

# Coronary Ectasia: Prevalence, Angiographic Characteristics and Clinical Outcome

Mohamed Ztati \*, Rim Zerhoudi, Imane Katif, Abdelkarim Ait Yahia, Mohammed El Jamili, Mustapha Elhattaoui

Cardiology Department - ERRAZI Hospital - Mohammed VI University Hospital, Marrakech.

\*Corresponding Author: Mohamed Ztati, Cardiology Department - ERRAZI Hospital - Mohammed VI University Hospital, Marrakech.

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## Abstract

**Background/Aim:** Coronary artery ectasia (CAE) is defined as diffuse dilation of the coronary arteries and differs from coronary aneurysms by being non-localized. The condition is rare, often associated with atherosclerosis, and poses significant management challenges due to its unclear pathophysiology and varied clinical presentations. The study aims to provide insights into the hospital's experience with CAE and evaluate its prognostic factors.

**Materials and methods:** This retrospective study evaluated 28 patients with CAE from 1692 individuals who underwent coronary angiography between January 2021 and January 2024. Patients with other heart conditions were excluded. CAE diagnosis was based on coronary artery dilation  $\geq 1.5$  times the adjacent normal segment. Data on clinical characteristics, myocardial perfusion, and stenosis were collected, and coronary lesions were classified using the Markis classification system.

**Results:** CAE was observed in 1.65% of the study population, with most patients being male smokers, averaging 55 years of age. Of the CAE patients, 74% had significant lesions, 59% had multi-vessel involvement, and 25% presented with spontaneous thrombosis. Left ventricular dysfunction was noted in 60% of patients. Pharmacologic treatments included aspirin for all, while 9 patients also received anticoagulation therapy. Percutaneous coronary intervention (PCI) was conducted in select cases with thrombus formation or significant stenosis.

**Conclusion:** CAE remains a poorly understood condition with a significant thrombotic risk. Atherosclerosis was the predominant cause, and management strategies, especially for thrombosis, remain individualized. Optimal treatment is still unclear, and large multicenter studies are needed to develop better treatment guidelines for CAE patients.

**Keywords:** coronary artery ectasia; atherosclerosis; thrombosis; percutaneous coronary intervention

## Introduction

Coronary artery ectasia (CAE) is characterized by the dilation of the coronary artery lumen. The first case of coronary aneurysm was reported by Charles Bougon in 1812 [1], yet this pathology remains poorly studied and poorly understood. Coronary ectasia is generally defined by consensus as a dilation of the coronary artery diameter exceeding 1.5 times its normal diameter [2, 5]. In contrast, the definition of coronary aneurysms is more variable: they are considered localized ectasias, with criteria varying based on the degree of dilation and the length of the affected segment (dilation exceeding 1.5 or 2 times the normal caliber, a pathological segment representing less than one-third or one-half of the vessel length, or simply described as localized) [3,4,5].

Some authors suggest that aneurysms and ectasias are merely angiographic variations of the same pathology. The most widely used classification for assessing their extent remains the one proposed by Markis et al. [5], with variants that include multi-vessel forms, encompassing both aneurysms and ectasias. CAE is a relatively rare and poorly understood condition. Although its pathophysiology remains unclear, atherosclerosis appears to be the underlying mechanism in most

cases [2,3]. Here we provide our hospital experience in managing such coronary heart disease with an overview and status report on CAE.

## Materials and methods:

This study was a retrospective study that evaluated patients who underwent coronary angiography and presented with varying degrees of coronary artery ectasia (CAE) between January 2021 and January 2024. Inclusion criteria were adults with a confirmed diagnosis of CAE based on coronary artery dilation  $\geq 1.5$  times the adjacent normal segment, and complete clinical and angiographic data availability [3,4]. Exclusion criteria included valvular heart disease, cardiomyopathy, and congenital heart defects to eliminate CAE as a secondary condition.

