

Fokus Herz Bern 2025

Koronaranomalien – etwas Beunruhigendes?

Freitag 16. Mai, 2025

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Koronaranomalie Team



Christoph Gräni

Leiter
Koronaranomalie-
Sprechstunde



Lorenz Räber

Invasive Evaluation



Matthias Siepe

Chirurgische Versorgung

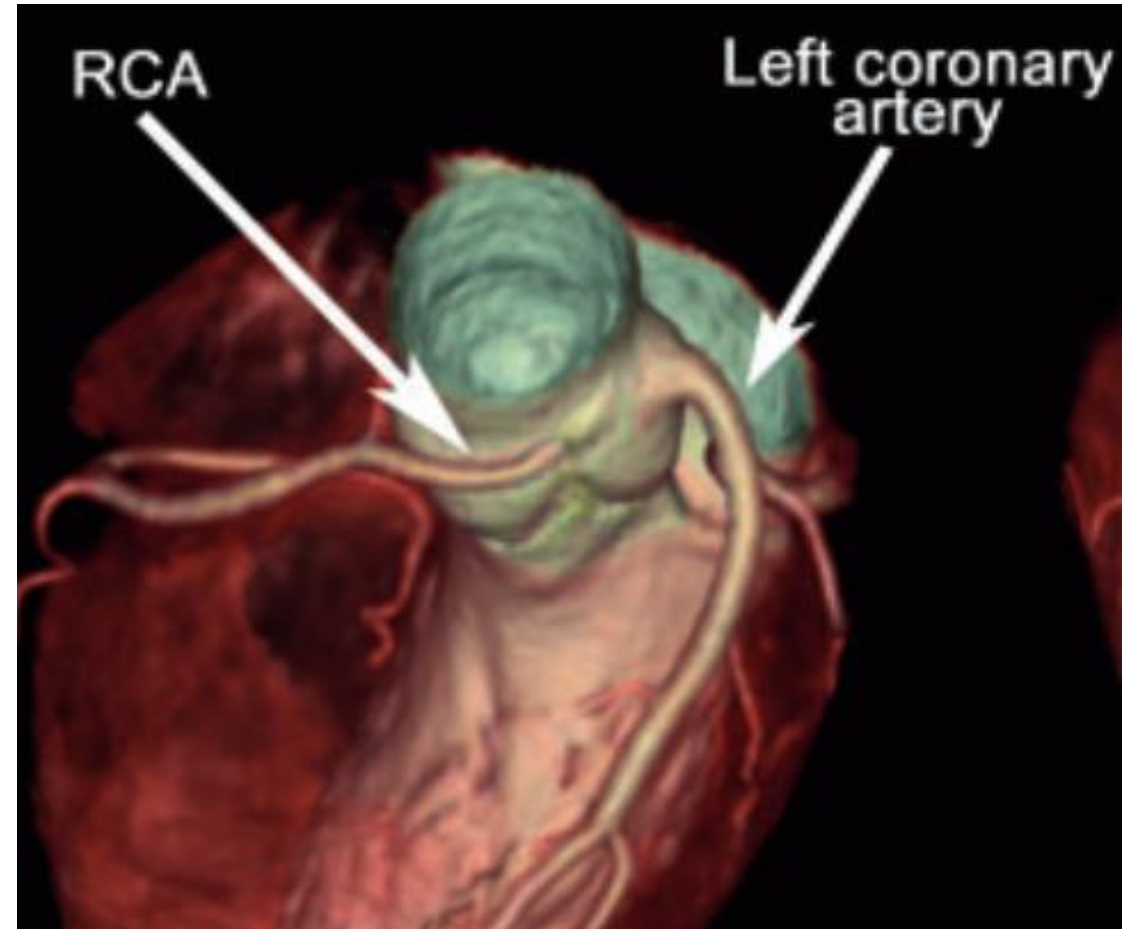
Background

Prevalence:

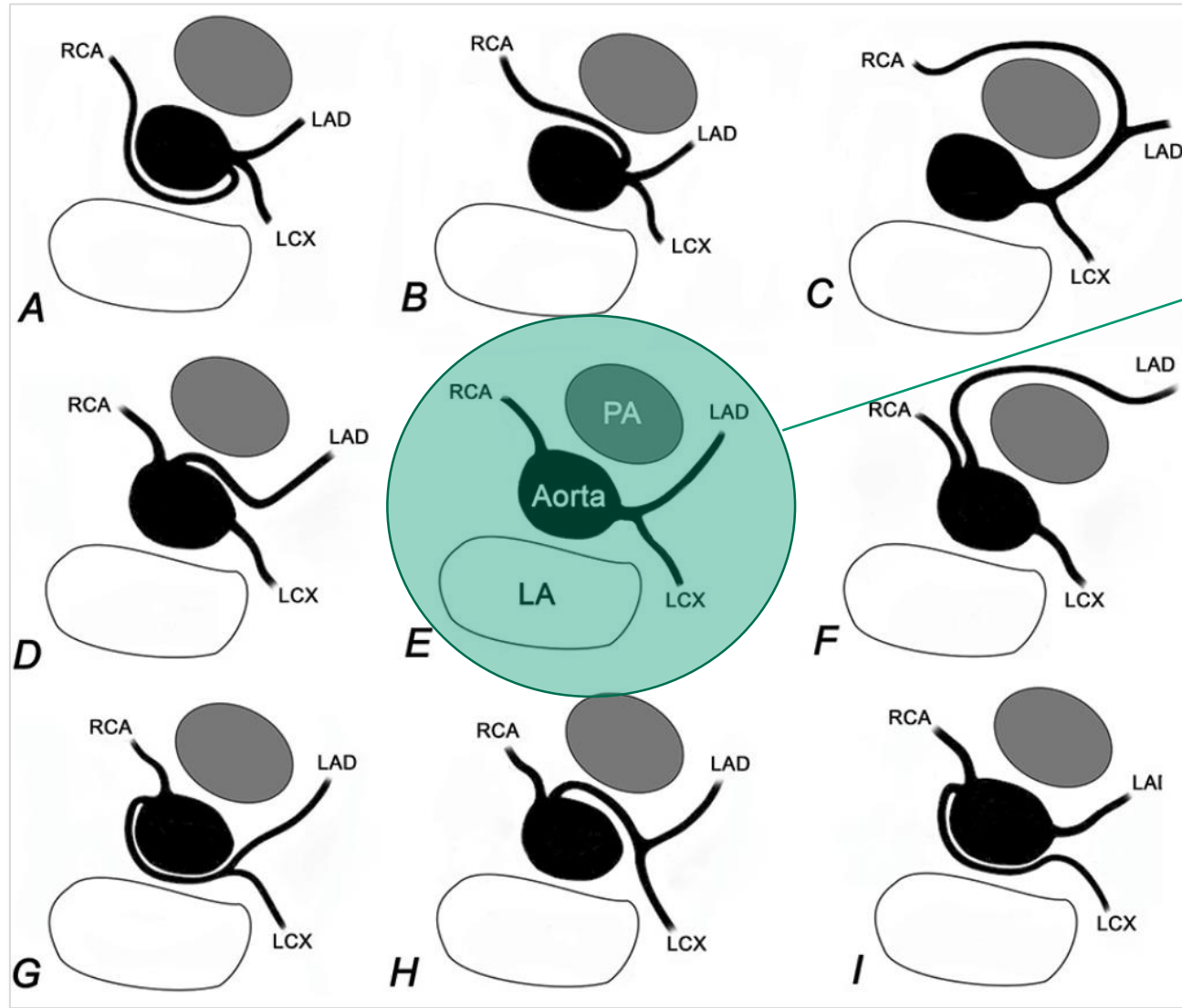
- 0.26% AAOCA:
Anomalous Aortic Origin of a Coronary Artery

Clinical presentation:

- Vary



Variants of AAOCA

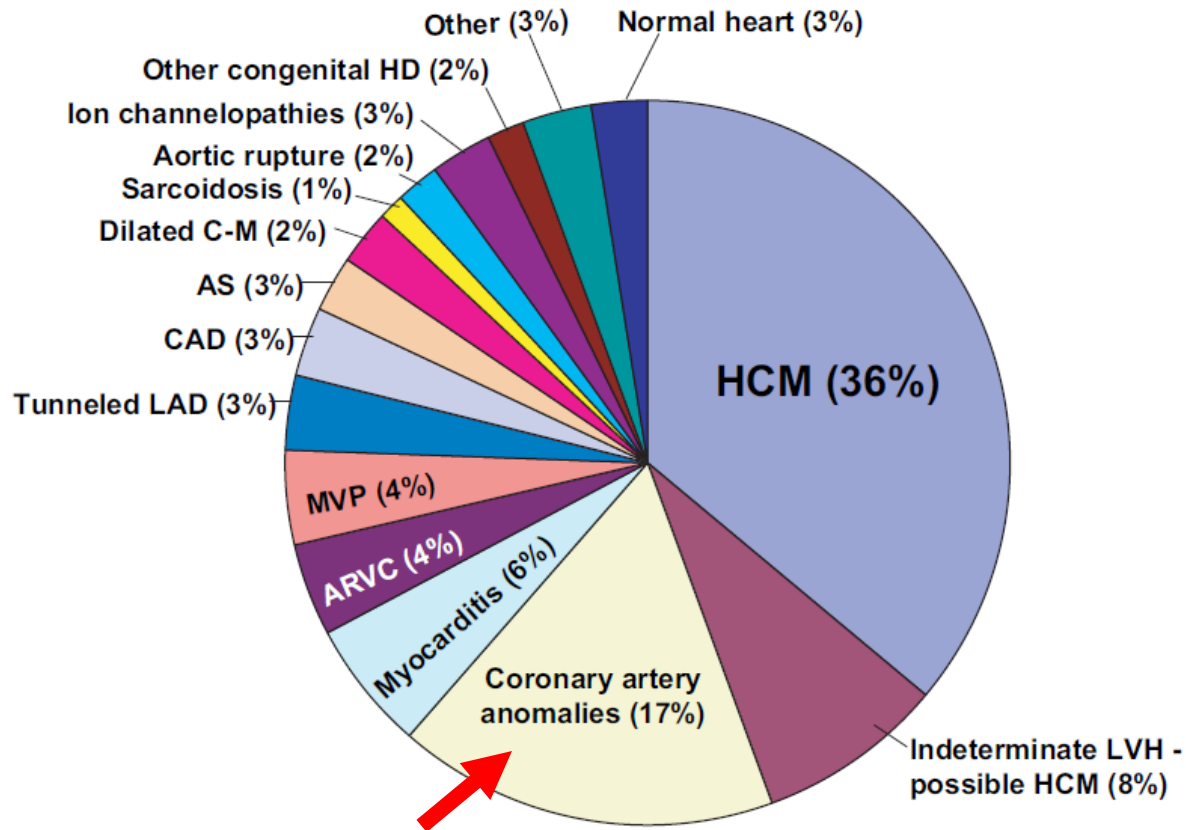


Normal

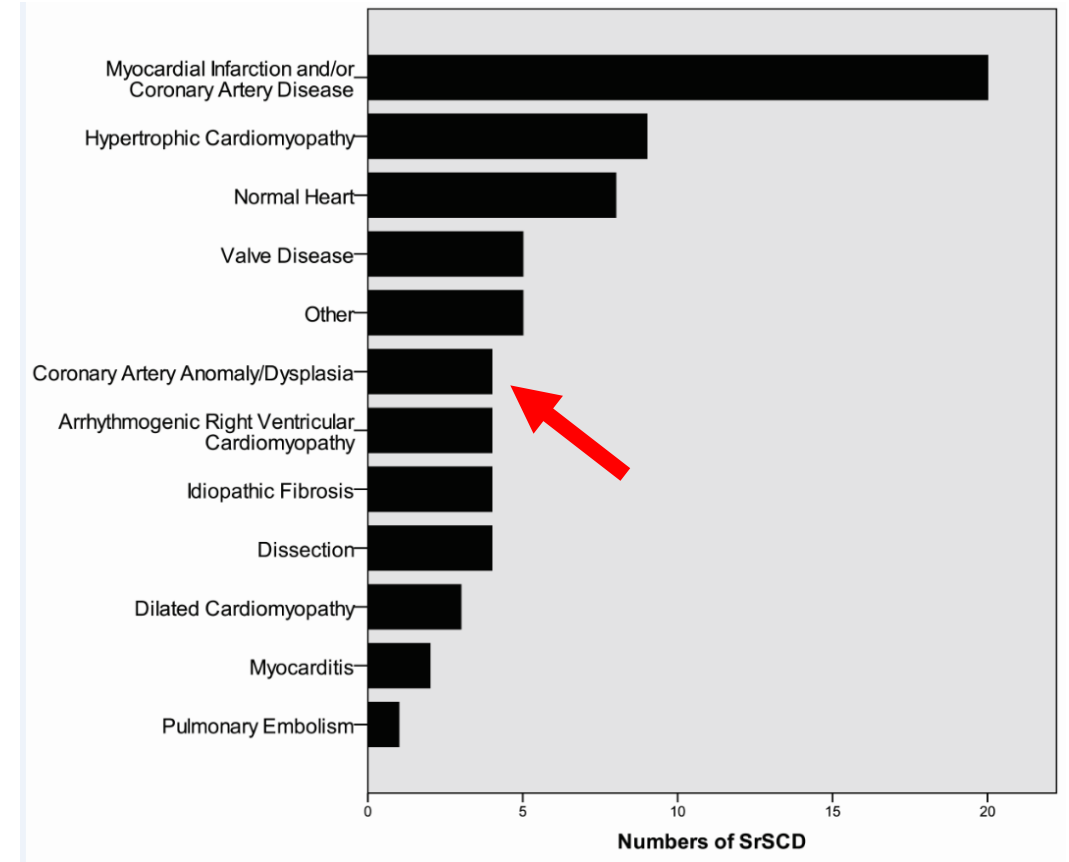
Gräni et al. Swiss Med Wkly. 2016

Are AAOCA dangerous?

