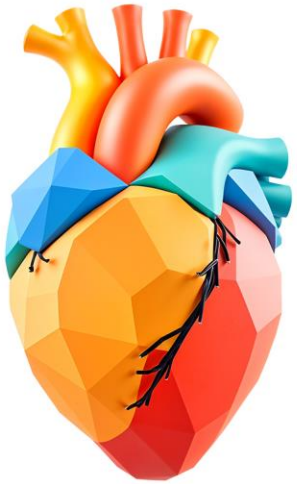

Critical Cardiovascular Care

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

Advanced heart failure in a patient with good LVEF

Nasim Naderi MD,FESC

Professor of cardiology-Heart failure specialist

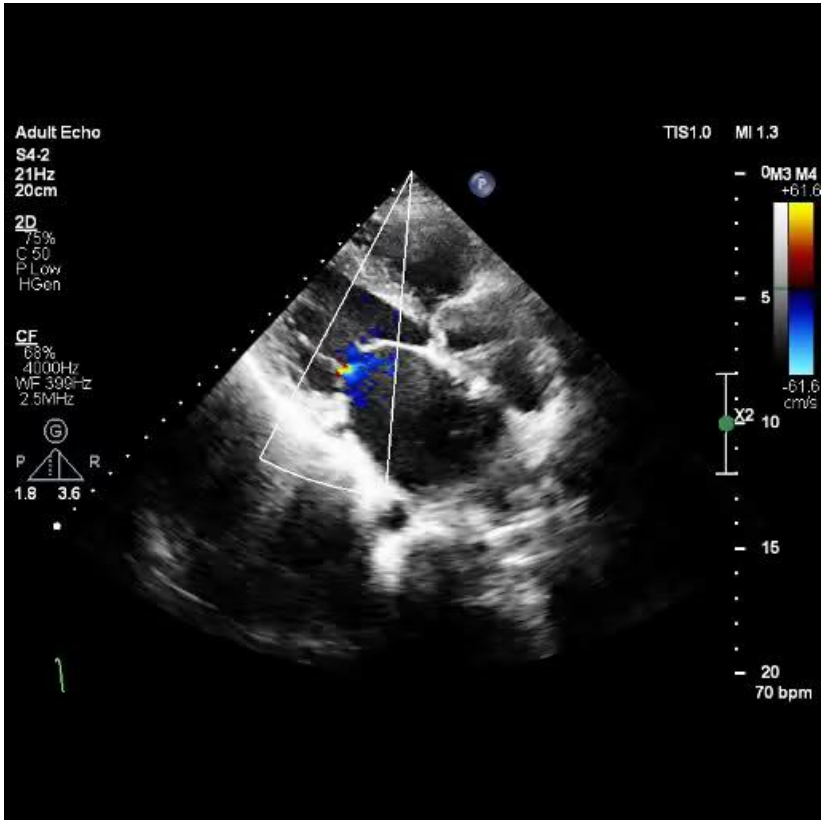
Rajaie Cardiovascular institute

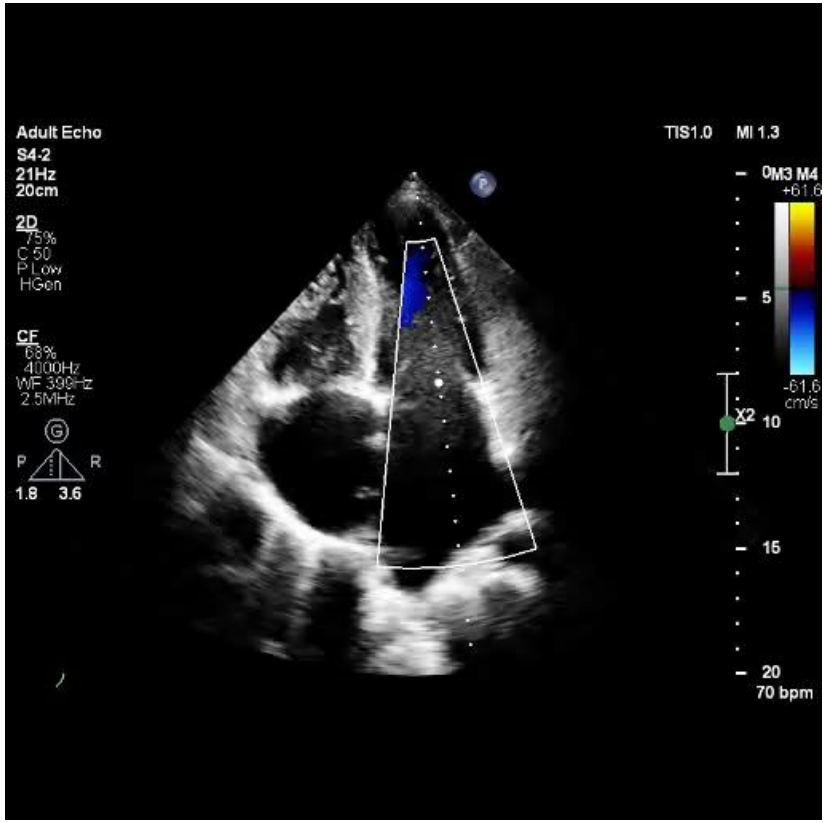
Case Presentation

- 32 years old lady
- History of hypertrophic heart syndrome and CHB since childhood
- No FHx
- PPM→ICD
- AF/pace rhythm

- Her symptoms were increased as frequent congestive symptoms at 32 years
- Easily controlled by increasing doses of diuretics
- Her congestive symptoms were increased more and more following several episodes of COVID-19

