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CORONARY ARTERY ANOMALIES

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ABSTRACT
Coronary artery anomalies (CAAs) is a heterogeneous set of condition with varied definition, morphogenesis, clinical presentation, diagnosticworkup, prognosis, and treatment.[1-6]Pathophysiologicalmechanisms and clinical prognoses for differentforms of CAAs is different[7]. Identification of the different CAA's is of utmost importance to predict the prognosis accurately. The present review focuses on the set of anomalies with the origin of a coronary artery from the opposite sinus. This is the subgroup of CAAs that has the mostpotential for clinical repercussions,specifically sudden deathin the young.

Keywords: NIL.

INTRODUCTION

Definition of Coronary Anomalies

Coronary artery anomalies are defined as a constellation of conditions, where there is a variation in one or all the coronary arteries in the origin, course, termination or intrinsic anatomy. Definition of a coronary artery anomaly has two basic steps: (1) Definition of the normal coronary anatomy qualitatively and quantitatively (Table 1)(2)Remaining features should be considered to define an anomaly of coronaryanatomy[7].

TABLE 1: Normal Features of the Coronary Anatomy in Humans

Feature	Range
No. of ostia	2 to 4
Location	right and left anterior sinuses (upper midsection)
Proximal orientation	45° to 90° off the aortic wall
Proximal common stem or trunk	only left (LAD and Cx)
Proximal course	direct, from ostium to destination
Mid-course	extramural (subepicardial)
Branches	adequate for the dependent myocardium

Essential territories	RCA (RV free wall), LAD (anteroseptal), OM (LV free wall)
Termination	capillary bed

LAD indicates left anterior descending artery; Cx, circumflex artery; RCA, right coronary artery; RV, right ventricular; OM, obtuse marginal artery; and LV, left ventricular.

The basic issue in the definition of a normal coronaryarteryis thenormal spectrum of variation. It is important to note here that any form observed in >1% of an unselected general population is considered to be a normal variant by common consensus[9]. However, the field waits a widely accepted endorsement of this view by representative professionalgroups[8]. The fact that myocardial bridges are present in more than 1% of the general population suggests that they may be a normal variant. A basic principle of coronary classification should be that the nature and name of a specific coronary artery are assigned, not according to the site of origin or proximal course, but according to the dependent territory[9] (Table 2).

TABLE 2: ESSENTIAL TERRITORIES OF CORONARY ARTERY

VESSEL	DEPENDENT TERRITORY	NOT ESSENTIAL
RCA	Right ventricular free wall	Posterior descending branch originate from the RCA Ostium of the RCA be located at the right anterior sinus of valsalva
LAD	Anterior inter-ventricular septum	Diagonal branch originate from LAD
LCX	Free wall of LV	Course in left atrioventricular groove Origin of obtuse marginal branch

There is no standardized and comprehensive classification scheme available for CAA's. A useful way to classify the CAA's is according to anomalies of origin, course and termination.

- Anomalies of origin
  - Origin from opposite sinus
  - Origin outside the r or l sinus
  - Separate origin lad lcx
  - origin near the original sinus : low, high, commissural
- Intrinsic anatomical anomalies of course
  - COSA
  - Aneurysms
  - Bridges
- Anomalies of termination
  - Av fistula

Incidence of Coronary Artery Anomalies

Various studies report the incidence of CAA's from 0.9% to 5.6% is general population[9]. In a prospective analyses performed in a continuous series of 1950 patients studied by coronary angiography, Angelini et al found that CAAs had a global incidence of 5.64%. They reported 0.92% incidence of anomalous origination of the RCA from the left sinus and the0.15% incidence of anomalous origination of the

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left coronary artery from the right sinus (for a total incidence of 1.07%for ACAOS)[9] (Table 3)

TABLE 3: Incidence of Coronary Anomalies and Patterns, as observed in a Continuous Series of 1950 Angiograms[9]

Variable	N (%)
Coronary anomalies (total)	110 (5.64)
Split RCA	24 (1.23)
Ectopic RCA (right sinus)	22 (1.13)
Ectopic RCA (left sinus)	18 (0.92)
Fistulas	17 (0.87)
Absent left main coronary artery	13 (0.67)
Circumflex arising from right sinus	13 (0.67)
LCA arising from right sinus	3 (0.15)
Low origination of RCA	2 (0.1)
Other anomalies	3 (0.27)
Coronary dominance patterns Dominant RCA	1641 (89.1)
Dominant LCA (circumflex)	164 (8.4)
Codominant arteries (RCA, circumflex)	48 (2.5)

