Anomalous Coronary Arteries: Understanding High-Risk Features

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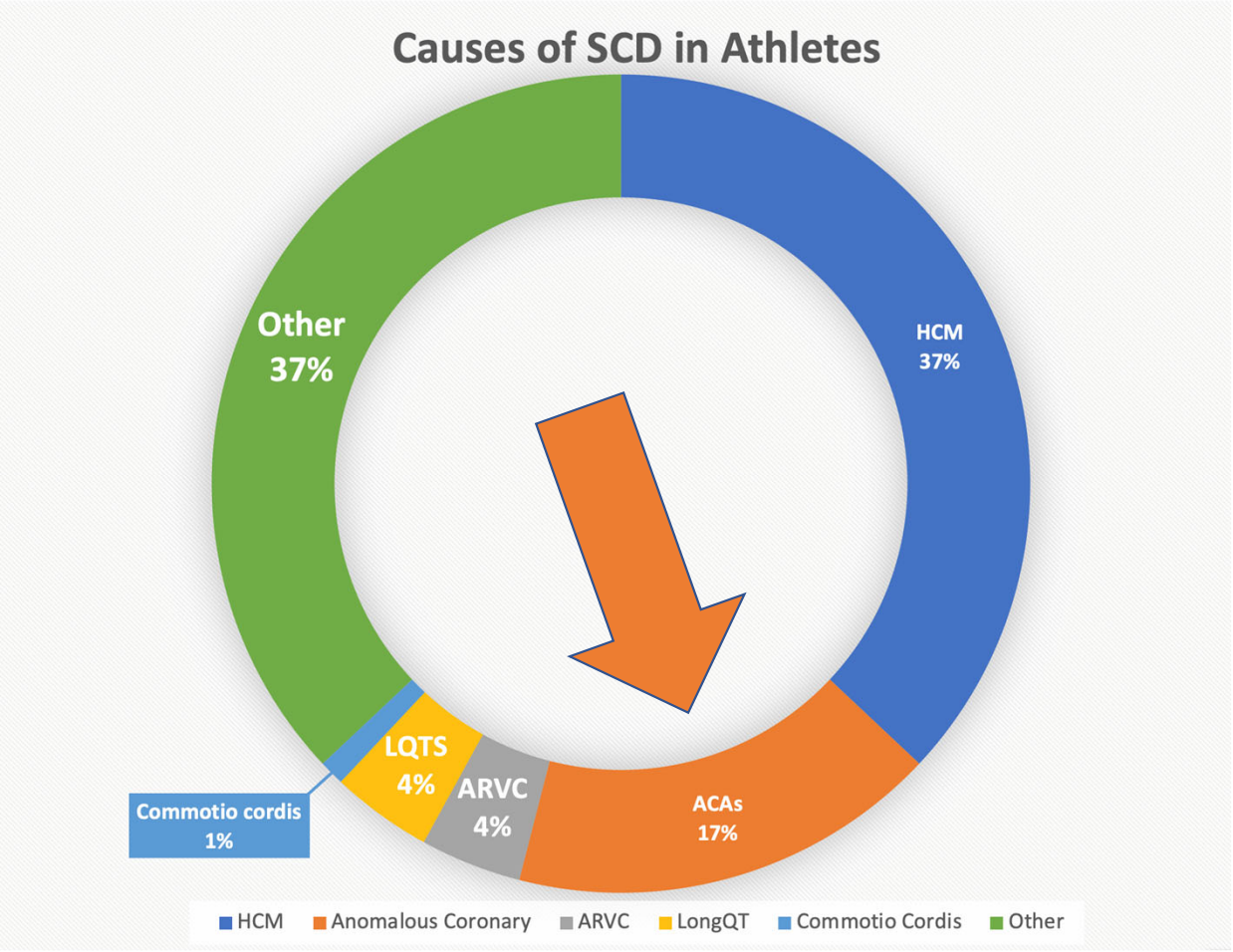
Learning Objectives

- Various types of anomalous coronary arteries
- High-risk anatomic features
- Dynamic factors contributing to ischemia
- Pros and cons of various imaging modalities

Why Anomalous Coronary Arteries Matter



Prevalence & Unpredictability

Landmark investigation in 1980s by Dr. Angelini on military recruit autopsies with SCD

2nd leading cause of SCD in athletes

Deaths often occur during exercise

Symptoms can be silent

~50% = initial presentation SCD

Young patients with exertional syncope

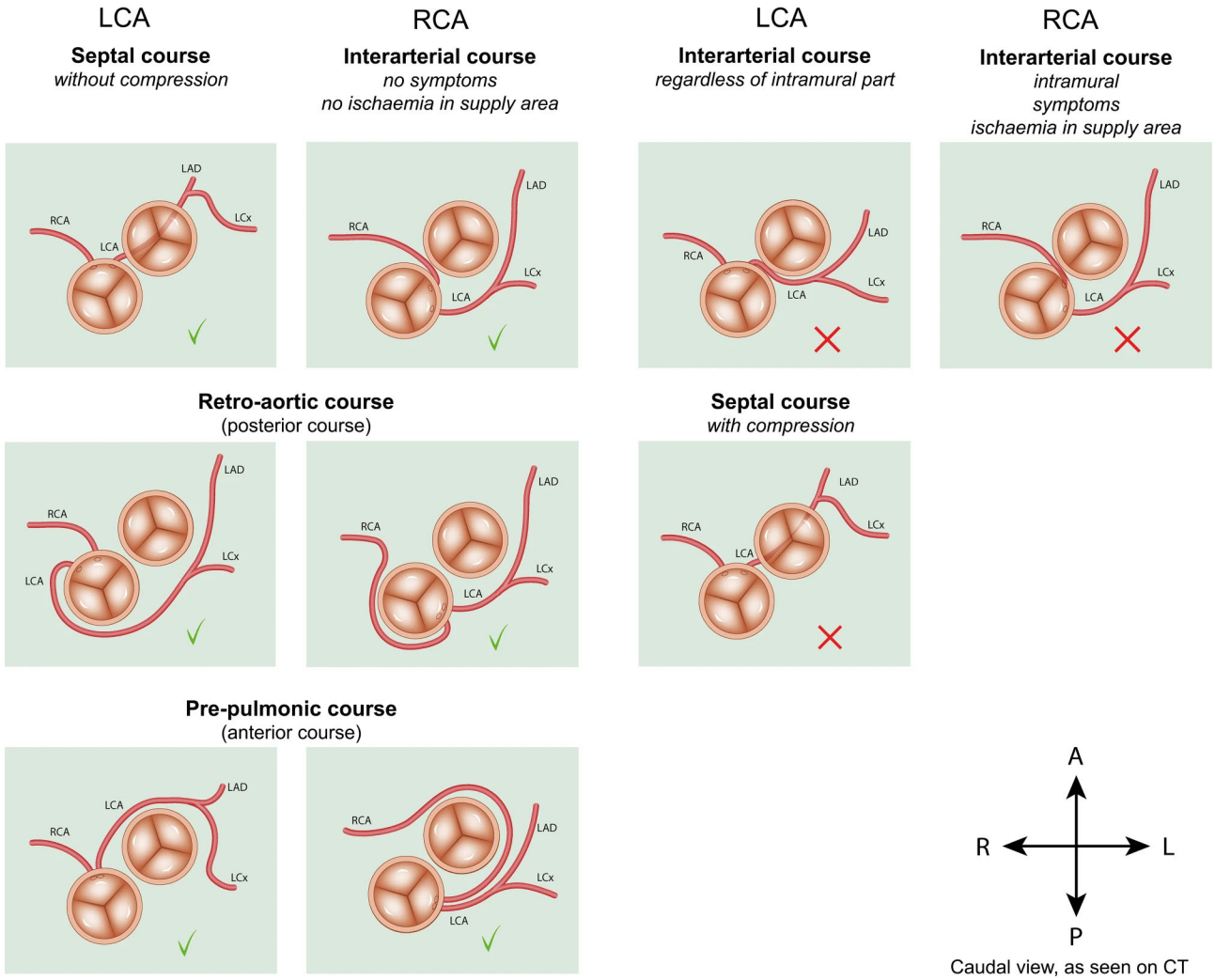
*Frommelt et al.

What is a variant? What is an anomaly?

- Epicardial artery distribution shows significant interindividual variability
  - Shepherds crook, type I-III LAD, dual LAD, split RCA, ….. etc.
    - Makes PCI more difficult, but are normal variants and not pathologic

- Anomalies are rare: often defined as <1% and may have a clinical significance
  - The true prevalence of *pathologic* congenital coronaries is difficult to ascertain (usually retrospective)

Anomalous Anatomy Is Highly Variable and Important



Caudal view, as seen on CT

ORIGIN

- Which great vessel?
  - AO vs PA
- Which cusp?
  - LCC vs RCC vs NCC
- Coronary branch
  - Right from left?
  - Left from right?

