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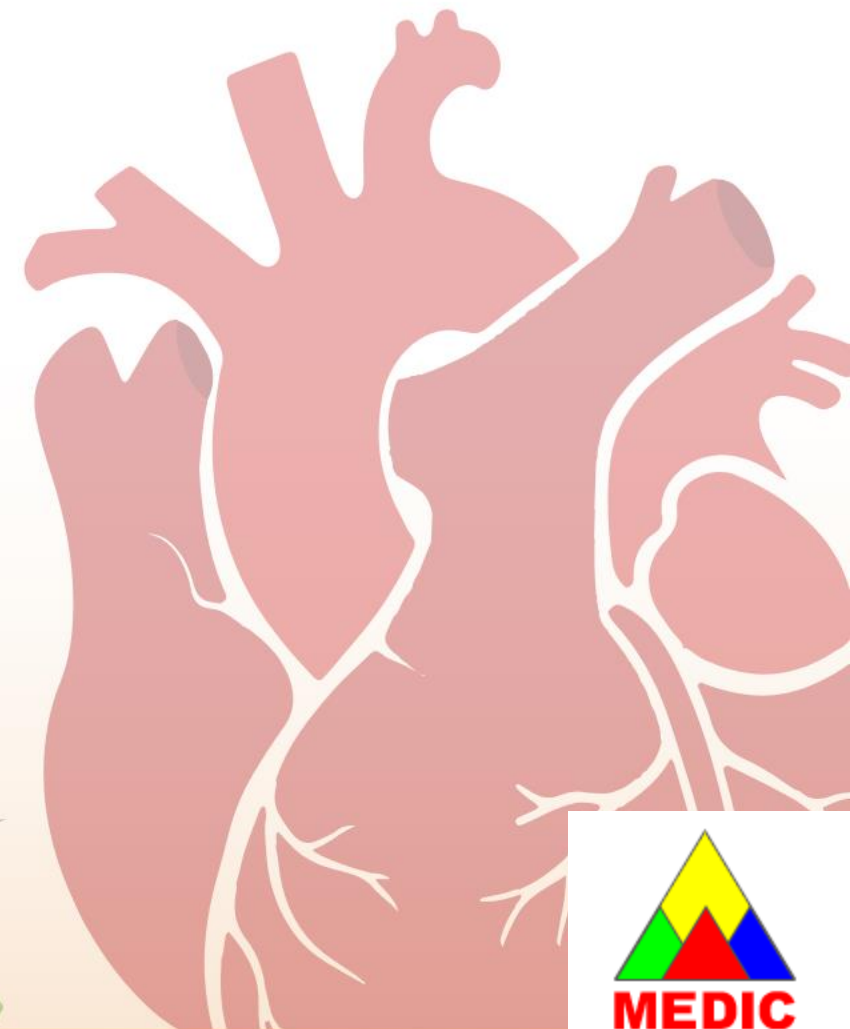


Multimodality Imaging in Vietnam: Addressing Past Diagnostic Limitations to Guide Current Treatment in Adult ALCAPA.

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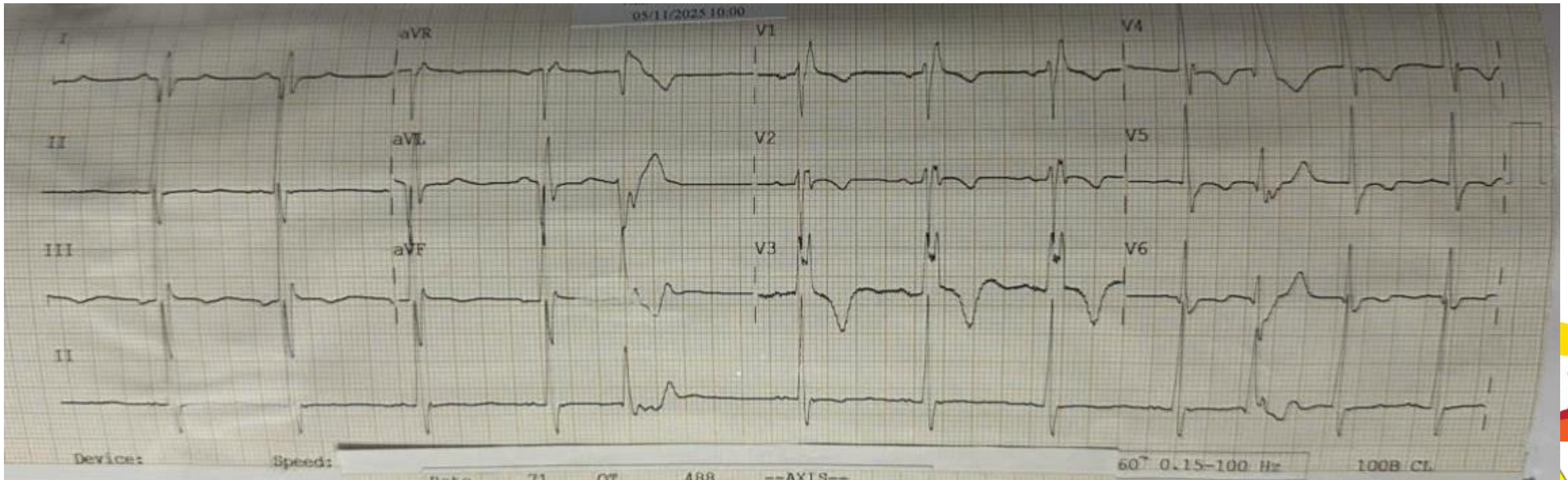