Starting in January 2021, patients diagnosed with coronary ectasia (CE) were identified and subjected to biannual clinical evaluations. CAE was characterized by a coronary artery dilation exceeding 20 mm in length and a diameter  $\geq 1.5$  times (3) at of the adjacent normal segment. In the absence of a normal segment for comparison, an estimated "normal" caliber was determined through consensus between a hemodynamic

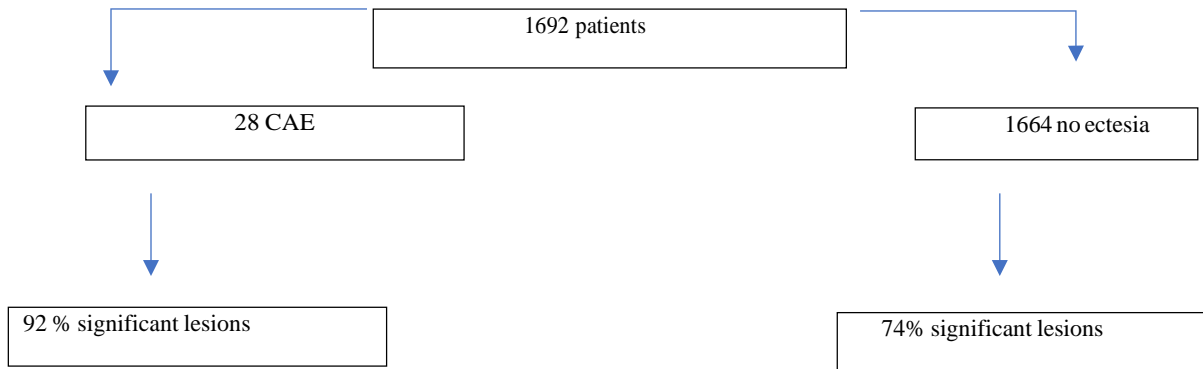
specialist and a lead physician. Study inclusion was based on a collective decision-making process.

Post-visual assessment, quantitative data were collected. For each case, the maximum diameter of the dilated artery and corresponding healthy segments were measured. Myocardial perfusion was quantified using the Thrombolysis in Myocardial Infarction (TIMI) classification and the TIMI Frame Count methodology. Stenosis was operationally defined as a luminal reduction of  $\geq 70\%$  relative to the normal reference segment [6].

A descriptive analysis of the study population was performed. Quantitative variables were presented as medians and ranges, while qualitative variables were expressed as counts and percentage.

**Results:**

Between January 2021 and January 2024, 1692 patients underwent coronary angiographies, among who 28 presented with CAE, which represents a prevalence of 1,65% of our total patients (**figure. 1**).



**Figure 1:** Analysis of patients including distribution with respect to presence or absence of coronary artery ectasia and angiographically significant stenosis

**Clinical characteristics:**

Characteristics of patients with and without CEA appear in: Most of those with CEA were men, smokers, with an average age of 55 years old markedly below that of patients without CE (62 years old).

Of the 27 patients, 22 suffered MI, and 16 of whom had STEMI. patients 59% had multi-vessel CAE. The laboratory data of all patients were also showed in **Table 1**.

<i>Patients With Cae (N= 27)</i>	
<b>AGE, MEAN (SD), (YEARS)</b>	55
<b>MALE, COUNT (%)</b>	237 (82.9)
<b>HYPERTENSION, COUNT (%)</b>	142 (49.7)
<b>DIABETES, COUNT (%)</b>	63 (22.0)
<b>CURRENT SMOKING, COUNT (%)</b>	147 (51.4)
<b>PRIOR MI, COUNT (%)</b>	101(35.3)
<b>HYPERLIPIDEMIA, COUNT (%)</b>	130 (45.5)
<b>CLINICAL PRESENTATION, COUNT (%)</b>	
STABLE ANGINA	6 (2.1)
UNSTABLE ANGINA	103 (36.0)
STEMI	22
NSTEMI	5
<b>LABORATORY DATA</b>	
HS-CRP, MEDIAN (Q1,Q3), (MG/L)	24,5
TC, MEAN (SD), (MMOL/L)	4.24 (1.11)
TG, MEAN (Q1,Q3), (MMOL/L)	1.44(1.07–1.95)
HDL, MEAN (Q1,Q3), (MMOL/L)	1.08 (0.93–1.28)
LDL, MEAN (SD), (MMOL/L)	2.78 (1.00)
CREATININE, MEDIAN (Q1,Q3), (MMOL/L)	79.00 (72.00–94.00)
GLYCATED HEMOGLOBIN, <u>MEAN (%)</u>	6,1
<b>MULTIVESSEL, COUNT (%)</b>	59

