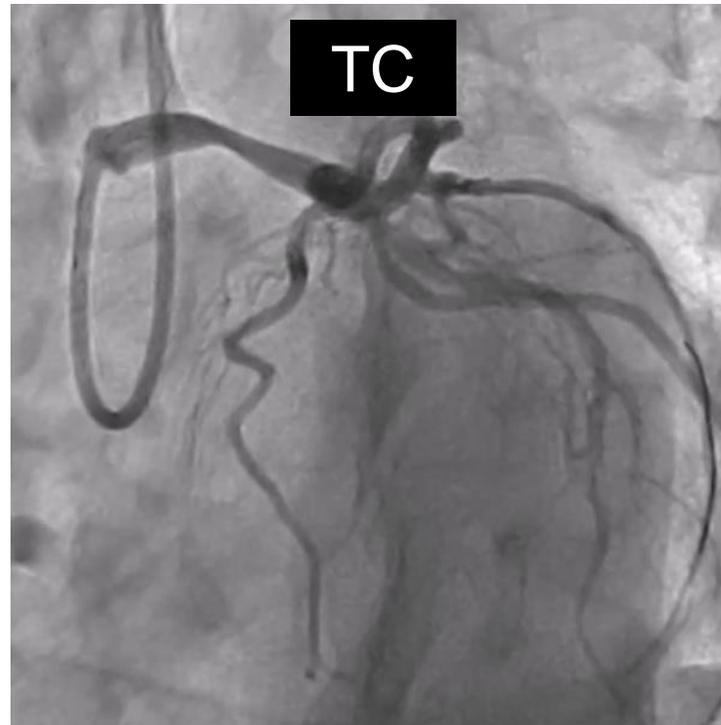
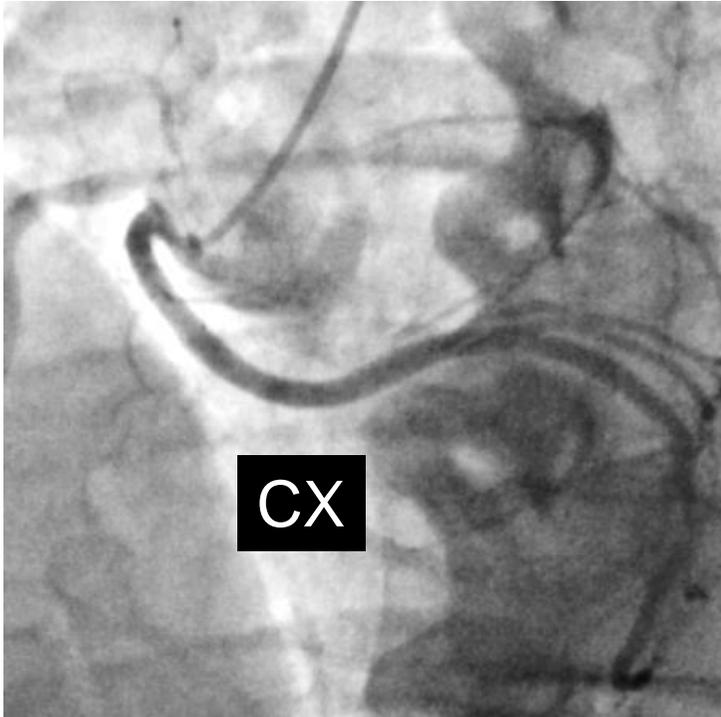
Prévalence selon le trajet ectopique

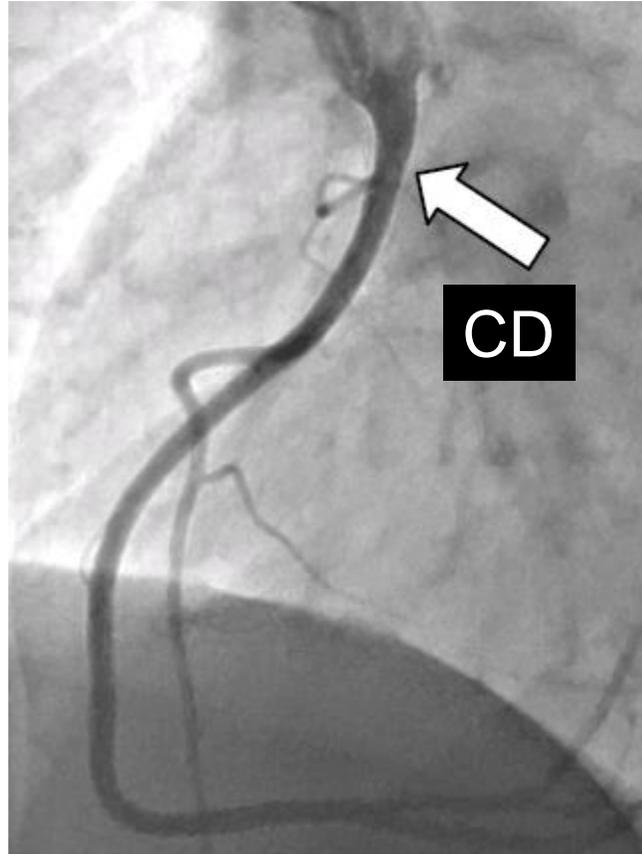
Prévalence selon le site de connexion

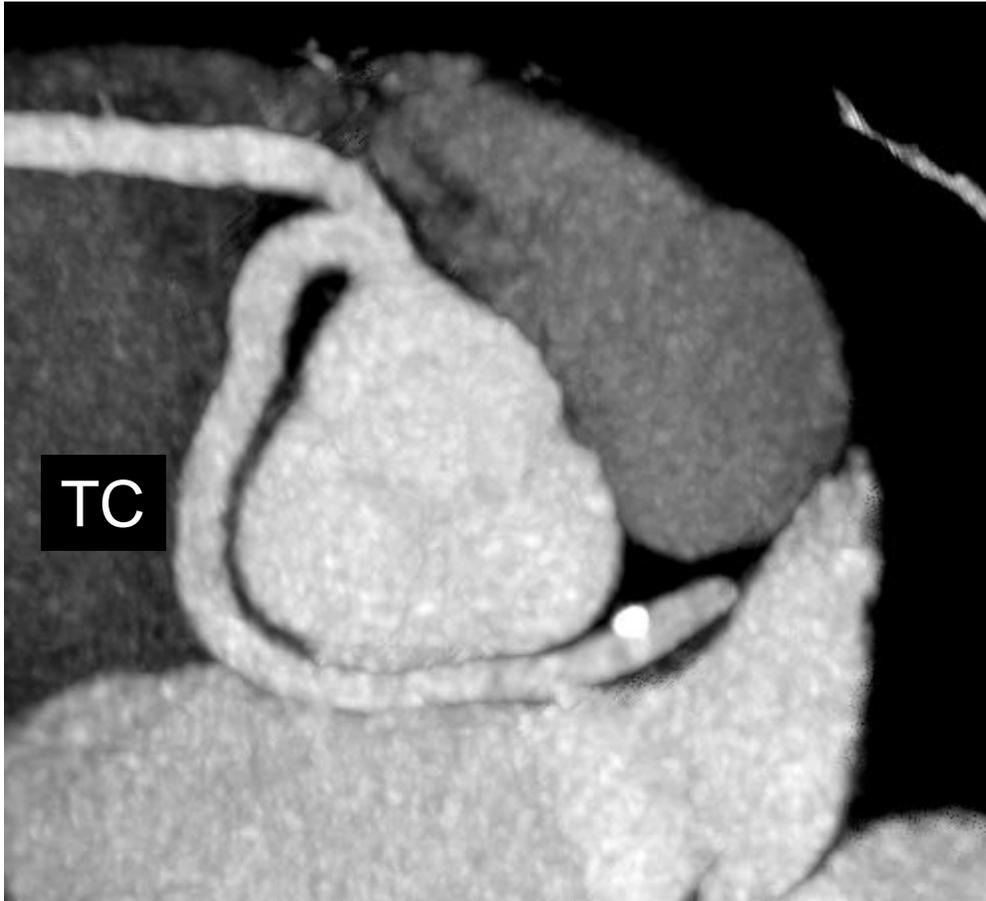
site de connexion	%
connexion dans sinus controlatéral	47.0
connexion dans artère controlatérale	43.5
connexion au-dessus jonction sinotubulaire	1.0
connexion anormale dans sinus habituel	1.0
artère coronaire unique	1.0
connexion avec artère pulmonaire	1.0
connexion dans sinus non coronaire	0.4
autres connexions anormales	0.1

* à partir des données du registre ANOCOR (ESC 2015)
472 patients – 496 anomalies coronaires

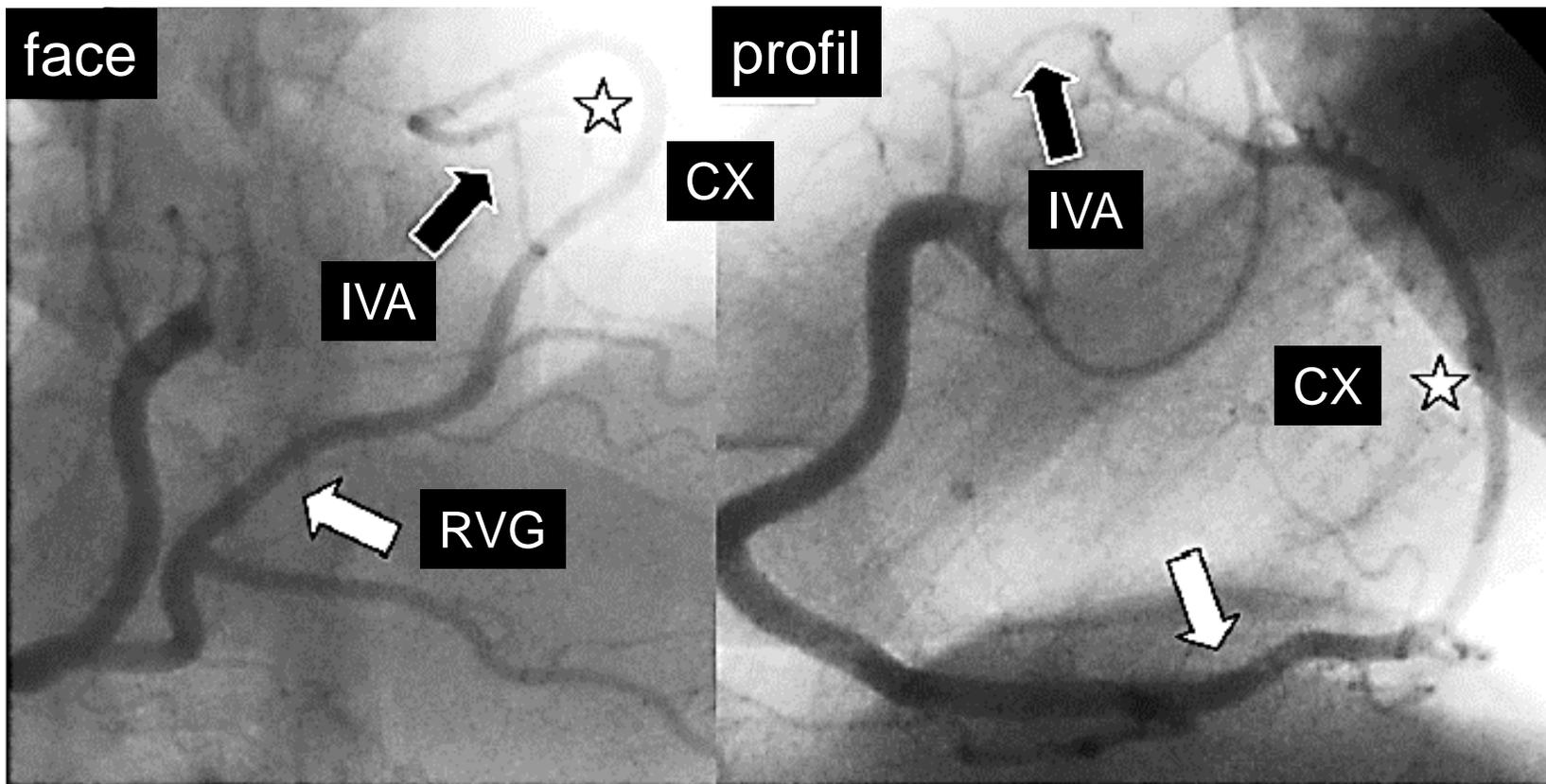






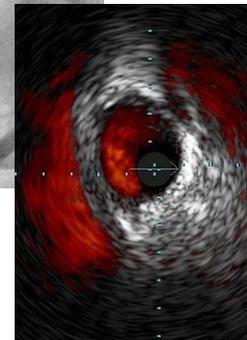
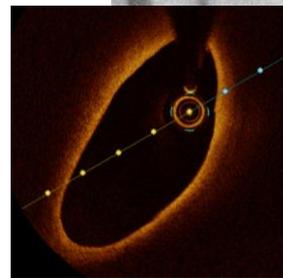
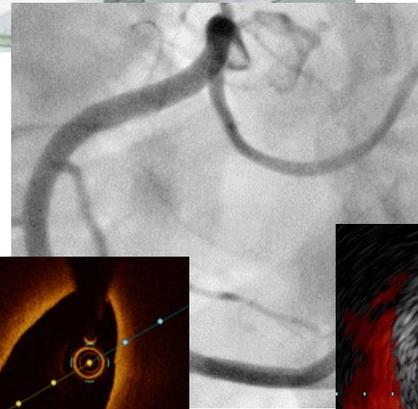
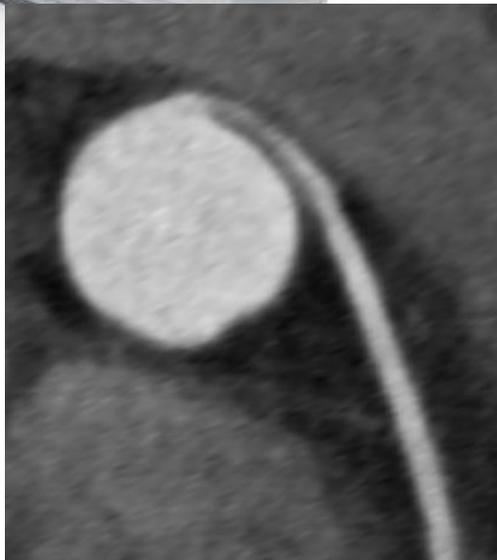
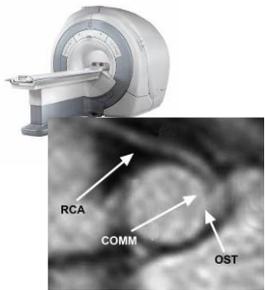


