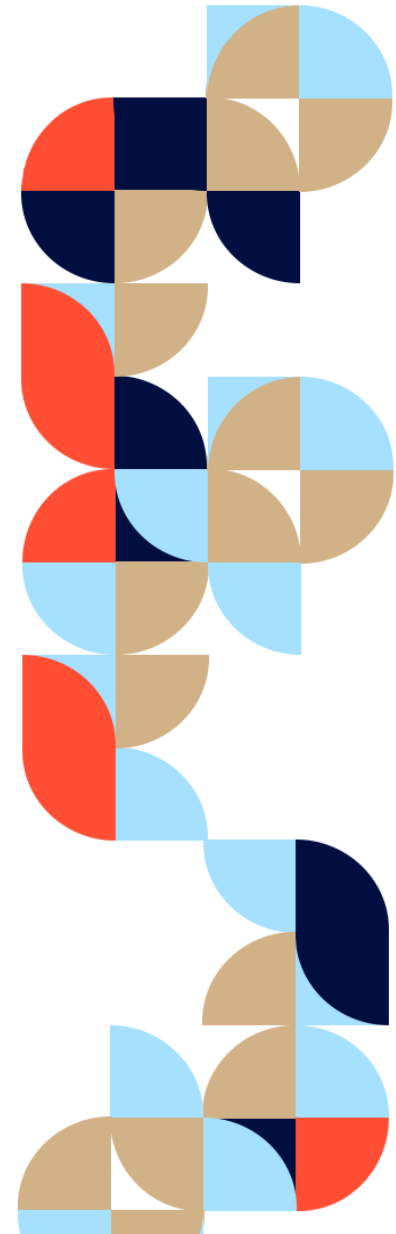


Avaliação da Doença de Fabry pela Ressonância Cardíaca: o que podemos valorizar no diagnóstico diferencial?

Marly Uellendahl MD, PhD

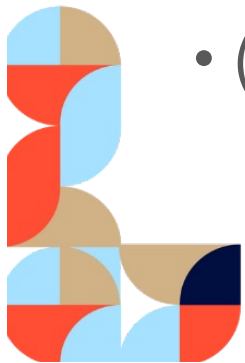
Fellow of the European Society of Cardiology
Doutora em Cardiologia pelo InCor – FMUSP
Coordenadora geral da SOCESP – Regional Osasco
Médica da área de Imagem Cardiovascular DASA- SP



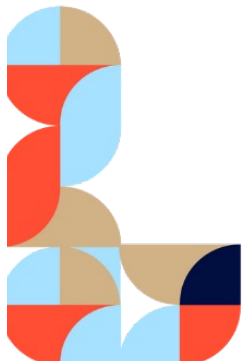
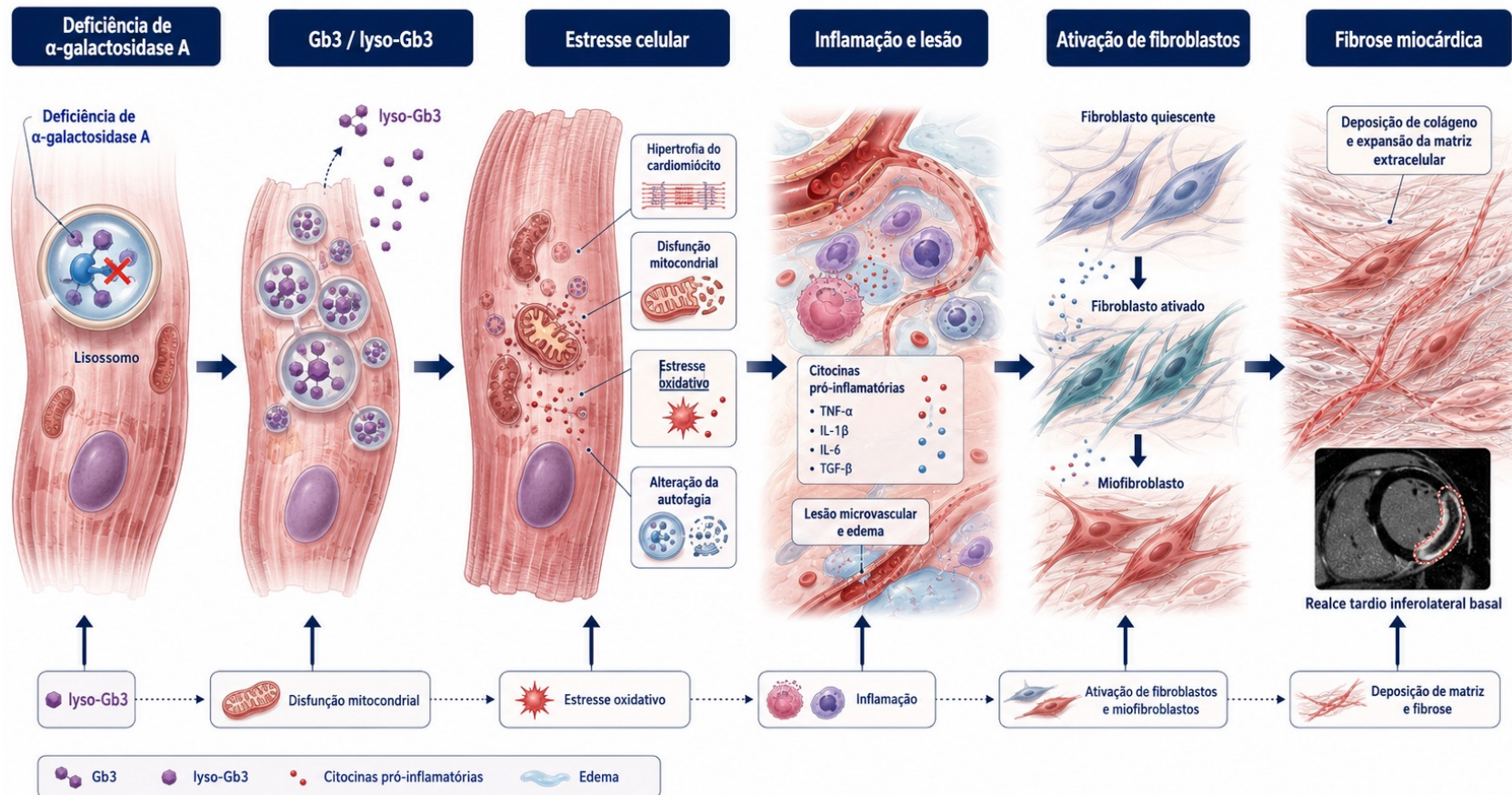
Declaração de Conflito de Interesse

