What to Recommend: Coronary Artery Anomalies

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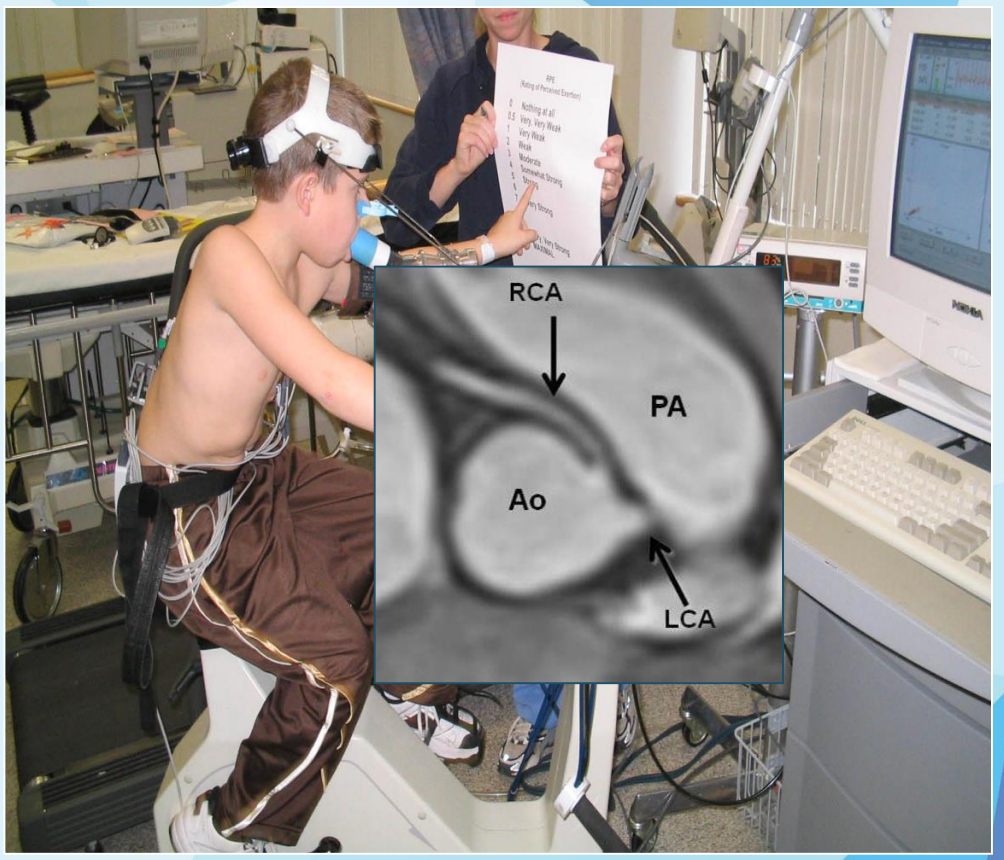


Children's Hospital of Philadelphia℠
Cardiac Center

Perelman School of Medicine
University of Pennsylvania

Cardiology 2018

Disclosures

- None

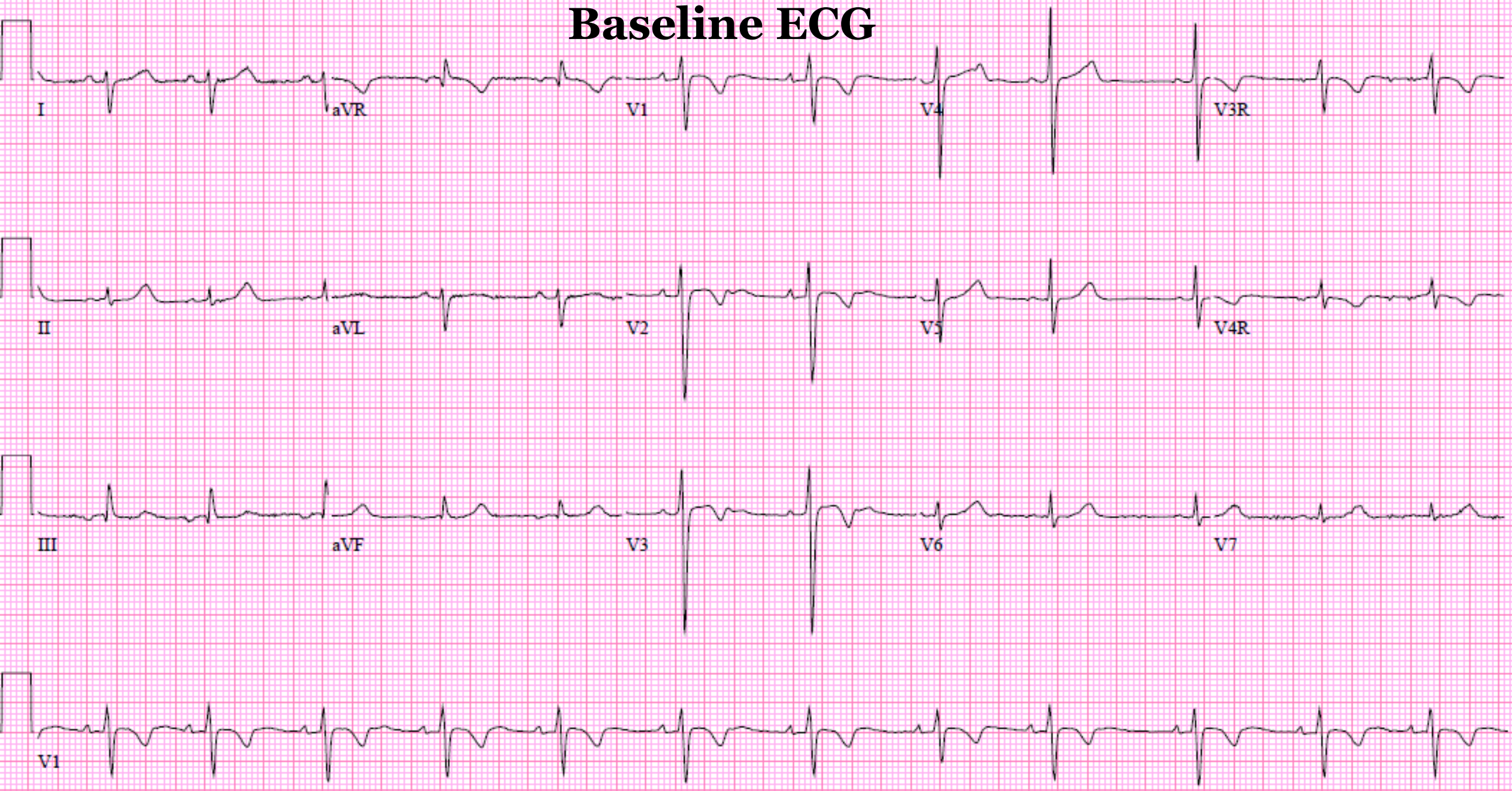
Objectives

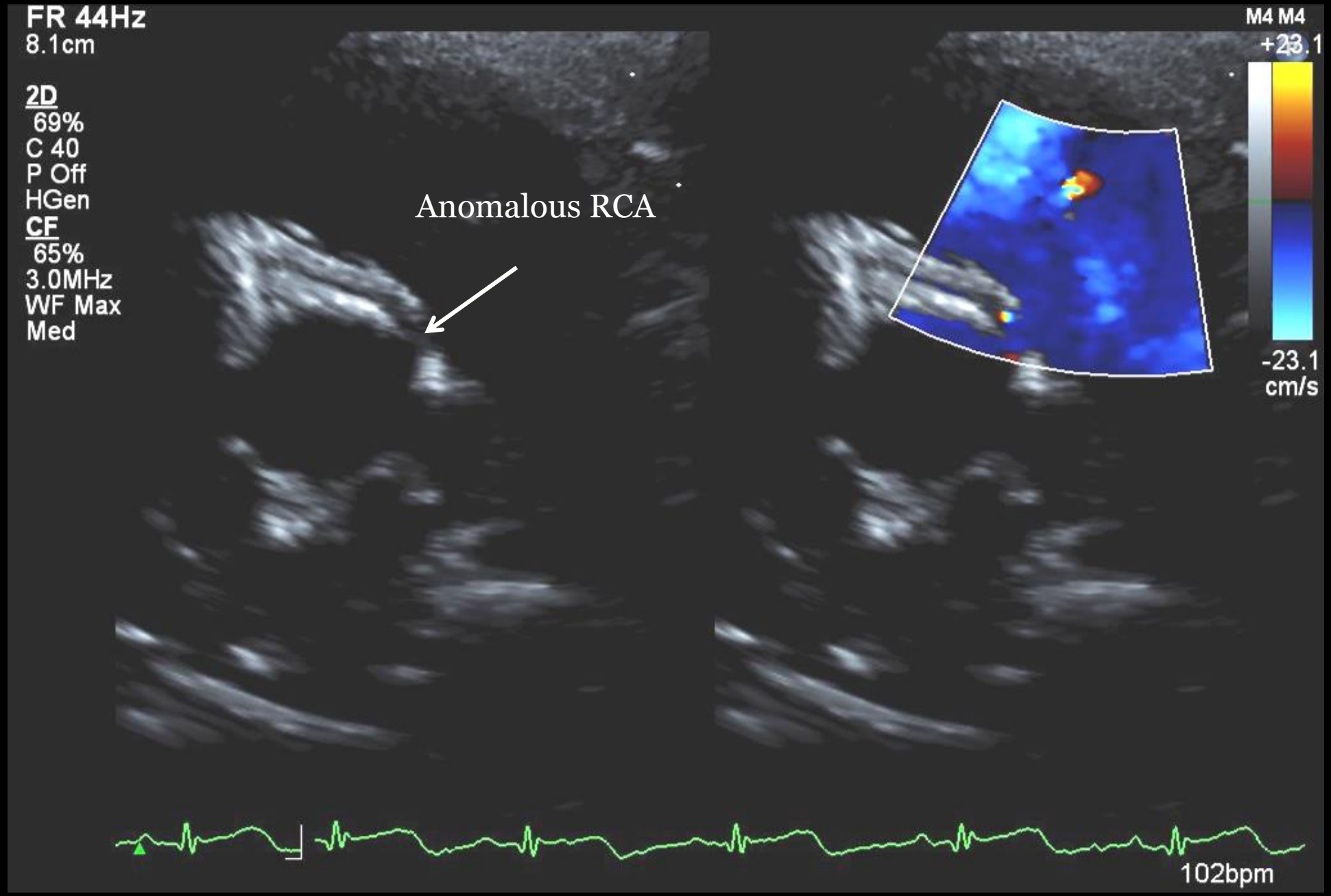
- Provide an overview of anomalous aortic origin of a coronary artery (AAOCA)
- Understand why there are concerns regarding sports activity in patients with AAOCA
- Review the use of exercise restriction in management of AAOCA
- Review recommendations for physical activity in patients with AAOCA