COURSE

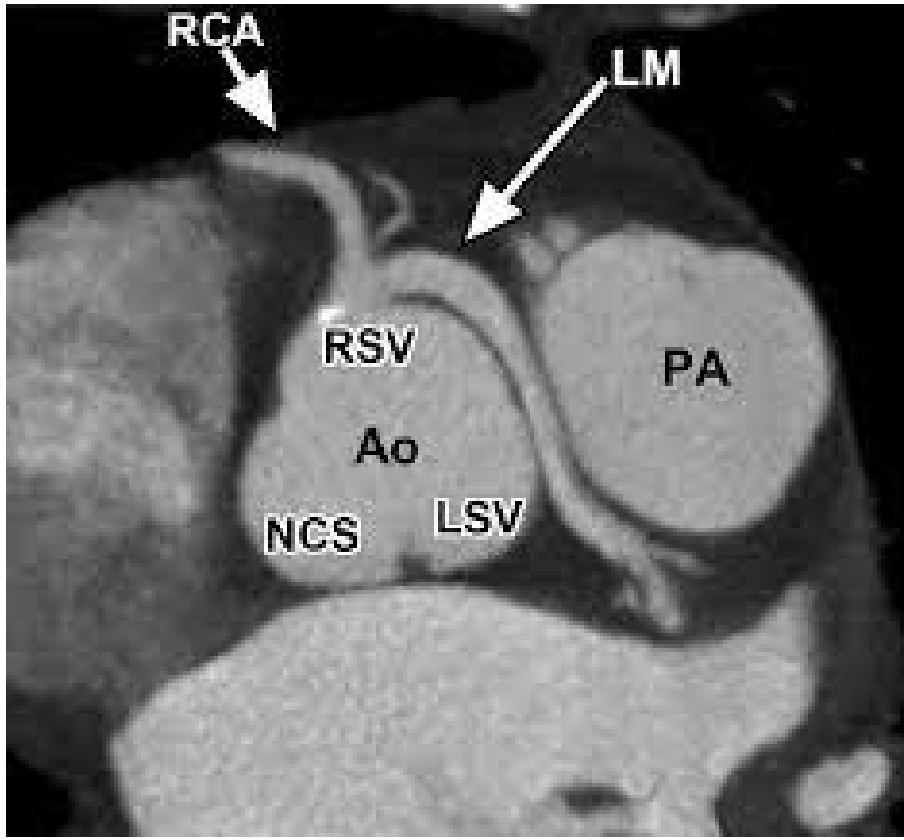
- Interarterial
- Pre-pulmonic
- Retroaortic
- Subpulmonic

TERMINATION

- Fistula
- Premature
- Ectopic

Interarterial Course - Red Herring?

- Historically, the interarterial course was considered the crucial abnormality for SCD
- Compression between the aorta and pulmonary artery?
  - The PA is low-pressure and unlikely to generate the counterforce to occlude a coronary artery
- However, the site closest to aortopulmonary proximity often has other high-risk abnormalities resulting in SCD.
  - The interarterial course may act only as a **surrogate** for other anatomical high-risk features
- Focus on additional features as well that may result in dynamic flow obstruction



Benign vs Malignant Course – Context Matters

High Risk Anatomy

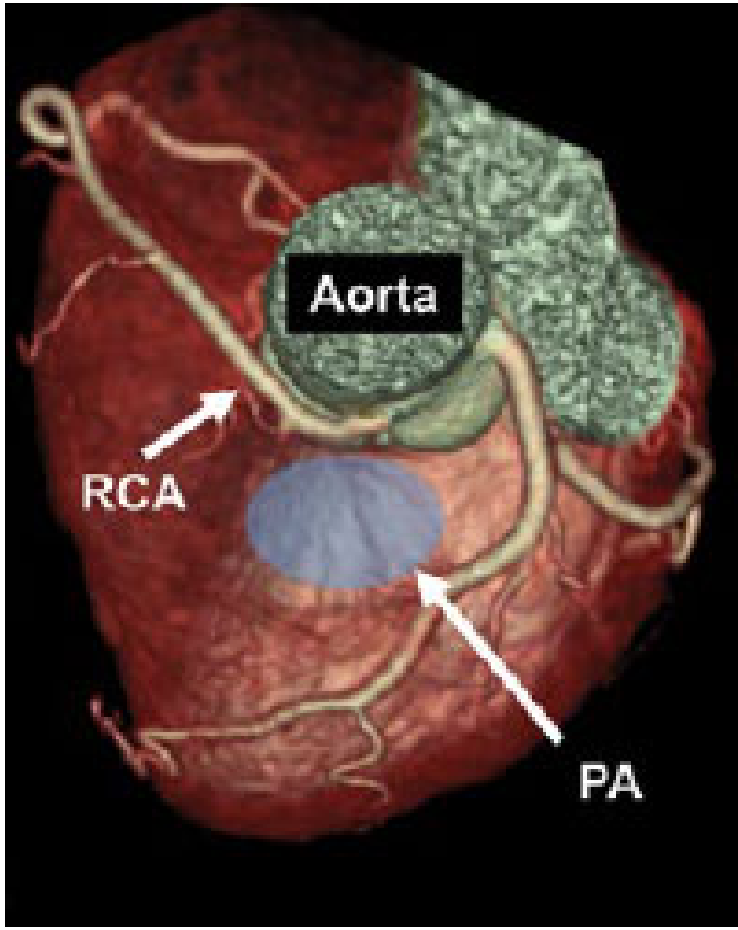
Consider myocardial territory being supplies

- Coronary from the opposite cusp
- Interarterial course
- Ostial morphology
- Intramural segment

Presentation

Signs, symptoms, or evidence of ischemia

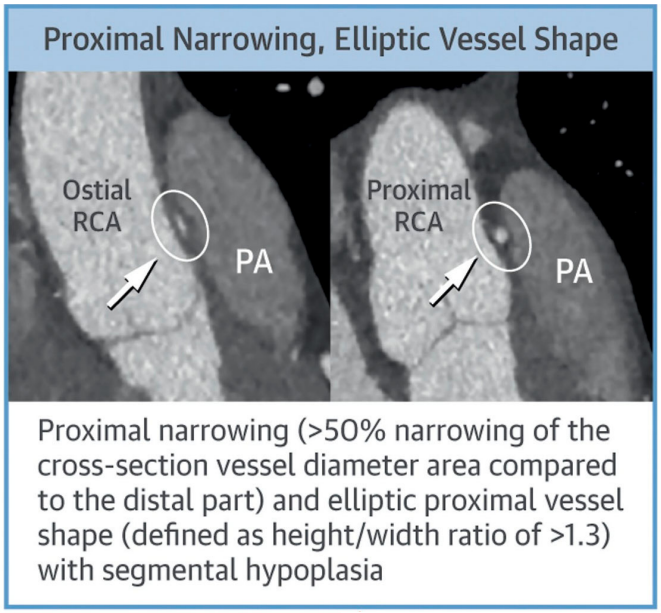
- Exertional Syncope
- Arrhythmias

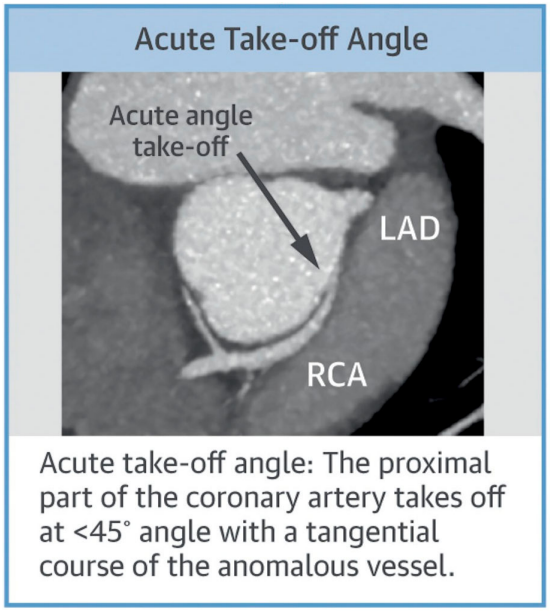


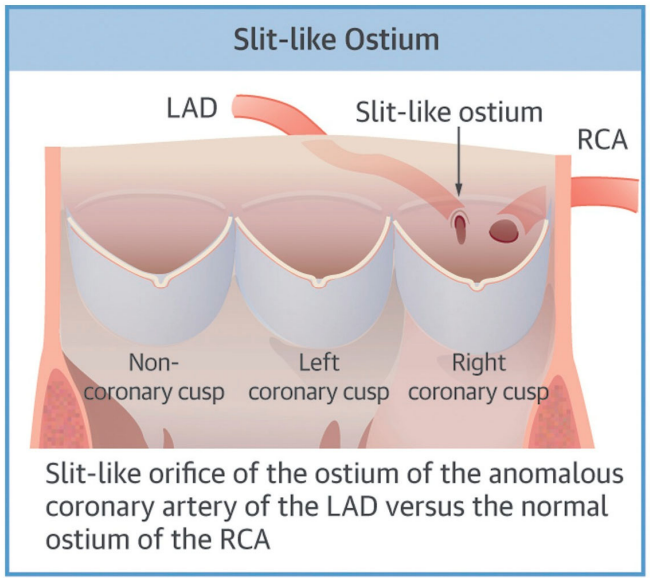
Ostial Morphology

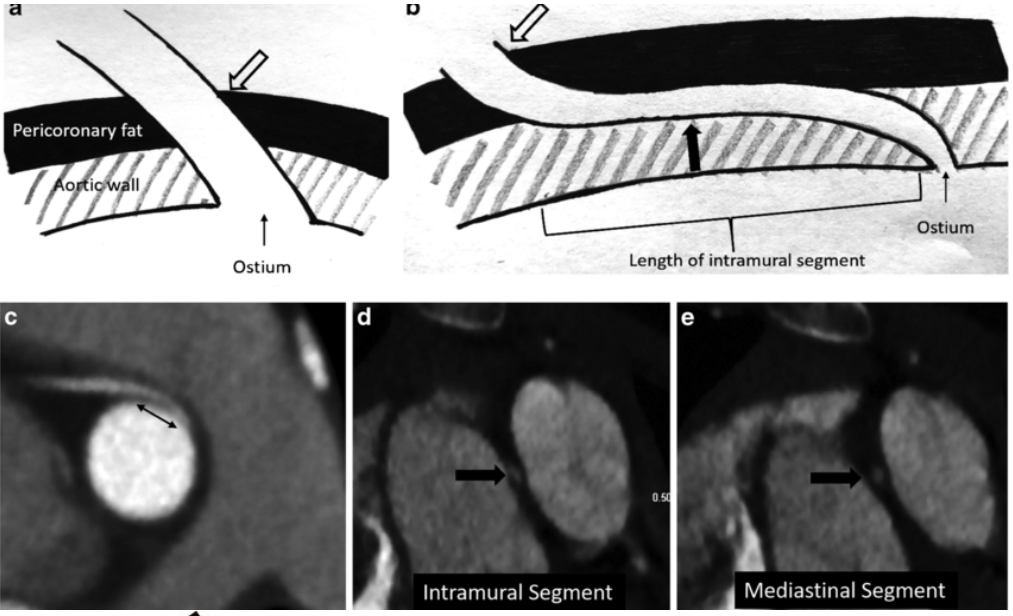
The shape of the ostium can influence flow dynamics