**Table 1:** Clinical and biological outcomes

**Echocardiography and coronary angiogram results:**

Transthoracic echocardiography was performed after in-hospital admission and at a median of 2 days after AMI. All images were analyzed

by a single investigator, who was blinded to all clinical data. The coronary angiography was performed using a radial artery approach or a femoral artery approach, and each image was interpreted by two independent cardiologists.

60% of the CAE patients admitted presented a LV dysfunction: (<50%), with only one patient exhibiting severe left ventricular dysfunction with an ejection fraction of 28%.

Most patients with ST-segmental elevated myocardial infarction (STEMI) received an emergency percutaneous coronary intervention (PCI) as part of reperfusion therapy within 12 h of the onset of symptoms. For most

patients with non-STEMI, initial antithrombotic therapy was instituted, and subsequent coronary angiography (delayed PCI) was performed within 48 hours.

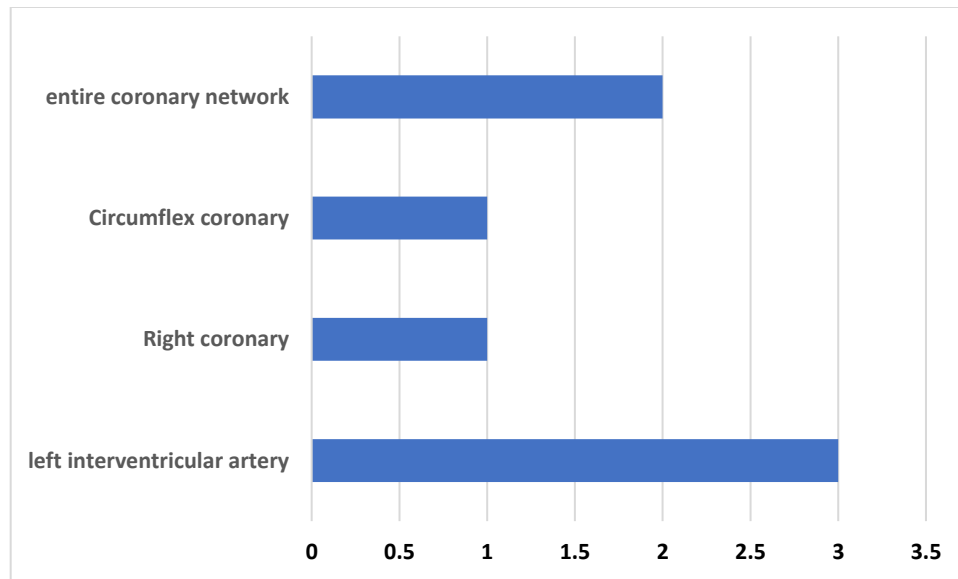
According to the Markis classification method CAE could be classified into four groups based on the extent of ectasia in the coronary arteries: Markis type I, diffuse ectasia of two or three vessels; Markis type II, diffuse disease in one vessel and localized disease in another vessel; Markis type III, diffuse ectasia of one vessel only; and Markis type IV, localized or segmental **ectasia (Table 2).**

Markis Classification	TYPE I	TYPE II	TYPE III	TYPE IV
Patients (%)	59%	9%	24%	8%

**Table 2:** Markis classification of the patients

Right coronary artery (RCA) lesions were the most common, accounting for 70% (n = 19 of CAEs) CAE was more likely to occur in the proximal segment (n = 15) of the three coronary arteries on coronary angiography. Regarding the shapes of CAE lesions, we found that spindle-shaped CAEs

were most common in all coronary vessels, while the diffuse CAEs is more frequently located in the RCA and the LCX arteries leading to inferior STEMI, and the scrotiform CAEs most occurred in the LAD artery. It showed that coronary tortuosity was observed in 38% of all coronary arteries (**Figure.2).**