ECHO


Adult Echo
S4-2
21Hz
20cm
2D
75%
C 50
P Low
HGen
CF
68%
4000Hz
WF 398Hz
2.5MHz
TIS1.0
MI 1.3
M3 M4
+61.6
5
-61.6
cm/s
10
15
20
70 bpm
1.8
3.6


Adult Echo
S4-2
21Hz
20cm
2D
75%
C 50
P Low
HGen
CF
68%
4000Hz
WF 398Hz
2.5MHz
TIS1.0
MI 1.3
M3 M4
+61.6
5
-61.6
cm/s
10
15
20
70 bpm
1.8
3.6

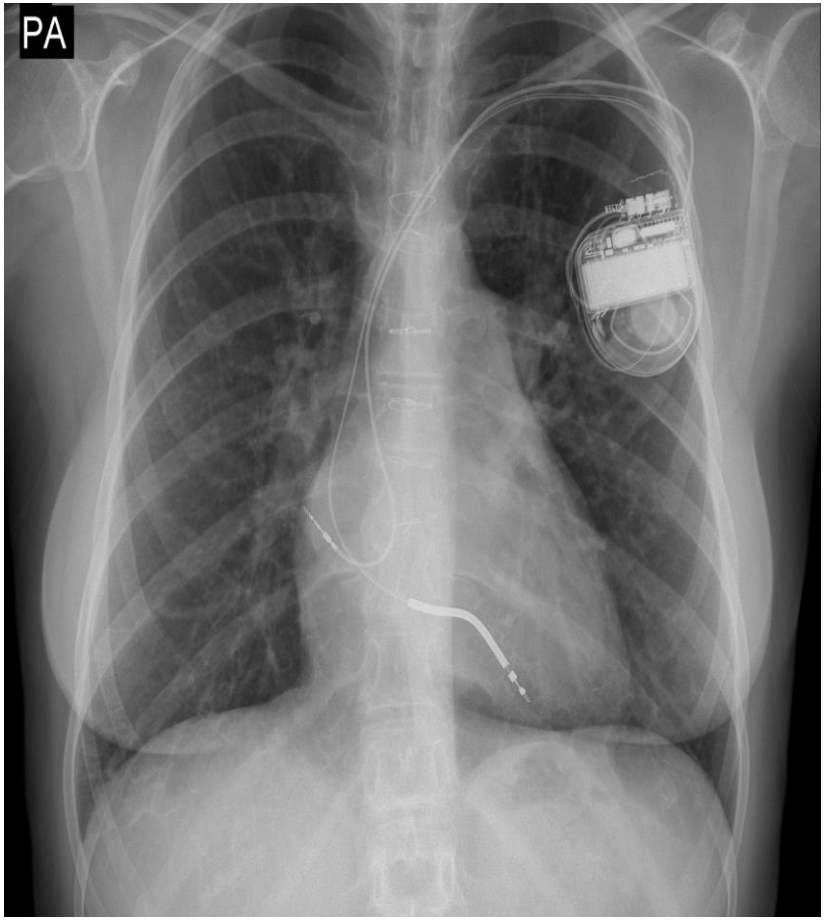

Adult Echo
S4-2
45Hz
18cm
2D
78%
C 50
P Low
HGen
TIS0.5
MI 1.3
0M3
5
X2
10
15
71 bpm
P
R
1.8
3.6

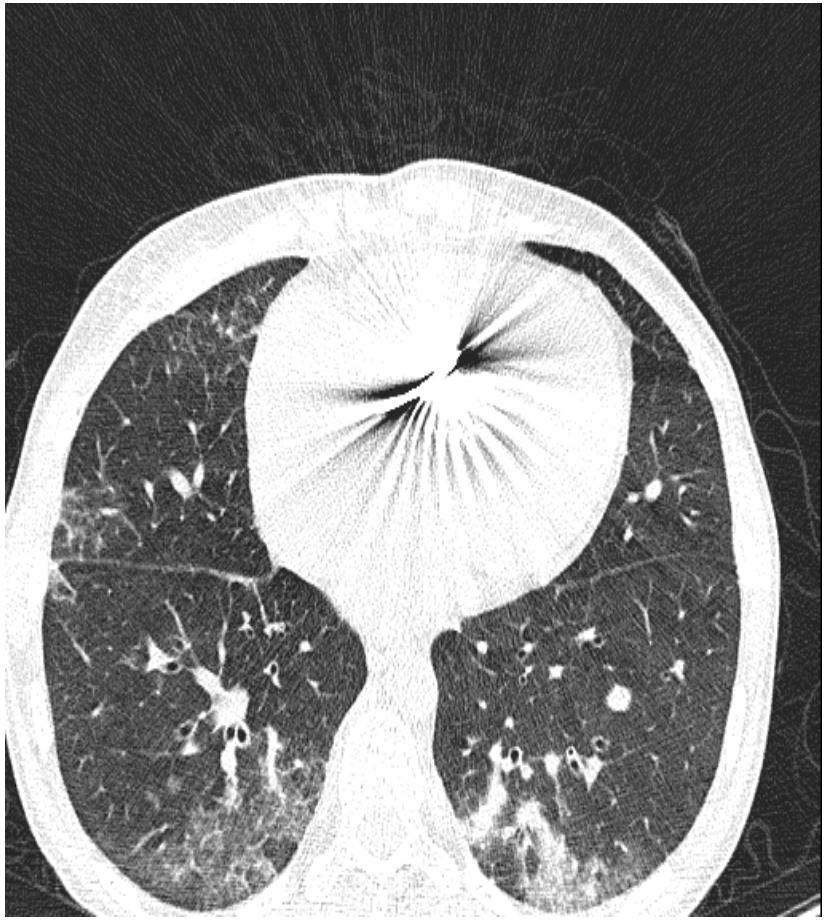
- The patient got listed
- Liver and Kidney were normal
- Surgery consult for HTX ??
- Pulmonologist consult ??

