

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

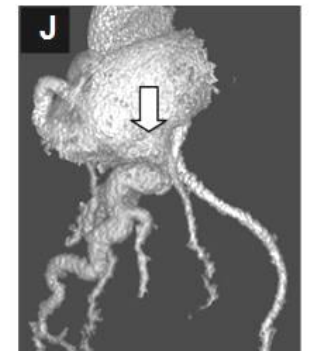
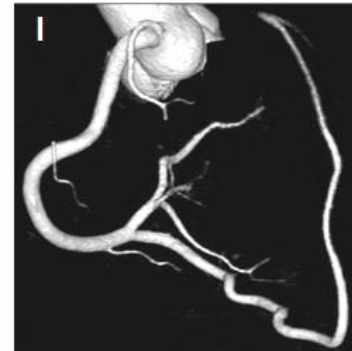
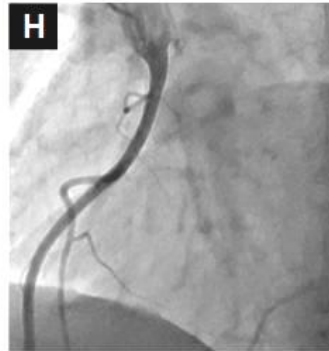
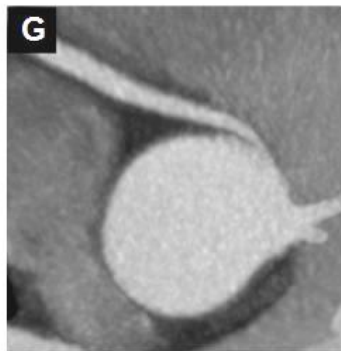
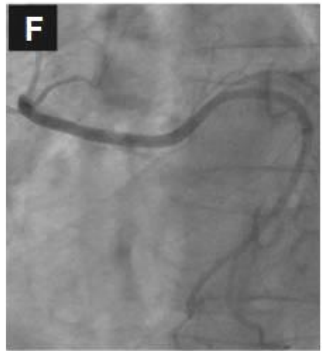
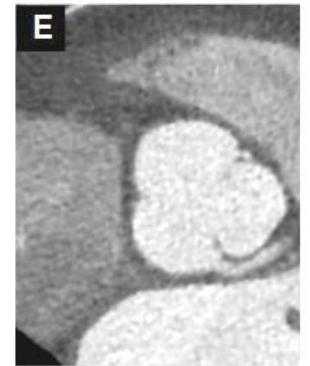
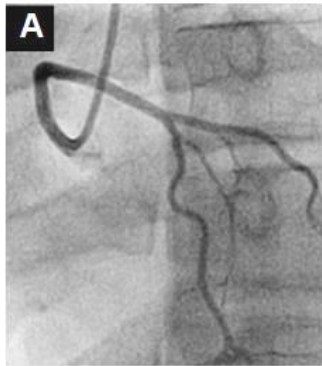
**Intervenant : Patrick Dupouy, Antony**

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

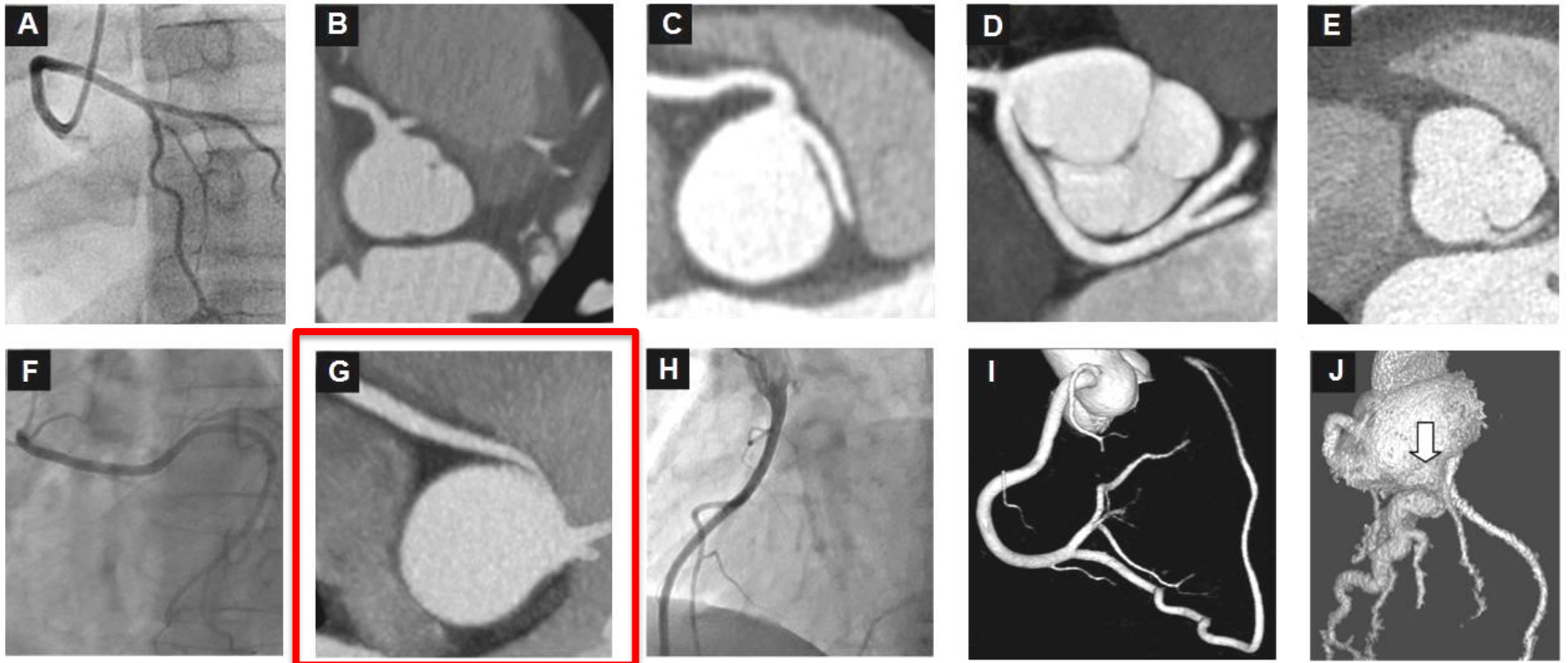
**Intervenant : Xavier Halna du Fretay, Saran**

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

# connexions anormales des coronaires



# connexions anormales des coronaires



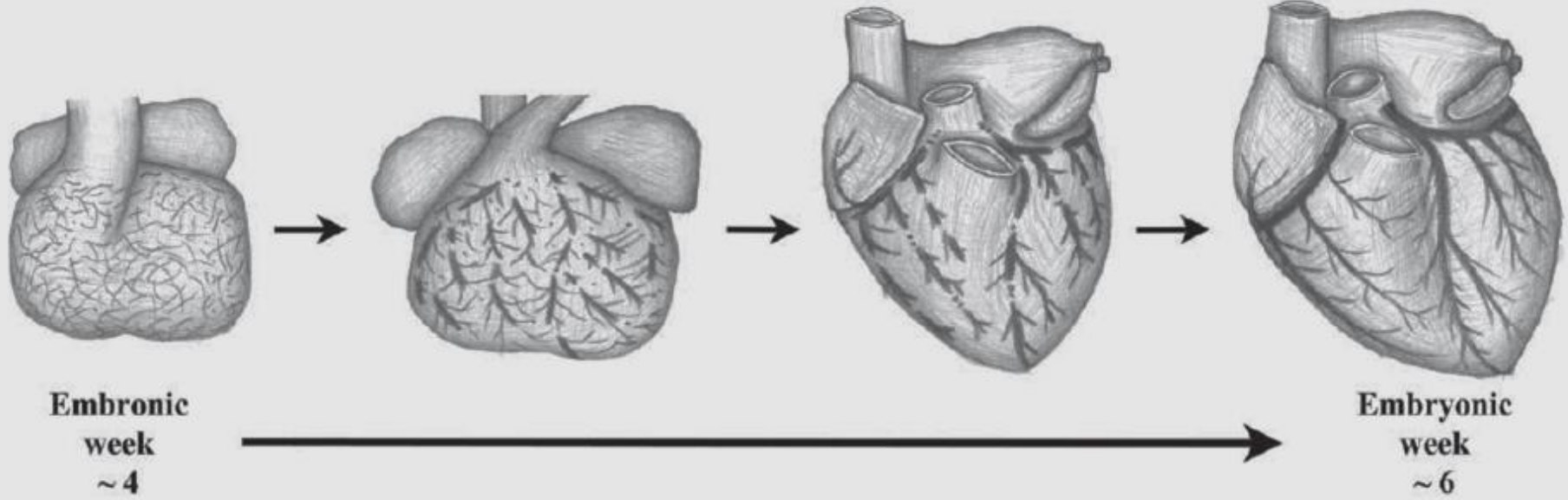
droite

# 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

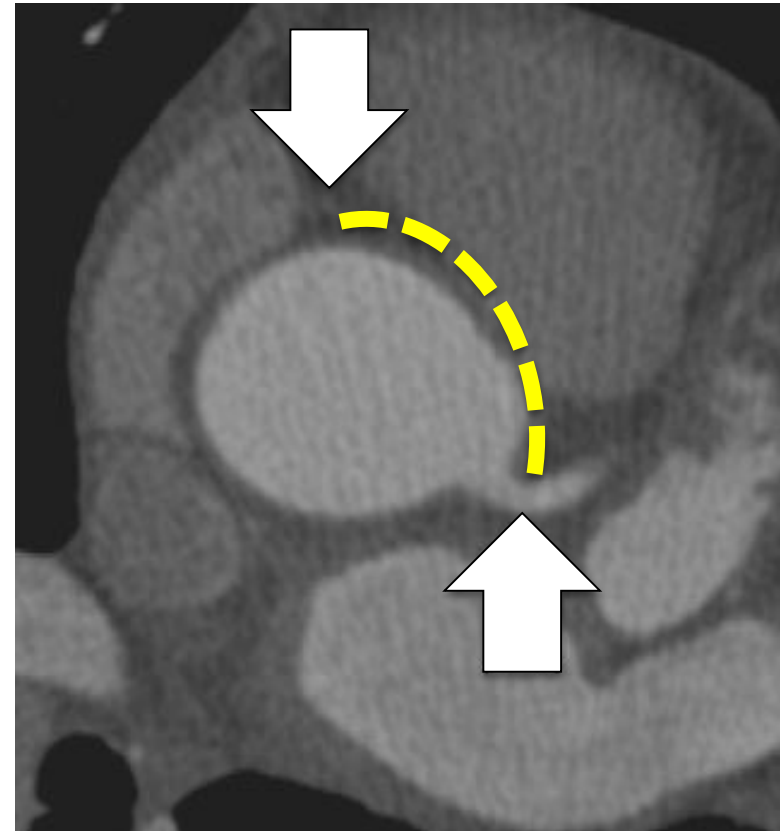
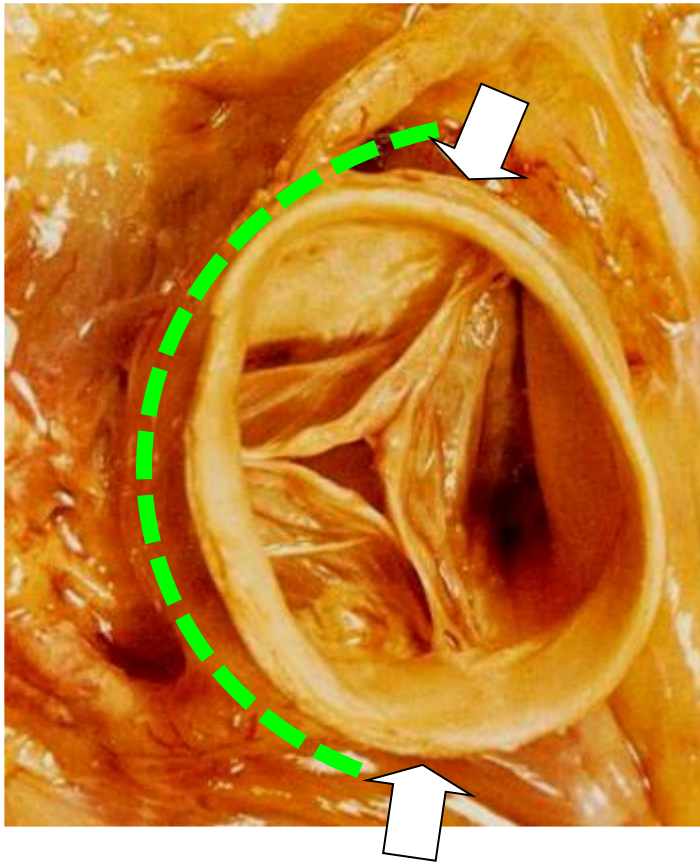
Développement de l'aorte des coronaires  
Anomalie d'origine

Développement vers l'aorte des coronaires  
Anomalie de connexion

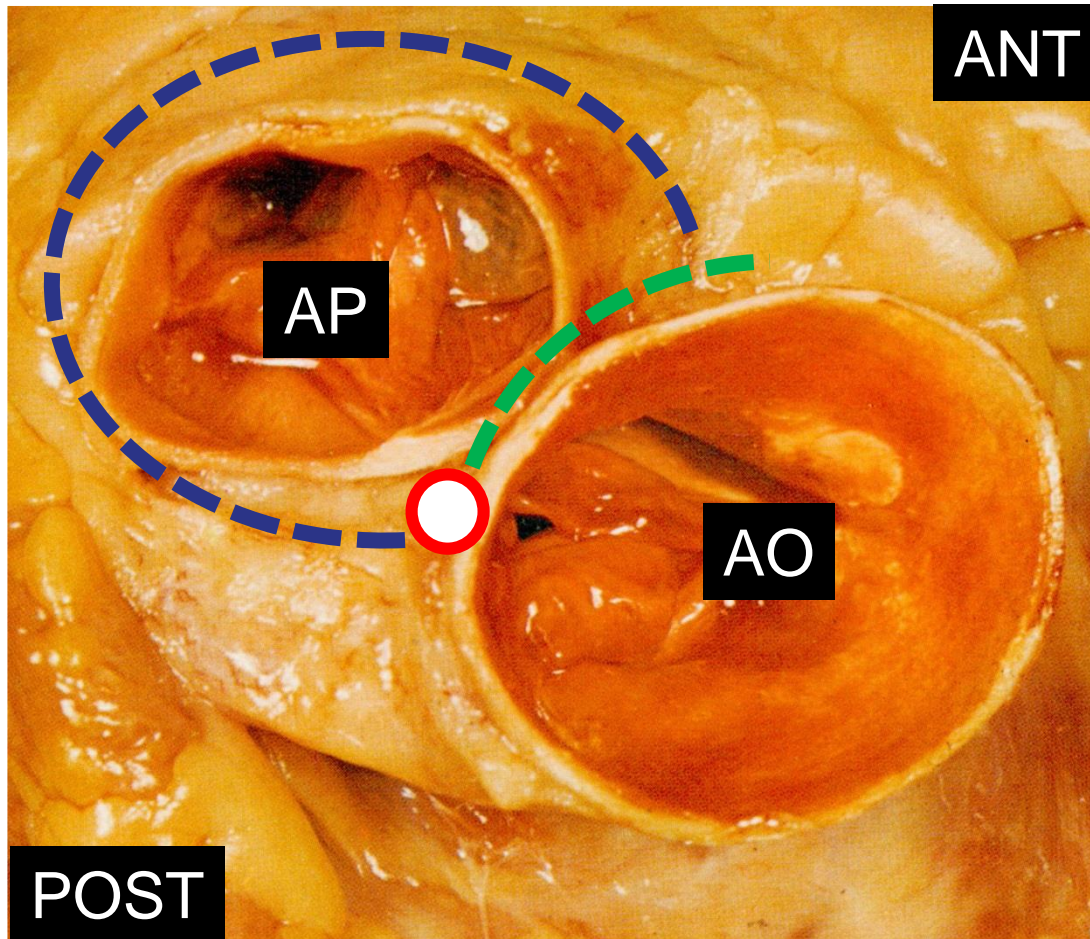


# sites habituels des connexions ectopiques droites

antérieur

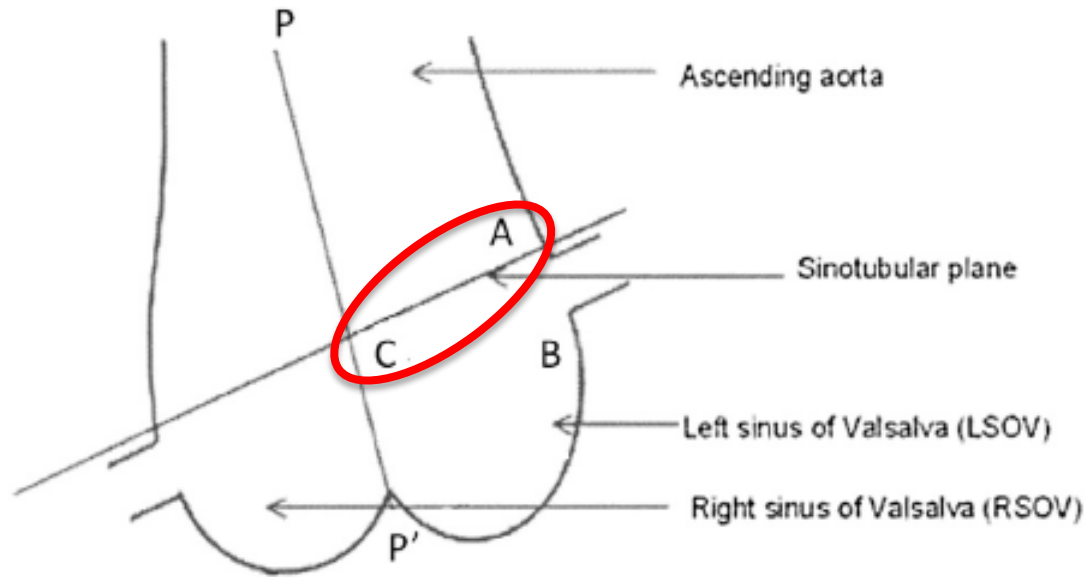


# trajets des coronaires droites ectopiques



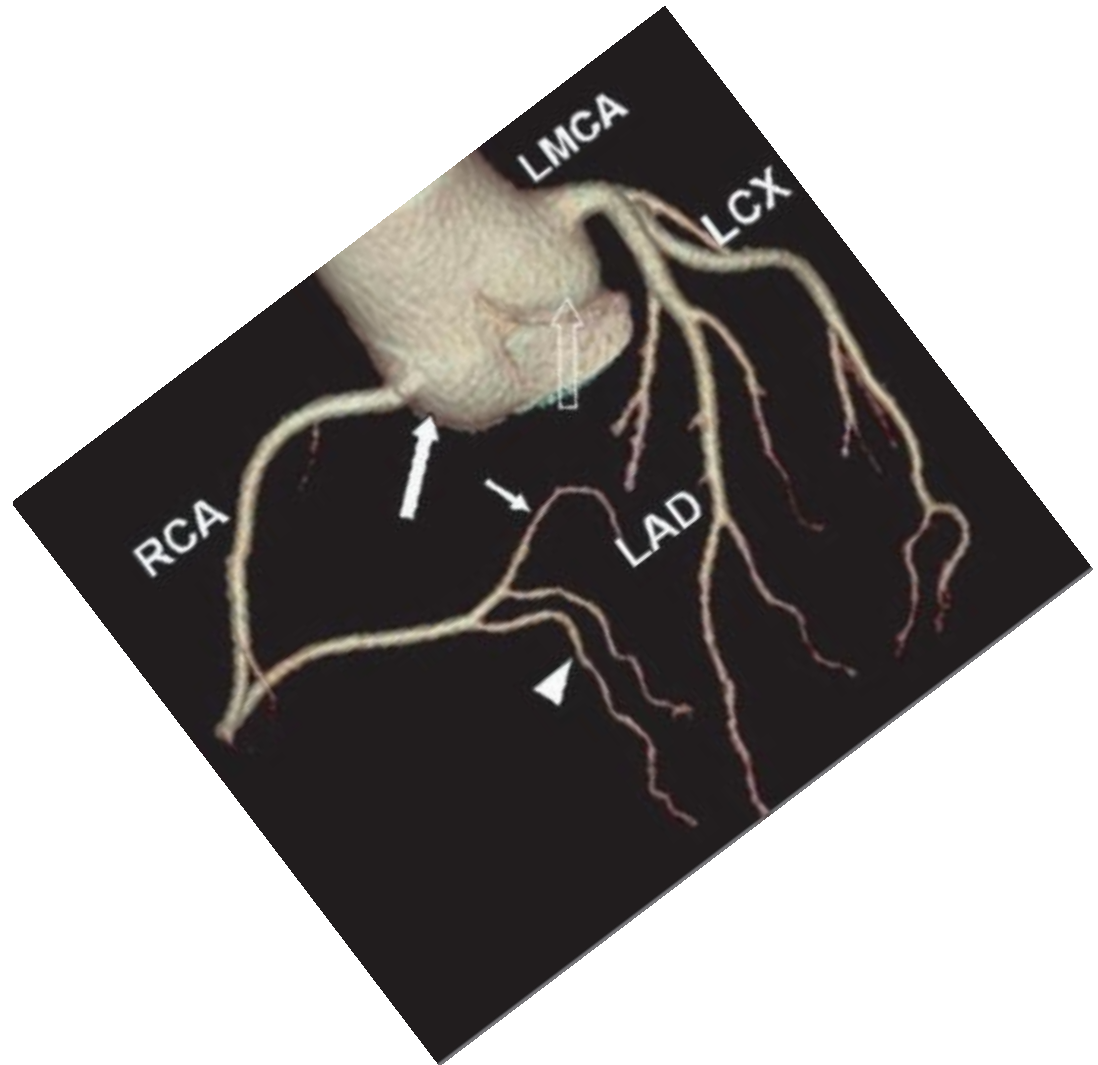
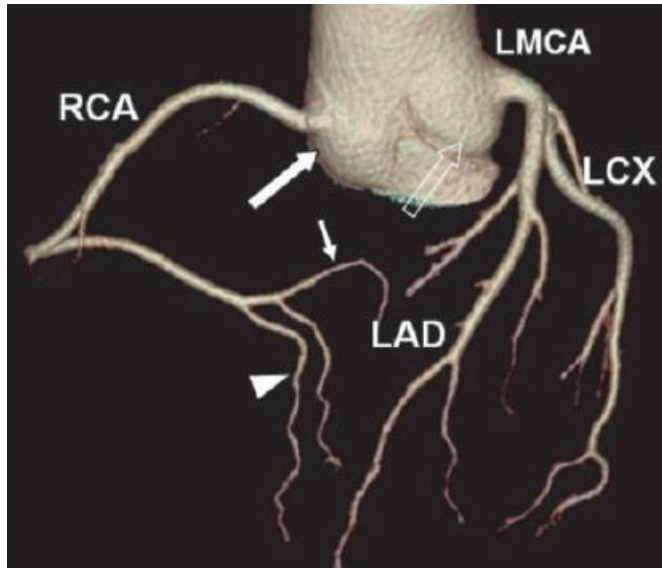
- ostium droit ectopique
- pré-pulmonaire
- pré-aortique