- De acordo com a Norma 1595/2000 do Conselho Federal de Medicina e a Resolução 96/2008 da Vigilância Sanitária, declaro que:

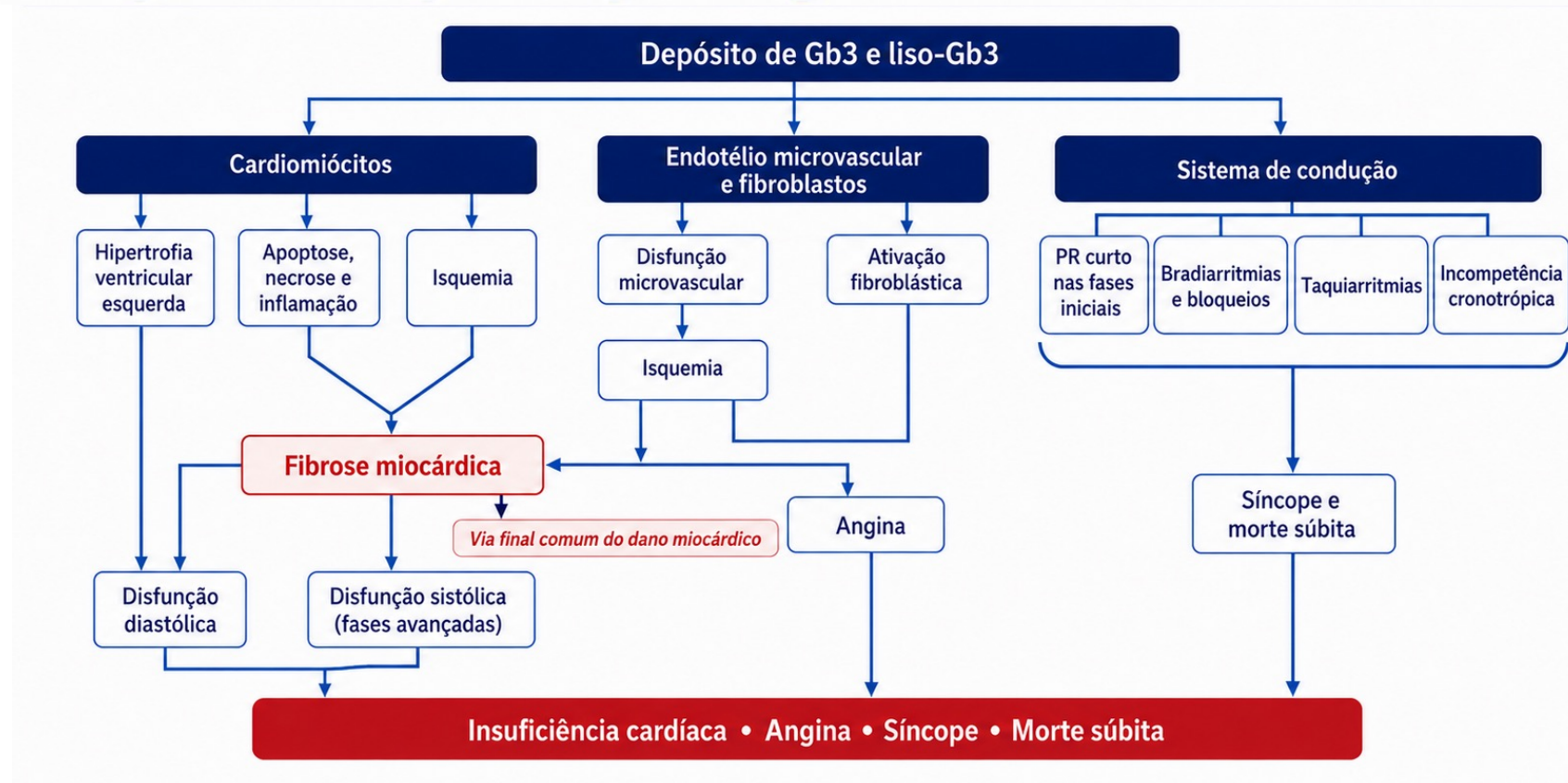
- (X) Escrevo material científico para a Takeda Brasil



Do acúmulo lisossômico à fibrose miocárdica



Doença de Fabry: fisiopatologia do acometimento cardíaco



Qual o diagnóstico?

