

ANOMALOUS AORTIC ORIGIN OF CORONARY ARTERY

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ANOMALOUS

1. Inconsistent with or deviating from what is usual, normal or expected.
2. Of uncertain nature or classification.
3. Synonyms: irregular, unusual, equivocal or paradoxical

Webster's Collegiate Dictionary

OGDEN JA 1970

BOX 38-1 Congenital variations of the coronary artery

MINOR CORONARY VARIATIONS

- High takeoff
- Multiple ostia
- Anomalous circumflex artery origin
- Anomalous anterior descending artery origin
- Absent proximal ostium/single ostium in other aortic sinus
- Hypoplastic proximal coronary artery
- Congenital proximal coronary artery
- Congenital distal stenosis
- Coronary artery from the posterior aortic sinus
- Ventricular origin of an accessory coronary artery

MAJOR CORONARY ANOMALIES

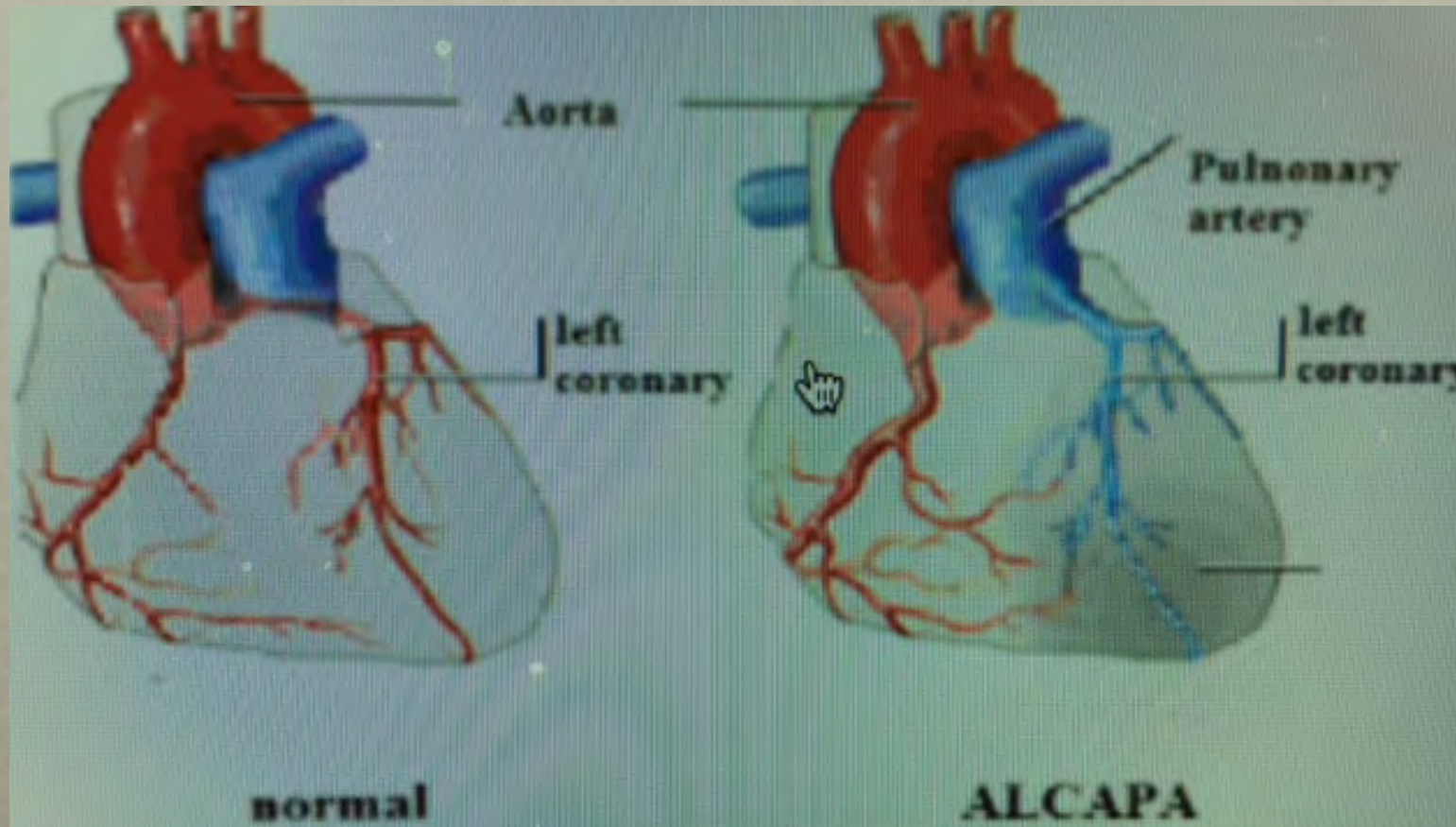
- Coronary "arteriovenous" fistula
- Anomalous origin from the pulmonary artery
 - Left coronary artery
 - Right coronary artery
 - Both coronary arteries

SECONDARY CORONARY ANOMALIES

- Secondary coronary "arteriovenous" fistula
- Variations in transposition of the great vessels
- Variations in truncus arteriosus
- Variations in tetralogy of Fallot
- Ectasia of coronary arteries in supravulvar aortic stenosis
- Mural coronary artery

From Ogden JA: Congenital anomalies of the coronary arteries, *Am J Cardiol* 25:474, 1970.

