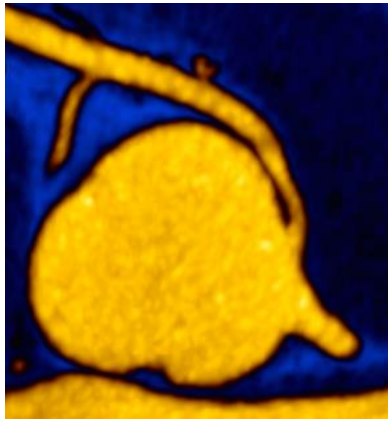
CHORUS SEOUL

November 2017
Seoul, Korea

anomalous connection of the right **cor**onary artery with interarterial course



Pierre Aubry on behalf of the ANOCOR Group

Bichat Hospital
Paris
France

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

anoCor

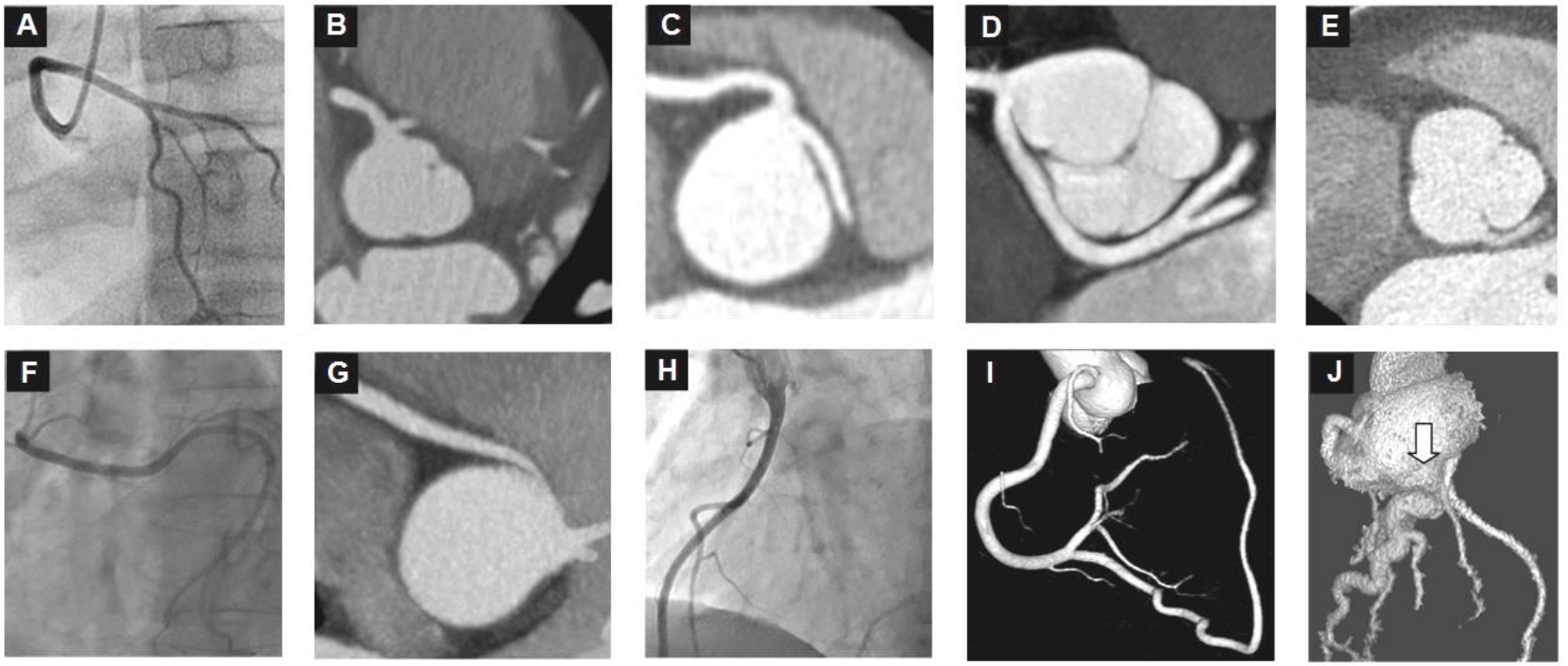
CHORUS SEOUL

November 2017
Seoul, Korea

Conflict of interest: nothing to report

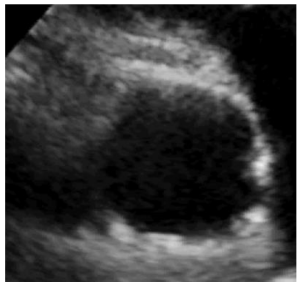
Anomalous connections of coronary arteries

Wide spectrum of anomalous connections

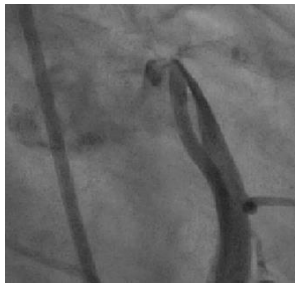


Anomalous connections of coronary arteries

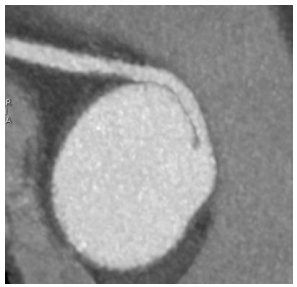
Prevalence with cardiovascular imaging



- Echocardiography 0.2/100



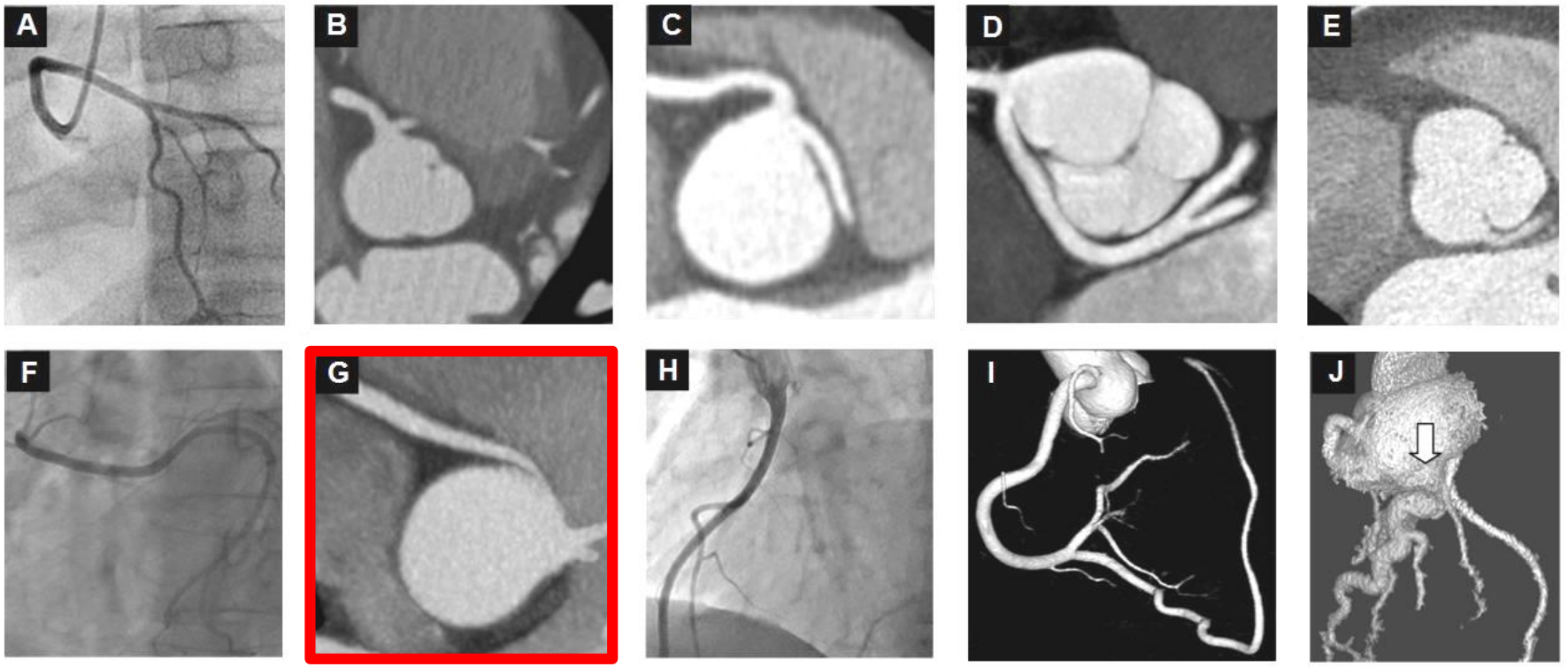
- Selective angiography 0.8/100



- CT scan angiography 1.2/100

anomalous connection of the RCA with interarterial course

Wide spectrum of anomalous connections



ANOCOR cohort

- n=472 patients
- n=496 abnormalities
- recruitment : 01/2010-01/2013
- 71 French investigators

Coordination

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

ASSISTANCE PUBLIQUE HÔPITAUX DE PARIS

Grant

GACI
Groupe Athérome et Cardiologie Interventionnelle de la Société Française de Cardiologie

anomalous connection of the RCA with interarterial course

- Not rare congenital anomaly
- Interarterial course in most of cases
- Very low risk of sudden death in youngs
- Possible ischemia/symptoms in adults
- Management remains debated
- Screening policy in athletes

anomalous connection of the RCA with interarterial course

PREVALENCE IN SELECTED POPULATION

ANOCOR Registry

TABLE 1 Demographic and angiographic characteristics of the ANOCOR cohort

Parameters	
Number of subjects, n	472
Mean age, y (SD)	63 (13)
Gender male, %	76.2
Invasive CA alone, n (%)	297 (62.9)
Computed tomography CA alone, n (%)	20 (4.3)
Invasive + computed tomography CA, n (%)	155 (32.8)
Total number of anomalous connections	496
Type of artery	
Left main, n (%)	60 (12.1)
Left anterior descending, n (%)	27 (5.4)
Circumflex, n (%)	235 (47.4)
Right, n (%)	165 (33.3)
Other, n (%)	9 (1.8)

Koutsoukis A. Congenit Heart Dis 2017

anomalous connection of the RCA with interarterial course

PREVALENCE IN YOUNG POPULATION

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

L-ACAOS = 2
R-ACAOS = 17

Total ACAOS = 19

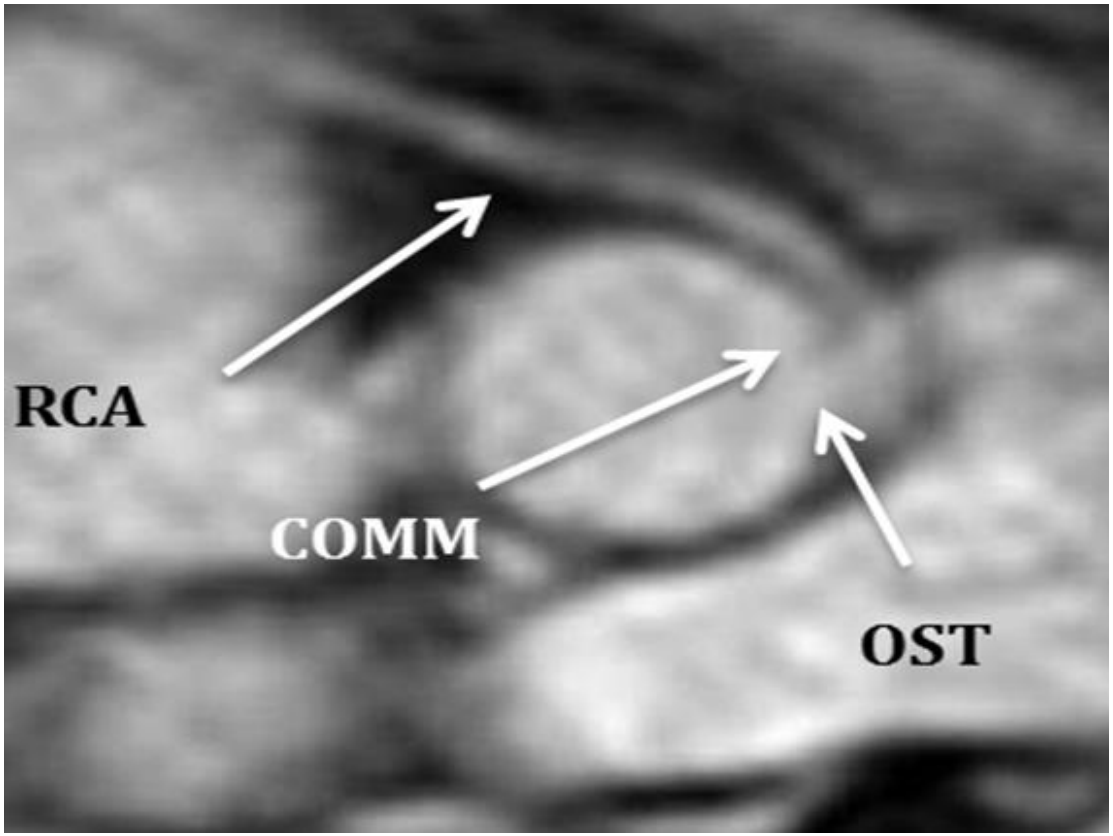
Prevalence L-ACAOS = 0.04%
Prevalence R-ACAOS = 0.32% ≈ 3/1000

Total prevalence ACAOS = 0.35% ≈ 4/1000

AMERICAN COLLEGE of CARDIOLOGY

Angelini P. ACC sessions 2017

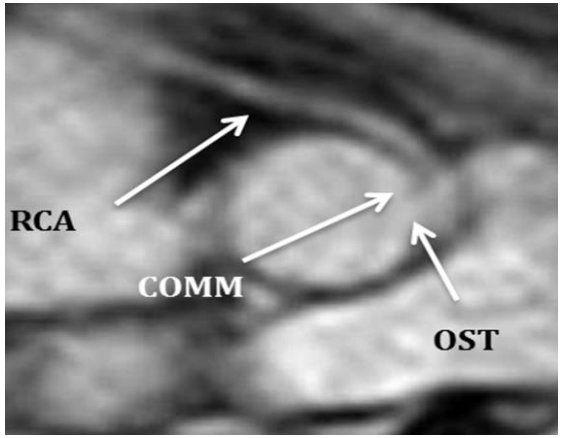
MRI-based study
n = 5.255
middle school children (mean age 13 years)
2010-2017

