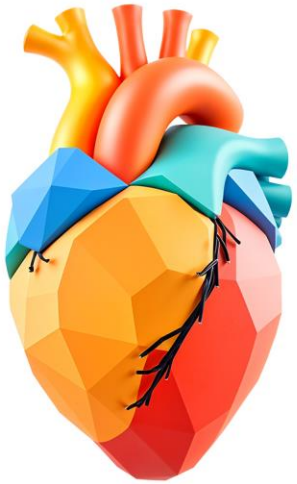

Critical Cardiovascular Care

دانشگاه علوم پزشکی و خدمات بهداشتی درمانی تهران

مرکز قلب تهران

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12th Annual Tehran Heart Center Congress

7th CRITICAL CARDIOVASCULAR CARE

دوازدهمین کنگره سالیانه مرکز قلب تهران

2025

۲۵ و ۲۶ بهمن ماه ۱۴۰۳

13 & 14 February

Tehran Heart Center

Tehran, Iran

CMR in Cardiomyopathies: does it change the diagnosis prognosis or outcome?

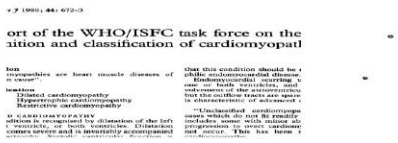
Dr Sahar Asl Fallah

Cardiologist, Fellowship in cardiovascular imaging

History of Cardiomyopathy

1980

World Health Organization (WHO) defined cardiomyopathies as "heart muscle diseases of unknown cause" to distinguish cardiomyopathy from cardiac dysfunction due to known cardiovascular entities such as hypertension, ischemic heart disease, or valvular disease.

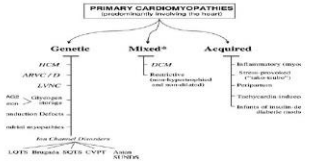

ort of the WHO/ISFC task force on the
ition and classification of cardiomyopat

1990

The **first genetic mutation** associated with hypertrophic cardiomyopathy (HCM) was identified in single base substitution in the **MYH7 gene** encoding ß-myosin heavy chain, a key component of the cardiac sarcomere

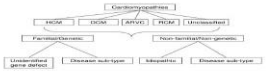
2006

American Heart Association (AHA) divided cardiomyopathies into primary or confined to the heart; genetic, mixed (genetic and nongenetic), and acquired, and secondary, as part of systemic diseases and previously referred to as specific cardiomyopathies.

