

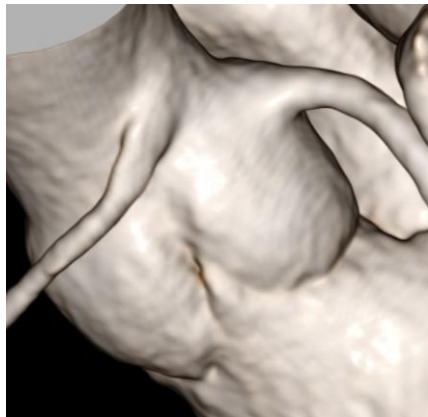
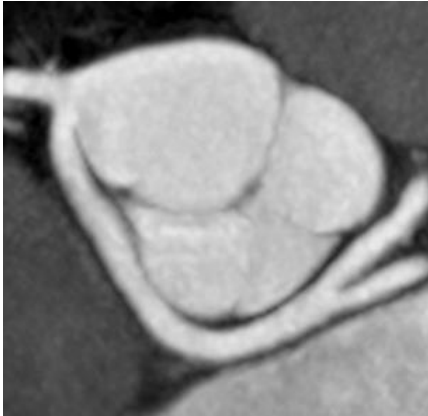
ANOmalties CORonaires congénitales (ANOCOR) et chirurgie

Pierre Aubry

Hôpital Bichat, Paris 75018

Centre Hospitalier, Gonesse 95500





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

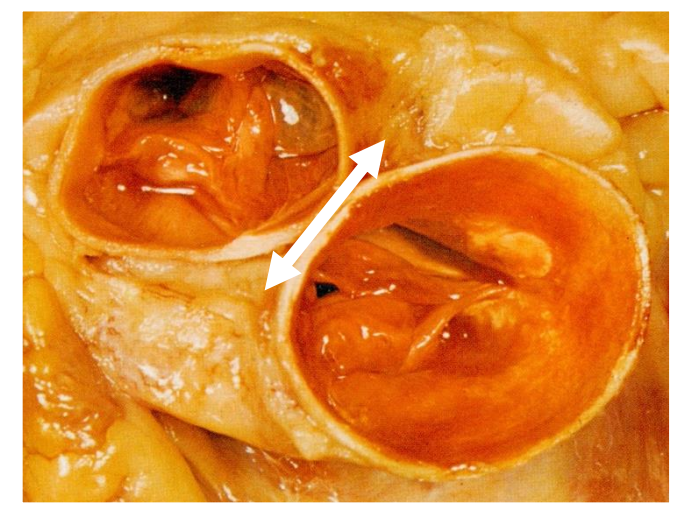
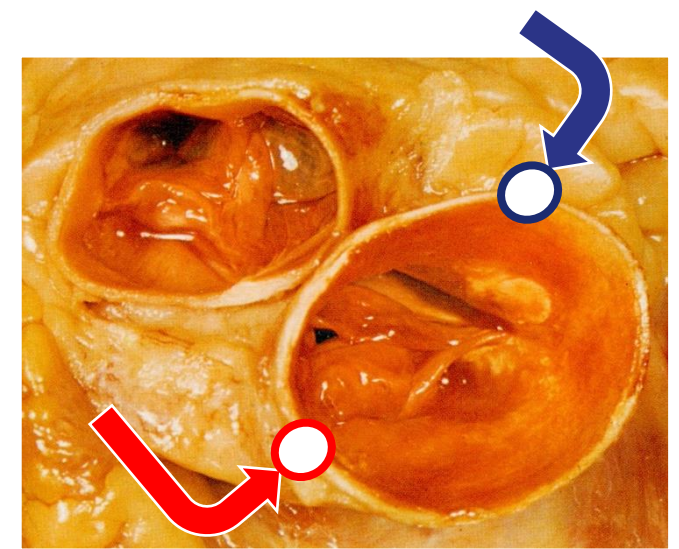
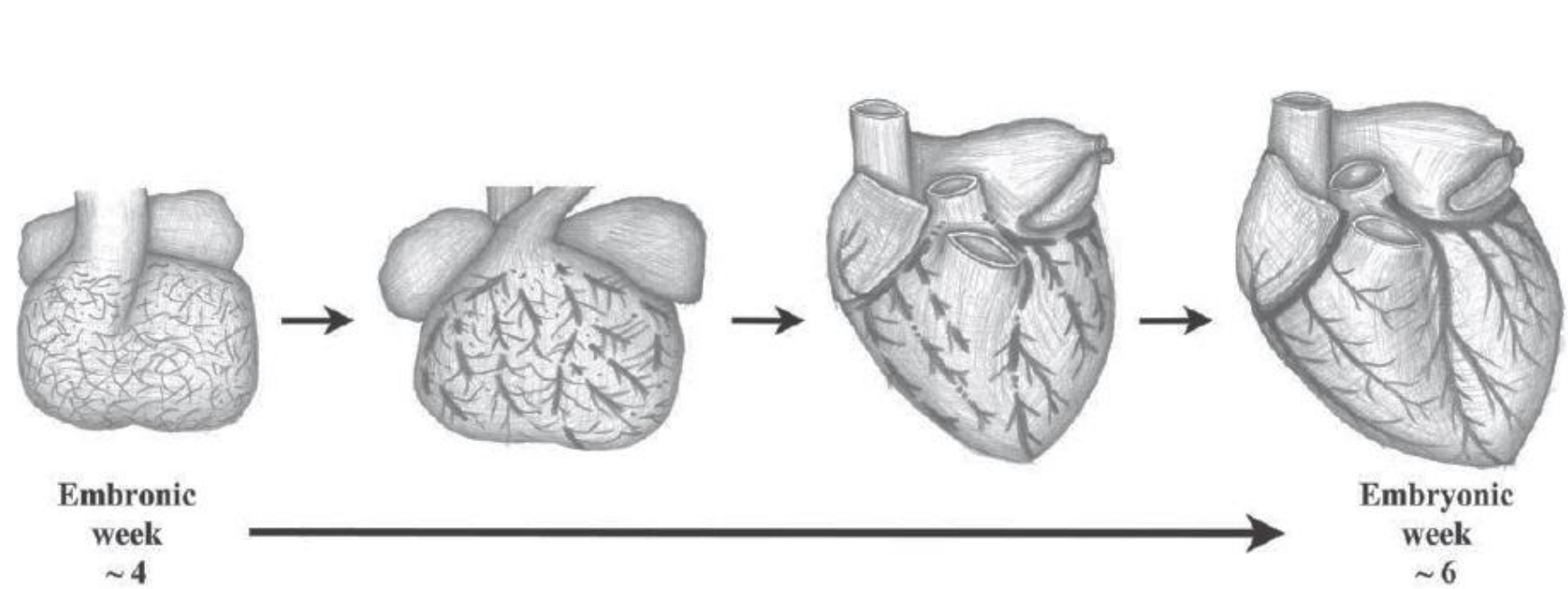
Origine anormale

Développement à partir de l'aorte

Connexion anormale

Développement vers l'aorte

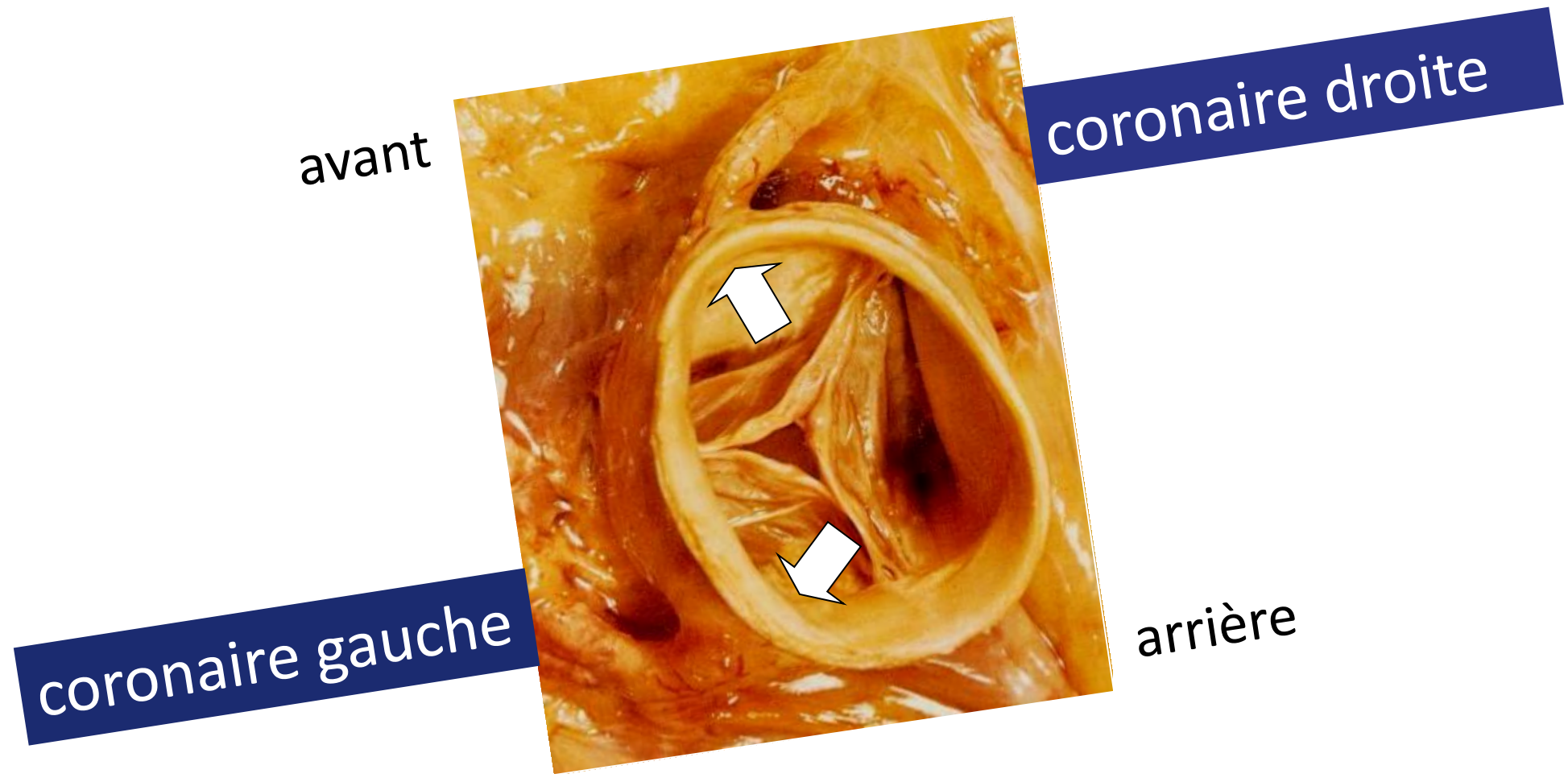
Embryologie et anatomie



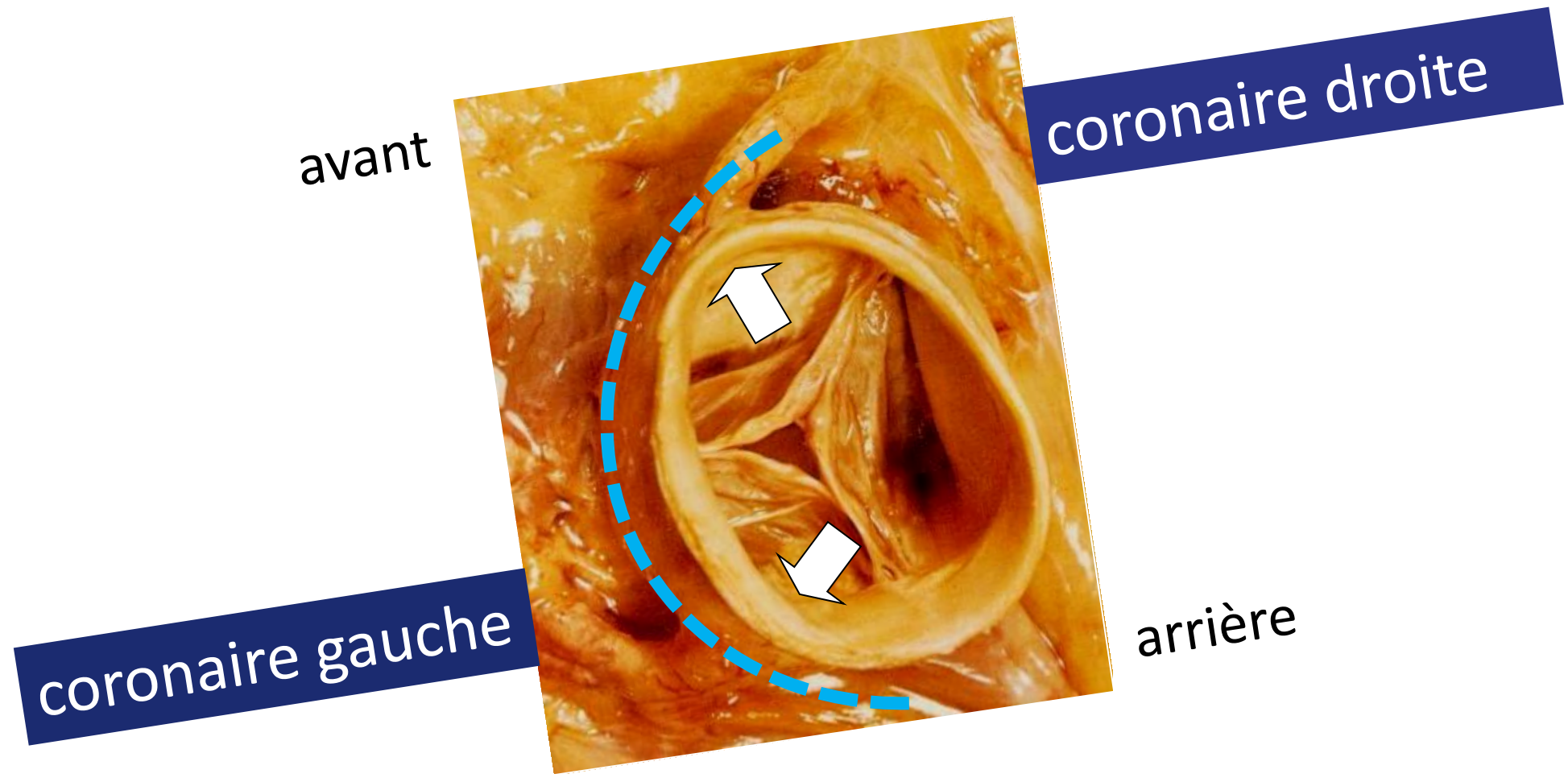
Development of coronary vessels during embryogenesis.

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

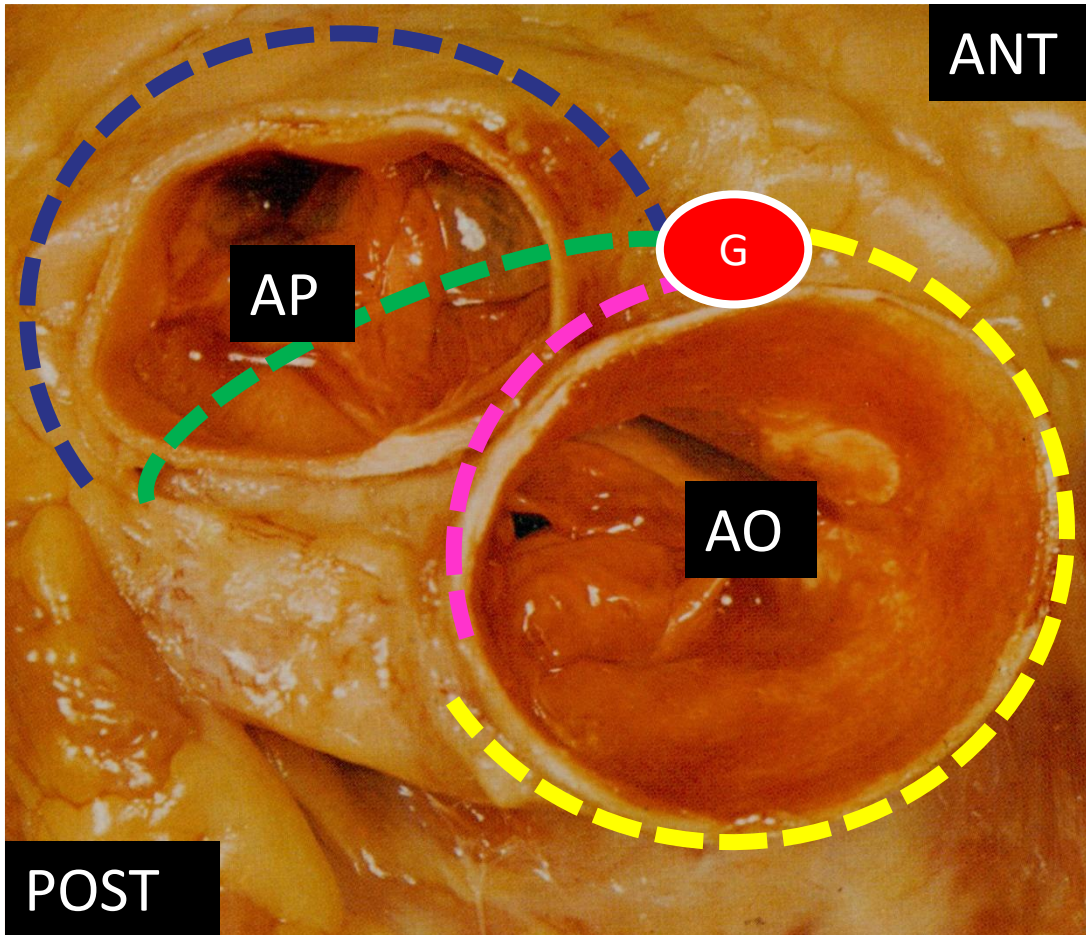
Connexions coronaires normales



Connexions coronaires anormales (---)

