Anomalous Aortic Origin of Coronary Arteries – Surgical Management

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BACKGROUND

- Anomalous aortic origin of a coronary artery with inter-arterial, intra-conal or intra-mural course (AAOCA) is a rare congenital anomaly:
  - Left main coronary arises from the right sinus (ALCA) OR
  - Right coronary artery arises from the left sinus (ARCA)
- Prevalence ranges from 0.1%-0.3%.
- ARCA ~ 6 times more prevalent than ALCA
- ALCA has a higher risk of sudden cardiac death

Taylor AJ et al. J Am Coll Cardiol 1992;20:640-7; Basso C et al. J Am Coll Cardiol 2000;35:1493-501; Maron BJ et al. JAMA 1996;276:199-204; Brothers JA et al., J Am Coll Cardiol 2007;50:2078-82

RISK OF SUDDEN DEATH

- In the United States, AAOCA is the 2nd leading cause of sudden cardiac death in children after hypertrophic cardiomyopathy.
- Sudden death most commonly occurs during or just after exercise, notably among otherwise healthy, young athletes.

Maron et al., Circulation 1980;62:218-9; Corrado et al., Am J Med 1990;89-588-96; Frescura C et al., Hum Pathol 1998;29:689-95.

DIAGNOSIS

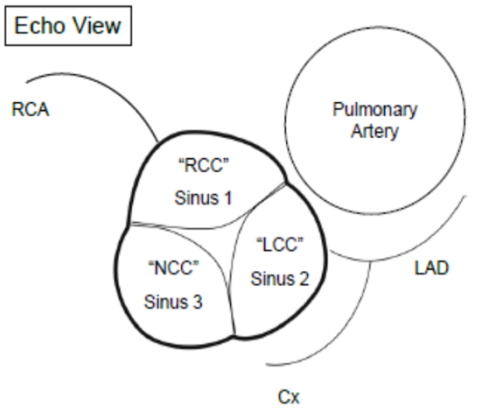
- Most patients are initially diagnosed by transthoracic echocardiography.
- If the diagnosis is unclear further diagnostic evaluation is warranted:
  - CT
  - MRI
  - Cardiac catheterization with coronary angiography
- Further testing to evaluate for ischemia:
  - Exercise stress test, stress echocardiogram, stress myocardial perfusion scan

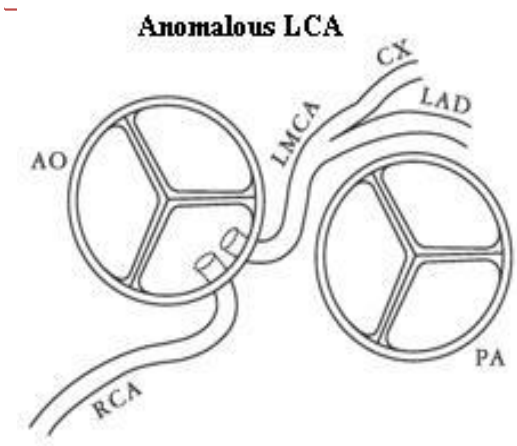
PRESENTING SYMPTOMS

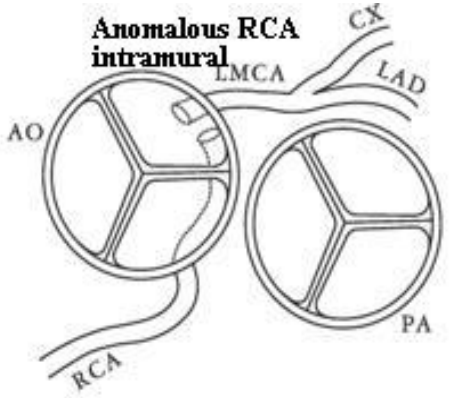
- The challenge is diagnosing AAOCA as patients often are asymptomatic
- Cardiovascular presenting symptoms, often occurring during or just after exertion, include:
  - Chest pain
  - Dizziness
  - Syncope
  - Ventricular arrhythmia
  - Myocardial infarction, aborted sudden death, or sudden death