**Figure 2.** Analysis of coronary involvement in patients with artery ectasia

A total of 44.08% CAE segments had more than two coronary collateral circulations. CAEs in the LCX artery were more likely to associate with coronary collateral circulation, although there were insignificant differences in RENTROP scores among the vessels. What's more, based on the qualitative comparative analysis, there existed insignificantly statistical difference in the minimum lesion diameter, maximum lesion diameter, or the diameter stenosis rate in the expansion section among the vessels.

For pharmacologic management of CAE: Proposed therapies include Antiplatelets as the standard treatment: aspirin was administered in all our patient [27]. Anticoagulants for secondary prevention: Warfarin has been proposed as a treatment since ectatic coronary arteries are prone to thrombosis, dissection, and spasm but was only given to 9 patients after a consultation of the heart team (patient with thrombus formation, multivessel ectasia high thrombotic burden.)

Lipid-lowering medications, ACE inhibitors and Beta-blockers was also administered

For non pharmacologic management : the Percutaneous treatment of CAE is a valuable option in patients with suitable anatomical and clinical

features. However, PCI of ectatic and aneurysmatic lesions presents several challenges that's why it was only performed in only 4 patients with Thrombus formation and in 8 patients with concomitant obstructive lesions or patients exhibiting symptoms or signs of myocardial ischemia despite adequate pharmacologic treatment.

The etiological investigation had demonstrated only 2 cases of inflammatory or connective tissue diseases : 1 behçet dease in a 42 years old man and a LED in a 34 years women , the Atherosclerosis was the predominant etiology in the remaining patients.

**CAE with thrombus formation (figure 3,4,5):**

7 (25%) of our patient presented with spontaneous thrombosis :57% had diffuse ectasia in one vessel (RCA), 28% had multivessel ectasia, and only 1 had diffuse ectasia in LVA and localized disease in RCA and CX.

4 patients had PCI: -The decision to attempt primary percutaneous coronary intervention (PCI) on the RCA was based on an electrocardiogram and coronary angiogram. Due to the angiographical evidence of a heavy thrombus burden, we attempted PCI using repetitive

coronary aspirations with a 6-French Export Catheter (Medtronic, Minneapolis, MN) for thrombus extraction.

However for 2 patients: the lesion was resistant to repetitive aspirations and the blood flow was only partially restored in the second obtuse marginal branch (OM2) with Thrombolysis In Myocardial Infarction

grade Consequently, the patient received tirofiban (10 µg/kg body weight) through guiding catheter after thrombus aspiration, and following 36 hours of intravenous infusion (0.15 µg/kg/min).

2 have benefited of CABG, and only 1 was put under medication therapy only : Warfarine, aspirine for the long run.

