

session pédagogique

Connexions anormales des coronaires : de A à Z

Pierre Aubry, 75018 Paris

Patrick Dupouy, 92160 Antony

Xavier Halna du Fretay, 45770 Saran

DÉCLARATION DE LIENS D'INTÉRÊT AVEC LA PRÉSENTATION

Intervenant : Pierre Aubry, Paris

Je n'ai pas de lien d'intérêt à déclarer

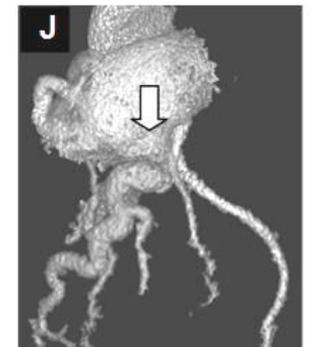
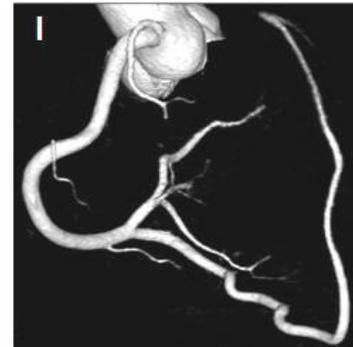
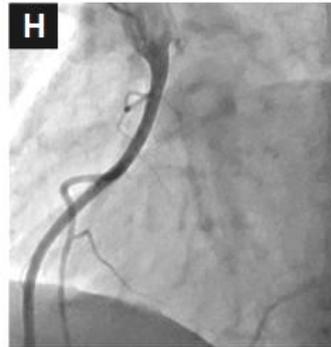
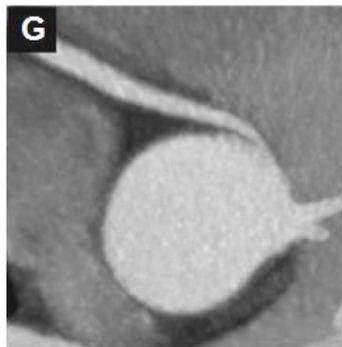
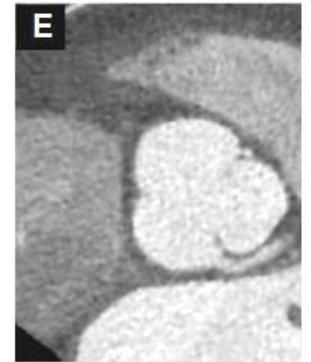
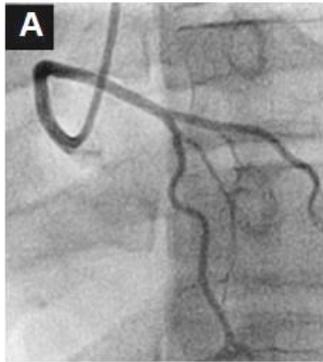
Intervenant : Patrick Dupouy, Antony

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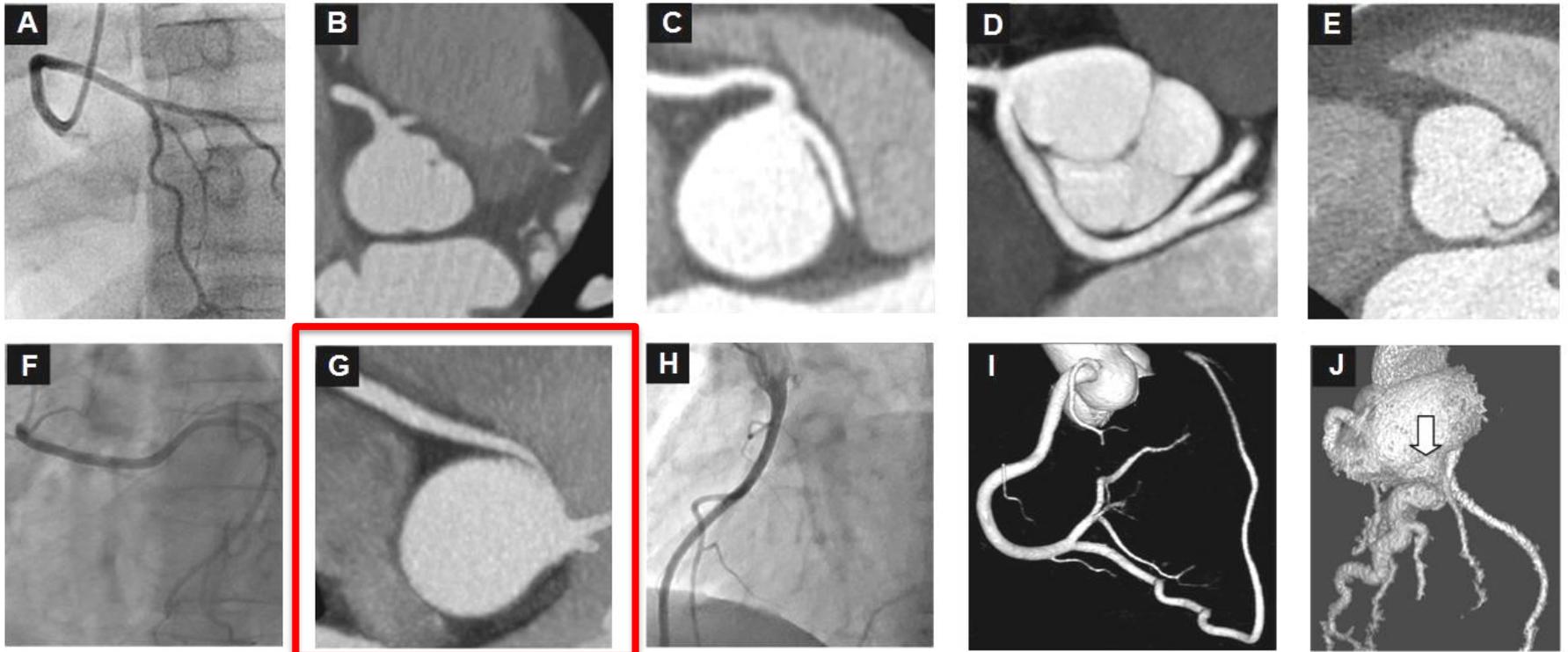
Intervenant : Xavier Halna du Fretay, Saran

Je n'ai pas de lien d'intérêt à déclarer

connexions anormales des coronaires



connexions anormales des coronaires



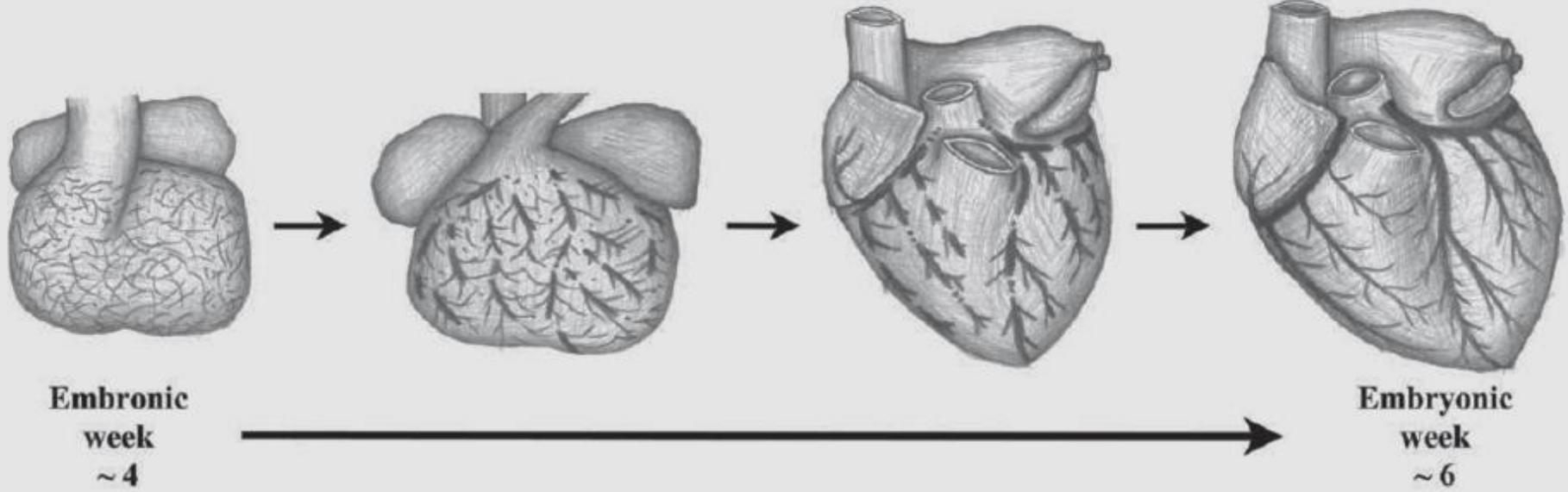
droite

objectifs pédagogiques

- Modes de présentation
- Outils diagnostiques
- Techniques de cathétérisme
- Bilan complémentaire
- Evaluation du risque
- Options thérapeutiques

Anatomie

embryologie



Development of coronary vessels during embryogenesis.

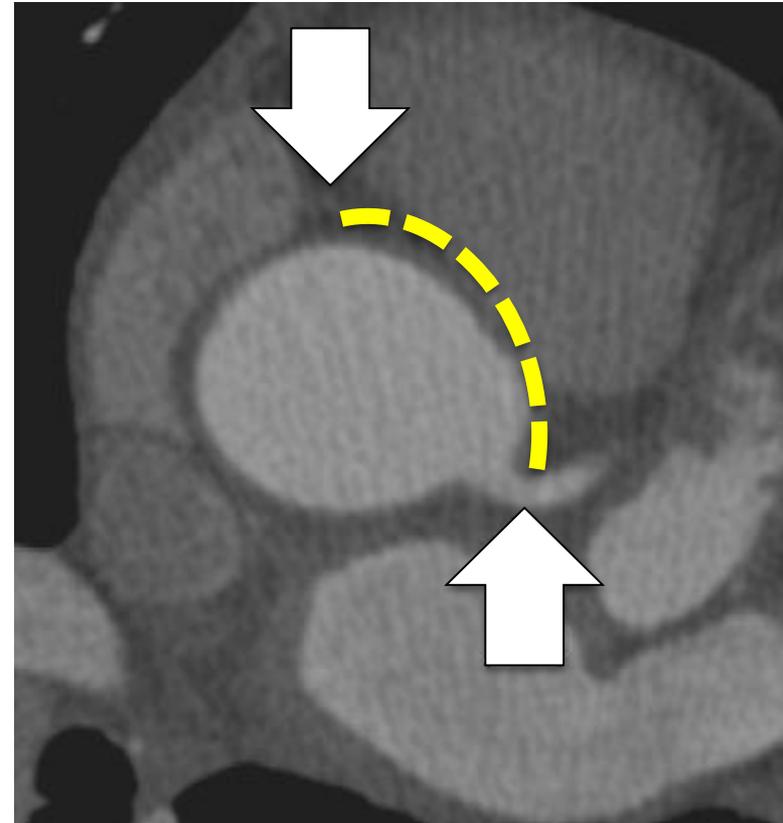
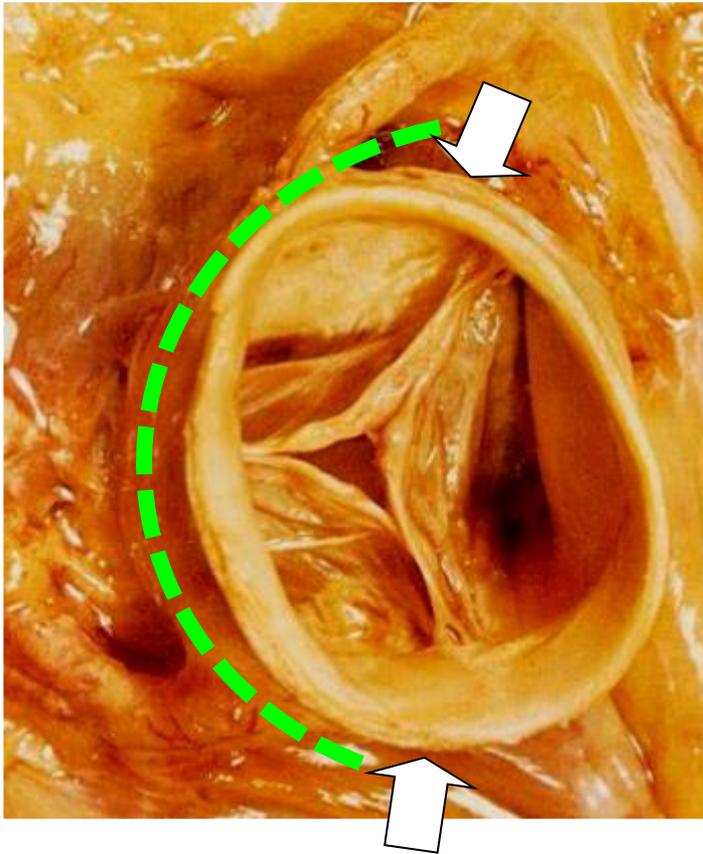
Lluri G. *Clin Cardiol* 2014
Bogers AJ. *Anat Embryol* 1989

Développement de l'aorte des coronaires
Anomalie d'origine

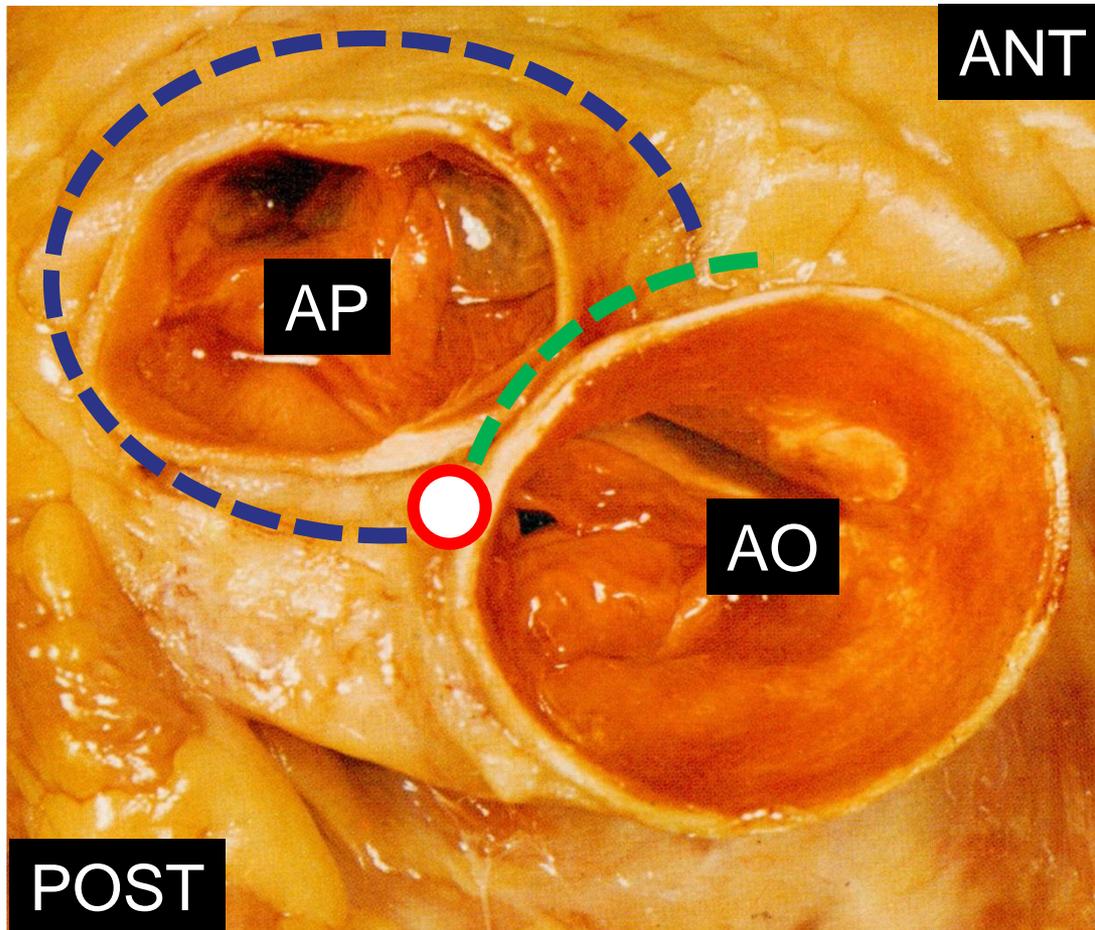
Développement vers l'aorte des coronaires
Anomalie de connexion

sites habituels des connexions ectopiques droites

antérieur



trajets des coronaires droites ectopiques



- ostium droit ectopique
- pré-pulmonaire
- pré-aortique

sites habituels de connexion ectopique droite

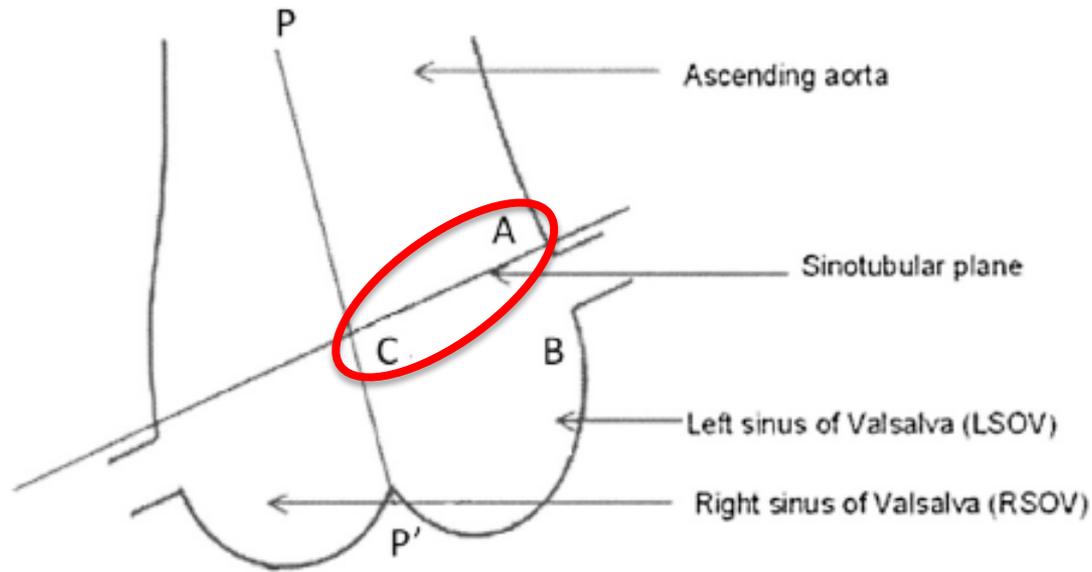
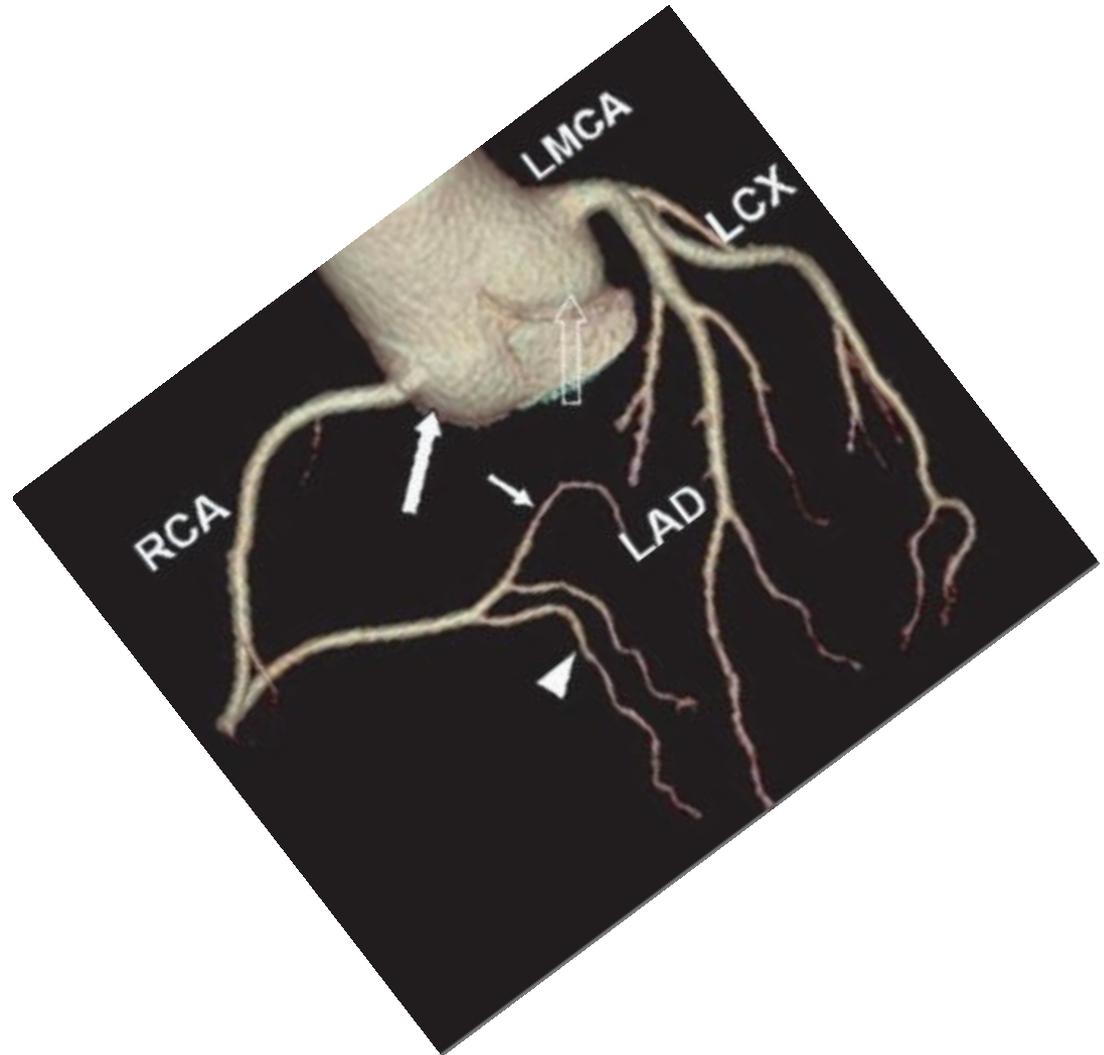
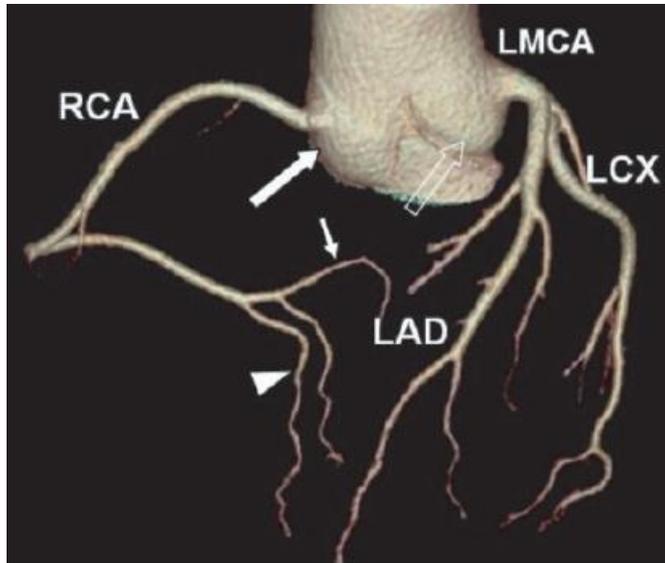
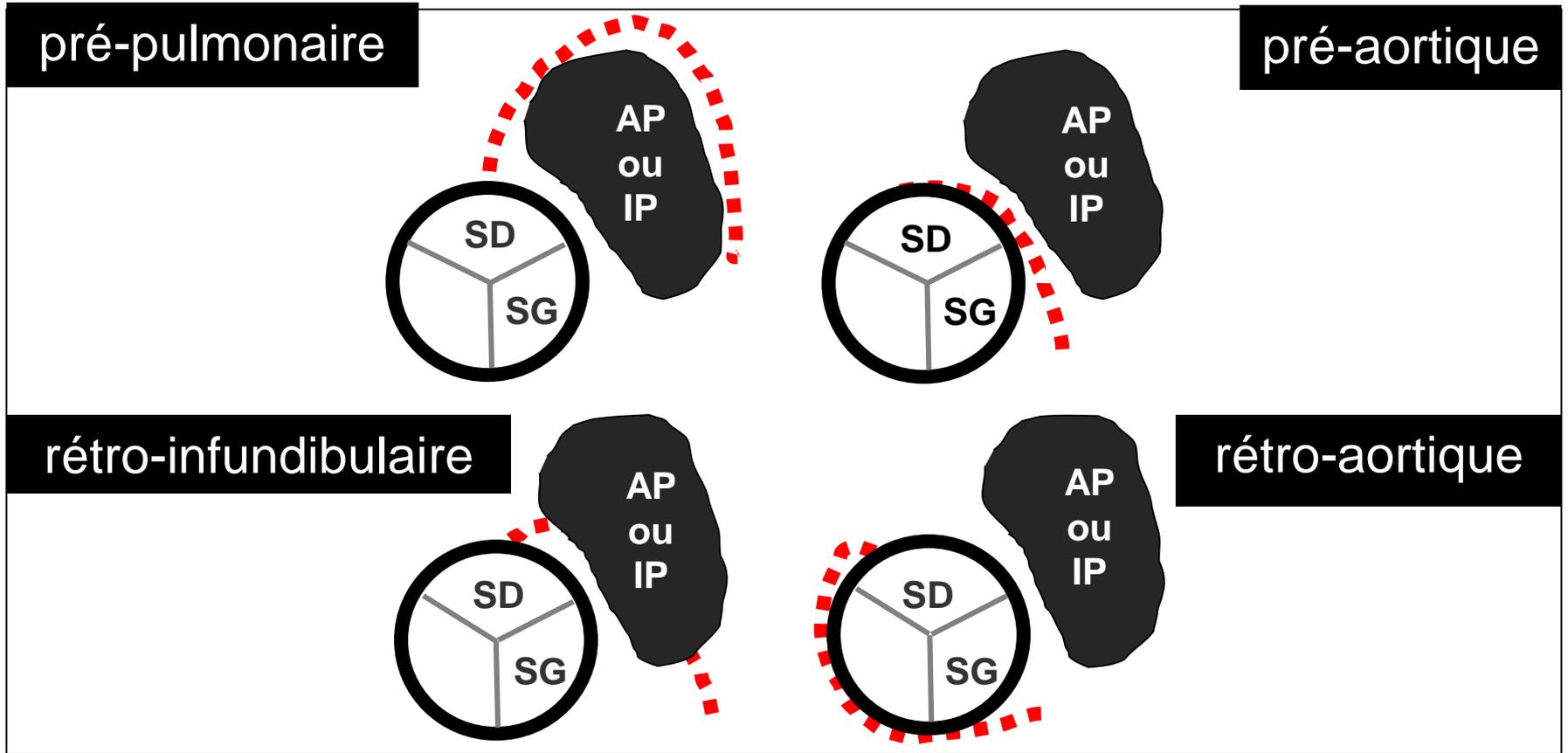


Fig. 1 – Origin of anomalous RCA from LSOV.
Representative diagram of aortic root and sinuses in LAO projection. P–P' indicates a hypothetical plane running through the midline. Sites A through C represents common sites for the origin of the anomalous artery.