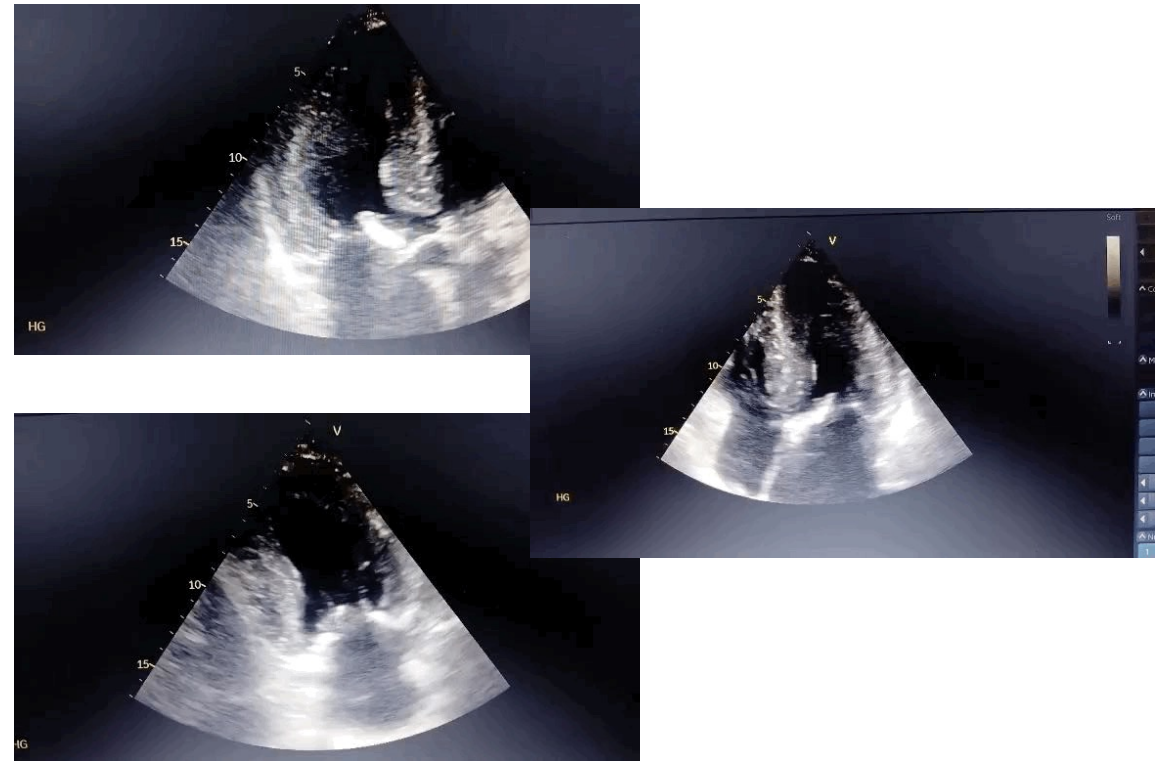
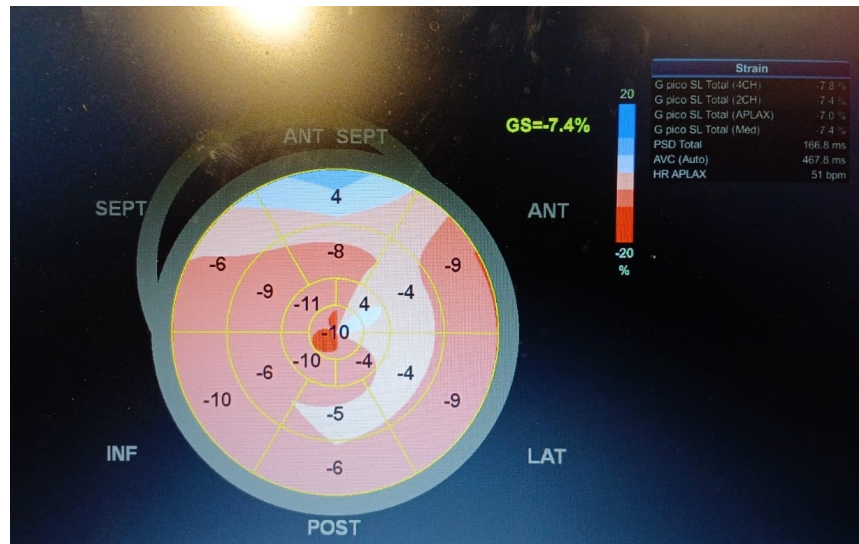
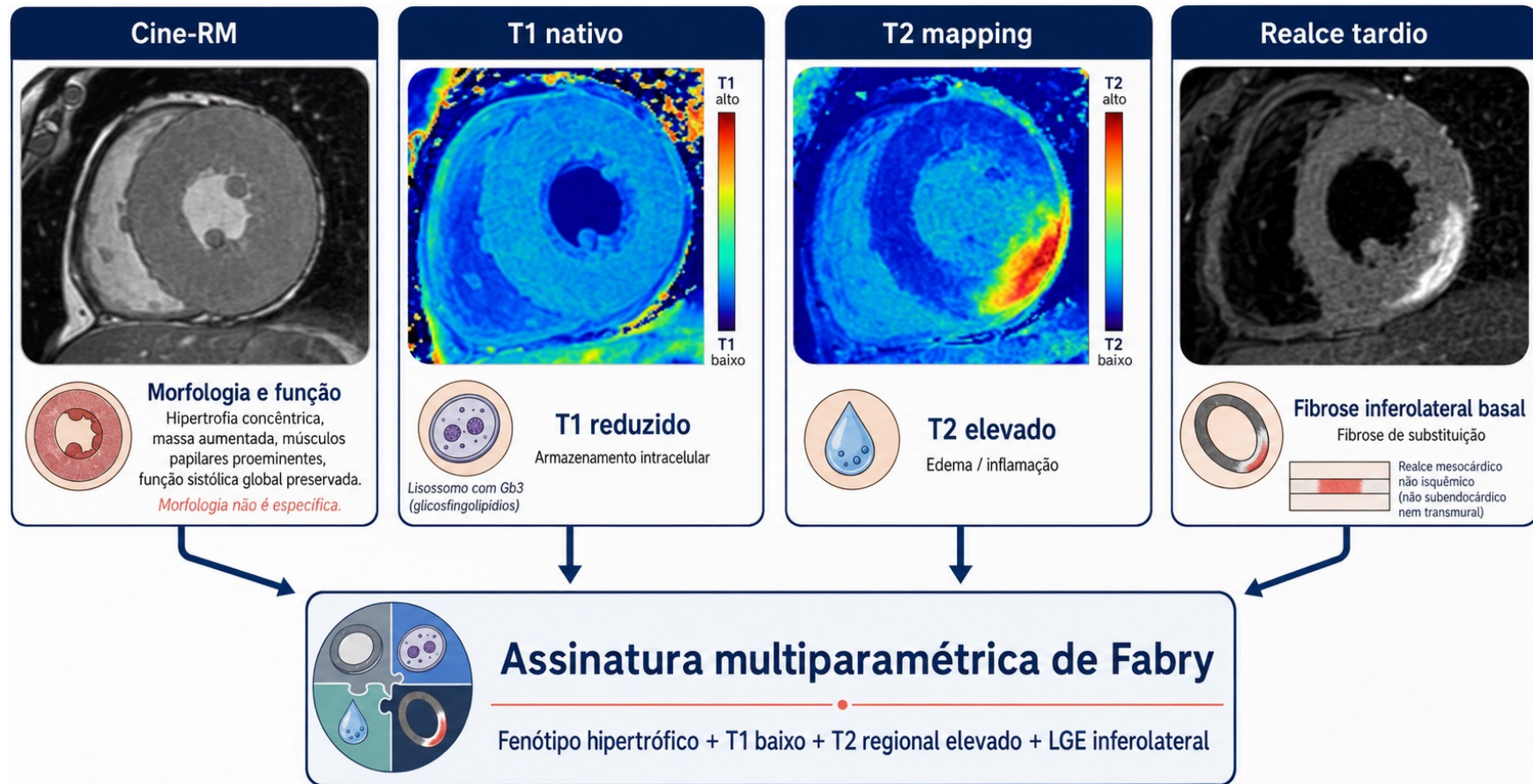