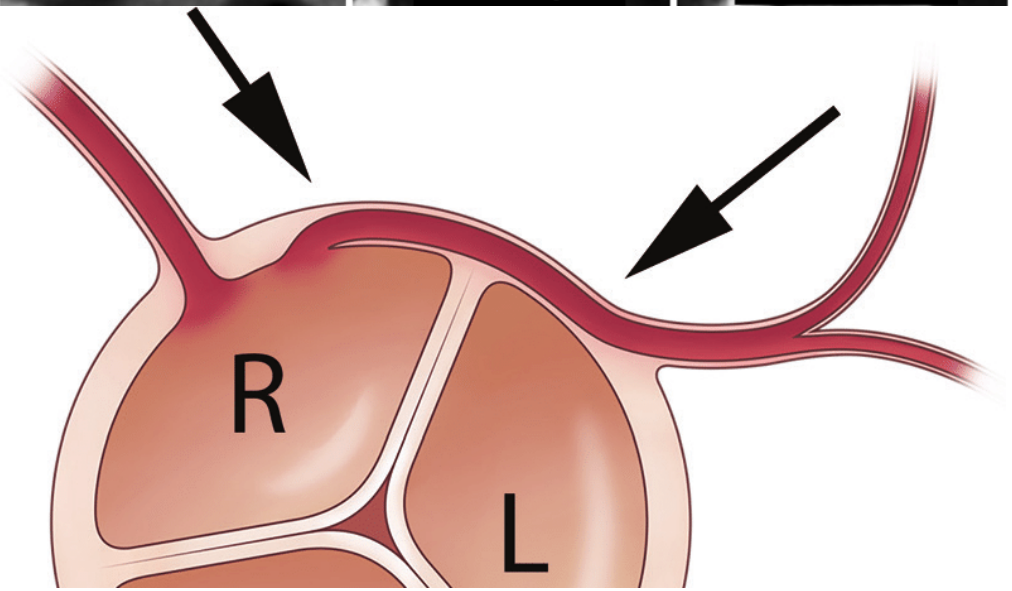
- Slit-like ostium
  - ≥50% reduction of the minimal lumen diameter
- Acute angle take-off
  - < 45◦
- Ostial ridge
- Ostial stenosis
- Eccentric ostium





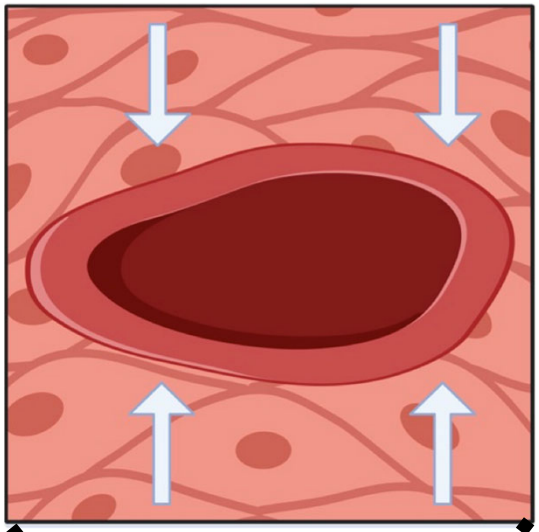






Intramural Segment

- Traverses the muscular aortic wall
- Potentially the “most malignant”
- Varies in length and thickness
  - Both diameter stenosis and length affect the hemodynamic significance
- The longer the segment the more it can become compromised
  - *Lateral compression*
- When combined with other features even higher risk



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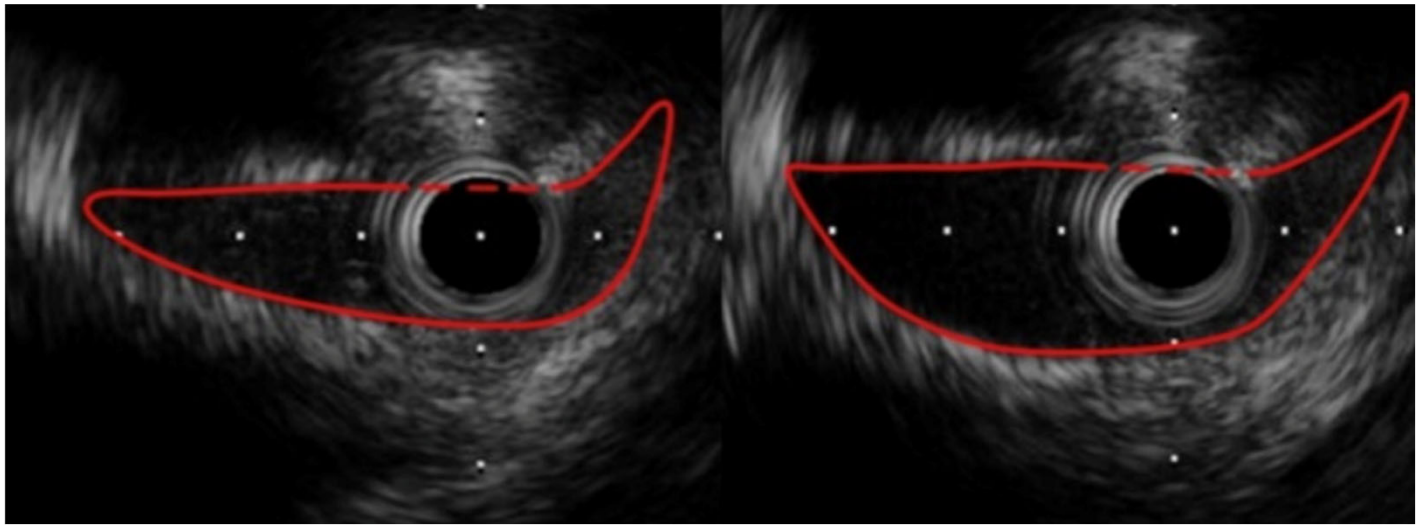
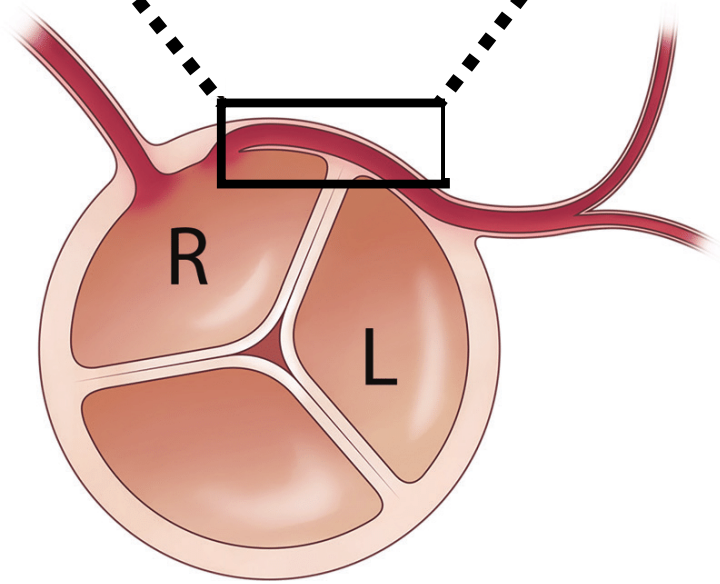
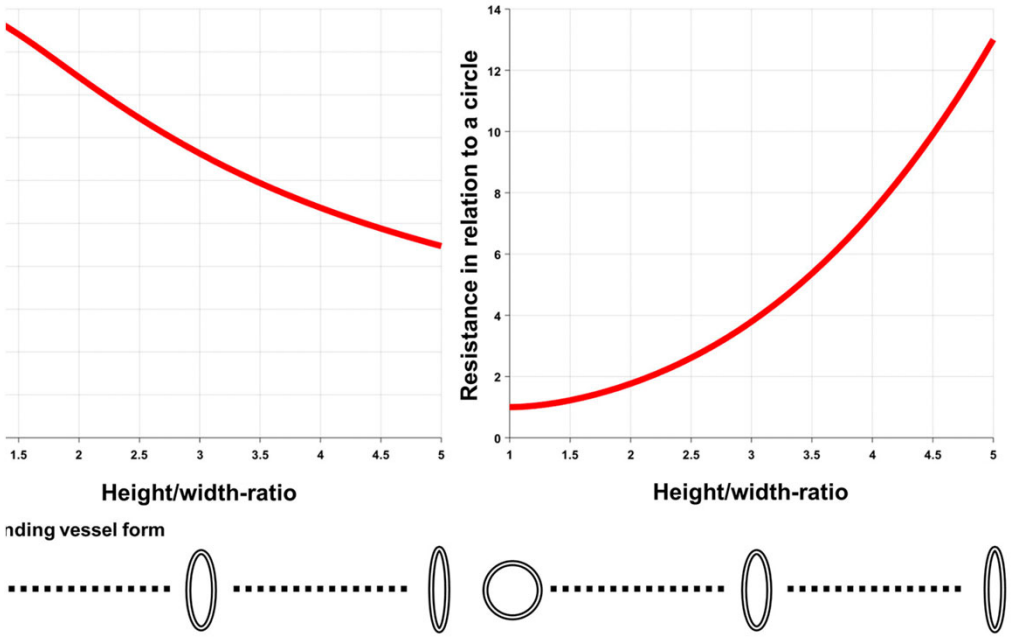
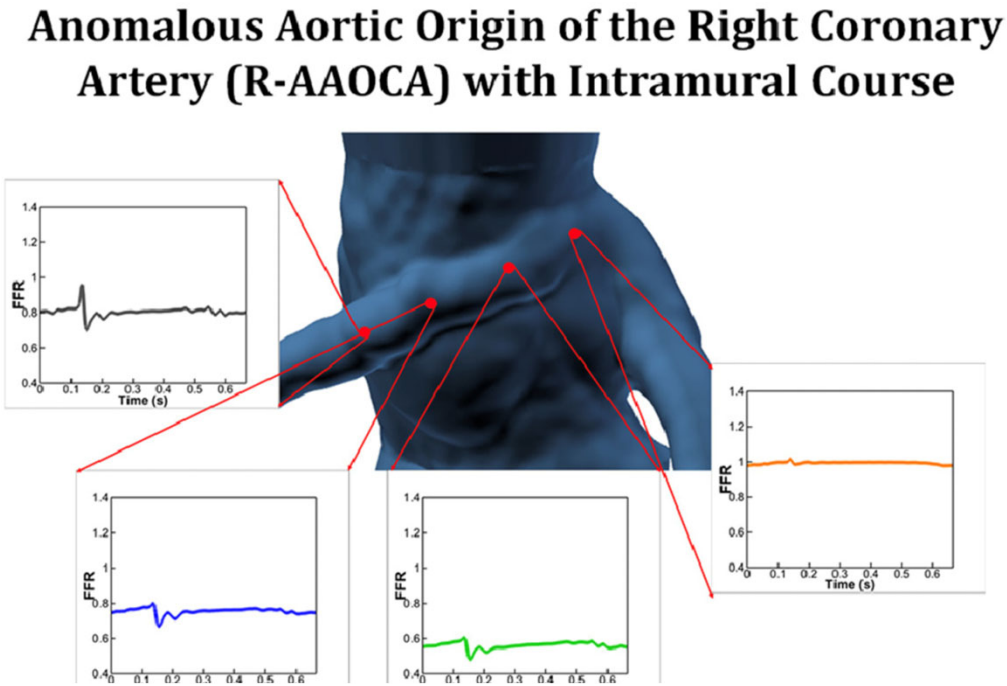




