Pronostic et prise en charge des anomalies de connexion et de tra

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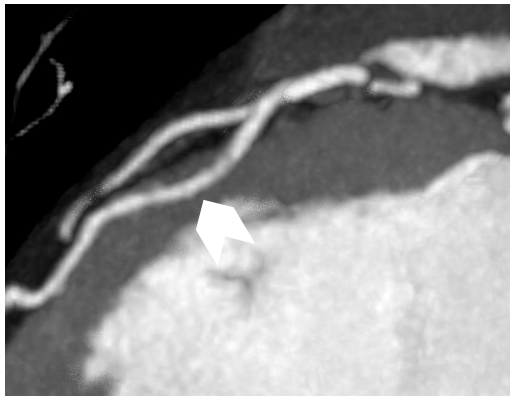
HÔPITAUX UNIVERSITAIRES PARIS NORD VAL DE SEINE Bichat - Claude-Bernard

ASSISTANCE PUBLIQUE HÔPITAUX DE PARIS

Intervenant: Pierre AUBRY

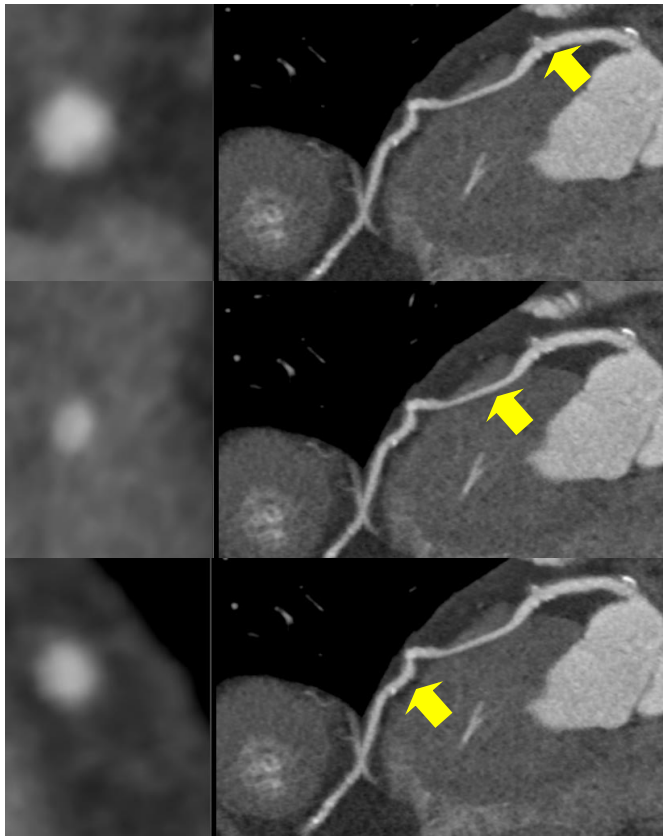
Je n’ai aucun lien d’intérêt

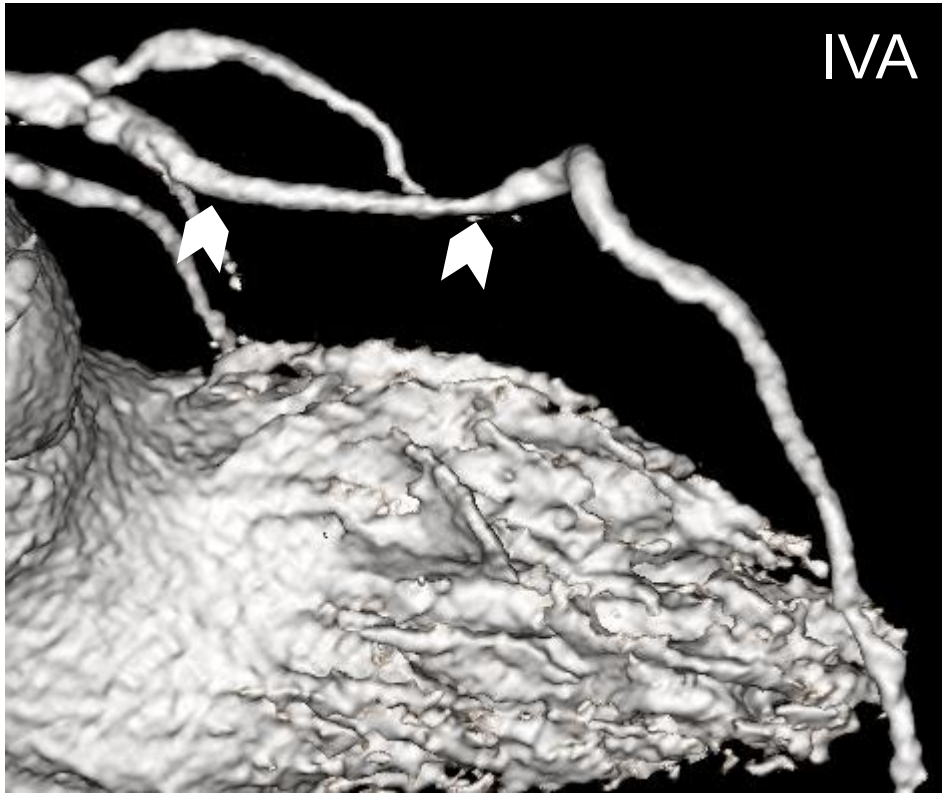


Á Anomalies sur le trajet

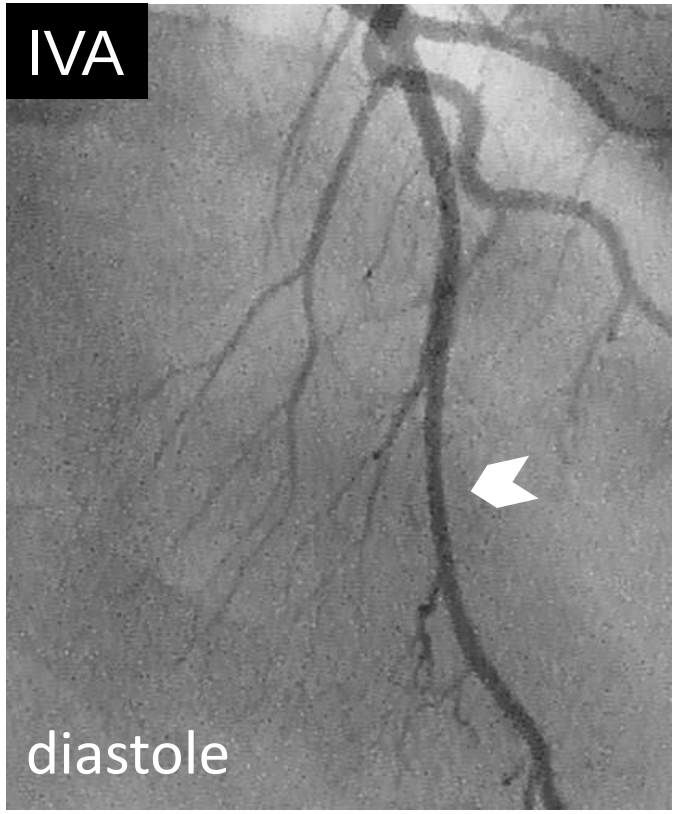
Á Anomalies de connexion

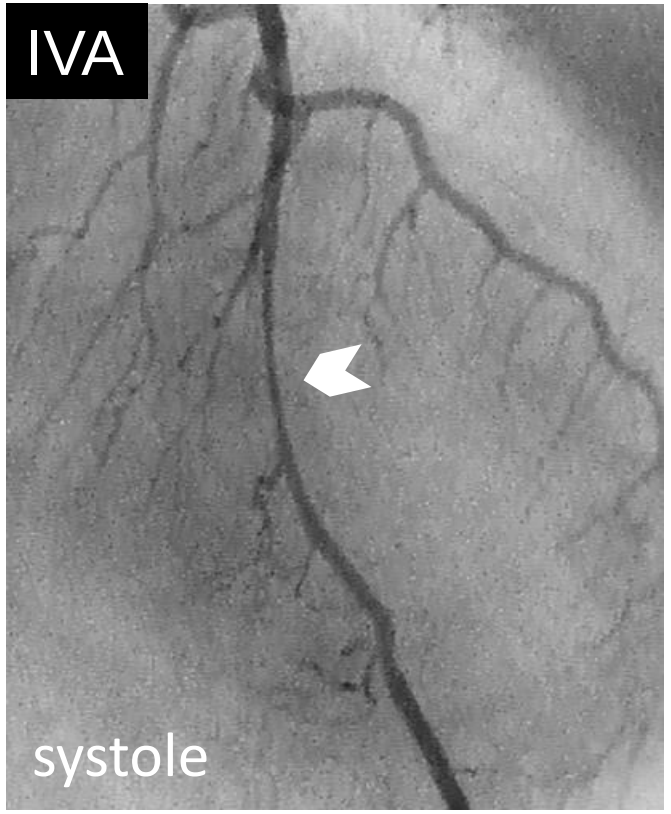
Trajet coronaire intramyocardique



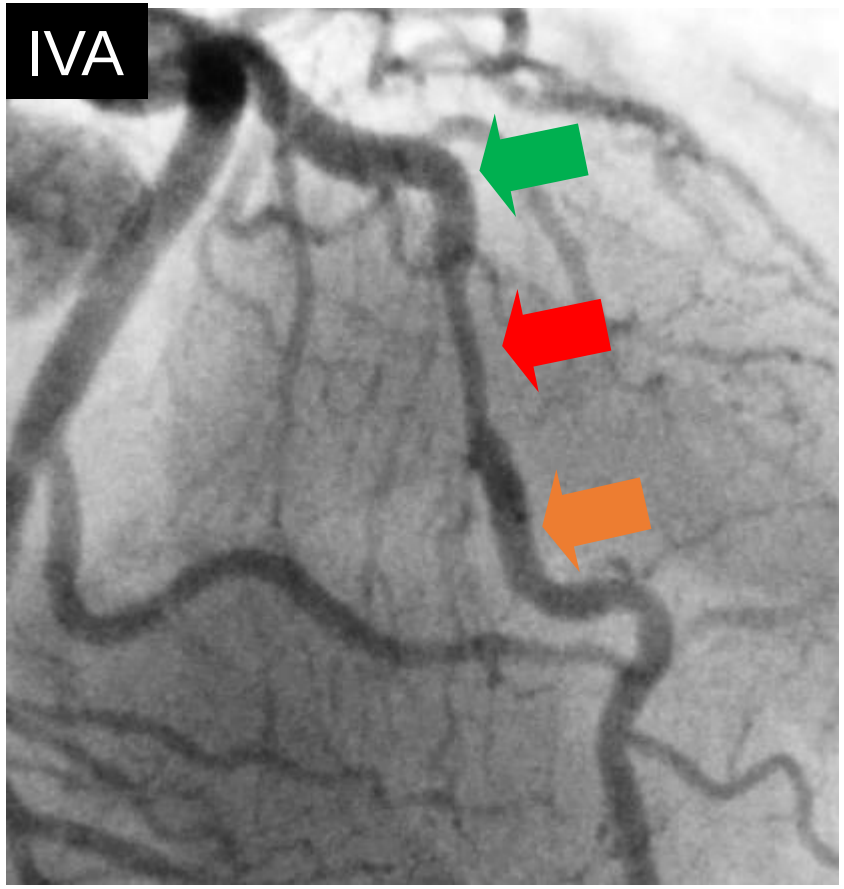


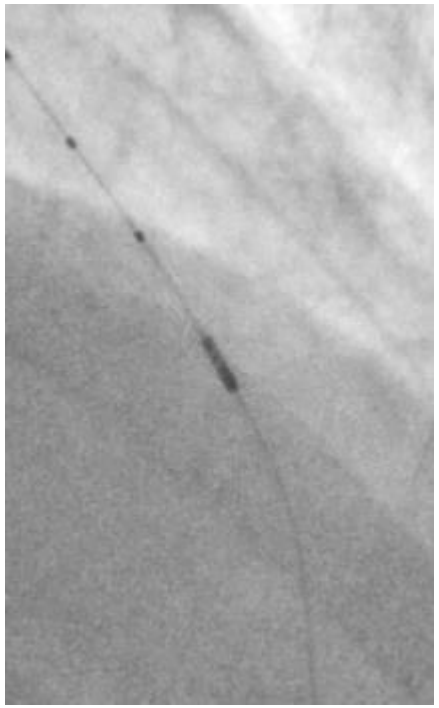
Trajet coronaire intramyocardique

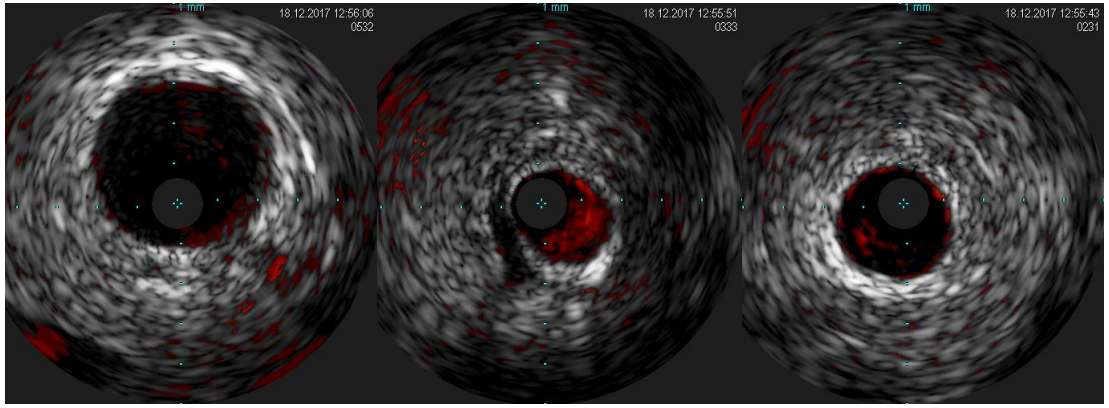




Trajet coronaire intramyocardique







Trajet coronaire intramyocardique

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

situation non rare

lien de causalité ?

Profondeur : ≥ 5 mm
Longueur : ≥ 25 mm

trajet intramyocardique incident

trajet intramyocardique pathologique

- Á Symptômes ischémiques ✓