Sports-related sudden cardiac death in the young: autopsies

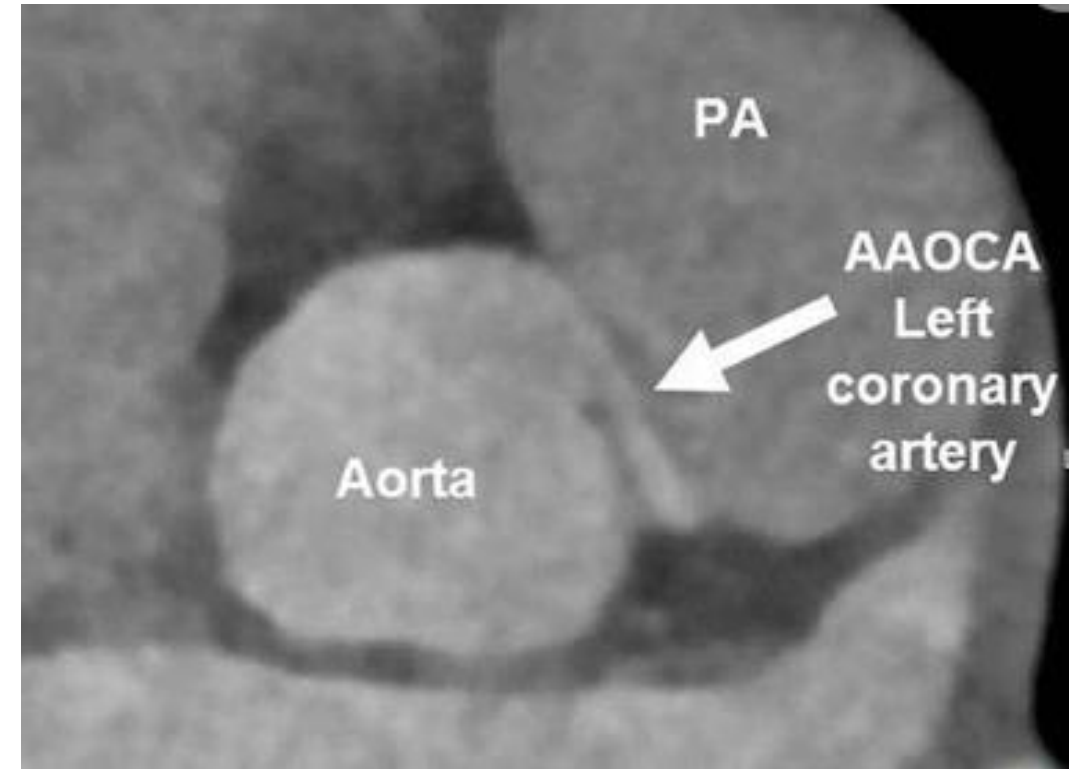
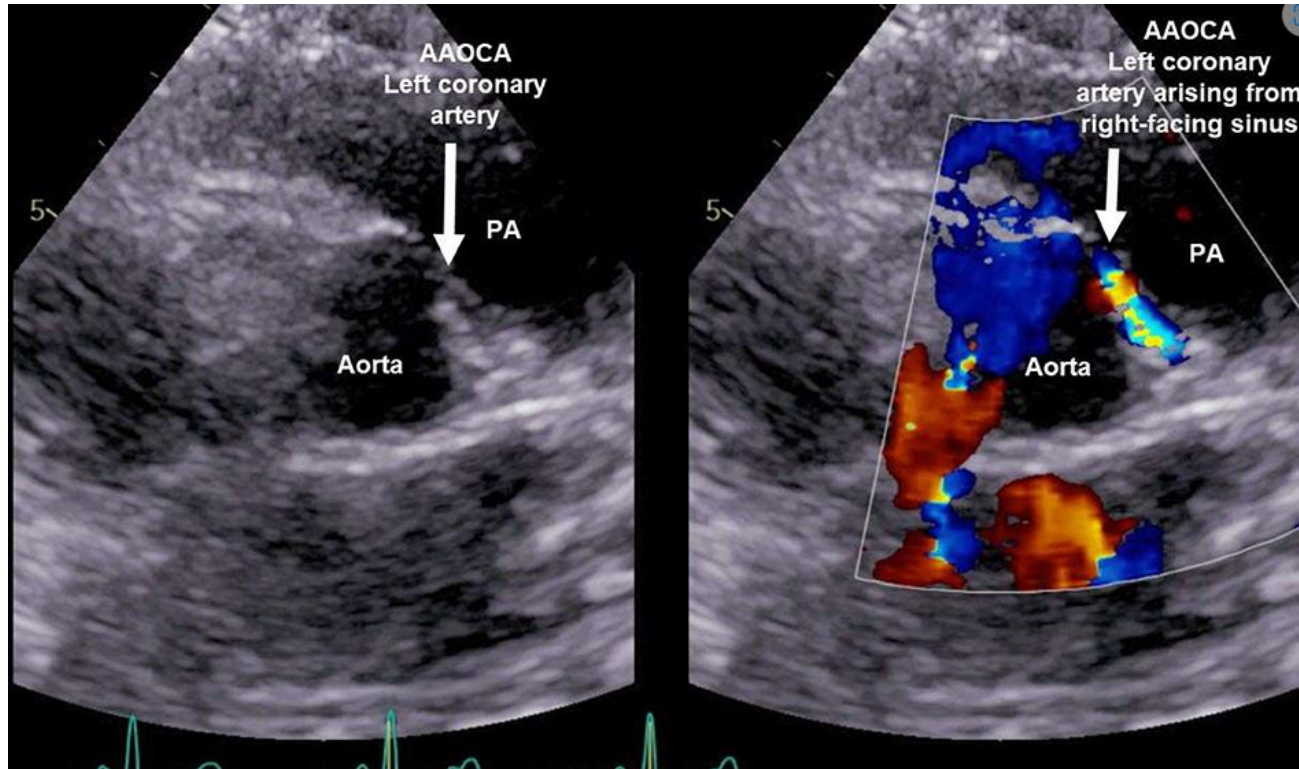


Maron BJ et al. Circulation. 2007



Gräni et al. Eur J Prev Cardiol. 2016

14 y/o, chest pain and dizzines while exercise, elevated Troponin



Borns J. Gräni C. et al. Front Cardiovasc Med. 2020

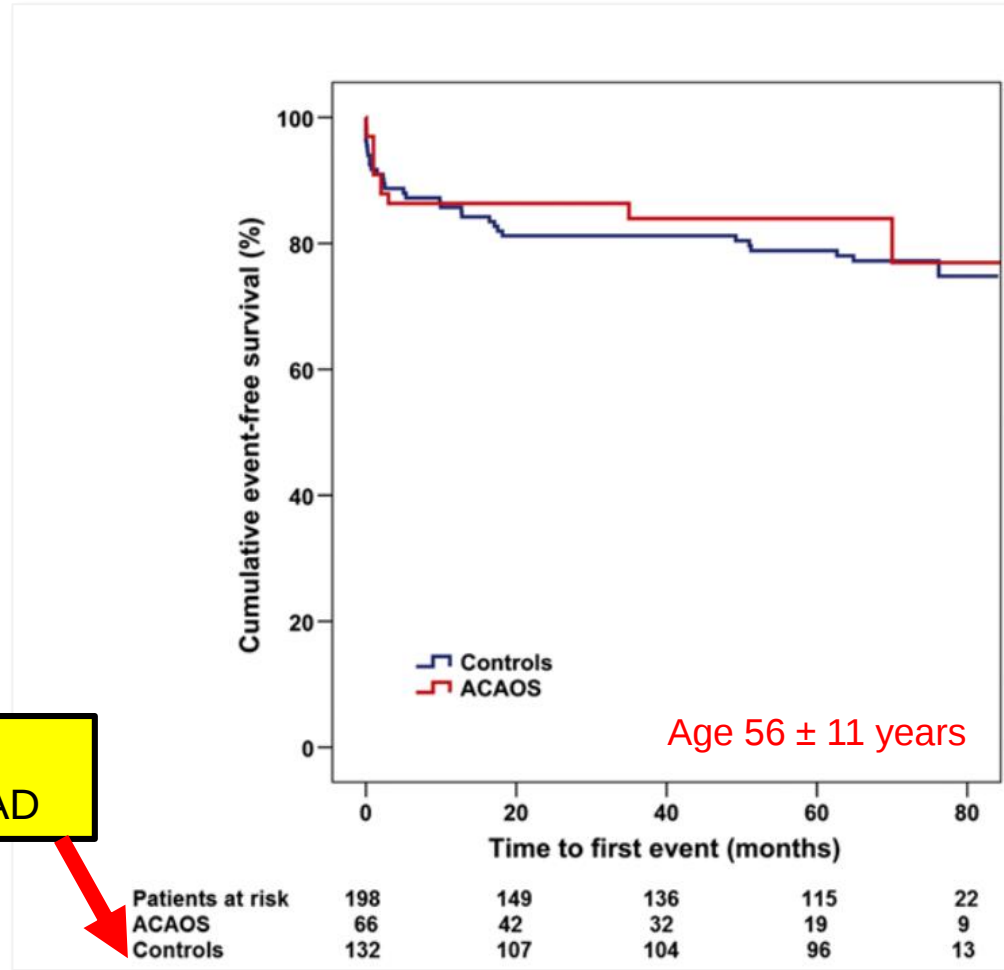
Issue in adults with AAOCA

- CCTA usage↑ according to US and EU Guidelines for CAD assessment

→ AAOCA increasingly diagnosed

→ Symptoms are not reliable

Middle aged/older individuals with AAOCA



Gräni C et al. Eur Heart J. 2017 Jul 1;38(25):2009-2016.

J-CONOMALY Registry - AAOCA

Parameter	Total (n=65)	SCA (n=30)	Non-SCA (n=35)	AMI (n=5)	Angina (n=23)	Syncope (n=7)	P-value
Age (years)	41±23	29±20	52±21	53±26	57±16	34±25	<0.0001
Male sex	48 (74)	26 (87)	22 (63)	3 (60)	14 (61)	5 (71)	0.05
R-AAOCA	47 (72)	20 (67)	27 (77)	2 (40)	21 (91)	4 (57)	0.35
L-AAOCA	18 (28)	10 (33)	8 (23)	3 (60)	2 (9)	3 (43)	0.35

TABLE 1 Clinical and Histopathological Features of SCD Victims With AOCA

Age, yrs	Sex	Cardiac Symptoms	Circumstances of Death	HW (g)	LV Fibrosis
ALCA					
17	M	—	Died during exertion	320	—
43	M	—	Died at rest	451	—
18	M	—	Died during exertion	316	—
58	F	Palpitations CP	Died during exertion	340	—
15	M	Syncope	Died during exertion	318	—
13	M	Syncope	Died during exertion	220	—
11	M	Syncope	Died in sleep	204	—
15	M	CP, syncope	Died during exertion	341	—
13	M	Syncope	Died during exertion	140	—
28	F	Palpitations	Died in sleep	431	—
16	M	—	Died during exertion	350	Posterior
ARCA					
39	M	—	Died at rest	410	Multiple locations
21	M	CP	Died during exertion	323	Multiple locations
14	M	—	Died in sleep	285	—
43	F	—	Died in sleep	558	—
20	M	—	Died at rest	478	Anterolateral
43	M	—	Died at rest	474	Posterior
23	M	—	Died during emotional stress	337	—
63	M	—	Died at rest	390	Posterior*
38	F	—	Died in sleep	323	—
18	M	—	Died in sleep	390	—
24	M	—	Died at rest	210	—

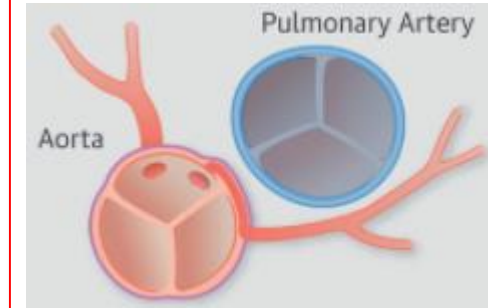
Anatomic high-risk features

Course
between
Aorta and A.
pulmonalis



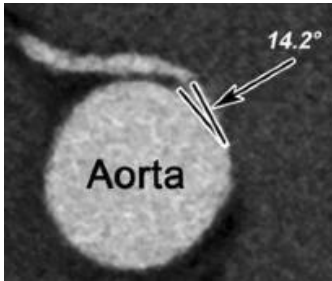
Interarterial course

Intramural course



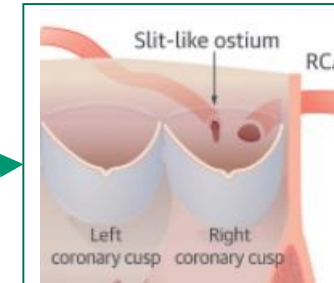
Course
within
tunica
media

Take off <45°



Acute take-off angle

Slit-like ostium



≥50% reduction
of the *ostial*
minimal lumen
diameter

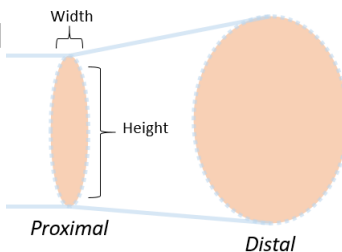
Elliptic vessel shape

Proximal narrowing



≥50% reduction of the
proximal minimal
lumen diameter/area
compared to the
normal distal reference

Elliptic proximal
vessel shape
defined as
height/width
ratio of >1.3



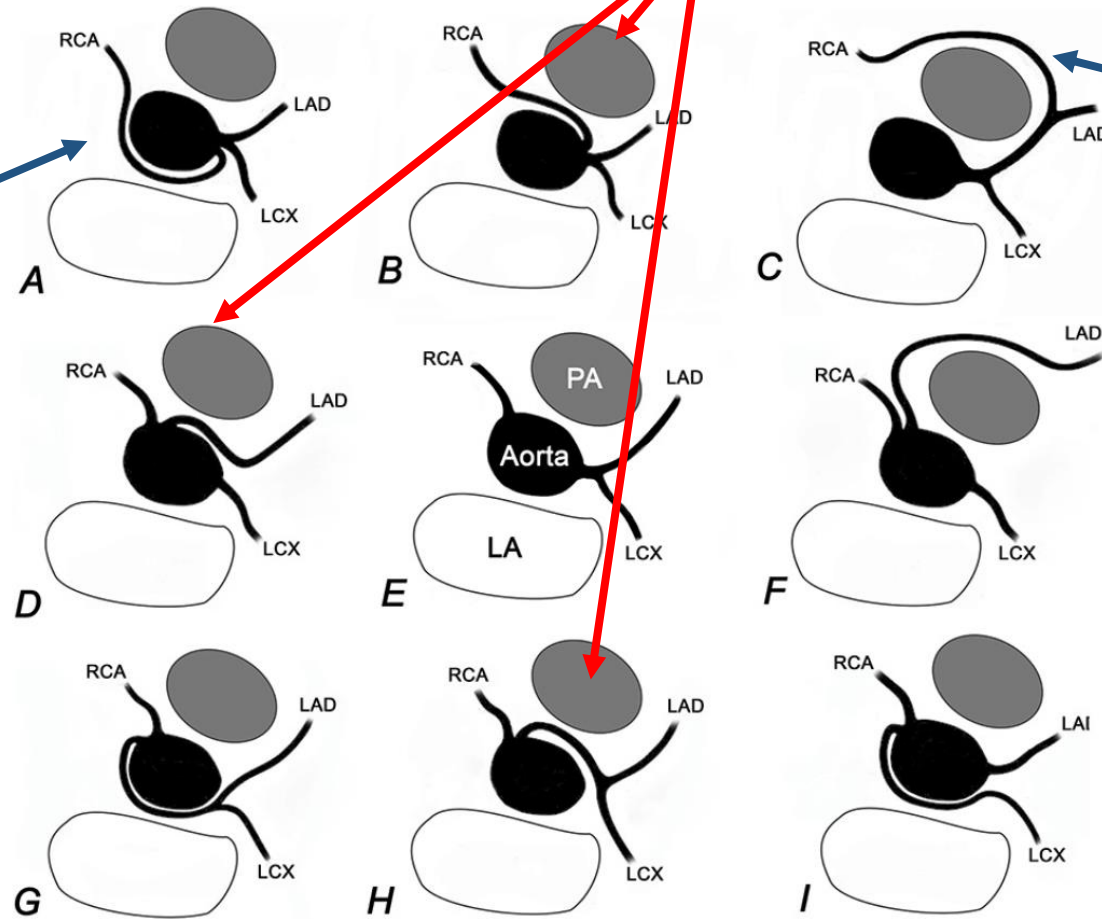
Gräni et al. JACC Cardiovasc Imaging 2017
Gräni et al. J Nucl Cardiol 2017

