

# HHP Care Model and Disease Management Webinar Series

## Heart Failure: Case Review

Thursday, December 9, 2021

5:30pm – 6:30pm

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## Moderator

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- You will be able to submit questions via the Q&A section.
  - Due to time constraints, any unanswered questions will be addressed this week and posted on the HHP website
- A recording of the meeting will be available tomorrow on the HHP website and intranet.

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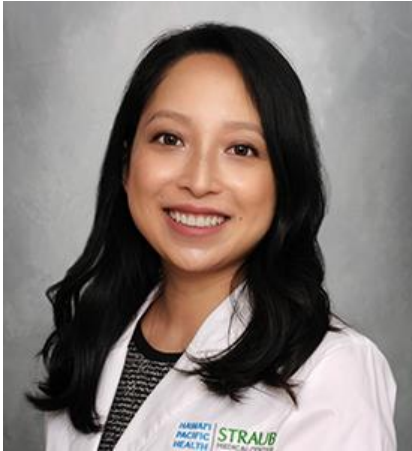


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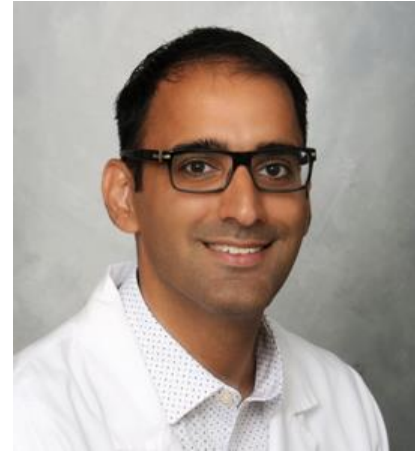
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# Heart Failure: Case Review



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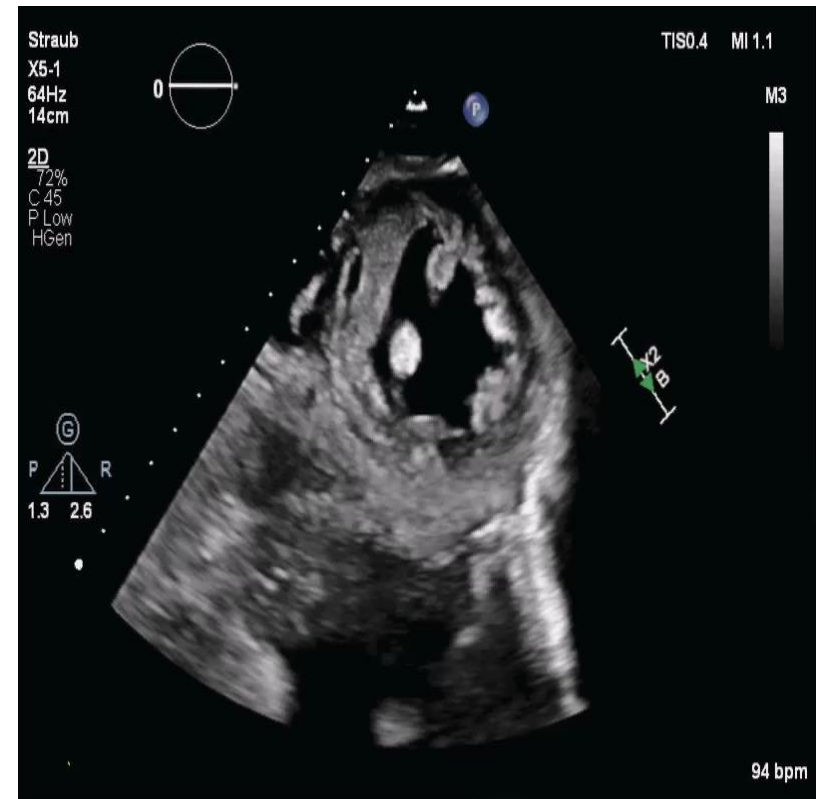
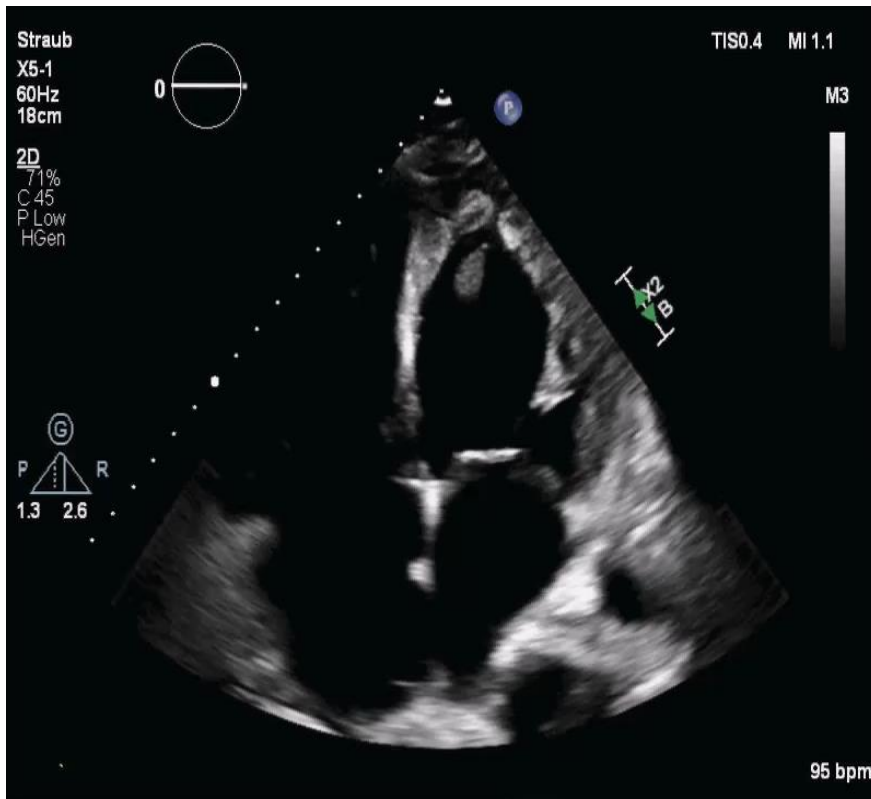
# Case 1