- Á Ischémie myocardique ✓
- Á Mort subite/arrêt cardiaque ✗

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THE PRESENT AND FUTURE

JACC STATE-OF-THE-ART REVIEW

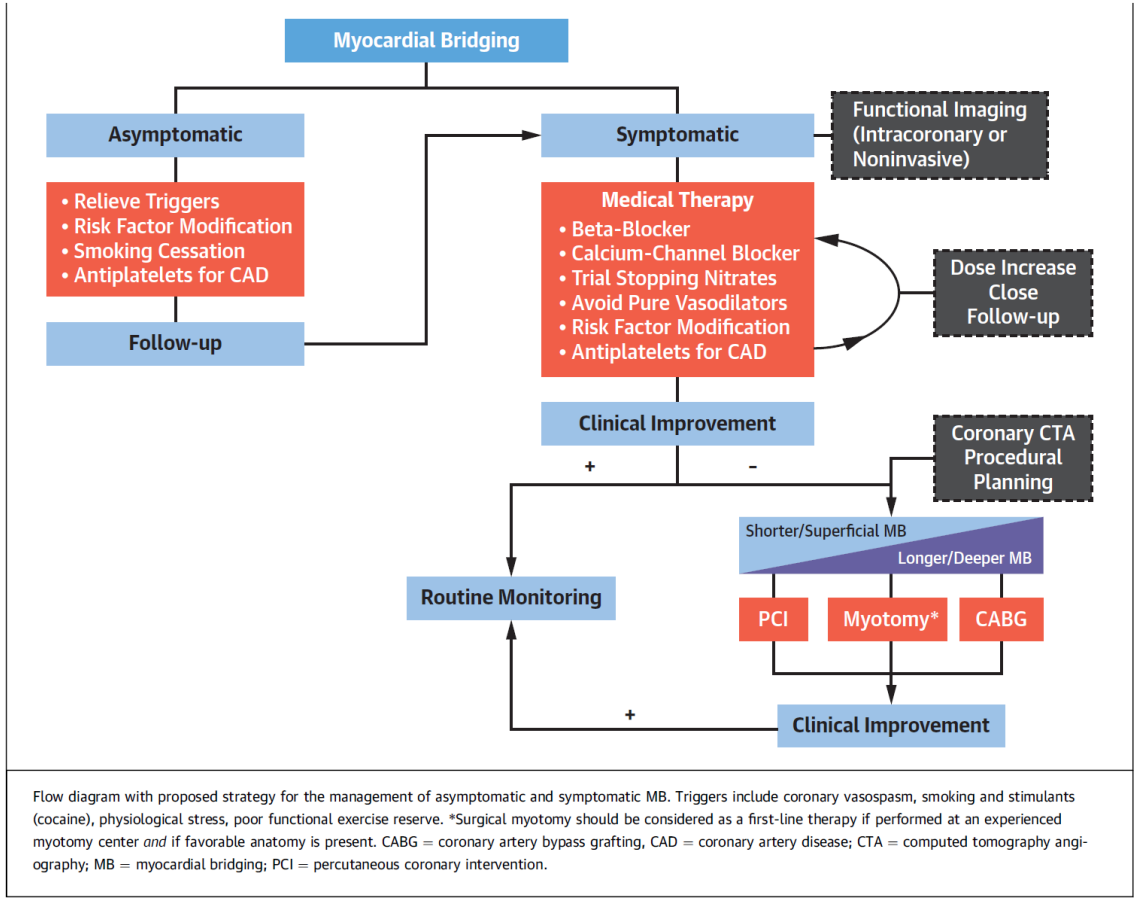
Myocardial Bridging: Diagnosis, Functional Assessment, and Management

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]

Anomalies coronaires congénitales de l'adulte

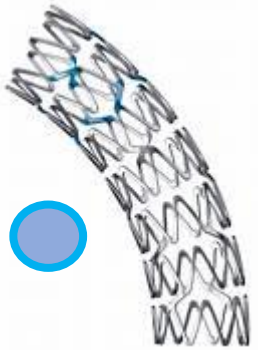
Trajet coronaire intramyocardique

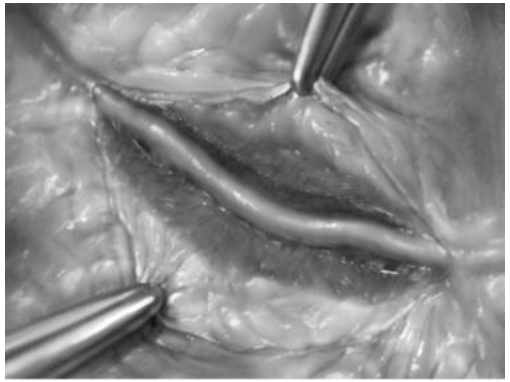


Flow diagram with proposed strategy for the management of asymptomatic and symptomatic MB. Triggers include coronary vasospasm, smoking and stimulants (cocaine), physiological stress, poor functional exercise reserve. *Surgical myotomy should be considered as a first-line therapy if performed at an experienced myotomy center *and* if favorable anatomy is present. CABG = coronary artery bypass grafting, CAD = coronary artery disease; CTA = computed tomography angiography; MB = myocardial bridging; PCI = percutaneous coronary intervention.