Artère coronaire unique

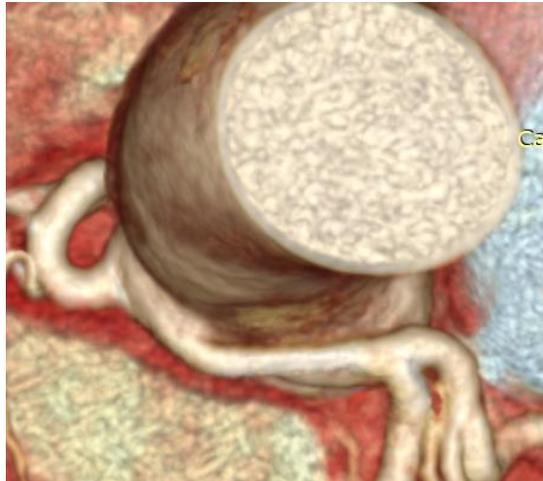


- Embryologie et anatomie
- Classification anatomique
- Prévalence
- **Imagerie**
- Ischémie myocardique
- Mort subite

Outils d'imagerie



Piège à éviter

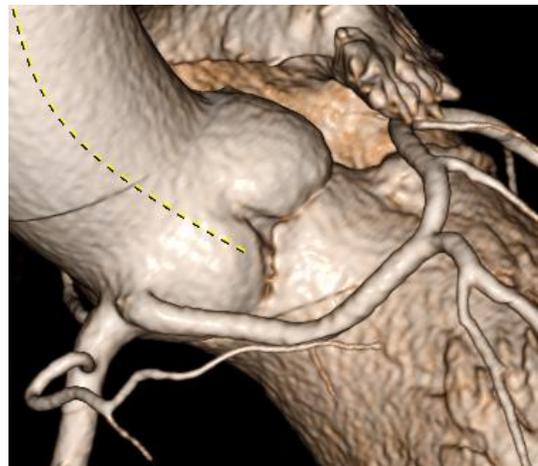


Trajet prépulmonaire

Trajet rétroaortique

Trajet interartériel

Trajet rétropulmonaire



Received: 11 October 2016 | Revised: 21 February 2017 | Accepted: 28 May 2017
DOI: 10.1111/chd.12504

ORIGINAL ARTICLE

WILEY Congenital Heart Disease

Interobserver variability in the classification of congenital coronary abnormalities: A substudy of the anomalous connections of the coronary arteries registry

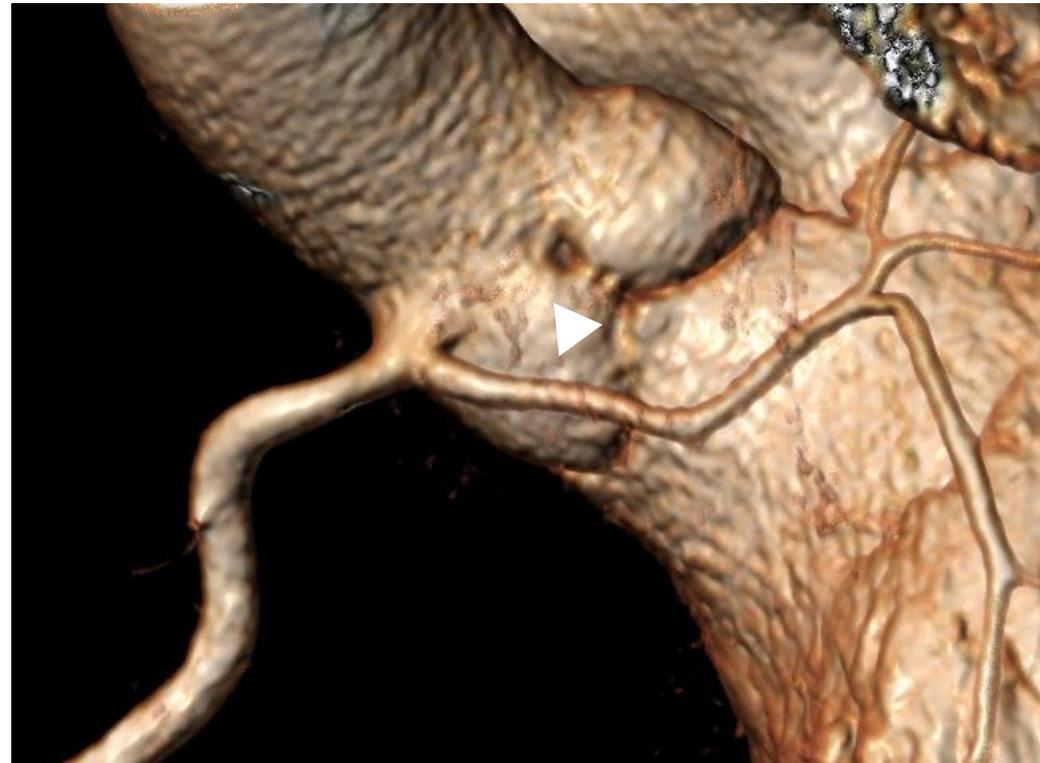
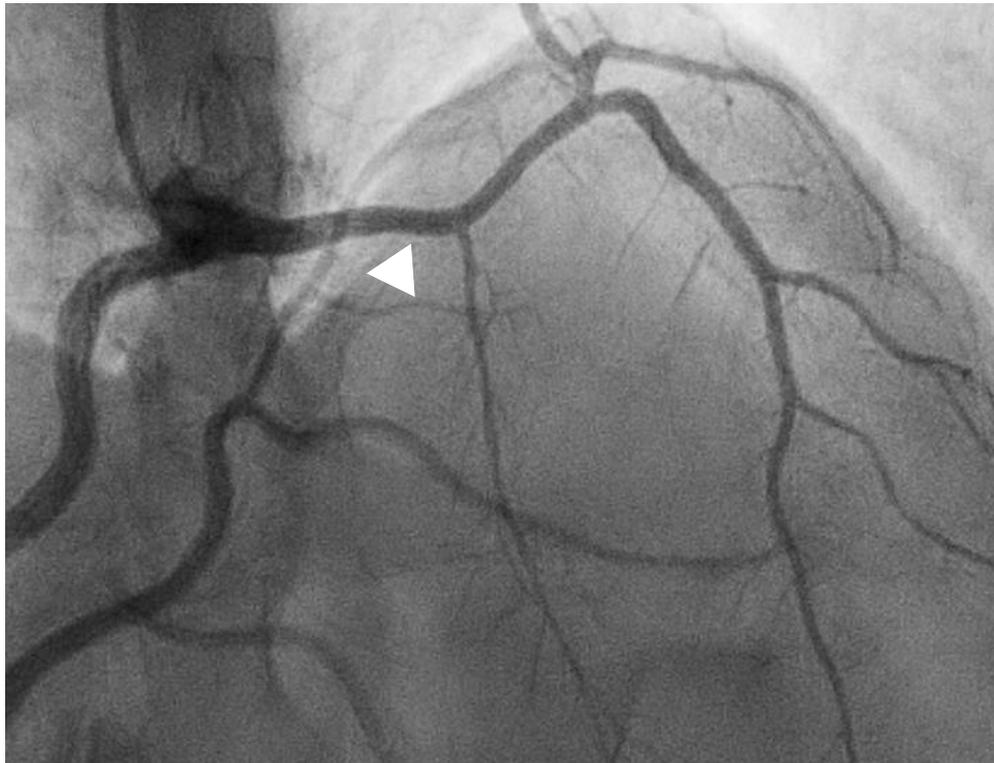
Athanasios Koutsoukis, MD¹ | Xavier Halna du Fretay, MD² | Patrick Dupouy, MD³ | Phalla Ou, MD, PhD⁴ | Jean-Pierre Laissy, MD, PhD⁴ | Jean-Michel Juliard, MD⁵ | Fabien Hyafil, MD⁶ | Pierre Aubry, MD⁵ | on behalf of the ANOCOR Investigators*

Congenital Heart Disease 2017

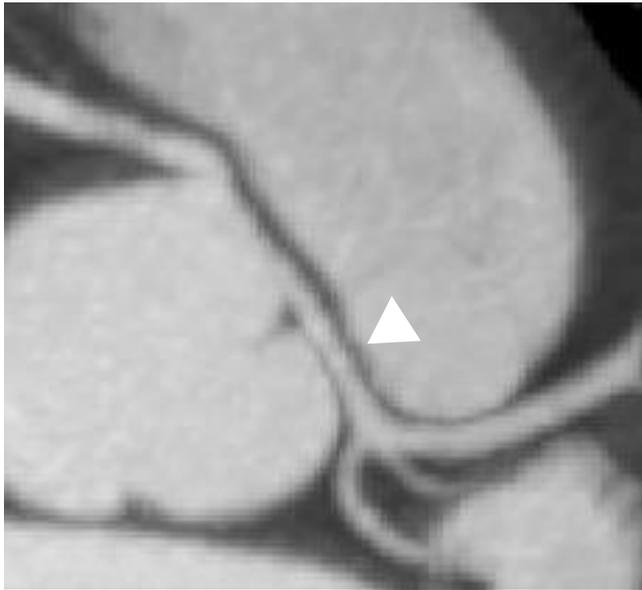
ANOCOR gauche avec trajet interartériel



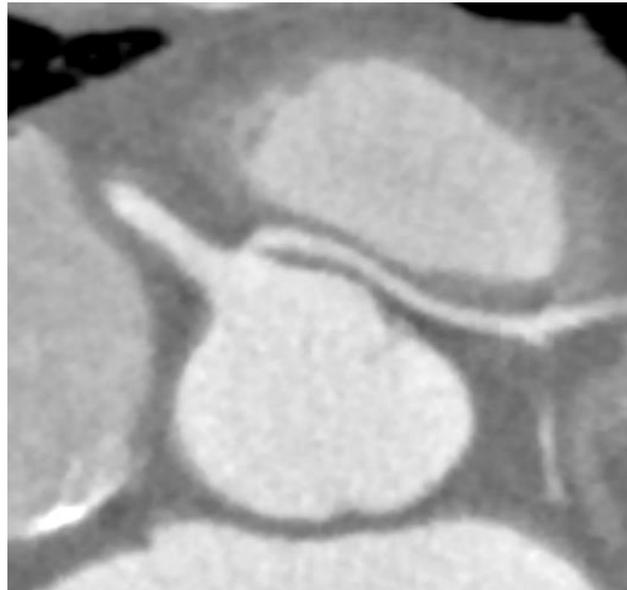
ANOCOR gauche avec trajet rétropulmonaire