# sites habituels de connexion ectopique droite

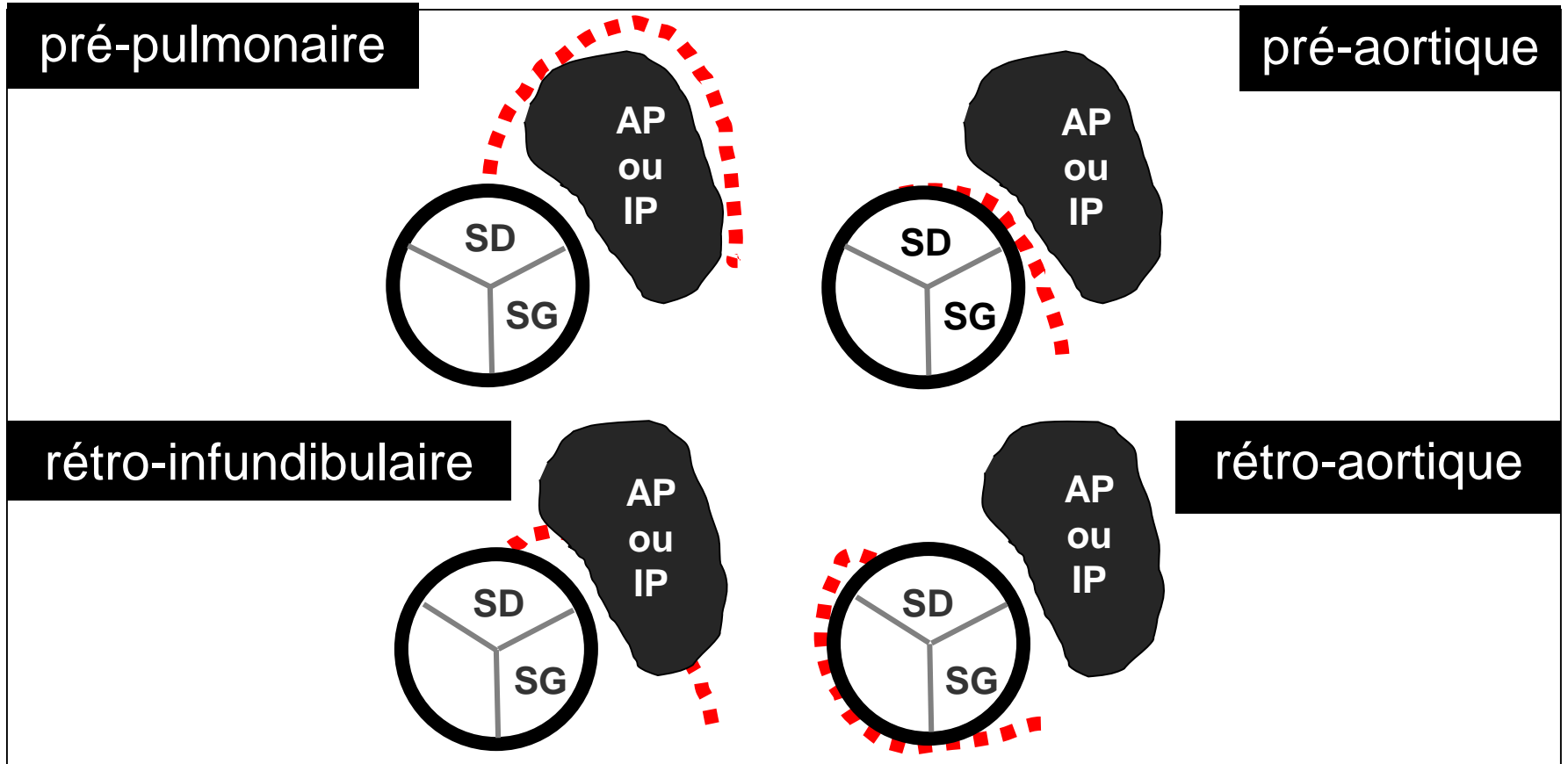


**Fig. 1 – Origin of anomalous RCA from LSOV.**  
Representative diagram of aortic root and sinuses in LAO projection. P–P' indicates a hypothetical plane running through the midline. Sites A through C represents common sites for the origin of the anomalous artery.

INDIAN HEART JOURNAL 66 (2014) 430–434

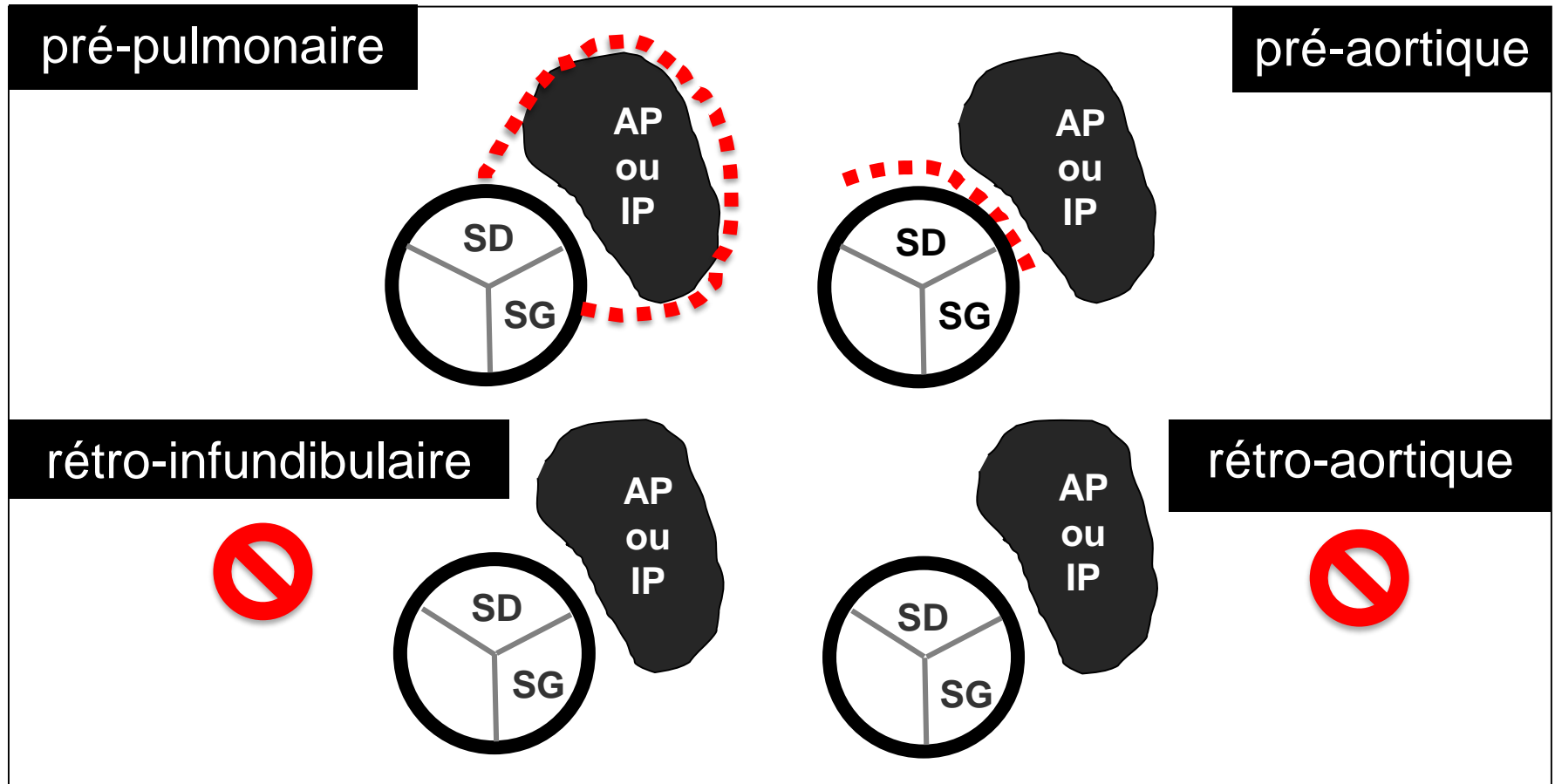


# tronc commun connecté dans sinus droit trajets possibles



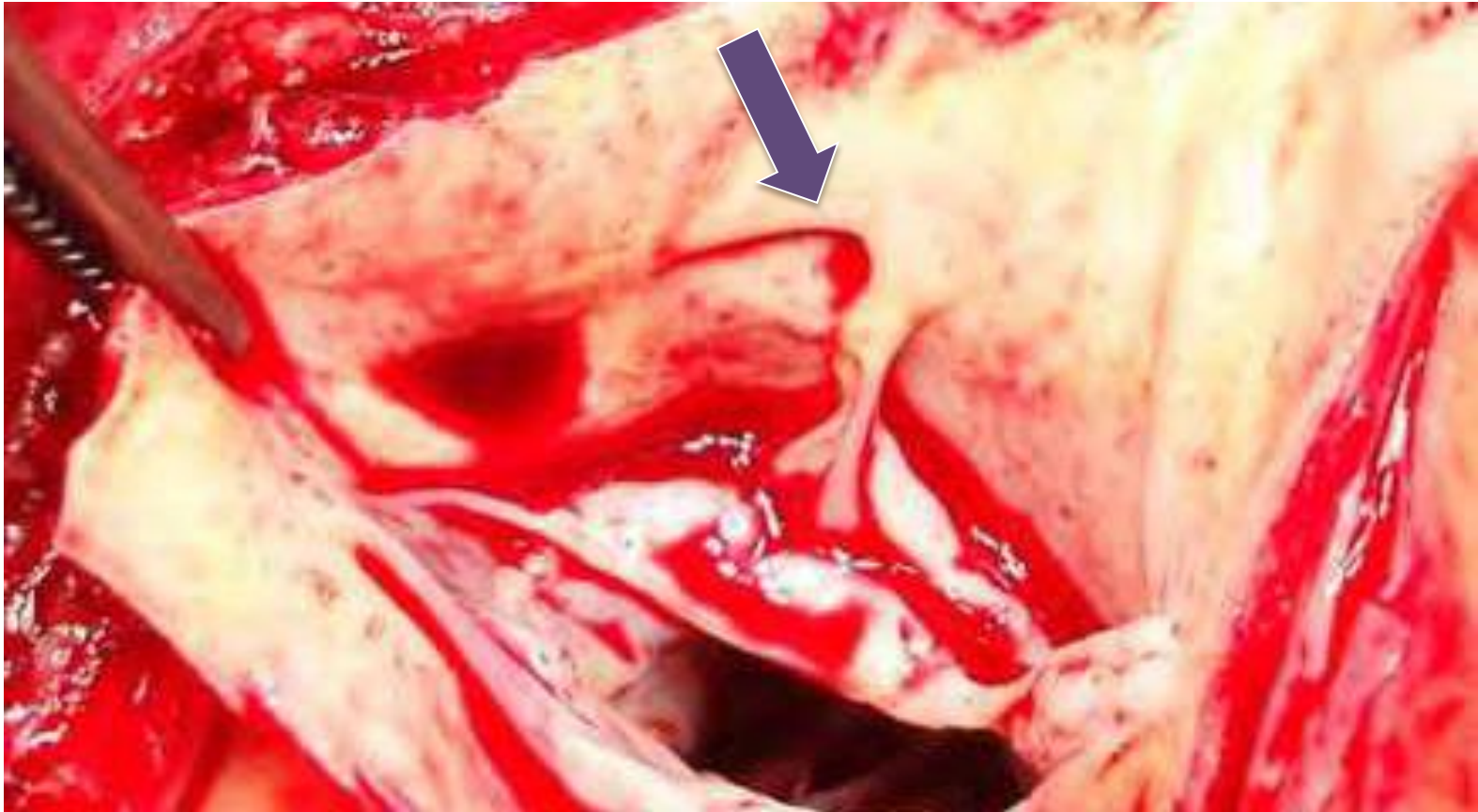
AP: artère pulmonaire, IP: infundibulum pulmonaire, SD: sinus droit, SG: sinus gauche

# coronaire droite connectée dans sinus gauche trajets possibles

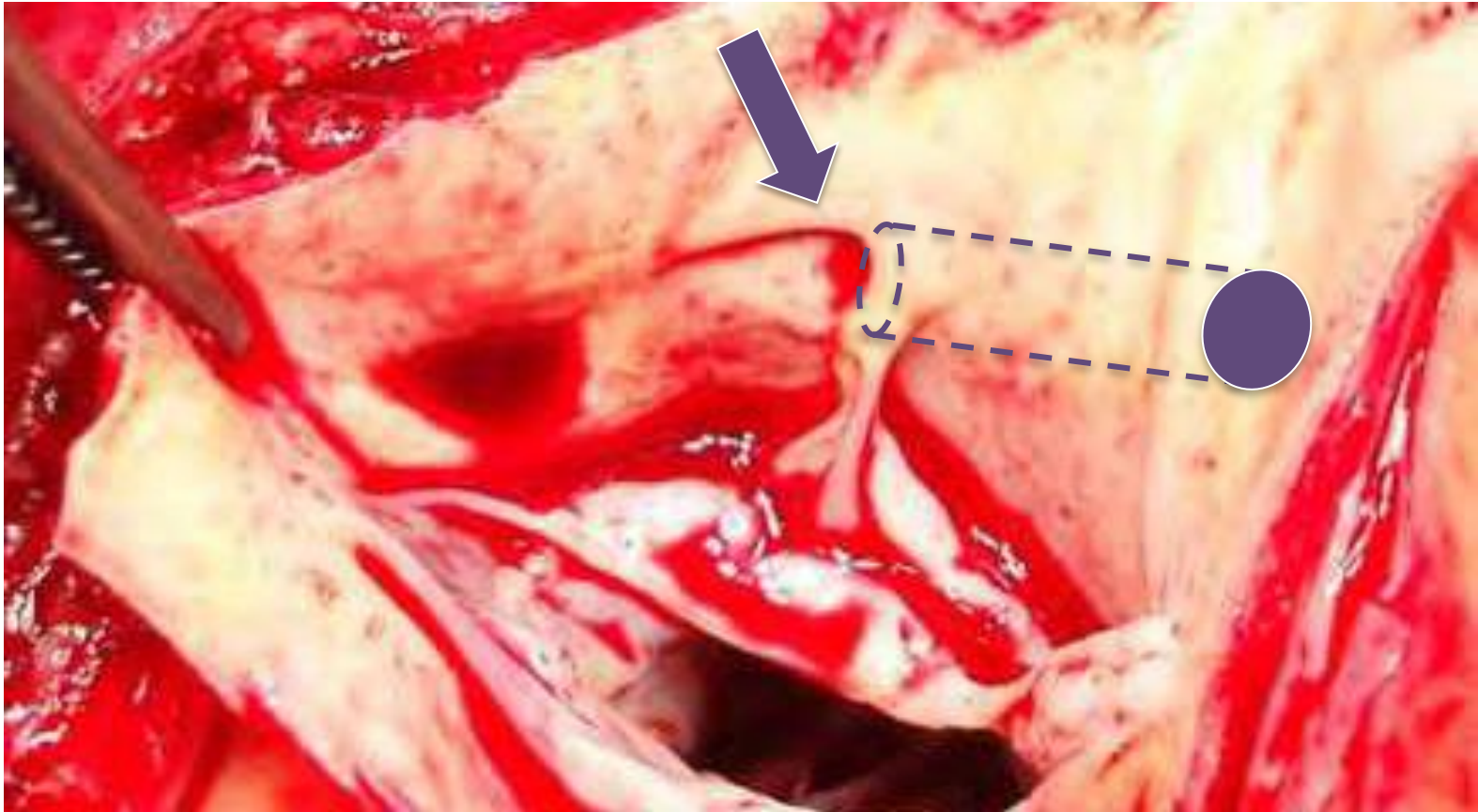


AP: artère pulmonaire, IP: infundibulum pulmonaire, SD: sinus droit, SG: sinus gauche

# connexion ectopique droite dans sinus gauche

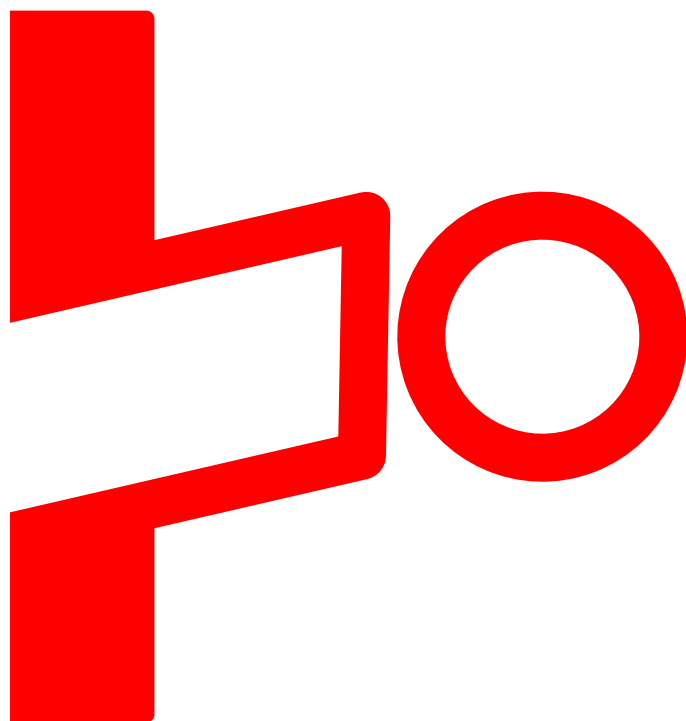


# connexion ectopique droite dans sinus gauche

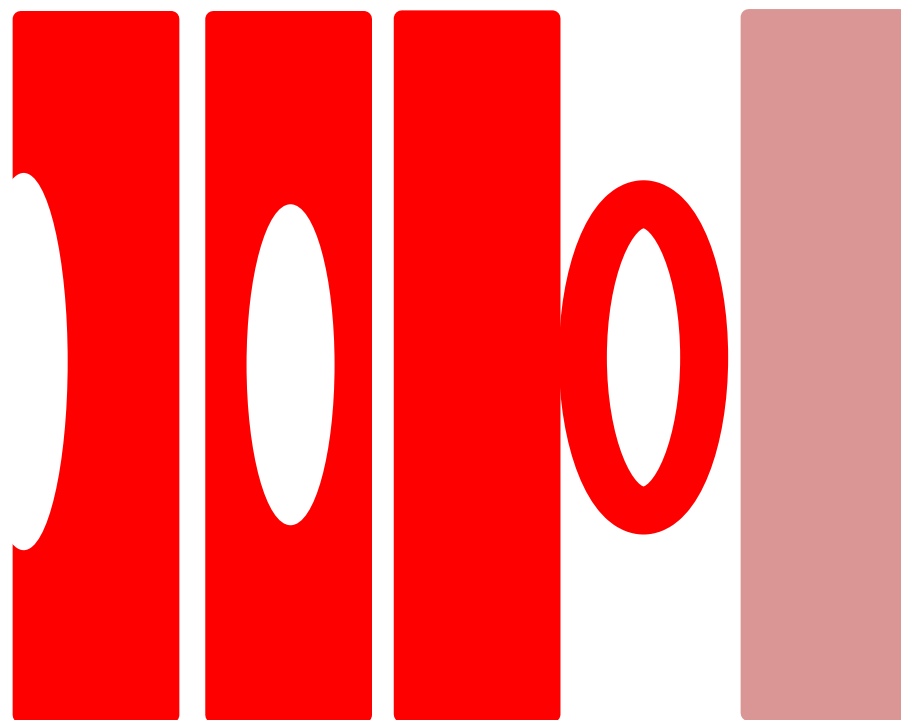




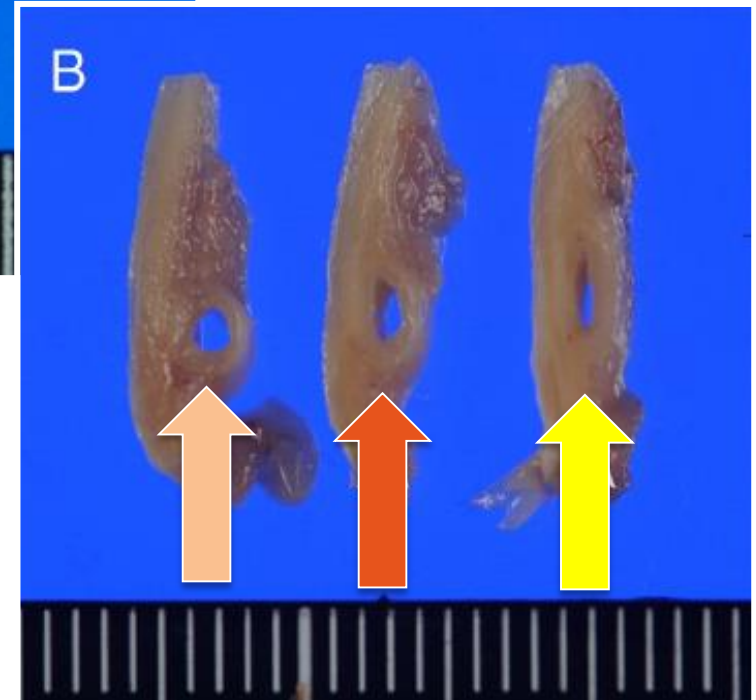
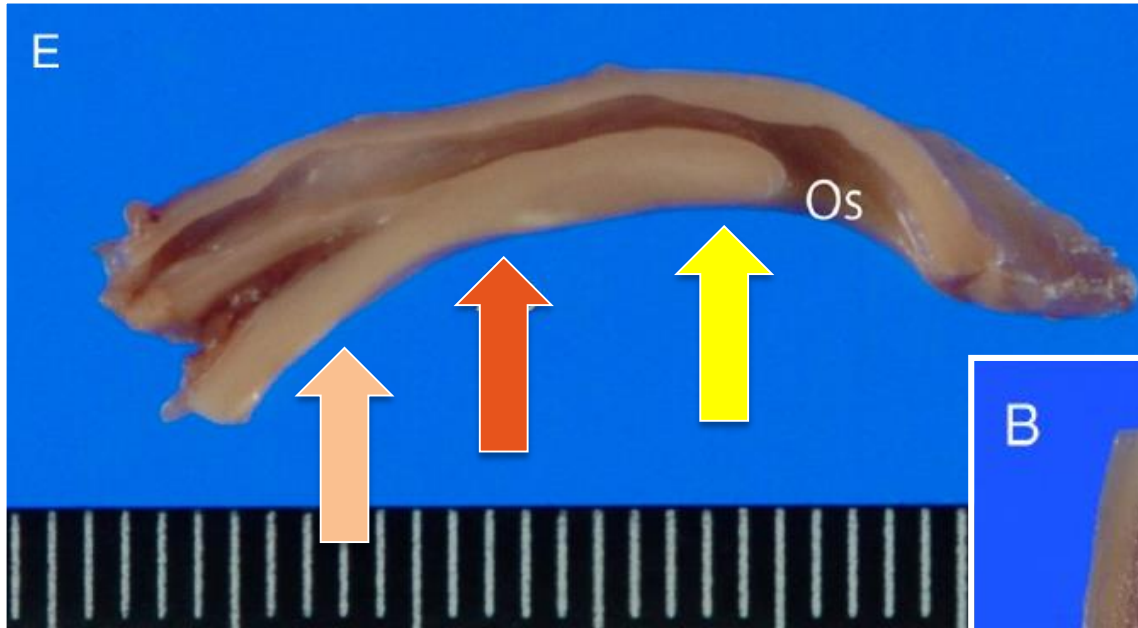
connexion extramurale



connexion intramurale

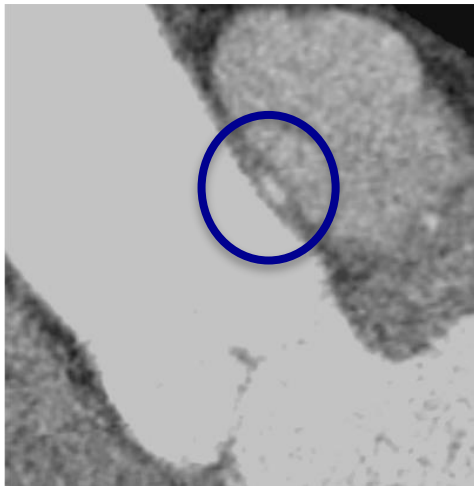
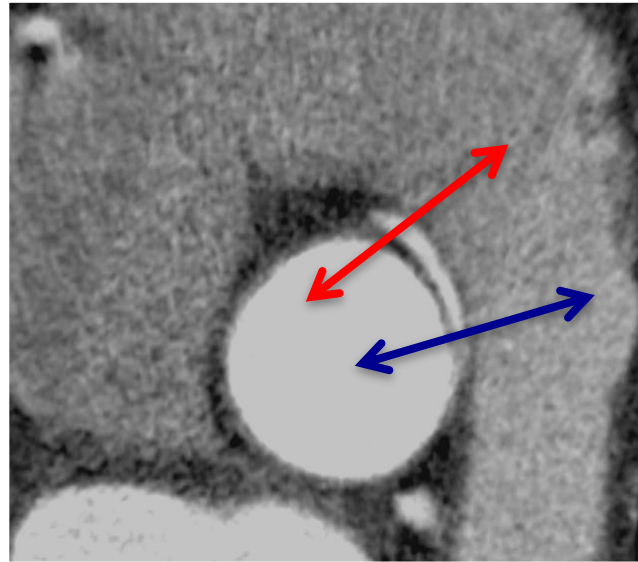