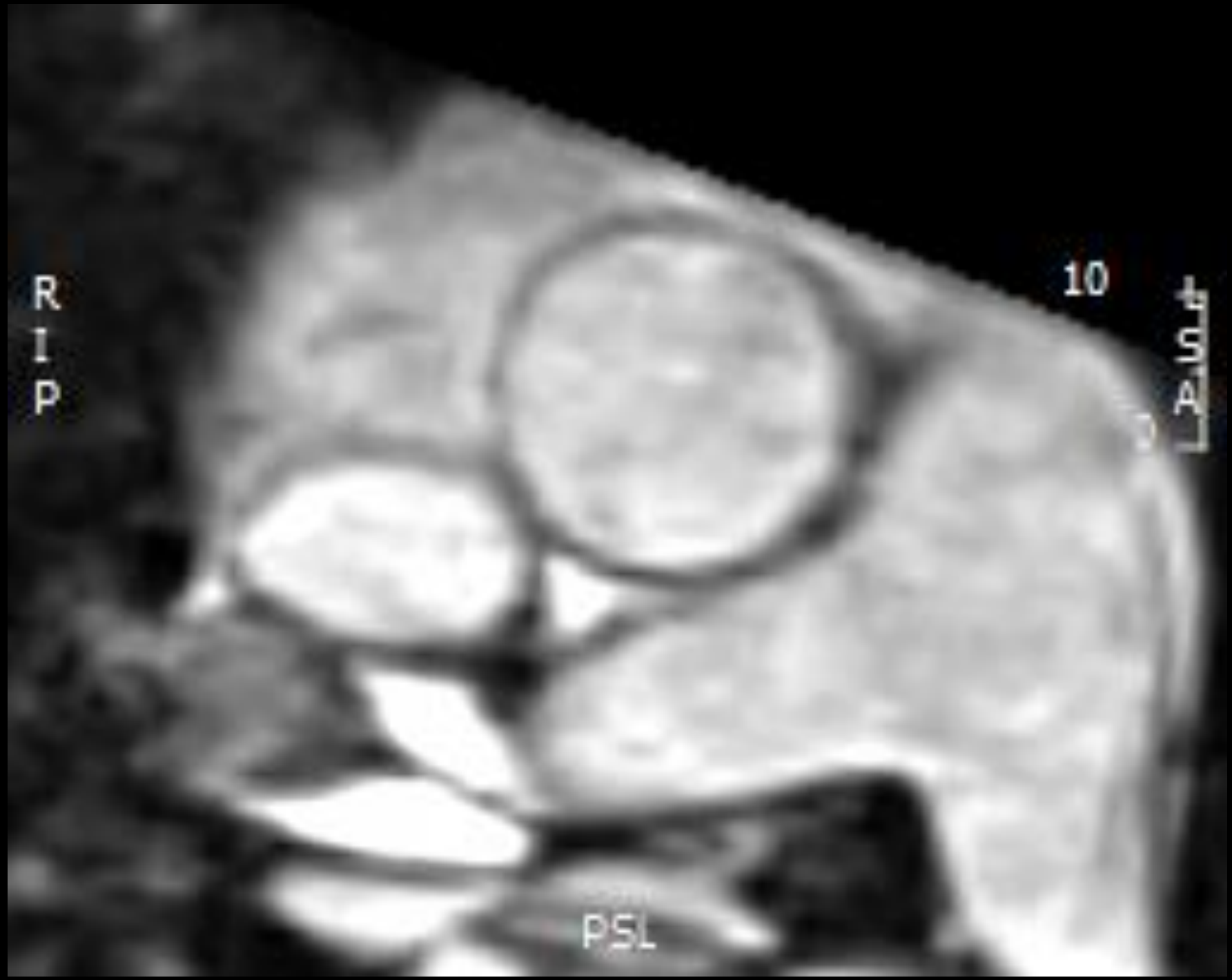
Clinical Case-Patient K.K.

- 14 yo competitive track athlete presents after experiencing dizziness after a 400 meter race
  - Sat down and drank Gatorade and water and felt better immediately
- Extremely hot and humid day
- Generally drinks 24-32 oz of fluid daily
- He denies chest pain, palpitations, syncope, or previous symptoms
- Family history significant for dad with unicuspid aortic valve and mom with recent-onset atrial fibrillation

Baseline ECG




FR 44Hz
8.1cm
2D
69%
C 40
P Off
HGen
CF
65%
3.0MHz
WF Max
Med
Anomalous RCA
M4 M4
+23.1
-23.1
cm/s
102bpm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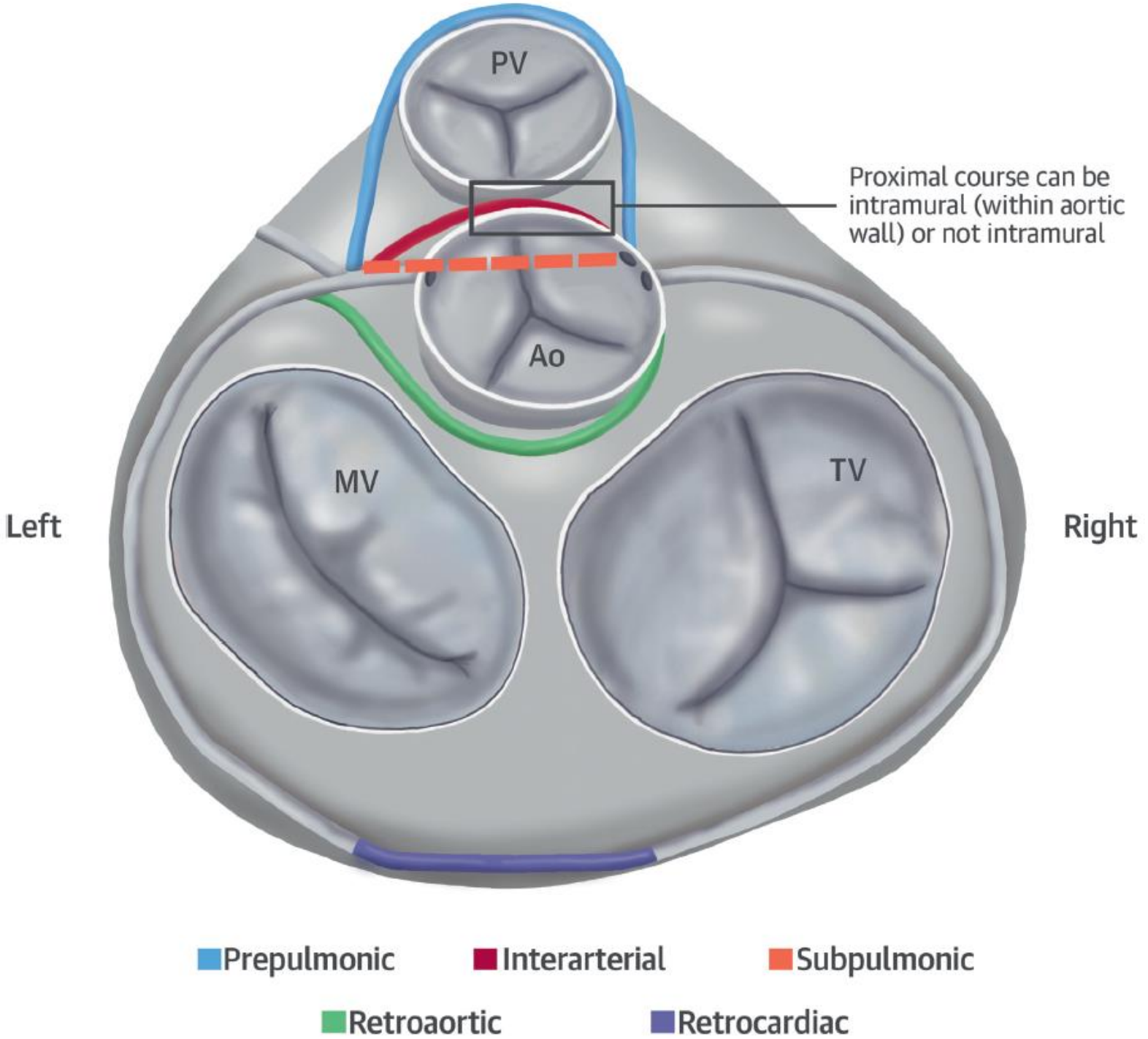
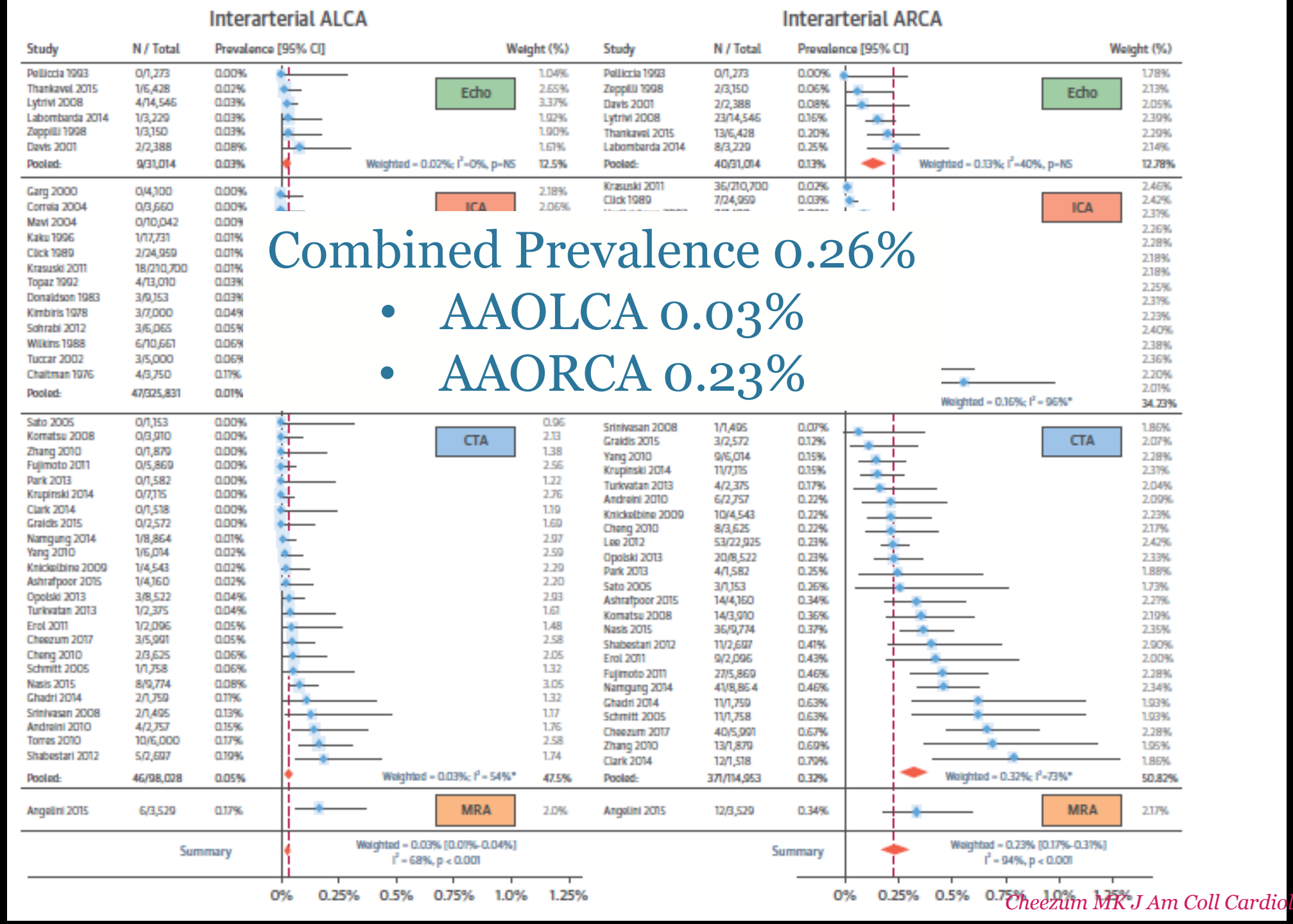
What is AAOCA?