42yoM with history of polysubstance abuse.  
Presented in May 2020 with progressive shortness of breath, left leg pain, edema, and discoloration

- Found to have acute limb ischemia s/p emergent thrombectomy

# Case 1

- TTE: LVEF 15-20%, large apical thrombi, severe pulmonary hypertension



# Case 1

- Hospitalization complicated by hypotension requiring pressors, large pleural effusion s/p thoracentesis, and bilateral DVT
- Meth and alcohol cessation strongly advised
- Discharge medications:
  - Lasix 20mg daily
  - Metoprolol tartrate 25mg BID
  - Lisinopril 5mg
  - Eliquis 5mg BID

# Case 1

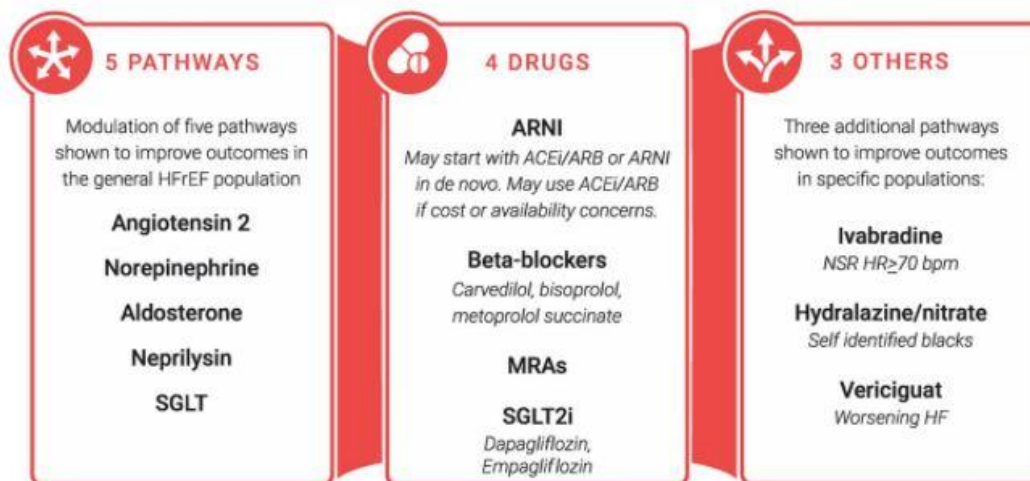
- Lisinopril switched to Entresto 24/26mg BID
- CTA with non-obstructive coronary artery disease
- TTE 12/20: LVEF 10-15%, no LV thrombus, depressed RV, moderate-severe tricuspid regurgitation, pulmonary hypertension