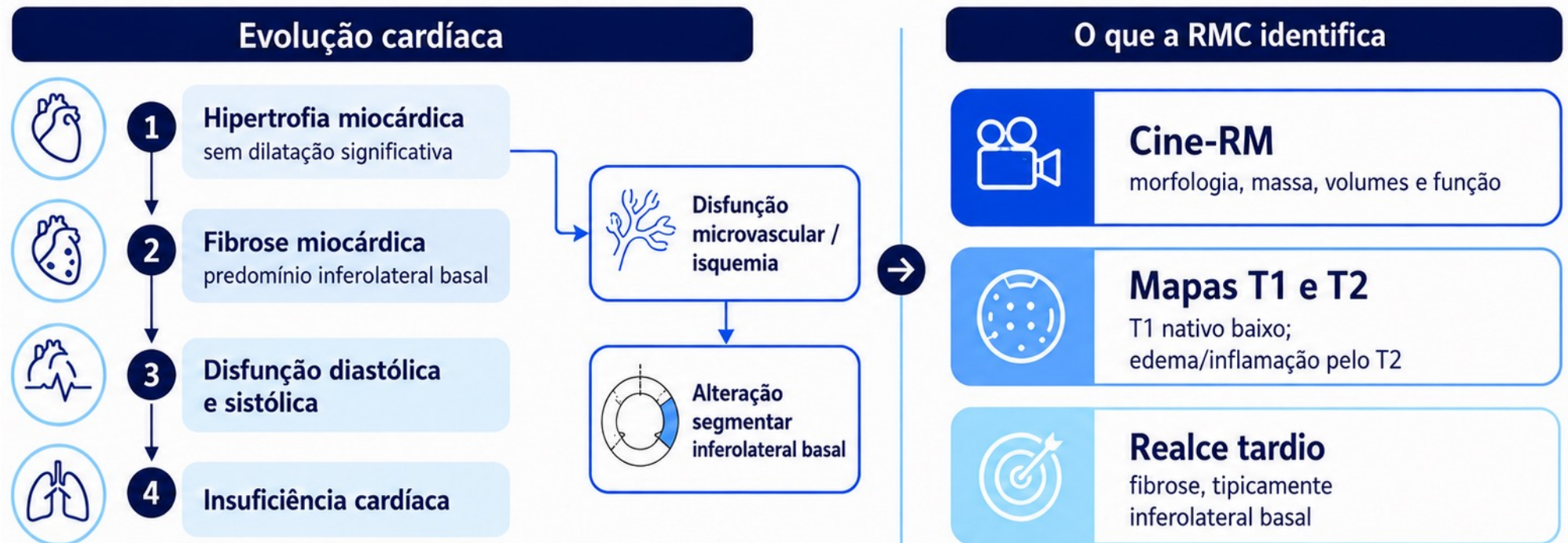
Imagem cedida como cortesia pelo Dr. Lício Volpolini