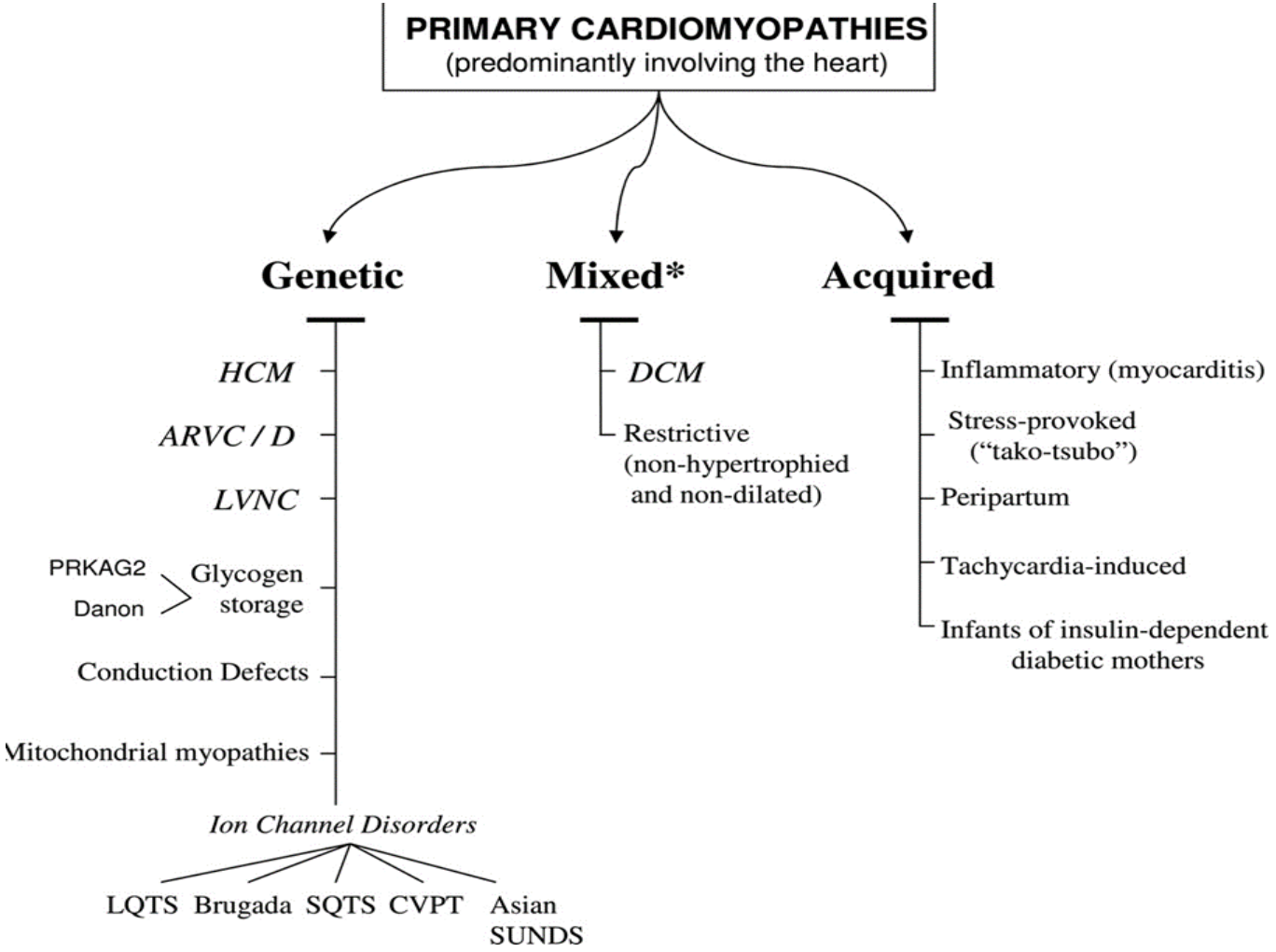


2008

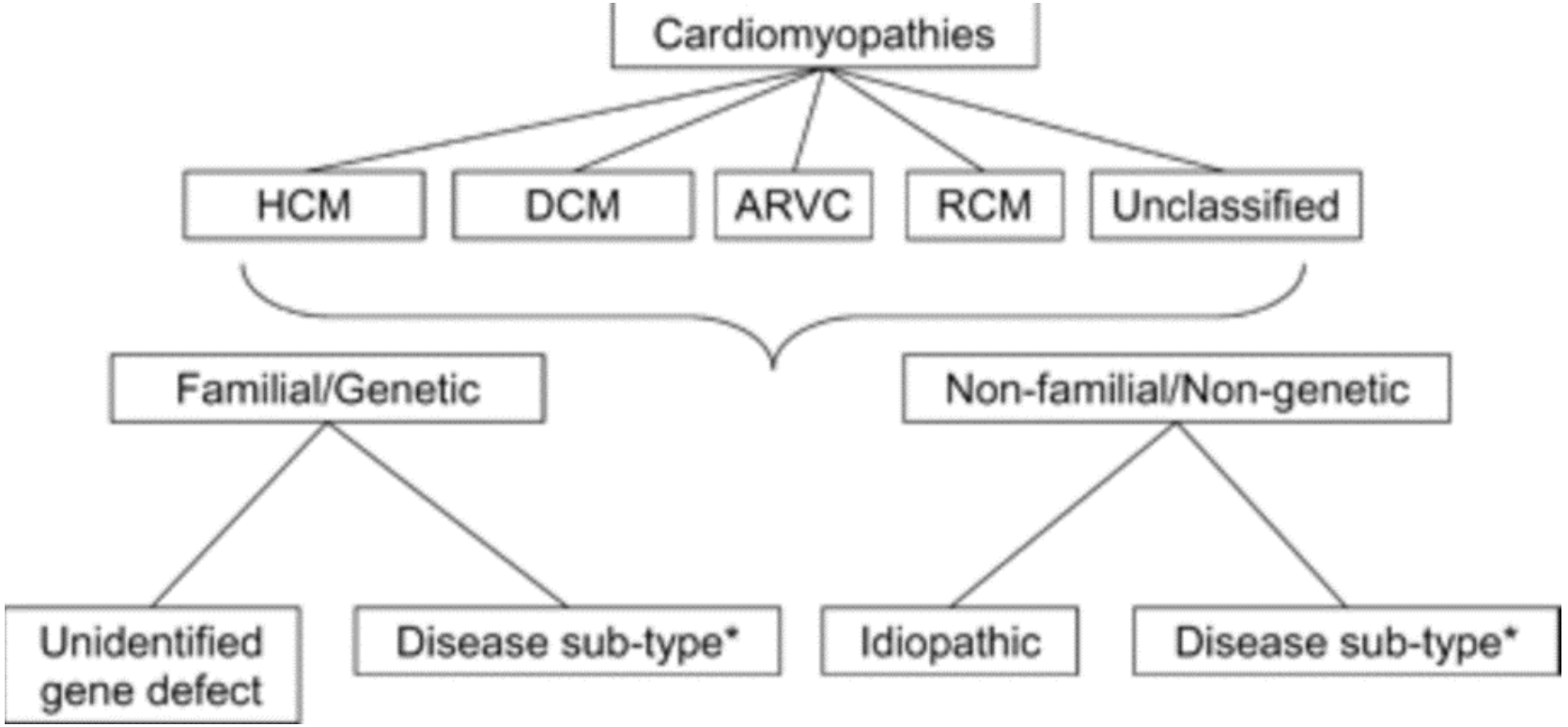
European Society of Cardiology (ESC) proposed a new definition of cardiomyopathy divided into phenotypes: dilated, hypertrophic, restrictive, arrhythmogenic right ventricular cardiomyopathy, and unclassified. The cardiomyopathies were then subclassified into familial and nonfamilial.



AHA 2006


PRIMARY CARDIOMYOPATHIES
(predominantly involving the heart)
Genetic
Mixed*
Acquired
HCM
ARVC / D
LVNC
PRKAG2
Danon
Glycogen storage
Conduction Defects
Mitochondrial myopathies
Ion Channel Disorders
LQTS
Brugada
SQTS
CVPT
Asian SUNDS
DCM
Restrictive (non-hypertrophied and non-dilated)
Inflammatory (myocarditis)
Stress-provoked ("tako-tsubo")
Peripartum
Tachycardia-induced
Infants of insulin-dependent diabetic mothers

ESC 2008


Cardiomyopathies
HCM
DCM
ARVC
RCM
Unclassified
Familial/Genetic
Non-familial/Non-genetic
Unidentified gene defect
Disease sub-type*
Idiopathic
Disease sub-type*

CMR

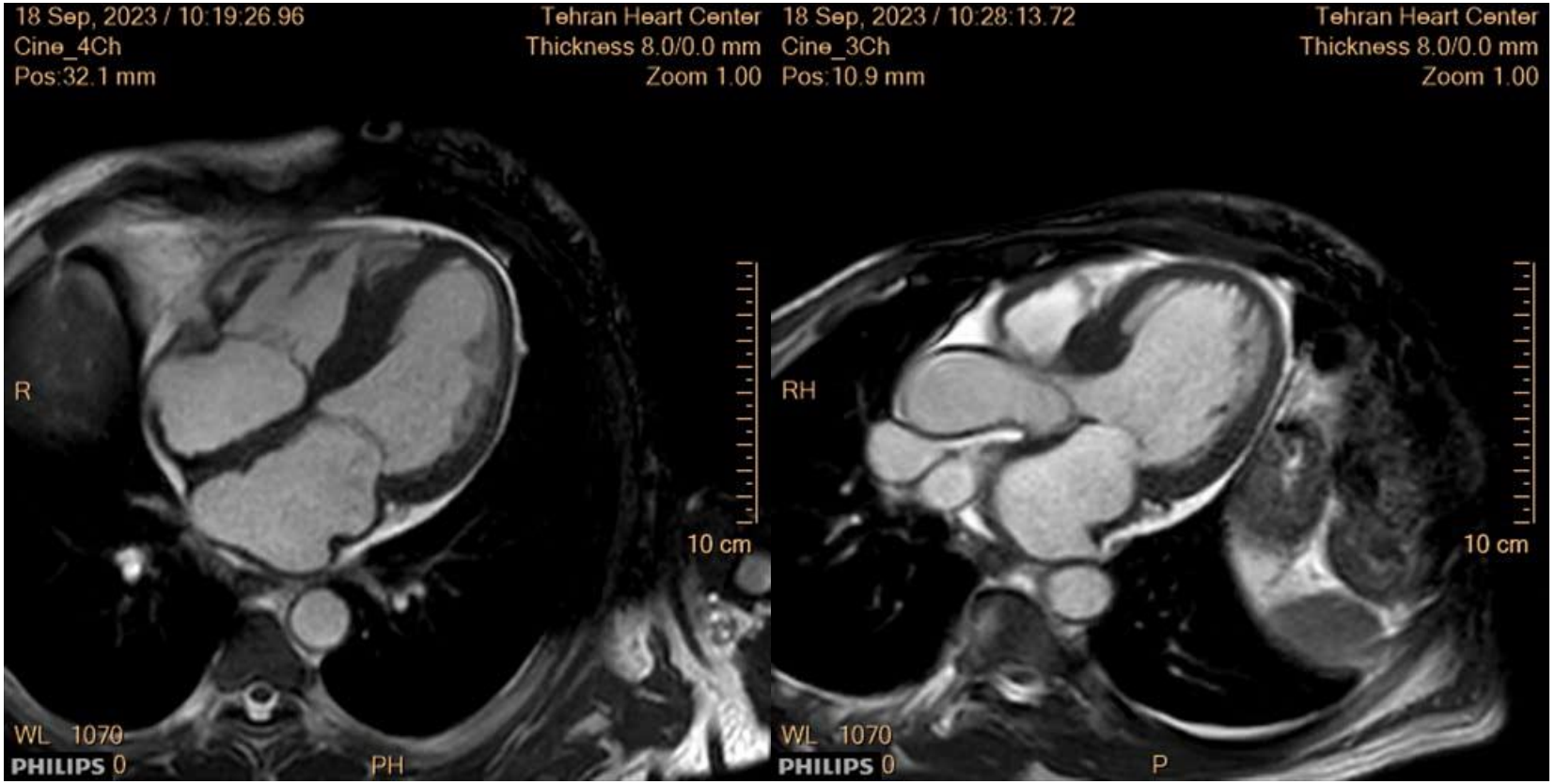
- **Reproducible measurements**
- **Morpho-functional assessment**
- **Myocardial tissue characterization.**
  - *Parametric mapping :*
    - *Quantification of T1/T2/T2* values*
    - *Extracellular volume (ECV).*
  - *Late gadolinium enhancement (LGE) and fibrosis*