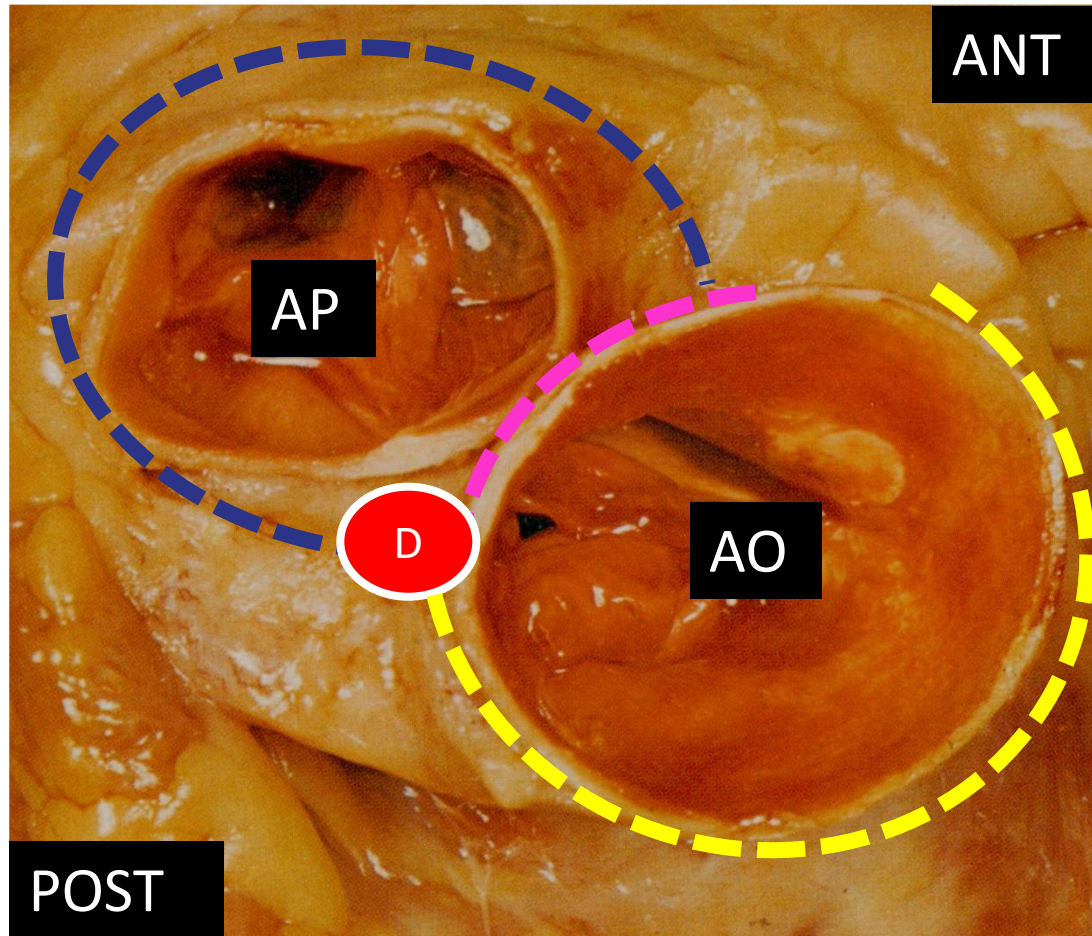


Trajets ectopiques pour la coronaire gauche



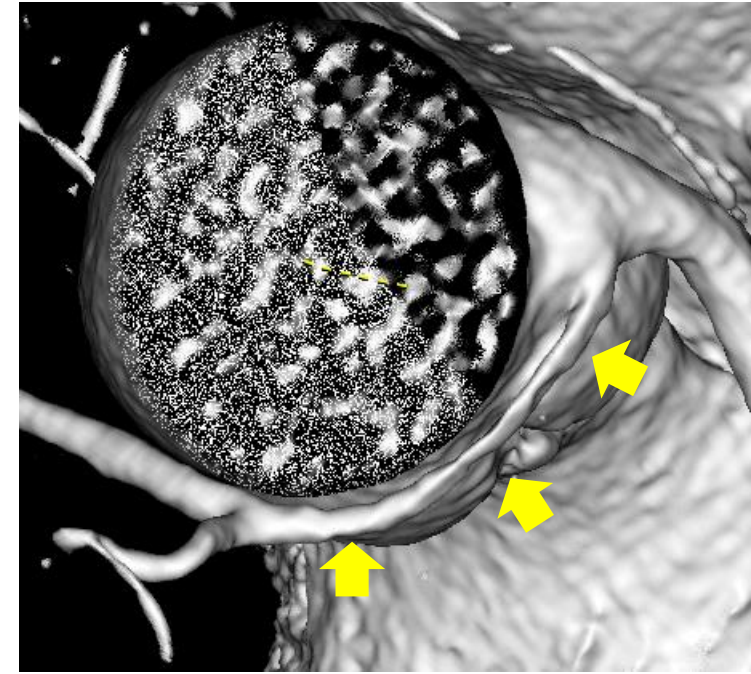
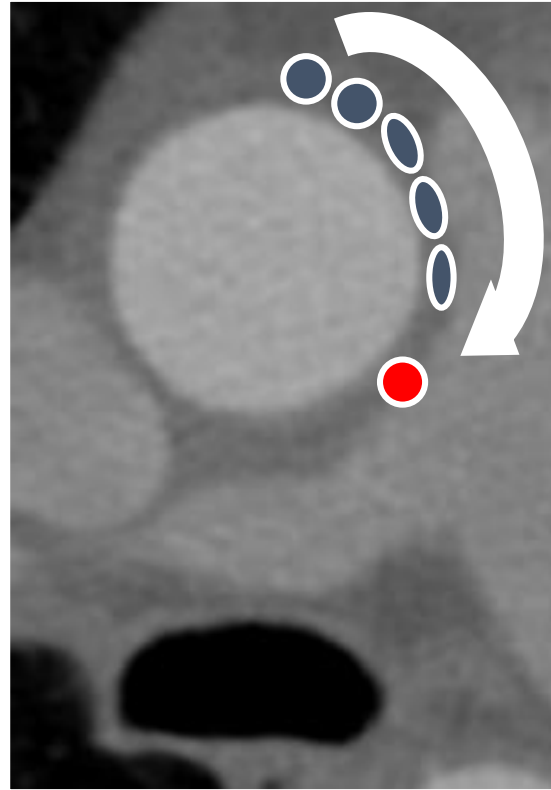
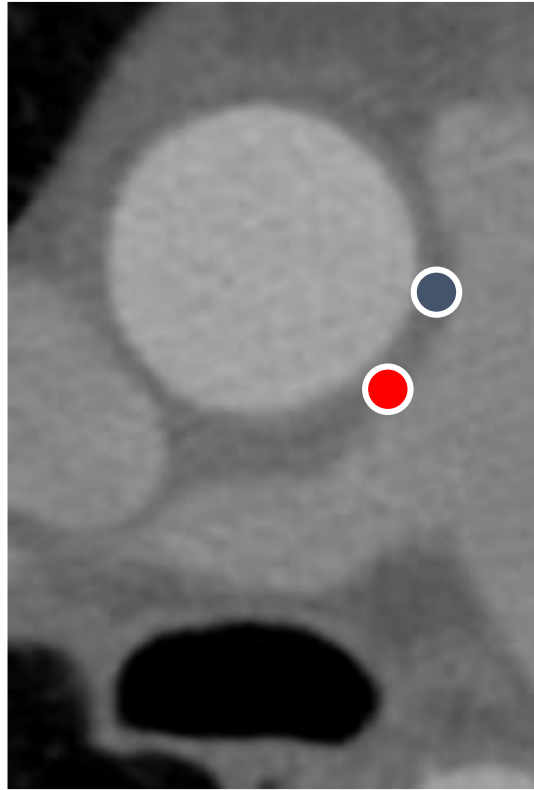
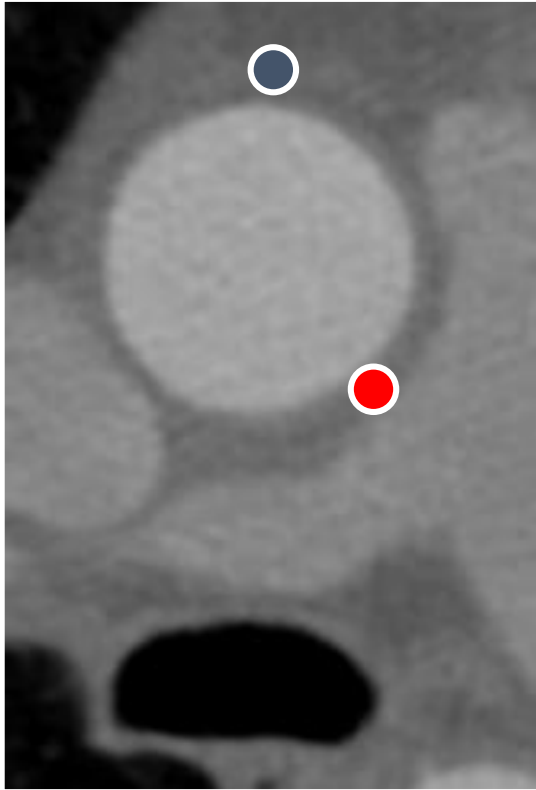
- prépulmonaire
- rétropulmonaire (intraseptal)
- interartériel (préaortique)
- rétroaortique

Trajets ectopiques pour la coronaire droite

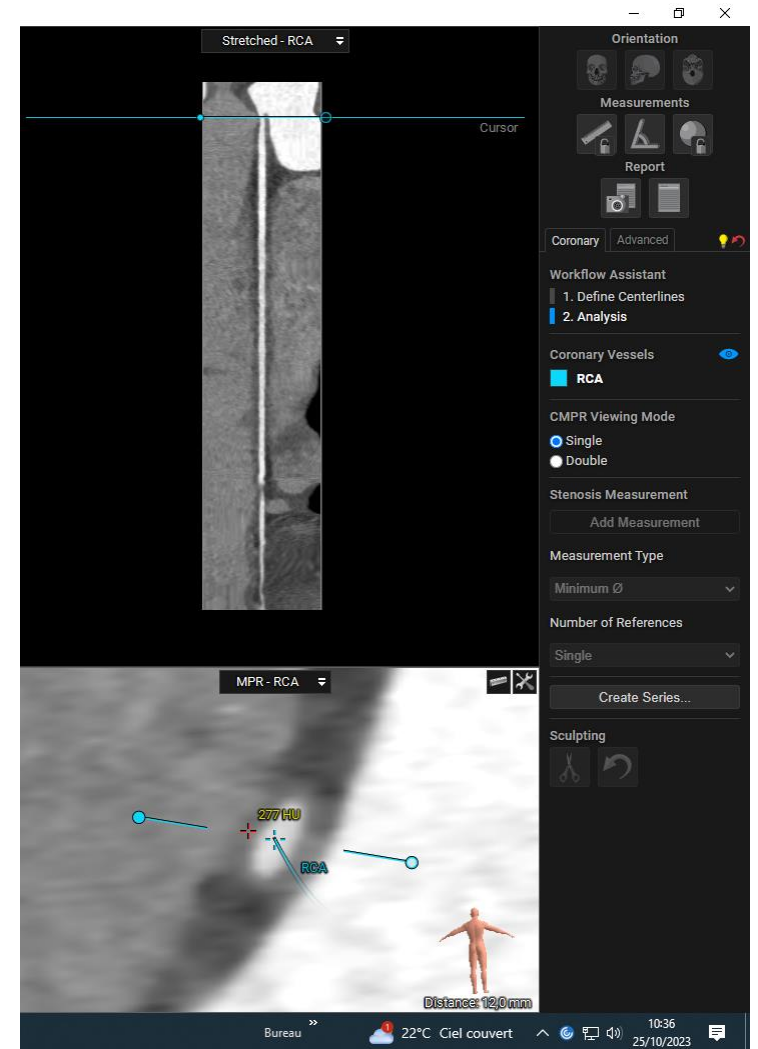
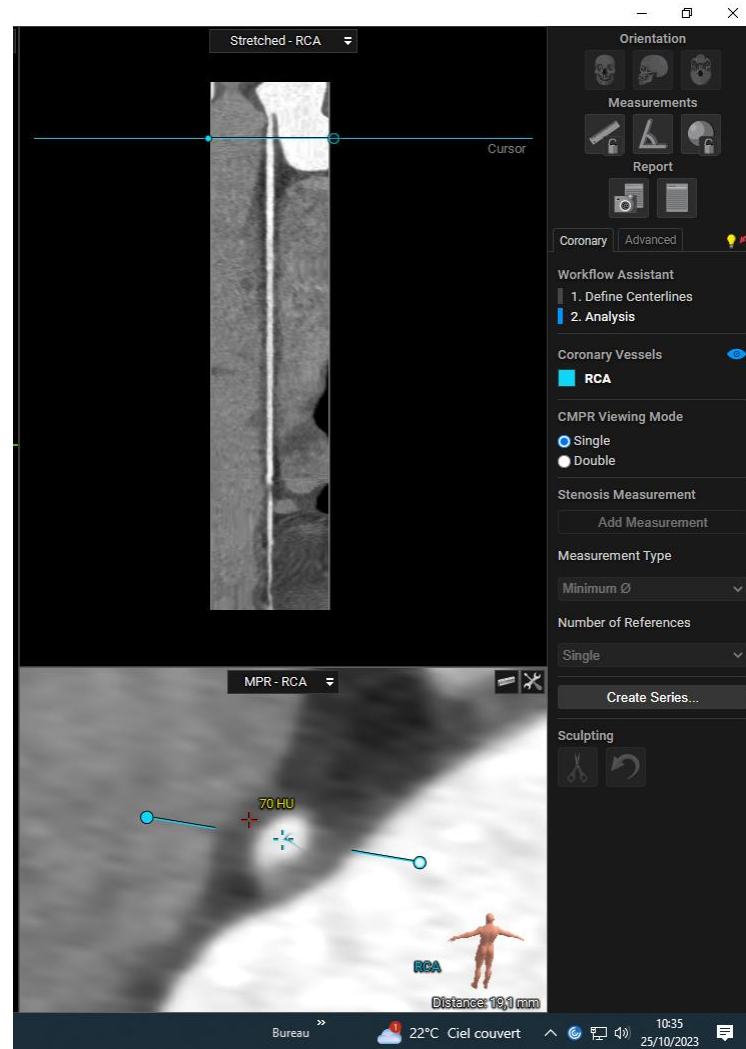
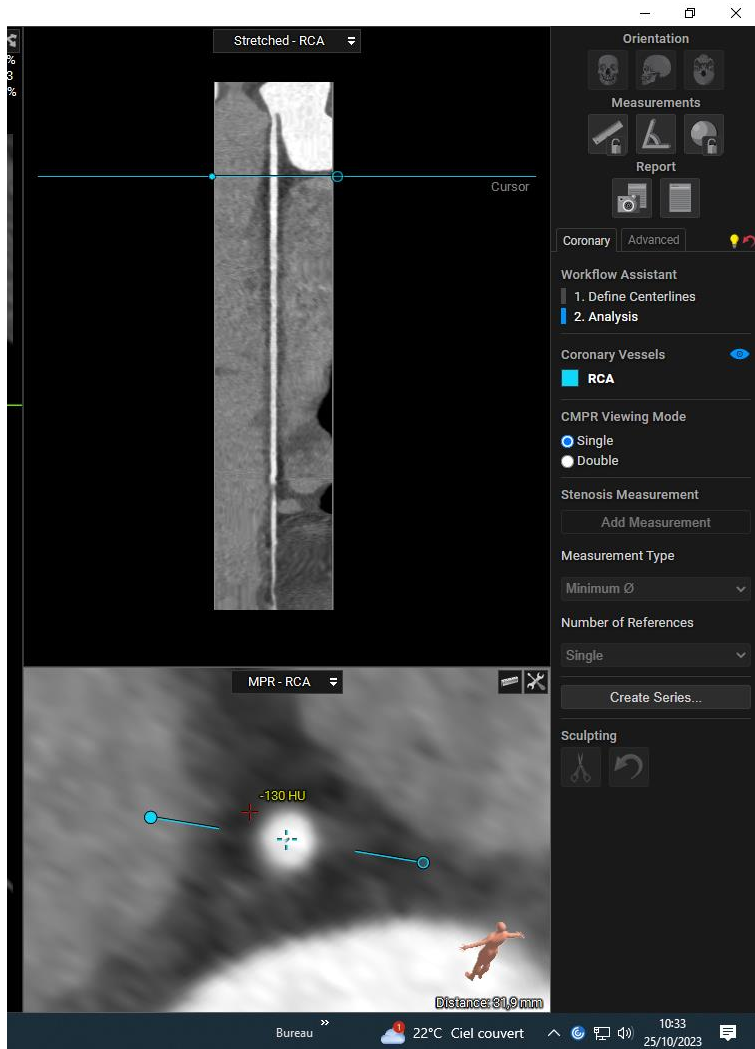


- prépulmonaire
- interartériel (préaortique)
- rétroaortique

Adaptation vasculaire



Adaptation vasculaire



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

Classification

- **Type d'artère**

Tronc
IVA
Circonflexe
Droite
Septale

- **Trajet**

Prépulmonaire
Rétropulmonaire
Interartériel
Rétroaortique
Normal

- **Site de connexion**

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

- **Risques**

Mort subite
Arrêt cardiaque
Arythmies ventriculaires
Ischémie myocardique
Absents

Classification

Liens de causalité :

- entre anomalie coronaire et symptomatologie
- entre anomalie coronaire et ischémie myocardique
- entre anomalie coronaire et arythmie ventriculaire
- entre anomalie coronaire et arrêt cardiaque

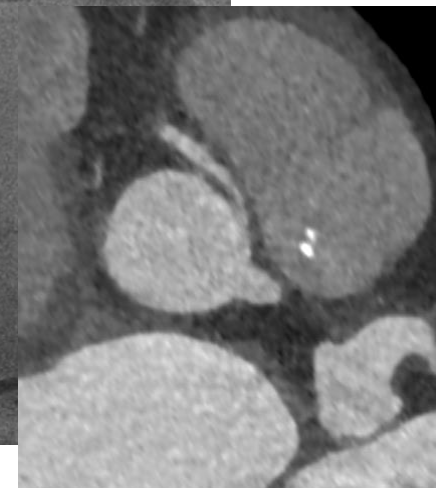
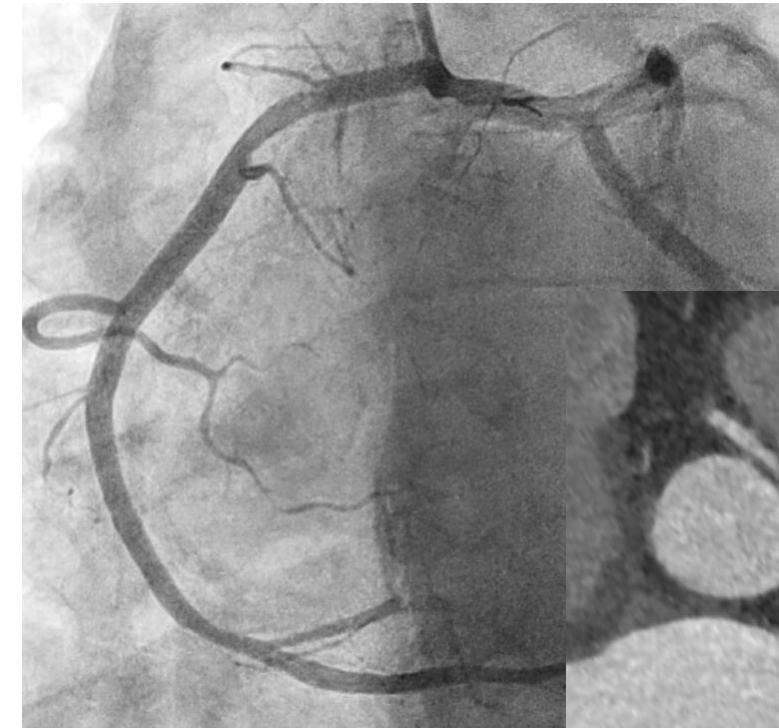
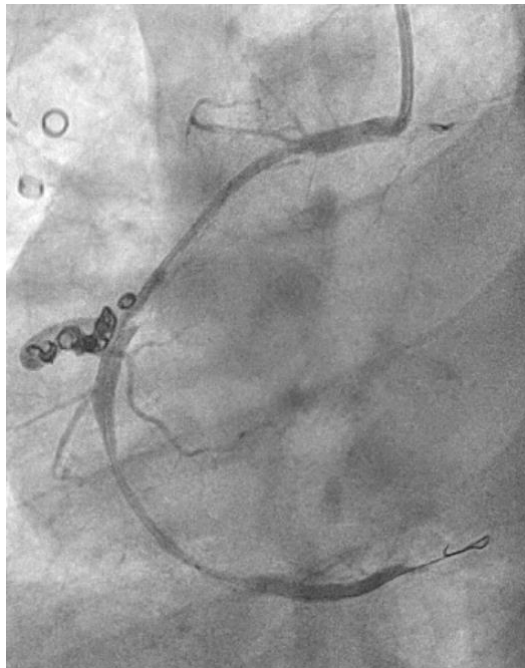
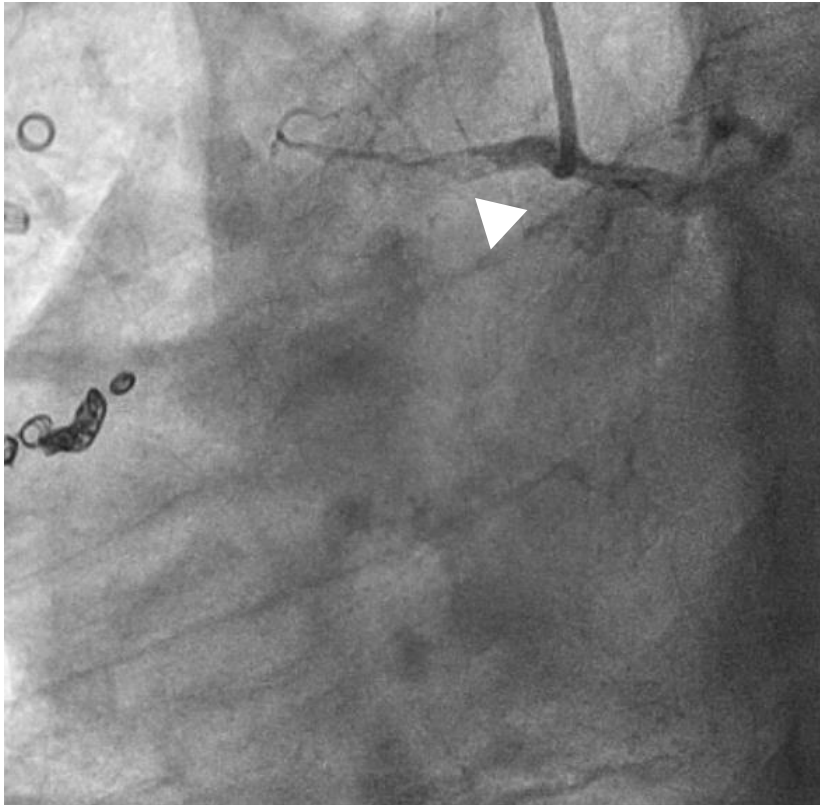
Absent

Possible/Probable

Certain

Classification

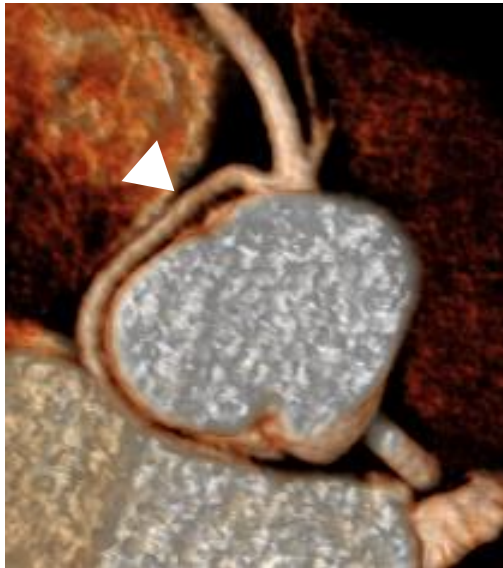
Homme de 35 ans – Maladie Rendu-Osler – MAV cérébrales/pulmonaires
SCA ST+ inférieur lors activité sportive



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

Prévalence selon l'artère coronaire **50%**

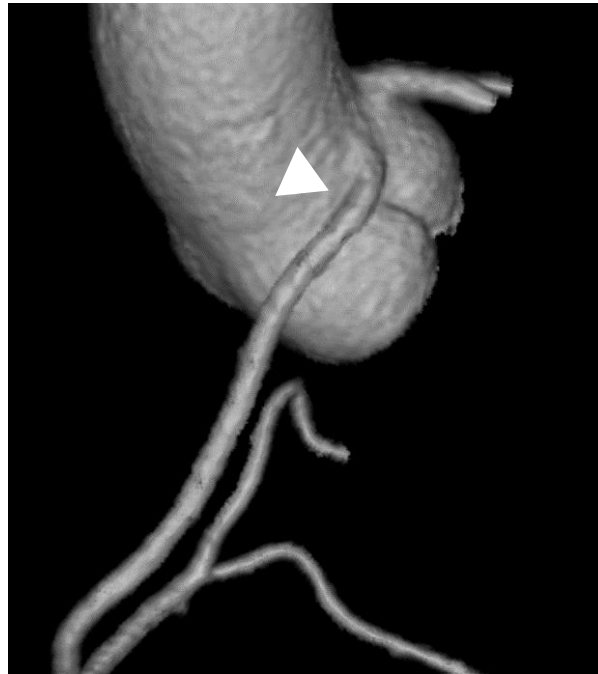
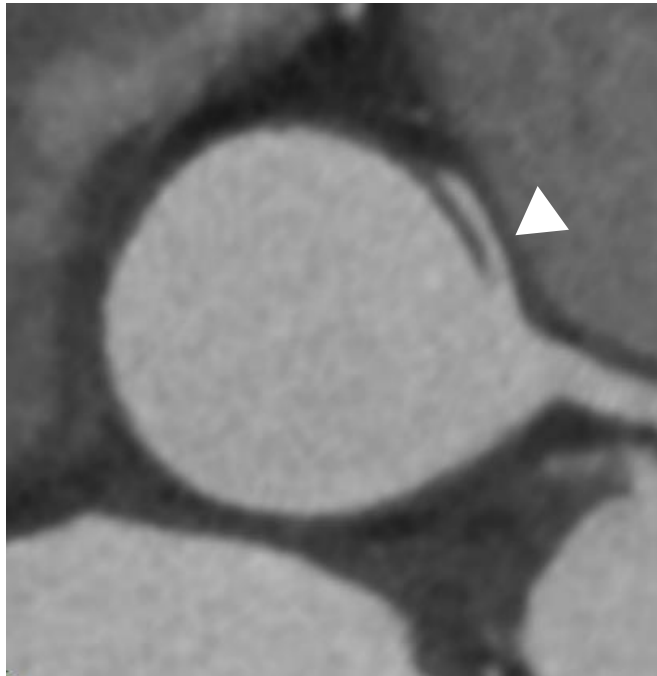
Circonflexe



