

IN 2018 HOW DO WE TREAT...

Anomalous Coronaries with a Malignant Course

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NO CONFLICT OF INTEREST
TO DECLARE

Definition and Incidence

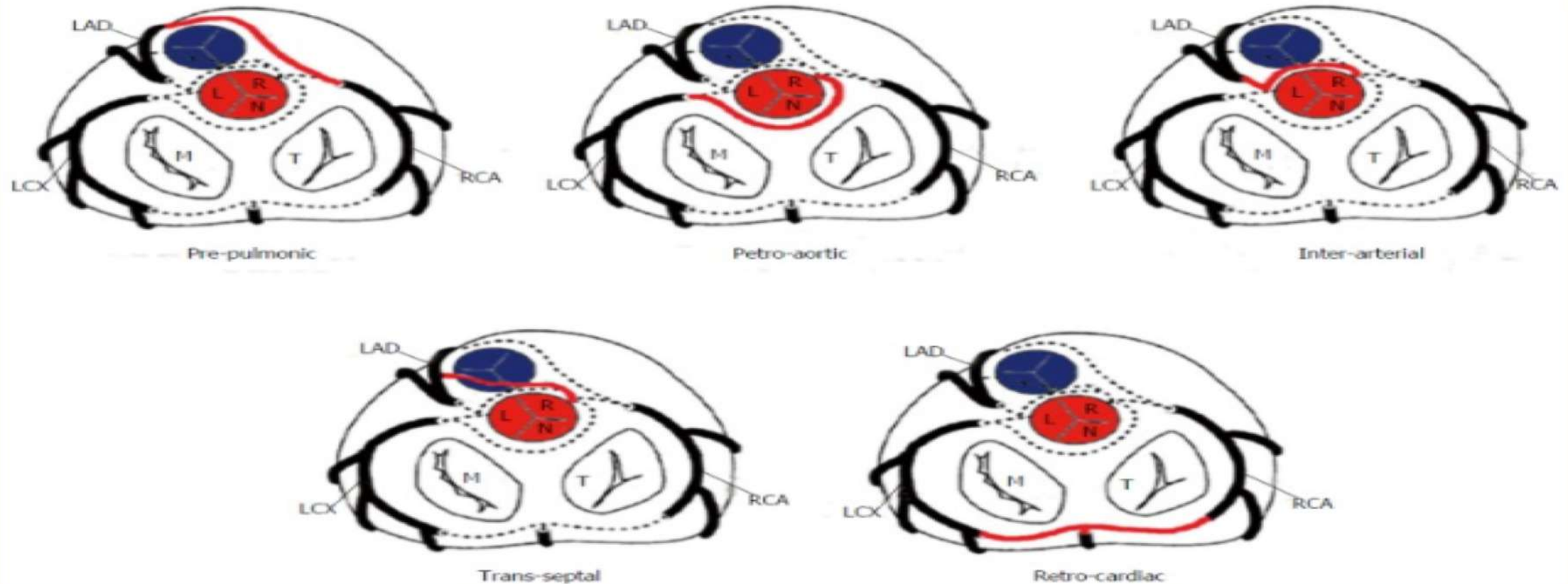
- **Coronary artery anomalies are defined as variations in coronary anatomy present in less than 1% of the population**
- **The true incidence is unknown**
 - **retrospective invasive angiographic studies: 0.3-5.6%**
 - **autopsy studies: 1%**

Both prone to referral bias

Angelini classification

- **Although very uncommon Coronary artery anomalies (CAA) are the second most common cause of exercise related deaths in competitive athletes accounting for 17%. (Commonest HOCCM: 36%)**
- **Sudden Cardiac Death (SCD) is the presentation in half of these cases.**
- **The risk of SCD in middle age or elderly individual with an incidentally discovered coronary anomaly is unclear, but is probably negligible.**

Sub-Types- Depending on the course



- **Benign:**

- Circumflex (LCx) from right sinus or right coronary artery (RCA) **incidence 0.37%**
- Separate left anterior descending artery (LAD) and LCx ostia **incidence 0.41%**