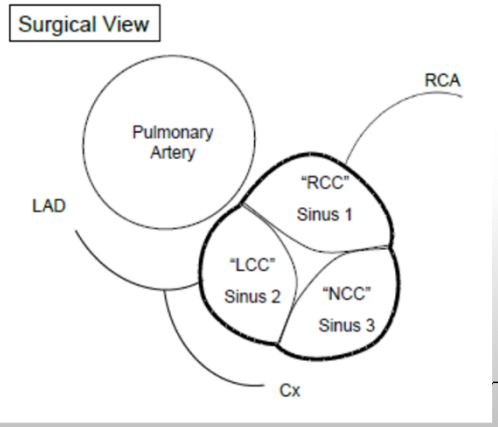
Romp RL et al., Ann Thorac Surg 2003;76:589-96; Erez E et al., Ann Thorac Surg 2006;82:973-7
Brothers JA et al., J Am Coll Cardiol 2007;50:2078-82

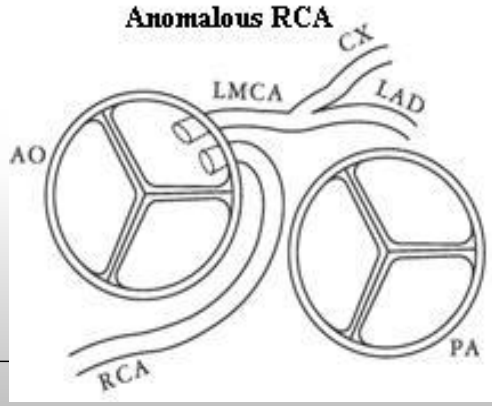
ANOMALOUS LCA AND RCA

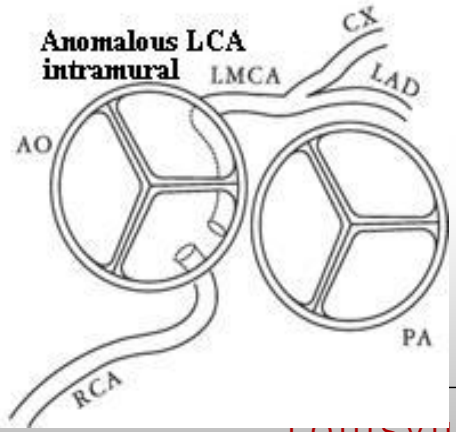




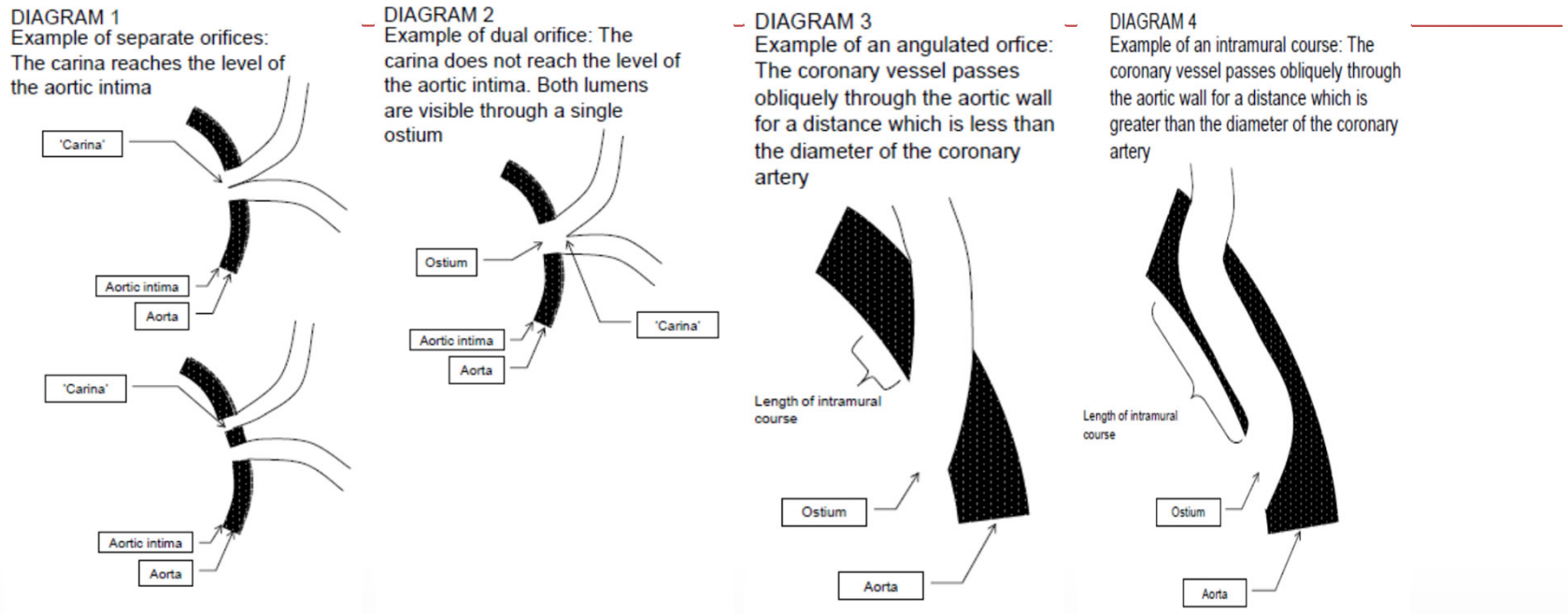








ANATOMICAL VARIANTS



- Other features: sinus of origin, slit-like takeoff, high ostial origin, interarterial course, intraconal/intraseptal/intramyocardial course

MORPHOLOGY AT SURGERY

Anomalous vessel	n	%
Left main	31	27%
Right	78	69%
LAD	2	2%
Both	2	2%

- CHSS AAOCA Registry

VESSEL COURSE

89% - Inter-arterial and Intra-mural

9% - Inter-arterial but not Intra-mural

2% - Neither Inter-arterial nor Intra-mural

PROXIMAL ANATOMY

Type	%
At or above level of commissure	88%
Below level of commissure	5%
Not stated	7%
Slit-like orifice	27%
Stenotic orifice	48%