Prévalence selon le trajet ectopique : rétroaortique **99%**

Prévalence selon l'artère coronaire **30%**

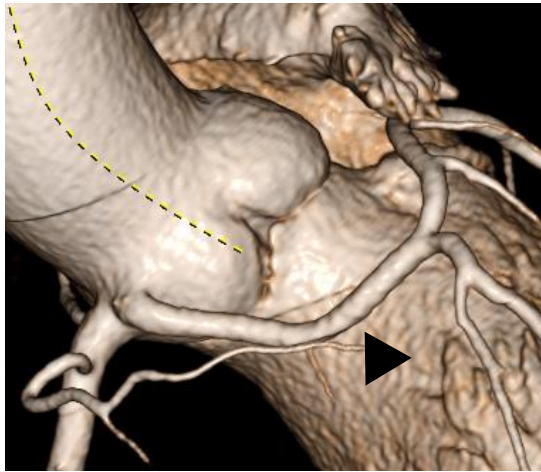
Droite



Prévalence selon le trajet ectopique : interartérielle **95%**

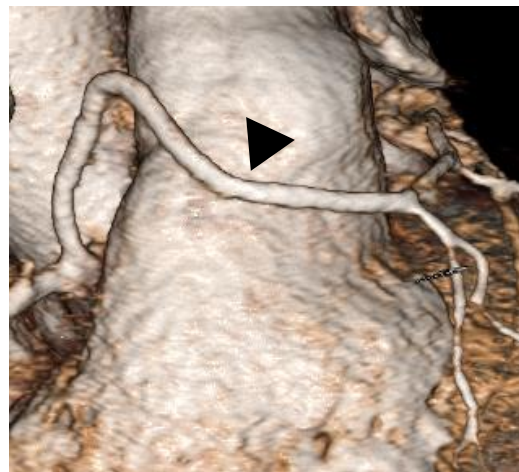
Prévalence selon l'artère coronaire **20%**

Tronc
IVA



Rétropulmonaire

45%



Prépulmonaire

30%



Rétroaortique

15%



Interartériel

10%

Prévalence selon le trajet ectopique : variable



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

Piège à éviter

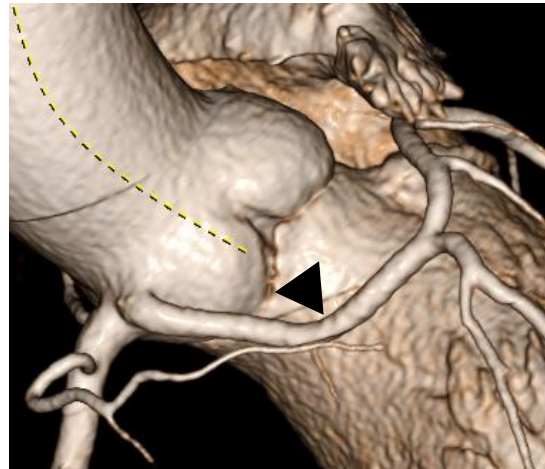


Trajet prépulmonaire

Trajet rétroaortique

Trajet interartériel

Trajet rétropulmonaire



Received: 11 October 2016 | Revised: 21 February 2017 | Accepted: 28 May 2017

DOI: 10.1111/chd.12504

ORIGINAL ARTICLE

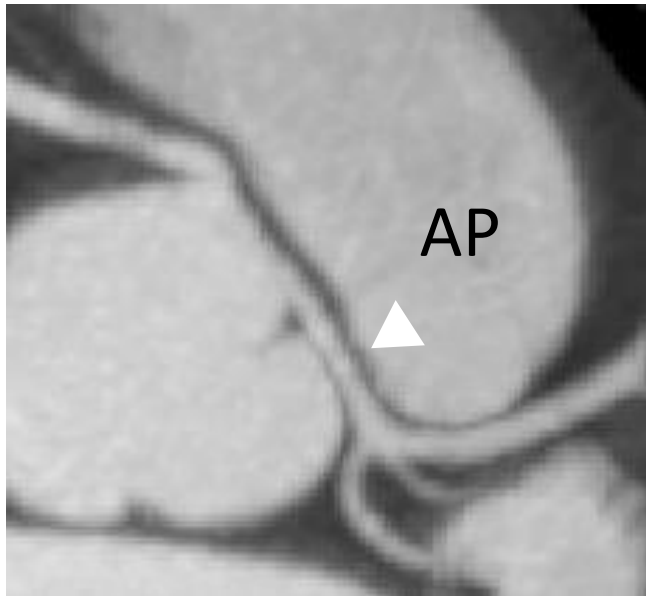
WILEY Congenital Heart Disease

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

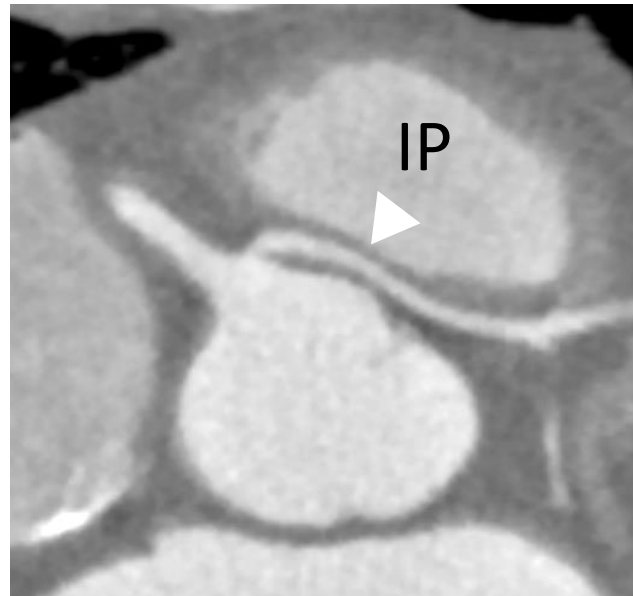
Athanasios Koutsoukis, MD¹ | Xavier Halna du Fretay, MD² | Patrick Dupouy, MD³ | Phalla Ou, MD, PhD⁴ | Jean-Pierre Laissy, MD, PhD⁴ | Jean-Michel Juliard, MD⁵ | Fabien Hyafil, MD⁶ | Pierre Aubry, MD⁵ | on behalf of the ANOCOR Investigators*

Congenital Heart Disease 2017

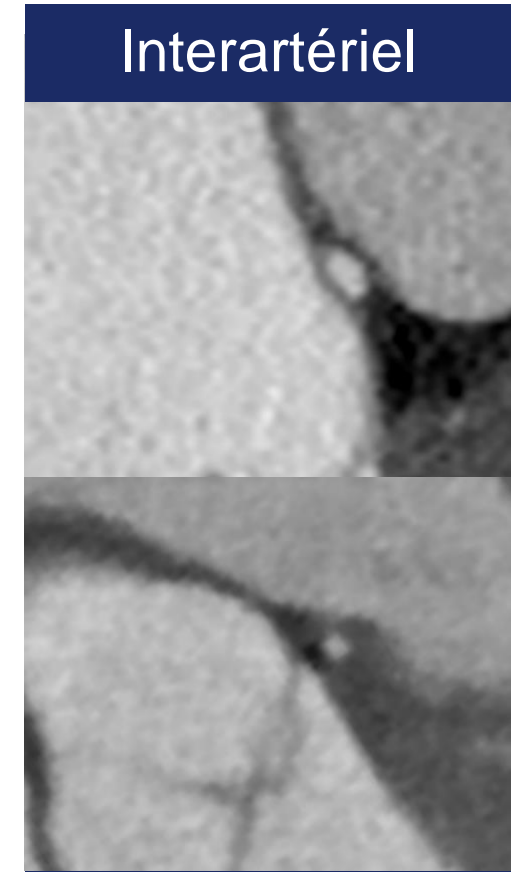
Formes anatomiques gauches à ne pas confondre



Trajet interartériel



Trajet rétropulmonaire

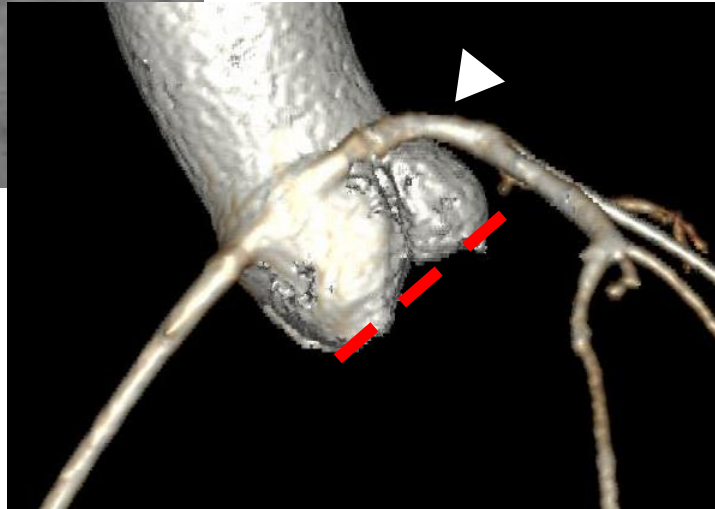
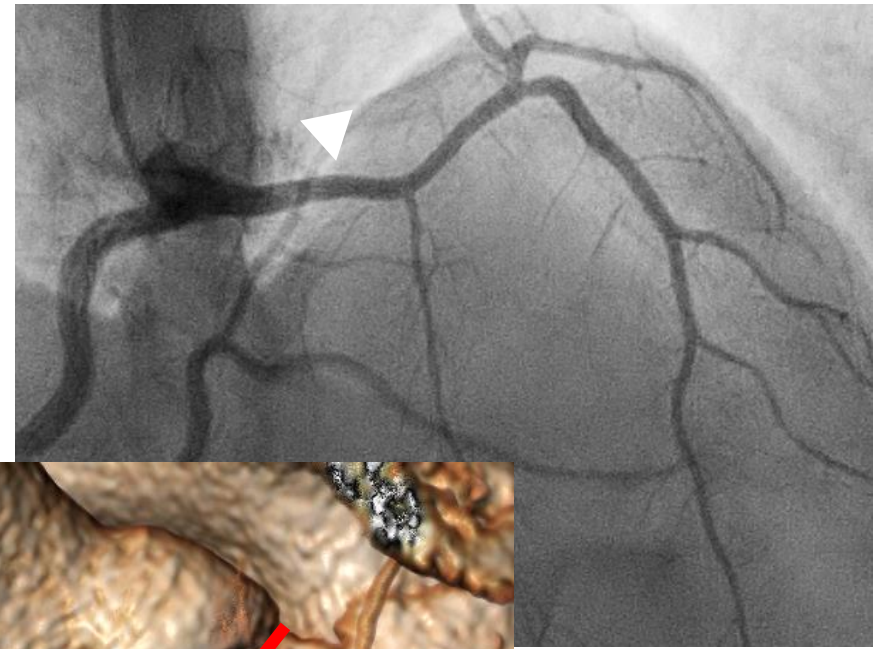


Interartériel

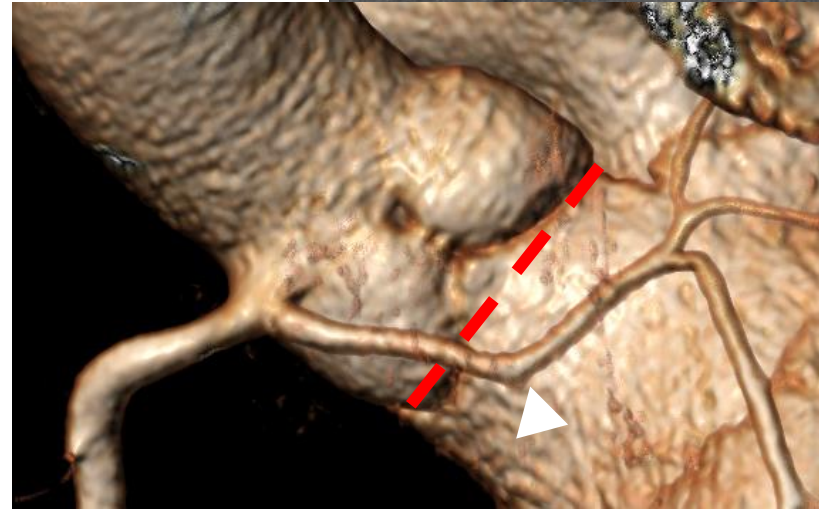
Rétropulmonaire



ANOCOR gauches

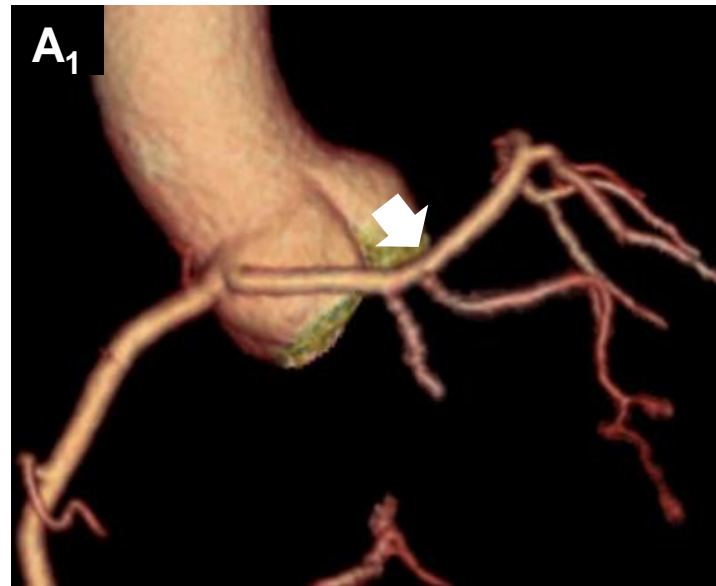


Trajet interartériel

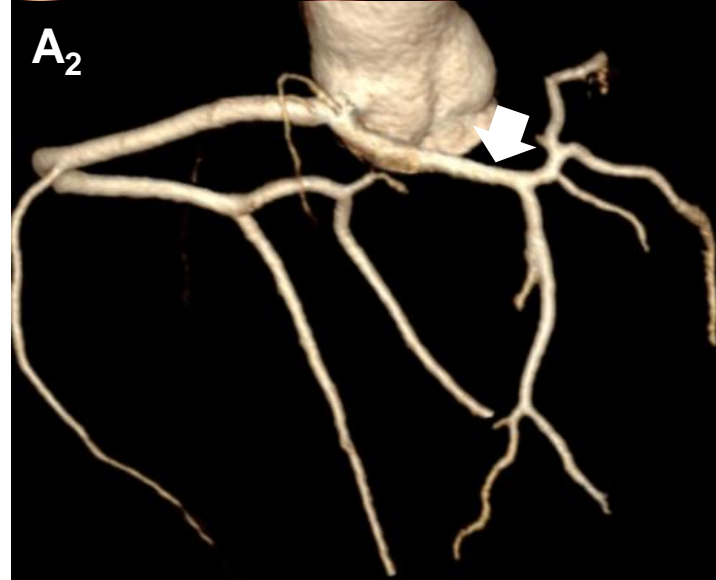


Trajet rétropulmonaire

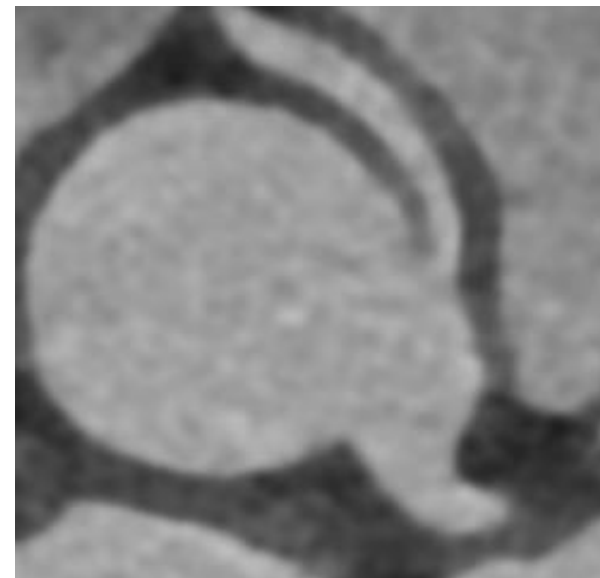
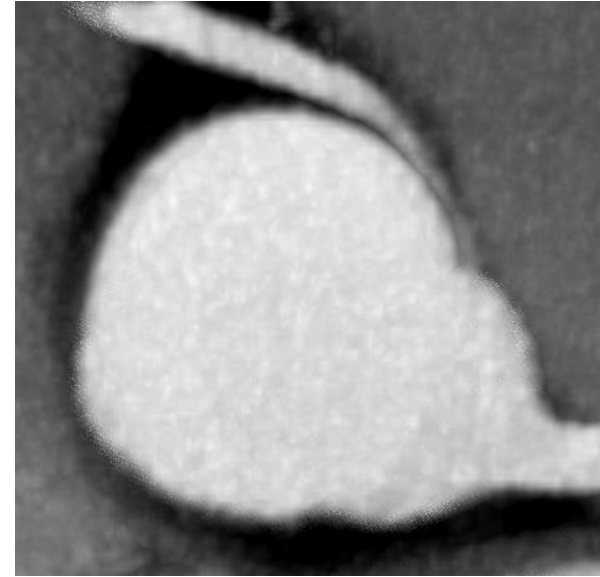
ANOCOR gauches

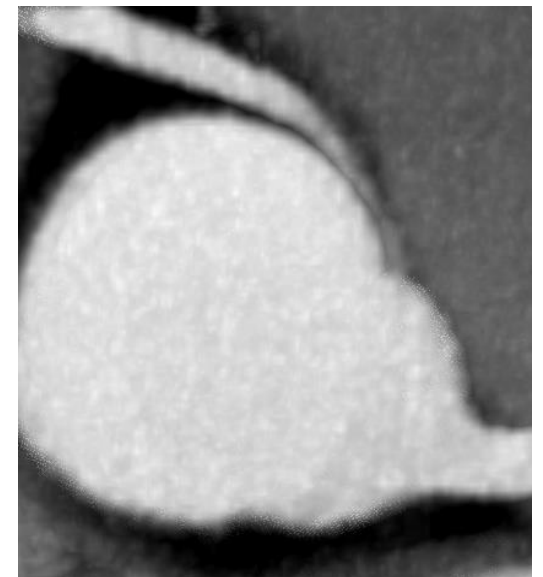
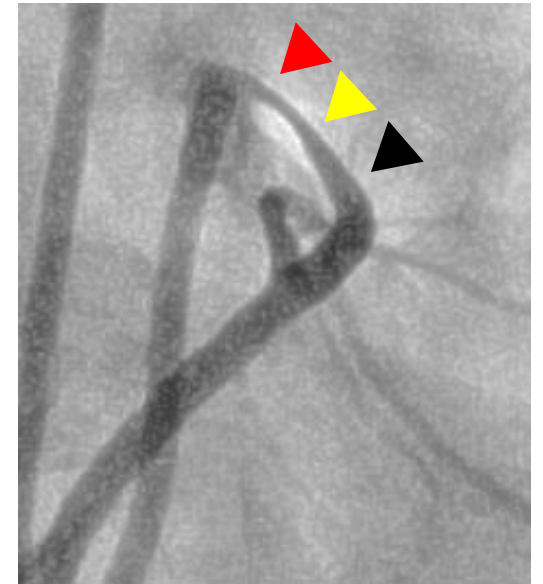
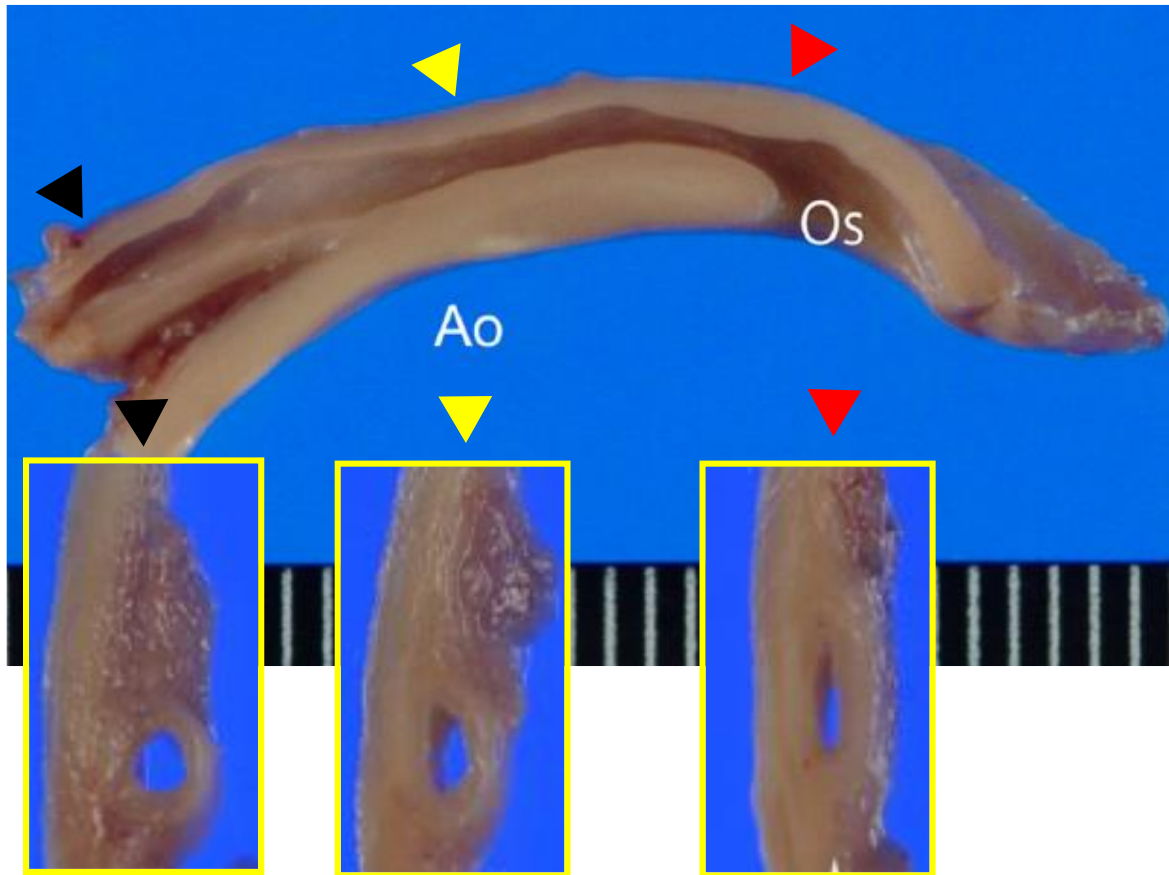