INDIAN HEART JOURNAL 66 (2014) 430–434

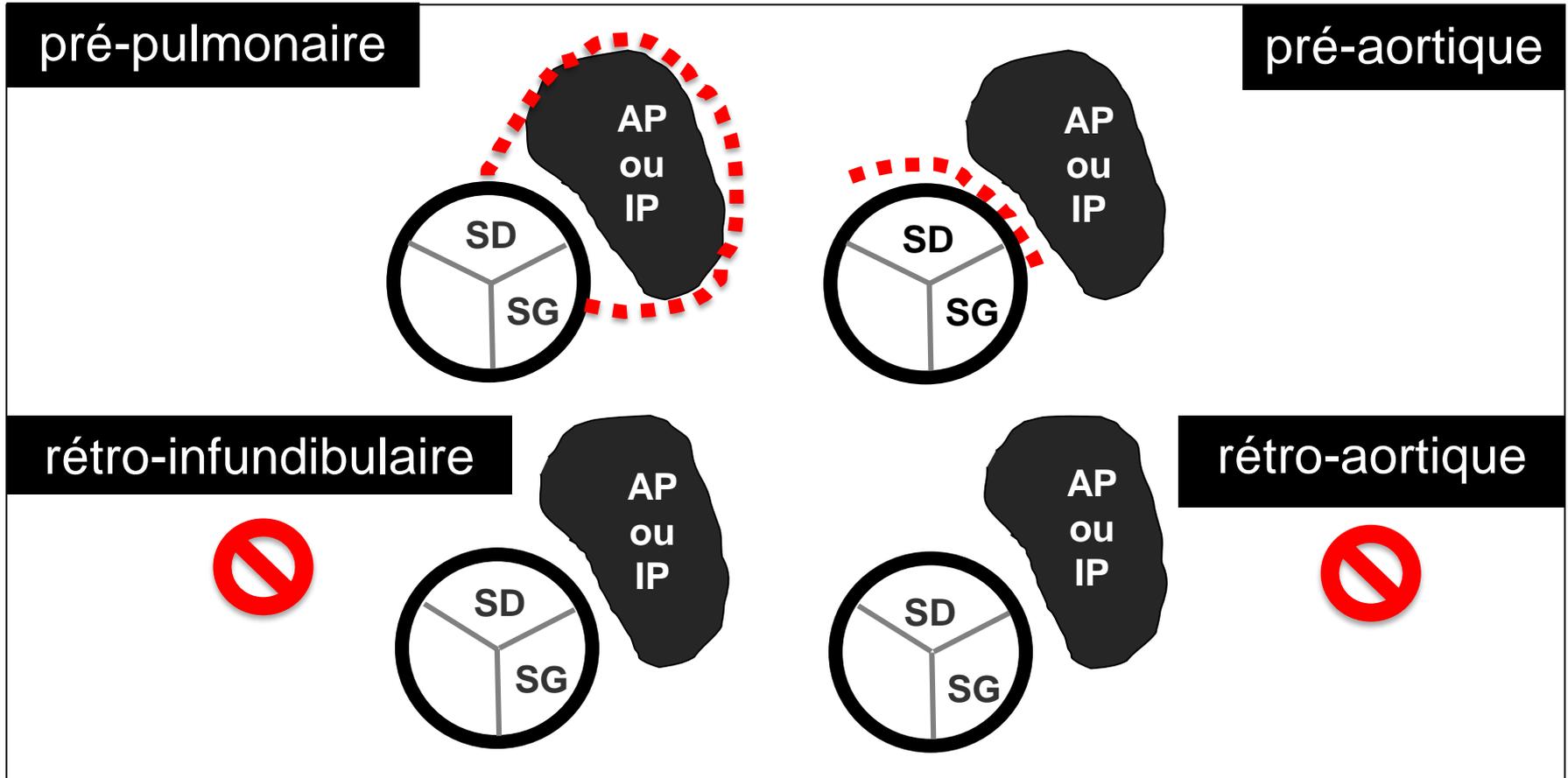


tronc commun connecté dans sinus droit trajets possibles



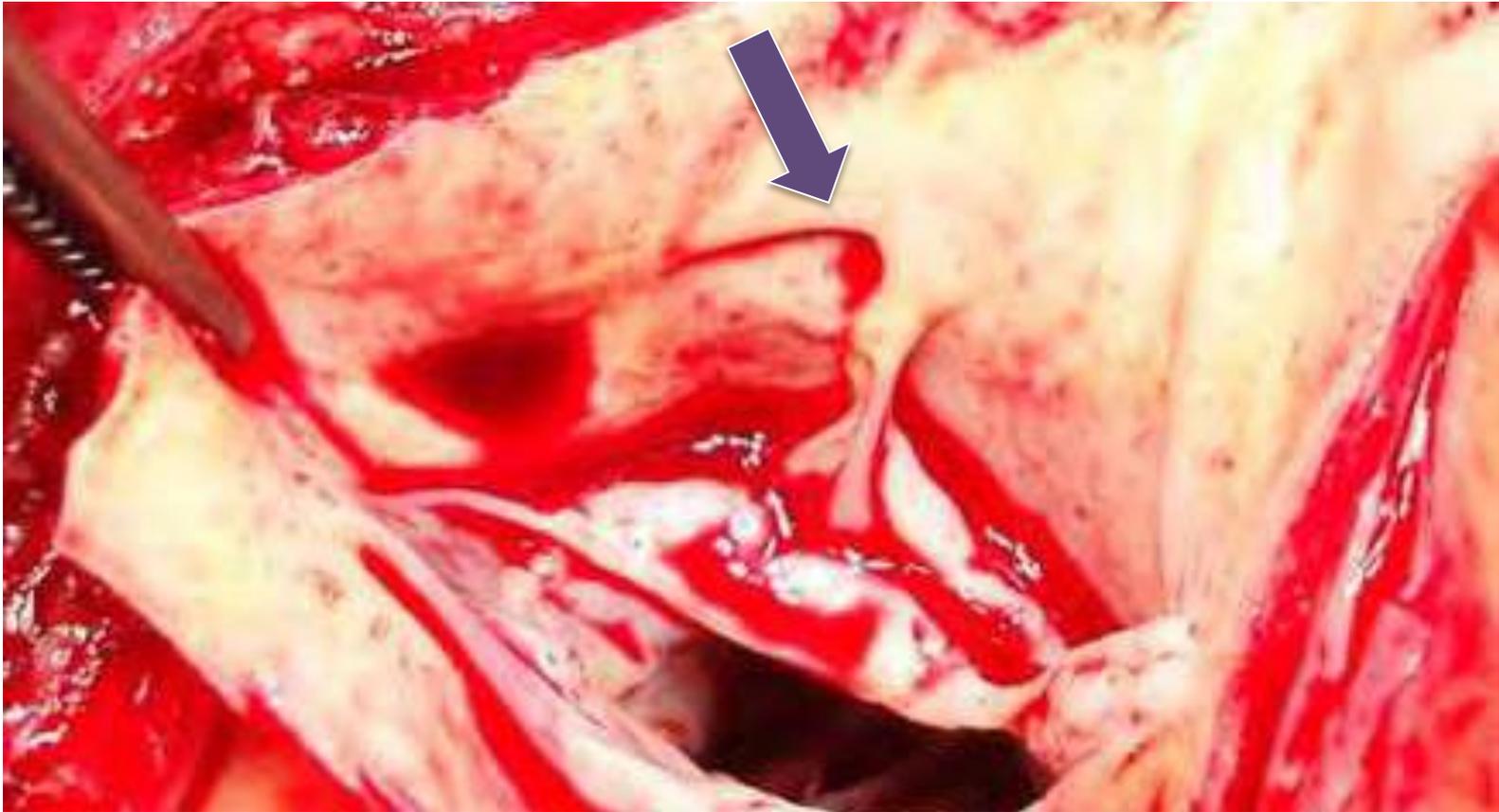
AP: artère pulmonaire, IP: infundibulum pulmonaire, SD: sinus droit, SG: sinus gauche

coronaire droite connectée dans sinus gauche trajets possibles

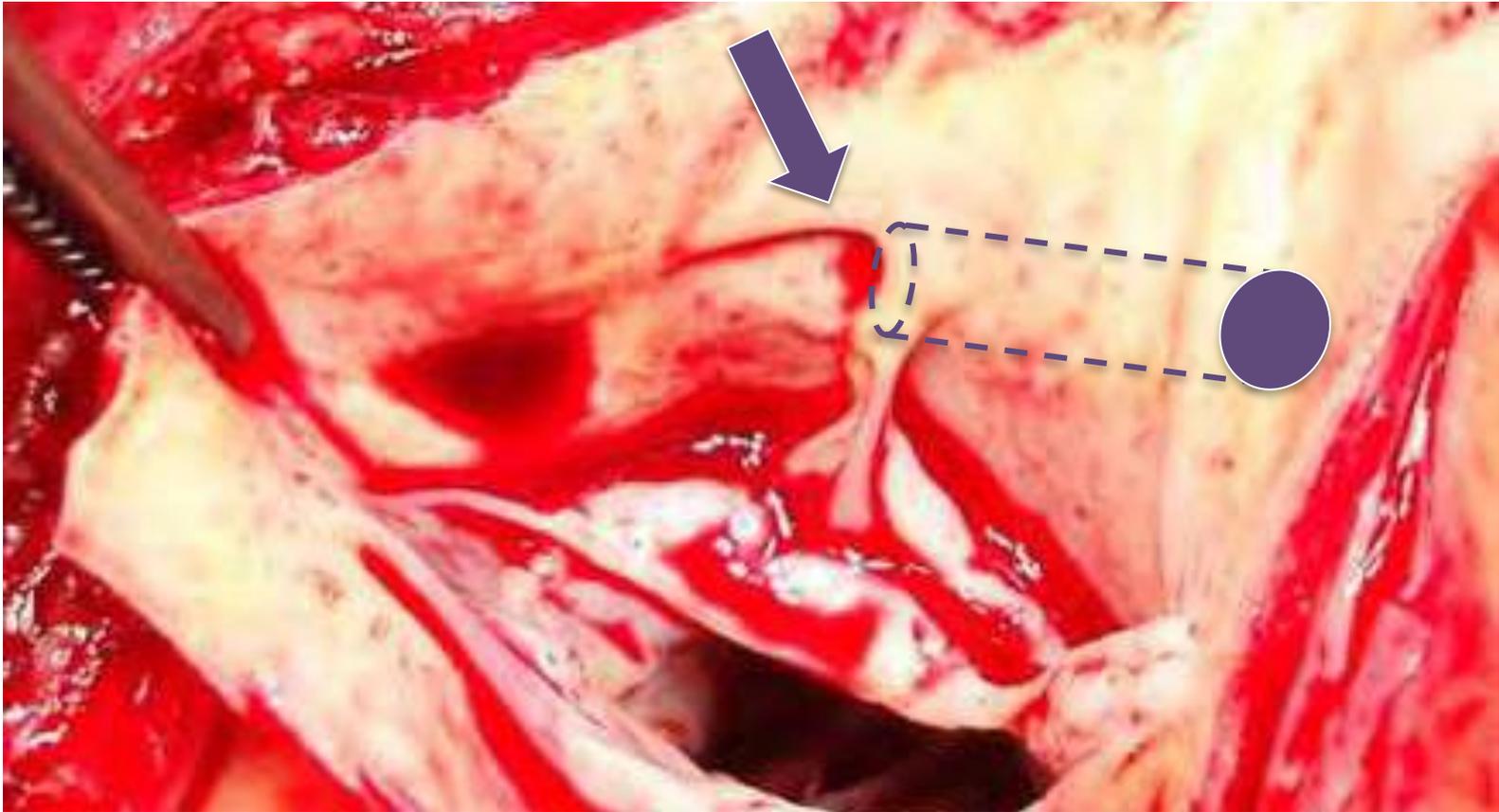


AP: artère pulmonaire, IP: infundibulum pulmonaire, SD: sinus droit, SG: sinus gauche

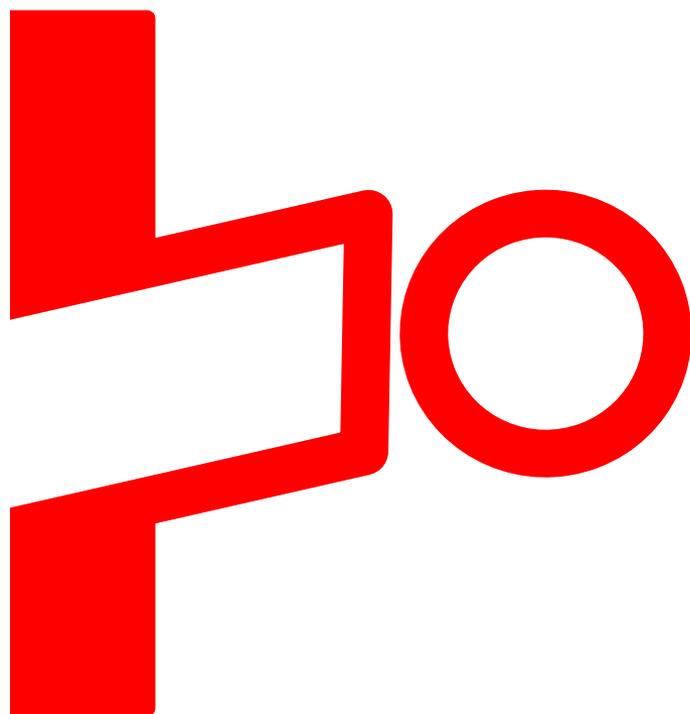
connexion ectopique droite dans sinus gauche



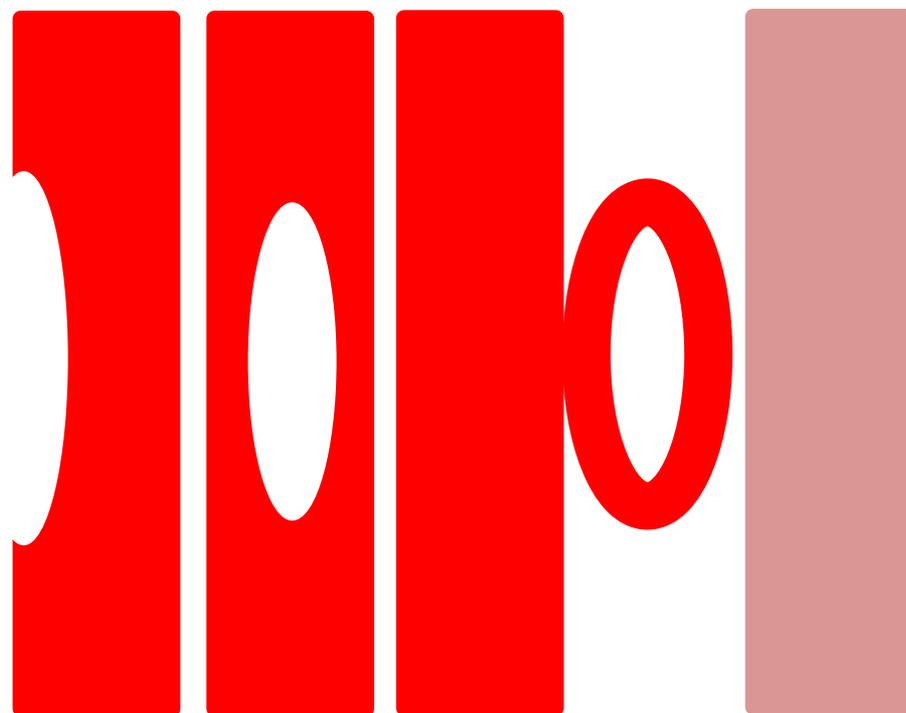
connexion ectopique droite dans sinus gauche



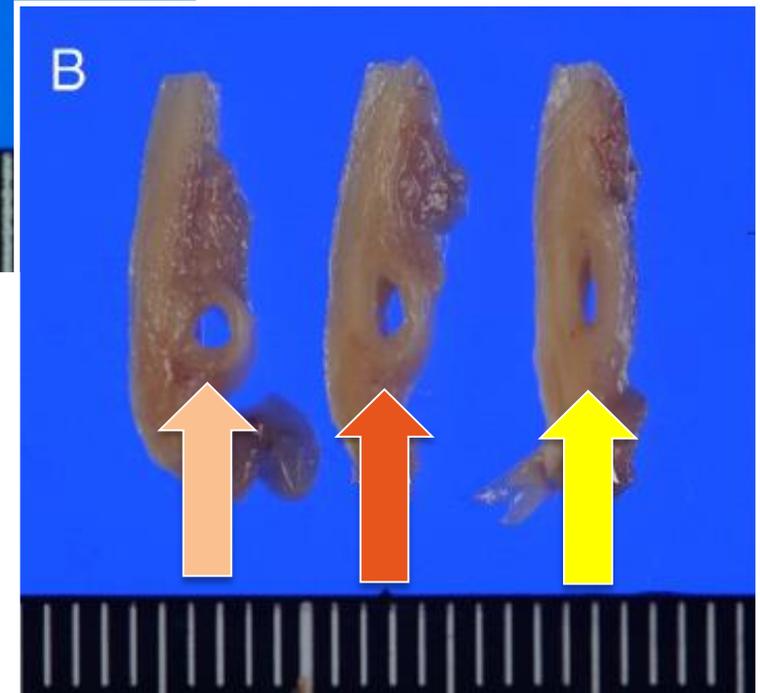
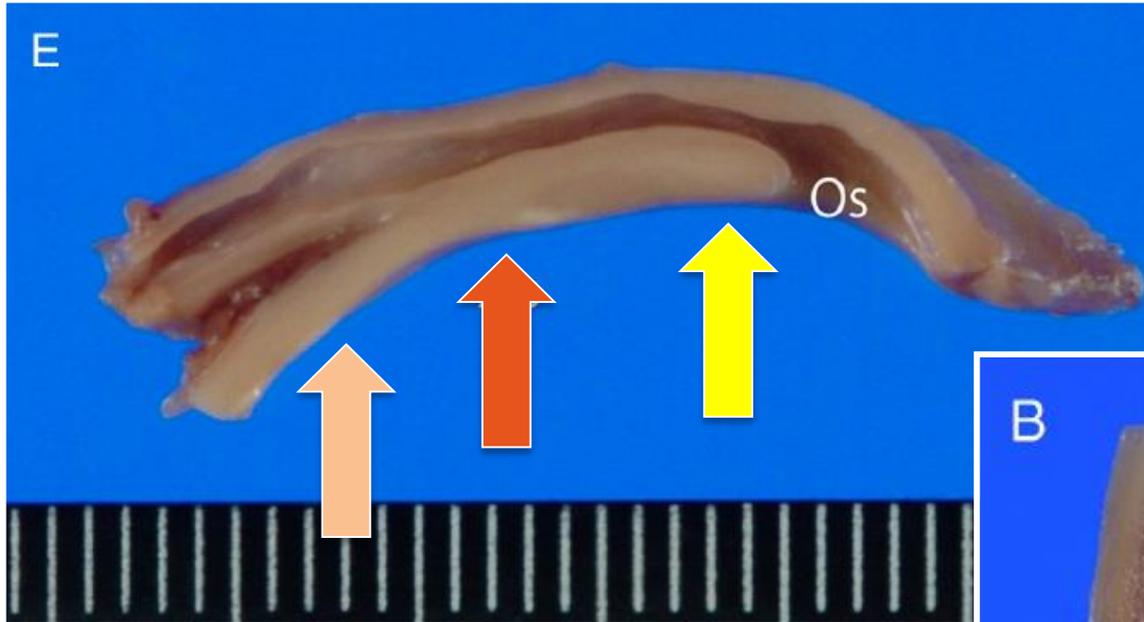
connexion extramurale



connexion intramurale

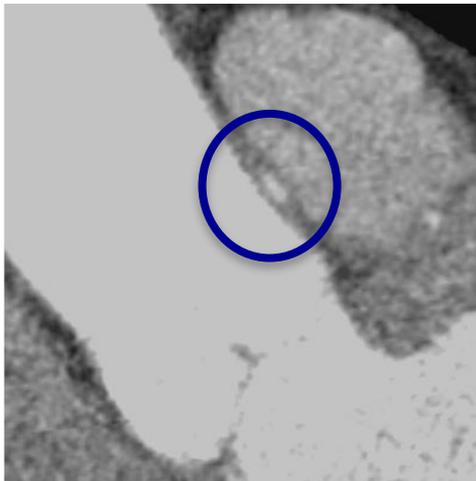
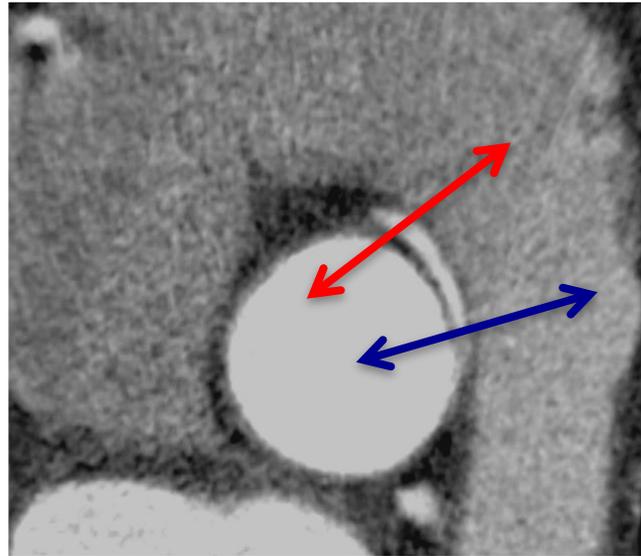