SURGICAL TECHNIQUES

Host of surgical options

Depends on

- Intra-mural course
- Proximal anatomy
- Associated CAD

CORONARY UNROOFING

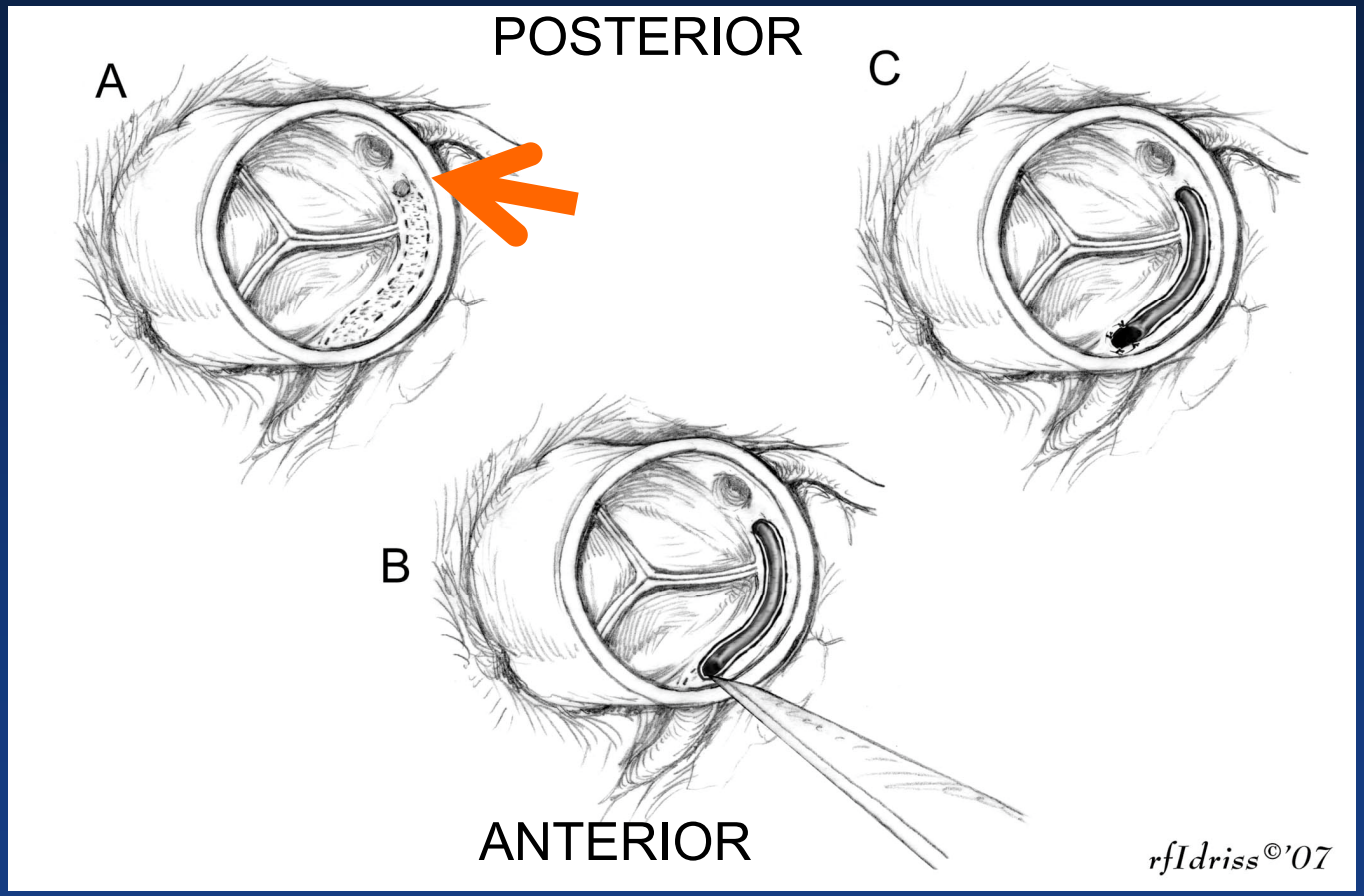
Most common procedure

Incision of common wall with tacking sutures

OR