Case

- **Patient:** 35-year-old male
- **Chief Complaint:** Progressive fatigue and reduced exercise tolerance.
- **History of Present Illness:** The patient reports progressively worsening fatigue over time. He currently experiences: Dyspnea with minimal exertion, fatigue even at rest. He was recently evaluated during a routine check-up and was diagnosed with heart failure.
- **Past Medical History:** Surgical closure of a ventricular septal defect (**VSD**) at the Vietnam Heart Institute in 2003, lost to follow-up since surgery.
- **Vital Signs:** Heart rate: 84 bpm, Blood pressure: 140/70 mmHg, Oxygen saturation: 96% on room air.
- **Physical Examination:** No cardiac murmur, Presence of premature ventricular contractions (**PVCs**). Lungs: clear to auscultation, no crackles. No other significant abnormalities detected

Laboratory + ECG

- ❖ **Laboratory Findings:** White blood cell count (WBC): $7.2 \times 10^3/\mu\text{L}$, Hematocrit (Hct): 41%
- ❖ **Electrocardiogram (ECG):** Sinus rhythm, Presence of premature ventricular contractions (PVCs), T-wave inversion in the inferior and lateral leads.



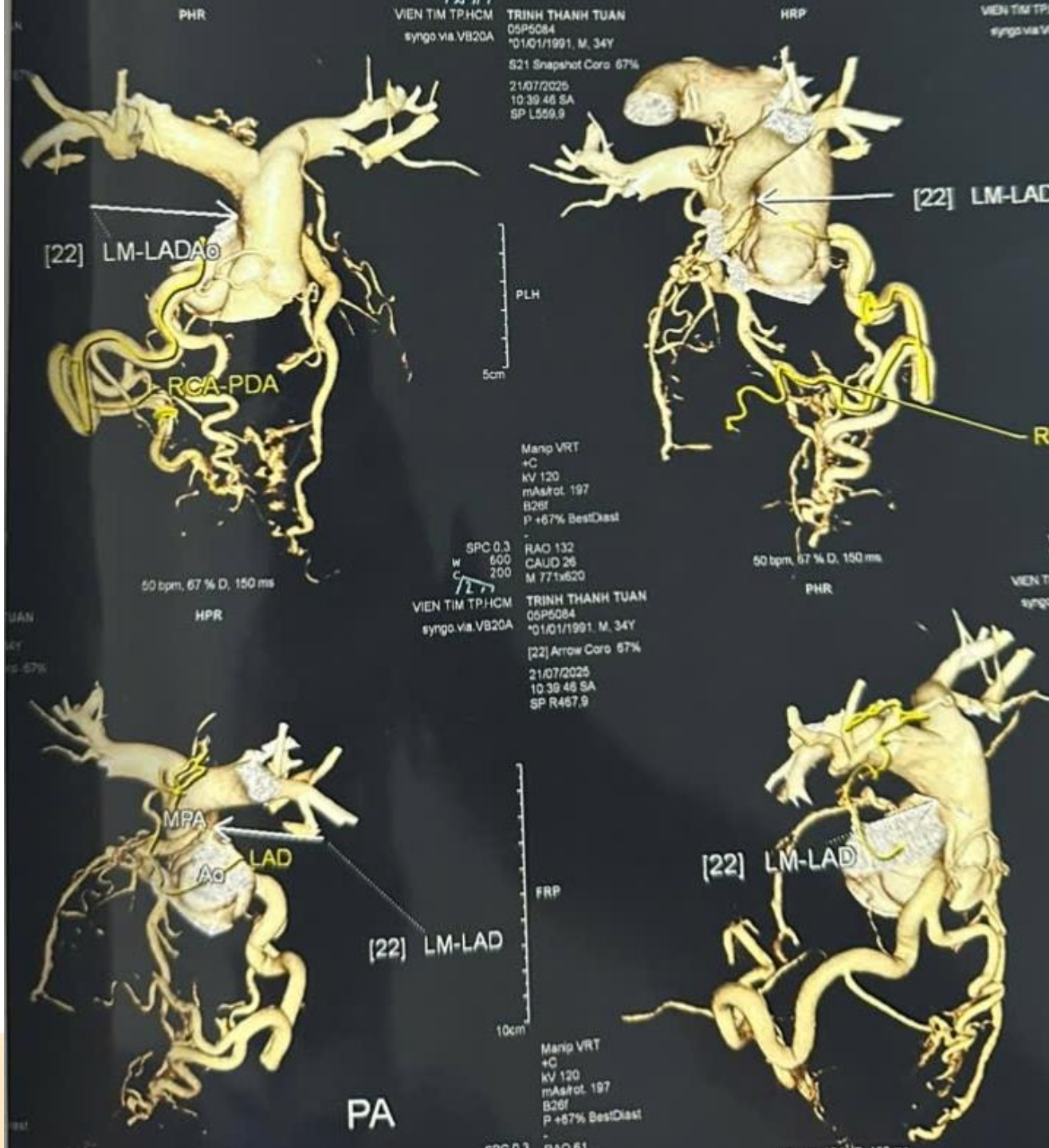
Suspicion of myocardial ischemia

Echocardiographic

- ❑ Dilated right coronary artery (RCA) at its origin (diameter: 6.8 mm);
left coronary artery (LCA) not visualized at its origin
- ❑ Evidence of retrograde flow within the interventricular septum
- ❑ Marked echogenicity of the anterolateral papillary muscle
- ❑ Global hypokinesia of the left ventricular myocardium
- ❑ Continuous flow entering the pulmonary artery at the bifurcation level
- ❑ Four-chamber cardiac dilatation
- ❑ No residual ventricular septal defect (VSD)
- ❑ Left ventricular ejection fraction (LVEF): 35%