ANOCOR droite avec passage intramural

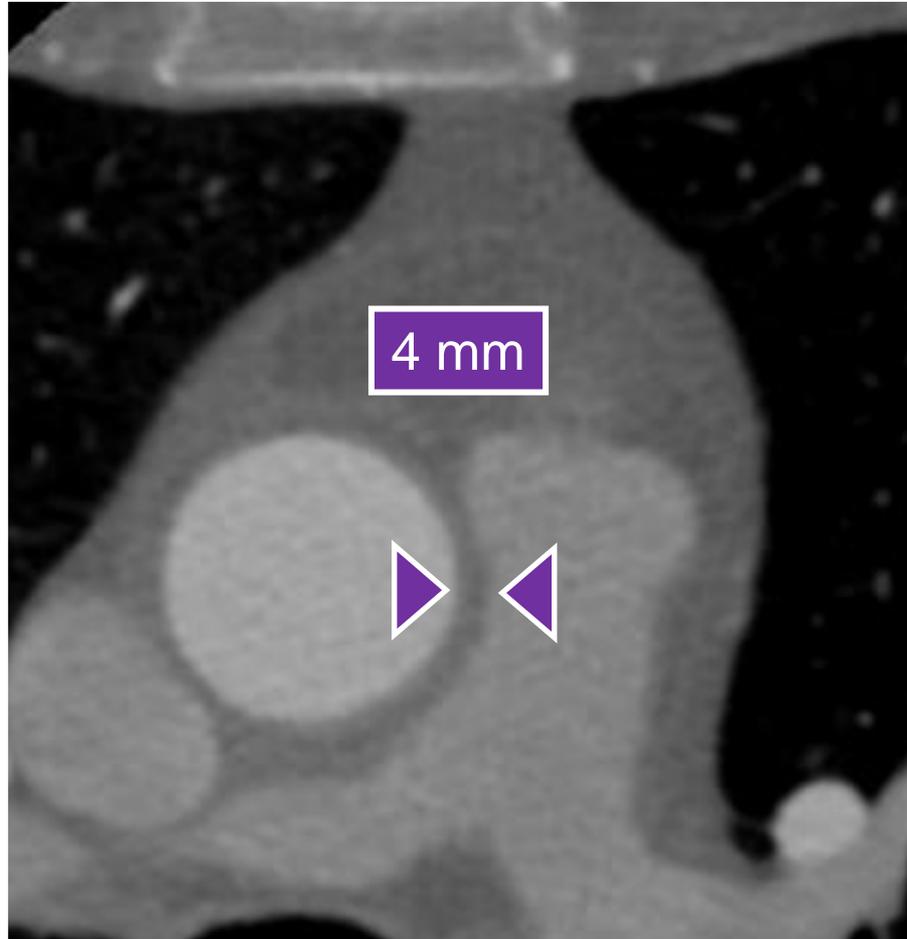


Hata Y et al. Cardiovasc Pathol 2014

ANOCOR droite avec passage intramural

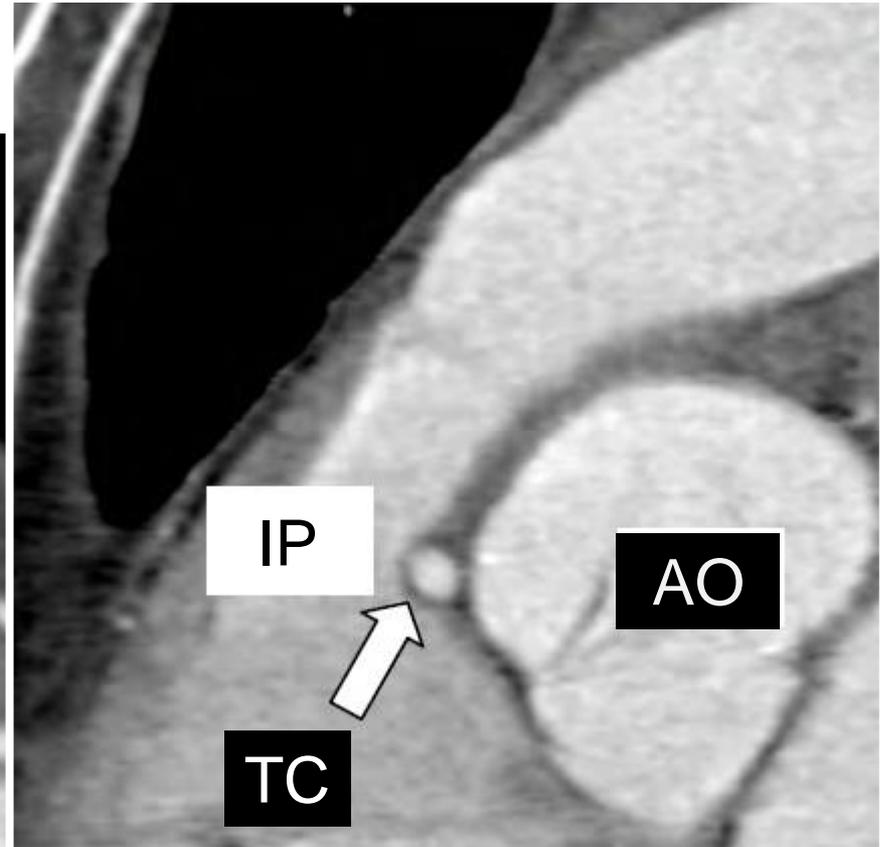
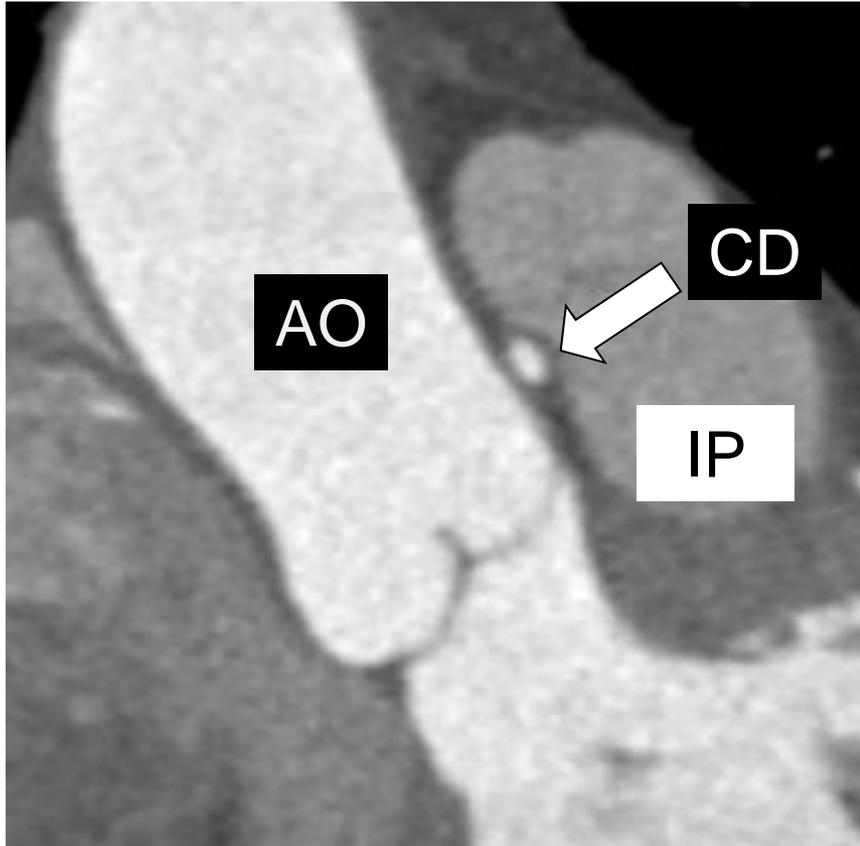


espace interartériel (aorte-artère pulmonaire)



anomalies de connexion proximale coronaire

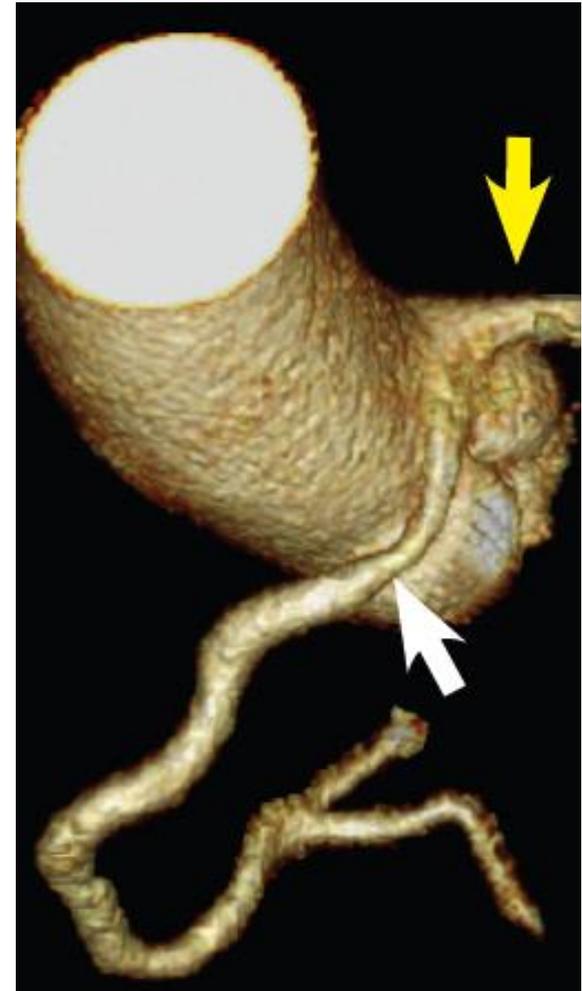
trajet préaortique



trajet rétroinfundibulaire

artère coronaire droite ectopique

- Connexions :
 - sinus gauche +++
 - artère controlatérale (tronc/IVA)
 - aorte ascendante
 - sinus postérieur
 - coronaire unique
- Trajets :
 - pré aortique +++
 - prépulmonaire
 - rétroaortique
 - normal



Modes de présentation

prévalence des connexions anormales en imagerie

≈1%

- échocardiographie (enfant) 2/1000 (0.2%)
- coronarographie 8/1000 (0.8%)
- scanner coronaire 12/1000 (1.2%)

mode de présentation

absence de symptôme	possible
douleur thoracique	possible
dyspnée	rare
palpitations	possible
lipothymie	possible
syncope	possible
SCA ST -	rare*
SCA ST +	très rare*
mort subite	rare

* en l'absence de maladie coronaire associée

mort subite et connexion anormale coronaire

- premier événement cardiovasculaire : **souvent**
- population en bonne santé et jeune : **généralement**
- lien avec une activité physique/sportive : **net**

Table 1. Causes of Sudden Death in 387 Young Athletes*

Cause	No. of Athletes	Percent
Hypertrophic cardiomyopathy	102	26.4
Commotio cordis	77	19.9
Coronary artery anomalies	53	13.7
Left ventricular hypertrophy of indeterminate causation†	29	7.5
Myocarditis	20	5.2
Ruptured aortic aneurysm (Marfan syndrome)	12	3.1
Arrhythmogenic right ventricular cardiomyopathy	11	2.8
Tunneled (bridged) coronary artery‡	11	2.8
Aortic valve stenosis	10	2.6
Atherosclerotic coronary artery disease	10	2.6
Dilated cardiomyopathy	9	2.3
Myxomatous mitral valve degeneration	9	2.3
Asthma (or other pulmonary condition)	8	2.1
Heat stroke	6	1.6
Drug abuse	4	1.0
Other cardiovascular cause	4	1.0
Long QT syndrome§	3	0.8
Cardiac sarcoidosis	3	0.8
Trauma causing structural cardiac injury	3	0.8
Ruptured cerebral artery	3	0.8

Maron B. J Am Coll Cardiol 2005

mort subite récupérée

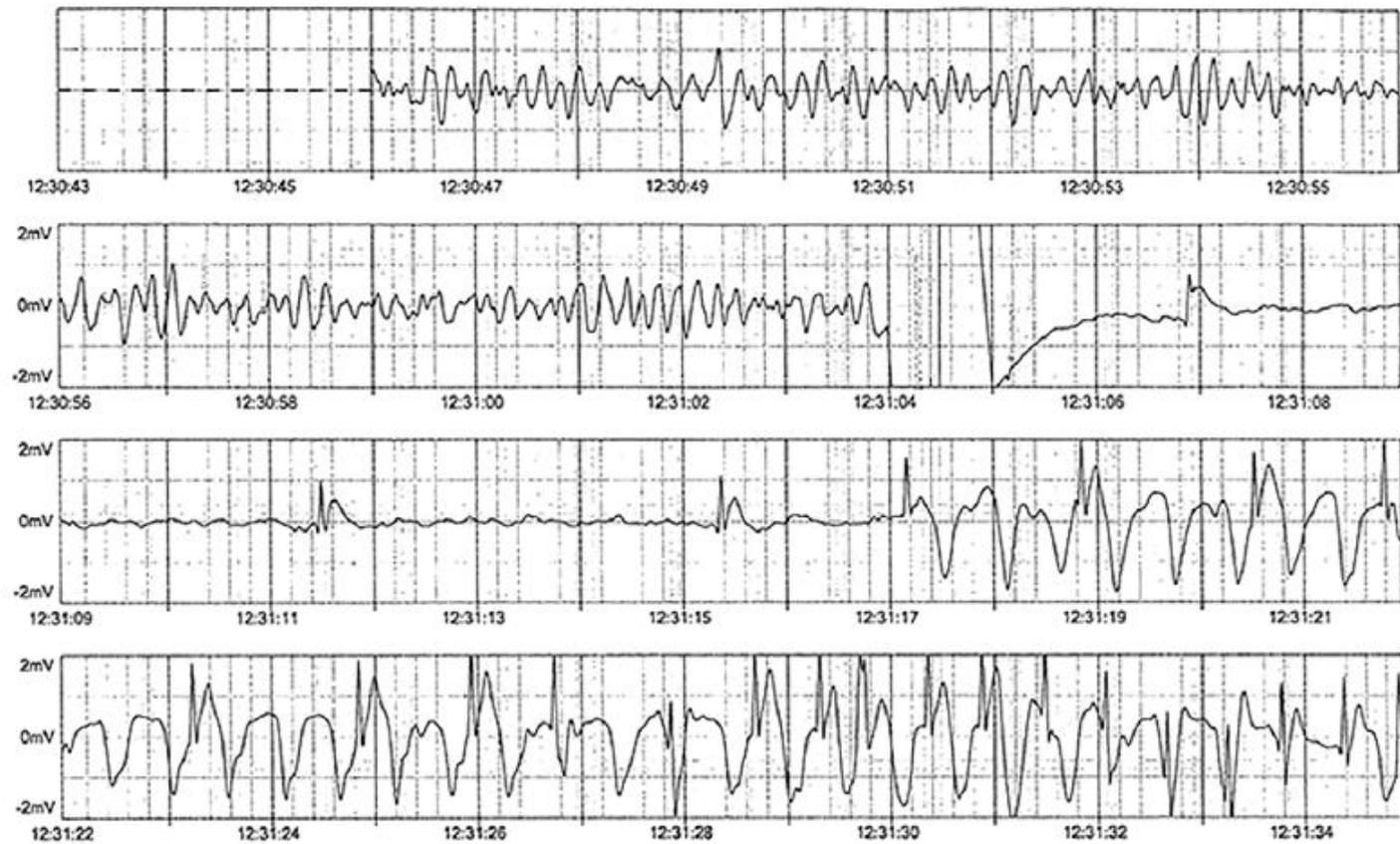


Figure 1. ECG recording from an automated external defibrillator

Shimizu T et al. Intern Med 2014

mécanismes de la fibrillation ventriculaire

- ischémie myocardique ?
- zones de fibrose myocardique ?
- seuil arythmogène bas ?
- hypotension post-effort ?
- association de plusieurs mécanismes ?
- autre mécanisme ?

anomalies de connexion proximale des artères coronaires

répartition selon l'artère coronaire

100 ANOCOR*

artère coronaire	%
tronc commun	12.0
artère interventriculaire antérieure	5.5
artère circonflexe	47.5
artère coronaire droite	33.0
autres artères	2.0

* à partir des données du registre ANOCOR (ESC 2015)



anomalies de connexion proximale des artères coronaires

répartition selon le trajet*

		%
coronaire droite	trajet préaortique	89.5
	autres trajets	10.5

* à partir des données du registre ANOCOR (ESC 2015)



Outils diagnostiques

QUAND DOIS-JE PENSER A UNE ANOCOR ?

- SYMPTOMATOLOGIE CARDIAQUE CHEZ PATIENT JEUNE
- PENDANT ACTIVITE SPORTIVE
- PAS DE FACTEURS DE RISQUE

- PENDANT LA CORONAROGRAPHIE
- JE NE TROUVE PAS FACILEMENT LA CORONAIRE DROITE

- PENDANT UN SCANNER

ACC/AHA Guideline

ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease

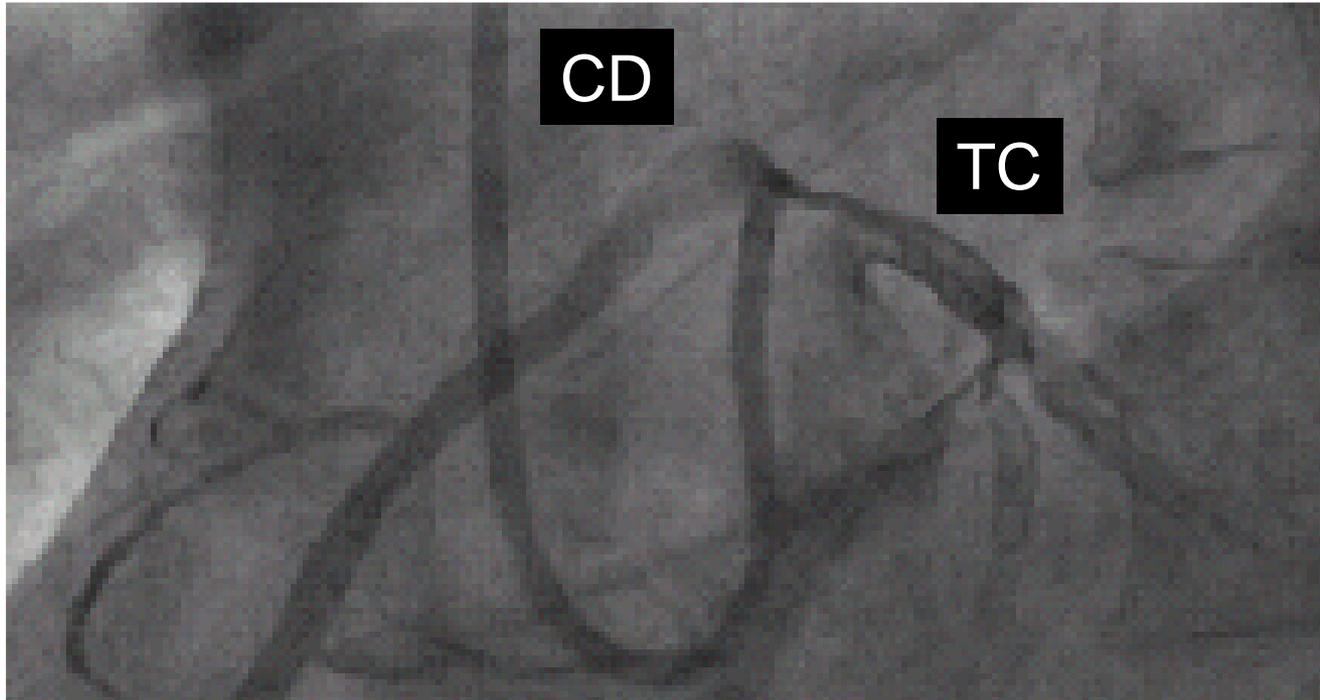
Circulation December 2, 2008

8.5. Recommendations for Congenital Coronary Anomalies of Ectopic Arterial Origin

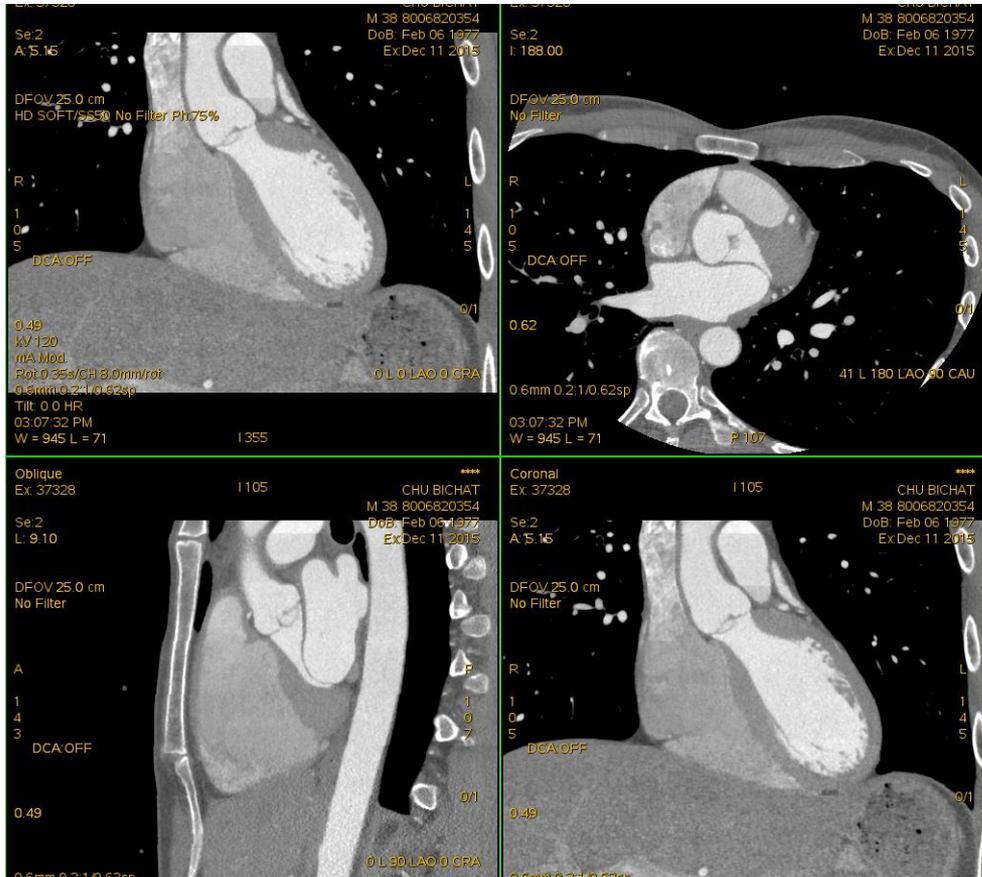
Class I

1. The evaluation of individuals who have survived unexplained aborted sudden cardiac death or with unexplained life-threatening arrhythmia, coronary ischemic symptoms, or LV dysfunction should include assessment of coronary artery origins and course. (*Level of Evidence: B*)
2. CT or magnetic resonance angiography is useful as the initial screening method in centers with expertise in such imaging. (*Level of Evidence: B*)

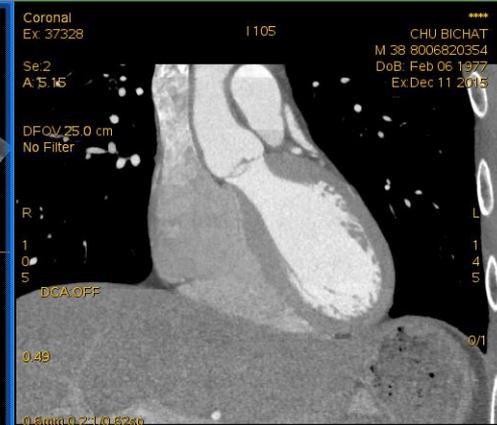
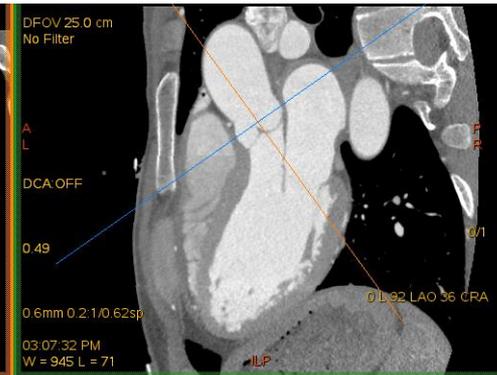
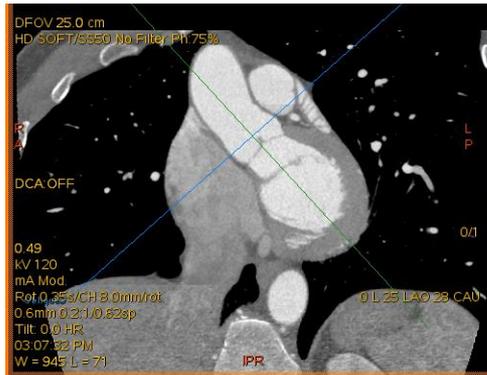
connexion droite dans le sinus controlatéral