# ANOCOR droite avec passage intramural

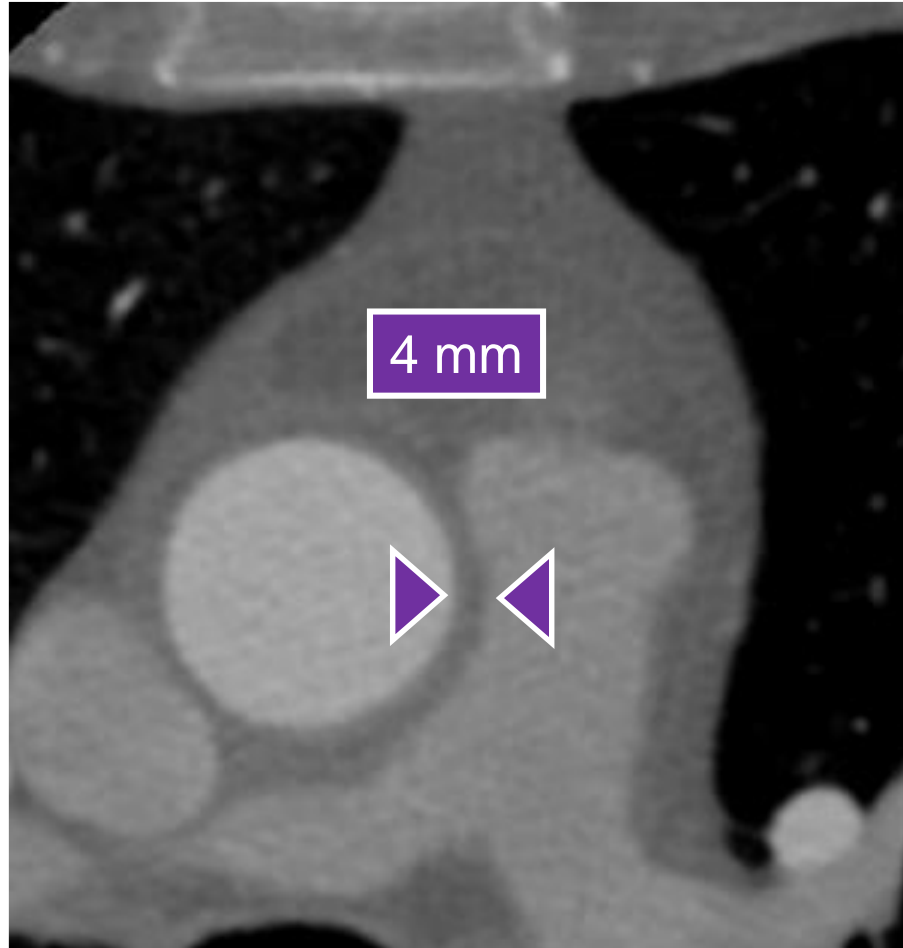


Hata Y et al. Cardiovasc Pathol 2014

# ANOCOR droite avec passage intramural

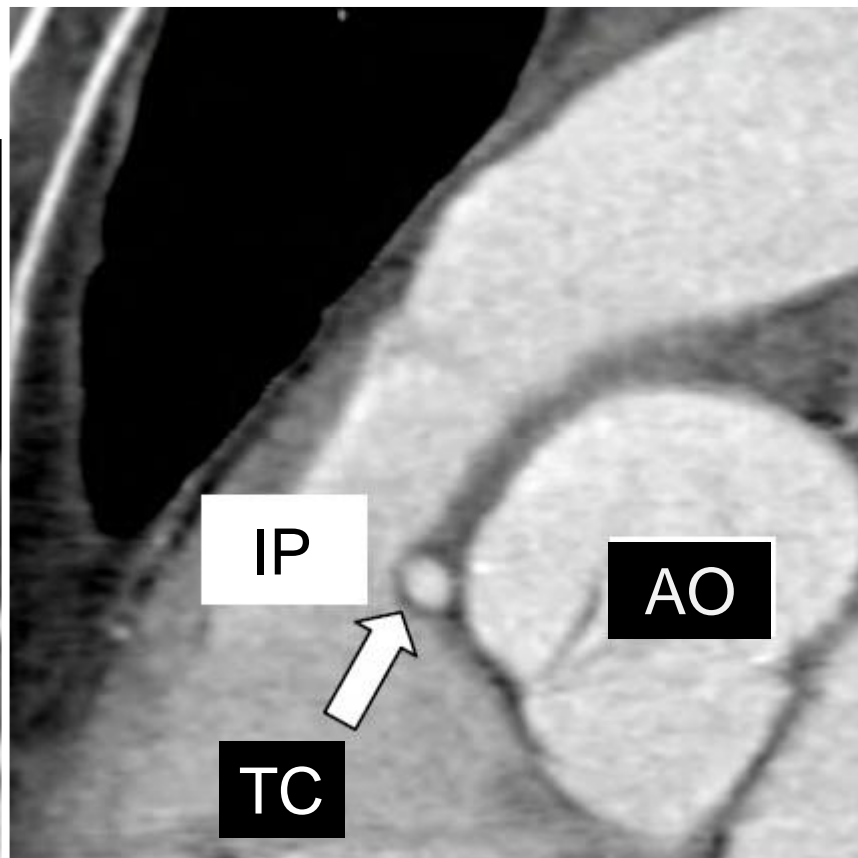
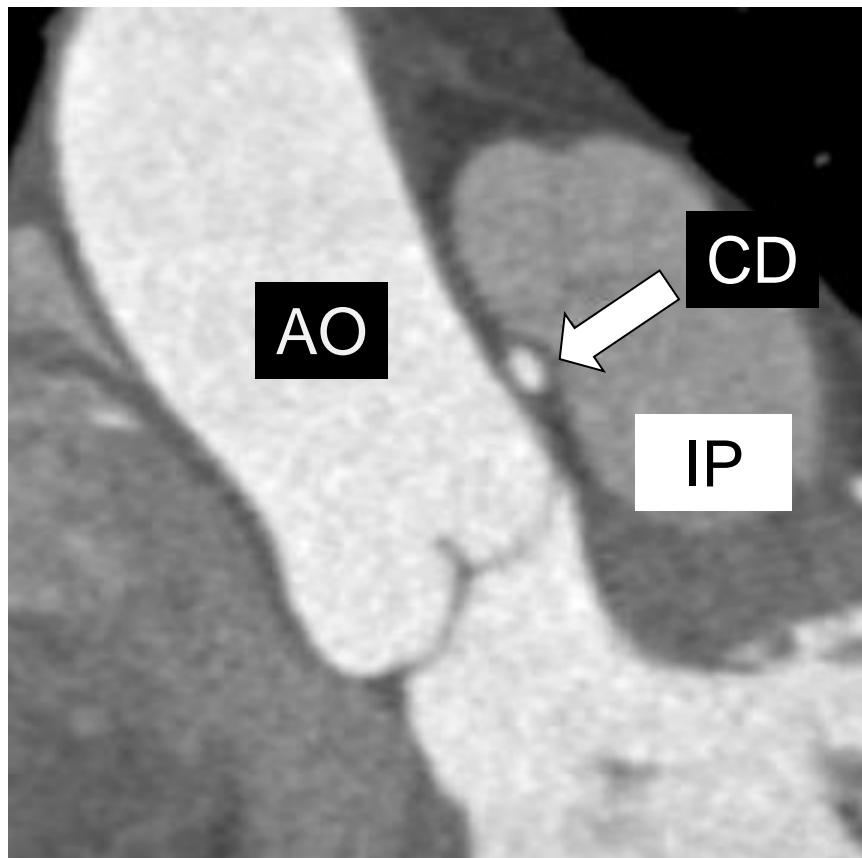


# espace interartériel (aorte-artère pulmonaire)



# anomalies de connexion proximale coronaire

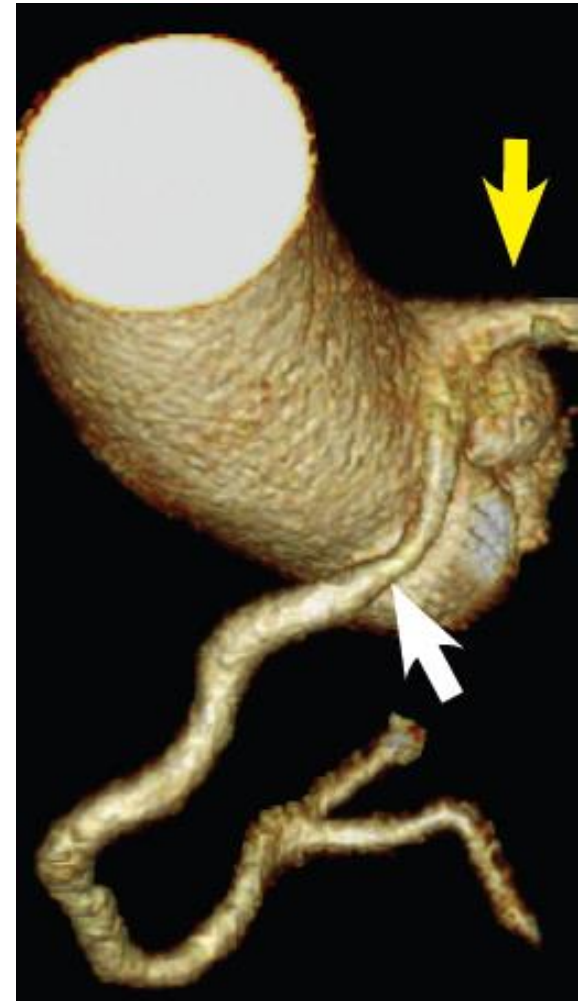
## trajet préaortique



## 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

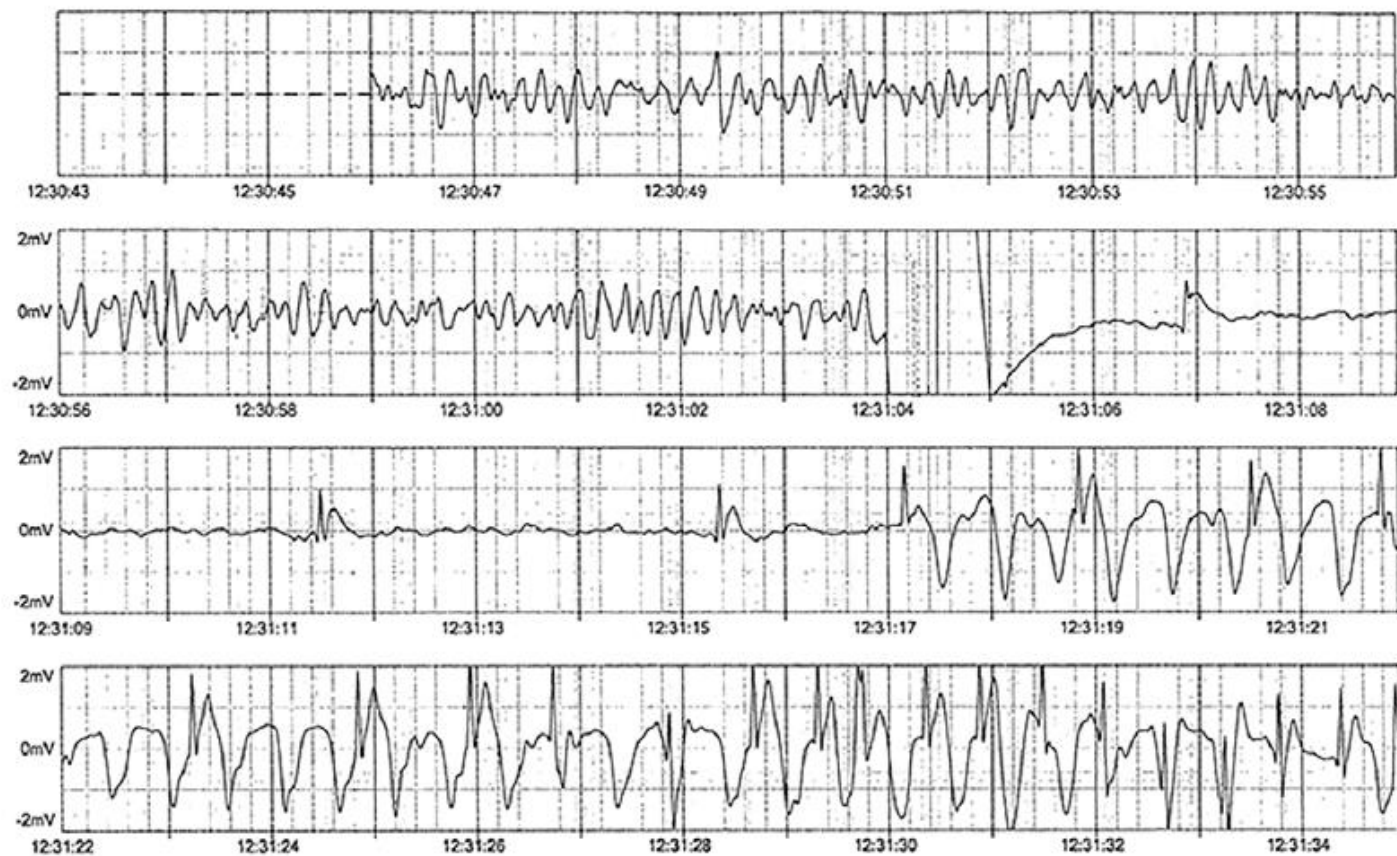
- premier événement cardiovasculaire : **souvent**
- population en bonne santé et jeune : **généralement**
- lien avec une activité physique/sportive : **net**

**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\*

<b>artère coronaire</b>	<b>%</b>
tronc commun	12.0
artère interventriculaire antérieure	5.5
artère circonflexe	47.5
artère coronaire droite	<b>33.0</b>
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

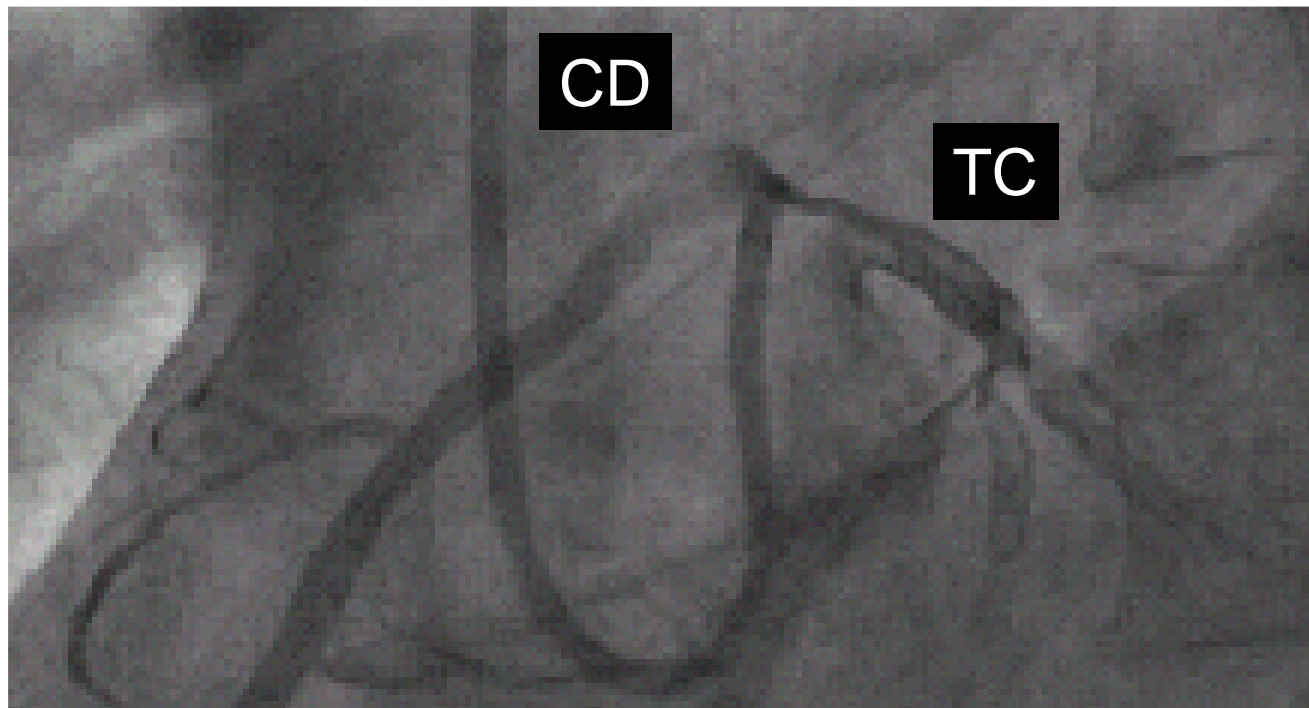
*Circulation* December 2, 2008

#### 8.5. Recommendations for Congenital Coronary Anomalies of Ectopic Arterial Origin

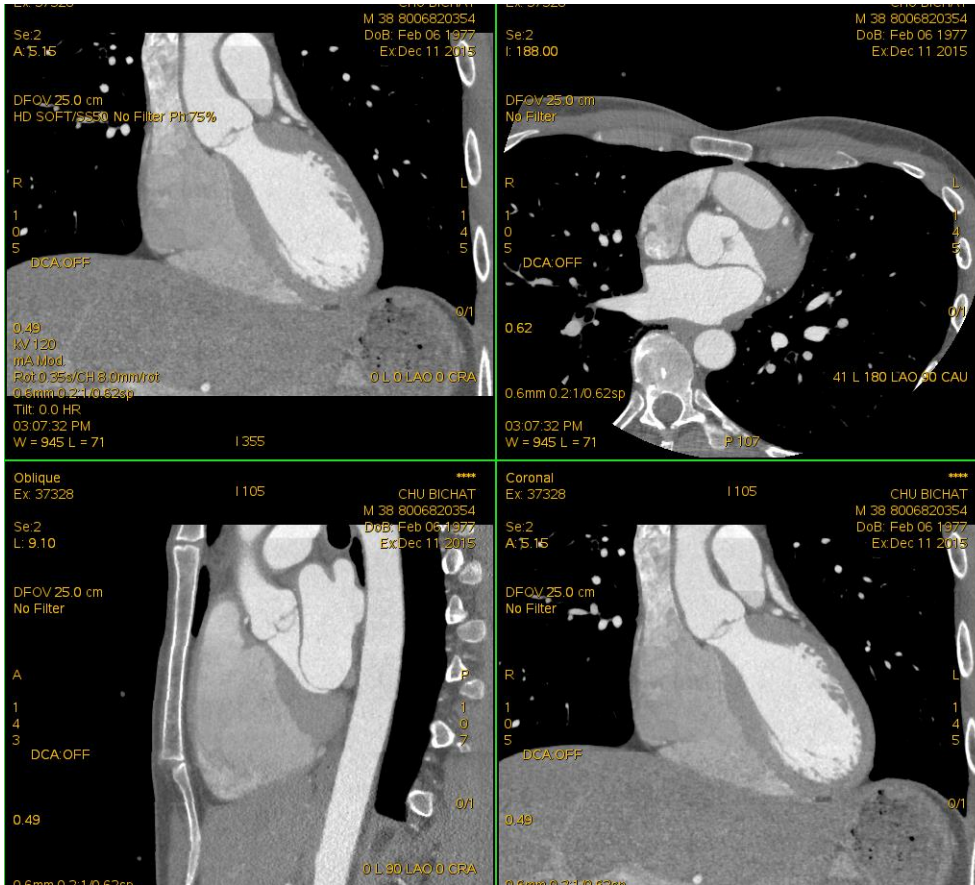
##### *Class I*

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*)