RMC: assinatura multiparamétrica de Fabry



Cardiomiopatia de Fabry: evolução e avaliação pela RMC

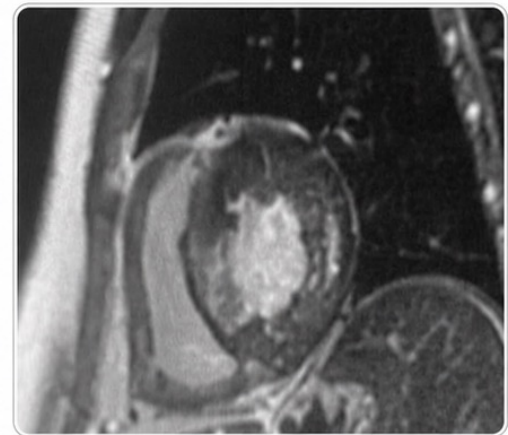
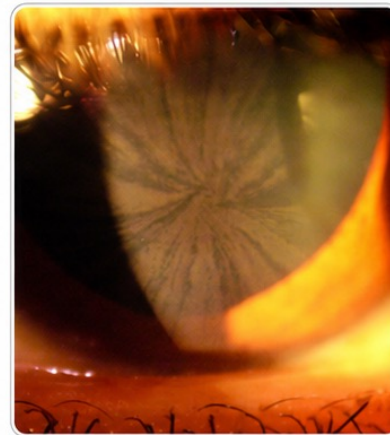
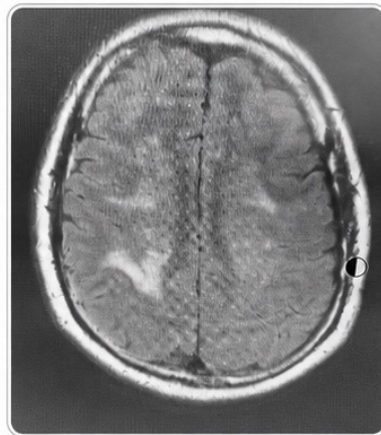
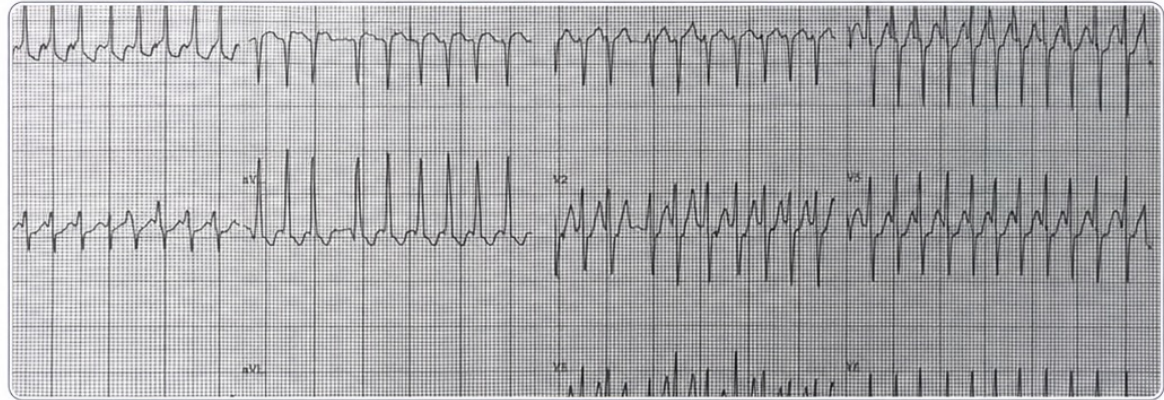


A RMC integra fenótipo, atividade inflamatória e fibrose — do diagnóstico precoce à estratificação prognóstica.



Caso clínico:

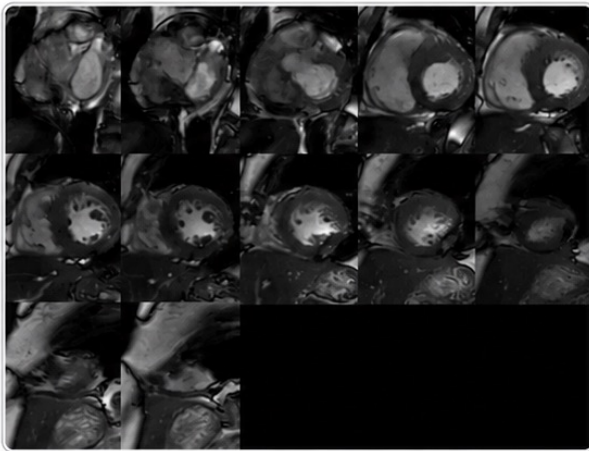
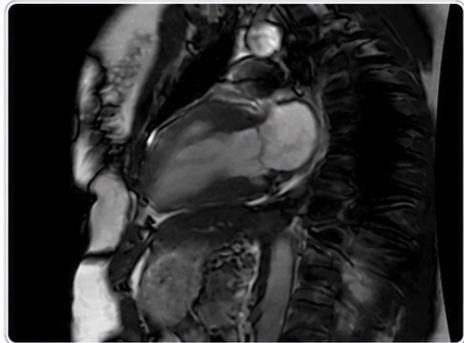
Homem, 49 anos,
no PS com taquiarritmia



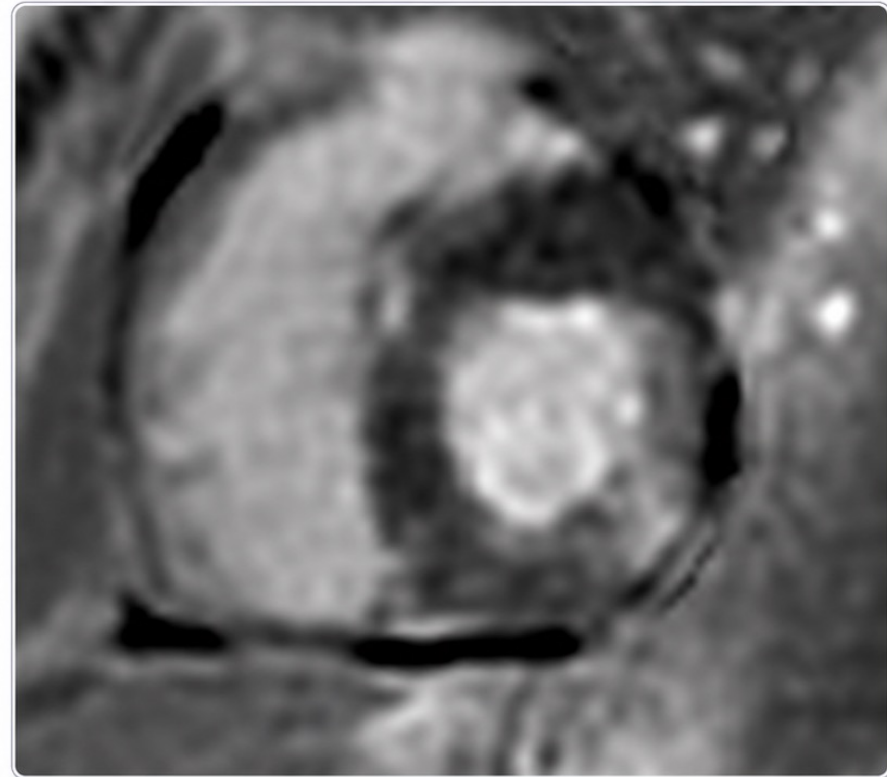
Imagens cedidas como cortesia pela Dra. Edileide Barros