In a study of 12457 patients by Yamakana et al[10], coronary artery anomalies were found in 112 patients (0.9% incidence), 100 patients (89.3%) had origin and distribution anomalies, and 12 patients (10.7%) had coronary artery fistulae. Separate origins of left anterior descending and left circumflex coronary artery from the left sinus of Valsalva was the most common anomaly (63.4%). The right coronary artery rising from the left coronary sinus of Valsalva was found in 10 (8.9%) patients. Anomalous origin of the left circumflex coronary artery from the right sinus of Valsalva was seen in 10 (8.9%) patients. The left main coronary artery from the right coronary sinus of Valsalva was found in 1 (0.89%) patient while an isolated single coronary artery was seen in 2 (1.78%) patients.

In comparison, Drory and colleagues[11] studied the incidence of CAAs in acontinuous series of 162 patients with sudden unexpected death. The incidence of CAA related sudden death was 0.6% (1 of 162 cases). This result suggests that extreme exercise plays a powerful role in such deaths.

The incidence at our center in a study of 9670 consecutive angiograms was 3.47%. Various anomalies and their incidence is summarized in Table 4.

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TABLE 4: Incidence of Coronary Anomalies and Patterns, as observed in a Continuous Series of 9,670 Angiograms at our centre

Type	N (%)
LMCA from RSV	1 (0.01)
Separate origins of LAD and LCX in LSV	21 (0.21)
LCX from RCA or RSV	35 (0.35)
RCA from High posterior RCS	67 (0.69)
RCA from LSV	33 (0.34)
Single coronary artery	4 (0.04)
Origin of Lcx from mid LAD	1 (0.01)
Intercoronary communications and	2 (0.02)
Ectopic origin from the pulmonary artery	1 (0.01)
Coronary artery fistulae	13 (0.13)
Myocardial Bridge LAD	158 (1.61)
Myocardial Bridge RCA	2 (0.02)
Myocardial Bridge LCX	1 (0.01)

Clinical significance and pathophysiology

Most CAA's are unable of interference with the coronary artery's function to provide adequate blood flow to the dependent myocardium. Only the CAA's capable of such interruption become of clinical significance by causing ischemia and rarely SCD. Most CAA's are free of ischemia (80%).Some CAAs may cause occasional ischemia, others (e.g.,anomalous origination of the left coronary artery from the pulmonary artery) obligatorily cause ischemia, and yet othersonly predispose the patient to have a misdiagnosis or complications(clotting, spasm, or atherosclerotic buildup) (Table 5).

TABLE 5: Clinical classification of CAA's

TYPE OF ISCHEMIA	**CORONARY ANOMALY**
Absence of ischemia	Majority of anomalies (split RCA, ectopic RCA from right cusp, ectopic RCA from left cusp
Episodic ischemia	Anomalous origin of a coronary artery from the opposite sinus (ACAOS); coronary artery fistulas
Obligatory ischemia	Anomalous left coronary artery from the pulmonary artery (ALCAPA)

There are the varied mechanisms of ischemia in CAA. Slit-like coronary orifice, acute angle of take-off from aorta, inter-arterial course between aorta and PA, spasm secondary to endothelial damage have all been thought to contribute to ischemia. Recently, aortic intramural intussusceptionhas been quoted as a

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predominant mechanism. The reasons for insistence on the intussusceptions of anomalous arteries are related to the following newly discovered mechanisms of stenos is:

1. Coronary hypoplasia. Intramural intussuscepted segment of the proximal ectopic artery is smaller in circumference than the more distal extramural vessel. Arteries that arise congenitally inside the aortic media likely cannot grow normally either before or afterbirth[12].
2. Lateral compression. The cross section of the intramural segment is characteristically not circular but ovoid. The lateral compression results in a smaller area than that possessed by a circle of the same circumference. This parameter can be quantified with the asymmetry ratio (the ratio of the smallest to the largest diameter in an IVUS cross section) [13].The smaller diameter is further compressed during each systole.