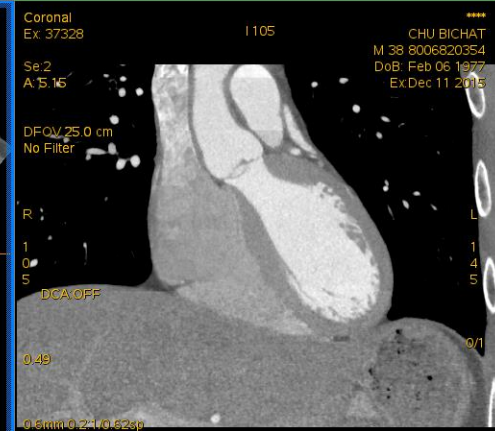
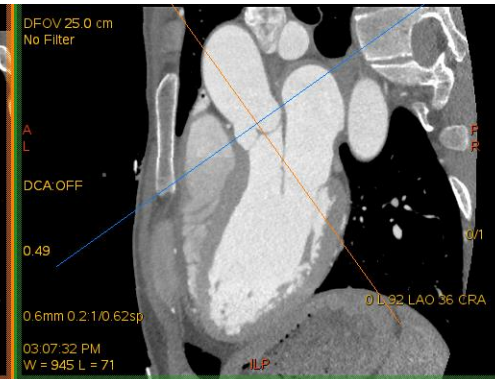
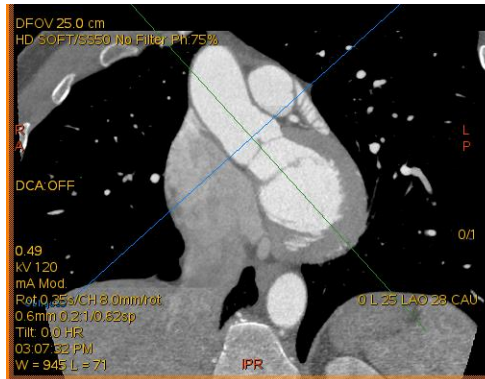
connexion droite dans le sinus controlatéral



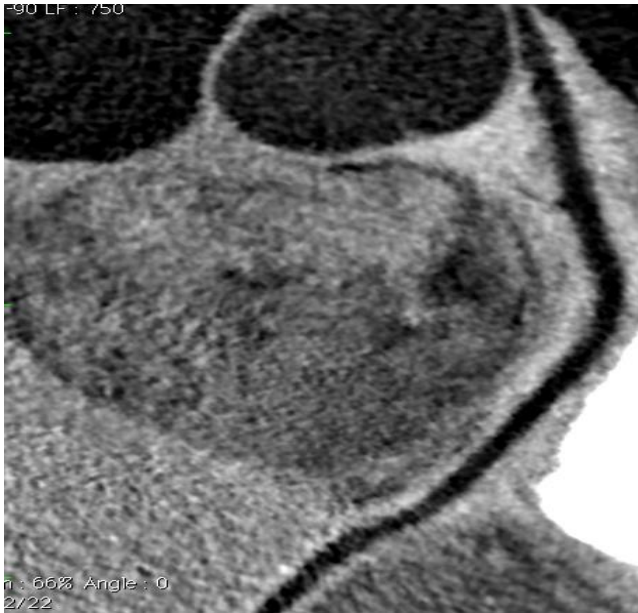
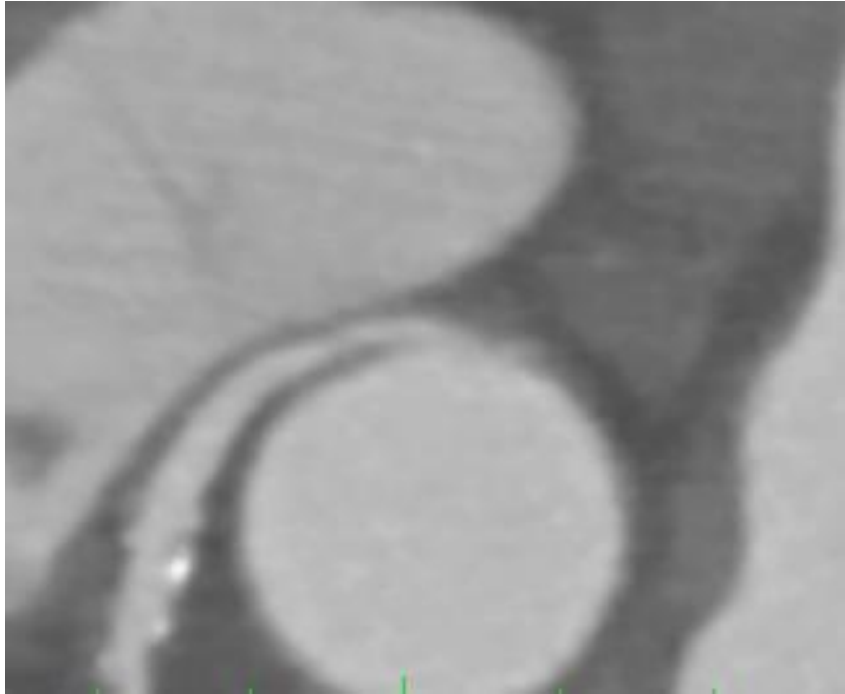
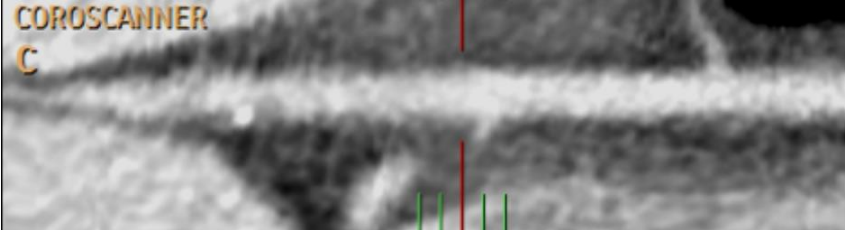
# imagerie dans la plan axial strict



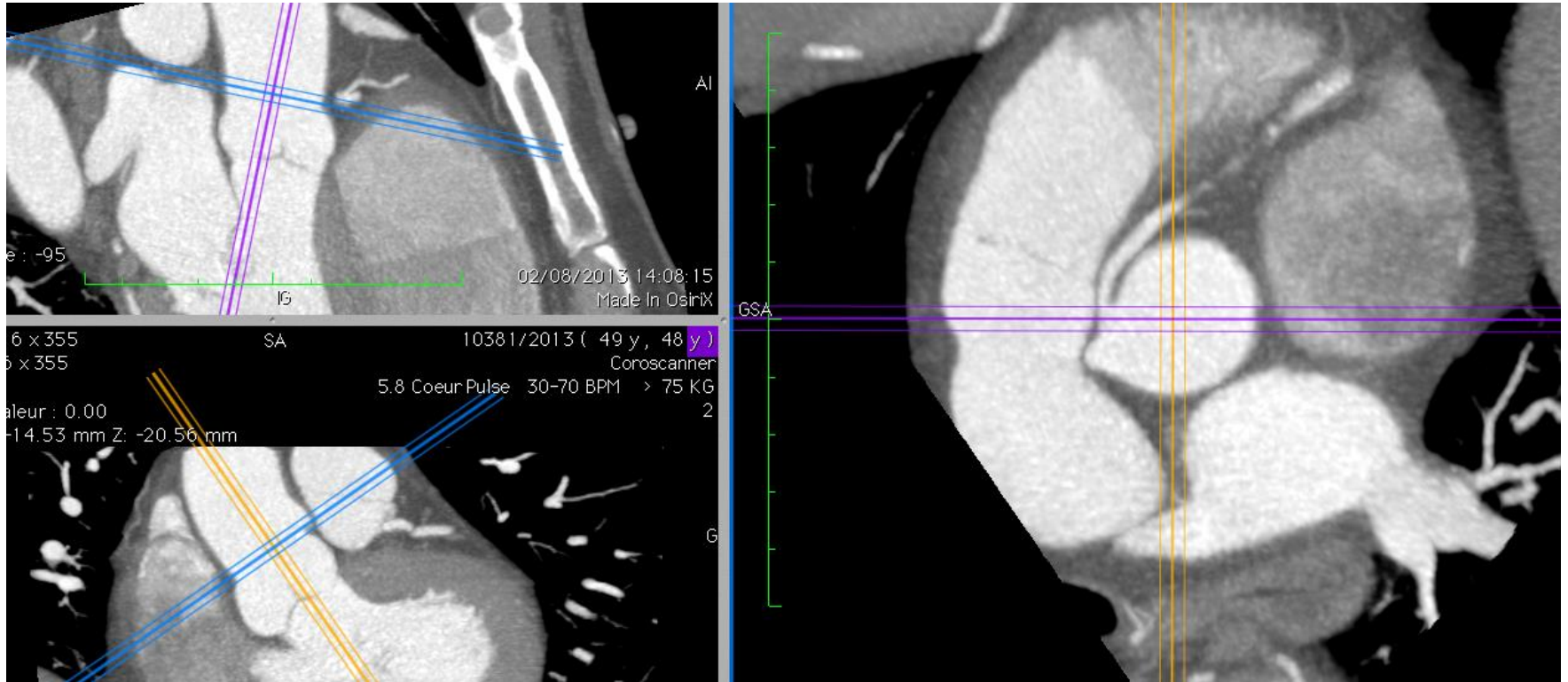
# imagerie dans le plan de l'anneau aortique



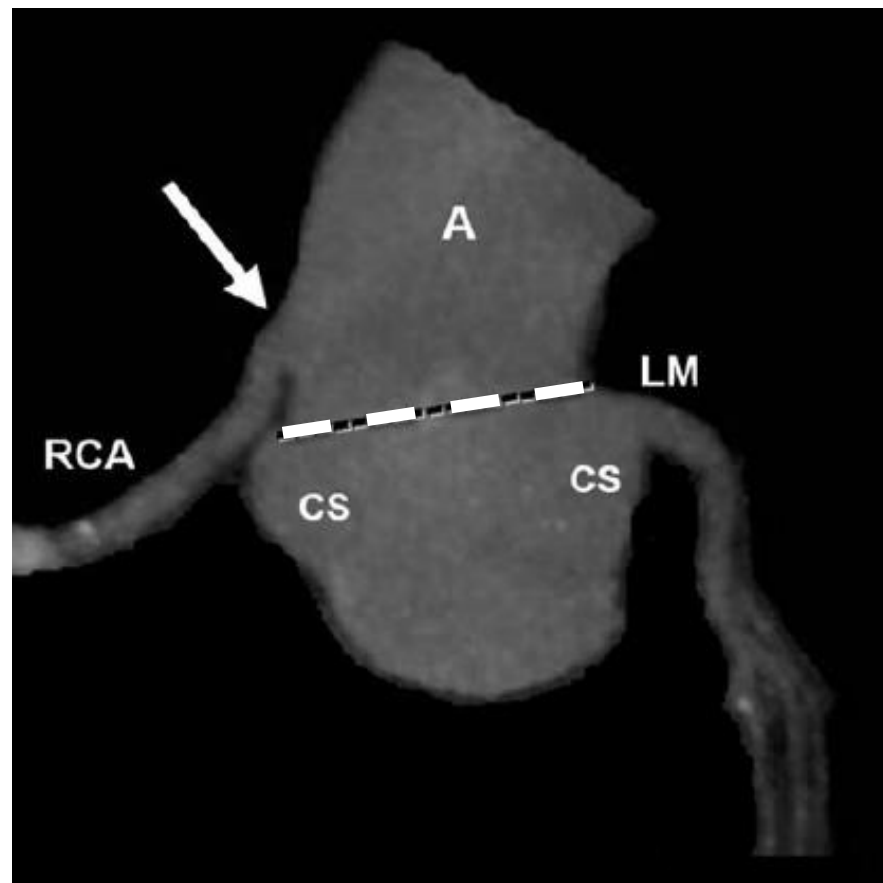
# connexion droite dans le sinus controlatéral



# connexion droite dans le sinus controlatéral



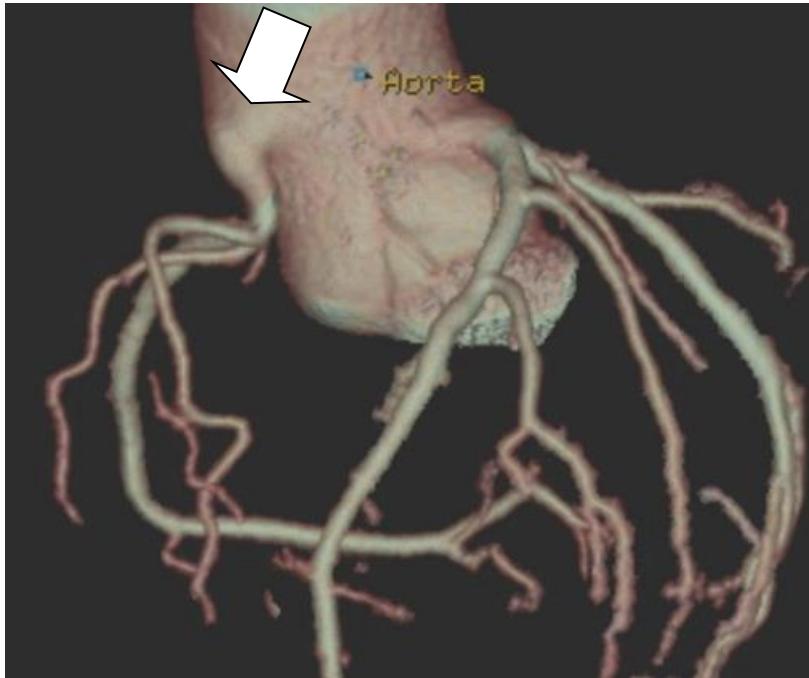
# connexion aortique haute



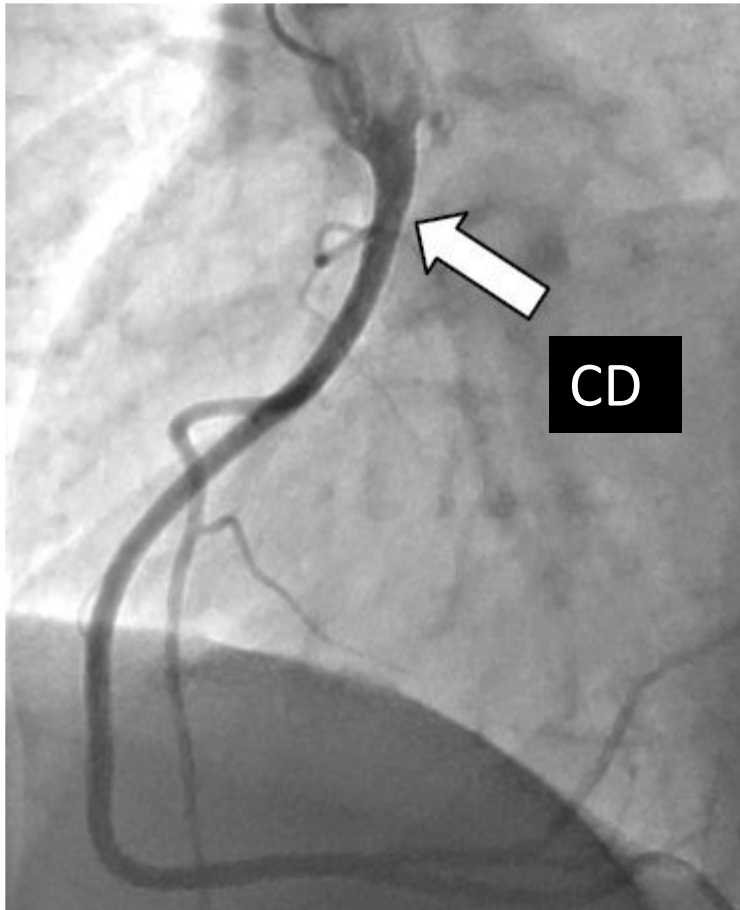
10 mm au dessus  
jonction sinotubulaire



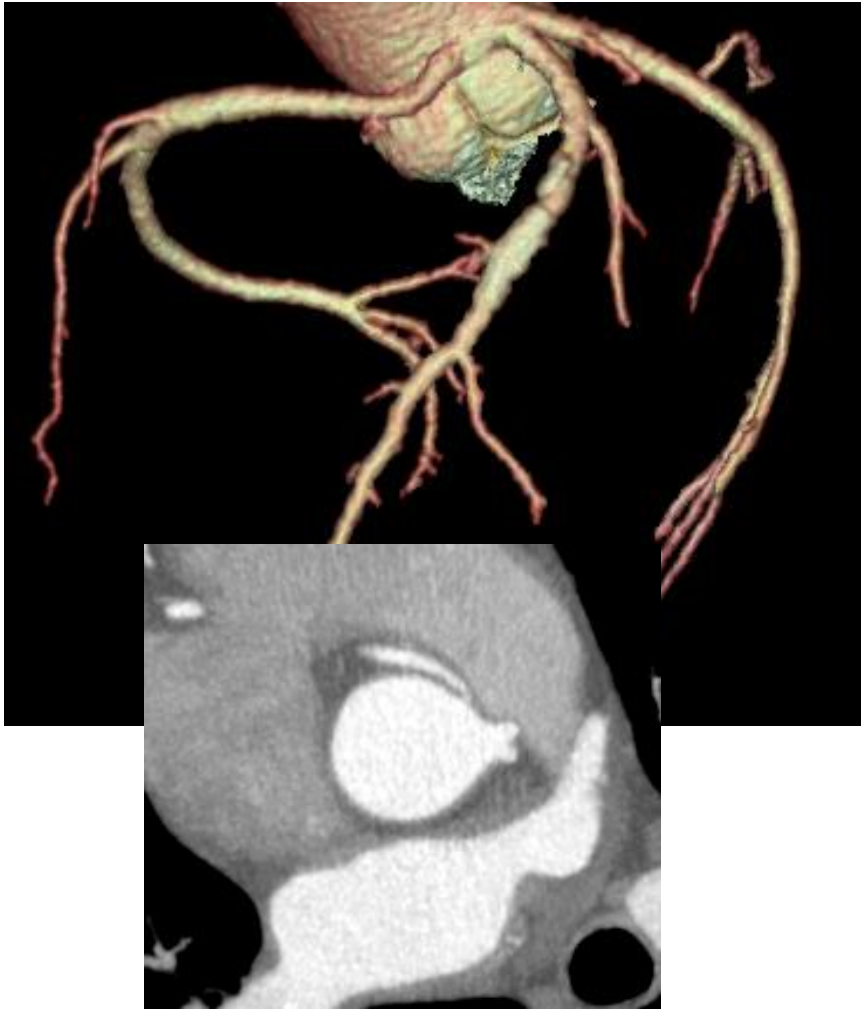
# connexion aortique haute coronaire droite



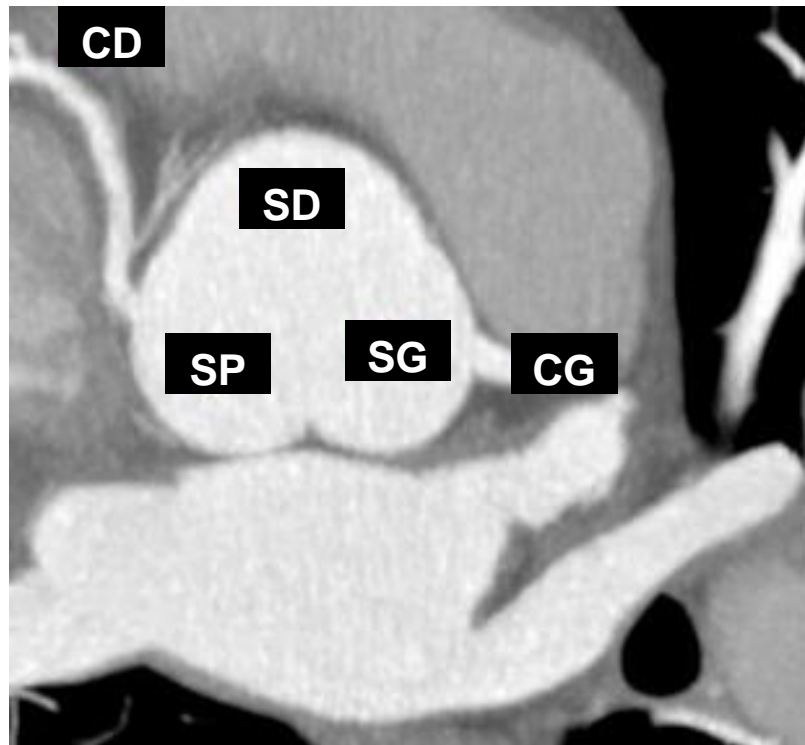
# connexion aortique haute



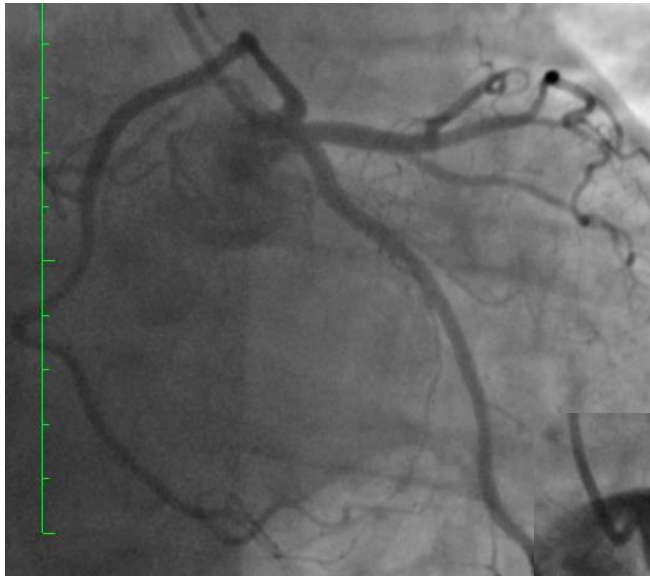
# connexion droite dans le sinus controlatéral



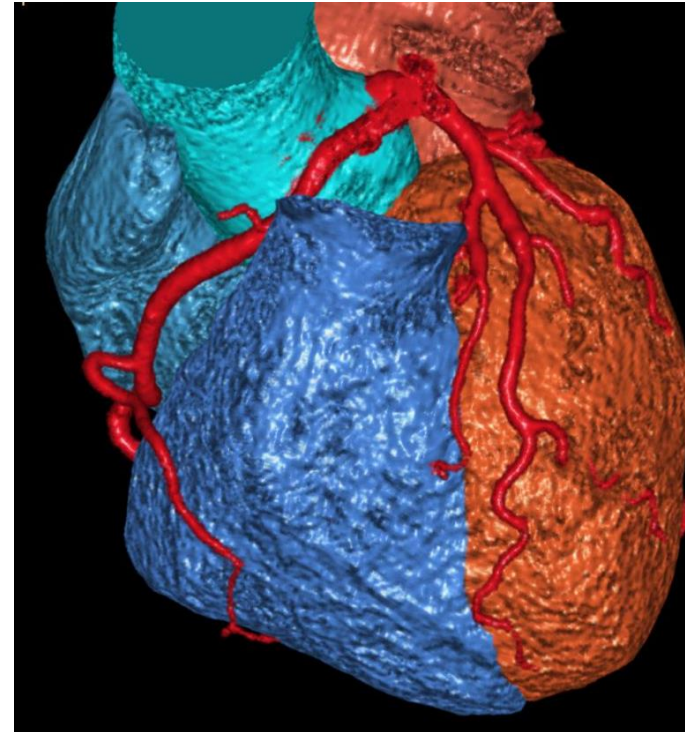
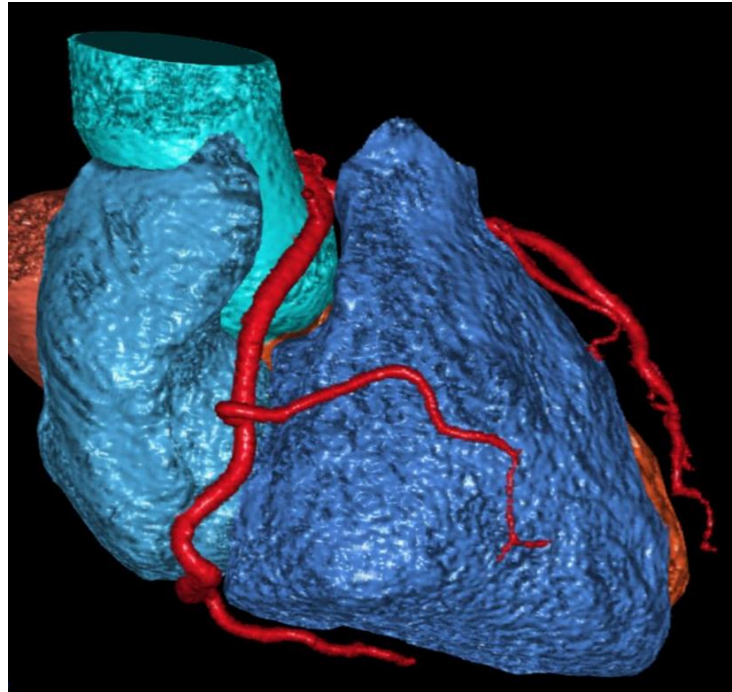
# connexion droite dans le sinus postérieur