- Anomalous Aortic Origin of a Coronary Artery
- When both coronary arteries arise from or above the same aortic sinus with a single ostium or two separate ostia
- Several different subtypes with varying degrees of risk
  - Vast majority are benign
  - Exercise recommendations are mainly for patients who have interarterial, commonly intramural AAOCA



Cheezum MK J Am Coll Cardiol 2017



Combined Prevalence 0.26%

- AAOLCA 0.03%
- AAORCA 0.23%

Cheezum MK J Am Coll Cardiol 2017

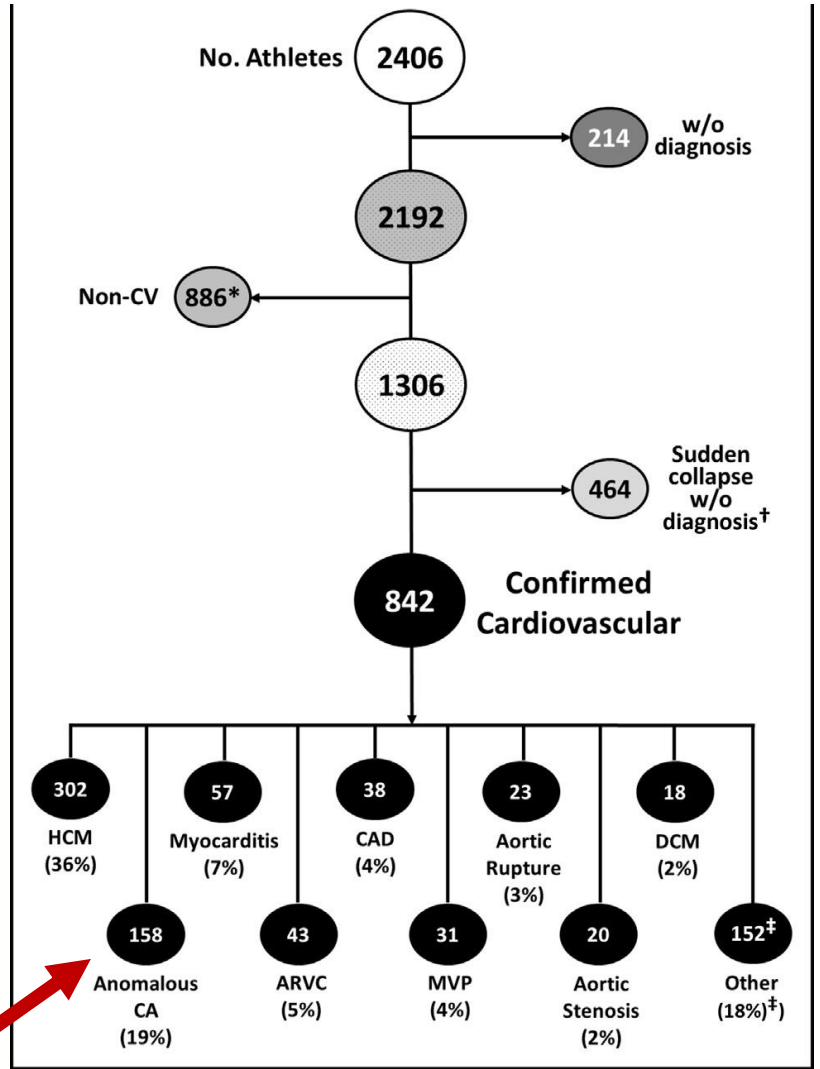
SCD Risk: General Population

- Two fairly recent studies looked at sudden cardiac death (SCD) in populations under age 35
  - 4 deaths due to AAOCA in a combined 34 million patient years
- In one study with 2 deaths from AAOCA noted that both were with vigorous activity and both were AAOLCA
- The risk of SCD from AAOCA in the non-competitive general population is too low to measure

Meyer et al Circ. 2012; Pilmer et al Heart Rhythm, 2014

Demographics and Epidemiology of Sudden Deaths in Young Competitive Athletes: From the United States National Registry

Barry J. Maron, MD, Tammy S. Haas, RN, Aneesha Ahluwalia, Caleb J. Murphy, BS, Ross F. Garberich,
Hypertrophic Cardiomyopathy Center, Minneapolis Heart Institute Foundation, Minn.



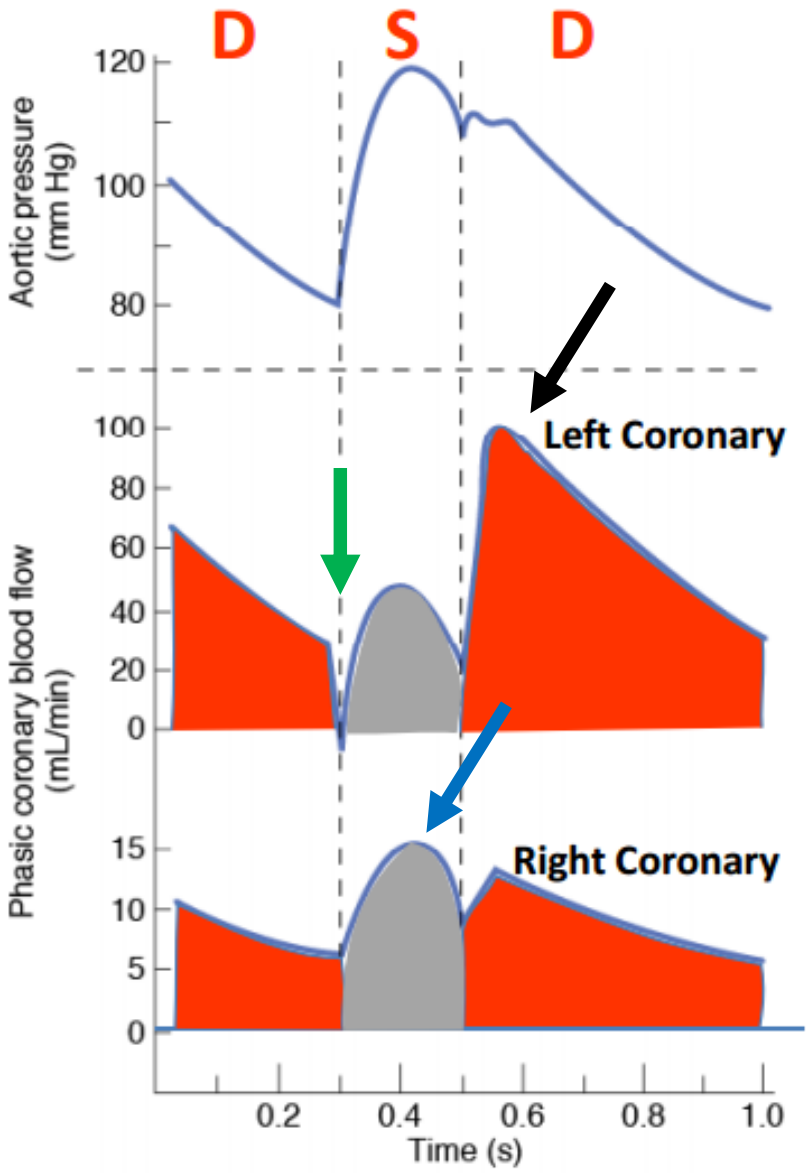
- 1980-2011
- Average age 19 years

Am J Med 2016

Why Are Our Patients with AAOCA at Risk for Ischemia/SCD with Vigorous Activity?