Aortic walldistensibility (degree of cross-sectional area enlargement associated with a certain increase in pressure) is a further important variable that depends on intrinsic anatomic changes inthe aortic wall. Conditions predisposing to an acute onset reduction in aortic compliance i.e. exercise, new onset hypertension, further predisposes to intramural intussusceptions leading to ischemia and SCD. When a carrier of ACAOS dies suddenly, in the absence of other lethal cardiovascular conditions, a low cardiac output and bradycardia or asystole typically occur early after extreme exercise, after which syncope and/or death ensues. Terminal ventricular fibrillation may also occur as a manifestation of critical ischemia or of reperfusionarrhythmia[14-16].

Apart from SCD other clinical manifestations of ACAOS dyspnea, palpitations, angina pectoris, dizziness, and syncope[16]. Whereas sudden death is usually associated with extreme exercise in young adults[17], the other manifestations of ACAOS are more frequently seen in older adults (specifically women) and are related to the onset of hypertension.

Outlines for Diagnostic and Treatment Protocols

Most patients are asymptomatic for a large portion of their lives, and an atypical chest-pain syndrome is the most common reason they are referred forcoronary angiography, which is when the diagnosis is typically made. For most types of coronary anomalies, the fundamental clinical approach should be masterly inactivity. CAA's known to be malignant, particularly ACAOS, should be adequately approached.

HOW TO APPROACH A PATIENT OF ACAOS?

Testing should sequentially include electrocardiography, Holter monitoring (basically to document atrial or ventricular arrhythmias as nonspecific markers of ACAOS), and focused expert echocardiography (transthoracic and, if needed, Trans esophageal) with Doppler interrogation to identify the coronary origin and proximal course[18-20], followed by CT coronary angiogram. Diagnosed cases of ACAOS should be subjected to stress testing to document ischemia. Additional obstructive coronary disease of atherosclerotic origin should be ruled out.