Findings are suggestive of an anomalous origin of the left coronary artery (suspected ALCAPA)

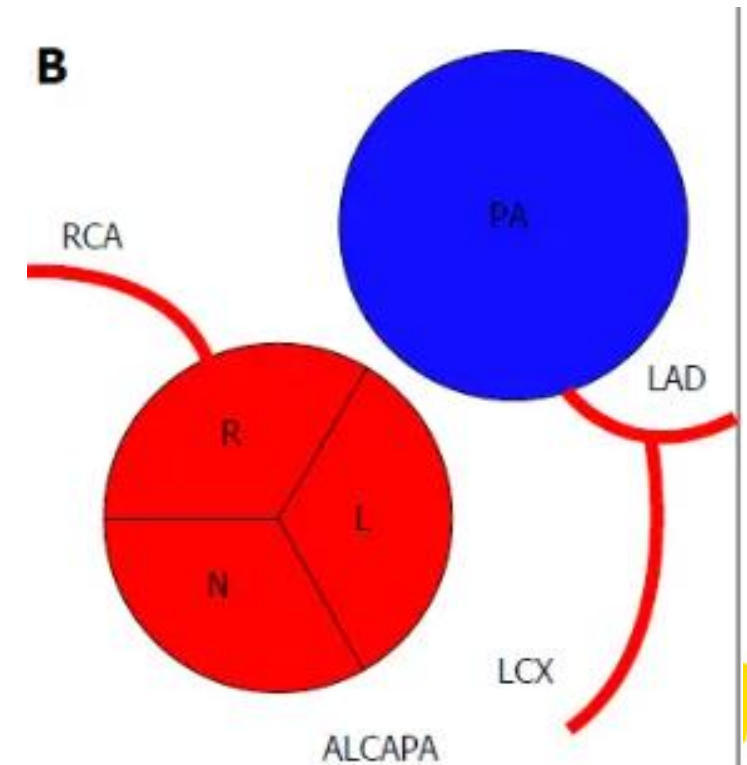
CTscan



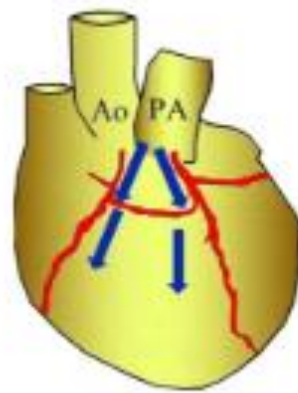
- ✓ Anomalous left coronary artery arising from the pulmonary artery (ALCAPA).
- ✓ Dilated left coronary artery without evidence of thrombosis.

Anomalous Left Coronary Artery from the Pulmonary Artery (ALCAPA)

- Mechanism: persistence of the pulmonary buds with concomitant involution of the aortic buds that are precursors of the coronary arteries.
- Pathogenesis:
 - *Coronary steal syndrome*
 - *Myocardial ischemia*
 - *Collateral circulation*



Radiopaedia.org – ALCAPA article



Neonatal Period

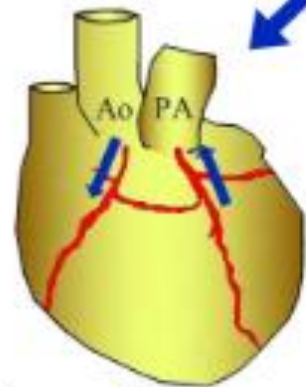
Pressure PA = Ao
Antegrade flow in LCA

↓
No symptoms

Decrease in PA pressure
Ability to develop collaterals
between RCA and LCA

2 TYPES OF ALCAPA

Infant Type

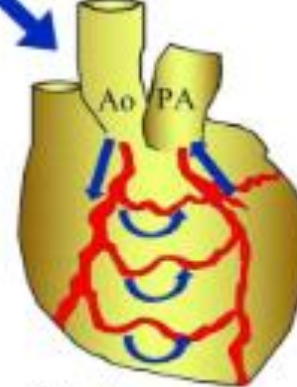


Pressure PA < Ao
Retrograde flow LCA
No collaterals between
RCA and LCA
RCA and LCA normal in size

↓
Ischemic cardiomyopathy
due to infarction

↓
90% death first year

Adult Type



Pressure PA < Ao
Retrograde flow LCA
Collaterals between RCA
and LCA
Marked dilatation of the
LCA and RCA due to the
increased longstanding
volume

↓
Chronic myocardial ischemia
Dysrhythmias

↓
Sudden death

Indications for Cardiac Magnetic Resonance Imaging (CMR)