ALCAPA



AAOCA

AAOCA

is the acronym for

Anomalous Aortic Origin of a
Coronary Artery



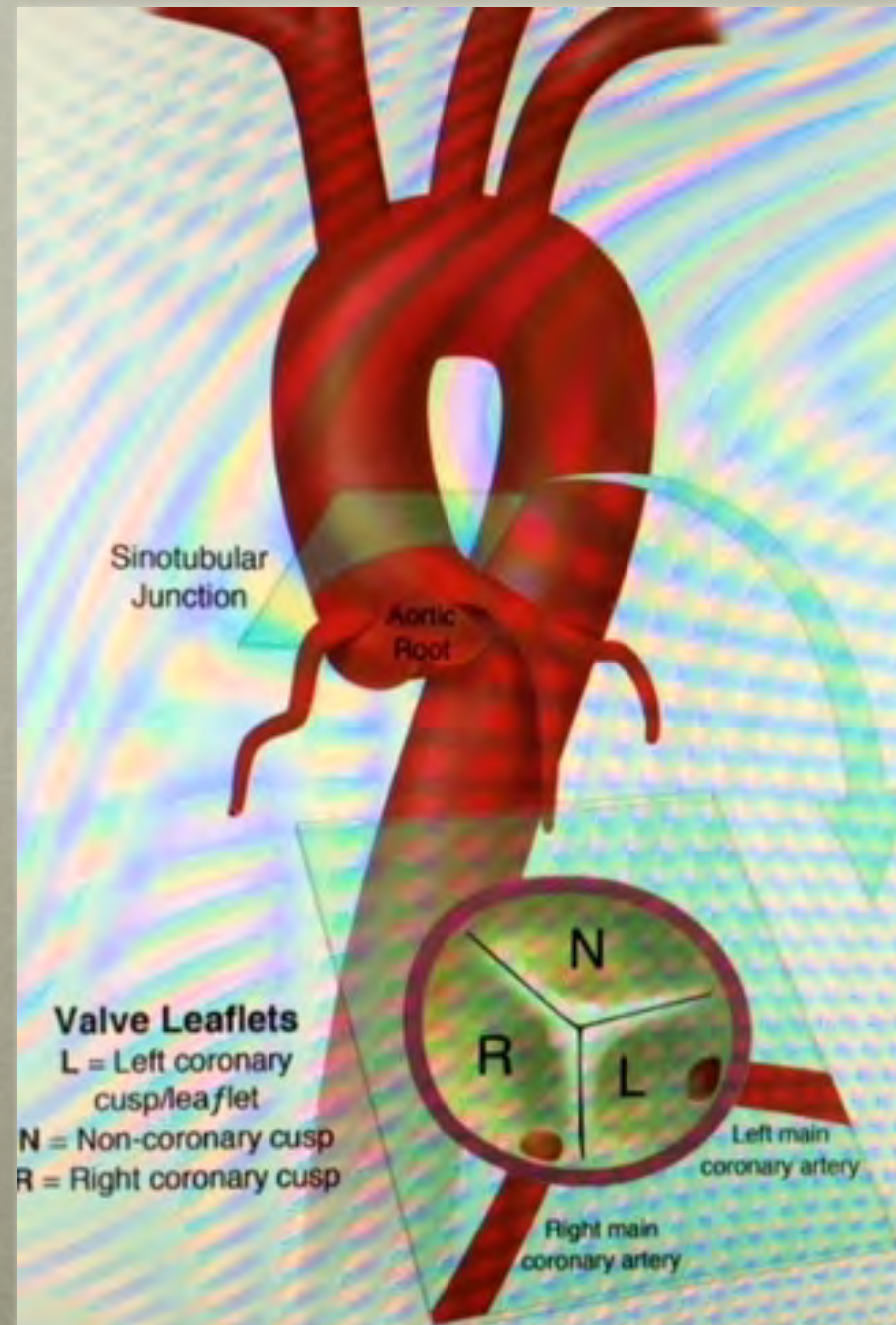
by allacronyms.com



AORTIC ROOT ANATOMY

3 sinuses

Each named for respective
coronary artery



AAOCA

TABLE 38-1

Classification of Anomalous Aortic Origins of Coronary Arteries (AAOC)

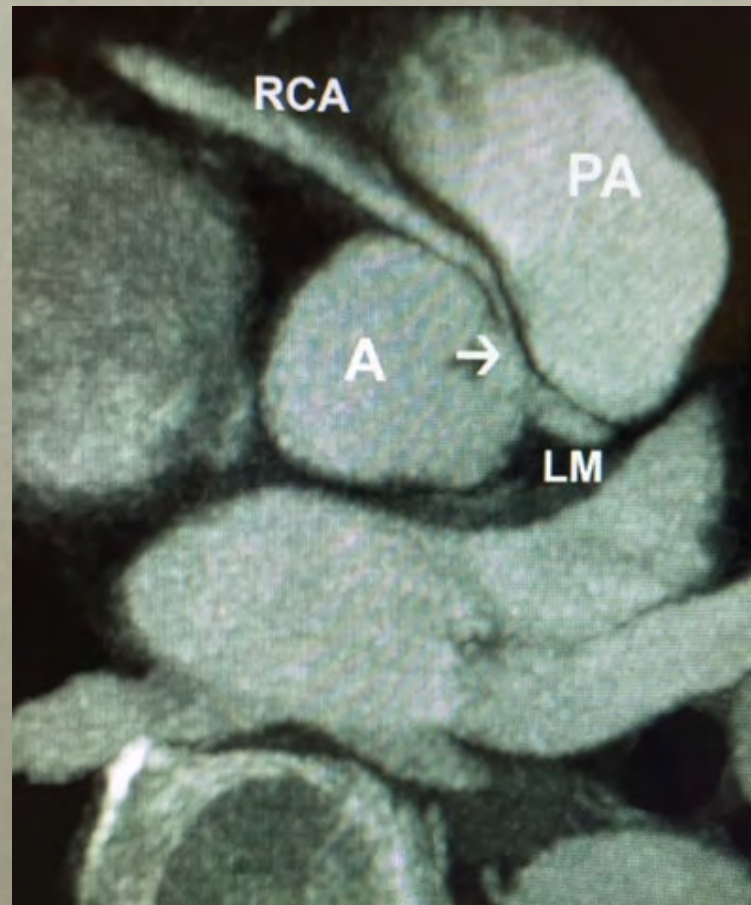
Type	Incidence
LMCA from RASV	21% ³⁹ of AAOC: Four different courses possible in relation to
RCA from LASV	6% to 27% ^{39,43} of AAOC: Can cross the aortic root anteriorly, pulmonary artery
CX from RASV or RCA	10% to 60% ⁵⁰ of AAOC (most common): Courses either ant vessels, not between, ⁵⁰ always retroaortic ⁵¹
Inverted coronary arteries	Rare

From Taylor AJ, Rogan KM, Virmani R: *J Am Coll Cardiol* 20:640, 1992.