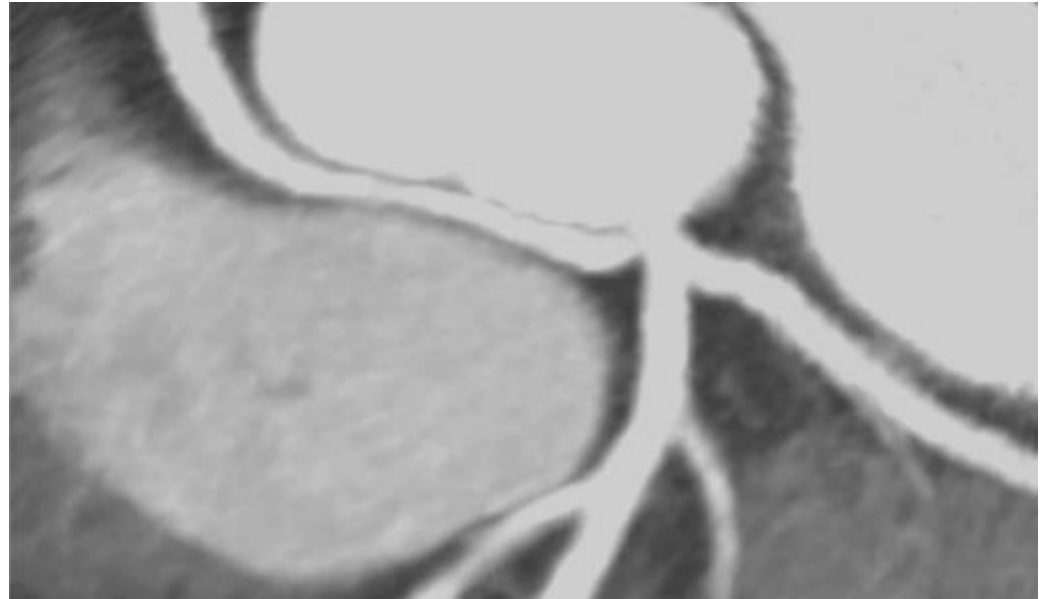
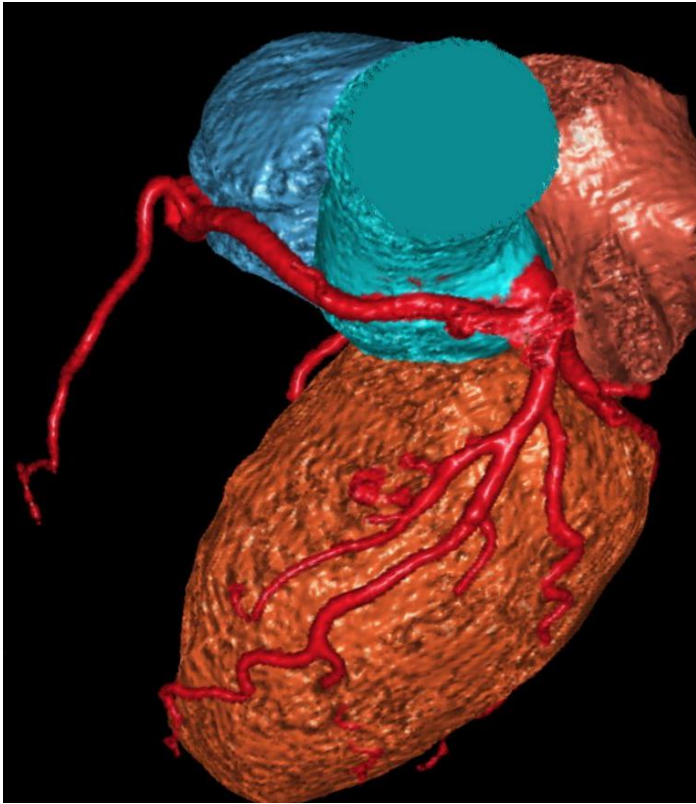
# connexion droite dans tronc commun



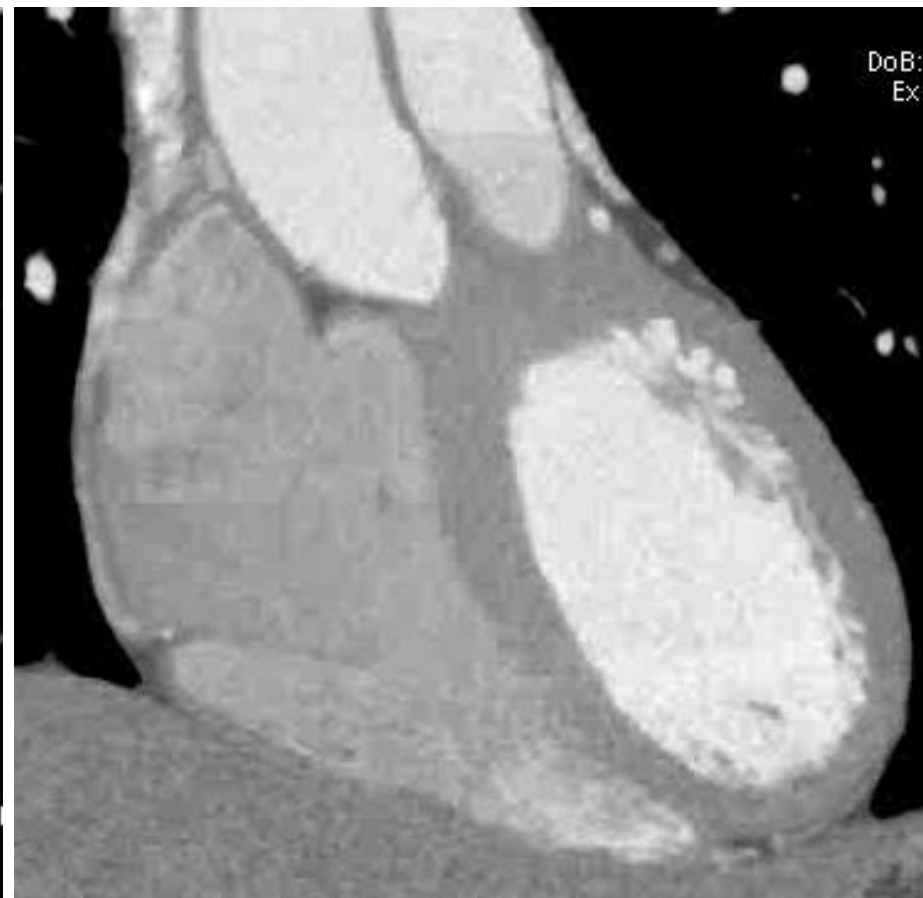
# connexion droite dans tronc commun avec trajet préaortique



# connexion droite dans tronc commun avec trajet préaortique

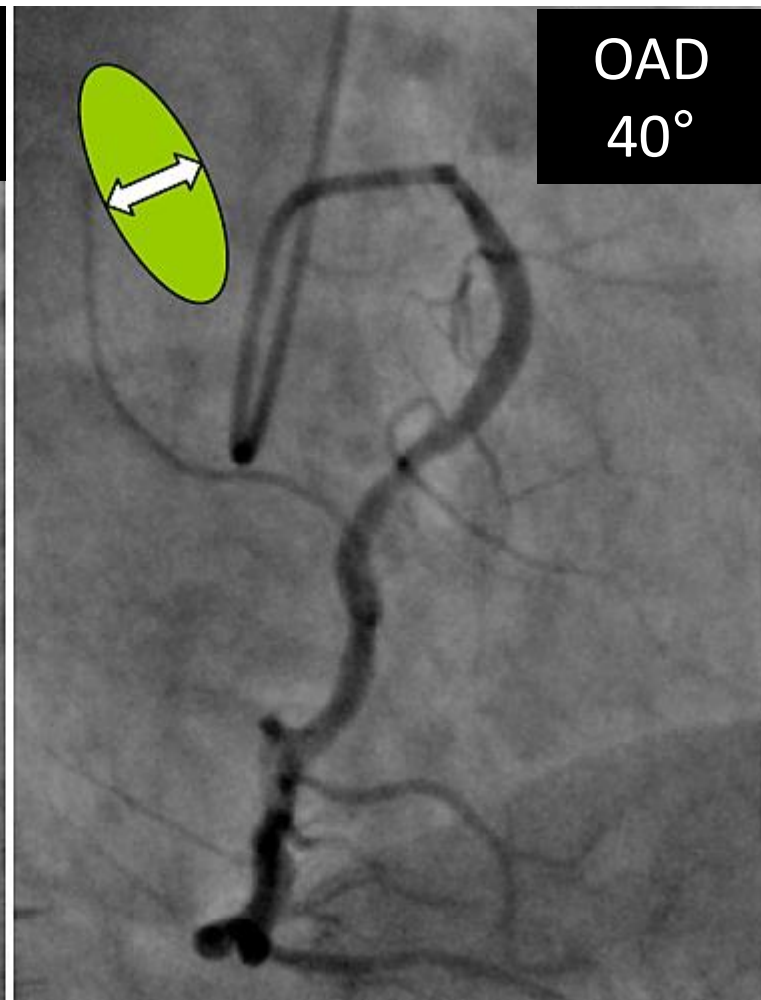
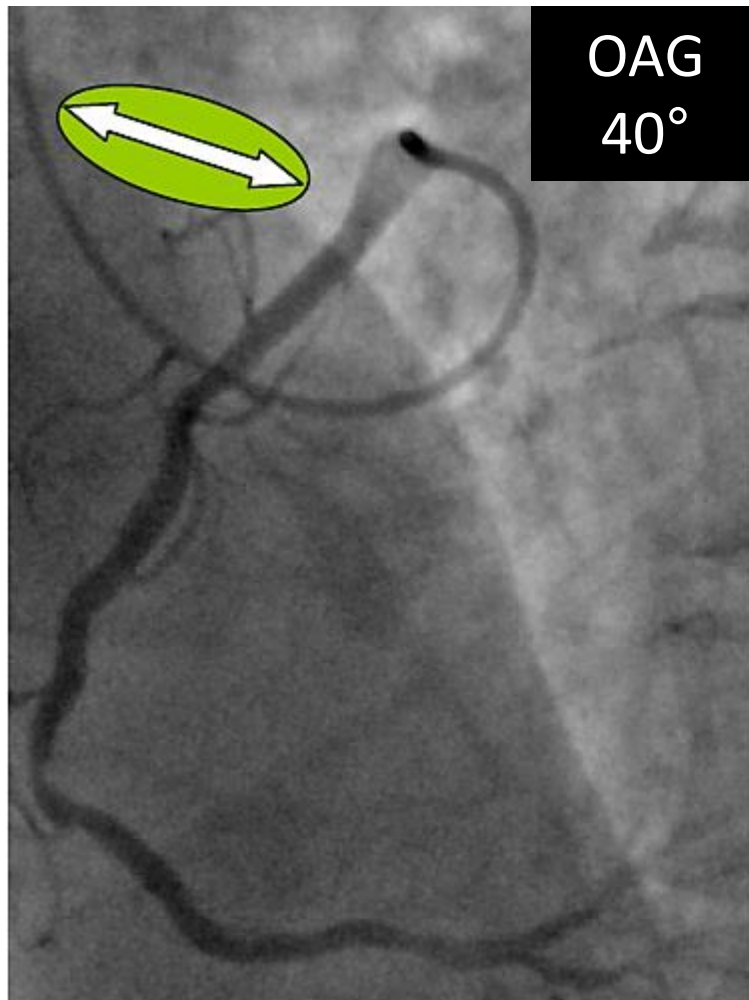


# connexion droite dans le sinus controlatéral

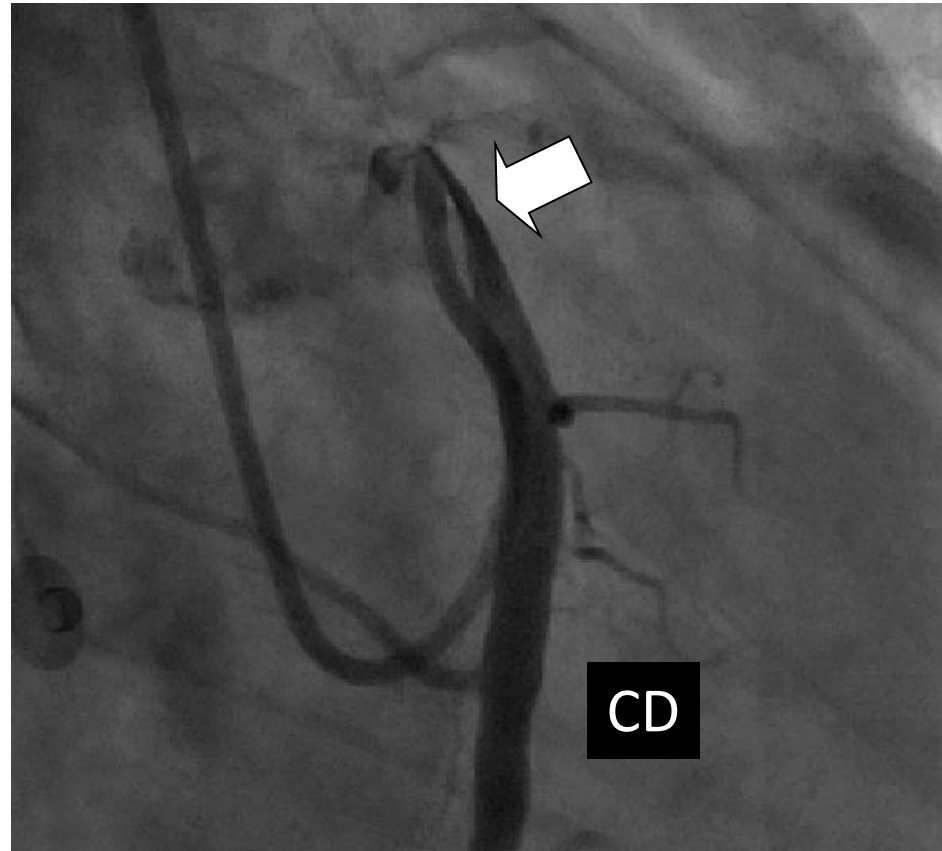




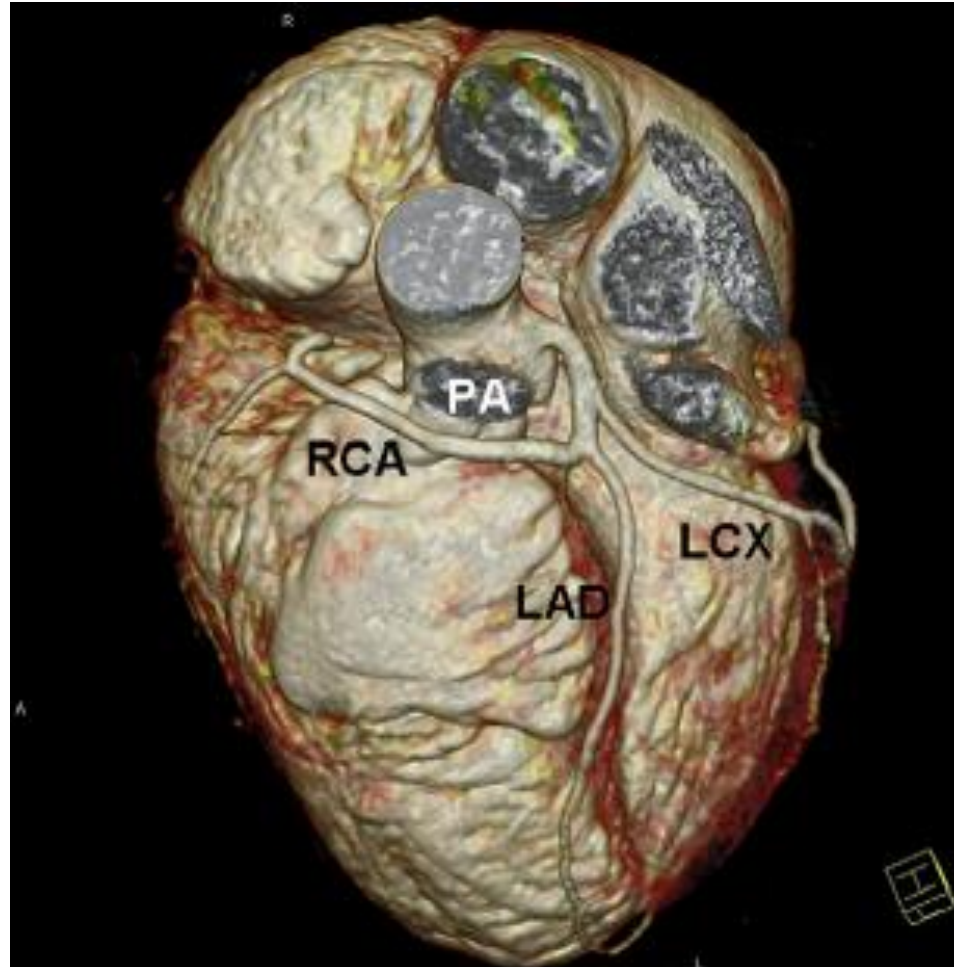
# coronaire droite avec probable passage intramural



