

ANOMalies CORonaires congénitales (ANOCOR)

Pierre Aubry

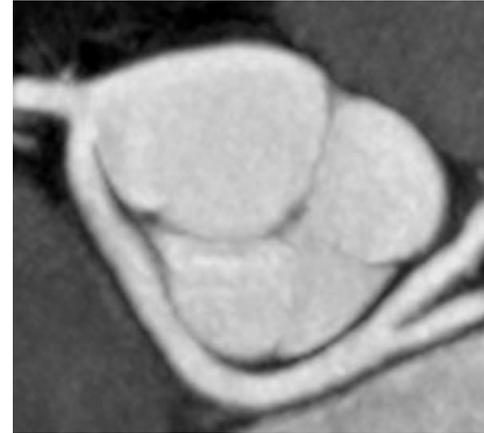
Hôpital Bichat, Paris 75018

Centre Hospitalier, Gonesse 95500

ANOMALIES CORONAIRES CONGENITALES

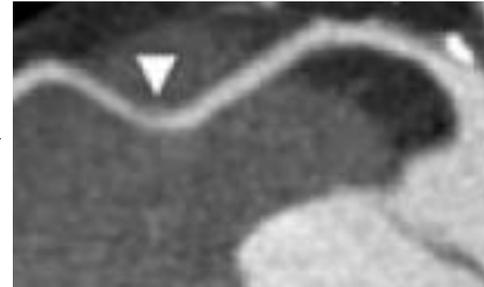


Connexion proximale



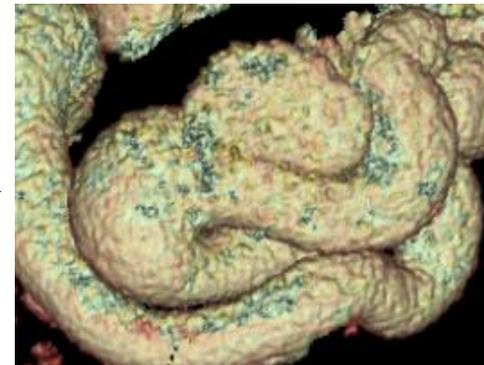
Connexion aortique anormale

Trajet



Trajet intramyocardique

Terminaison



Fistule coronaire

Groupe ANOCOR

Groupe de travail multidisciplinaire sur les ANOMalies CORonaires congénitales

Contact : pcaubry@yahoo.fr

2010 - ...

CA Pierre Aubry (Paris)

CA Olivier Boudvillain (Paris)

CA Patrick Dupouy (Melun)

CA Xavier Halna du Fretay (Saran)

CA Athanasios Koutsoukis (Le Plessis Robinson)

RA Jean-Pierre Laissy (Gonesse)

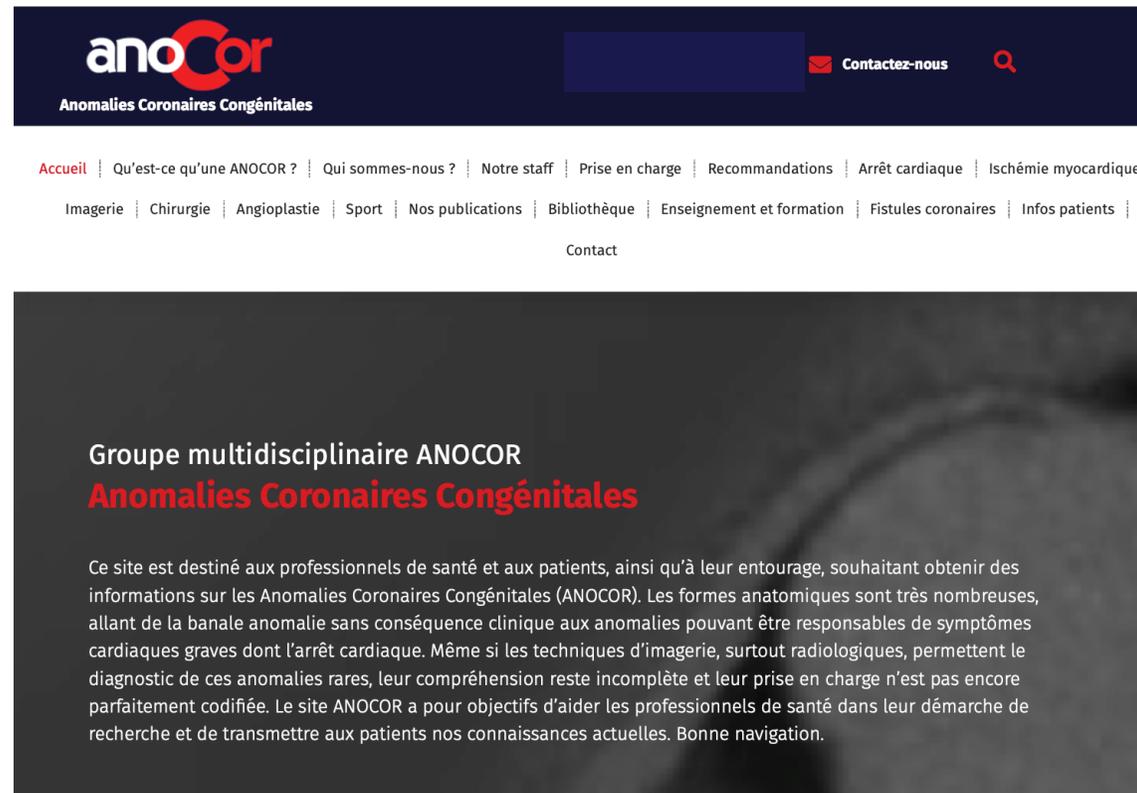
RA Phalla Ou (Paris)

CH

Groupe ANOCOR

Groupe de travail multidisciplinaire sur les ANOMalies CORonaires congénitales

Contact : pcaubry@yahoo.fr



Site ANOCOR

<https://www.anocor.fr>



Groupe ANOCOR

Groupe de travail multidisciplinaire sur les ANOMalies CORonaires congénitales

Staffs

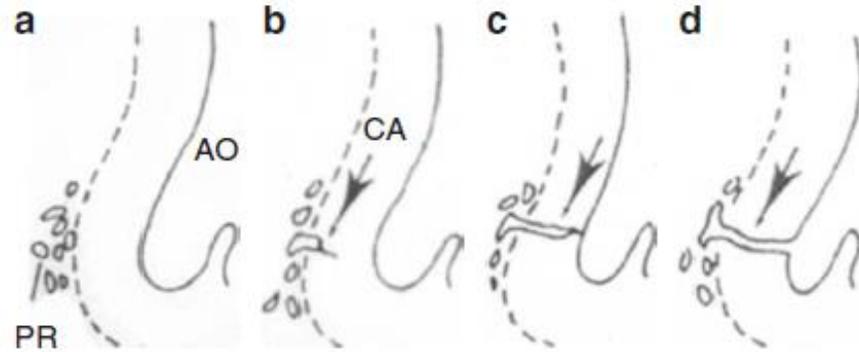
- Récupération des éléments du dossier
- Analyse du dossier
- Avis adressé au cardiologue référent
- Consultation avec le patient (exceptionnelle)

≈ 1.600 dossiers (2010-2024)

- Embryologie et anatomie
- Classification
- Prévalence
- Imagerie
- Ischémie myocardique
- Mort subite
- Dépistage
- Prise en charge
- Chirurgie
- Angioplastie
- Activités sportives

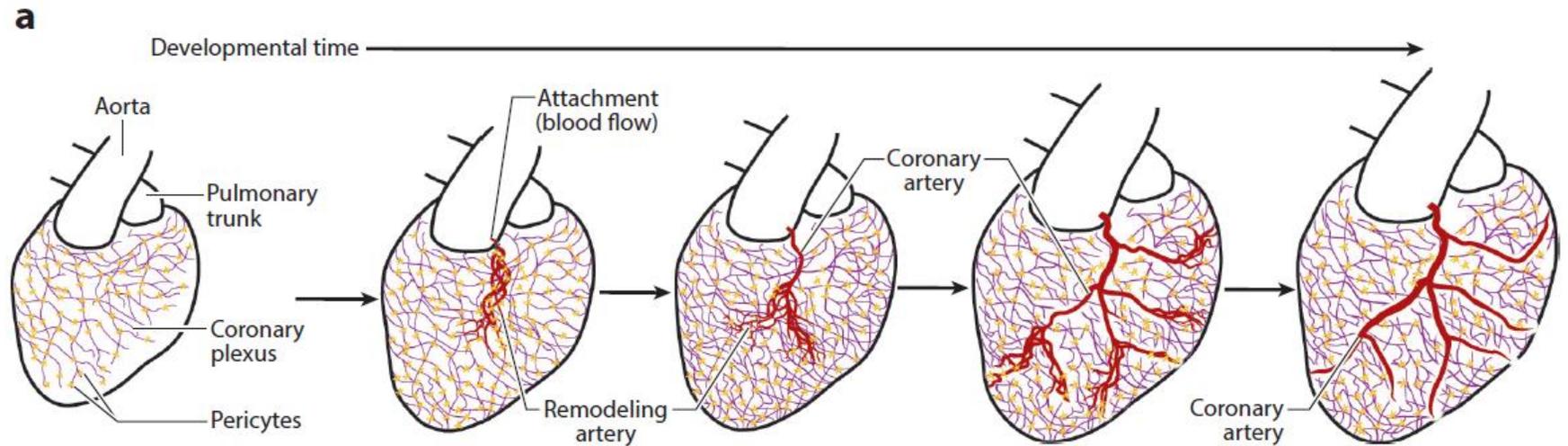
Embryologie et anatomie

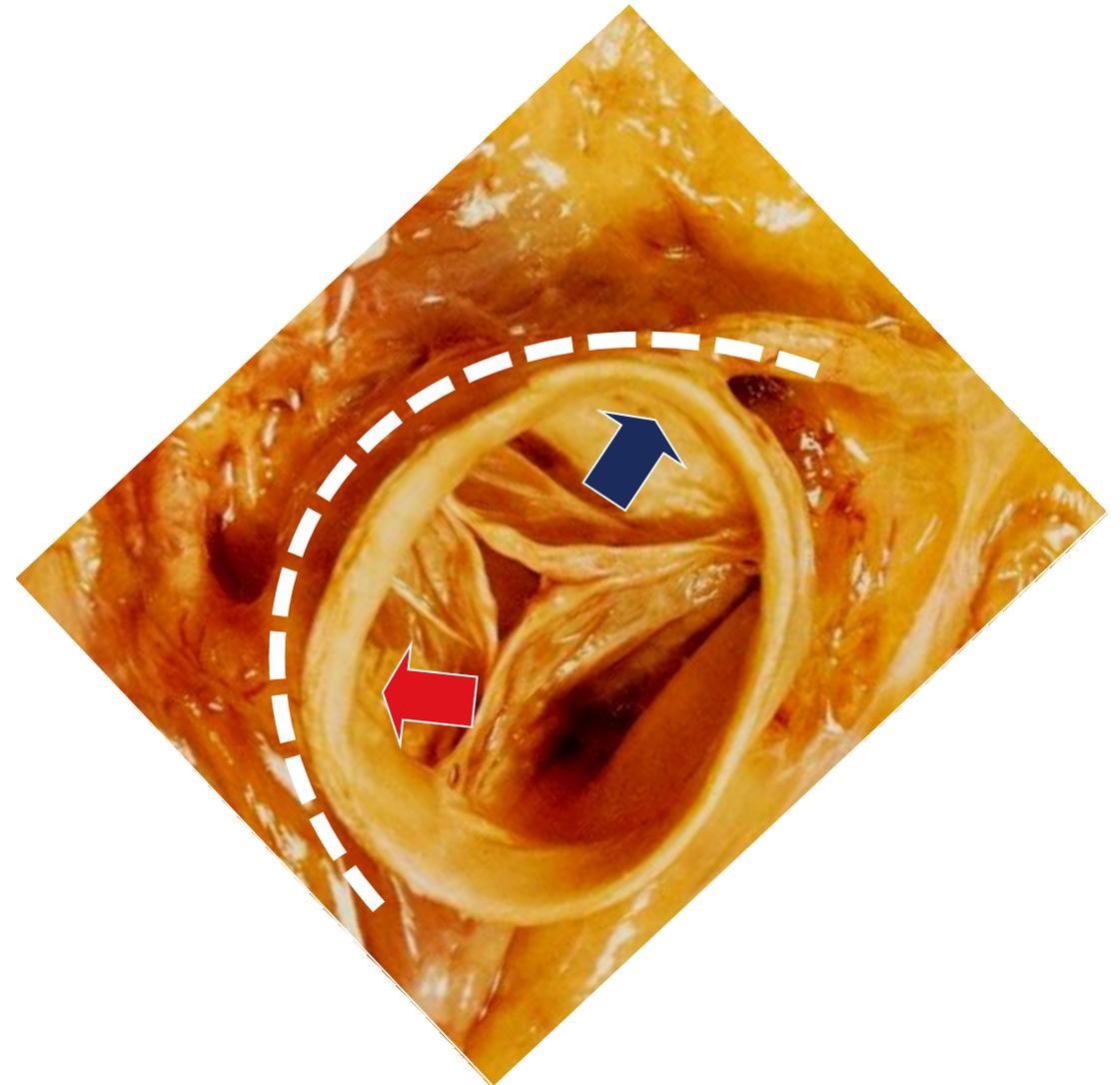
Fig. 2.9 Ingrowth Developmental Hypothesis with cells of the peritruncal ring penetrating the aortic wall. (a–e) show the progression of the ingrowth of epicardial cells through the aortic wall until the endocardium



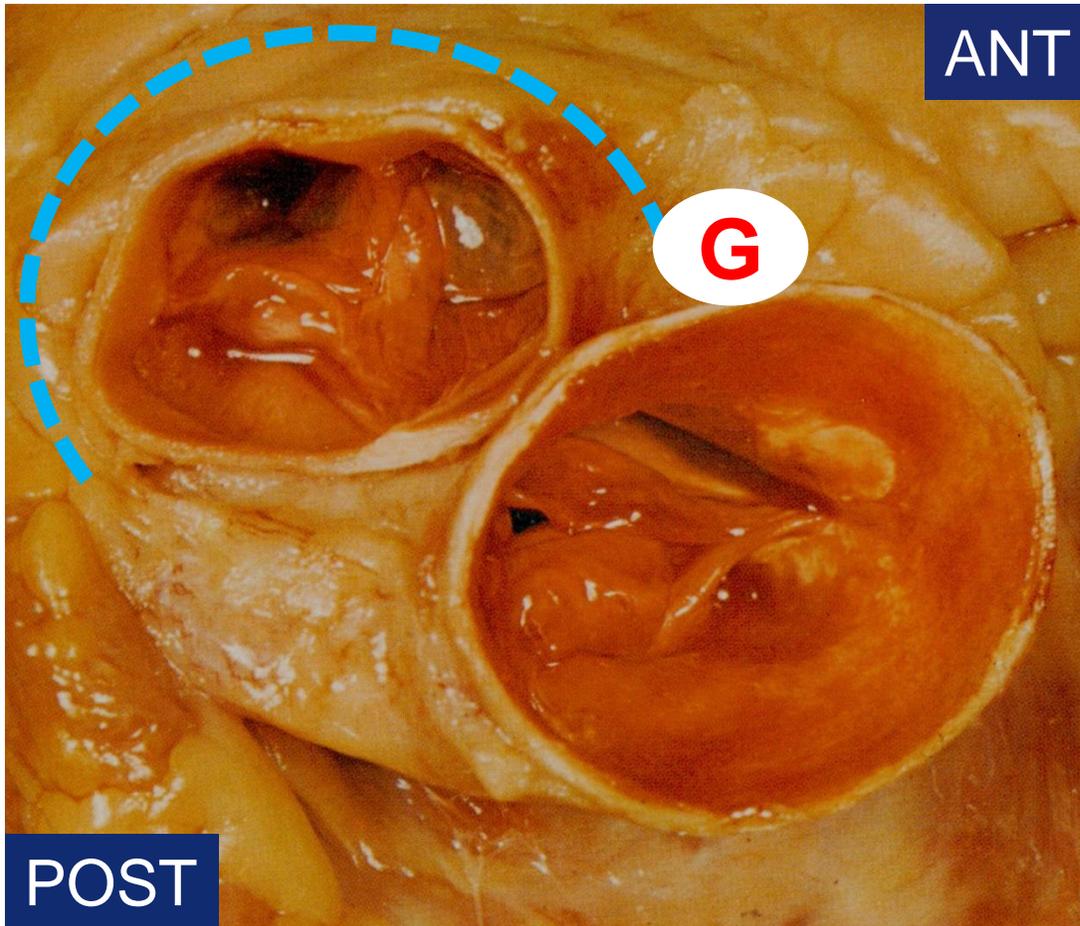
Bogers AJC Anat Embryol 1989

*Sharma B
Annu Rev Physiol 2017*



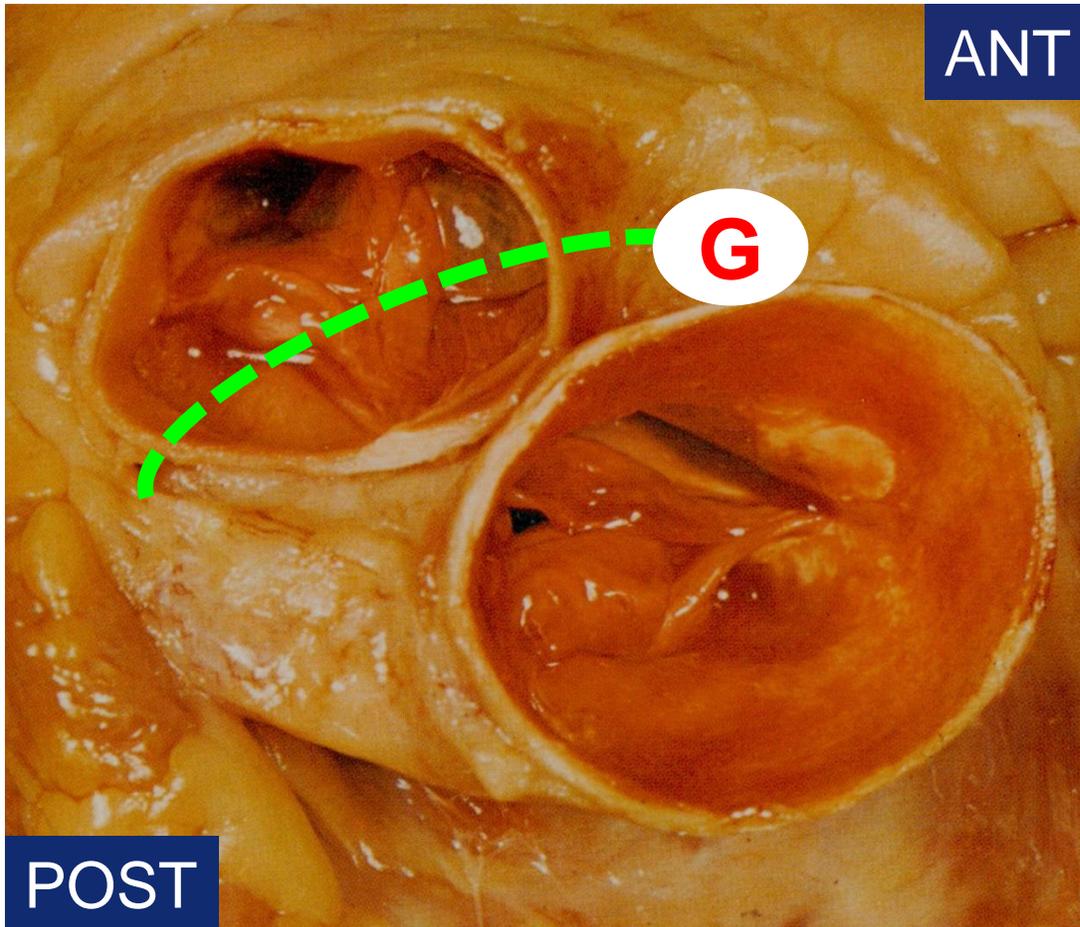


Trajets ectopiques possibles pour la coronaire gauche



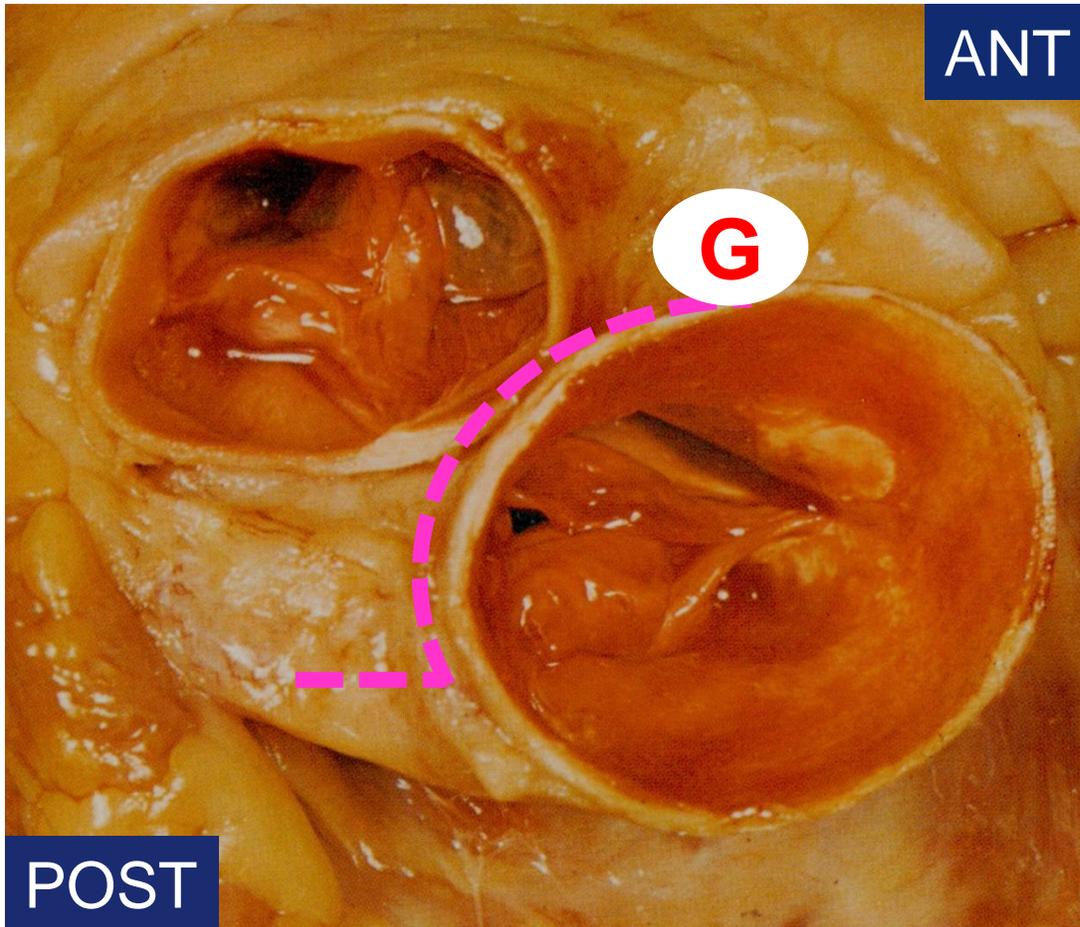
- — — — — prépulmonaire
- — — — — rétropulmonaire (intraseptal)
- — — — — interartériel (préaortique)
- — — — — rétroaortique

Trajets ectopiques possibles pour la coronaire gauche



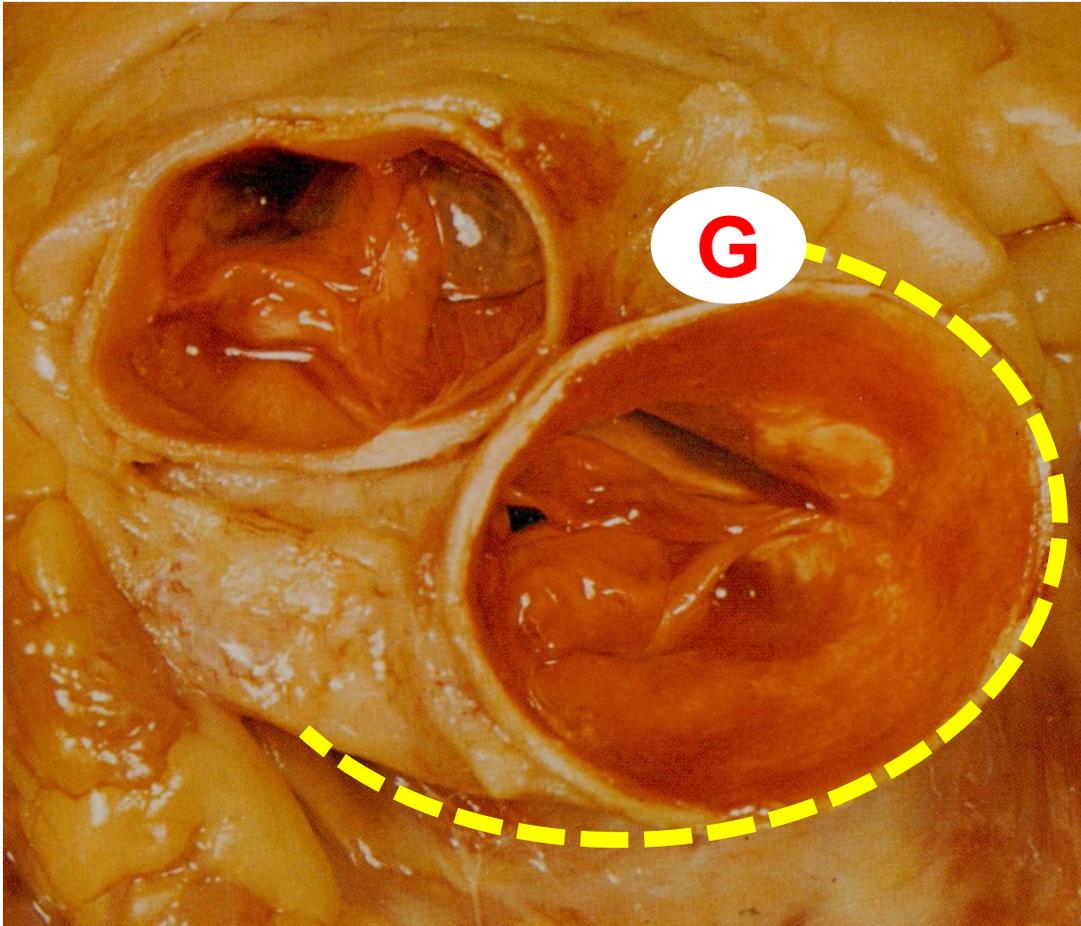
- — — — prépulmonaire
- — — — rétropulmonaire (intraseptal)
- — — — interartériel (préaortique)
- — — — rétroaortique

Trajets ectopiques possibles pour la coronaire gauche



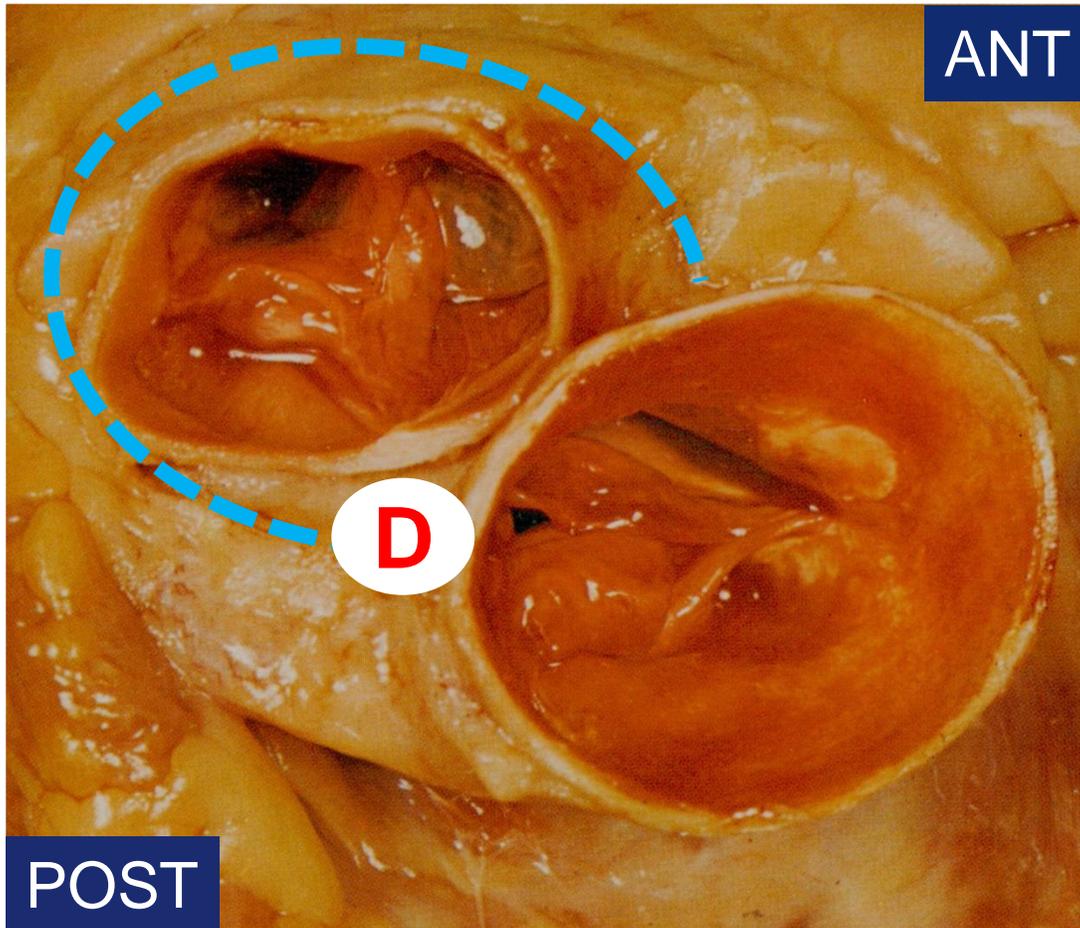
- — — prépulmonaire
- — — rétropulmonaire (intraseptal)
- — — interartériel (préaortique)
- — — rétroaortique

Trajets ectopiques possibles pour la coronaire gauche



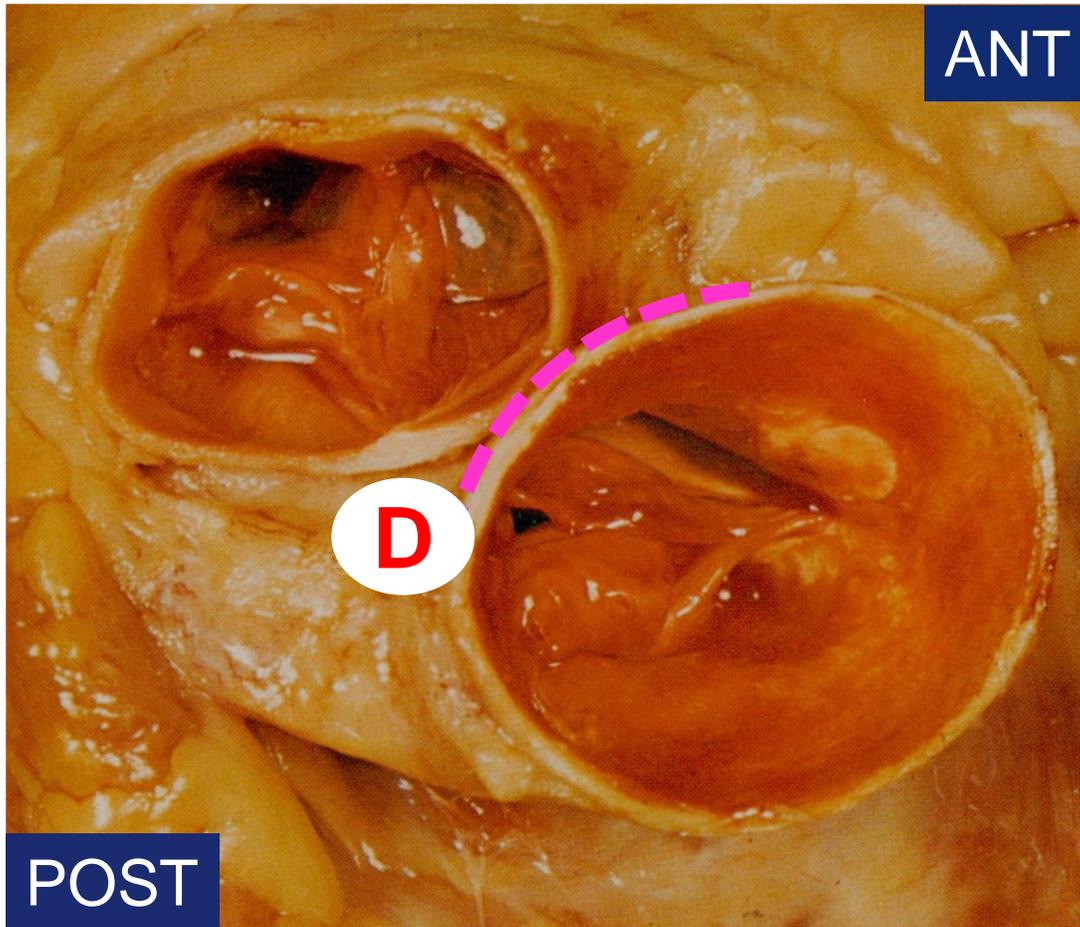
- — — — — prépulmonaire
- — — — — rétropulmonaire (intraseptal)
- — — — — interartériel (préaortique)
- — — — — rétroaortique