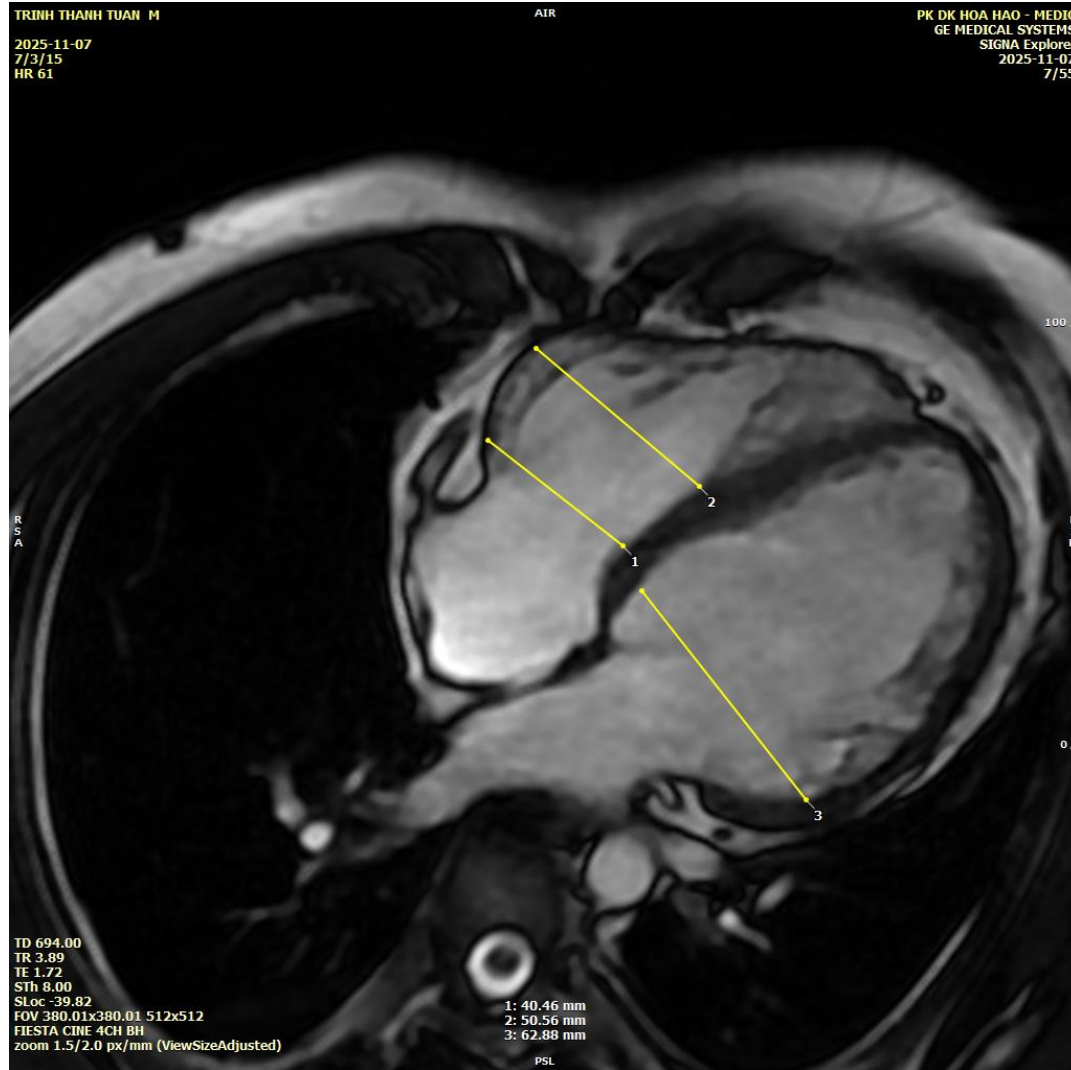
Recommendations for the management of patients with anomalous coronary arteries

Recommendations	Class ^a	Level ^b
Non-pharmacological functional imaging (e.g. nuclear study, echocardiography, or CMR with physical stress) is recommended in patients with coronary anomalies to confirm/exclude myocardial ischaemia.	I	C

- ❖ CMR is useful for assessment of ventricular function, myocardial fibrosis (LGE), and ischaemia in congenital heart disease.”

ESC Guidelines 2020 (Baumgartner et al., *Eur Heart J* 2021)