Case 1

- A 64 year old man
- Worsening dyspnea from 6 months ago
- Episodes of paroxysmal palpitation

- Echo : Suspected HCM

➢ Recommendation: CMR for definite Diagnosis

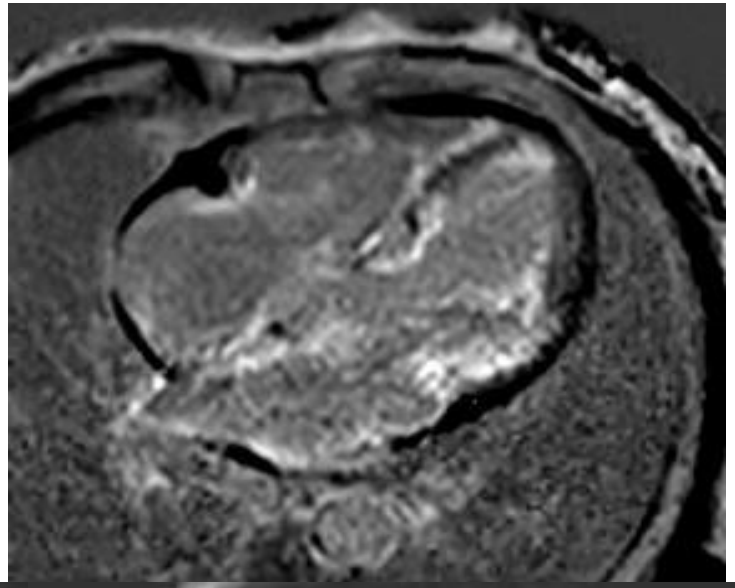


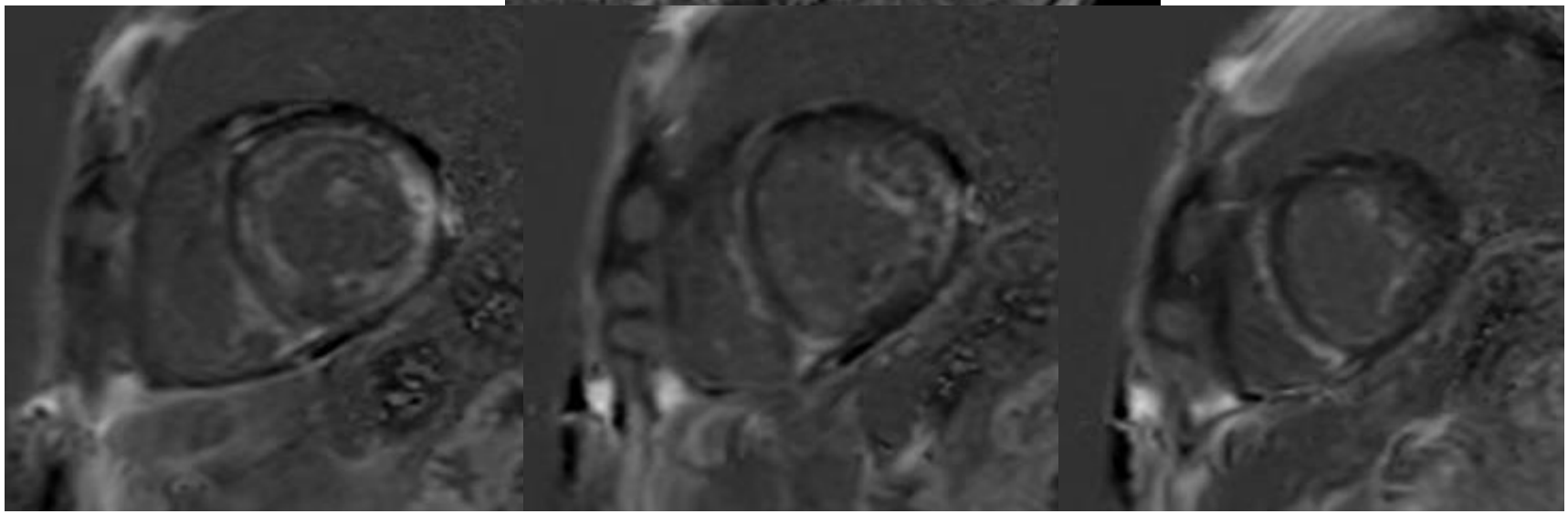
LVEF: 56%
LVEDVI: 100cc/m2

Max septal thickness: 16mm in mid infero-septal wall
LV mass index: 52gr/m2

Tissue characteristics

- **Abnormal myocardial nulling pattern:** reversed nulling.
- **Global Subendocardial to mid wall LGE** more prominent in basal segments
- **Bi-atrial** wall and **interatrial** septal LGE.
- Significantly **increased myocardial T1 value**: 1200ms





Cardiac Amyloidosis

- **Multi-systemic disorder:** accumulation of insoluble amyloid fibrils in the extracellular space

- **Cardiac involvement**: Accumulation of two types of protein:

✓ Immunoglobulin light-chain (AL): by small B-cell

Cardiac cause of death: within a few months

✓ Transthyretin (TTR)

- Hereditary form (ATTRm): Mutations in TTR

- Non-hereditary form: misfolding of wild-type transthyretin (ATTRwt) "senile systemic amyloidosis": late onset , more often in men

- **ATTR-CM** : *better prognosis (overall survival 4–5 years from diagnosis) than the AL-CM (<6 months)*

- **The main hypertrophic pattern in ATTR-CM** :

  Asymmetrical LV hypertrophy (present in 79% of patients)

  - *Sigmoid septum in 55%*
  - *Reverse septal contour in 24% of patients with ATTR-CM*

- **The main hypertrophic pattern in AL-CM :**

  - *Symmetrical and concentric*

LGE in amyloid cardiomyopathy:

- **Diffuse and subendocardial in early stages**
- **Transmural in advanced stages of the disease**
- **Diffuse infiltration of the myocardium leading to:**

- Abnormal nulling of the myocardium

- Significant Increase in T1 and ECV

Case 2

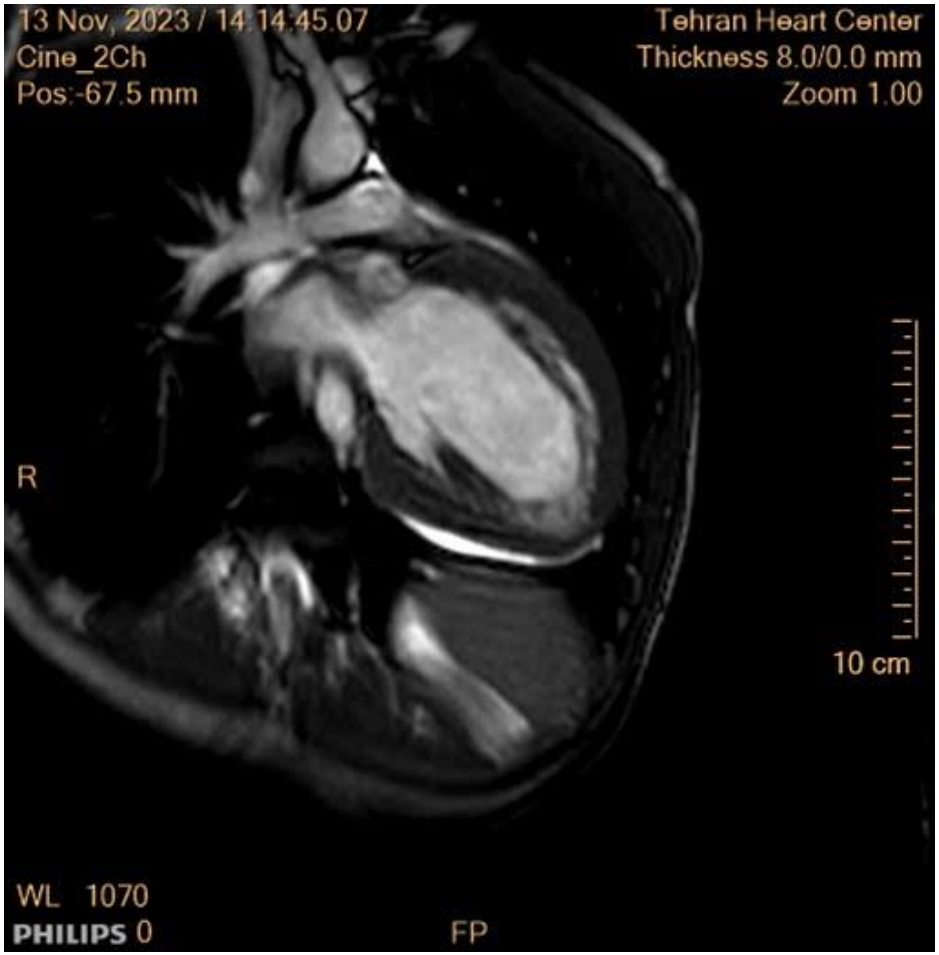
- A 37 year-old man with prolonged weakness, dyspnea during exercise and atypical chest pain
- Past medical history : none
- Echo:

  Severe LV dysfunction

  Significant concentric LVH

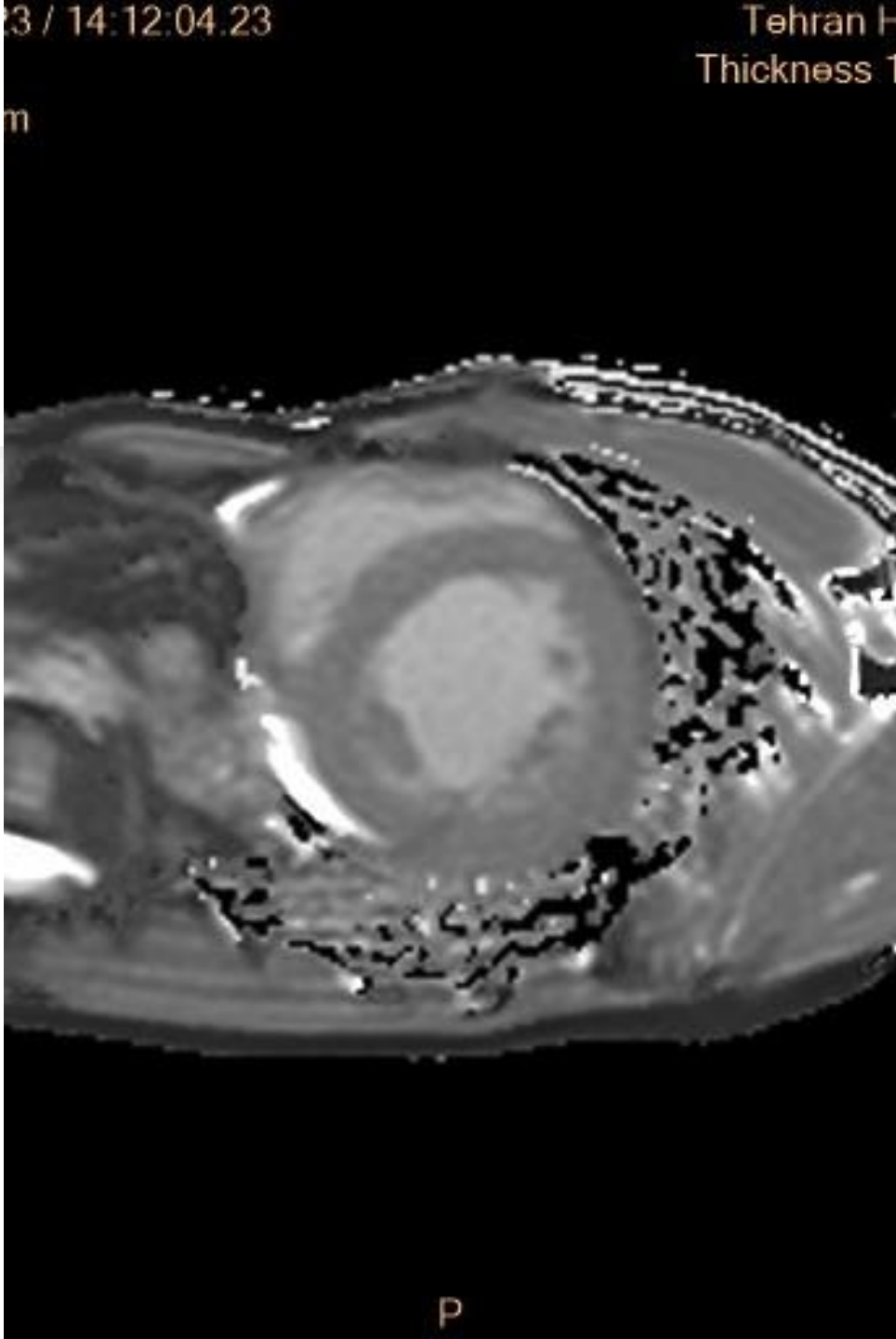
➢ CMR for evaluation of CMP

- ***LVEF: 20 %, LVEDVI: 129 ml/m2***
- ***Concentric LVH. Max septal thickness:14mm increased total and indexed mass.***
- ***RVEF: 38 %, RVEDVI: 78 ml/m2, RVH in RV free wall***