Lateral Compression

- Exercise physiology increases aortic wall stress due to increased blood volume and pressure changes
  - elliptic deformation [defined as height/width ratio of >1.3
- Decreased cross-sectional area during increased myocardial oxygen demand
- Flow Resistance = $\Delta P = \frac{8\mu LQ}{\pi r^4}$





Flow Resistance with Cross Sectional Area

Two Tier Concept: The Perfect Storm

Tier 1: Anatomic Factors
Origin and Course
Intramural Segment
Ostial Morphology
Tier 2: Dynamic Factors
Flow Dynamics
Increased cardiac output
Increased dP/dt

1. **High-risk anatomic features**
2. **Increased Cardiac Output:**
    1. Shear stresses increases
    2. Aorta expansion
3. **Altered Vascular Dynamics:**
    1. Changed great vessel compliance
    2. Elliptical malformation → ostium or proximal segment compression
4. **Increased Myocardial Oxygen Demand:**
    1. Compression leads to myocardial ischemia
5. **Arrhythmias**

Combined, these create *ischemia*

Not all modalities are created equal

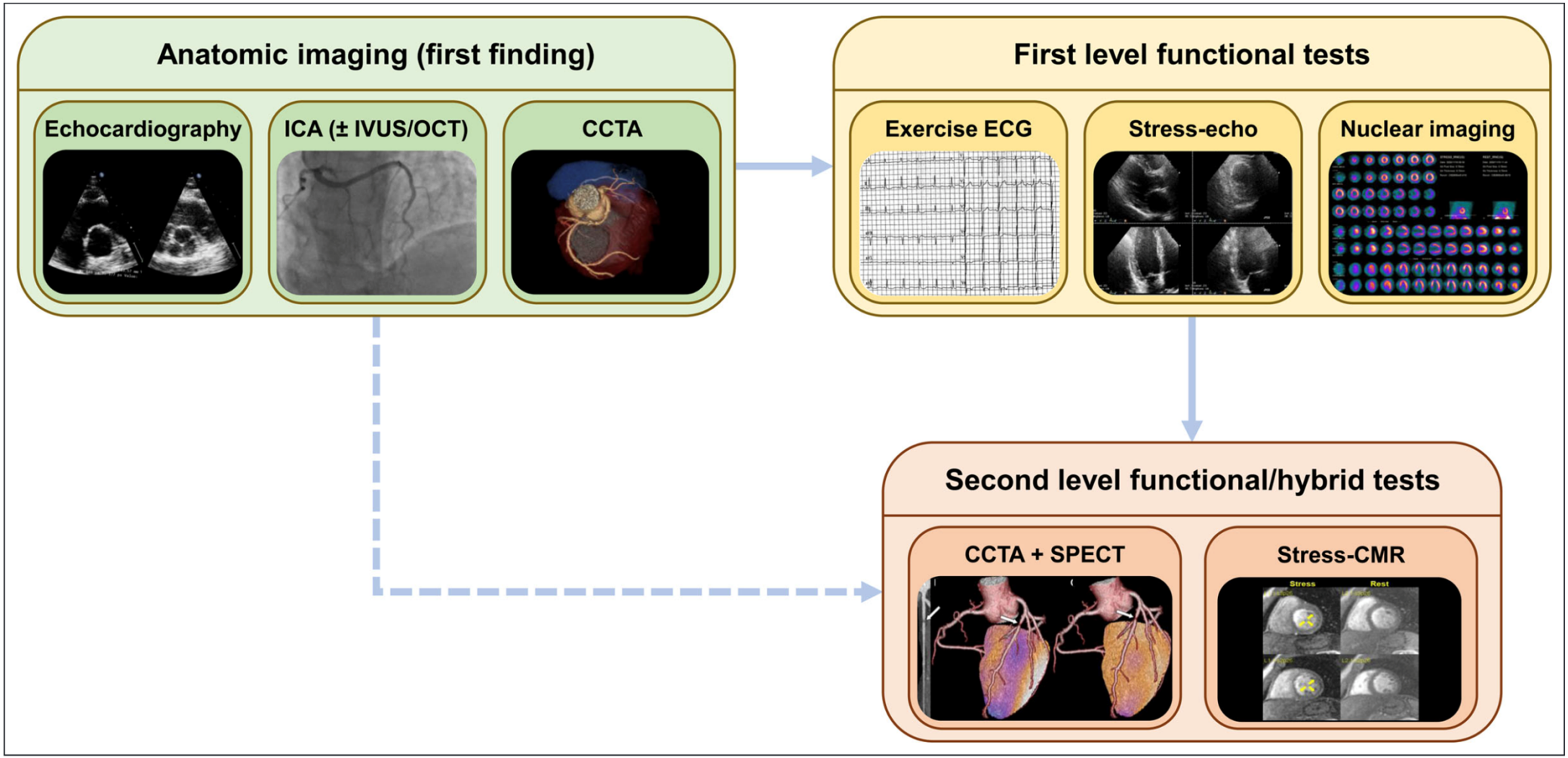
- Not the same ischemic pathophysiology as fixed atherosclerotic disease
- *Dynamic process* resulting from vascular stress occurring during exercise

- Optimal diagnostic modality = detects anomaly **and** anatomical high-risk features, ischemia, evidence of myocardial fibrosis/scar
- Pure vasodilators (i.e., adenosine or regadenoson) **are not able to provoke the dynamic components** (i.e., dynamic lateral compression of the intramural course) and, thus, are prone to provide false negative results.
- Ideally use physical exercise or dobutamine to increase both heart rate and stroke volume

Proposed Stress Protocols

Stress Testing Protocol	Dose	Increase in Coronary Blood Flow	Increase in Cardiac Output
Physical Exercise	85% max heart rate	↑↑	↑↑
Adenosine	1.40 mg/kg/min	↑↑↑	↑↑↑
Regadenoson	0.4mg bolus	↑↑	-
Norepinephrine	0.01 µg/kg/min	↑↑	-
Dobutamine	40 µg/kg/min	↑	↑↑
Dobutamine + Volume Expansion (Saline: 1.5–3 l, Atropine: 1 mg)	40 µg/kg/min	↑↑↑	↑↑↑