Trajets ectopiques possibles pour la coronaire droite



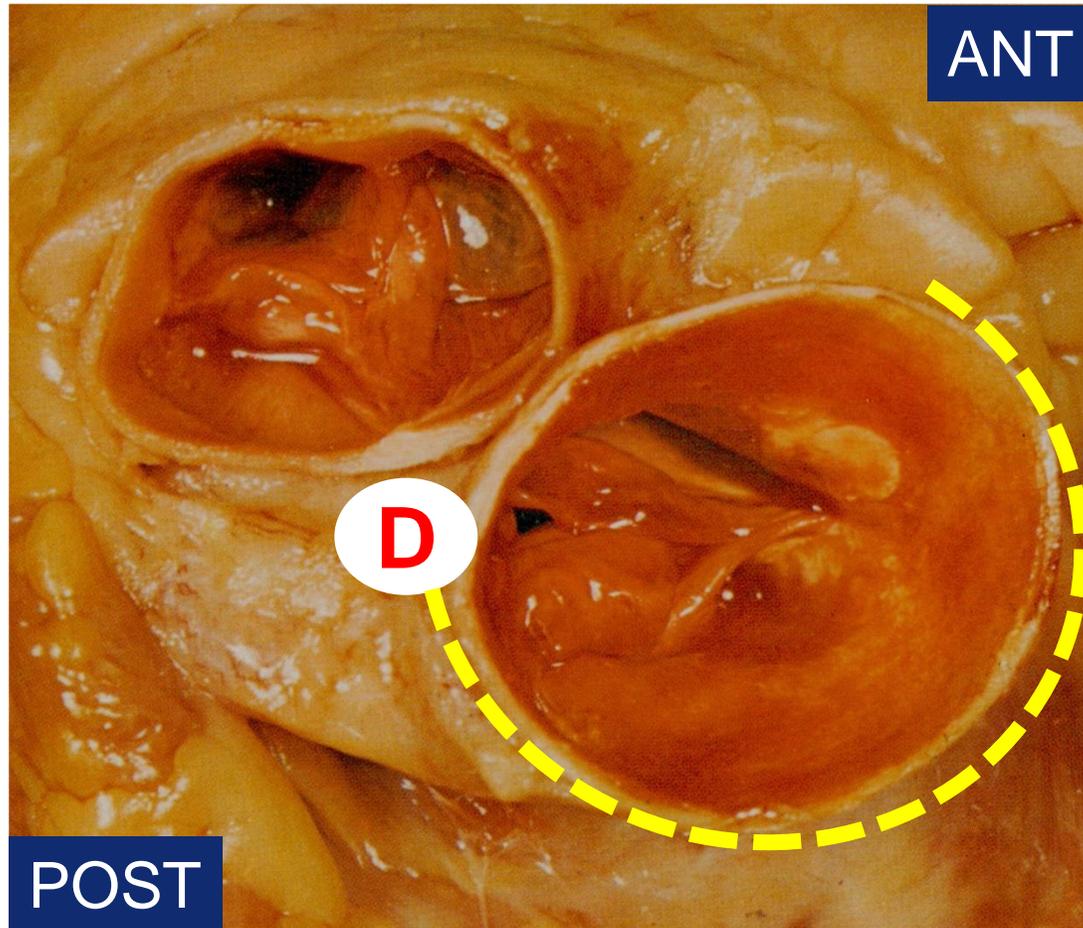
- — — prépulmonaire
- — — interartériel (préaortique)
- — — rétroaortique

Trajets ectopiques possibles pour la coronaire droite



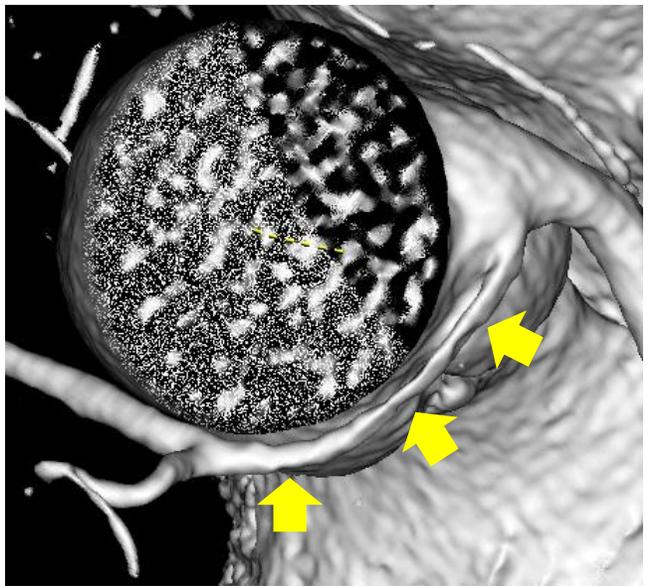
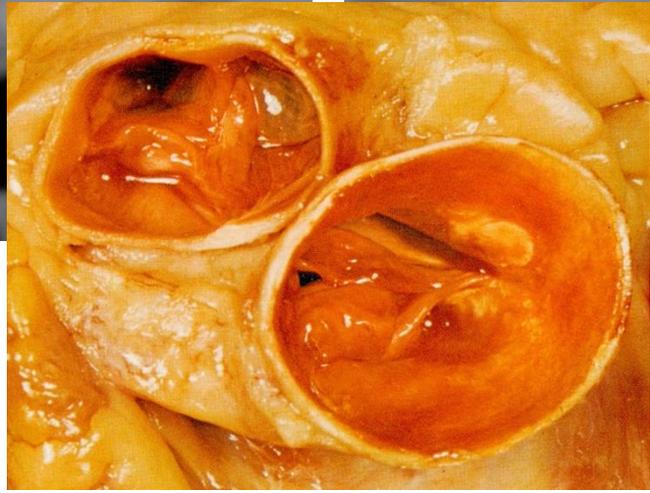
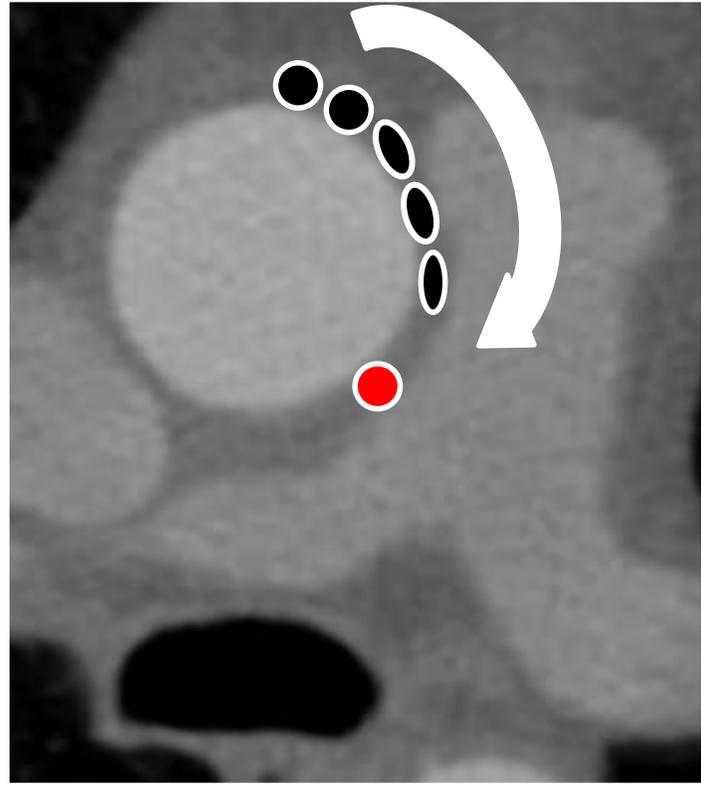
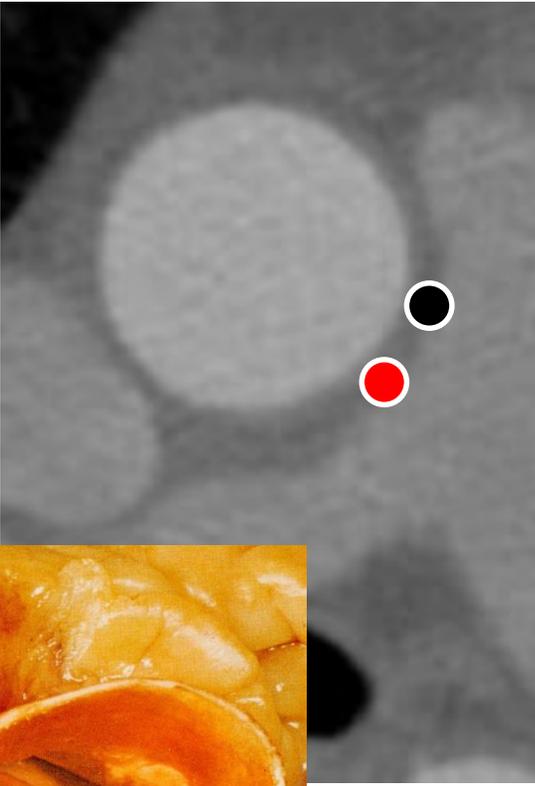
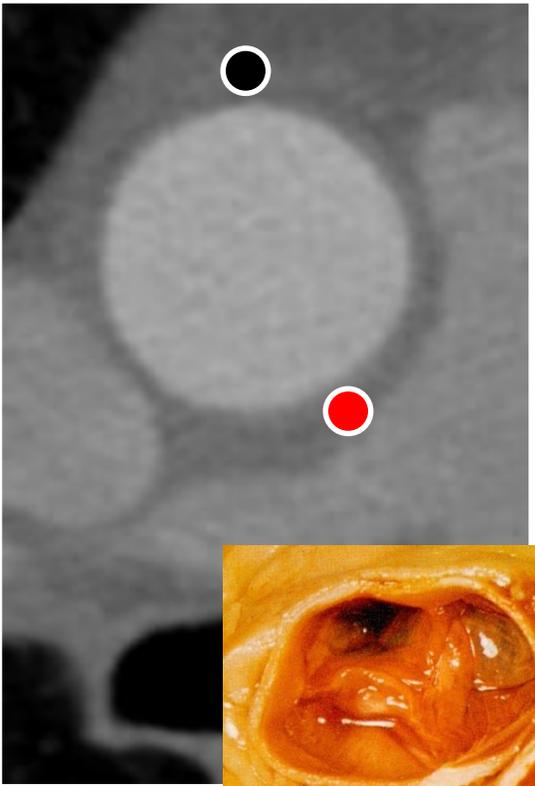
- — — prépulmonaire
- — — interartériel (préaortique)
- — — rétroaortique

Trajets ectopiques possibles pour la coronaire droite

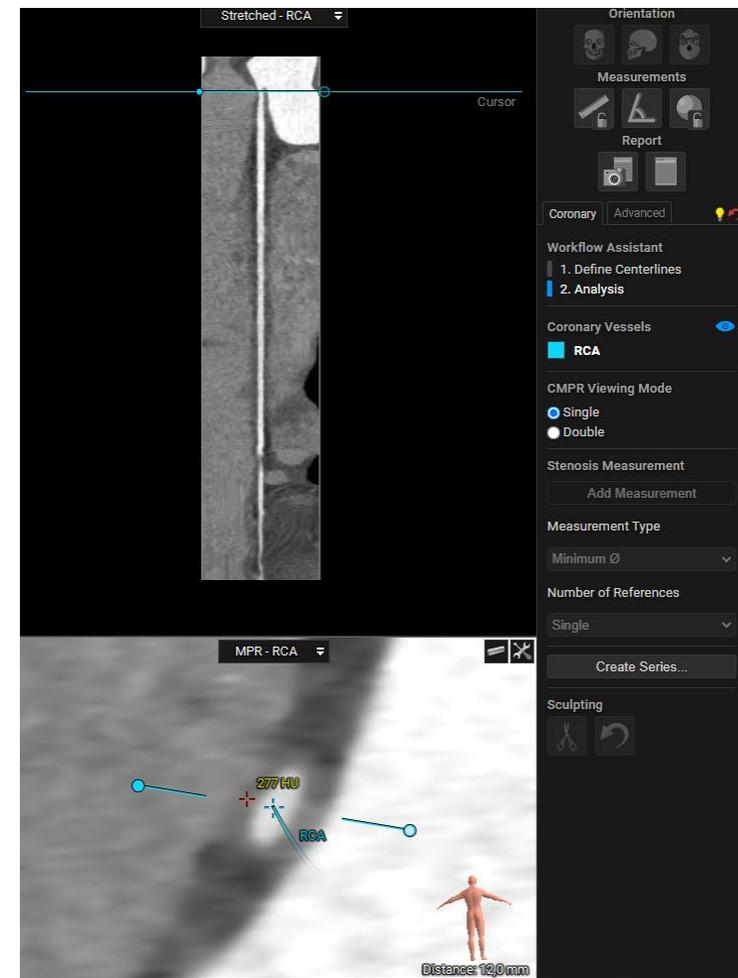
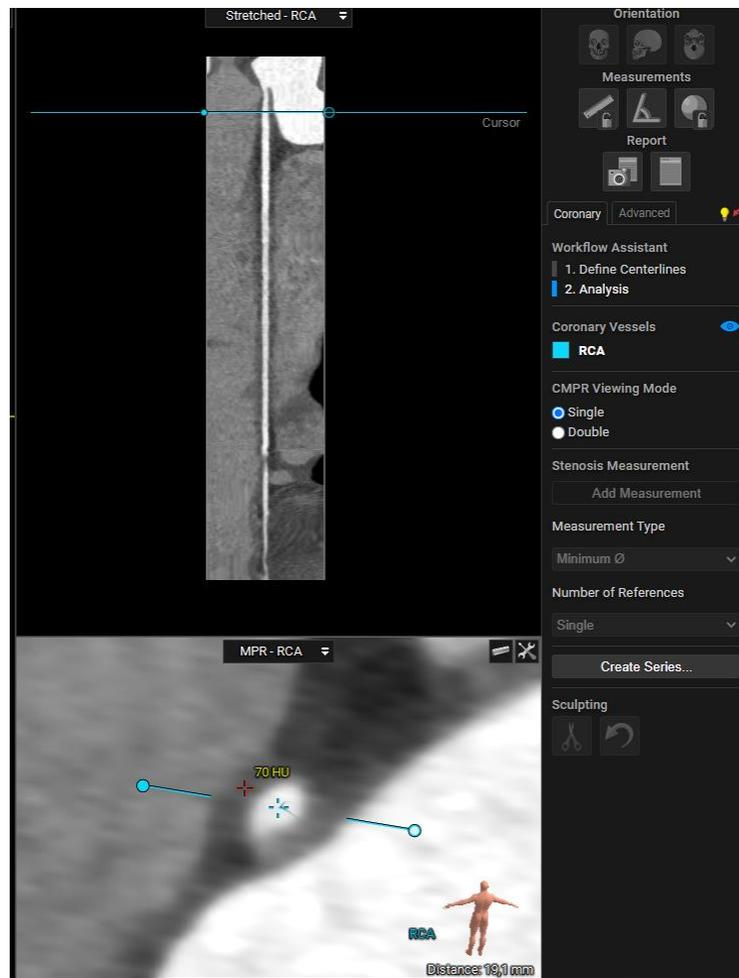
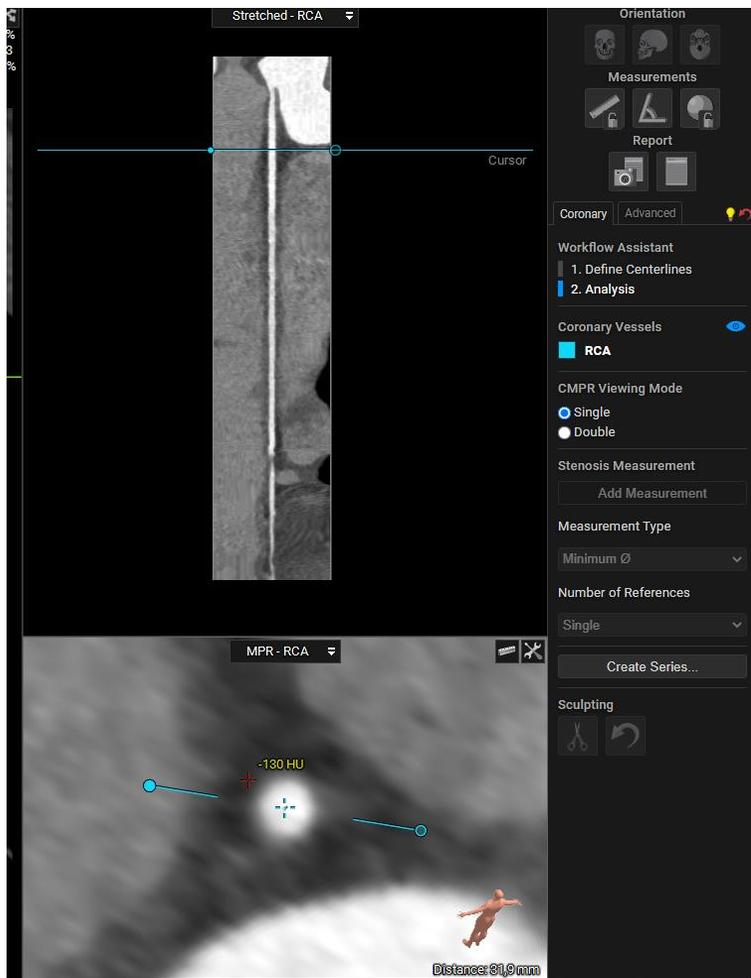


- — — prépulmonaire
- — — interartériel (préaortique)
- — — rétroaortique

Adaptation vasculaire

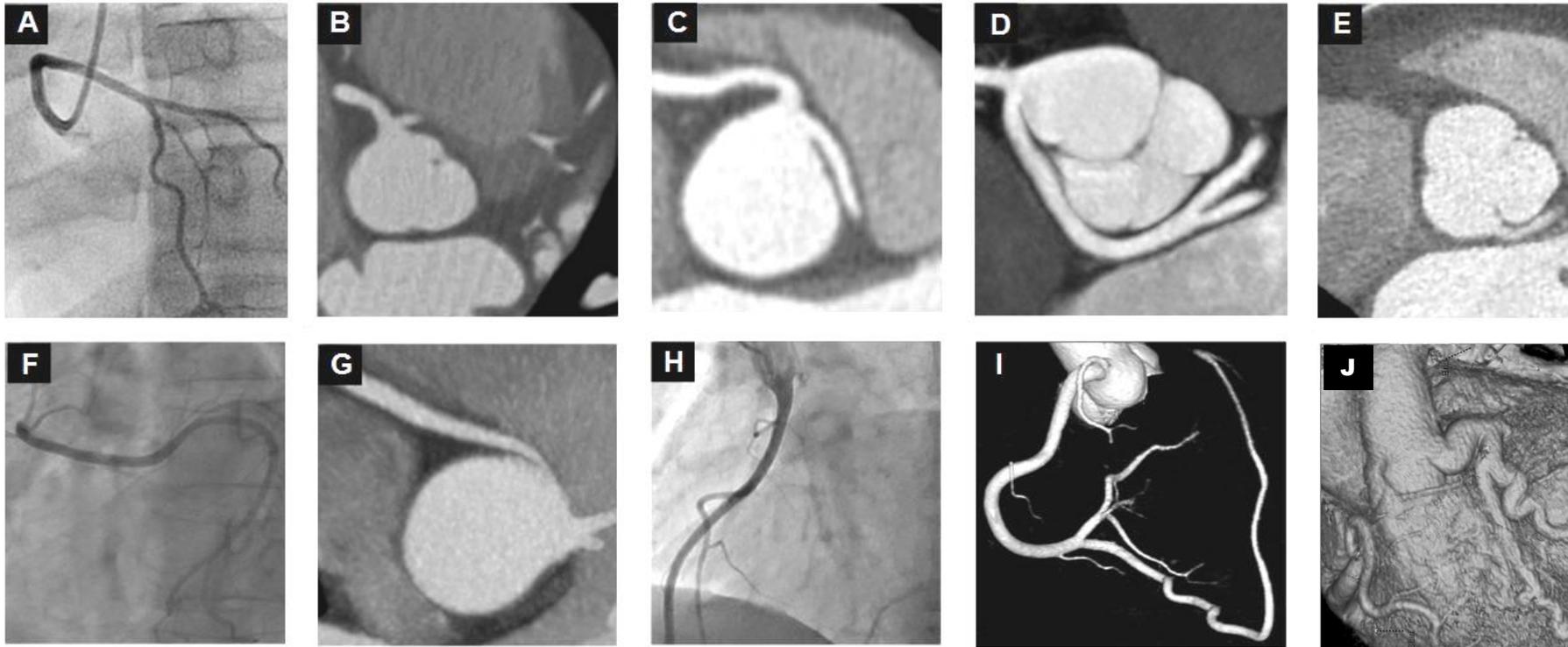


Adaptation vasculaire



- Embryologie et anatomie
- **Classification**
- Prévalence
- Imagerie
- Ischémie myocardique
- Mort subite
- Dépistage
- Prise en charge
- Chirurgie
- Angioplastie
- Activités sportives

Nombreuses formes anatomiques



Classification anatomique

- Type d'artère

1

Tronc
IVA
Circonflexe
Droite
Septale

- Site de connexion

2

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

- Trajet

3

Prépulmonaire
Rétropulmonaire
Interartériel
Rétroaortique
Normal

Passage myocardique
Passage intramural

4

Classification clinique

- Risques

5

Mort subite
Arrêt cardiaque
Arythmies V
Ischémie myocardique
Absents

- Liens de causalité

6

Entre anomalie coronaire et symptomatologie
Entre anomalie coronaire et ischémie myocardique
Entre anomalie coronaire et arythmie ventriculaire
Entre anomalie coronaire et arrêt cardiaque

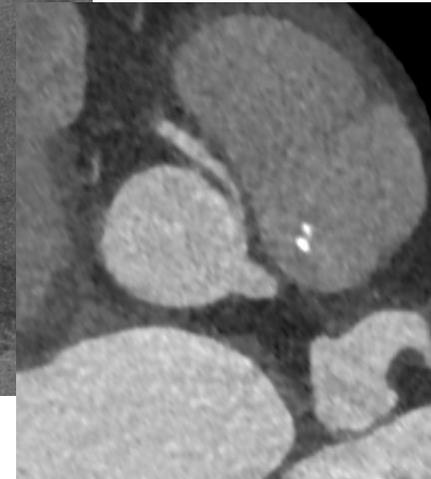
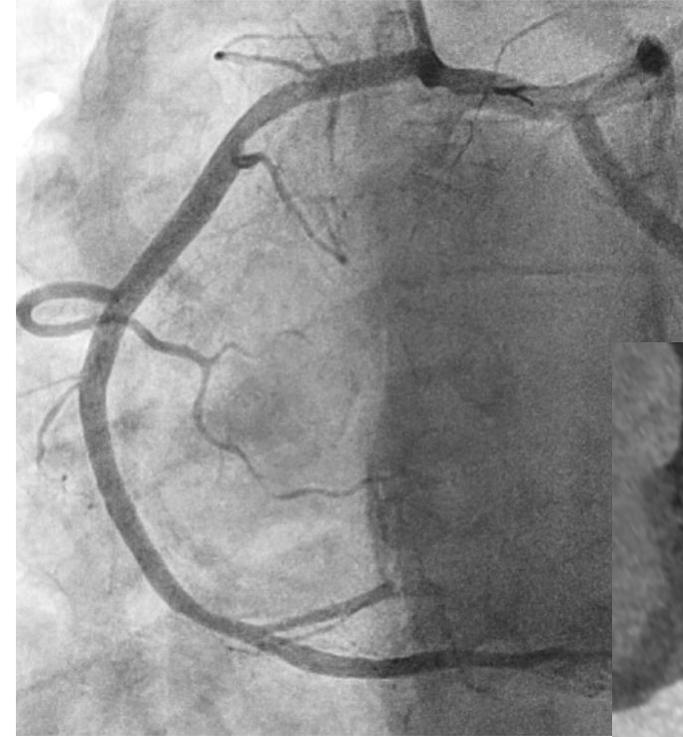
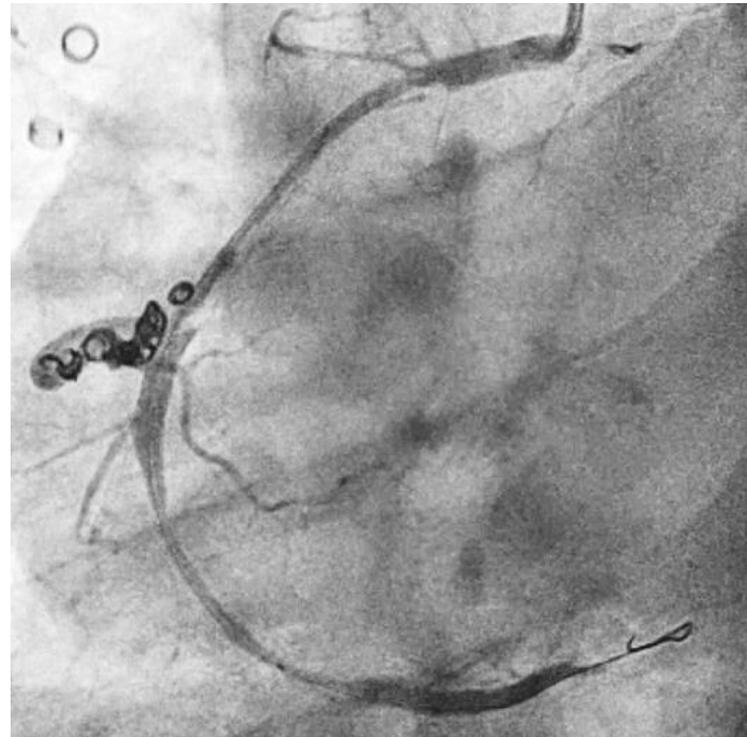
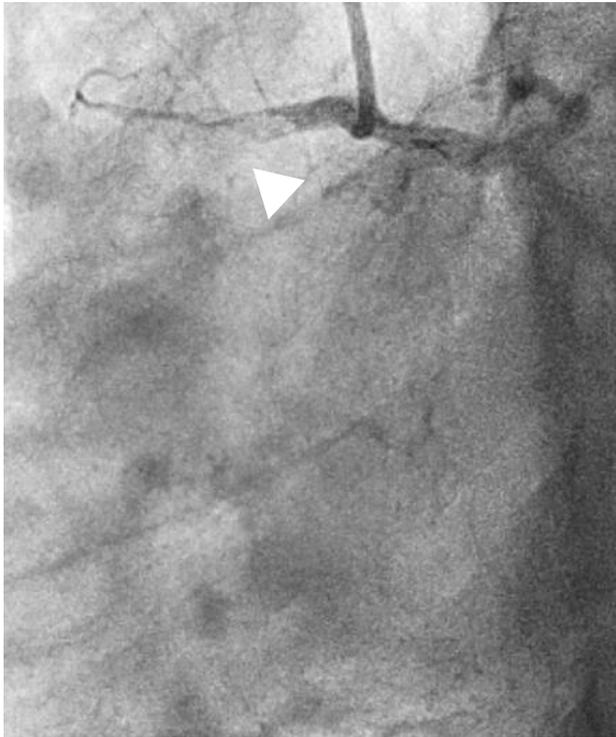
Absent

Possible/Probable

Certain

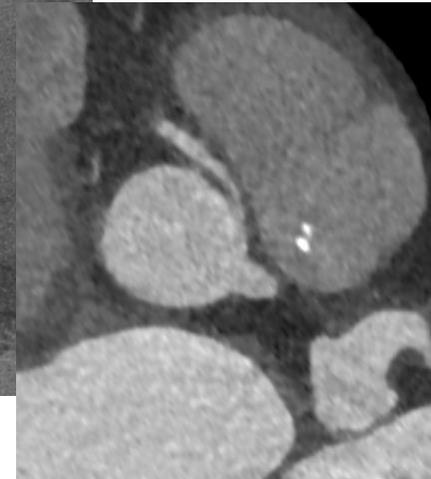
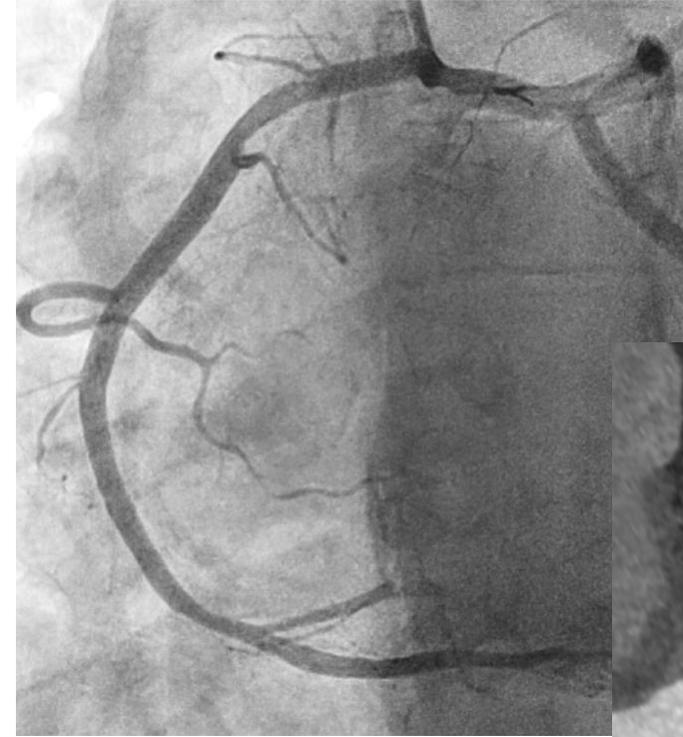
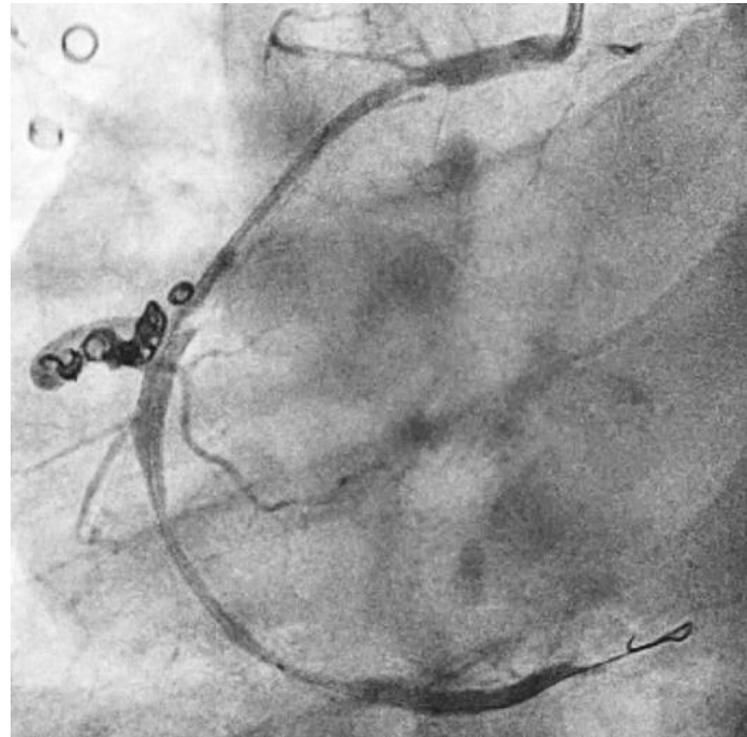
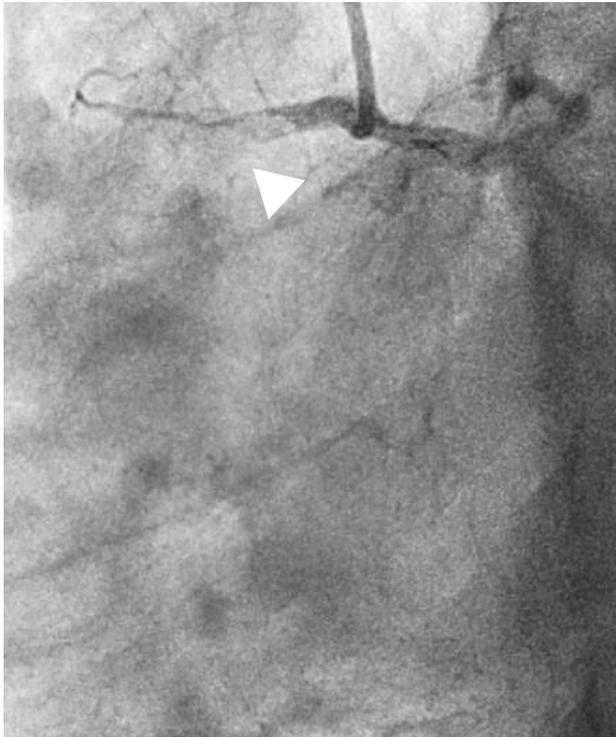
Classification