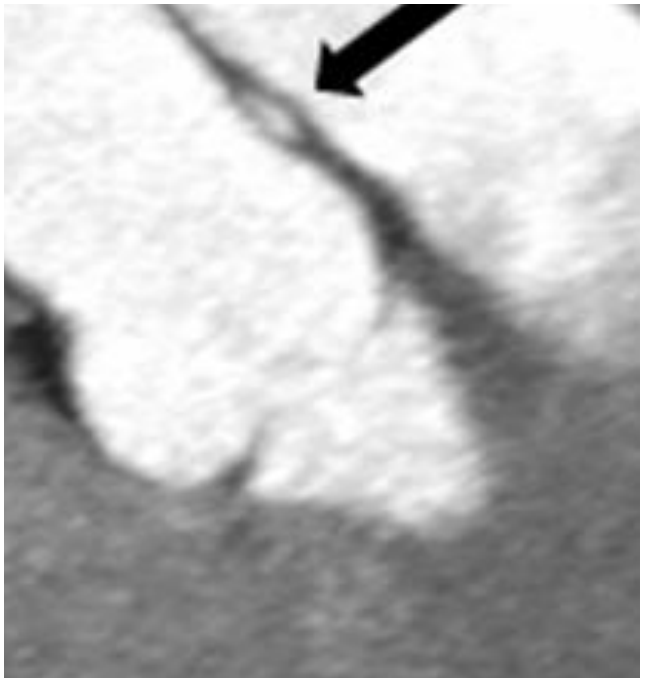


AMERICAN COLLEGE of CARDIOLOGY

Angelini P. ACC sessions 2017

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





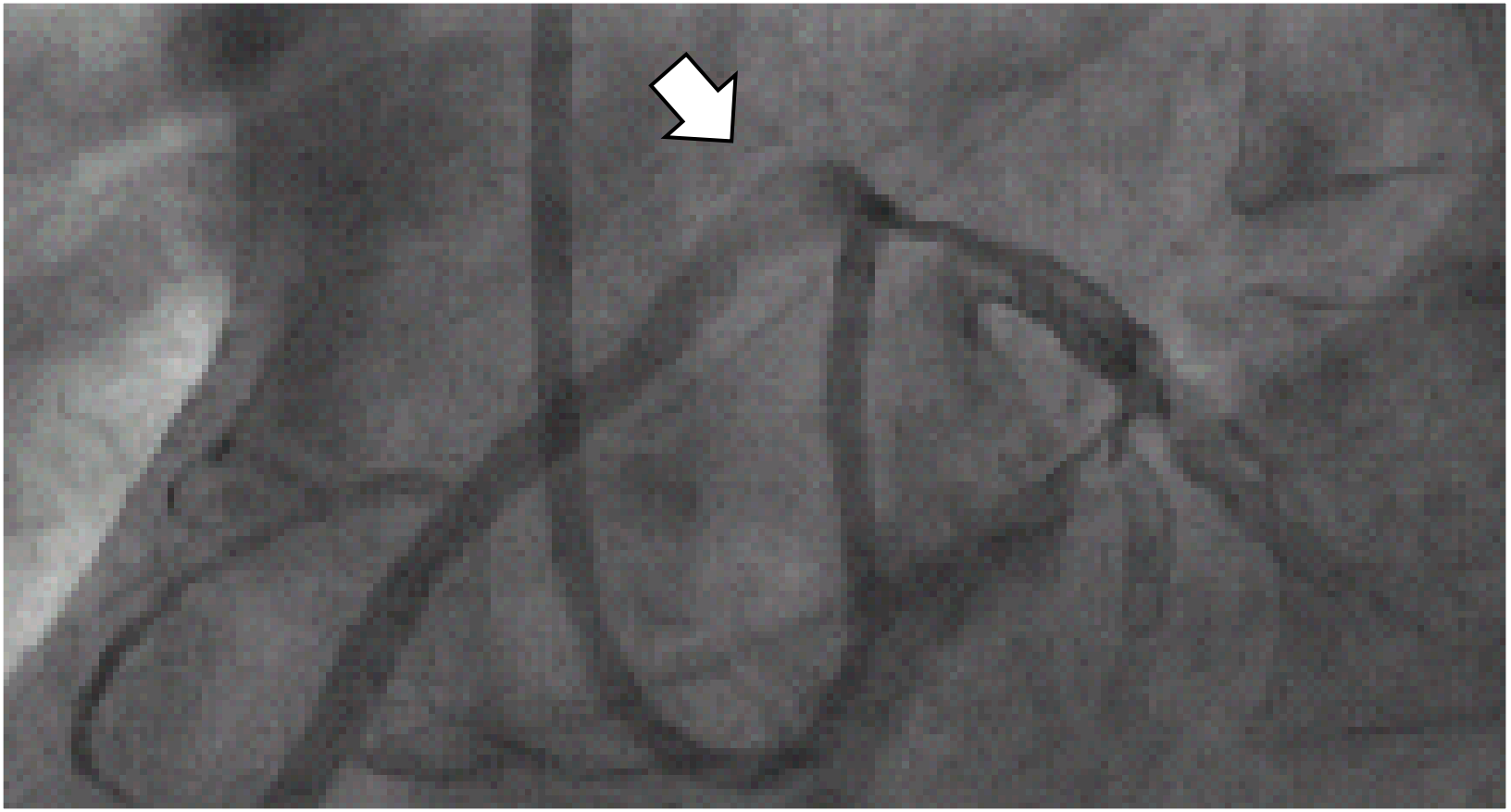
AMERICAN COLLEGE of CARDIOLOGY

Angelini P. ACC sessions 2017

CHD with risk of sudden cardiac death (estimation)

CONGENITAL HEART DISEASE	PREVALENCE cases per 100 000
Anomalous connections of coronary arteries	**350 (0.35%)**
Hypertrophic cardiomyopathy	200 (0.20%)
Wolf-Parkinson-White syndrome	150 (0.15%)
Long QT syndrome	50 (0.05%)
Idiopatic dilated cardiomyopathy	40 (0.04%)
Arrhythmogenic right ventricular cardiomyopathy	40 (0.04%)
Brugada syndrome	20 (0.02%)
Catecholaminergic polymorphic ventricular tachycardia	10 (0.01%)

anomalous connection of the right coronary artery



CHD with risk of sudden cardiac death (estimation)

CONGENITAL HEART DISEASE	PREVALENCE cases per 100 000
Anomalous connection of right coronary artery	**300 (0.30%)**
Hypertrophic cardiomyopathy	200 (0.20%)
Wolf-Parkinson-White syndrome	150 (0.15%)
Long QT syndrome	50 (0.05%)
Anomalous connection of left coronary artery	**40 (0.04%)**
Idiopatic dilated cardiomyopathy	40 (0.04%)
Arrhythmogenic right ventricular cardiomyopathy	40 (0.04%)
Brugada syndrome	20 (0.02%)
Catecholaminergic polymorphic ventricular tachycardia	10 (0.01%)

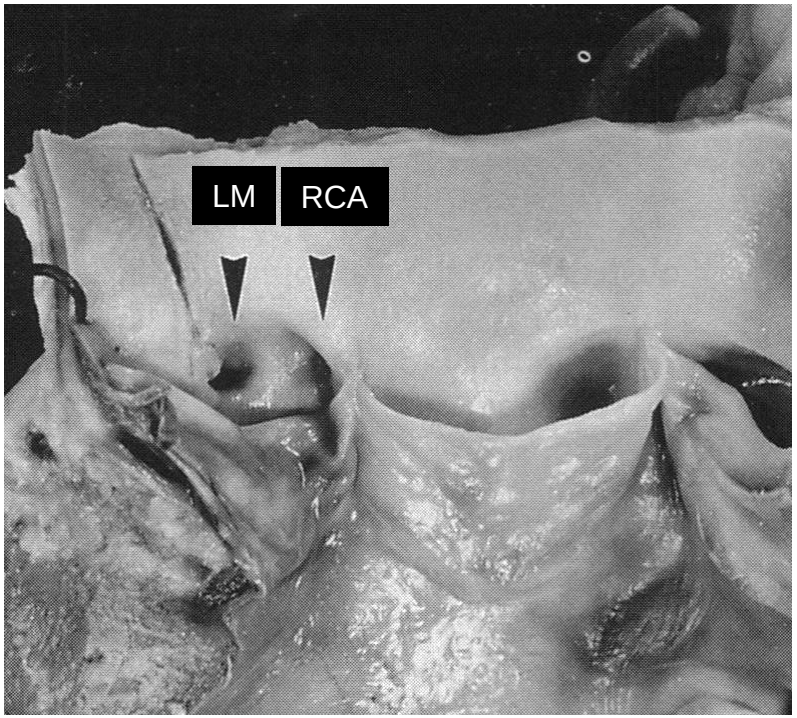
anomalous connection of the RCA with interarterial course