Trajet rétropulmonaire



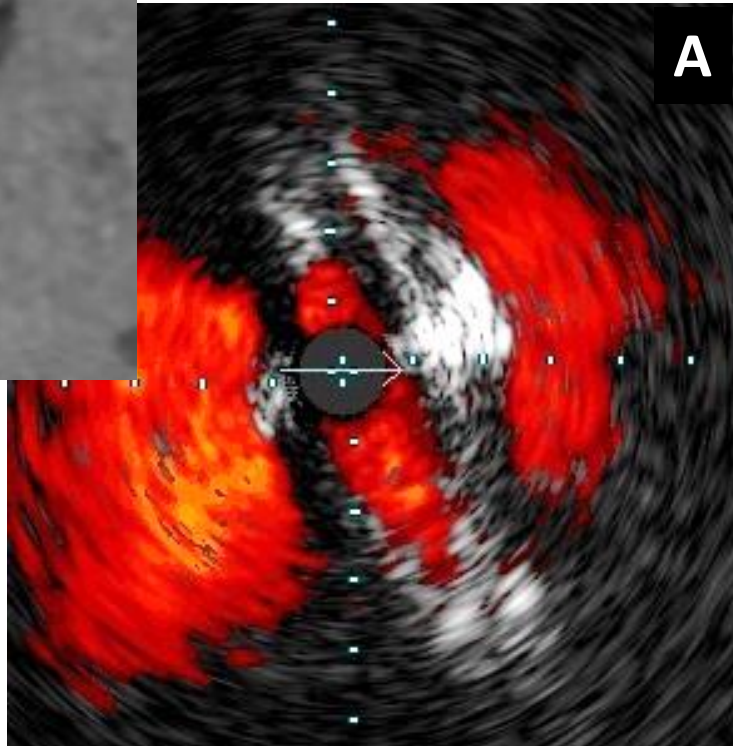
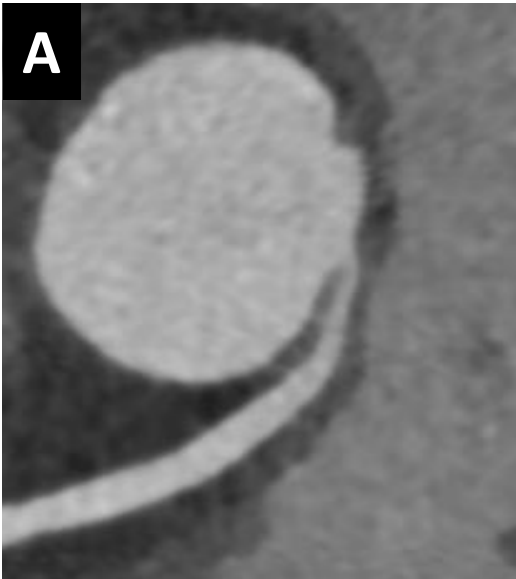
Passage aortique intramural



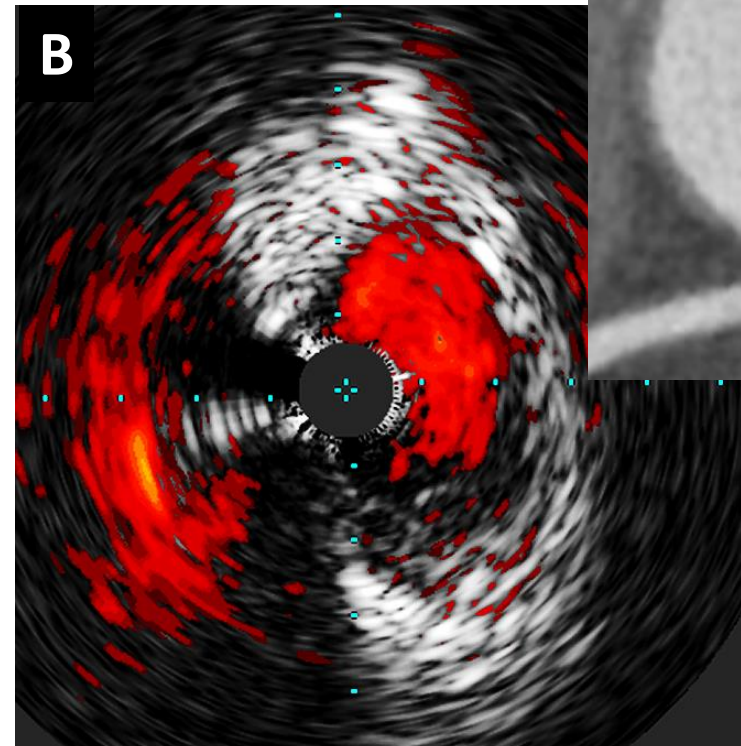


Right ANOCOR with an intramural course
Hata Y et al. Cardiovasc Pathol. 2014.

Echographie endocoronaire

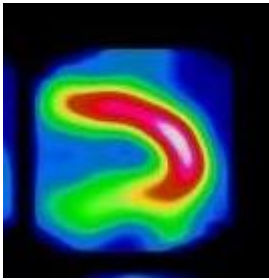


Passage intramural



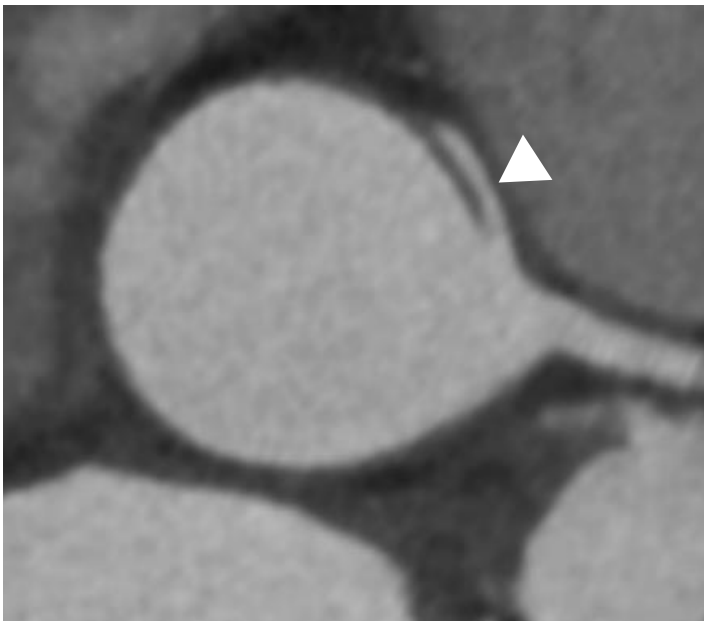
Passage juxtamural

- Mort subite/arrêt cardiaque récupéré
 - Symptômes d'allure ischémique (angor/syncope/dyspnée)
 - Ischémie myocardique (test avec imagerie)
- Embryologie et anatomie
 - Classification
 - Prévalence
 - Imagerie
 - Ischémie myocardique
 - Mort subite
 - Prise en charge
 - Chirurgie
 - Angioplastie

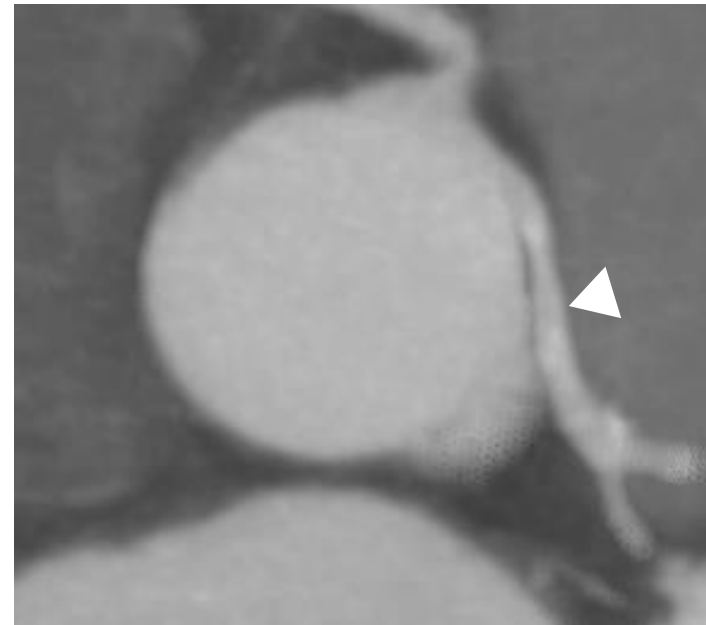


ANOCOR à risques

Trajet interartériel

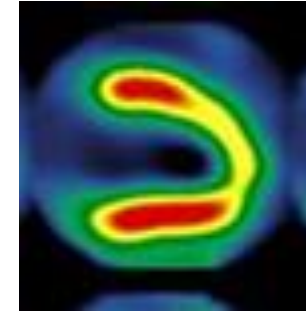


ANOCOR droite

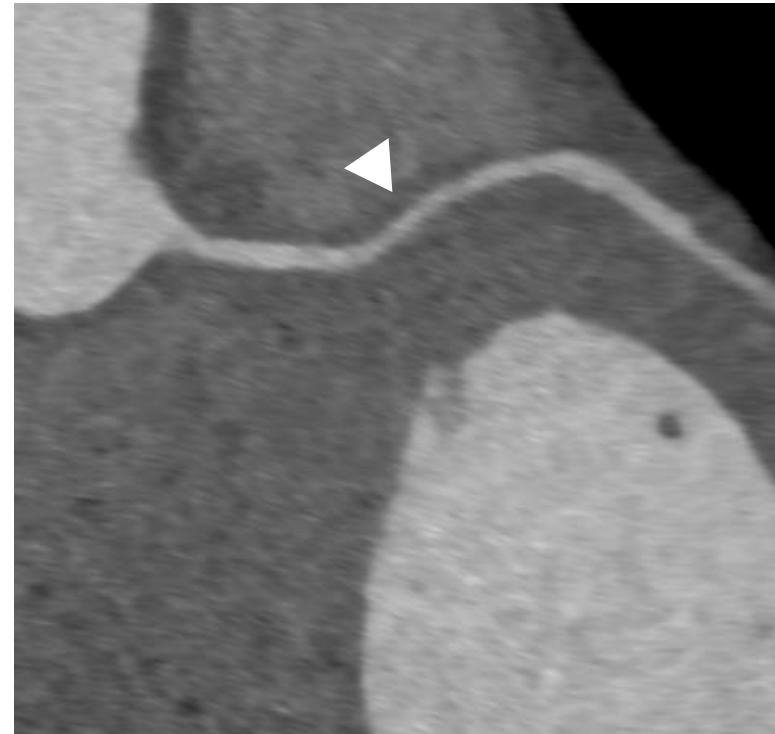


ANOCOR gauche

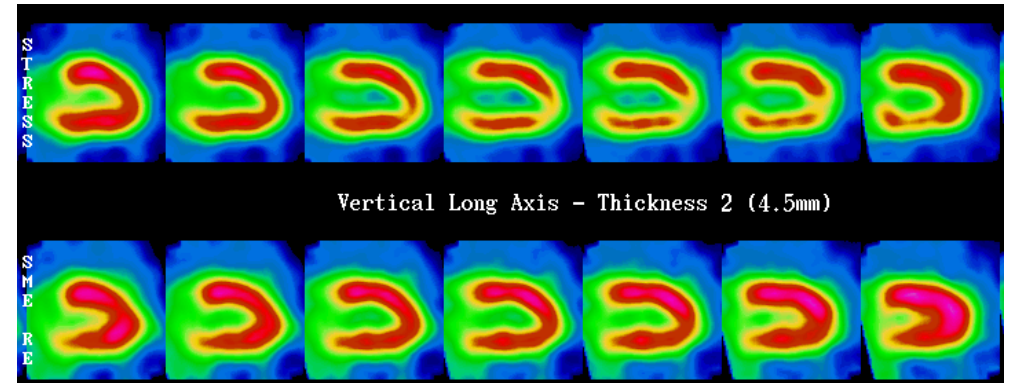
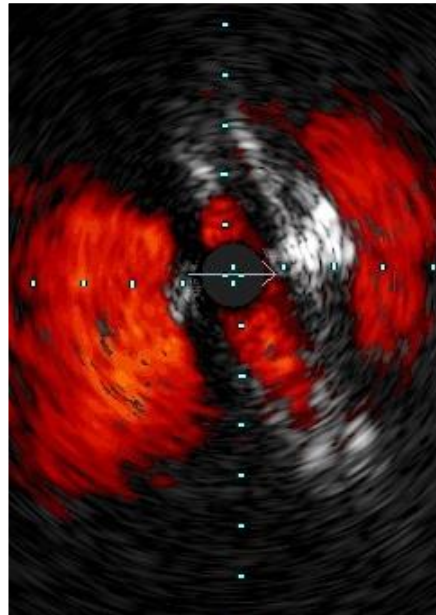
ANOCOR à risque



Trajet rétropulmonaire avec passage intramyocardique



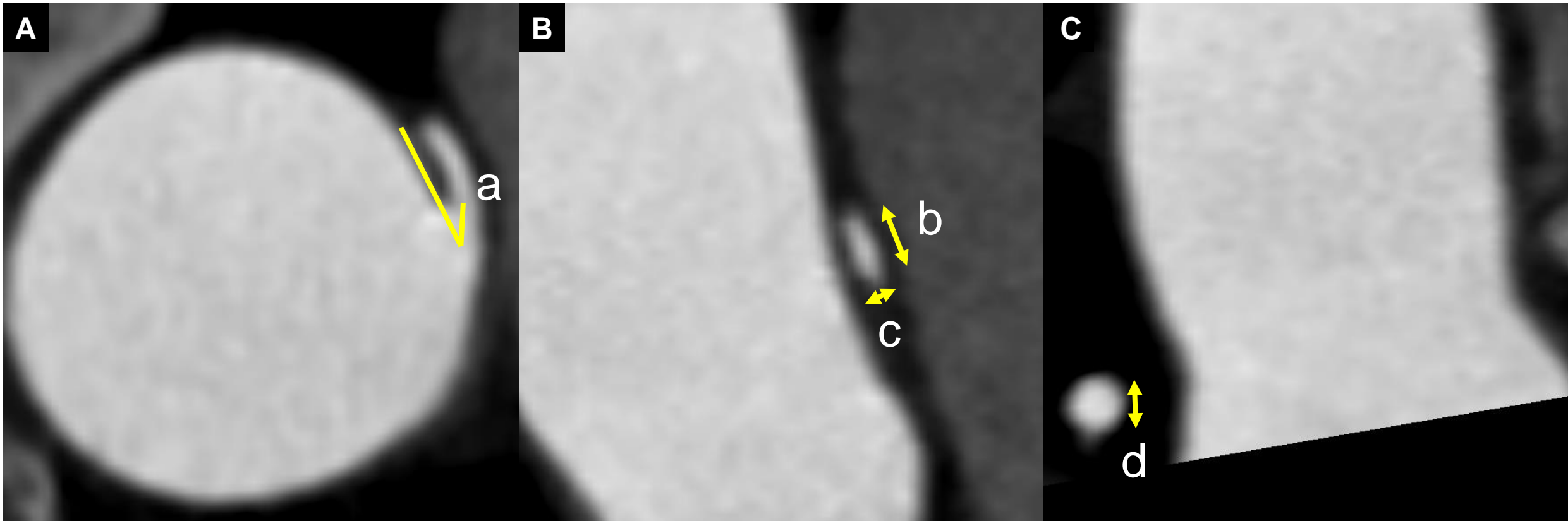
Ischémie myocardique documentée



Prévalence \approx 15%

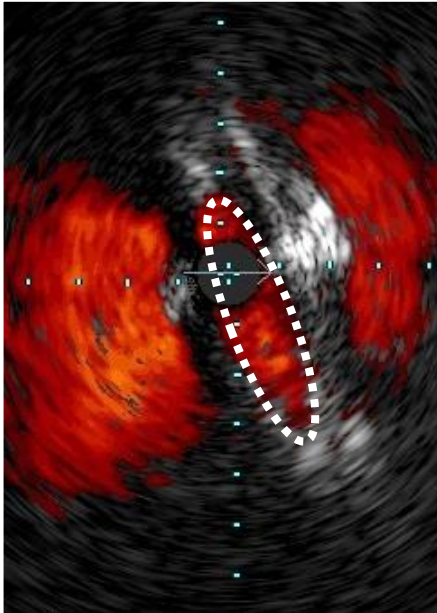
Réduction de diamètre : $(1 - c/d) \times 100 = 40-70\%$

Réduction de surface : $(1 - \text{surface proximale}/\text{surface distale}) \times 100 = 40-70\%$

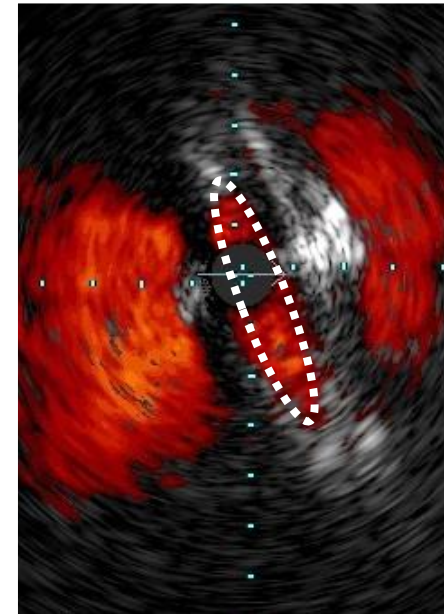
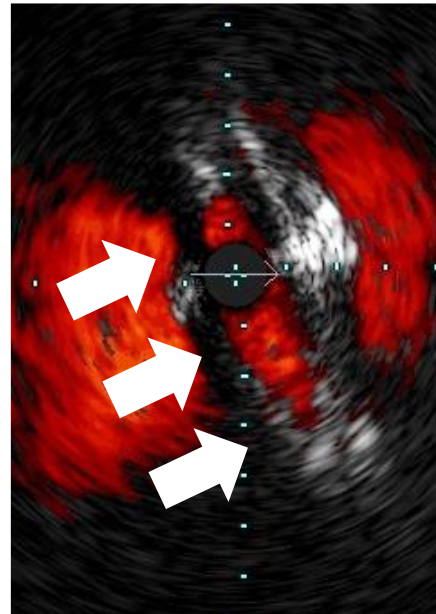


Two-Tier Concept

Fixed Component



Dynamic Component



Anomalies coronaires congénitales de l'adulte

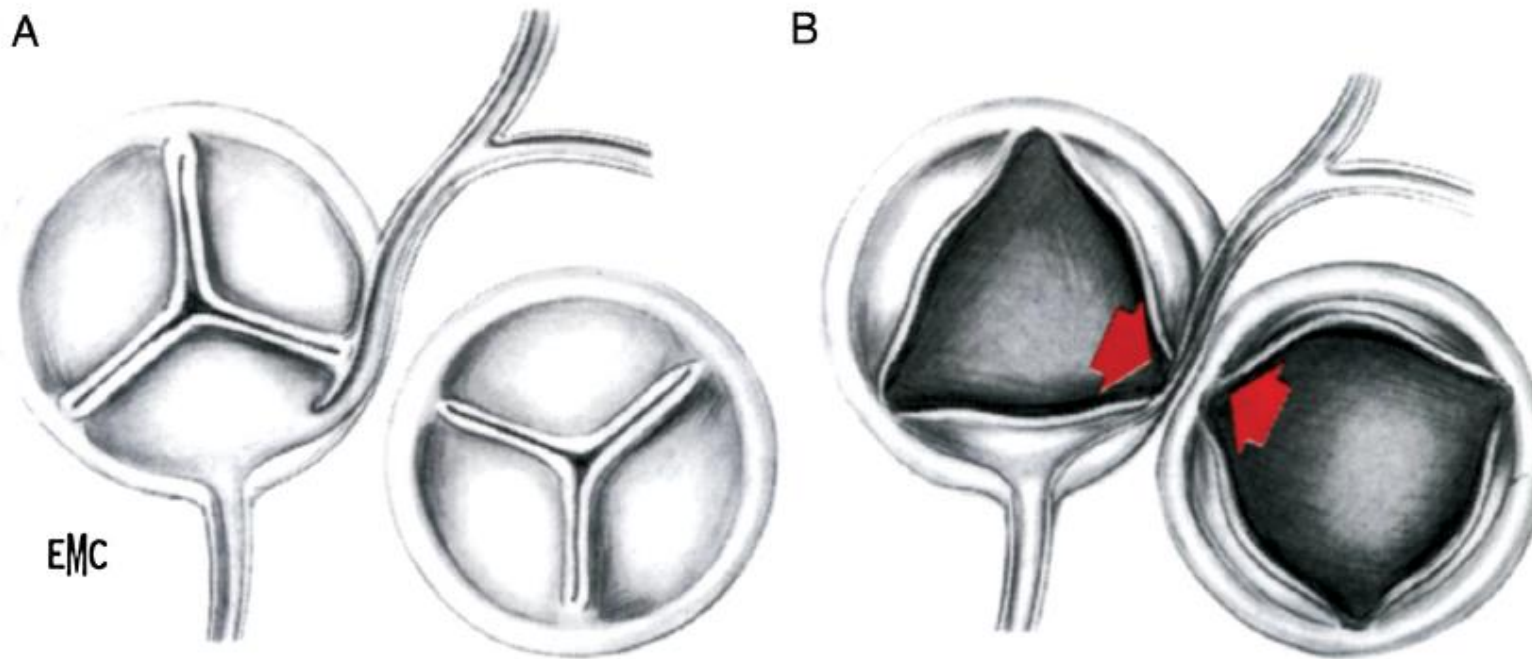
Recherche d'ischémie myocardique

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.