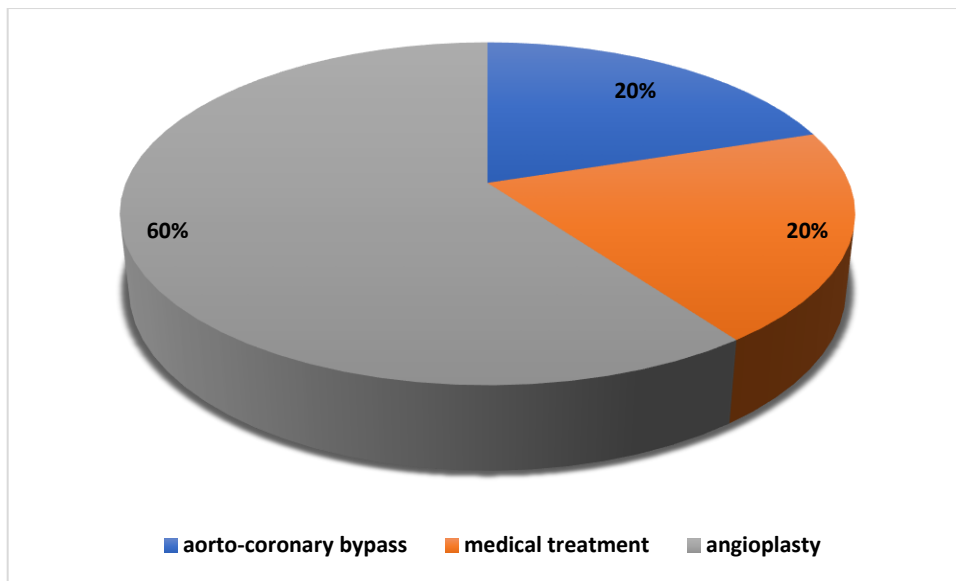


Figure 3: Treatment of CAE with thrombus formation

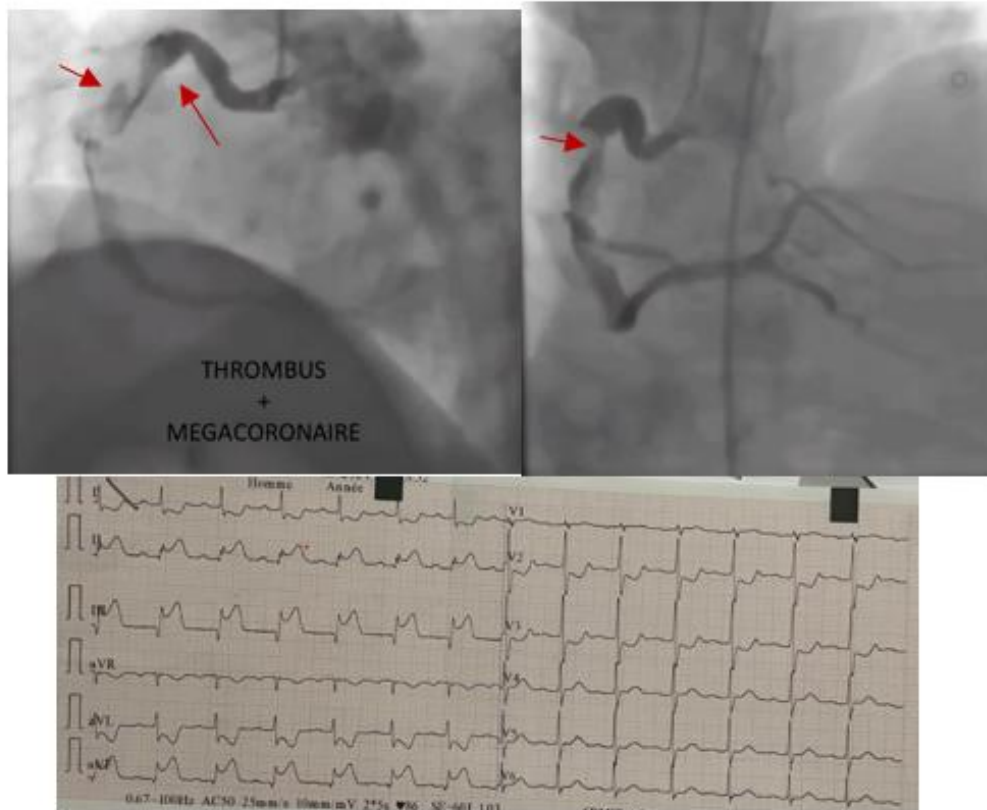
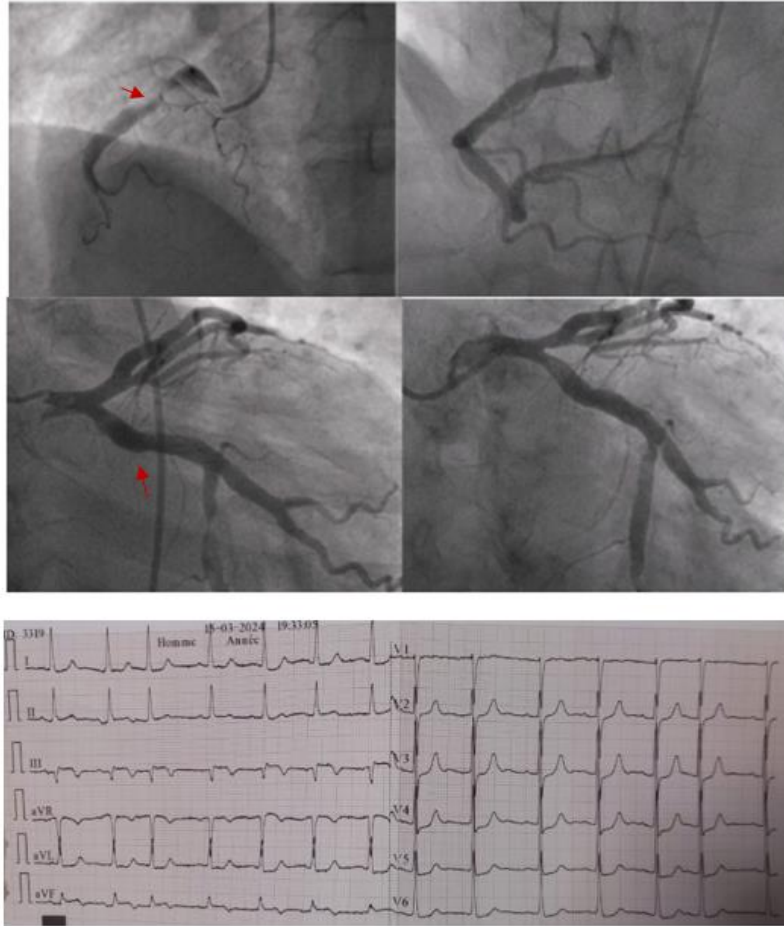