Variants of AAOOCA

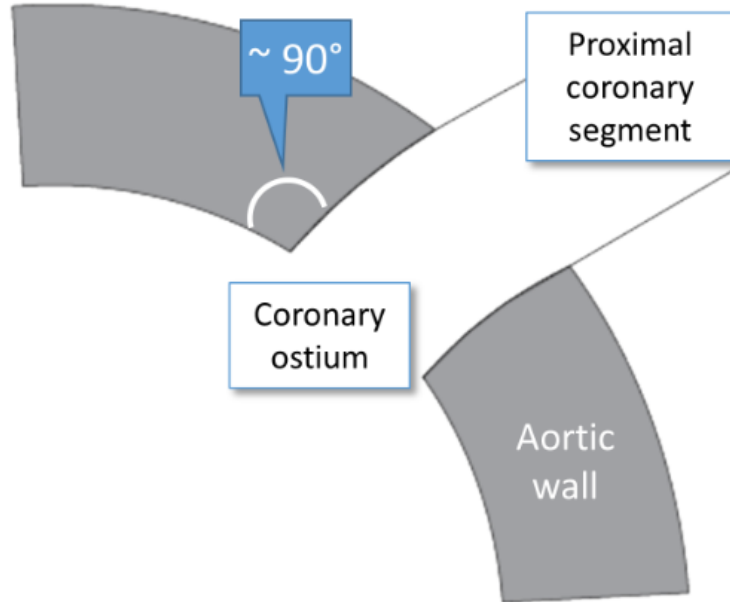
**Interarterial
Course = «Malignant»?**

**Retroaortic
course**

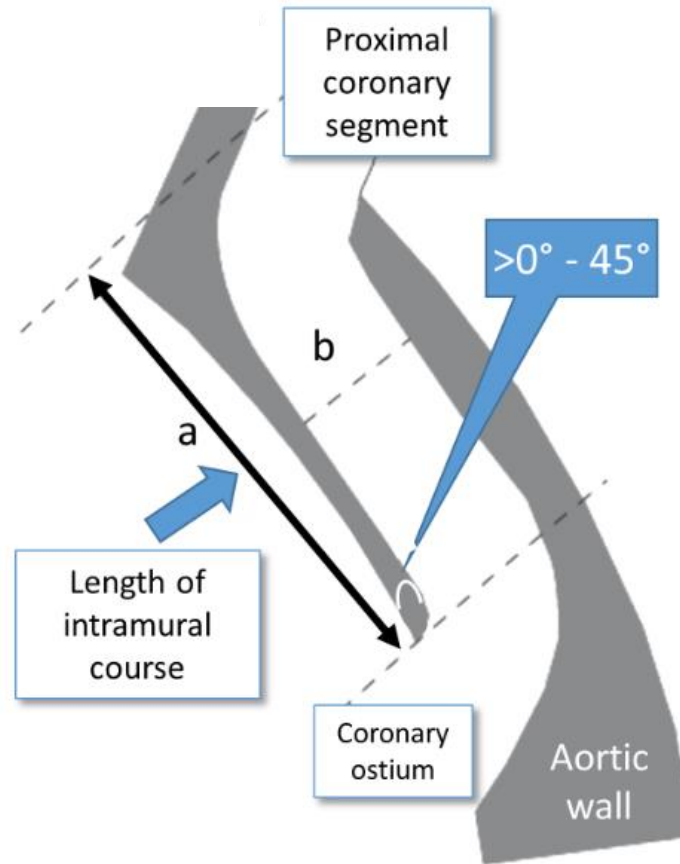
**Pre-pulmonic
course**



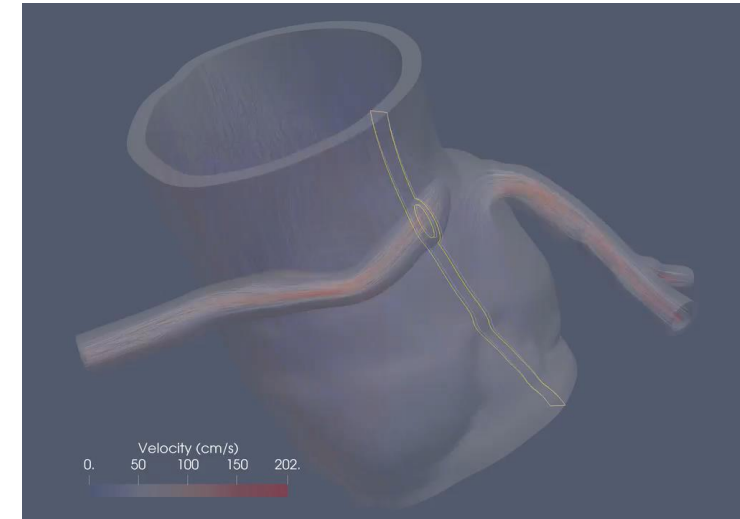
Intramural course



Normal



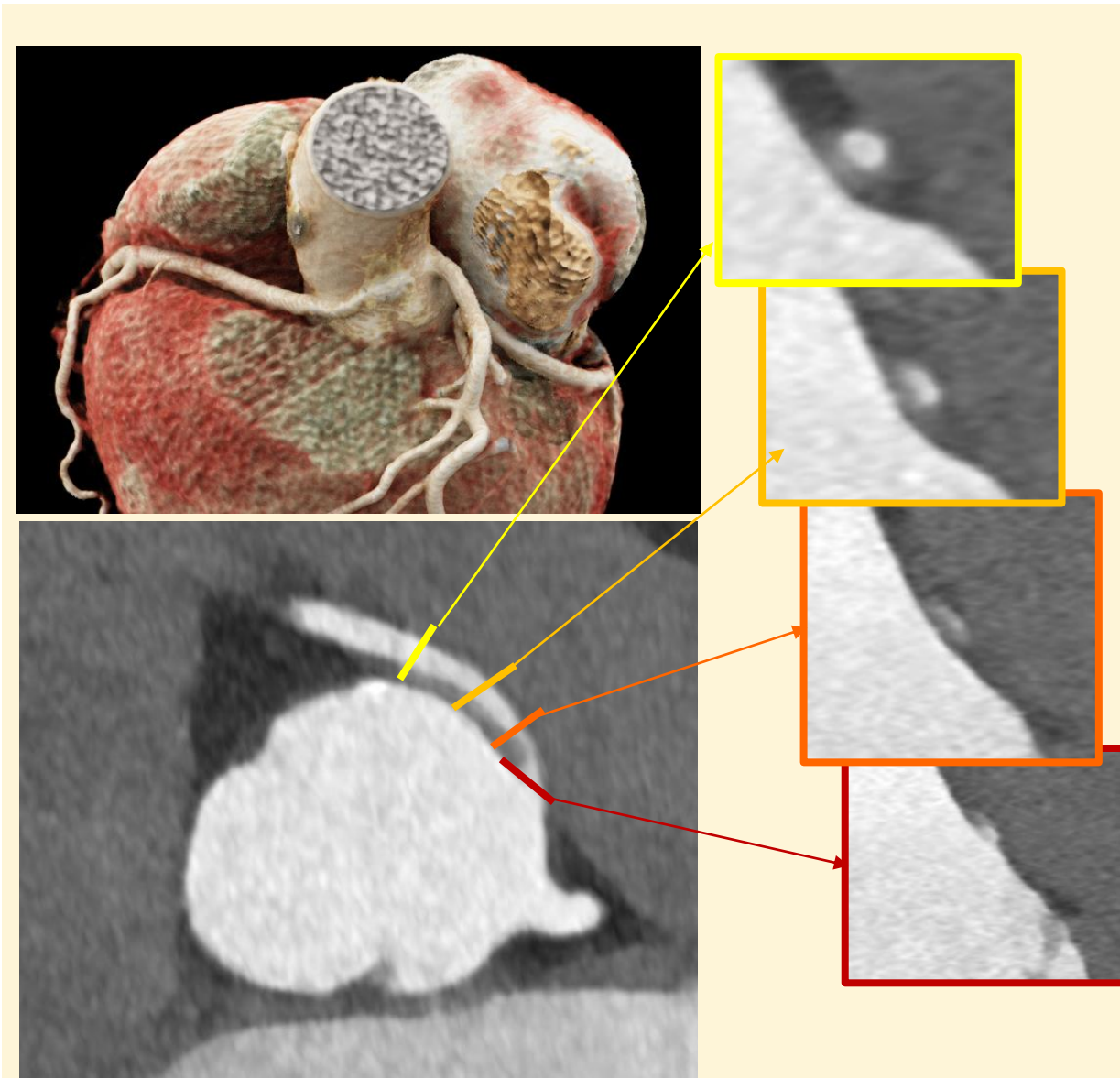
Intramural course



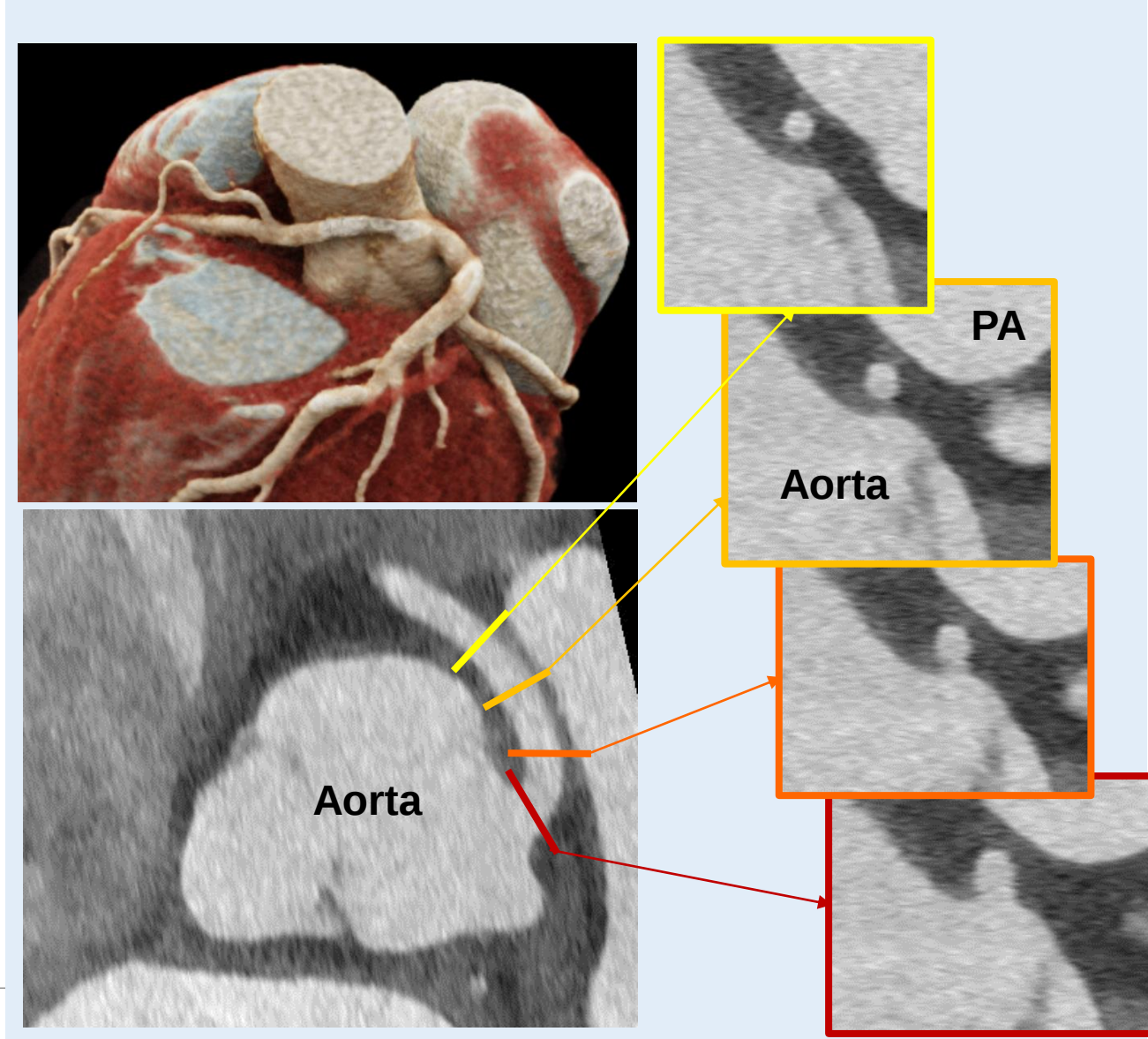
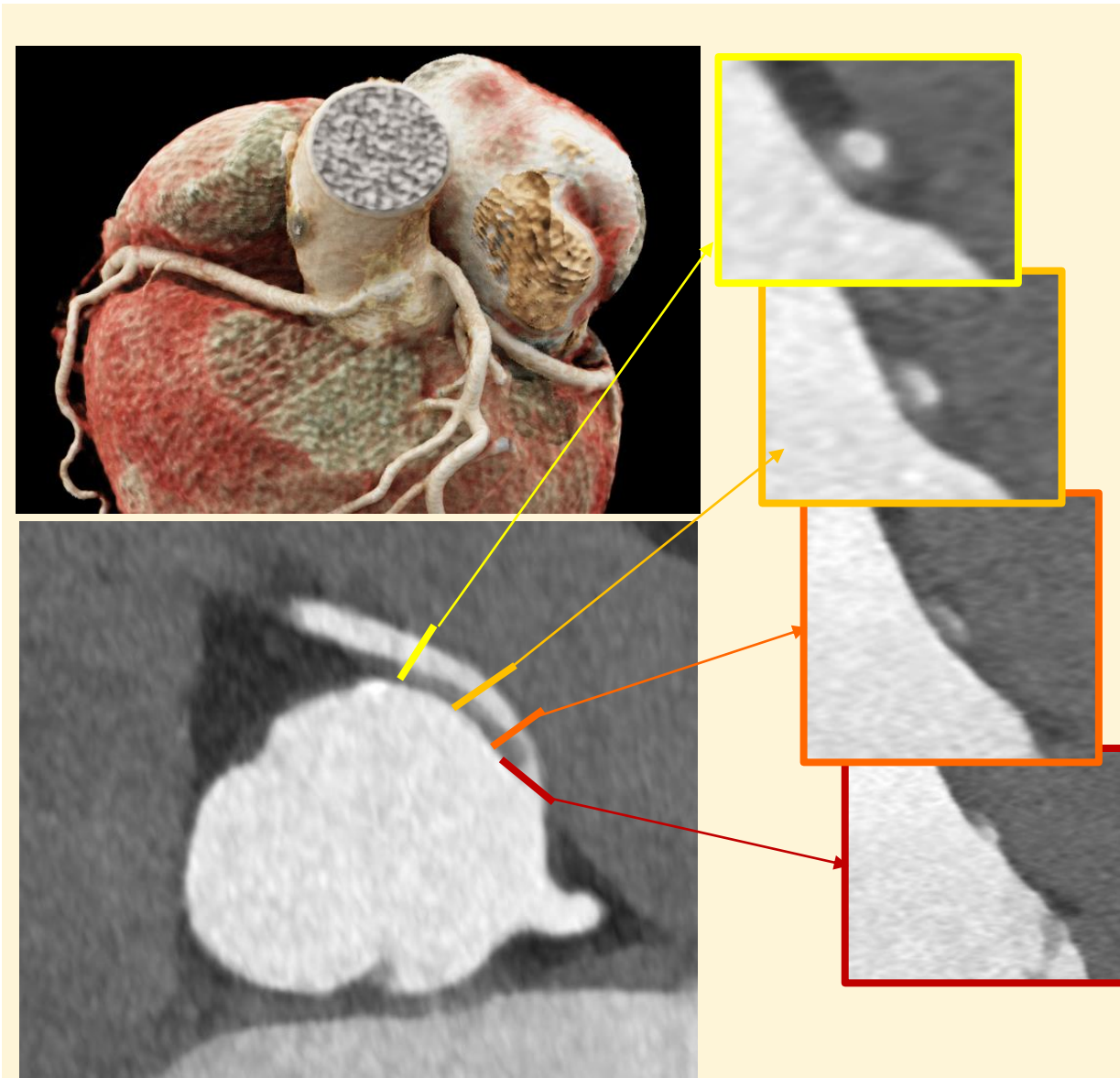
Bigler & Gräni C et al. J Am Heart Assoc 2022