Exteriorization with tacking sutures

Unroofing of an Intramural Coronary Artery



COMMISSURE TAKEDOWN

~ 33% patients will need commissural takedown

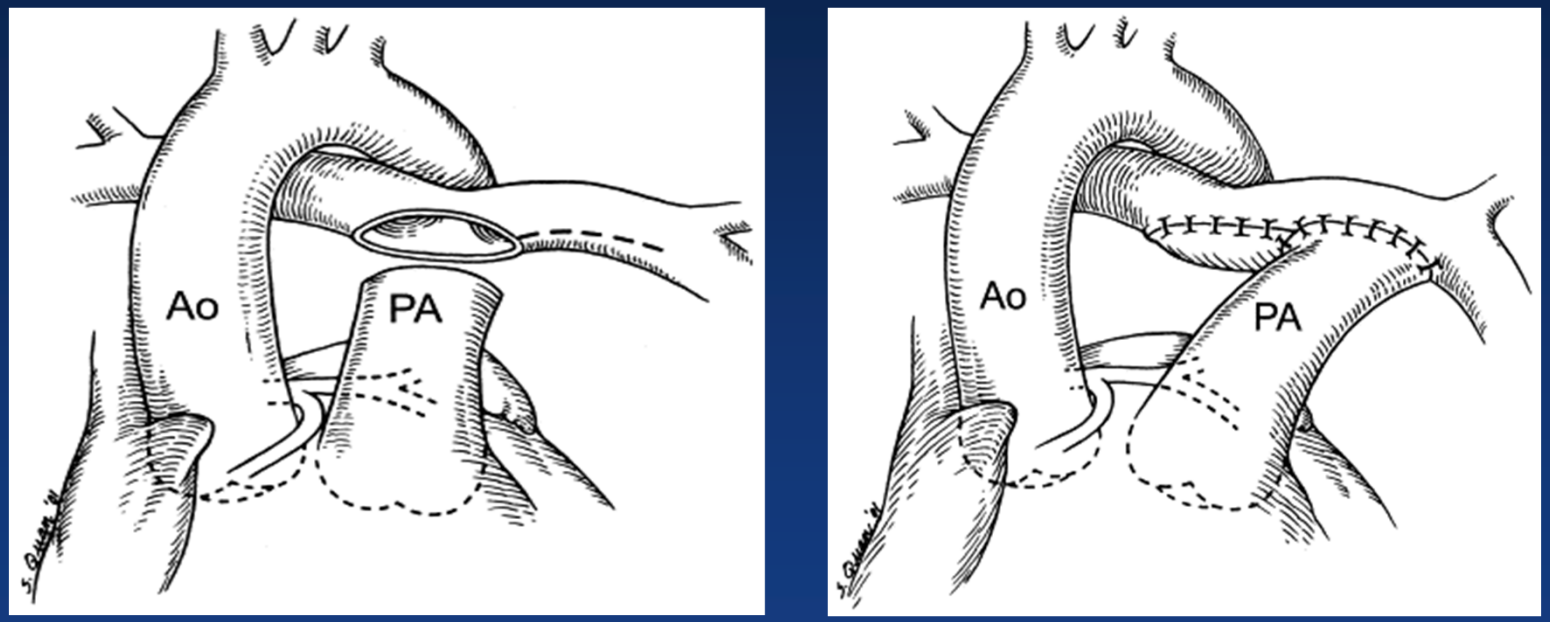
~ 90% of these patients will do well with simple resuspension of the commissures