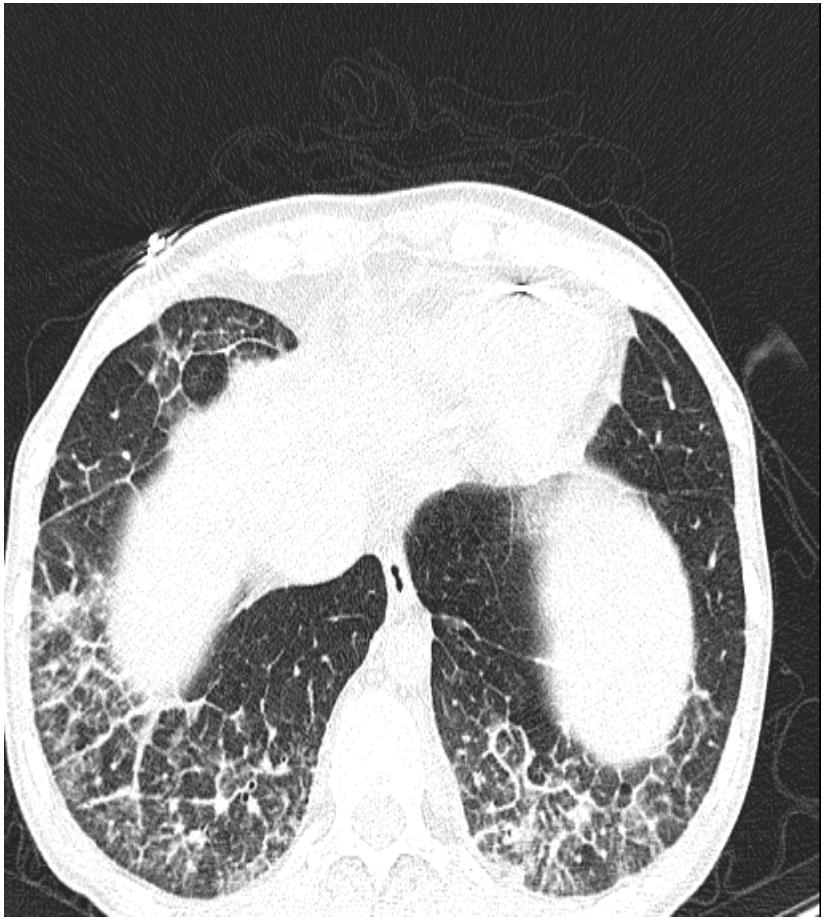
RHC

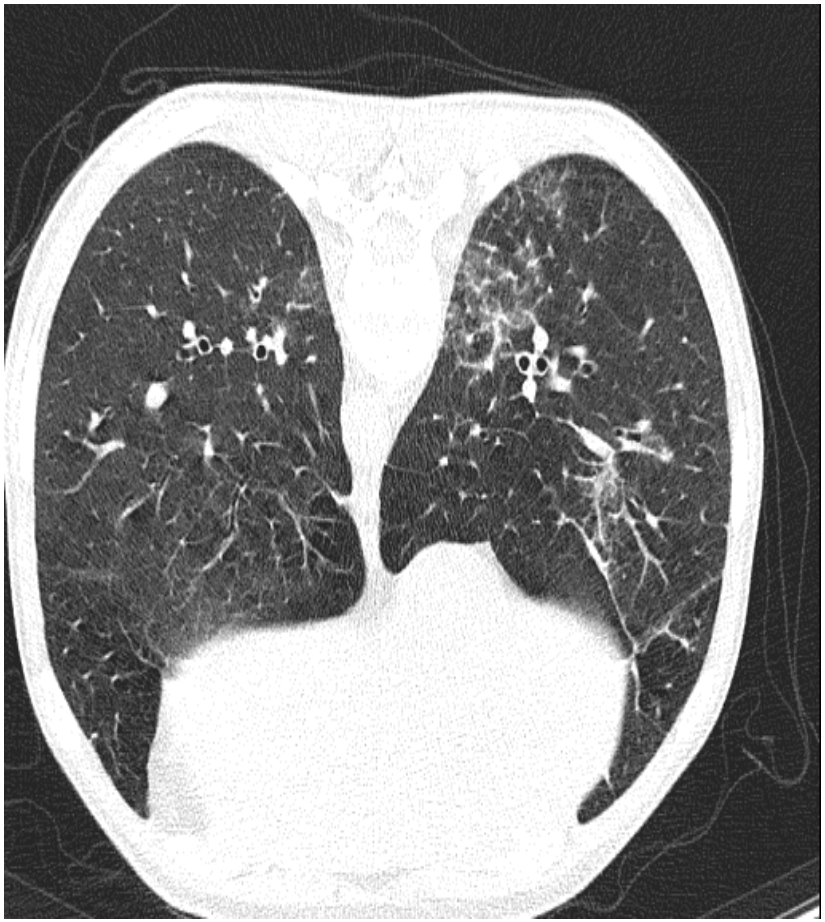
- CO:2.66 lit/min
- CI=1.65 Lit/min/BSA
- CVP=20 mmHg
- PAP=50/25(32)
- PCWP=19
- Systemic O2 Sat=95%
- Mixed venous O2 sat=49%
- PVR=4.88
- SVR=25.9
- BP=113/77(89)
- HR=70 (pace rhythm)