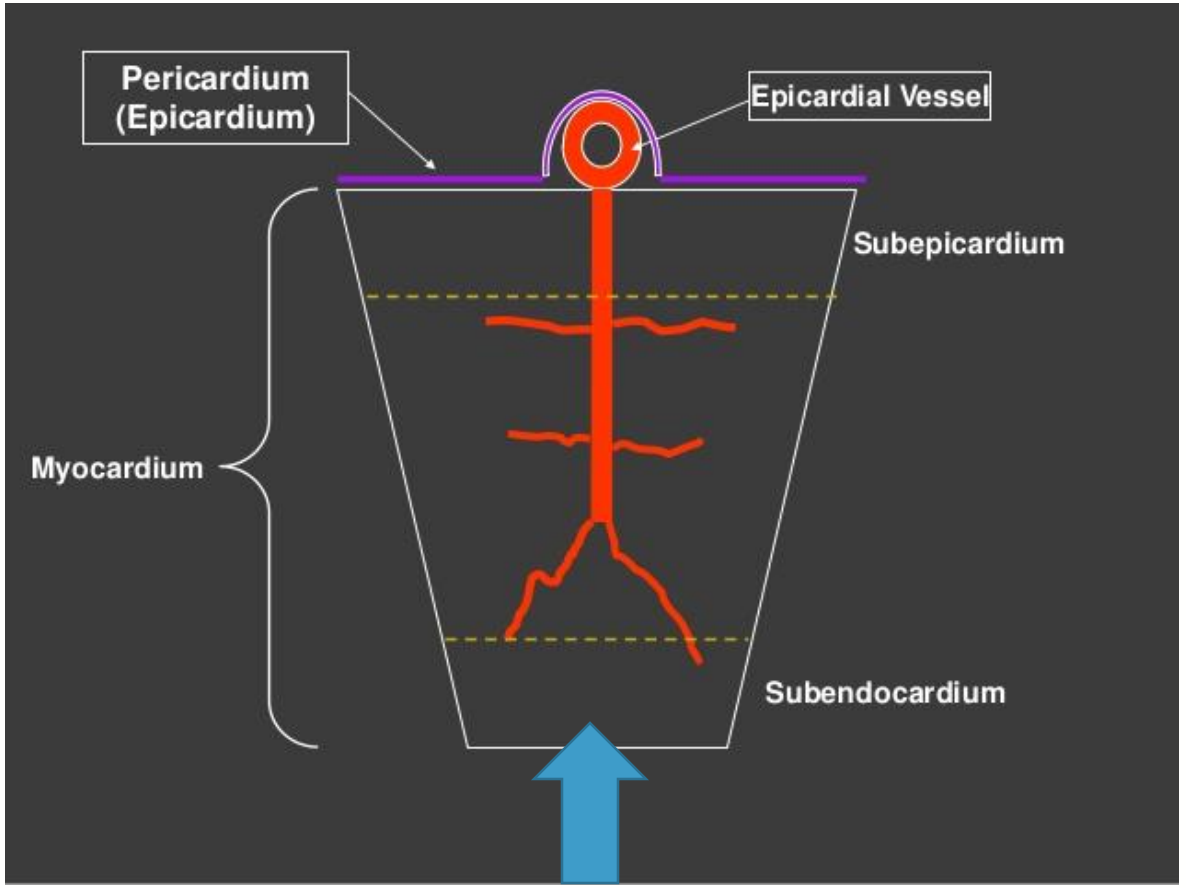
Phasic Coronary Flow in the Coronary Arteries

- Coronary blood flow changes during the cardiac cycle
- 70-85% of coronary flow to the **left ventricle** occurs during *diastole*
- Coronary flow to the **right ventricle** is primarily in *systole*



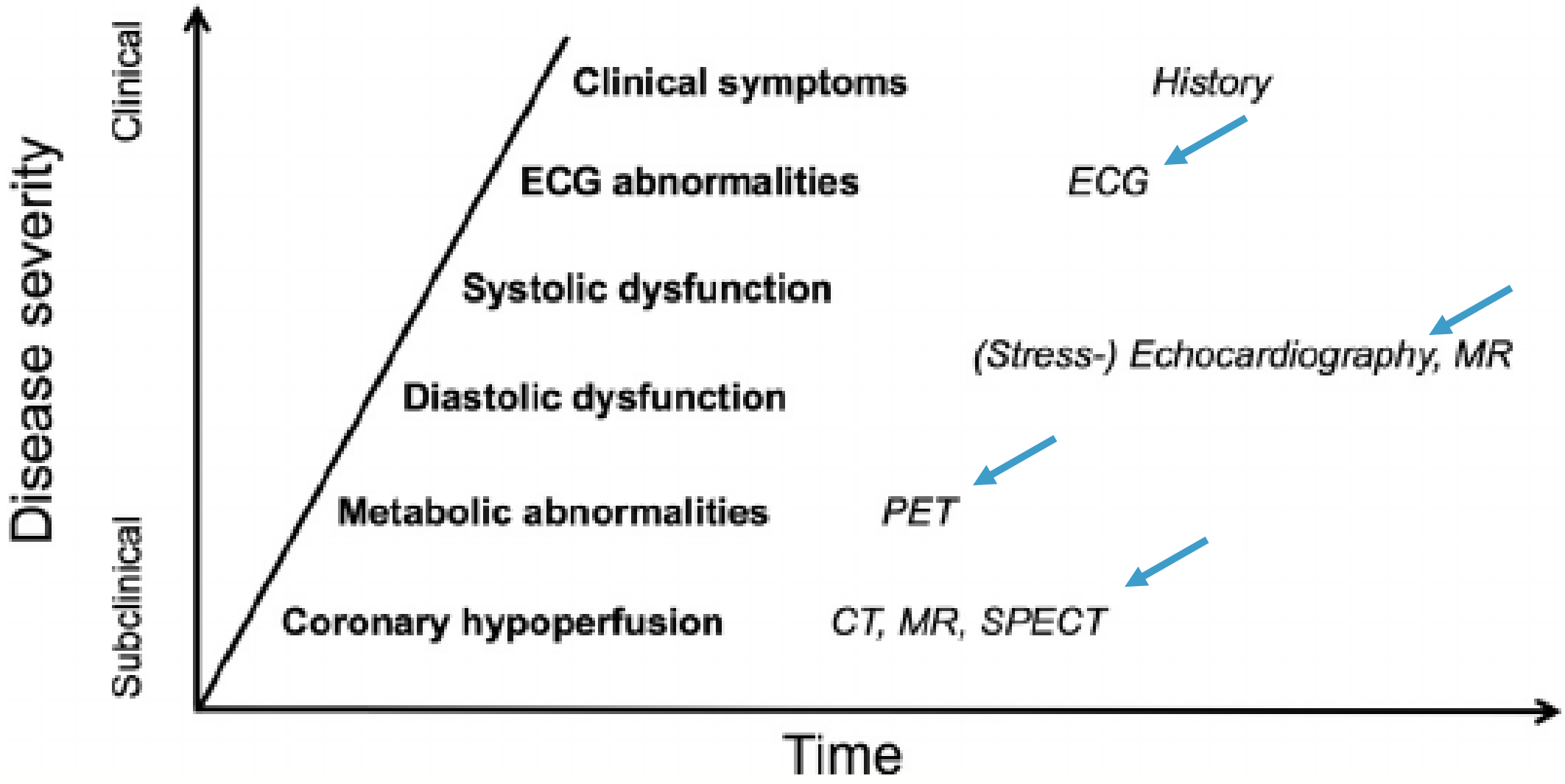
Subendocardium and Ischemia

- Extravascular compressive forces subendocardium > subepicardium
  - Preferential dilation and increased blood flow in the subendocardial vessels due to increased wall stress under normal resting conditions
  - Epicardial coronary stenosis or narrowed orifices are associated with ⬇ subendocardial:subepicardial flow ratio, notably during exercise