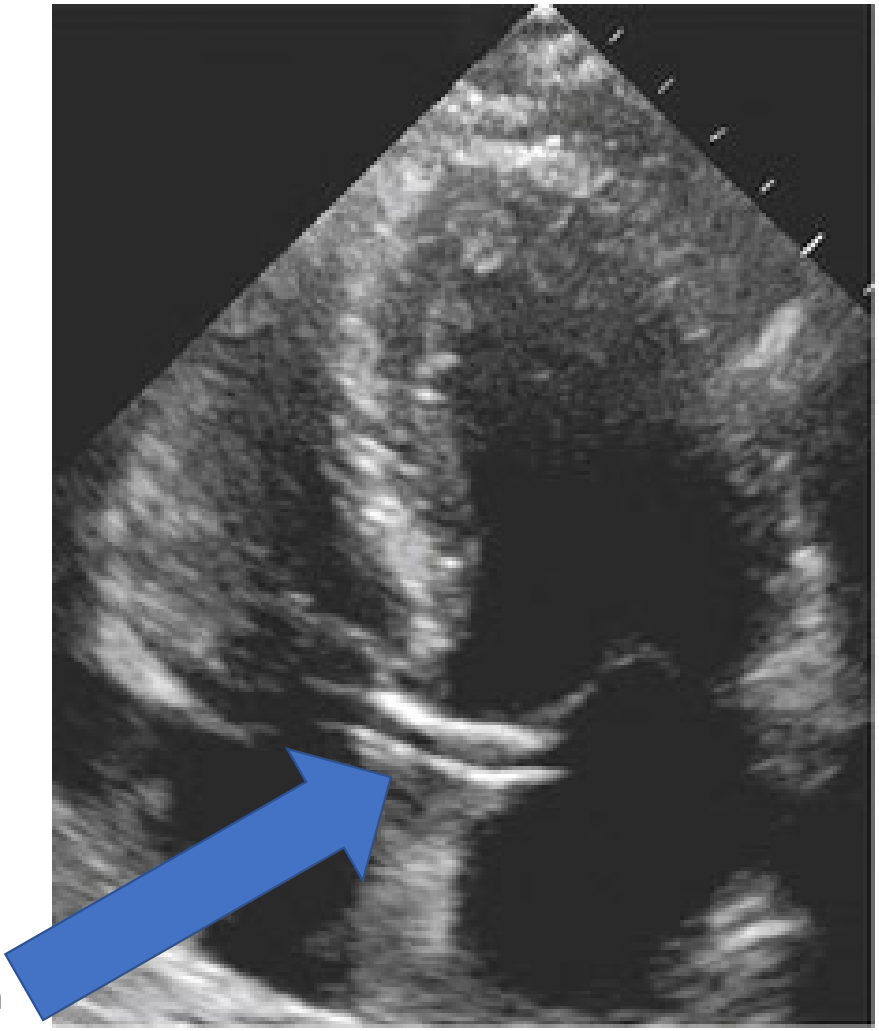
Hyperemia does not equate to physiologic stress



Gentile et al

Transthoracic Echocardiography

- Can visualize the origin and proximal course non-invasively without radiation
- Usually made in short-axis view in the plane of the aortic root
  - Can include focused color Doppler of the aortic wall to identify an intramural course
- Eval concomitant congenital heart defects
- General good acoustic windows in children, making Good initial eval in pediatric populations
- However, it is not very good in adults with significant interobserver variability

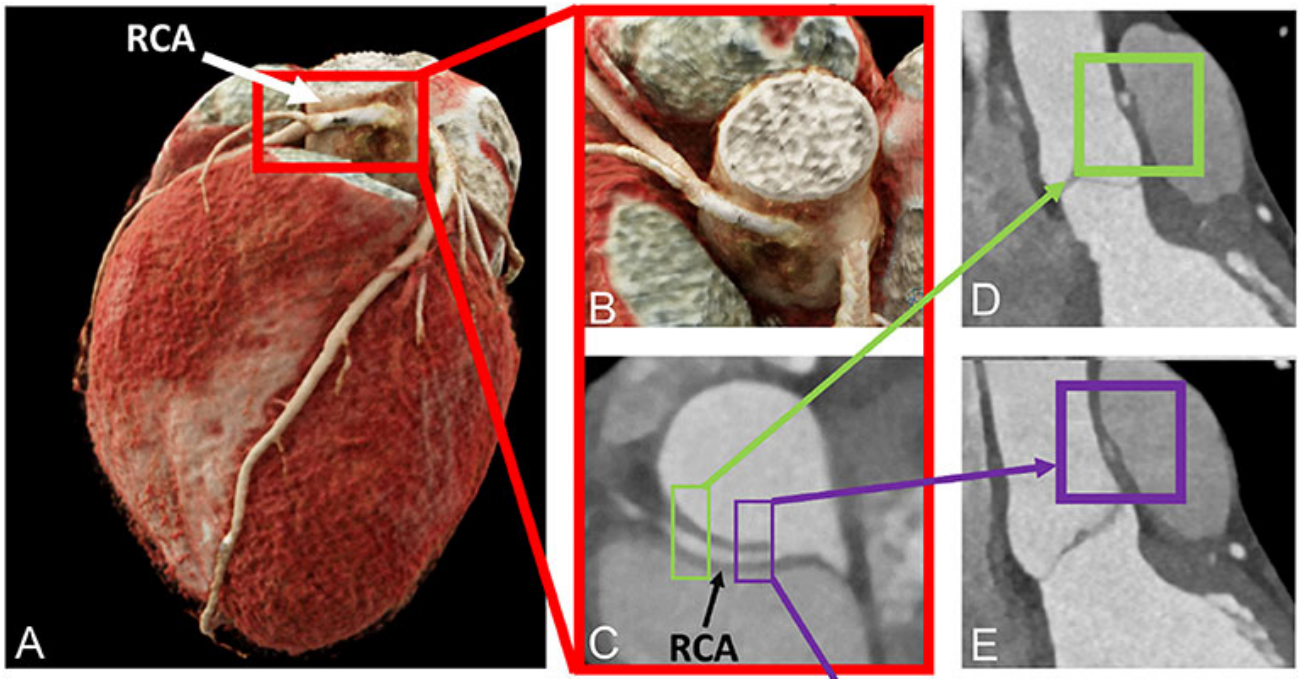


The “RAC” Sign

Witt et al

Coronary Computed Tomography Angiography

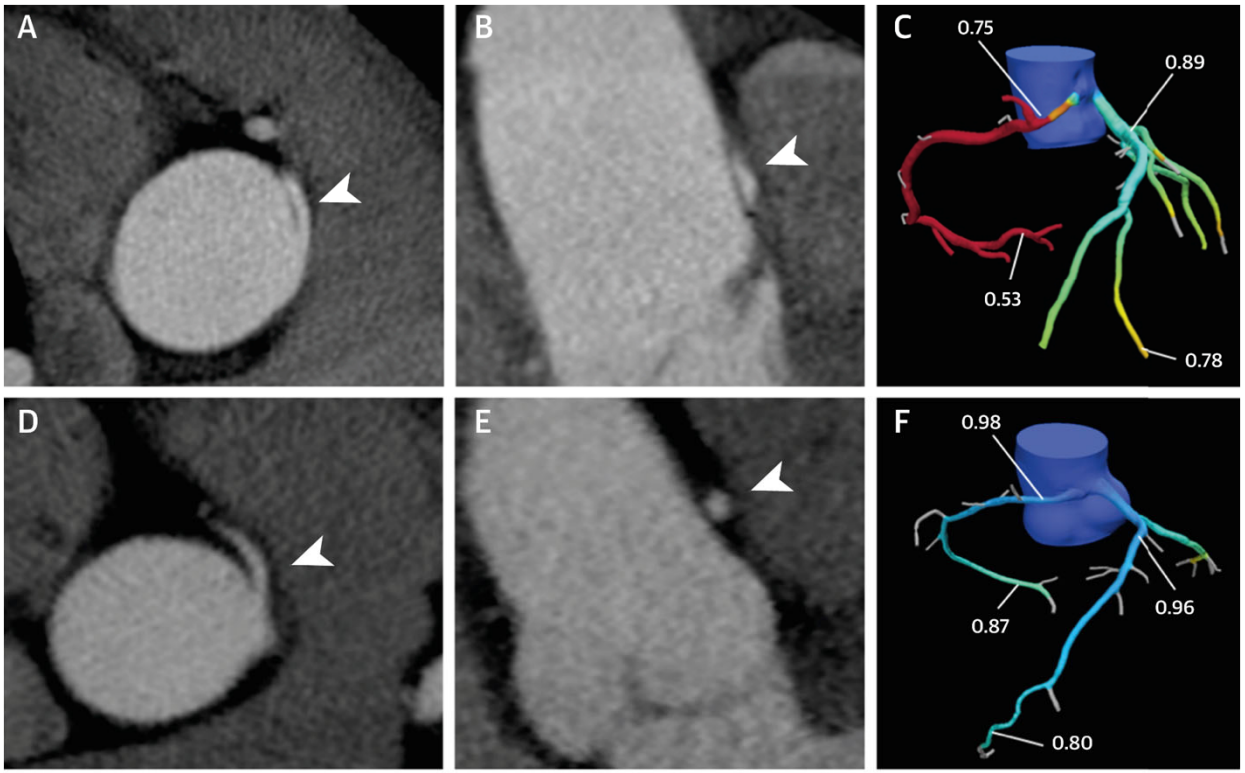
- Provides the **best** spatial resolution (0.25 x 0.25 mm)
  - detailed anatomic eval
  - Acute angle take off, intramural course, slit like ostium
- Advanced post-processing methods enable the detailed evaluation
- Full coronary course evaluation including concomitant atherosclerotic CAD
- Modern tech = low radiation (often just 0.5–3 mSv)



CT-FFR

Primarily used in the evaluation of *fixed* CAD lesions and the diagnostic value remains unclear for *dynamic* lesions

Some promising trials for utilization, but further work is needed



Coronary Computed Tomography Angiography-Derived Fractional Flow Reserve in Patients with Anomalous Origin of the Right Coronary Artery from the Left Coronary Sinus

Chun Xiang Tang, PhD,[1,*] Meng Jie Lu, MSc,[1,*] Joseph Uwe Schoepf, MD,[1,2] Christian Tesche, MD,[2] Maximilian Bauer, MD,[2] John Nance, MD,[2] Parkwood Griffith, BS,[2] Guang Ming Lu, MD,[1] and Long Jiang Zhang, MD, PhD[1]

Physiological Evaluation of Anomalous Aortic Origin of a Coronary Artery Using Computed Tomography–Derived Fractional Flow Reserve

Julien Adjedj, MD, PhD [†]; Fabien Hyafil, MD [†]; Xavier Halna du Fretay, MD; Patrick Dupouy, MD; Jean-Michel Juliard, MD; Phalla Ou, MD, PhD; Jean-Pierre Laissy, MD, PhD; Olivier Muller, MD, PhD; William Wijns, MD, PhD; Pierre Aubry, MD