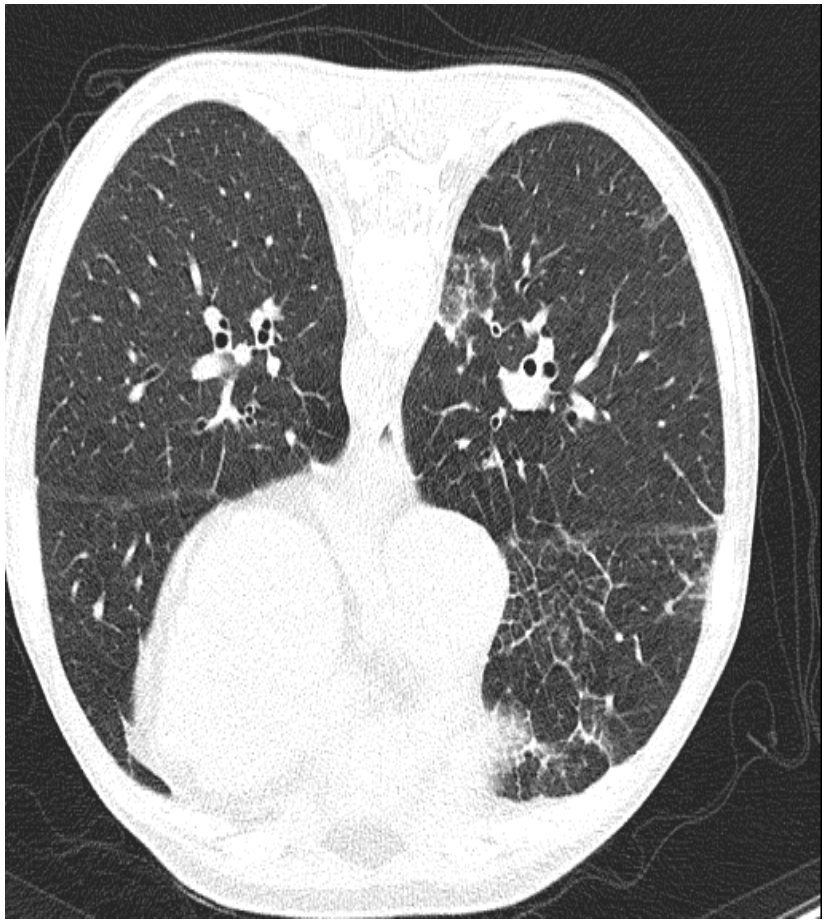
Chest CT

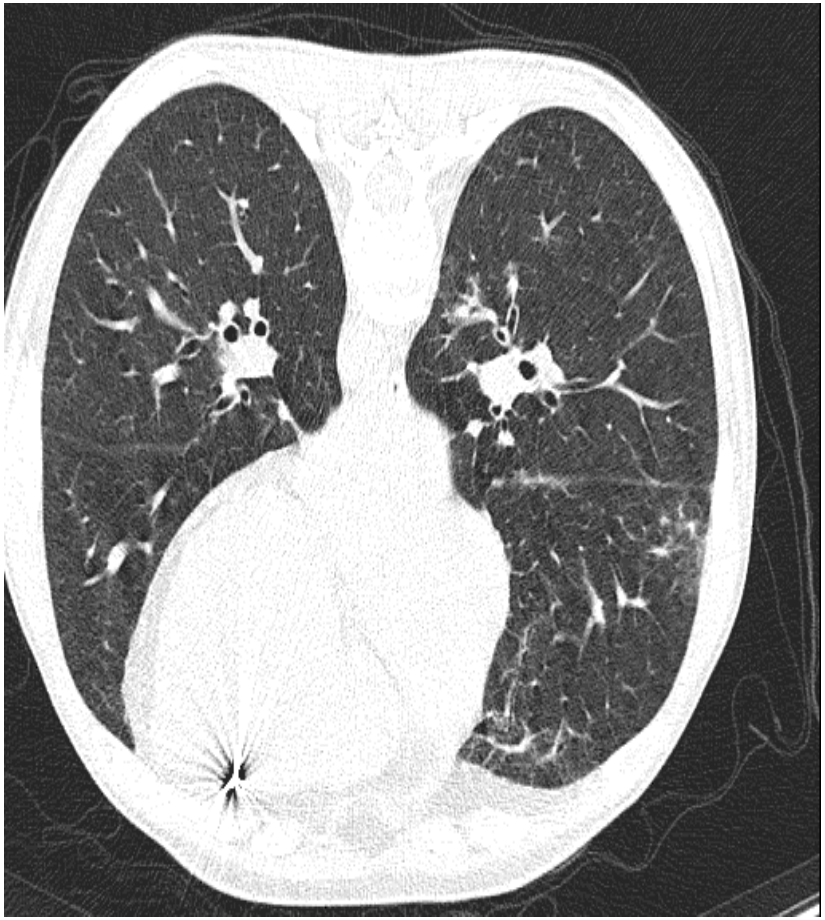


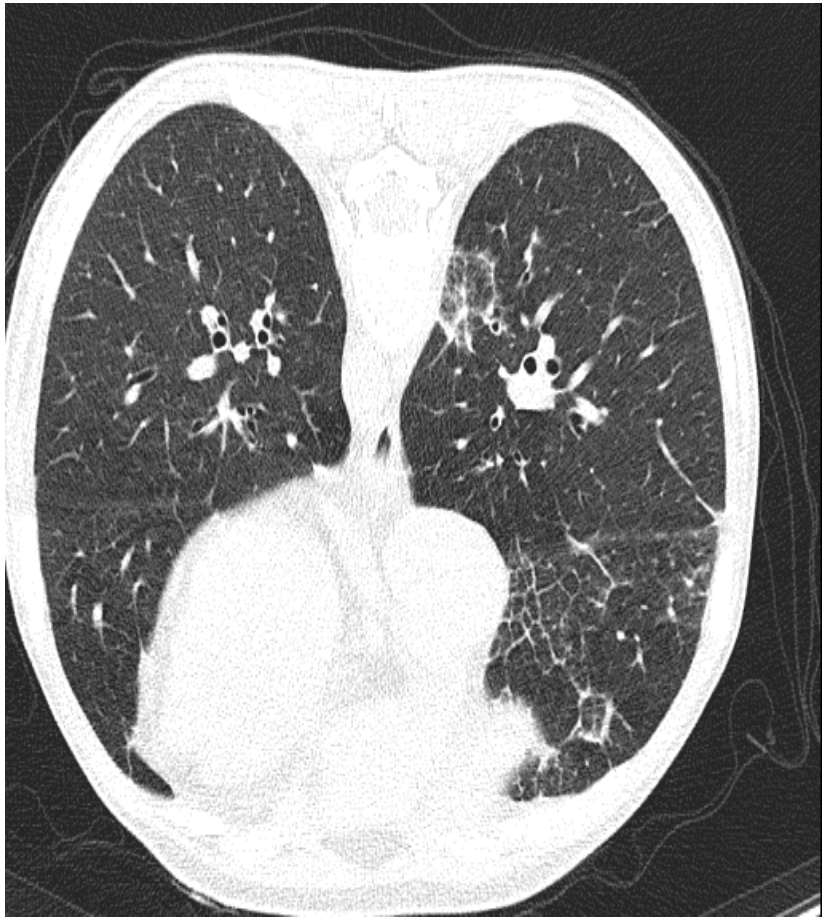




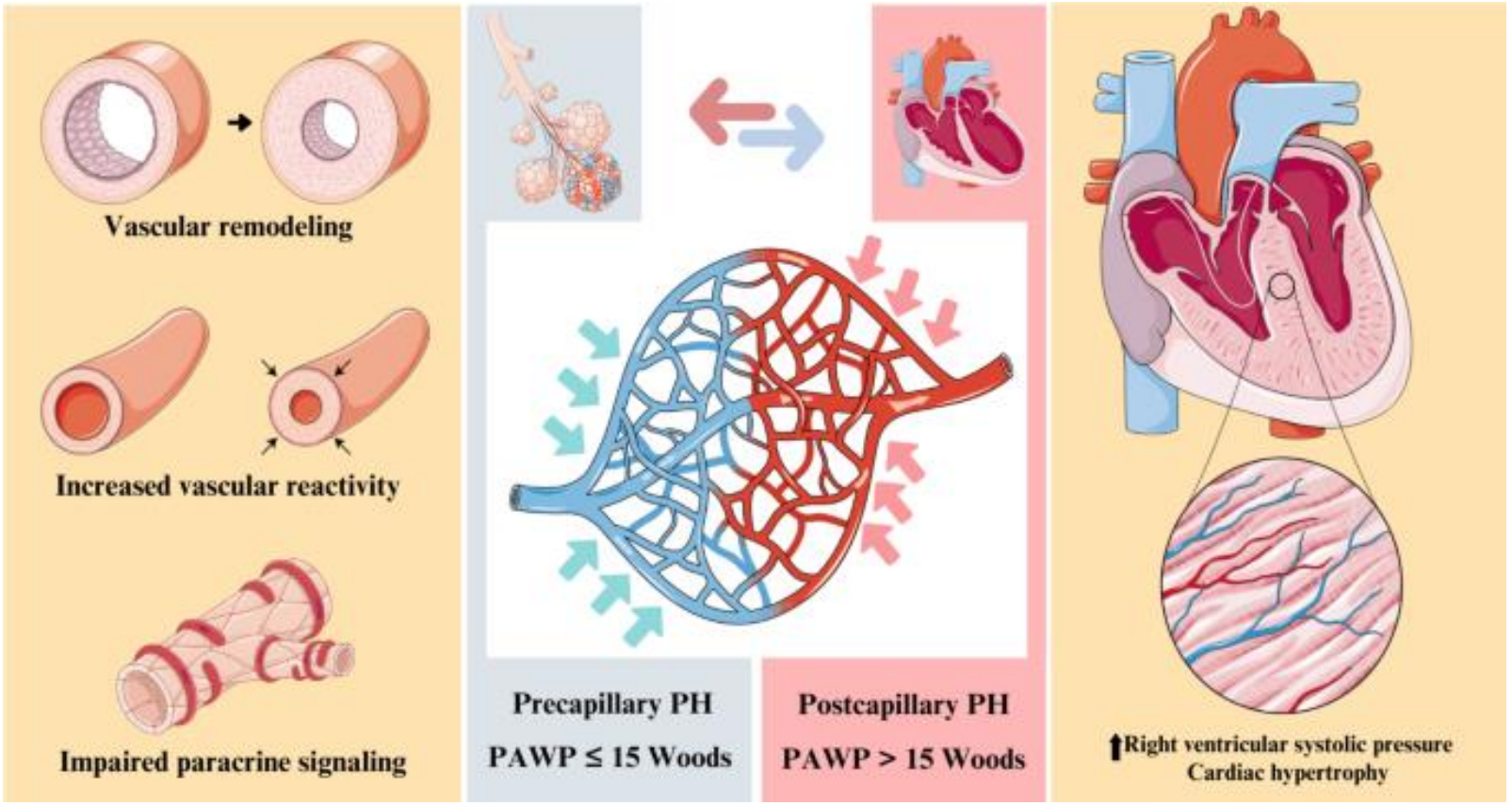




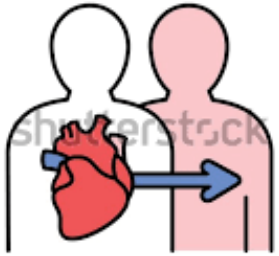




What about High PVR???



The patient Got Transplanted



- **Early post op. Coarse**

JACC: HEART FAILURE VOL. 11, NO. 11, 2023