RISK OF SUDDEN CARDIAC DEATH

anomalous connection of the RCA with interarterial course

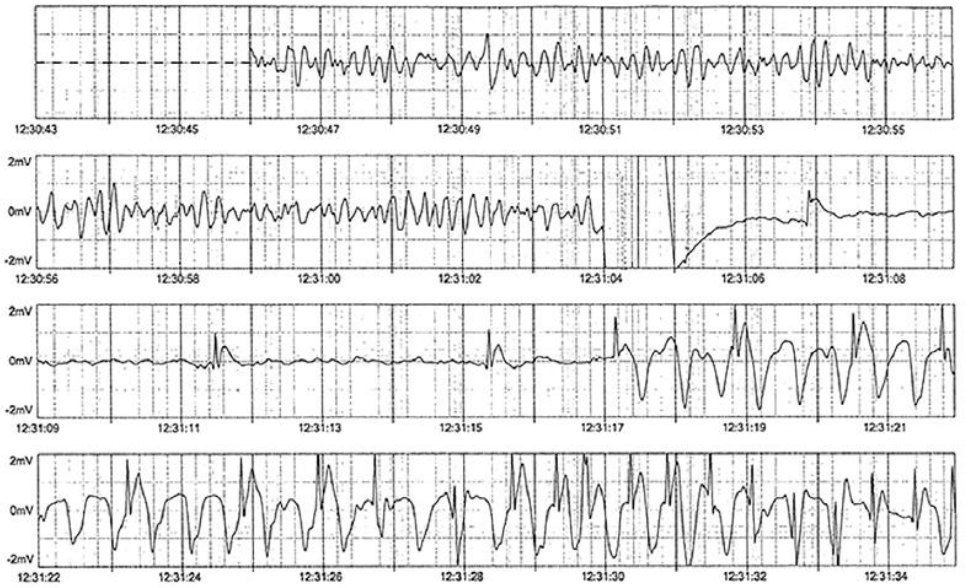
Risk of sudden cardiac death

sudden cardiac death



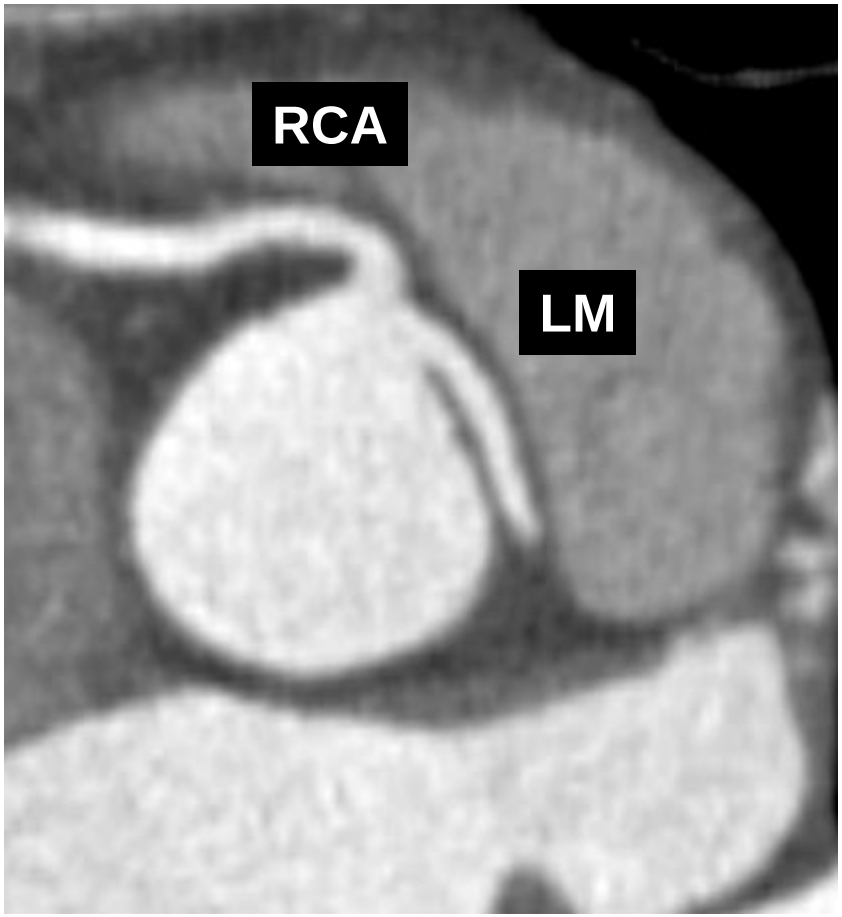
Carrado D. Br Heart J 1992

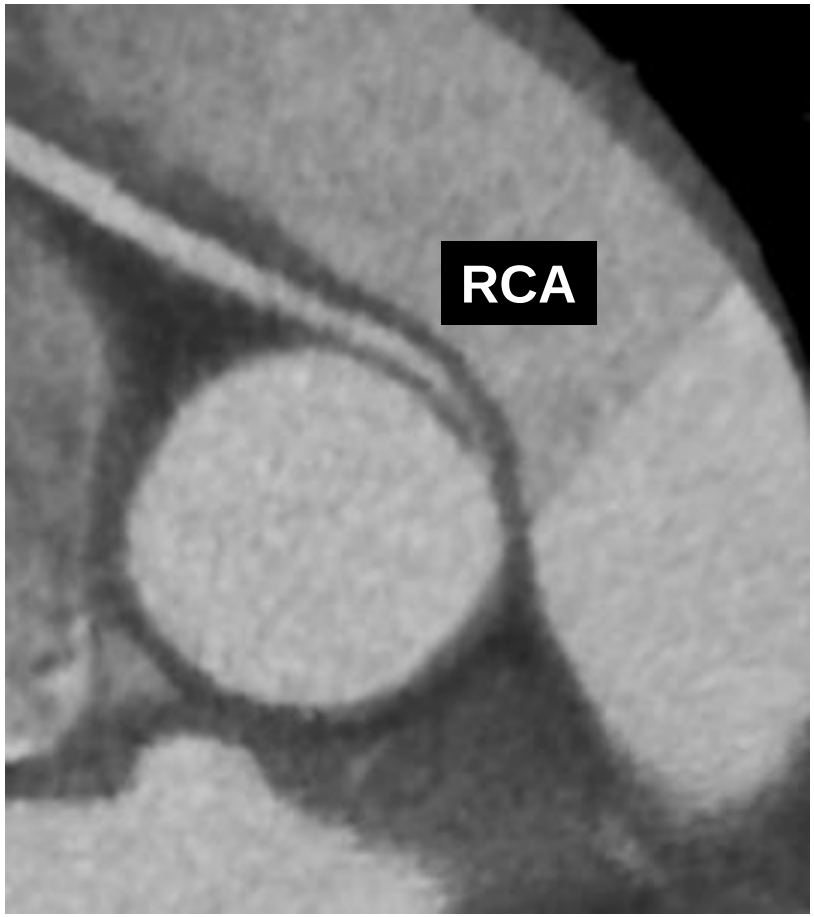
aborted cardiac arrest



Shimizu T. Intern Med 2014

Anatomic features with risk of SCD

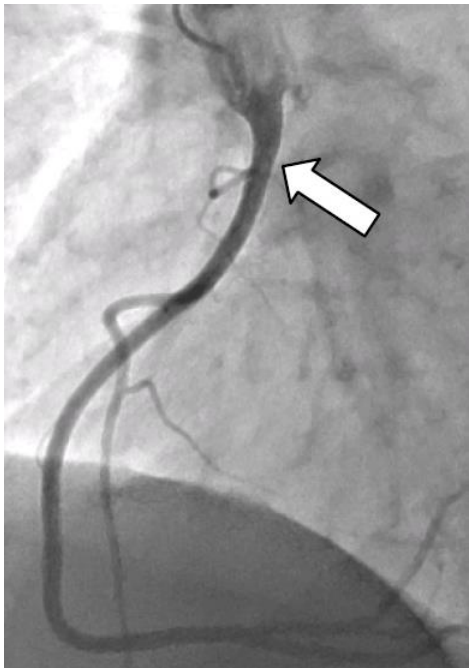


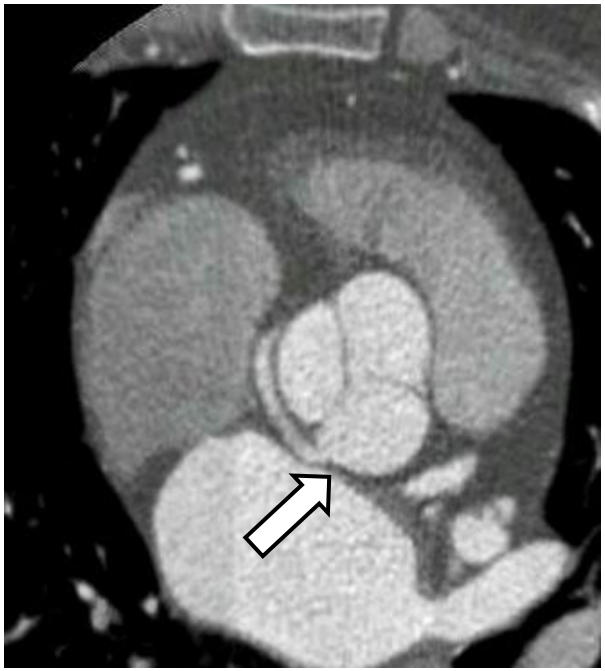


anomalous connection from the opposite sinus with interarterial course

Registry

Anomalous connection of RCA	165
Interarterial course	148 (90%)
Other courses	17 (10%)





Aubry P. Eur Heart J 2015

Risk of sudden cardiac death in athletes

- 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 (coronary anomaly)

Corrado D. JAMA 2006

Sudden cardiac death and ACAOS

annual risk of sudden cardiac death (estimation*)

L-ACAOS	**0.1%**
R-ACAOS	**0.005%**

total annual incidence = 0.2%

* general population

CHD with risk of sudden cardiac death (estimation)

CONGENITAL HEART DISEASE	SCD cases per 100 000 / year
Catecholaminergic polymorphic ventricular tachycardia	1500
Hypertrophic cardiomyopathy	1000-2000
Brugada syndrome	1000
Long QT syndrome	500-1000
Idiopatic dilated cardiomyopathy	500-1000
Arrhythmogenic right ventricular cardiomyopathy	500-1000
Wolf-Parkinson-White syndrome	100
Anomalous connection of left coronary artery	**100**
Anomalous connection of right coronary artery	**5**

Sudden cardiac death and ACAOS

annual risk of sudden cardiac death (estimation*)

L-ACAOS	**0.3%**
R-ACAOS	**0.01%**

* population with ages 12-35 years
Brothers J. J Thorac Cardiovasc Sur 2010

Registry

12 aborted SCD (2.5%)

2 ACAOS-related SCD (0.4%)

number	age	artery	connection	course	significant CAD
1	50	Cx	contralateral artery	retroaortic	present
2	75	Cx	contralateral artery	retroaortic	present
3	72	Cx	contralateral artery	retroaortic	present
4	16	LM	pulmonary artery	normal	absent
5	53	Cx	contralateral artery	retroaortic	present
6	48	Cx	contralateral artery	retroaortic	absent
7	57	CX	contralateral artery	retroaortic	present
8	60	RCA	ascending aorta	preaortic	present
9	31	RCA	contralateral sinus	preaortic	absent
10	60	RCA	contralateral sinus	preaortic	present
11	30	RCA	contralateral sinus	preaortic	absent
12	44	CX	contralateral sinus	retroaortic	absent

Aubry P. Arch Cardiovasc Dis 2016

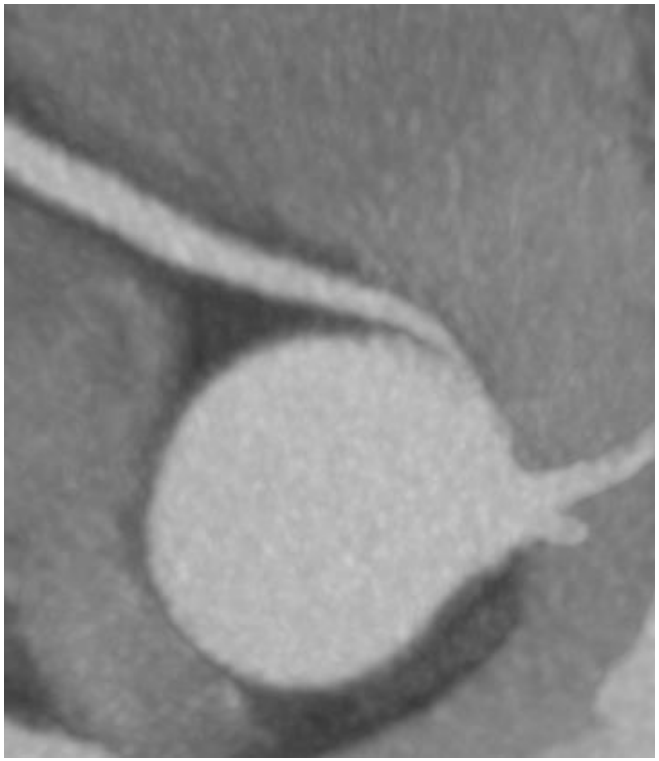
anomalous connection of the RCA with interarterial course

mechanism(s) of ventricular fibrillation

The exact mechanism of sudden cardiac death associated with
anomalous connection of coronary artery **is not known**

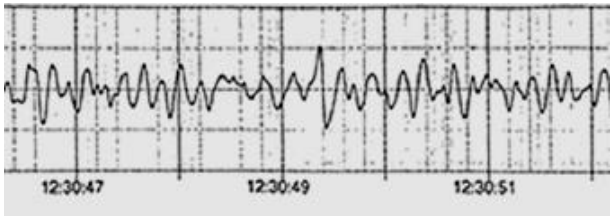
anomalous connection of the RCA with interarterial course

mechanism(s) of ventricular fibrillation



?

24/07/2012
12.28 AM

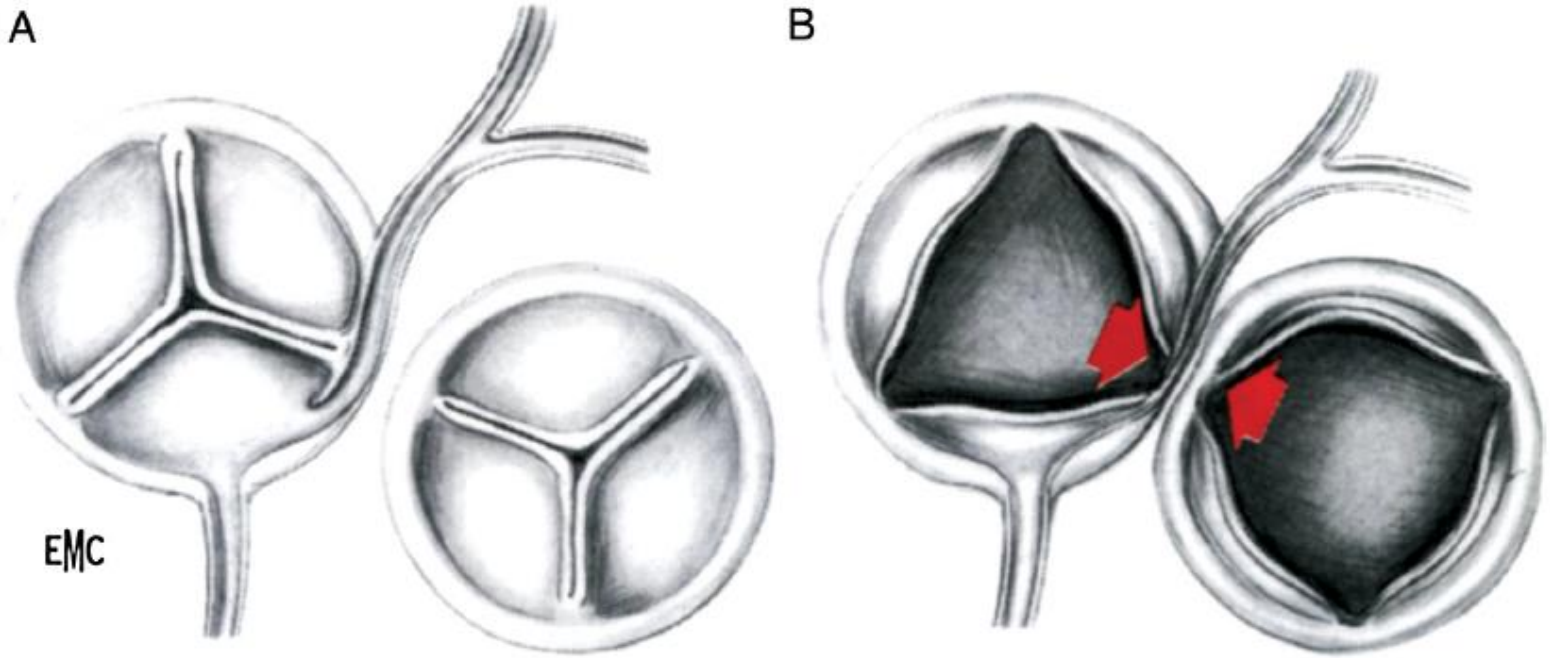


mechanism(s) of the ventricular fibrillation

- myocardial ischemia
- limitation of coronary reserve
- myocardial fibrosis area
- abnormal arrhythmic response to ischemia
- hypotension post exercise
- combination of mechanisms
- fortuitous association
- …

anomalous connection of the RCA with interarterial course

Exertional dynamic compression



Raisky O, Vouhé P. EMC 2007

Never demonstrated

2015 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

The Task Force for the Management of Patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death of the European Society of Cardiology (ESC)

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

Anomalous connections of coronary arteries : **not mentioned**

Priori SG. Eur Heart J 2015

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

A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society

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

Recommendations for Surgery and Revascularization Procedures in Patients With Ischemic Heart Disease

References that support the recommendations are summarized in Online Data Supplement 11.

COR	LOE	Recommendations
I	B-NR	1. Patients with sustained VA and survivors of SCA should be evaluated for ischemic heart disease, and should be revascularized as appropriate (1-4).
I	C-EO	2. In patients with anomalous origin of a coronary artery suspected to be the cause of SCA, repair or revascularization is recommended.

Al-Khatib SM, et al.

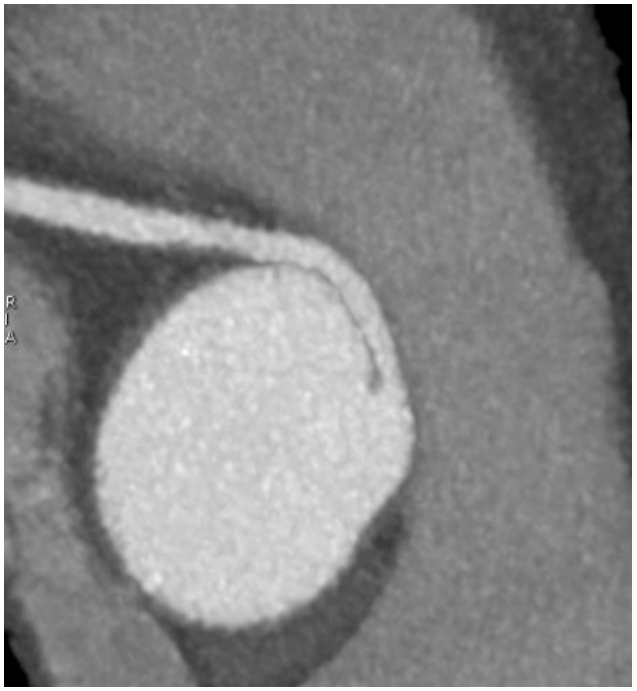
(*Circulation*. 2017;000:e000–e000. DOI: 10.1161/CIR.0000000000000548.)