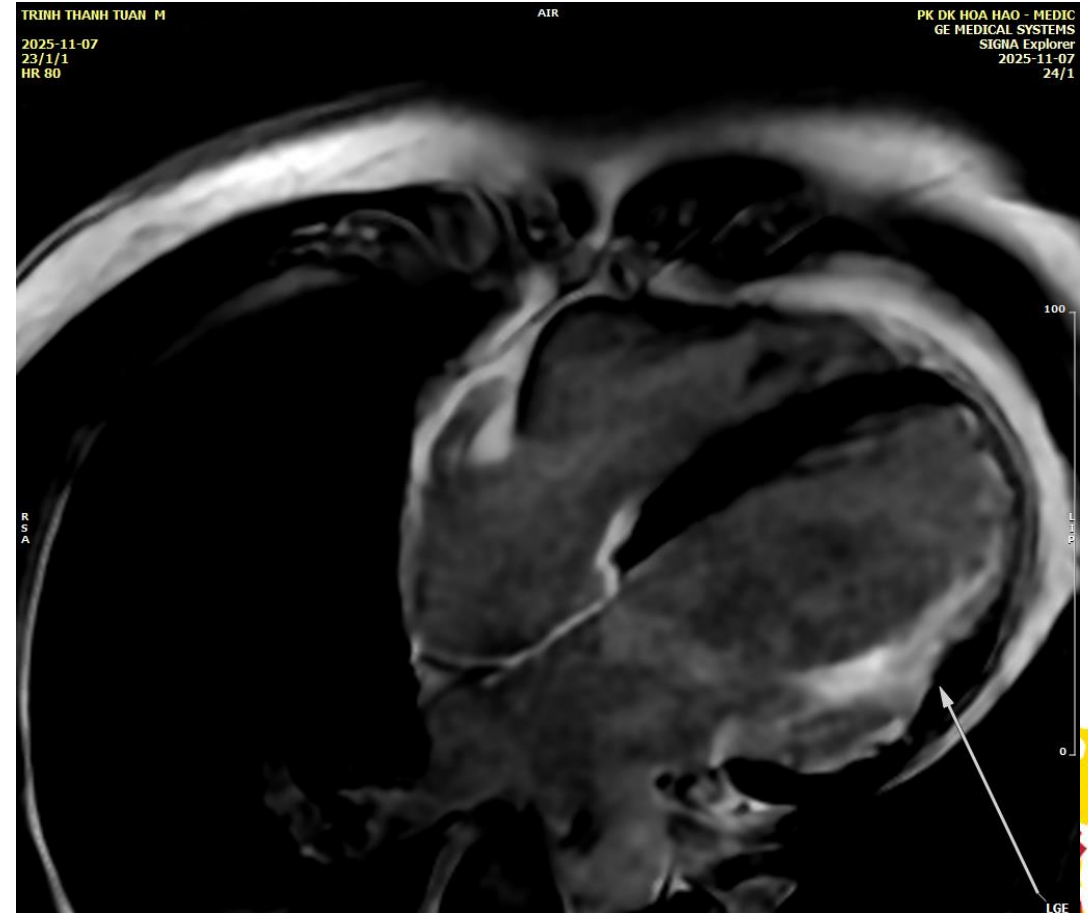
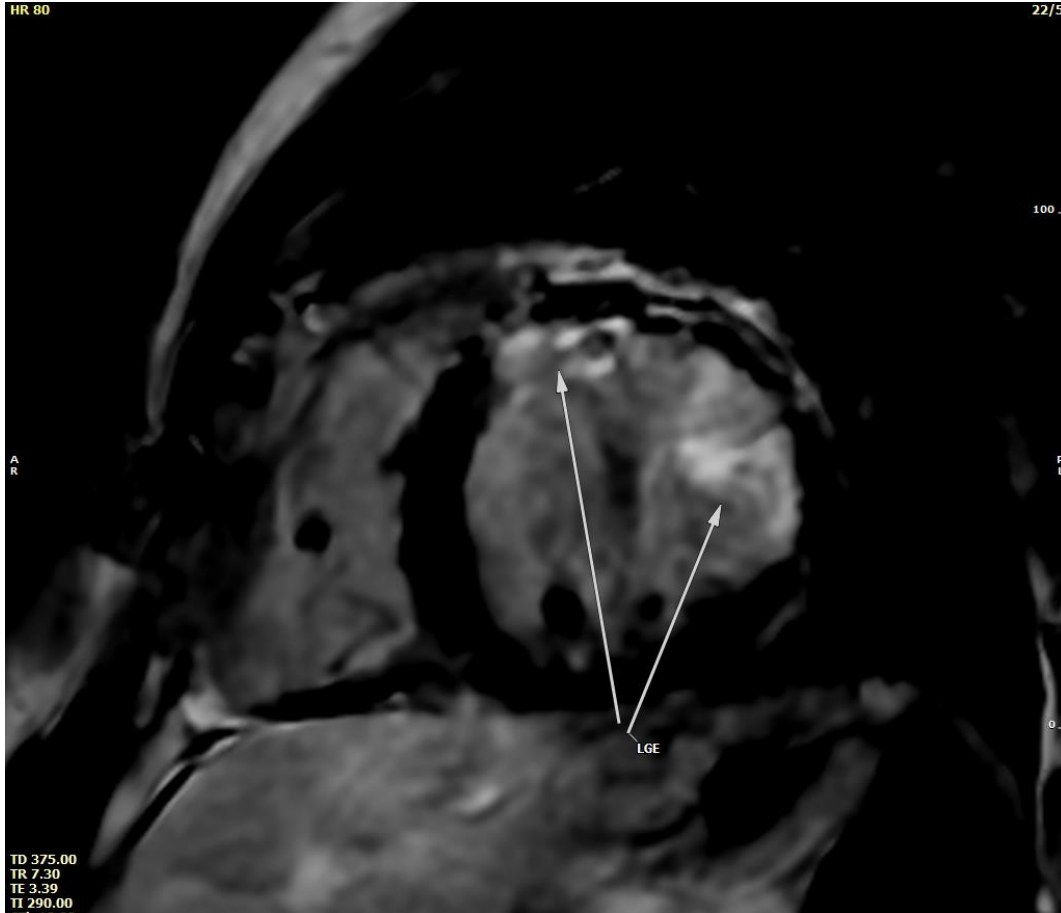
Cardiac MRI (CMR) Findings



- Left ventricular (LV) dilatation and increased LV volumes (LVDd = 63mm; LVEDVi = 133ml/m²).
- Severely reduced left ventricular systolic function, preserved right ventricular function (LVEF = 29%; RVEF = 56%).
- Moderate mitral regurgitation (RF = 21%).