TABLE 6: APPROACH TO A PATIENT WITH R-ACAOS

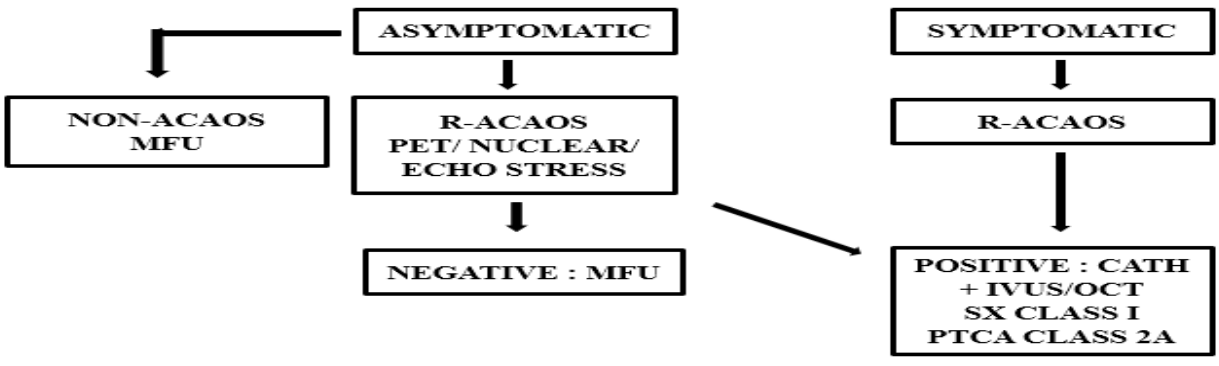

TABLE 7: APPROACH TO A PATIENT WITH L-ACAOS

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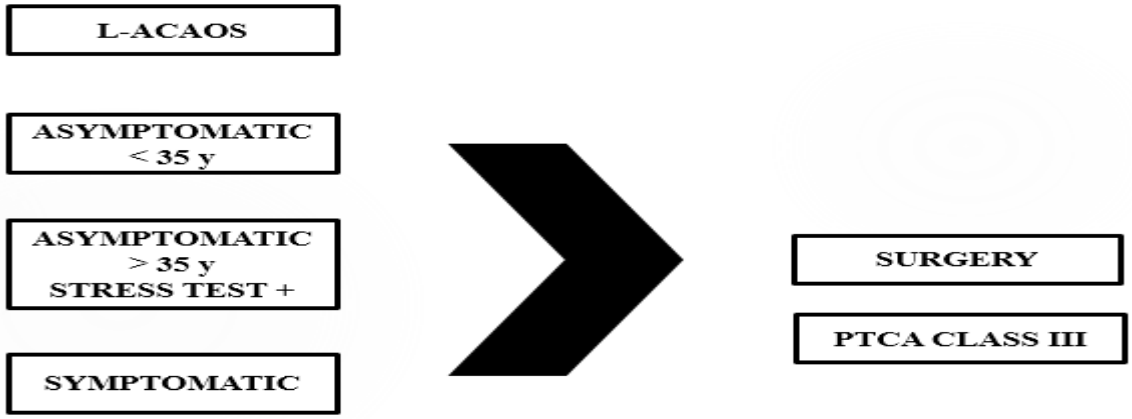


Treatment Options

Symptomatic patients of ACAOS have 3 treatment options: medical treatment, coronary angioplasty withstent deployment, and surgical repair. Intervention may be justified in some cases to prevent sudden death and improve the quality of life despite non availability of long term follow up data.

Medical treatment includes heart rate control with beta blockers an activity restriction. Beta blocker therapy is probably as effective as restriction of activity (avoidance of severe exertion) in these patients[39].

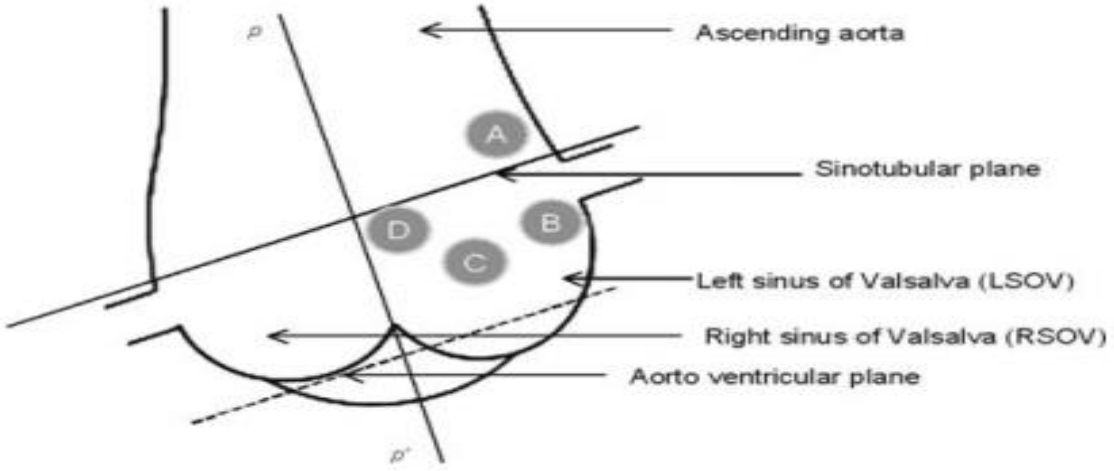
PERCUTANEOUS INTERVENTION

PCI is feasible in symptomatic patients with R-ACAOS or with evidence of ischemia on stress testing. In patients of L-ACAOS surgery is preferred. In the setting of primary PCI, stent implantations have been done in both the subset of patients.PCI of right-ACAOS is indicated in the presence of (1) disabling symptoms and/or a high risk of sudden death, (2) area stenos is more severe than 50% with respect to the distal normal vessel on IVUS, (3) a large dependent myocardial territory (more than a third of the total), and (4) reversible ischemia, as documented by anuclear stress test[42].

PCI in R-ACAOS

The operator faces many technical problems during angioplasty of lesions in ACAOS. The commonly encountered problems include ostial identification, optimal guide catheter selection, support during PCI and hardware tracking in the anomalous artery.

Sarkar et al have classified the anomalous R-ACAOS ostia according to relation to left sinus into 4 types[21].



A rough guide to catheter selection is given according to the type of ostia[21] (Table 8).