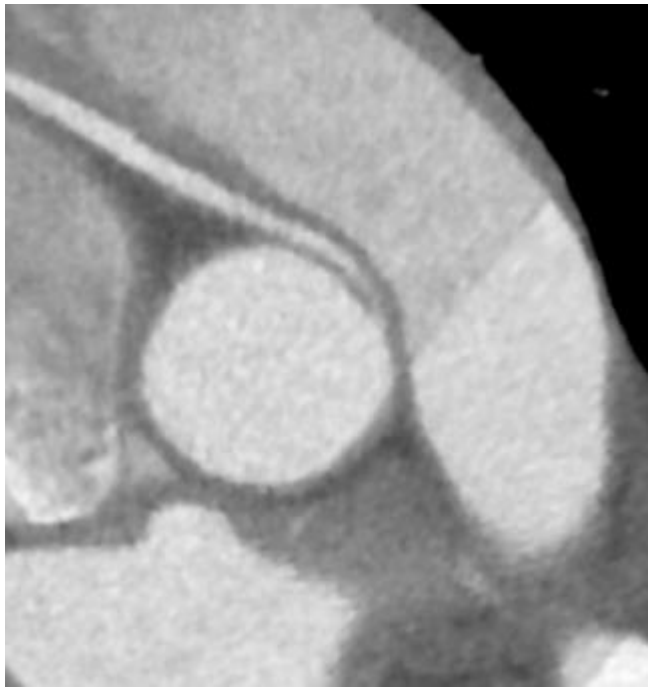
anomalous connection of the RCA with interarterial course

ISCHEMIA/SYMPTOMS

possible in patients >35 year-old

anomalous connection of the RCA with interarterial course

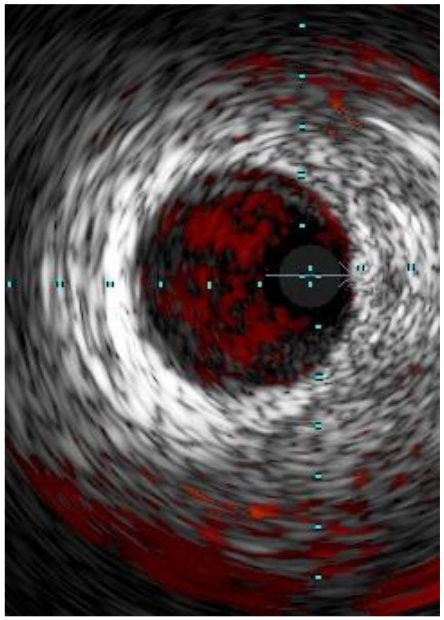




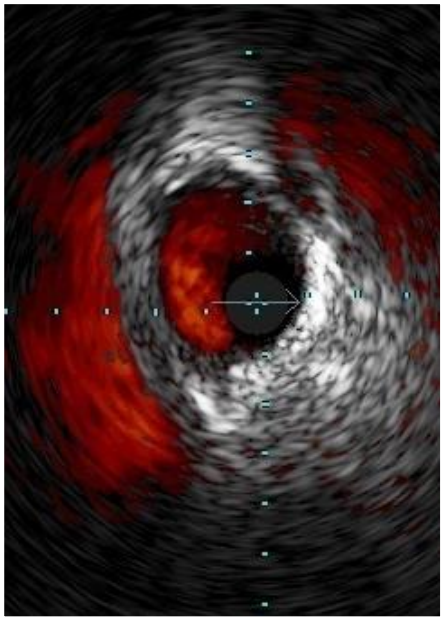
anomalous connection of the RCA with interarterial course

IVUS evaluation of R-ACAOS

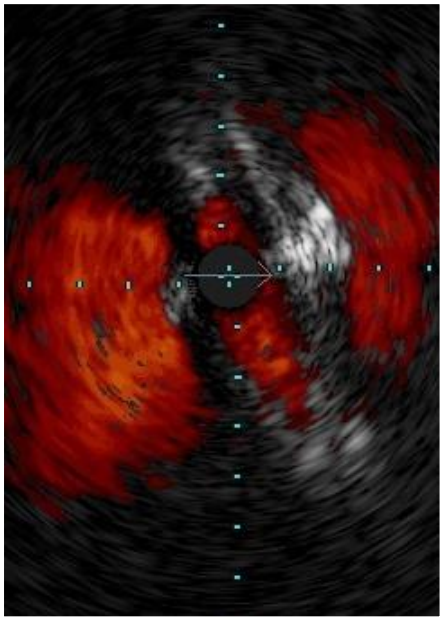
extramural



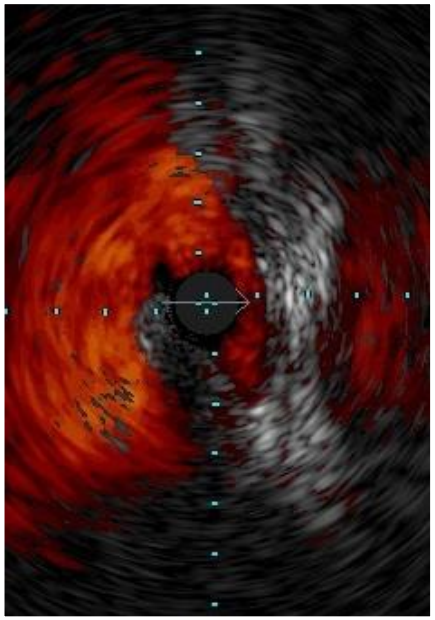
juxtamural



intramural

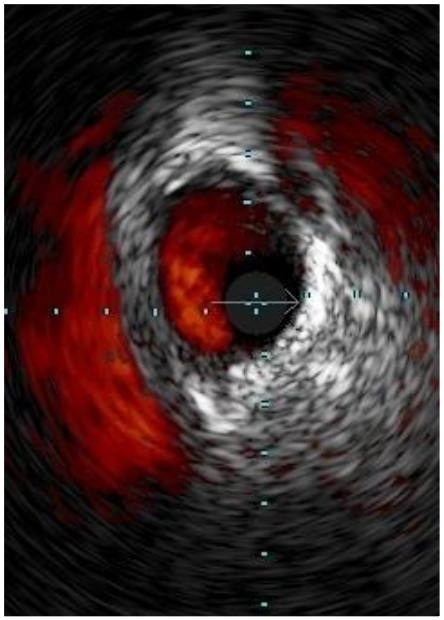


ostium

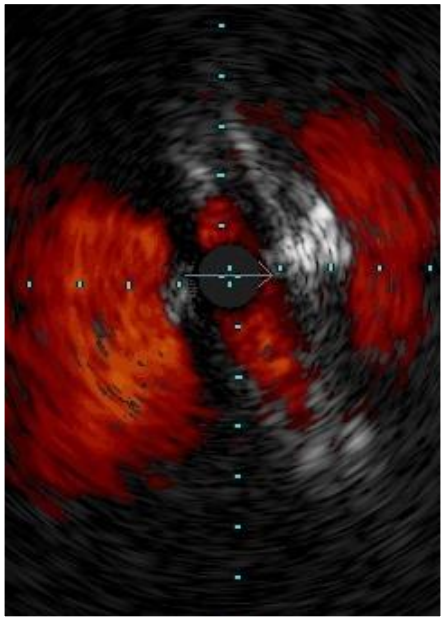


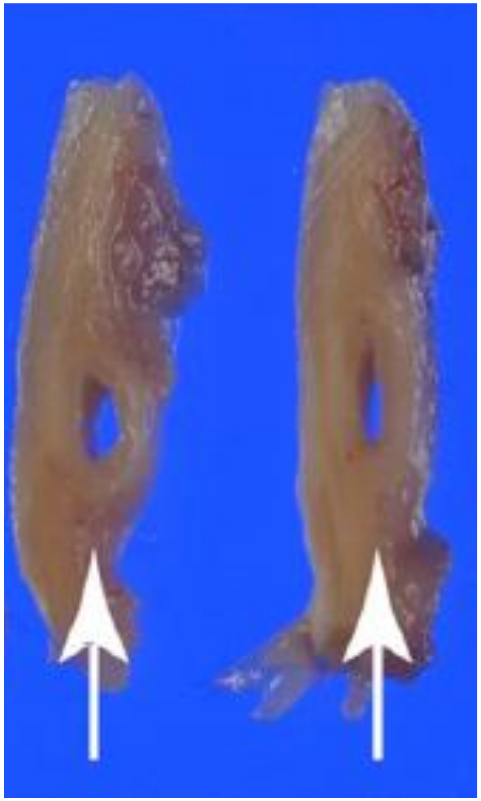
IVUS evaluation of R-ACAOS

juxtamural



intramural

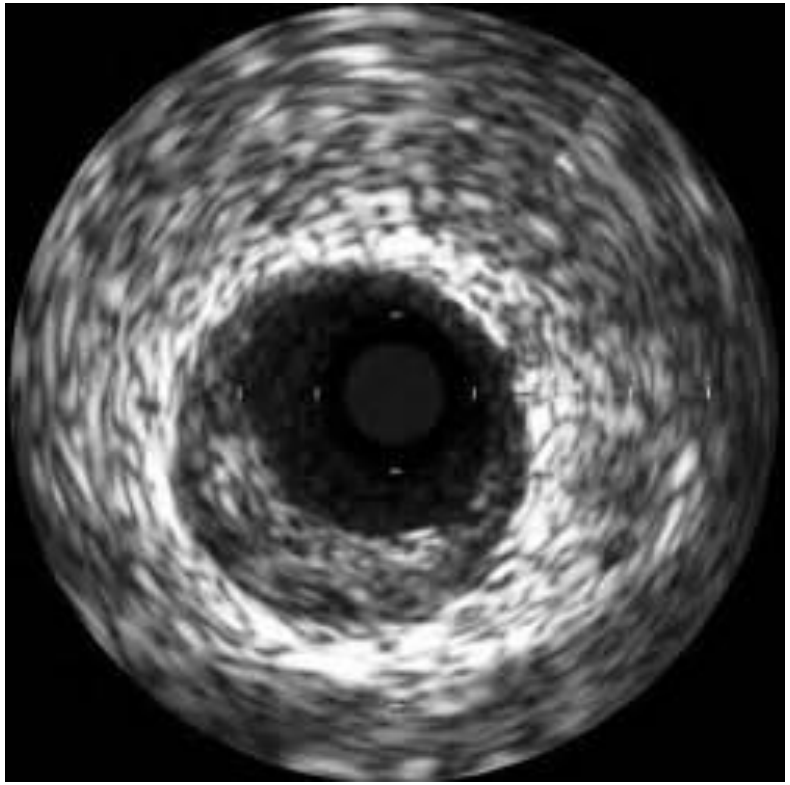




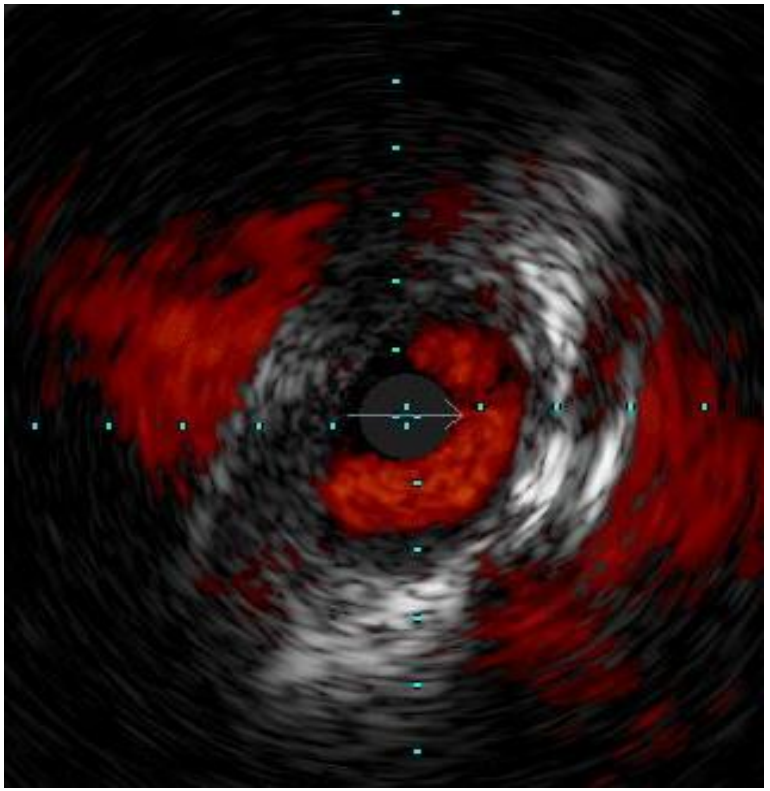
R-ACAOS with intramural course
Hata Y et al. Cardiovasc Pathol 2014