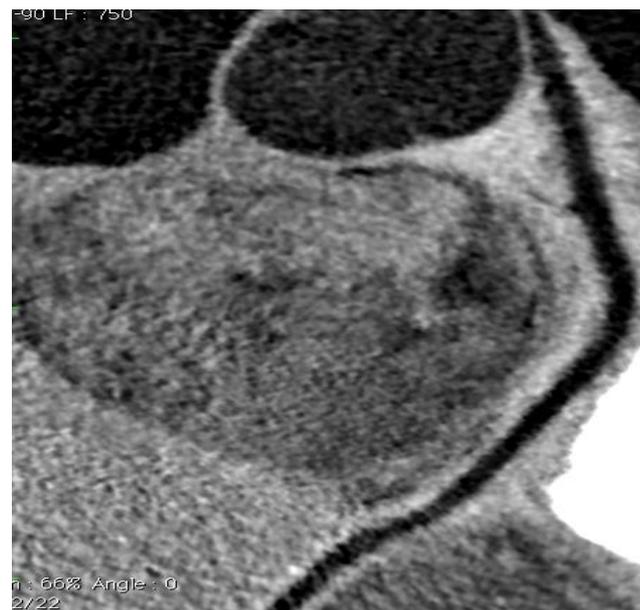
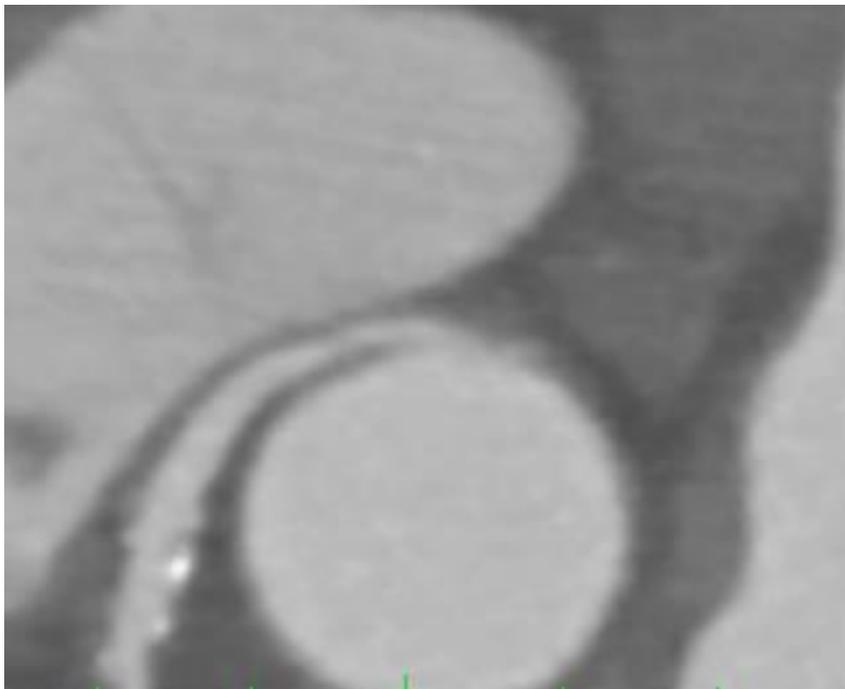
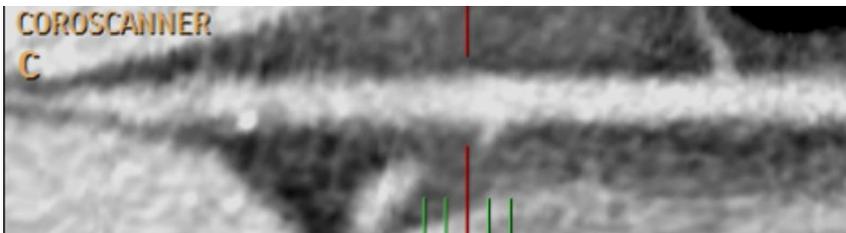
imagerie dans la plan axial strict



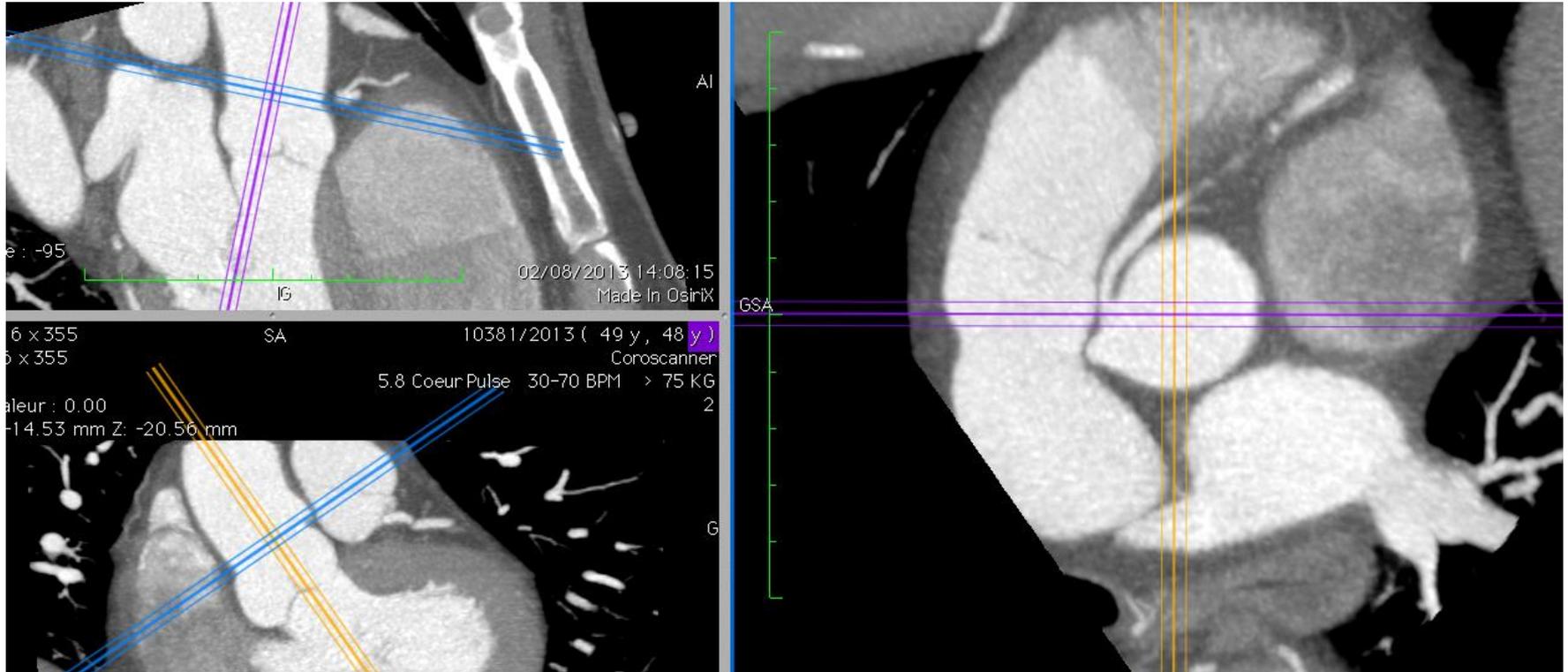
imagerie dans le plan de l'anneau aortique



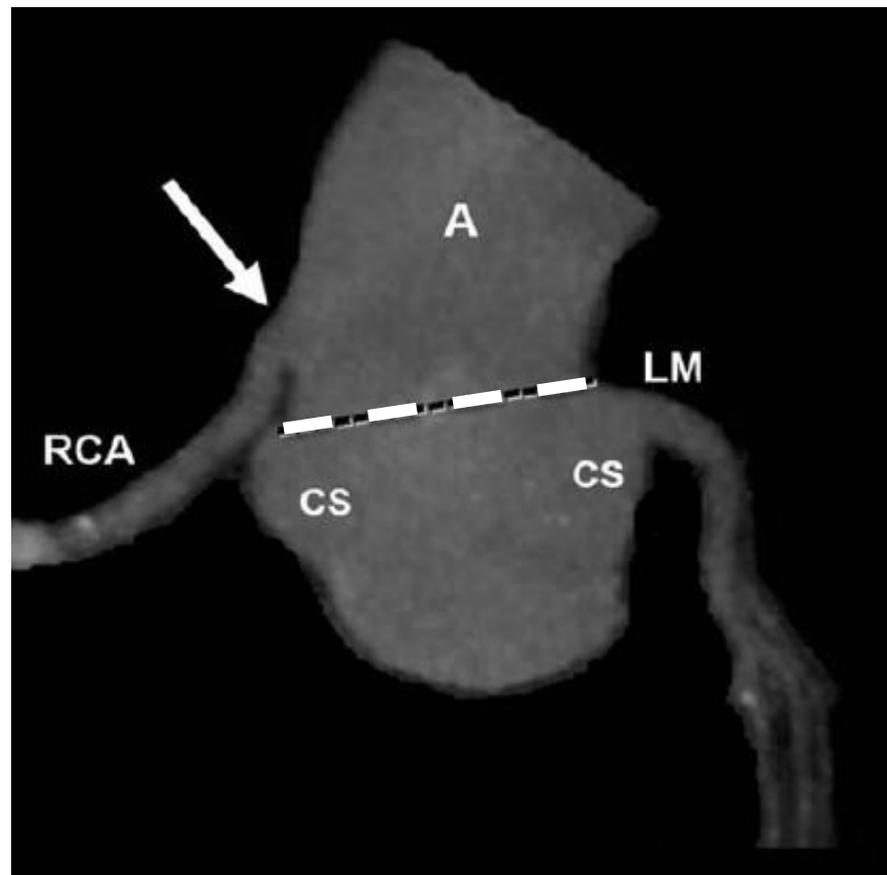
connexion droite dans le sinus controlatéral



connexion droite dans le sinus controlatéral

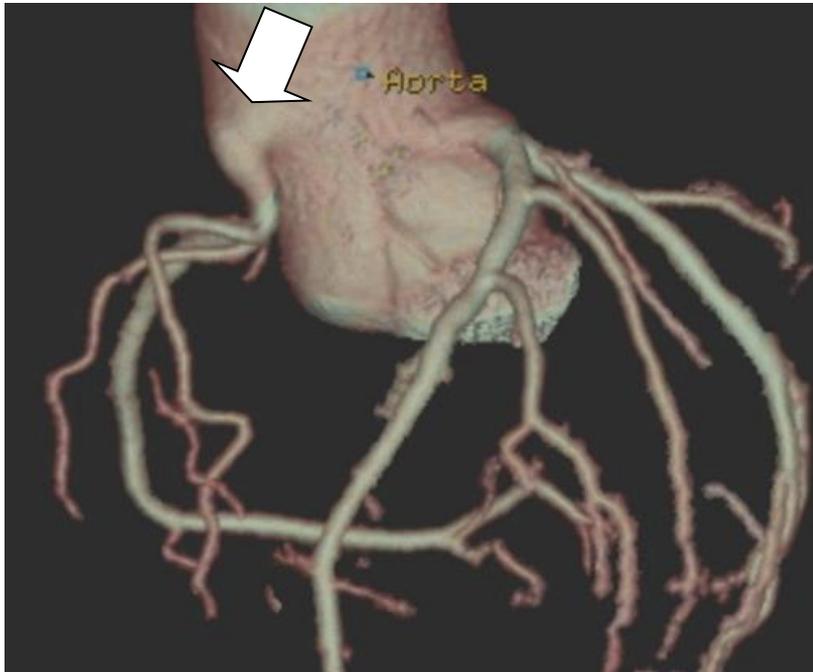


connexion aortique haute

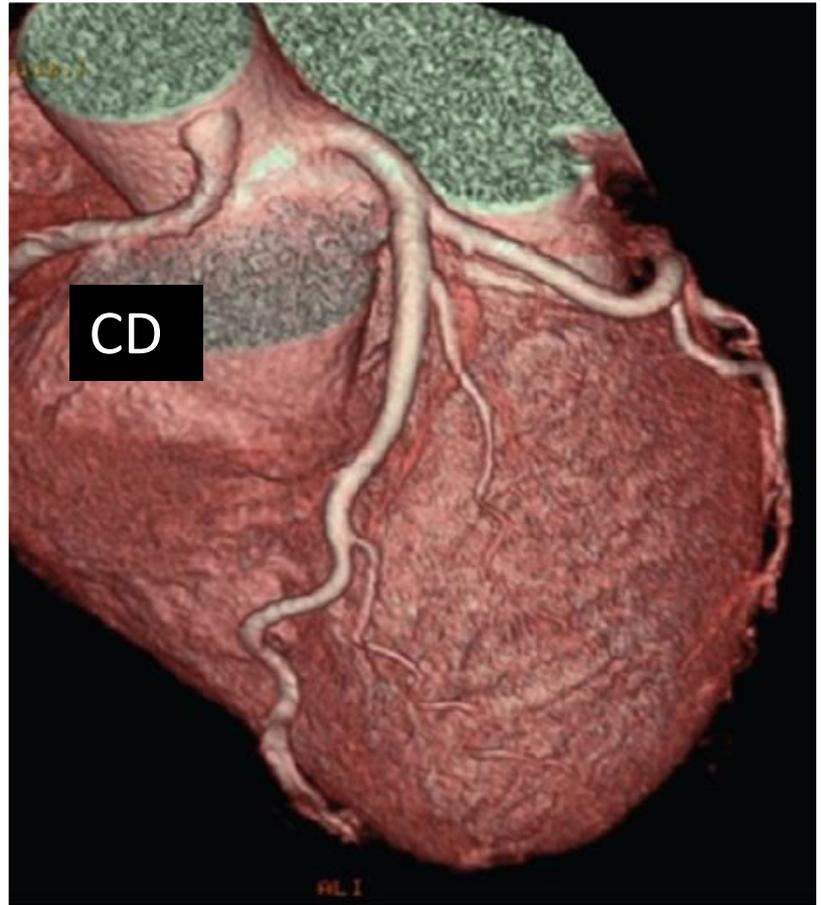
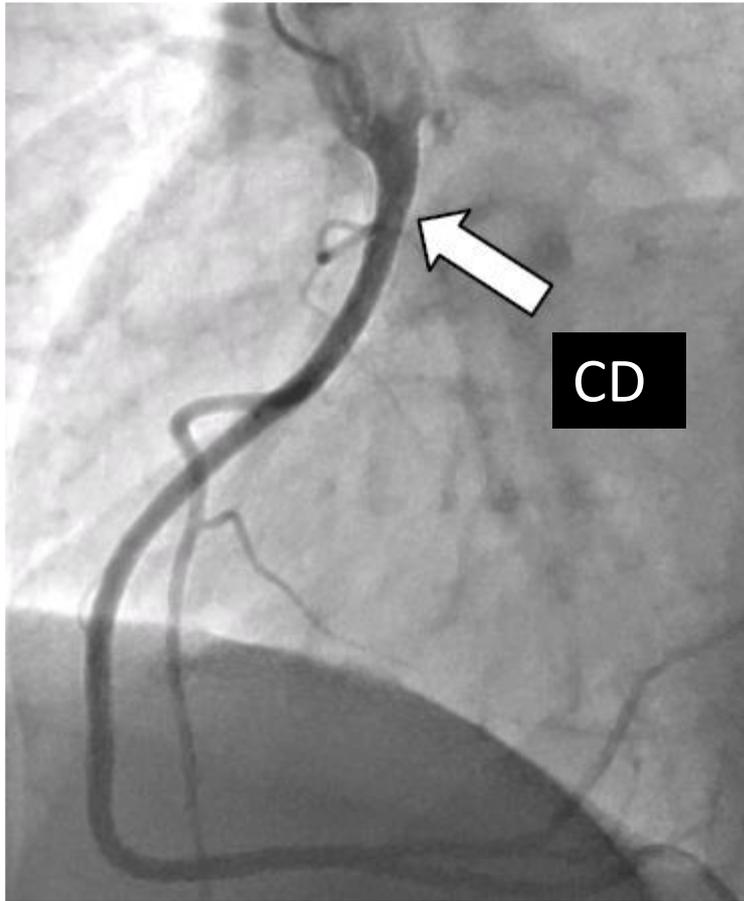


10 mm au dessus
jonction sinotubulaire

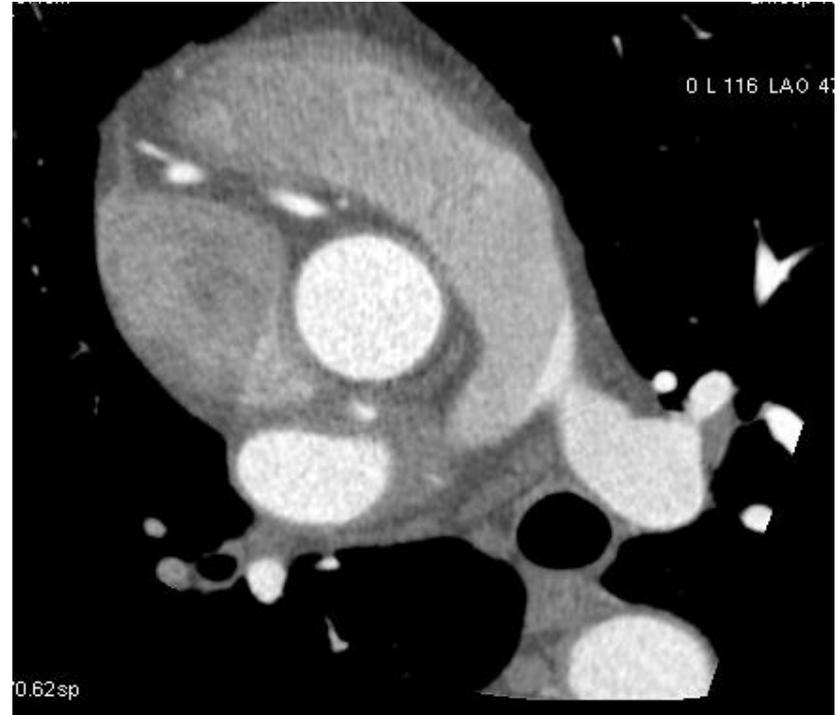
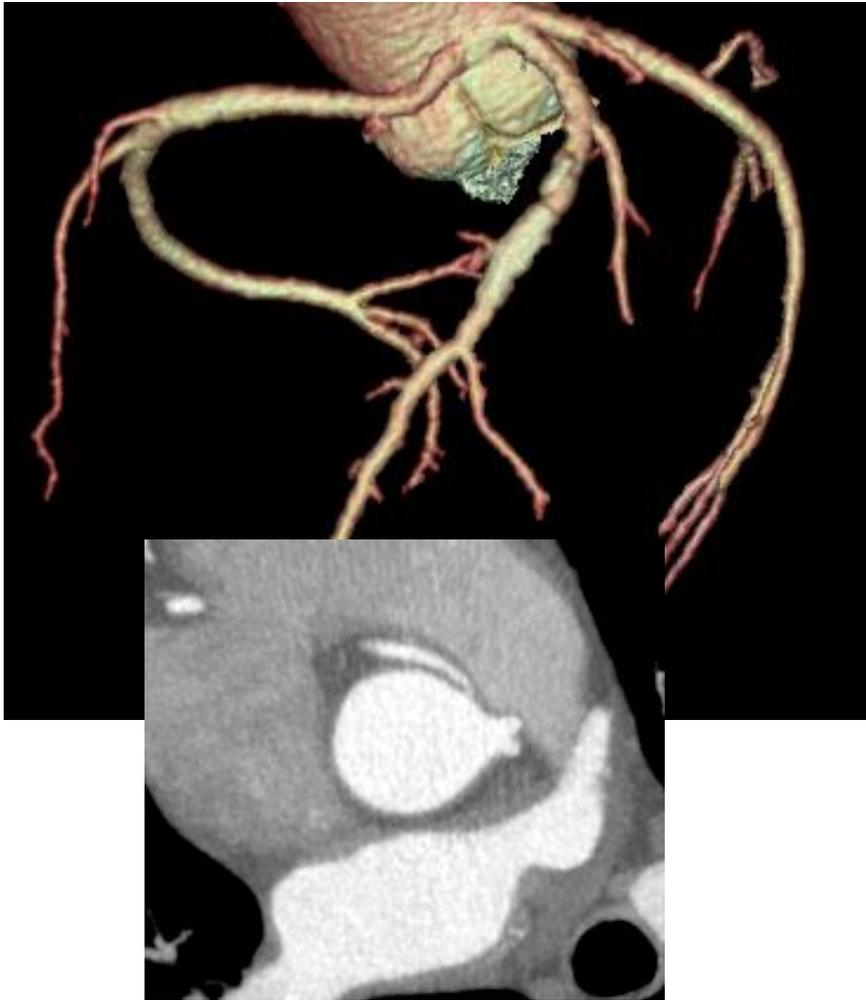
connexion aortique haute coronaire droite



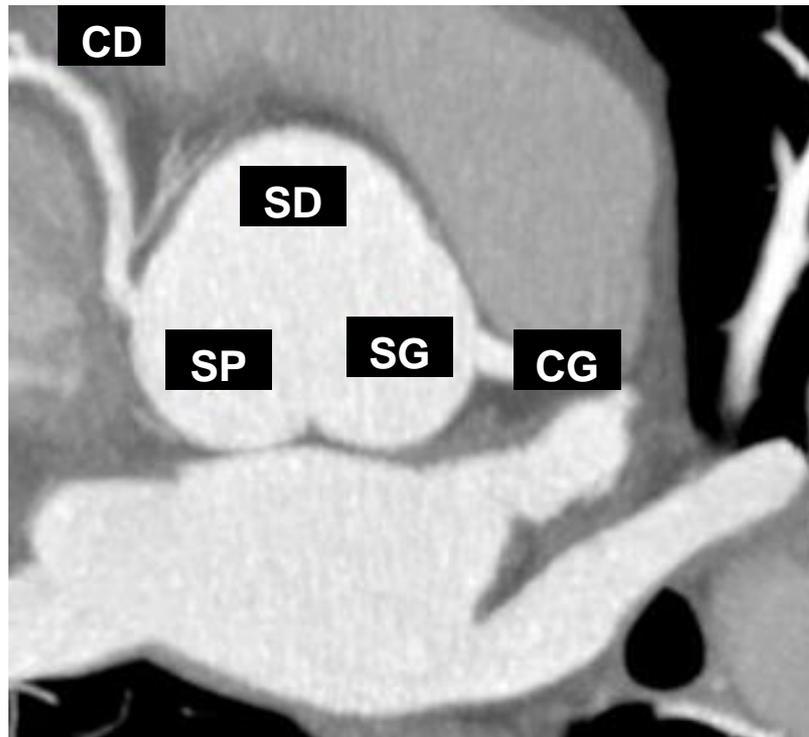
connexion aortique haute



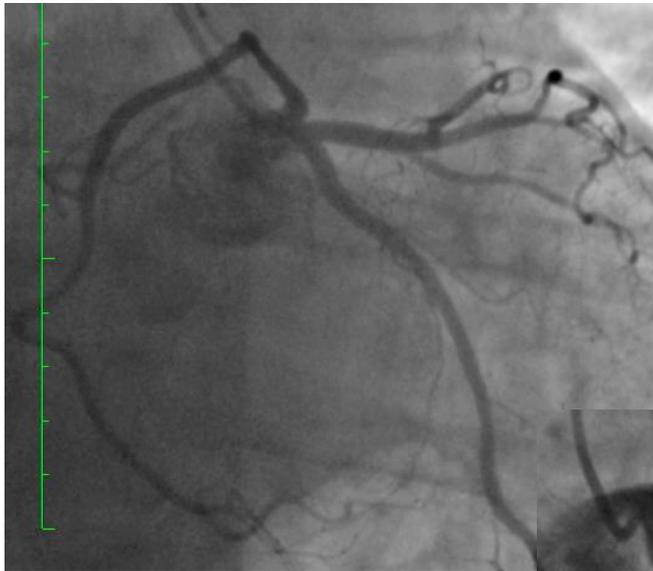
connexion droite dans le sinus controlatéral