Sterheim D et al. JACC 2021

- Adenosine testing
- Acetylcholine testing
- Dobutamine testing

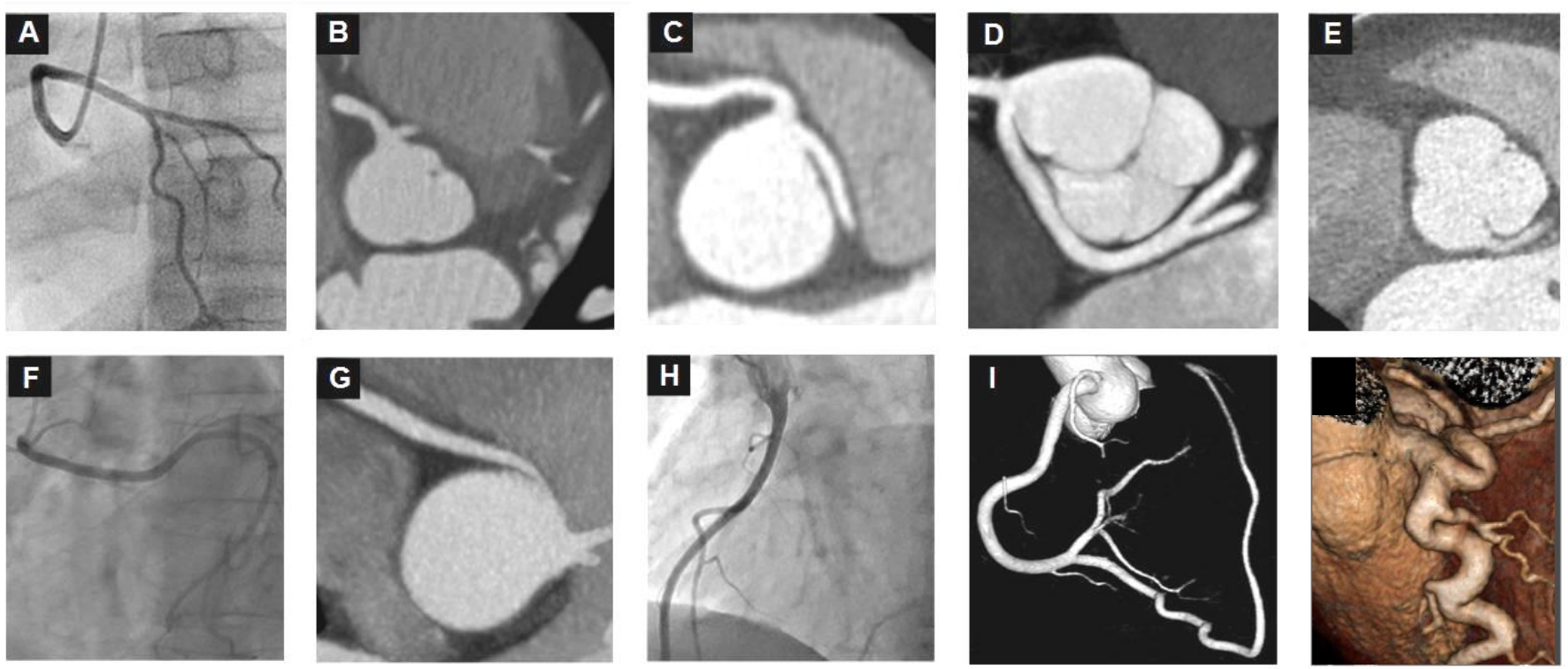




Á Anomalies sur le trajet

Á Anomalies de connexion

Nombreuses formes anatomiques



Pronostic

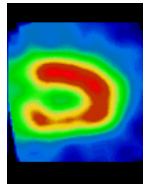
- Symptômes ischémiques
- Syndromes coronariens aigus
- Ischémie myocardique
- Cardiomyopathie ischémique (FEVG/nécrose)
- Arrêt cardiaque récupéré

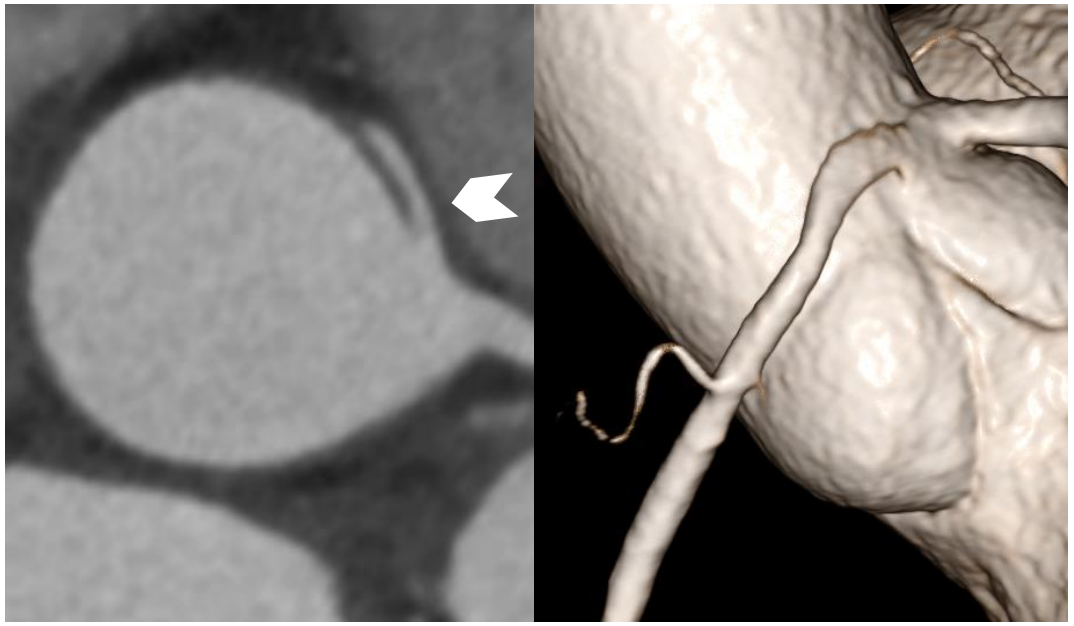
Anomalies de connexion aortique

Pronostic

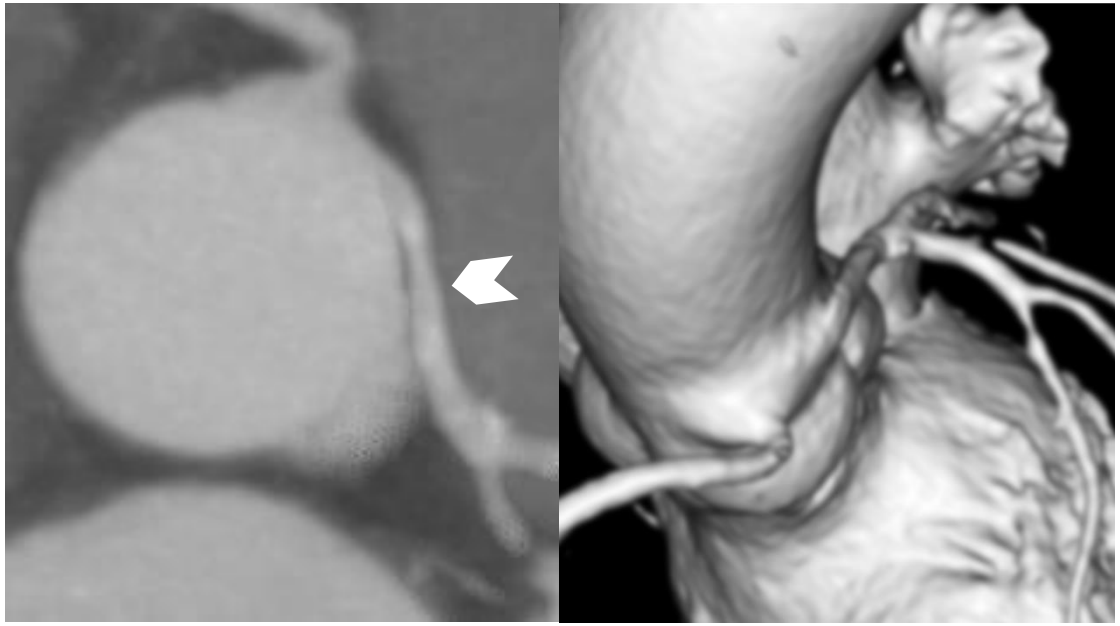
- Á Symptômes ischémiques ✓
- Á Syndromes coronariens aigus ✗
- Á Ischémie myocardique ✓
- Á Cardiomyopathie ischémique (FEVG/nécrose) ✓
- Á Arrêt cardiaque récupéré ✓