Cardiac MRI (CMR) Findings

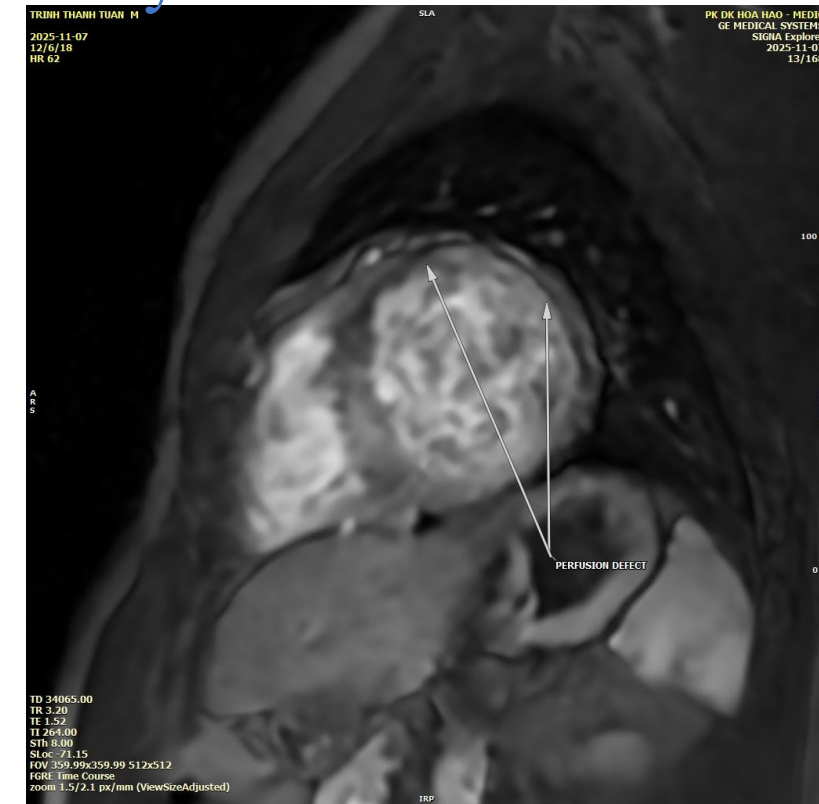
LGE



- Subendocardial Late Gadolinium Enhancement (LGE) involving the anterior and lateral walls of the left ventricle, and the entire anterolateral papillary muscle. The total fibrotic/scar burden is 24g, representing 21% of the total myocardial mass.