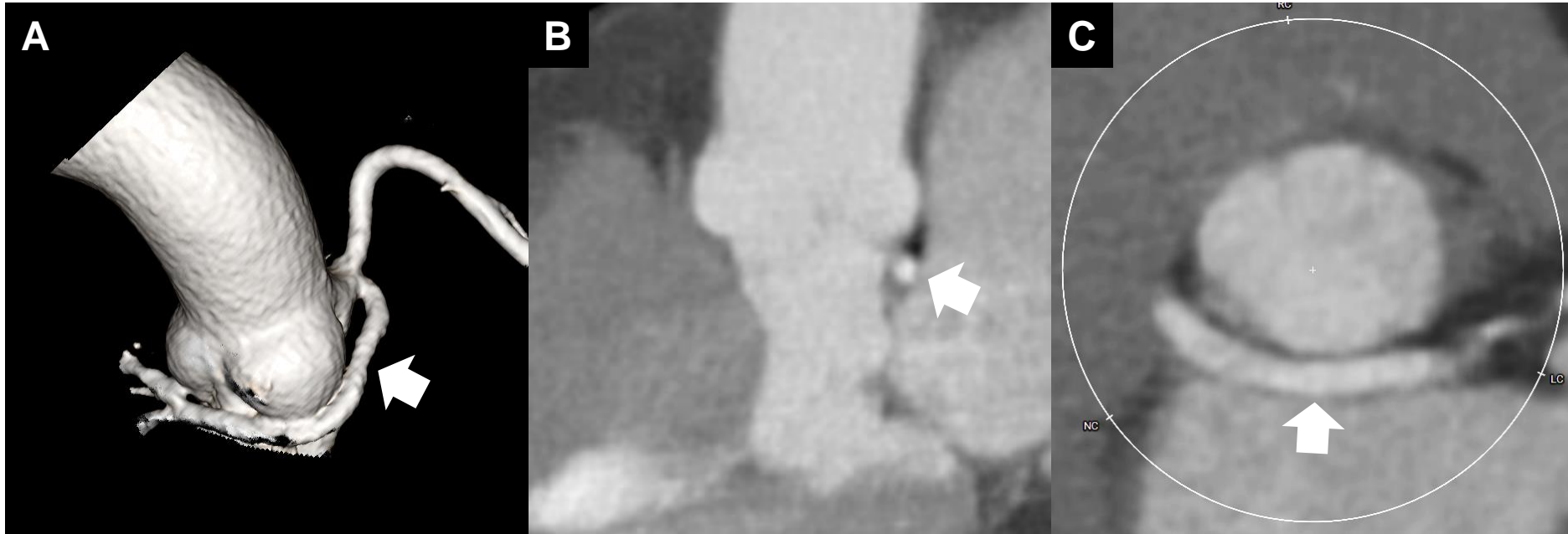
Exertional dynamic compression



Raisky O, Vouhé P. EMC 2007

Never demonstrated

External compression



Aortic valve surgery



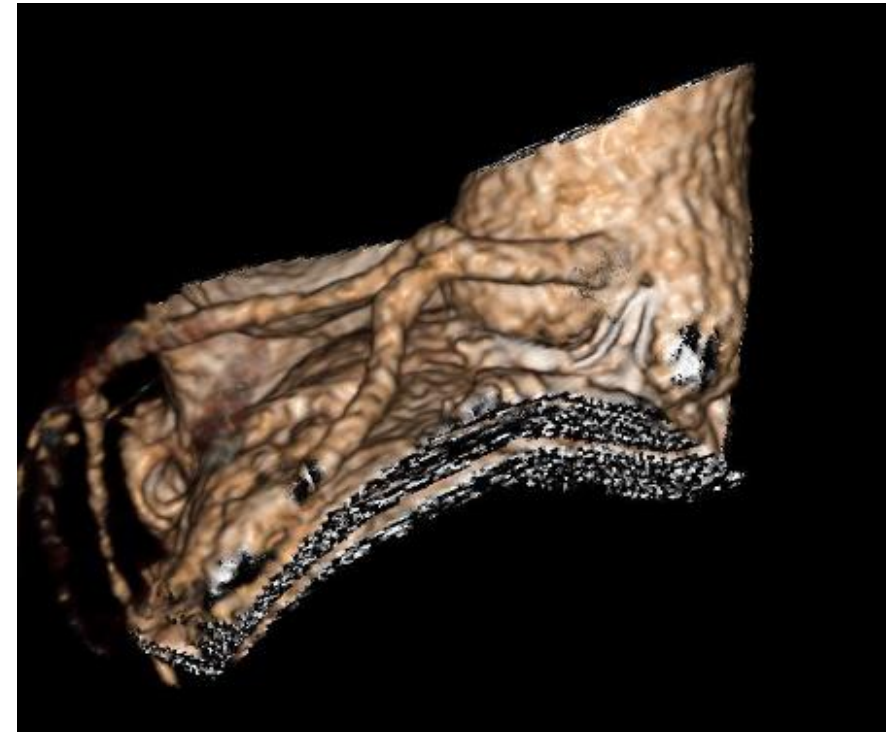
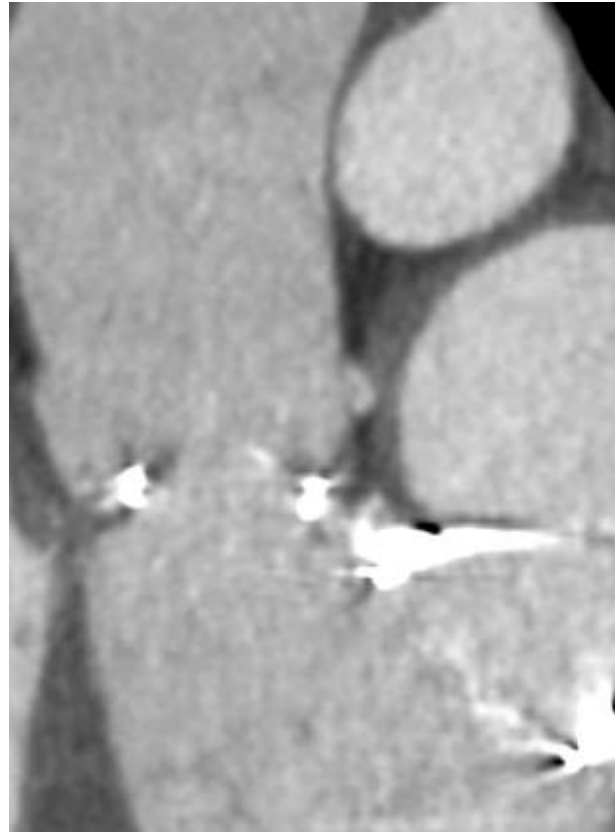
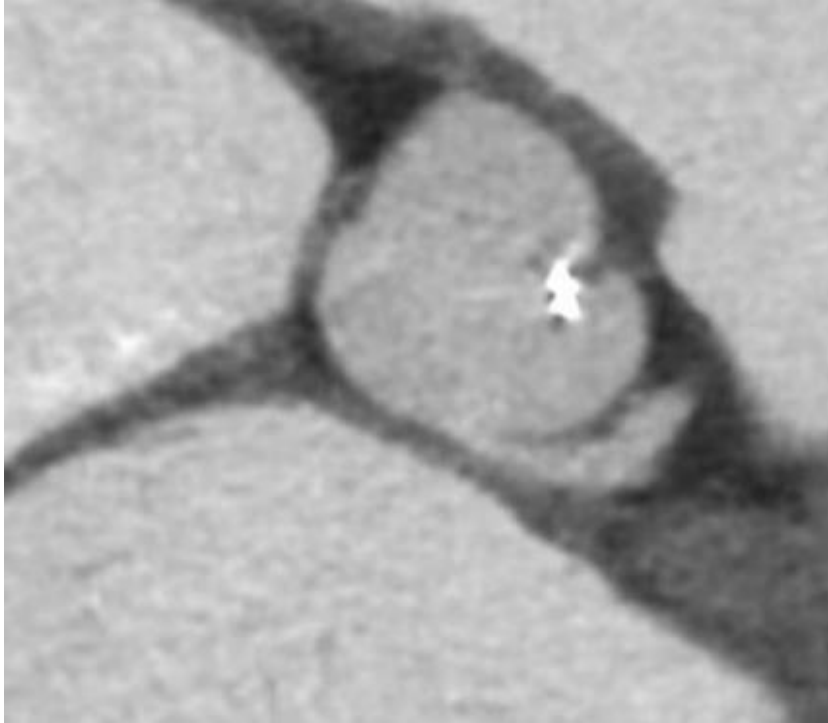
Transcatheter aortic valve implantation

Cas clinique 1

Mme P.

- Femme de 28 ans
- Valvulopathie mitro-aortique post RAA : IM 3/4, IA 3/4
- VG dilaté, FEVG 60%
- Pas de coronarographie/scanner coronaire pré opératoire
- Staff médicochirurgical : remplacement aortique/mitral avec bioprothèses (2015)
- TDR ventriculaires sortie de CEC : reprise avec nouvelle bioprothèse aortique
- ECMO
- Evolution favorable : FEVG 50%

Scanner coronaire

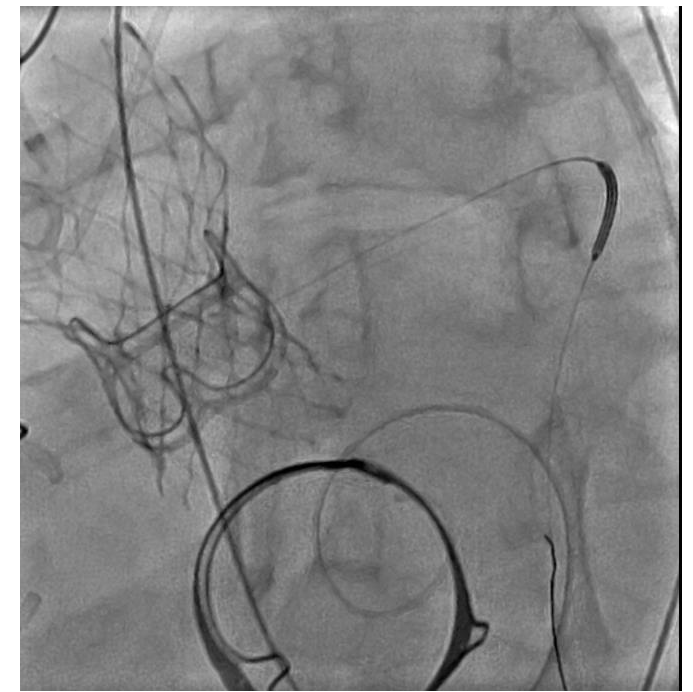
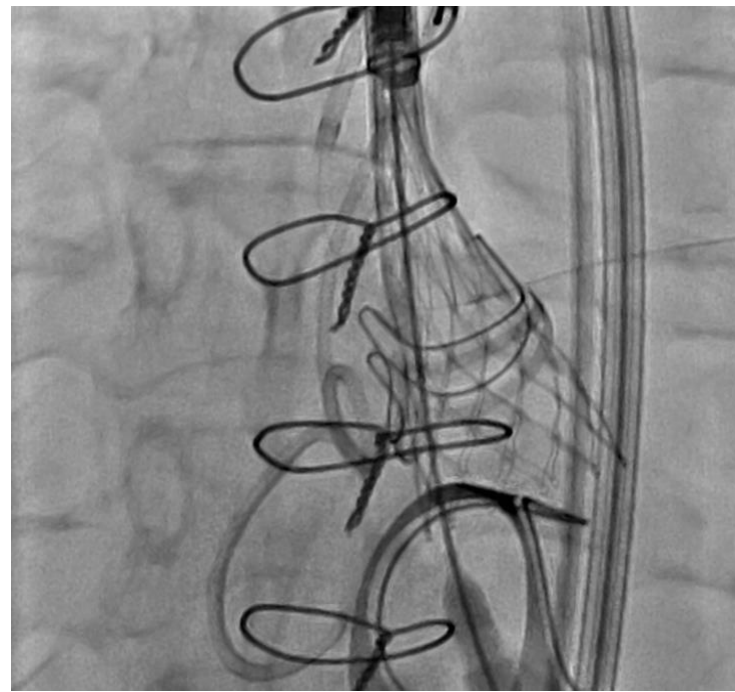
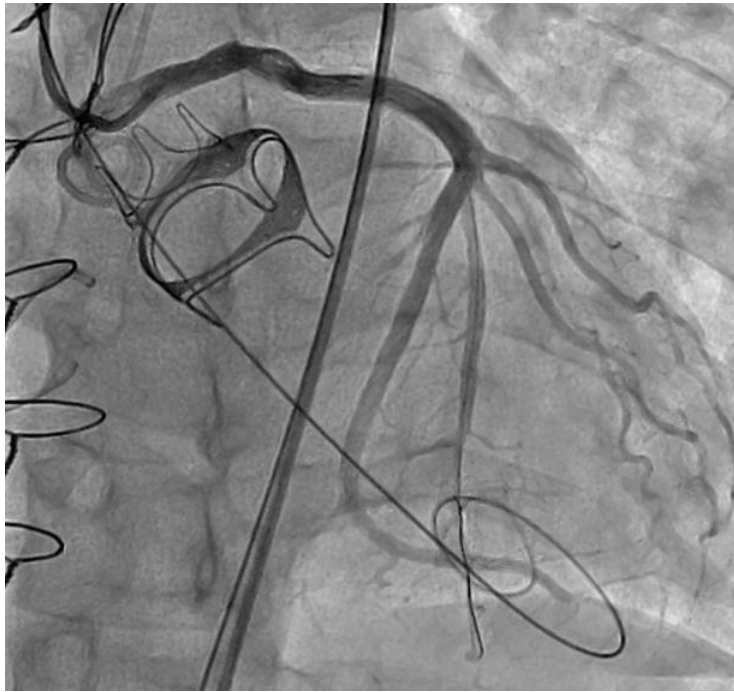


Cas clinique 1

Mme P.

- Dégénérescence deux bioprothèses (sténoses serrées)
- Staff médicochirurgical : TAVI/TMVI

TAVI



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

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

* Anomalie de connexion avec trajet interartériel

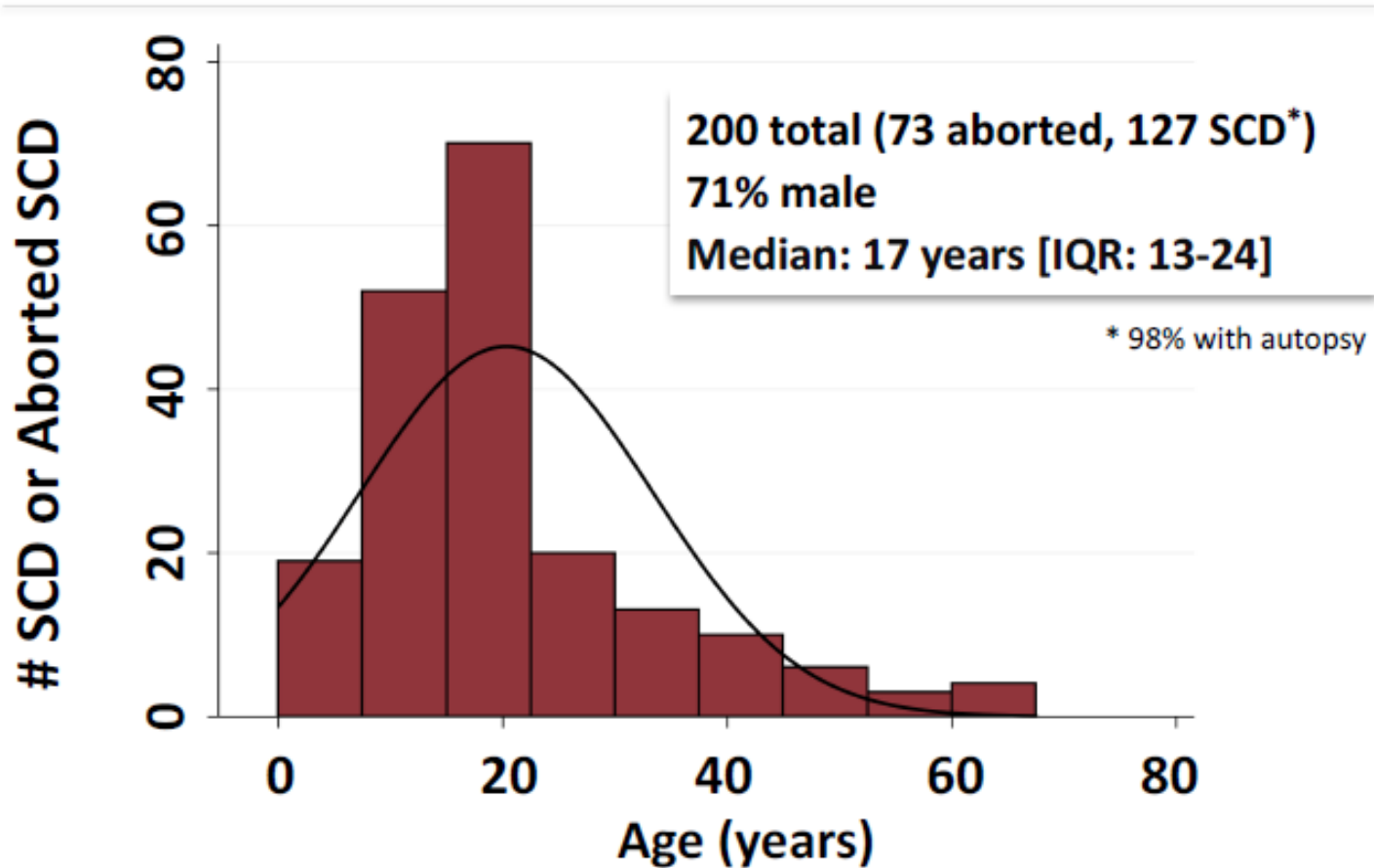
** Nombre de cas à la naissance (estimations)

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

* Anomalie de connexion avec trajet interartériel

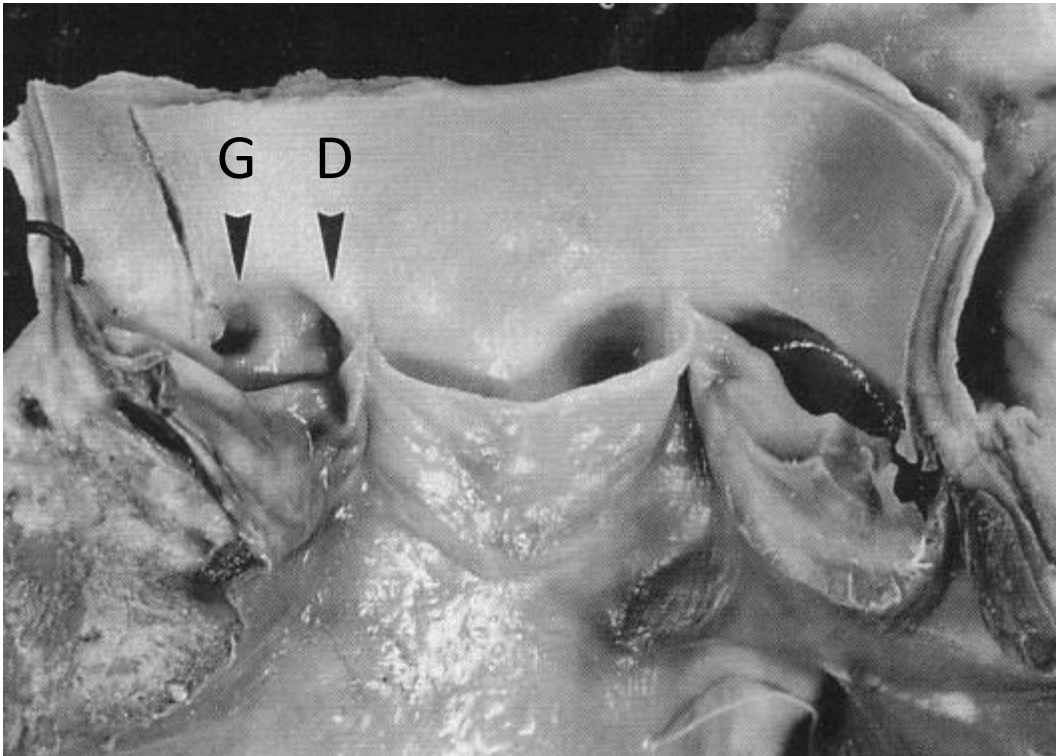
** Incidence annuelle de mort subite (estimations)

Age of SCD or Aborted SCD Attributed to AAOCA



1990

Mort subite



Corrado et al. Br Heart J. 1992.

2010

Arrêt cardiaque récupéré

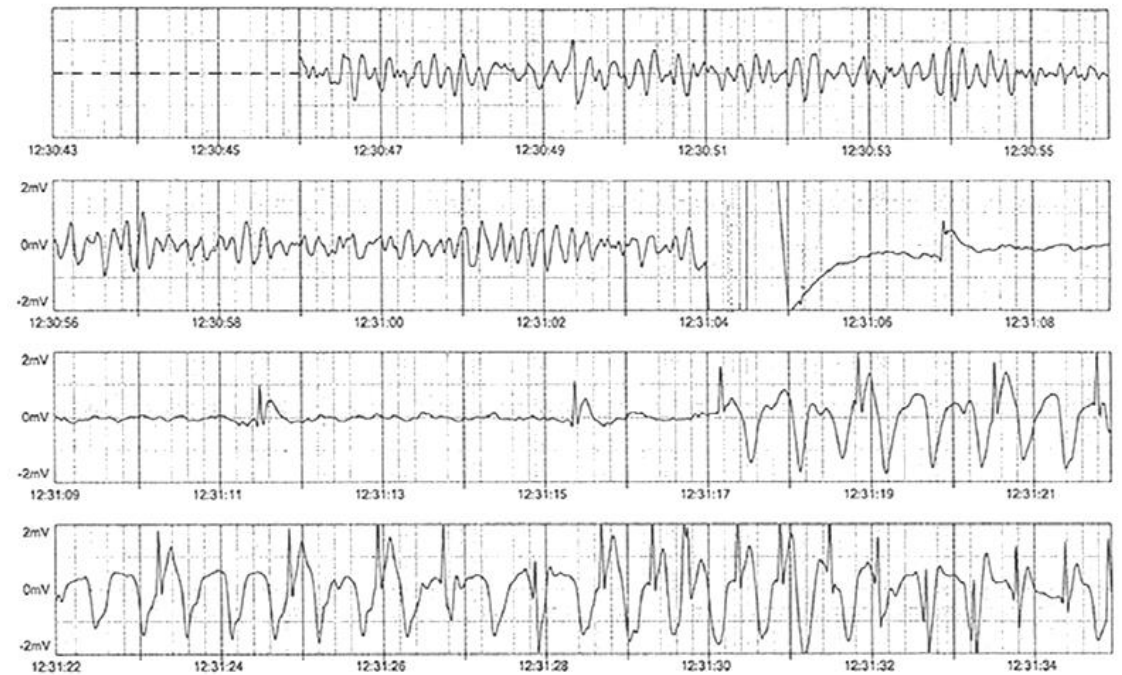
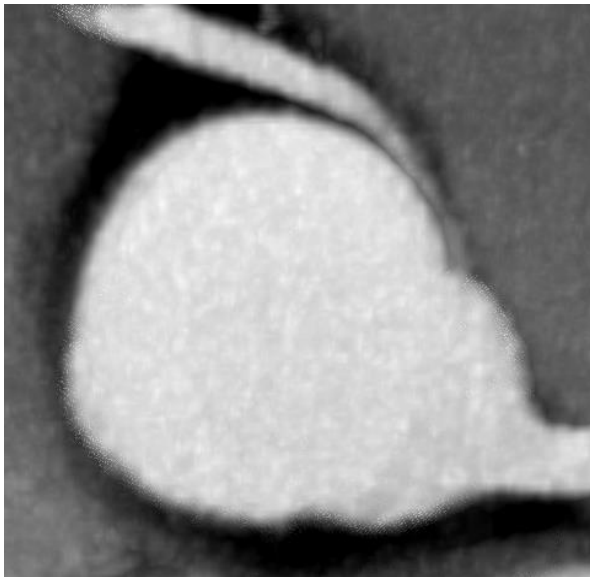
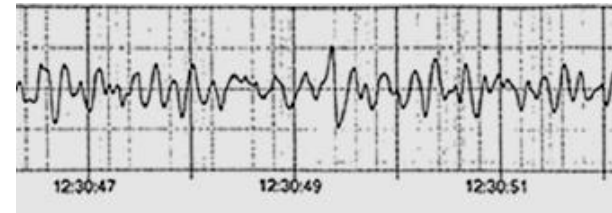