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



Classification

Homme de 35 ans – Maladie Rendu-Osler – MAV cérébrales/pulmonaires
SCA ST+ inférieur lors activité sportive
Diagnostic d'embolie paradoxale sur MAV pulmonaires



- Embryologie et anatomie
- Classification
- **Prévalence**
- Imagerie
- Ischémie myocardique
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- Chirurgie
- Angioplastie
- Activités sportives

Prévalence



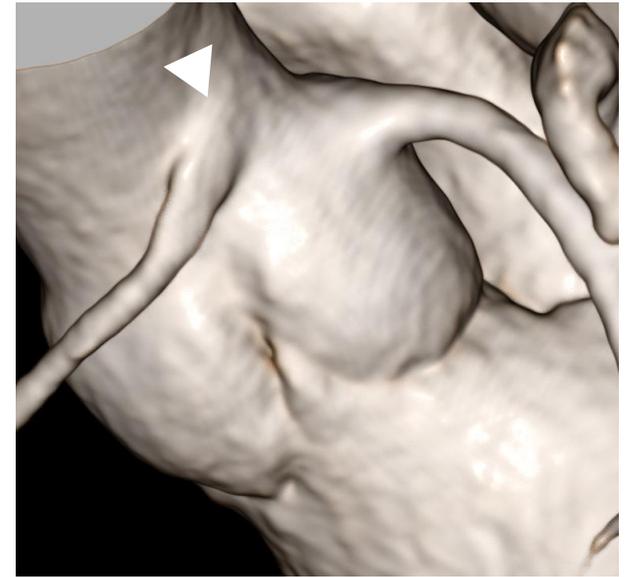
Echocardiographie

0.2%



Coronarographie

0.8%



Scanner

1.0%

Prévalence selon l'artère



Circonflexe

- sinus droit
- coronaire droite

Prévalence angiographique

• **4/1000**

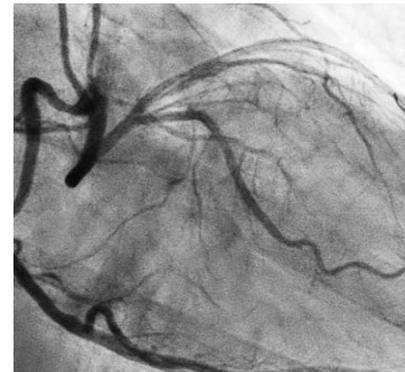
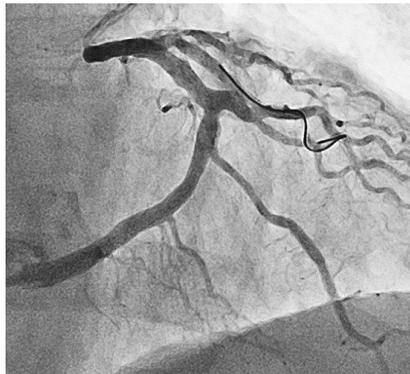
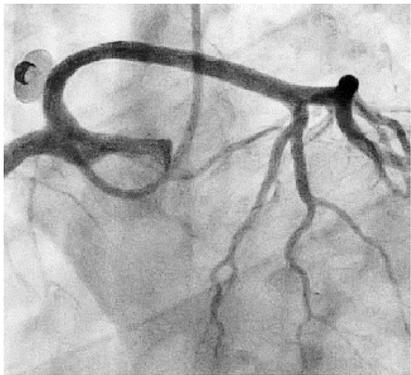


Coronaire droite

- sinus gauche

Prévalence angiographique

• **3/1000**



Tronc commun/IVA

- sinus droit
- coronaire droite

Prévalence angiographique

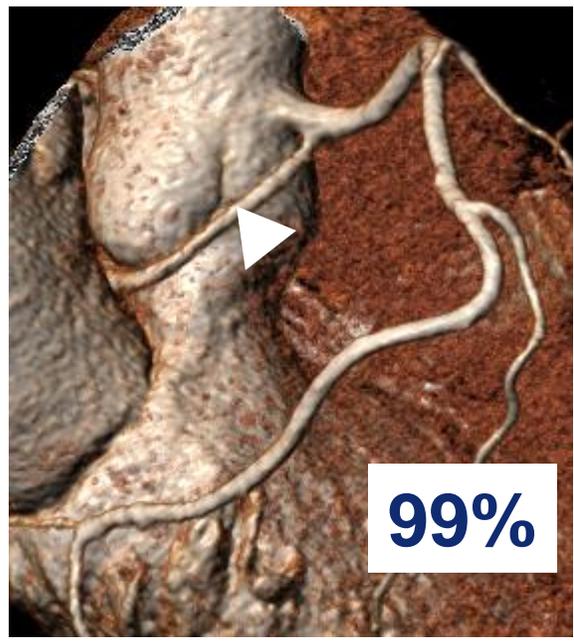
• **1/1000**

Prévalence selon le trajet

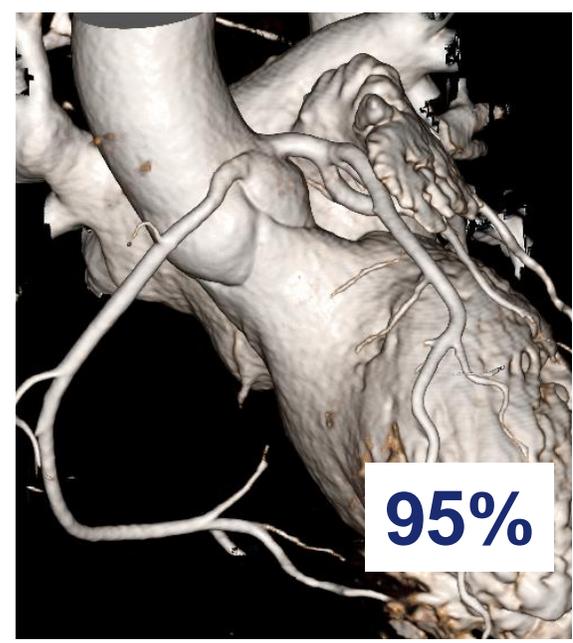
Circonflexe



Coronaire droite



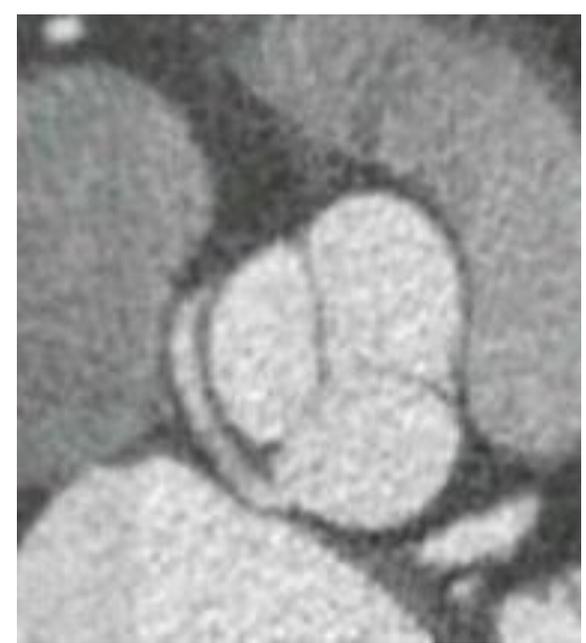
Rétroaortique



Interartériel



Prépulmonaire

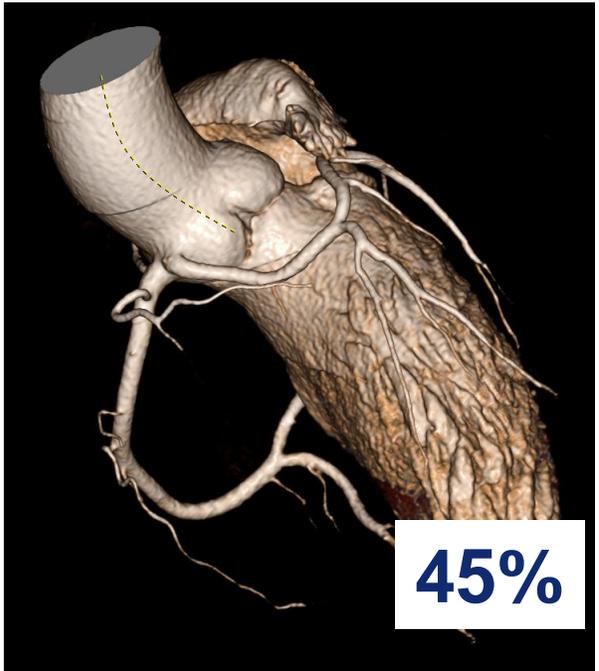


Rétroaortique

Prévalence selon le trajet



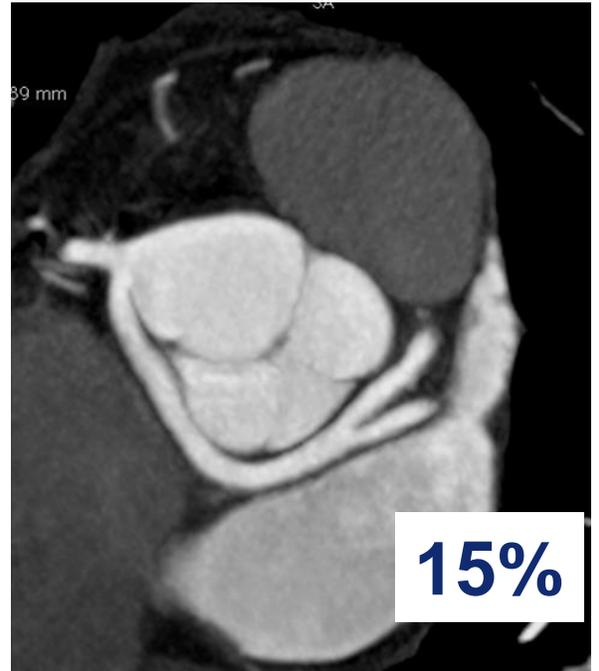
Prépulmonaire



Rétropulmonaire



Interartériel



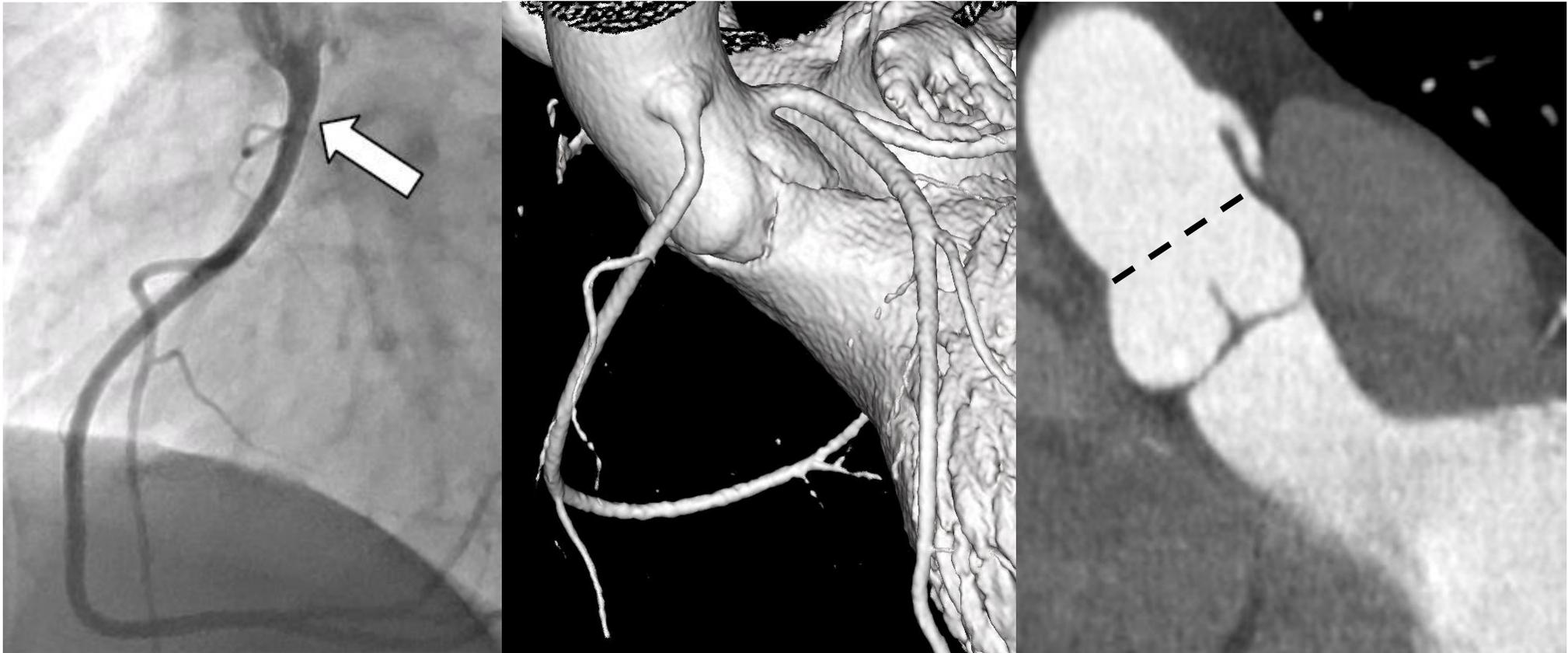
Rétroaortique

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*

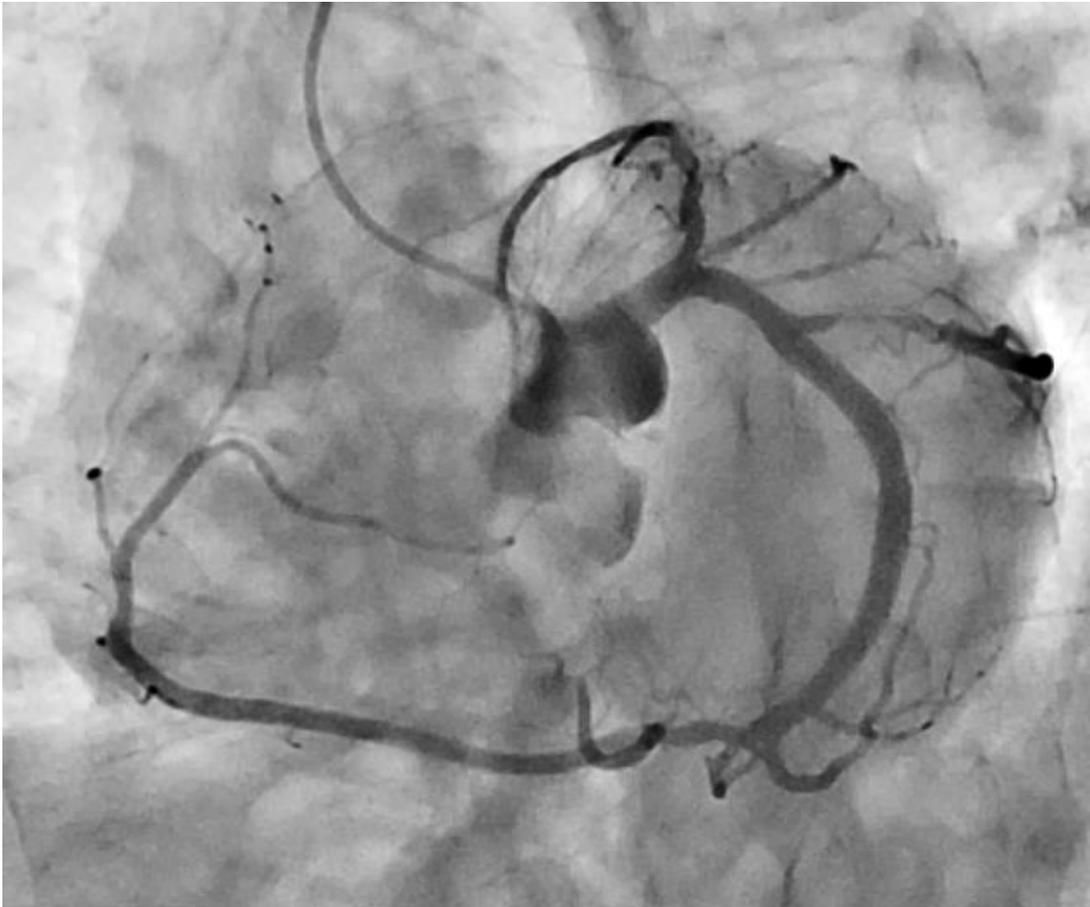
Connexion coronaire droite dans aorte thoracique



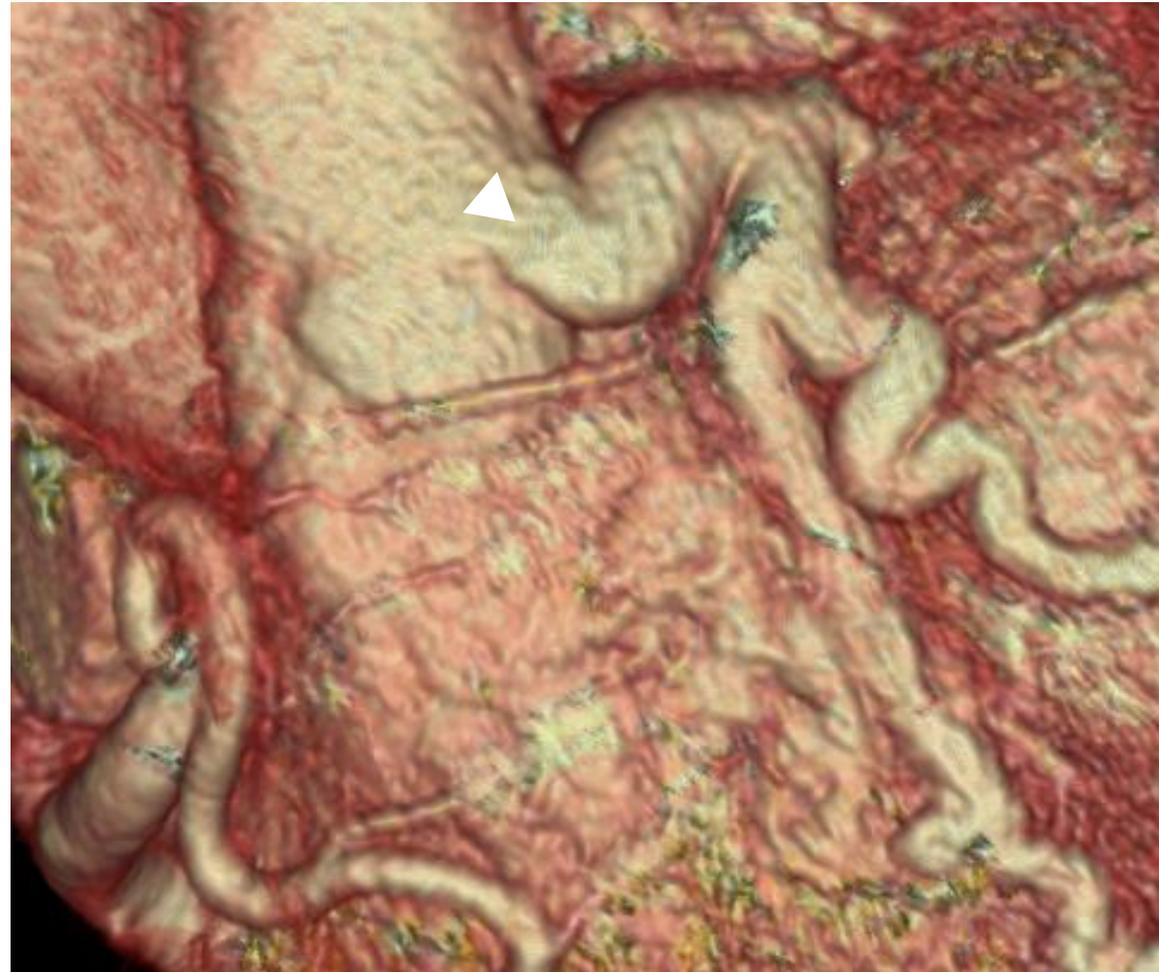
Connexion tronc commun dans sinus non-croanaire



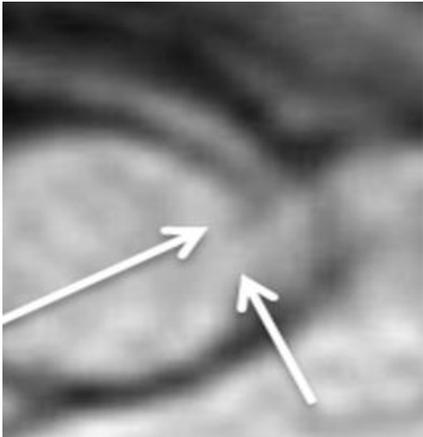
Artère coronaire unique



Connexion IVA pulmonaire



MRI-based study (n = 5.255)
 Middle school children (mean age 13 years)
 2010-2017



Clinical Investigation

High-Risk Cardiovascular Conditions in Sports-Related Sudden Death:

Prevalence in 5,169 Schoolchildren Screened via Cardiac Magnetic Resonance

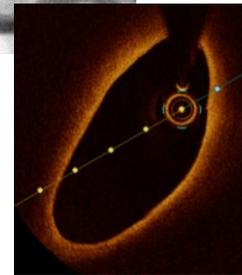
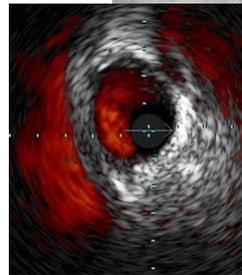
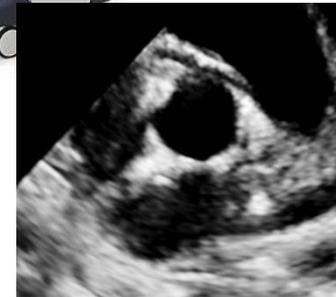
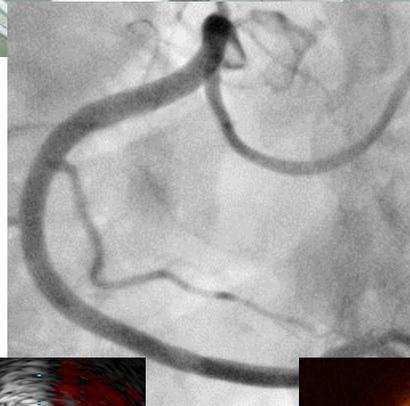
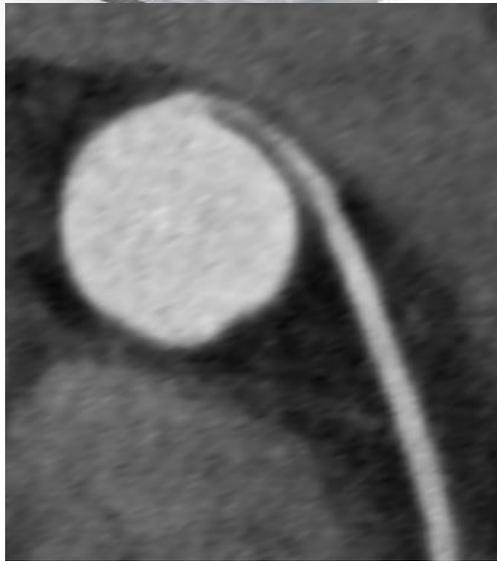
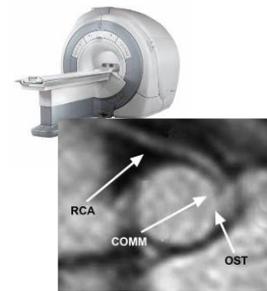
Angelini P Texas Heart Journal 2018

- | | |
|------------------------------|---|
| ▪ L-ACAOS-IA = 2 | ▪ Prevalence L-ACAOS-IA = 0.04% |
| ▪ R-ACAOS-IA = 17 | ▪ Prevalence R-ACAOS-IA = 0.32% |
| ▪ Total ACAOS-IA = 19 | ▪ Total prevalence ACAOS-IA = 3/1000 |

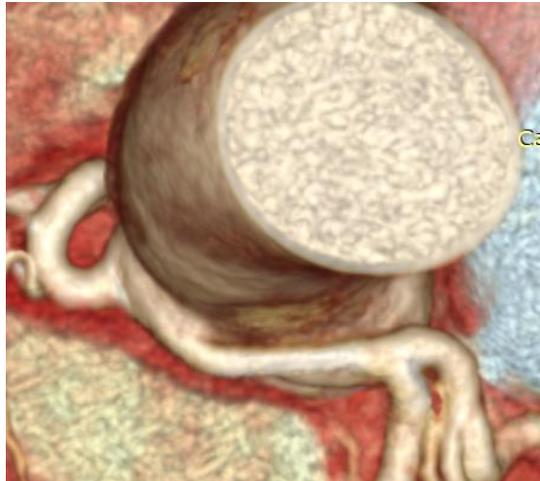
ACAOS: anomalous coronary artery from opposite sinus; IA: interarterial.

- Embryologie et anatomie
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- Prévalence
- **Imagerie**
- Ischémie myocardique
- Mort subite
- Dépistage
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Outils d'imagerie



Piège à éviter

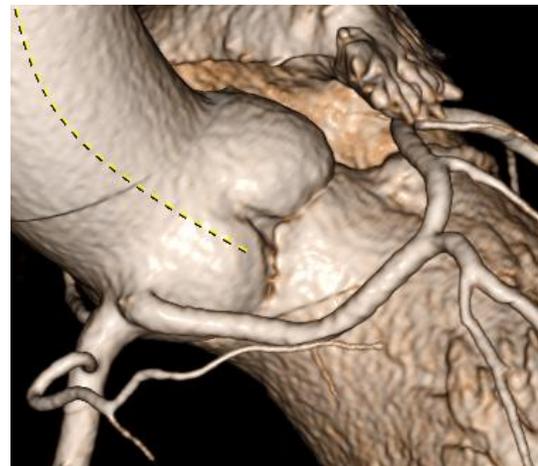


Trajet prépulmonaire

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*

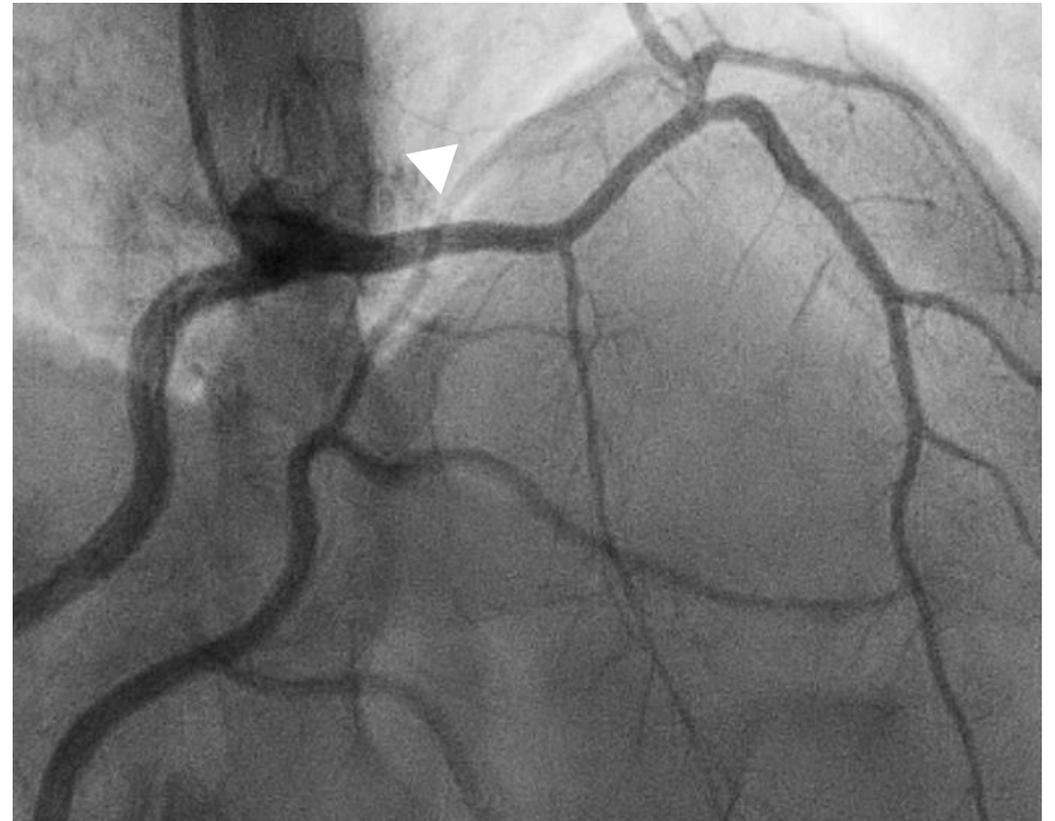
Koutsoukis A Congenital Heart Disease 2017