Anomalies de connexion aortique à risque



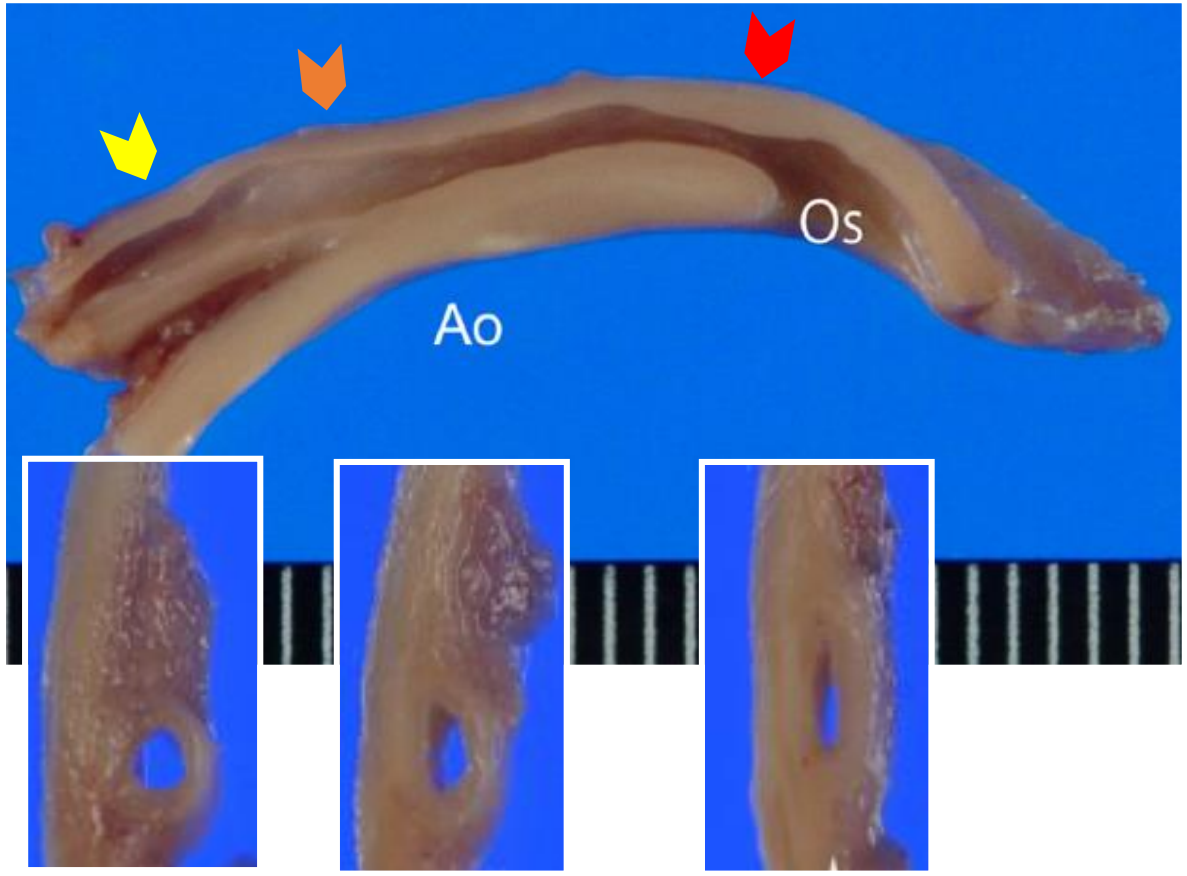


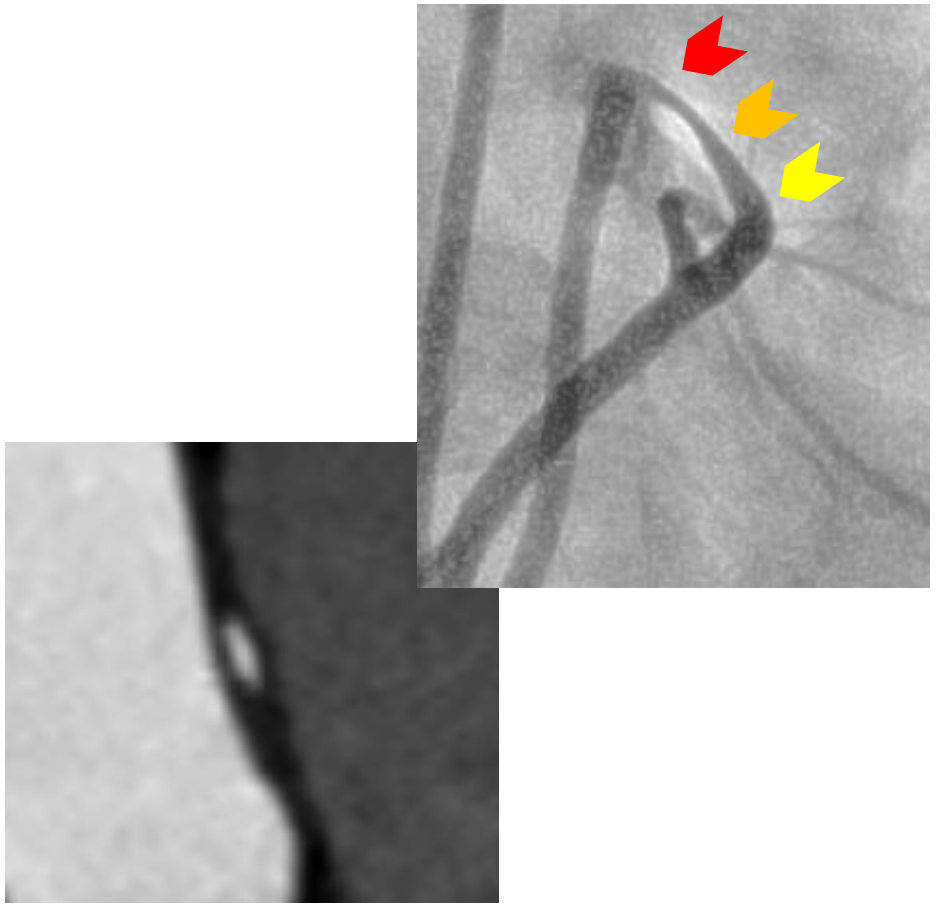
ANOCOR droite interartérielle



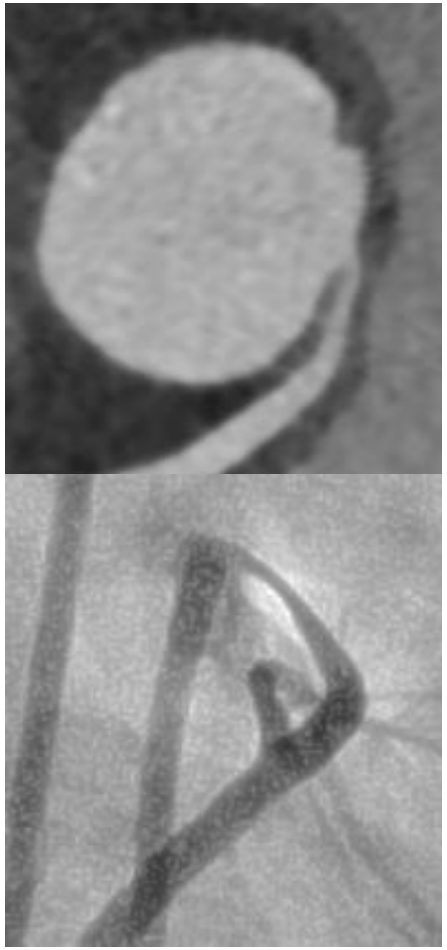
ANOCOR gauche interartérielle

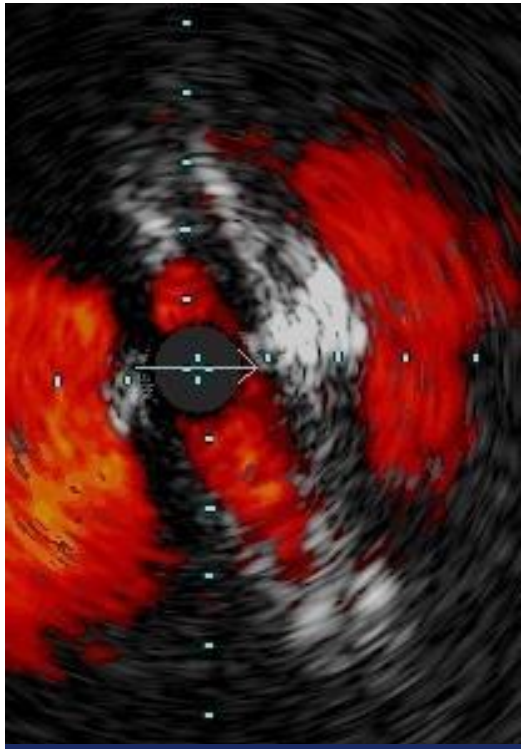
Passage intramural aortique



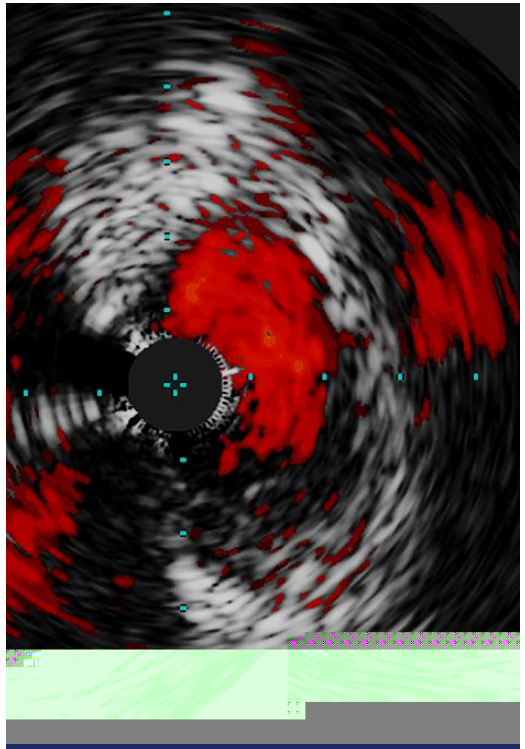


Anomalies coronaires congénitales de l'adulte

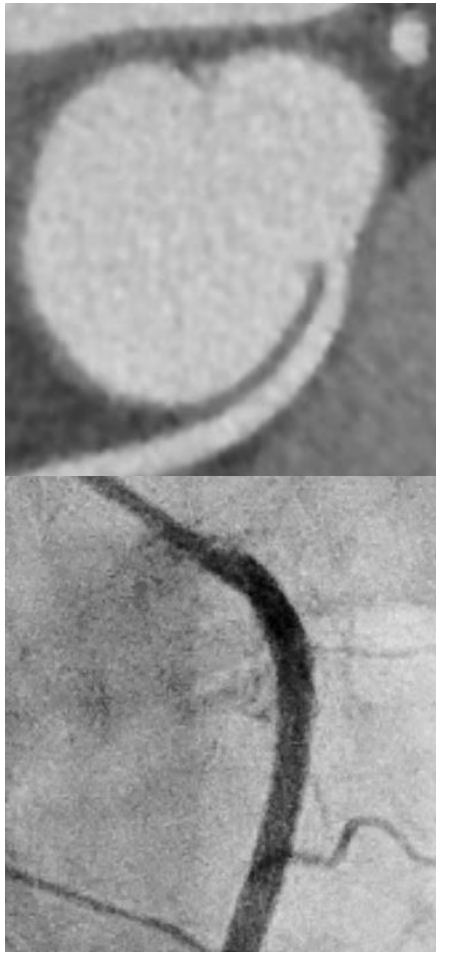




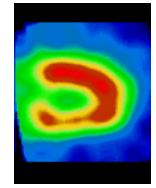
Passage intramural +

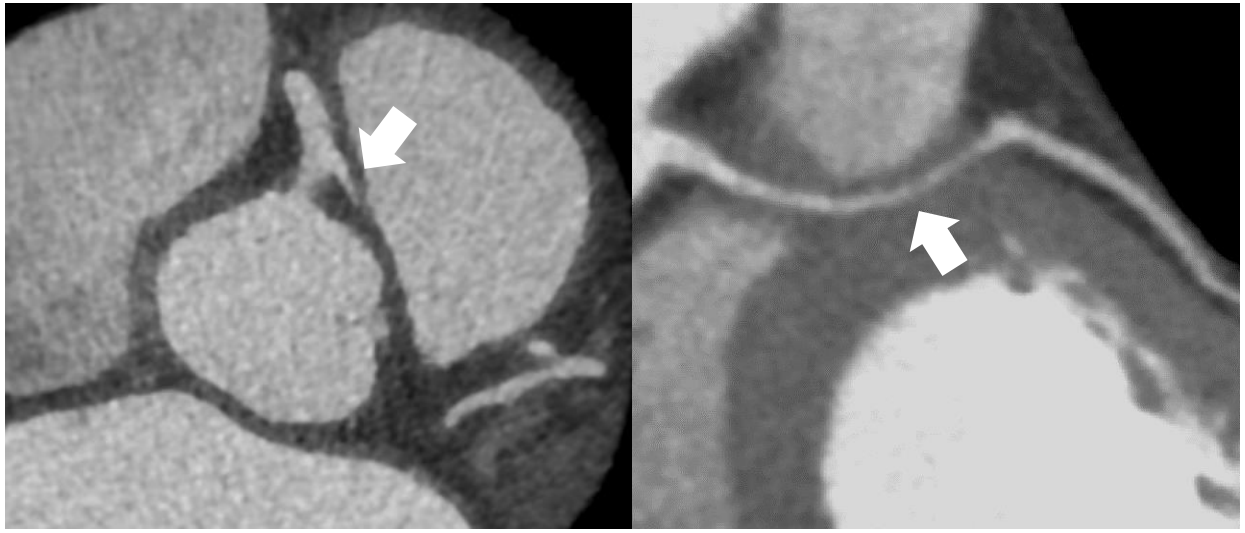


Passage intramural -

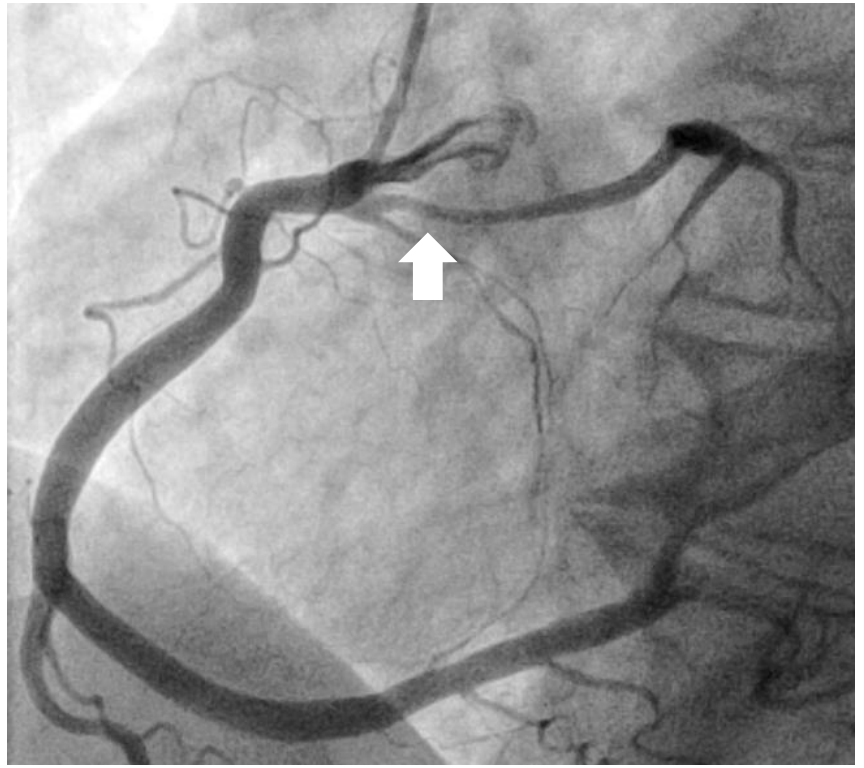


Anomalies de connexion aortique à risque

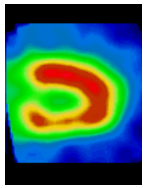




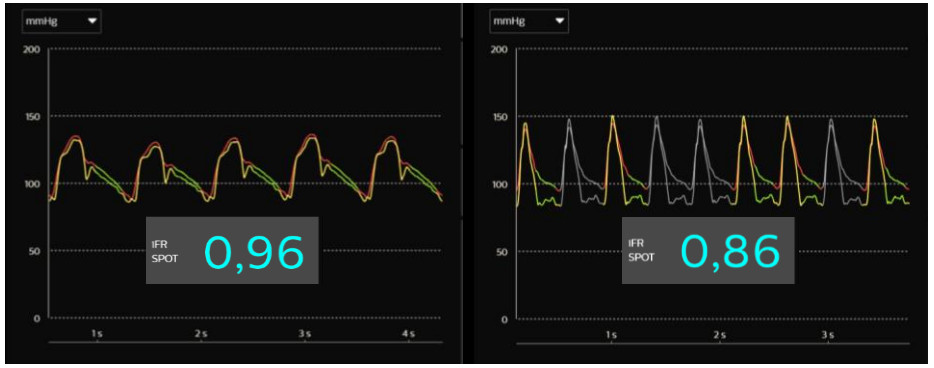