Formes anatomiques gauches à ne pas confondre

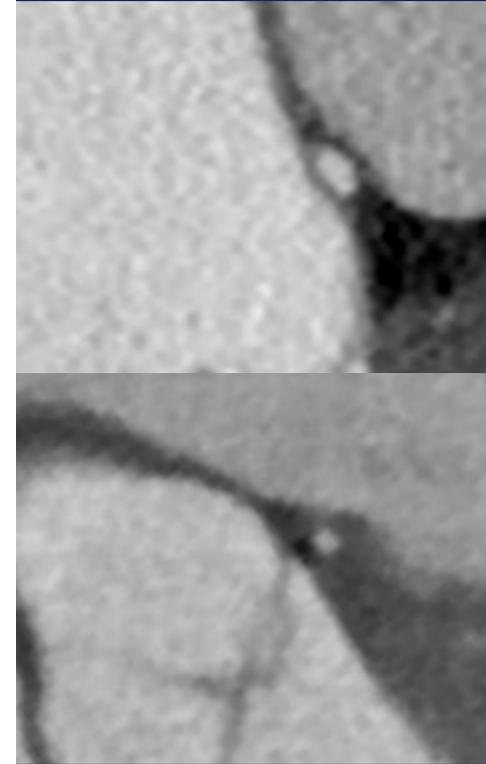


Trajet interartériel



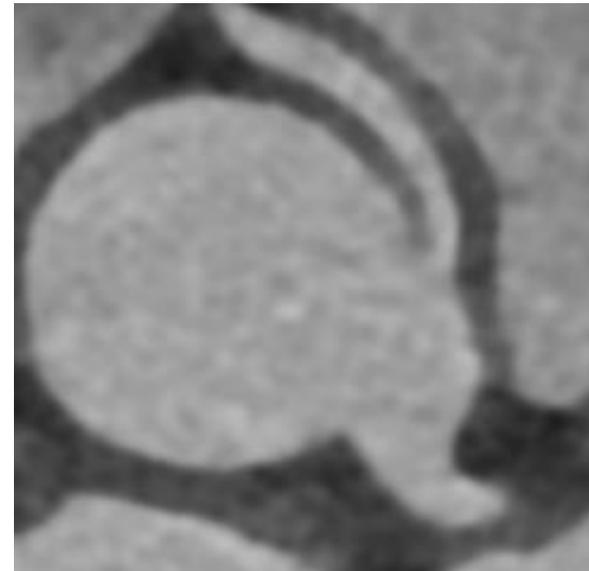
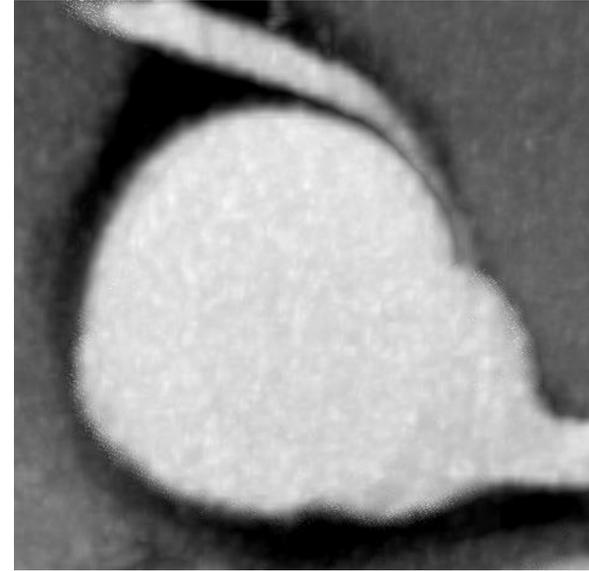
Trajet rétropulmonaire

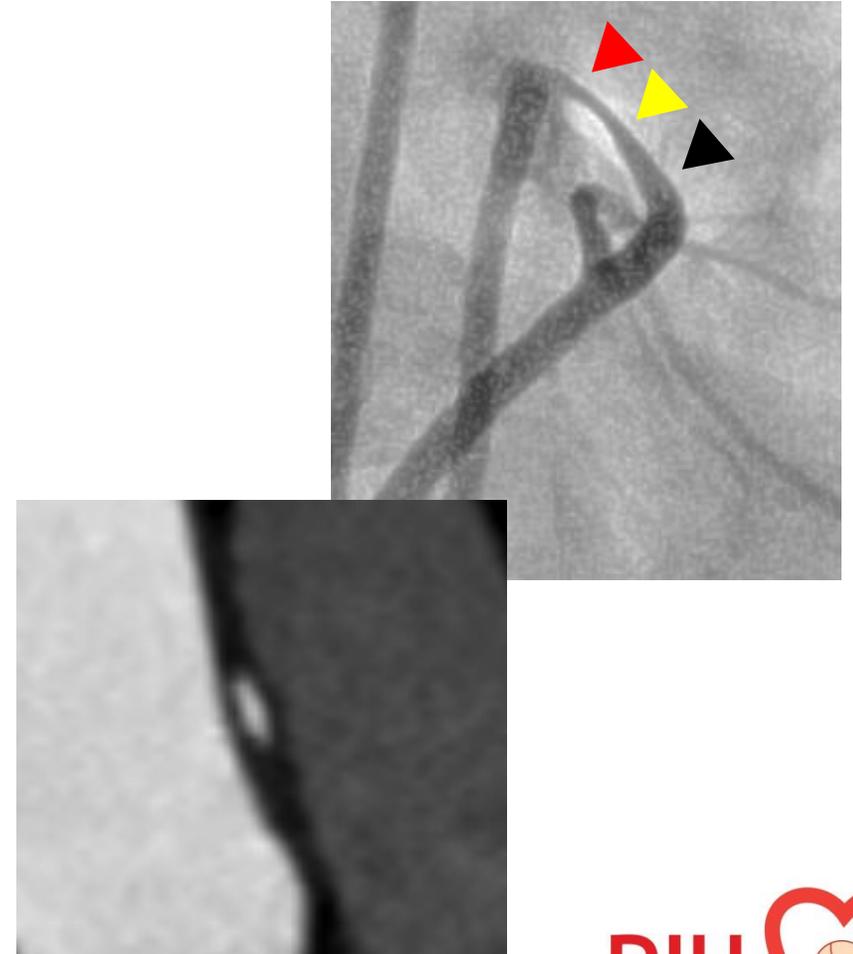
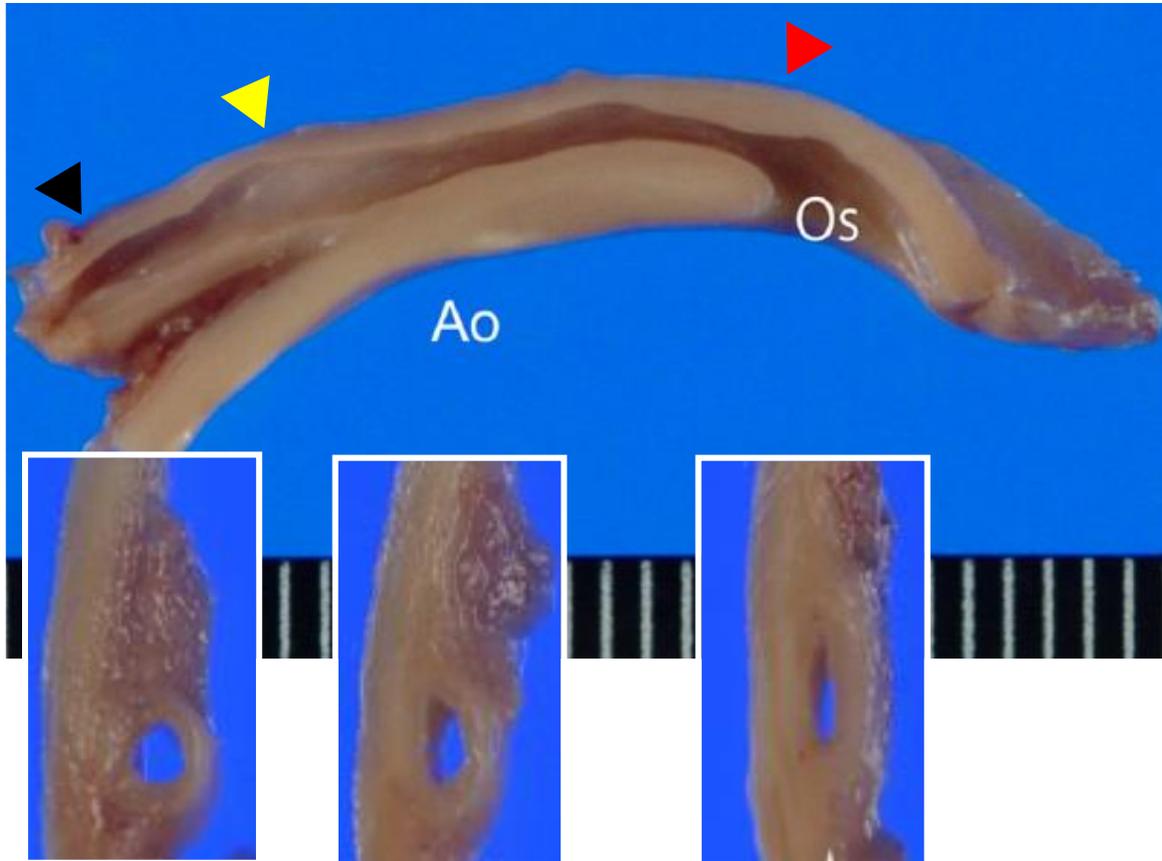
Interartériel



Rétropulmonaire

Passage aortique intramural
Passage aortique juxtamural

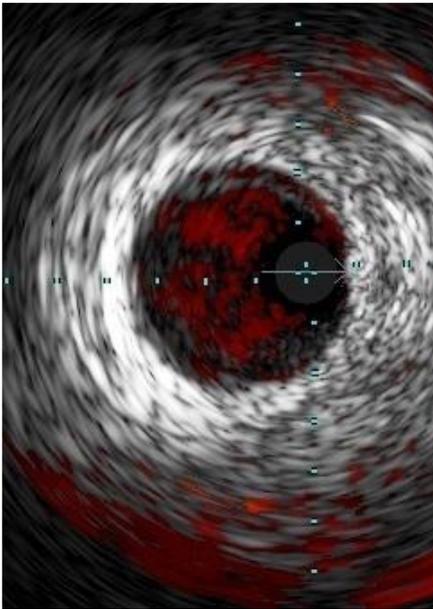




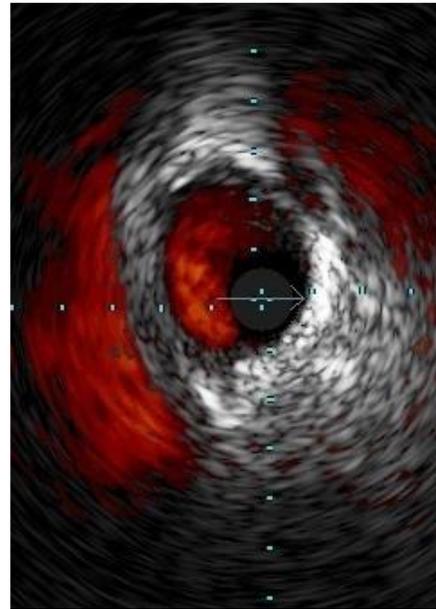
Right ANOCOR with an intramural course
Hata Y et al. Cardiovasc Pathol. 2014.