PUBLISHED BY ELSEVIER

STATE-OF-THE-ART REVIEW

Advanced Heart Failure Therapies for Hypertrophic Cardiomyopathy

State-of-the-Art Review and an Updated Analysis From UNOS

Lusha W. Liang, MD, MSc,* Heidi S. Lumish, MD,* Lorenzo R. Sewanan, MD, PhD, Yuichi J. Shimada, MD, MPH, Mathew S. Maurer, MD, Shepard D. Weiner, MD, Gabriel Sayer, MD, Nir Uriel, MD, MSc, Kevin J. Clerkin, MD, MSc

- Advanced heart failure among patients with HCM is underappreciated
- 1 in 15 patients with HCM will progress to advanced heart failure.

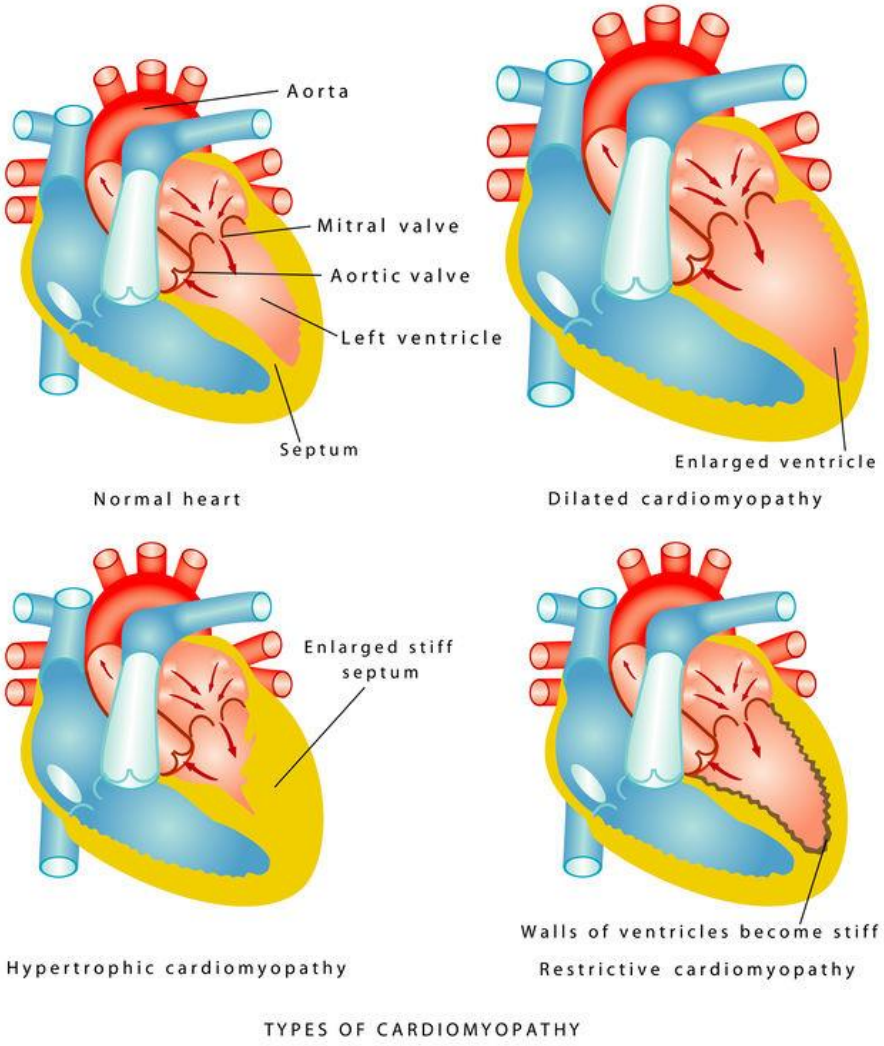


TABLE 1 OPTN/UNOS Review Board Guidance for Hypertrophic/Restrictive Cardiomyopathy Exception Requests