# Case 1

- Hospitalized July 2021 with decompensated heart failure, hypotension and new onset afib with RVR
- Actively using meth
- Started on dobutamine 5mcg/kg/min and Lasix gtt with 25lb diuresis
- Extensive counseling
- Discharge medications:
  - Bumex 1mg BID
  - Metoprolol XL 25mg
  - Entresto 24/26mg BID
  - Spironolactone 25mg
  - Jardiance 10mg

# Guideline Directed Medical Therapy

## Principles and Pathophysiologic Targets of HFrEF Pharmacotherapy



	Initiation and optimization of medication dosing				
CDMMT	Day 1	Days 7-14	Days 14-28	Days 21-42	After day 42
ARNI	Initiate at low dose	Continue	Titrate, as tolerated	Titrate, as tolerated	Maintenance or additional titration of the 4 foundational therapies
$\beta$ -Blocker	Initiate at low dose	Titrate, as tolerated	Titrate, as tolerated	Titrate, as tolerated	Consideration of EP device therapies or transcatheter mitral valve repair
MRA	Initiate at low dose	Continue	Titrate, as tolerated	Continue	Consideration of add-on medications or advanced therapies, if refractory
SGLT2i	Initiate	Continue	Continue	Continue	Manage comorbidities

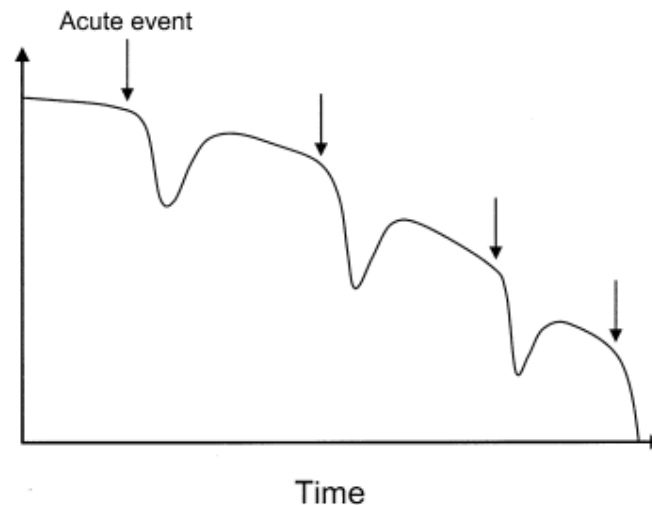
# Simultaneous or Rapid Sequence Initiation

## Benefits with initiating BB+ARNI+MRA+SGLT2i:

- Rapid improvement in health status (within 1-8 weeks)
- Rapid reduction in HF hospitalization (within 2-4 weeks)
- Rapid reduction in HF rehospitalization (within 2-4 weeks)
- Rapid reduction in mortality (within 2-4 weeks)
- Improvement in LVEF (within 12 weeks)

# Hospitalization

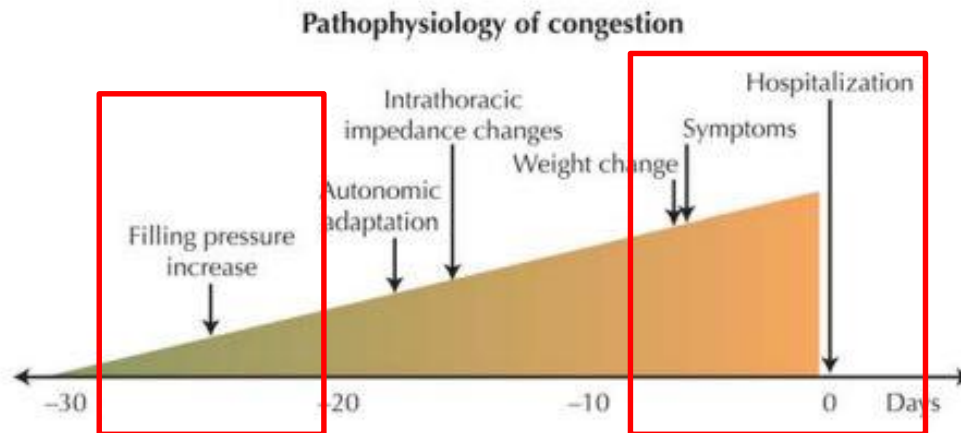
- HF is associated with the highest 30-day readmission rate (20-25%)
- 50% of patients will be admitted at least once within 1 year of diagnosis
- Hospitalization has a clear prognostic significance
  - High 30-day and 5-year mortality