CX, Circumflex coronary artery; LASV, left aortic sinus of Valsalva; LMCA, left main coronary artery; RASV, right aortic sinus of Valsalva; RCA, right coronary artery.

AAOCA

- Very contemporary
- Still being defined
- Controversies exist
- Different referral centers treat differently
- Catheter based angiogram use to be the gold standard
- CT angiogram may be now
- Some say Echo



AAOCA

Normal Origins



Abnormal Origin
RCA



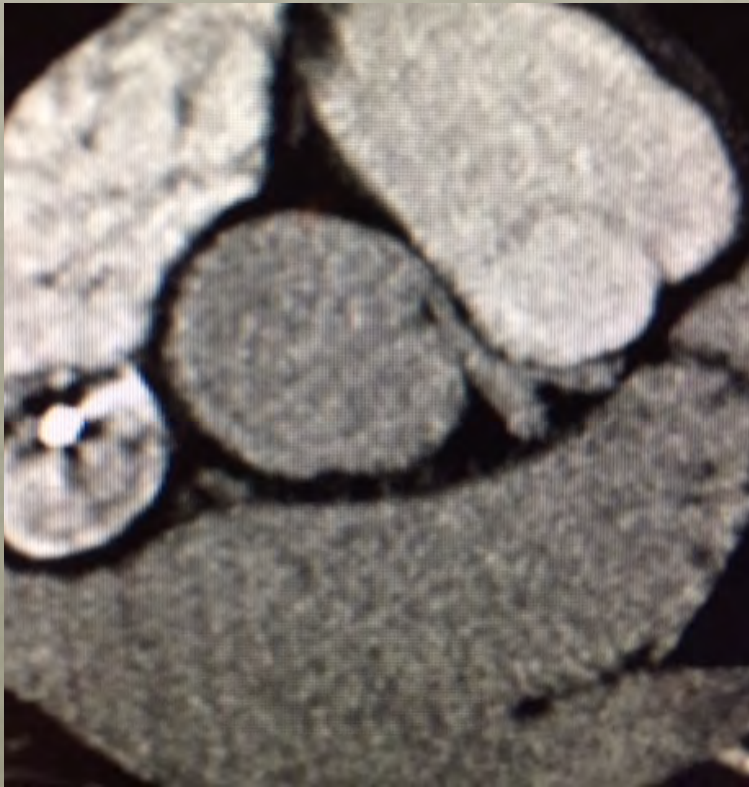
AAOCA

- Left from right sinus
- Right from left sinus
- Rarely from non sinus



AAOCA

LAAOCA



RAAOCA



AAOCA FACTS

- Some types associated with sudden cardiac death(SCD)
- In the U.S., AAOCA is the second leading cause of SCD in children, after hypertrophic cardiomyopathy.
- SCD most commonly occurs during or just after exercise, notably among otherwise healthy young athletes

Maron et al, Circulation 1980;62: 218-219

Corrado et al, Am J Med 1990; 89: 588-96

Frescura C et al, Hum Pathol 1998; 29:689-95

AAOCA FACTS

Approx. 500,000 people USA living with AAOCA

AAORCA (1.3%) 6x>prevalence than AAOLCA(0.047%)

Cath Cardiovasc Diagn 1990;21: 28-40

SCD children 0.8-6.2/100,000 per year

Coronary anomaly account for 13%

Circ 2009; 119: 1085-92

Ralph S Mosca, M.D.

AAOCA FACTS

No patient with SCD from AAOCA <10 yrs or >30 yrs*

*2 patients(1-9 yrs) SCD while being treated medically

Incidence of SCD with AAORCA rare approx 15 cases from 1975-2015

Ped Cardiol 2009;30: 911-921

Screening of active military recruits (n=6,300,000)

6,000-12,000 affected

21 deaths due to AAOCA-all AAOLCA(risk 0.17-0.35%)

Ann Int Med 2004; 141: 829-34

Ralph Mosca M.D.