mechanism(s) of myocardial ischemia

CAD

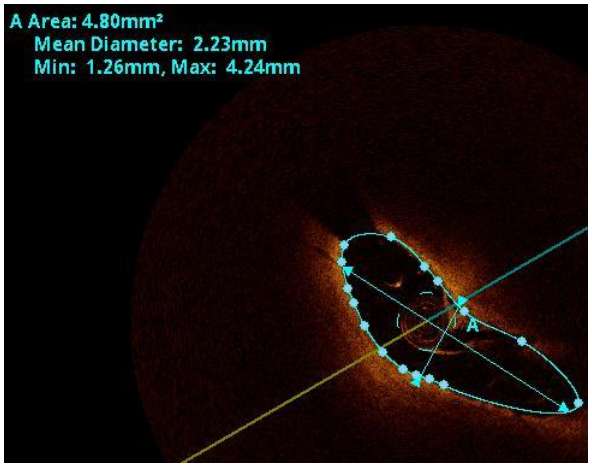


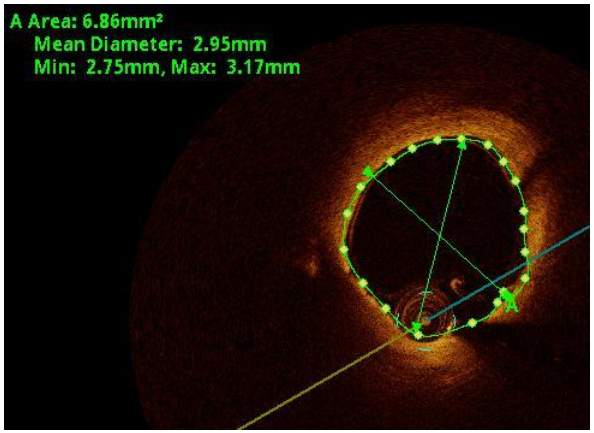
R-ACAOS

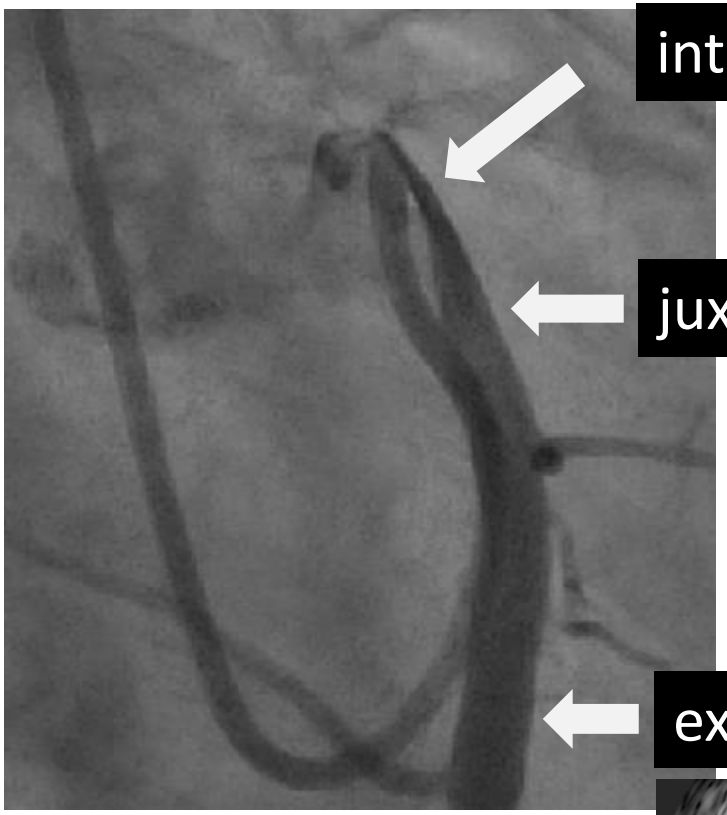


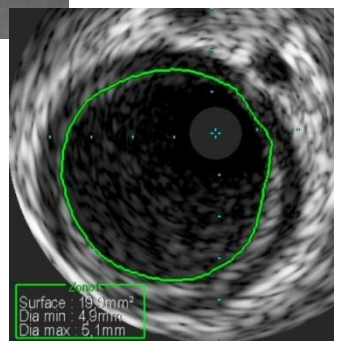
anomalous connection of the RCA with interarterial course

IVUS evaluation of R-ACAOS







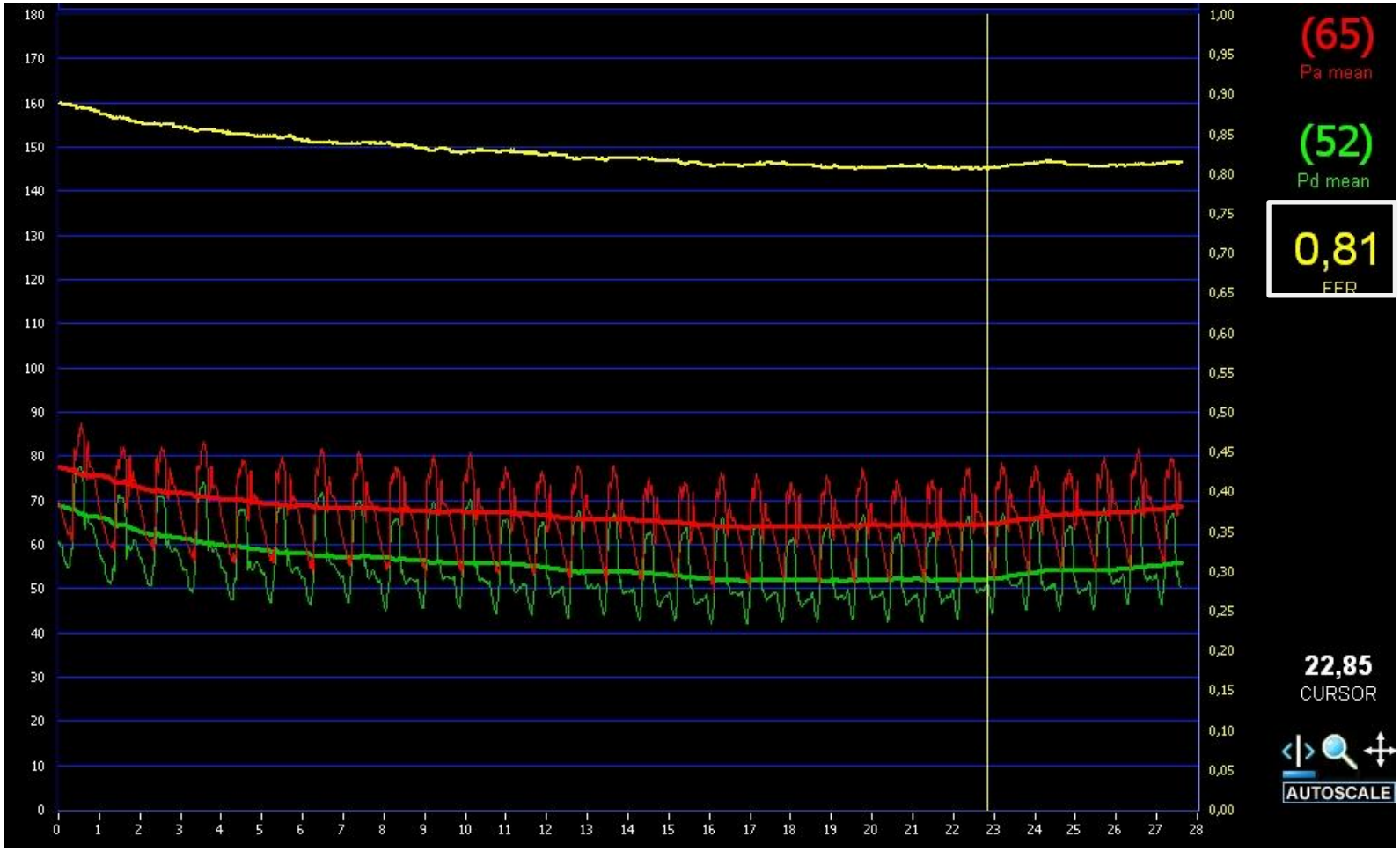


minimal lumen area: 4.8 mm^2

lumen area reduction: 75%

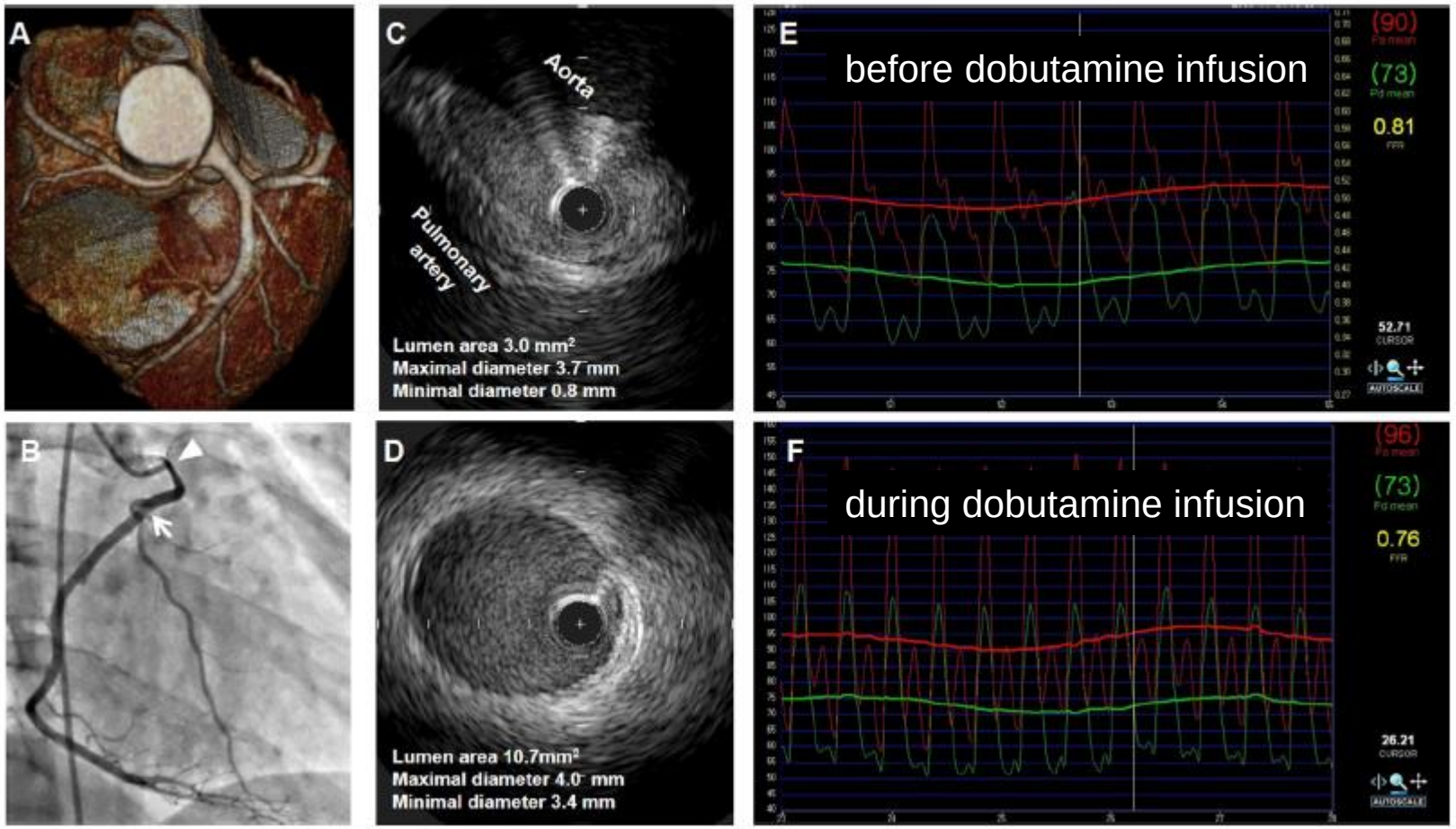
anomalous connection of the RCA with interarterial course

Fractional Flow Reserve (FFR)



Physiological and clinical relevance of anomalous right coronary artery originating from left sinus of Valsalva in adults

Sang Eun Lee,[1] Cheol Woong Yu,[2] Kyungil Park,[3] Kyung Woo Park,[1] Jung-Won Suh,[4] Young-Seok Cho,[4] Tae-Jin Youn,[4] In-Ho Chae,[4] Dong-Ju Choi,[4] Ho-Jun Jang,[5]