Jiang et al. JTCVS Tech. 2022

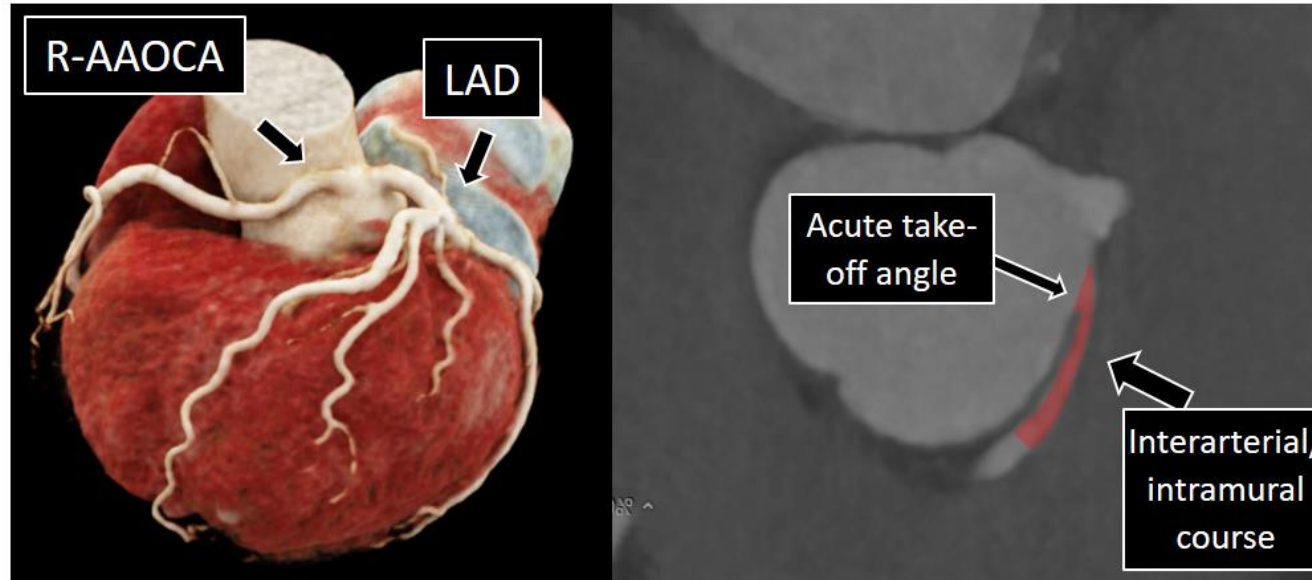
AAOCA with interarterial course **with & without intramural course**



AAOCA with interarterial course **with & without intramural course**



False concept: scissor like compression of interarterial course



HR Rest	HR Stress	Max HR (220 - Age)	BP Rest	BP Stress	PAP Rest	PAP Stress	CoP Rest	CoP Stress
67	150	168	116/64	142/63	22/11	47/24	100/59	95/60

Schütze, Räber, Gräni et al. Eur Heart J Case Rep 2024

Pathophysiology

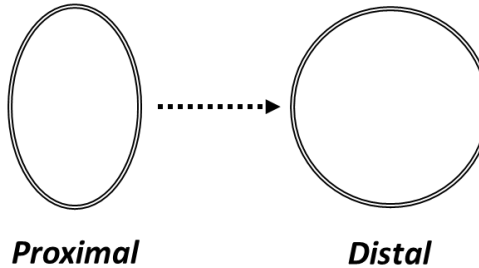
Physical or Dobutamine

Adenosine/Regadenoson

Pathophysiology in patients with AAOCA

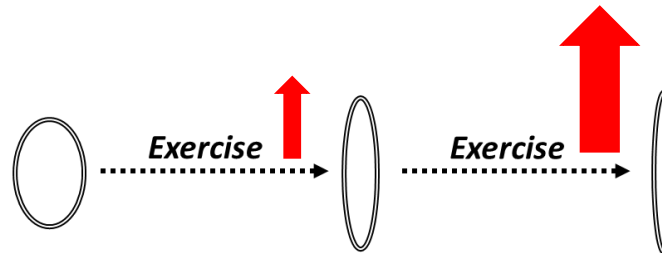
Fixed

- Slit-like ostium
- Proximal narrowing



Dynamic

- Acute take-off angle
- Intramural course with lateral compression



Most important factors for dynamic compression:

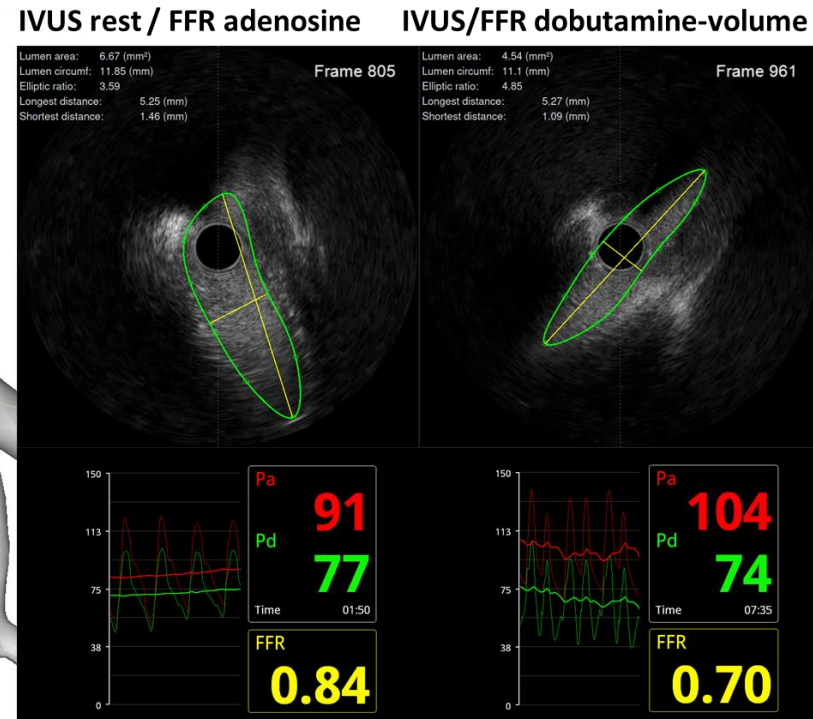
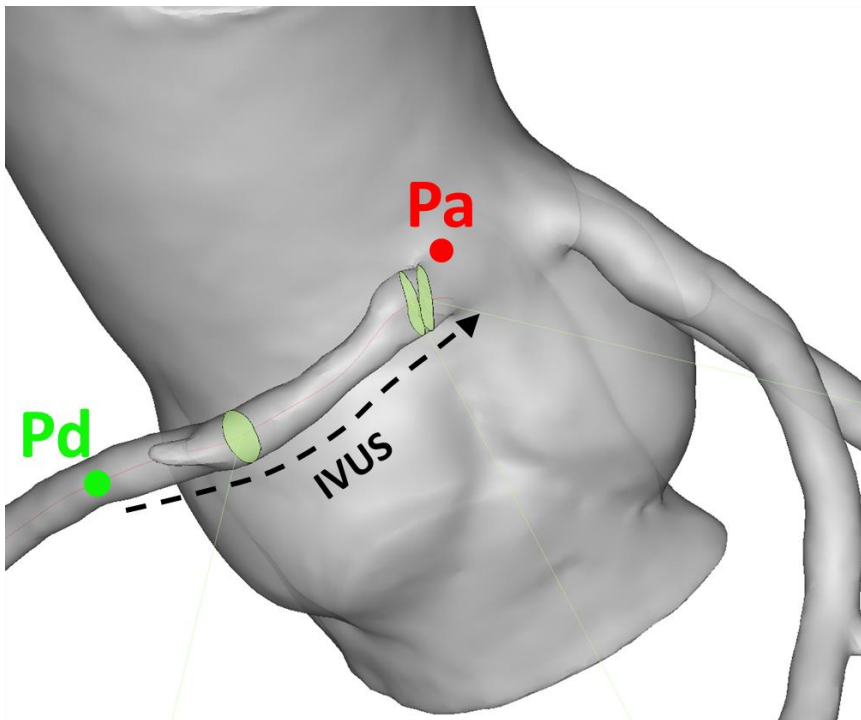
- Increased stroke volume
- Blood pressure (systole/diastole)
- Heart rate

Other factors to consider

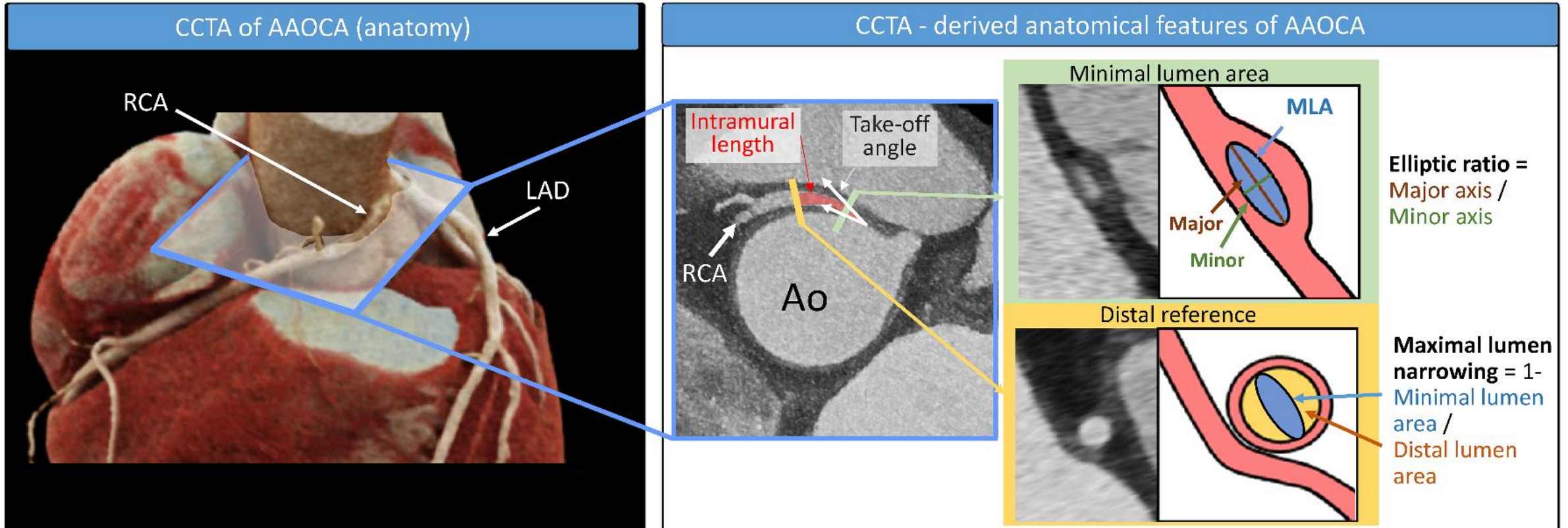
- Pre-load
- Afterload (Microvascular Resistance)
- Stiffness of the Aorta

Invasive assessment

- Dobutamin-volume: Assessment of **fixed** + **dynamic** stenosis
 - $FFR_{Dobutamine}$ and $IVUS_{Dobutamine}$
 - Max. Dobutamine $40\mu\text{g/kg}$ per body weight per minute + $\sim 3'000\text{ml}$ ringer lactate + 1mg atropine