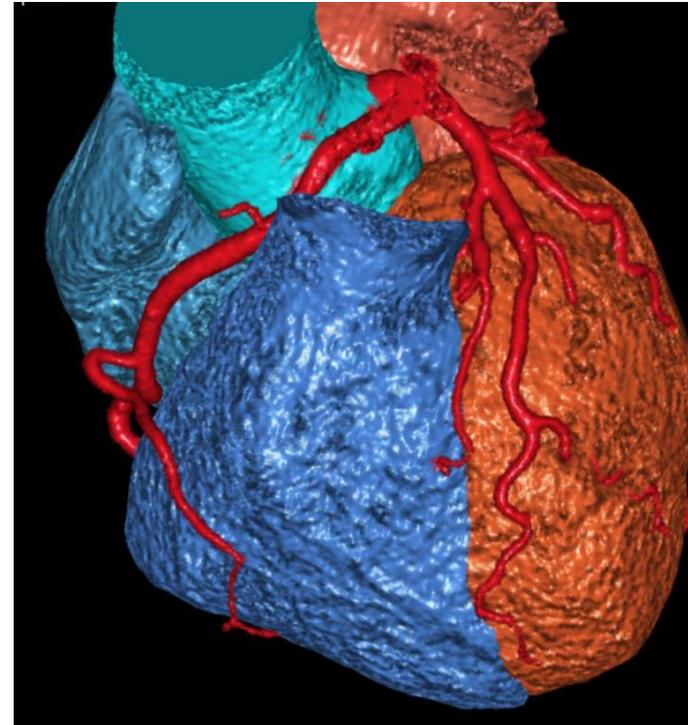
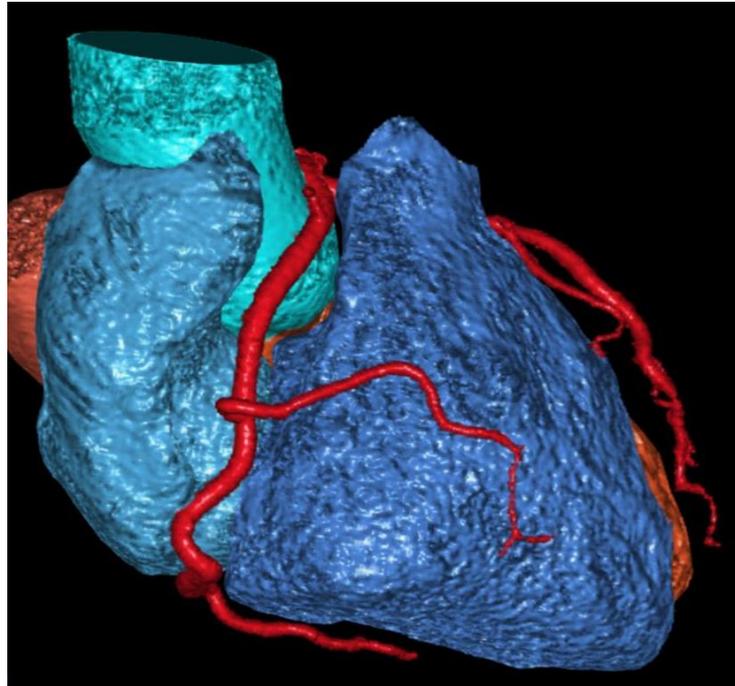
connexion droite dans le sinus postérieur



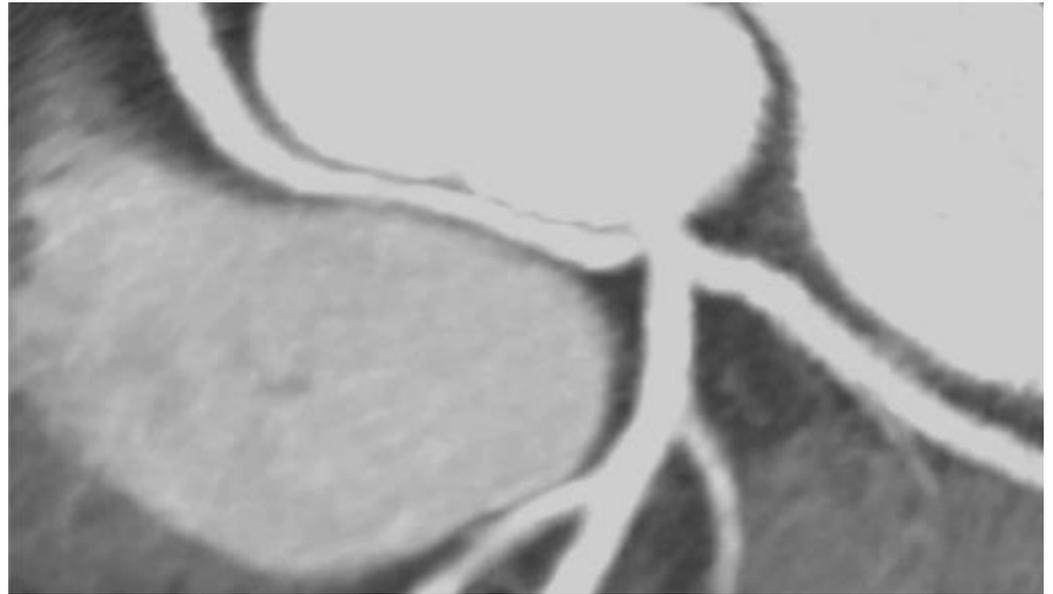
connexion droite dans tronc commun



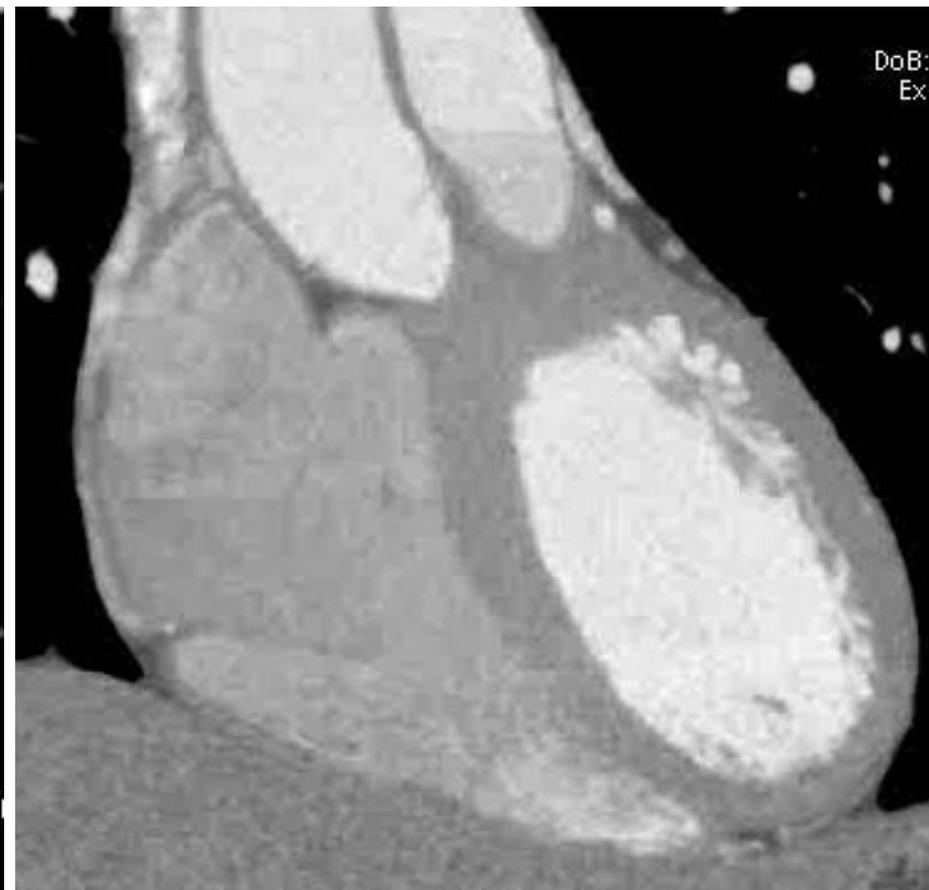
connexion droite dans tronc commun avec trajet préaortique



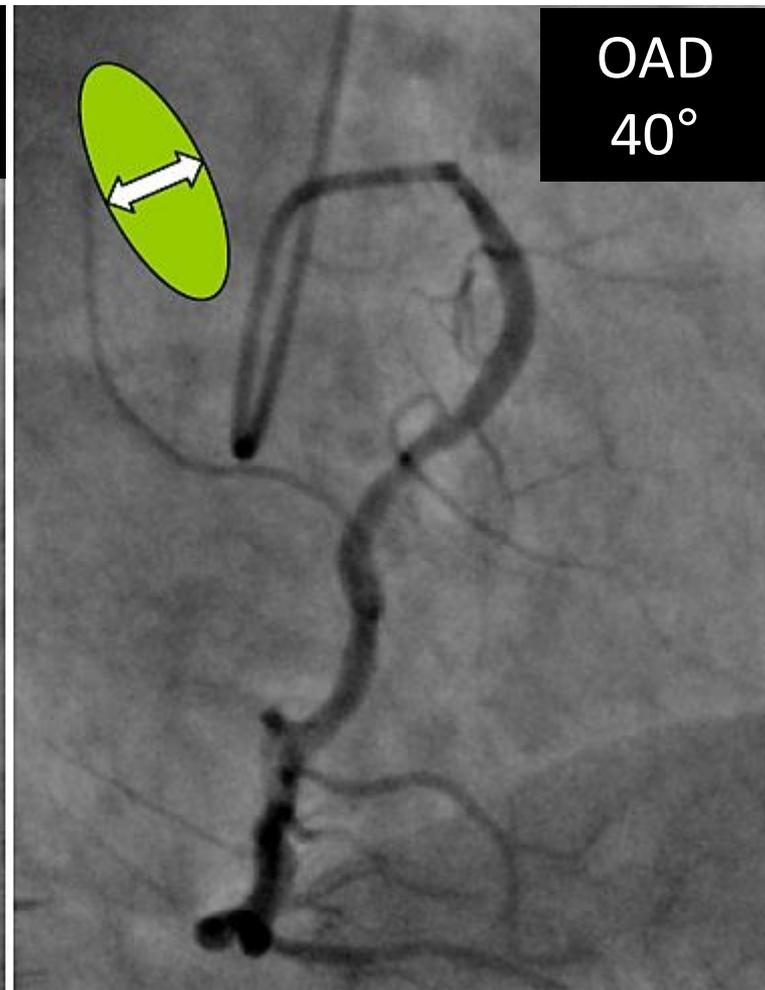
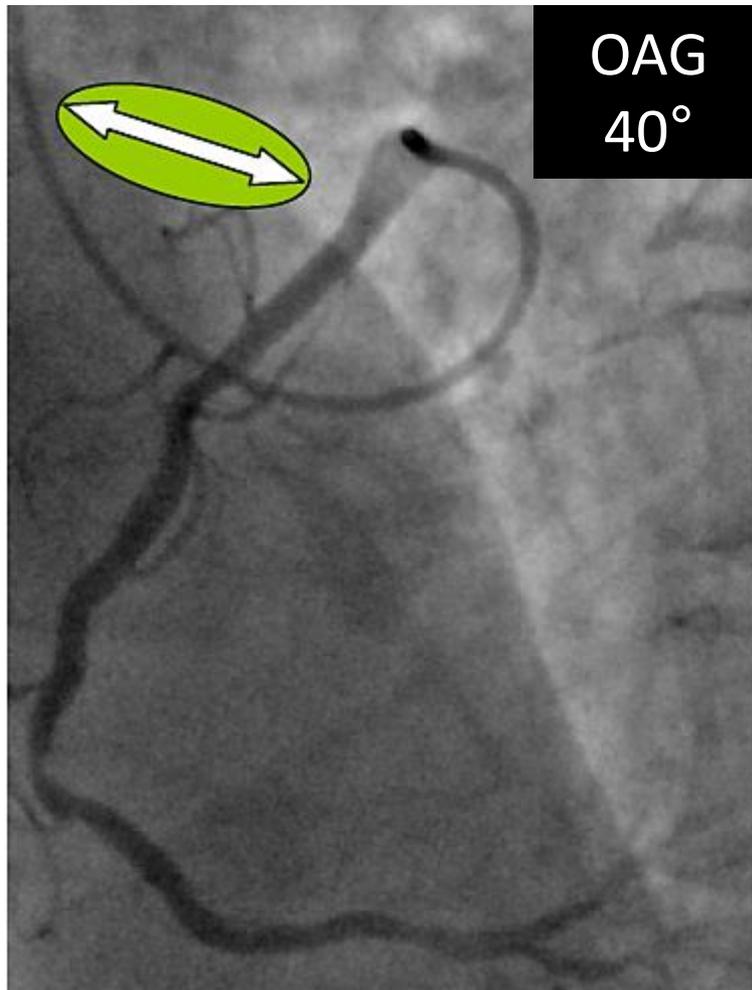
connexion droite dans tronc commun avec trajet préaortique



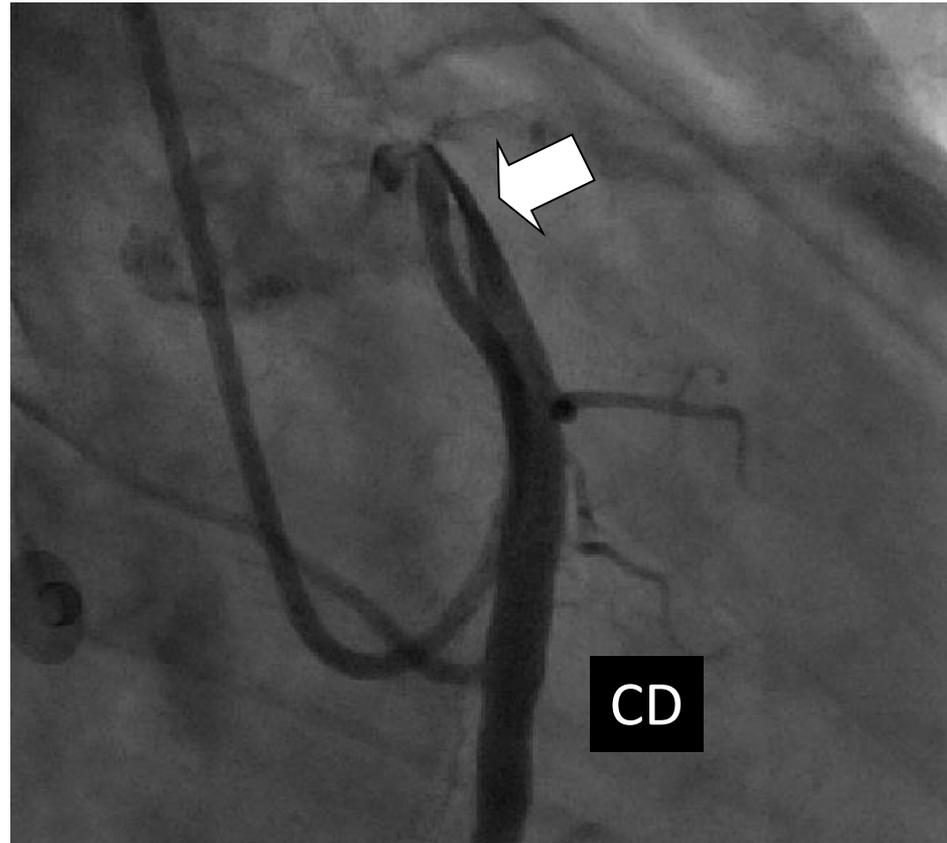
connexion droite dans le sinus controlatéral



coronaire droite avec probable passage intramural



connexion droite dans le sinus controlatéral
déformation coronaire

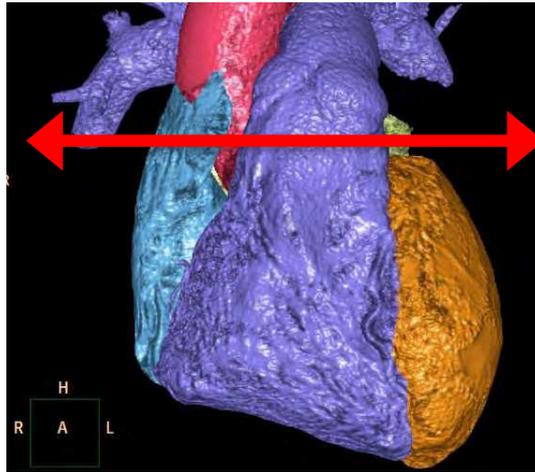


Techniques de cathétérisme

inversion des coupes



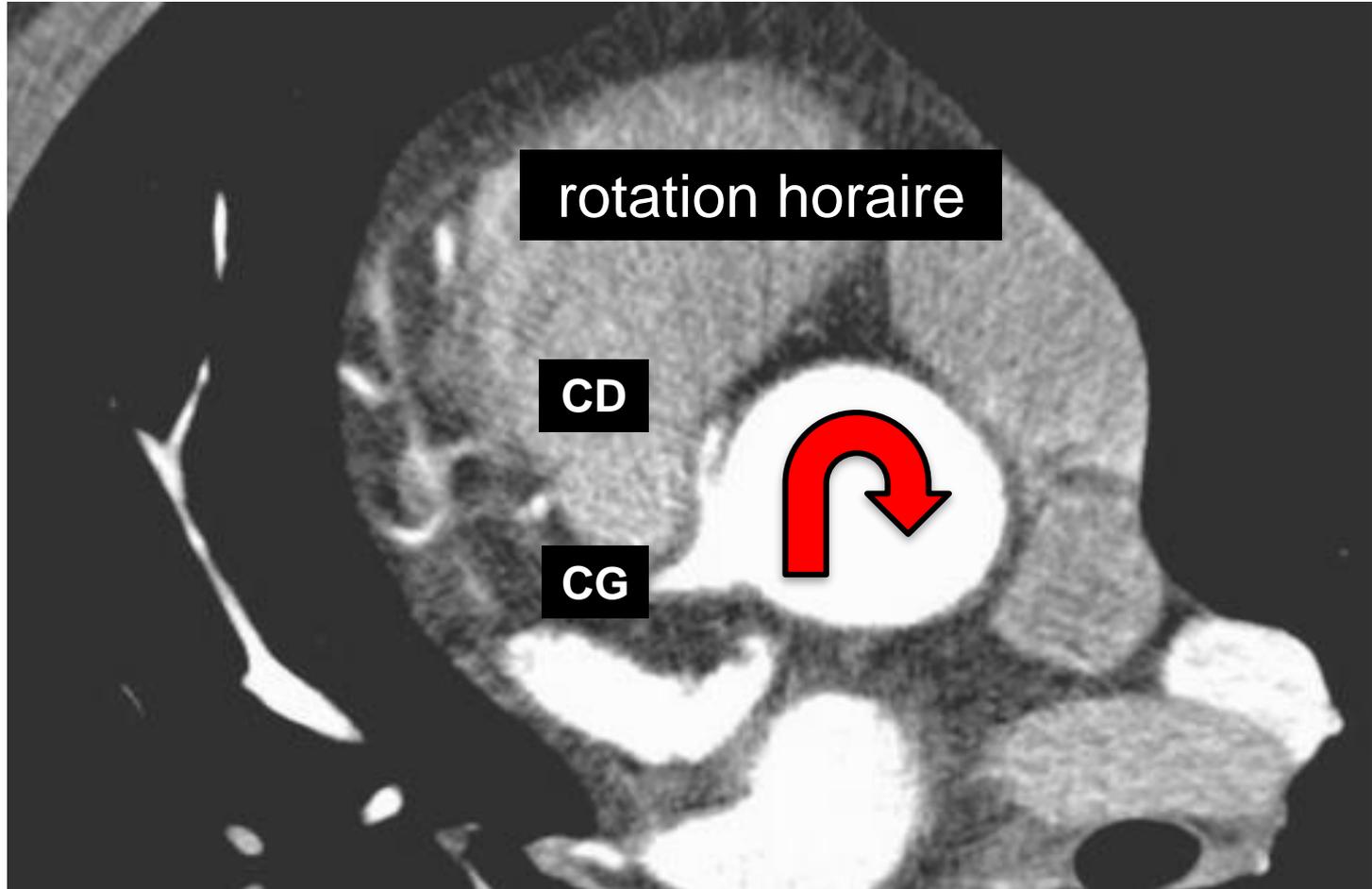
cardiologue



radiologue



manipulation des cathéters

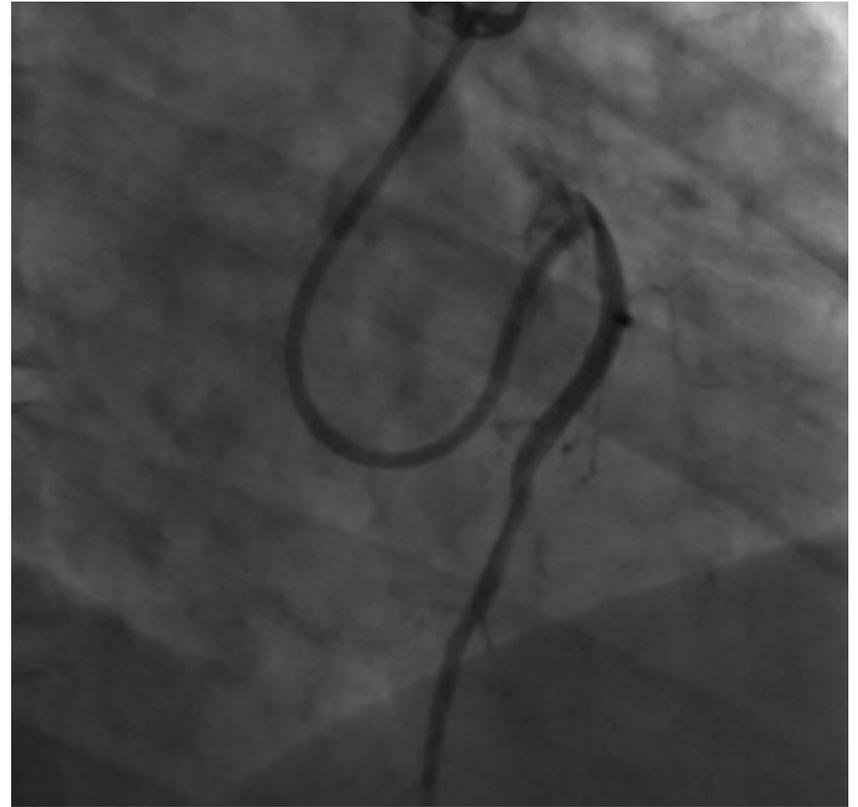


manipulation des cathéters



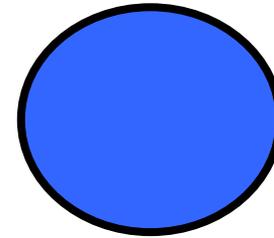
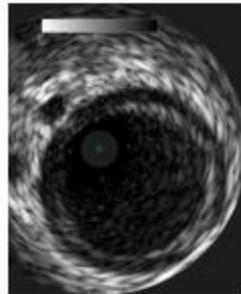
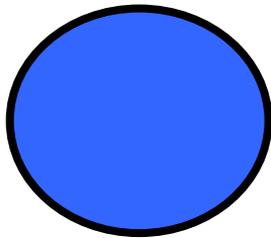
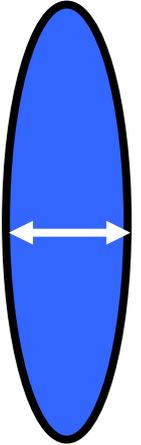
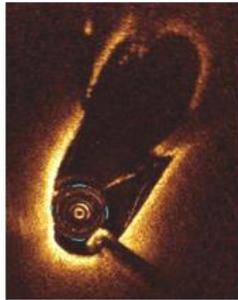
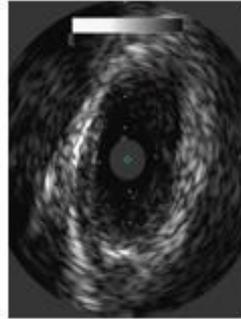
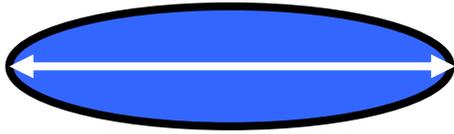
Push et rotation horaire

manipulation des cathéters

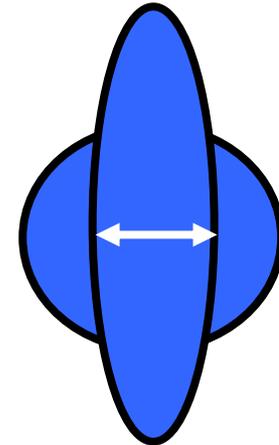
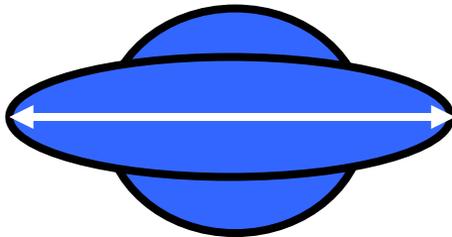
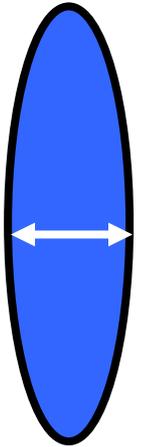
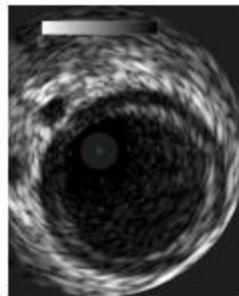
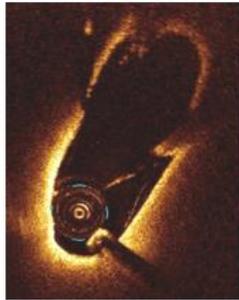
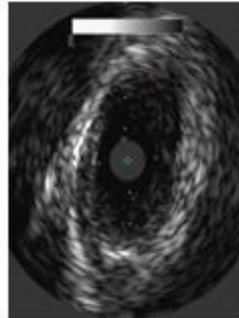
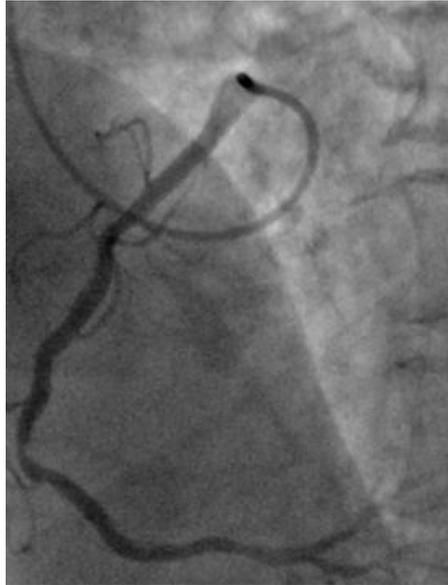
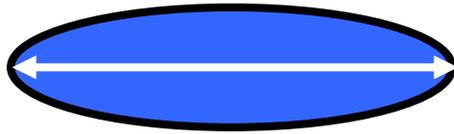


Utilisation du même cathéter

coronaire droite avec probable passage intramural



coronaire droite avec probable passage intramural



cathéters pour connexion droite dans sinus gauche

- Sonde diagnostique ou cathéter-guide
- Sonde JR : rarement efficace
- Sondes JL 3.5/4 : possible mais ...
- Sondes AL : le plus souvent AL0.75/1/2/3
- Sonde EBU 3.5/4 (voie radiale)
- Sonde AR : plutôt non

cathéters pour connexion droite dans sinus gauche

marque de cathéters



Adroit Cordis



Launcher Medtronic

Coronary intervention in anomalous origin of the right coronary artery (ARCA) from the left sinus of valsalva (LSOV): A single center experience

Kalaichelvan Uthayakumaran ^{a,*}, Vijayakumar Subban ^a,
 Anitha Lakshmanan ^b, Balaji Pakshirajan ^a, Ramkumar Solirajaram ^c,
 Jaishankar Krishnamoorthy ^c, Ezhilan Janakiraman ^c,
 Ulhas M. Pandurangi ^c, Latchumanadhas Kalidoss ^c,
 Mulasari Ajit Sankaradas ^d

Table 1 – Summary of 17 cases of PCI in Anomalous RCA originating from LSOV. The type of Take off, sequence of catheter tried and the successful catheter for engagement presented.