ANOCOR gauche rétropulmonaire

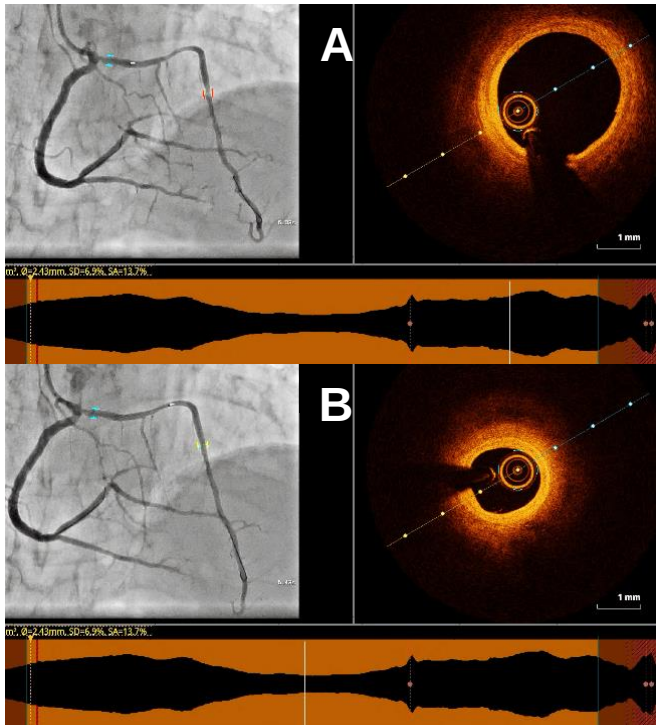


Anomalies de connexion aortique à risque



ANOCOR gauche rétropulmonaire



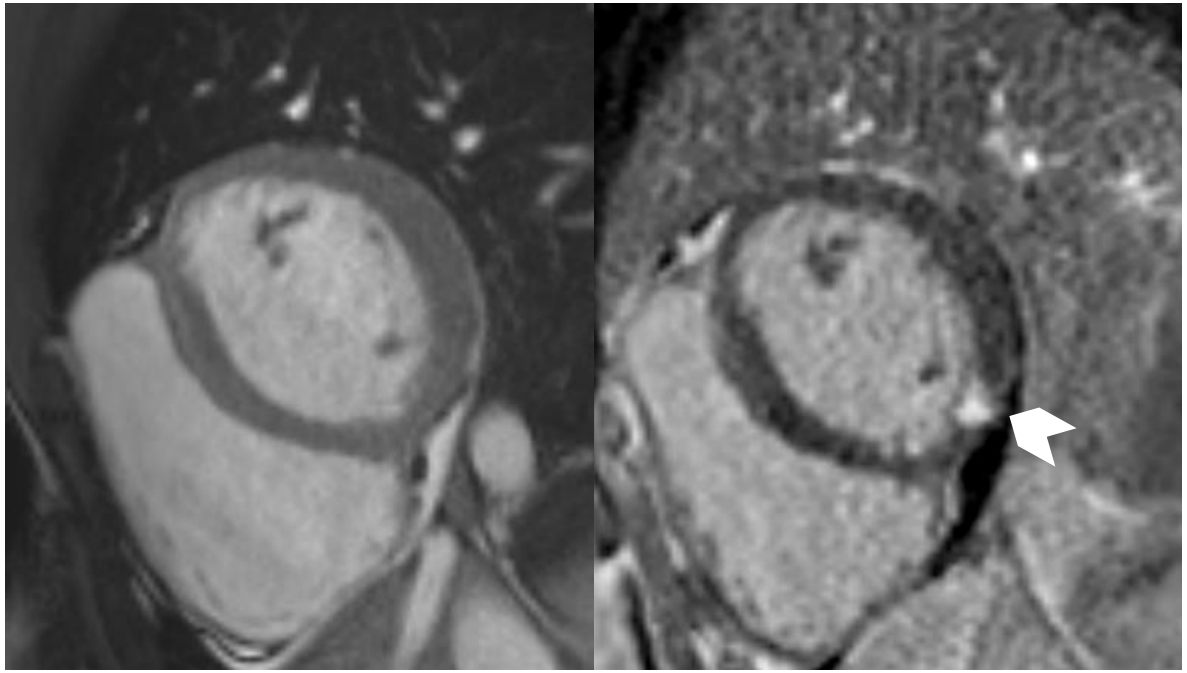


Anomalies de connexion aortique non à risque

ANOCOR prépulmonaire

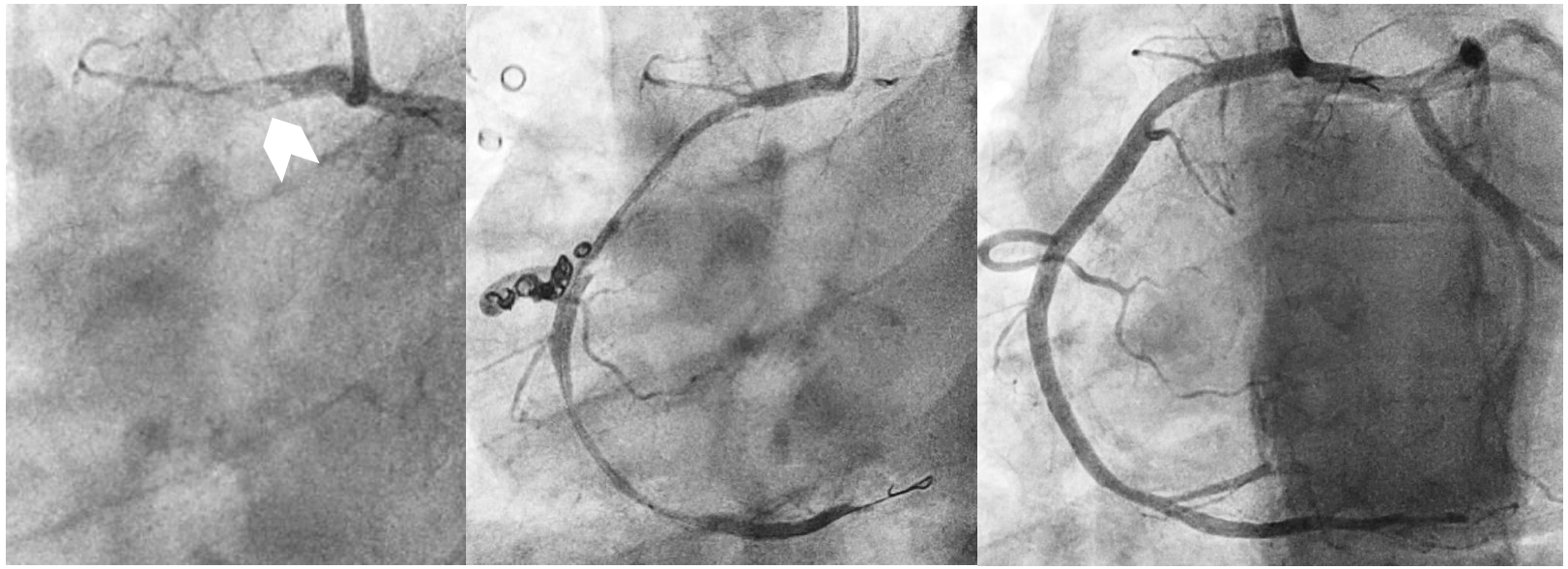
ANOCOR rétroaortique

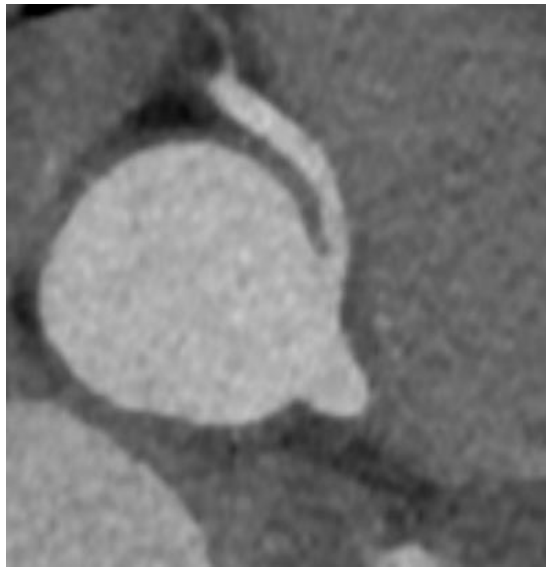
Place de l'IRM cardiaque



Lien de causalité avec présentation initiale

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





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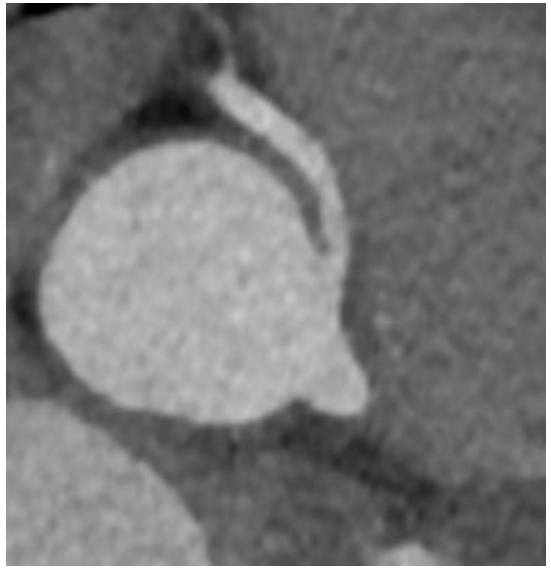
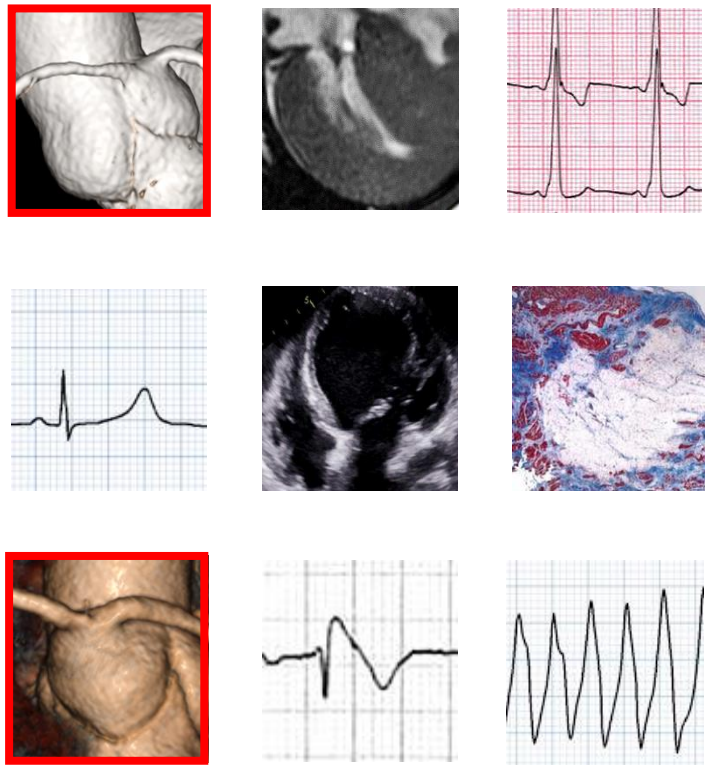


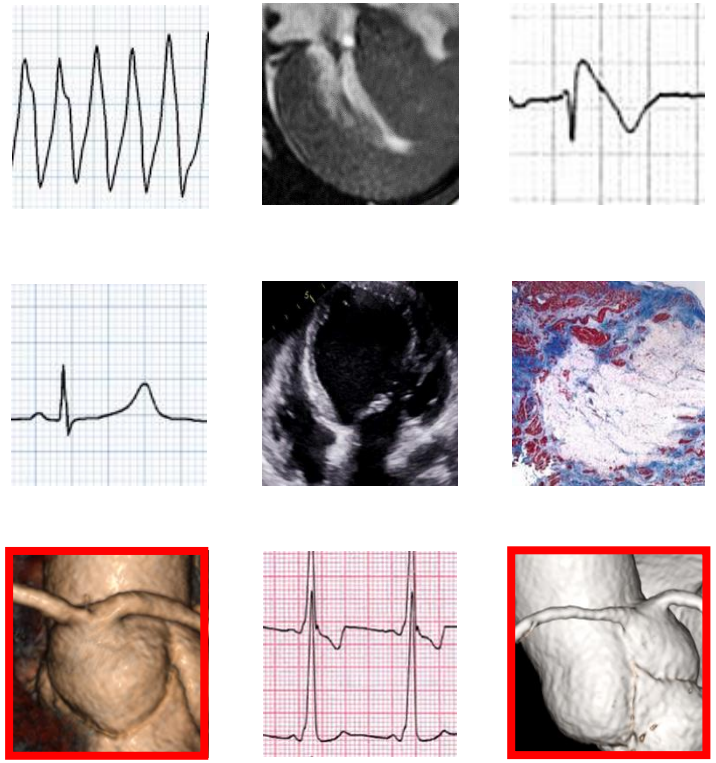
Toujours vérifier le lien de causalité

Cardiopathies congénitales à risque



Cardiopathie	Prévalence*
ANOCOR* droite avec trajet interartériel	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 avec trajet interartériel	0.03%
Syndrome de Brugada	0.02%
Tachycardie ventriculaire catécholergique	0.01%

*Prévalence de l'anomalie (estimations)



Cardiopathies congénitales à risque

Cardiopathie	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 avec trajet interartériel	0.2%
Syndrome pré-excitation ventriculaire	0.1%
ANOCOR droite avec trajet interartériel	0.02

*Incidence annuelle de mort subite (estimations)

:

1. Studies suggest that ARCA is one of the more common types of AAOCA.
2. The incidence of SCD associated with ARCA has been reported to be relatively low, but exact figures may vary.