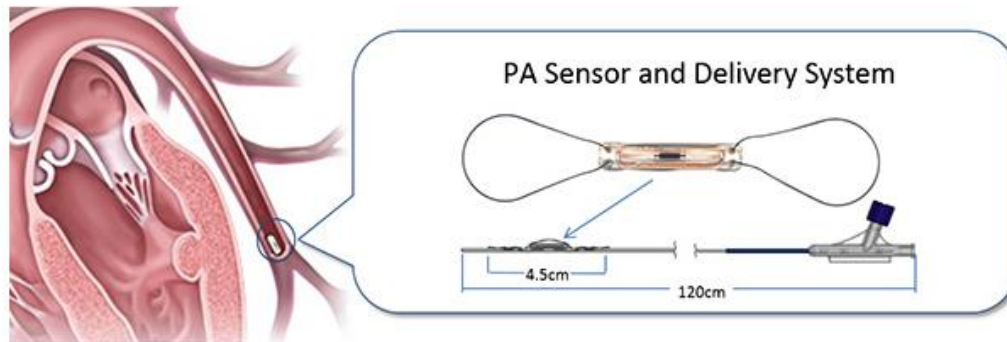
# Hemodynamic Monitoring

- We rely on physical markers to predict clinical deterioration
- Hemodynamic-guided strategy to proactively monitor for worsening heart failure prior to symptom onset



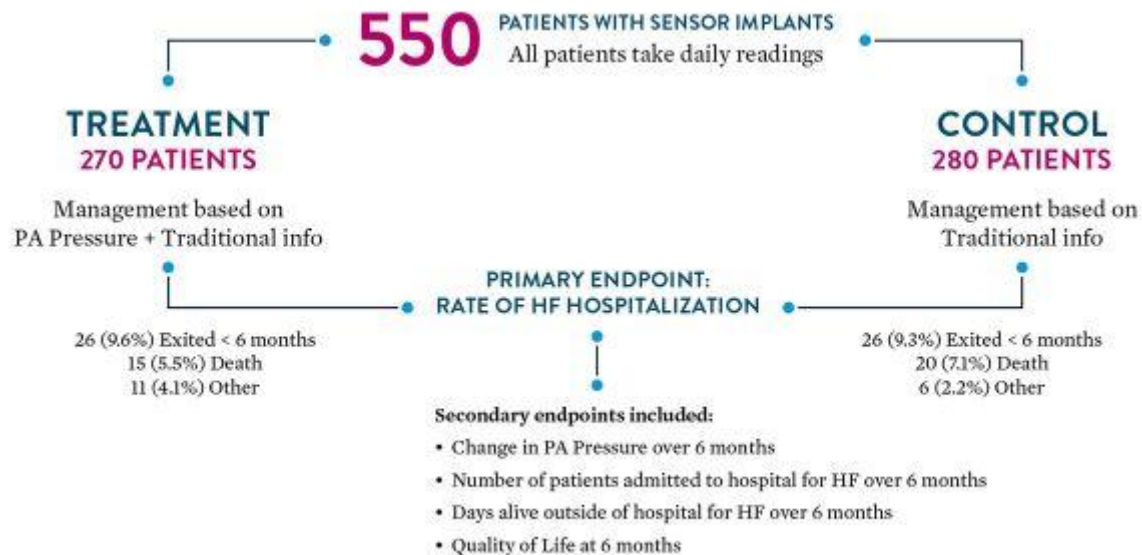
# CardioMEMS

- Implantable, battery-free sensor, implanted into the distal pulmonary artery
- Microelectromechanical system to continuously measure pulmonary systolic, diastolic, and mean pressures
- Data transmitted wirelessly to an online portal