Echographie endocoronaire

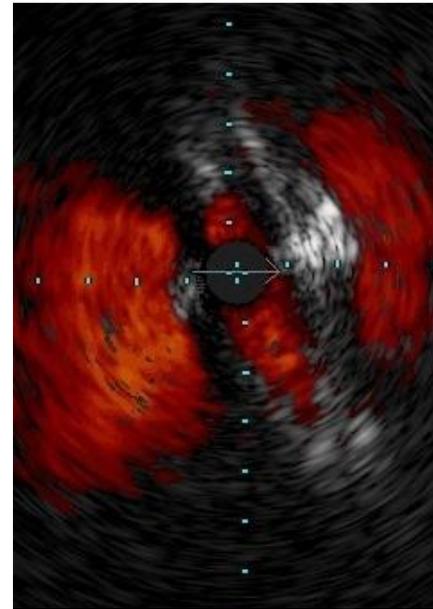
extramural



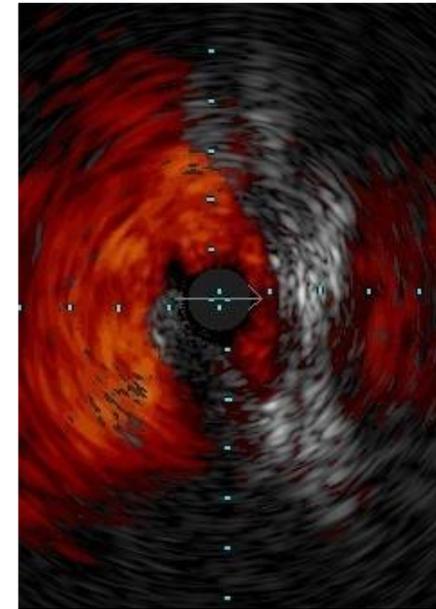
juxtamural



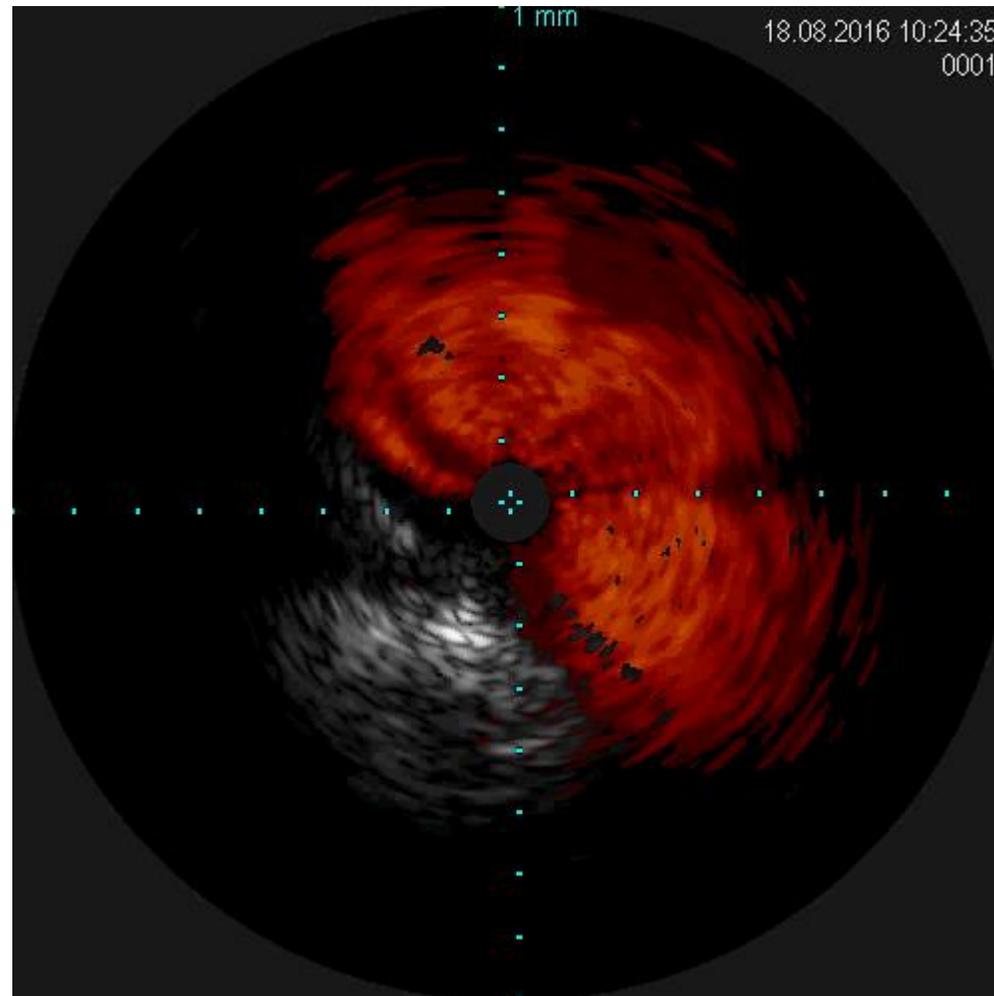
intramural



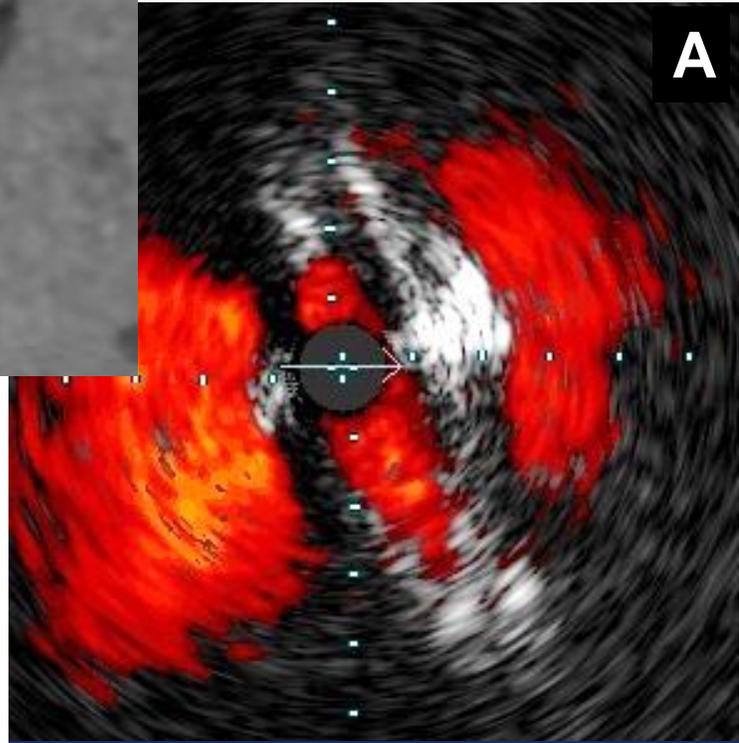
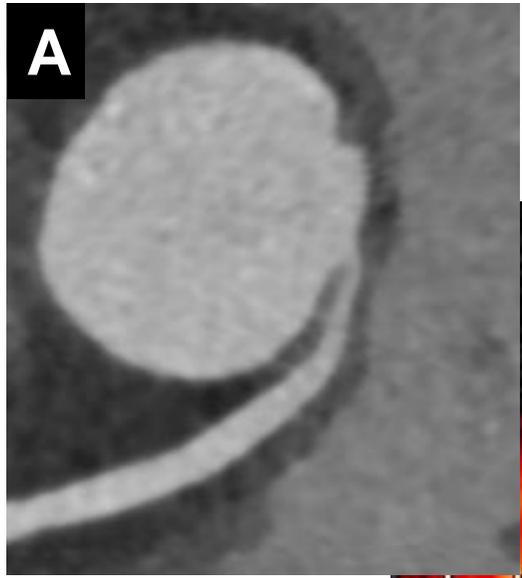
ostium



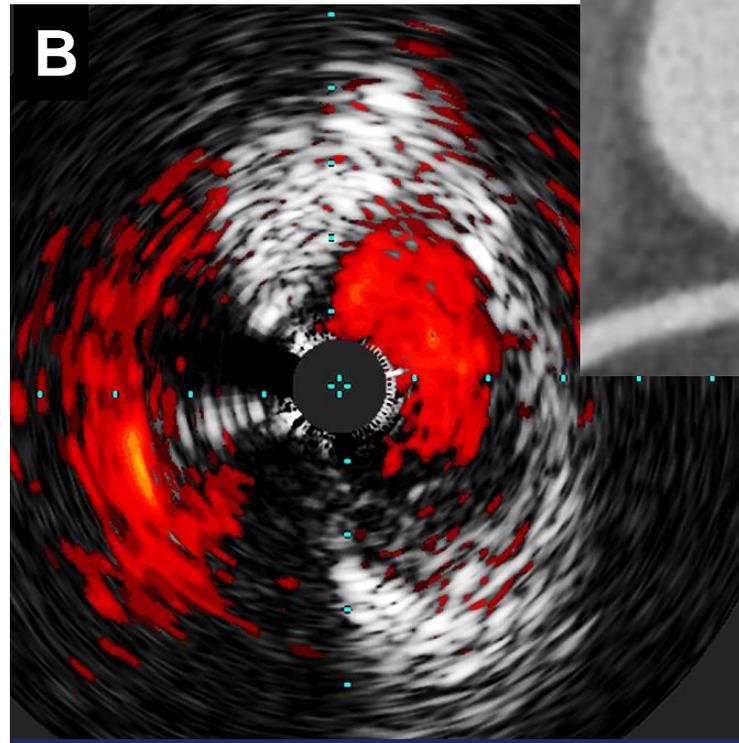
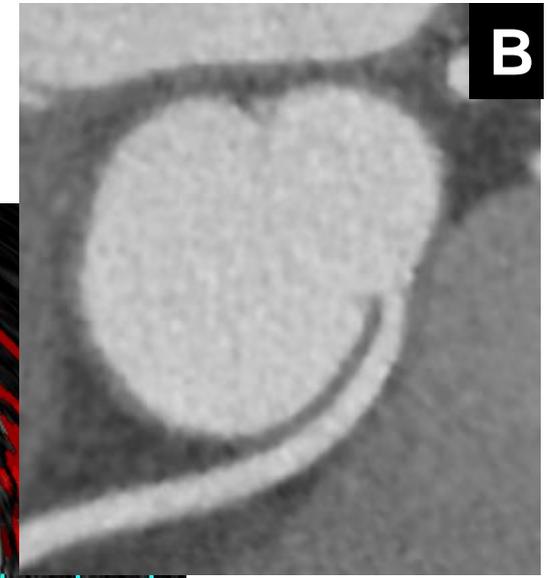
Echographie endocoronaire



Echographie endocoronaire

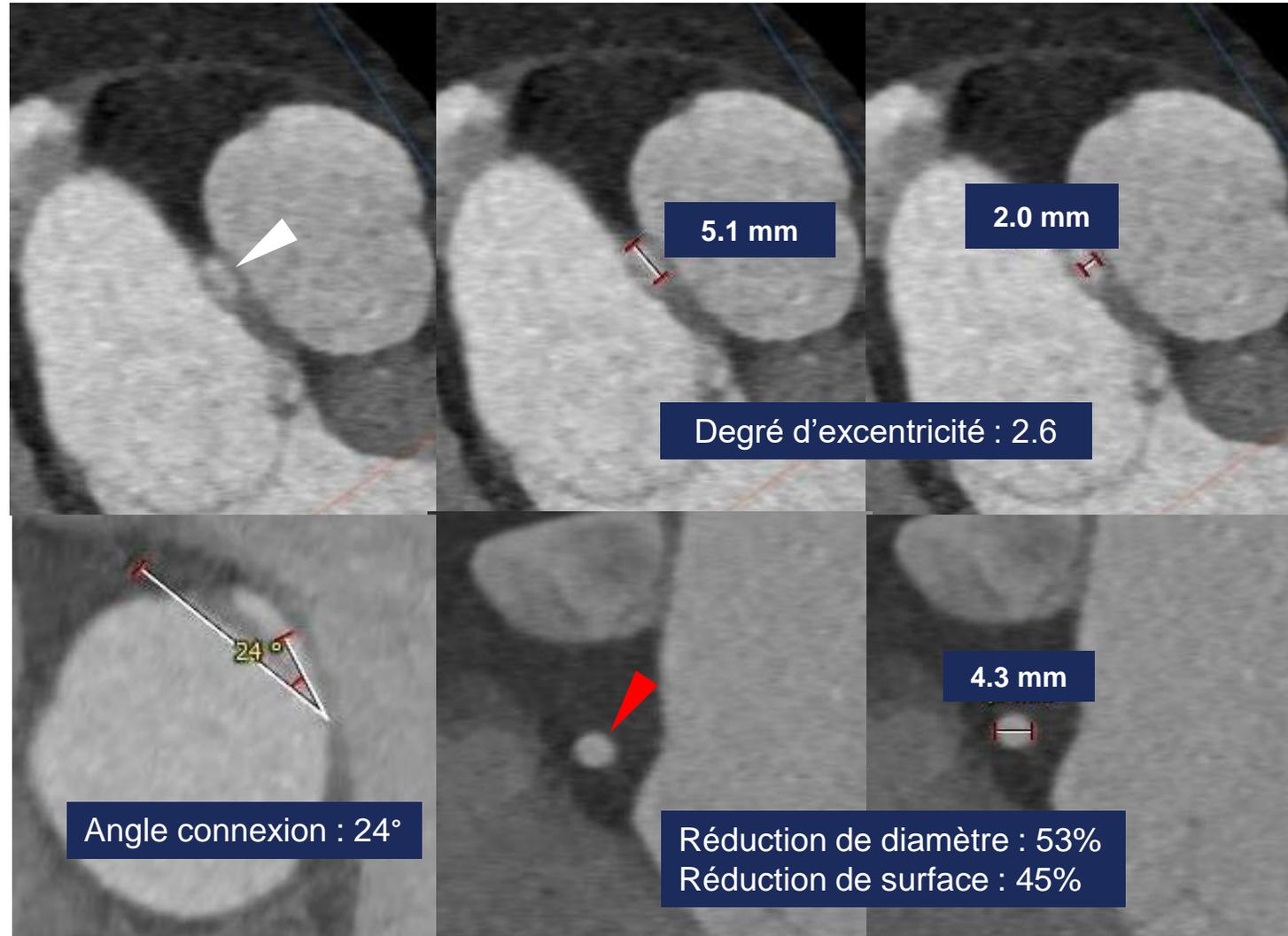
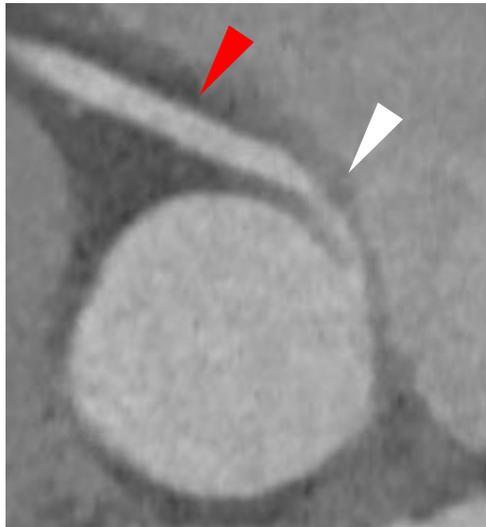