Cine-RM



Realce tardio e mapa T1

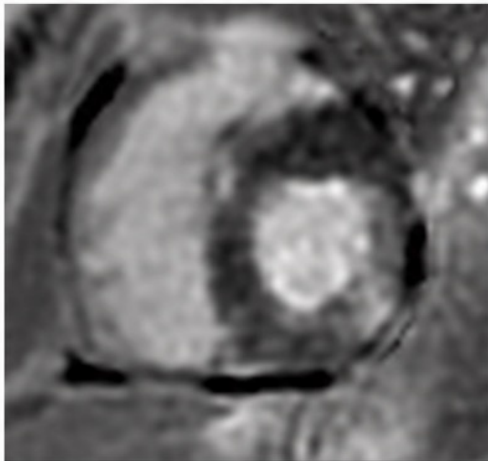


Imagens de arquivo pessoal



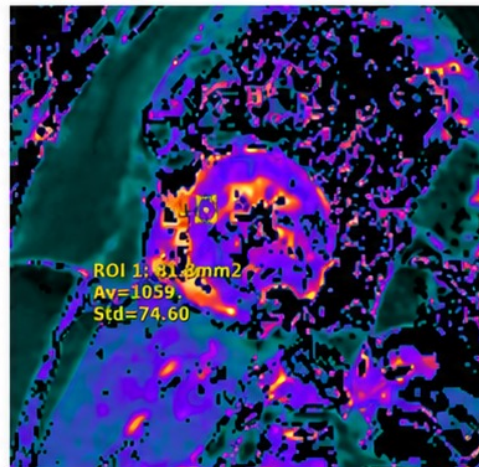
RMC multiparamétrica na doença de Fabry

REALCE TARDIO



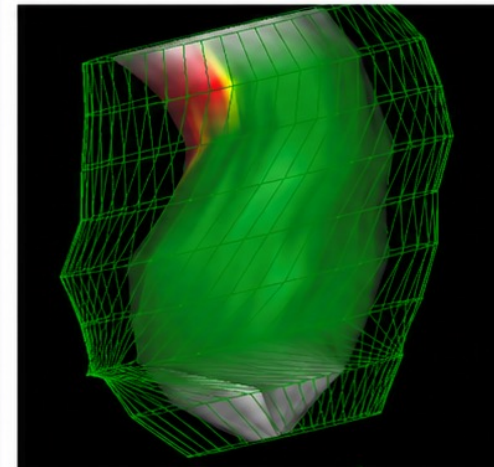
Fibrose inferolateral basal

MAPA T1 NATIVO



Caracterização do depósito miocárdico

STRAIN MIOCÁRDICO

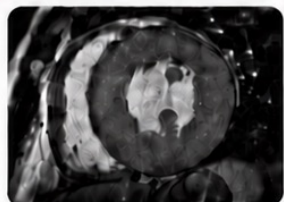
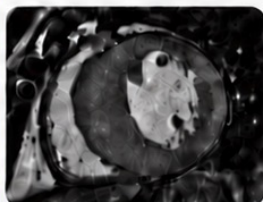
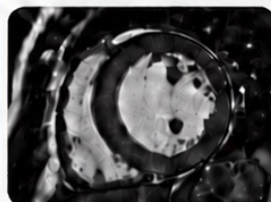
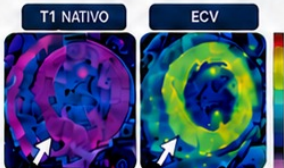
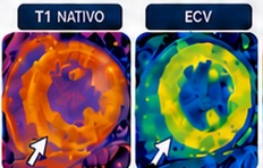
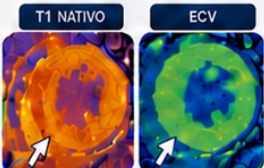
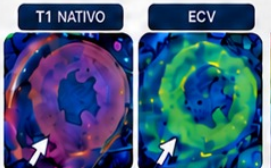
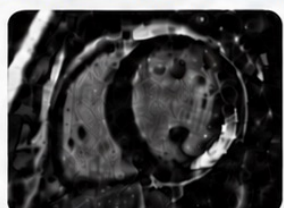
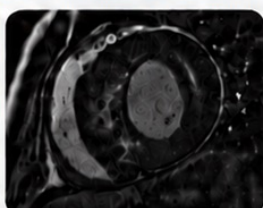
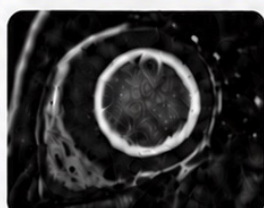
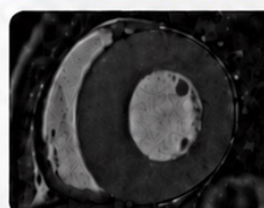
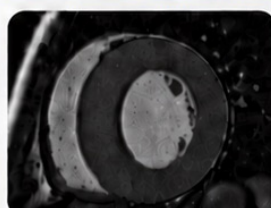