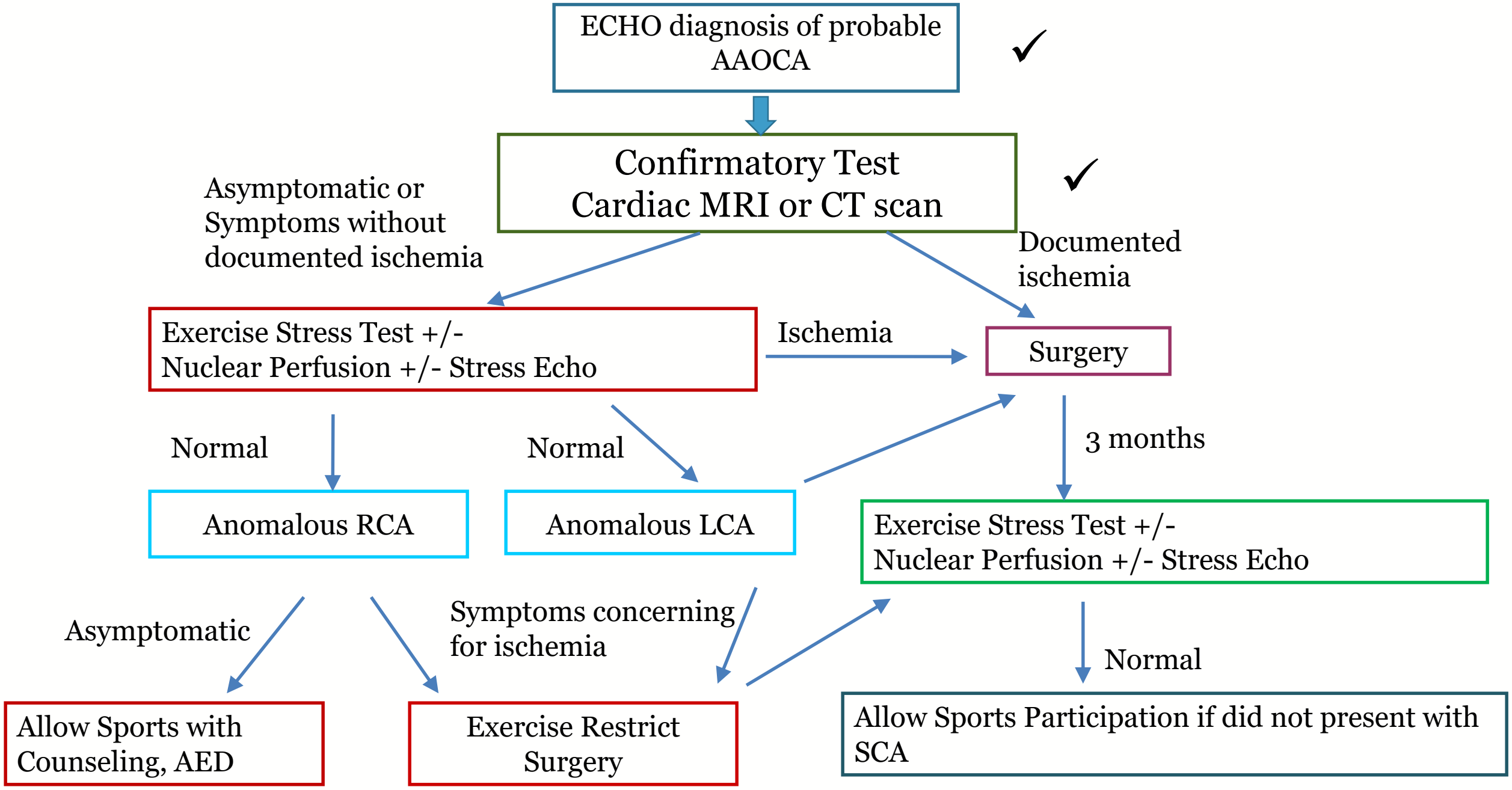


What Provocative Tests Should I Use to Evaluate for Ischemia/Arrhythmia?

Ischemic Cascade

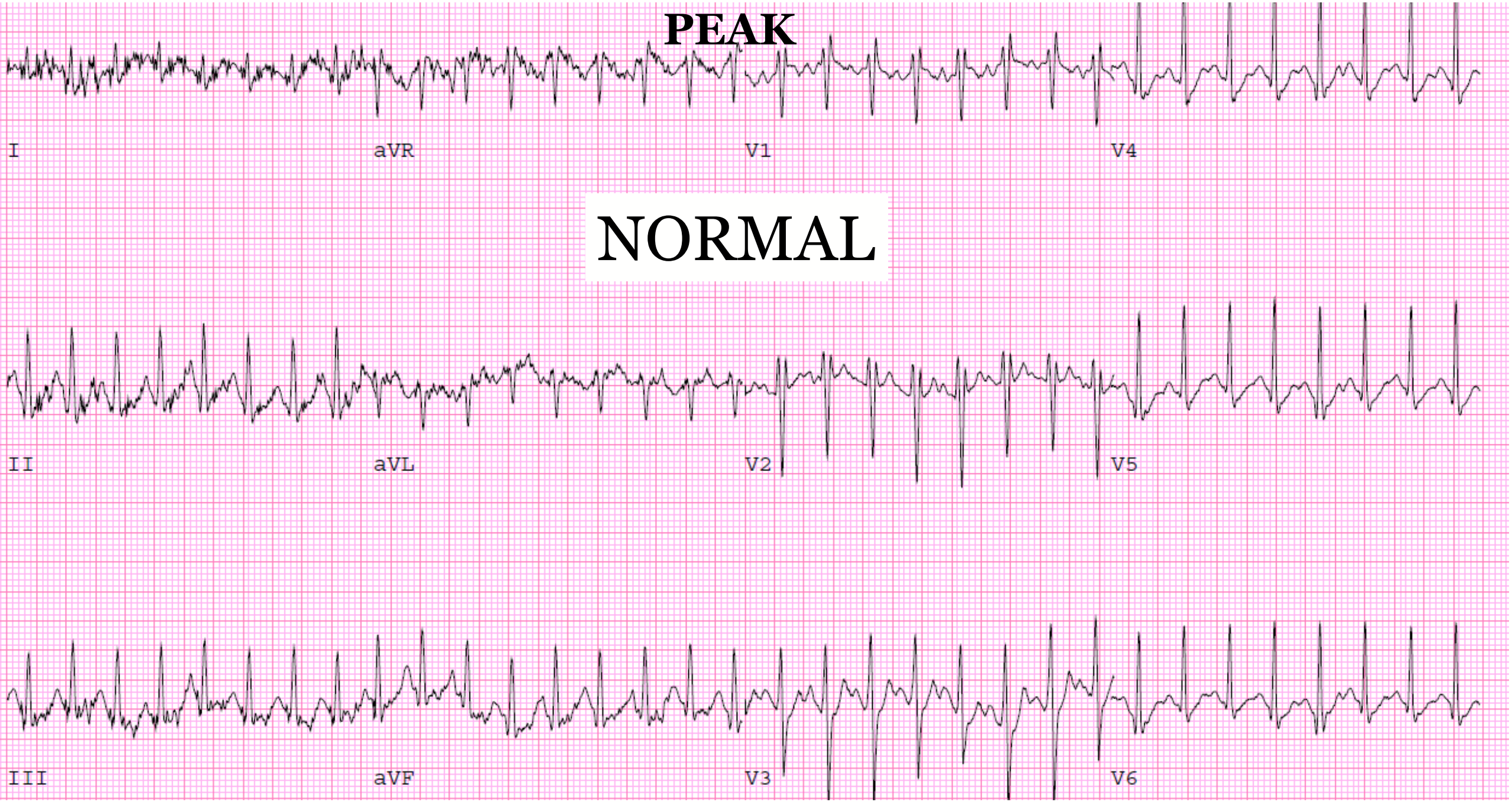


Renker et al. Radiol Clin N Am 2015



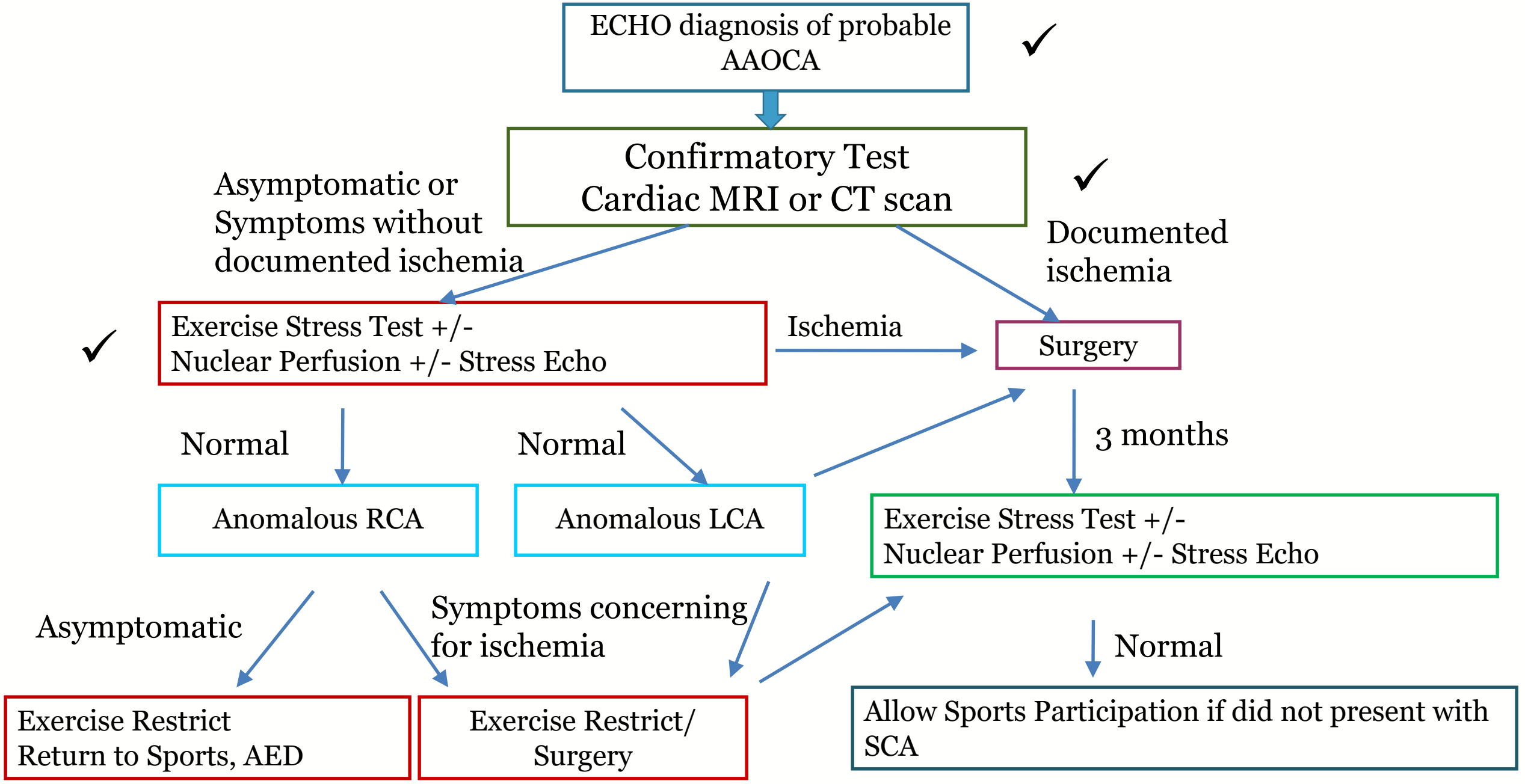
Van Hare et al., J Am Coll Cardiol 2015; Brothers et al., JTCVS 2017

PEAK



NORMAL

Testing is Done: What About Exercise Restriction?