Figure 1. ECG recording from an automated external defibrillator

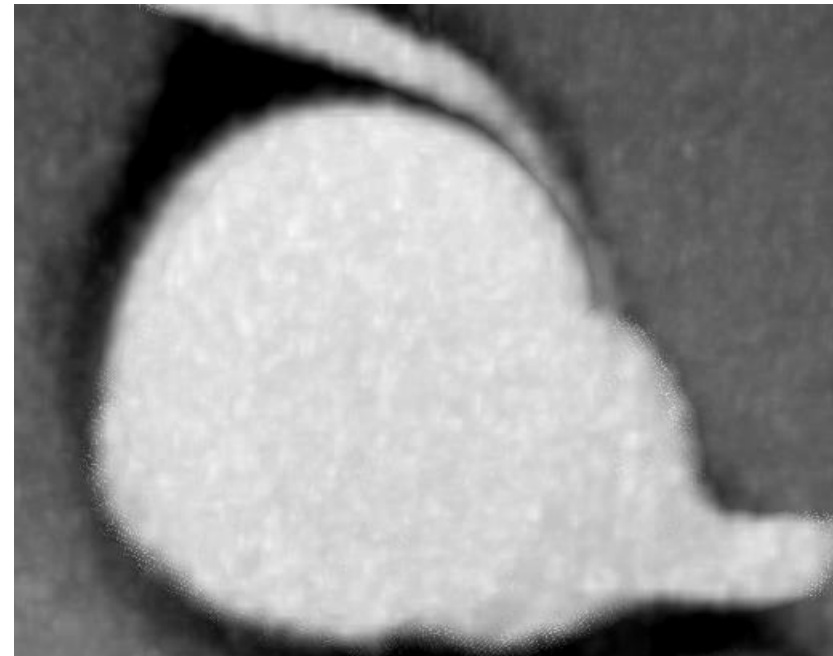
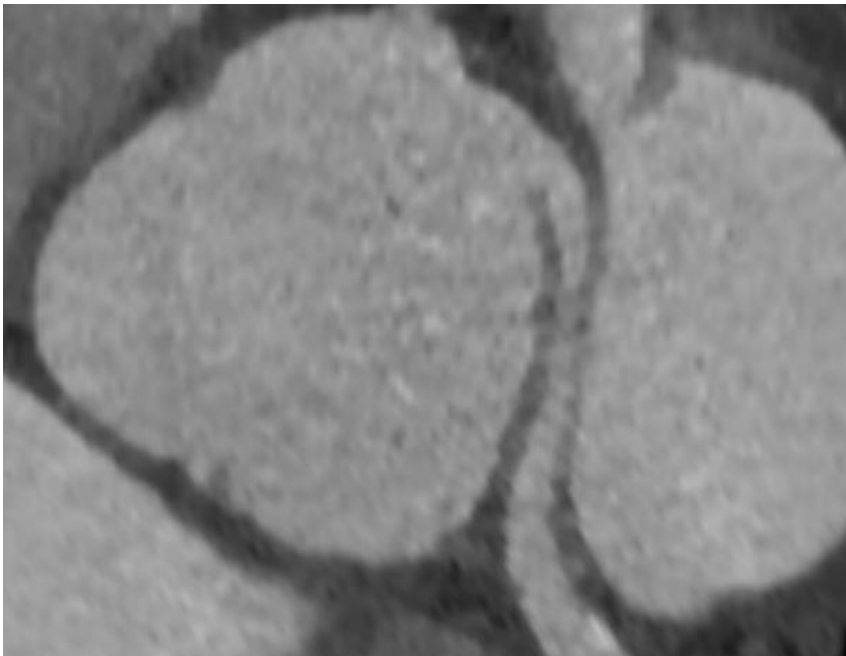
Mécanisme(s) de la fibrillation ventriculaire



24/07/2022
vers 12.20



Sur risque de mortalité pour les formes gauches (x 10)



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

Management of AAOCA at risk

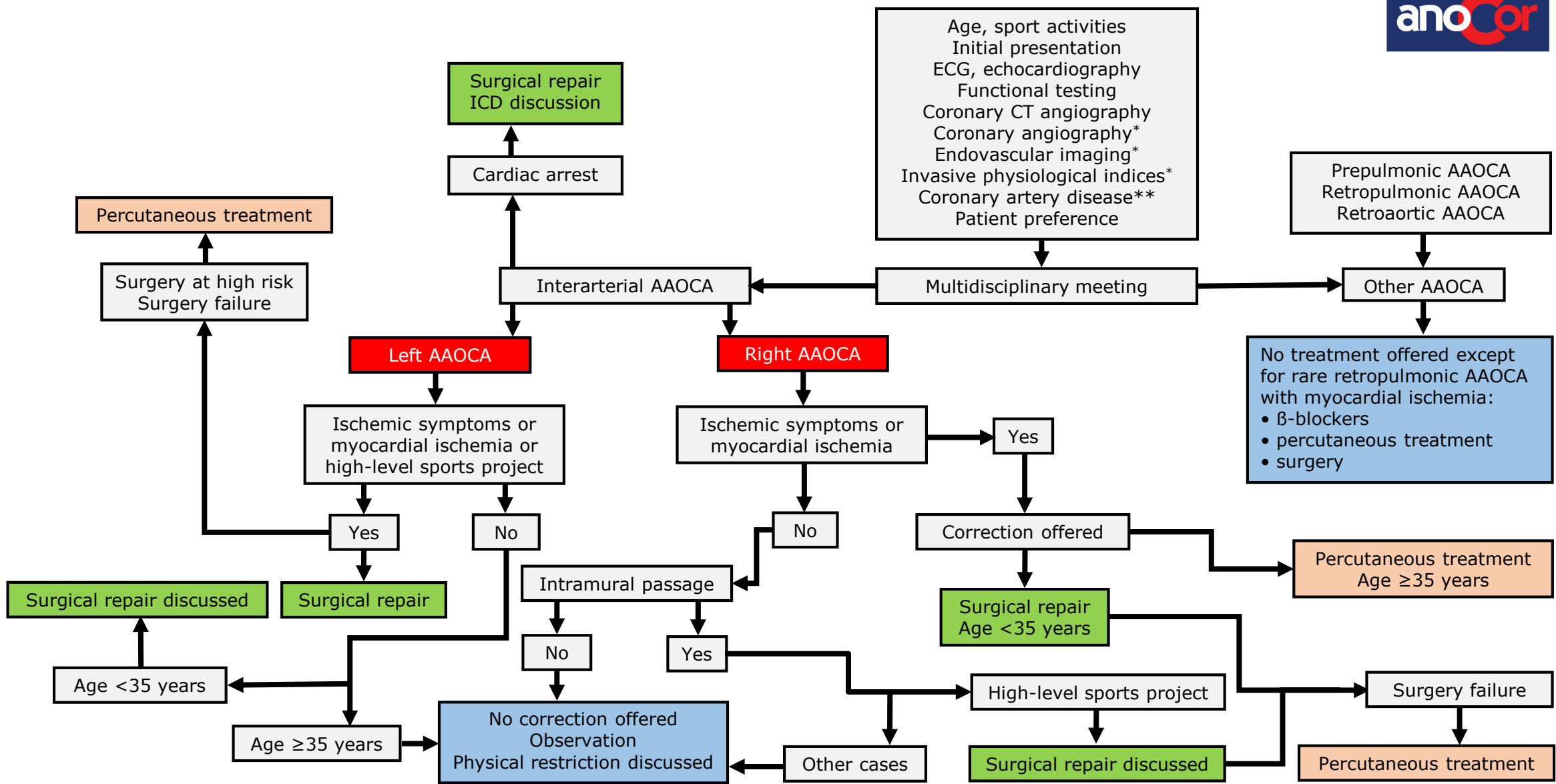
Decision-making

- Age < 35 or ≥ 35 years
- Left AAOCA vs. Right AAOCA
- History of aborted cardiac arrest
- Ischemic symptoms/relationship with exertion
- Induced myocardial ischemia with imaging
- Anatomic characteristics (CT scan/angio/IVUS/OCT)
- Physiological assessment (iFR, FFR)
- Sports profile/Patient choice

No AAOCA risk-SCD score

Surgery/PCI/Medical/Observation/Physical restriction

Algorithm for people ≥12 year-old with AAOCA



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. et al. 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. et al. Eur Heart J. 2020.

Guidelines

Guidelines

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.

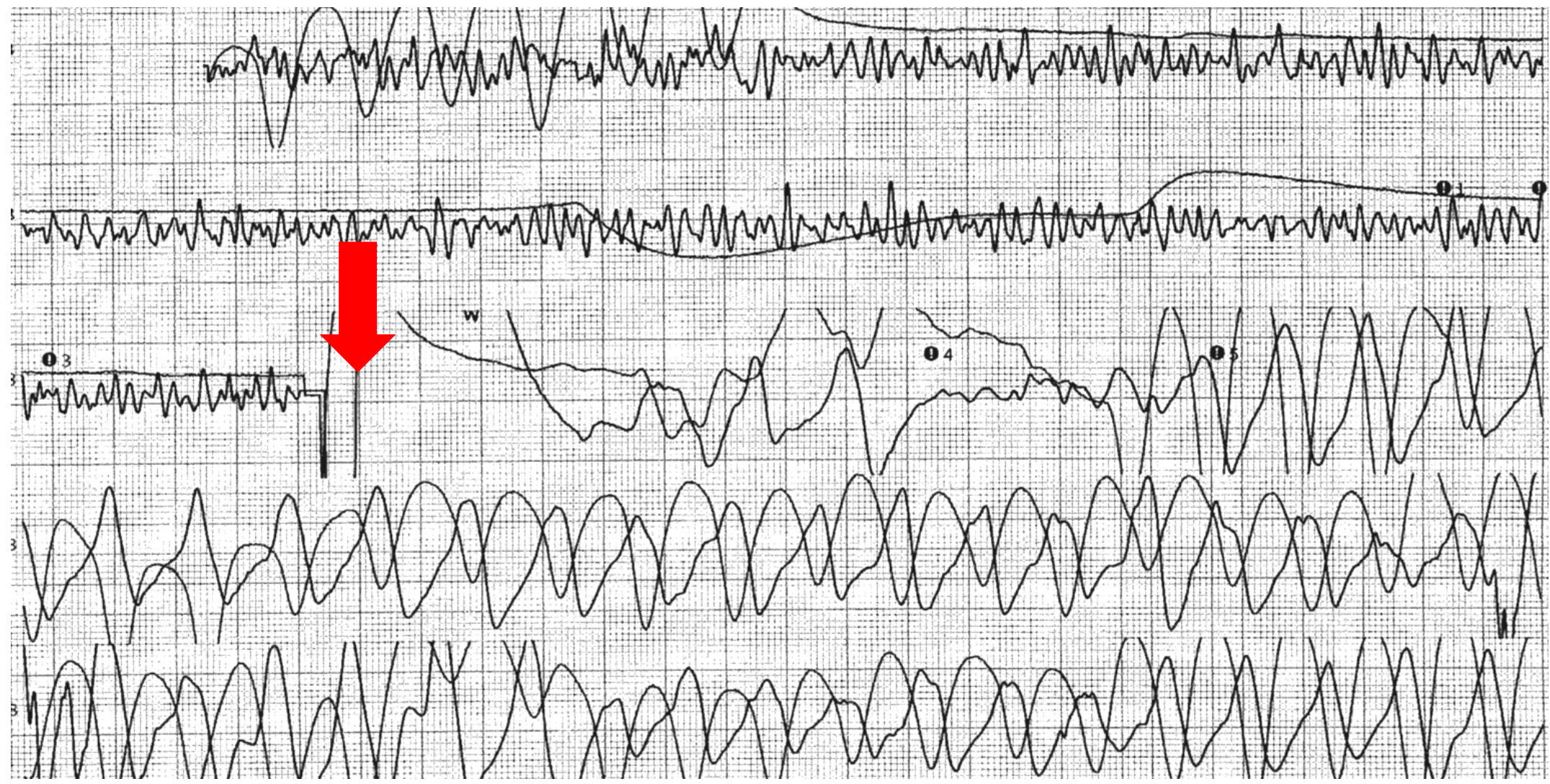
SCA: sudden cardiac arrest

Cas clinique 2

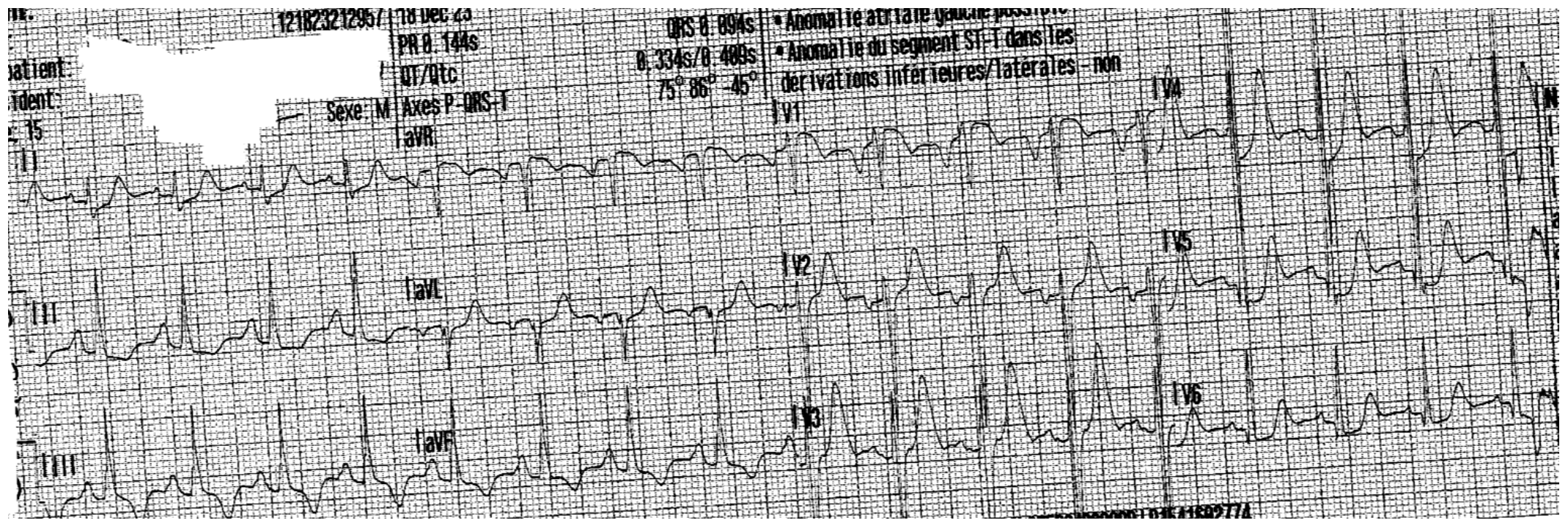
Jeune N.

- Jeune garçon de 15 ans
- Pas d'antécédents cardiovasculaires
- Pratique sportive régulière
- Basket-ball niveau départemental
- Arrêt cardiaque 18/12/2023 début match de basket
- Aucun prodrome
- Rythme choquable
- No flow > 1 min , low flow < 15 min
- Rythme sinusal, sous-décalage ST inférolatéral
- Pic troponine à 6.000 ng/L
- Coronarographie

Tracé DAE



Tracé ECG post ACR



Coronarographie



Scanner coronaire

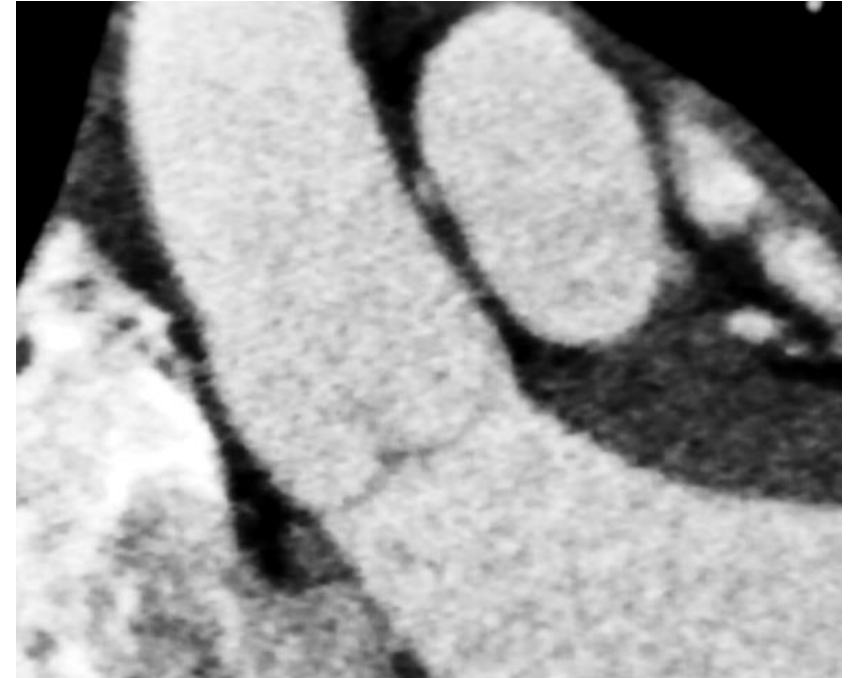
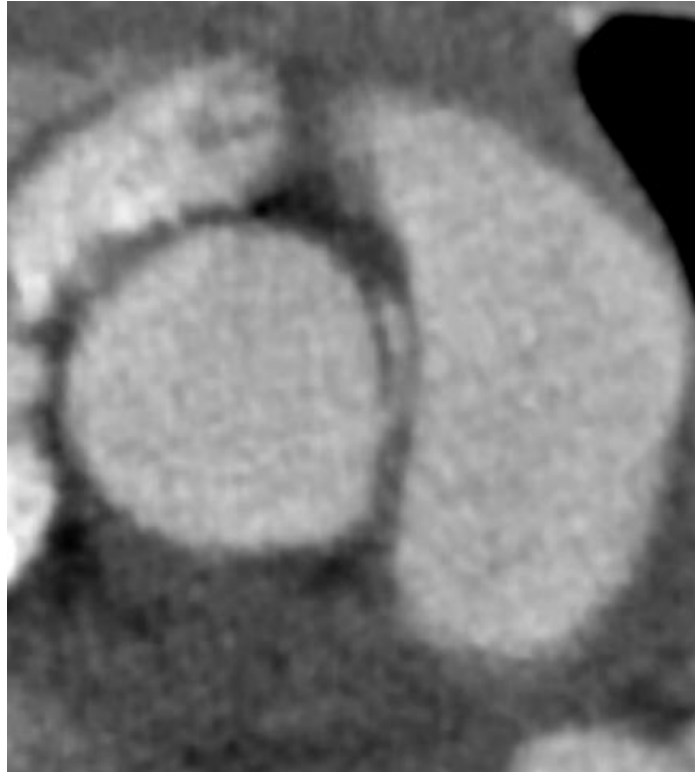
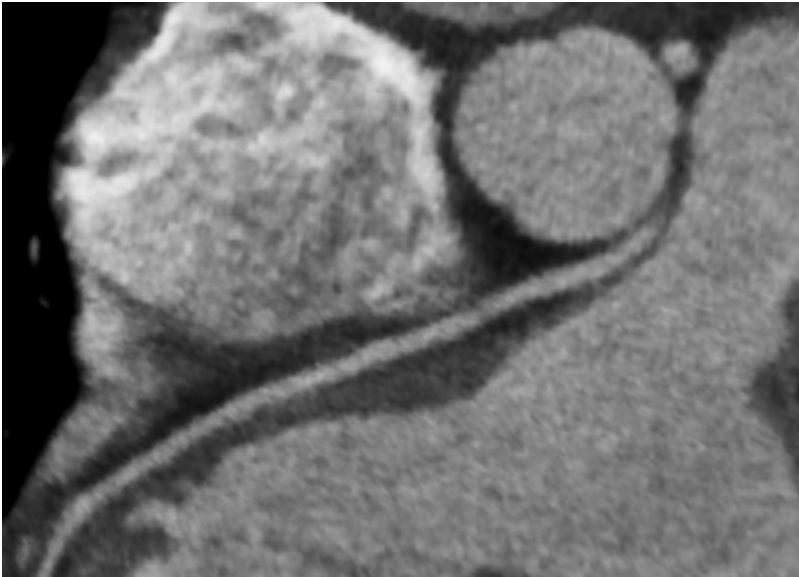
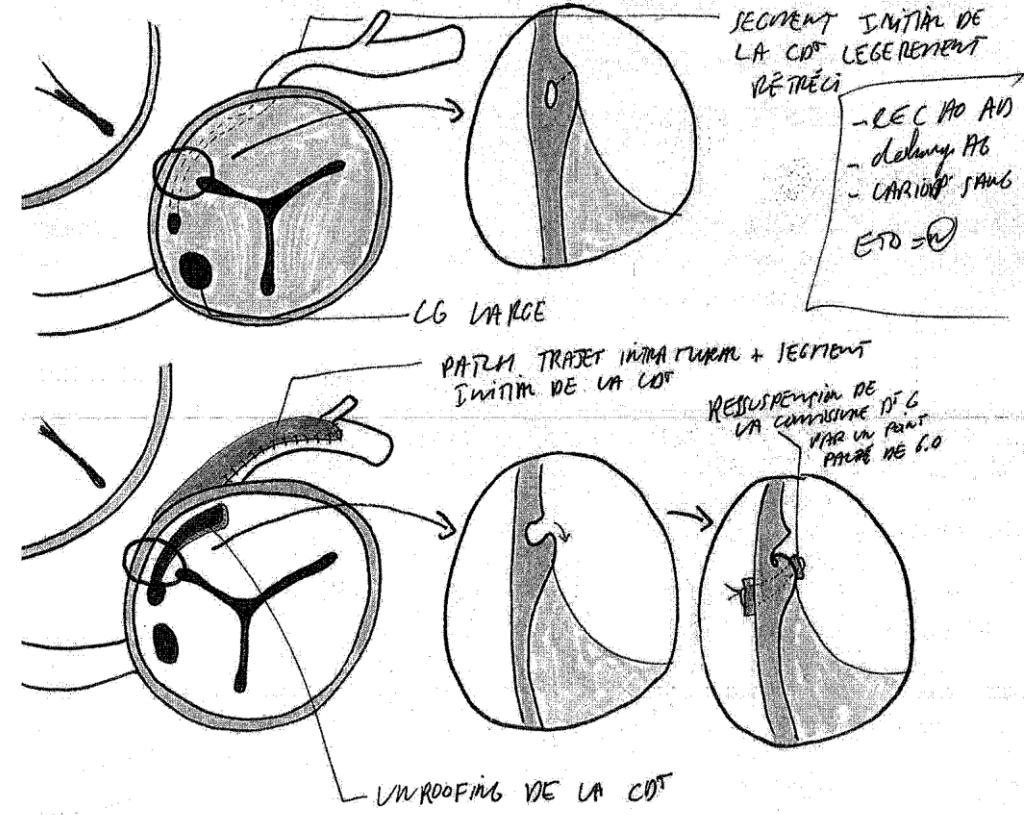