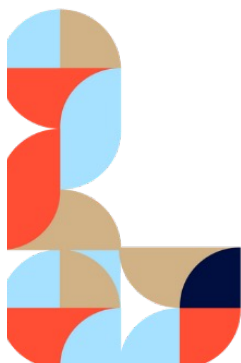
Disfunção regional subclínica

Estrutura + tecido + função: diferentes estágios da cardiomiopatia de Fabry

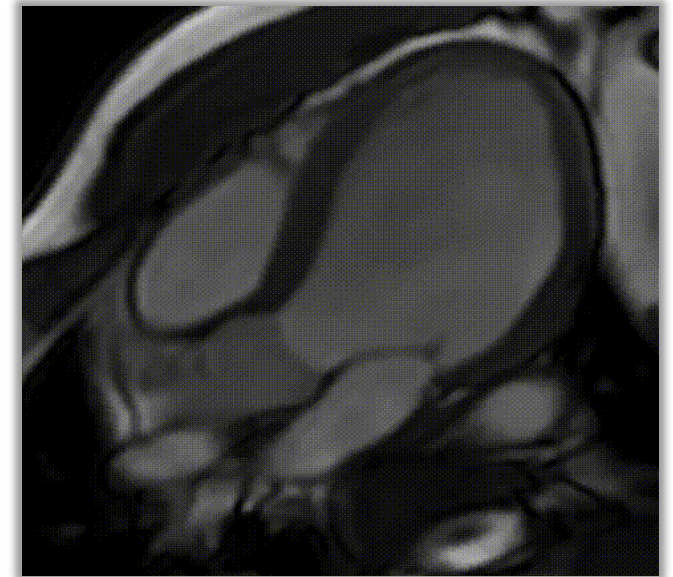
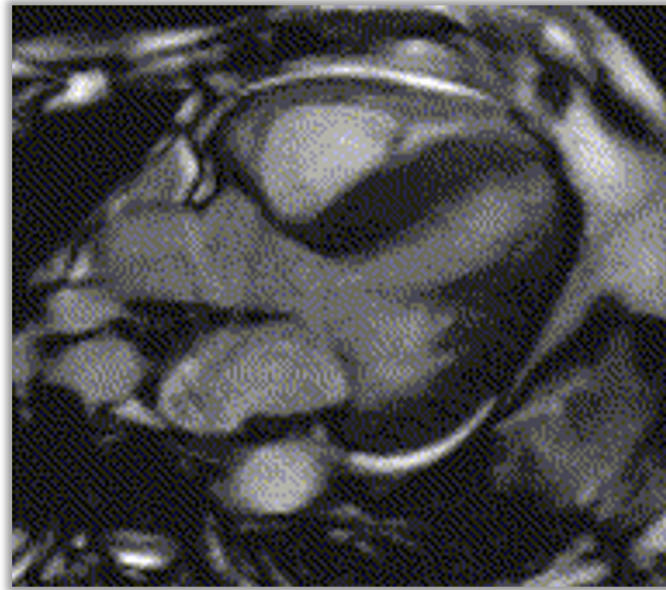


Diagnóstico diferencial dos fenótipos hipertróficos pela RMC

	FABRY	CMH SARCOMÉRICA	AMILOIDOSE	CARDIOPATIA HIPERTENSIVA	CORAÇÃO DE ATLETA
 1) CINE-RM / MORFOLOGIA (eixo curto)	 HVE geralmente concêntrica; músculos papilares proeminentes	 Hipertrofia assimétrica ou apical; criptas podem ocorrer	 Espessamento concêntrico, frequentemente biventricular	 HVE leve e cavidades aumentadas	
 2) T1 NATIVO E ECV	T1 NATIVO ECV  T1 ↓ ECV normal no início T1 pode pseudonormalizar com fibrose	T1 NATIVO ECV  T1 normal/↑ ECV ↑ variável	T1 NATIVO ECV  T1 ↑↑ ECV ↑↑ Anulação miocárdica difícil	T1 NATIVO ECV  T1 normal/↑ discreto ECV normal/↑	T1 NATIVO ECV  T1 e ECV geralmente normais ou discretamente reduzidos
 3) PADRÃO DE REALCE TARDIO (LGE)	 Mesocárdio inferolateral basal	 Segmentos hipertrofiados e inserções do VD	 Subendocárdico difuso ou transmural	 Ausente ou focal inespecífico	 Habitualmente ausente



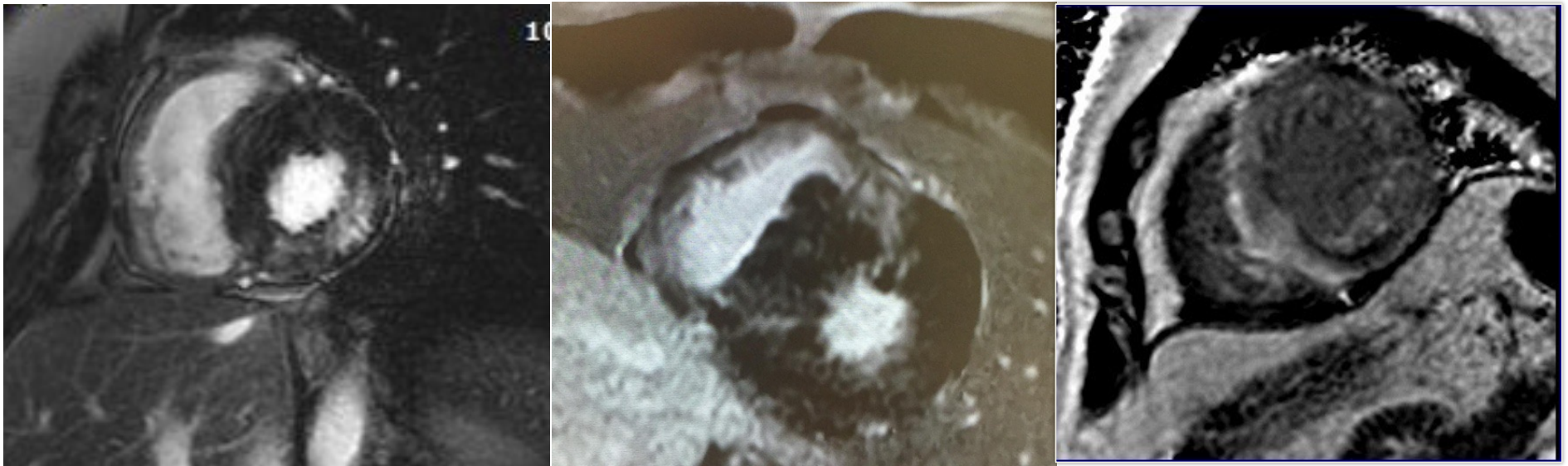
Cine-RM diferencia as hipertrofias?