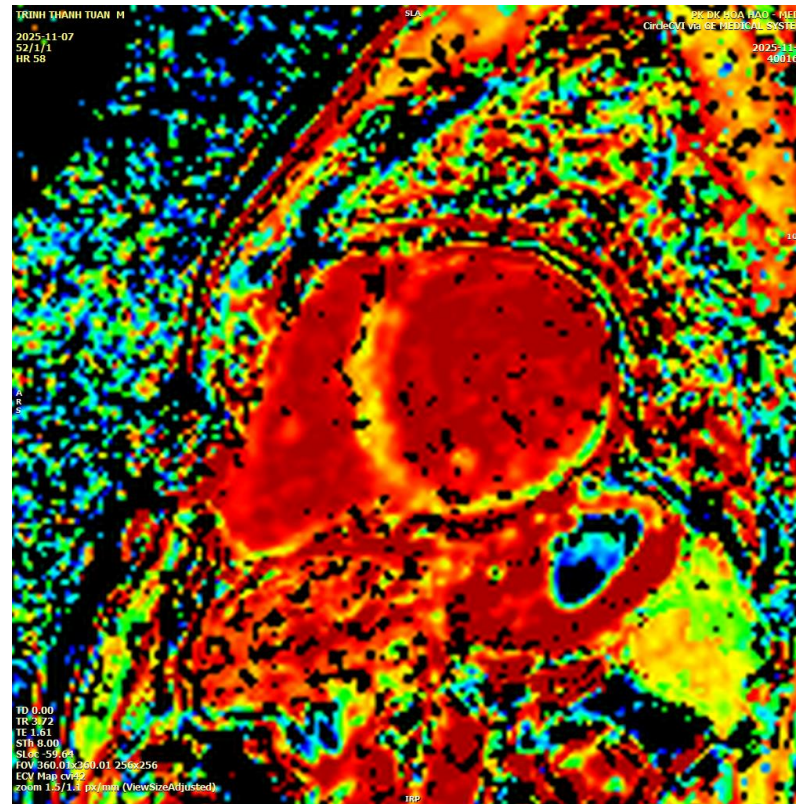
Cardiac MRI (CMR) Findings

Early Perfusion



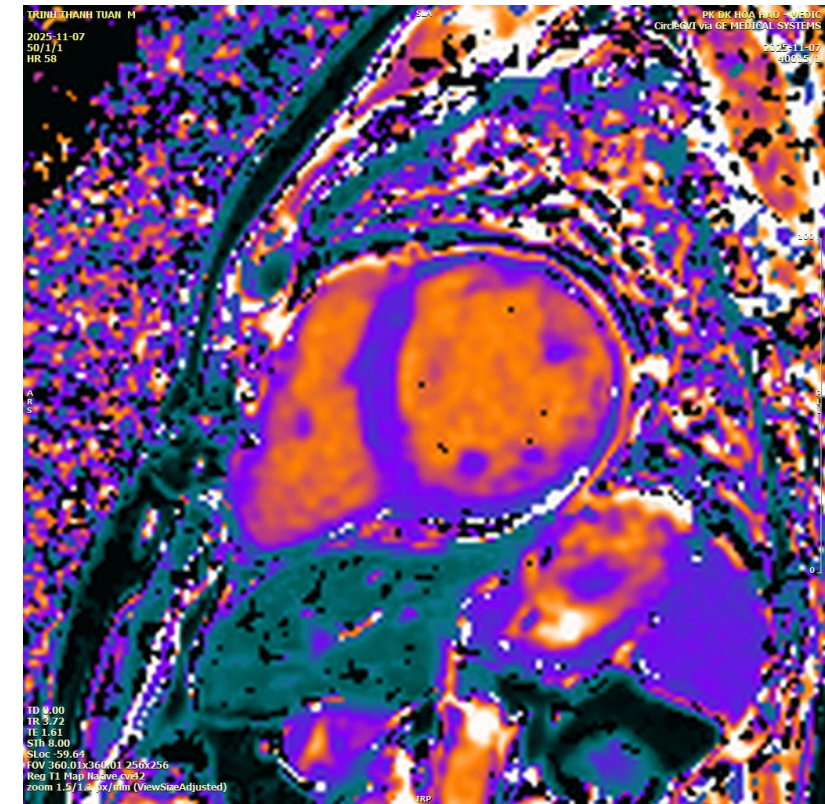
- Perfusion defects involving the anterior and lateral walls

ECV



- Extracellular Volume fraction: $ECV = 42\%$

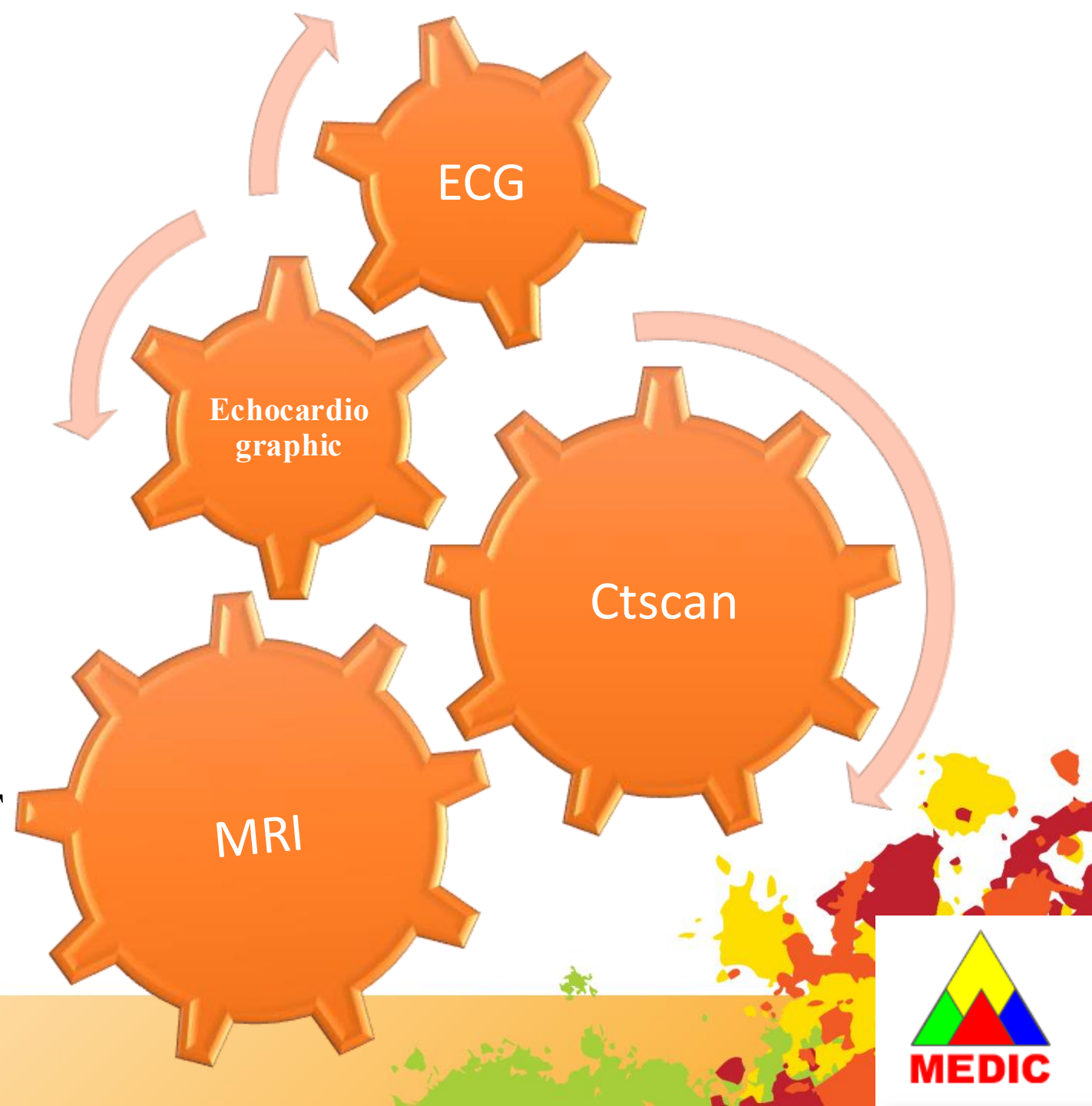
T1



- Native T1 relaxation time = 1243 ms

Discussion

- Echocardiography: Suggested coronary artery anomalies, reduced ejection fraction (EF), and no residual ventricular septal defect (VSD).
- ECG: Suggested myocardial ischemia.
- Cardiac CT: Confirmed the diagnosis of ALCAPA.
- Cardiac MRI: Assessed reduced EF and myocardial scarring/fibrosis, guiding the treatment plan.