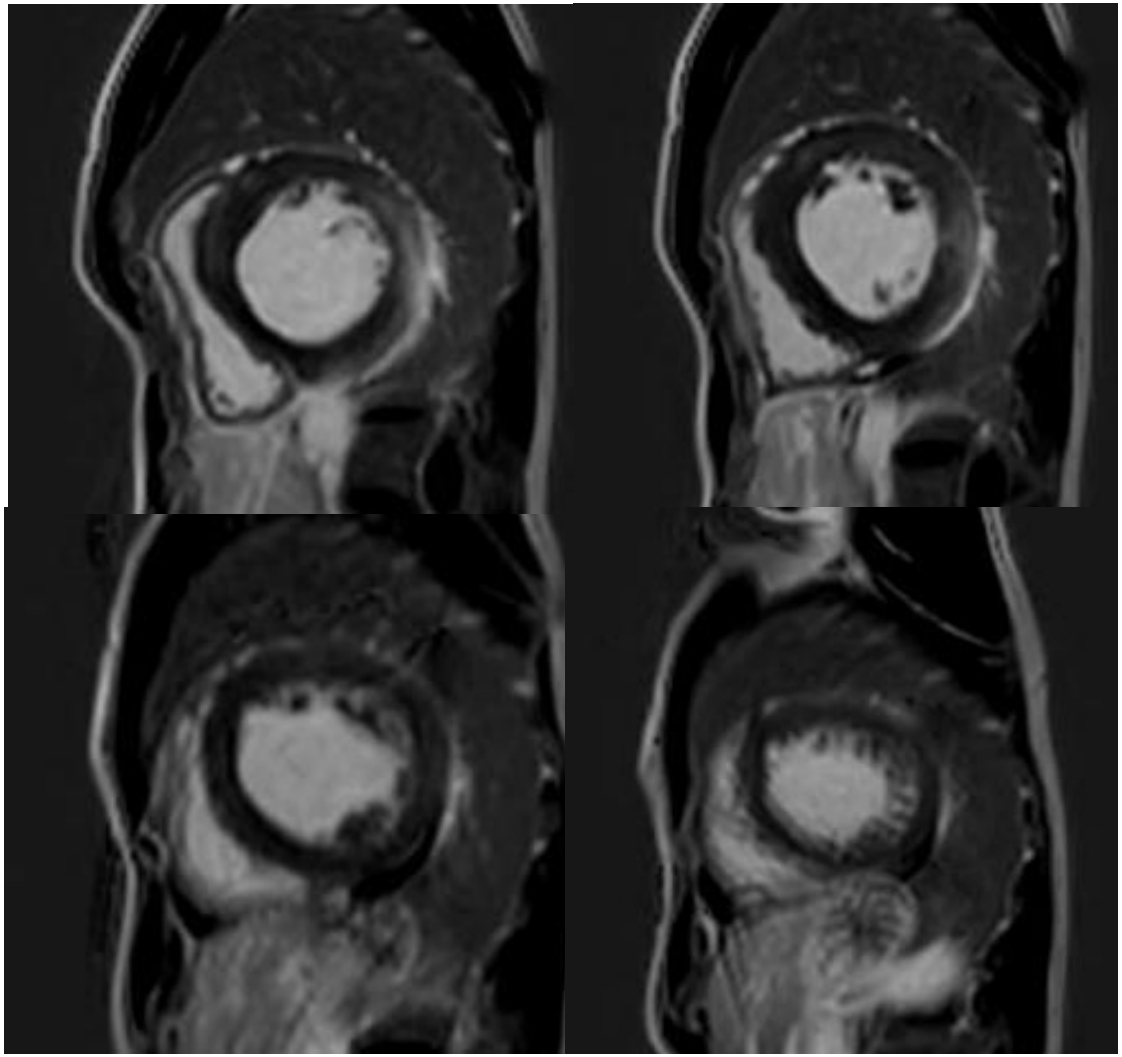


Myocardial T1 mapping

- <u>Abnormally Reduced</u> Global Myocardial T1 relaxation time: 860ms (normal range in our center: 900-1050)