Lee SE, *et al*. *Heart* 2016;**102**:114–119. doi:10.1136/heartjnl-2015-308488

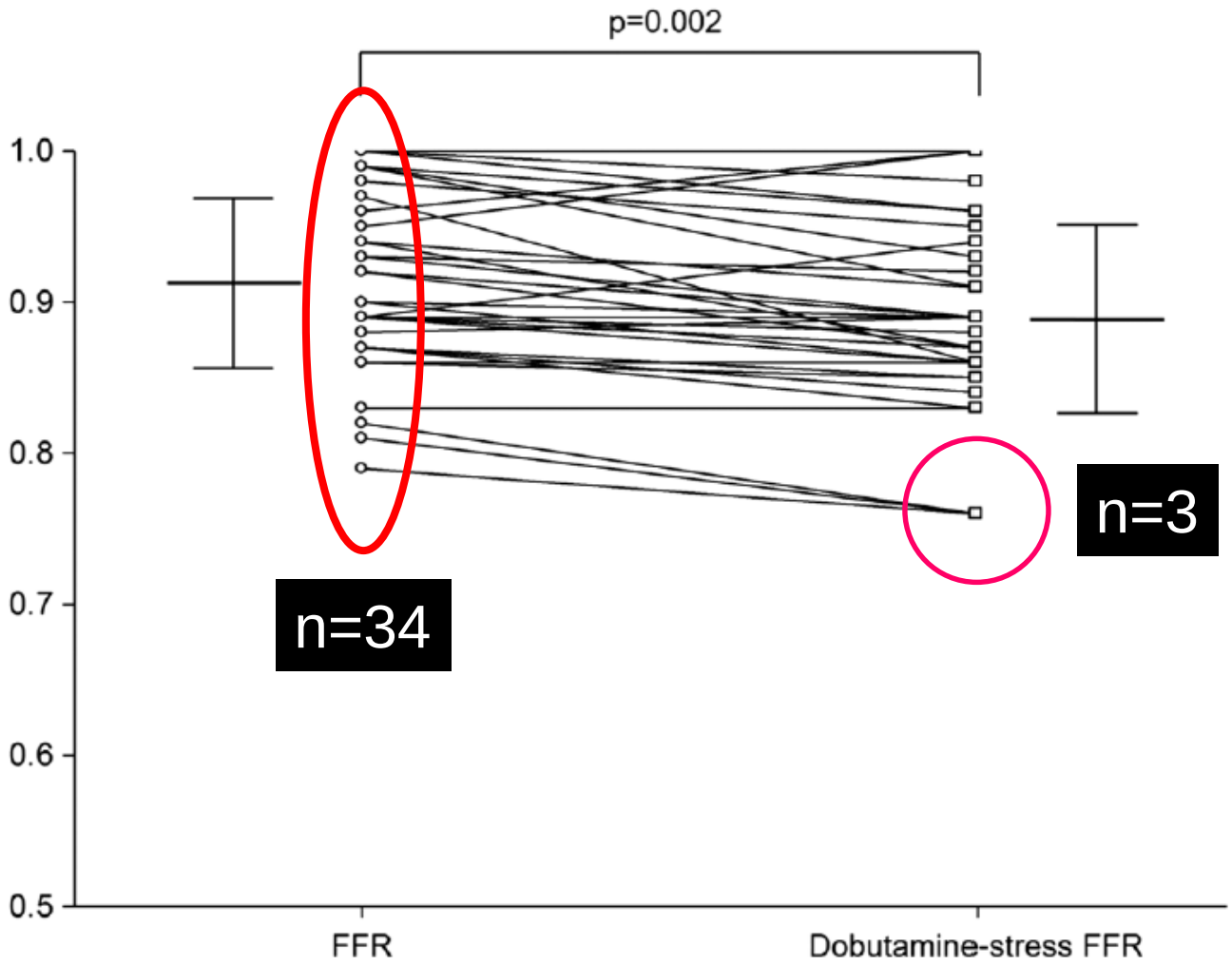
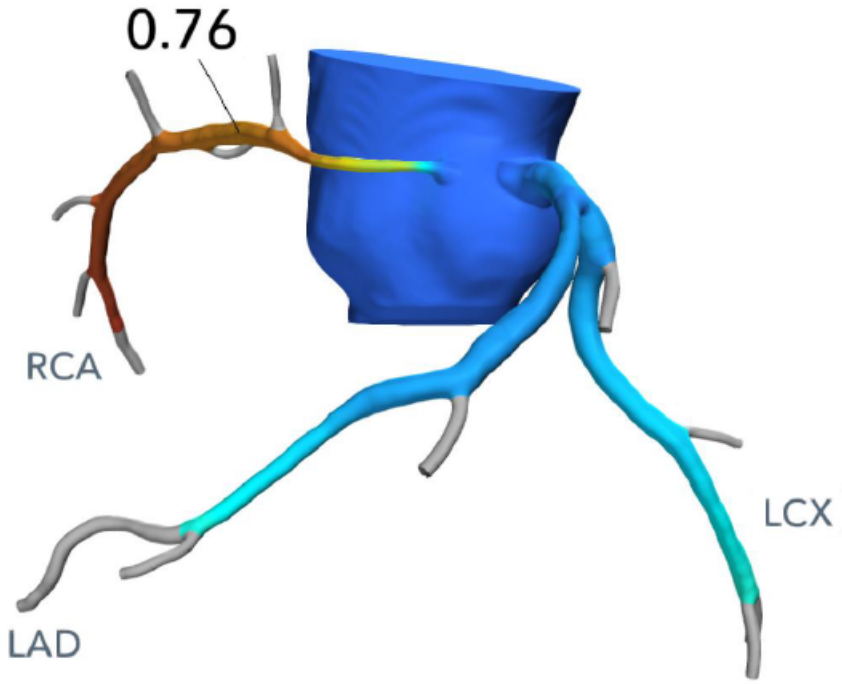


Figure 2 Changes in fractional flow reserve (FFR) with dobutamine infusion. p Value from Wilcoxon signed rank test.

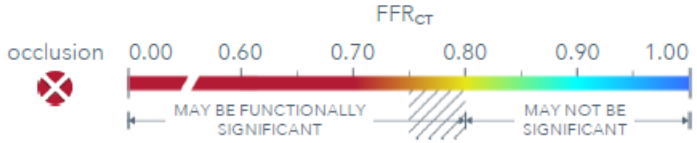
Lee SE, *et al*. *Heart* 2016;**102**:114–119. doi:10.1136/heartjnl-2015-308488

HeartFlow® FFR$_{CT}$

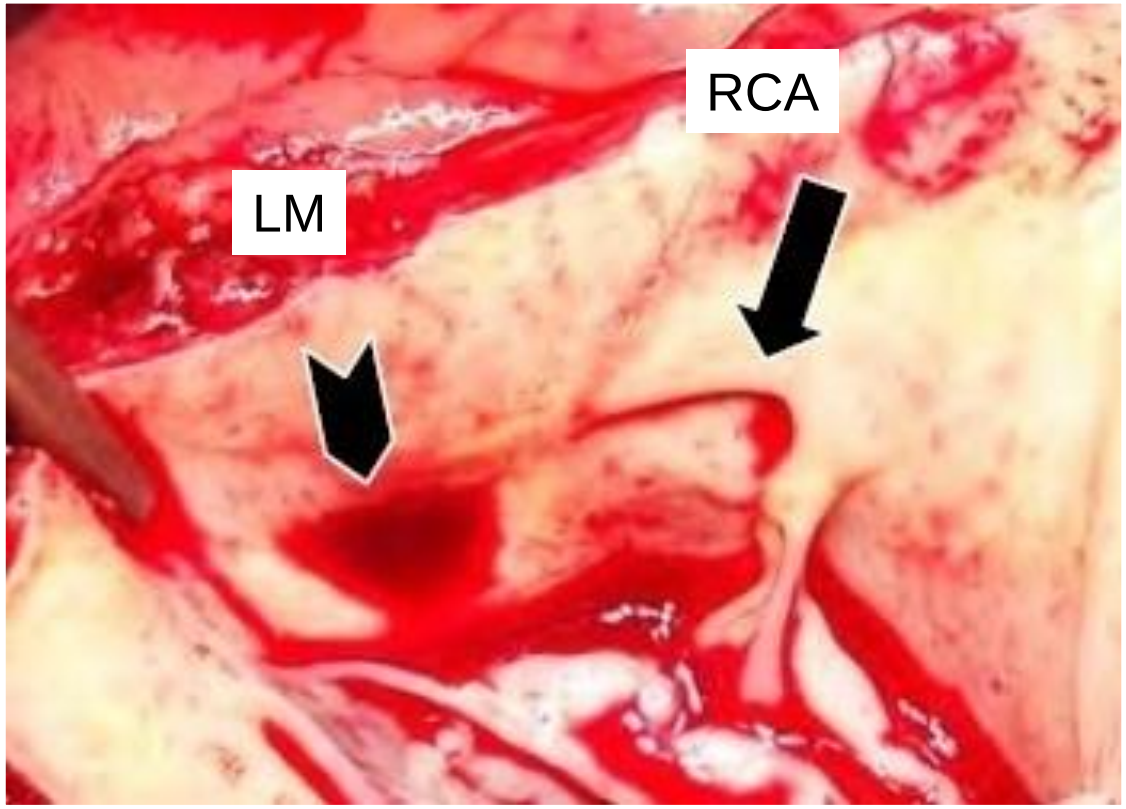
R-ACAOS



FFR$_{CT}$ values are specified distal to modeled stenoses > 30%.



anomalous connection of the RCA

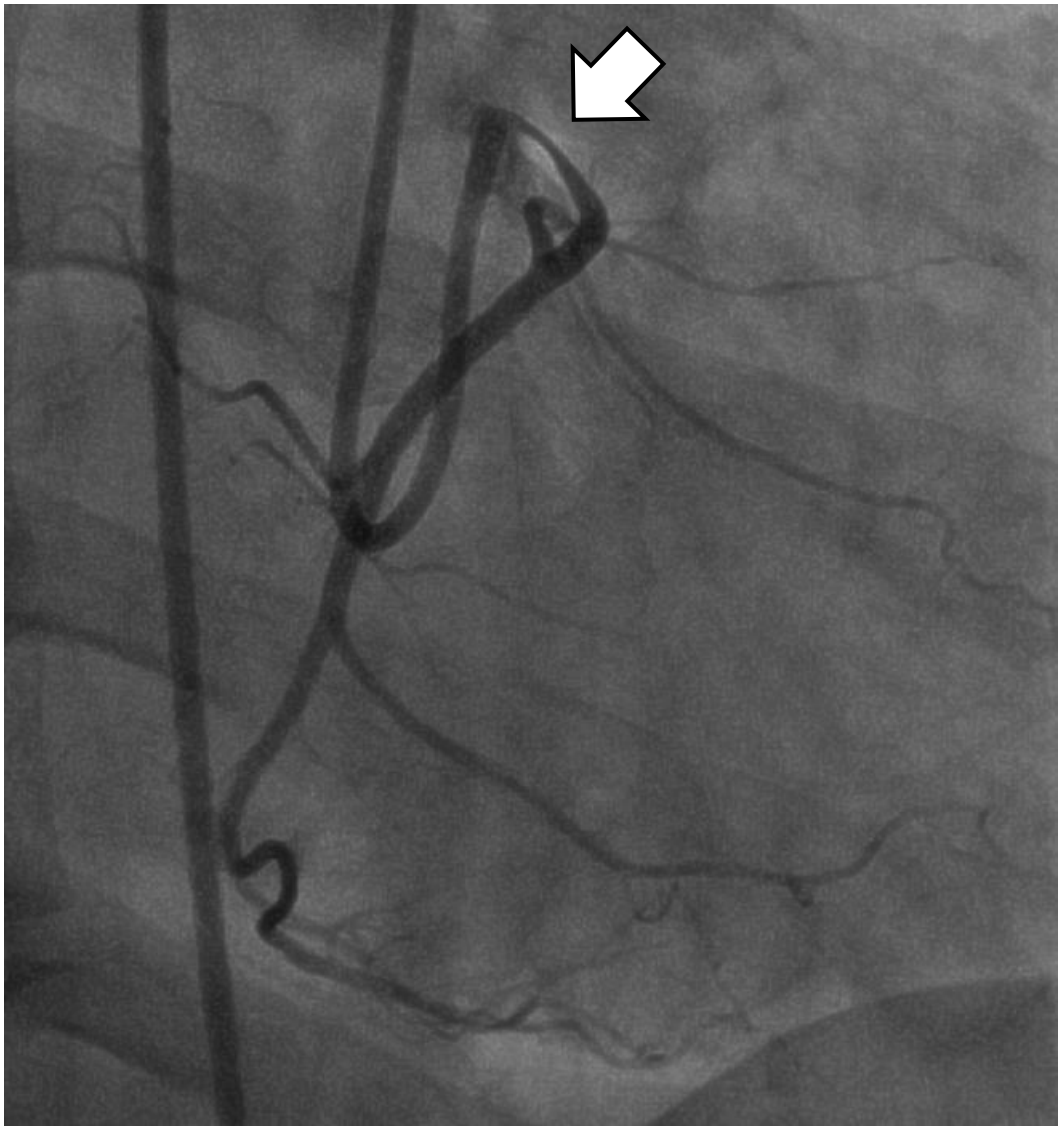


Hata Y Cardiovasc Pathol 2014

PCI in anomalous connections without CAD

- accurate diagnosis of the anomalous connection
- identification of abnormalities requiring correction
- place of PCI?

PCI in anomalous connections of the coronary arteries (ANOCOR)



right anomalous connection without CAD

anomalous connection of the RCA with interarterial course

CURRENT MANAGEMENT

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

8.5. Recommendations for Congenital Coronary Anomalies of Ectopic Arterial Origin

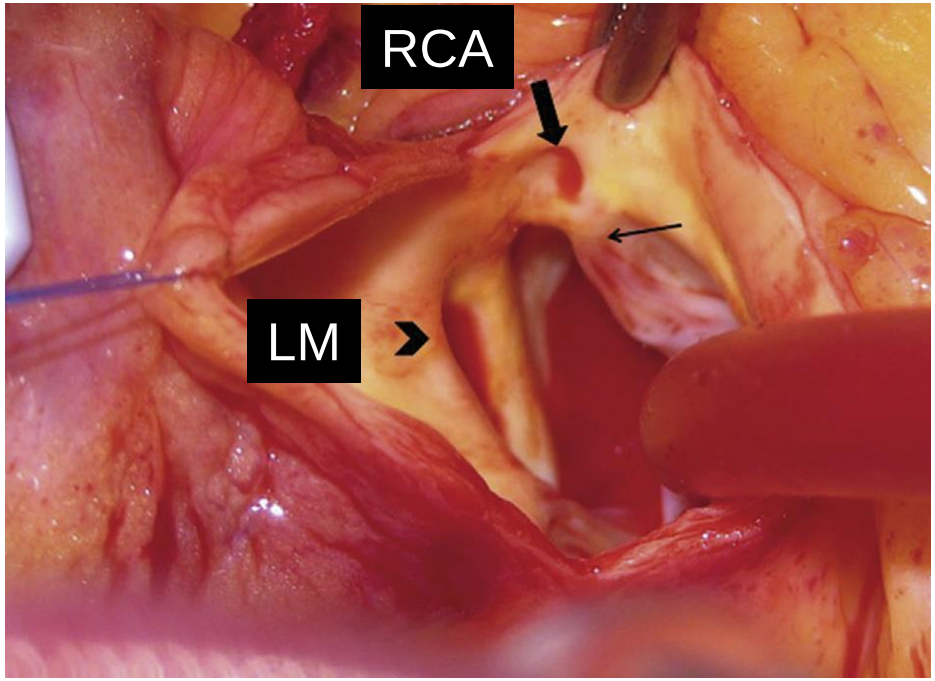
Circulation December 2, 2008

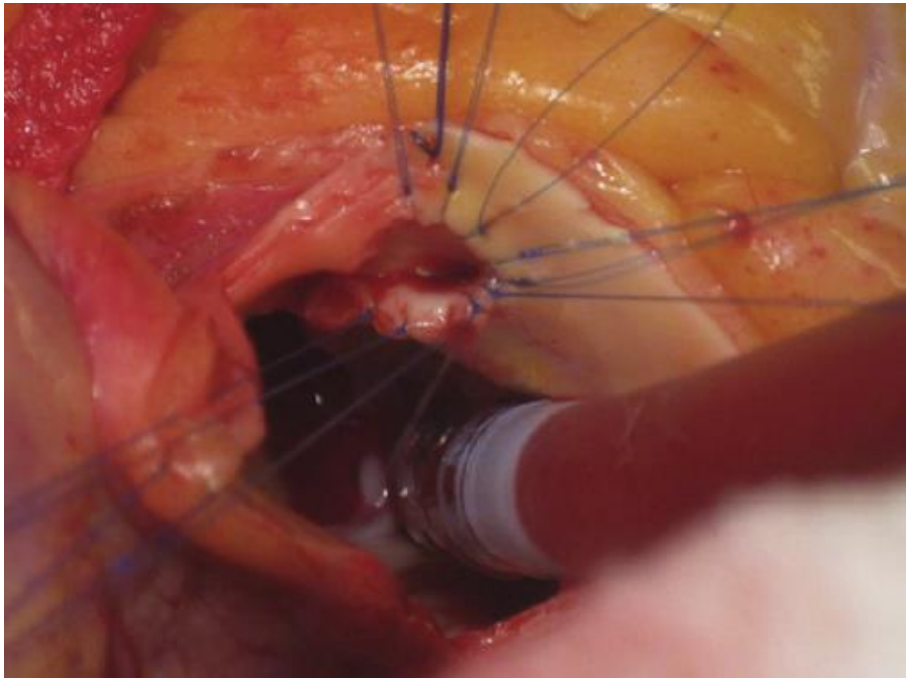
CLASS I

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*)

