

ALCAPA SYNDROME

Anomalous Left Coronary Artery
from Pulmonary Artery

Conflict of interest

I have nothing to declare

History

A 14 years old girl .

Chief complaint:

Chest pain and dyspnea for 2 days before admission.

Misdiagnosed as a case of musculoskeletal chest pain , after deterioration , she referred to heart center for further evaluation and treatment .

CAD risk factors:

No CAD risk factors, no family history of premature coronary artery disease or congenital heart disease.

PHYSICAL EXAMINATION :

Sinus tachycardia; PR=120 b/m.

BP 110/70.

Clear chest.

ECG:

Sinus tachycardia, ST depression in lateral and inferior leads.

CHEST X RAY: Normal cardiothoracic ratio.

Investigations :

Hb =13g

WBC=11,500

Elevated serum Troponin

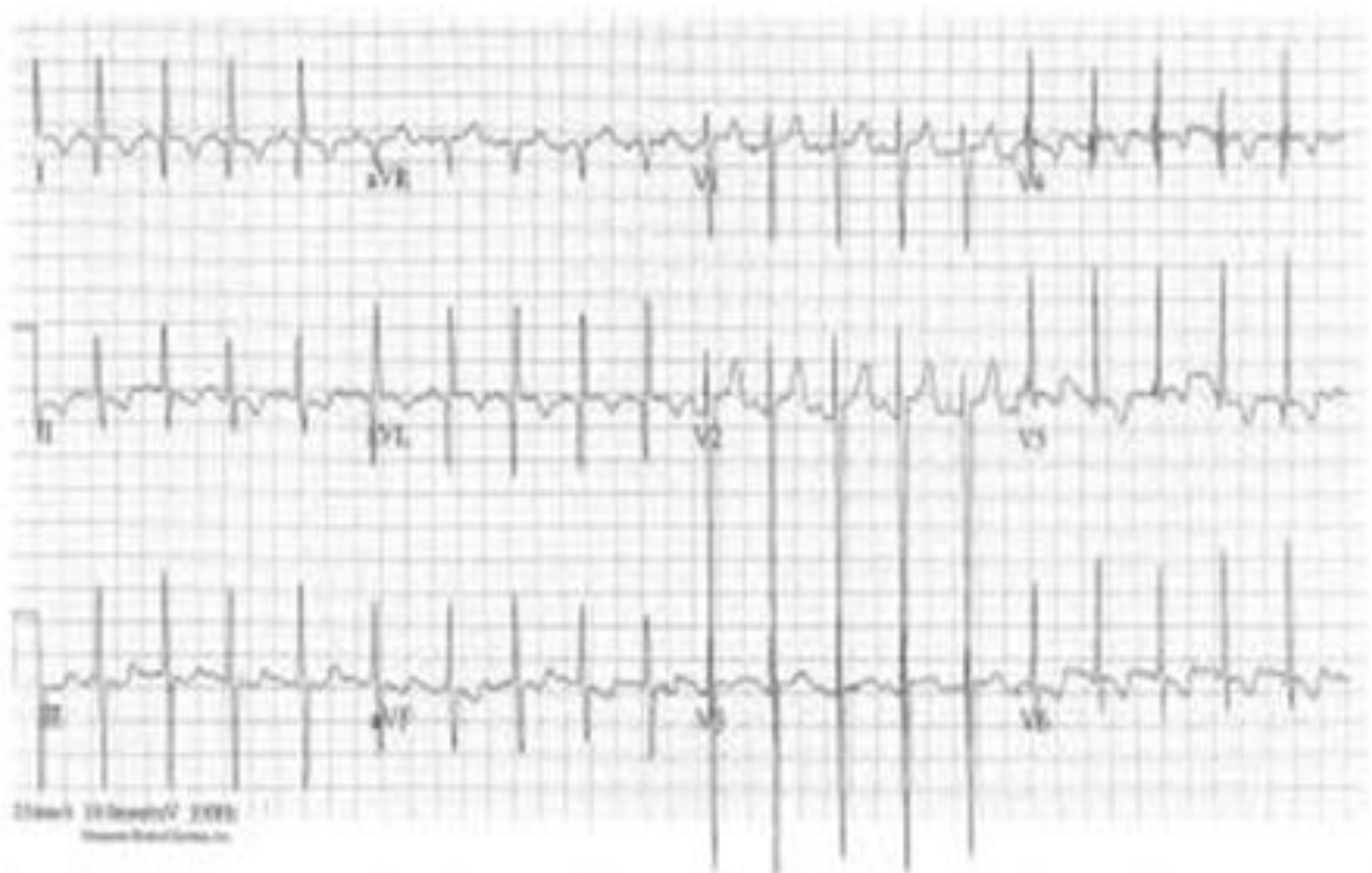
Blood Urea=35mg

Serum creatinine =0.9mg

Blood sugar=120mg

Normal lipids profile

ECG



ECHO:

LVEF=60%.

Anterior hypokinesia.