Patient no	Type of take off	Sequence of catheters	Successfully cannulated catheter	Amt of contrast (ml)	Fluoro time
1	A	JR 3.5,AR2; AL1; JL 4.0; JL 5.0	JL 5.0	210	17.5
2	C	JR 3.5; AR2; AL1	AL 1	120	12.2
3	B	JR 3.5; AR 2.0; AL 1; JL 4.0; JL 5.0; EBU 3.0	EBU 3.5	320	63.3
4	C	JR 3.5; AR 2.0; AL 1.0; JL 5.0; AL 2.0	AL -2	220	17.3
5	A	JR 3.5; AL 1.0; JL 4.0	JL 4.0	130	12.2
6	A	JR 3.5; JL 4.0; JL 5.0	JL 5.0	150	13.5
7	C	JR 3.5; AR 2; JL 5.0; AL 1	AL 1	200	17.8
8	B	JR 3.5; AR 2.0; AL 1; JL 5.0; EBU 3.5	EBU 3.5	280	23.8
9	A	JR 3.5; AR 2; AL 1; JL 4.0; JL 5.0	JL 5.0	270	22.5
10	C	JR 3.5; AR 2.0; JL 5.0; AL 1	AL1	180	15.4
11	C	JR 3.5; AR 2.0; AL1; AL 2.0, JL 4.0	JL 4.0	300	51.3
12	A	JR 3.5; AL 1; JL 4.0	JL 4.0	150	12.5
13	B	JR 3.5; AR 2.0; AL 1, JL 4.0	JL 4.0	180	21.5
14	A	JR 3.5; AL 1; JL 4.0; JL 5.0	JL 5.0	200	17.4
15	A	JR 3.5; JL 4.0; JL 5.0	JL 5.0	170	14.2
16	C	JR 3.5; AR 2.0; JL 5.0; AL 1	AL 1	180	15.6
17	A	JR 3.5; AL 1; JL 4.0; JL 5.0	JL 5.0	200	15.7

connexion aortique haute coronaire droite



cathétérisme des coronaires droites ectopiques

Si connexion sinus gauche

- Sondes : AL, EBU, JL
- Canulation ostium gauche
- Recherche OAG 40°
- Push et rotation horaire
- Opacification OAD 40°, OAG 40°
- Si cathétérisme difficile: cathé-guide 6F et guide 0.014

Si connexion aortique haute

- Sondes : AL, MP
- Recherche OAG 40° au dessus du sinus droit
- Rotation anti-horaire

Bilan complémentaire

Paolo Angelini,^{1,2*} MD, Carlo Uribe,² MD, Jorge Monge,² MD, Jonathan M. Tobis,³ MD,
MacArthur A. Elayda,⁴ MD, PhD, and James T. Willerson,¹ MD

Catheterization and Cardiovascular Interventions 86:199–208 (2015)

ANOCOR droites
n=67

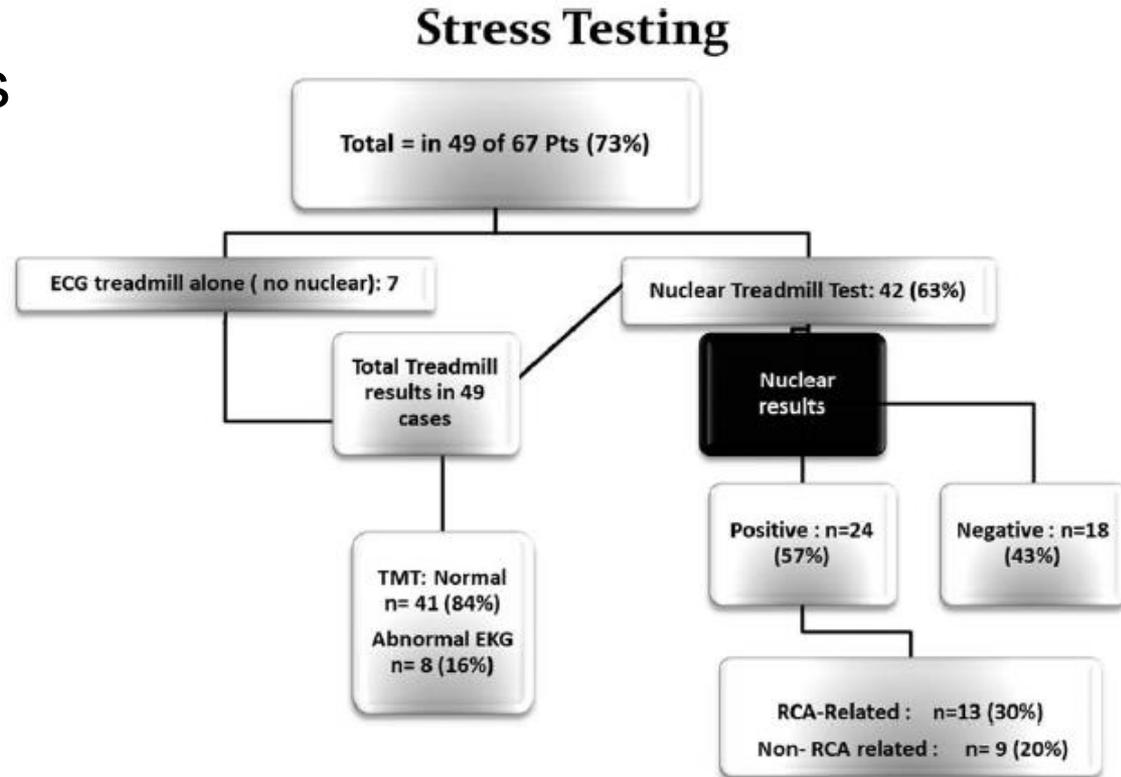


Fig. 3. Stress testing clinical evaluation studies for severity of ischemia. Note that, out of 67 patients, electrocardiographic treadmill test results were available for 49 patients and nuclear treadmill stress test results for 42 patients. ECG, electrocardiography; TMT, treadmill test; RCA, right coronary artery.

Clinical Features and Prognosis of Japanese Patients With Anomalous Origin of the Coronary Artery

Bunji Kaku, M.D., Masami Shimizu, M.D., Hiroyuki Yoshio, M.D.
 Hidekazu Ino, M.D., Sumio Mizuno, M.D.*, Honin Kanaya, M.D.**
 Shozo Ishise, M.D.*** and Hiroshi Mabuchi, M.D.

TABLE II POSITIVE EXERCISE STRESS TESTS IN PATIENTS WITHOUT ORGANIC CORONARY STENOSIS

(n=33)

<i>Type</i>	<i>Treadmill test or Master's 2-step test</i>	<i>Excercise ²⁰¹TL SPECT</i>	<i>Complications</i>
<i>RCA from the left sinus of Valsalva</i>	10/22	3/4	<i>Exercise-induced hypotension</i> 2 <i>Ventricular tachycardia</i> 2
<i>LCx from the right sinus of Valsalva</i>	3/5	1/2	
<i>LMCA from the posterior sinus of Valsalva</i>	1/4	0/2	<i>Exercise-induced hypotension</i> 1
<i>LMCA from the right sinus of Valsalva</i>	1/1		
<i>LAD and LCx from the right sinus of Valsalva</i>	1/1	0/1	
<i>Total</i>	16/33 (48.5%)	4/9 (44.4%)	5/33 (15.2%)

Jpn Circ J 1996; **60**: 731–741

Sudden Death

Clinical Profile of Congenital Coronary Artery Anomalies With Origin From the Wrong Aortic Sinus Leading to Sudden Death in Young Competitive Athletes

Cristina Basso, MD, PhD,* Barry J. Maron, MD, FACC,† Domenico Corrado, MD,‡ Gaetano Thiene, MD*

Padua, Italy and Minneapolis, Minnesota

Table 3. Previously Published Reports of Wrong Sinus Coronary Artery Anomalies in Persons Aged ≤ 35 Years in Whom ECG Data Were Available

Reference (#)	Year	Age, Gender	Anomaly	Symptoms	12-lead ECG	Exercise Stress Test	Myocardial Perfusion Scintigraphy	Athlete	Outcome ECG
Cheitlin et al. (11)	1974	14 M	LMCA	Syncope†	Normal	Normal	0	0	Alive (with surgery)
Pedal et al. (26)	1976	10 F	LMCA	Syncope	Normal	Normal	0	0	Sudden death
Benge et al. (27)	1980	25 M	RCA	Syncope	T wave inversion (II, III, aVF, V3-V6)	Normal	Not diagnostic	0	Alive
Mustafa et al. (28)	1981	12 M	LMCA	Syncope	Normal	Normal	Normal	0	Alive (with surgery)
Brandt et al. (29)	1983	35 M	RCA	Angina†	Inferior subendocardial MI	Normal	0	0	Alive (with surgery)
Donaldson et al. (30)	1983	* M	LMCA	Angina†	Normal	Positive (ventricular tachycardia)	0	0	Alive (with surgery)
Donaldson et al. (30)	1983	* M	LMCA	Angina†	Normal	Positive (subendocardial ischemia)	0	0	Alive (with surgery)
Donaldson et al. (30)	1983	* M	LMCA	Syncope†	Normal	Positive (subendocardial ischemia)	0	0	Alive (with surgery)
Barth and Roberts (31)	1986	14 M	LMCA	Syncope/angina†	Normal	Normal	0	0	Sudden death
Vander Sande et al. (32)	1989	14 F	LMCA	Syncope†	Normal	First degree atrioventricular block	0	0	Sudden death
Maron et al. (33)	1991	21 M	LMCA	Syncope†	LVH, T wave inversion (lateral leads)	Positive (sinus bradycardia, syncope)	0	0	Alive (with surgery)
Corrado et al. (14)	1992	22 M	RCA	Palpitations	Normal (PVC)	Normal	0	+	Sudden death
Amarasena et al. (34)	1993	18 M	LMCA	Syncope†	T wave inversion (I, aVL, V2-V3)	Normal	0	0	Alive (with surgery)
Jureidini et al. (35)	1994	12 M	LMCA	Syncope†	Normal	Normal	0	0	Sudden death
Van Son et al. (36)	1996	9 M	LMCA	Angina, syncope†	ST-T changes (II, aVF, V5-V6)	Normal	Normal	0	Alive (with surgery)
Phoon et al. (37)	1997	11 M	LMCA	Angina†	ST-T changes; PVC	Normal	Normal	0	Alive (with surgery)
Zeppilli et al. (38)	1998	17 M	RCA	None	Normal (PVC)	Normal	Normal	+	Alive
Zeppilli et al. (38)	1998	18 M	RCA	Dyspnea†	Incomplete RBBB	Normal	Positive (ischemia)	+	Alive

Sudden Death

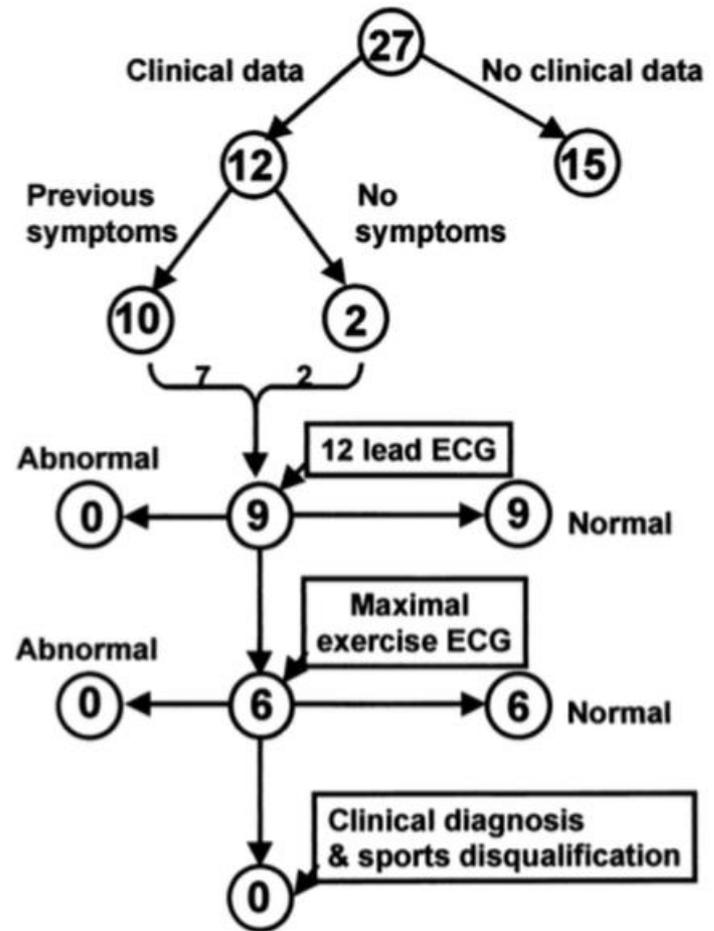
Clinical Profile of Congenital Coronary Artery Anomalies With Origin From the Wrong Aortic Sinus Leading to Sudden Death in Young Competitive Athletes

Cristina Basso, MD, PhD,* Barry J. Maron, MD, FACC,† Domenico Corrado, MD,‡ Gaetano Thiene, MD*

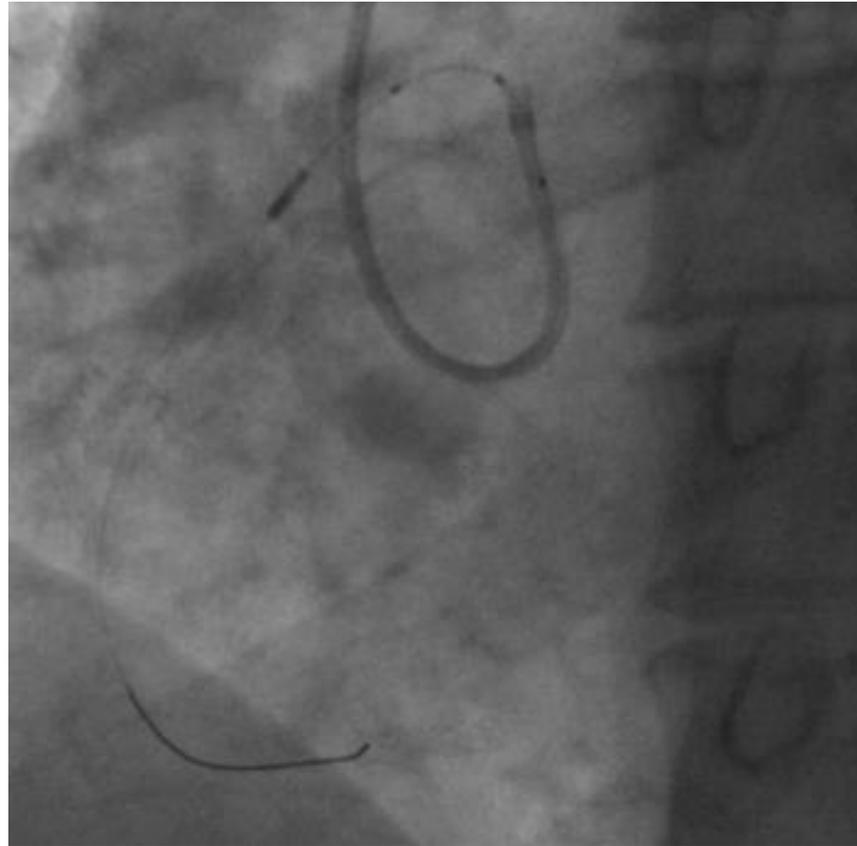
Padua, Italy and Minneapolis, Minnesota

Table 2. Demographic and Clinical Data in 12 Athletes With Wrong Sinus Coronary Artery Origin and Clinical Manifestations or Diagnostic Testing During Life

Age at Death	Gender	Nation	Race	Sport	Level	Site	Activity	Circumstances of Death	Prior Symptoms	Time From Symptoms to Sudden Death
11	M	Italy	W	Soccer	JHS	Field	Game	During effort	No	—
12	M	U.S.	B	Basketball	JHS	Field	Practice	During effort	Chest pain	7 days
12	M	U.S.	W	Hockey	JHS	Hotel	Sedentary	After effort	Syncope*, chest pain*	15 mo
12	M	U.S.	B	Basketball	JHS	Field	Practice	During effort	Syncope*†	14 mo
14	M	Italy	W	Soccer	JHS	Field	Game	During effort	No	—
15	M	Italy	W	Soccer	JHS	Field	Game	During effort	Syncope*	11 mo
15	F	U.S.	W	Tracksprint	HS	Field	Practice	During effort	Dizziness, palpitations*	15 mo
15	M	U.S.	B	Basketball	JHS	Field	Practice	During effort	Syncope†, chest pain	24 mo
16	M	U.S.	B	Basketball	HS	Field	Game	During effort	Chest pain*	8 mo
22	M	Italy	W	Soccer	Pro	Field	Game	During effort	Palpitations	12 mo
29	M	Italy	W	Rugby	Pro	Field	Practice	During effort	Palpitations	13 mo
32	F	Italy	W	Running	Pro	Field	Game	During effort	Chest pain*	9 mo

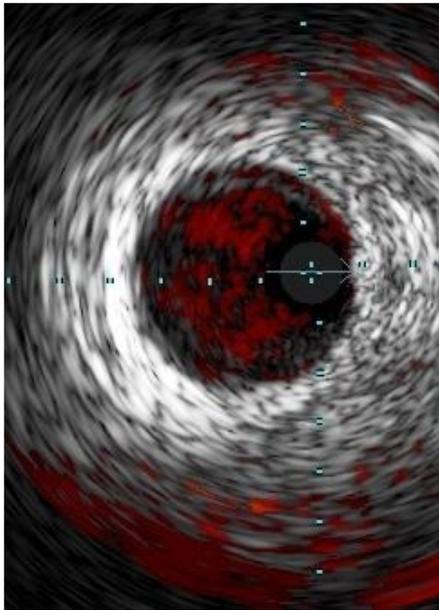


échographie endocoronaire

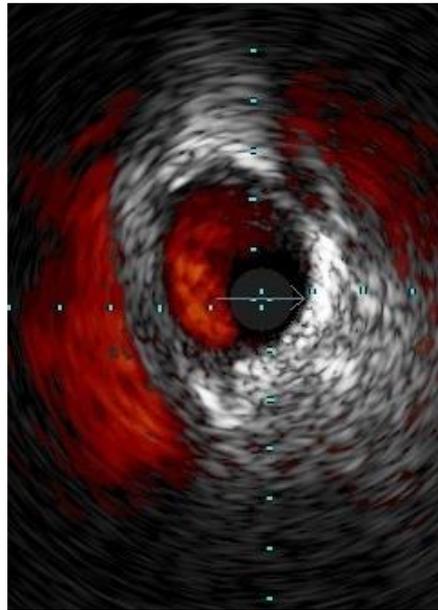


échographie endocoronaire ANOCOR droite avec passage intramural

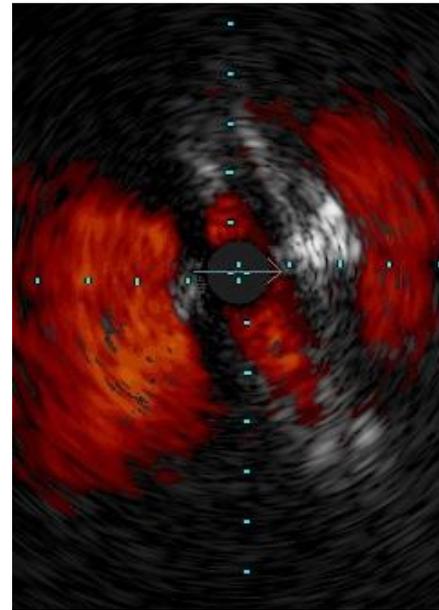
extramural



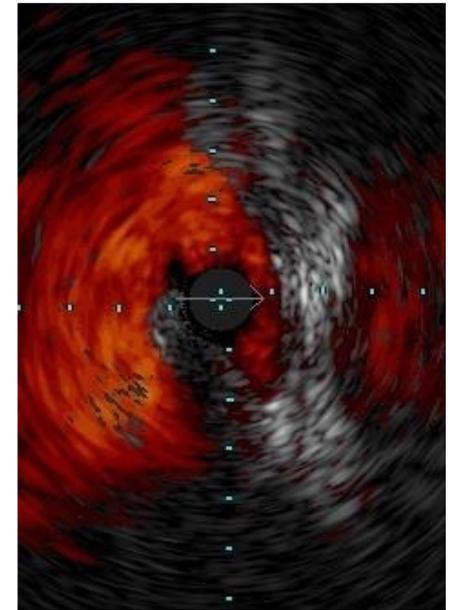
juxtamural



intramural



ostium



Paolo Angelini,^{1,2*} MD, Carlo Uribe,² MD, Jorge Monge,² MD, Jonathan M. Tobis,³ MD,
MacArthur A. Elayda,⁴ MD, PhD, and James T. Willerson,¹ MD

Catheterization and Cardiovascular Interventions 86:199–208 (2015)

ANOCOR droites
n=66

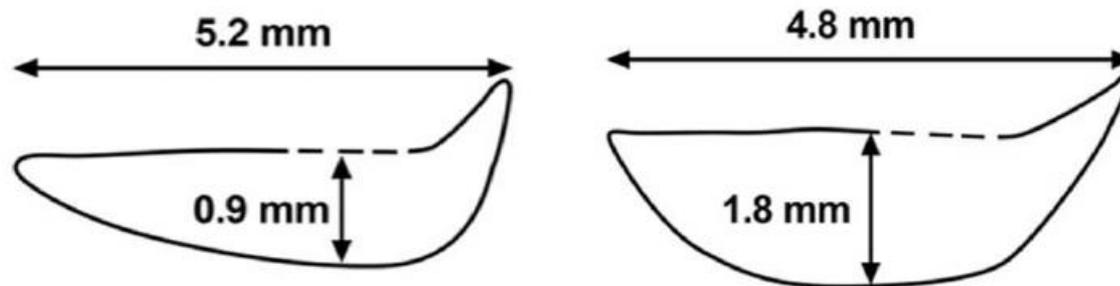
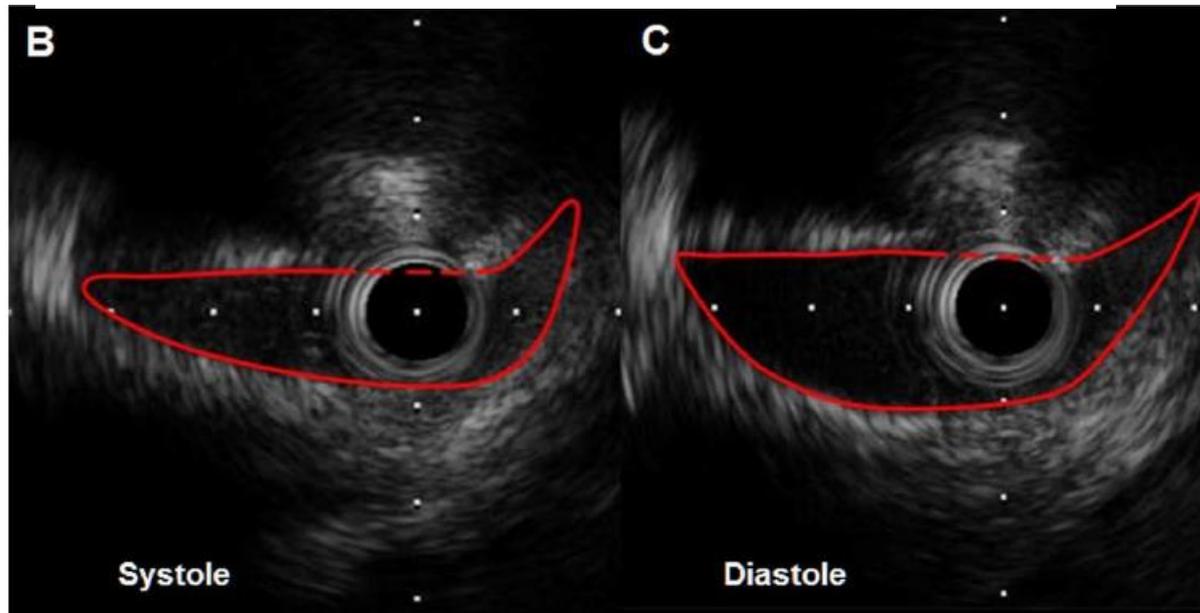
TABLE IV. Quantitative Measurements From Intravascular Ultrasonography

	Mean \pm SD	Range
Reference distal segment of RCA		
Cross-sectional area (mm ²)	12.1 \pm 4.2	4.5–28.0
Diameter (mm)	3.9 \pm 0.7	2.4–6.0
Intramural segment of RCA (worst stenosis)		
Cross-sectional area (mm ²)	6.0 \pm 2.4	1.6–16.2
Minimal diameter (mm)	1.6 \pm 0.7	0.9–4.8
Maximal diameter (mm)	4.4 \pm 1.3	2.4–8.6
Minimal/maximal diameter index (mm)	0.40 \pm 0.17	0.14–0.99
Degree of stenosis (%AS)	49 \pm 18	4–83

%AS, percent area of stenosis; RCA, right coronary artery.