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TABLE 8: Guide to catheter selection in R-ACAOS[21]

TYPE A	FL, FCL
TYPE B	FCL, VL
TYPE C	VL, FCL
TYPE D	AL, JR

Appropriate guide selection is the crux to a successful PCI in R-ACAOS. An ideal guide will provide excellent support and co-axial engagement. However, in view of the aberrant anatomy generally guide support is poor.

Various alternative methods are used to improve support during the procedure:

1. Two wires can be used. Use of extra-support wires further help in improving support.
2. Balloon anchorage to one of the side branches in stabilizing the system and providing support.
3. Manual under sizing of Judkins left catheter helps in making the catheter more co-axial and improving support.
4. Steam deformation helps in under sizing the catheters and maintain the shape during the procedure.
5. Special catheters like Ikari left maybe used when routine catheters fail to provide adequate support

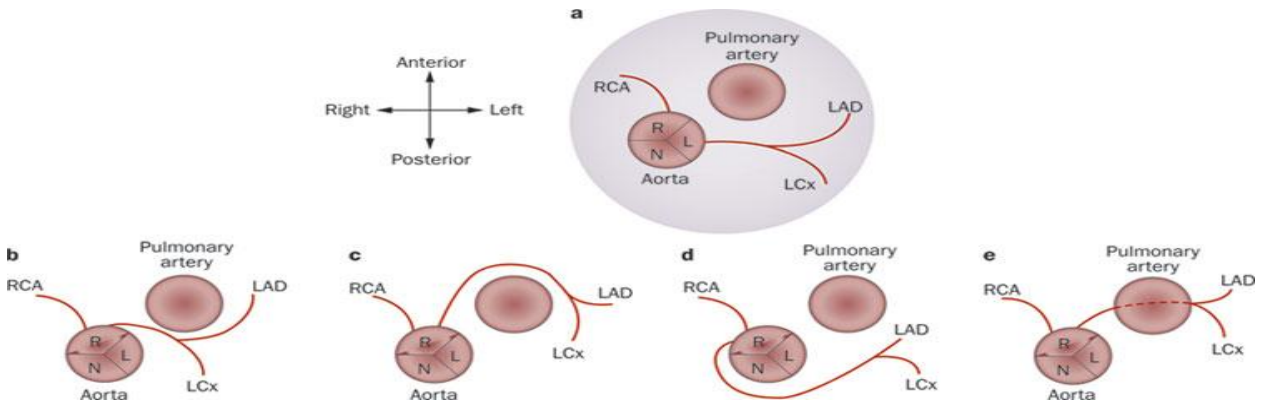
Immediate and late results are improved if full luminal restoration, to match the area of the distal vessel, is attained at the intramural segment and for about 4 mm beyond it. Drug-eluting stents offer the best probability to avoid restenosis, but definitive data need to be collected regarding this off-label use of stents. Moreover, restenosis appears to be rare, and, if it does occur, is related to in-stent fibro cellular growth, not stent compression.

PCI in L-ACAOS

These cases should be routinely treated with surgical intervention. However, PCI have been done in the setting of STEMI.

There are 4 well defined sub-types according to the course of the anomalous artery:

- Inter-arterial : SCD 50%
- Anterior free-wall course
- Retro-aortic course
- Septal course-most common



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Identification of the coronary course during cardiac catheterization involves angiogram in RAO projection to adequately separate aorta and pulmonary artery.

A "DOT" is seen in retro-aortic and interarterial course. Identification of this is important as these two groups have extremely high risk of SCD, and should undergo surgical repair. An "EYE" is seen in septal and anterior course, both of which have much lower risk of SCD, and can be undertaken for PCI.

Surgical correction, which is especially recommended for left-ACAOS that involves a large territory at risk, may consist of (1) direct reimplantation of the ectopic artery at the aortic root (2) unroofing of the intramural coronary segment, from the ostium to the exit point, off the aortic wall; or (3) osteoplasty, which creates a new ostium at the end of the ectopic artery's intramural segment. [22-24]

Conclusions

Coronary artery anomalies comprise a very heterogeneous group with diverse pathophysiology and clinical presentation. Most of the anomalies are benign. However, a few malignant anomalies are known to cause SCD. IVUS is the

Preferred means to evaluate the mechanisms responsible forischemia in potentially significant CAAs, especially ACAOS. R-ACAOS patients can undergo PCI. However, surgery is preferred in cases of L-ACAOS.

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