Figure 4: This case involves a 67-year-old patient with risk factors including hypertension, diabetes, dyslipidemia, and active smoking at 20 pack-years, presenting with a non-complicated inferior STEMI, 12 hours post-onset of pain. Emergency coronary angiography revealed significant stenosis of the right coronary artery with flow slowdown and an extensive thrombus extending along the entire artery, which is ectatic with a diameter measuring 8 mm.

The patient received dual antiplatelet therapy (DAPT) with loading doses of clopidogrel and aspirin, along with a heparin bolus in the catheterization

lab, agrastat, and thromboaspiration. After 48 hours of Tirofiban, a follow-up angiogram showed restored TIMI 3 flow and significant arterial



**Figure 5:** A 58-year-old patient with no cardiovascular risk factors presented with an inferior STEMI 36 hours after symptom onset. Coronary angiography revealed a 90% stenosis in the circumflex artery and a thrombotic stenosis in the right coronary artery (TIMI 2 flow). Following 48 hours of Agrastat treatment, TIMI 3 flow was restored, and the circumflex artery lesion was treated with an active stent.

### Discussion:

Coronary artery ectasia (CAE) is a rare abnormal aneurysmal dilatation of the coronary arteries. Its prevalence ranges from 0.85 % to 5.3 % according to prior studies [7] which match our rate of 1.5%. It is described as segmental dilatation with a diameter 1.5 times that of the normal coronary artery next to it [8] and is differentiated from coronary artery aneurysms by its dilatation that exceeds 1/3 the length of the coronary vessel.

Atherosclerosis is considered to be the most frequent (50%) cause of CAE, it represented 92 % of the patients in our study. Only 10% to 20% of cases of CAE have been reported resulting from inflammatory or connective tissue diseases, the rate in our study was clearly below this with only 7% of our patient that was diagnosis with auto immune disease.

A study by Liu et al demonstrated that in patients with CAE and atherosclerosis followed for 1 to 16 years, CAE progression was minimal, while atherosclerosis progressed and the extent of ectasia was related to the degree of stenosis. The results indicate that prevention and treatment of atherosclerotic changes may be of greater clinical importance than treatment of ectatic changes [9].