~ 10% of these patients will need an aortic valve replacement

OTHER TECHNIQUES

- Coronary artery reimplantation – reserved for Inter-arterial but not Intra-mural course
- Pulmonary artery relocation – Inter-arterial but not intra-mural course. Accompanied with unroofing for Inter-arterial and Intra-mural course

Pulmonary Artery Relocation



LESS COMMON TECHNIQUES

- Simple osteoplasty, no unroofing (3%)
- Coronary artery bypass grafting (2%)
- Ostial window (1%)
- Side-to-side anastomosis from outside the aorta without unroofing (1%)

OPERATIVE OUTCOMES

Post-operative ischemia – 2%

Coronary artery dissection – 1%

Operative mortality – 2%

OUR OUTCOMES

2020 – 2022

Total AAOCA surgeries performed – 7

Mean Age – 52 +/- 10 years

Male Sex – 5 (72%)

Unroofing procedure – 7 (100%)

Isolated unroofing procedure – 3 (43%)

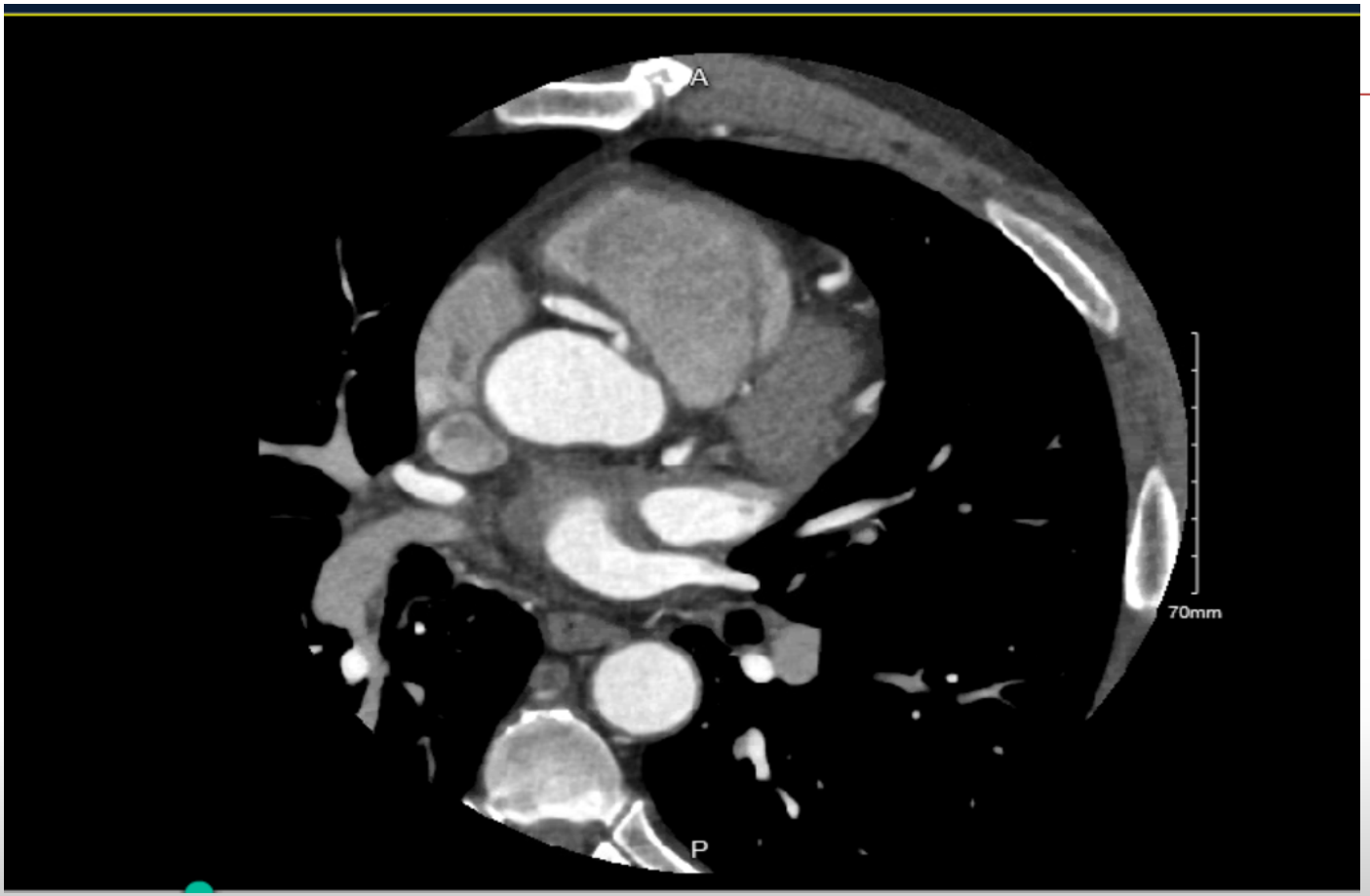
Associated procedures – CABG (n=2, 29%), AVR (n=1, 14%), MVR (n=1, 14%)