Formes anatomiques gauches à ne pas confondre

Tronc avec trajet interartériel

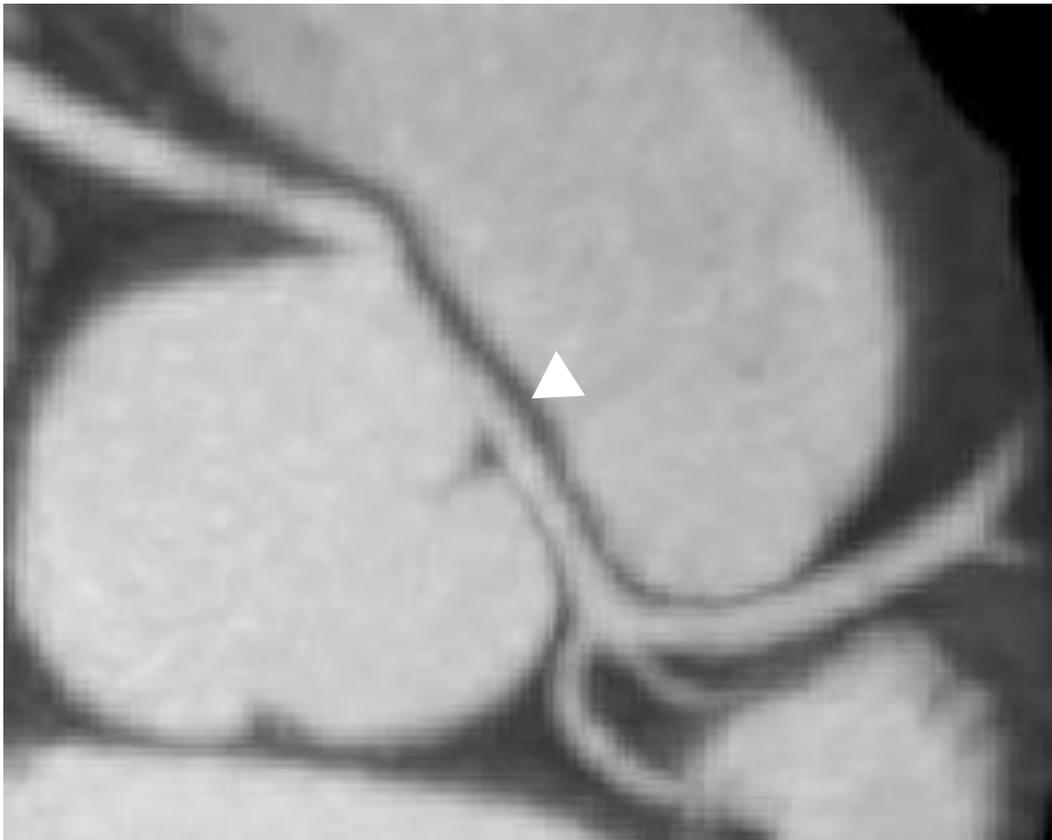


Tronc avec trajet rétropulmonaire



Formes anatomiques gauches à ne pas confondre

Tronc avec trajet interartériel

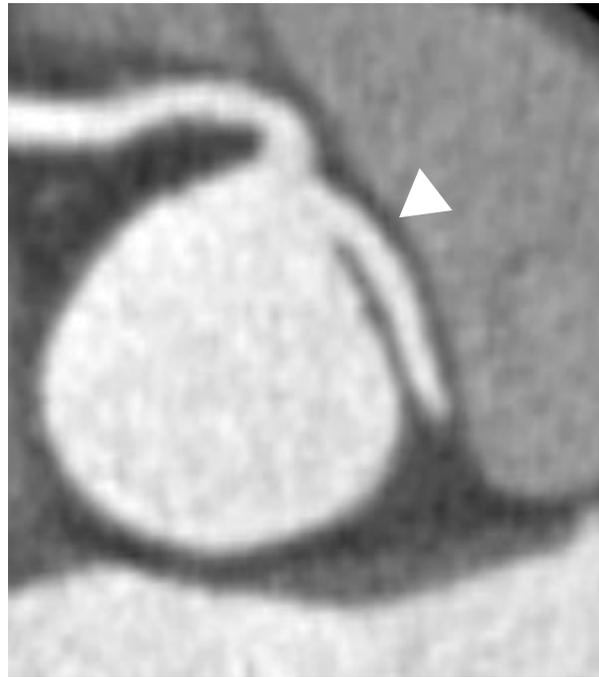


Tronc avec trajet rétropulmonaire

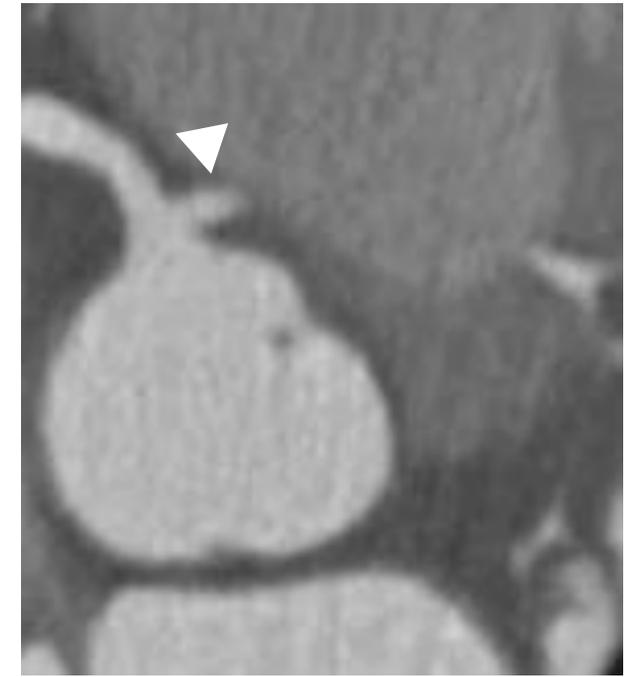
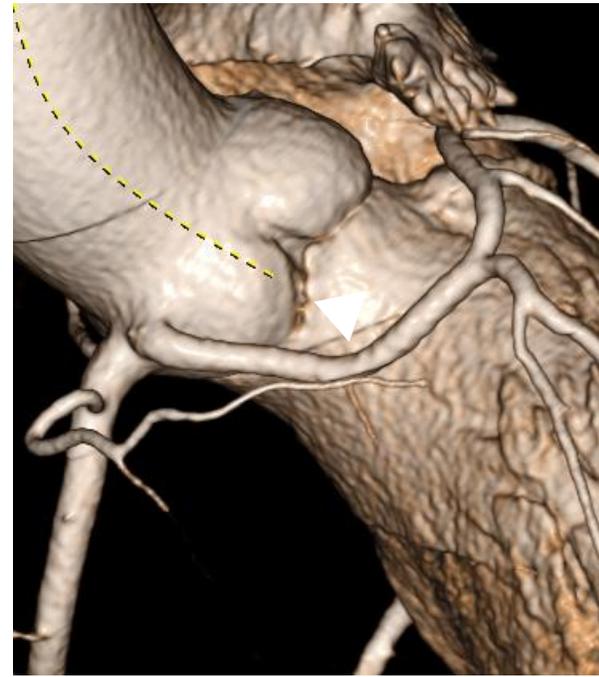


Formes anatomiques gauches à ne pas confondre

Tronc avec trajet interartériel

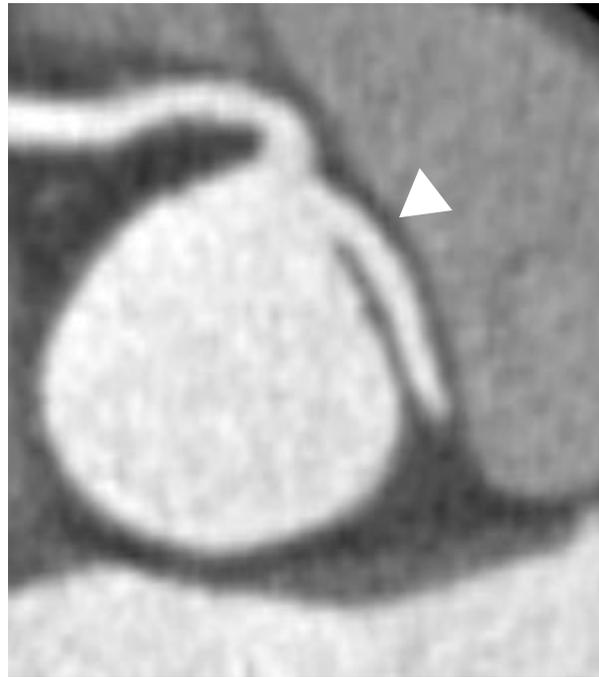


Tronc avec trajet rétropulmonaire

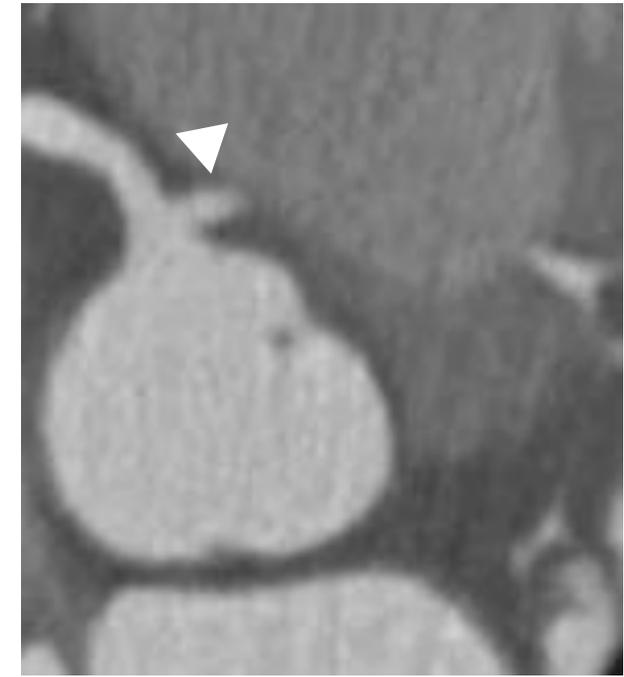
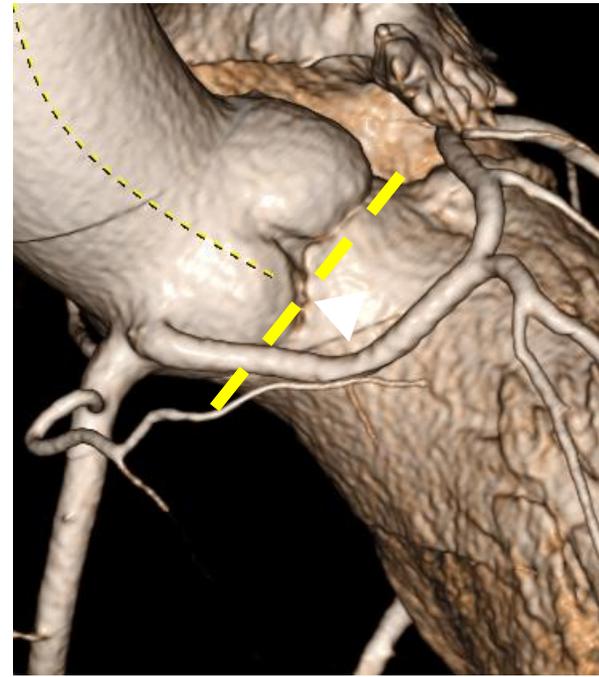


Formes anatomiques gauches à ne pas confondre

Tronc avec trajet interartériel



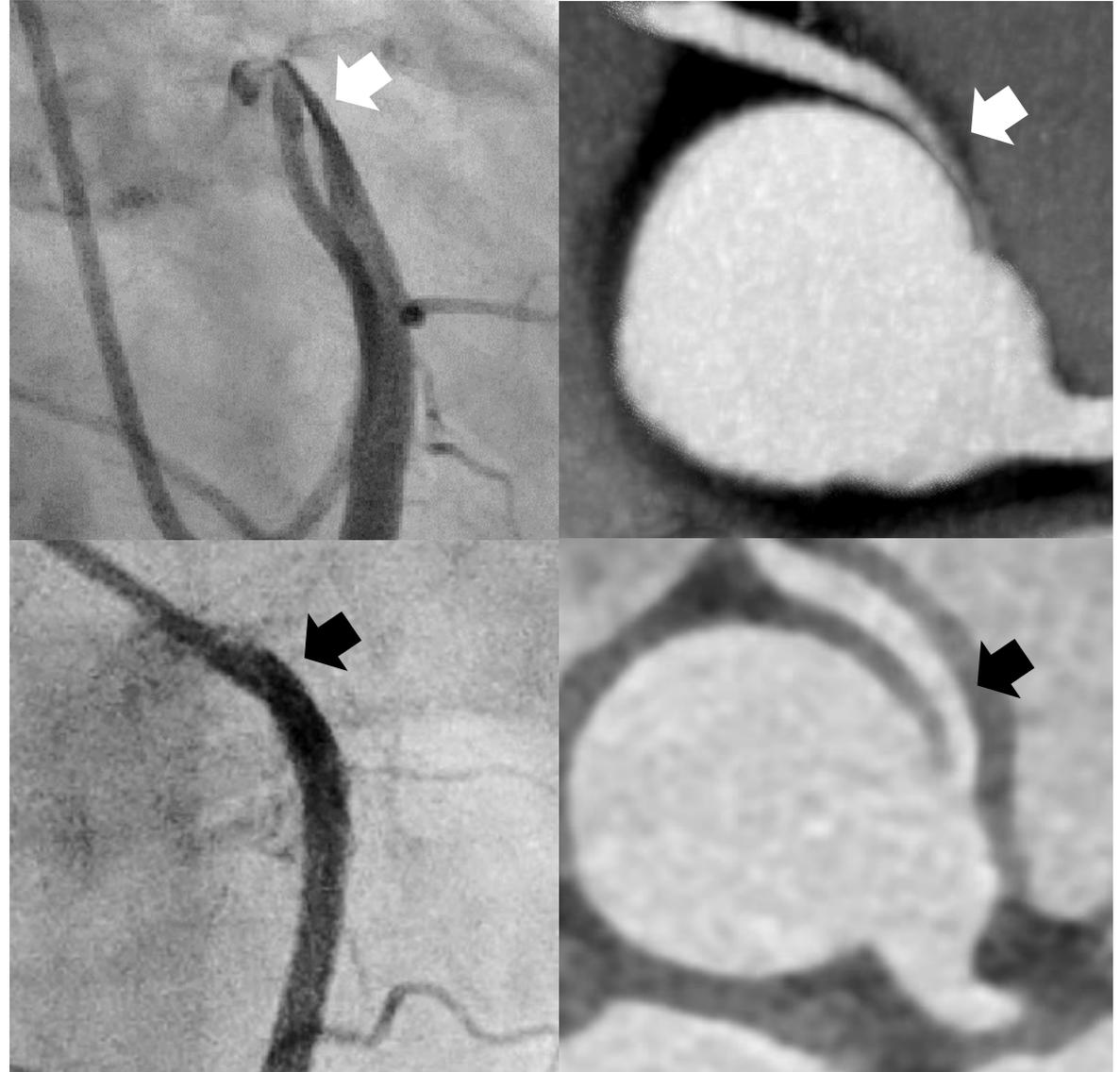
Tronc avec trajet rétropulmonaire



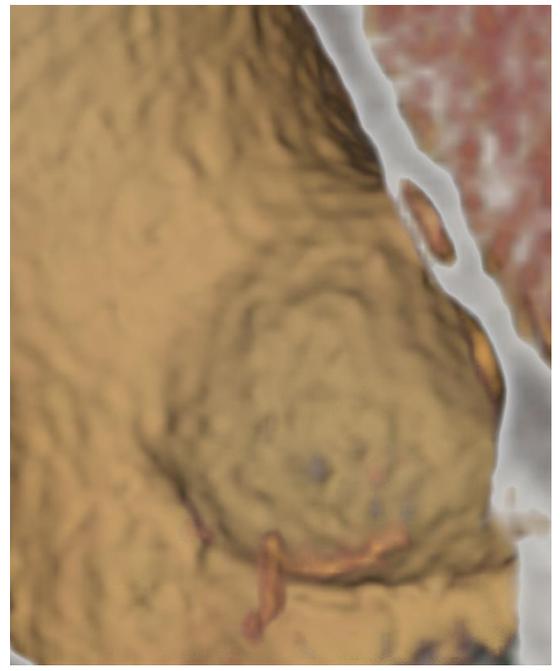
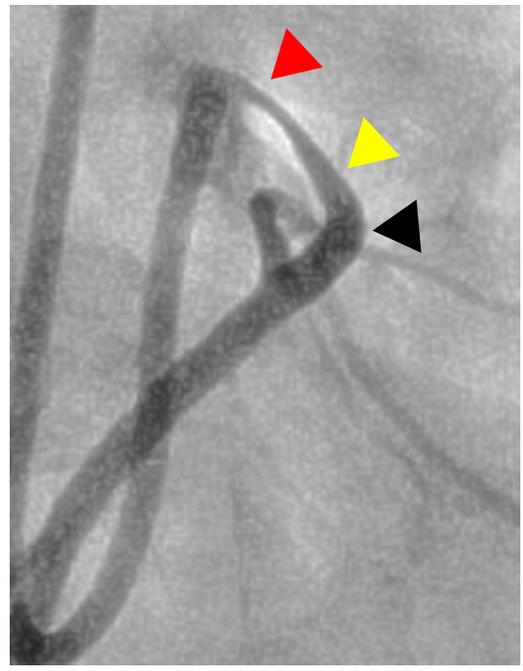
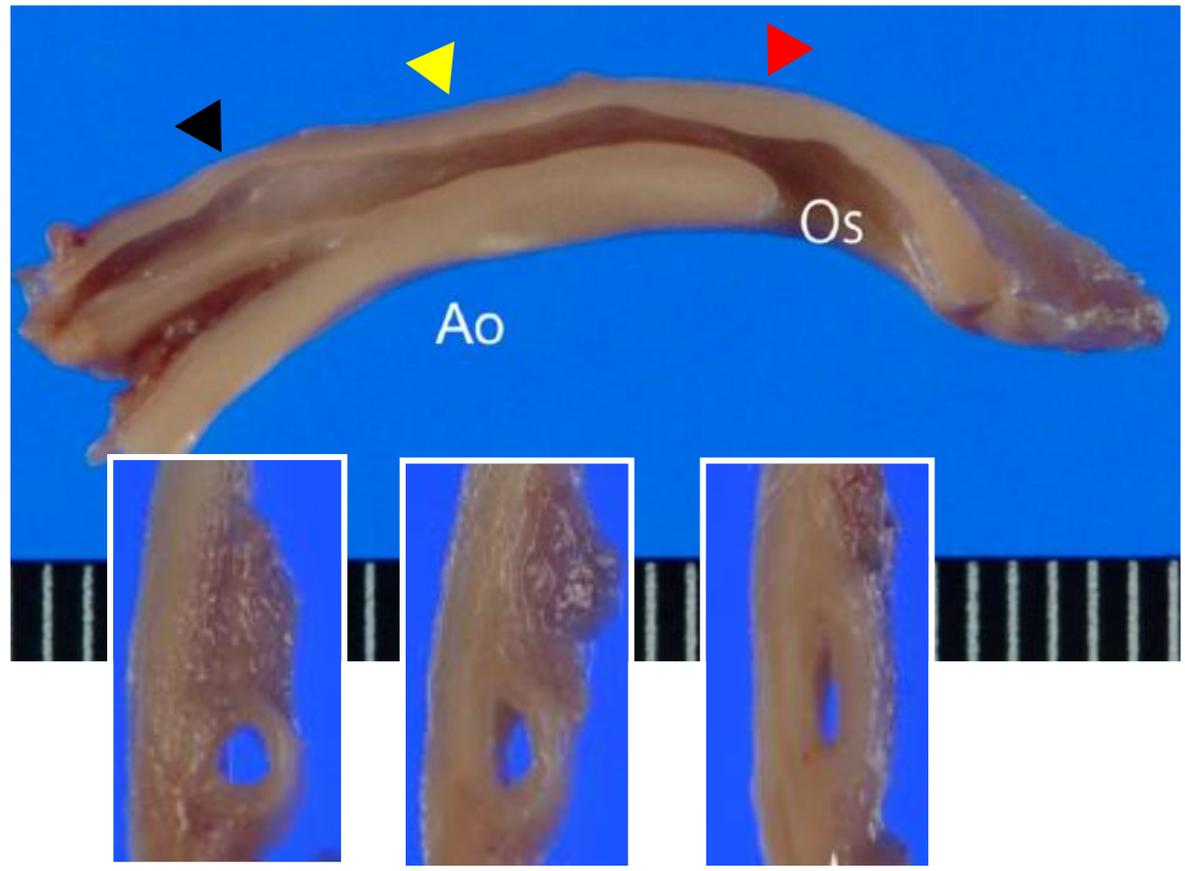
Passage aortique intramural 

Anomalie avec trajet interartériel

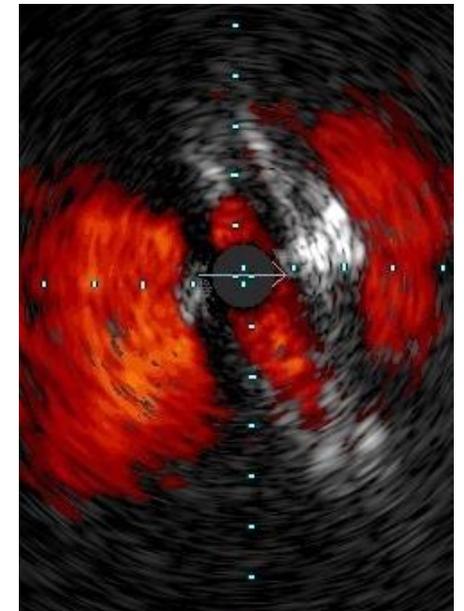
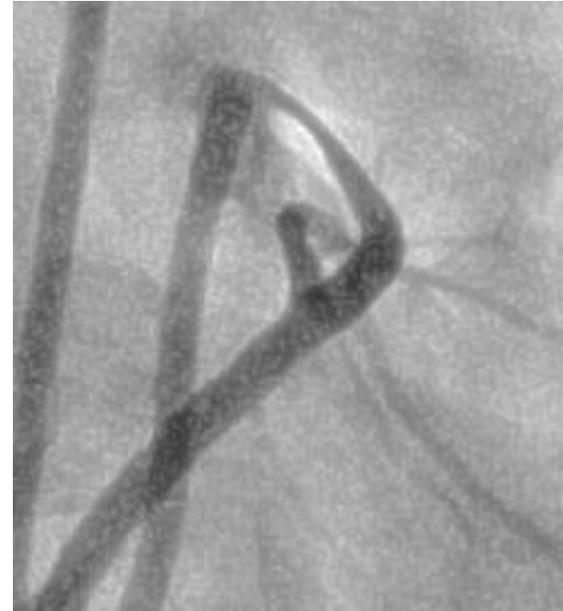
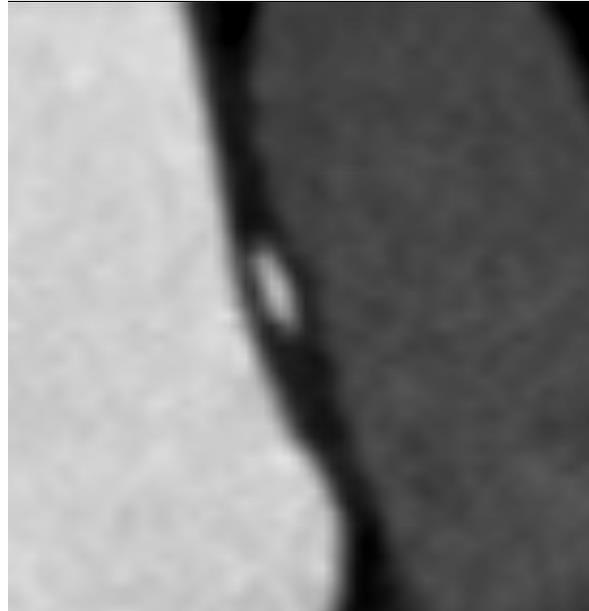
Passage aortique intramural 



Passage intramural aortique

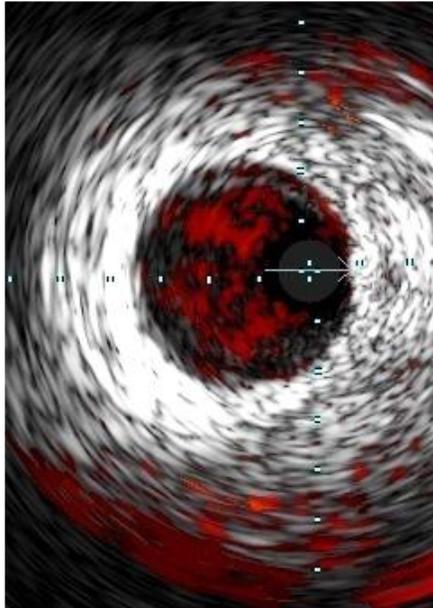


Passage intramural aortique

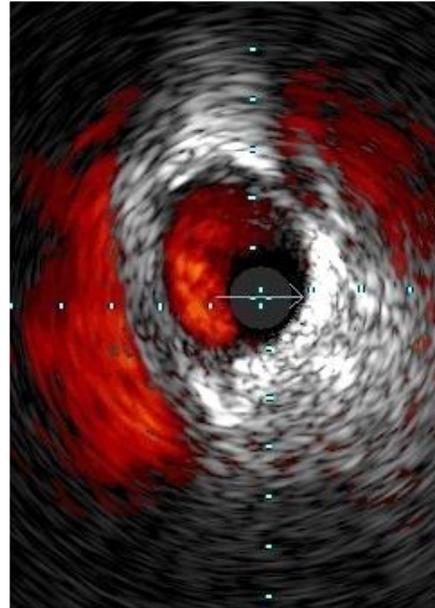


Passage intramural aortique

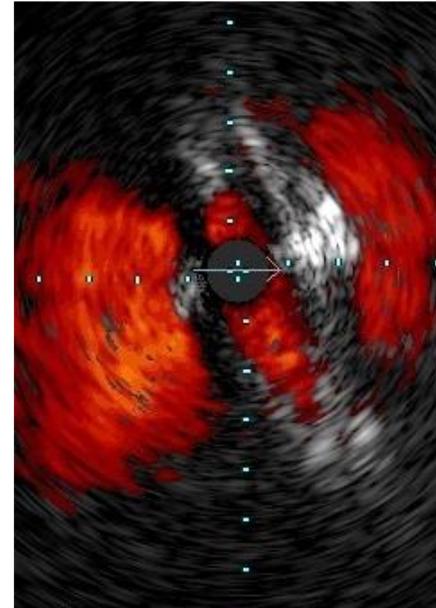
Normal



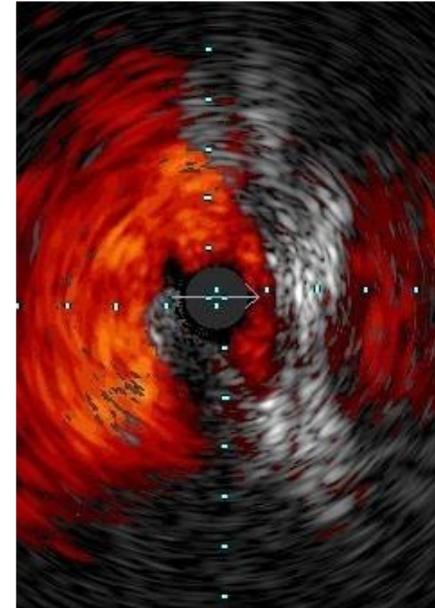
Juxta aortique



Intramural



Ostium

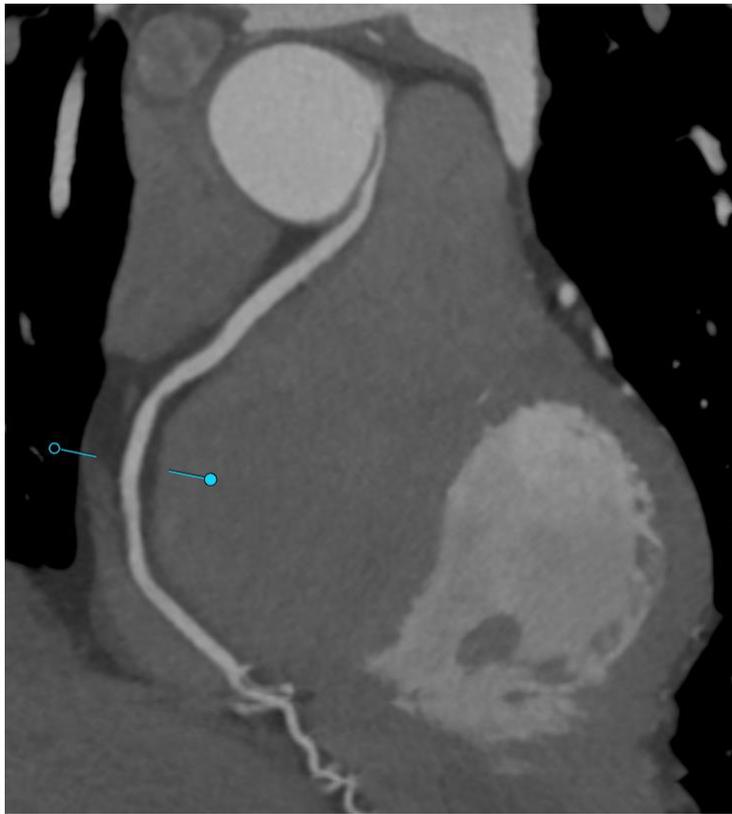


Echographie endocoronaire

Analyse tomographique coronaire droite avec trajet interartériel



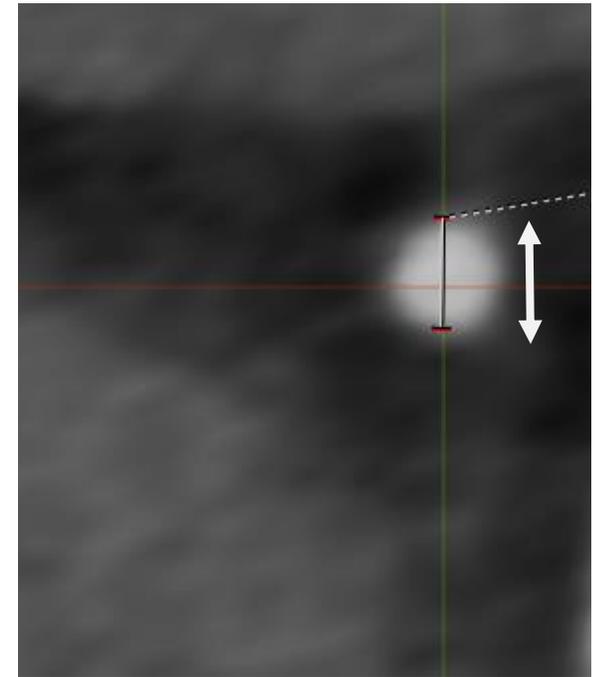
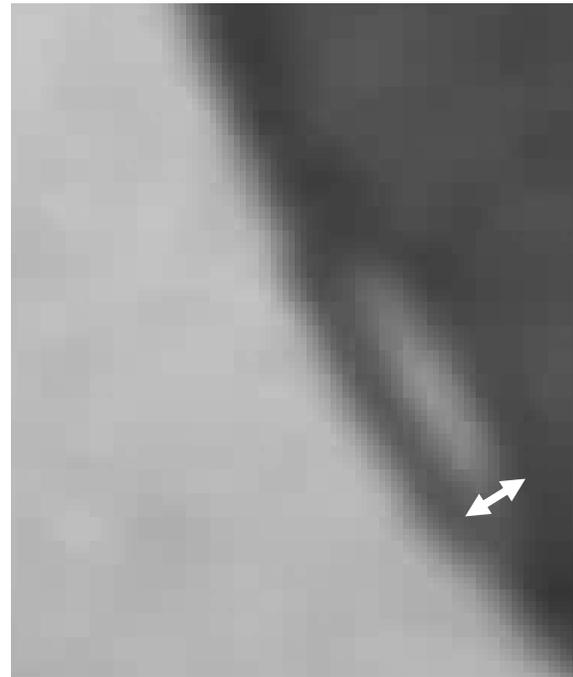
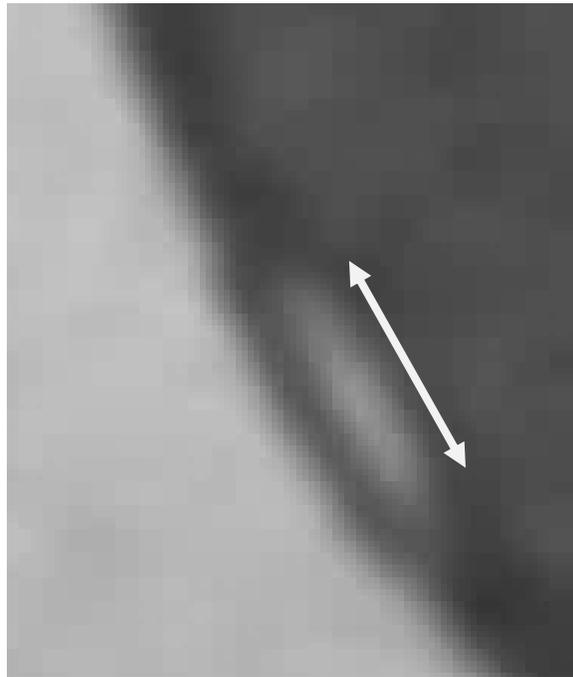
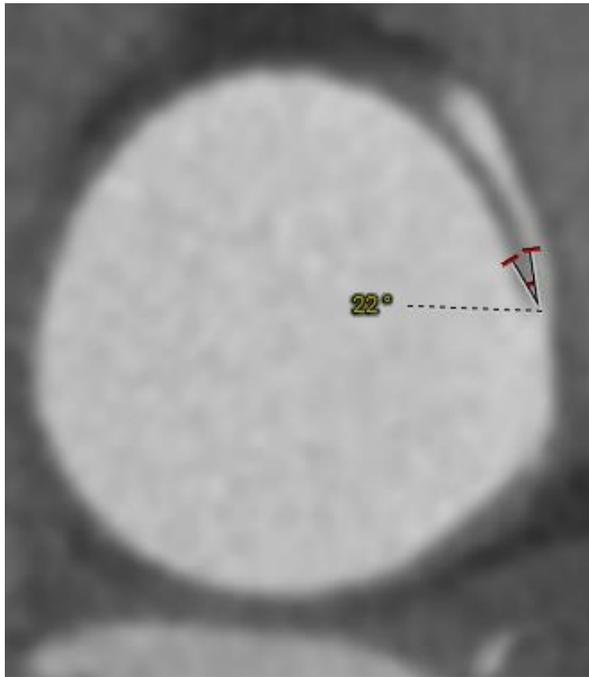
Volume rendering VR)