- ***Subepicardial to mid wall LGE in basal to mid infero-lateral wall.***

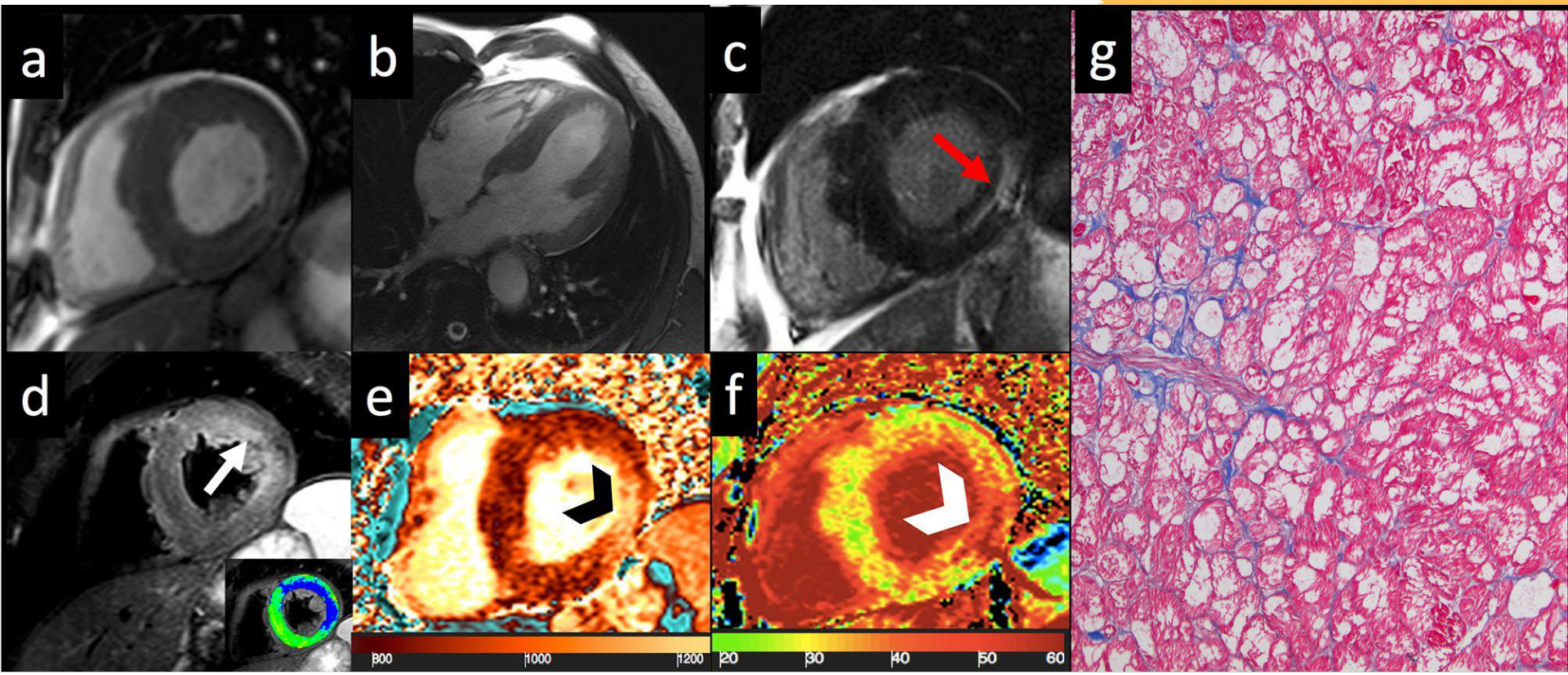


Anderson-Fabry CMP

- X-linked recessive multisystemic lysosomal storage disease
- Mutation in the α-galactosidase gene : lysosomal **accumulation of glycosphingolipids** in several organs
- Cardiac, cerebral, and renal impairment
- **LV hypertrophy that mimics HCM**, with different degrees in wall thickening.
- **Myocardial storage leads to :**

  - *Progressive systolic and diastolic dysfunction*

  - *Arrhythmias or heart failure*



severe reduction in global T1 (reddish brown color)

Hematoxylin and eosin histology (**g**, ×200) shows cardiomyocytes hypertrophy, caused by large cytoplasmic and perinuclear vacuoles, containing myelin bodies

Galea N, Polizzi G, Gatti M, Cundari G, Figuera M, Faletti R. Cardiovascular magnetic resonance (CMR) in restrictive cardiomyopathies. La radiologia medica. 2020 Nov;125:1072-86.

❑**Myocardial T1 values** : global shortening

✓ Myocardial storage of glycosphingolipids: very low T1 of fat tissue

✓ Different from other infiltrative cardiomyopathies with normal or elongated nT1 values

❑**LGE** : in up to 50% of AFD patients typically in inferolateral wall

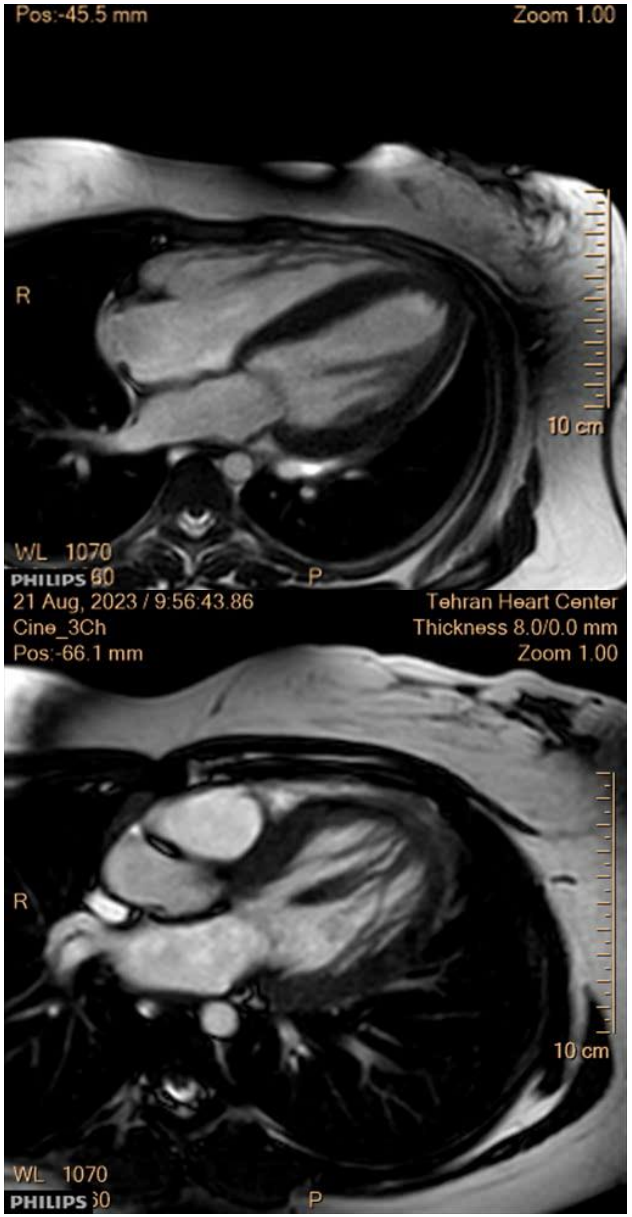
- the best predictor of cardiac events
- preceding the development of LVH

Continuous enzyme replacement therapy (ERT) : Dramatic improvement in clinical cardiac symptoms in a substantial number of patients.

Case 3

- 13 year-old girl with dizziness and dyspnea in exercise
- LVH with LVOT gradient at rest(20mmHG) and suspected HOCM in echo
- Referred for CMR

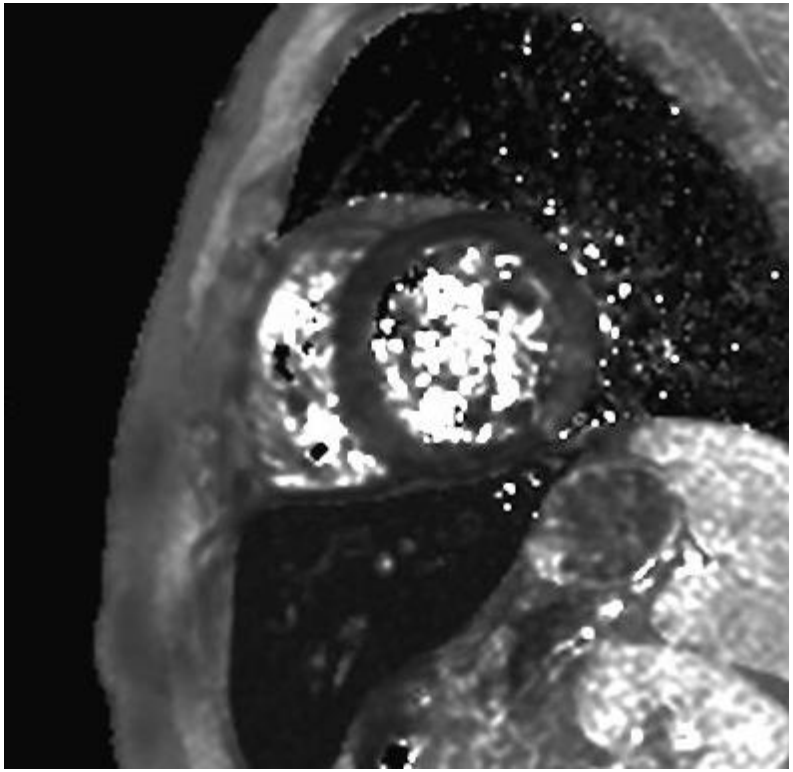
- *Normal LV size with supra-normal systolic function. EF: 67%, LVEDVI: 79ml/m2*
- *Increased total and indexed LV mass Maximum myocardial thickness = 13.5 mm*
- *Evidence of subaortic flow acceleration below the AV, narrow LVOT(8mm) suggestive for tunnel shaped subaortic stenosis*
- *No myocardial edema or fibrosis*
- *Normal T1 and T2*