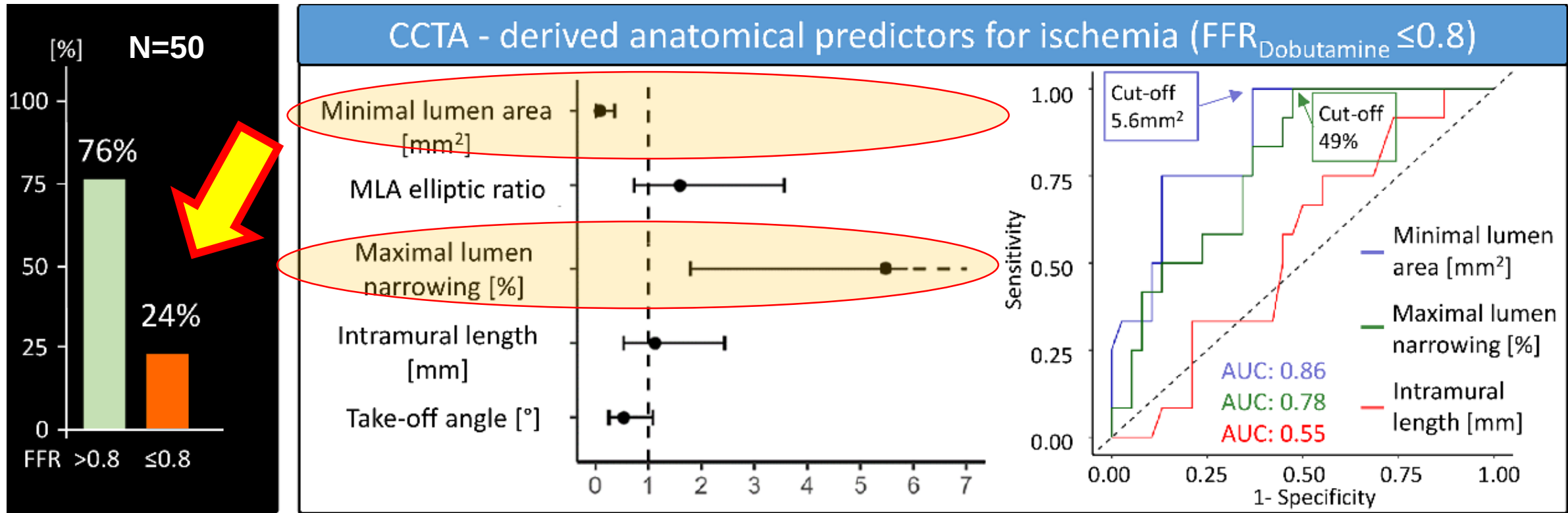


CCTA predictors for hemodynamic relevance in R-AAOCA-IA-IM



Stark, Räber, Gräni et al. Circulation. 2025

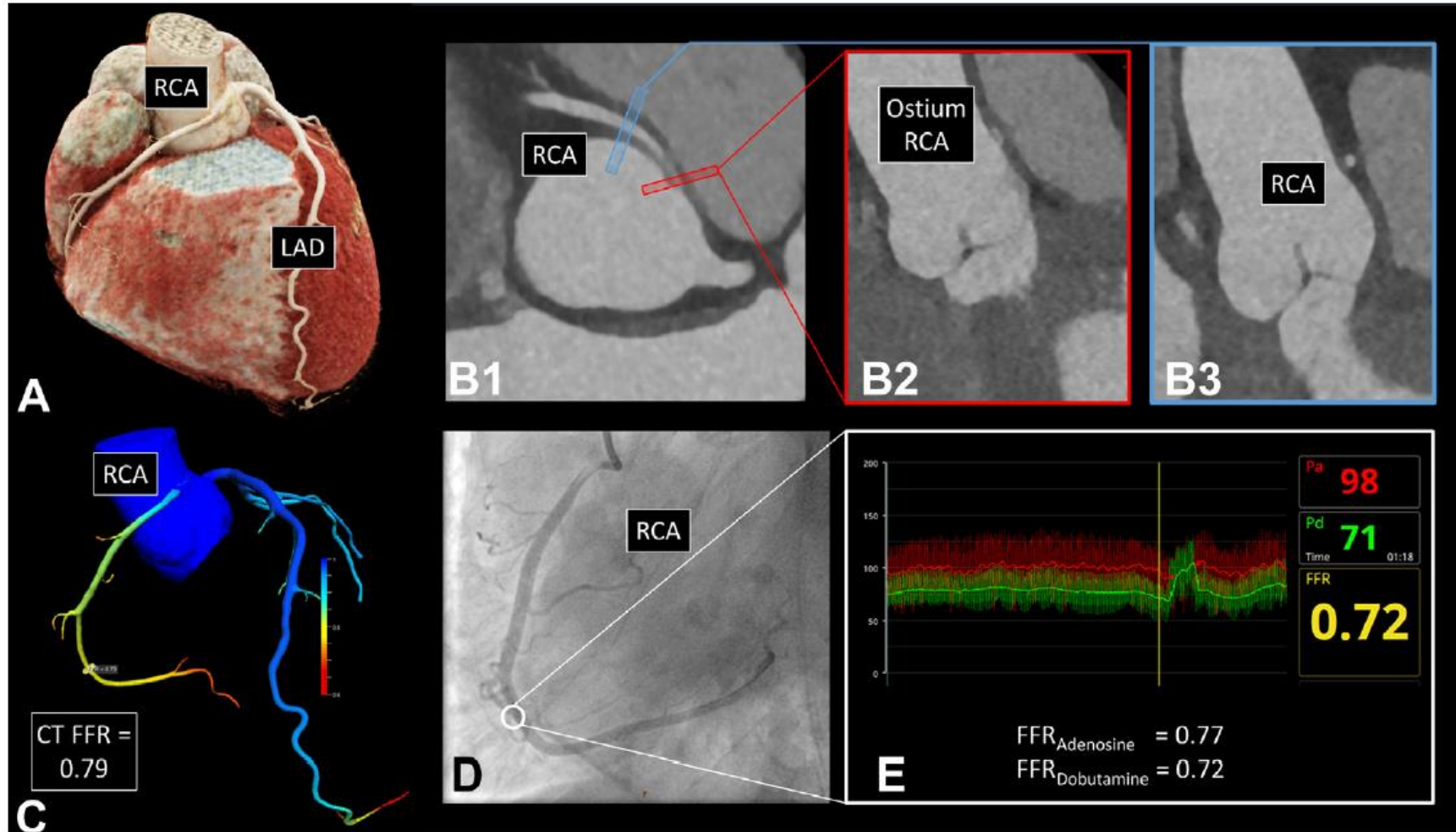
CCTA versus Dobutamin FFR reference in R-AAOCA-IA-IM



→ Not all “malignant” (hemodynamically relevant)

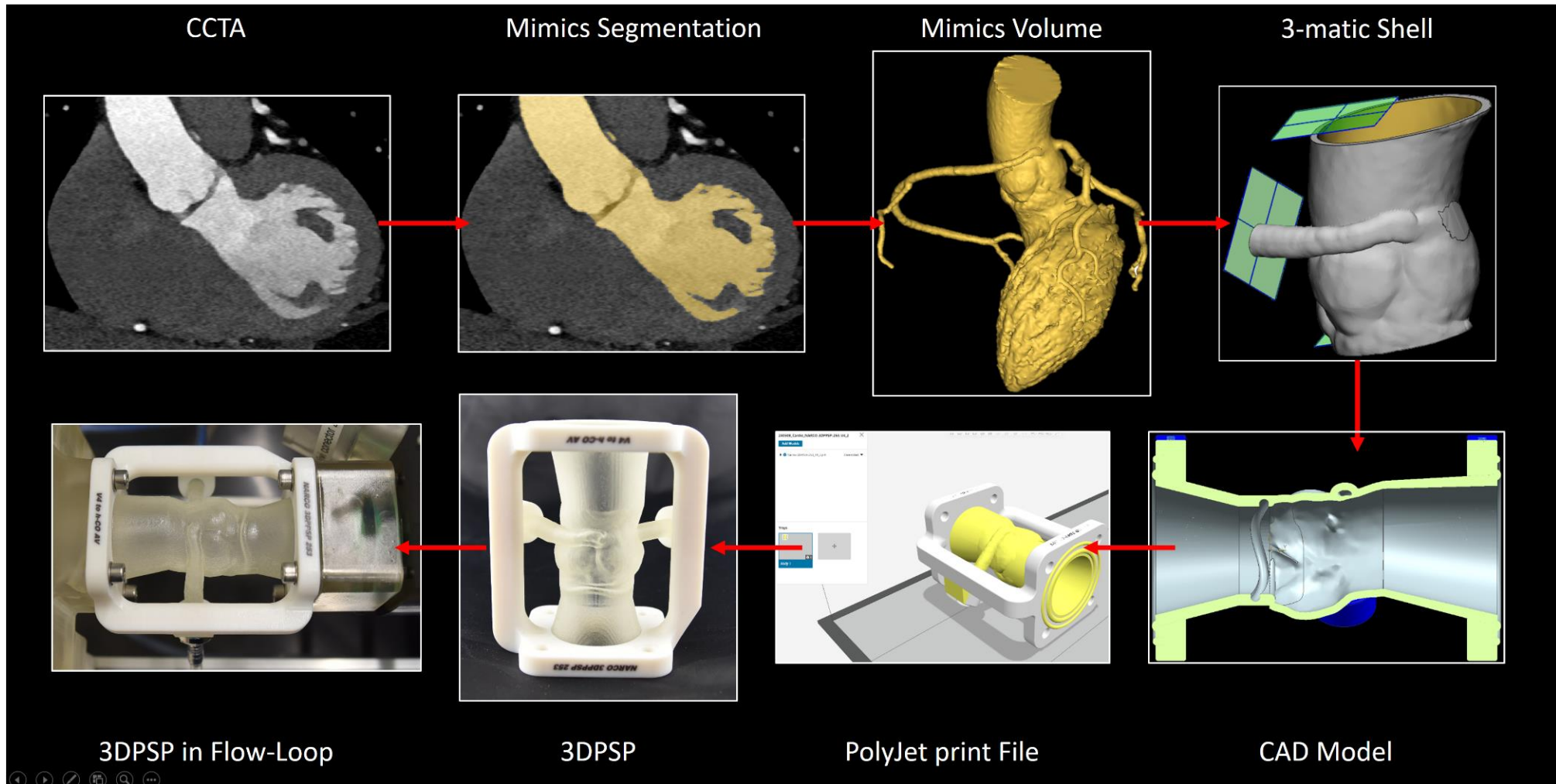
Outlook: Digital Twin and Artificial Physical Twin

Computational fluid dynamics

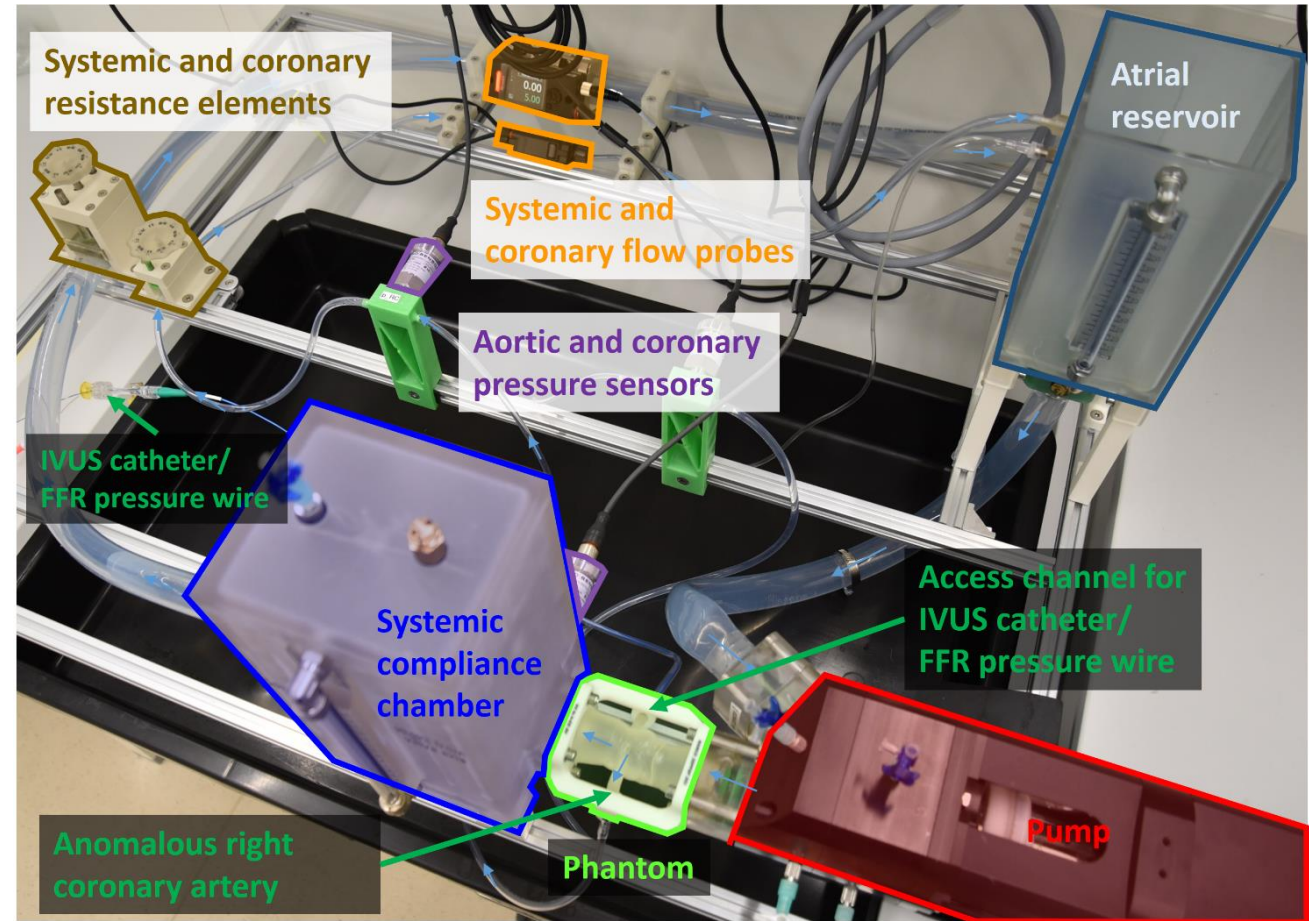
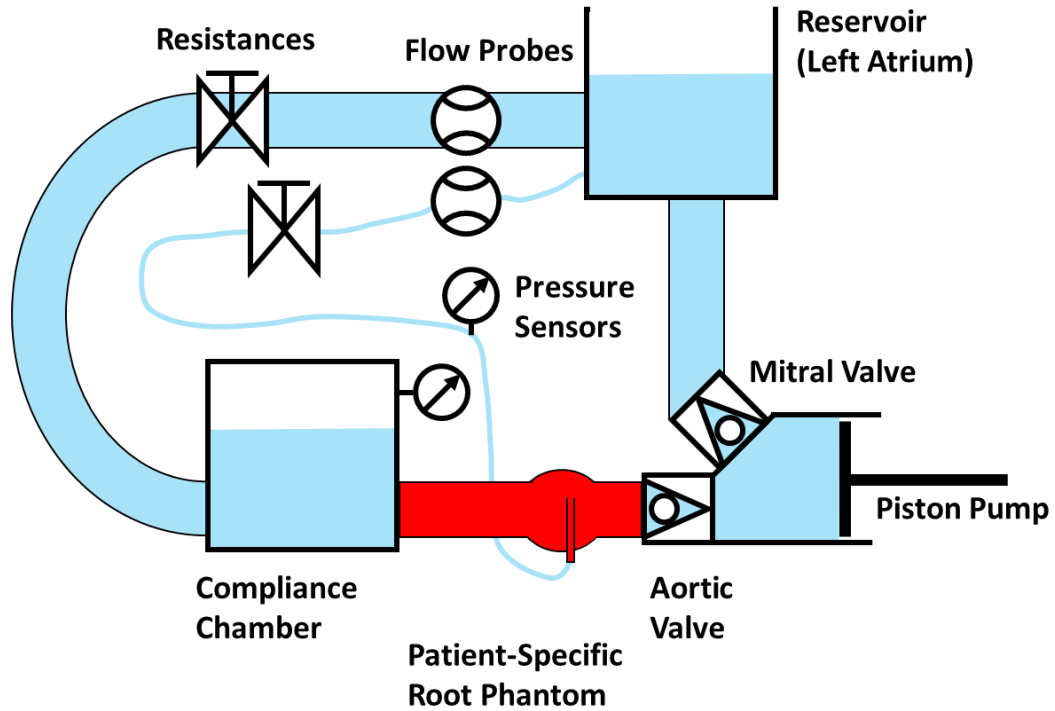