Passage intramural

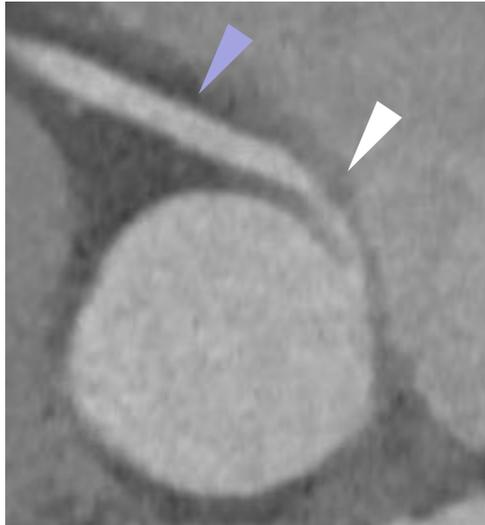


Passage juxtamural

Analyse tomographique



Définition tomographique d'un passage intramural aortique



- Degré d'excentricité (grand axe/petit axe) ≥ 2.0

- Angle de connexion $\leq 30^\circ$

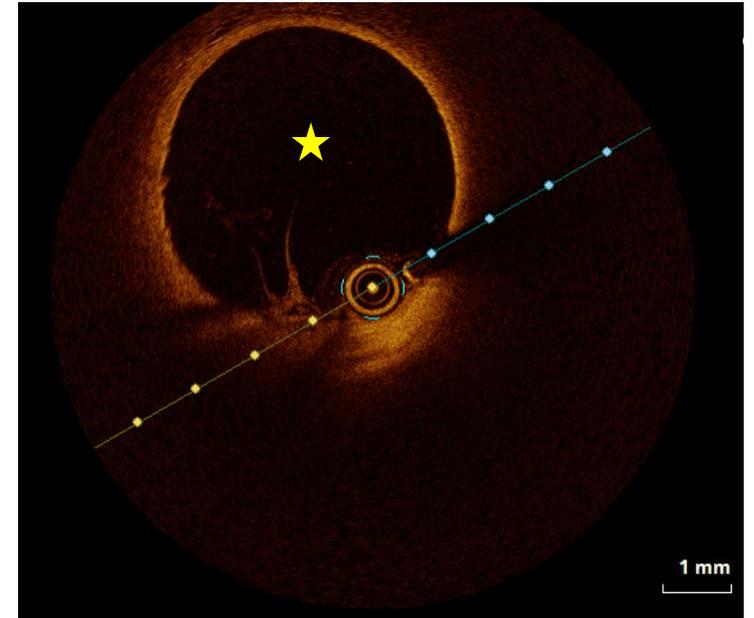
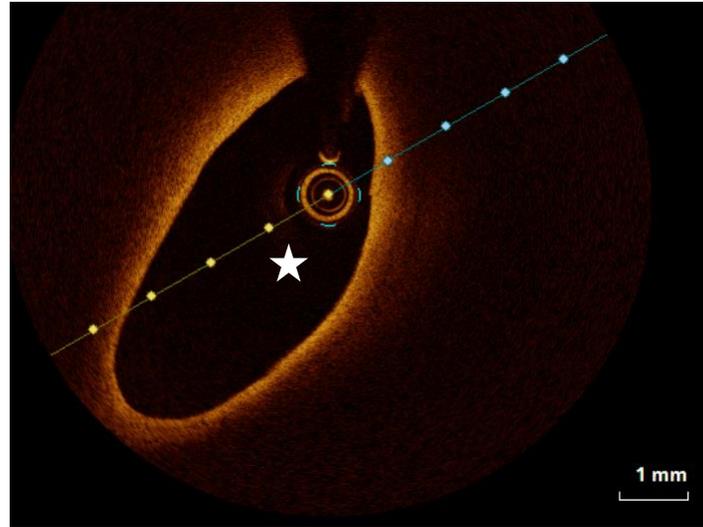
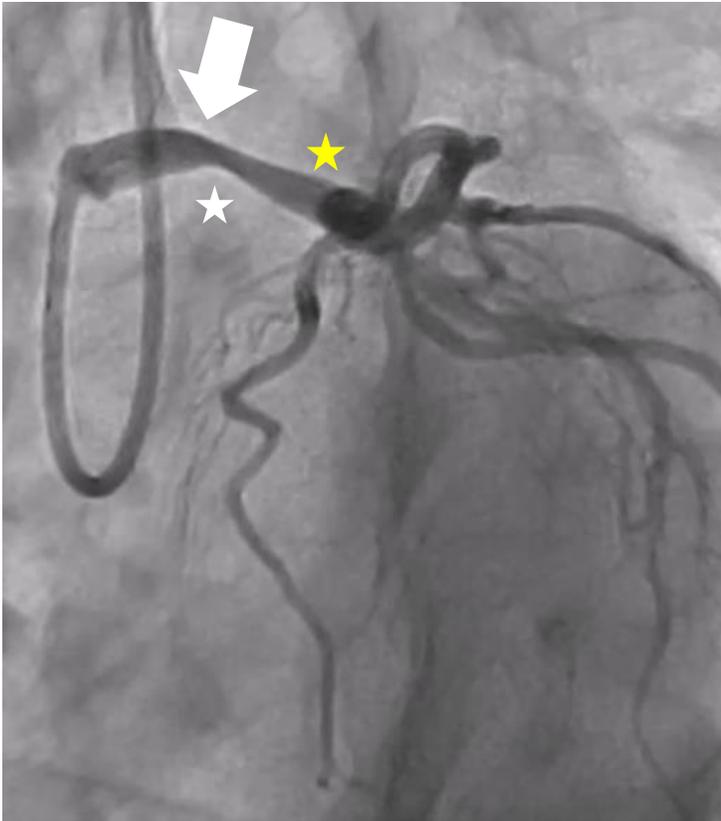
- Réduction de diamètre $\geq 50\%$

Trois critères présents* : passage intramural aortique certain

Deux critères présents* : passage intramural aortique incertain

Zéro ou un critère présent : passage intramural aortique absent

* Petit axe < 2.0 mm

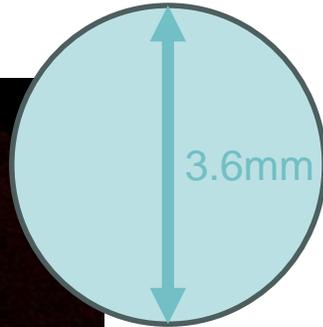
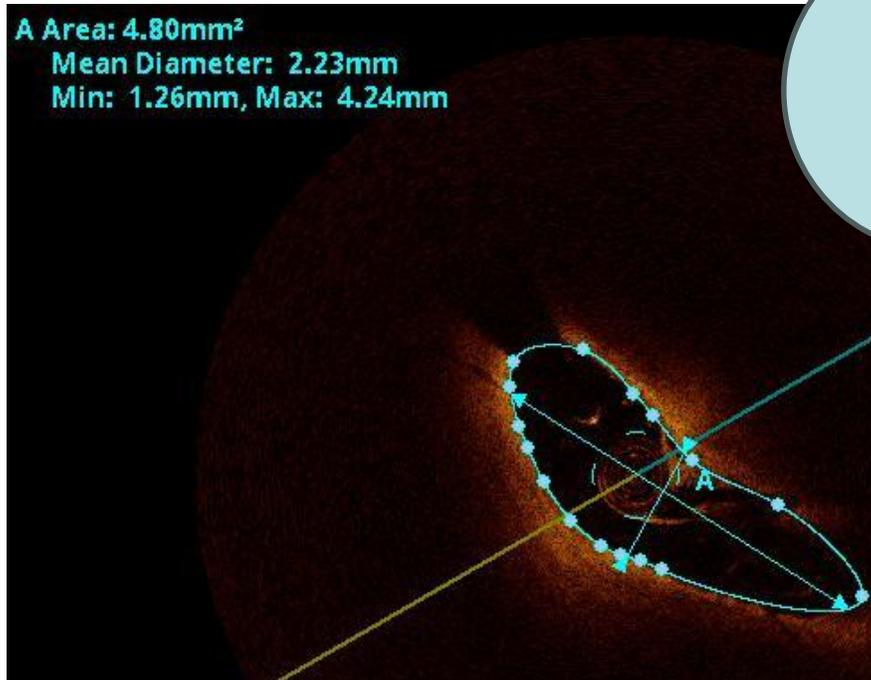


- Embryologie et anatomie
- Classification anatomique
- Prévalence
- Imagerie
- **Ischémie myocardique**
- Mort subite

Ischémie myocardique

- Mort subite/arrêt cardiaque récupéré
- Symptômes d'allure ischémique (angor/syncope/dyspnée)
- Ischémie myocardique (avec imagerie)

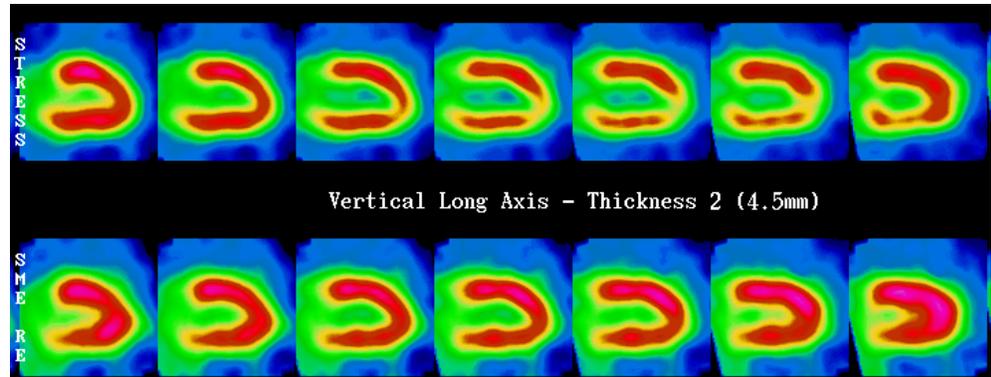
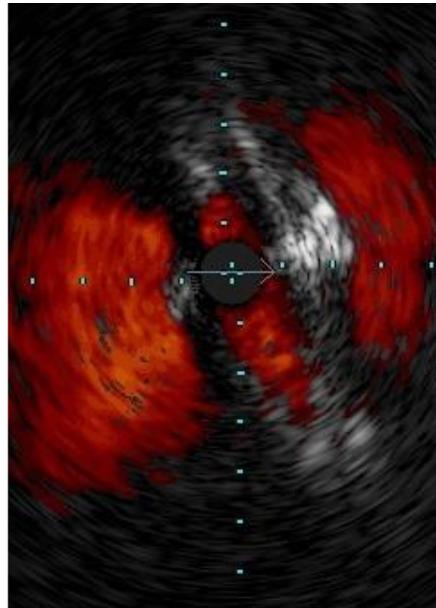
ANOCOR droites ou gauches avec trajet interartériel Ischémie myocardique peu fréquemment documentée



Diamètre luminal normal : 3.6 mm
Diamètre luminal minimal : 1.3 mm
Réduction de diamètre luminal : **64%**

Surface luminale normale : 10.2 mm²
Surface luminale minimale : 4.8 mm²
Réduction de surface luminale : **53%**

Ischémie myocardique documentée

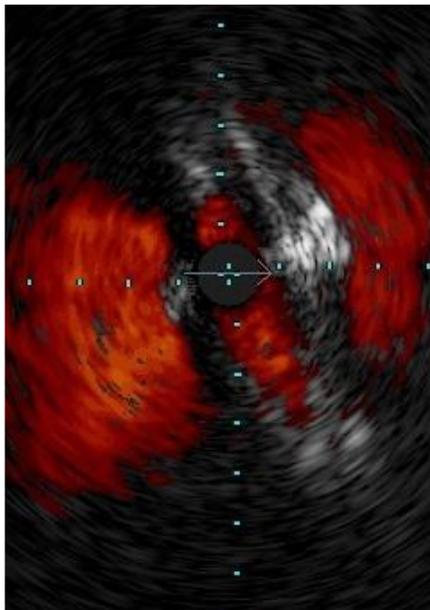


Prévalence \approx 10%

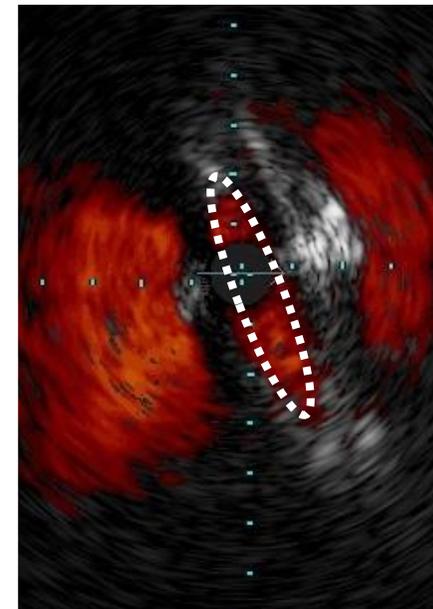
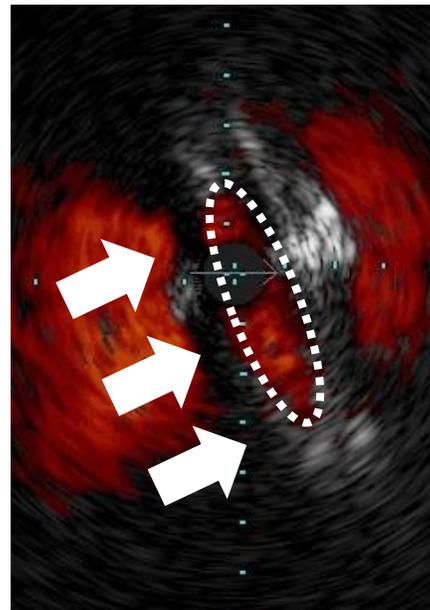
Ischémie myocardique

Two-Tier Concept

Fixed Component



Dynamic Component



Recherche d'ischémie myocardique

TABLE 3 | Overview of possible stress protocols in assessing patients with ACAOS.

	Physical exercise		Adenosine	Regadenoson	Norepinephrine	Dobutamine	Dobutamine + volume challenge
Protocol/dose	85% of max. HR	100% of max. HR	140 µg/kg/min	Bolus: 400 µg	0.01 µg/kg/min	40 µg/kg/min	40 µg/kg/min + saline: 1.5–3 l+ atropine: 1 mg
Applied in	Non-invasive testing	Non-invasive testing	Non-invasive / invasive testing	Non-invasive testing	Invasive testing	Non-invasive / invasive testing	Invasive testing
Increase in coronary blood flow to detect relevant fixed stenosis	+++	+++++	+++	+++	++	+++	+++
Increased heart minute volume to provoke dynamic lateral compression	++	+++++	-	-	+++	++	+++
Reproducibility of symptoms	+++	+++++	-	-	++	++	+++
Tolerability	+++	+++	++	+++	++	++	++

HR, heart rate.