Case 4

- A 37 year-old man with underlying hemochromatosis
- Recent dyspnea
- Echo: Severe LV systolic dysfunction
- CMR for evaluation of CMP and Iron overload

- ***Severely enlarged LV size with severely reduced systolic function. EF: 26 %, LVEDVI: 139 ml/m2,***
- ***Mildly enlarged RV size with moderately reduced systolic function. EF: 33 %, RVEDVI: 97 ml/m2***
- ***Myocardial T2* : 8ms***
- ***Severe iron overload in the heart***



Iron overload CMP

- **Hereditary:** hemochromatosis
- **Iatrogenic**: Frequent blood transfusions
- **<u>CMP: The leading cause of death in iron overloaded patients</u>**
- **Iron deposition in the myocardium result in**:

  -Dysfunction, originally diastolic and then systolic
- **Symptoms:** From asymptomatic patients to irreversible heart failure in severely overloaded patients with dilated cardiomyopathy

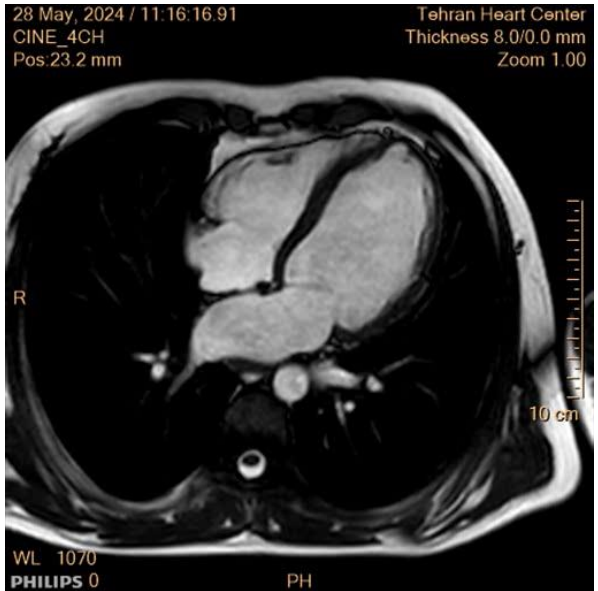
- **CMR** : the most powerful tool to detect and quantify cardiac iron overload by myocardial T2* quantification

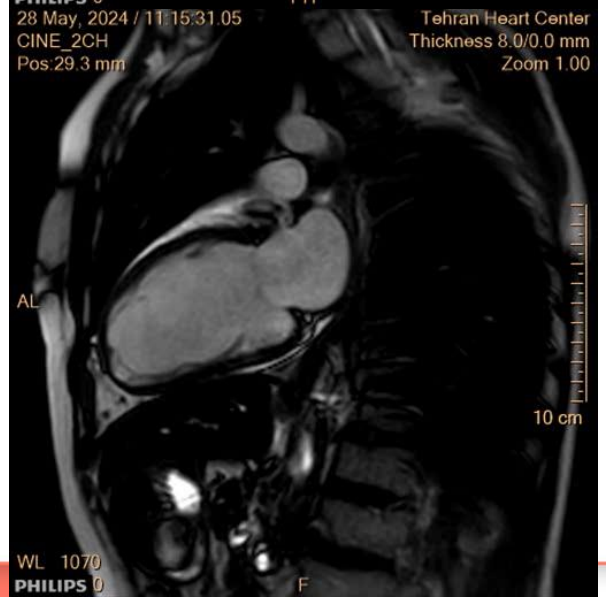
- In iron overloaded hearts the paramagnetic effect of iron is responsible for changes in MR signal intensity, shortening T1 and T2 relaxation times

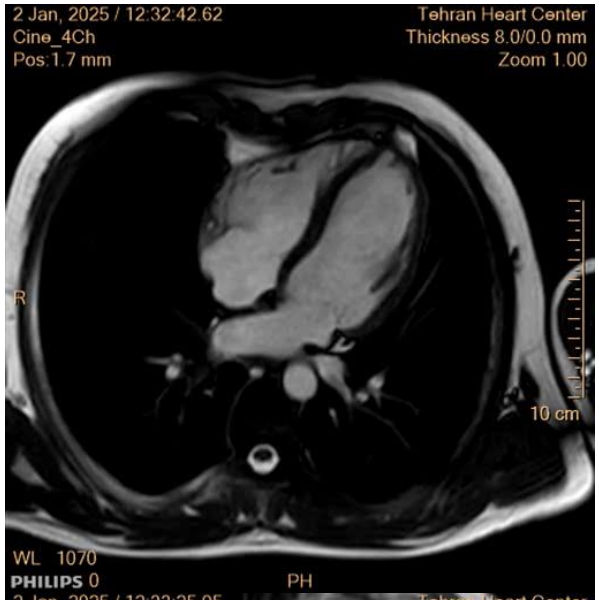
- T2* value can be measured by Gradient echo sequences (which are more susceptible to magnetic field inhomogeneity)

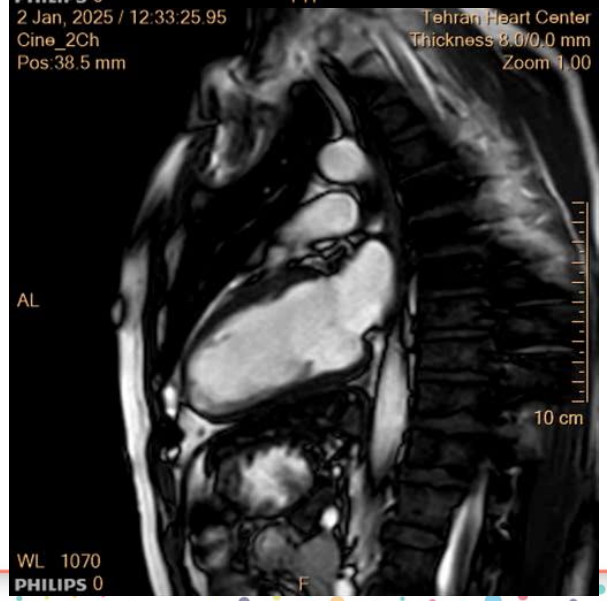
- **T2*>20 ms** :Green zone: **No iron overload**, with low risk for congestive heart failure.
- **T2* from 10 to 20 ms :** Yellow zone : Mild to moderate iron overload and patients are at intermediate risk of cardiac failure.
- **T2*<10 ms** : Red zone: Severe iron overload and patients are at high risk of cardiac decompensation, needing prompt intensification of iron chelation therapy
- **T2* monitoring**: A crucial role in monitoring the iron overload status during the chelation therapy