connexion droite dans le sinus controlatéral  
déformation coronaire



# connexion droite dans artère interventriculaire antérieure avec trajet prépulmonaire

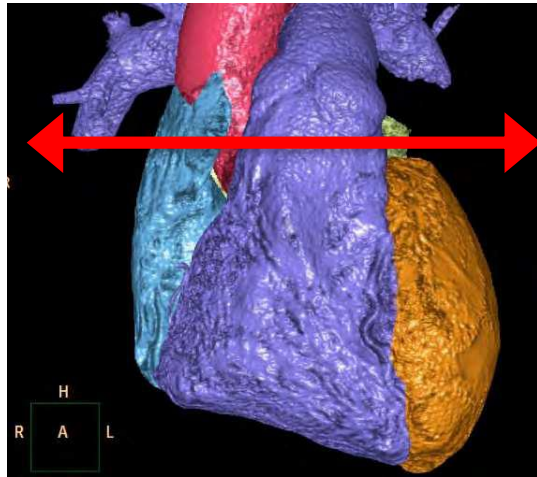


# Techniques de cathétérisme

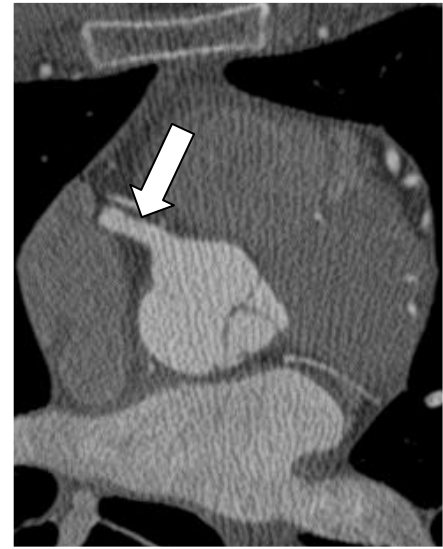
# inversion des coupes



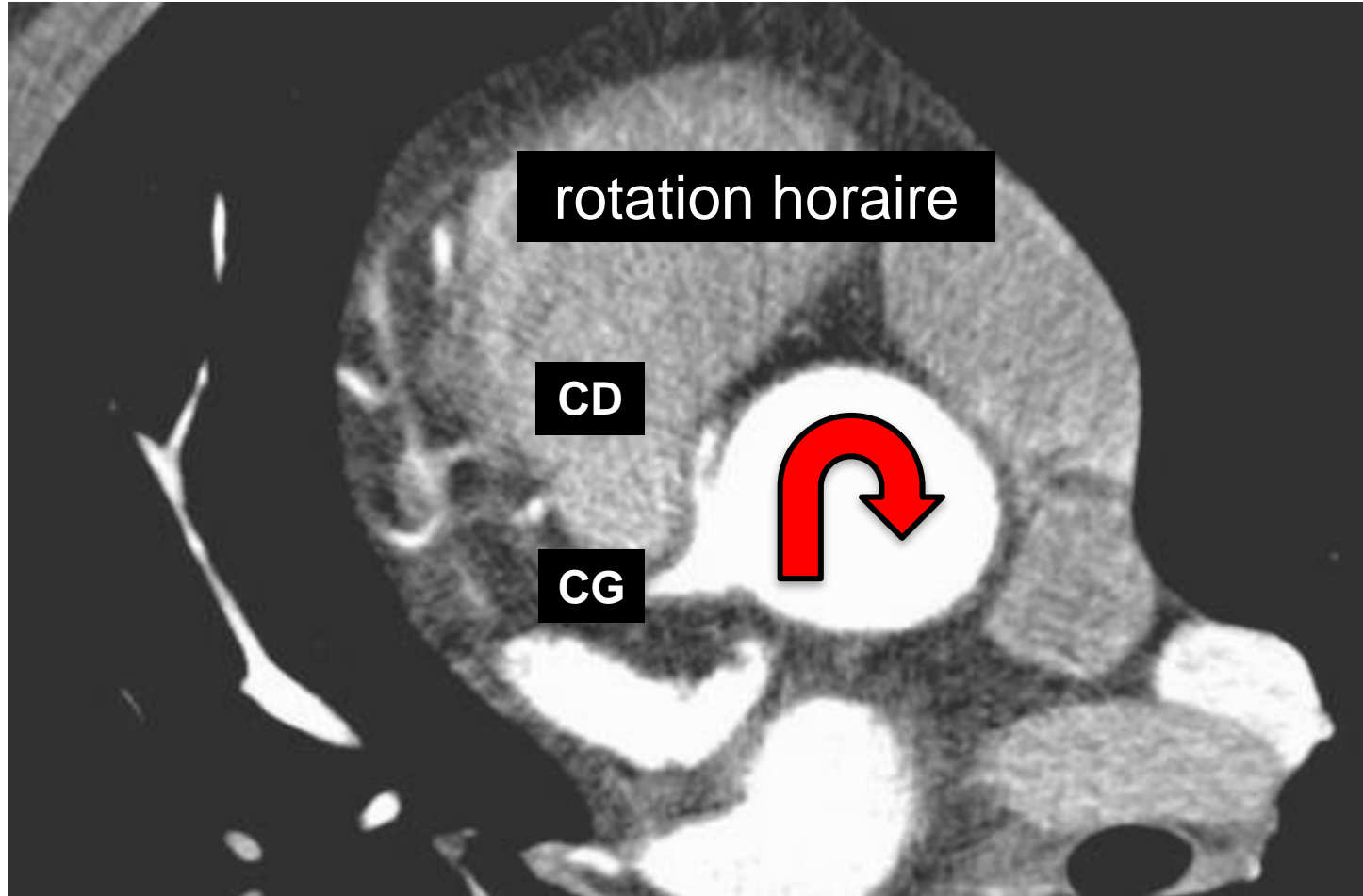
cardiologue



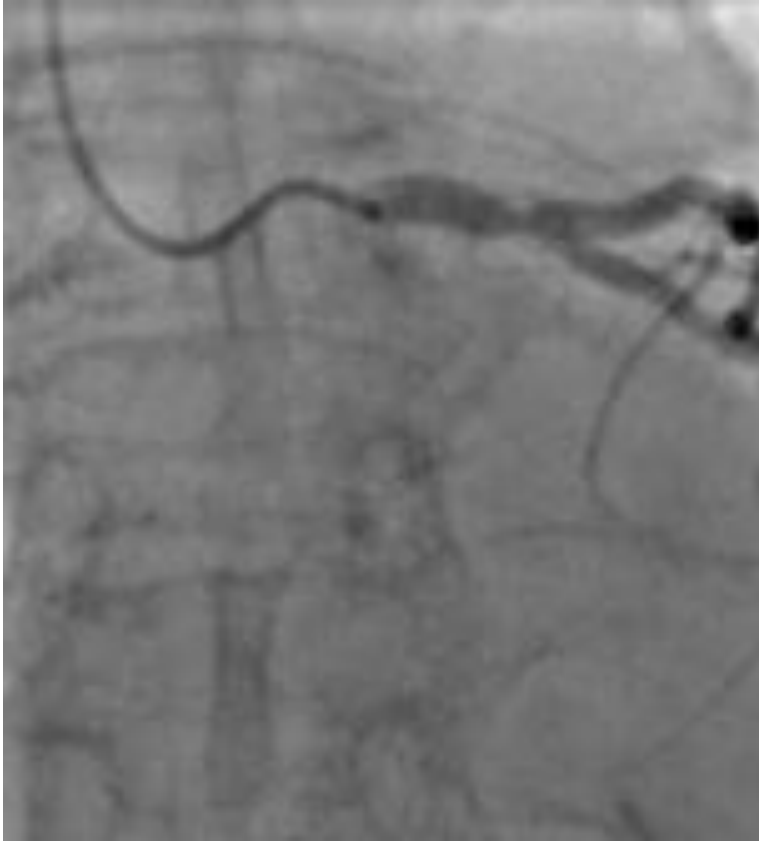
radiologue



# manipulation des cathéters

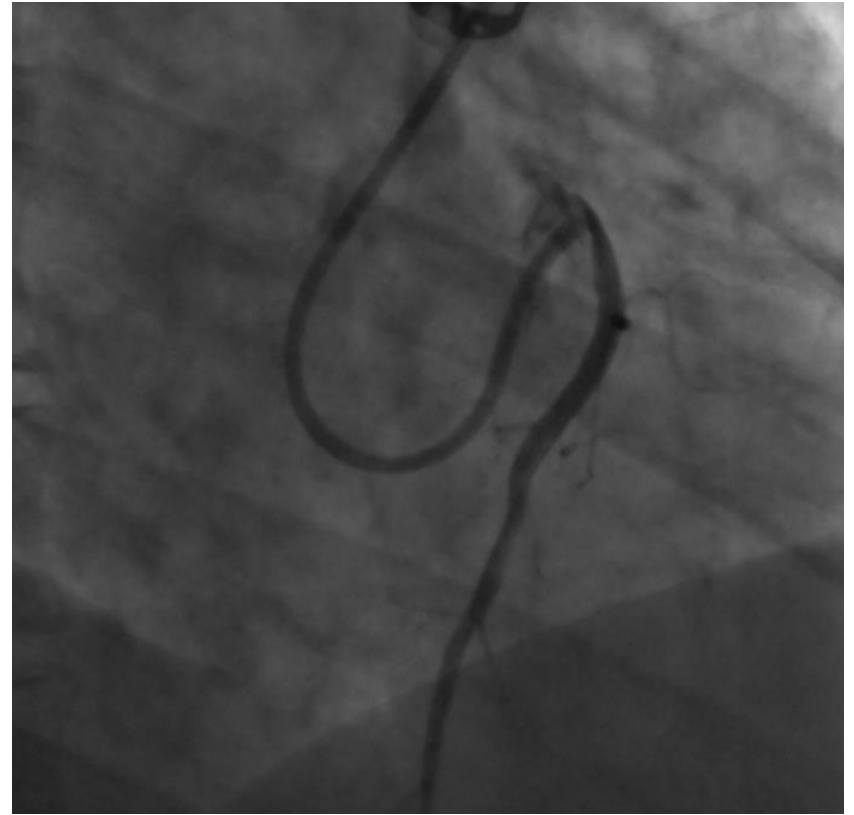


# manipulation des cathéters



Push et rotation horaire

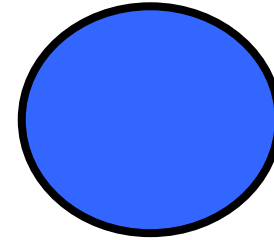
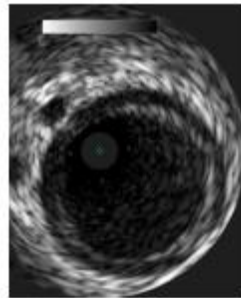
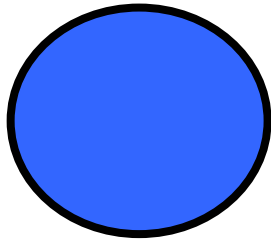
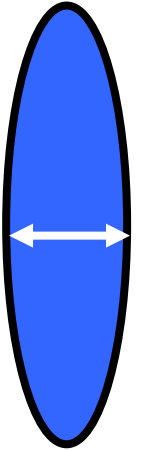
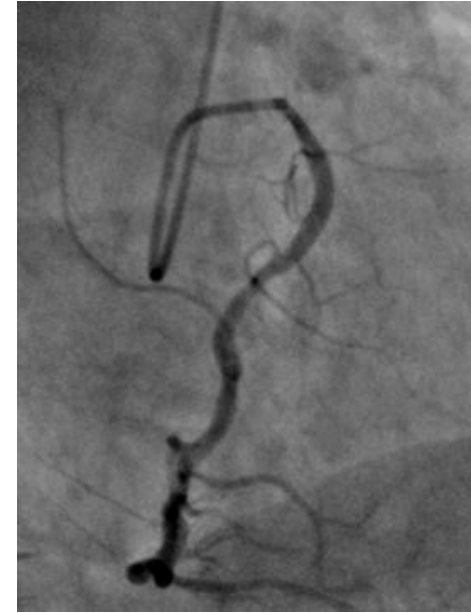
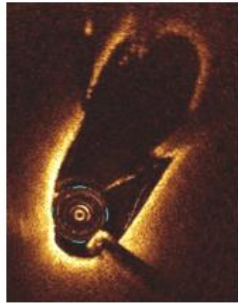
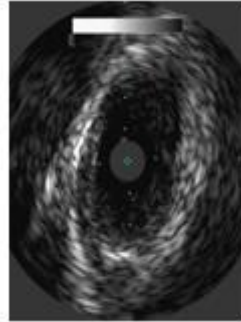
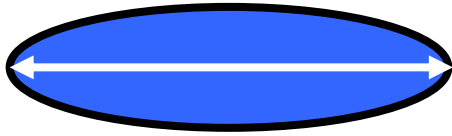
# manipulation des cathéters



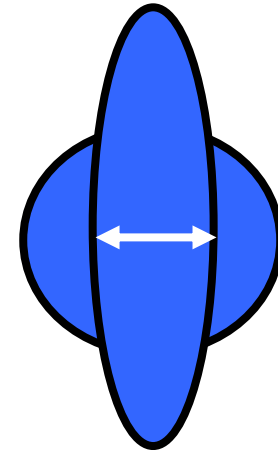
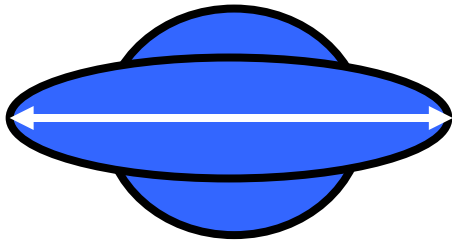
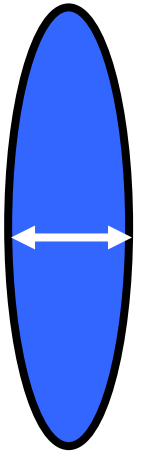
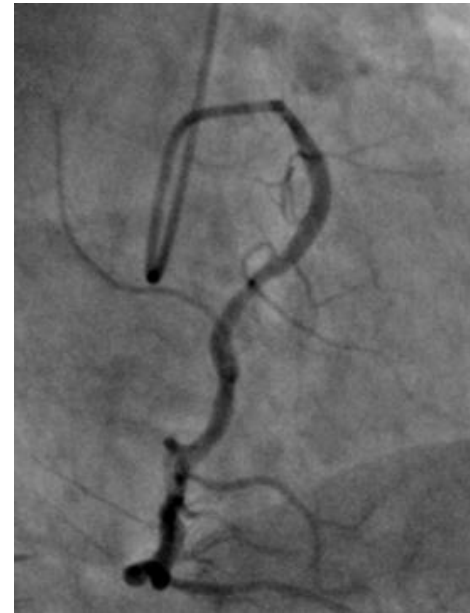
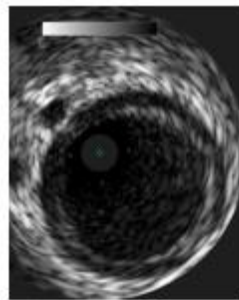
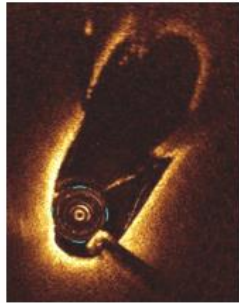
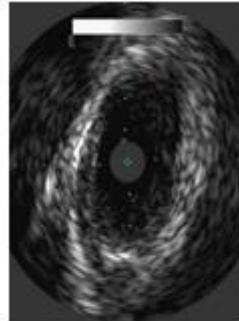
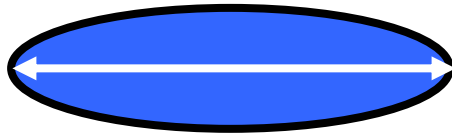
Utilisation du même cathéter



# 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 <sup>a,\*</sup>, Vijayakumar Subban <sup>a</sup>,  
Anitha Lakshmanan <sup>b</sup>, Balaji Pakshirajan <sup>a</sup>, Ramkumar Solirajaram <sup>c</sup>,  
Jaishankar Krishnamoorthy <sup>c</sup>, Ezhilan Janakiraman <sup>c</sup>,  
Ulhas M. Pandurangi <sup>c</sup>, Latchumanadhas Kalidoss <sup>c</sup>,  
Mullasari Ajit Sankaradas <sup>d</sup>

**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,<sup>1,2\*</sup> MD, Carlo Uribe,<sup>2</sup> MD, Jorge Monge,<sup>2</sup> MD, Jonathan M. Tobis,<sup>3</sup> MD,  
MacArthur A. Elayda,<sup>4</sup> MD, PhD, and James T. Willerson,<sup>1</sup> MD

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

ANOCOR droites  
n=67

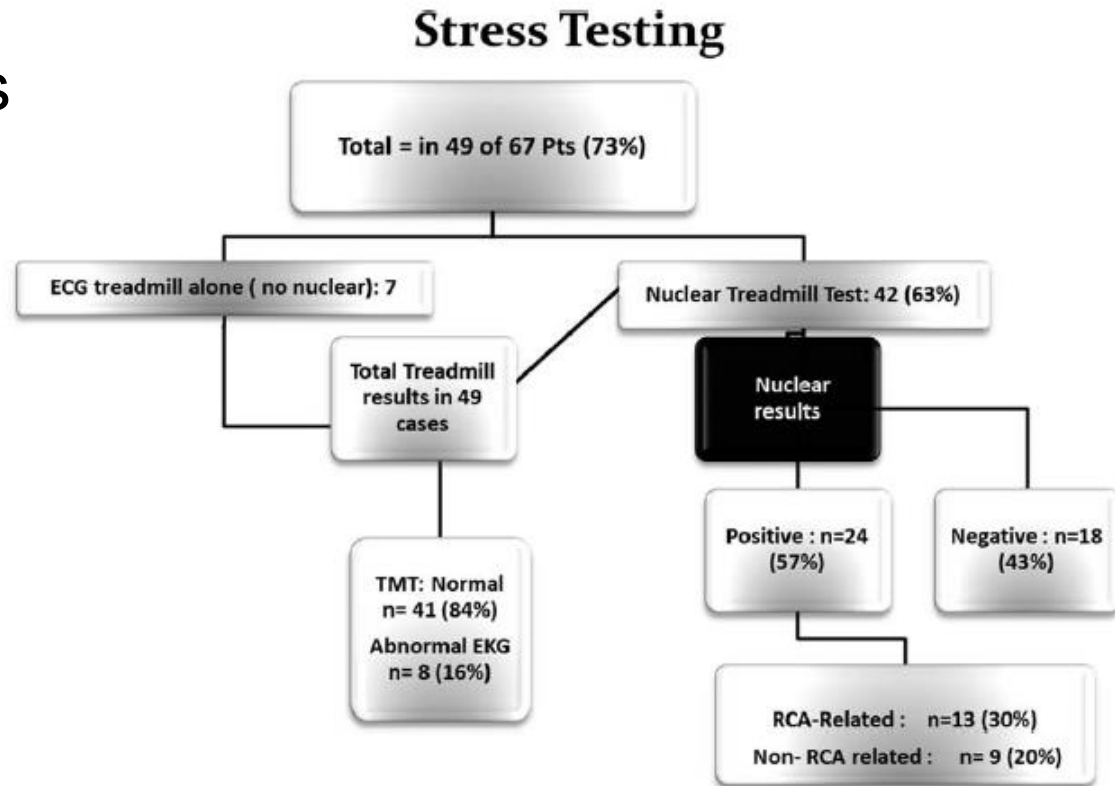


Fig. 3. Stress testing clinical evaluation studies for severity of ischemia. Note that, out of 67 patients, electrocardiographic treadmill test results were available for 49 patients and nuclear treadmill stress test results for 42 patients. ECG, electrocardiography; TMT, treadmill test; RCA, right coronary 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 <sup>201</sup>TL SPECT</i>	<i>Complications</i>
<i>RCA from the left sinus of Valsalva</i>	<i>10/22</i>	<i>3/4</i>	<i>Exercise-induced hypotension 2 Ventricular tachycardia 2</i>
<i>LCx from the right sinus of Valsalva</i>	<i>3/5</i>	<i>1/2</i>	
<i>LMCA from the posterior sinus of Valsalva</i>	<i>1/4</i>	<i>0/2</i>	<i>Exercise-induced hypotension 1</i>
<i>LMCA from the right sinus of Valsalva</i>	<i>1/1</i>		
<i>LAD and LCx from the right sinus of Valsalva</i>	<i>1/1</i>	<i>0/1</i>	
<i>Total</i>	<i>16/33 (48.5%)</i>	<i>4/9 (44.4%)</i>	<i>5/33 (15.2%)</i>

*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

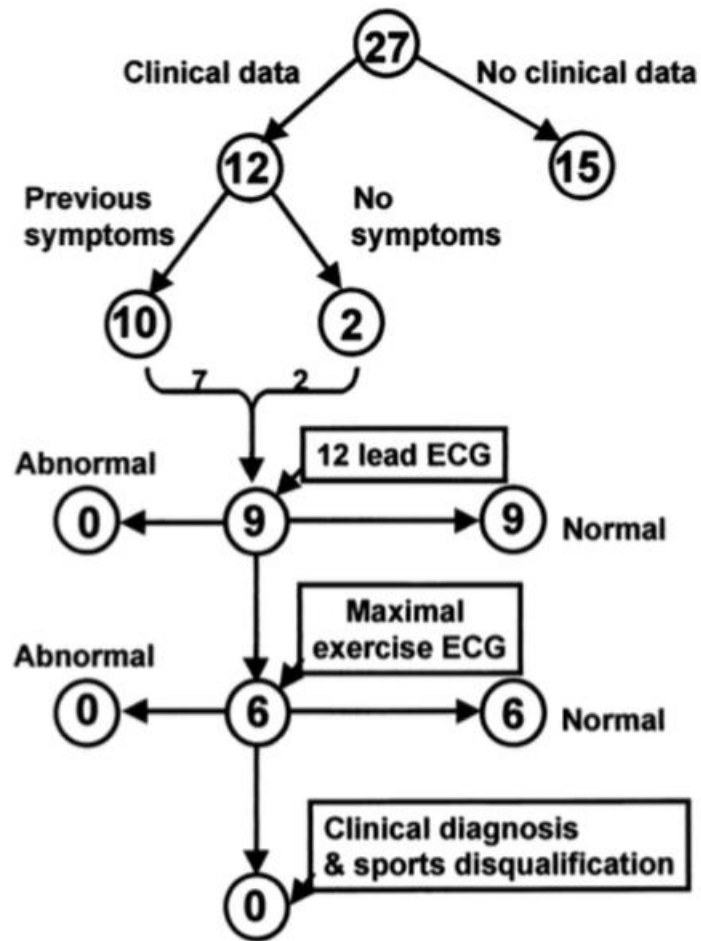
# Clinical Profile of Congenital Coronary Artery Anomalies With Origin From the Wrong Aortic Sinus Leading to Sudden Death in Young Competitive Athletes

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

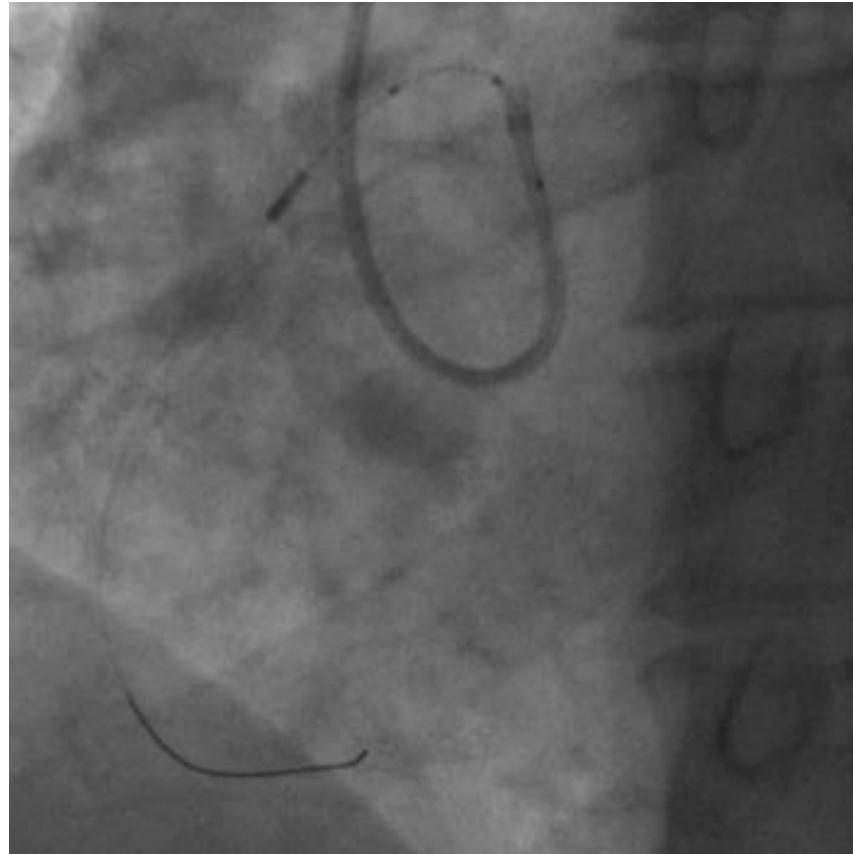
Padua, Italy and Minneapolis, Minnesota

**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

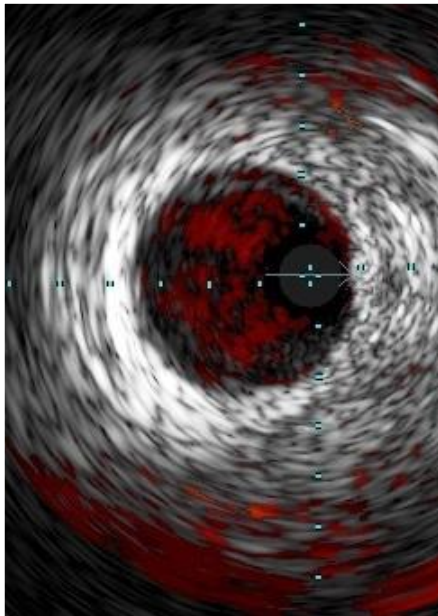


# échographie endocoronaire

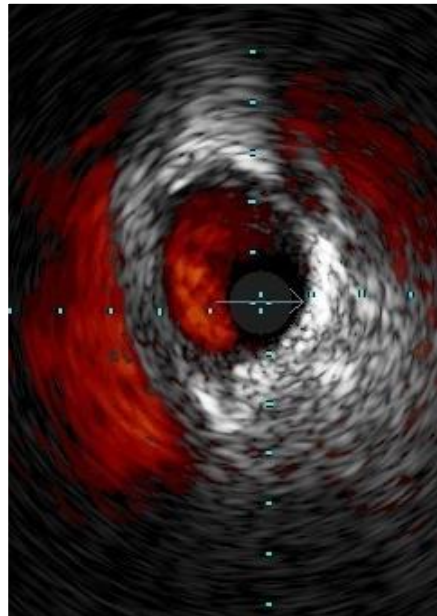


# échographie endocoronaire ANOCOR droite avec passage intramural

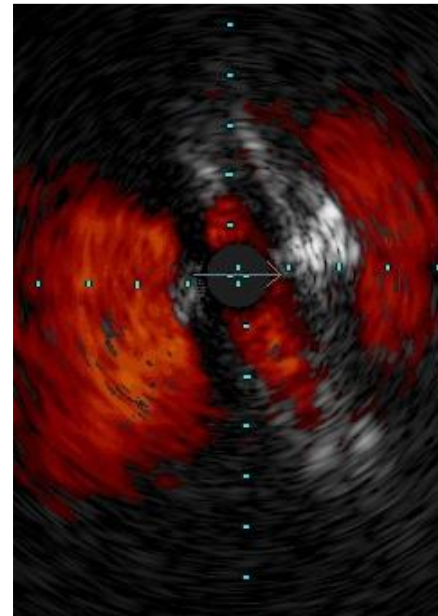
extramural



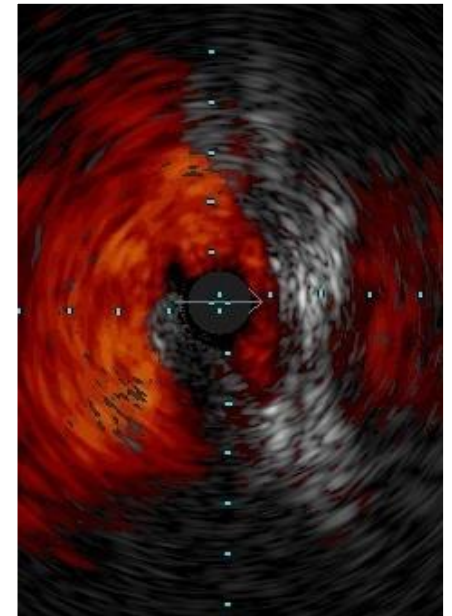
juxtamural



intramural



ostium



Paolo Angelini,<sup>1,2\*</sup> MD, Carlo Uribe,<sup>2</sup> MD, Jorge Monge,<sup>2</sup> MD, Jonathan M. Tobis,<sup>3</sup> MD,  
MacArthur A. Elayda,<sup>4</sup> MD, PhD, and James T. Willerson,<sup>1</sup> MD