:

1. ALCAPA, also known as Bland-White-Garland syndrome, is a rare but serious congenital anomaly.
2. SCD is a recognized complication of untreated ALCAPA, particularly in infants, where it can occur early in life due to myocardial ischemia.

OpenAI GPT-3

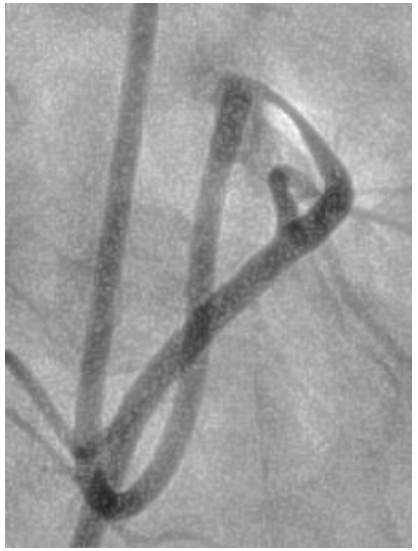
:

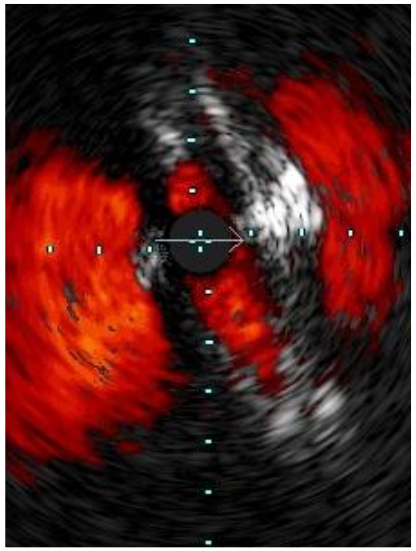
1. The incidence of SCD associated with ALCA can vary.
2. Some studies have suggested that ALCA may carry a higher risk of SCD compared to other types of AAOCA, especially when the anomalous artery takes an interarterial course between the aorta and pulmonary artery.

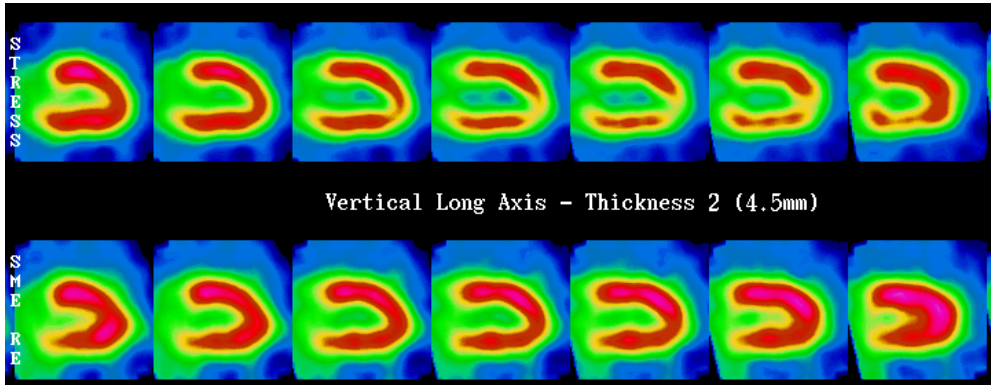
:

1. Incidence rates of SCD for less common variants of AAOCA may be less well-documented.
2. The risk of SCD in these cases may depend on factors such as the specific anatomical course of the anomalous artery and the presence of associated cardiovascular abnormalities.

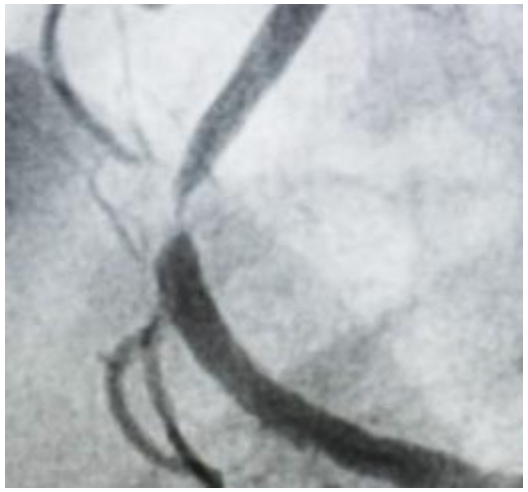
Ischémie myocardique documentée

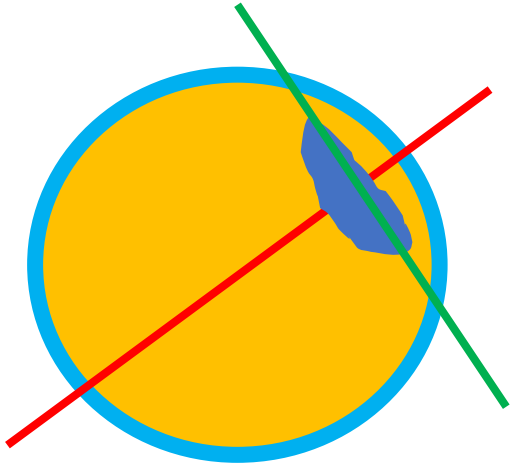




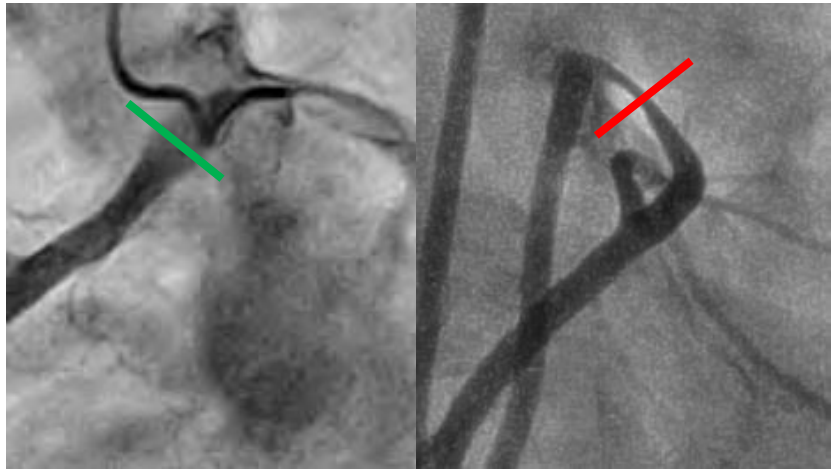


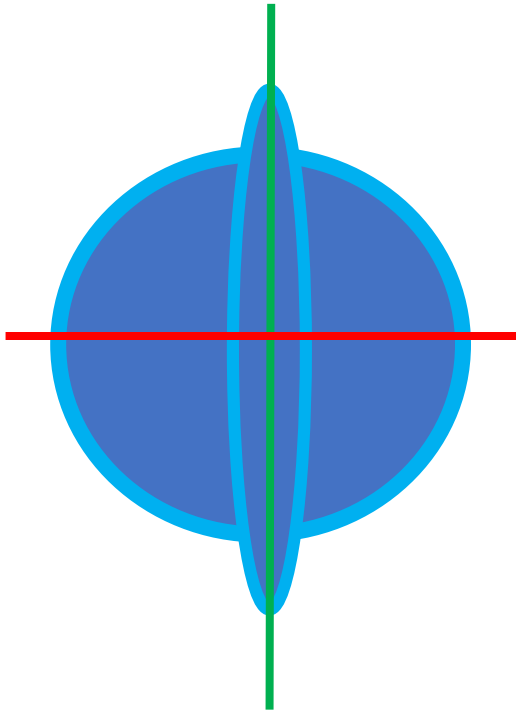
Prévalence ≈ 15%





Réduction de surface : > 70%



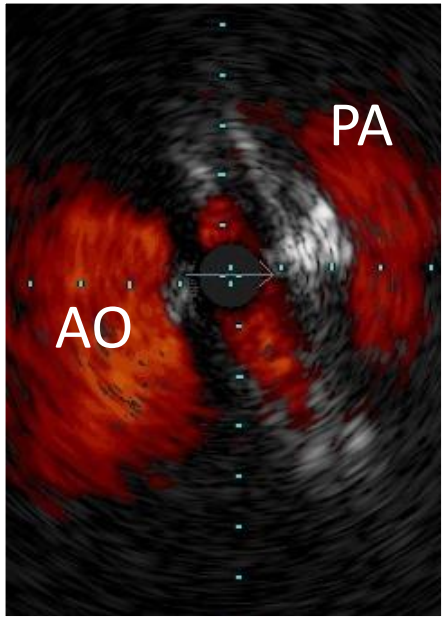


Réduction de surface: 35-70%

Two-Tier Concept

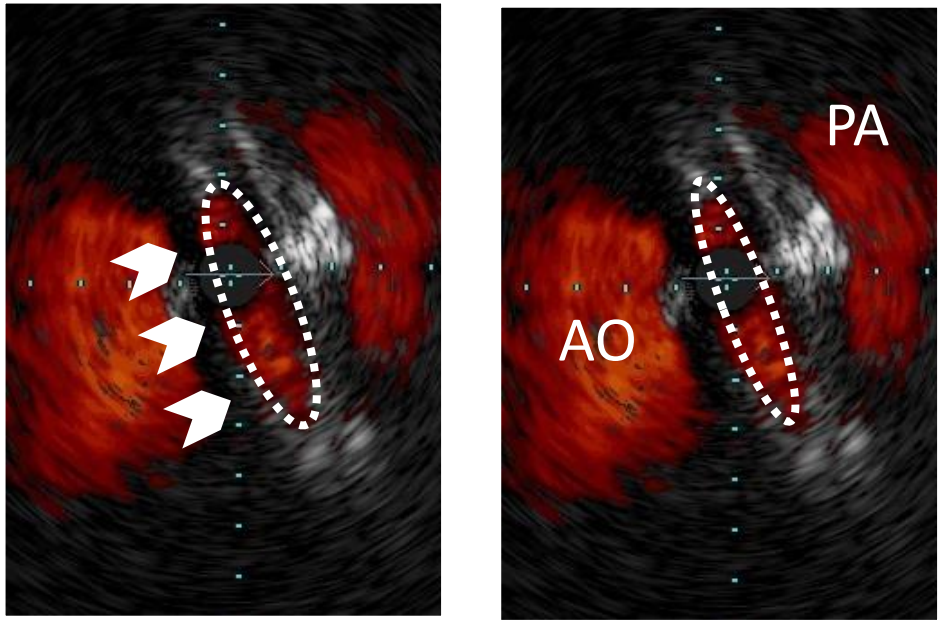
Bigler MR. et al. Front Cardiovasc Med. 2021.