Status 2 Exception Criteria	Status 3 Exception Criteria
1. Continuous hemodynamic monitoring with a pulmonary artery catheter 2. The candidate is on maximally tolerated inotropic dosages 3. Two markers of hemodynamic instability or 1 marker of hemodynamic instability and 1 marker of end-organ dysfunction Hemodynamic instability: • Systolic BP <90 mm Hg • LVEDP, RVEDP, or PCWP >20 mm Hg • Cardiac index ≤2.2 L/min/m^2 • Mixed venous saturation <50% • Transpulmonary gradient ≥15 mm Hg • Pulmonary vascular resistance ≥2.5 WUs End-organ dysfunction: • Arterial lactate >2.5 mmol/L • Increase in serum creatinine >50% above baseline • Increase in total bilirubin >50% above baseline • AST or ALT >2 times upper limit of normal	1. Has either: • Invasive pulmonary artery catheter • Daily hemodynamic monitoring to measure cardiac output and left ventricular filling pressures 2. Is supported by continuous inotropic infusion to improve end-organ perfusion/function 3. Prior to initiation of inotropes, demonstrated evidence of decompensated heart failure, as evidenced by *at least* 2 of the following: • Systolic blood pressure <90 mm Hg • Left or right atrial pressure, left or right ventricular end-diastolic pressure, or pulmonary capillary wedge pressure >20 mm Hg • TPG ≥15 mm Hg • PVR ≥2.5 WUs • Cardiac index <1.8 L/min

ALT = alanine aminotransferase; AST = aspartate aminotransferase; BP = blood pressure; LVEDP = left ventricular end-diastolic pressure; OPTN = Organ Procurement and Transplantation Network; PCWP = pulmonary capillary wedge pressure; PVR = pulmonary vascular resistance; RVEDP = right ventricular end-diastolic pressure; TPG = transpulmonary gradient; UNOS = United Network for Organ Sharing.

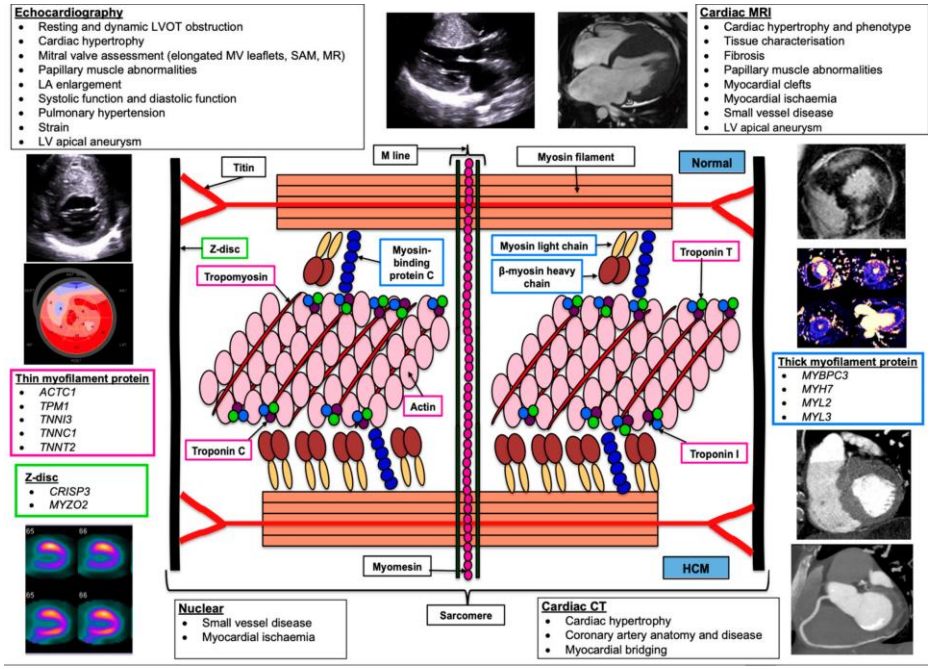
TABLE 2 Hemodynamics of Candidates at the Time of Listing for Heart Transplantation Since Adoption of the 2018 Heart Allocation System in the United States

	HCM	All Others	*P* Value
PA systolic, mm Hg	40 (32-50)	40 (30-51)	0.41
PA diastolic, mm Hg	20 (14-25)	20 (14-26)	0.94
PCWP, mm Hg	19 (13-24)	18 (12-25)	0.11
PVR, WU	2.25 (1.54-3.33)	2.16 (1.43-3.14)	0.009
Cardiac index, L/min/m^2	1.97 (1.68-2.34)	2.08 (1.73-2.49)	<0.001

Values are median (IQR).

HCM = hypertrophic cardiomyopathy; PA = pulmonary artery; other abbreviations as in Table 1.