Imagens de arquivo pessoal



Realce tardio pode diferenciar os fenótipos hipertróficos!

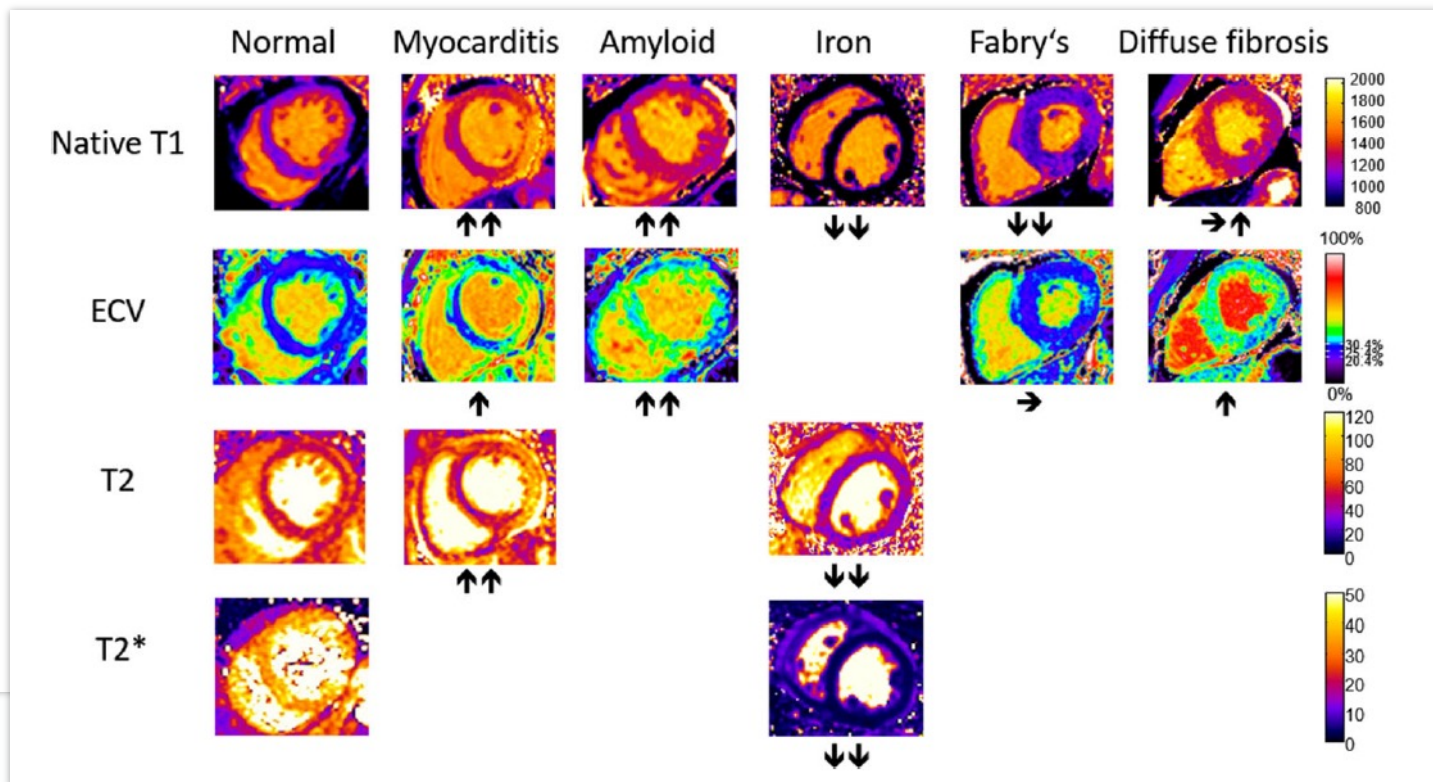


Imagens de arquivo pessoal



Messroghli et al. *Journal of Cardiovascular Magnetic Resonance* (2017) 19:75
DOI 10.1186/s12968-017-0389-8

Journal of Cardiovascular
Magnetic Resonance



RMC como classe I na avaliação das cardiomiopatias!



ESC

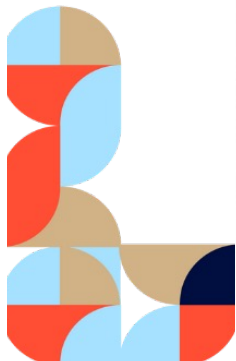
European Heart Journal (2023) 00, 1–124
European Society of Cardiology <https://doi.org/10.1093/eurheartj/ehad194>

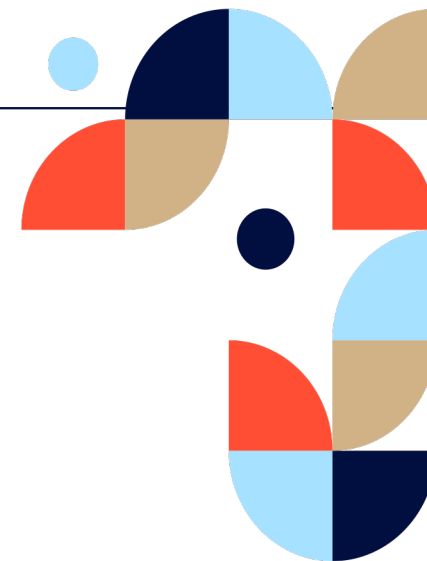
ESC GUIDELINES

2023 ESC Guidelines for the management of cardiomyopathies

Developed by the task force on the management of cardiomyopathies of the European Society of Cardiology (ESC)

Mensagens Finais





Obrigada!

mauellendahl@gmail.com



+55 11 993260206



Nesta apresentação foram utilizadas figuras criadas com o apoio de IA generativa (Chat GPT/OpenAI) concebidas e editadas pela autora.