Recherche d'ischémie myocardique

Homme de 40 ans

Sportif +

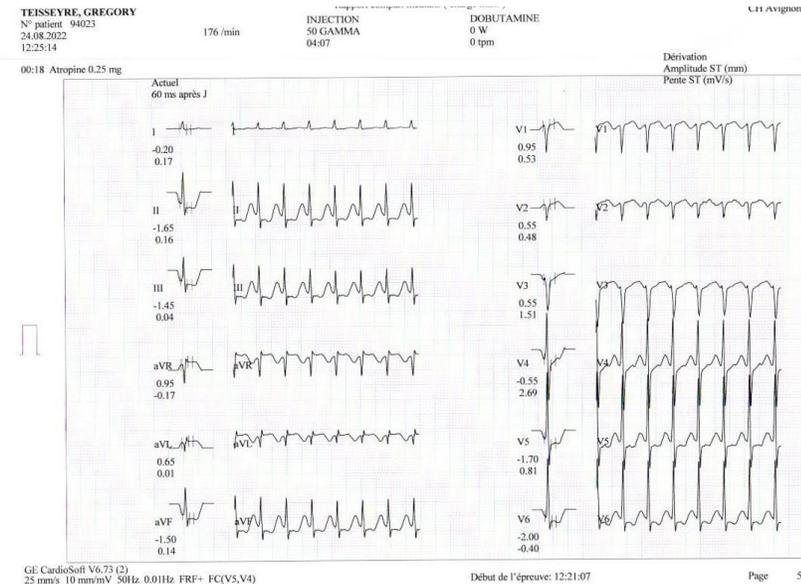
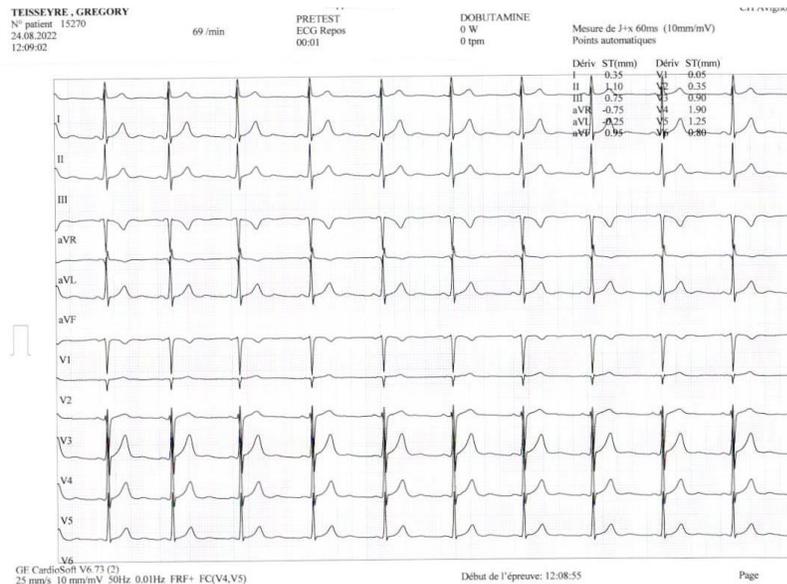
Pas d'antécédents cardiaques

Découverte fortuite ANOCOR gauche avec trajet interartériel

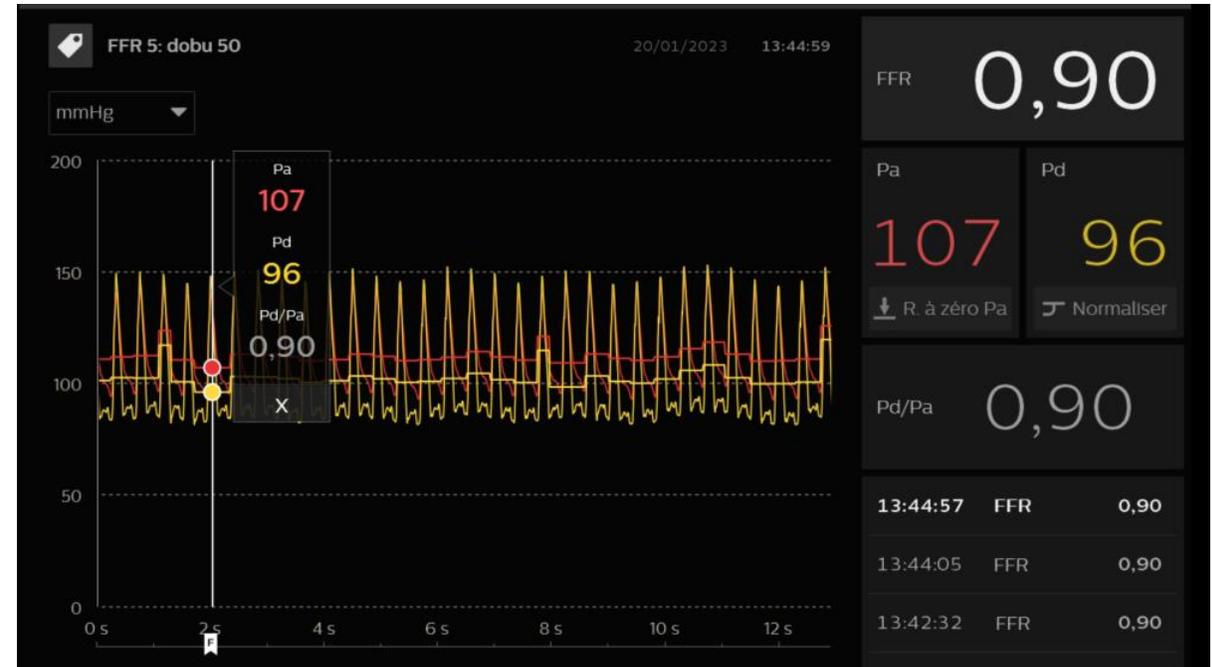
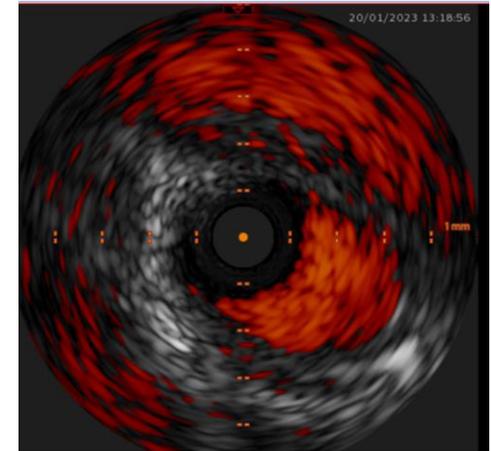
Scintigraphie myocardique d'effort : normale à 100% FMT

Echocardiogramme d'effort : normale à 80% FMT

Echocardiogramme sous dobutamine : lipothymie sans angor à 50 gamma/kg/min



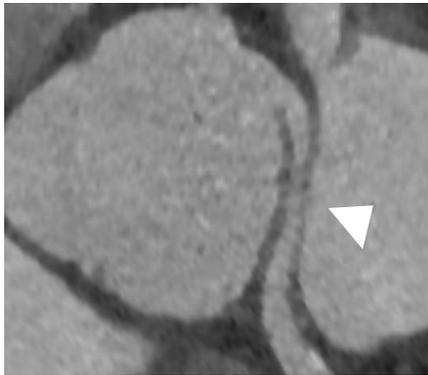
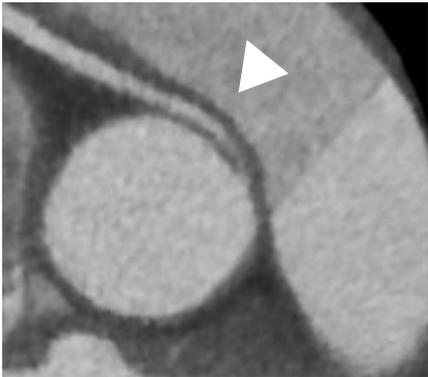
ANOCOR droite FFR sous dobutamine