Multiplanar reconstructions (MPR)



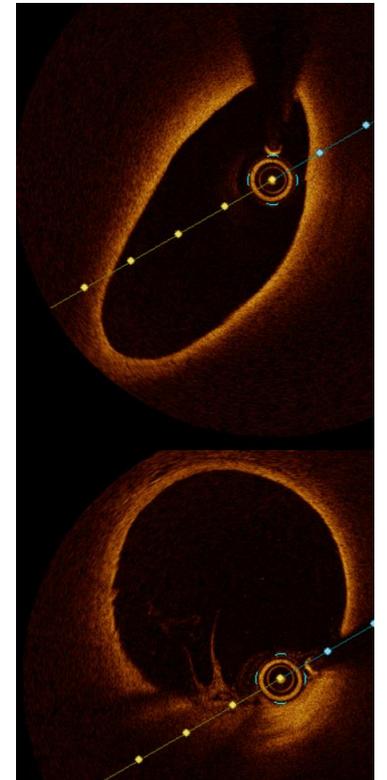
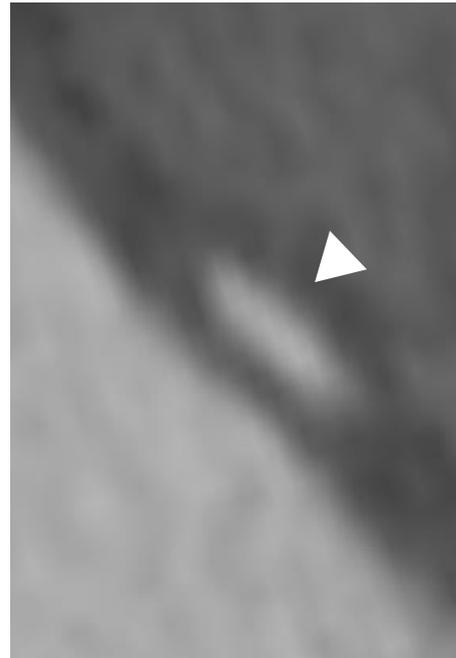
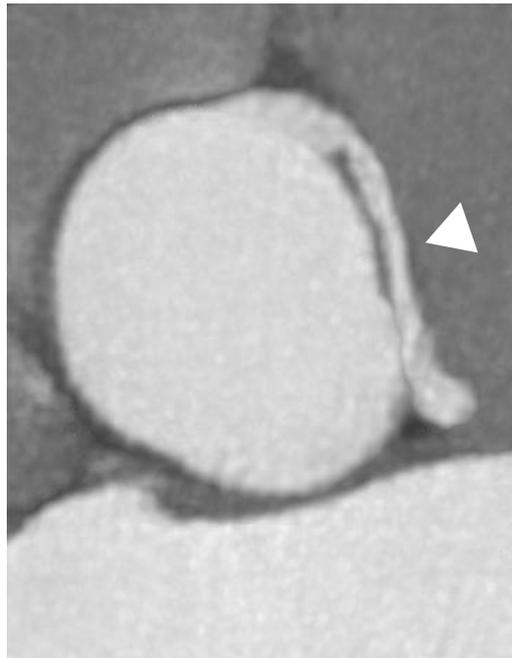
Analyse tomographique coronaire droite avec trajet interartériel



Analyse tomographique d'un passage intramural aortique

- Degré d'excentricité (grand axe/petit axe) ≥ 2.0
 - Angle de connexion $\leq 35^\circ$
 - Réduction de diamètre $\geq 50\%$
 - Réduction de surface $\geq 50\%$
-
- **≥ 3 critères présents** : passage intramural aortique certain
 - **2 critères présents** : passage intramural aortique incertain
 - **< 2 critères présents** : passage intramural aortique absent

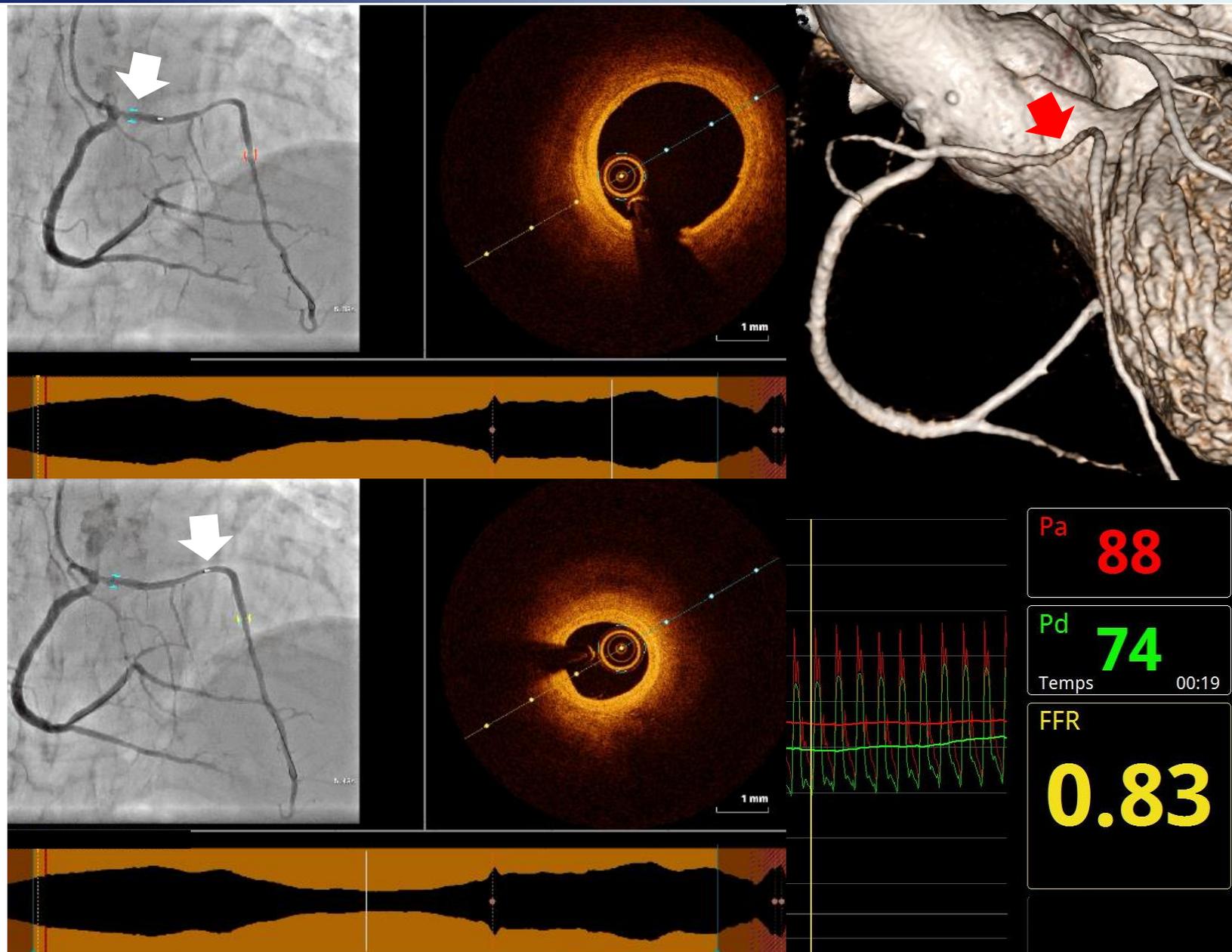
Tronc commun avec trajet interartériel



Tronc commun avec trajet rétropulmonaire

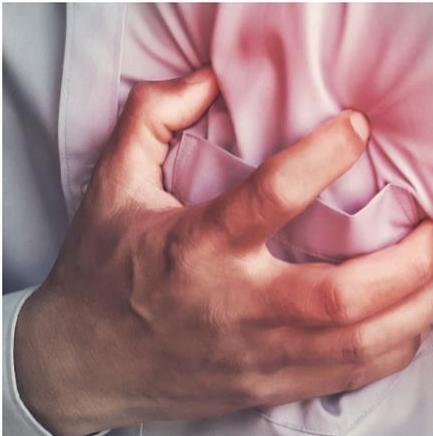


Tronc commun
avec trajet
rétropulmonaire

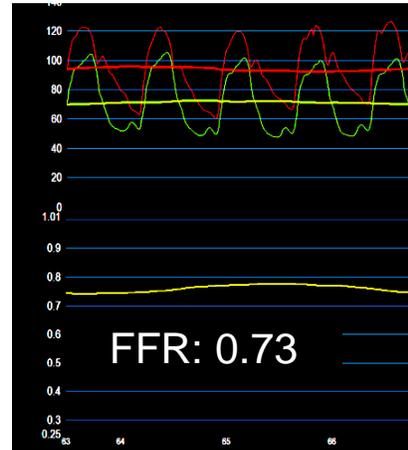
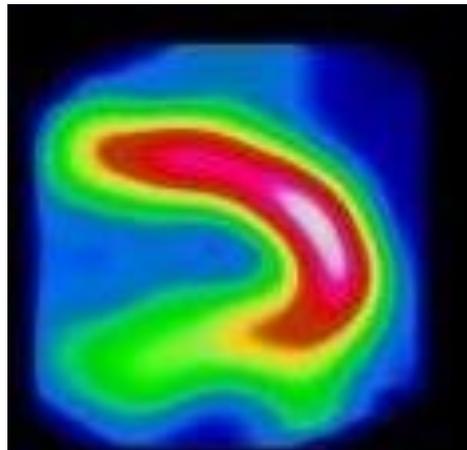


- Embryologie et anatomie
- Classification
- Prévalence
- Imagerie
- **Ischémie myocardique**
- Mort subite
- Dépistage
- Prise en charge
- Chirurgie
- Angioplastie
- Activités sportives

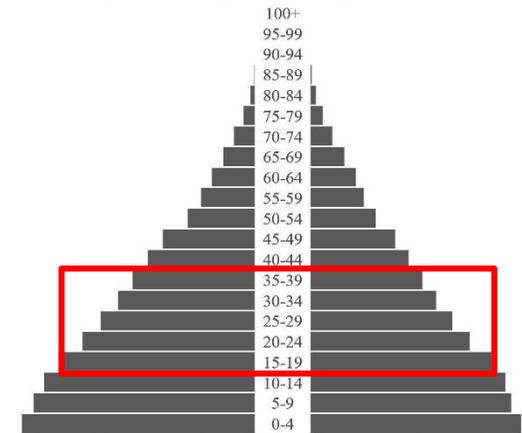
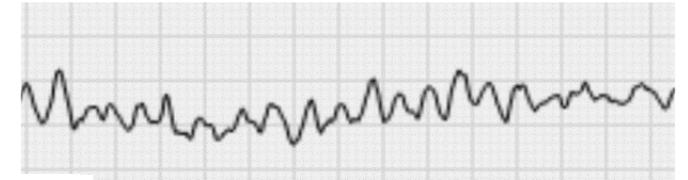
Symptômes ischémiques



Ischémie myocardique



ACR/MS



Syncope d'effort

Texas Heart Institute Journal

Case Reports

Repeated Syncope During Exercise as a Result of Anomalous Origin of Left Coronary Artery With Intramural Aortic Course in a Teenage Boy

Amioka N Tex Heart Inst J 2022

- Garçon de 17 ans
- Activité sportive (course à pied)
- Syncope d'effort
- Test d'effort
- ANOCOR gauche
- Pas d'arythmie ventriculaire

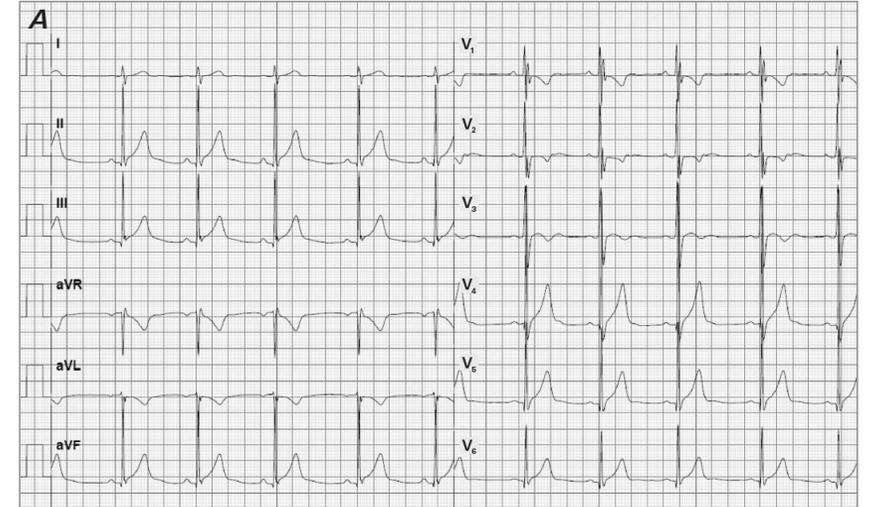
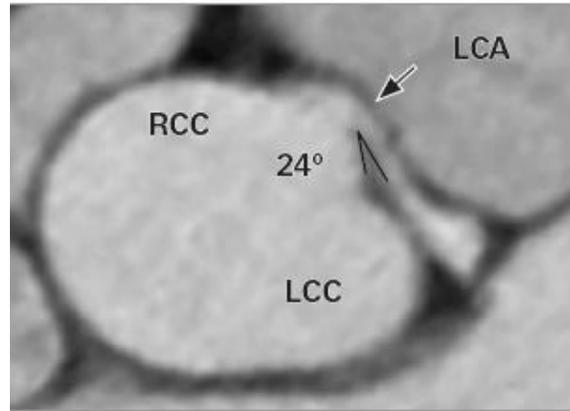
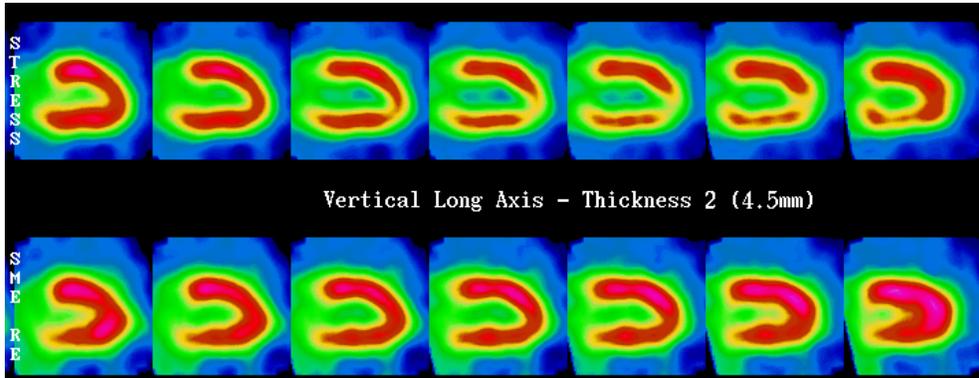


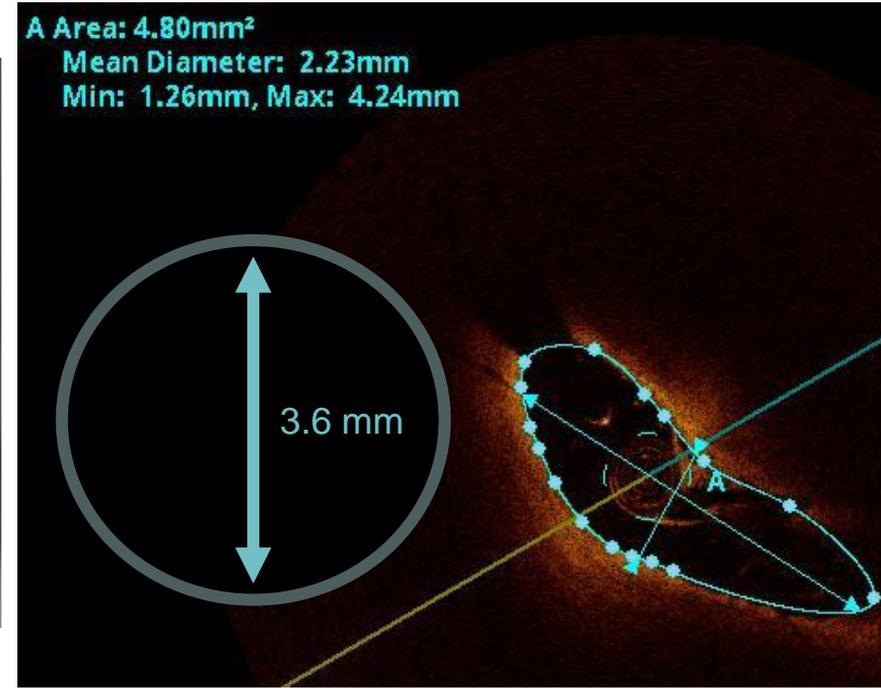
Fig. 1 Findings of an ECG before and after a treadmill exercise stress test. **A)** Before exercise, there was no specific finding on the ECG other than incomplete right bundle branch block. **B)** On the exercise stress test, the ECG showed ST-segment elevation at lead aVR and ST-segment depression at other leads accompanied by a decrease in blood pressure (99/56 mm Hg to 68/38 mm Hg), chest pain, and faintness.

ECG, electrocardiogram.

Ischémie myocardique documentée



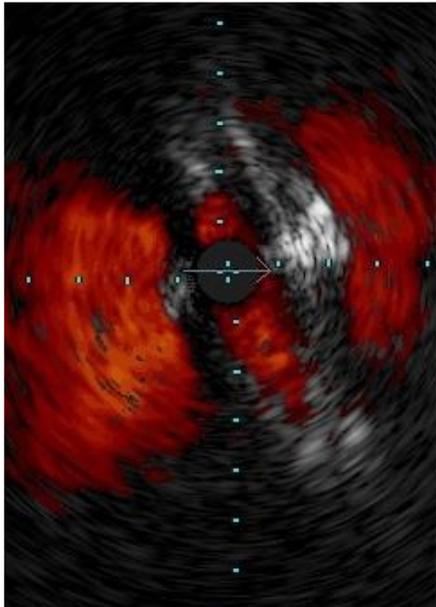
Prévalence ≈ 10-15%



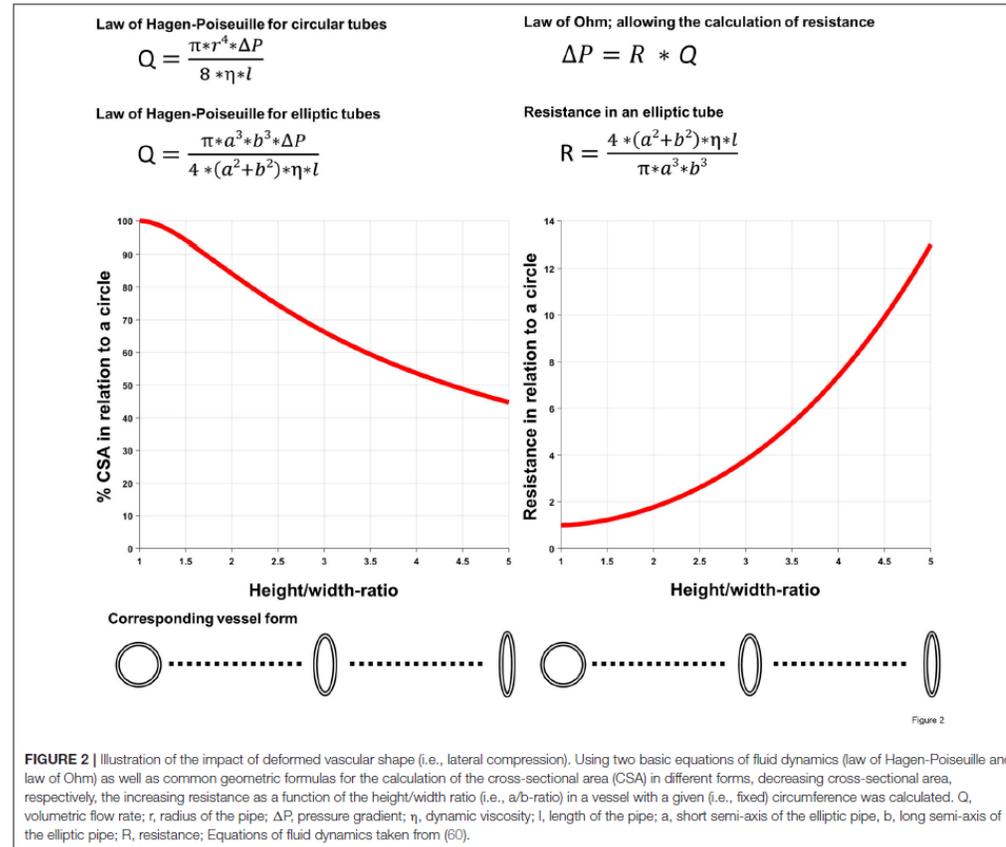
Diamètre luminal normal : 3.6 mm
Diamètre luminal minimal : 1.3 mm
Réduction de diamètre luminal : **64%**

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

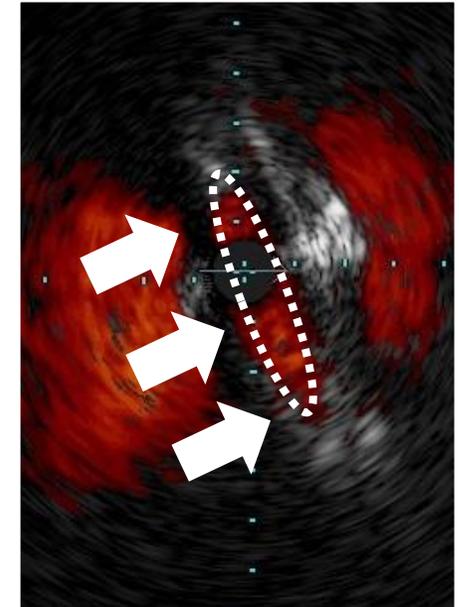
Two-Tier Concept Fixed Component



Ischémie myocardique



Two-Tier Concept Dynamic Component



Recherche d'ischémie myocardique

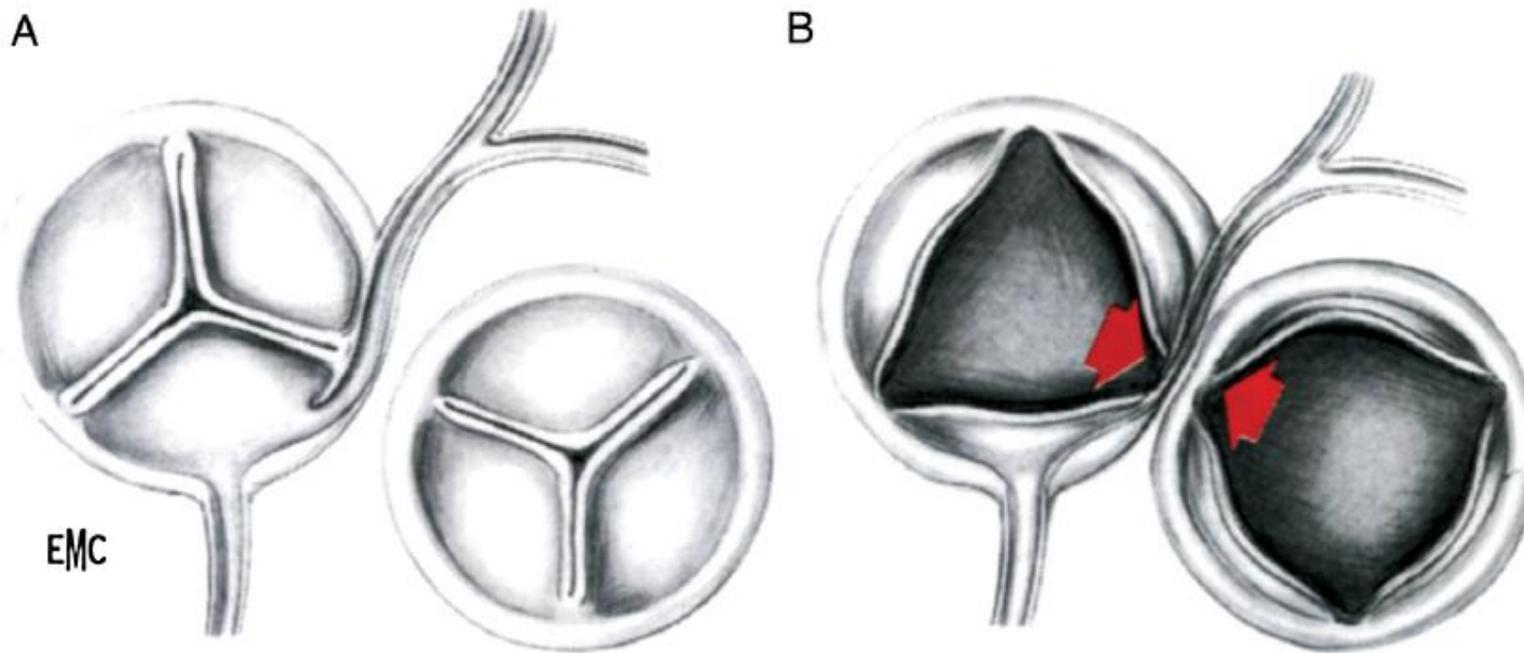
TABLE 3 | Overview of possible stress protocols in assessing patients with ACAOS.

	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.



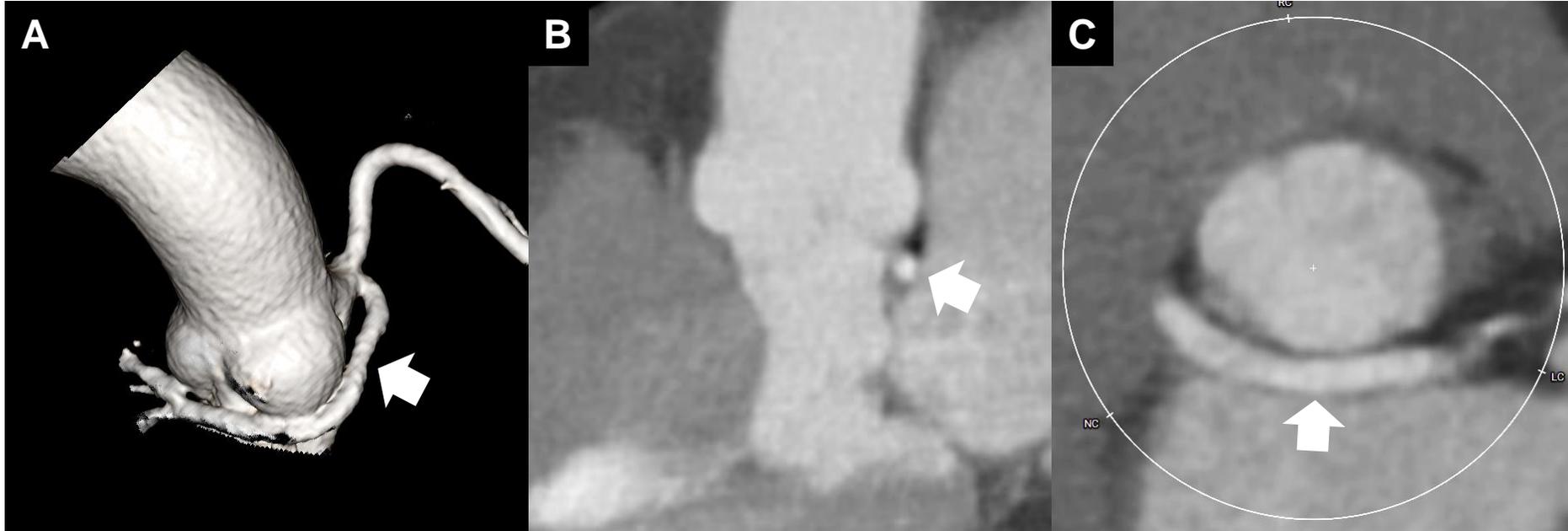
Exertional dynamic compression



Raisky O, Vouhé P. EMC 2007.