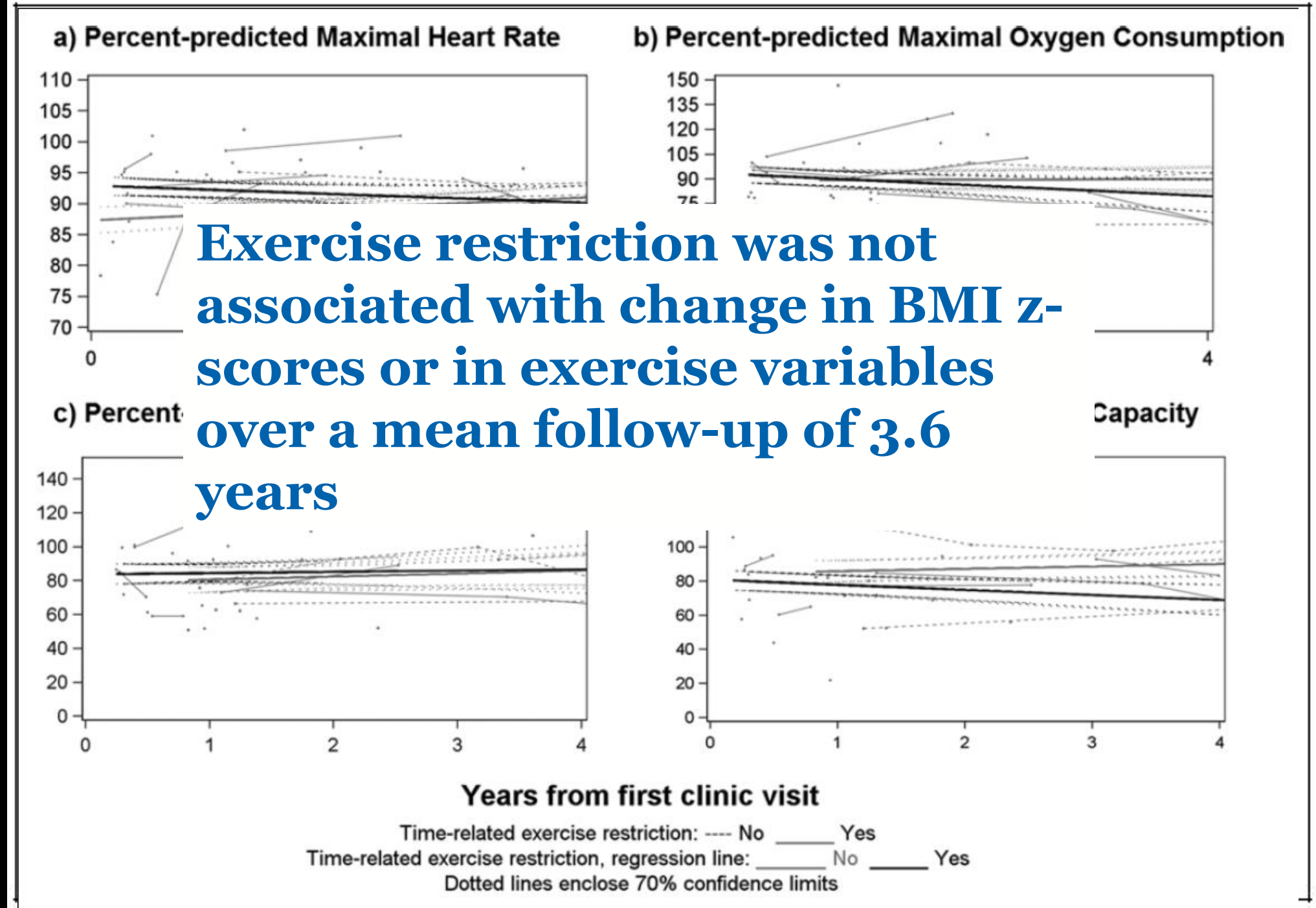
Van Hare et al., J Am Coll Cardiol 2015; Brothers et al., JTCVS 2017

Effects of Exercise Restriction on Patients With Anomalous Aortic Origin of a Coronary Artery

Matthew D. Elias, MD[1], James Meza, MD[2], Brian W. McCrindle, MD[2], Julie A. Brothers, MD[1], Stephen Paridon, MD[1], and Meryl S. Cohen, MD[1]

- Aim to identify association of exercise restriction with changes over time in BMI and exercise performance
- Retrospective review of 72 patients from AAOCA CHSS Registry
  - Median age at presentation 12.6 yrs (10.1-15.8)
  - Mean follow-up 3.6 +/- 3 years
  - 71% with AAORCA
  - 54% had surgery
  - 82% exercise-restricted on presentation

World J Pedatr Cong Heart Surg 2016



Exercise restriction was not associated with change in BMI z-scores or in exercise variables over a mean follow-up of 3.6 years

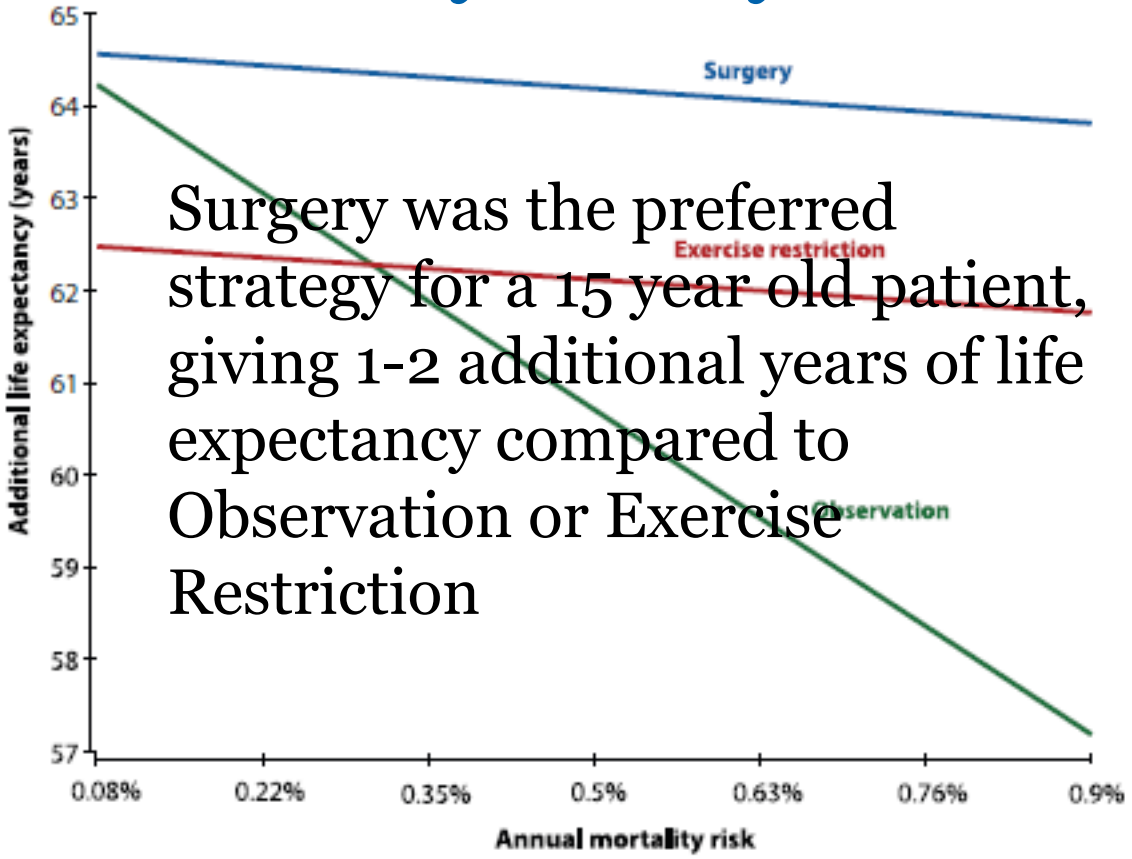
Decision analysis to define the optimal management of athletes with anomalous aortic origin of a coronary artery

Carlos M. Mery, MD, MPH,[a,b] Keila N. Lopez, MD, MPH,[c] Silvana Molossi, MD, PhD,[a,c] S. Kristen Sexson-Tejtel, MD, PhD,[a,c] Rajesh Krishnamurthy, MD,[a,d] E. Dean McKenzie, MD,[a] Charles D. Fraser, Jr, MD,[a,b] and Scott B. Cantor, PhD[e]

- 2 separate decision analysis models: one for AAOLCA and one for AAORCA
- 3 strategies compared: observation, exercise restriction, and surgery
- Probabilities and health utilities estimated based on current literature