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

ANOCOR droites  
n=66

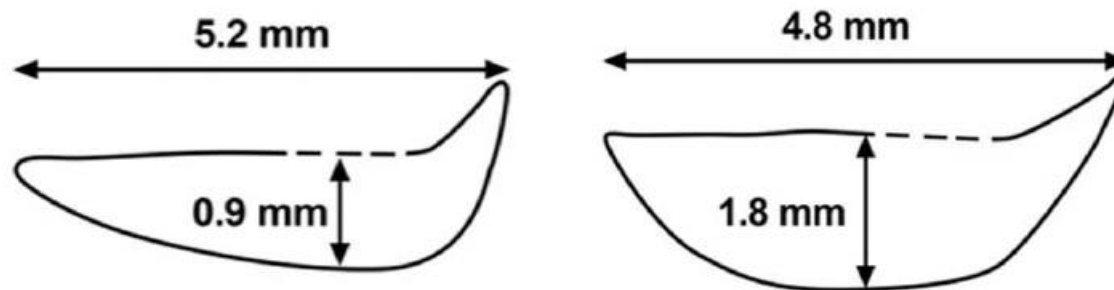
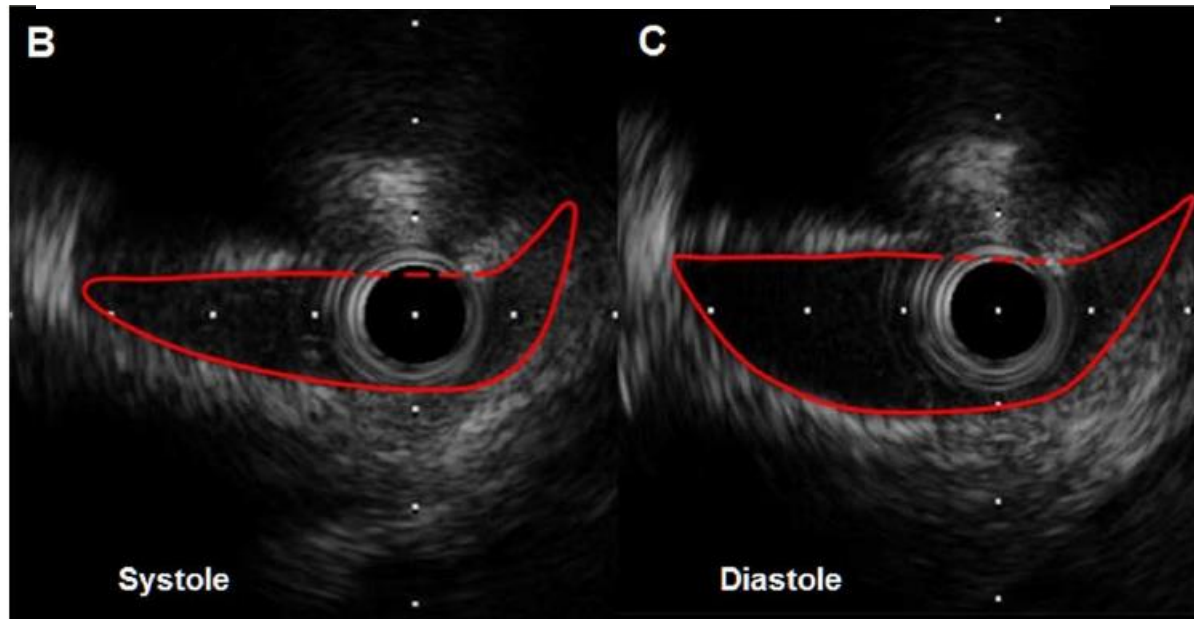
**TABLE IV. Quantitative Measurements From Intravascular Ultrasonography**

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

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

Paolo Angelini,<sup>1,2\*</sup> MD, Carlo Uribe,<sup>2</sup> MD, Jorge Monge,<sup>2</sup> MD, Jonathan M. Tobis,<sup>3</sup> MD,  
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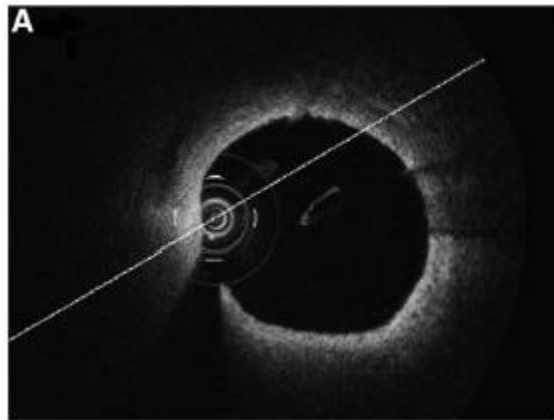
Catheterization and Cardiovascular Interventions 86:199–208 (2015)





# Optical Coherence Tomography endocoronaire ANOCOR droite avec probable passage intramural

Paolo Angelini,<sup>1,2\*</sup> MD, Carlo Uribe,<sup>2</sup> MD, Jorge Monge,<sup>2</sup> MD, Jonathan M. Tobis,<sup>3</sup> MD,  
MacArthur A. Elayda,<sup>4</sup> MD, PhD, and James T. Willerson,<sup>1</sup> MD

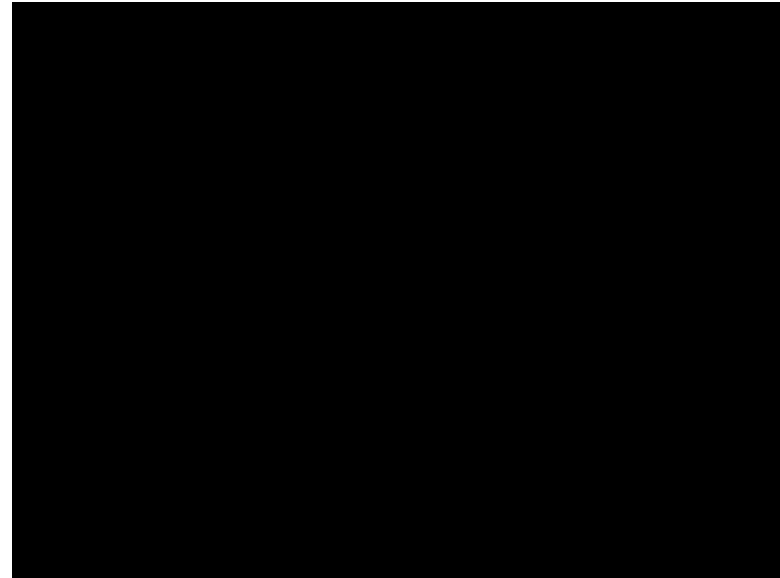


Catheterization and Cardiovascular Interventions 86:199–208 (2015)

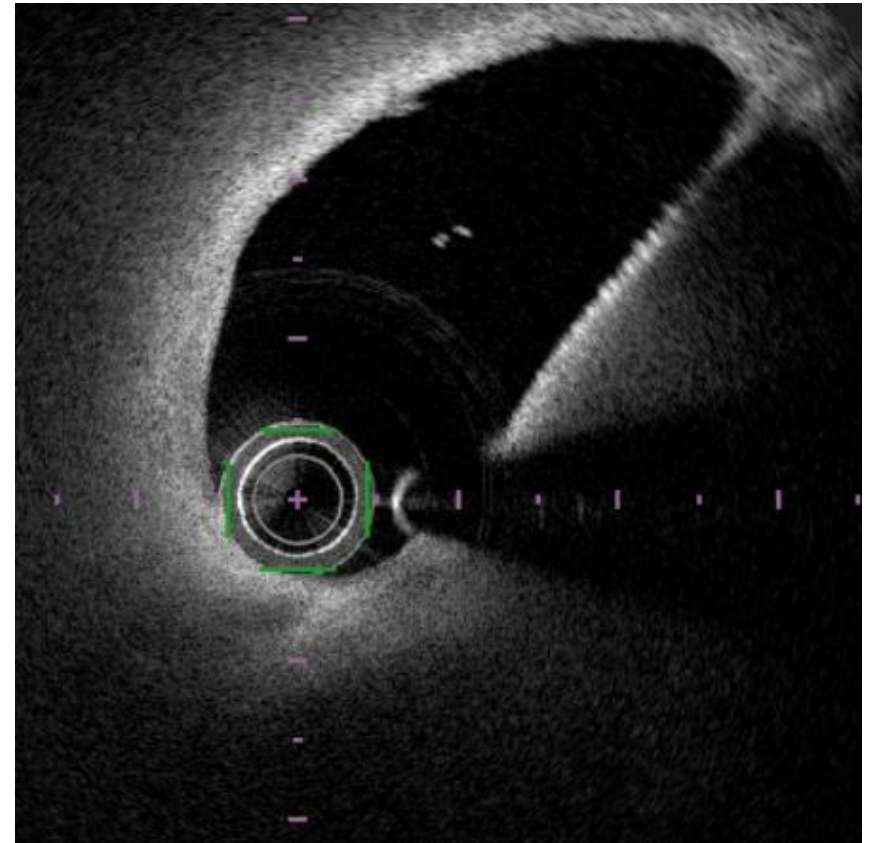
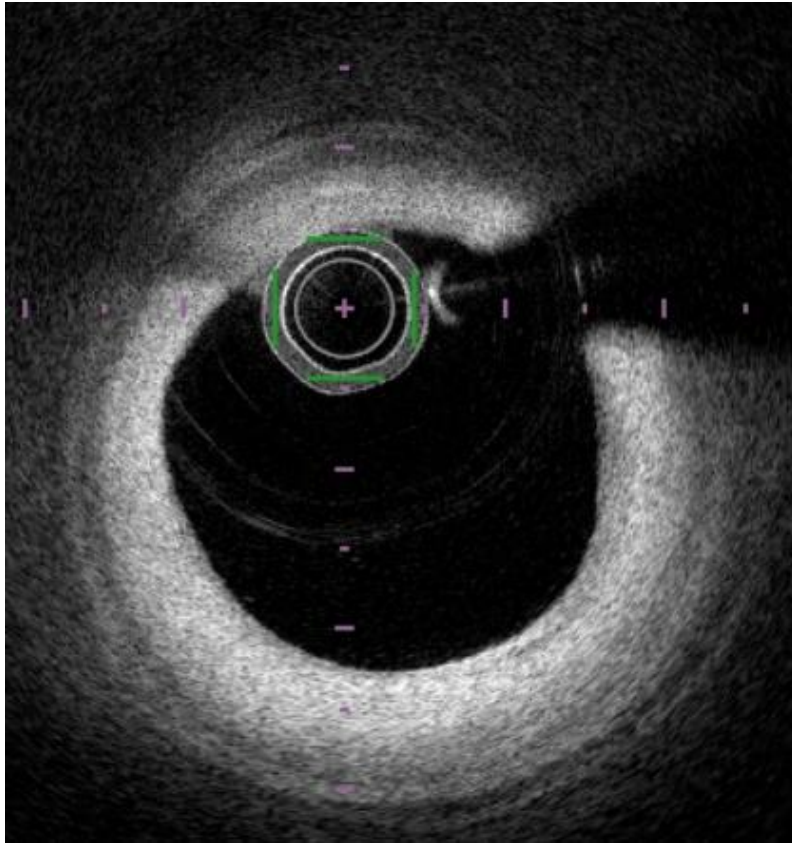
Fig. 2. Optical coherence tomography (OCT) images. With this modality, the inner lumen can be crisply imaged, but only if the operator can advance the guiding catheter selectively enough to eliminate most of the luminal blood, thereby enabling infrared laser imaging. (A) The reference distal cross-sectional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



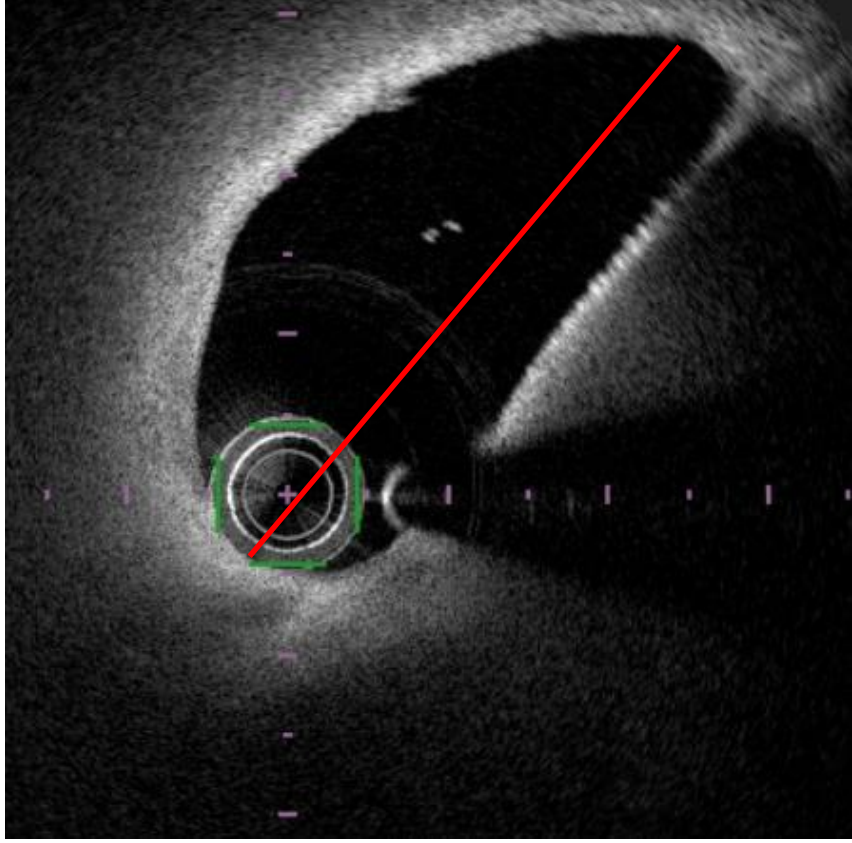
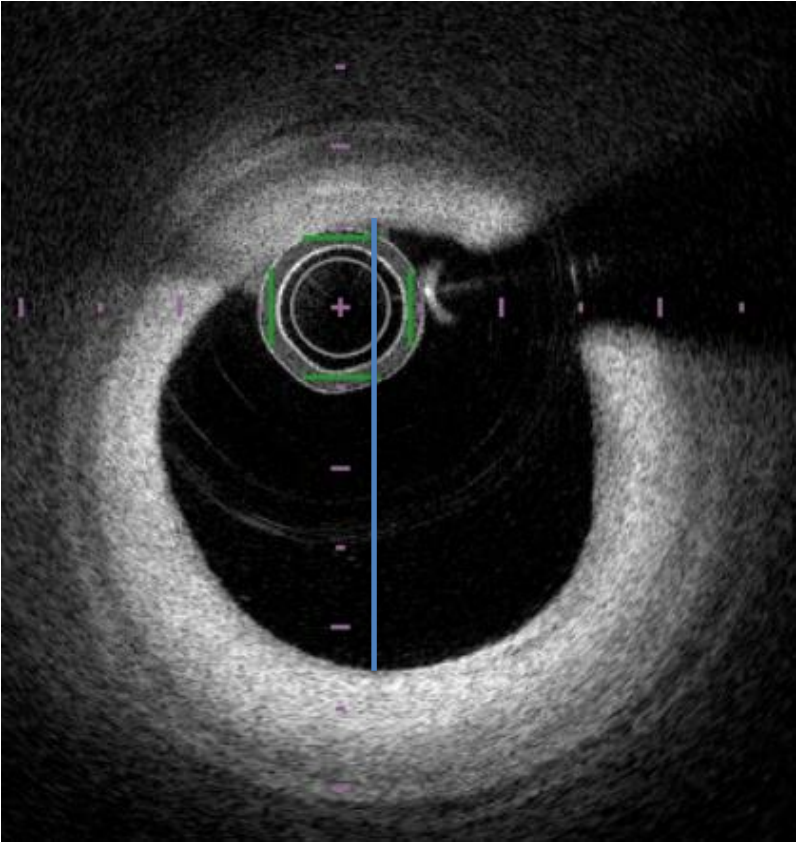
# *Optical Coherence Tomography* endocoronaire ANOCOR droite



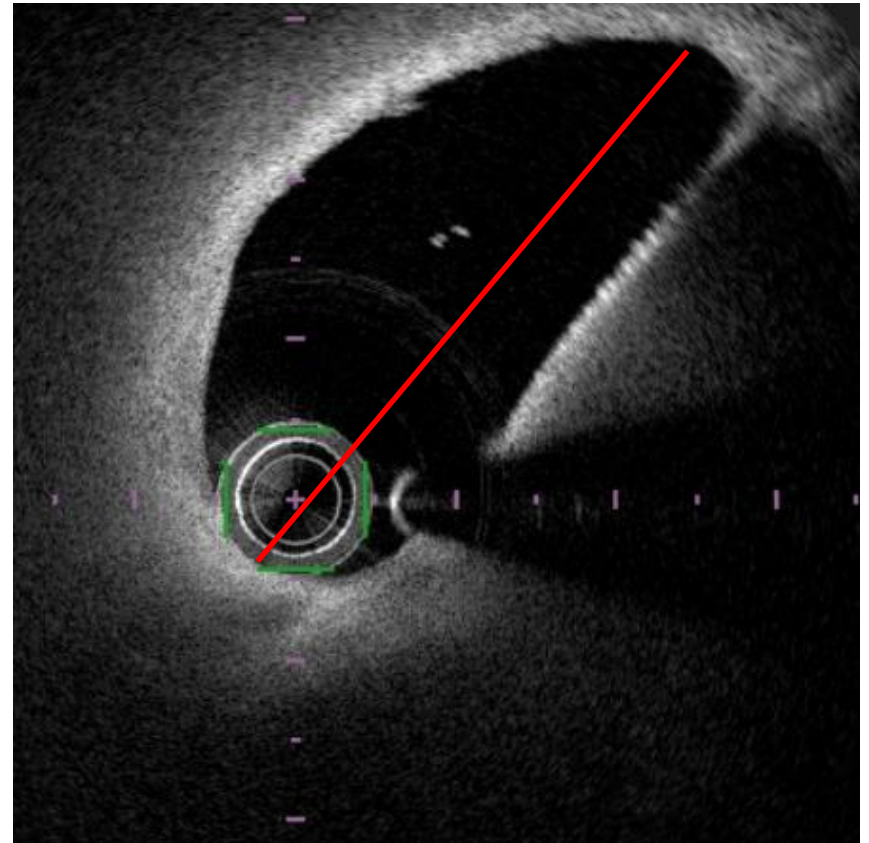
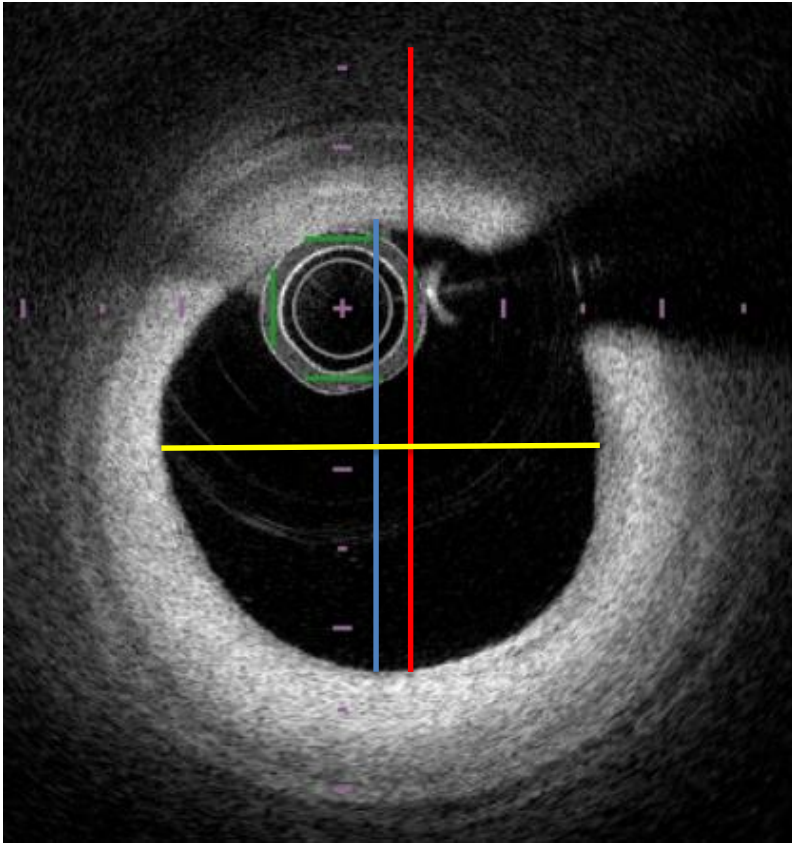
# Optical Coherence Tomography endocoronaire ANOCOR droite