Never demonstrated

Compression extrinsèque d'un trajet rétroaortique

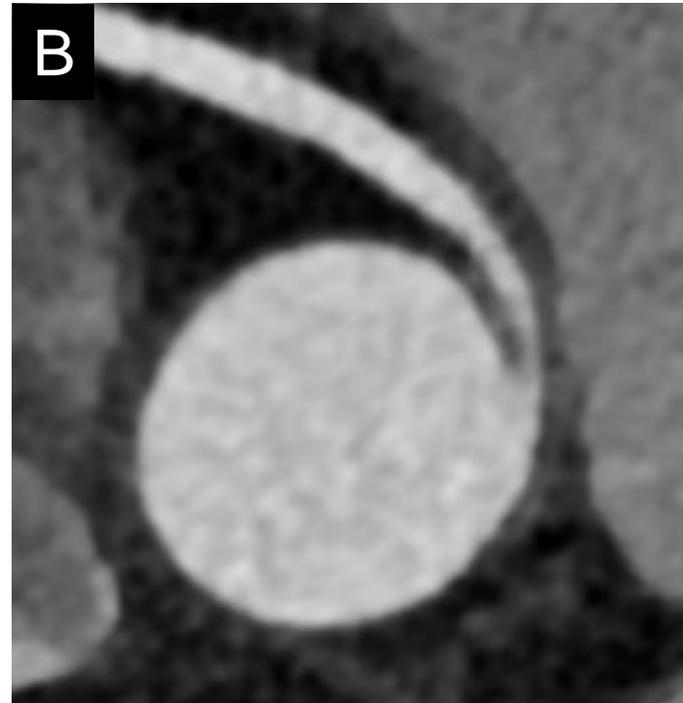
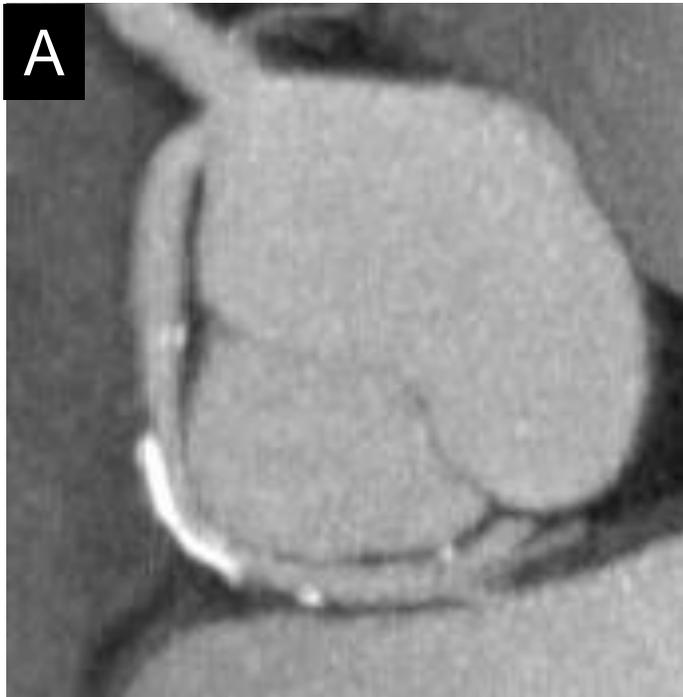


Chirurgie aortique valvulaire



Transcatheter aortic valve implantation (TAVI)

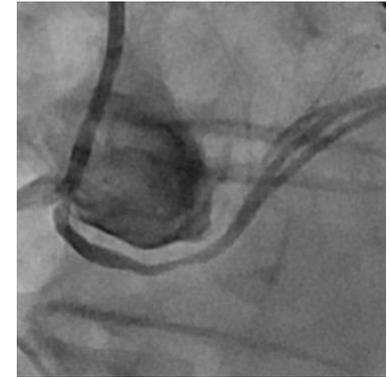
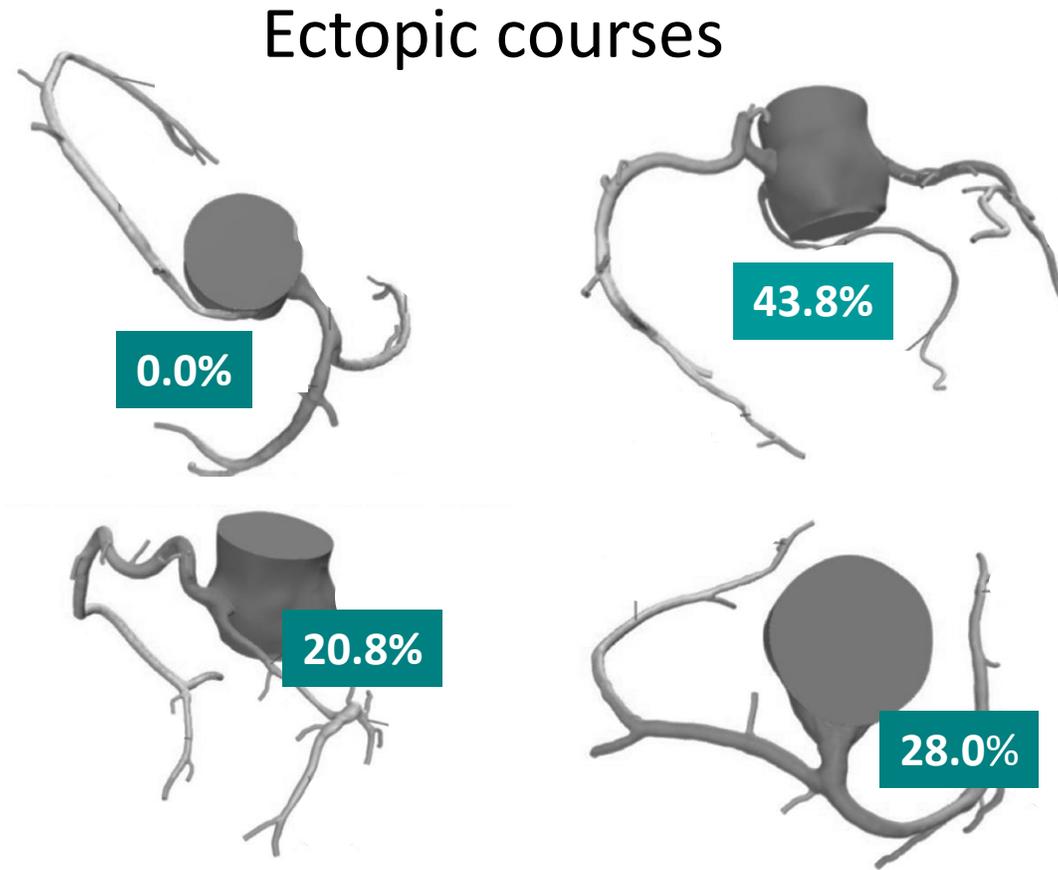
Athérome coronaire et trajets ectopiques



Prevalence and location of coronary artery disease



**AAOCA (RCA)
with interarterial course
N = 122**



**AAOCA (Cx artery)
with retroaortic course
N = 219**

**AAOCA (LM artery)
with subpulmonic course
N = 24**

**AAOCA (LM artery)
with prepulmonic course
N = 25**

- Embryologie et anatomie
- Classification
- Prévalence
- Imagerie
- Ischémie myocardique
- **Mort subite**
- Dépistage
- Prise en charge
- Chirurgie
- Angioplastie
- Activités sportives

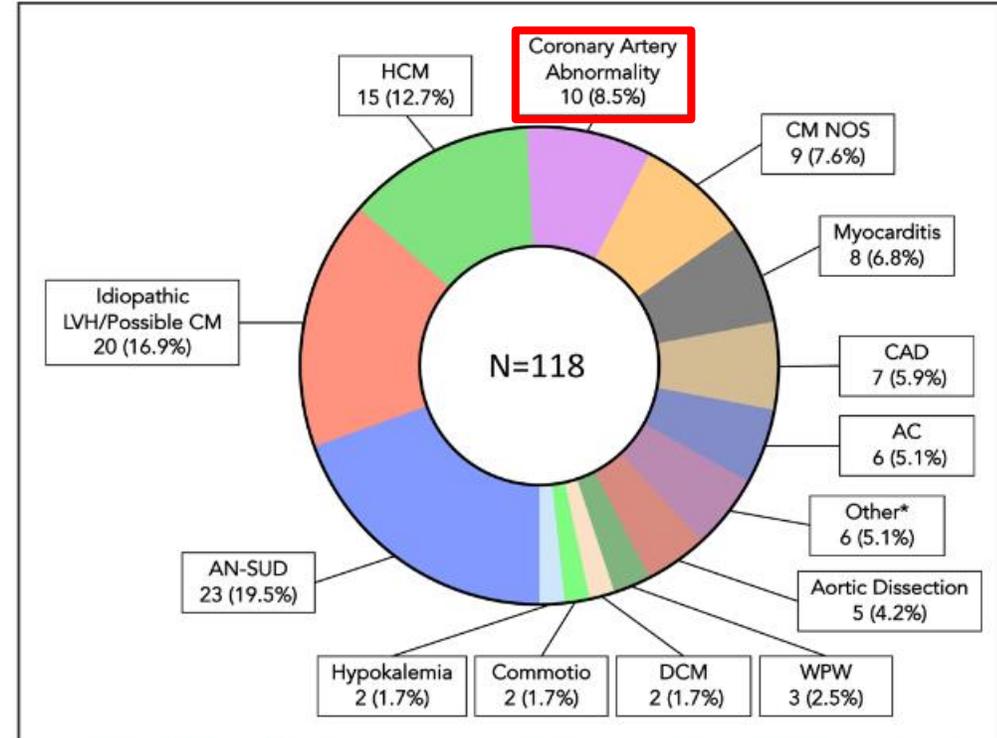


Figure 3. Causes of sudden cardiac death or findings on cardiac autopsy among National Collegiate Athletic Association athletes (n=118).

*Other: 1 each of long QT syndrome, complications of congenital heart disease, idiopathic left ventricular hypertrophy (LVH)/possible sickle cell trait, Kawasaki disease, complications of congenital heart disease, idiopathic left ventricular hypertrophy (LVH)/possible sickle cell trait, Kawasaki disease, complications after heart transplant, and sudden cardiac death in individual with pacemaker for idiopathic atrioventricular block. AC indicates arrhythmogenic cardiomyopathy; AN-SUD, autopsy-negative sudden unexplained death; CAD, coronary artery disease; CM, cardiomyopathy; DCM, dilated cardiomyopathy; HCM, hypertrophic cardiomyopathy; NOS, not otherwise specified; and WPW, Wolff-Parkinson-White syndrome.

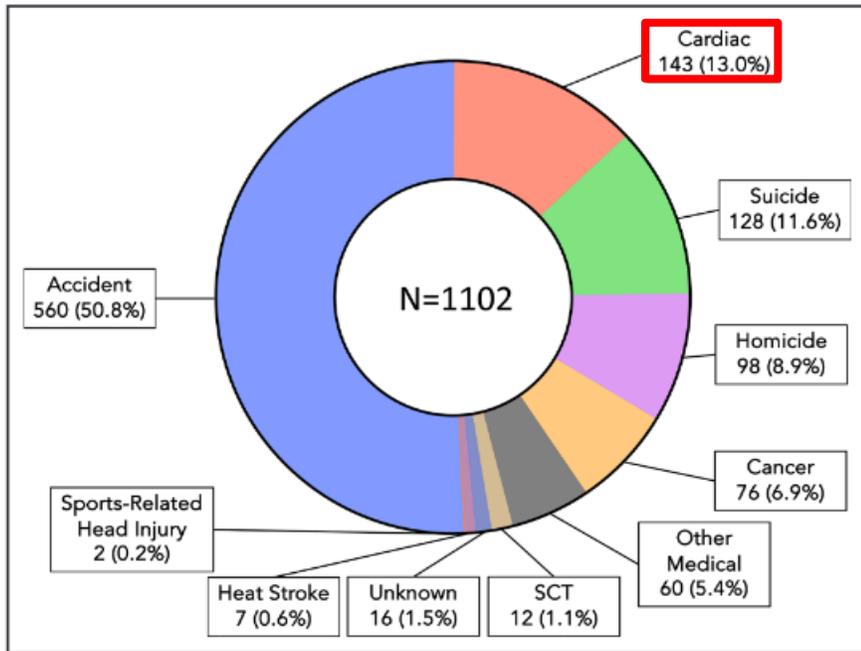


Figure 1. Causes of death among National Collegiate Athletic Association athletes (n=1102). SCT indicates sickle cell trait.

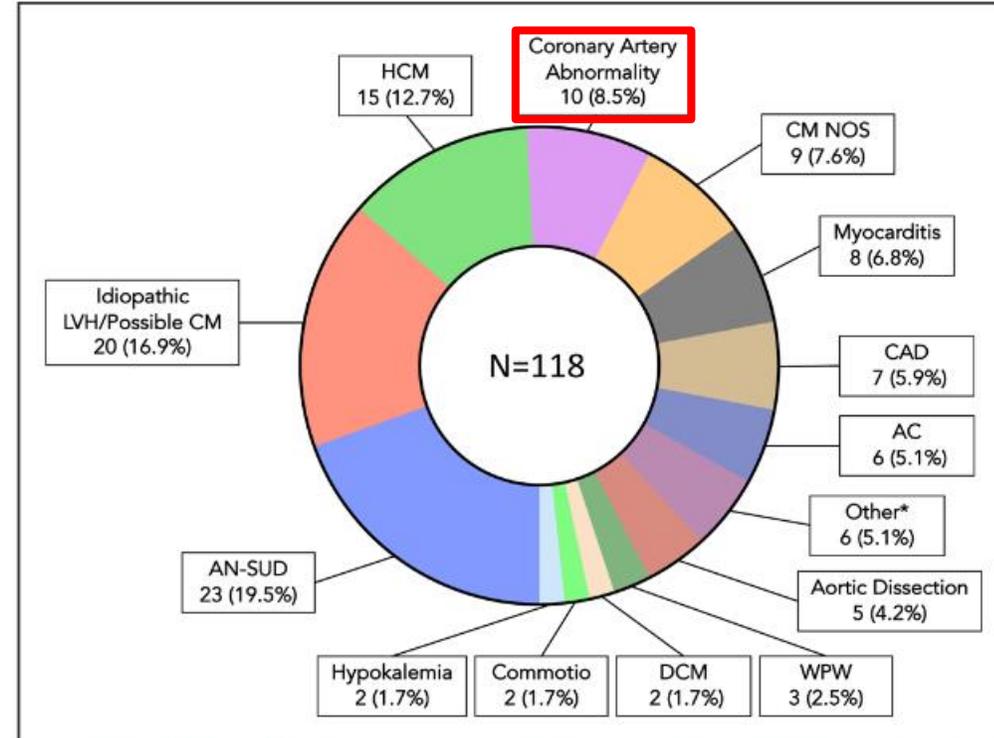


Figure 3. Causes of sudden cardiac death or findings on cardiac autopsy among National Collegiate Athletic Association athletes (n=118).

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Q. Please give me the incidence of sudden cardiac death for each AAOCA.

1. Anomalous Origin of the Right Coronary Artery (ARCA):

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.

2. Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA):

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.

3. Anomalous Origin of the Left Coronary Artery from the Right Sinus (ALCA):

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.

4. Other Anomalous Origins (e.g., Anomalous Origin of Circumflex 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.

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

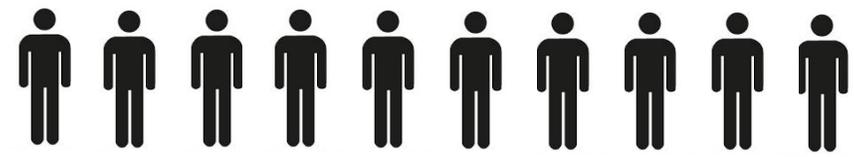
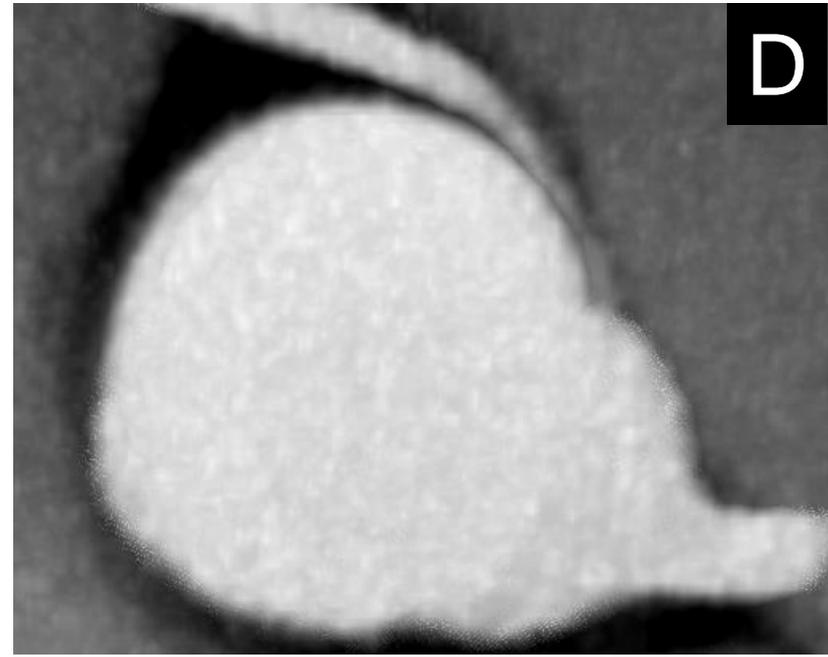
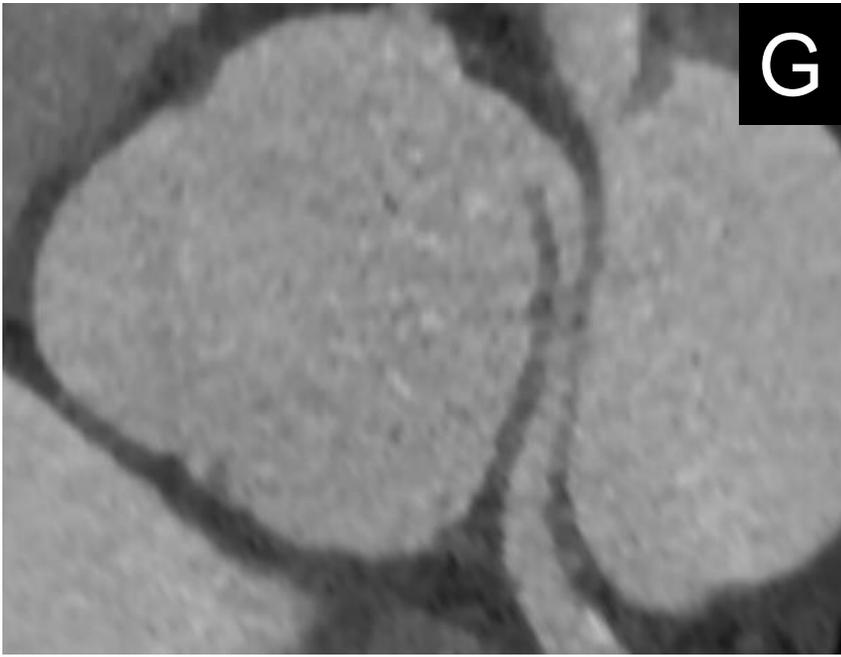
** Prévalence à la naissance (estimations)

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%

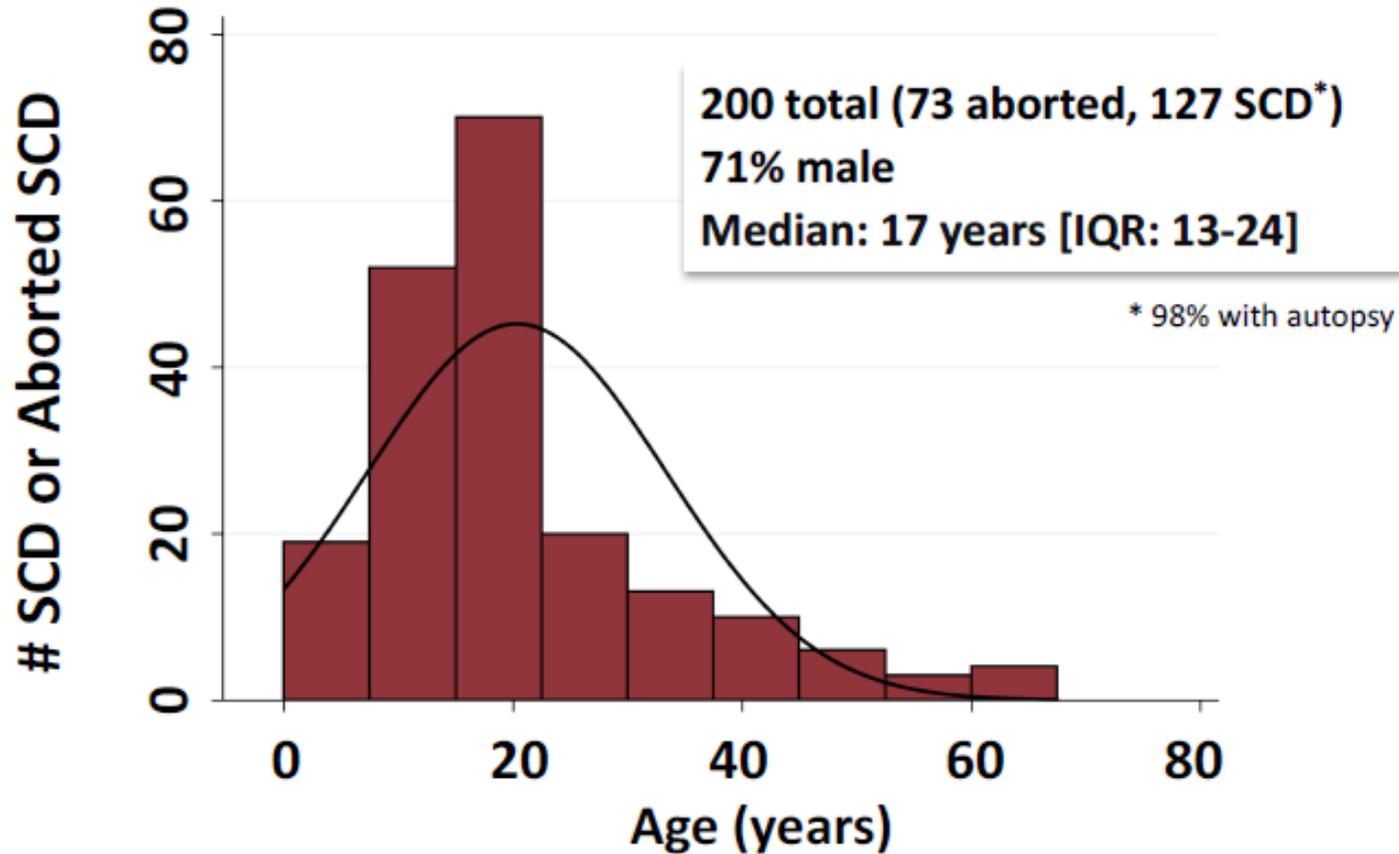
* Anomalie de connexion avec trajet interartériel

** Incidence annuelle de mort subite (estimations)

Sur-risque de mortalité pour les formes gauches X 10



Age of SCD or Aborted SCD Attributed to AAOCA



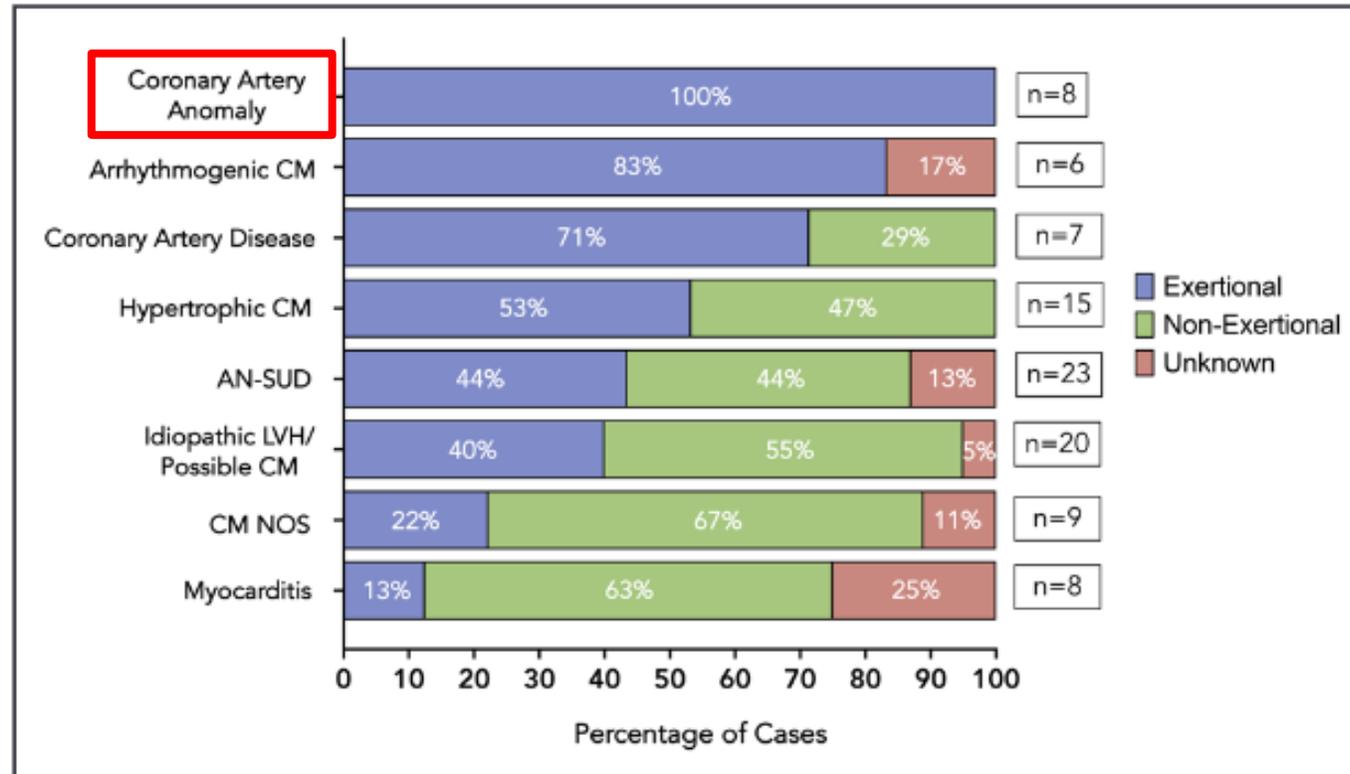
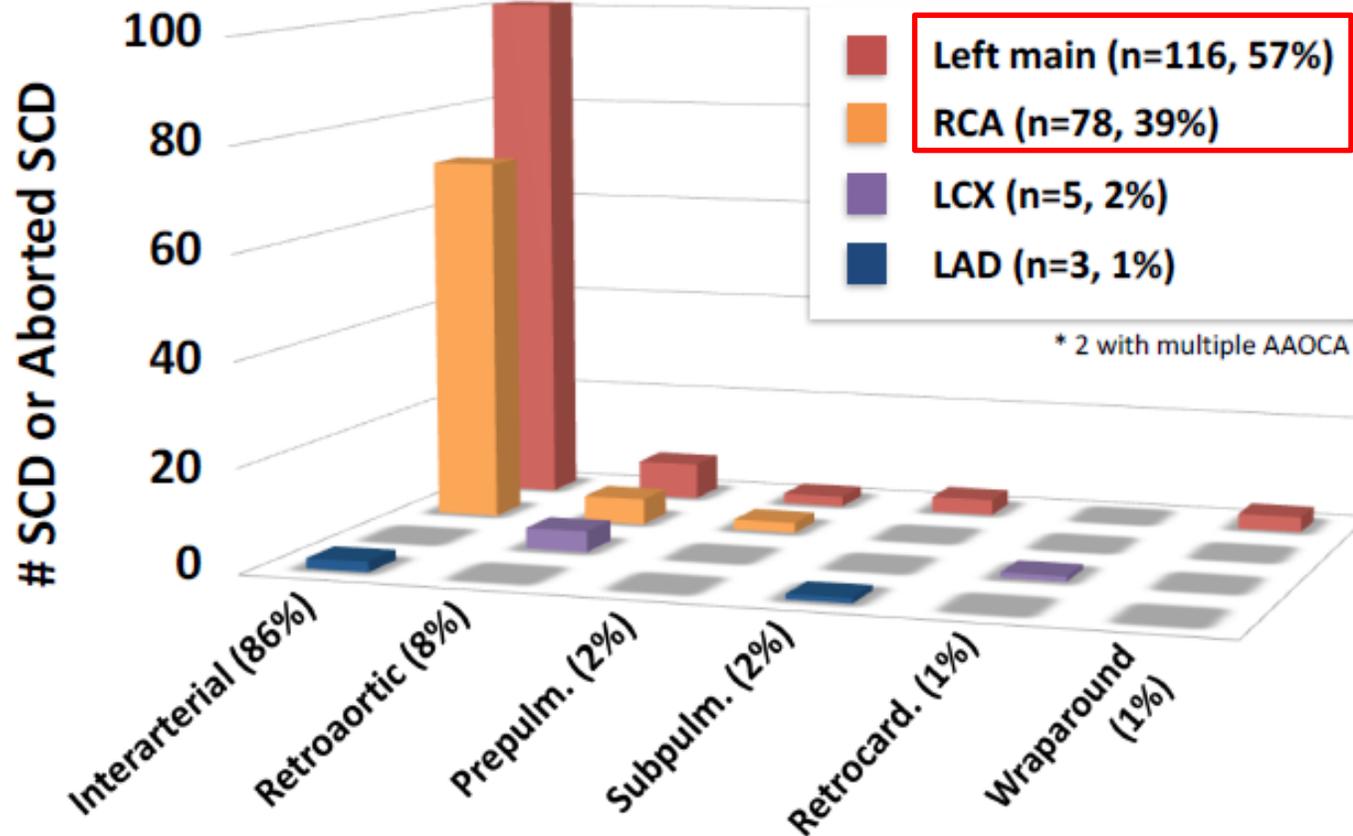


Figure 4. Exertional status at time of death by common causes of sudden cardiac death.

AN-SUD indicates autopsy-negative sudden unexplained death; CM, cardiomyopathy; LVH, left ventricular hypertrophy; NOS, not otherwise specified; and SCD, sudden cardiac death.

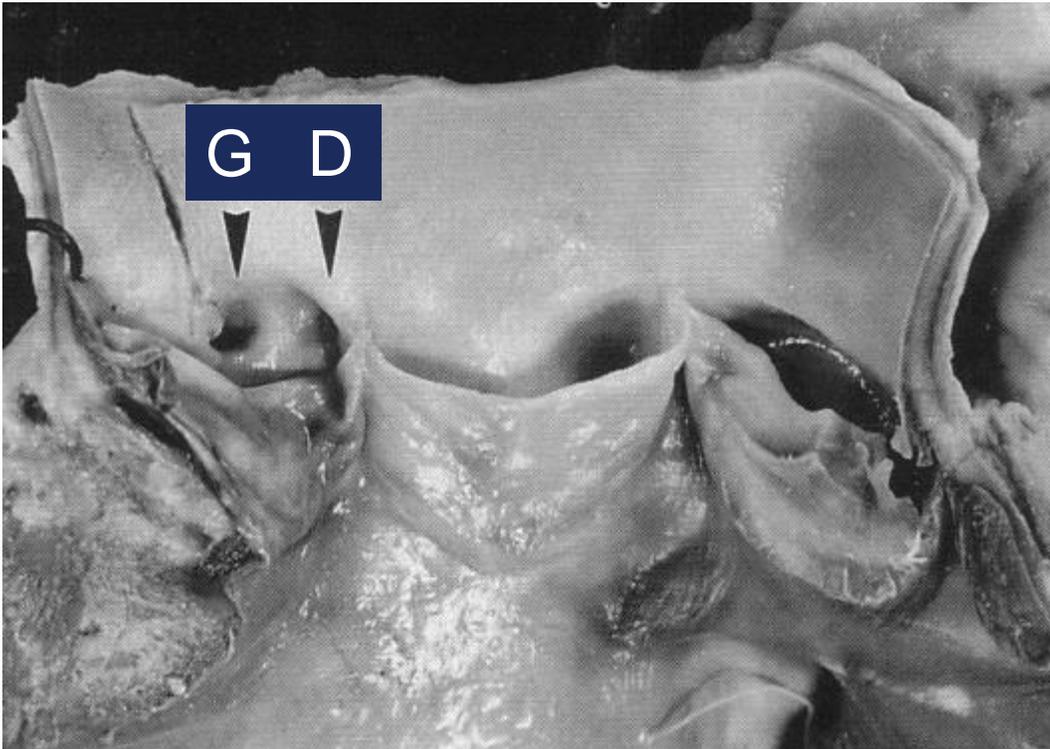
AAOCA Anatomy in SCD/Aborted SCD Patients



Cause de la mort subite : rythmique

1990

Mort subite



Corrado et al. Br Heart J. 1992.