Schéma opératoire

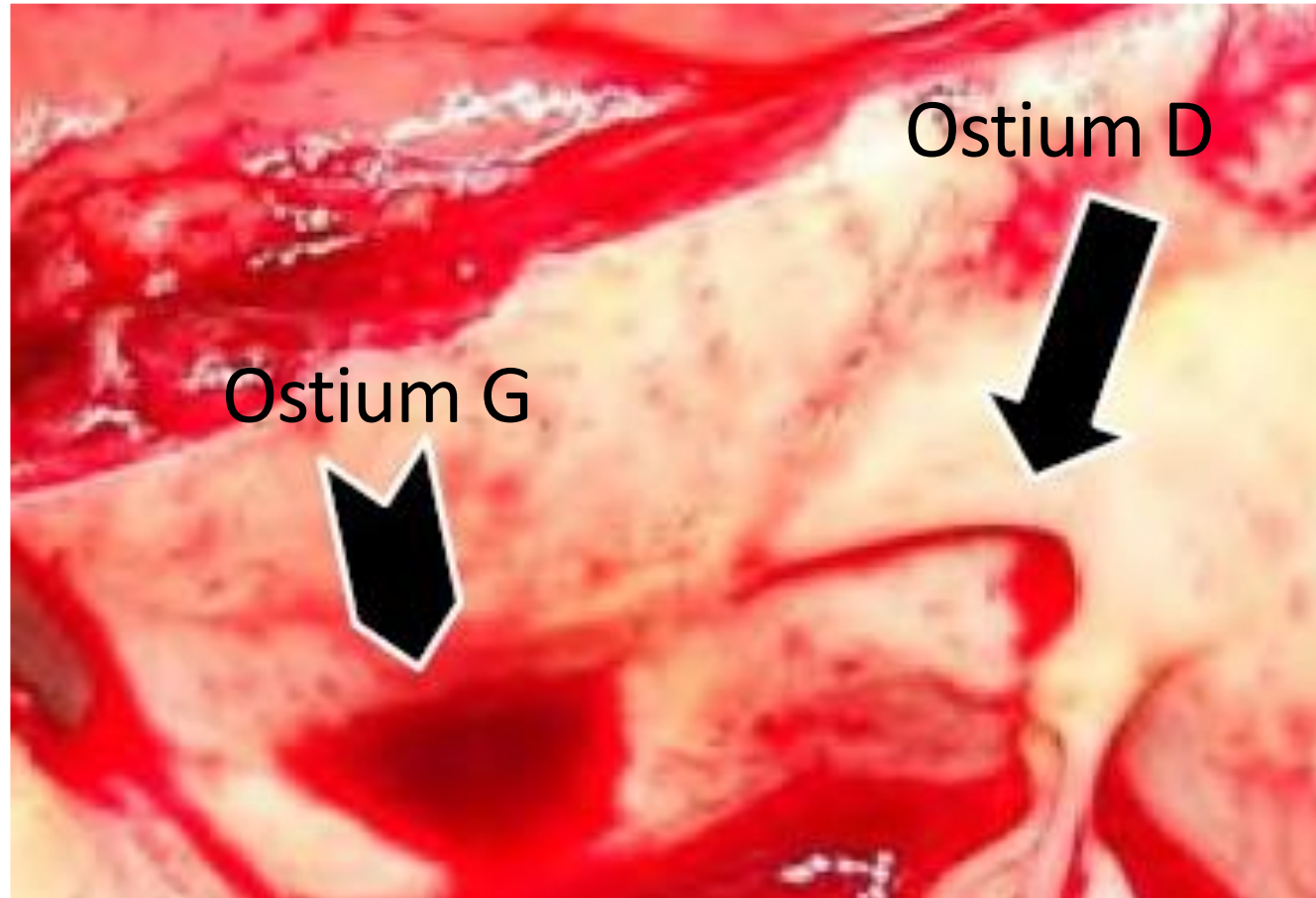
Docteur Gaudin
CHU NECKER

INTERVENTION : NEO - OSTIUM CORONARIEN DROIT
 DIAGNOSTIC : ANOMALIE DE NAISSANCE DE LA CORONARIEN DROITE NEUIS LE SIENS GAUCHE AVEC TRAJET INTRA-MURAL / ACR RECUPERE / BASTON BARR



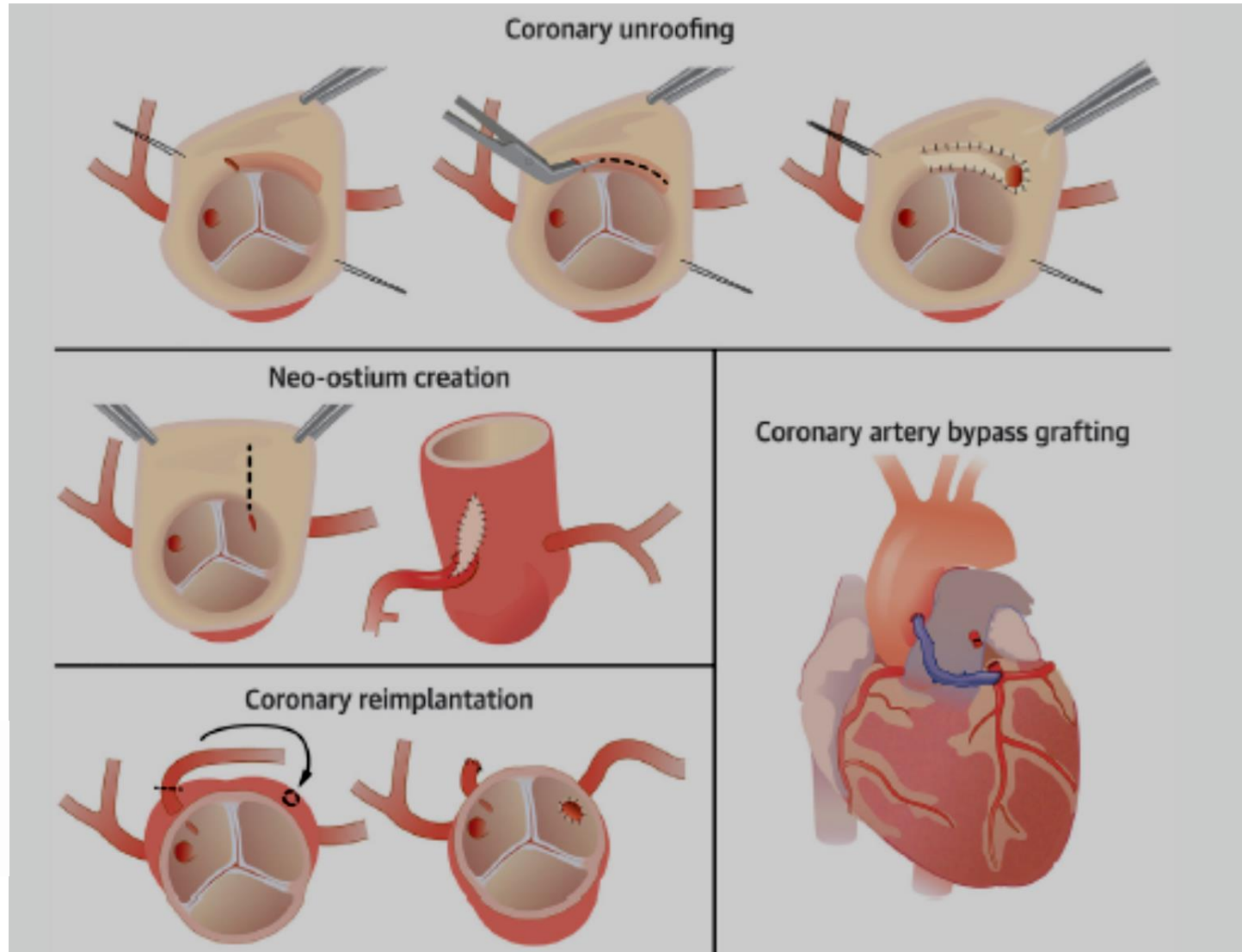
CEC : 112 Clampage aortique: 72min Perfusion cérébrale :
 Codage :
 CIM : Q245 CCAM : D0EA 001 DIAG : CHIR :

ANOCOR droite



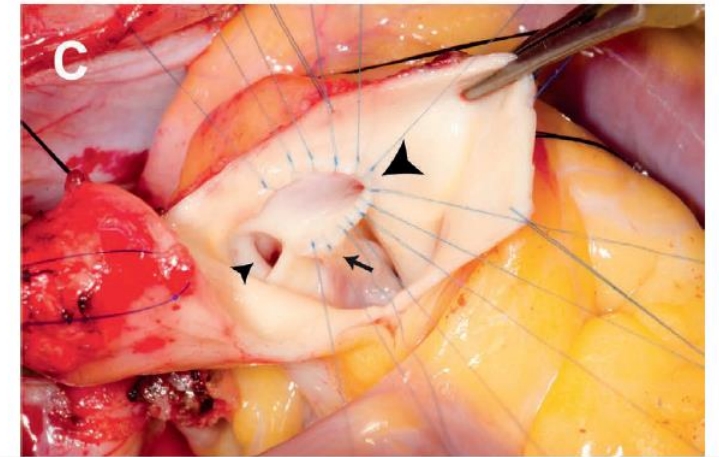
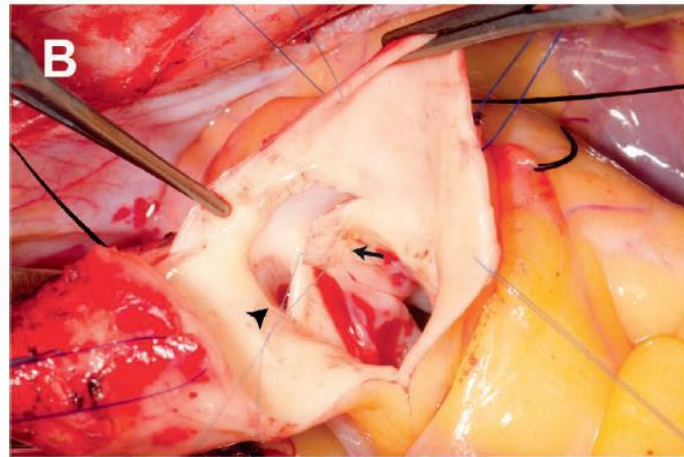
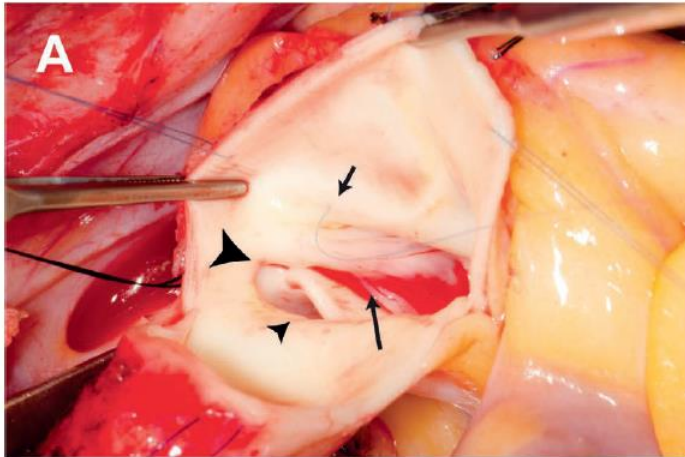
Techniques chirurgicales

ANOCOR avec trajet interartériel



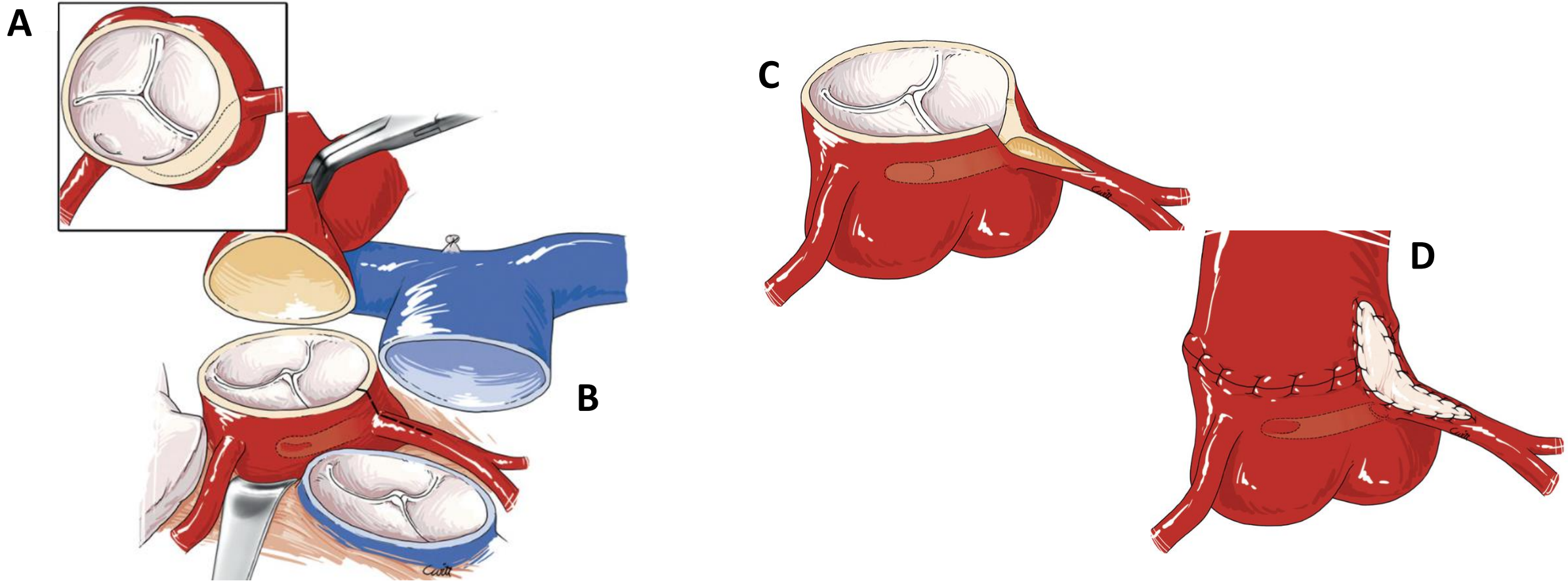
ANOCOR avec trajet interartériel

Unroofing technique

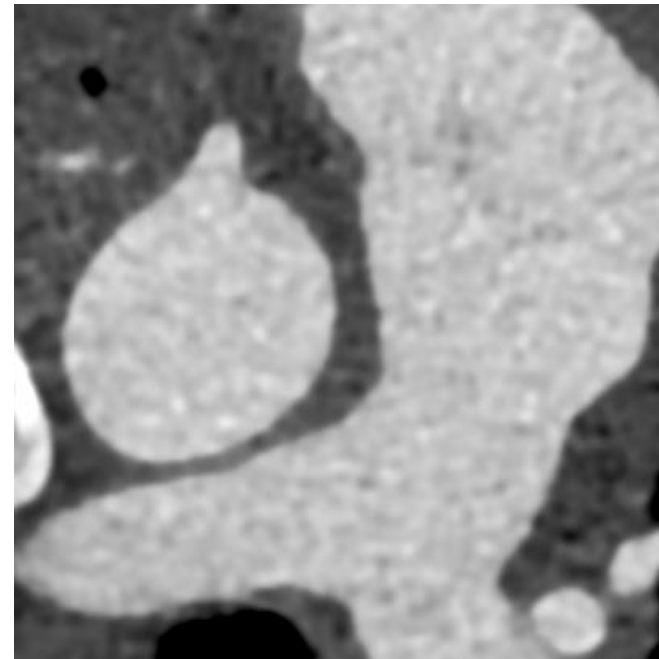
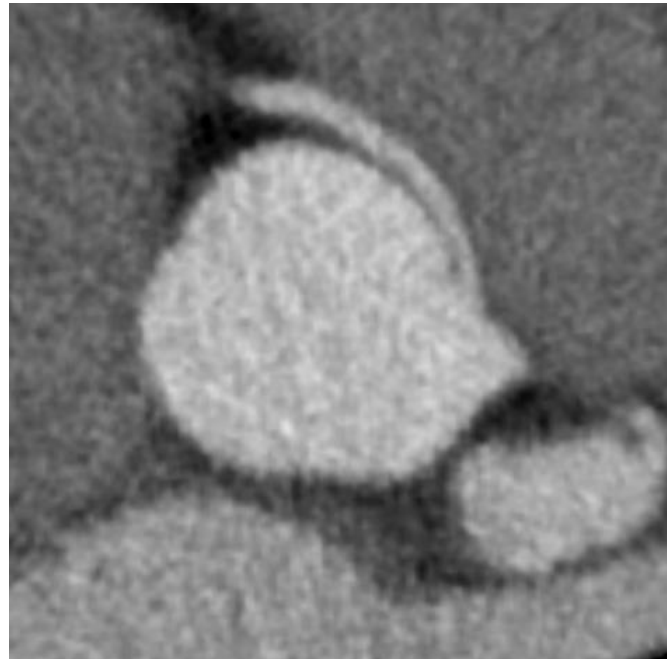


ANOCOR avec trajet interartériel

Création nouvel ostium



Scanner coronaire post chirurgie

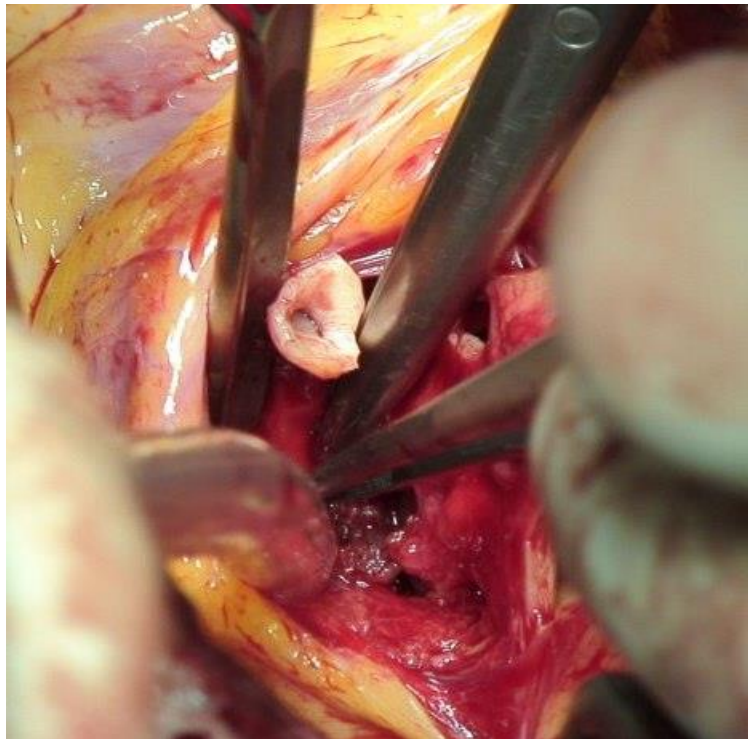


Création nouvel ostium

ANOCOR avec trajet interartériel

Réimplantation coronaire

Rarement adapté



Cas clinique 3

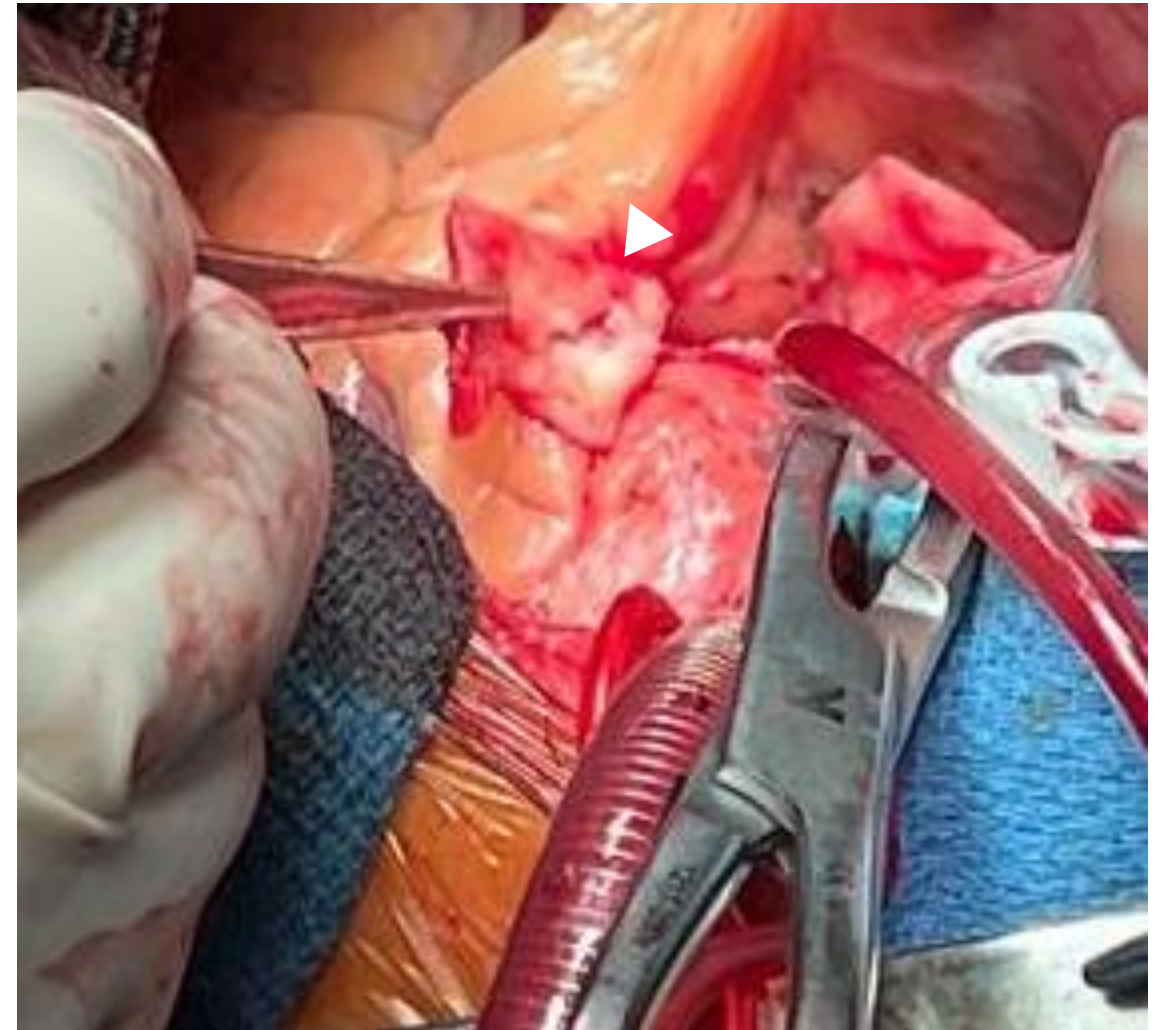
Mme P.