Bigler & Gräni et al. JACC Case Rep 2022
Stark & Gräni et al. J Cardiovasc Dev Dis 2023

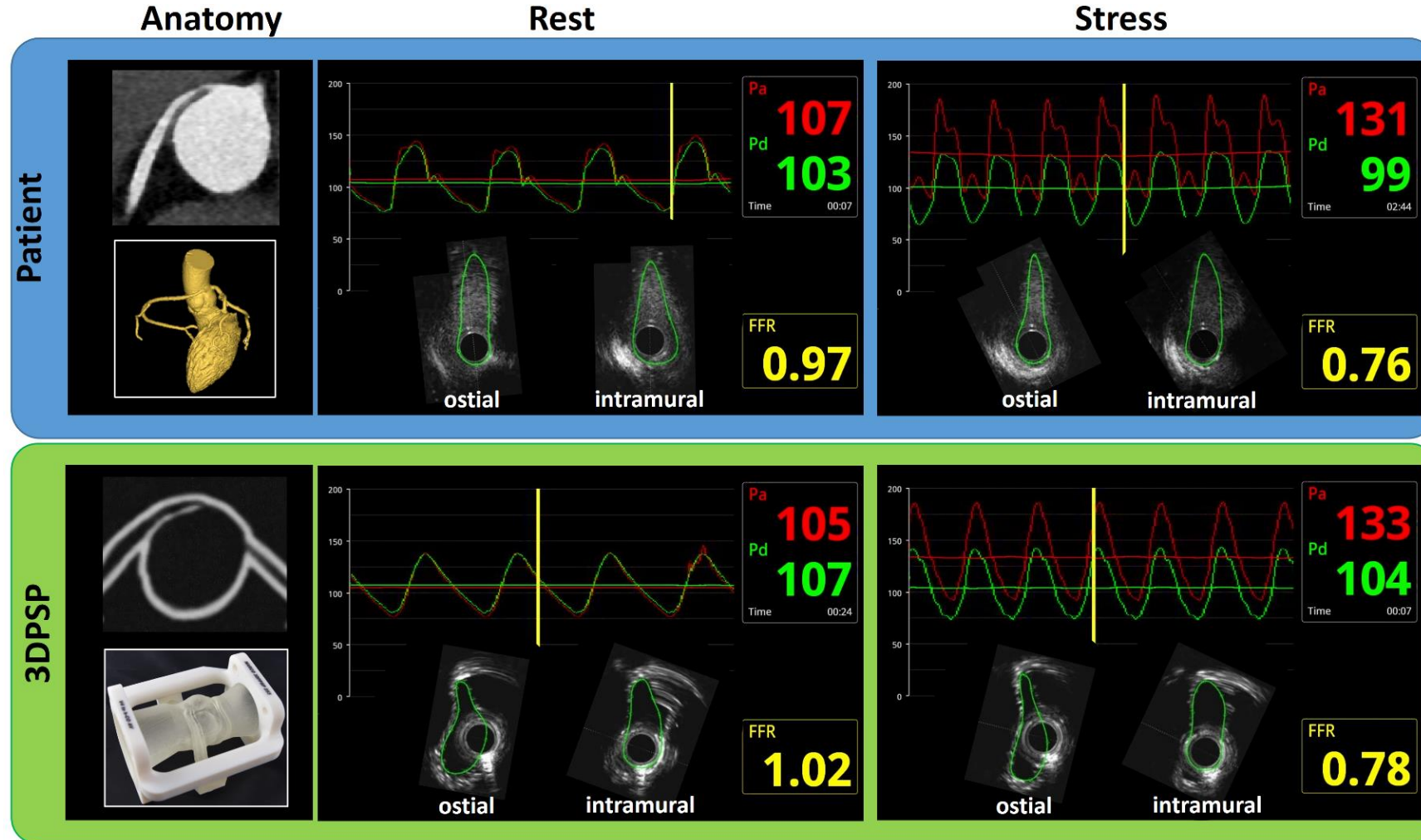
3D printed flow loop AAOCA



3D printed flow loop AAOCA



3D printed flow loop AAOCA



nature communications



Article

<https://doi.org/10.1038/s41467-025-58362-9>

AI-based detection and classification of anomalous aortic origin of coronary arteries using coronary CT angiography images

Received: 24 September 2024

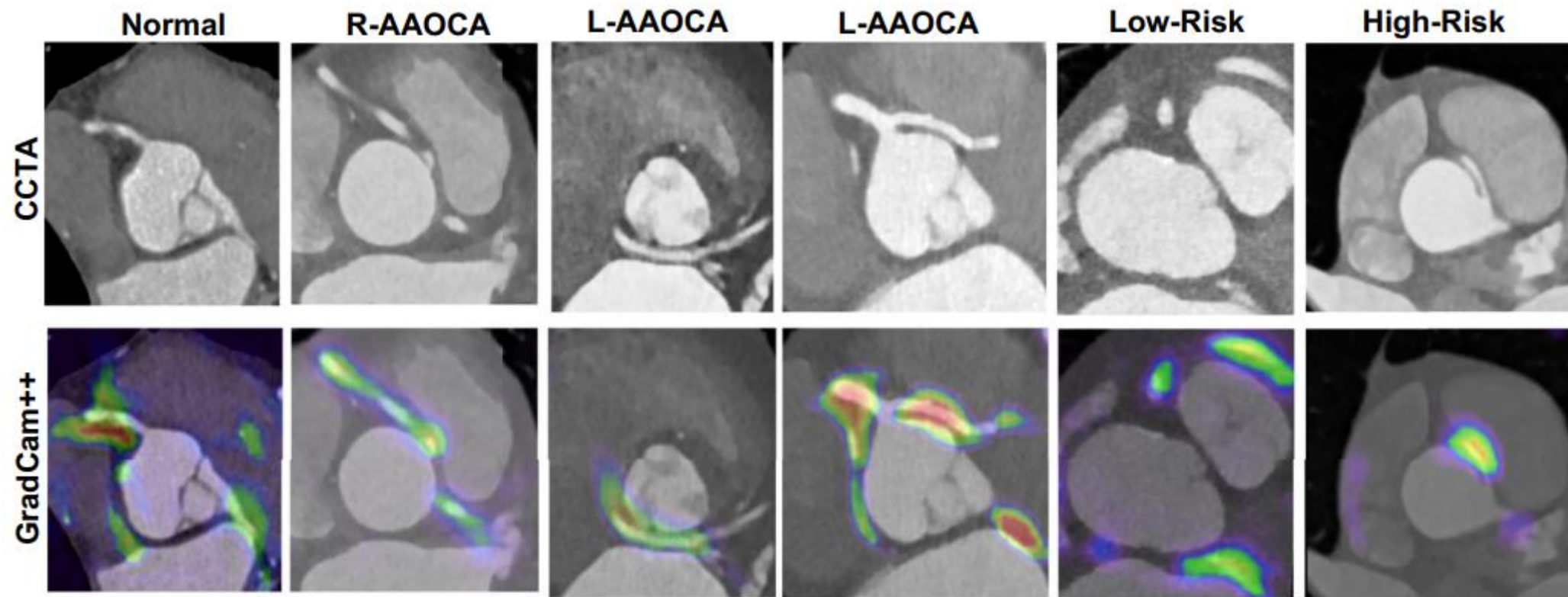
Accepted: 5 March 2025

Published online: 01 April 2025



Check for updates

Isaac Shiri¹, Giovanni Baj ^{1,5}, Pooya Mohammadi Kazaj ^{1,5}, Marius R. Bigler¹, Anselm W. Stark¹, Waldo Valenzuela ², Ryota Kakizaki¹, Matthias Siepe³, Stephan Windecker¹, Lorenz Räber¹, Andreas A. Giannopoulos⁴, George CM. Siontis¹, Ronny R. Buechel ⁴ & Christoph Gräni ¹ ✉



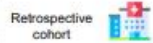
Open access data set with segmentation for developing and evaluation of segmentation model



1203 3D-CT images with the label for aorta and LV

Data with the label for AAOCA

Training & validation dataset



536 Patients

1567 3D-CCTA

Prospective cohort

359 Patients

1066 3D-CCTA

Data without label for AAOCA

External testing dataset

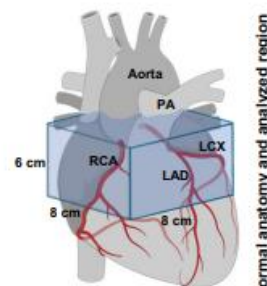
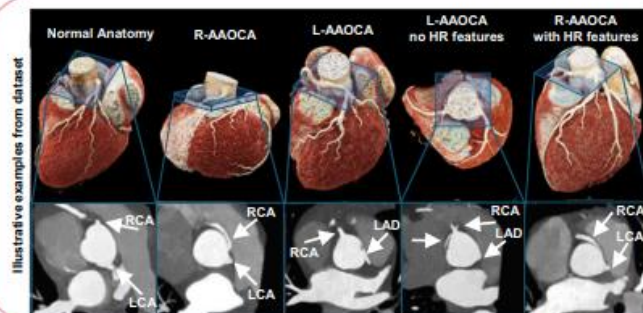
483 Patients

497 3D-CCTA

External clinical evaluation dataset

998 Patients

998 3D-CCTA



Normal anatomy and analyzed region

Segmentation

Automatically segment the aorta and LV to detect the intersection for cropping purposes



Anomaly Detection

Classification of anomaly versus the normal cases to detect the AAOCA

Class 0: Normal
Class 1: AAOCA



Origin Classification

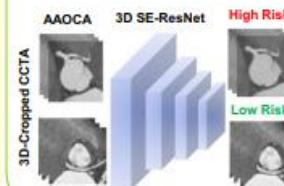
Classification of the anomalous vessel

Class 0: R-AAOCA
Class 1: L-AAOCA

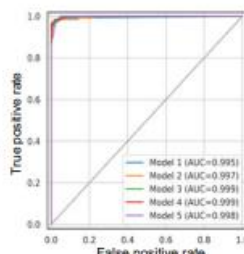


Risk Classification

Low-Risk: pre-pulmonic, retro-aortic, high take off
High-Risk: inter-arterial and sub-pulmonic



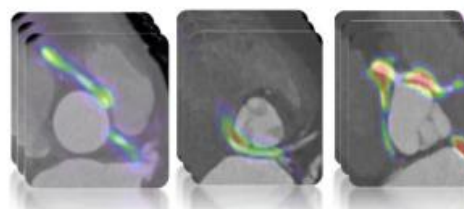
t-SNE in Anomaly detection



ROC curve external testing

True label	Predicted label	
	0	1
0	358	0
1	6	133

External testing



Interpretability using GradCam++

Conclusions

- ❖ AAOCA numbers in adults increase, CAD more important, but AAOCA can become relevant later in life
- ❖ Thorough anatomical assessment & ischemia testing important
- ❖ Invasive testing preferred over noninvasive stress (CAVE: evidence needed!) → use physical or dobutamine testing
- ❖ R-AAOCA-IA-IM: 25% hemodynamic relevant → Mortality is low, mostly «innocent» bystanders
- ❖ Future role: computational fluid dynamics / 3D print for ischemia simulation & intervention planning in AAOCA

AAOCA Therapy

Assess hemodynamic relevance of ACAOS

- $FFR_{Dobutamine} \leq 0.80$ or
- $IVUS_{Dobutamine} > 50\%$ Minimal Lumen Area reduction or
- Ischemia noninvasive imaging (max. physical / dobutamine)

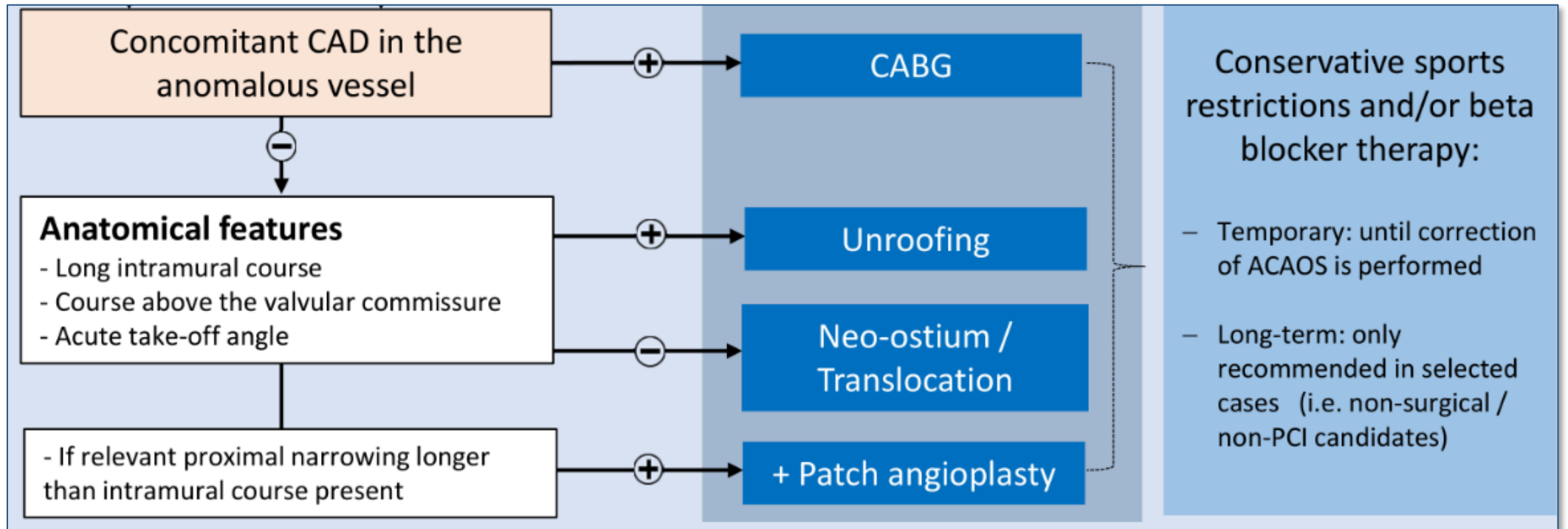


Table 2:

Surgical data

	n = 71 (100%)
Types of AAOCA correction, n (%)	
Unroofing	51 (72)
Reimplantation	20 (28)
Additional neo-ostium patchplasty	9 (13)
Patients who underwent concomitant procedures, n (%)	28 (39)
Concomitant procedures, n (%)	30
Aortocoronary bypass grafting (of the non anomalous coronary artery)	2 (3)
Aortic valve replacement	9 (13)
Valve-sparing aortic root replacement	3 (4)
Aortic root replacement with composite graft	5 (7)
Aortic valve repair	1 (1)
Aortic arch replacement	3 (4)
Supracoronary ascending aorta replacement	1 (1)
Mitral valve repair	4 (6)
Atrial myxoma removal	1 (1)
Atrial septum defect closure	1 (1)

AAOCA: anomalous aortic origin of coronary artery.