2010

Arrêt cardiaque récupéré

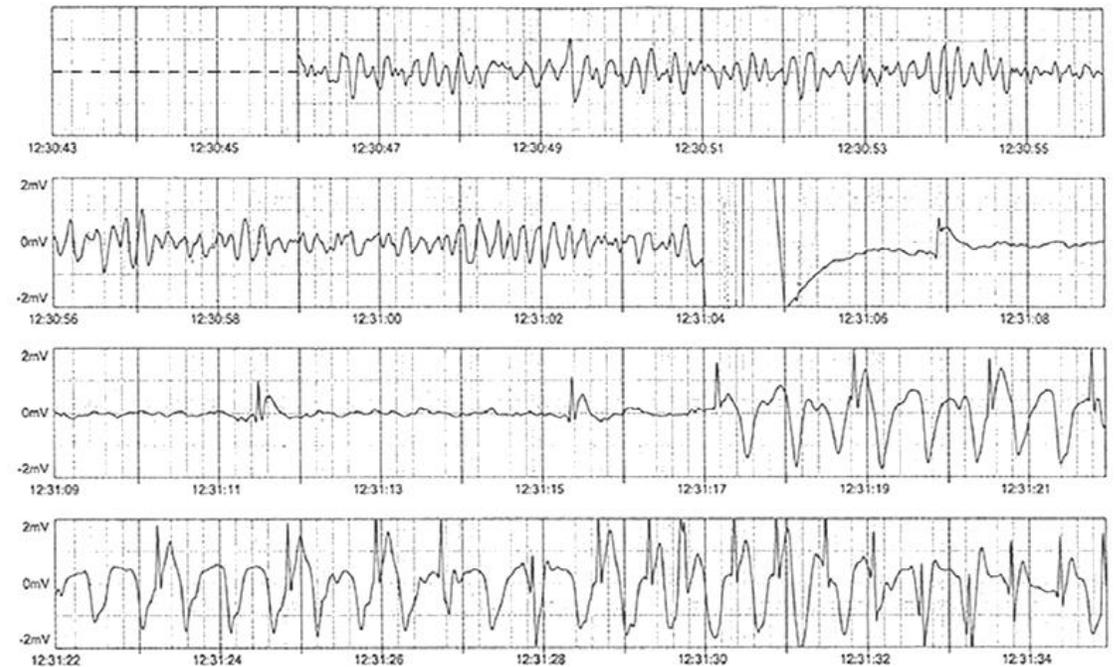
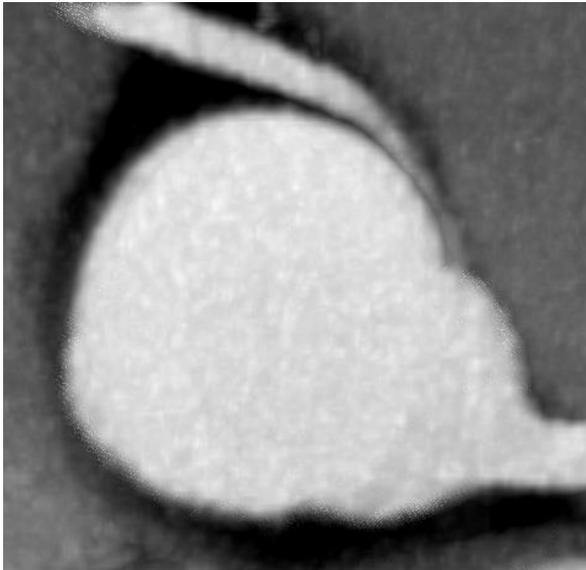
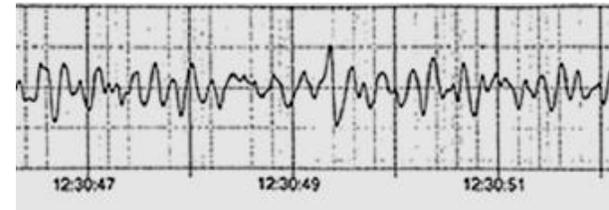


Figure 1. ECG recording from an automated external defibrillator

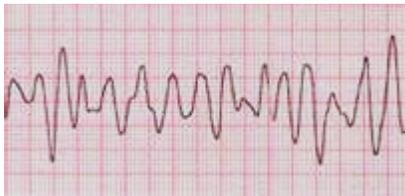
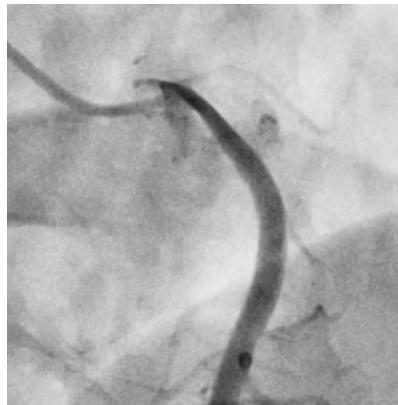
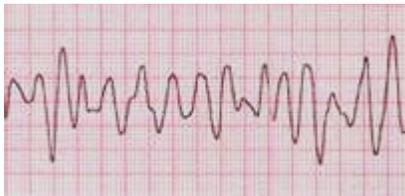
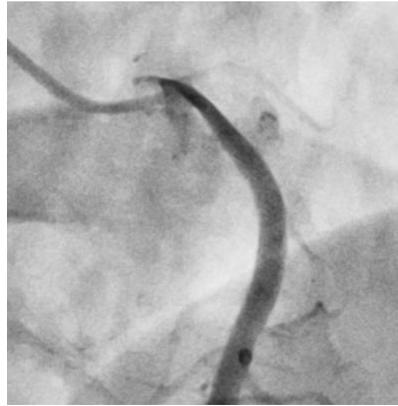
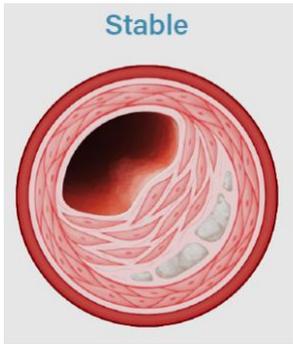
Mécanisme(s) de la fibrillation ventriculaire



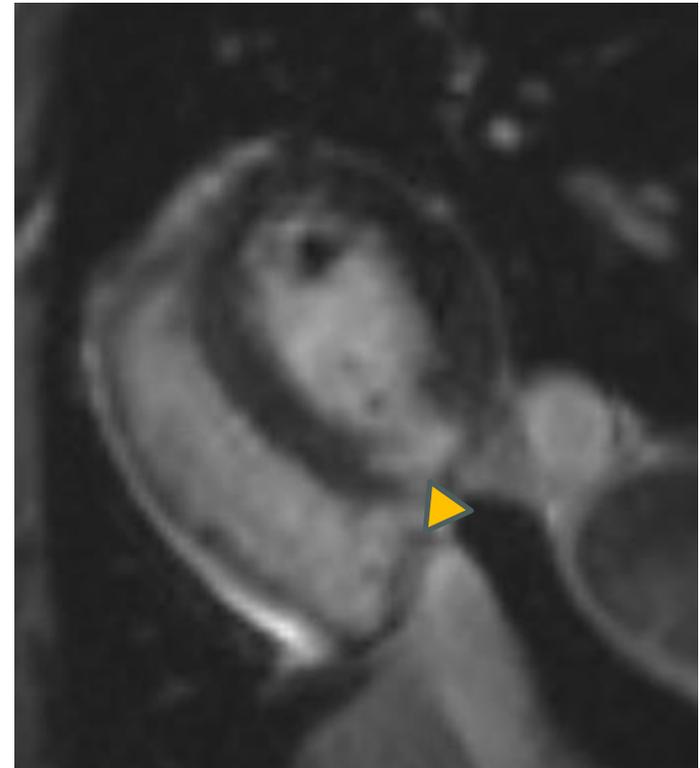
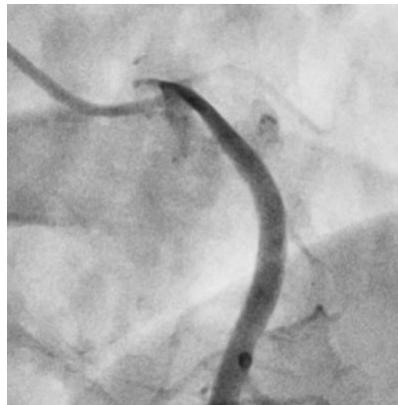
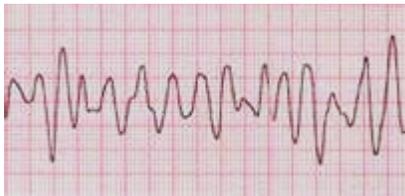
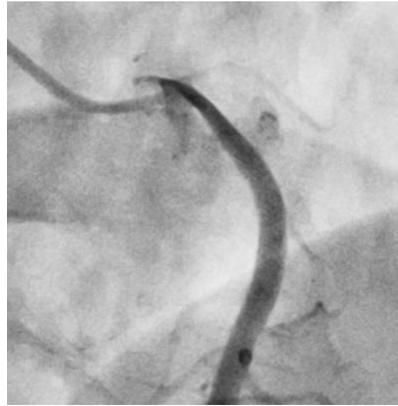
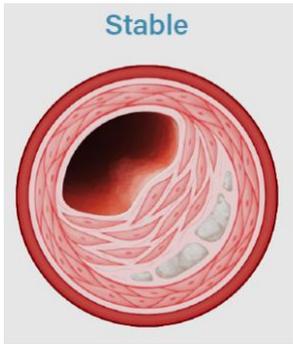
24/07/2022
vers 12.30



Mécanisme(s) de la fibrillation ventriculaire



Mécanisme(s) de la fibrillation ventriculaire



Stratification du risque de mort subite

- ANOCOR gauche / ANOCOR droite
- Trajet interartériel
- Passage intramural aortique
- Symptomatologie d'allure ischémique
- Ischémie myocardique documentée avec imagerie
- Activité physique sportive d'intensité élevée
- Age < 35 ans

Mais... **pas encore de score de risque validé**

Mort subite ou arrêt cardiaque récupéré sur ANOCOR

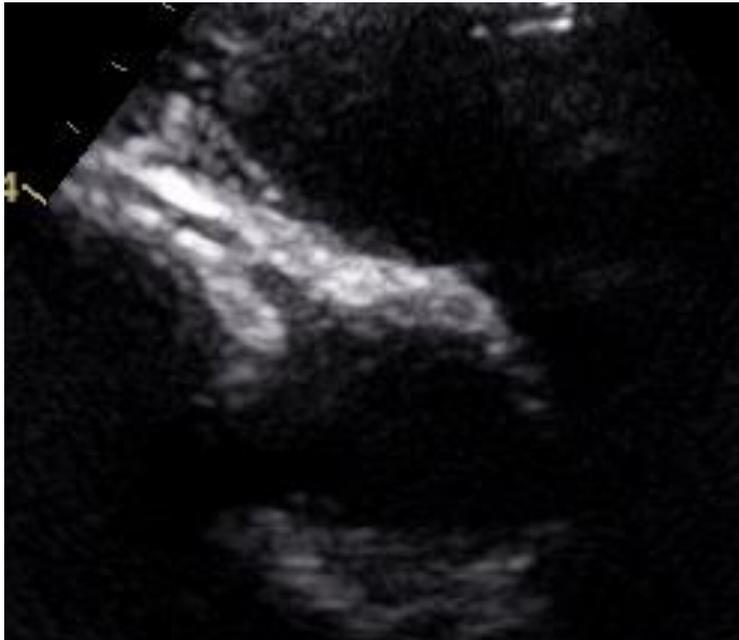
- Anomalie cardiaque congénitale présente à la naissance
- Anomalie cardiaque génétique : oui / non
- Caractère héréditaire : oui / non
- Dépistage familial : oui / non

- Embryologie et anatomie
- Classification
- Prévalence
- Imagerie
- Ischémie myocardique
- Mort subite
- **Dépistage**
- Prise en charge
- Chirurgie
- Angioplastie
- Activités sportives

Faut-il dépister les ANOCOR chez le jeune sportif ?

Echocardiographie transthoracique

coronaire droite normale

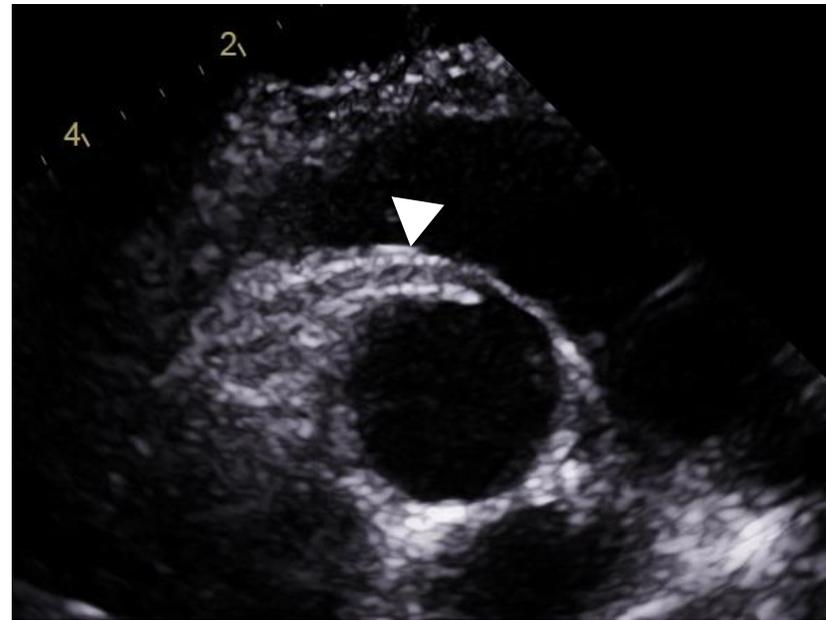


coronaire gauche normale



Faut-il dépister les ANOCOR chez le jeune sportif ?

Echocardiographie transthoracique



ANOCOR droite

Faut-il dépister les ANOCOR chez le jeune sportif ?

Exemple de dépistage d'ANOCOR

Prévalence : 3/1.000

Population : 7.000

Test : échocardiogramme transthoracique

Sensibilité : 90%

Spécificité : 90%

- 2 faux négatifs
- 6.298 vrais négatifs
- 682 faux positifs
- 18 vrais positifs

Faut-il dépister les ANOCOR chez le jeune sportif ?

Explorations Fonctionnelles Cardio-Vasculaires

Laboratoire d'échocardiographie

Echodoppler cardiaque transthoracique

Echographe : GE Vivid 9

Réalisé le 22/07/2021

M. né le : 01/01/2004

Motif : bilan d'aptitude sportive, précordialgies atypiques

Ventricule gauche non dilaté, non hypertrophié de fonction systolique normale et de cinétique homogène.

Profil et pressions de remplissage gauches normaux.

Pas de valvulopathie significative.

Aorte de taille normale.

Cavités droites non dilatées.

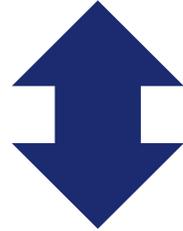
Pression artérielle pulmonaire systolique et pression de l'oreillette droite normales.

Pas d'épanchement péricardique.

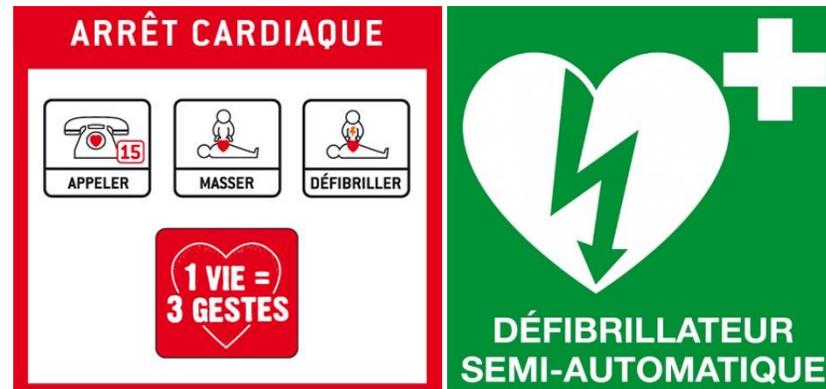
Au total : examen dans les limites de la normale.



Proactive strategy with preparticipation screening

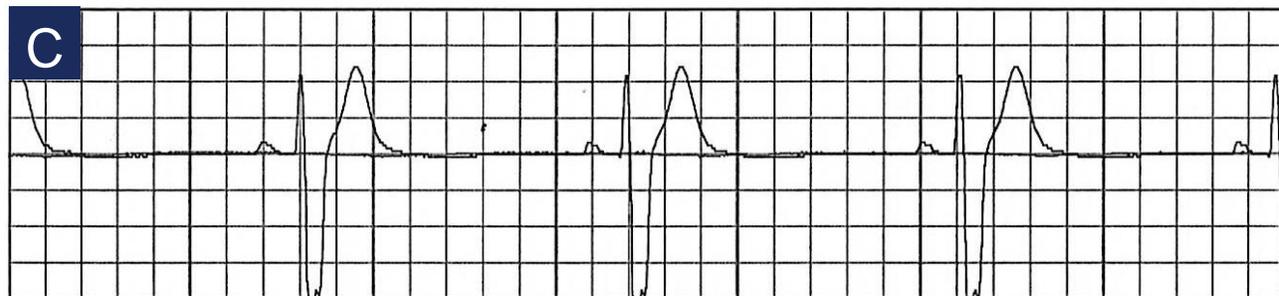
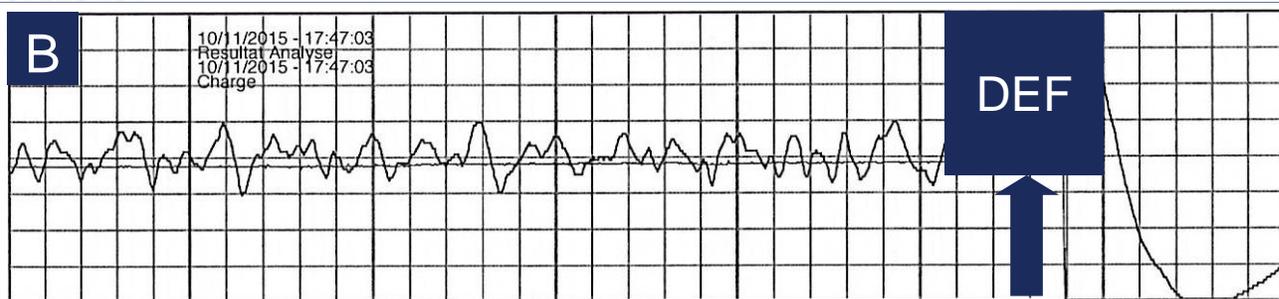


Reactive strategy with CPR/AED



Sudden cardiac arrest – High school sports activities

15-year old boy – Left coronary anomaly



- Embryologie et anatomie
- Classification
- Prévalence
- Imagerie
- Ischémie myocardique
- Mort subite
- Dépistage
- **Prise en charge**
- Chirurgie
- Angioplastie
- Activités sportives

Guidelines for cardiac arrest

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. ^{c,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

CA, cardiac arrest; VA, ventricular arrhythmia.

^aClass of recommendation.

^bLevel of evidence.

^cHigh-risk anatomy is defined as interarterial course, slit-like shaped orifice, high orifice, acute-angle take-off, and intramural course and its length.

Guidelines for cardiac arrest

2017 AHA/ACC/HRS Guideline for Management of Patients With Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death: Executive Summary

4.3. Surgery and Revascularization Procedures in Patients With Ischemic Heart Disease

Recommendations for Surgery and Revascularization Procedures in Patients With Ischemic Heart Disease		
References that support the recommendations are summarized in Online Data Supplement 11.		
COR	LOE	Recommendations
I	B-NR	1. Patients with sustained VA and survivors of SCA should be evaluated for ischemic heart disease, and should be revascularized as appropriate (1-4).
I	C-EO	2. In patients with anomalous origin of a coronary artery suspected to be the cause of SCA, repair or revascularization is recommended.



???

SCA: sudden cardiac arrest

2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease: Executive Summary

COR	LOE	Recommendations
Therapeutic		
I	B-NR	1. <u>Surgery</u> is recommended for AAOCA from the left sinus or AAOCA from the right sinus for symptoms or diagnostic evidence consistent with coronary ischemia attributable to the anomalous coronary artery. ^{S4.4.5.2-1-S4.4.5.2-3}
IIa	C-LD	2. <u>Surgery</u> is reasonable for anomalous aortic origin of the left coronary artery from the right sinus in the absence of symptoms or ischemia. ^{S4.4.5.2-4-S4.4.5.2-6}
IIa	C-EO	3. <u>Surgery</u> for AAOCA is reasonable in the setting of ventricular arrhythmias.
IIb	B-NR	4. <u>Surgery</u> or continued observation may be reasonable for asymptomatic patients with an anomalous left coronary artery arising from the right sinus or right coronary artery arising from the left sinus without ischemia or anatomic or physiological evaluation suggesting potential for compromise of coronary perfusion (eg, intramural course, fish-mouth-shaped orifice, acute angle). ^{S4.4.5.2-4-S4.4.5.2-6}

Stout KK Circulation 2019.

2020 ESC Guidelines for the management of adult congenital heart disease

Anomalous aortic origin of the coronary artery		
<u>Surgery</u> 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
<u>Surgery</u> should be considered in <i>asymptomatic</i> patients with AAOCA (right or left) and evidence of myocardial ischaemia.	IIa	C
<u>Surgery</u> should be considered in <i>asymptomatic</i> patients with AAOLCA and no evidence of myocardial ischaemia but a high-risk anatomy. ^c	IIa	C
<u>Surgery</u> may be considered for symptomatic patients with AAOCA even if there is no evidence of myocardial ischaemia or high-risk anatomy. ^c	IIb	C
<u>Surgery</u> 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
<u>Surgery</u> is not recommended for AAORCA in asymptomatic patients without myocardial ischaemia and without high-risk anatomy. ^c	III	C

Baumgartner H Eur Heart J 2020.

Guidelines

Guidelines

2020 ESC Guidelines for the management of adult congenital heart disease

ANOCOR interartérielle symptomatique et avec ischémie myocardique

ANOCOR interartérielle asymptomatique et avec ischémie myocardique

ANOCOR interartérielle gauche asymptomatique et sans ischémie myocardique

ANOCOR interartérielle symptomatique et sans ischémie myocardique

ANOCOR interartérielle droite asymptomatique et sans ischémie myocardique

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 should be considered in asymptomatic patients with AAOCA (right or left) and evidence of myocardial ischaemia.	IIa	C
Surgery should be considered in asymptomatic 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 asymptomatic patients with AAOLCA without myocardial ischaemia and without high-risk anatomy ^c when they present at young age (<35 years).	IIb	C
Surgery is not recommended for AAORCA in asymptomatic patients without myocardial ischaemia and without high-risk anatomy. ^c	III	C

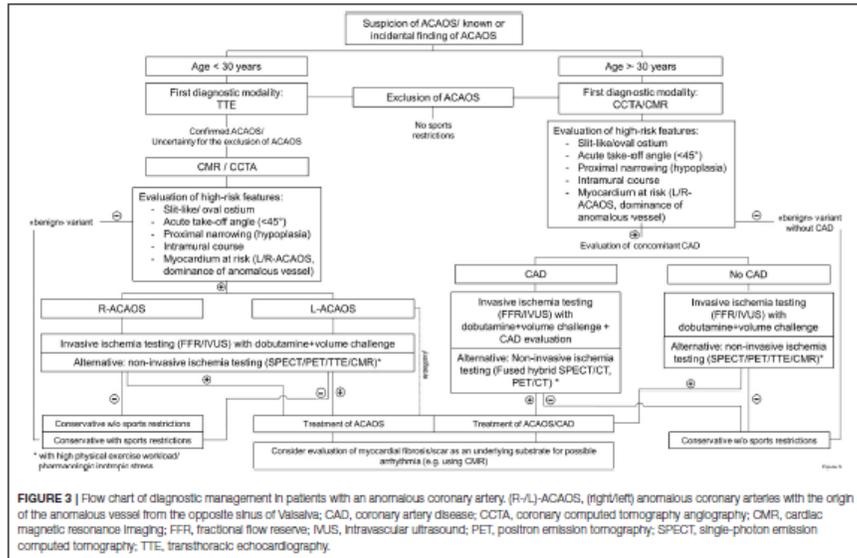


FIGURE 3 | Flow chart of diagnostic management in patients with an anomalous coronary artery. (R-/L-)ACAOS, (right/left) anomalous coronary arteries with the origin of the anomalous vessel from the opposite sinus of Valsalva; CAD, coronary artery disease; CCTA, coronary computed tomography angiography; CMR, cardiac magnetic resonance imaging; FFR, fractional flow reserve; IVUS, intravascular ultrasound; PET, positron emission tomography; SPECT, single-photon emission computed tomography; TTE, transthoracic echocardiography.

Bigler MR Front Cardiovasc Med 2021.

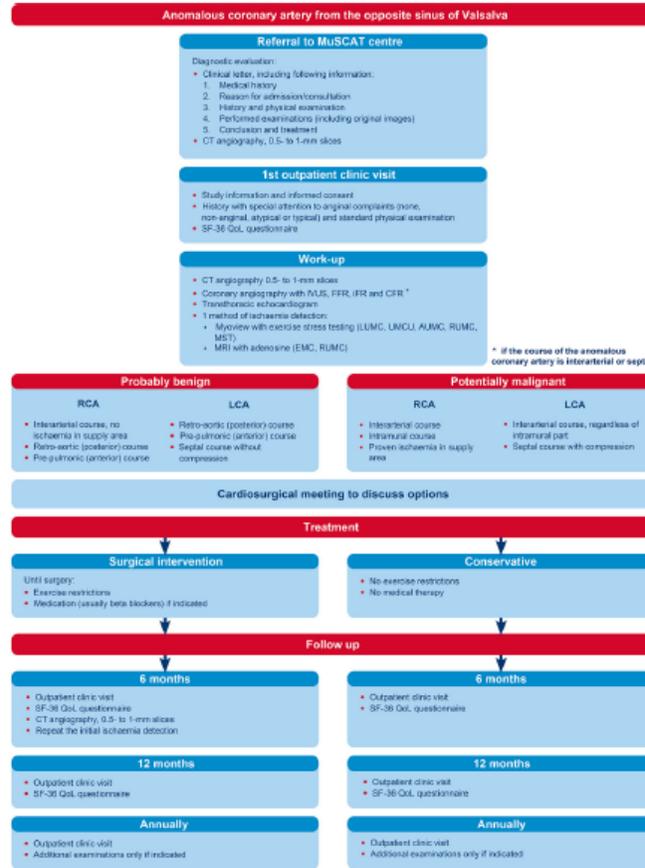


Fig. 2 Flowchart of study protocol. IVUS intravascular ultrasound, FFR fractional flow reserve, IFR instantaneous wave-free ratio, CFR coronary flow reserve, LUMC Laiden University Medical Centre, UMCU University Medical Centre Utrecht.

AUMC Amsterdam University Medical Centre, EMC Erasmus University Medical Centre, RUMC Radboud University Medical Centre, MST Medisch Spectrum Twente, RCA right coronary artery, LCA left coronary artery, QoL quality of life

Koppel CJ Neth Heart J 2022.

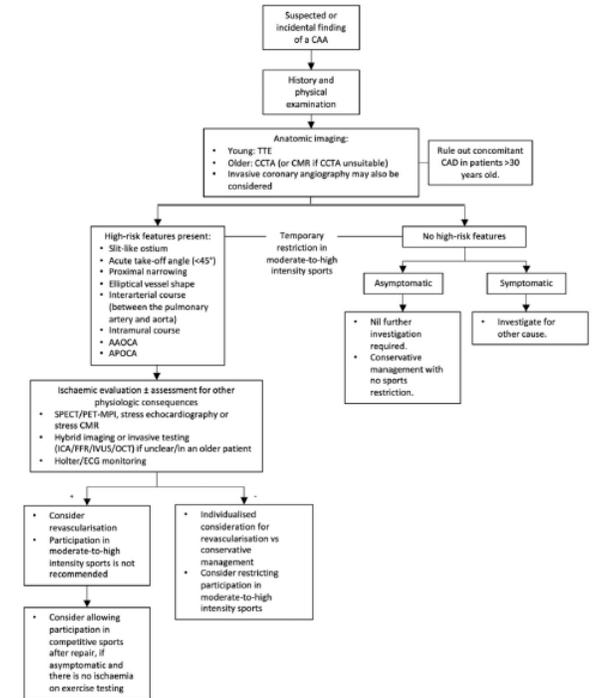


FIG 3. Proposed management algorithm for CAAs. APOCA, anomalous pulmonary origin of a coronary artery; AAOCA, anomalous aortic origin of a coronary artery; CAA, coronary artery anomaly; CAD, coronary artery disease; CCTA, coronary computed tomography angiography; CMR, cardiac magnetic resonance; ECG, electrocardiogram; MPI, myocardial perfusion imaging; SPECT, single-photon emission computed tomography; TTE, transthoracic echocardiogram. (Color version of figure is available online.)

Lau W Curr Probl Cardiol 2023.

- Embryologie et anatomie
- Classification
- Prévalence
- Imagerie
- Ischémie myocardique
- Mort subite
- Screening
- Prise en charge
- **Chirurgie**
- Angioplastie
- Activités sportives

Techniques chirurgicales



Main Options for the Treatment of AAOCA in Adult Patients

Coronary unroofing

Neo-ostium creation

Coronary artery bypass grafting

Coronary reimplantation

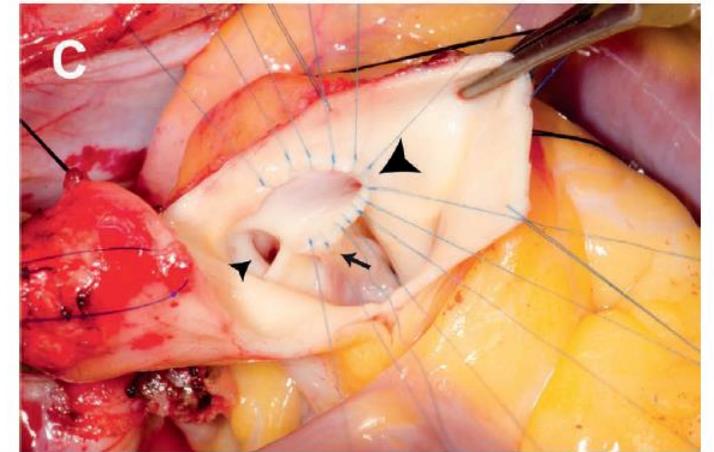
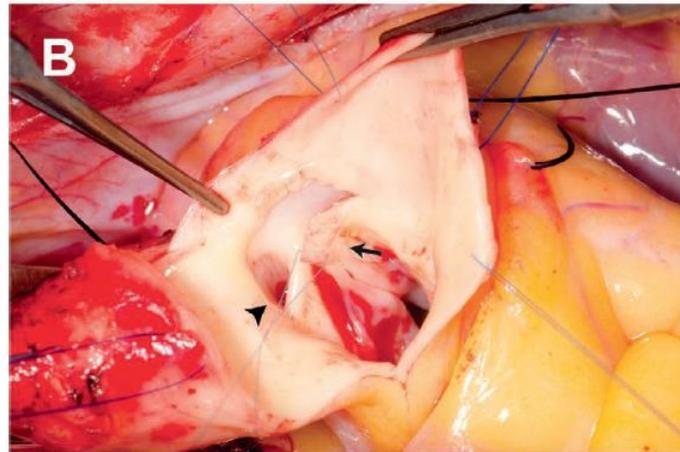
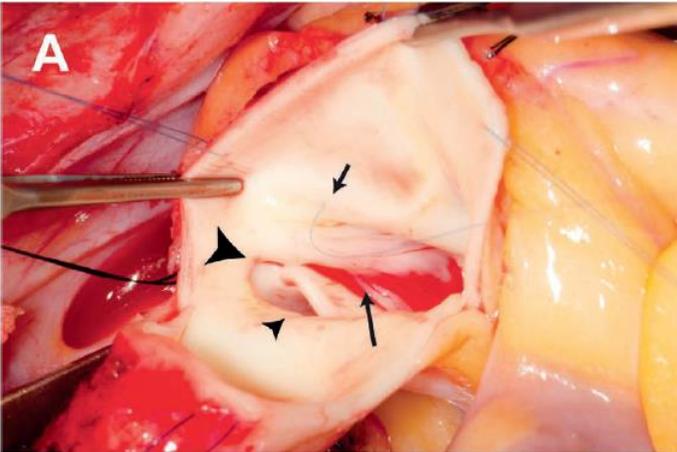
External unroofing of a transeptal AAOCA

VISUAL ABSTRACT Main surgical techniques for treatment of anomalous aortic origin of the coronary arteries in adult patients. Coronary unroofing, neo-ostium creation, coronary artery bypass grafting, coronary reimplantation, and external unroofing of a transeptal coronary artery with anomalous aortic origin. (AAOCA, anomalous aortic origin of the coronary arteries.)



Techniques chirurgicales

Unroofing technique

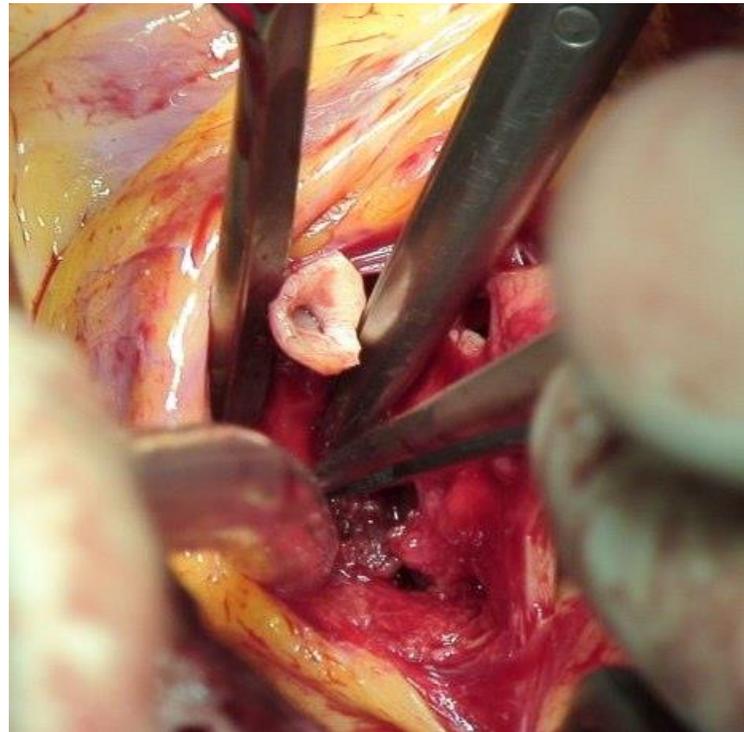


Molossi S MD Cardiovasc J 2019.