Role of FFR-CT for the Evaluation of Patients With Anomalous Aortic Origin of Coronary Artery

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Warda Ferrag, François Scalbert, Julien Adjedj, Patrick Dupouy, Phalla Ou, Jean-Michel Juliard, Reza Farnoud, Ahmed A. Benadji, Xavier Halna Du Fretay, William Wijns, Pierre Aubry, and Fabien Hyafil

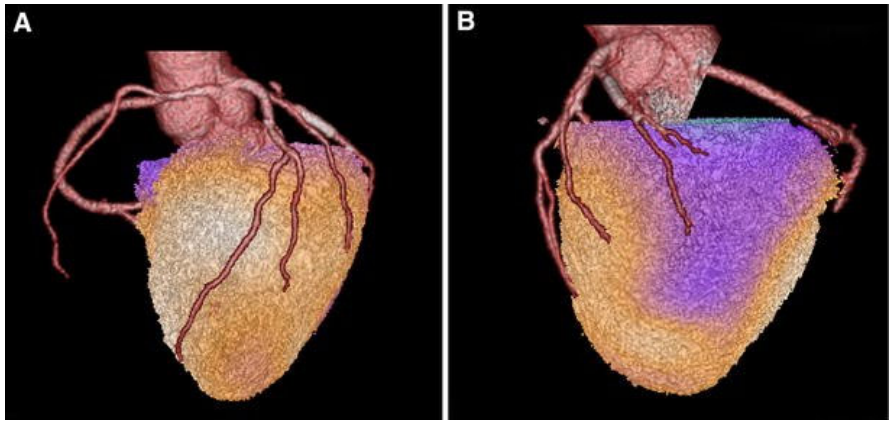
J Am Coll Cardiol Img. 2021 May, 14 (5) 1074–1076

SPECT

- Established techniques for stratification and assessment of myocardial perfusion in the setting of ***fixed*** CAD
  - As opposed to dynamic coronary anomalies
- Multiple studies have shown a favorable assessment of the hemodynamic relevance of anomalous coronaries with favorable diagnostic performance.
- Use caution when using pure vasodilators, as they may **not provoke the dynamic components and** provide false negative results.
- Concern that limited spatial resolution may miss small subendothelial ischemia

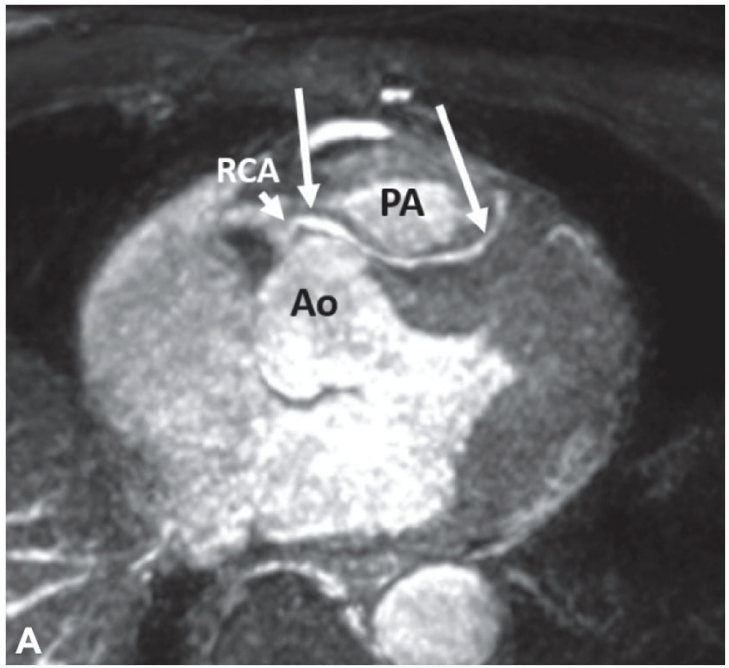
Hybrid CCTA/SPECT myocardial perfusion imaging findings in patients with anomalous origin of coronary arteries from the opposite sinus and suspected concomitant coronary artery disease

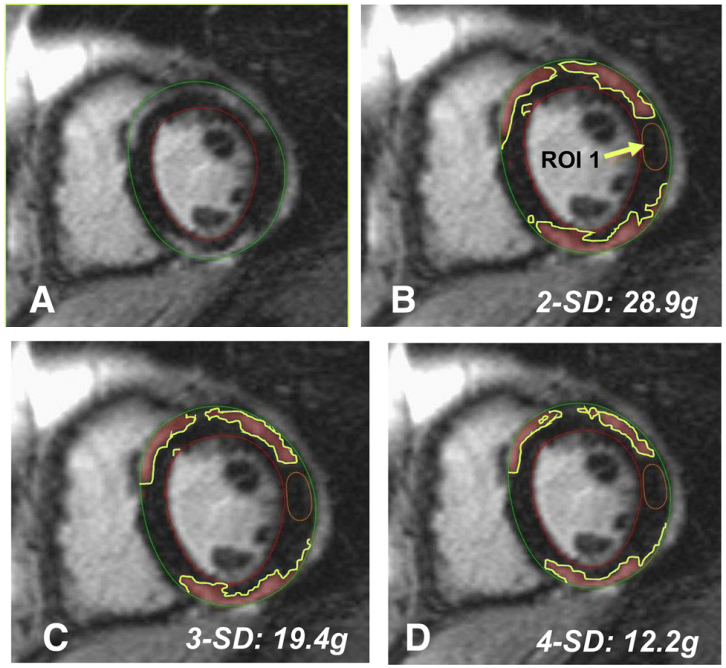
Christoph Gräni MD, Dominik C. Benz MD, Christian Schmied MD, Jan Vontobel MD, Fran Mikulicic MD, Mathias Possner MD, Olivier F. Clerc MD, Julia Stehli MD, Tobias A. Fuchs MD, Aju P. Pazhenkottil MD, Oliver Gaemperli MD, Ronny R. Buechel MD & Philipp A. Kaufmann MD



Cardiac Magnetic Resonance

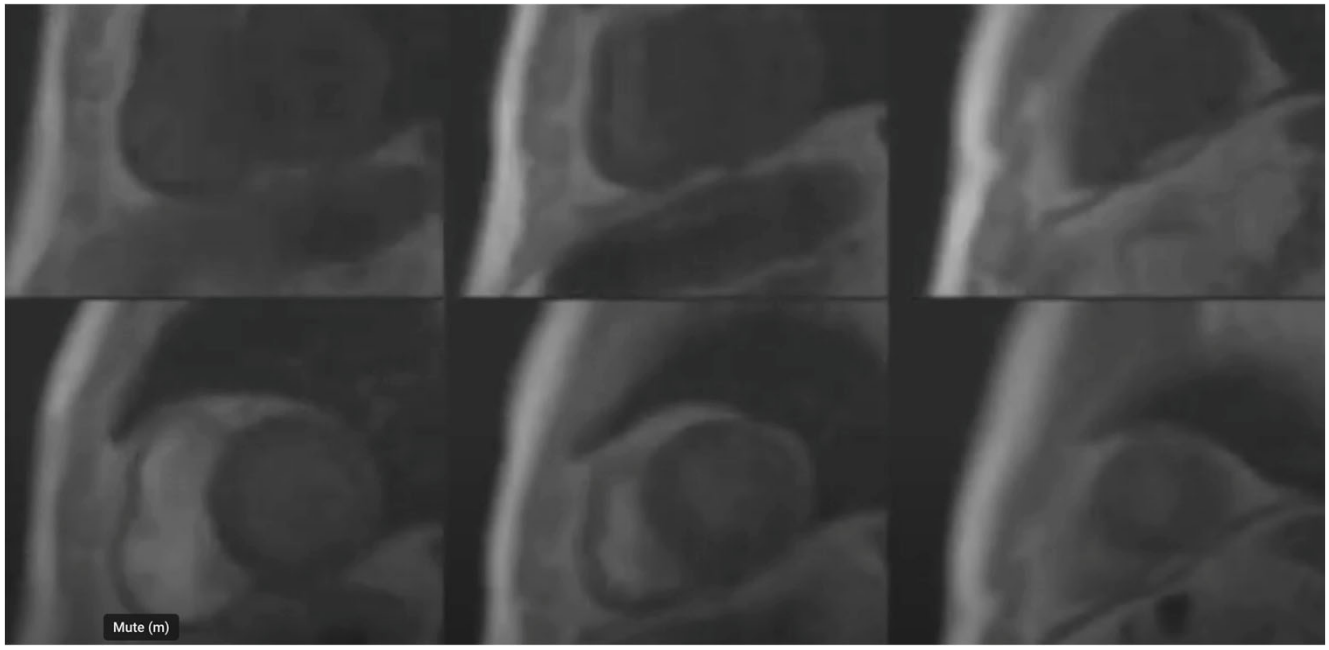
- Tomographic imaging at high spatial resolution, ~1 x 1 x 1 mm [lower than CCTA]
- Allows functional ischemic testing to investigate the hemodynamic relevance by pharmacologic inotropic stress (i.e., dobutamine)
- Also assesses possible underlying arrhythmogenic myocardial fibrosis (LGE).
  - Expression of recurrent minor myocardial ischemia → substrate for ventricular tachyarrhythmia





Stress CMR

- Pros:
  - Detects ischemia, lacks radiation and has the capability to evaluate myocardial perfusion, wall motion, and tissue characterization in a single examination.
- Cons:
  - Claustrophobia. CMR stress tests can be uncomfortable (especially dobutamine)
  - Reduced spatial resolution compared to CCTA