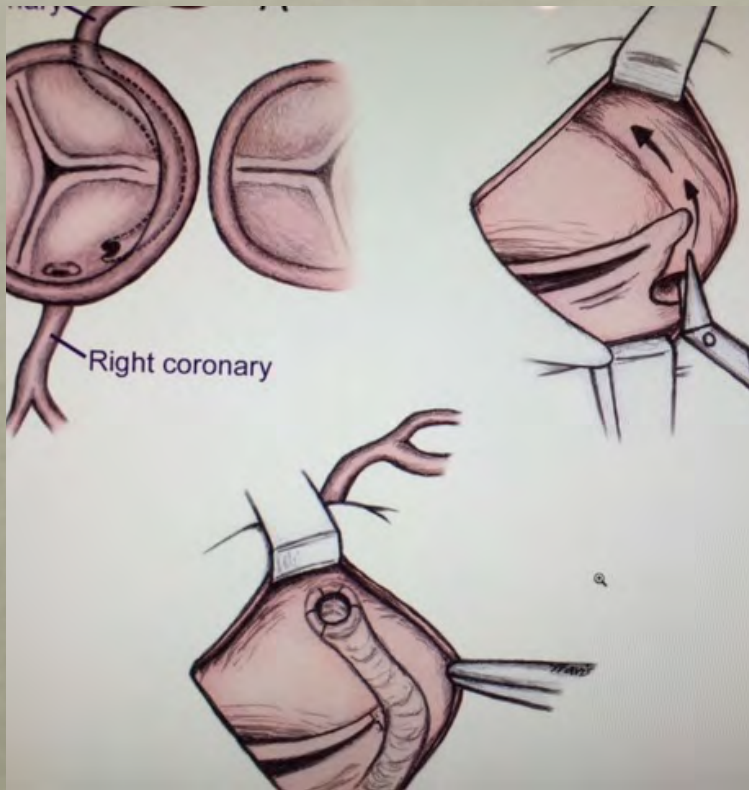
AAOCA FACTS

Autopsy Study Data

Author	N	AAORCA	AAOLCA	SCD- Right	SCD- Left
Chietlin- 1974	51	18	33	0/18 (0%)	9/33 (27%)
Taylor- 1992	30	21	9	4/21 (19%)	8/9 (89%)
Kragel- 1988	32	25	7	8/25 (32%)	5/7 (71%)
Taylor- 1997	101	52	49	13/52 (25%)	28/49 (57%)
Frescura- 1998	11	7	4	4/7 (57%)	4/4 (100%)
TOTALS	225	123	102	29/123 (24%)	54/102 (53%)

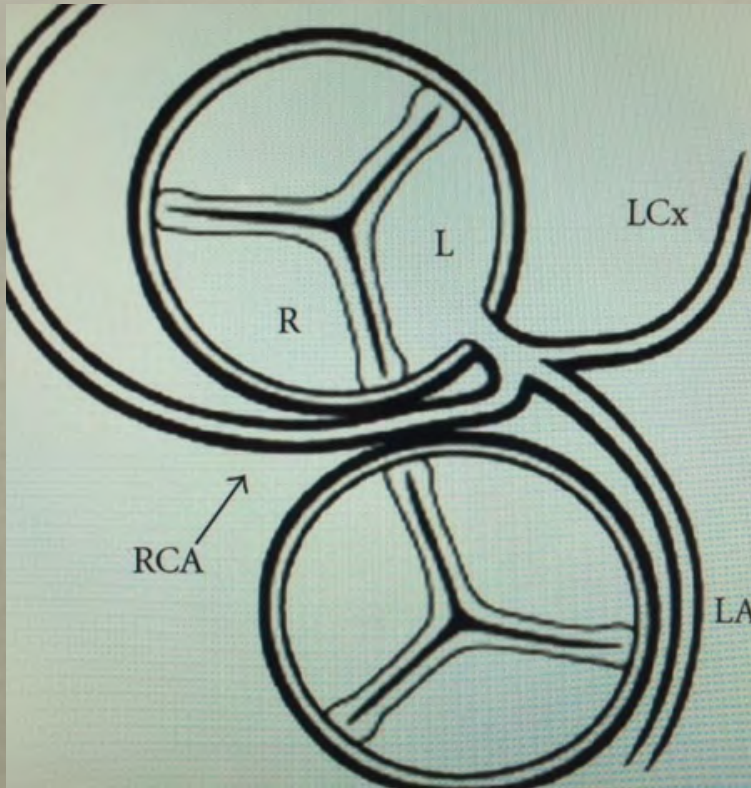
SURGICAL TREATMENT

Intramural

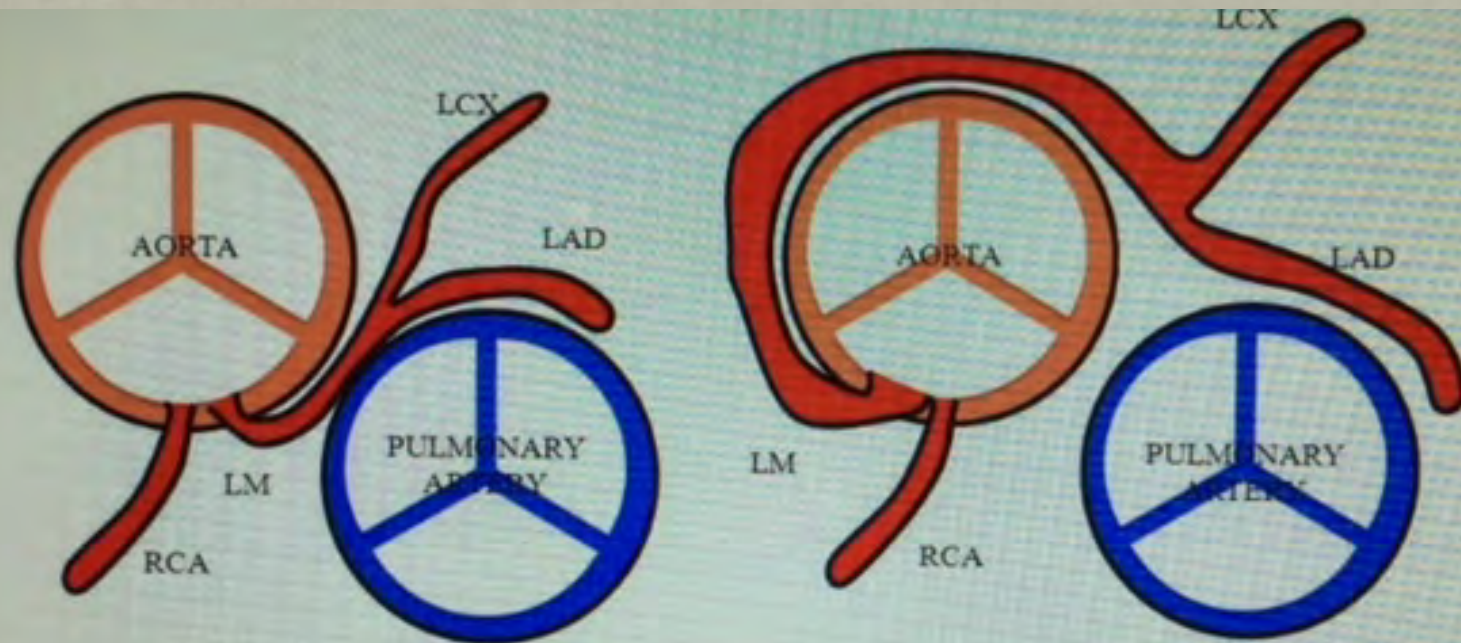


SURGICAL TREATMENT

Interarterial



NOT SURGICAL VS SURGICAL



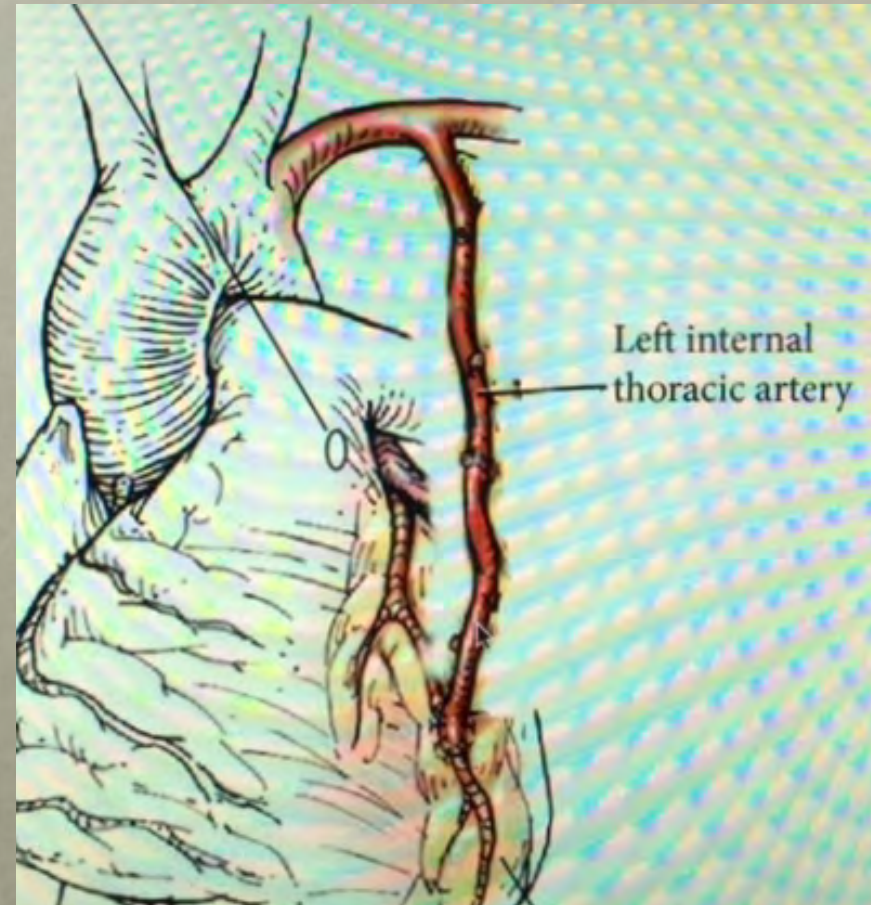
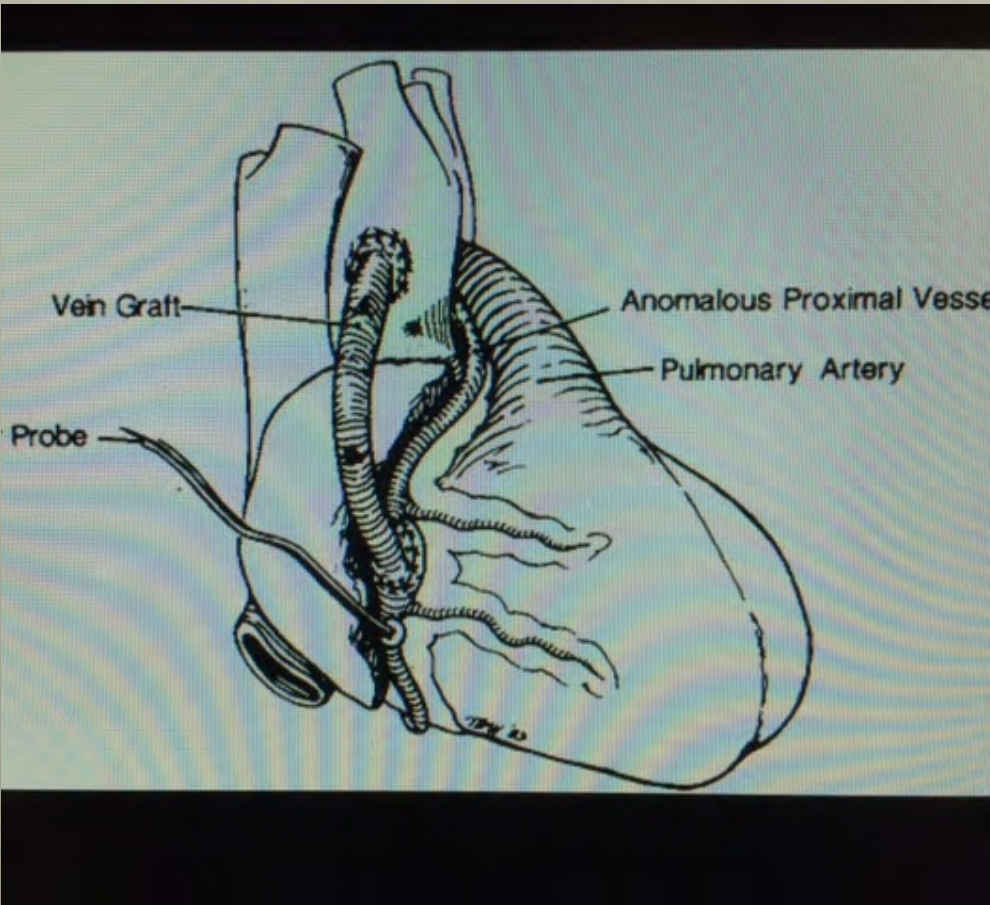
Anomalous LCA from right
sinus - Inter-arterial Course

Increased risk of sudden death

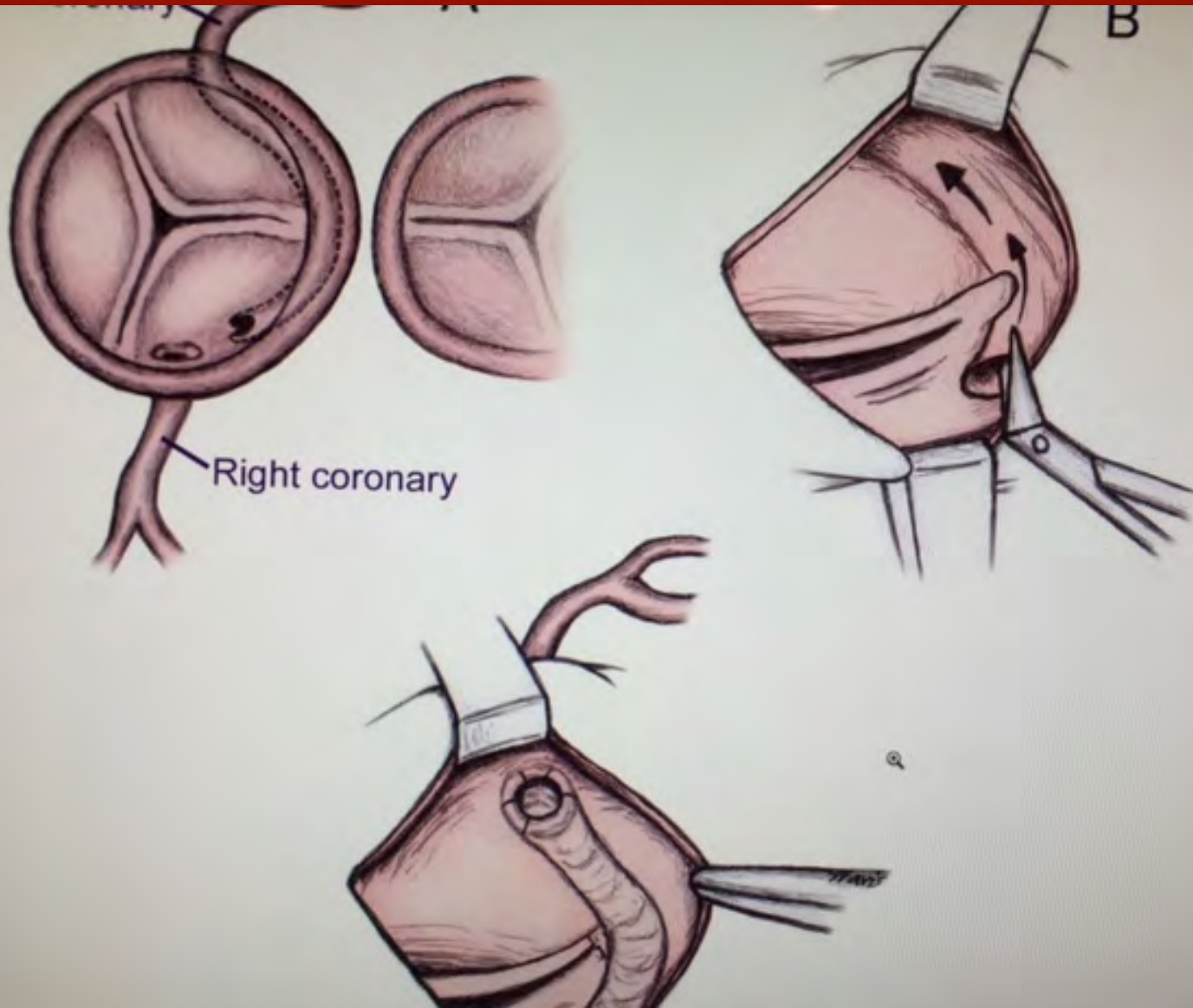
Anomalous LCA from right
sinus - Retro-aortic course

Prognosis benign

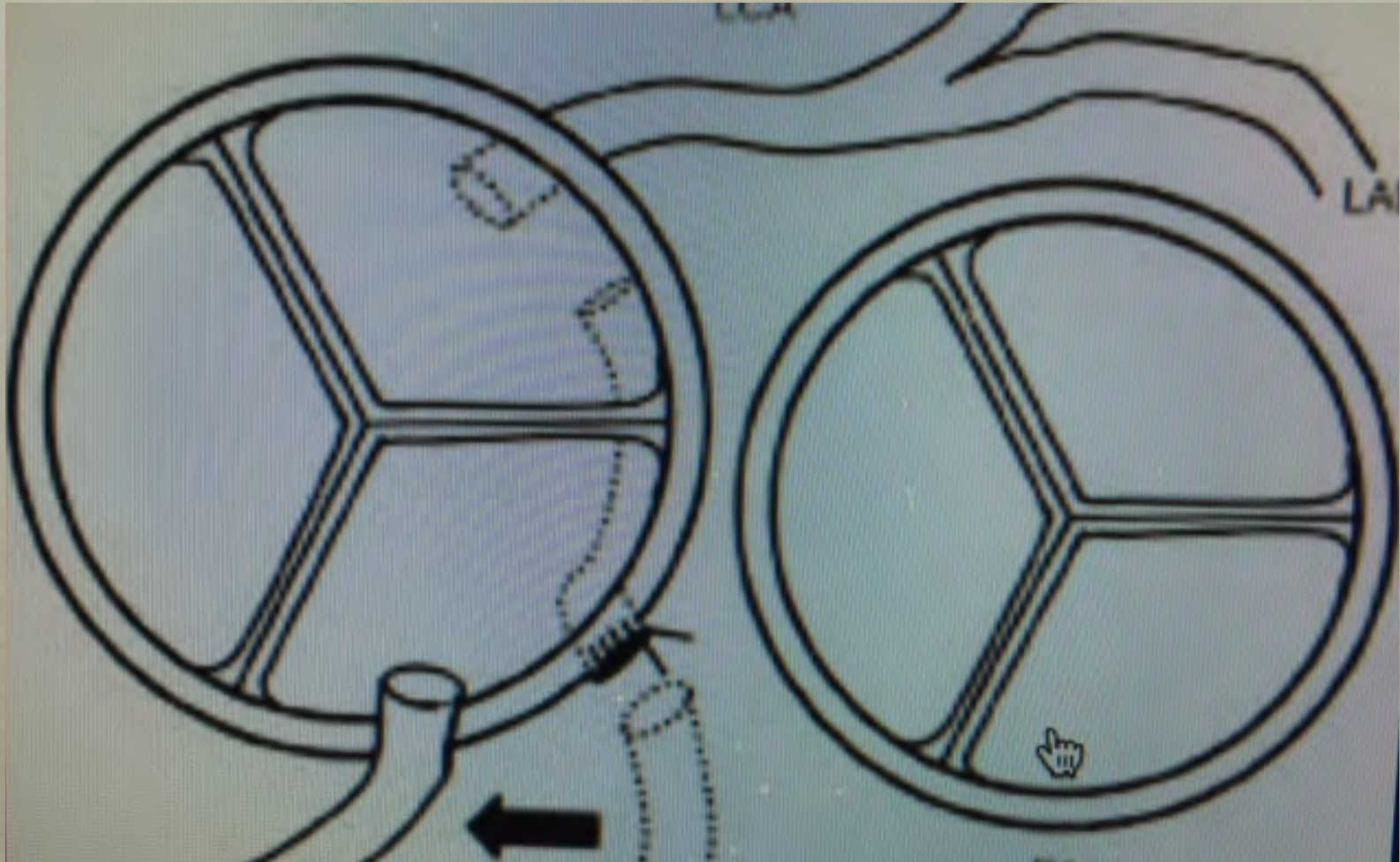
SURGICAL APPROACHES



UNROOFING



TRANSLOCATION



FROMMELT ET AL

Ten-year experience with surgical unroofing of anomalous aortic origin of a coronary artery from the opposite sinus with an interarterial course

Peter C. Frommelt, MD, FACC,^a David C. Sheridan, MD,^a Stuart Berger, MD,^a Michele A. Frommelt, MD, FACC,^a and James S. Tweddell, MD^b

Background: Anomalous aortic origin of a coronary artery from the opposite sinus with an intramural course between the great arteries (AAOCA) is associated with ischemia and sudden cardiac death in children, and surgical unroofing has been used to alleviate that risk.

Methods: The cardiology database was reviewed to identify all patients with AAOCA who underwent surgical unroofing 10 years.

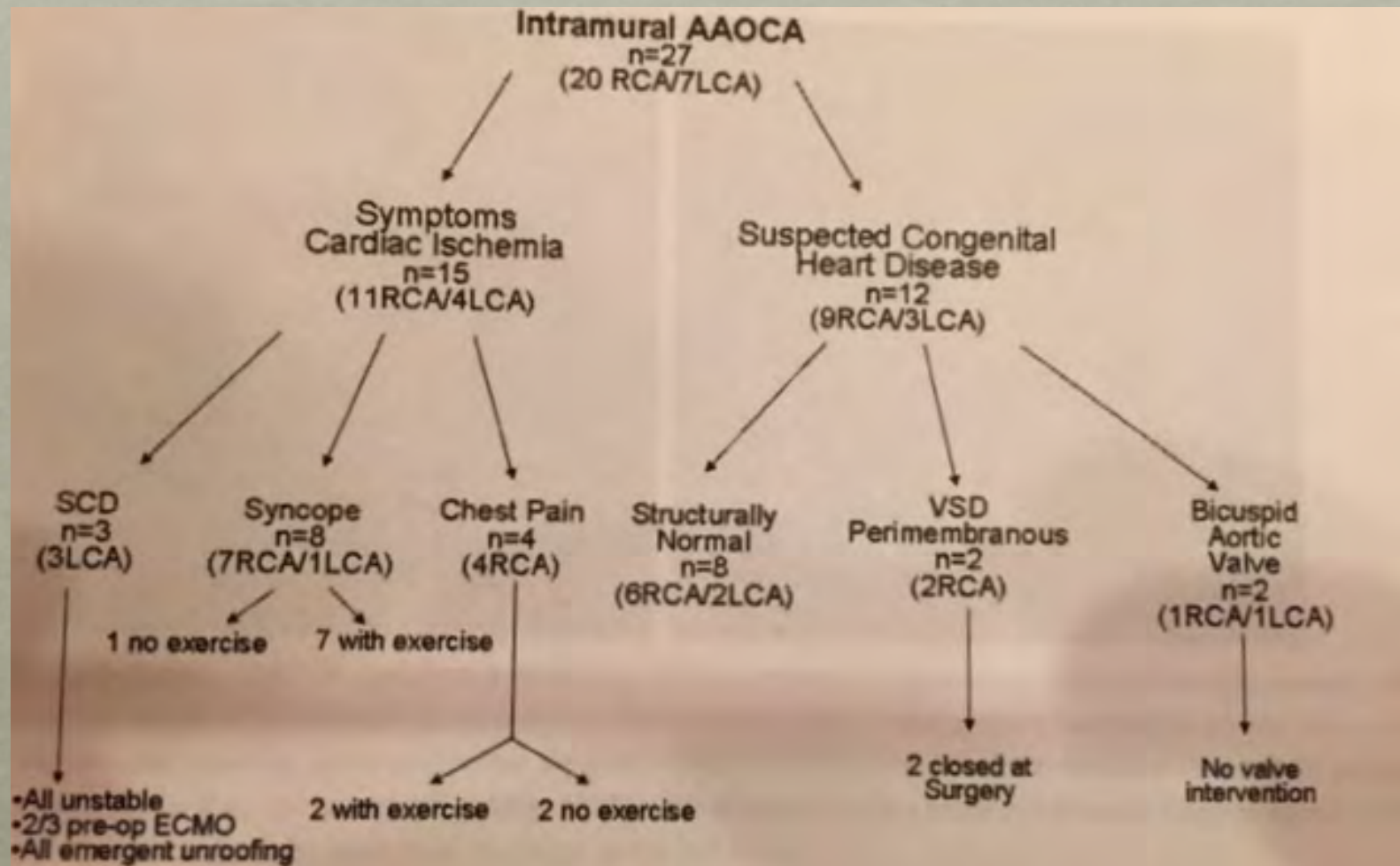
Results: From March 1999 to September 2009, 27 patients with a mean age of 12.6 ± 3.5 years (range, 4–22 years) had surgical unroofing of AAOCA of the left coronary from the right sinus (left AAOCA, 7/27, 26%) and of the right coronary from the left sinus (right AAOCA, 20/27, 74%). Of these, 26 had diagnoses made by transthoracic echocardiography. Symptoms included resuscitated sudden cardiac death in 3, syncope in 8, and chest pain in 4. No symptoms were present in 12 cases of serendipitous diagnosis. Unroofing of the intramural portion was successfully performed in all cases. A slitlike coronary orifice was described at surgical inspection in 12 patients, 7 of whom had right AAOCA and no symptoms. All patients after unroofing have patent coronary flow by Doppler and normal echocardiography and exercise treadmill testing at mean follow-up of 1.8 years. None have activity restrictions.

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3	11	M	RCA	None	TTE		VSD	Yes	Unroofing; AoV resusp; VSD closure
4	16	M	LCA	SCD with exercise	TTE	TEE	None	No	Unroofing; AoV resusp
5	20	M	LCA	SCD with exercise	Cath		None	No	Unroofing; AoV resusp
6	4	F	RCA	None	TTE	Cath	VSD	No	Unroofing; AoV resusp; VSD closure
7	10	M	RCA	Nonexercise CP	TTE	Cath	None	Yes	Unroofing
8	14	M	RCA	CP with exercise	TTE	Cath	None	Yes	Unroofing
9	13	M	RCA	None (benign PVCs)	TTE		None	No	Unroofing
10	7	M	RCA	None (heart murmur)	TTE		None	Yes	Unroofing
11	16	M	RCA	CP with exercise	TTE		None	Yes	Unroofing
12	13	F	RCA	Nonexercise syncope	TTE		None	Yes	Unroofing
13	9	M	LCA	None (heart murmur)	TTE		None	No	Unroofing
14	13	F	RCA	Syncope/CP with exercise	TTE	MRI	None	Yes	Unroofing
15	15	F	RCA	None (heart murmur)	TTE		None	No	Unroofing
16	16	M	RCA	Syncope with exercise	TTE		None	No	Unroofing
17	13	F	RCA	Sleep apnea	TTE		None	No	Unroofing
18	14	F	RCA	None	TTE		Bicuspid AoV	Yes	Unroofing
19	16	M	RCA	Nonexercise CP	TTE		None	Yes	Unroofing
20	16	M	RCA	Syncope with exercise	TTE	TEE	None	No	Unroofing
21	10	F	RCA	Syncope after exercise	TTE	CT	None	No	Unroofing
22	9	M	RCA	Syncope with exercise	TTE	Cath	None	Yes	Unroofing
23	7	F	RCA	None (heart murmur)	TTE	CT	None	Yes	Unroofing
24	11	M	LCA	None (abnormal ECG for LVH)	TTE		None	No	Unroofing
25	15	M	RCA	None (SVT)	TTE		None	No	Unroofing
26	14	M	LCA	SCD with exercise	TTE		None	No	Unroofing
27	11	F	RCA	Syncope with exercise	TTE		None	No	Unroofing

AOCA, Aortic cross-clamp origin of coronary artery; Dx, diagnosis; CHD, congenital heart disease; F, female; LCA, left coronary artery; TTE, transthoracic echocardiogram; Cath, catheterization; MRI, magnetic resonance imaging; CT, computed tomography; ECG, electrocardiogram; SVT, supraventricular tachycardia; TEE, transesophageal echocardiogram.

FROMMELT ET AL



FROMMELT ET AL

- 27 patients with mean age 12.6 years
- Diagnosis of intramural course by echocardiography
- Treatment exclusively by unroofing
- 2/3 preop SCD on ecmo
- Left intramural course much longer than right
- All patients treated regardless of whether right, left asymptomatic, etc if intramural

NOW FOR THE MOVIE