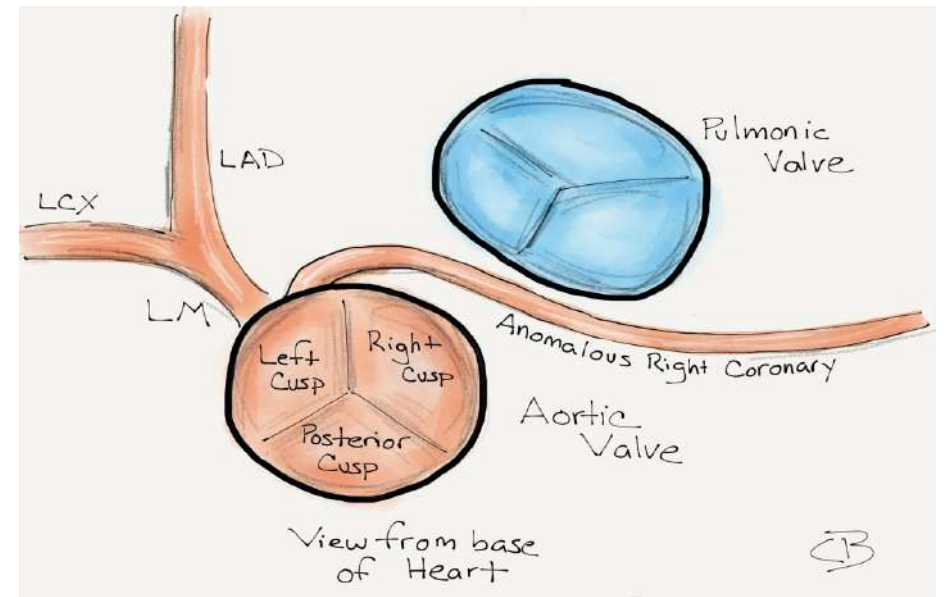
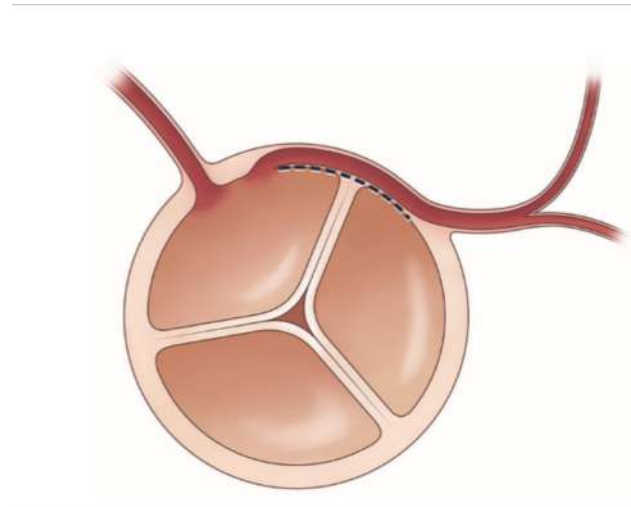
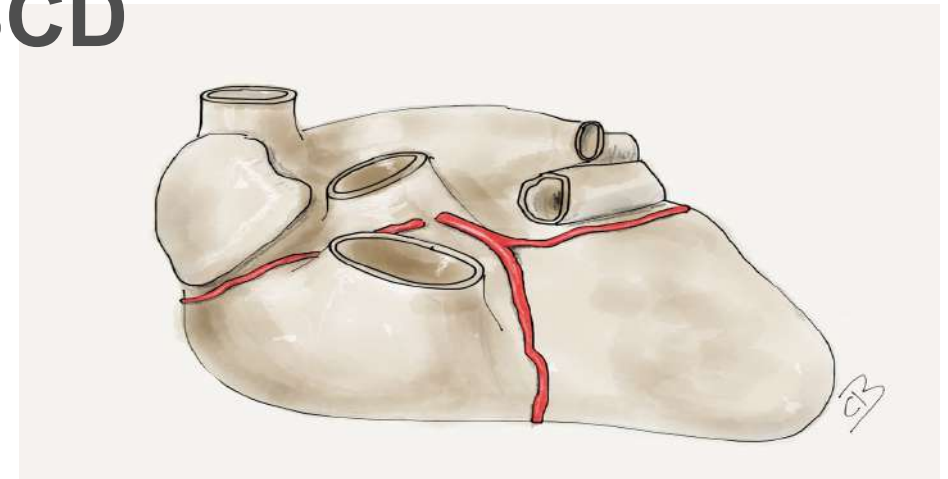
- **Potentially serious:**