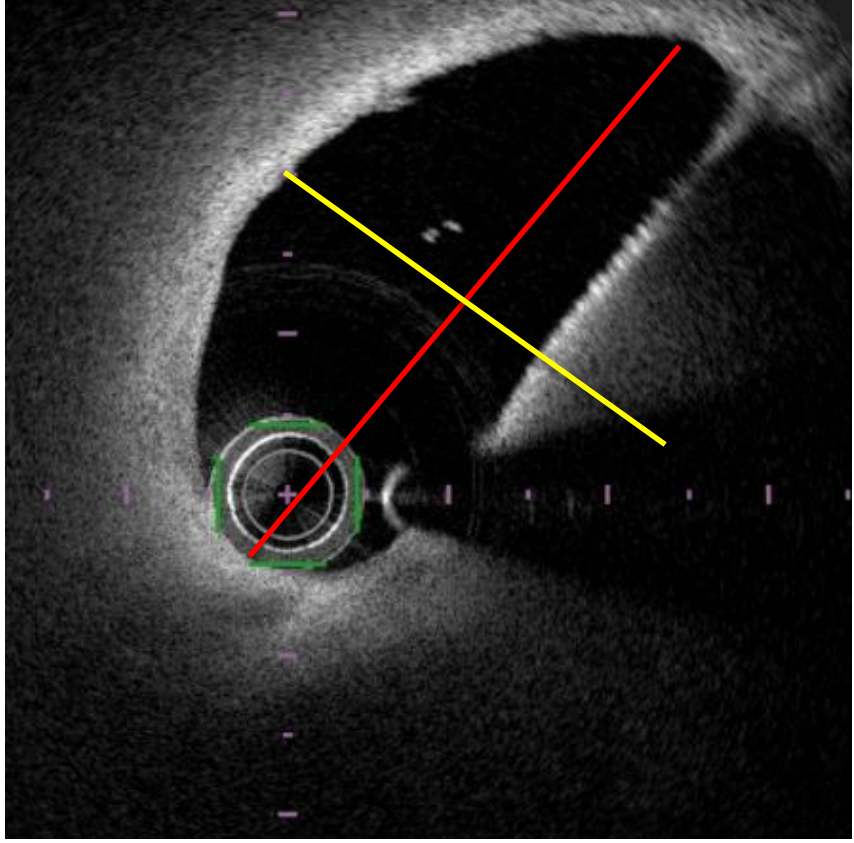
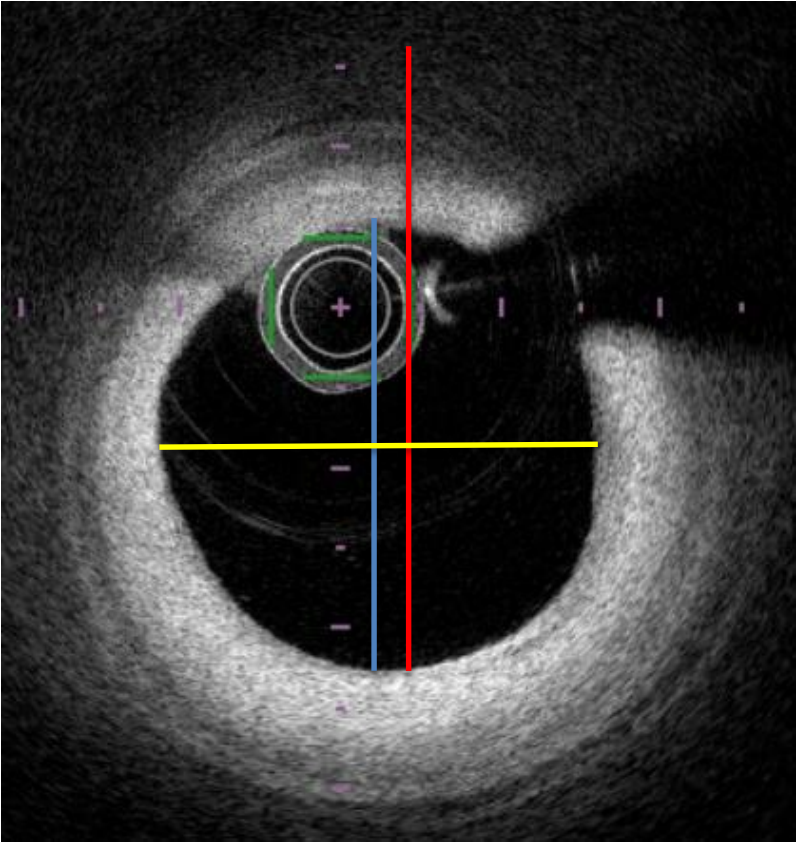
# Optical Coherence Tomography endocoronaire ANOCOR droite



# Optical Coherence Tomography endocoronaire ANOCOR droite

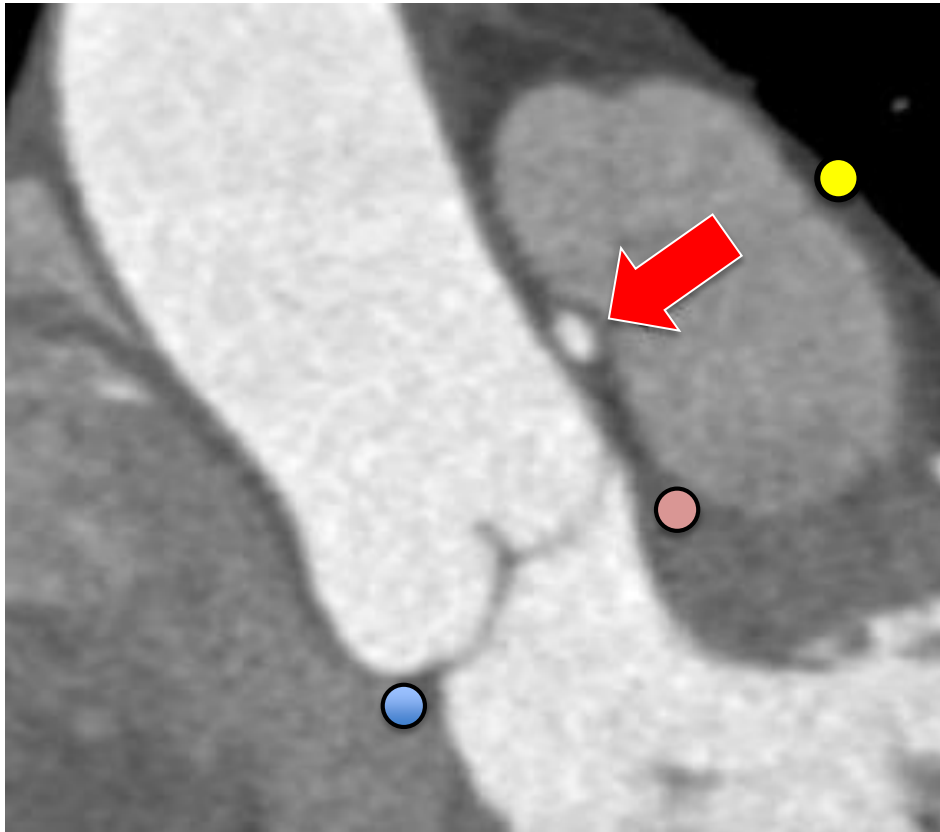


# Optical Coherence Tomography endocoronaire ANOCOR droite



# 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

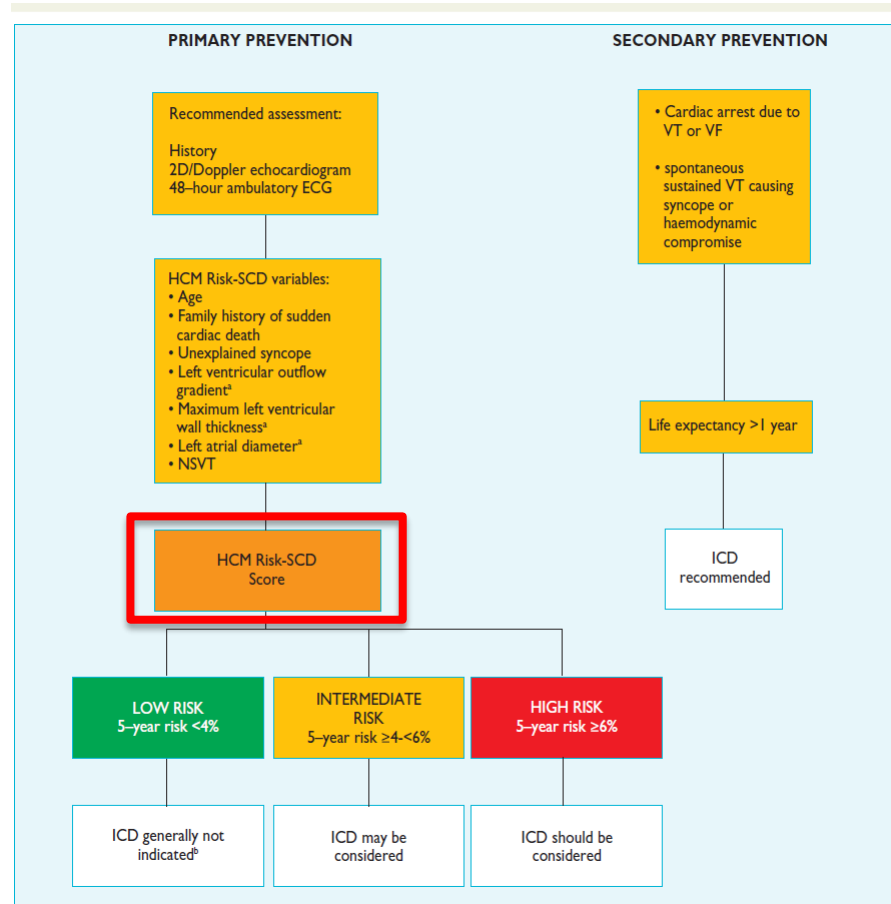
1.9 deaths/100.000 person-years

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

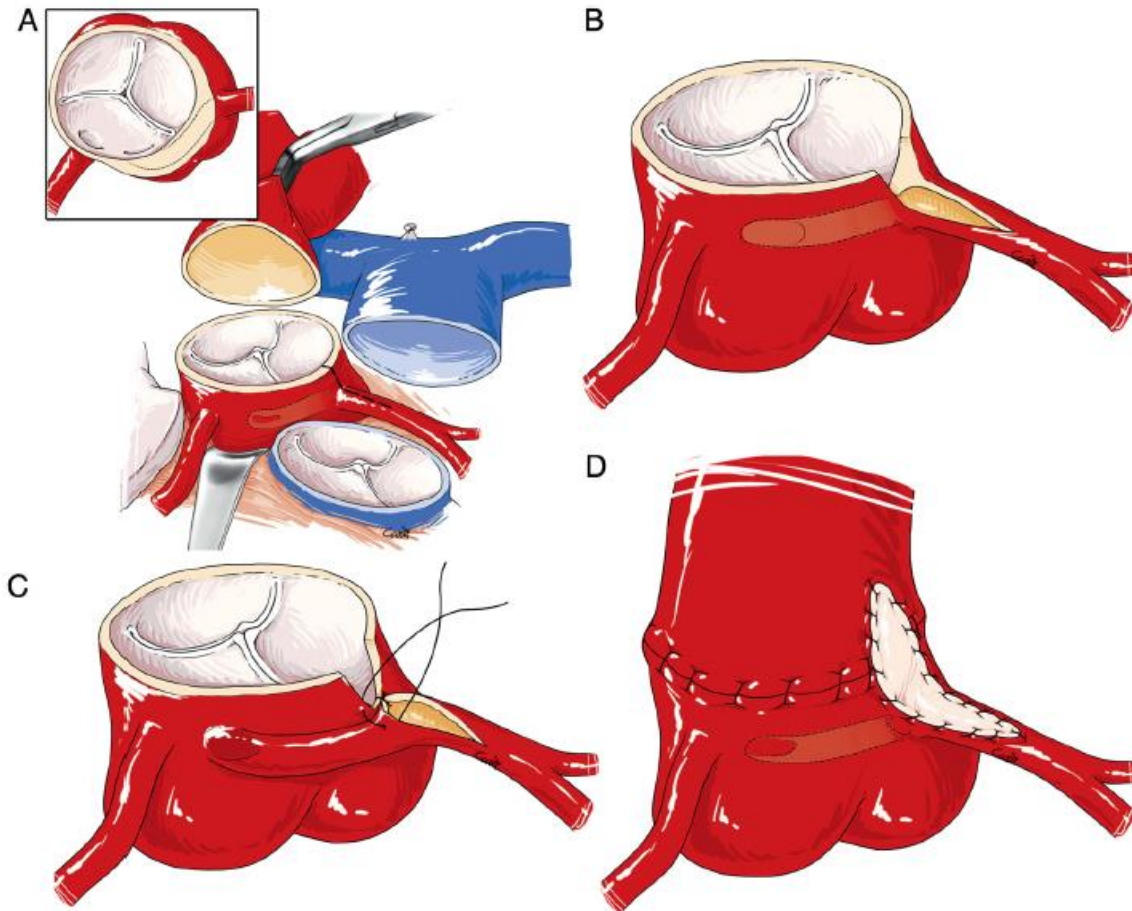
### ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease

*Circulation* December 2, 2008

#### *Class IIa*

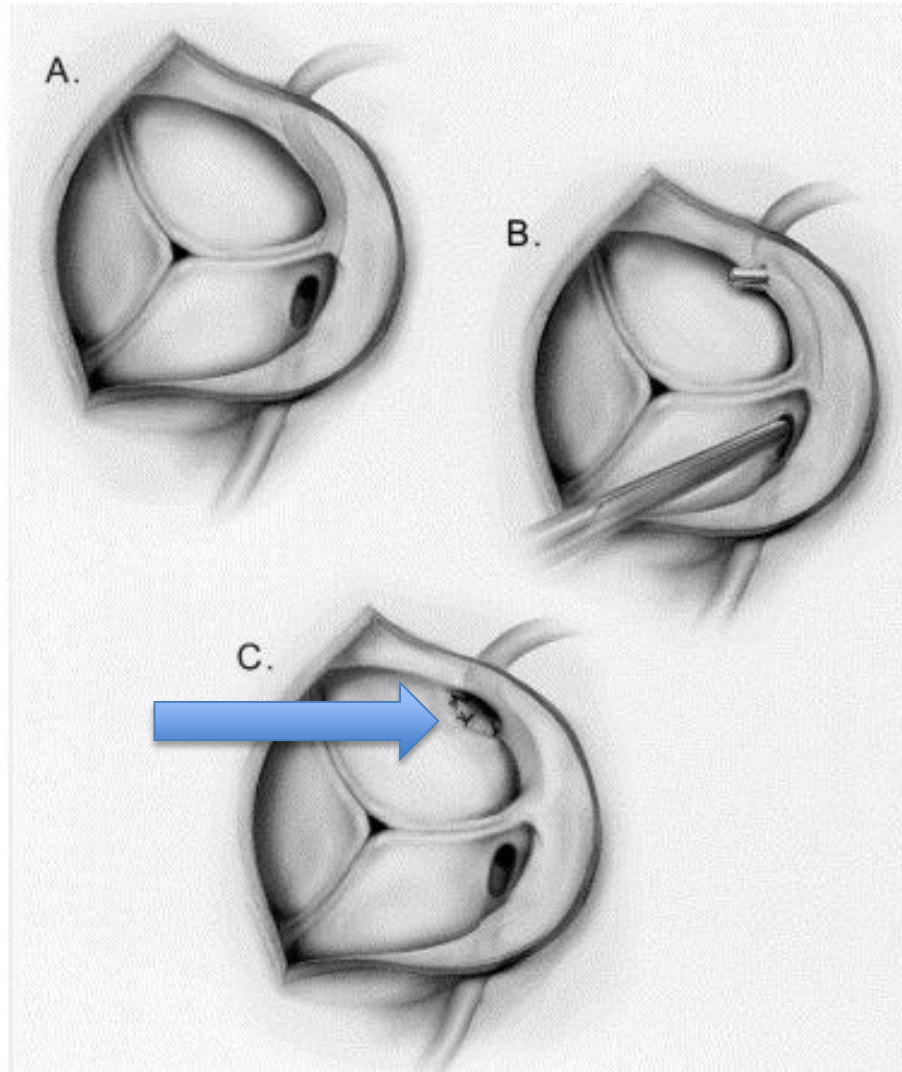
1. Surgical coronary revascularization can be beneficial in the setting of documented vascular wall hypoplasia, coronary compression, or documented obstruction to coronary flow, regardless of inability to document coronary ischemia. (*Level of Evidence: C*)
2. Delineation of potential mechanisms of flow 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



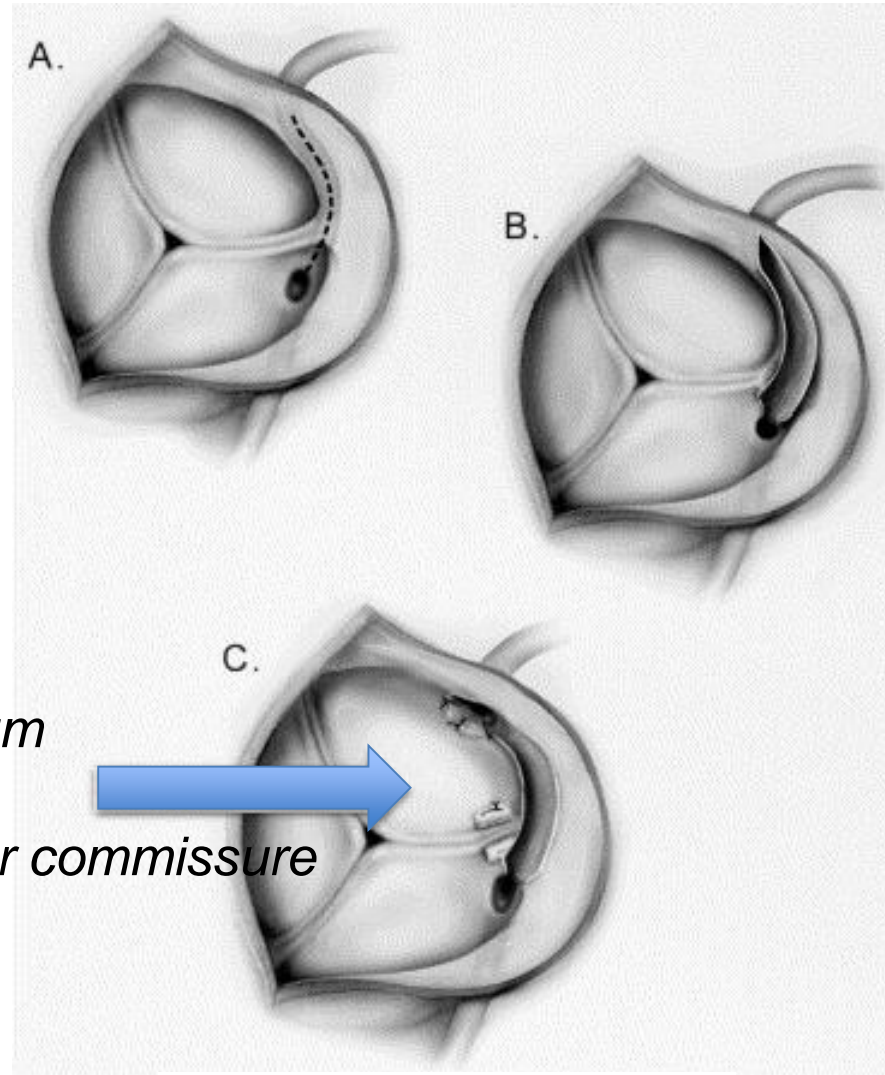
Gaudin R et al. Multimed Man Cardiothorac Surg 2014

# correction chirurgicale : création néoostium



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

# correction chirurgicale : *unroofing*



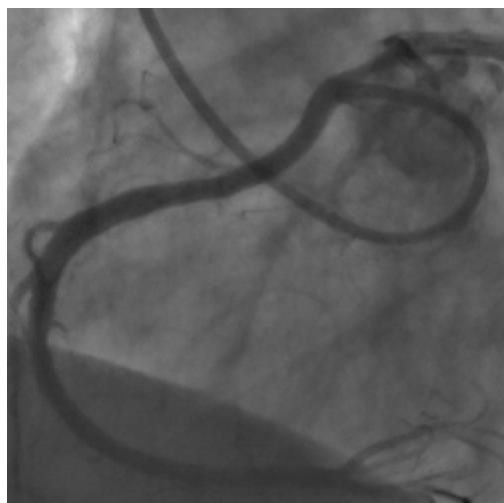
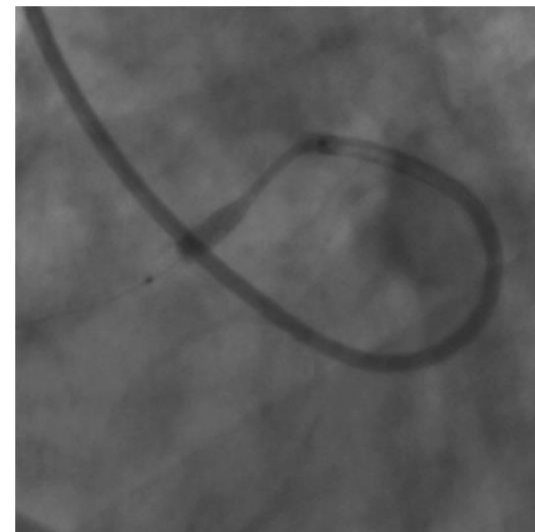
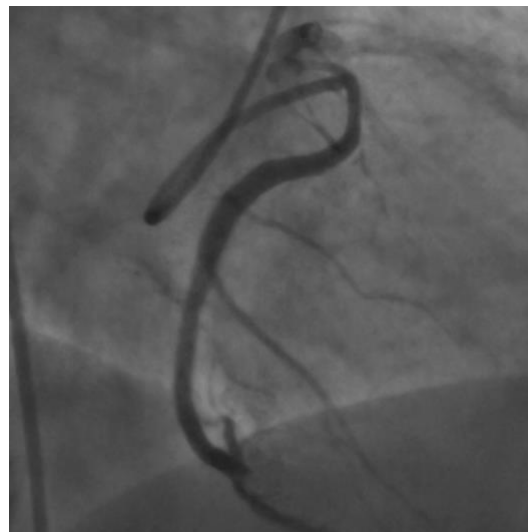
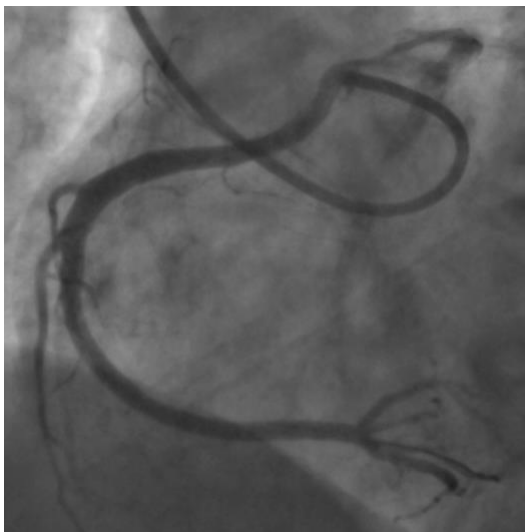
*création néo ostium*

*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

Paolo Angelini,<sup>1,2\*</sup> MD, Carlo Uribe,<sup>2</sup> MD, Jorge Monge,<sup>2</sup> MD, Jonathan M. Tobis,<sup>3</sup> MD, MacArthur A. Elayda,<sup>4</sup> MD, PhD, and James T. Willerson,<sup>1</sup> MD

**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)

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

## 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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