Fixed Component



Reduction in lumen area : 35-70%

Dynamic Component



Strenuous Exertion

Wall stress ⬆

Reduction in lumen area : > 70%

Bigler MR. et al. Front Cardiovasc Med. 2021.

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.

ESC European Society of Cardiology
European Heart Journal (2022) **43**, 3997–4126
https://doi.org/10.1093/eurheartj/ehac262

ESC GUIDELINES

2022 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

Developed by the task force for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death of the European Society of Cardiology (ESC)

Endorsed by the Association for European Paediatric and Congenital Cardiology (AEPC)

Zeppenfeld K. Eur Heart J. 2022.

Recommendation Table 25 — Recommendations for sudden cardiac death prevention in patients with coronary anomalies

Recommendations	Class[a]	Level[b]
Treatment		
Surgery is recommended in patients with anomalous aortic origin of a coronary artery with CA, syncope suspected to be due to VAs, or angina when other causes have been excluded.[585,586,588]	I	C
Surgery should be considered in asymptomatic patients with anomalous aortic origin of a coronary artery and evidence of myocardial ischaemia or abnormal aortic origin of the left coronary artery with high-risk anatomy.[c,585,586,588]	IIa	C

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CA, cardiac arrest; VA, ventricular arrhythmia.
[a]Class of recommendation.
[b]Level of evidence.
[c]High-risk anatomy is defined as interarterial course, slit-like shaped orifice, high orifice, acute-angle take-off, and intramural course and its length.

2020 Guidelines for Adult Congenital Heart Disease

ESC European Society of Cardiology

Anomalous aortic origin of the coronary artery

Surgery is recommended for AAOCA in

patients with typical angina symptoms who

Baumgartner H et al. Eur Heart J. 2020.

Paramètres décisionnels

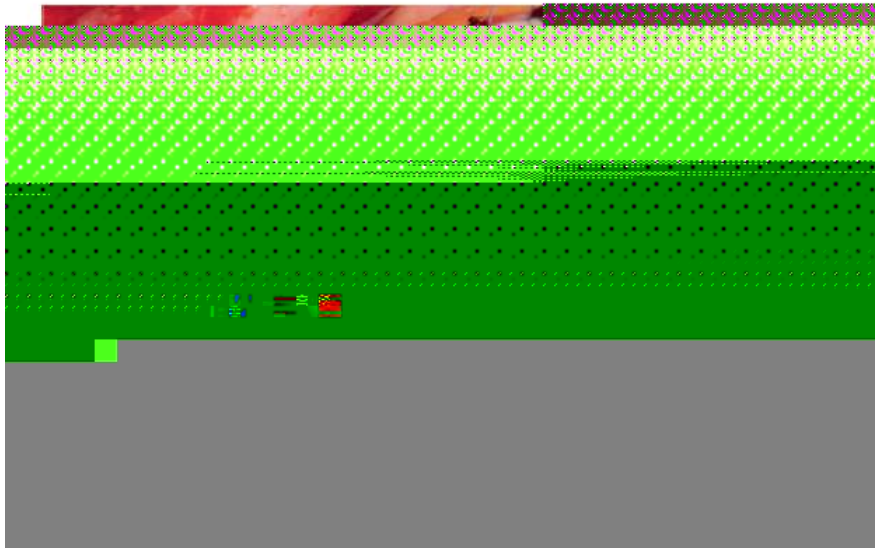
Stratification

‡ Arrêt cardiaque récupéré
‡ Age < 35 ans
‡ Symptomatologie d'allure ischémique
‡ Ischémie myocardique documentée avec imagerie
‡ Trajet interartériel
‡ Formes gauches/formes droites
‡ Passage intramural aortique
‡ Activité physique sportive d'intensité élevée
‡ Préférence du patient

...

Gaudino M. et al. Ann Thorac Surg. 2023.

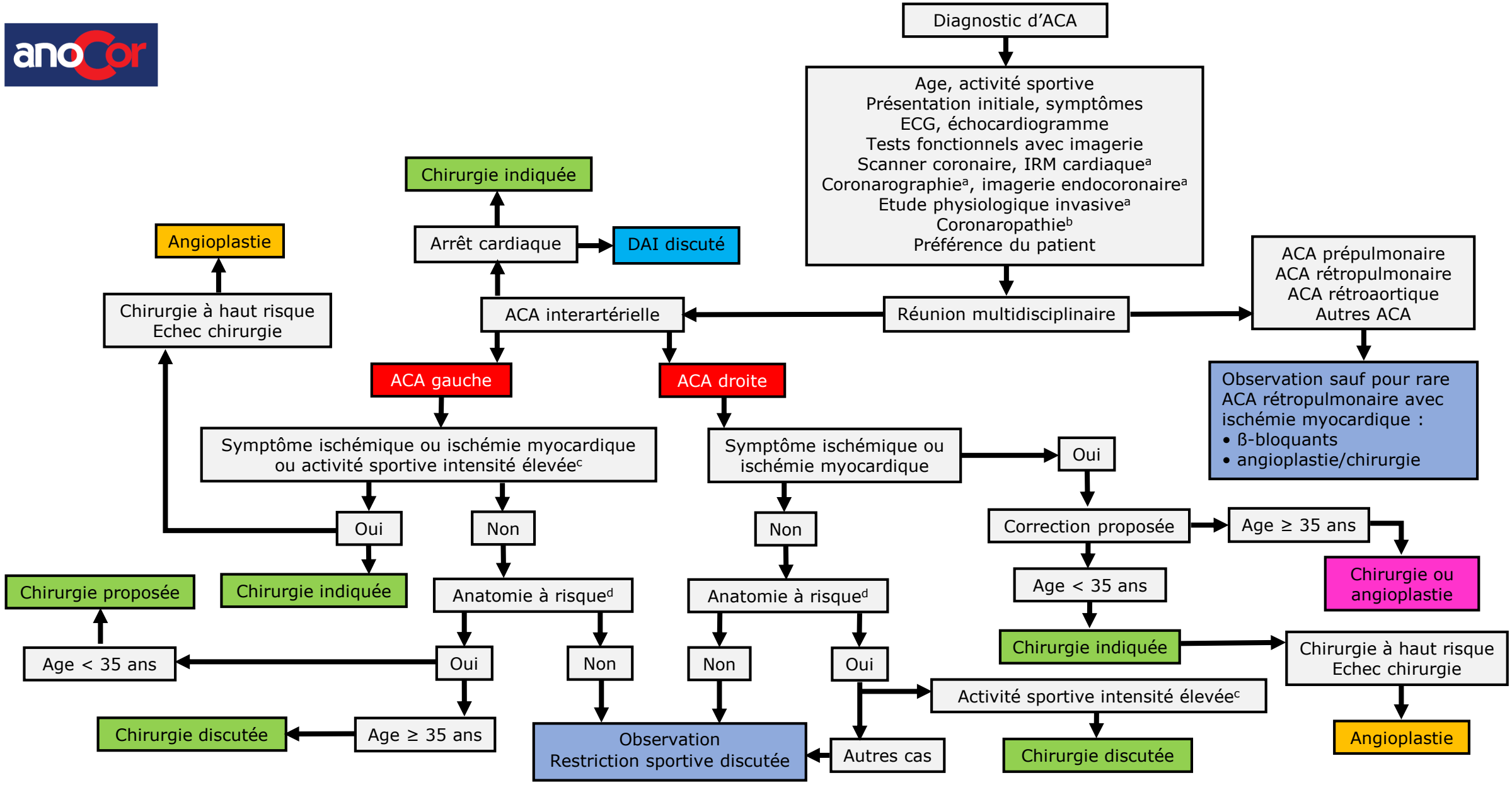
Unroofing technique



Molossi S. et al. MD Cardiovasc J. 2019.

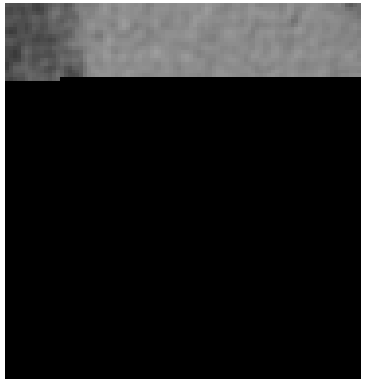
Création nouvel ostium

Vouhé P. Bulletin Académie Nationale Médecine. 2014.



[a] Optionnel.
[b] Stratégie de revascularisation à adapter en cas de maladie coronaire associée.
[c] Activité sportive d'intensité élevée selon la classification des recommandations européennes.
[d] Angle de connexion < 45, forme elliptique de l'ostium, réduction de diamètre ≥ 50% ou passage intramural aortique.
ACA : anomalie de connexion aortique. ANOCOR : anomalies coronaires congénitales. DAI : défibrillateur automatique implantable. ECG : électrocardiogramme. IRM : imagerie par résonance magnétique.

Angioplastie *de novo*



64 ans/vélo/lipothymie d'effort/ANOCOR droite/test d'effort +

Angioplastie post chirurgie

55 ans/ectasie aorte/ANOCOR droite/RVA + Bentall/réimplantation coronaire

Groupe de travail multidisciplinaire sur les anomalies coronaires congénitales

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