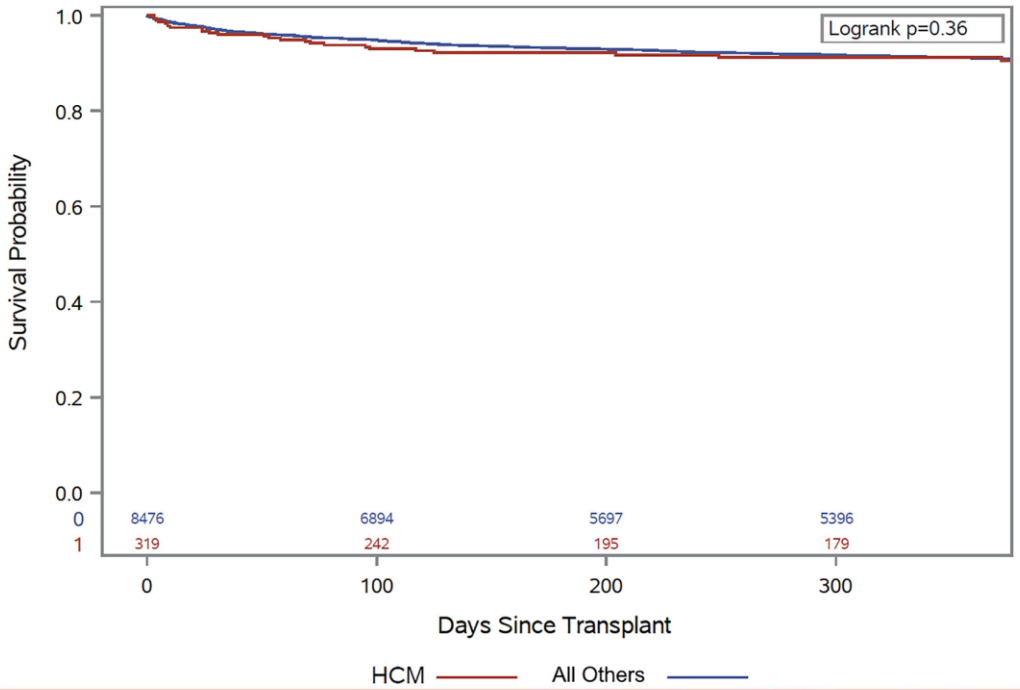
- The diagnosis of advanced heart failure among patients with HCM requires the integration of multiple diagnostic tests
- echocardiography,
- cardiac magnetic resonance,
- right heart catheterization,
- CPET



Journal of Clinical Medicine, *13*(3), 842.
https://doi.org/10.3390/jcm13030842

- LVADs ???
- patients with smaller left ventricles (LVEDD <5 cm) have poor outcomes.

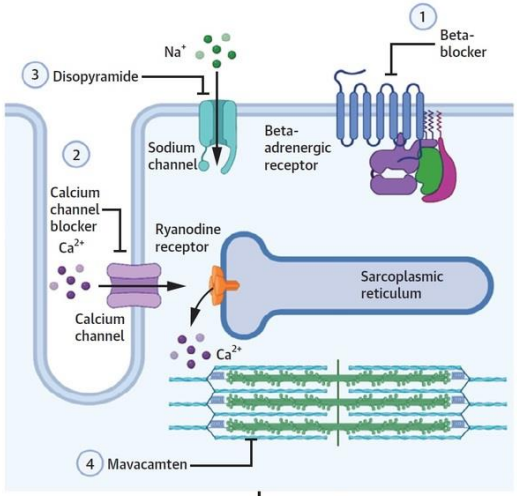
- Heart transplantation is the best heart replacement therapy for patients with hypertrophic heart syndrome
- Post transplantation outcomes are excellent.



- Latent precapillary PH places patients at an increased risk post-transplantation right ventricular dysfunction.
- Following transplantation, the donor heart is exposed to the pretransplantation medications that remain in the recipient's system.

CENTRAL ILLUSTRATION Therapeutics for HCM Across the Disease Spectrum

Medical Management or Septal Reduction Therapy



Progression to Advanced Heart Failure

LVAD | **Heart Transplant**

Potential Challenges

- Arrhythmia from suction events
- Device thrombosis
- Low flow from cannula obstruction

Potential Challenges

- Post-transplant right ventricular dysfunction due to elevated PVR
- Post-transplant graft dysfunction due to pretransplant negative inotropic medications

Liang LW, et al. J Am Coll Cardiol HF. 2023;11(11):1473-1480.

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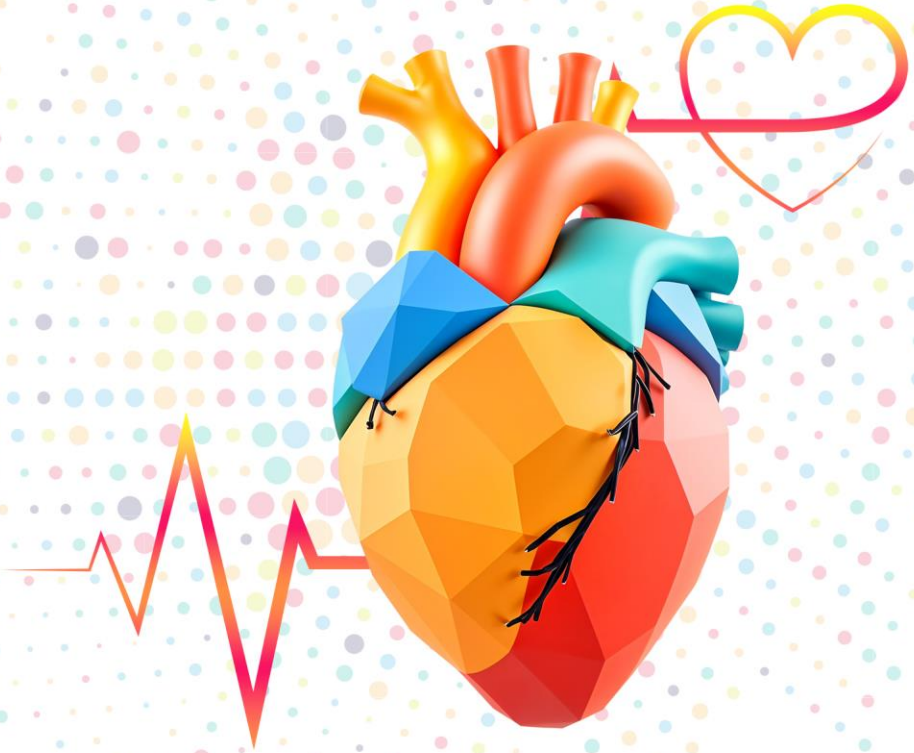

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