anomalous connection of the RCA with interarterial course

Unroofing with creation of neo-ostium





Feins EN et al. Ann Thorac Surg 2016

Weaknesses of surgical repair

- Guidelines focused on young people
- Very few patients with history of sudden death
- No randomized controlled studies
- Lack of long-term data after correction
- Possible failure (stenosis/aneurysm/thrombosis)

http://dx.doi.org/10.1016/j.jtcvs.2016.06.066

Expert consensus guidelines: Anomalous aortic origin of a coronary artery

Julie A. Brothers, MD,[a] Michele A. Frommelt, MD,[b] Robert D. B. Jaquiss, MD,[c] Robert J. Myerburg, MD,[d] Charles D. Fraser, Jr, MD,[e] and James S. Tweddell, MD[f]

J Thorac Cardiovasc Surg 2017;153:1440-57

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,[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

- retrospective study with 42 ectopic RCA
- mean age 48±12 years (12-73)
- PCI with IVUS guidance (BMS/Cypher/Taxus/Promus stents)
- indications for angioplasty:

  symptoms/ischemia

  or intensive sport practice

  or IVUS surface reduction >50%
- angiographic success (100%)
- no in-hospital MACE
- angiographic restenosis (4/42)

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

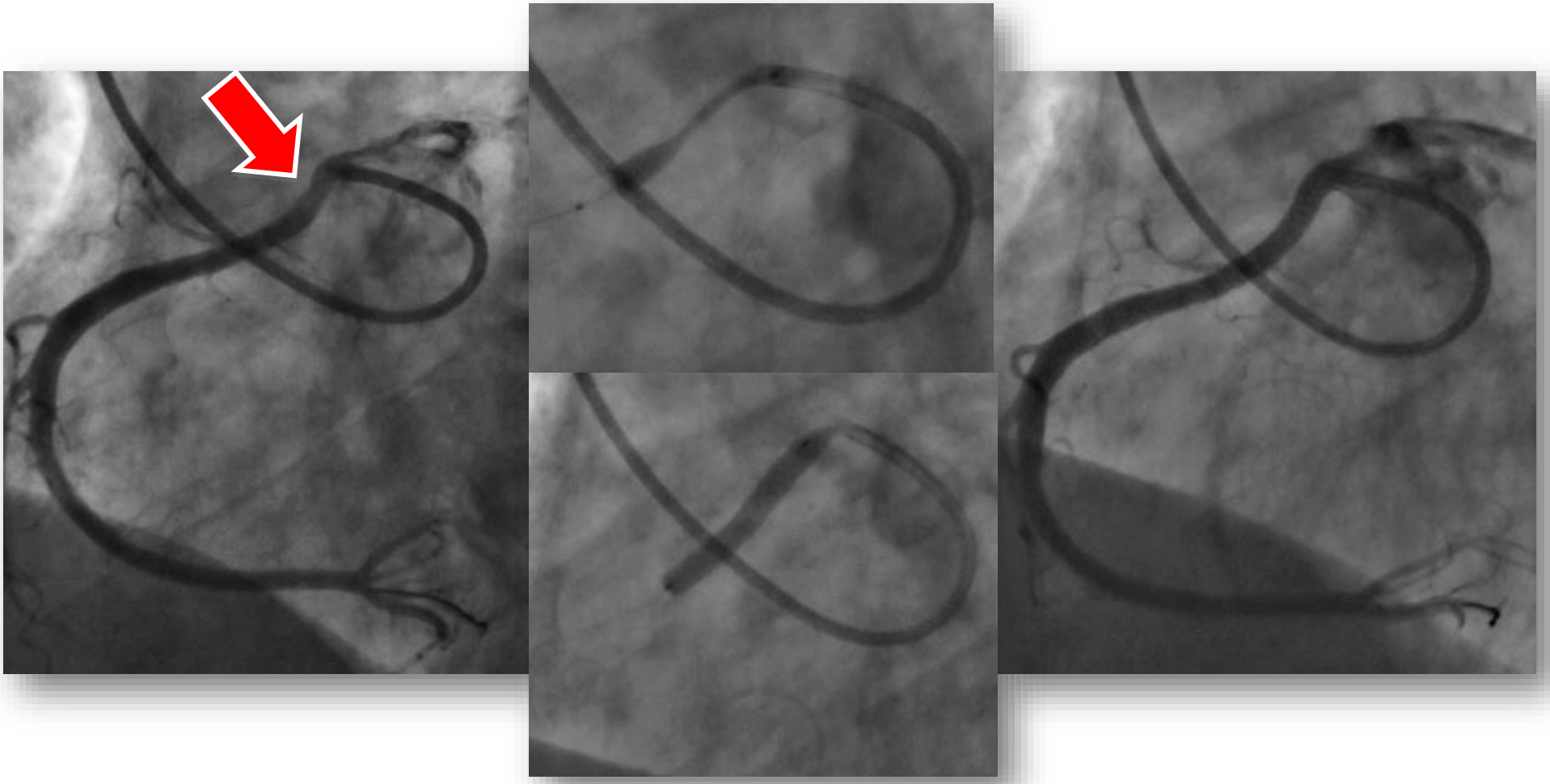
ANOCOR stenting registry (2015)
multidisciplinary team

selected population

- right anomalous connection
- age >35 year-old
- no history of aborten sudden death
- angina and/or documented ischemia
- pre aortic course with/without intramural pathway
- no significant CAD associated

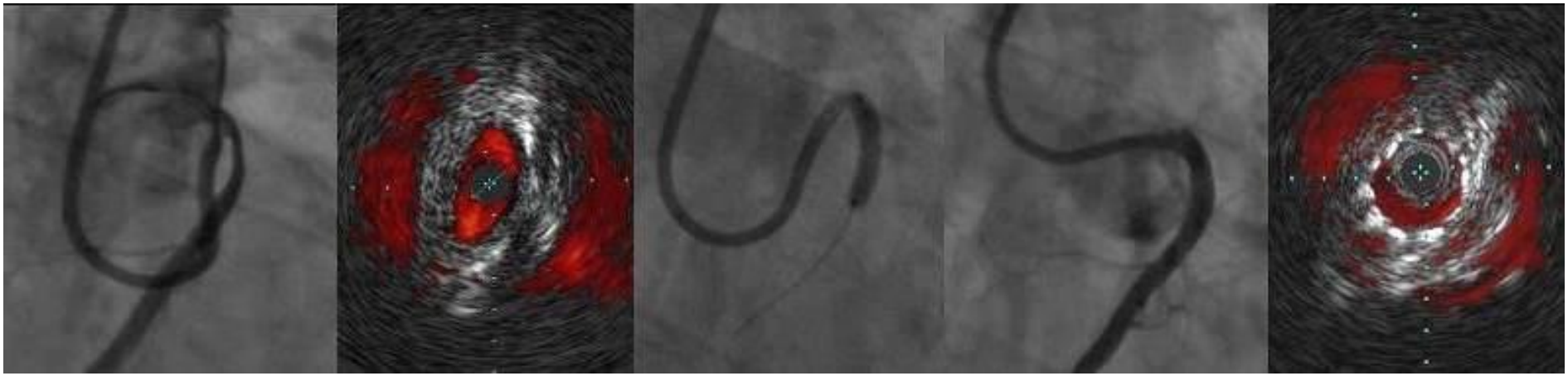
PCI in anomalous connections of the coronary arteries (ANOCOR)

stenting of ectopic right coronary artery

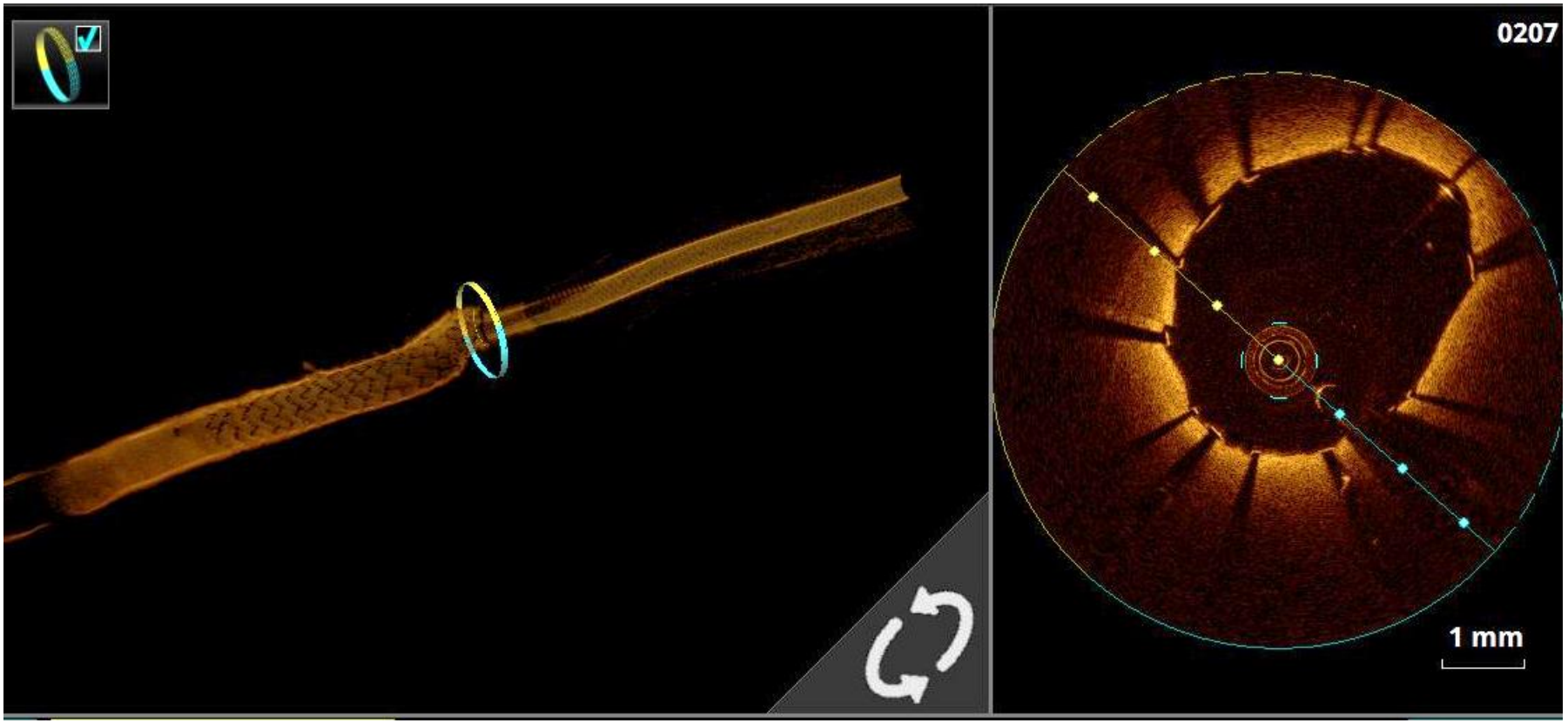


PCI in anomalous connections of the coronary arteries (ANOCOR)