- Anomalous left coronary artery from the pulmonary artery (ALCAPA)
- Anomalous right coronary artery from the pulmonary artery (ARCAPA) **exceptionally rare (0.0008-0.008%)**
- Coronary artery fistula
- Anomalous coronary artery from the opposite sinus of Valsalva (left main or LAD from right sinus or RCA from left sinus) **Commonest cause of SCD**

- **Interaortic RCA from L Sinus is 3X more common than Interaortic LCA from R Sinus but has a weaker association with SCD**
- **SCD is preceded by symptoms in only 50% of the subjects**
 - **Chest pain**
 - **Syncope**
 - **Breathlessness**

Features Associated with SCD

- **Interarterial Course**
- **Intramural Segment-** proximal part of an anomalous coronary artery contained within the aortic wall. Coronary artery sharing the aortic wall media without an intervening adventitia
- **Acute angle take off**
-



Mechanism of Ischaemia & SCD

Intense exercise → expand the great vessels → compress the artery between the aorta and the PA → Ischaemia

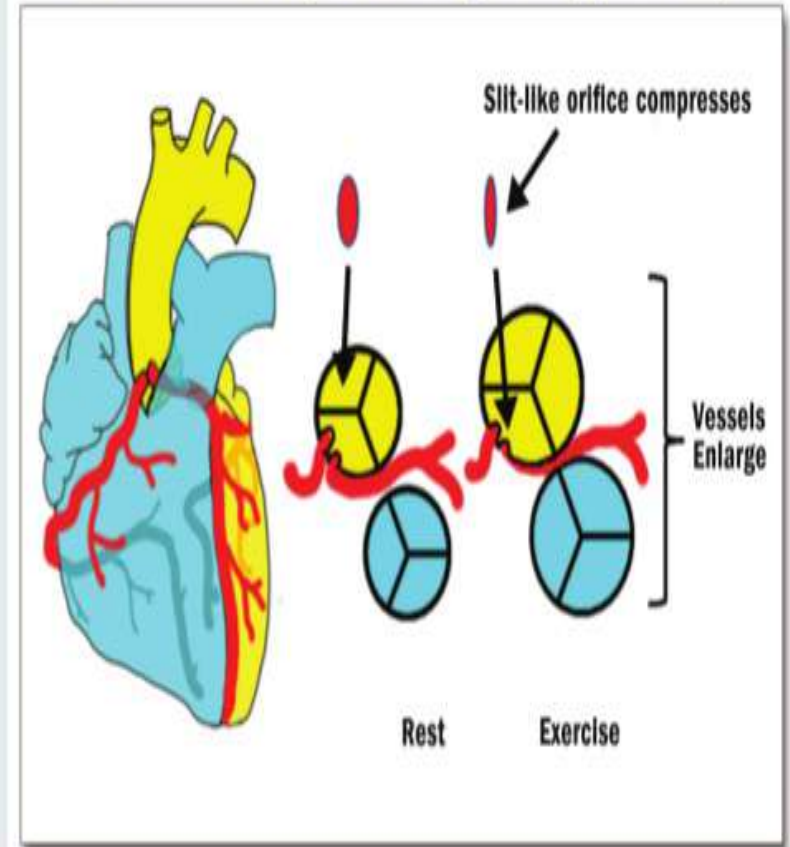
However, in the absence of enlarged PA or PAH, PA is not likely to exert enough pressure to occlude an interarterial coronary artery.

The acute angle takeoff of the intramural coronary artery, can result in a slitlike coronary artery ostium.