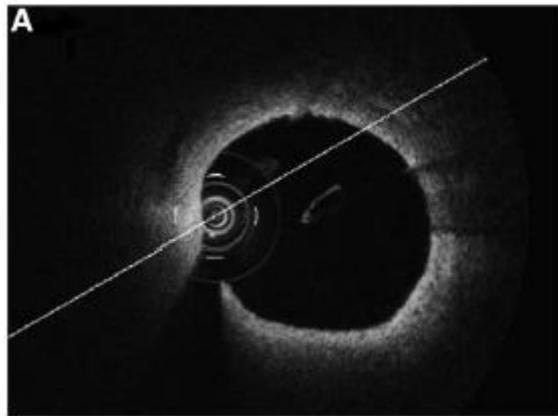
Paolo Angelini,^{1,2*} MD, Carlo Uribe,² MD, Jorge Monge,² MD, Jonathan M. Tobis,³ MD,
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Catheterization and Cardiovascular Interventions 86:199–208 (2015)



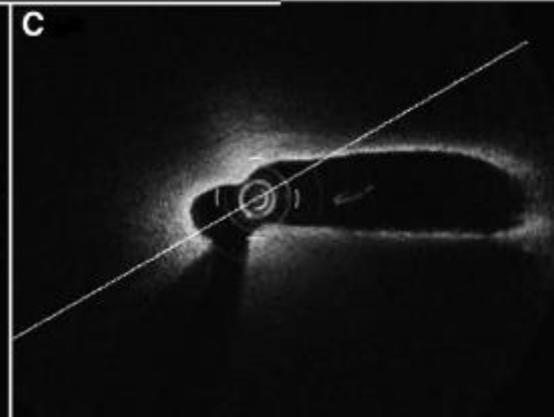
Optical Coherence Tomography endocoronaire ANOCOR droite avec probable passage intramural

Paolo Angelini,^{1,2*} MD, Carlo Uribe,² MD, Jorge Monge,² MD, Jonathan M. Tobis,³ MD,
MacArthur A. Elayda,⁴ MD, PhD, and James T. Willerson,¹ MD

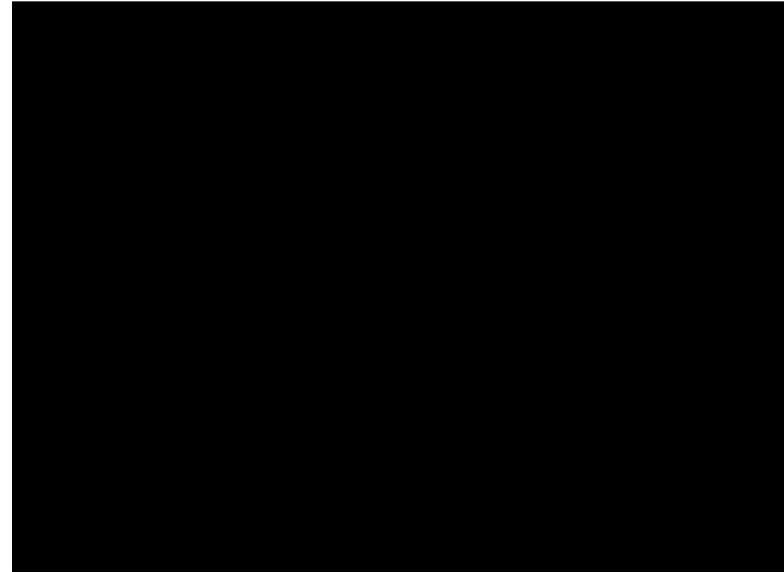


Catheterization and Cardiovascular Interventions 86:199–208 (2015)

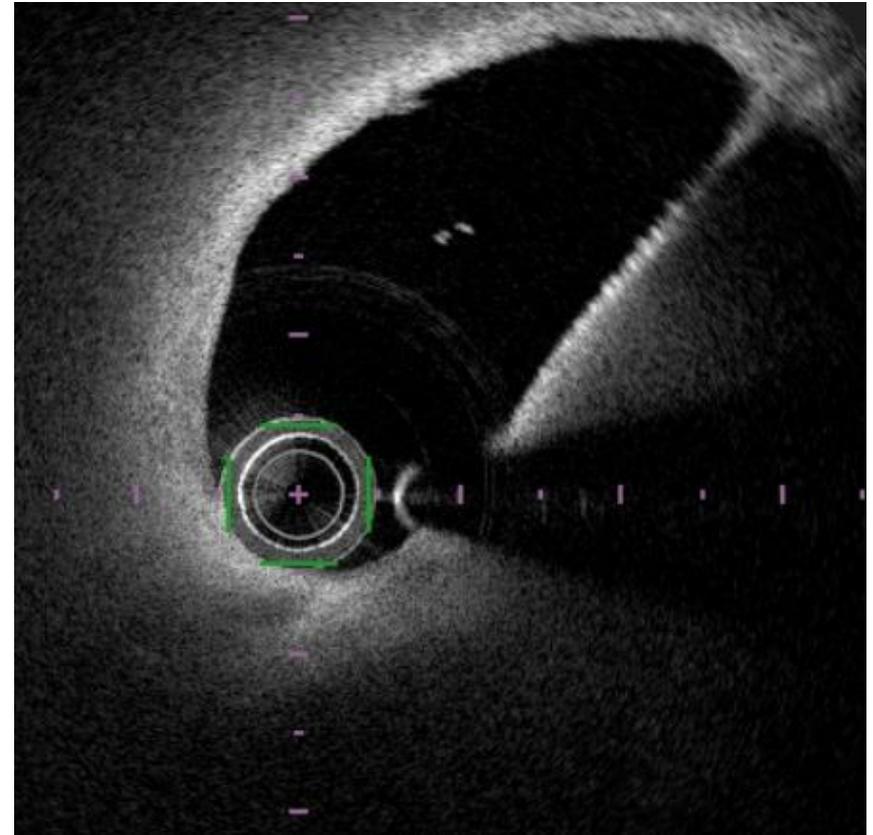
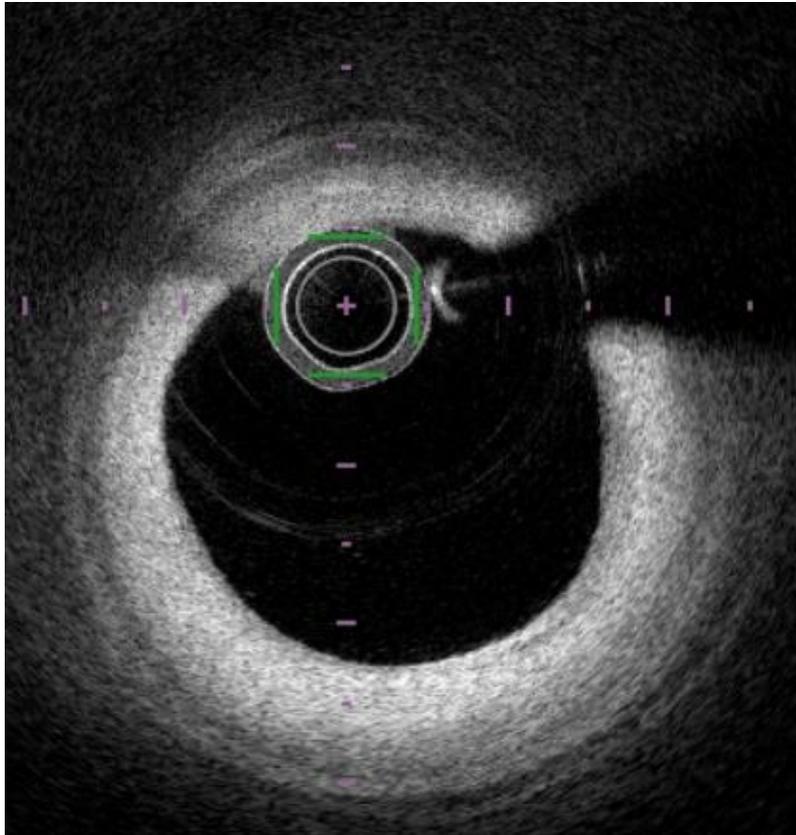
Fig. 2. Optical coherence tomography (OCT) images. With this modality, the inner lumen can be crisply imaged, but only if the operator can advance the guiding catheter selectively enough to eliminate most of the luminal blood, thereby enabling infrared laser imaging. (A) The reference distal cross-sectional area (CSA) is shown (diameters 3.8×4.0 mm; CSA 15.42 mm^2) for contrast with the proximal systolic (B) (diameters, 8.7×5.3 mm; CSA, 4.6 mm^2 ; 70% stenosis) and diastolic images (C) (diameters, 6.2×10.3 mm; CSA, 6.4 mm^2 ; 58% stenosis).



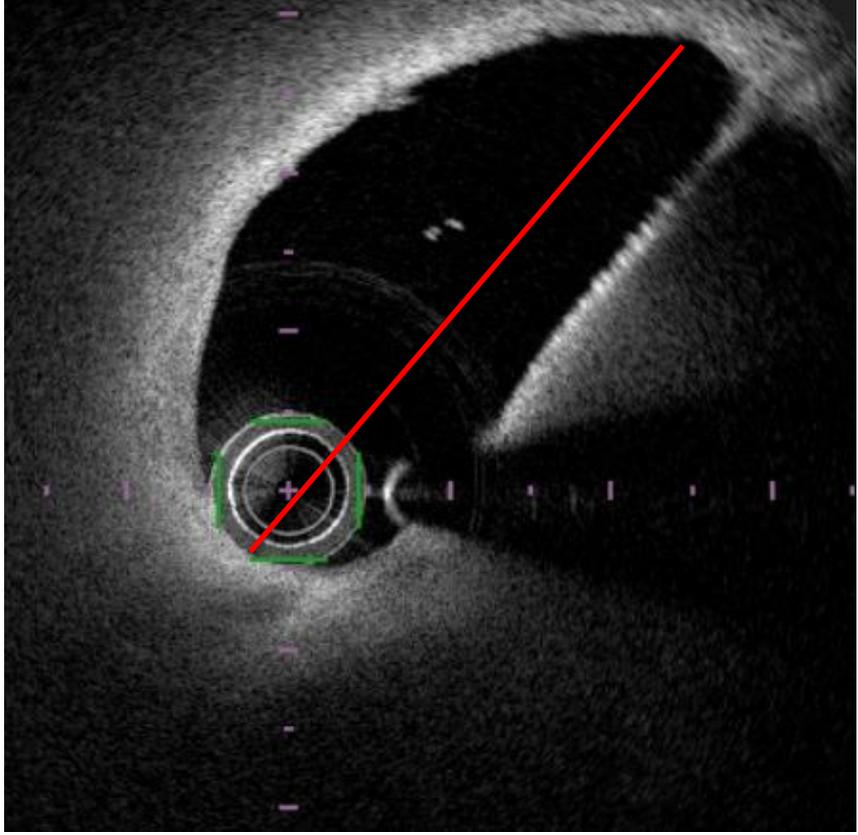
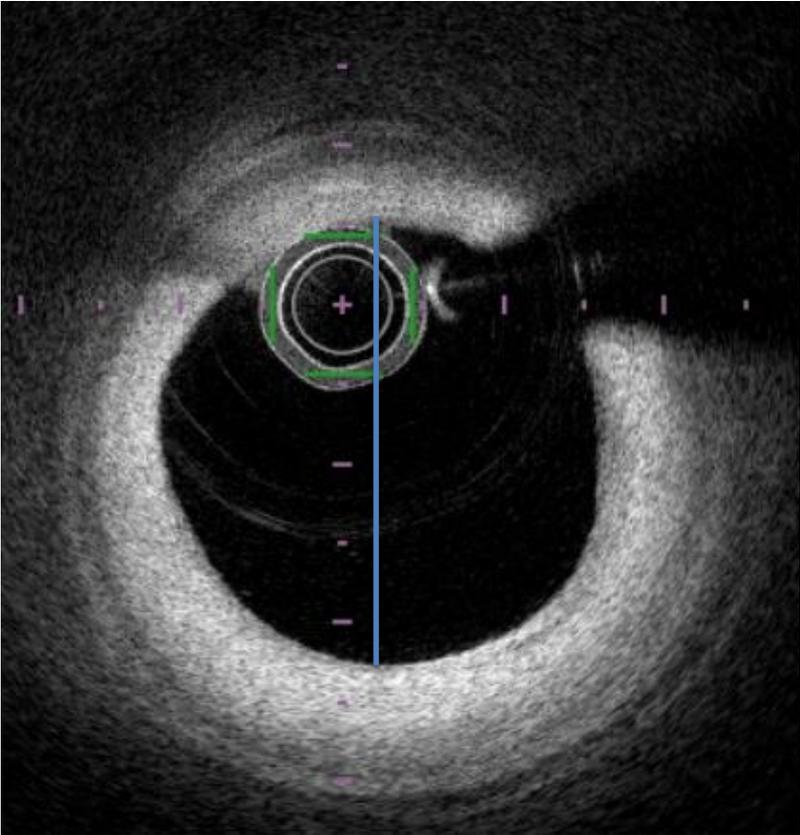
Optical Coherence Tomography endocoronaire ANOCOR droite



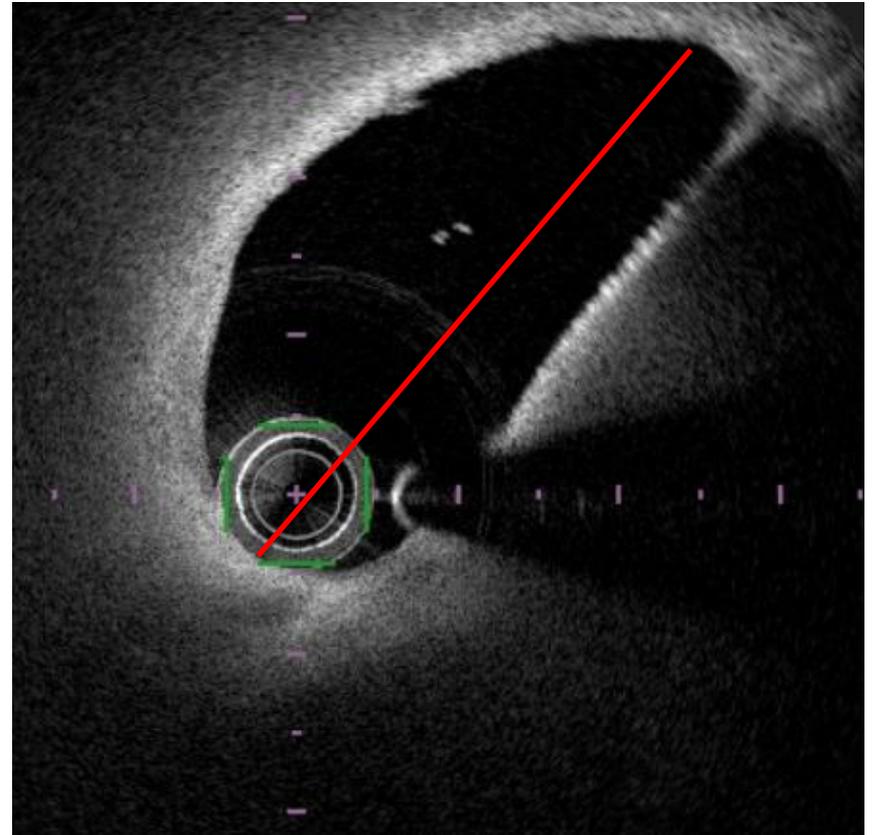
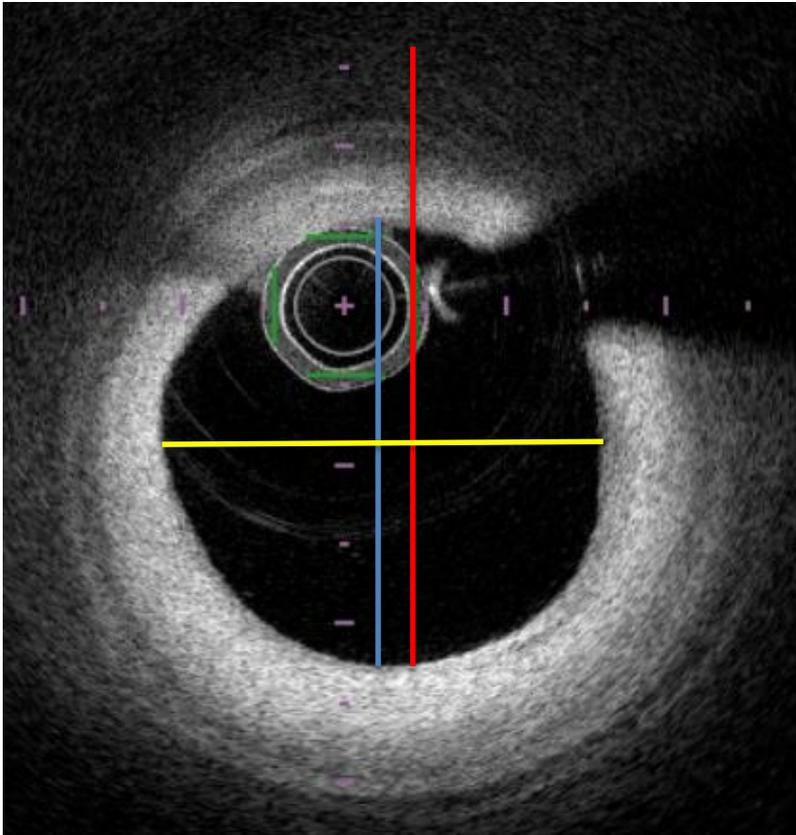
Optical Coherence Tomography endocoronaire ANOCOR droite



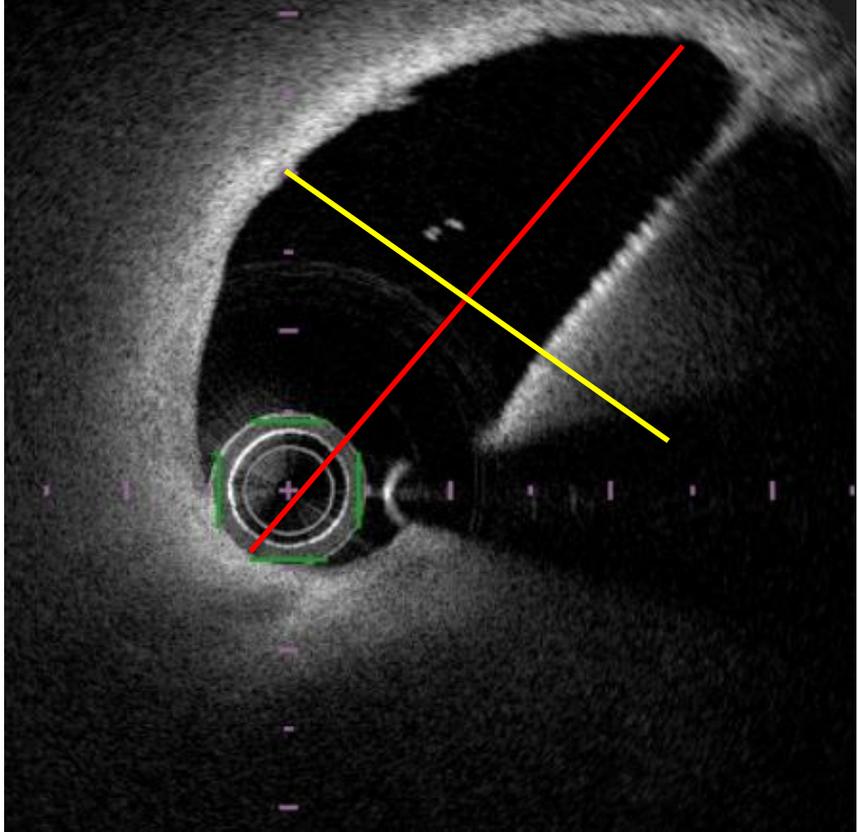
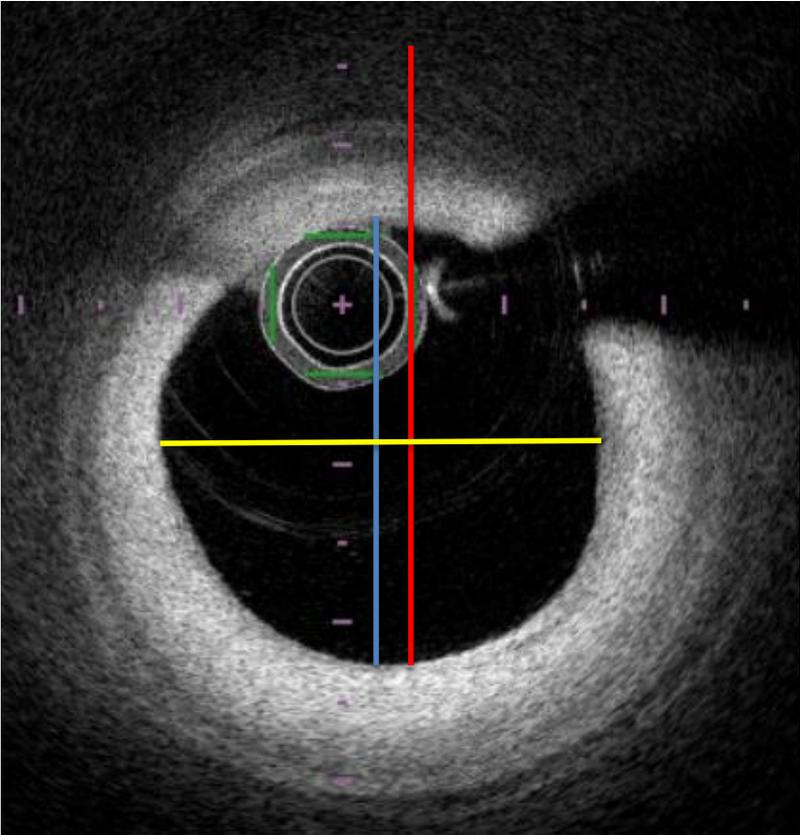
Optical Coherence Tomography endocoronaire ANOCOR droite



Optical Coherence Tomography endocoronaire ANOCOR droite

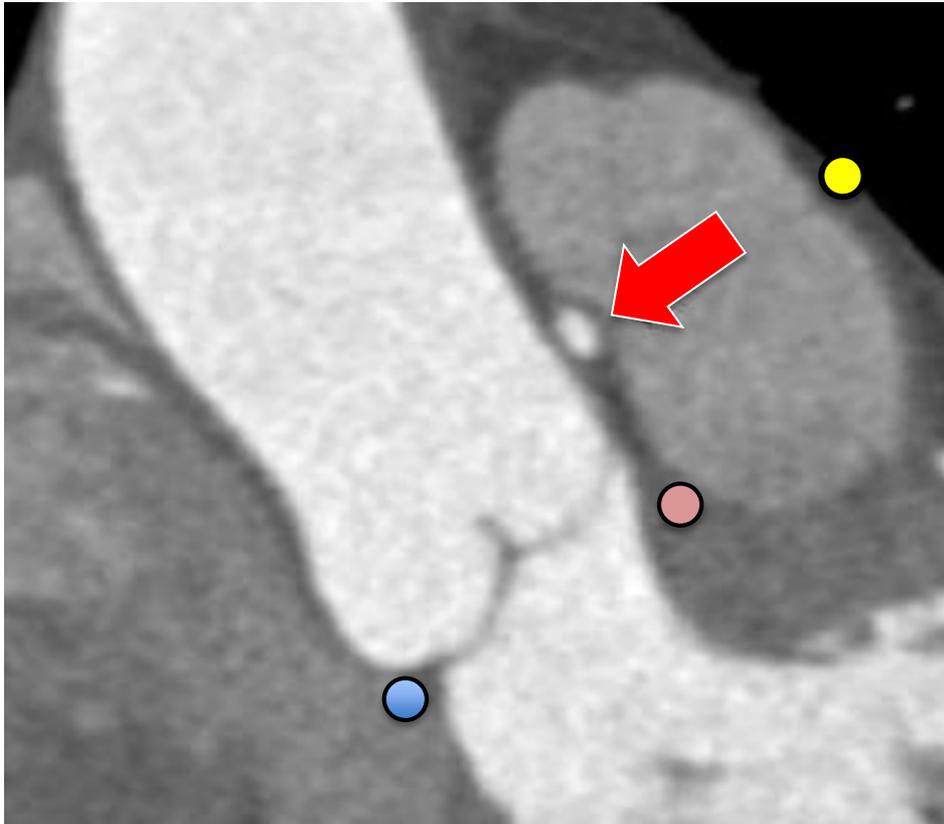


Optical Coherence Tomography endocoronaire ANOCOR droite



Evaluation des risques

patients à risque de mort subite
question du patient : suis-je exposé ?