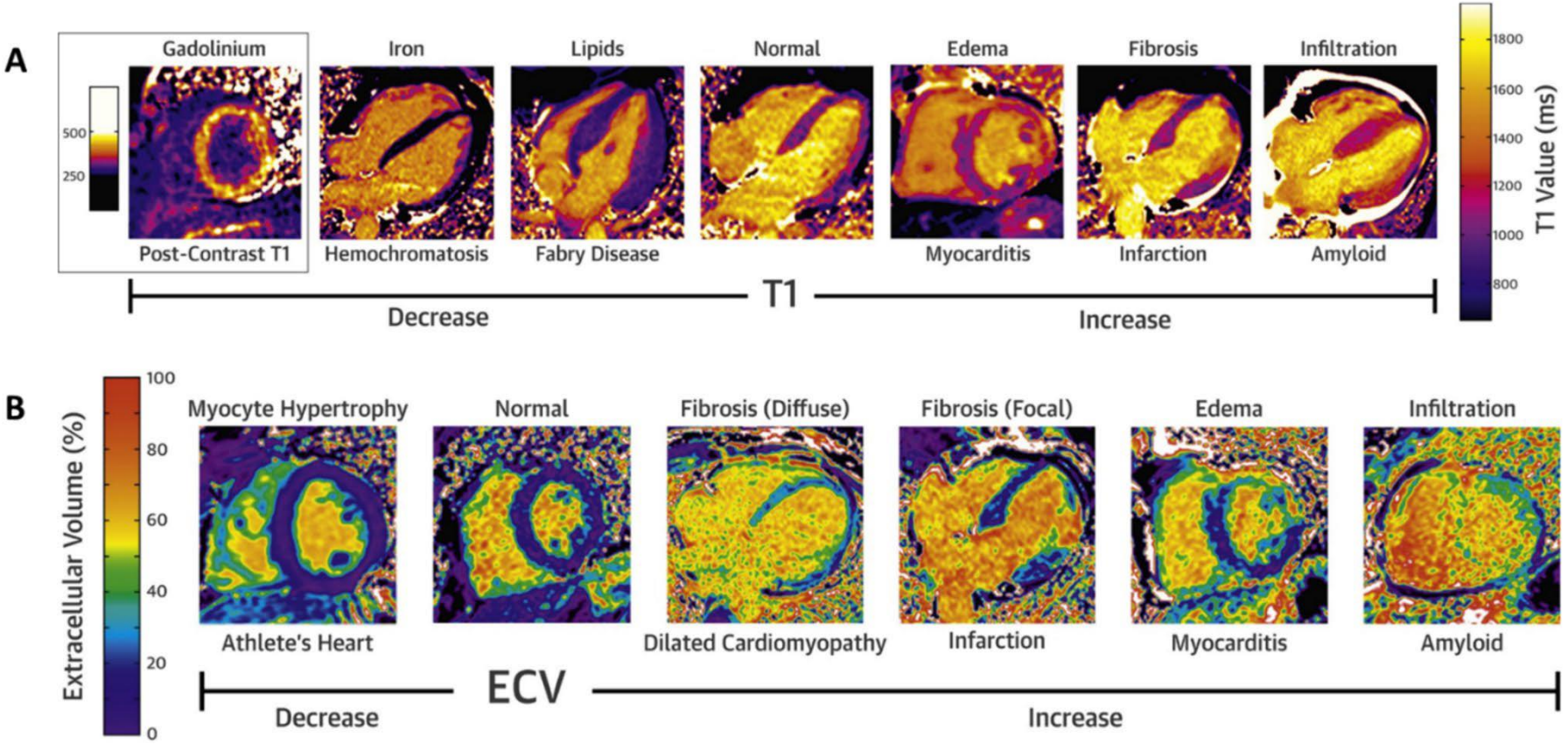
F/U CMR after 6 months of Iron chelation










A
Gadolinium
Iron
Lipids
Normal
Edema
Fibrosis
Infiltration
500
250
Post-Contrast T1
Hemochromatosis
Fabry Disease
Myocarditis
Infarction
Amyloid
T1
Decrease
Increase
1800
1600
1400
1200
1000
800
T1 Value (ms)
B
Extracellular Volume (%)
100
80
60
40
20
0
Myocyte Hypertrophy
Normal
Fibrosis (Diffuse)
Fibrosis (Focal)
Edema
Infiltration
Athlete's Heart
Dilated Cardiomyopathy
Infarction
Myocarditis
Amyloid
ECV
Decrease
Increase

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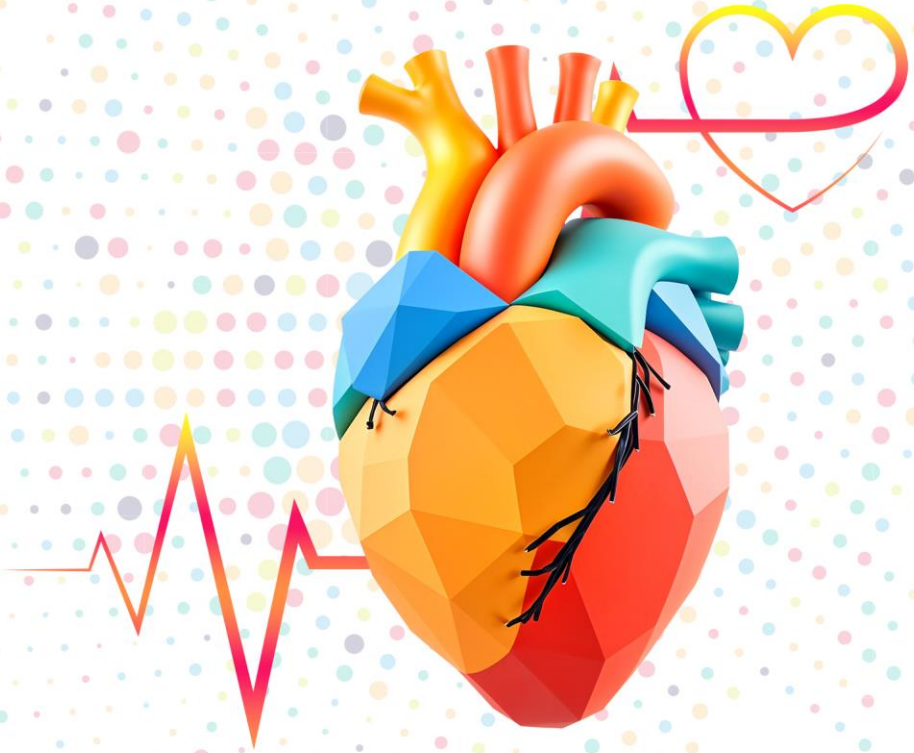

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