JOURNAL ARTICLE

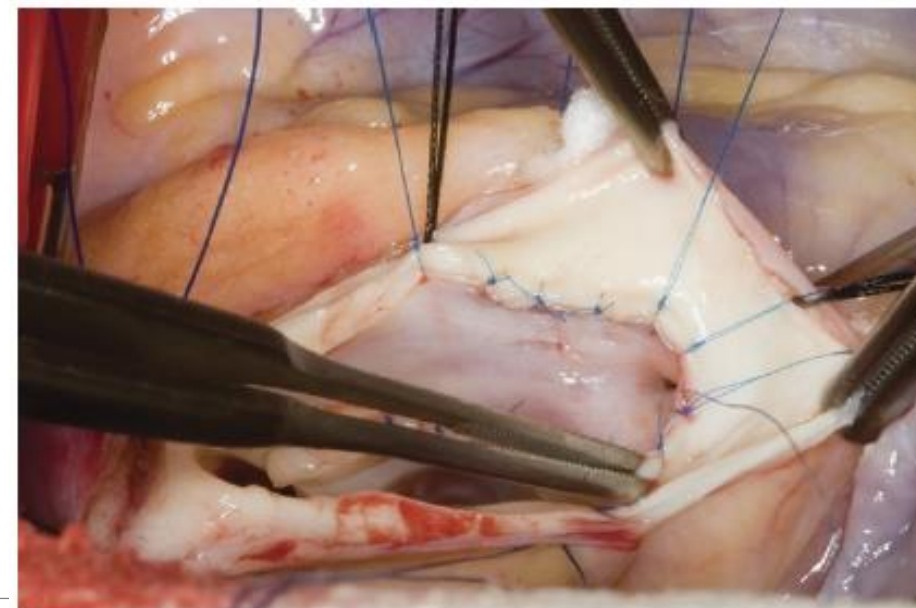
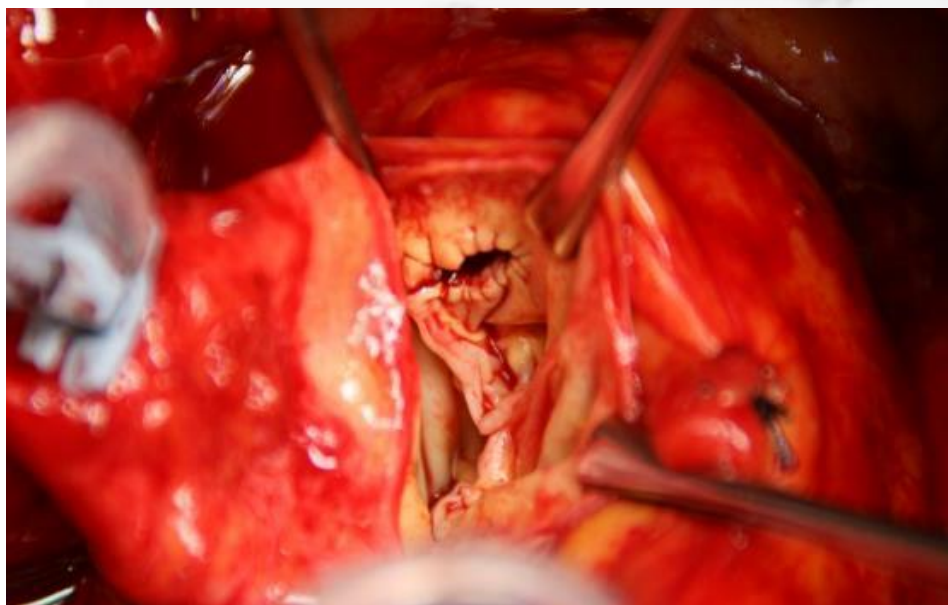
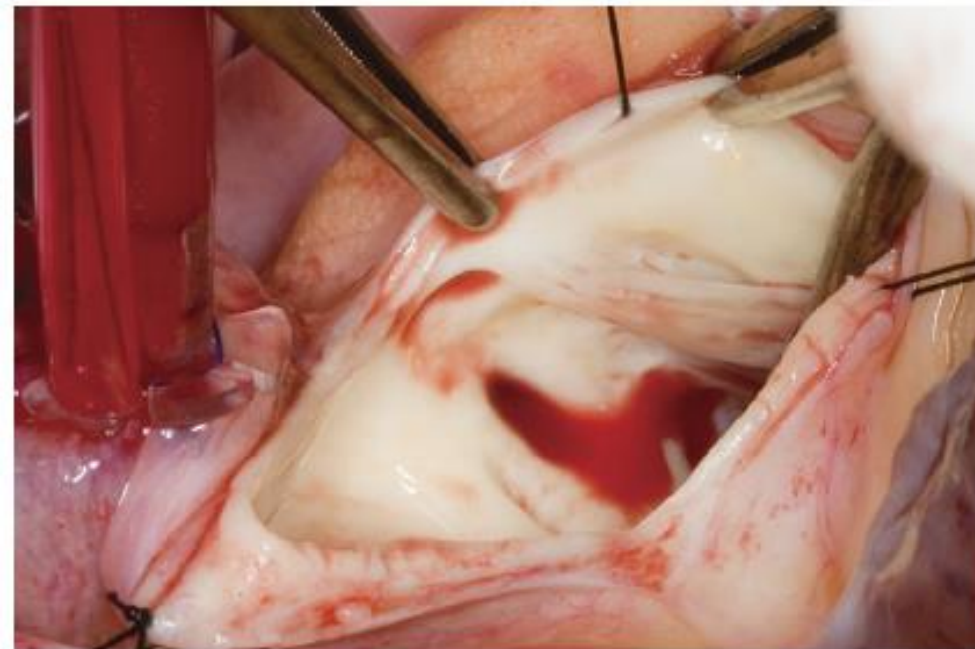
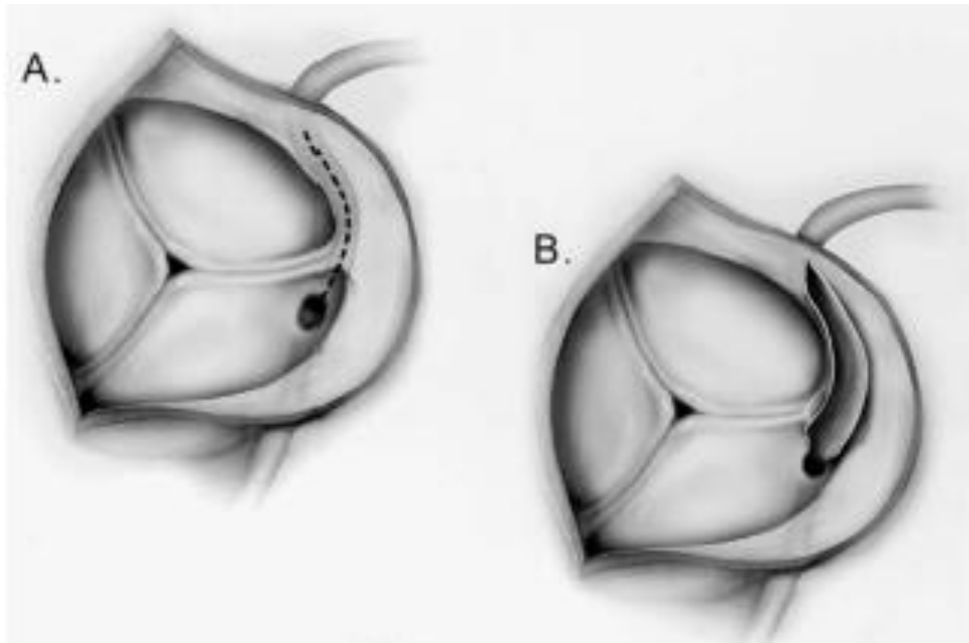
Safety and efficacy of surgical correction of anomalous aortic origin of coronary arteries: experiences from 2 tertiary cardiac centres FREE

Fabio Pregaldini ✉, Hannah Widenka, Mohamed Barghout, Christoph Gräni, Martin Czerny, Fabian A Kari, Salome Chikvatia, Alexander Kadner, Matthias Siepe

European Journal of Cardio-Thoracic Surgery, Volume 63, Issue 5, May 2023, ezad075,
<https://doi.org/10.1093/ejcts/ezad075>

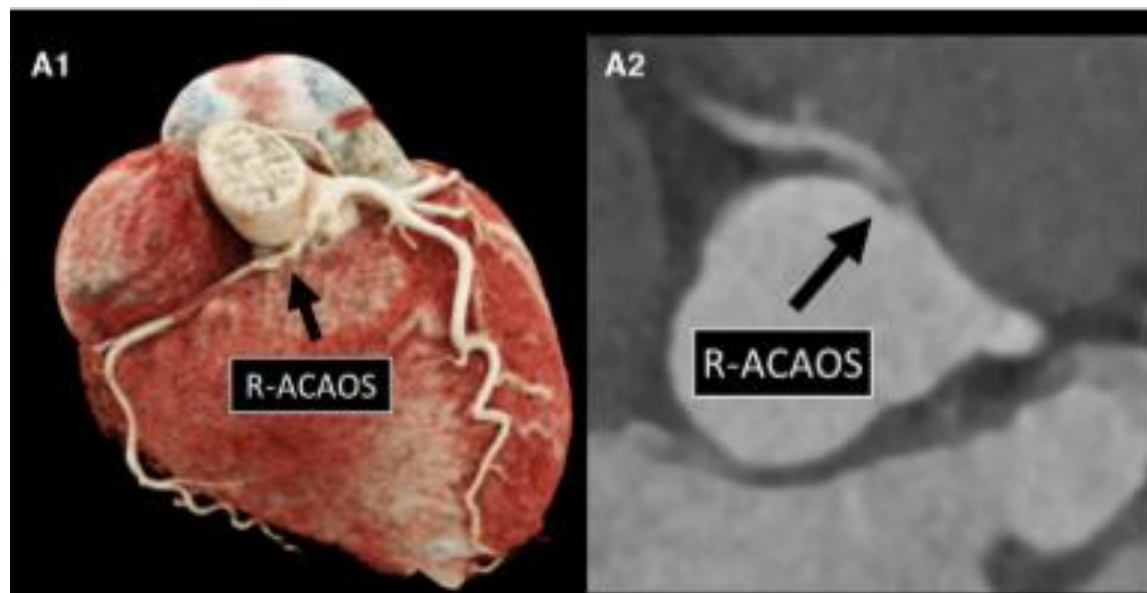
Published: 15 March 2023 **Article history** ▼