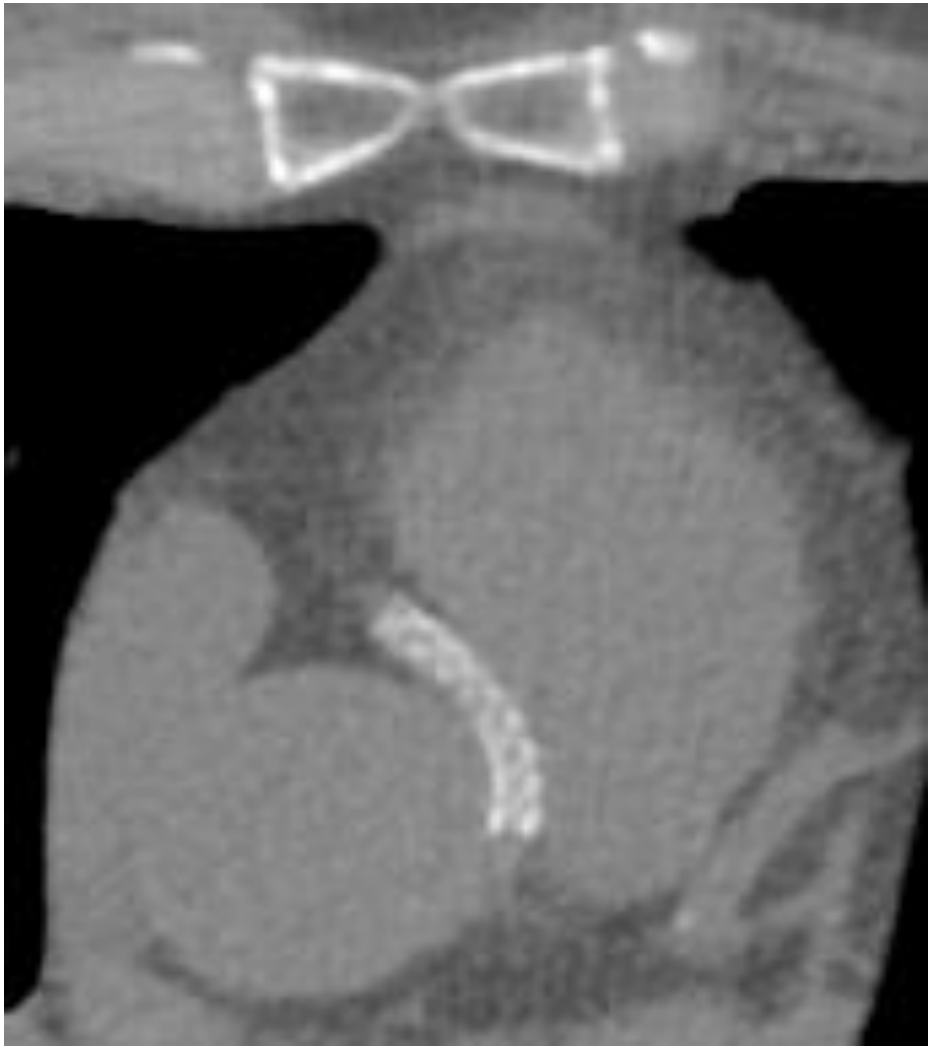
stenting of ectopic right coronary artery

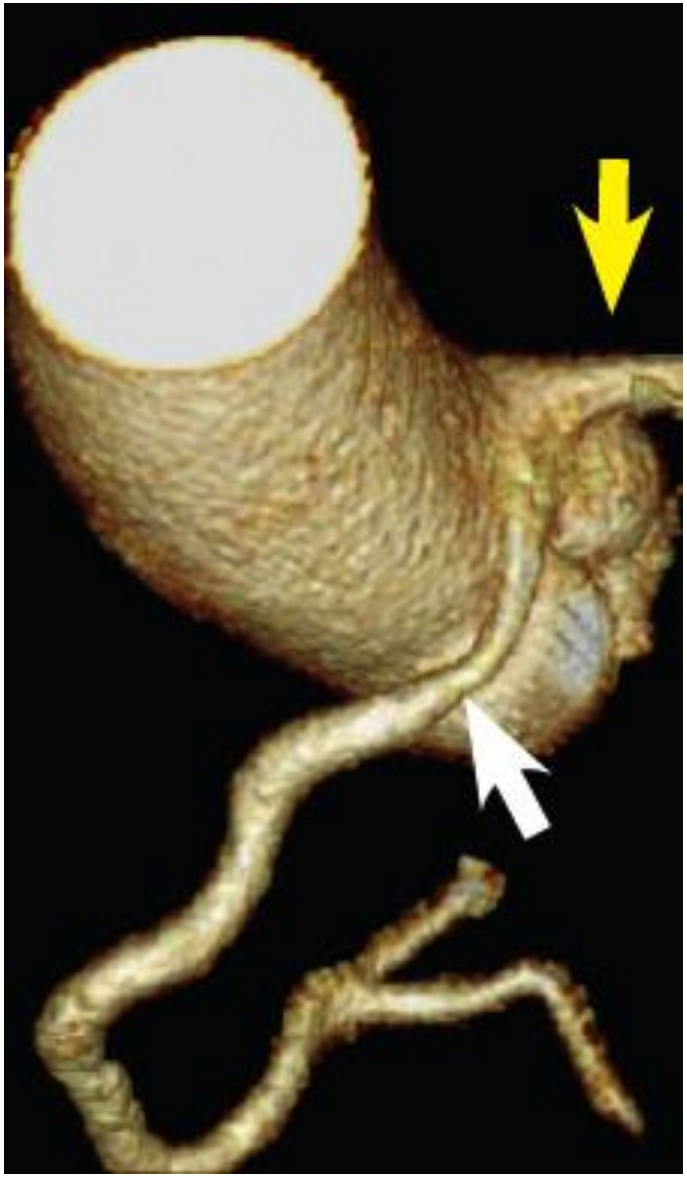


OCT evaluation of R-ACAOS stenting



CT scan at 6 months





anomalous connection of the RCA with interarterial course

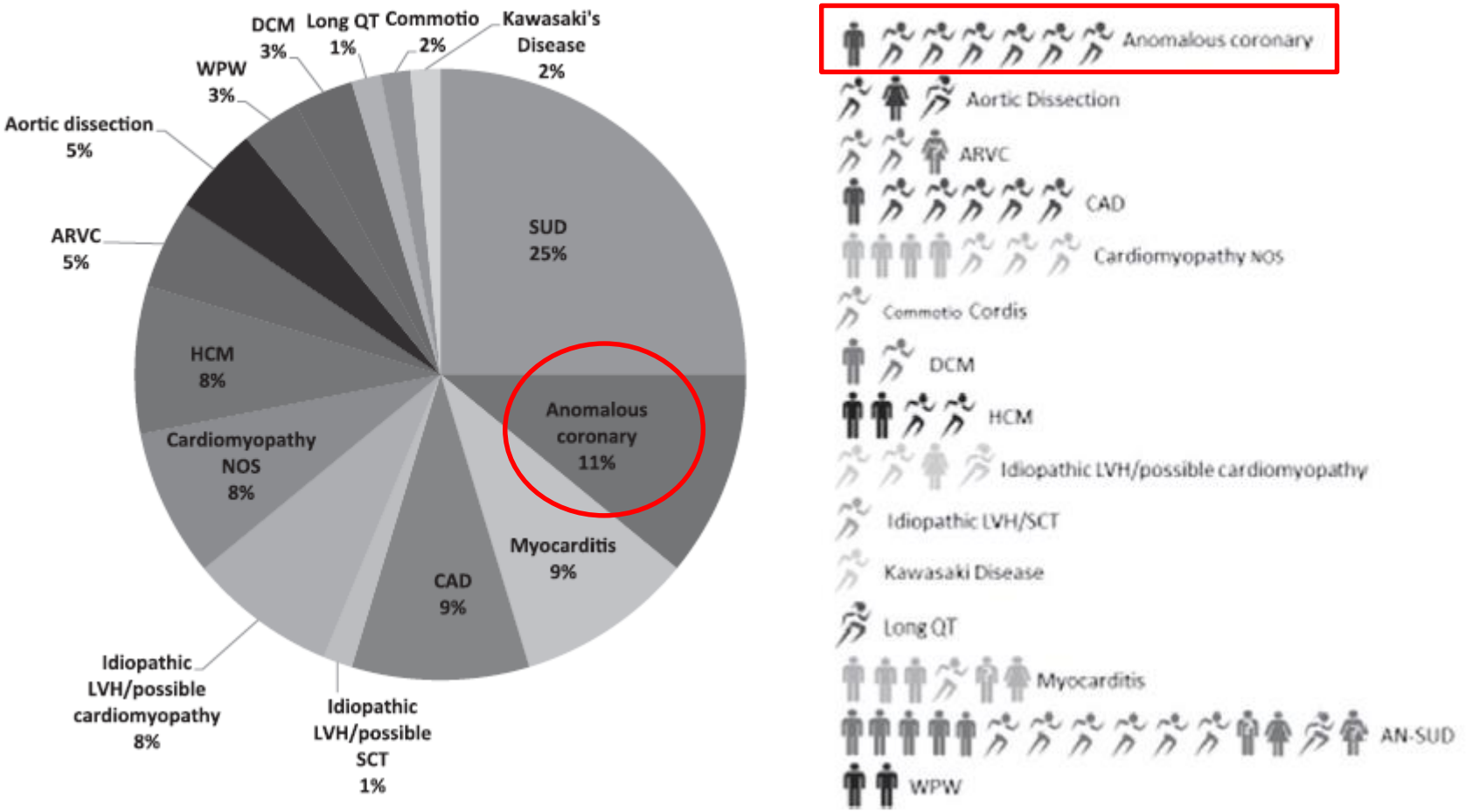
How to manage?

- Age <35 or ≥35 year-old
- History of aborted cardiac arrest
- Symptoms / relation with exertion
- Non invasive documented myocardial ischemia
- Anatomic features (CT/angio/IVUS)
- FFR (invasive / non-invasive)
- Competitive activities

Surgery/PCI/observation/exercise restriction

Incidence, Cause, and Comparative Frequency of Sudden Cardiac Death in National Collegiate Athletic Association Athletes

A Decade in Review



Harmon KG. Circulation 2015

European Journal of Preventive Cardiology

EHRA Position paper

Pre-participation cardiovascular evaluation for athletic participants to prevent sudden death: Position paper from the EHRA and the EACPR, branches of the ESC. Endorsed by APHRS, HRS, and SOLAECE

European Journal of Preventive Cardiology
2017, Vol. 24(1) 41–69

DOI: 10.1177/2047487316676042
ejpc.sagepub.com
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Coronary congenital abnormalities

Mons L. Eur J Prev Cardiol 2017

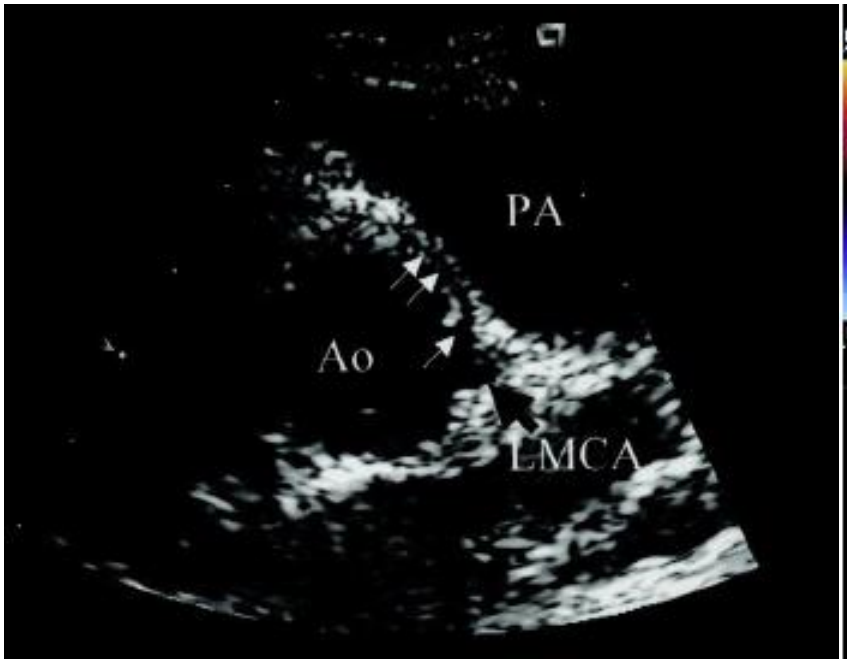
PRE-PARTICIPATION CARDIOVASCULAR EVALUATION

- History
- Physical examination

- History
- Physical examination
- ECG

anomalous connection of the RCA with interarterial course

R-ACAOS (transthoracic echocardiography)



Frommelt PC. J Am Coll Cardiol 2003

PRE-PARTICIPATION CARDIOVASCULAR EVALUATION

Cost-efficacy of pre-participation evaluation

Table 4 Studies assessing cost-efficacy of PPE

Study	Target population	Cost/life-year	Cost/life saved	Comments
Fuller (2000)[172]	High School Athletes	H&P $84 000 ECG: $44 000	N/R	Screened once at study entry
Maron *et al.* (2007)[173]	High school and middle school athletes	N/R	$3 400 000	Approximate estimation in a non-dedicated paper
Wheeler *et al.* (2010)[174]	High school and college athletes (14–22 years)	H&P: 199 000$ H&P&ECG: $76 100	N/R	Screened once at study entry
Halkin *et al.* (2012)[171]	Registered high school, college, and professional participants	N/R	$10 600 000–$14 400 000	Annual screening
Leslie *et al.* (2012)[175]	High school (≥14 years) freshmen participating in organized sports.	$91 000	N/R	Screened once at study entry
Schoenbaum *et al.* (2012)[176]	Athletes ≥14 years	Adding ECG to H&P: +$68 000	$900 000	
Corrado *et al.* (2013)[177]	Young (12–35 yo) athletes	~$67 000	~$1 350 000	Approximate estimation in a non-dedicated paper
Assanelli *et al.* (2015)[178]	European and Algerian athletes seeking a sports medical certificate	Europe: 4071 $PPP Algeria: 582 $PPP	N/R	Estimation on prospectively collected data

All costs in US dollars or purchasing-power-parity-adjusted US dollars.
N/R, Not reported.

Mons L. Eur J Prev Cardiol 2017

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

AHA/ACC SCIENTIFIC STATEMENT

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

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

George F. Van Hare, MD, FACC, *Chair**
Michael J. Ackerman, MD, PhD, FACC*
Juli-anne K. Evangelista, DNP, APRN, CPNP-AC, FACC*
Richard J. Kovacs, MD, FAHA, FACC*
Robert J. Myerburg, MD, FACC*
Keri M. Shafer, MD*
Carole A. Warnes, MD, FACC*
Reginald L. Washington, MD, FAHA*

anomalous connection of the RCA with interarterial course

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)*.**

Van Hare GF. J Am Coll Cardiol 2015

R-ACAOS

- Prevalence in general population: **0.3%**
- Presence of interarterial course: **90%**
- Association with sudden cardiac death: **yes**
- Risk of sudden cardiac death: **0.005%/year**
- Vulnerability periods: **<35 year-old and sport**
- Mechanism of sudden cardiac death: **ventricular fibrillation**
- Mechanism(s) of ventricular fibrillation: **?**
- Primary prevention of sudden cardiac death: **?**
- Place of screening in athletes: **?**
- Ischemia/symptoms in patients >35 year-old: **possible**
- Management of patients >35 year-old: **?**
- Place of PCI: **?**

Thank you

pcaubry@yahoo.fr