Mild mitral regurgitation

No pericardial effusion.

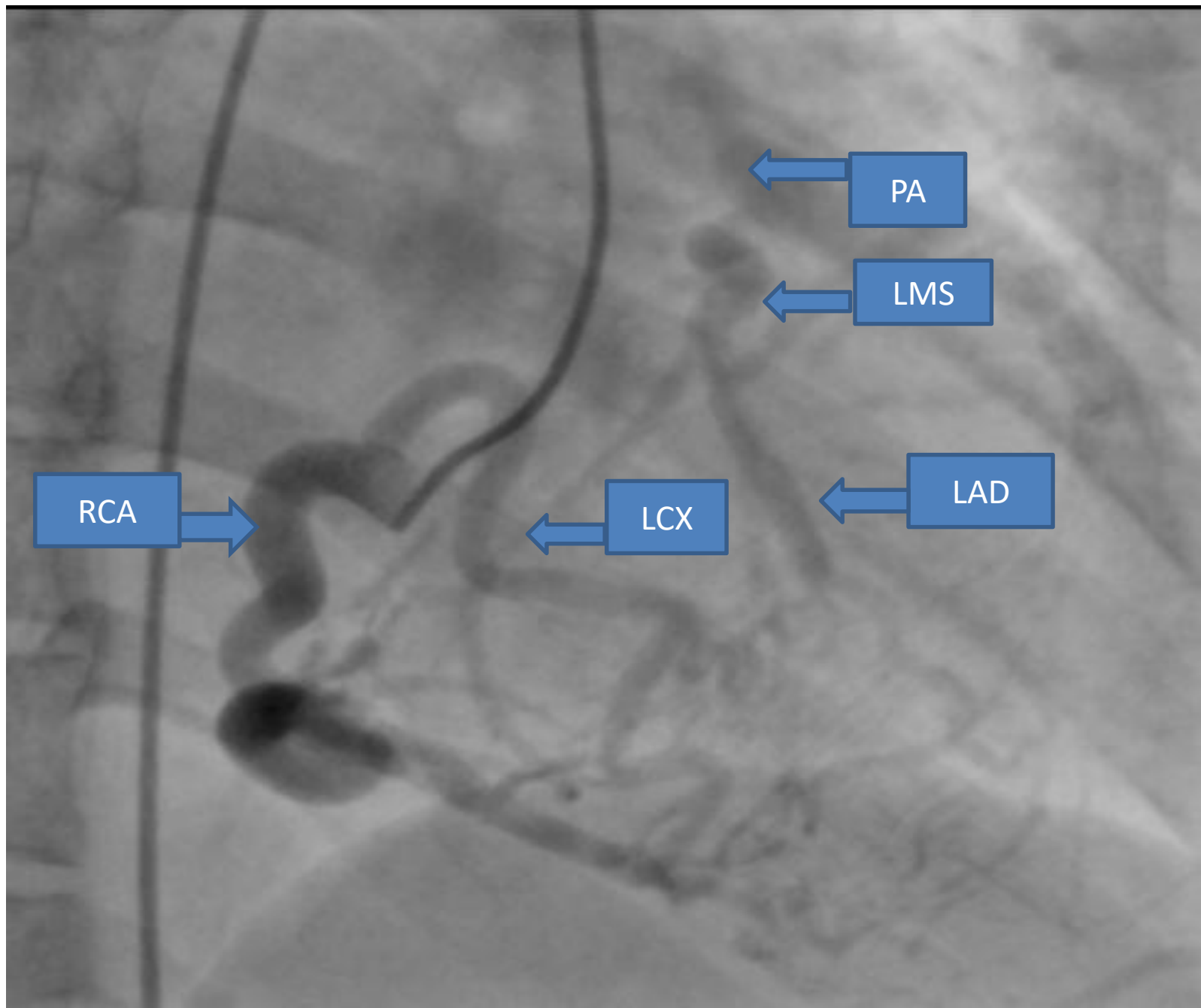
Left coronary angiography:

Absent left main stem ostium in the left coronary sinus .

RCA angiography:

Big right coronary artery with retrograde flow via collaterals from RCA to left anterior descending and circumflex arteries to left main stem artery to pulmonary artery.

Left main stem originated from pulmonary artery.



Medical treatment:

Conservative measures including :

Bed rest.

ECG monitoring.

Pain relief.

Beta blockers.

Surgery:

Ligation of left coronary artery ostium with left internal mammary artery graft to left anterior descending artery.

Follow up:

She had no major complications or ischemic symptoms two years after operation.

Follow up echo showed normal left ventricle systolic function and no valve abnormality.

Home message

ALCAPA syndrome is a very rare coronary anomaly (1 of 300,000 live births--0.4% of congenital cardiac anomaly) that needs high index of suspicion for diagnosis.

LV dysfunction in childhood and adolescence may be due to ischemia caused by coronary anomaly rather than classical myocarditis or cardiomyopathy.

Early surgical intervention is a definitive and life saving therapy .

Thank you for your attention