	Echocardiography	CCTA	CMR	SPECT
Physical characteristics				
Spatial resolution	++	+++	++	+
Temporal resolution	++/+++*	++	++	+
Anatomy of coronary arteries				
Proximal	+++	++++	++++	-
Distal	++	++++	++	-
Assessment of vascular territories	-	+++	++	-
Anatomic high-risk features in ACAOS				
Interarterial course	++	++++	++++	-
Fixed components				
Slit-like ostium	+	++++	++	-
Proximal narrowing	++	+++	++	-
Dynamic components				
Take-off angle	++	++++	++++	-
Elliptic shape	++	+++	++	-
Intramural course	++	++++	+++	-
Physiologic high-risk consequences in ACAOS				
Ischemia	++°	+**	++++	+++°
Scar	+	++	++++	+++

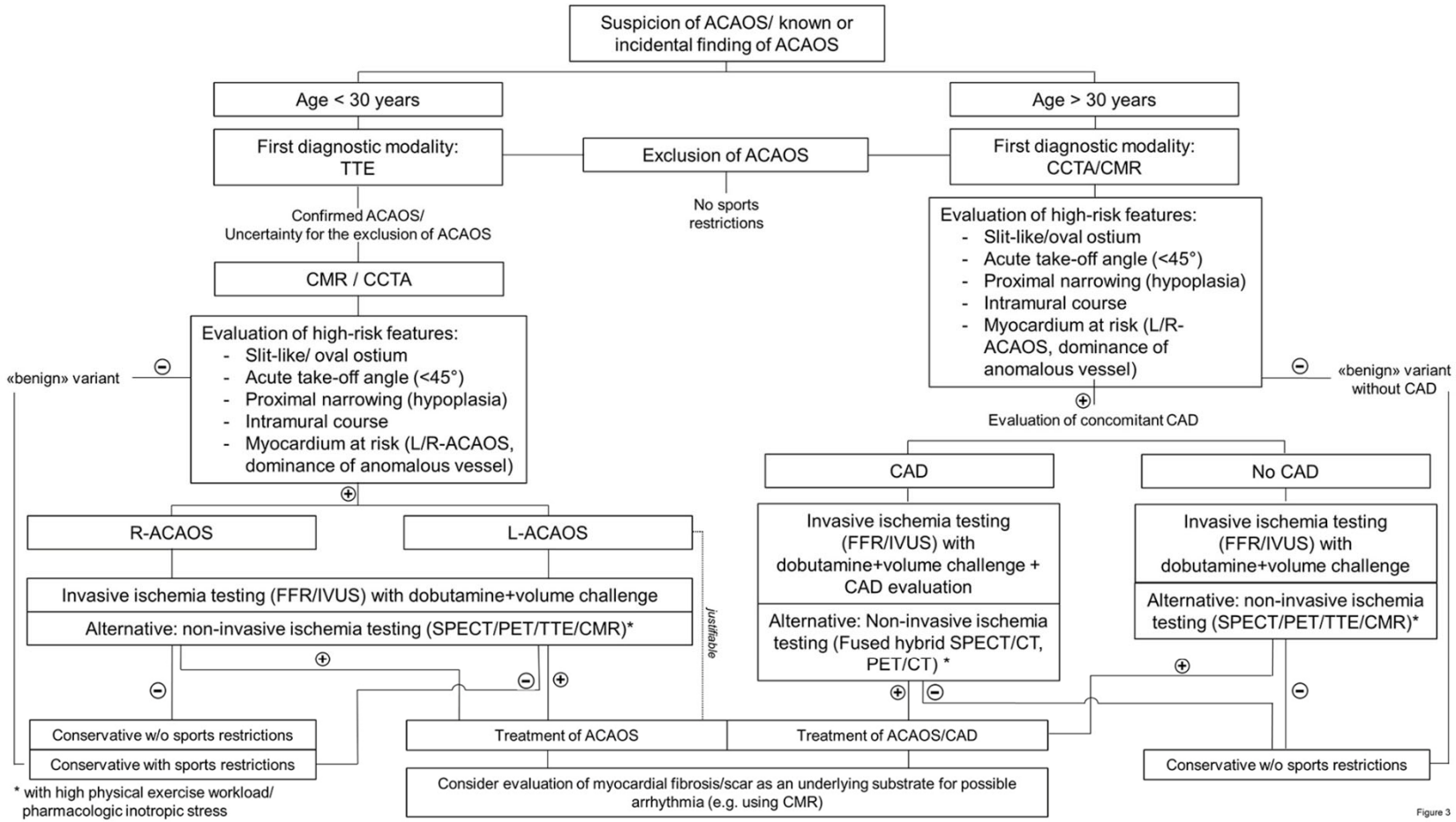
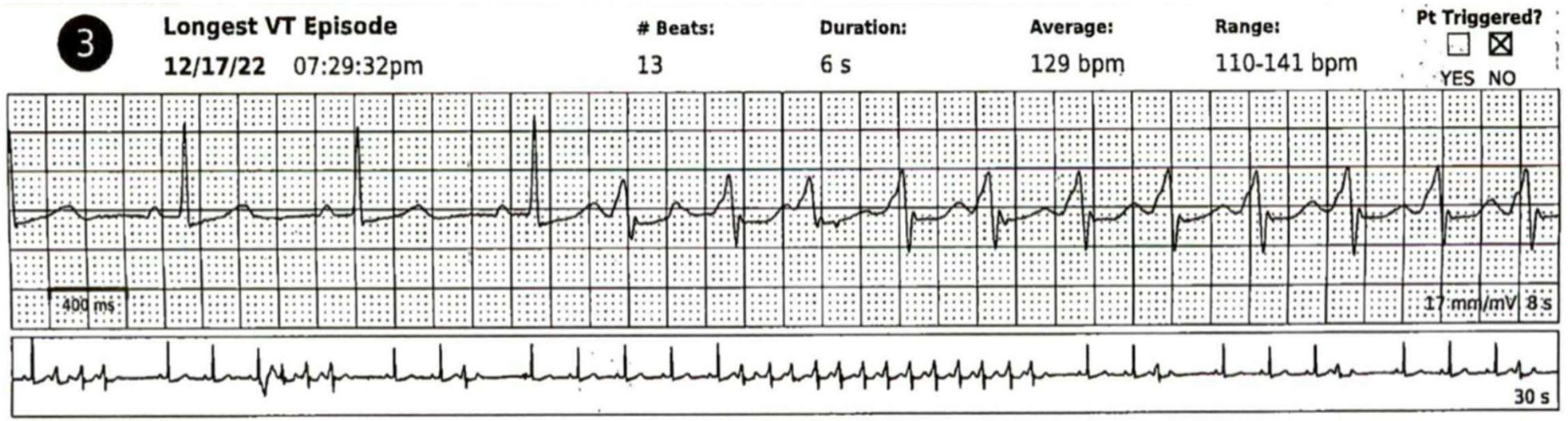
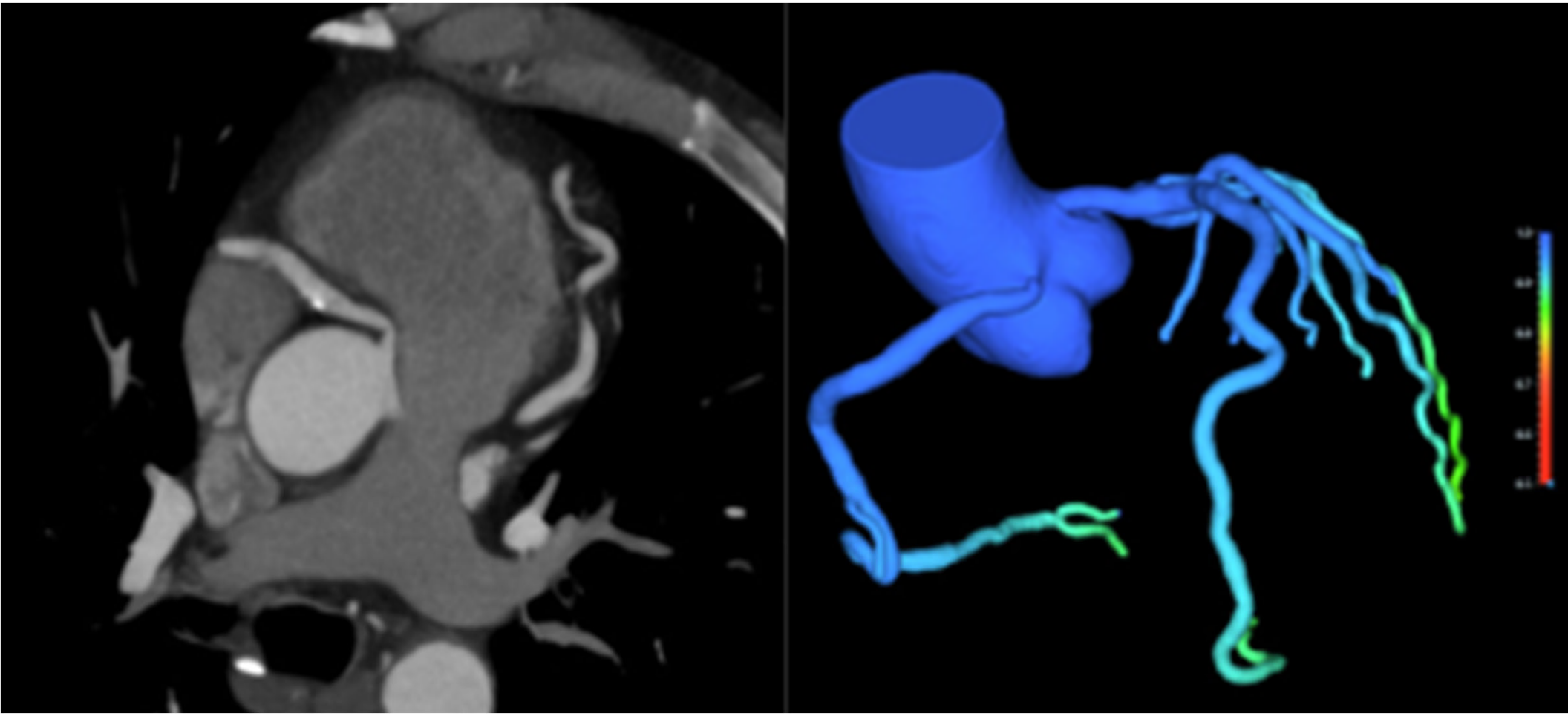