Discussion

- ❖ Dual-coronary-supply reconstruction is recommended for patients with ALCAPA to restore hibernating myocardial activity and allow for full recovery of left ventricular function.
- ❖ Myocardial fibrosis as a substrate for malignant ventricular arrhythmias and sudden cardiac death => Providing critical prognostic information.

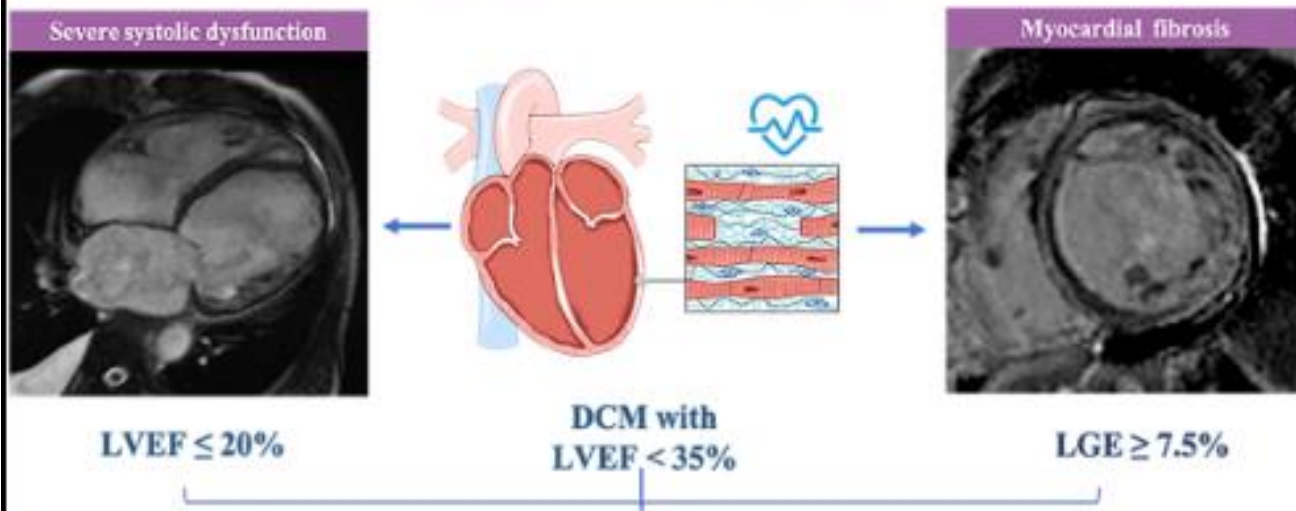


Development cohort (70%, n = 890)

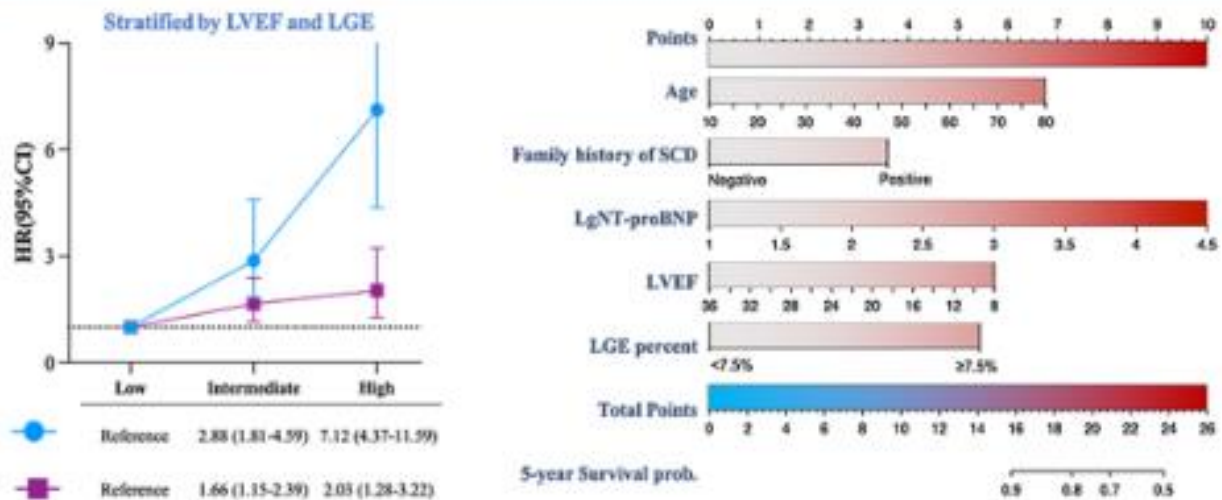


Internal validation cohort (30%, n = 382)

Follow-up: 86.3 months (IQR: 72.5-106.6)



Cardiac MRI-based risk stratification for clinical decision making



Thank you for your attention!