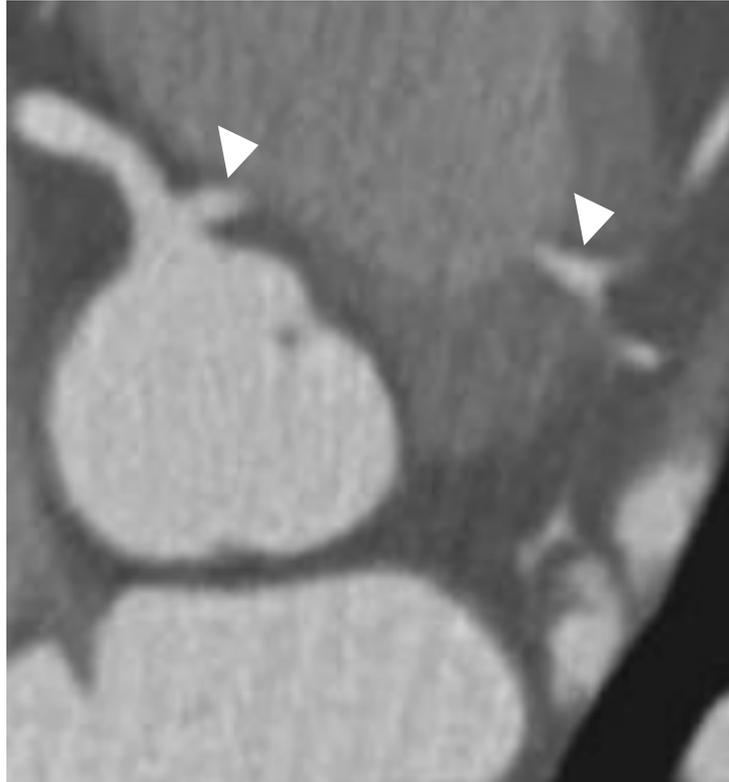
ANOCOR à risque d'ischémie myocardique

Connexions aortiques

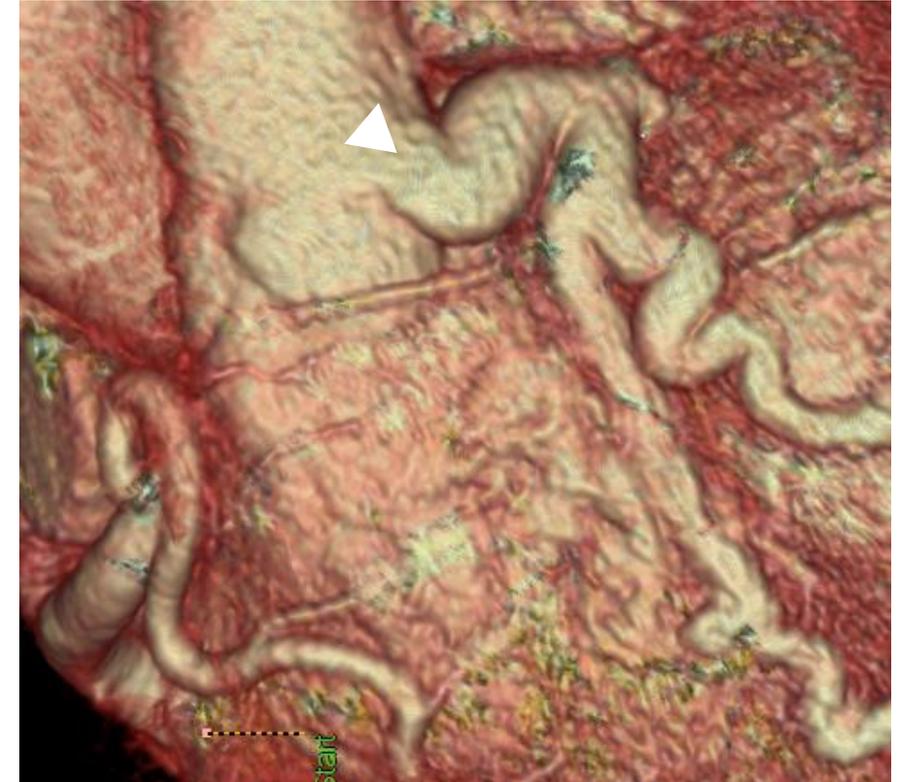
Trajet interartériel



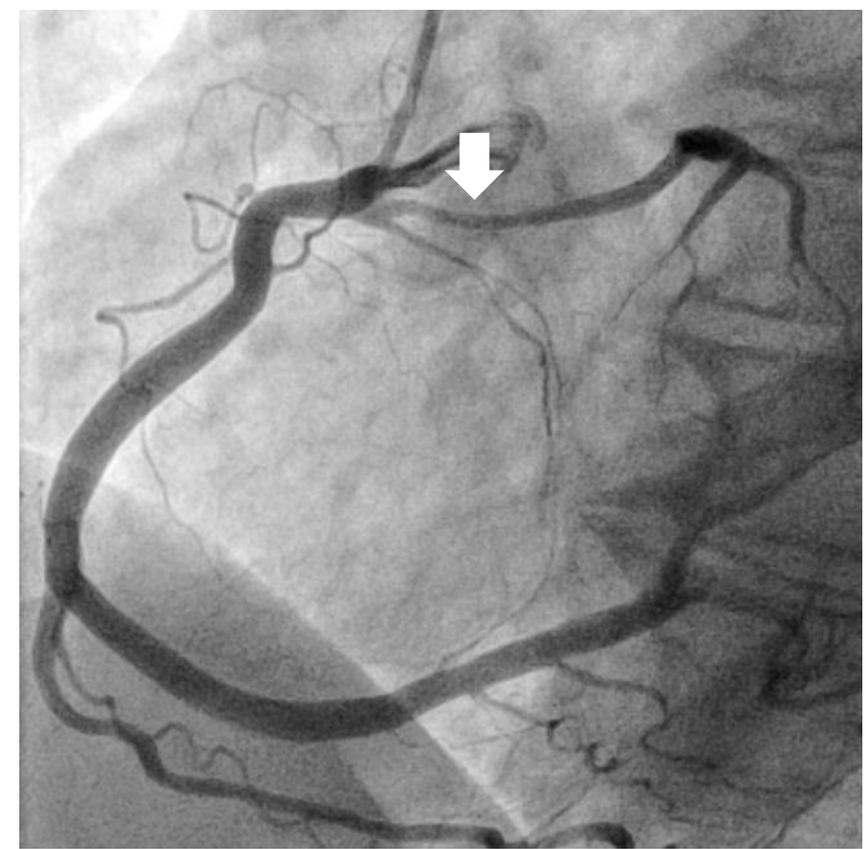
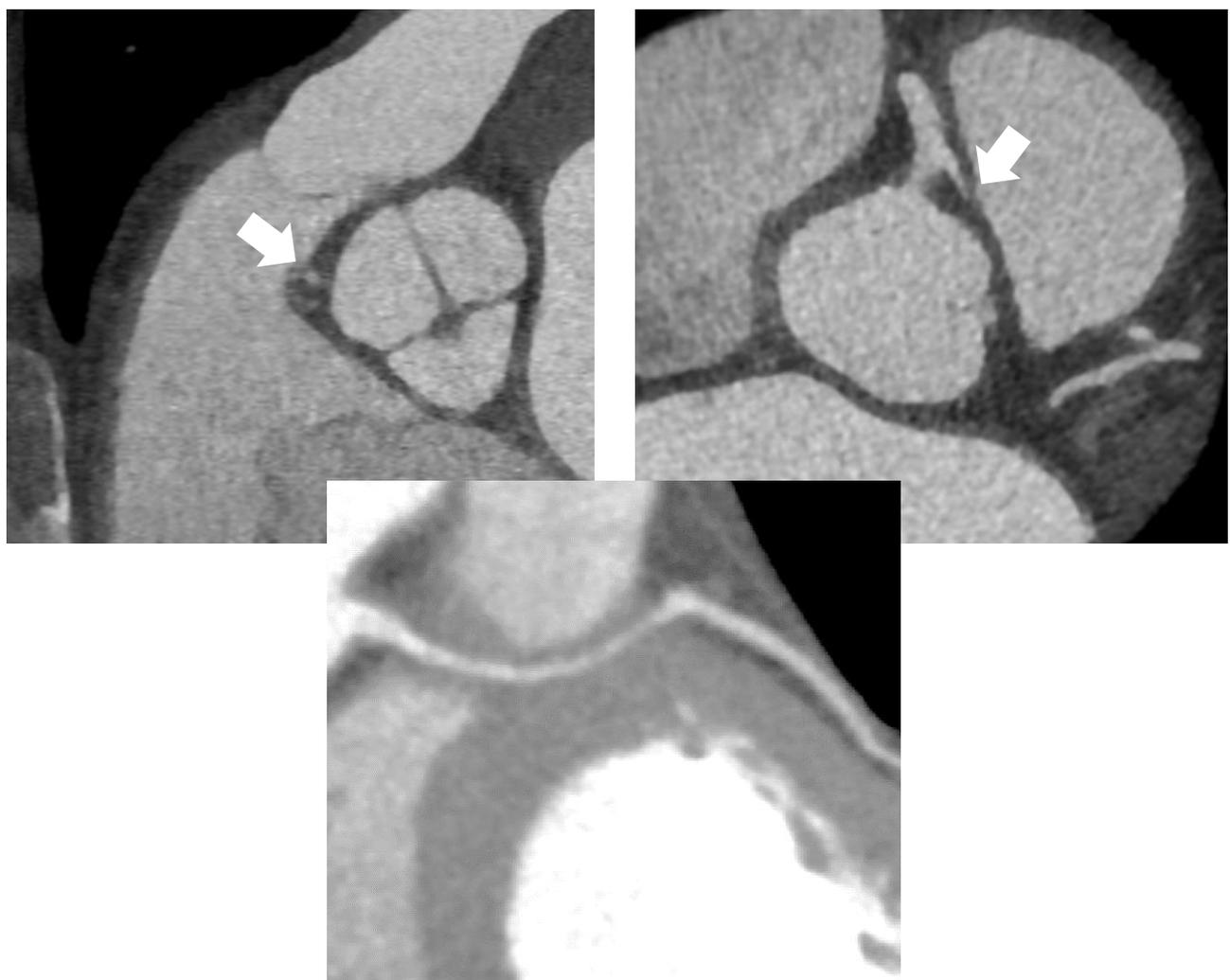
Trajet rétropulmonaire



Connexions pulmonaires

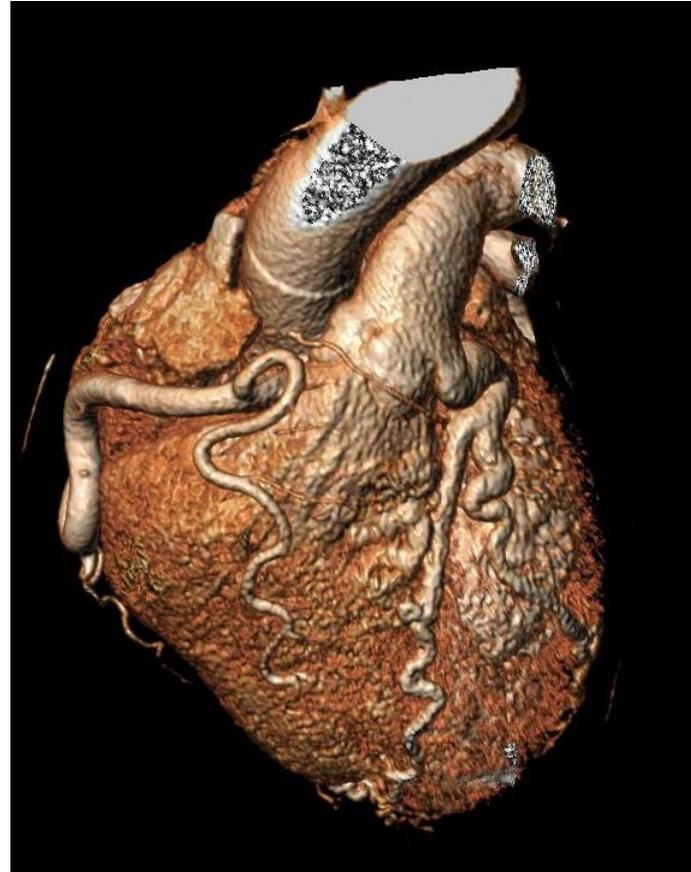
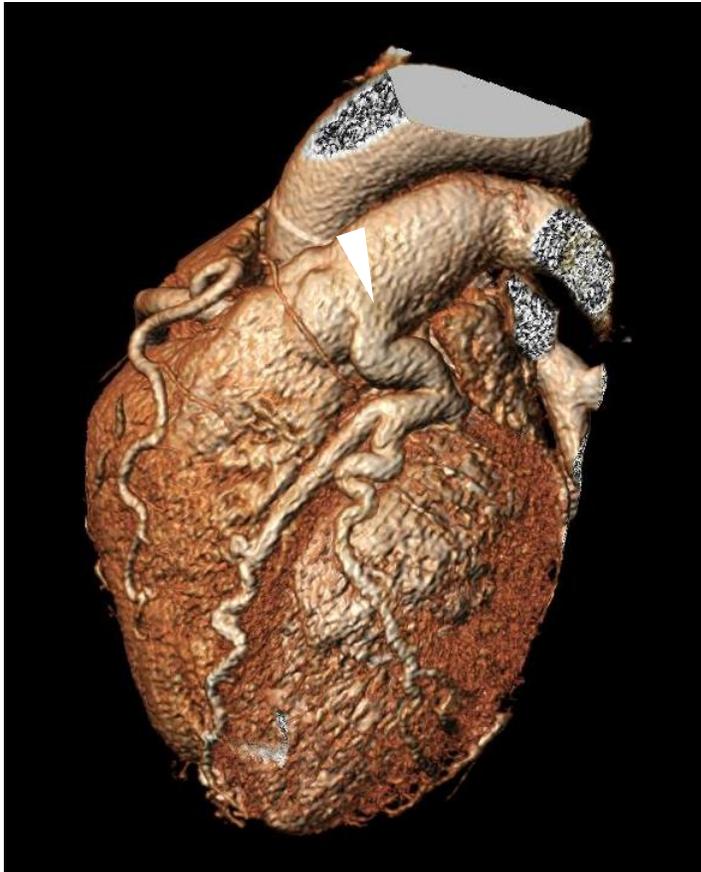


Trajet rétropulmonaire du tronc gauche



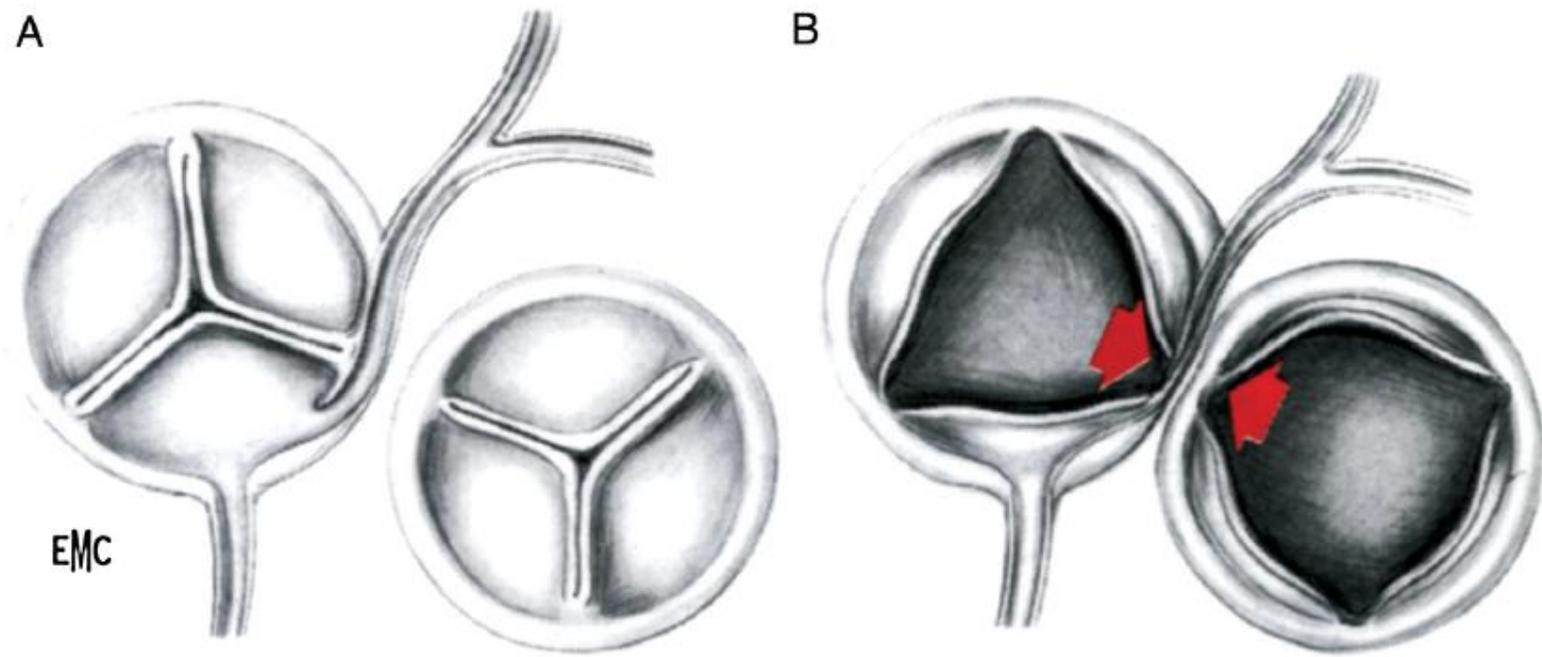
ANOCOR à risque d'ischémie myocardique

Connexions pulmonaires



- Découverte très rare chez l'adulte
- Droite > IVA > Tronc commun > Circonflexe
- Cardiomyopathie ischémique
- VG dilaté et hypokinétique
- Insuffisance mitrale possible
- Séquelle possible de nécrose
- Hypoperfusion myocardique chronique