FIGURE 3 | Flow chart of diagnostic management in patients with an anomalous coronary artery. (R-/L)-ACAOS, (right/left) anomalous coronary arteries with the origin of the anomalous vessel from the opposite sinus of Valsalva; CAD, coronary artery disease; CCTA, coronary computed tomography angiography;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

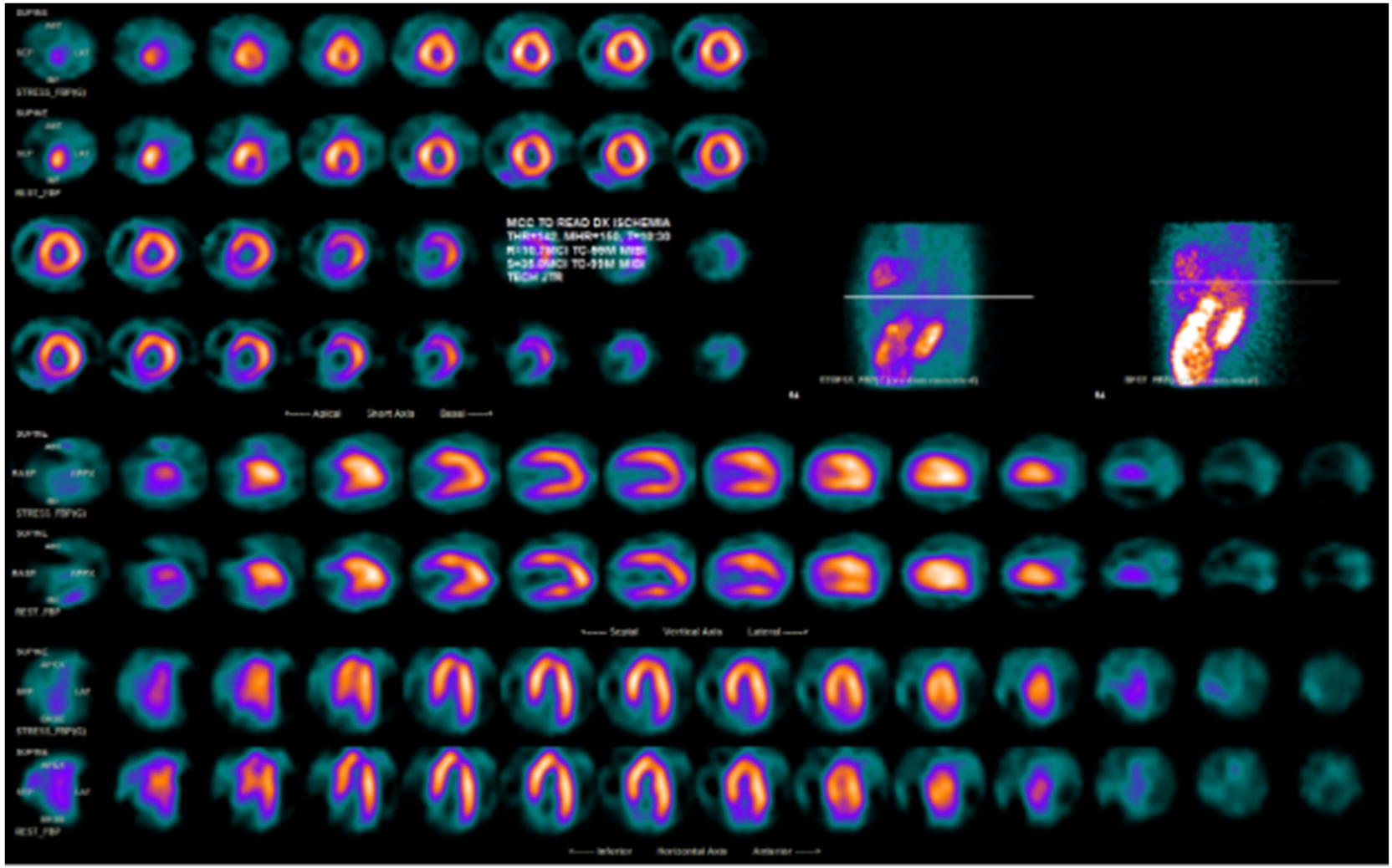
Case Presentation

- A 52-year-old male was advised to go to the emergency department after an abnormal result on his mobile cardiac outpatient telemetry monitor
- 348 runs of NSVT, the fastest being 200bpm, the longest episode was 47 seconds with 100 polymorphic VT complexes

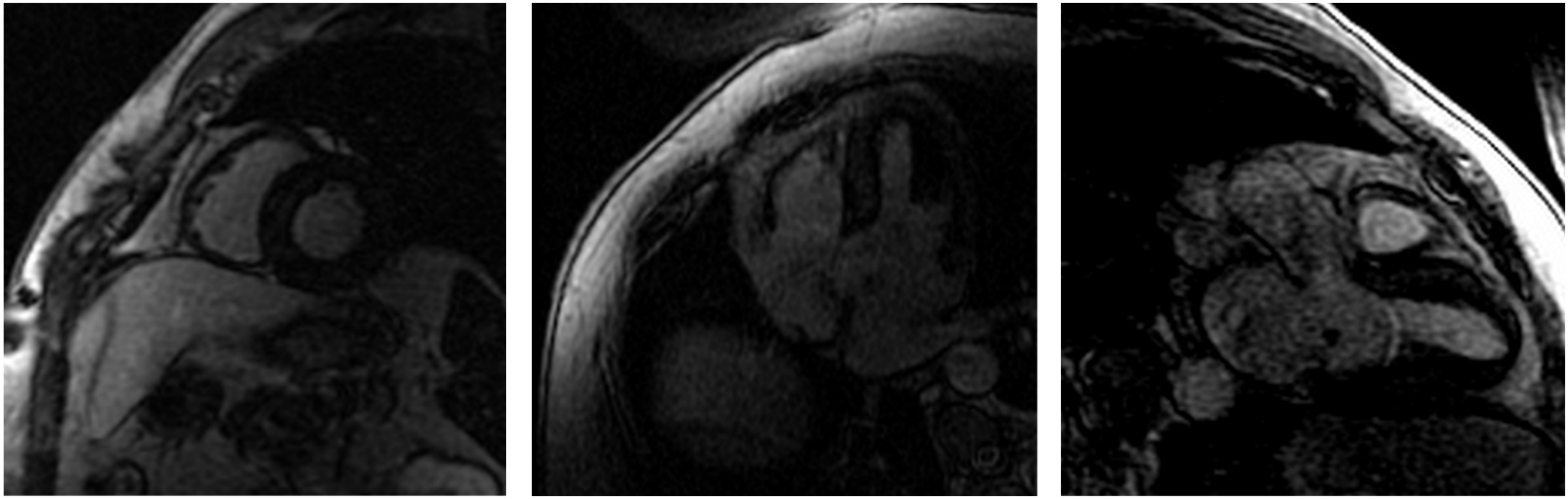




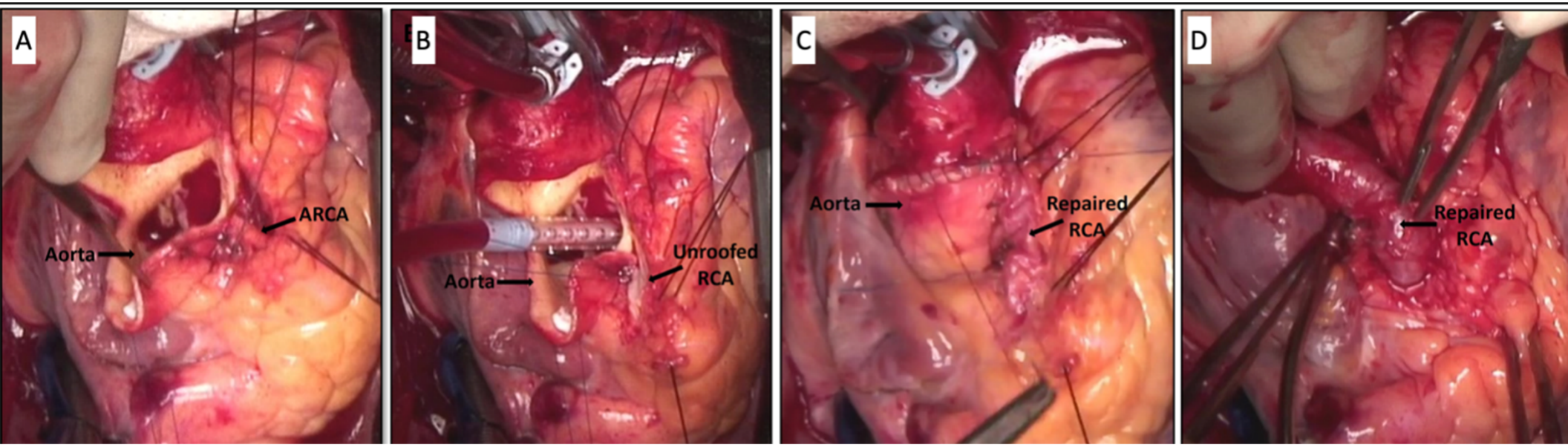
Anomalous RCA from LCC. Negative CT-FFR



Lexiscan SPECT unremarkable



CMR with no focal scar



Unroofed and repaired right coronary artery

Great outcome, no complications, and no recurrent arrhythmia on follow-up

Thank you!