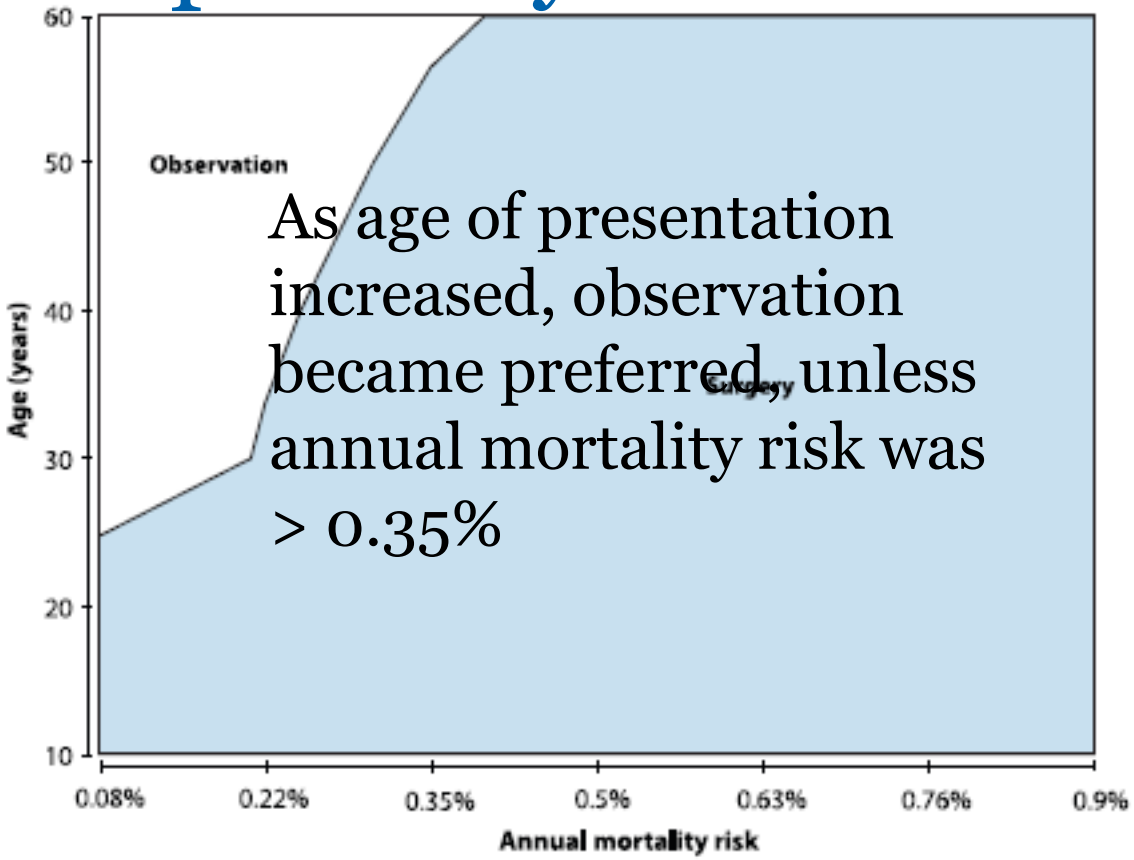
J Thorac Cardiovasc Surg 2016

Sensitivity Analysis for Life Expectancy: AAOLCA



Surgery was the preferred strategy for a 15 year old patient, giving 1-2 additional years of life expectancy compared to Observation or Exercise Restriction

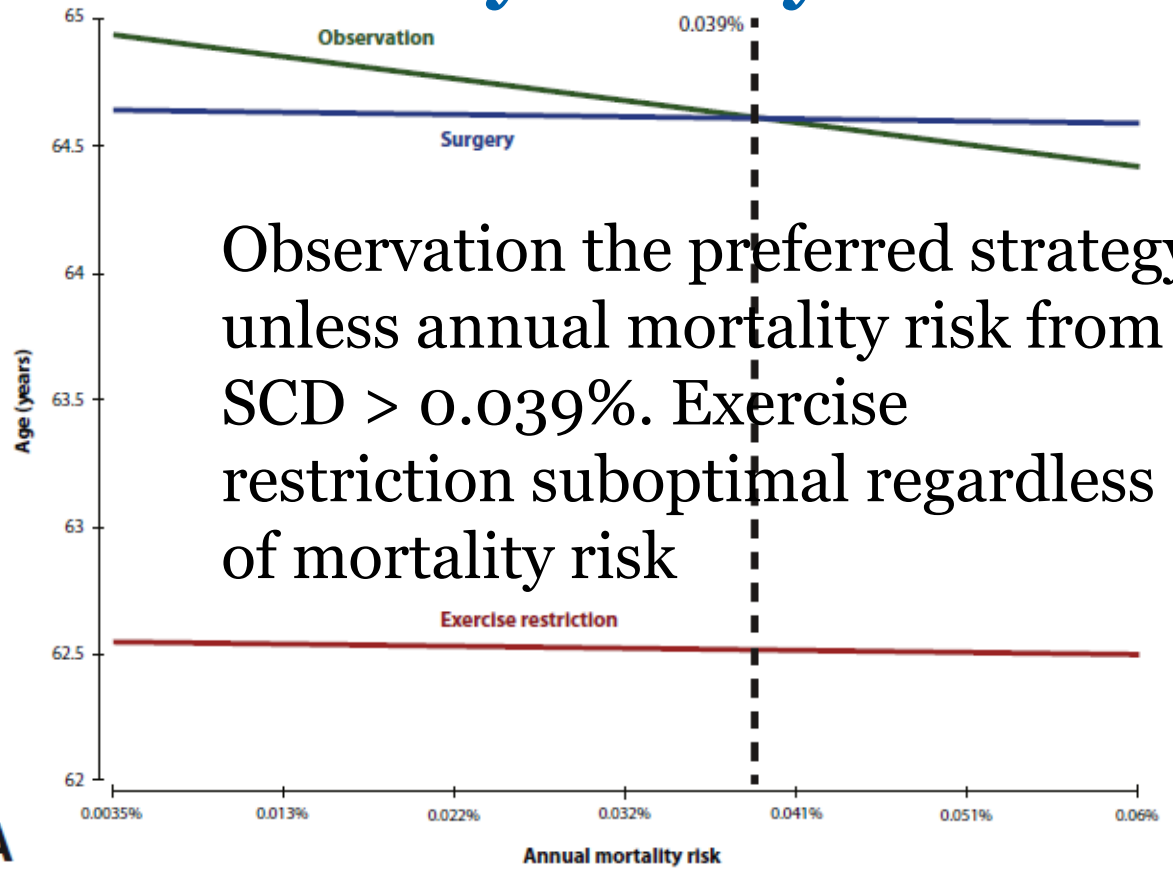
B



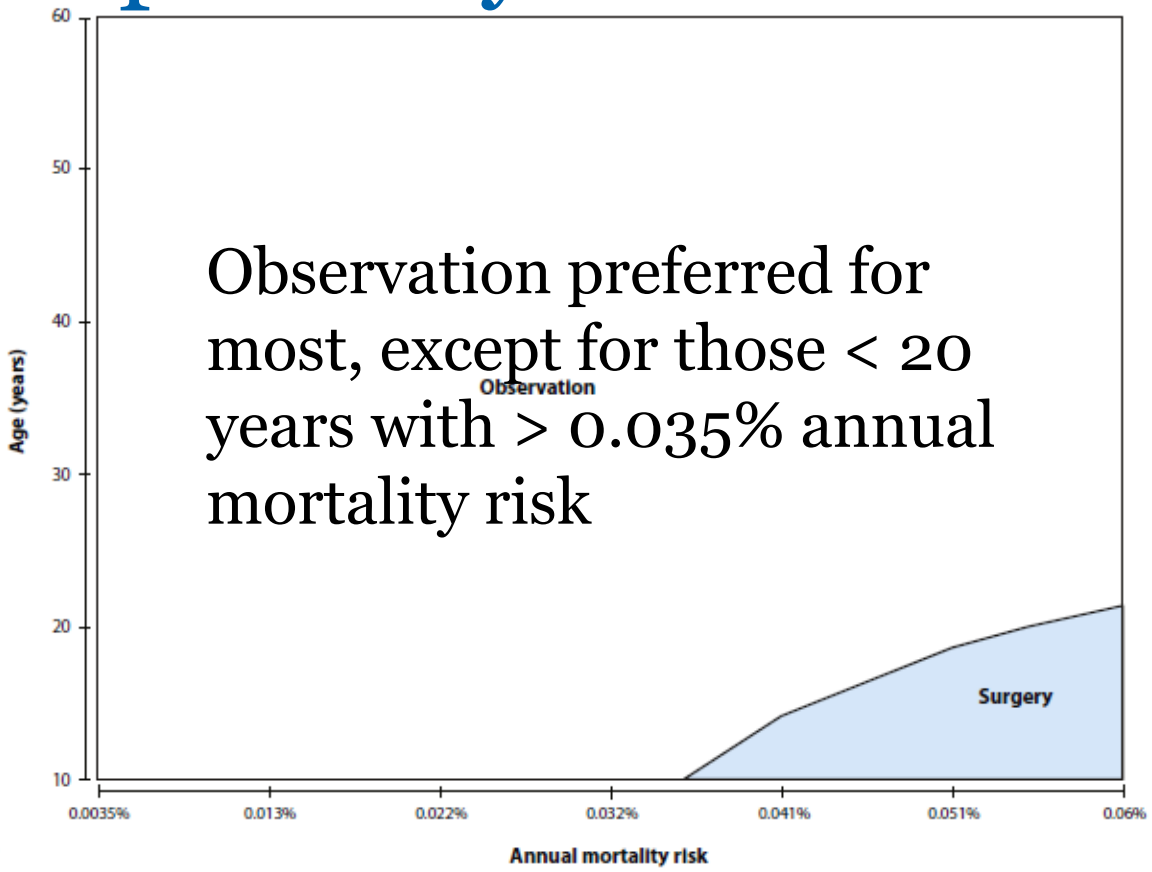
As age of presentation increased, observation became preferred, unless annual mortality risk was > 0.35%

A. 1-way sensitivity analysis showing future life expectancy for each of the strategies for a 15 yo patient while varying the annual risk of SCD
B. 2-way sensitivity analysis shows the optimal strategy while simultaneously varying age at presentation and annual risk. COLORED area represents range where surgery is optimal; white area where observation is optimal