Unroofing



Unroofing

Vorher

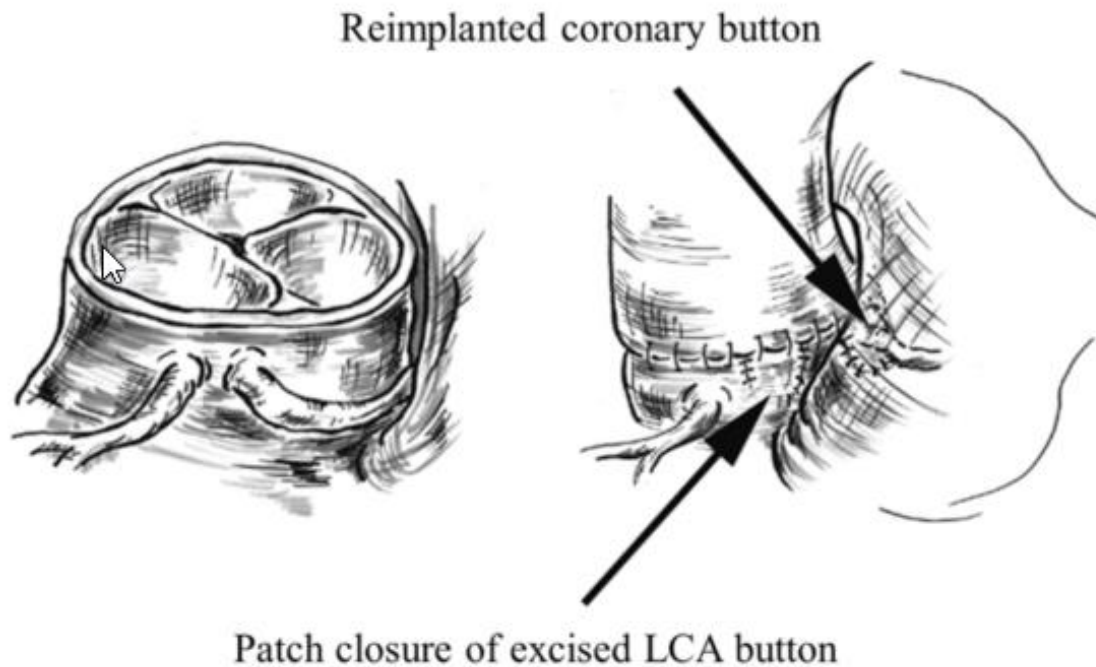


Nachher



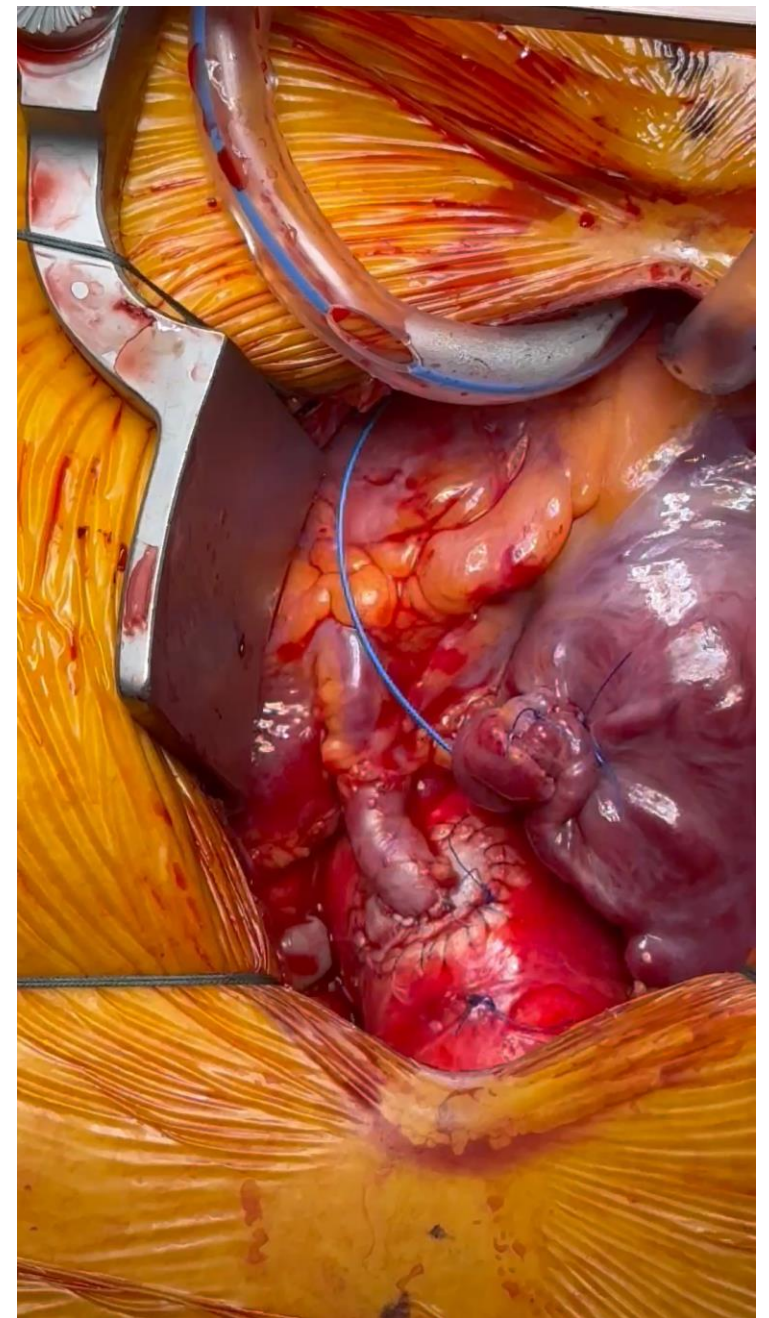
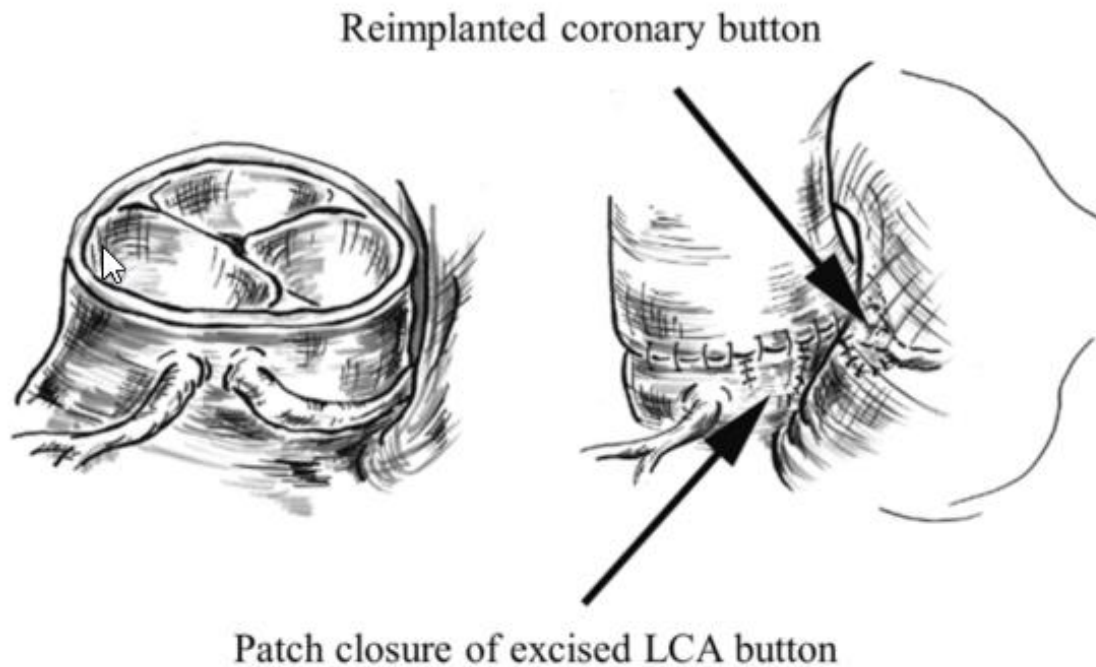
Reimplantation

B Coronary translocation / reimplantation



Reimplantation

B Coronary translocation / reimplantation



Bigler & Siepe & Gräni C et al. J Am Heart Assoc 2022

Safety and efficacy of surgical correction of anomalous aortic origin of coronary arteries: experiences from 2 tertiary cardiac centres

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Symptom evolution after correction for AAOCA

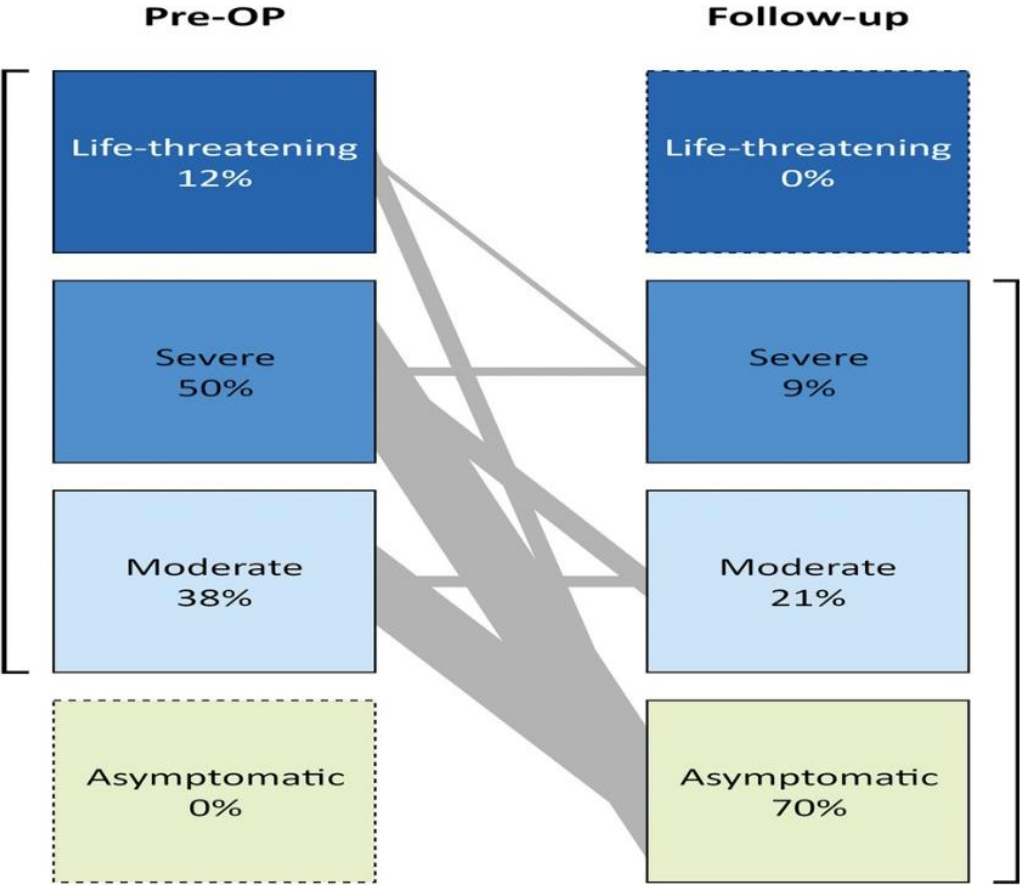


Table 4:

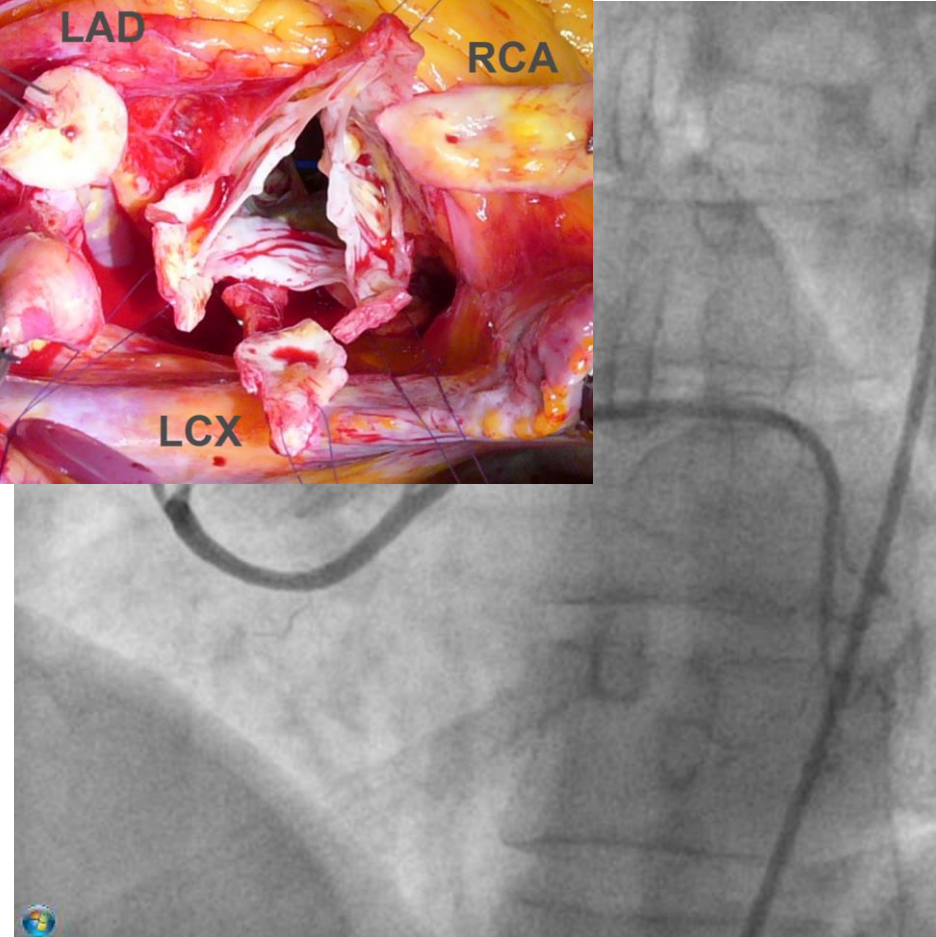
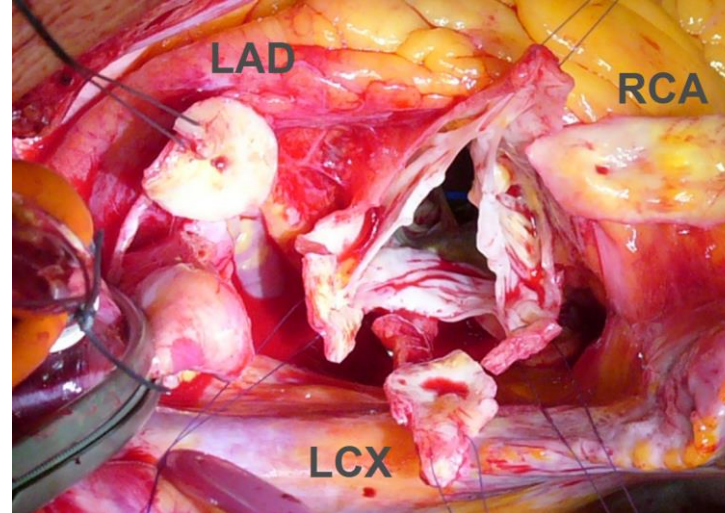
n=71

Postoperative complications

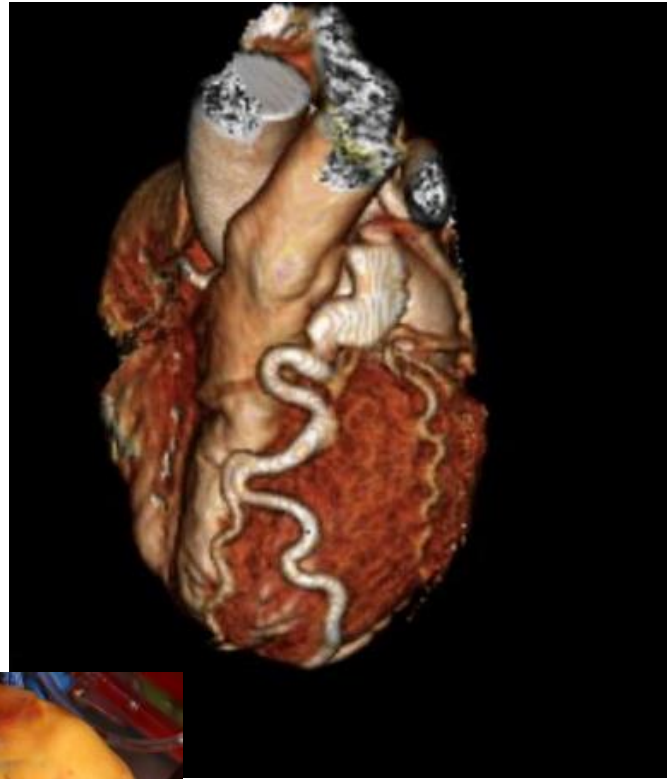
	Early (<30 days)	Late (>30 days)
Cardiovascular death	0	0
Death other than cardiovascular	0	3
Unknown cause of death	0	2
Myocardial infarction	3	0
Stroke	1	0
Reoperation		
Coronary artery bypass grafting	3	0
Pericardial drain due to tamponade	1	0
Sternal wound infection	0	2
Pacemaker implantation	1	0
Cardiovascular mechanical support		
ECMO	4	0
IABP	2	0

ECMO: extracorporeal membrane oxygenation; IABP: intra-aortic balloon pump.

Beispiel für Notwendigkeit dergleichzeitigen Mit-Behandlung: LCx from RC-Sinus beim Wurzelaneurysma



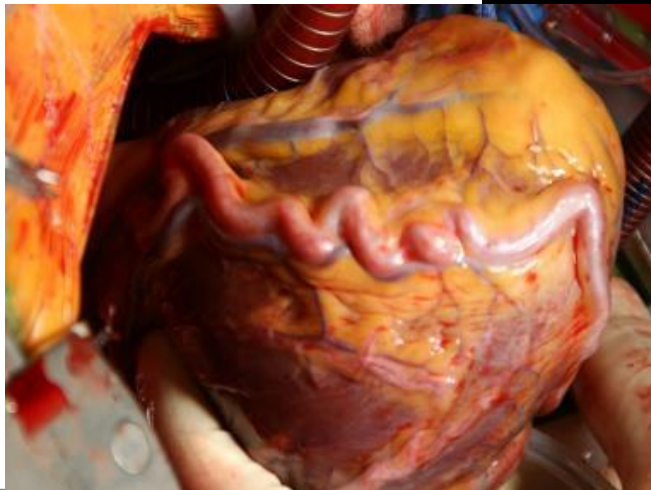
Koronarfisteln



Häufig Zufallsdiagnose

Selten relevant mit Symptomen

Interventionell und
chirurgisch leicht behandelbar



Vielen Dank für die Aufmerksamkeit

INSELGRUPPE

European Coronary Artery Anomaly Conference

Friday, January 16th, 2026

Topics:

- AAOCA – From Anatomy to Disease
- The Pathologist's View of AAOCA
- Surgical Strategies in AAOCA
- From Imaging to Artificial Twin in AAOCA
- The view of the patient

Venue:

Neues Anna Seiler Haus Inselspital Bern,
Freiburgstrasse 20, 3010 Bern, Switzerland

Organizing Committee:

Prof. Dr. Dr. med. Christoph Gräni

Prof. Dr. med. Matthias Siepe

www.kardiologie.insel.ch

Registration via QR-code:

