# Insight Into Right Coronary Artery Anomaly Using 256-row Cardiac CT

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## **Disclosures**

No relevant conflicts to disclose



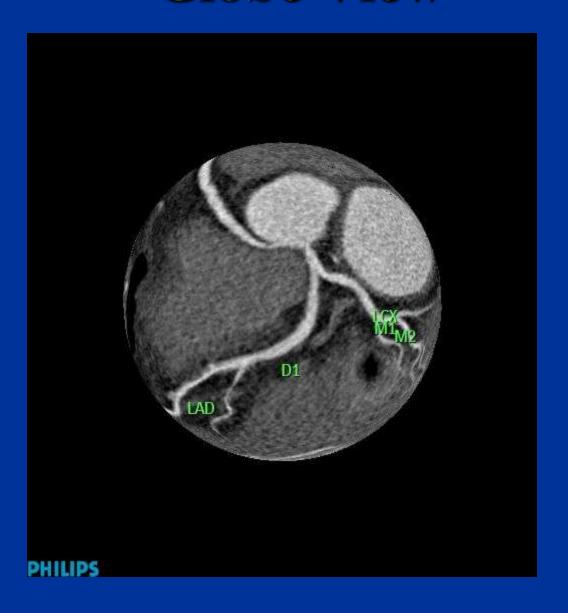
# **Axial Imaging**



## "Crime Scene"?



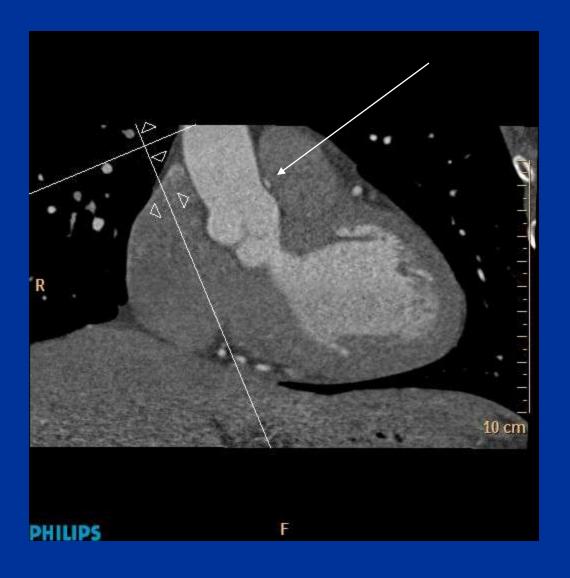
# **Globe View**



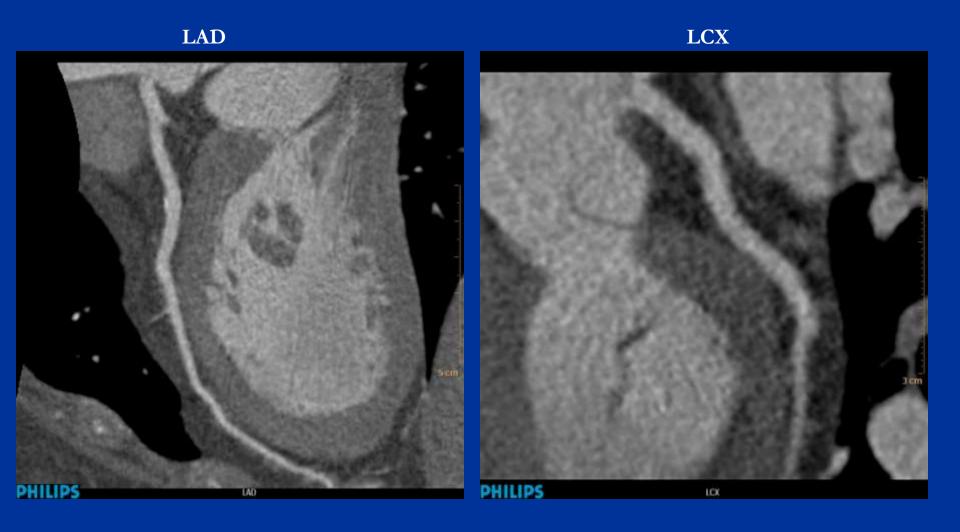
## Curved MPR of the Anomalous RCA



# "Pressure"



# Left Coronary Artery Branches



#### TABLE 2. Classification of Coronary Anomalies in Human Hearts

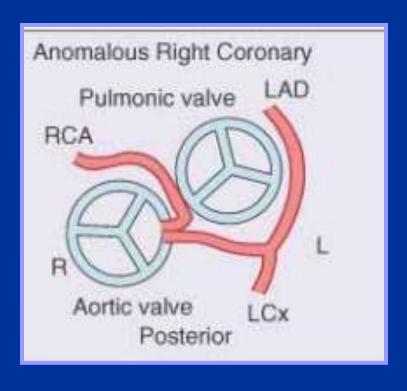
- A. Anomalies of origination and course
- 1. Absent left main trunk (split origination of LCA)
- Anomalous location of coronary estium within aertic root or near proper aertic sinus of Valsalva (for each artery)
- a. High
- b. Low
- c. Commissural
- Anomalous location of coronary estium outside normal "coronary" aortic sinuses
- a. Right posterior aortic sinus
- b. Ascending aorta
- c. Left ventricle
- d. Right ventricle
- e. Pulmonary artery
- (1) LCA that arises from posterior facing sinus
- (2) Cx that arises from posterior facing sinus
- (3) LAD that arises from posterior facing sinus
- (4) RCA that arises from anterior right facing sinus
- (5) Ectopic location (outside facing sinuses) of any coronary artery from pulmonary artery
  - (a) From anterior left sinus
- (b) From pulmonary trunk
- (c) From pulmonary branch
- f. Aortic arch
- g. Innominate artery
- h. Right carotid artery
- i. Internal mammary artery
- j. Bronchial artery
- k. Subclavian artery
- I. Descending thoracic aorta
- Anomalous location of coronary ostium at improper sinus (which may involve joint origination or "single" coronary pattern)
- a. RCA that arises from left anterior sinus, with anomalous course
- (1) Posterior atrioventricular groove or retrocardiac
- (2) Retroaortic
- (3) Between aorta and pulmonary artery (intramural)
- (4) Intraseptal
- (5) Anterior to pulmonary outflow
- (6) Posteroanterior interventricular groove (wraparound)
- b. LAD that arises from right anterior sinus, with anomalous course
  - Between aorta and pulmonary artery (intramural)
- (2) Intraseptal
- (3) Anterior to pulmonary outflow
- (4) Posteroanterior interventricular groove (wraparound)
- c. Cx that arises from right anterior sinus, with anomalous course
- (1) Posterior atrioventricular groove
- (2) Retroaortic
- d. LCA that arises from right anterior sinus, with anomalous course

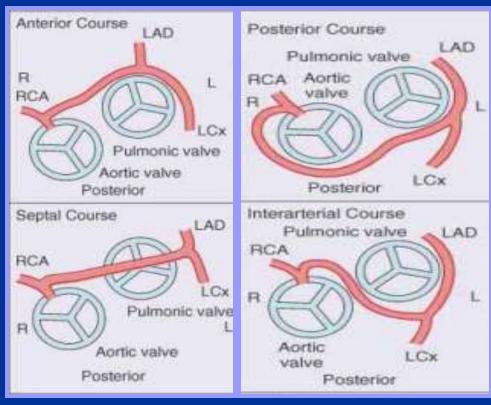
#### Continued

- (1) Posterior atrioventricular groove
- (2) Retroaortic
- (3) Between aorta and pulmonary artery
- (4) Intraseptal
- (5) Anterior to pulmonary outflow
- (6) Posteroanterior interventricular groove
- 5. Single coronary artery (see A4)
- B. Anomalies of intrinsic coronary arterial anatomy
- 1. Congenital ostial stenosis or atresia (LCA, LAD, RCA, Cx)
- 2. Coronary ostial dimple
- 3. Coronary ectasia or aneurysm
- 4. Absent coronary artery
- 5. Coronary hypoplasia
- 6. Intramural coronary artery (muscular bridge)
- 7. Subendocardial coronary course
- 8. Coronary crossing
- Anomalous origination of posterior descending artery from the anterior descending branch or a septal penetrating branch
- 10. Split RCA
- a. Proximal+distal PDs that both arise from RCA
- b. Proximal PD that arises from RCA, distal PD that arises from LAD
- c. Parallel PDs ×2 (arising from RCA, Cx) or "codominant"
- 11. Split LAD
- a. LAD+first large septal branch
- b. LAD, double (parallel LADs)
- 12. Ectopic origination of first septal branch
  - a. RCA
  - b. Right sinus
  - c. Diagonal
  - d. Ramus
  - e, Cx
- C. Anomalies of coronary termination
- 1. Inadequate arteriolar/capillary ramifications
- 2. Fistulas from RCA, LCA, or infundibular artery to:
- a. Right ventricle
- b. Right atrium
- c. Coronary sinus
- d. Superior vena cava
- e. Pulmonary artery
- . . .
- f. Pulmonary vein
- g. Left atrium h. Left ventricle
- i. Multiple, right+left ventricles
- D. Anomalous anastomotic vessels
- LCA indicates laft coronary artery; LAD, left descending coronary artery; RCA, right coronary artery; Cx indicates circumflex; and PD, posterior descending branch. Adapted from Angelini P et al <sup>10</sup> with permission from Lippincott, Williams & Wilkins. Copyright 1999.

## **Anomalies That May Cause Ischemia**

## ACAOS





Zipes: Braunwald's Heart Disease: A Textbook of Cardiovascular Medicine, 7th ed

# Proposed Mechanisms of Decreased Coronary Flow in ACAOS

- Stretch and compression of intramural segment by AV commissure
- Spasm or thrombosis at anomalous site
- On IVUS imaging, Angelini demonstrated ACAOS involves "intramural proximal intussusception of the ectopic CA at the aortic root wall"
  - Hypoplasia of the intussuscepted segment
  - Lateral luminal compression during systole
  - Segment length: the longer the segment, the more severe the stenosis

#### **Annals of Internal Medicine**

ARTICLE

## Sudden Death in Young Adults: A 25-Year Review of Autopsies in Military Recruits

Robert E. Eckart, DO; Stephanie L. Scoville, DrPH; Charles L. Campbell, MD; Eric A. Shry, MD; Karl C. Stajduhar, MD; Robert N. Potter, DVM, MPH; Lisa A. Pearse, MD, MPH; and Renu Virmani, MD

Background: Sudden death among military recruits is a rare but devastating occurrence. Because extensive medical data are available on this cross-sectional and diverse population, identification of the underlying causes of sudden death may promote health care policy to reduce the incidence of sudden death.

Objective: To determine the causes of nontraumatic sudden death among a cohort of military recruits.

Design: Retrospective cohort study using demographic and autopsy data from the Department of Defense Recruit Mortality Registry.

Setting: Basic military training.

Patients: All nontraumatic sudden deaths from a monitored 6.3 million men and women age 18 to 35 years.

Measurements: Descriptive analysis, crude mortality rates of causes of sudden death, and frequency of events as a function of cause of death.

Results: Of 126 nontraumatic sudden deaths (rate, 13.0/100 000 recruit-years), 108 (86%) were related to exercise. The most com-

mon cause of sudden death was an identifiable cardiac abnormality (64 of 126 recruits [51%]); however, a substantial number of deaths remained unexplained (44 of 126 recruits [35%]). The predominant structural cardiac abnormalities were coronary artery abnormalities (39 of 64 recruits [61%]), myocarditis (13 of 64 recruits [20%]), and hypertrophic cardiomyopathy (8 of 64 recruits [13%]). An anomalous coronary artery accounted for one third (21 of 64 recruits) of the cases in this cohort, and, in each, the left coronary artery arose from the right (anterior) sinus of Valsalva, coursing between the pulmonary artery and aorta.

Limitations: This cohort underwent a preenlistment screening program that included history and physical examination; this may have altered outcomes.

Conclusions: Cardiac abnormalities are the leading identifiable cause of sudden death among military recruits; however, more than one third of sudden deaths remain unexplained after detailed medical investigation.

Ann Intern Med. 2004;141:829-834.
For author affiliations, see end of text.
See editorial comment on pp 882-884.

www.annals.org

Table 3. Nontraumatic Sudden Deaths with an Identifiable Cardiac Abnormality during Recruit Training, 1977–2001 (n = 64)

Cardiac Abnormality	Sudden Deaths, n (%)*
Cardiomyopathy	23 (36)
Myocarditis	13 (20)
Hypertrophic cardiomyopathy	8 (13)
Idiopathic dilated cardiomyopathy	1 (2)
Right ventricular dysplasia	1 (2)
Coronary artery pathology	39 (61)
Anomalous coronary artery	21 (33)
Atherosclerotic coronary artery disease	10 (16)
Coronary artery hypoplasia	3 (5)
Coronary aneurysm	2 (3)
Intramyocardial coronary bridge	2 (3)
Coronary dissection	1 (2)
Miscellaneous cardiac findings	2 (3)
Bicuspid aortic valvular stenosis	1 (2)
Embolic myocardial infarction	1 (2)

<sup>\*</sup> Some numbers have been rounded

# Rare Case Reports: Anomalous RCA & Sudden Death

Forensic Sci Int. 2001 Dec 1;123(2-3):254-6.

Anomalous origin of the right coronary artery from the left sinus of Valsalva: report of two cases.

Bunai Y, Akaza K, Tsujinaka M, Nagai A, Nakamura I, Ohya I.

Source

Department of Legal Medicine, Gifu University School of Medicine, 40 Tsukasamachi, Gifu 500-8705, Japan. bunaiy@cc.gifu-u.ac.jp

Abstract

Anomalous origin of the right coronary artery (RCA) from the left sinus of Valsalva (LSV) is rare and has only recently been recognized as a cause of sudden cardiac death in otherwise healthy individuals. In this report, we describe two cases of anomalous origin of the RCA from the LSV. In the first case, a previously healthy 11-year-old girl died suddenly and unexpectedly. An autopsy revealed no abnormalities, except for anomalous origin of the RCA from the LSV. In the second case, a 21-year-old woman died by drowning in a bath, probably after an episode of syncope brought about by anomalous origin of the RCA from the LSV.

<u>I Thorac Cardiovasc Surg.</u> 2011 Mar 23. [Epub ahead of print]

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

Frommelt PC, Sheridan DC, Berger S, Frommelt MA, Tweddell JS.

Source

Division of Pediatric Cardiology, Department of Pediatrics, Children's Hospital of Wisconsin, Medical College of Wisconsin, Milwaukee, Wis.

Abstract

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, 4-20 years) had surgical unroofing of AAOCA of the left **coronary** from the **right** sinus (left AAOCA, 7/27, 26%) or 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.

#### **CONCLUSIONS:**

AAOCA is frequently characterized by an intramural course, which can be reliably identified by echocardiography. This form can be safely repaired by unroofing the intramural segment without early morbidity. Symptoms of possible ischemia are common but not always correlated with **coronary** ostial findings at surgery.

### Long-Term Outcome and Impact of Surgery on Adults With Coronary Arteries Originating From the Opposite Coronary Cusp

Richard A. Krasuski, MD; Dari Magyar, MD; Stephen Hart, BS; Vidyasagar Kalahasti, MD; Richard Lorber, MD; Robert Hobbs, MD; Gosta Pettersson, MD; Eugene Blackstone, MD

Background—An anomalous coronary artery from the opposite sinus of Valsalva may increase sudden death risk in children and young adults, and surgical intervention is often recommended. The impact of this lesion when recognized in the adult and its management are ill defined.

Methods and Results—We reviewed 210 700 cardiac catheterizations performed over a 35-year period at a single institution and identified 301 adults with an anomalous coronary artery from the opposite sinus of Valsalva, either anomalous right coronary artery from the left cusp or anomalous left main coronary artery from the right cusp. Patients were stratified by the pathway of the anomalous artery and the chosen treatment. Among the 301 patients with anomalous coronary artery from the opposite sinus of Valsalva (0.14% of the cohort), 79% had anomalous right coronary artery from the left cusp, and 18% had an interarterial course (IAC). Patients with IAC were younger (52±13 versus 59±13 years; P=0.001) and more likely to undergo surgical intervention (52% versus 27%; P<0.001), but mortality was not increased with IAC. Among the 54 patients with IAC, 28 underwent surgical repair with no perioperative deaths. Patients evaluated since 2000 were significantly more likely to be referred for surgery (P=0.004). Surgical patients were more likely to have abnormal stress tests (90% versus 43%; P=0.01) and had more extensive atherosclerosis but less diabetes melfitus (0% versus 23%; P=0.01). Long-term survival at 10 years appeared similar in both groups.

Conclusions—In this single-center cohort study of patients with an anomalous coronary artery from the opposite sinus of Valsalva, surgical management appears to have been favored recently. Despite no perioperative mortality, a positive impact on long-term survival was not observed. The impact of surgery in older adults with anomalous coronary arteries arising from the opposite coronary sinus with IAC deserves further study. (Circulation, 2011;123:154-162.)

### **Cleveland Clinic Experience**

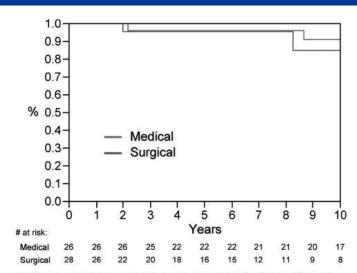
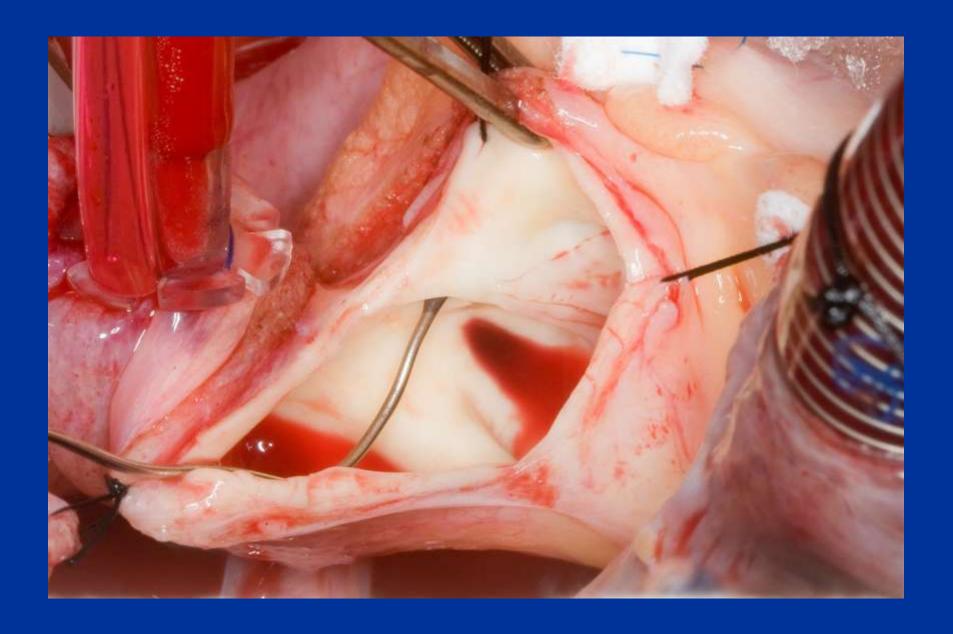


Figure 4. Survival free from death based on initial medical or surgical management in patients with ACAOS passing between the great vessels. At 10-year follow-up, there was no difference in survival (*P*=0.65).

# Thank You



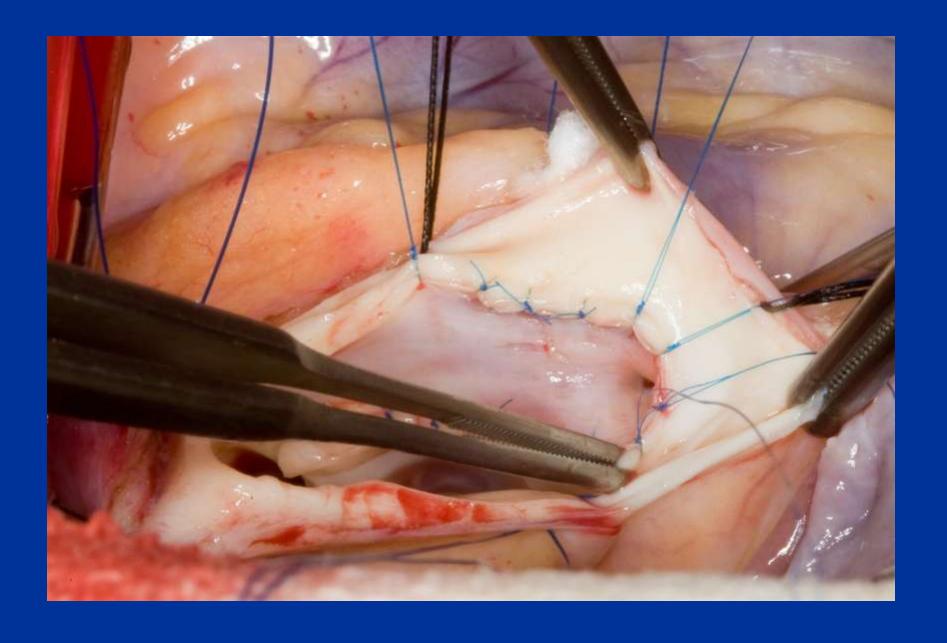
**Common Ostia with Intramural Course** 



**Cannulate RCA** 



**Unroof intramural origin** 



RCA exiting aortic wall