Sensitivity Analysis for Life Expectancy: AAORCA



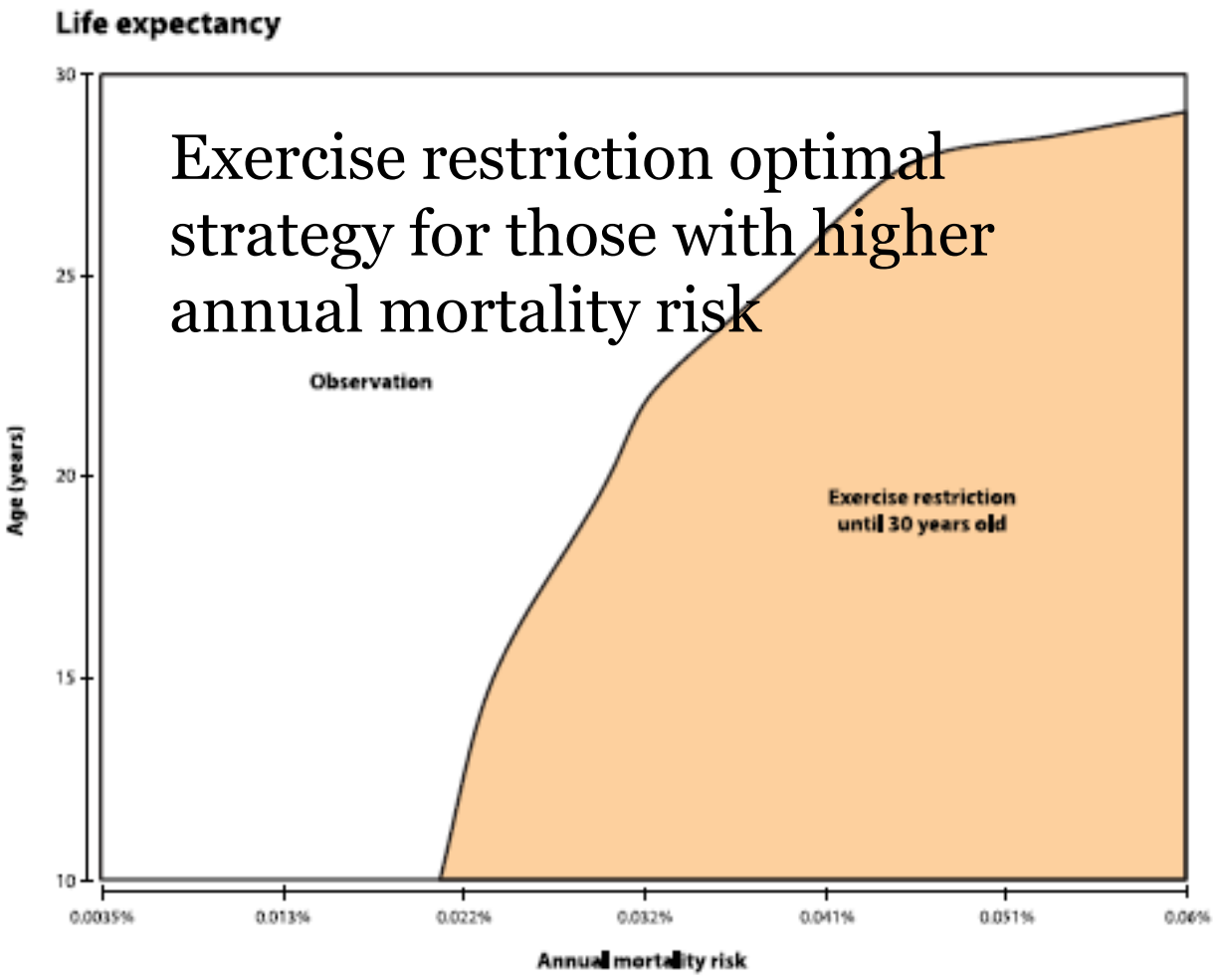
Observation the preferred strategy unless annual mortality risk from SCD > 0.039%. Exercise restriction suboptimal regardless of mortality risk

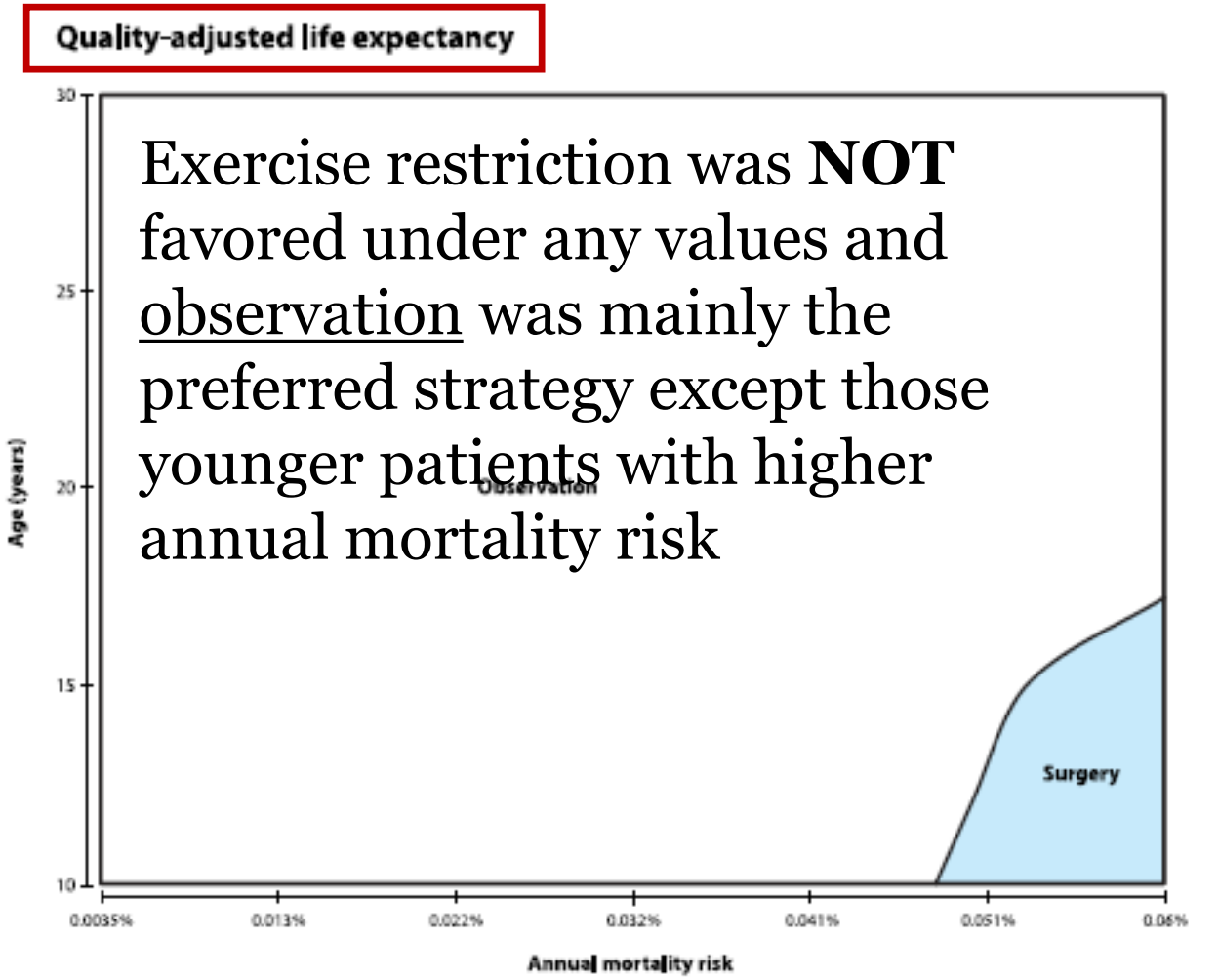


Observation preferred for most, except for those < 20 years with > 0.035% annual mortality risk

A. 1-way sensitivity analysis showing future life expectancy for each of the strategies for a 15 yo patient while varying the annual risk of SCD
B. 2-way sensitivity analysis shows the optimal strategy while simultaneously varying age at presentation and annual risk. COLORED area represents range where surgery is optimal; white area where observation is optimal

Sensitivity Analysis for Patients 10-30 years with AAORCA, Exercise Restricted until 30 years





So…I Don't Exercise Restrict but… My Patient Wants to Do Competitive Sports

Asymptomatic AAORCA

- Patients with AAORCA should be evaluated for inducible ischemia
  - Maximal exercise test with imaging (nuclear or stress echocardiography)
  - Other assessments (e.g., stress CMR, etc) may be done at the discretion of the institution/practitioner
- Without concerning symptoms of ischemia (e.g., syncope with exercise) or a positive provocative test for ischemia, participation in competitive athletics may be considered
  - Counseling concerning the risk of sudden cardiac death
- An AED should be present at every practice and game

Brothers JA et al., J Thorac Cardiovasc Surg 2017;
Van Hare et al., J Am Coll Cardiol 2015

Who Should Receive Surgery?

- Any patient (AAORCA or AAOLCA) with symptoms of chest pain or syncope suspected to be due to ventricular arrhythmias and/or ischemia, or a history of sudden cardiac arrest (SCA), should be exercise-restricted and offered surgery
- Any patient with interarterial, intramural AAOLCA, even without symptoms, should be offered surgery
  - Age 10 years or older, ? Earlier

Brothers JA et al., JTCVS 2017

AAOCA	Daily physical activity	Recreational sports	Competitive sports	Other
Healthy Children	60 min/day MVPA	Unrestricted	Unrestricted	
Asymptomatic AAORCA with normal CPET with imaging	Like healthy	Like healthy	Unrestricted, after counseling family about SCD risk	Annual CPET if competitive sports
<u>Before surgery</u>: Interarterial AAOLCA, and AAORCA with symptoms, arrhythmias, or signs of ischemia on CPET	Low intensity	Low intensity	Restricted, possible exception of class IA sports	
<u>3 months after surgery</u>: Interarterial AAOLCA and AAORCA who are asymptomatic and CPET without ischemia or cardiac arrhythmias	Like healthy	Like healthy	Unrestricted*	*Consider continued restriction at least 1 year for presentation with exertional syncope or SCA

MVPA=Moderate or vigorous physical activity; AAORCA=anomalous right coronary artery; AAOLCA=anomalous left coronary artery; CPET=Cardiopulmonary Exercise Test

Van Hare et al., J Am Coll Cardiol 2015
Brothers et al., J Thorac Cardiovasc Surg 2017

Patient K.K.

- All testing normal
- Exercise restriction while work-up was ongoing
- After much discussion with the family regarding risk, decision made to allow him to return to sports
  - AED at every sporting event and practice
  - Needs to drink at least 64 oz/day + more when practice/races are outside in the heat

Summary

Exercise in AAOCA

- Most patients with AAOCA can do some sort of physical activity, even if it is low intensity
- Exercise restriction is almost never the way to manage a patient with AAOCA, except during ongoing clinical work-up or awaiting surgery
  - Consider return to competitive sports 3 months post-operatively with normal exercise test and presentation NOT exertional syncope or SCA
- Most AAORCA patients can participate in competitive sports
  - Discussion of risks
  - AED present at practices and competitions
- We still have a lot to learn with AAOCA!

Thank You For Your Time