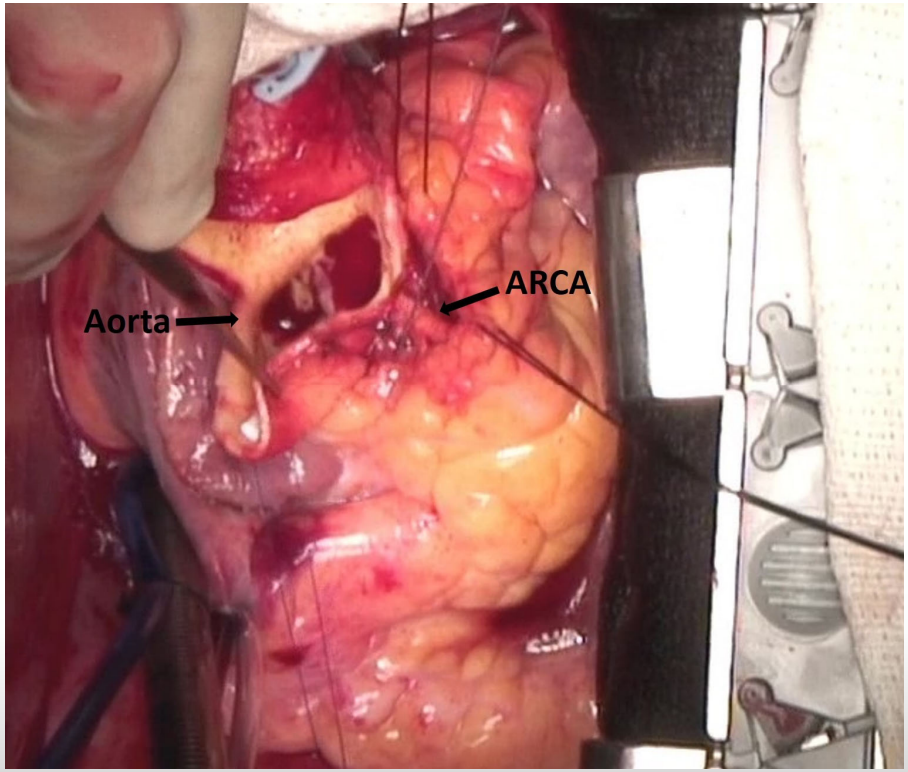
Operative mortality – 0

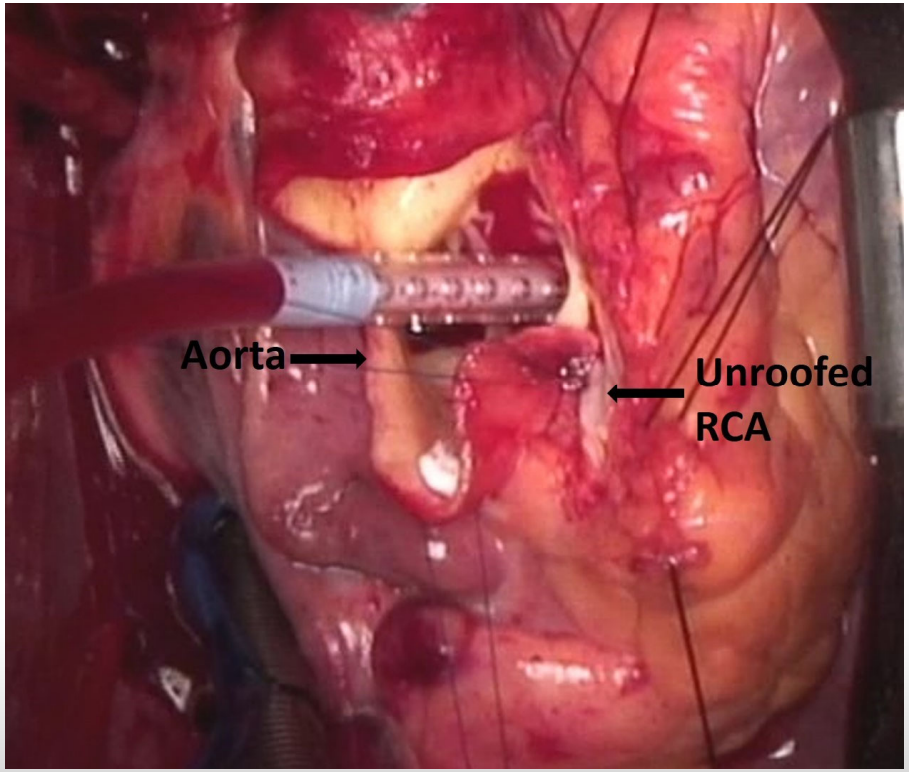
Post-operative complications: Stroke – 0, Renal failure – 0, PPM implantation – 1 (14%)

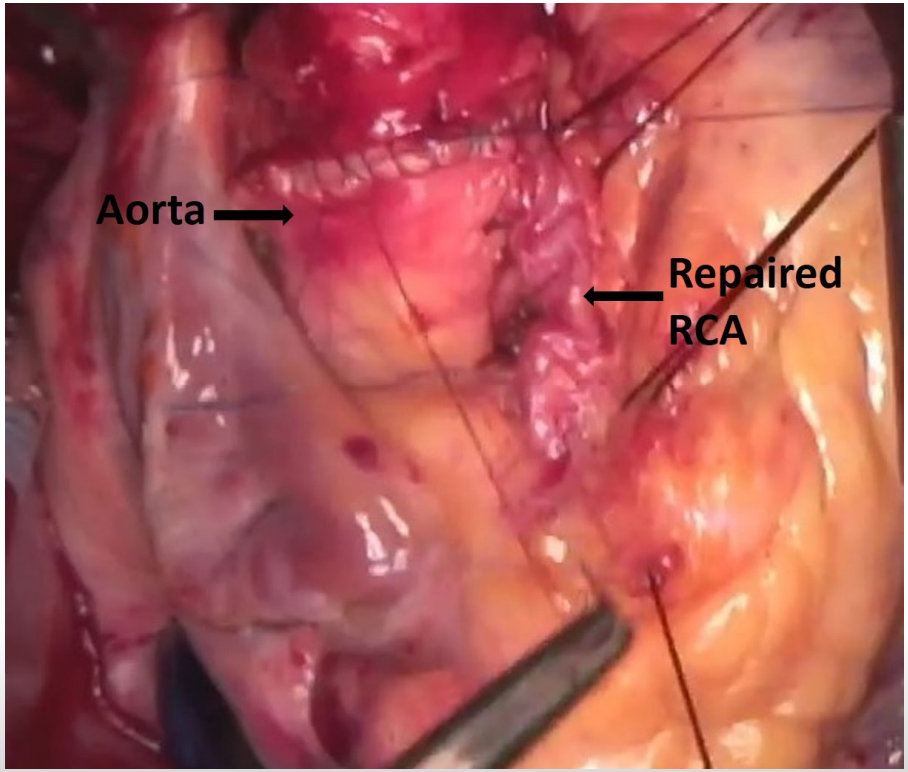
CASE

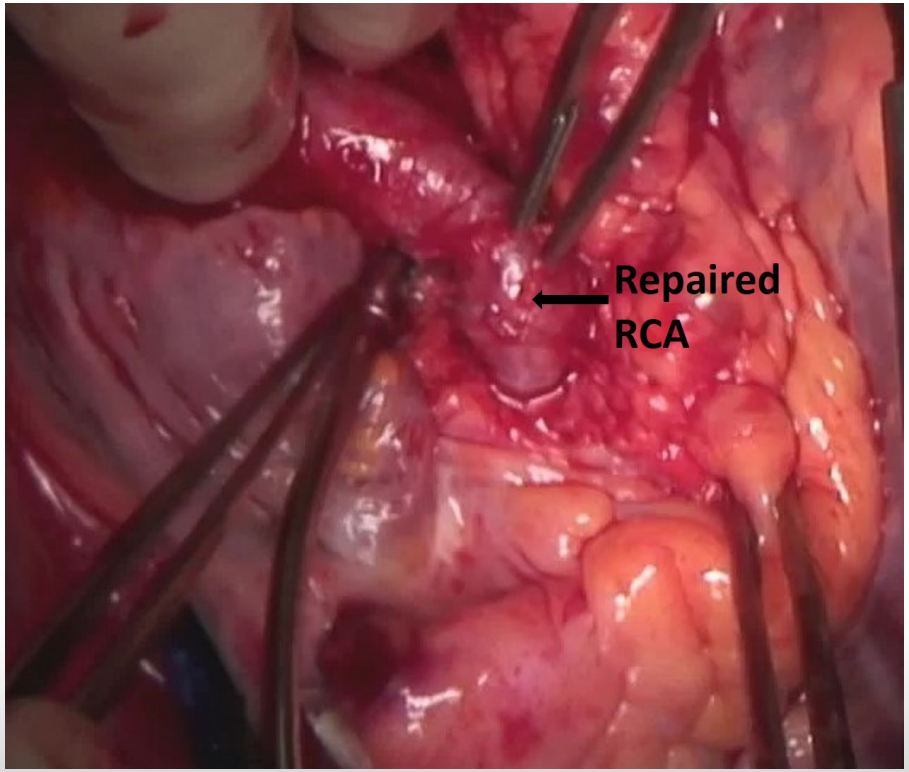
- 53/M, visiting Louisville on Holter monitor
- 348 runs of Vtach
- Asked to come to Heart Hospital
- Anomalous RCA from LCC at LCC/RCC commissure
- Acute angulation of RCA


A
70mm
P


Aorta
ARCA


Aorta
Unroofed
RCA


Aorta
Repaired
RCA


Repaired
RCA

TAKE HOME MESSAGE

Unroofing of an intramural segment is by far the most common procedure…

but unique morphological variants sometimes require more creative repair strategies

QUESTIONS AND DISCUSSION