- Femme de 59 ans
- SCA ST- troponine +
- Dilatation aortique : 55 mm (sinus), junction (46 mm), tube (48 mm)
- FEVG 45%
- Coronarographie/scanner coronaire : ANOCOR droite
- IRM cardiaque : rehaussement tardif inférieur
- Staff médicochirurgical : remplacement aortique/reimplantation coronaire/VMA

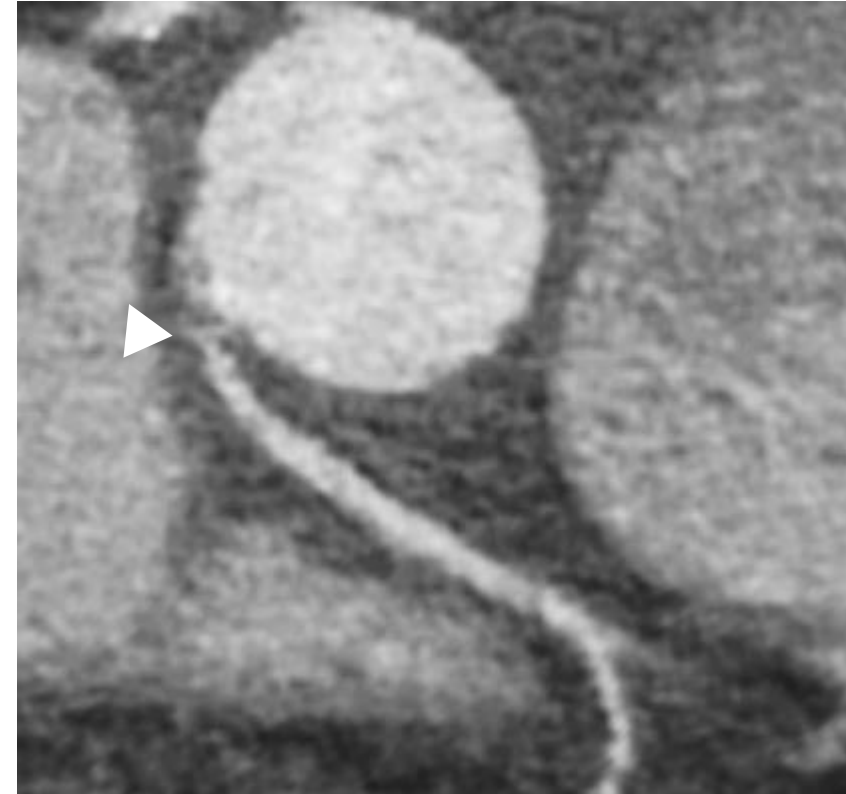
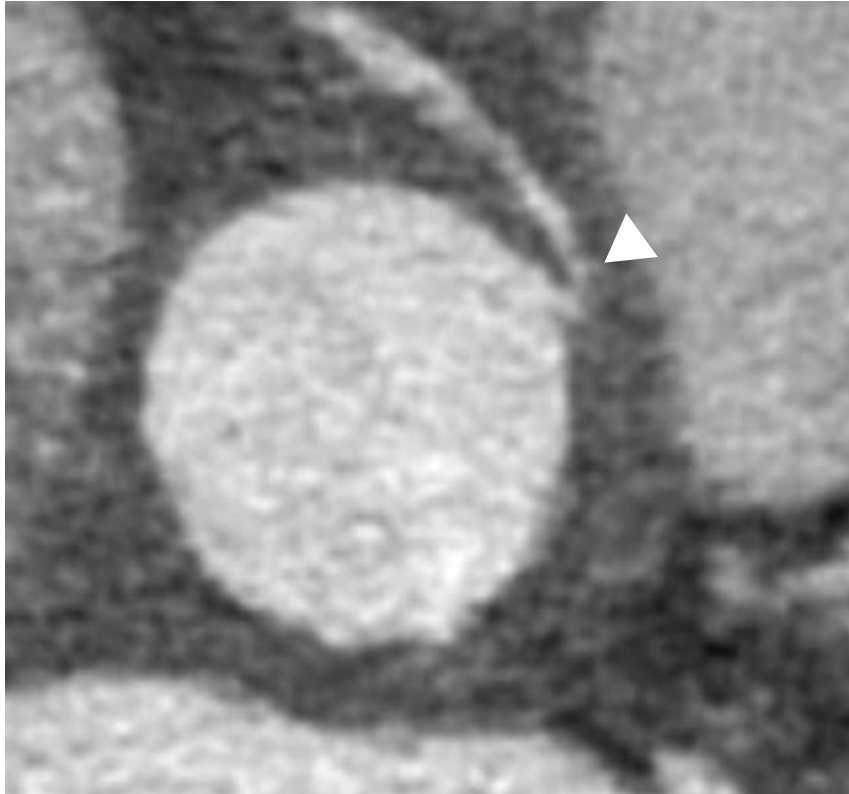
ANOCOR droite



Intervention

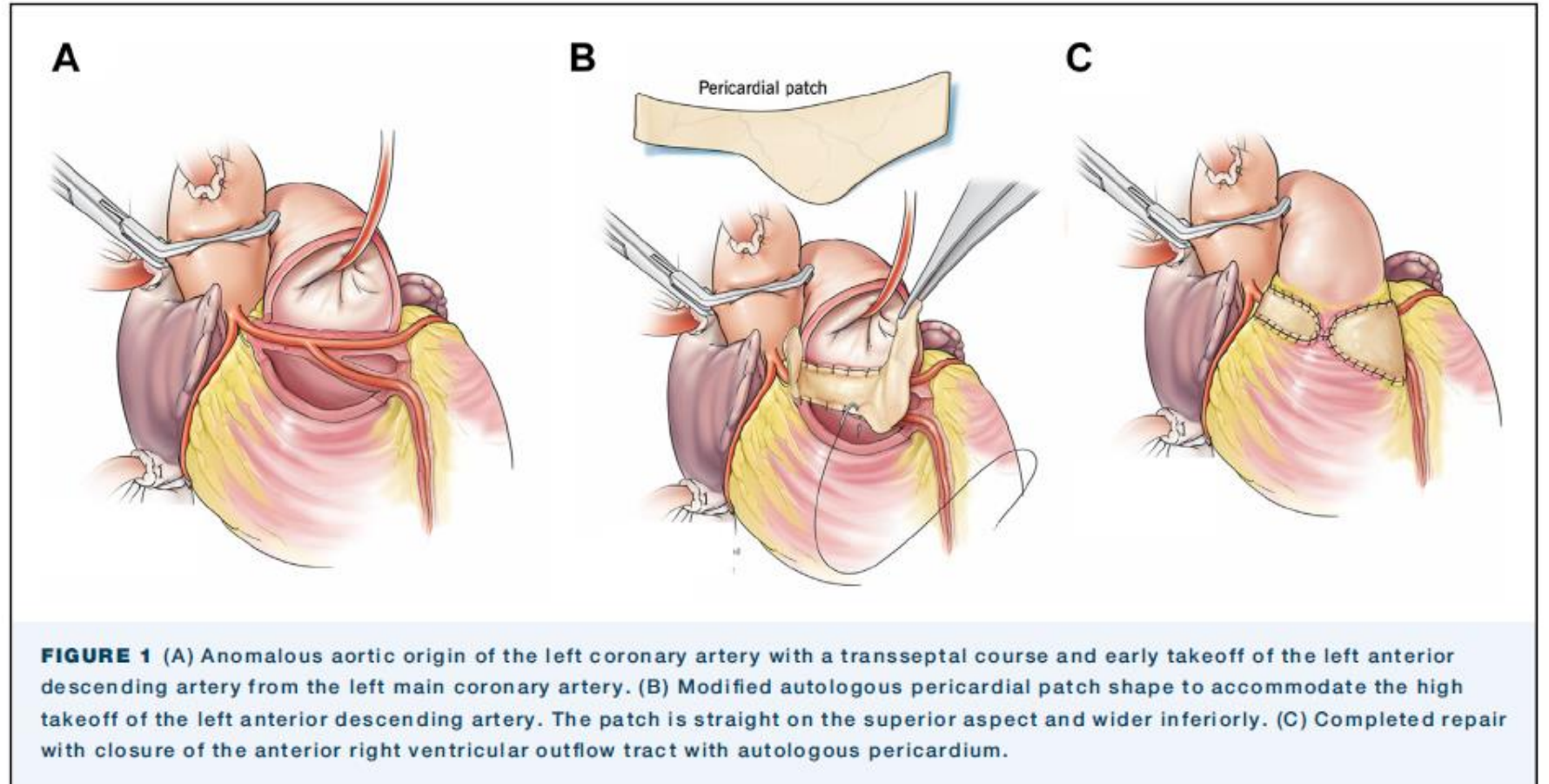


Scanner coronaire post opératoire



Techniques chirurgicales

ANOCOR avec trajet rétropulmonaire







ANOCOR 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, Pontaller 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

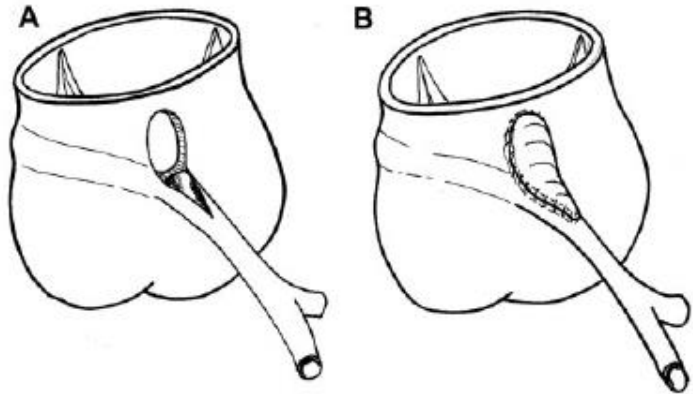
Maïra Gaillard^a, Margaux Pontaller ^a, Pichoy Danial^a, Anne Moreau de Bellaing^a, Régis Gaudin ^a,
Leonora du Puy-Montbrun^a, Bari Murtuza^a, Ayman Haydar^a, Sophie Malekzadeh-Milani ^b,
Damien Bonnet ^b, Pascal Vouhé^a and Olivier Raisky^{a,*}

- 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%)

ANOCOR et chirurgie

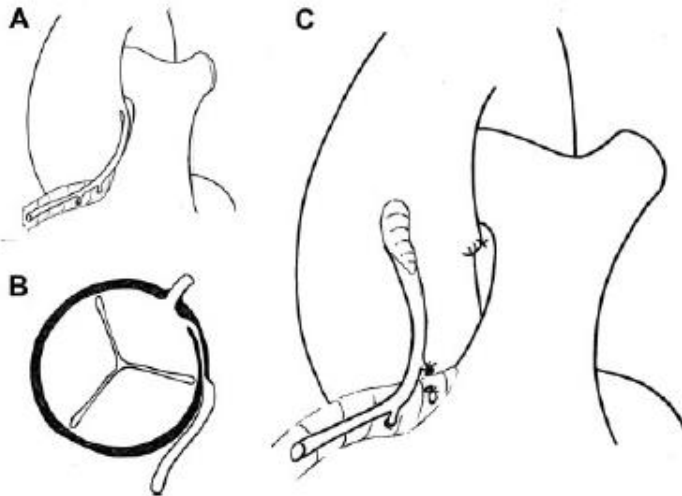
Anomalous aortic origin of coronary arteries: an alternative to the unroofing strategy

Anatomical repair



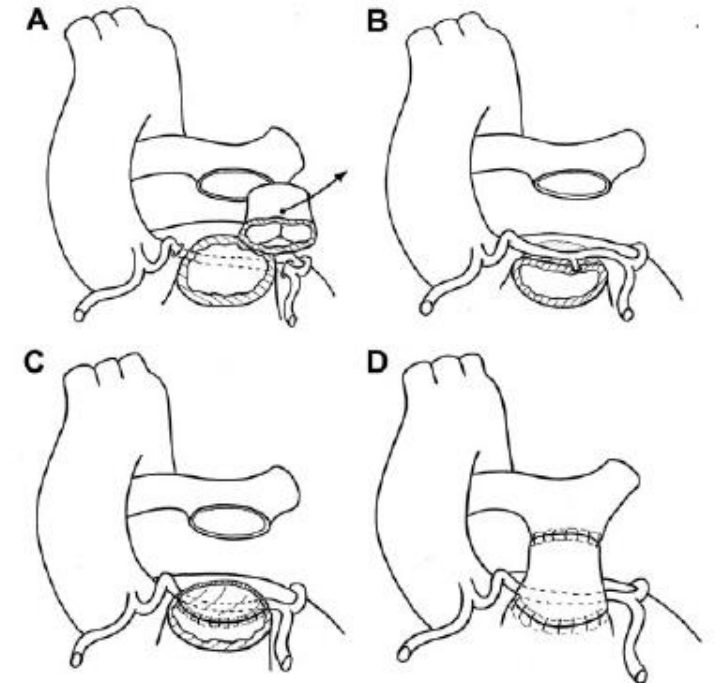
N=37 (60.7%)

Reimplantation



N=19 (31.1%)

Intraseptal relief



N=5 (8.2%)

ANOCOR et chirurgie

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

Early outcomes

Postoperative death : 0

PCI : 2 (3%) : 1 coronary thrombosis, 1 patch plication

Redo surgery : 1 (1.5%) : cardiac arrest/coronary thrombosis/ECMO

Late outcomes

Mean follow-up (years) : 38 months (1-15 years)

Death : 0

PCI : 2 (3%) : 1 recurrent restenosis, 1 scar stenosis

Redo surgery : 2 (3.5%) : 1 CABG, 1 surgical revision (patch aneurysm)

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

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

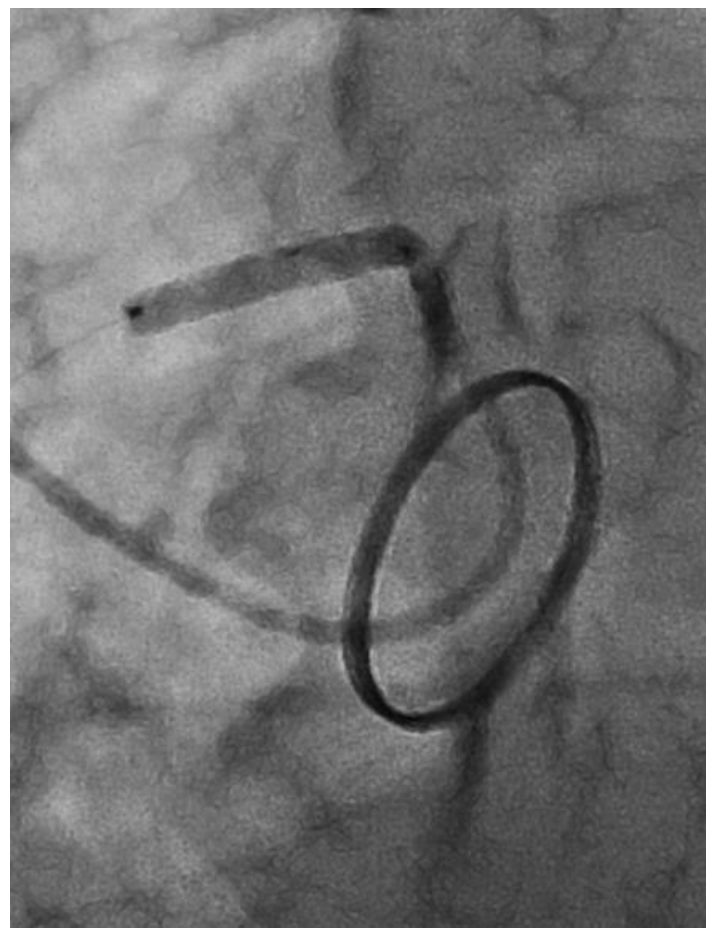
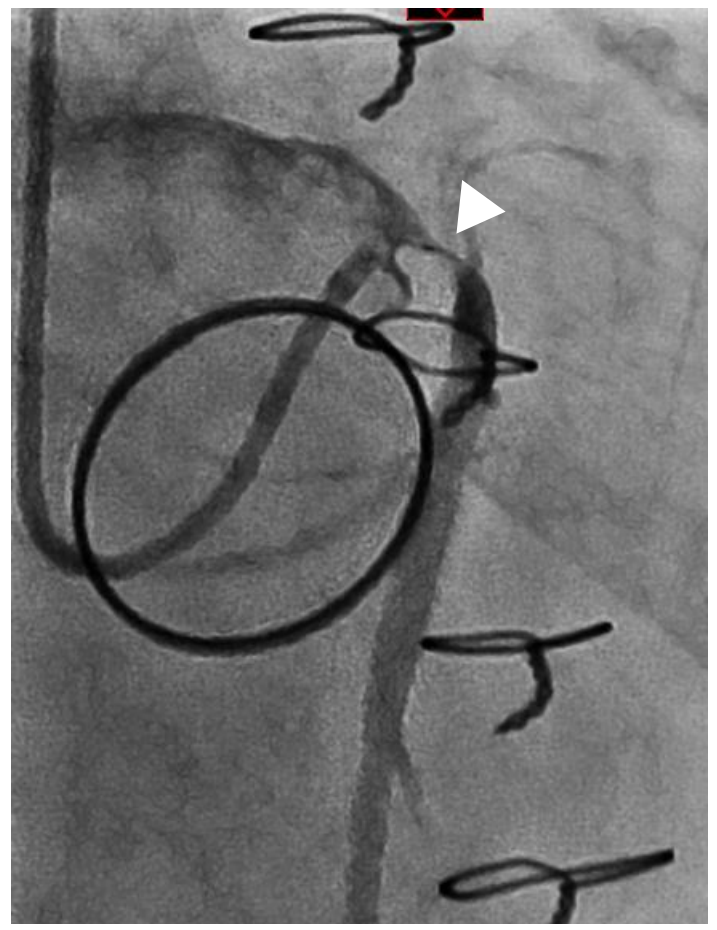
Angioplastie coronaire droite ectopique



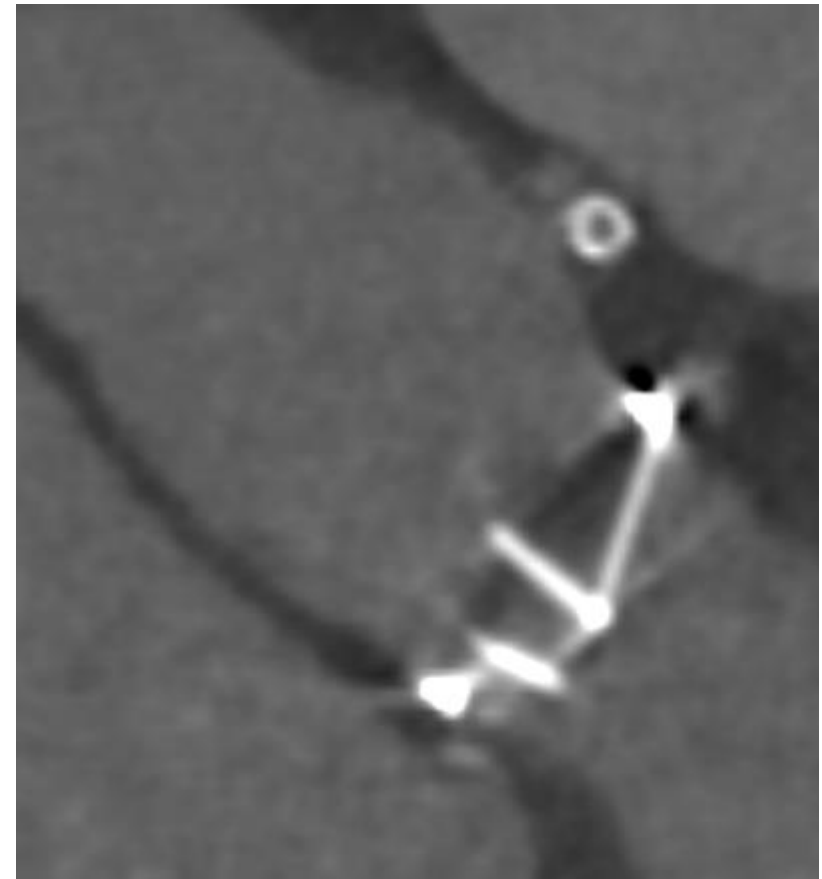
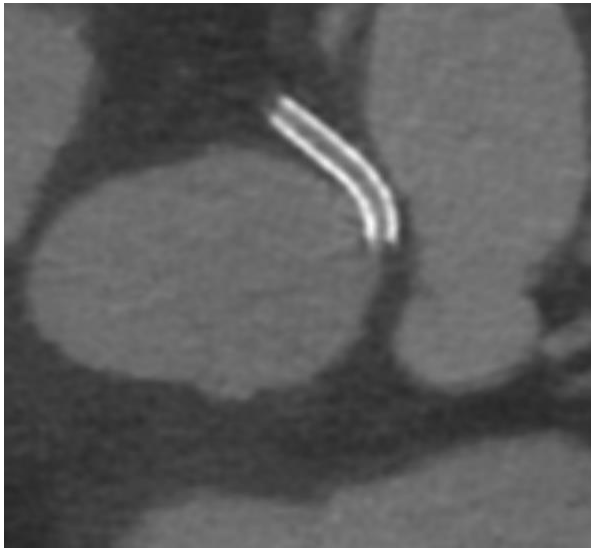
Est-ce possible ?

Cas clinique 3

Angioplastie coronaire



Scanner coronaire post angioplastie



ANOCOR 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
- Quelques échecs : thrombose précoce, sténose cicatricielle, anévrisme, insuffisance aortique

Groupe de travail multidisciplinaire sur les ANOMalies congénitales des artères CORonaires

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[Imagerie](#) | [Chirurgie](#) | [Angioplastie](#) | [Sport](#) | [Nos publications](#) | [Bibliothèque](#) | [Enseignement et formation](#) | [Fistules coronaires](#) | [Infos patients](#) |

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Groupe multidisciplinaire ANOCOR

Anomalies Coronaires Congénitales

Ouvert février 2024

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