CAE commonly presents with stable angina. However, STEMI can be caused by distal embolisation or a thrombus occluding an ectatic segment of coronary artery, as in this case [7]. Local thrombus formation due to stagnant local blood flow has been suggested as a cause of coronary

thrombosis in severe CAE presenting with acute myocardial infarction [10]. 25% of our CAE patients developed a spontaneous thrombus,

Clinical observations have demonstrated that thrombosis, embolization, and rupture of the involved segments are the leading causes of AMI and sudden death in patients with CAE [11]. In recent years, optical coherence tomography (OCT) has emerged as the most accurate method for intracoronary evaluation. Owing to a resolution of 10 to 20  $\mu\text{m}$ , OCT is more accurate than intravascular ultrasound (IVUS) and is useful for the evaluation and characterization of plaque features, both in stable and acute coronary artery disease, unfortunately it wasn't performed in our center due to its unaffordability.

The optimal management of CAE has not been firmly established [12]. Proposed therapies include aspirin and chronic anticoagulation. A comprehensive review and meta-analysis by Moghadam et al [13] revealed that patients with CAE had a significantly higher mean platelet volume than healthy individuals, indicating that platelet thrombotic effects may play a role in the genesis of CAE. As a result, antiplatelet drugs like aspirin may be useful in the treatment of this condition. All of our 27 patients were put under aspirin. Due to the susceptibility of ectatic coronary arteries to thrombosis, dissection, and spasm, anticoagulants such as Warfarin have been recommended as a treatment. Case reports for the efficacy of novel anticoagulants like rivaroxaban, dabigatran, and apixaban are also published which shows efficacy of novel oral anticoagulants (NOAC) in the treatment of the thrombosed ectatic coronary arteries [14]. 72% of our patient were put under anticoagulation

treatment on the other hand Nitrates can increase epicardial dilation, there by exacerbating myocardial ischaemia and causing angina pectoris in patients with CAE. [13] As such, nitrates should be avoided in patients with isolated CAE.

For those that present a spontaneous thrombus Percutaneous treatment of CAE is a valuable option. However, PCI of ectatic and aneurysmatic lesions presents several challenges, starting from lack of specific indications. PCI has several technical challenges that must be carefully balanced into decision-making process when considering the optimal revascularization strategy. In the setting of an ACS with an EIRA, high thrombus burden with distal embolization and microvascular damage substantially increases the technical complexity of PCI, with a high risk of procedural failure and adverse events at long term [15]. Given the high thrombus load, several reports showed a wider use of glycoprotein IIb/IIIa inhibitors and thrombus aspiration in patients treated with primary PCI of an EIRA, ATL is then recommended only in patient with CEA diameter < 5 mm, for the 7 patients that presented with similar diagnosis, only 1 had an ATL, the other 6 were put under medical therapy on the long run + glycoprotein IIb/IIIa inhibitors with a satisfactory angiographic control.

In absence of specific recommendations, management strategies are still individually tailored according to clinical presentation, anatomical features, and procedural complexity.

### Limitations:

A significant limitation of the current evidence on the management of CAE is that most studies have focused on symptomatic patients, often presenting with acute coronary syndrome (ACS). Conversely, there is a notable lack of data regarding asymptomatic patients with incidentally detected CAE and no evidence of significant coronary artery disease (CAD).

### Conclusion:

Coronary artery ectasia (CAE), characterized by a diffuse or focal dilation of an epicardial coronary artery, is observed in up to 5% of patients undergoing coronary angiography. The clinical presentations of CAE are varied, ranging from asymptomatic individuals to high-risk patients with acute coronary syndrome (ACS) Increasing evidence indicates a heightened risk of long-term adverse events in patients with CAE, particularly those with ACS. Managing these patients is complex, with treatment options encompassing optimal antithrombotic therapy, surgical interventions, and percutaneous procedures such as drug-eluting stent (DES) implantation, covered stent exclusion, or stent-assisted coil embolization.

Despite advances in understanding CAE in recent years, critical issues concerning its natural history and optimal treatment remain unresolved. Large multicenter studies are essential to provide clinicians with comprehensive guidance for managing this complex patient population.

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