Exertional dynamic compression

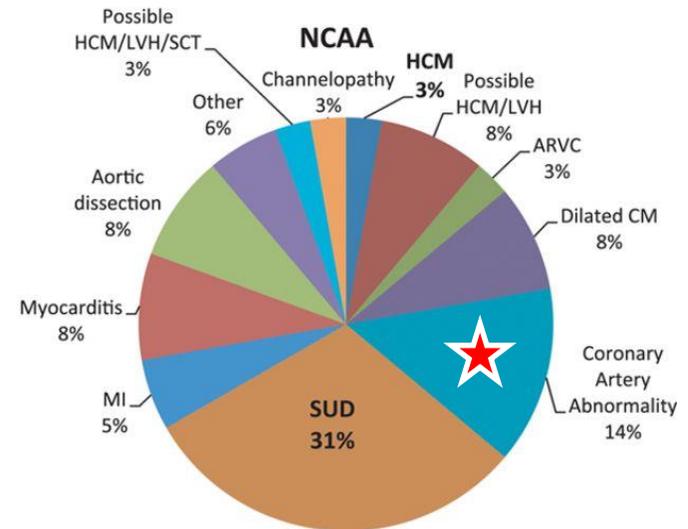
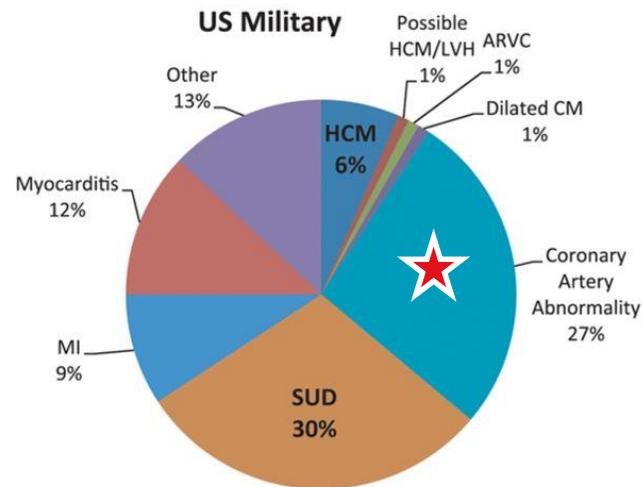
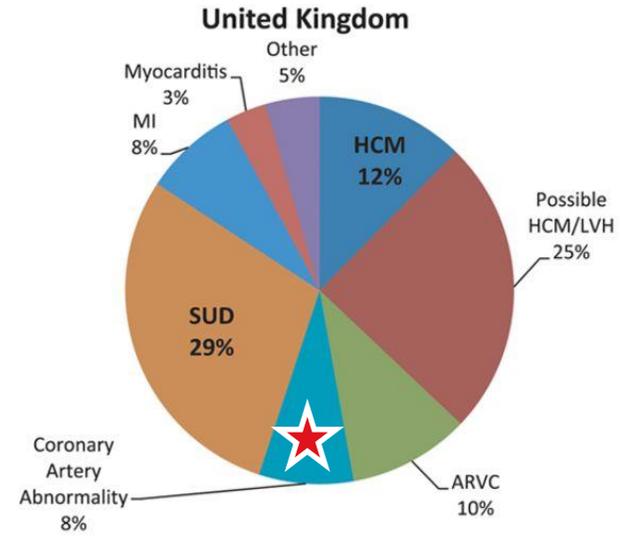
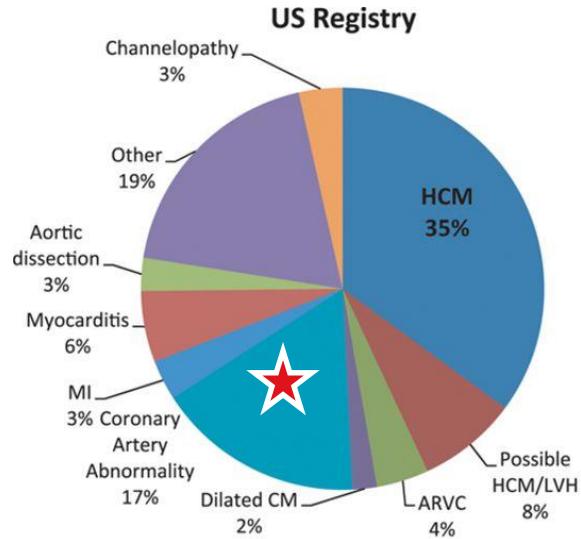


Raisky O, Vouhé P. EMC 2007

Never demonstrated

- Embryologie et anatomie
- Classification
- Prévalence
- Imagerie
- Ischémie myocardique
- **Mort subite**

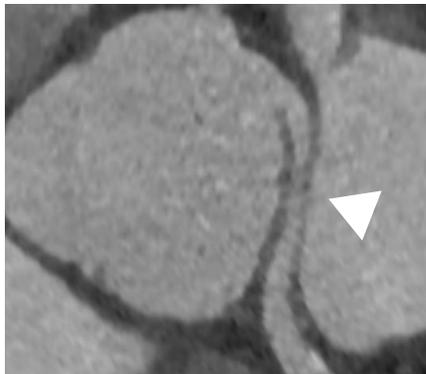
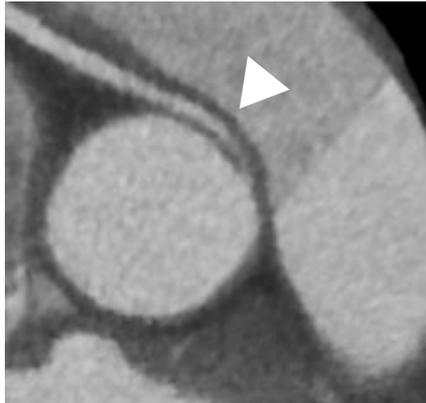
Sudden death
in 0-35 years of age



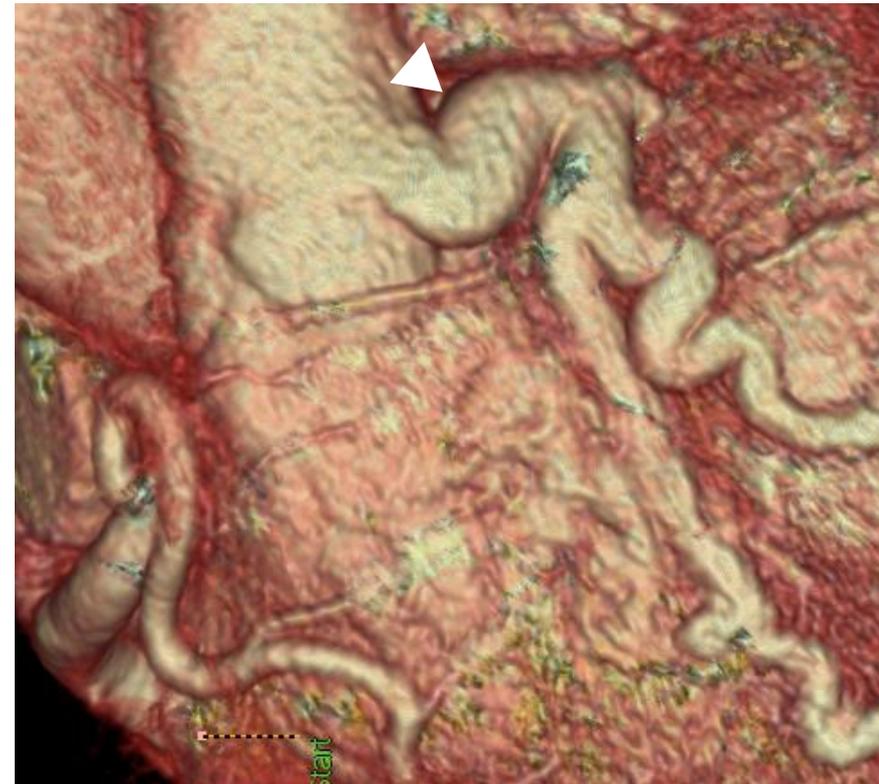
ANOCOR à risque de mort subite

Connexions aortiques

Trajet interartériel



Connexions pulmonaires



Cardiopathie congénitale à risque de mort subite	Prévalence**
ANOCOR* droite	0.3%
Cardiomyopathie hypertrophique	0.2%
Syndrome pré-excitation ventriculaire	0.15%
Syndrome de QT long	0.05%
Cardiomyopathie dilatée idiopathique	0.04%
Dysplasie ventriculaire droite arythmogène	0.04%
ANOCOR* gauche	0.03%
Syndrome de Brugada	0.02%
Tachycardie ventriculaire catécholergique	0.01%

* Anomalie de connexion avec trajet interartériel

** Nombre de cas à la naissance (estimations)

Cardiopathie congénitale à risque de mort subite	Incidence annuelle**
Tachycardie ventriculaire catécholergique	1.5%
Cardiomyopathie hypertrophique	1-2%
Syndrome de Brugada	1%
Syndrome de QT long	0.5-1%
Cardiomyopathie dilatée idiopathique	0.5-1%
Dysplasie ventriculaire droite arythmogène	0.5-1%
ANOCOR* gauche	0.2%
Syndrome pré-excitation ventriculaire	0.1%
ANOCOR* droite	0.02%

* Anomalie de connexion avec trajet interartériel

** Incidence annuelle de mort subite (estimations)

2020 Guidelines for adult congenital heart disease

Anomalous aortic origin of the coronary artery		
Surgery is recommended for AAOCA in patients with typical angina symptoms who present with evidence of stress-induced myocardial ischaemia in a matching territory or high-risk anatomy. ^c	I	C
Surgery is not recommended for AAORCA in asymptomatic patients without myocardial ischaemia and without high-risk anatomy. ^c	III	C

^cHigh-risk anatomy includes features such as an intramural course and orifice anomalies (slit-like orifice, acute-angle take-off, orifice >1 cm above the sinotubular junction).

Surgery should be considered in <i>asymptomatic</i> patients with AAOCA (right or left) and evidence of myocardial ischaemia.	IIa	C
Surgery should be considered in <i>asymptomatic</i> patients with AAOLCA and no evidence of myocardial ischaemia but a high-risk anatomy. ^c	IIa	C
Surgery may be considered for symptomatic patients with AAOCA even if there is no evidence of myocardial ischaemia or high-risk anatomy. ^c	IIb	C
Surgery may be considered for <i>asymptomatic</i> patients with AAOLCA without myocardial ischaemia and without high-risk anatomy ^c when they present at young age (<35 years).	IIb	C

Baumgartner H et al. Eur Heart J. 2020.

Informations dans le compte-rendu de coronarographie

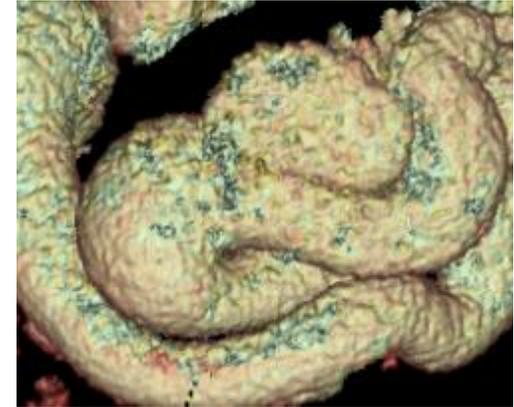
- Connexions anormales proximales
 - artère concernée
 - site de connexion
 - trajet probable par rapport à AO/AP
 - athérome coronaire associé
- Connexions anormales distales
 - artère concernée
 - type proximal (A) ou distal (B)
 - site de connexion
 - taille : petite/intermédiaire/large
 - unique/multiple
 - morphologie (tortuosités/calcifications)
- Trajet intramyocardique
 - artère concernée
 - présent/possible
 - écrasement systolique < ou > 50%

Conclusions

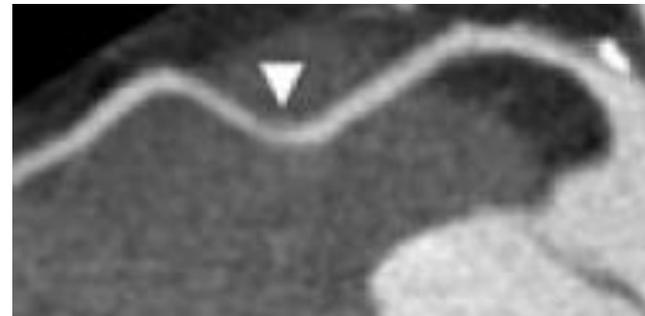


- Connexions anormales proximales
 - nombreuses formes anatomiques
 - trajet interartériel à reconnaître
 - aide de l'imagerie en coupe

- Connexions anormales distales
 - très rares
 - hétérogénéité anatomique
 - retentissement très variable
 - aide de l'imagerie en coupe



- Anomalies sur le trajet
 - ponts musculaires
 - prévalence élevée
 - généralement bénignes



Groupe de travail multidisciplinaire sur les ANOMalies congénitales des artères CORonaires

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Xavier Halna du Fretay (Saran)
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Phalla Ou (Paris)

staff ANOCOR



<http://anocor.fr>

Groupe multidisciplinaire ANOCOR

Anomalies Coronaires Congénitales

Ouverture début 2024

Ce site est destiné aux professionnels de santé et aux patients, ainsi qu'à leur entourage, souhaitant obtenir des informations sur les Anomalies Coronaires Congénitales (ANOCOR). Les formes anatomiques sont très nombreuses, allant de la banale anomalie sans conséquence clinique aux anomalies pouvant être responsables de symptômes cardiaques graves dont l'arrêt cardiaque. Même si les techniques d'imagerie, surtout radiologiques, permettent le diagnostic de ces anomalies rares, leur compréhension reste incomplète et leur prise en charge n'est pas encore parfaitement codifiée. Le site ANOCOR a pour objectifs d'aider les professionnels de santé dans leur démarche de recherche et de transmettre aux patients nos connaissances actuelles. Bonne navigation.