Intense exercise → expand the great vessels → stretching the artery in the aortic wall → flaplike closure → Ischaemia

Intramural course with high take off likely to predispose to SCD not inter-arterial course

Postulated Cause of Ischemia Due to Slit-Like Orifice in Patients with Anomalous Origin of Coronary From Opposite Cusp



Bashore, T.M. ACCSAP8, 2012.

Investigation/Diagnosis



- **Cardiac CT**
- **CMR**
- **Stress testing**
- **IVUS with stress**
- **Echo**

26% associated with other abnormalities such as bicuspid valve

Treatment Option

- **Medical Treatment and Observation**
 - **Asymptomatic**
 - **Not intramural**
 - **Anomalous RCA from L sinus**
 - **Age > 35**
 - **No inducible ischaemia**
- **??ICD**
- **Surgery**

ACC/AHA Adult Congenital Guidelines for Management of Anomalous Coronary Origin

Class I

- Surgical coronary revascularization for:
 - Anomalous LM from R sinus with course between Aorta and PA
 - Documented ischemia with anomalous coronary course between the great arteries
 - Anomalous RCA from L sinus with course between the great vessels and documented ischemia

Warnes CA, et al. Circulation, 118:714, 2008.

Surgical Option

Unroofing 28%

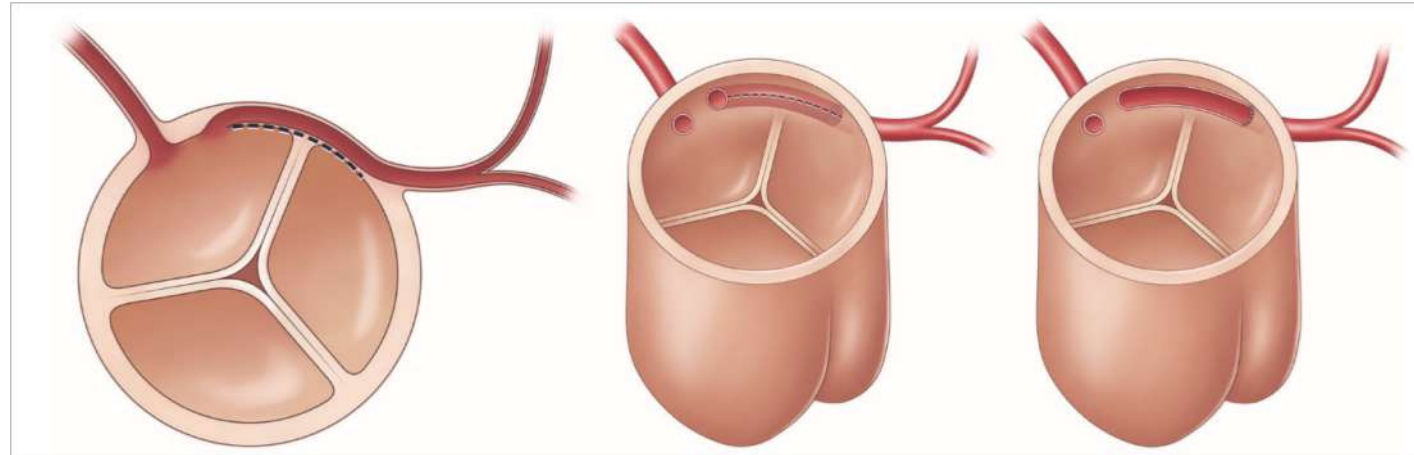
Preferred option with intramural course
No restenosis risk
Complication AR

CABG 38.5%

Problem with competitive flow
CA can be ligated
Good option with presence of atherosclerotic disease

Reimplantation

High risk of ostial restenosis



Summary

- Anomalous CA with an interarterial course although rare is important cause of SCD in particular in young athletes.
- There is significant uncertainty about which are malignant, require follow up or corrective intervention.
- Certain imaging features may confer a higher risk of SCD, such as the LCA involvement, slitlike orifice, intramural segment, and acute angle of origin.
- CT angio and CMR are excellent noninvasive tests for a comprehensive anatomic evaluation.
- Treatment options can be tailored depending on anatomical findings, mode of presentation and presence or absence of inducible ischaemia.

Thank You