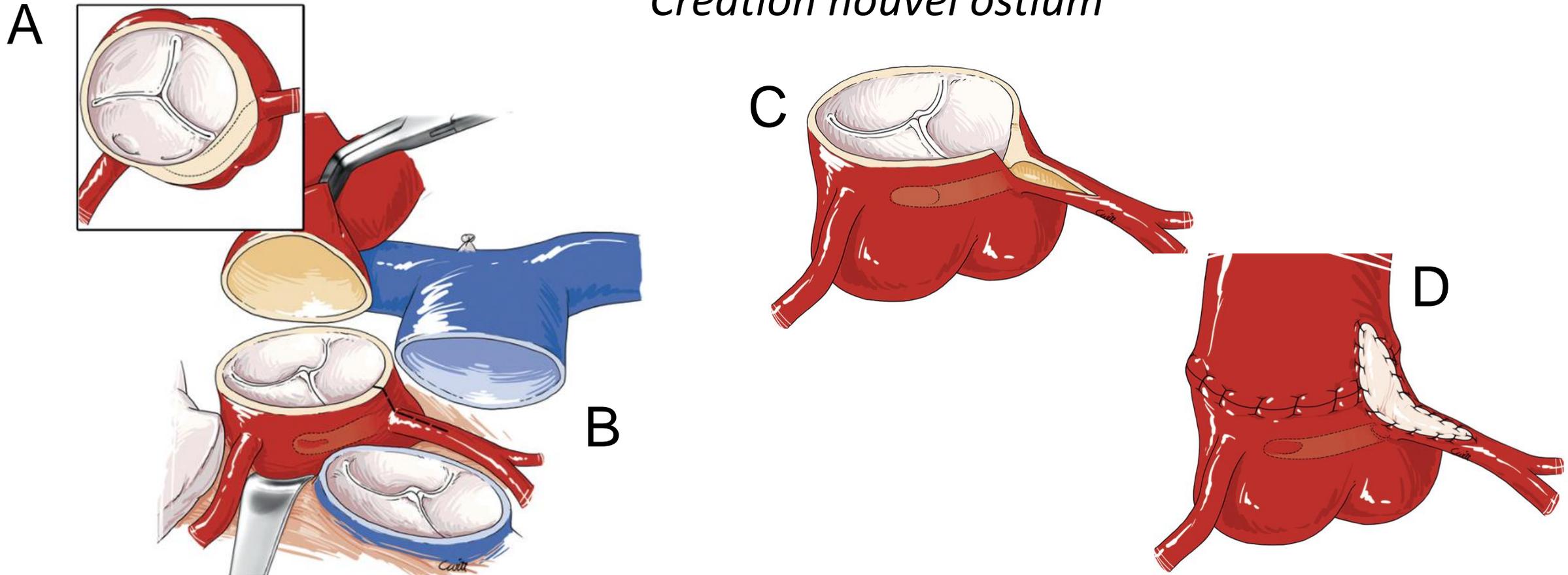
Techniques chirurgicales

Réimplantation coronaire

Pas adapté en général

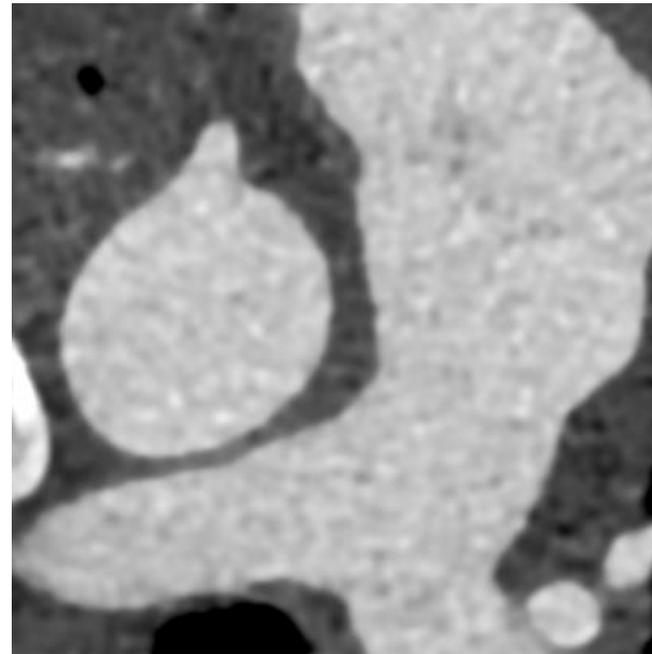
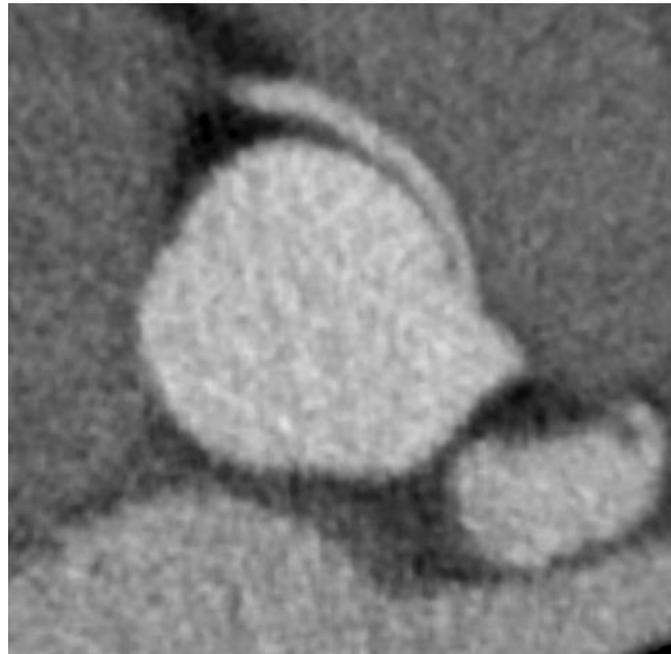


Techniques chirurgicales
Création nouvel ostium



Techniques chirurgicales

Création néo-ostium



Anomalies coronaires et chirurgie

- Recommandations : **souvent ciblées sur une population jeune**
- Décisions thérapeutiques : **indiquées sans tenir compte de l'âge**
- Etudes randomisées contrôlées : **aucune**
- Histoire naturelle et corrigée : **mal connue à long terme**
- Effet sur le risque de mort subite : **?**
- Correction chirurgicale : **techniques spécialisées**
- Echecs : **thrombose précoce, sténose cicatricielle, anévrisme, insuffisance aortique**

Anomalies coronaires et chirurgie

European Journal of Cardio-Thoracic Surgery 58 (2020) 975–982
doi:10.1093/ejcts/ezaa129 Advance Access publication 23 June 2020

ORIGINAL ARTICLE

Cite this article as: Gaillard M, Pontailier M, Danial P, Moreau de Bellaing A, Gaudin R, du Puy-Montbrun L et al. Anomalous aortic origin of coronary arteries: an alternative to the unroofing strategy. Eur J Cardiothorac Surg 2020;58:975–82.

Anomalous aortic origin of coronary arteries: an alternative to the unroofing strategy

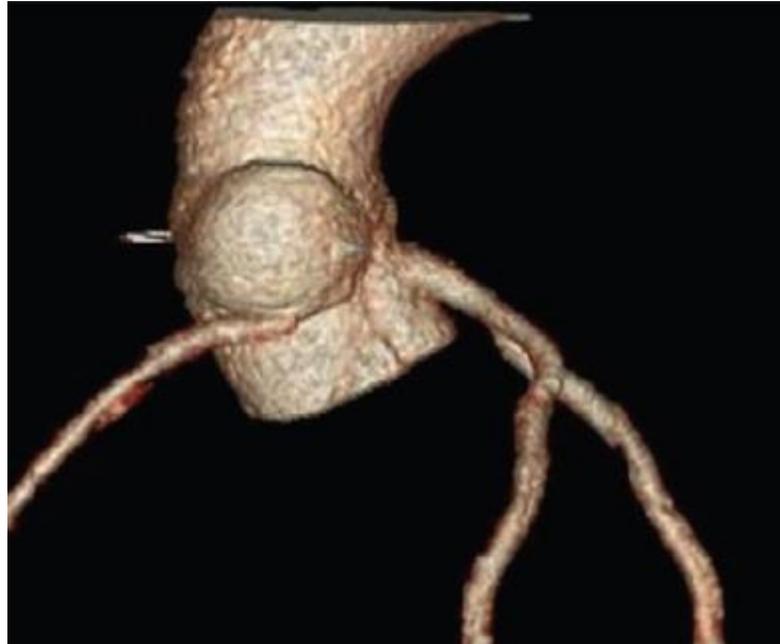
Maira Gaillard^a, Margaux Pontailier ^a, Pichoy Danial^a, Anne Moreau de Bellaing^a, Régis Gaudin ^a,
Leonora du Puy-Montbrun^a, Bari Murtuza^a, Ayman Haydar^a, Sophie Malekzadeh-Milani ^b,
Damien Bonnet ^b, Pascal Vouhé^a and Olivier Raisky^{a,*}

- N = 61
- 2005-2019
- Age (years - median): 14.7 (3.7 – 66.1)
- AAOCA right: 40 (66%) – AAOCA left : 21 (34%)
- Interarterial course: 56 (92%) – subpulmonic course : 5 (8%)
- Aborted sudden cardiac death: 5 (8%)

- PCI : 3 patients (5%)
- Redo surgery : 3 patients (5%)

Anomalies de connexion coronaire et chirurgie

1 year after surgery



- Embryologie et anatomie
- Classification
- Prévalence
- Imagerie
- Ischémie myocardique
- Mort subite
- Dépistage
- Prise en charge
- Chirurgie
- **Angioplastie**
- Activités sportives

Anomalous origins of coronary artery and PCI

Six-Month Success of Intracoronary Stenting for Anomalous Coronary Arteries Associated With Myocardial Ischemia

Doorey AJ Am J Cardiol 2000.

N=14

Origin of the Right Coronary Artery from the Opposite Sinus of Valsalva in Adults: Characterization by Intravascular Ultrasonography at Baseline and After Stent Angioplasty

Angelini P Cathet Cardio Interv 2015.

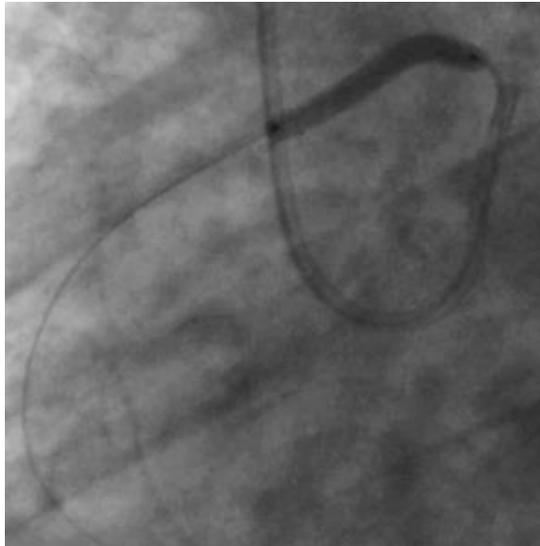
N=42

Place of Angioplasty for Coronary Artery Anomalies With Interarterial Course

Aubry P FCVM. 2021.

N=17

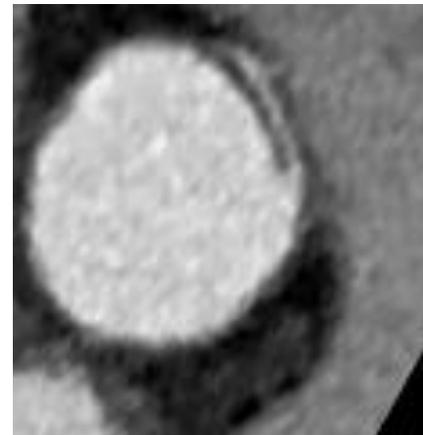
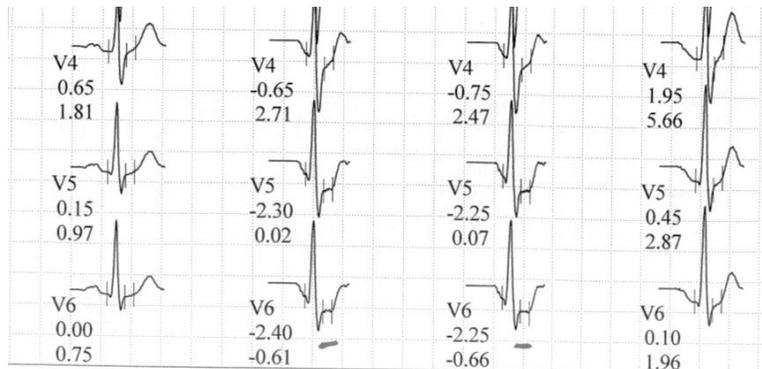
Angioplastie coronaire droite ectopique



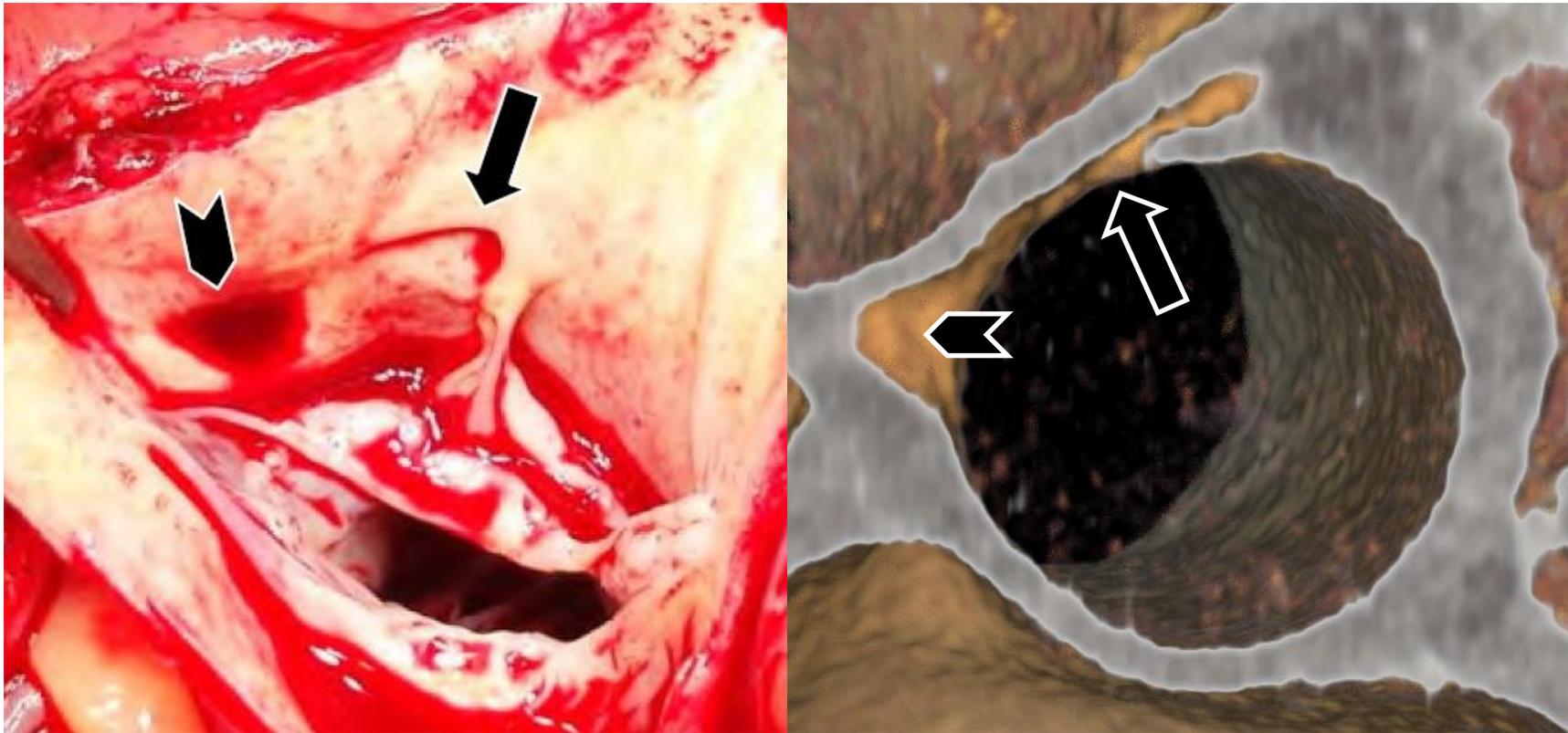
Est-ce possible ?

ANOCOR stenting registry n = 40 (2014-2024)

- 64-year old man
- Intensive sport activities (biking >100 km)
- Dizziness during exercise following by syncope
- Exercice stress test: asymptomatic (250 watts), positif ECG
- CTCA: right coronary AAOCA



Connexion coronaire droite dans sinus gauche

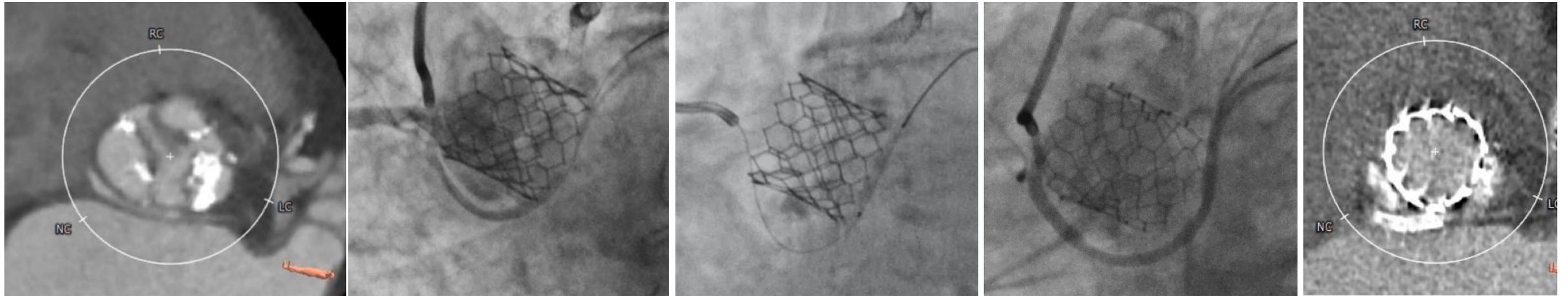




Angioplastie coronaire droite ectopique

Effets	Angioplastie
Symptômes d'allure ischémique	
Ischémie myocardique	
Réduction risque de mort subite	?
Resténose intrastent	
Dissection aortique	
Déformation structure du stent	

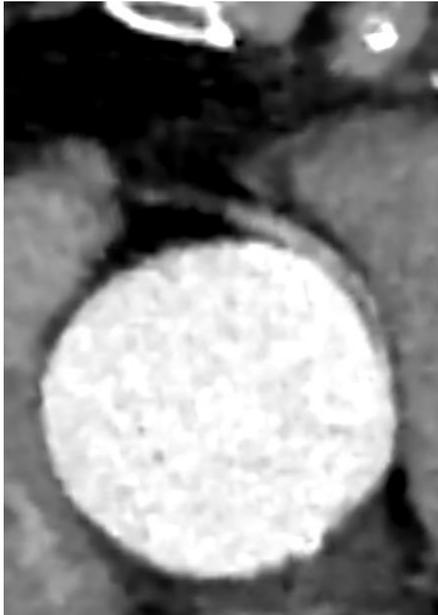
Cas rare mais... à connaître



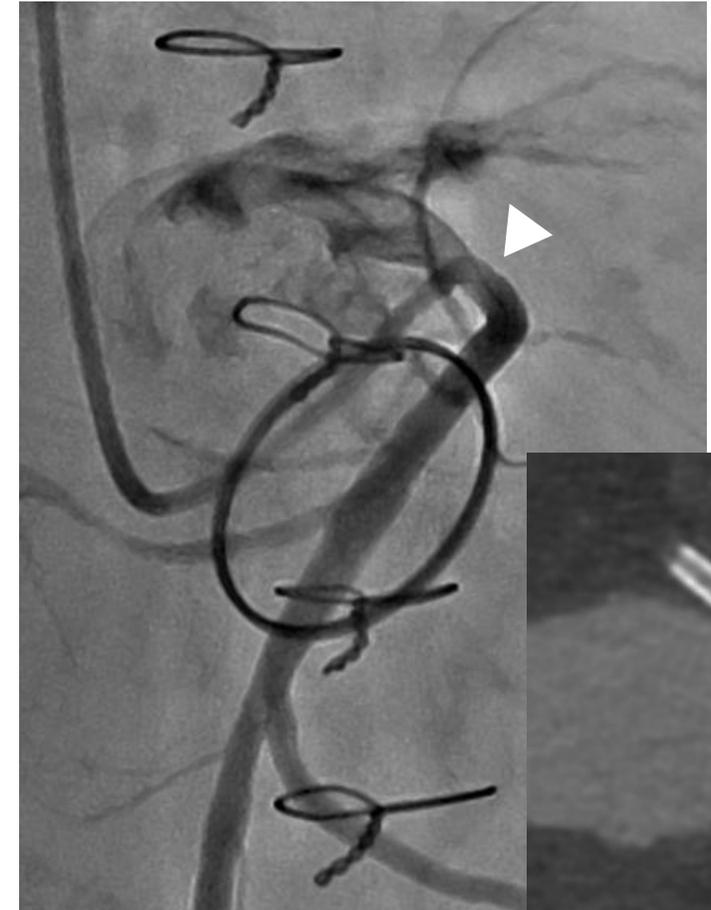
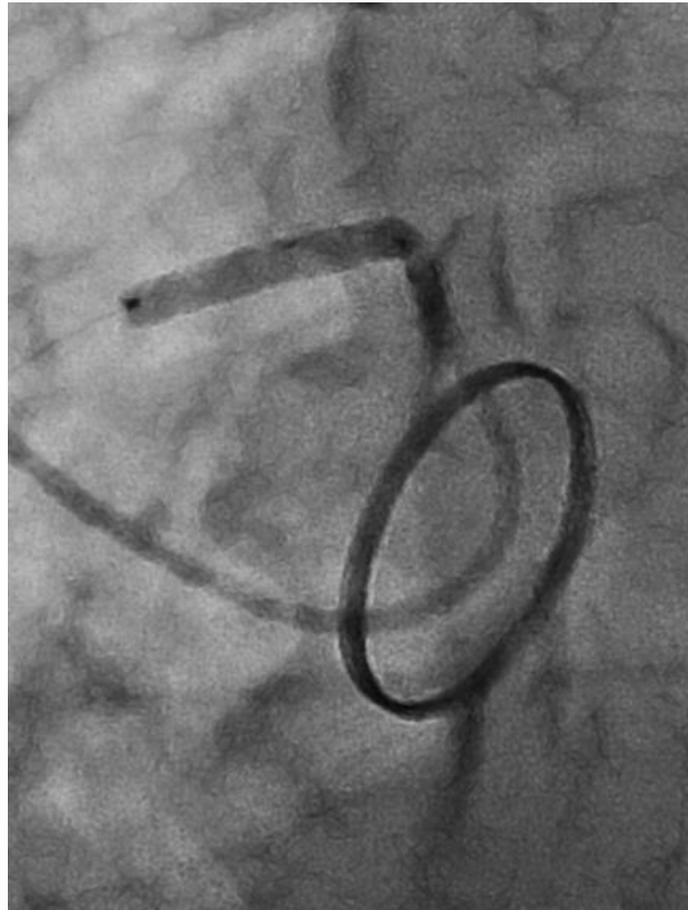
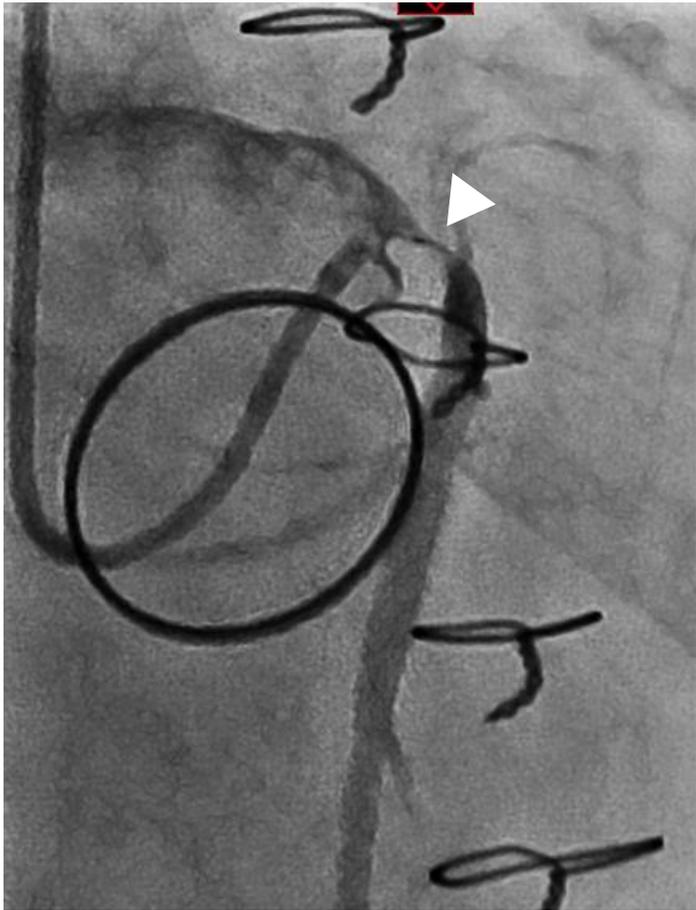
Etat de choc
Sous-décalage ST latéral

Courtesy of Dr Cavalerie (Toulouse)

Anomalie coronaire droite et chirurgie aortique



Anomalie coronaire droite et chirurgie aortique



- Embryologie et anatomie
- Classification
- Prévalence
- Imagerie
- Ischémie myocardique
- Mort subite
- Dépistage
- Prise en charge
- Chirurgie
- Angioplastie
- **Activités sportives**

2020 Guidelines on sports cardiology and exercise in patients with cardiovascular disease

Recommendations for exercise in young individuals/athletes with anomalous origins of coronary arteries

Recommendations	Class ^a	Level ^b
When considering sports activities, evaluation with imaging tests to identify high-risk patterns and an exercise stress test to check for ischaemia should be considered in individuals with AOCA.	IIa	C
In asymptomatic individuals with an anomalous coronary artery that does not course between the large vessels, does not have a slit-like orifice with reduced lumen and/or intramural course, competition may be considered, after adequate counselling on the risks, provided there is absence of inducible ischaemia.	IIb	C
After surgical repair of an AOCA, participation in all sports may be considered, at the earliest 3 months after surgery, if they are asymptomatic and there is no evidence of inducible myocardial ischaemia or complex cardiac arrhythmias during maximal exercise stress test.	IIb	C
Participation in most competitive sports with a moderate and high cardiovascular demand among individuals with AOCA with an acutely angled take-off or an anomalous course between the large vessels is not recommended. ^c	III	C

© ESC 2020

AOCA = anomalous origin of coronary arteries.

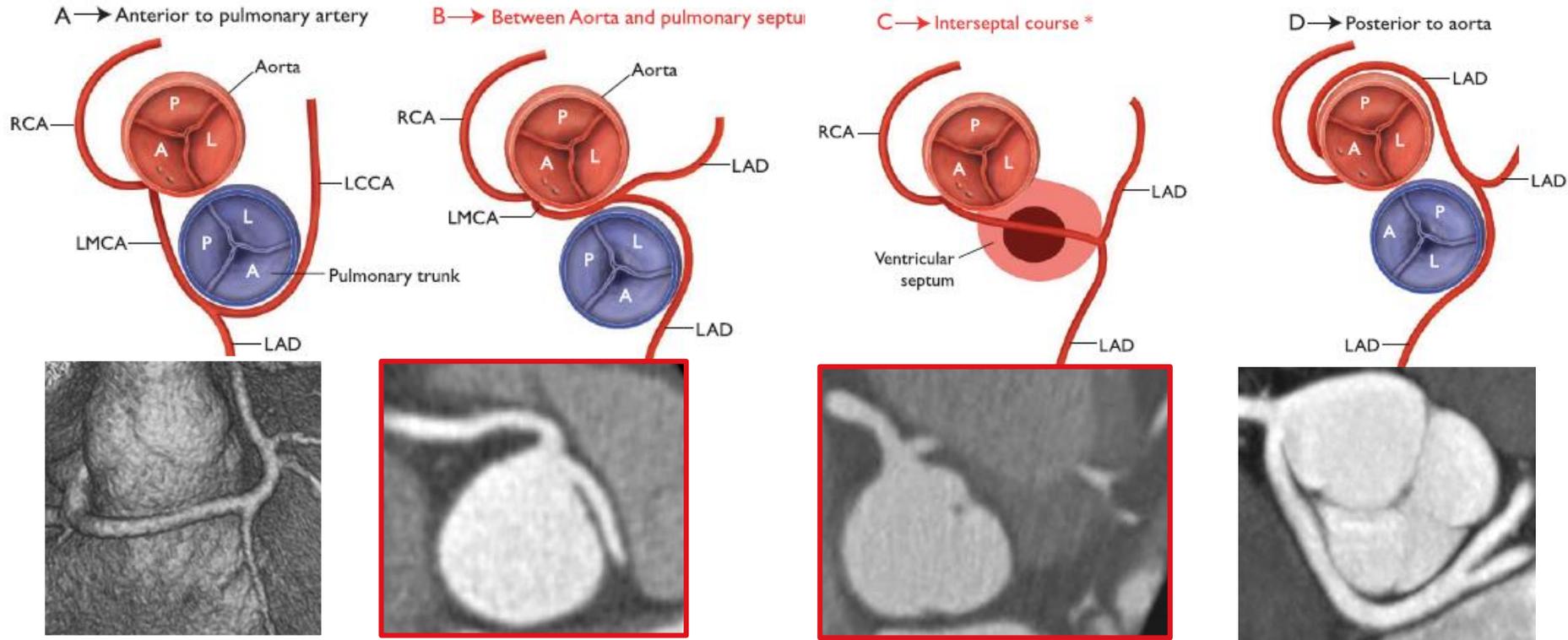
^aClass of recommendation.

^bLevel of evidence.

^cThis recommendation applies whether the anomaly is identified as a consequence of symptoms or discovered incidentally, and in individuals <40 years of age.

2020 Guidelines on sports cardiology and exercise in patients with cardiovascular disease

* Associated with sudden cardiac death



2. Athletes with an anomalous origin of a right coronary artery from the left sinus of Valsalva should be evaluated by an exercise stress test. For those without either symptoms or a positive exercise stress test, permission to compete can be considered after adequate counseling of the athlete and/or the athlete's parents (in the case of a minor) as to risk and benefit, taking into consideration the uncertainty of accuracy of a negative stress test (*Class IIa; Level of Evidence C*).

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<http://dx.doi.org/10.1016/j.jacc.2015.09.036>

AHA/ACC SCIENTIFIC STATEMENT

**Eligibility and Disqualification
 Recommendations for Competitive Athletes
 With Cardiovascular Abnormalities:
 Task Force 4: Congenital Heart Disease**



A Scientific Statement From the American Heart Association and American College of Cardiology

George F. Van Hare, MD, FACC,
*Chair**
 Michael J. Ackerman, MD, PhD,
 FACC*

Juli-anne K. Evangelista, DNP,
 APRN, CPNP-AC, FACC*
 Richard J. Kovacs, MD, FAHA, FACC*
 Robert J. Myerburg, MD, FACC*

Keri M. Shafer, MD*
 Carole A. Warnes, MD, FACC*
 Reginald L. Washington, MD, FAHA*

Restriction sportive

Éléments décisionnels

- ANOCOR gauche / ANOCOR droite
- Forme anatomique interartérielle
- Age
- Symptomatologie d'allure ischémique
- Ischémie myocardique documentée
- Type de sport
- Niveau de sport
- Pratique en compétition
- Projet sportif
- Avis patient
- Avis entourage familial si besoin
- Avis cardiologue du sport

	Skill 	Power 	Mixed 	Endurance 
LOW	Golf (buggy)	Shot putting (recreational)	Soccer (adapted)	Jogging
	Golf (18 holes walking)	Discus (recreational)	Basketball (adapted)	Long distance walking
	Table tennis (double)	Alpine skiing (recreational)	Handball (adapted)	Swimming (recreational)
	Table tennis (single)	Alpine skiing (recreational)	Volleyball	Speed walking
MEDIUM	Shooting	Short distance running	Tennis (double)	Mid/long distance running
	Curling	Shot putting	Ice-Hockey	Style dancing
	Bowling	Discus	Hockey	Cycling (road)
	Sailing	Alpine skiing	Rugby	Mid/long distance swimming
HIGH	Yachting	Judo/karate	Fencing	Long distance skating
	Equestrian	Weight lifting	Tennis (single)	Pentathlon
		Wrestling	Waterpolo	Rowing
		Boxing	Soccer (competitive)	Canoeing
		Basketball (competitive)	X-country skiing	
		Handball (competitive)	Biathlon	
			Triathlon	

■ Low intensity
 ■ Medium intensity
 ■ High intensity

Restriction sportive

- Aucune
- Sur activité sportive d'intensité basse
- Sur activité sportive d'intensité moyenne
- Sur activité sportive d'intensité élevée
- Sur la pratique en compétition
- En attendant une correction de l'anomalie coronaire
- Après la correction de l'anomalie coronaire

Information éclairée du patient

Pratique sportive dans un environnement adapté



Cas complexe à gérer

- Jeune sportif (football) de 15 ans
- Centre de formation (pôle espoirs)
- Echocardiogramme systématique
- Découverte ANOCOR droite avec trajet interartériel
- Pas de symptomatologie d'allure ischémique
- Pas d'ischémie myocardique documentée

Que proposer ?

Merci



The screenshot shows the ANOCOR website header and navigation menu. The header includes the ANOCOR logo, the text "Anomalies Coronaires Congénitales", a search bar, and a "Contactez-nous" button with an envelope icon. The navigation menu lists various topics: Accueil, Qu'est-ce qu'une ANOCOR?, Qui sommes-nous?, Notre staff, Prise en charge, Recommandations, Arrêt cardiaque, Ischémie myocardique, Imagerie, Chirurgie, Angioplastie, Sport, Nos publications, Bibliothèque, Enseignement et formation, Fistules coronaires, and Infos patients. A "Contact" link is also visible below the menu.

Groupe multidisciplinaire ANOCOR **Anomalies Coronaires Congénitales**

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.