- pré-pulmonaire
- rétro-infundibulaire
- pré-aortique
- rétro-aortique

prévalence angiographique des anomalies à risque *connexion ectopique avec trajet préaortique*

cohorte ANOCOR*

472 patients ≥ 15 ans - 496 ANOCOR
janvier 2010 - janvier 2013

$$\frac{151}{496} = 30\%$$

*Aubry P et al. Anomalous connections of the coronary arteries: a prospective observational cohort of 472 adults. The ANOCOR registry. Eur Heart J 2015;36 suppl 1:1138.



anomalies de connexion proximale coronaire

prévalence de l'anomalie en population générale

estimations

▪ prévalence ANOCOR globale	1/1.000 (0.01%)
▪ prévalence ANOCOR à risque	0.3/1.000 (0.03%)
▪ prévalence CIA	0.5/1.000 (0.05%)

marathon de Paris 2016 : 57.000 inscrits
17 participants potentiels avec ANOCOR à risque

population à risque de mort subite
risque de mort de subite

prévalence

incidence

=

numérateur

dénominateur

risque de mort subite

question du patient : à quel risque suis-je exposé ?

$$\text{fréquence annuelle} = \frac{\text{n événements}/100 \text{ patients}^*}{360 \text{ jours}}$$

* patients ayant une anomalie de connexion identifiée à risque de mort subite

mort subite et anomalies congénitales coronaires

12-35 years athletic population

follow-up period of 26 years

2.938.270 person-years of observation

55 deaths

91% during sports activity

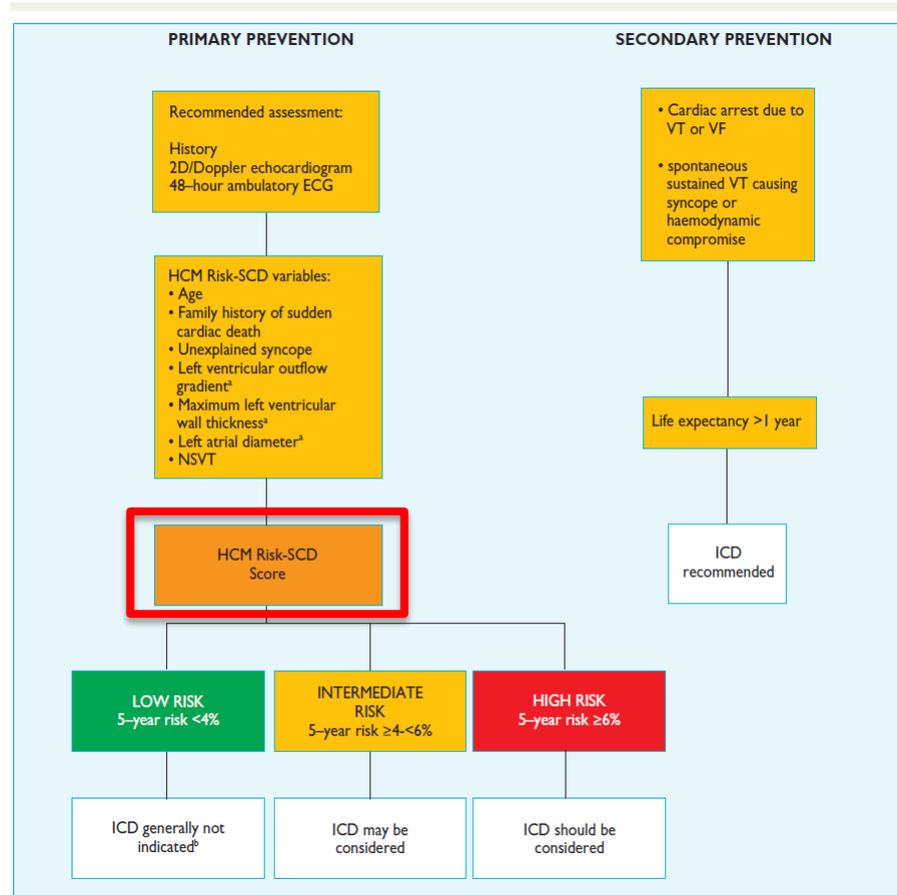
1.9 deaths/100.000 person-years

0.48 deaths/100.000 person-years (cardiomyopathy)

0.24 deaths/100.000 person-years (congenital coronary anomaly)

Corrado et al. JAMA 2016

2014 ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy



Options thérapeutiques

management des ANOCOR sans ischémie

ACC/AHA Guideline

ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease

Circulation December 2, 2008

When the patient has an anomalous right coronary artery and no evidence of ischemia, management is more controversial. A conservative approach in this situation may be reasonable. Given the not uncommon occurrence of anomalous coronary origins and their potential for a devastating outcome, it is imperative that improved data are generated regarding diagnosis, follow-up, and longer-term outcomes.

anomalies de connexion proximale coronaire

éléments décisionnels actuels pour le management

- âge < 30, 30-50, > 50 ans
- symptômes + lien avec l'effort
- antécédent de mort subite récupérée
- présence d'une ischémie myocardique
- anatomie (imagerie)
- souhait d'activité physique/sportive intense

anomalies de connexion proximale coronaire

possibilités pour le management

- chirurgie
- angioplastie
- médicaments
- restriction physique/sportive
- rien

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

Circulation December 1, 2015

- 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*).**

- 3. After successful surgical repair of an anomalous origin from the wrong sinus, athletes may consider participation in all sports 3 months after surgery if the patient remains free of symptoms and an exercise stress test shows no evidence of ischemia or cardiac arrhythmias (*Class IIb; Level of Evidence C*).**

- 6. Nonoperated athletes with an anomalous origin of a right coronary artery from the left sinus of Valsalva who exhibit symptoms, arrhythmias, or signs of ischemia on exercise stress test should be restricted from participation in all competitive sports, with the possible exception of class IA sports, before a surgical repair (*Class III; Level of Evidence C*).**

ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease

8.5. Recommendations for Congenital Coronary Anomalies of Ectopic Arterial Origin

CLASS I

Circulation December 2, 2008

3. Surgical coronary revascularization should be performed in patients with any of the following indications:
 - a. Anomalous left main coronary artery coursing between the aorta and pulmonary artery. (*Level of Evidence: B*)
 - b. Documented coronary ischemia due to coronary compression (when coursing between the great arteries or in intramural fashion). (*Level of Evidence: B*)
 - c. Anomalous origin of the right coronary artery between aorta and pulmonary artery with evidence of ischemia. (*Level of Evidence: B*)

ACC/AHA Guideline

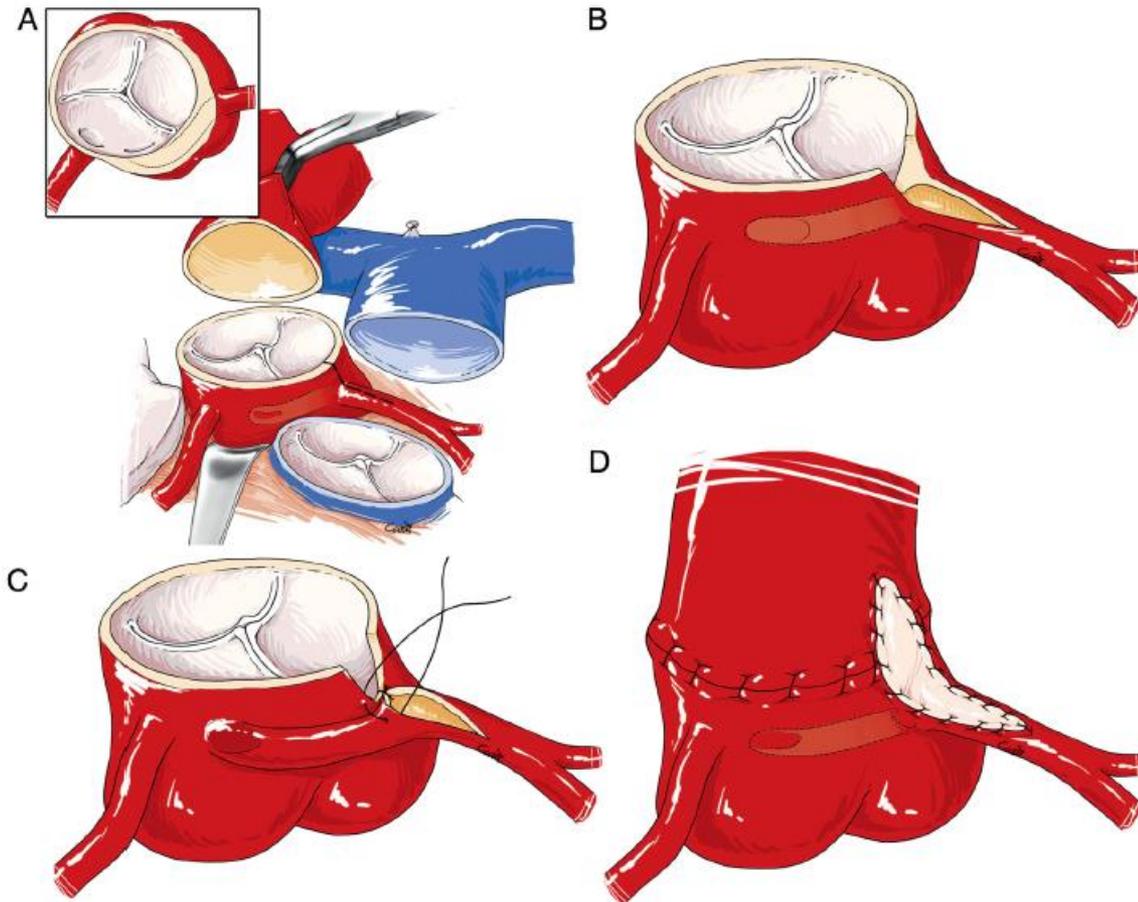
ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease

Circulation December 2, 2008

Class IIa

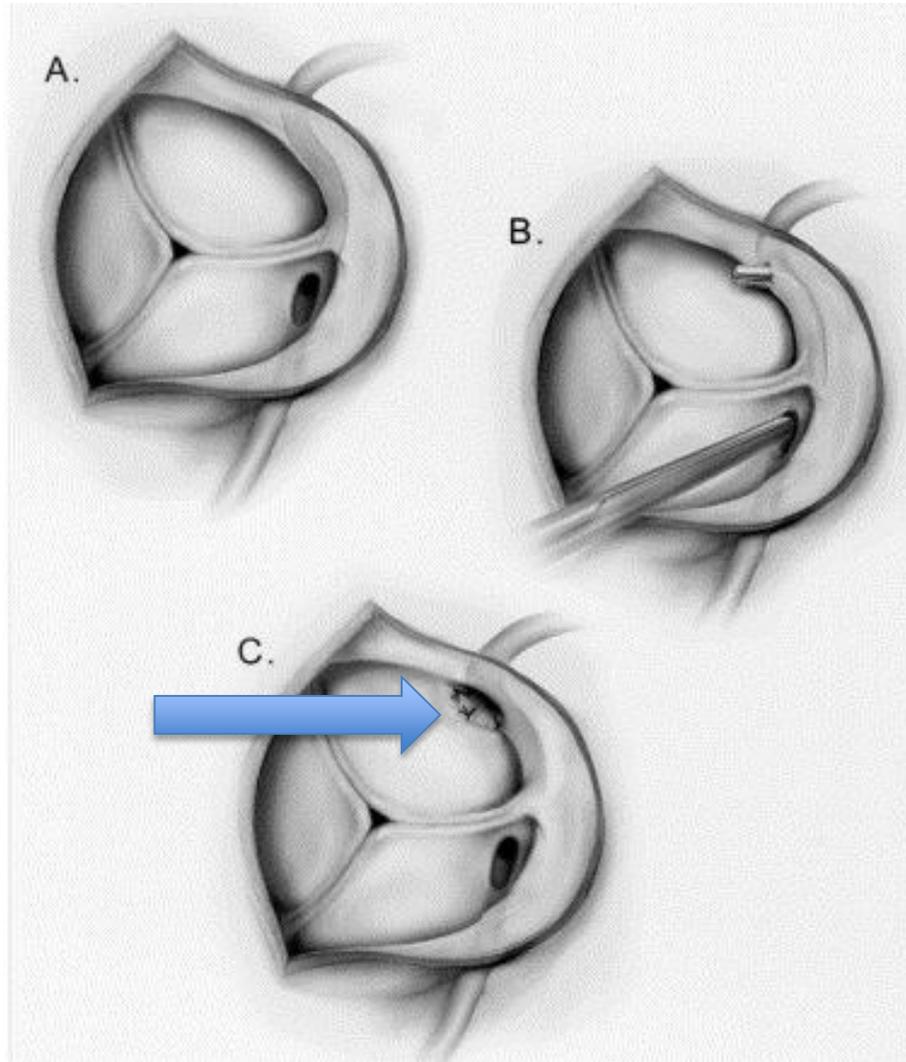
1. Surgical coronary revascularization can be beneficial in the setting of documented vascular wall hypoplasia, coronary compression, or documented obstruction to coronary flow, regardless of inability to document coronary ischemia. (*Level of Evidence: C*)
2. Delineation of potential mechanisms of flow restriction via intravascular ultrasound can be beneficial in patients with documented anomalous coronary artery origin from the opposite sinus. (*Level of Evidence: C*)

correction chirurgicale : réimplantation



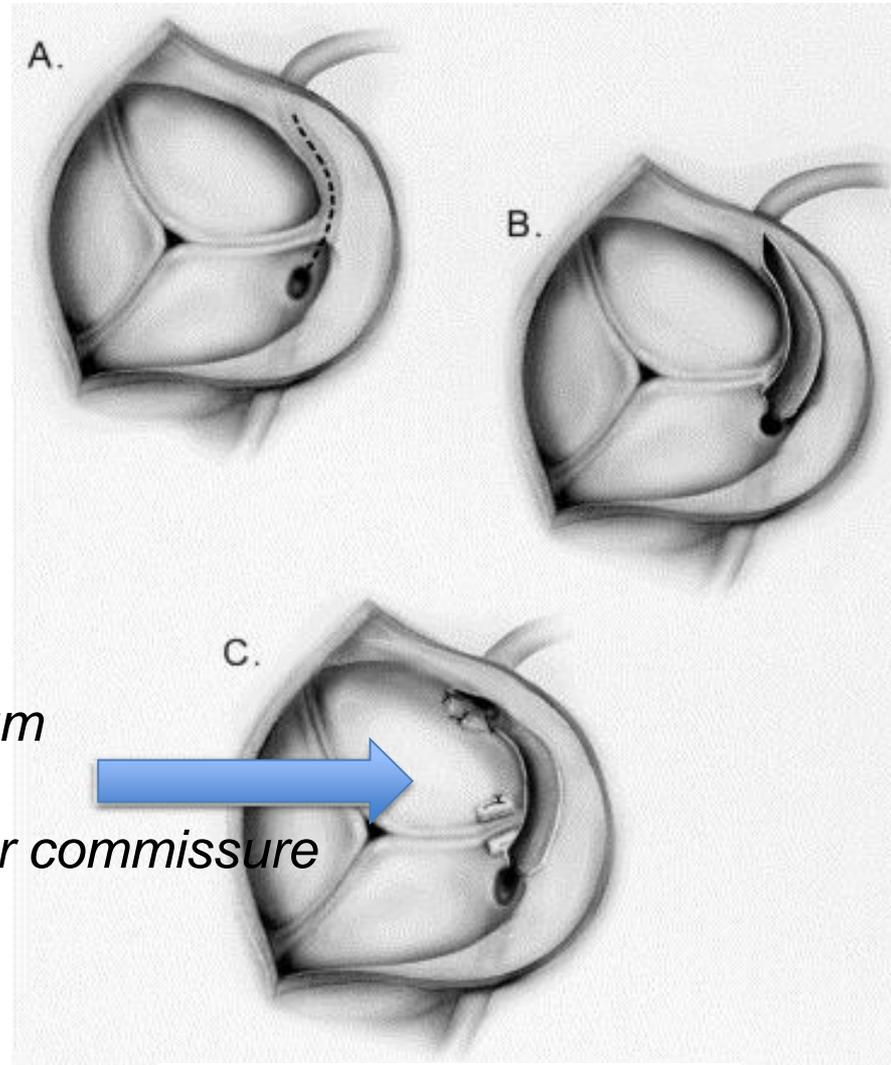
Gaudin R et al. Multimed Man Cardiothorac Surg 2014

correction chirurgicale : création néoostium



(Ann Thorac Surg 2003;76:589-96)

correction chirurgicale : *unroofing*

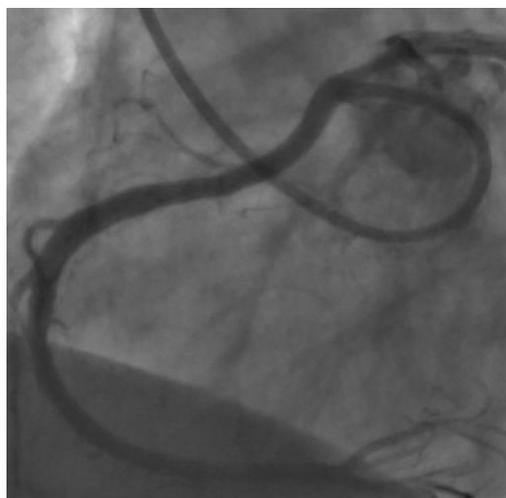
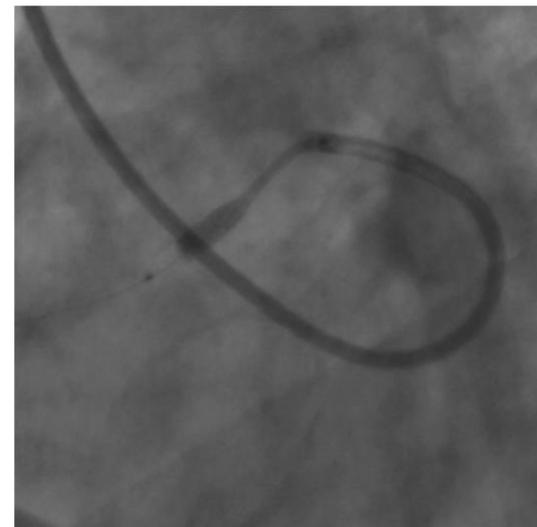
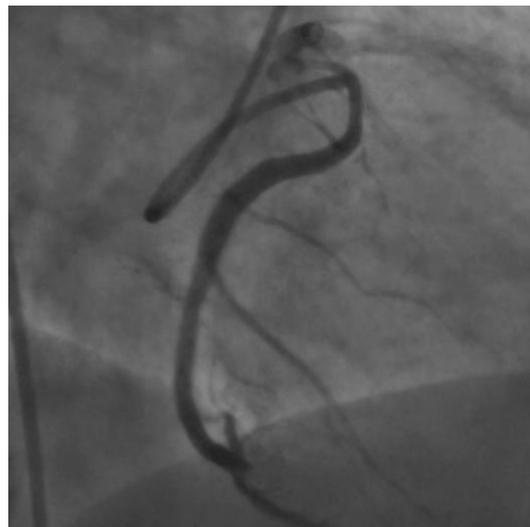
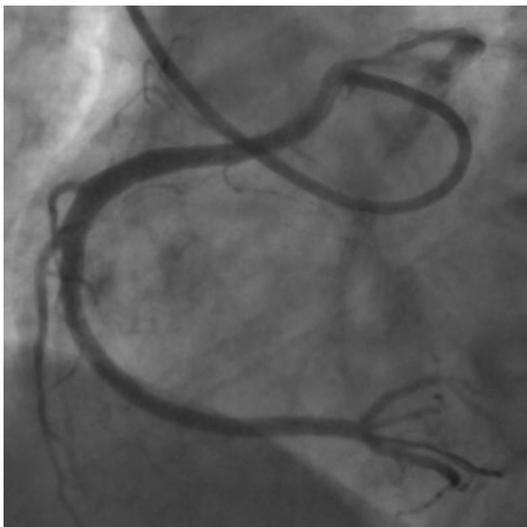


création néo ostium

geste éventuel sur commissure

(Ann Thorac Surg 2003;76:589-96)

traitement interventionnel par stenting



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

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TABLE I. Presenting Symptoms

Category	No. (%) of patients
Asymptomatic	8 (12%)
Chest pain	58 (87%)
Atypical	14 (24%)
Mild	3 (4%)
Moderate	8 (12%)
Severe	3 (4%)
Typical (CCS)	44 (76%)
I	5 (7%)
II	8 (12%)
III	21 (31%)
IV	10 (15%)
Shortness of breath (NYHA)	36 (54%)
I	15 (22%)
II	17 (25%)
III	4 (6%)
IV	0 (0)
Neurologic ischemic symptoms	24 (36%)
Dizziness	8 (12%)
Syncope	15 (22%)
Syncope causing collapse	1 (1%)

CCS, Canadian Cardiovascular Society; NYHA, New York Heart Association.

- Etude rétrospective avec 67 ANOCOR droites
- Age 48 ± 12 an (12-73)
- 42 angioplasties (BMS/Cypher/Taxus/Promus)
- Indication angioplastie :
 - symptômes
 - souhait sport intensif
 - réduction surface > 50%
- Procédure guidée par IVUS
- Succès angiographique (100%)
- Resténose angiographique (4/42)

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ACC/AHA Guideline

ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease

Circulation December 2, 2008

- Réimplantation plutôt que pontage (flux compétitif)
- Angioplastie (peu de cas publiés et un suivi limité)
- Attitude non interventionniste en cas d'ANOCOR droite sans ischémie

ANOCOR RISK

cohorte observationnelle prospective
anomalies de connexion proximale avec trajet préaortique
début du recrutement : courant 2017





informations

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