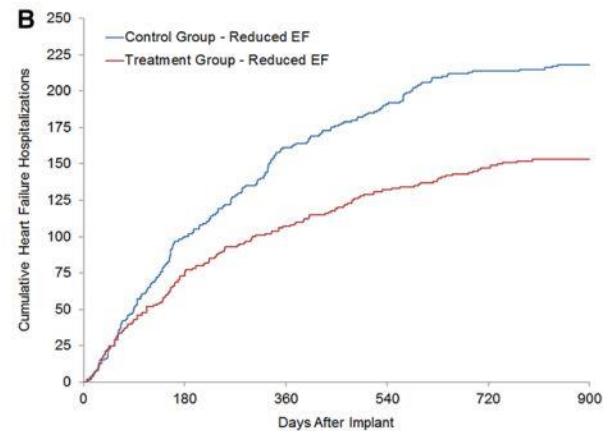
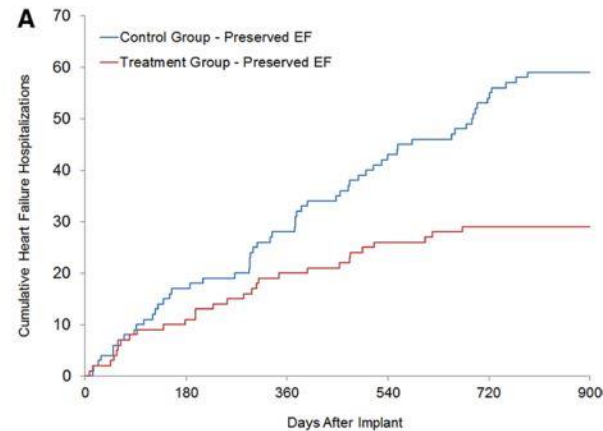
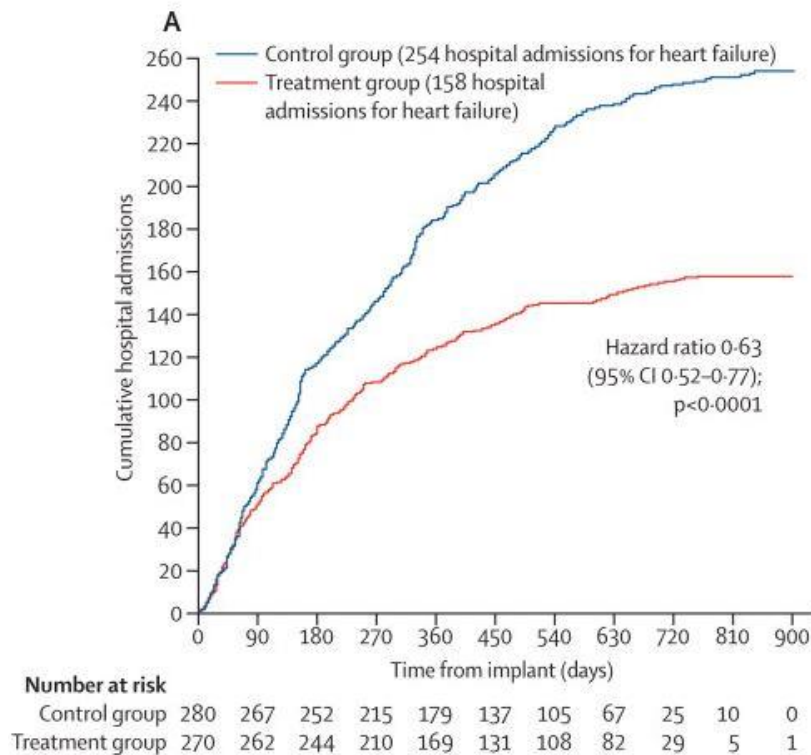
# CHAMPION Trial

- HF patients (HFrEF and HFpEF) with NYHA class III symptoms, and HF hospitalization in past 12 months



# CHAMPTION Trial

- HF-related hospitalization was reduced by 37% in the treatment group



# CardioMEMS

- Indications:
  - NYHA class III HF (both HFrEF and HFpEF)
  - One HF hospitalization in past 12 months
- Patient selection:
  - Difficult to manage volume status
  - Challenging physical assessment
  - Compliance with HF medical care
  - Live far from clinic

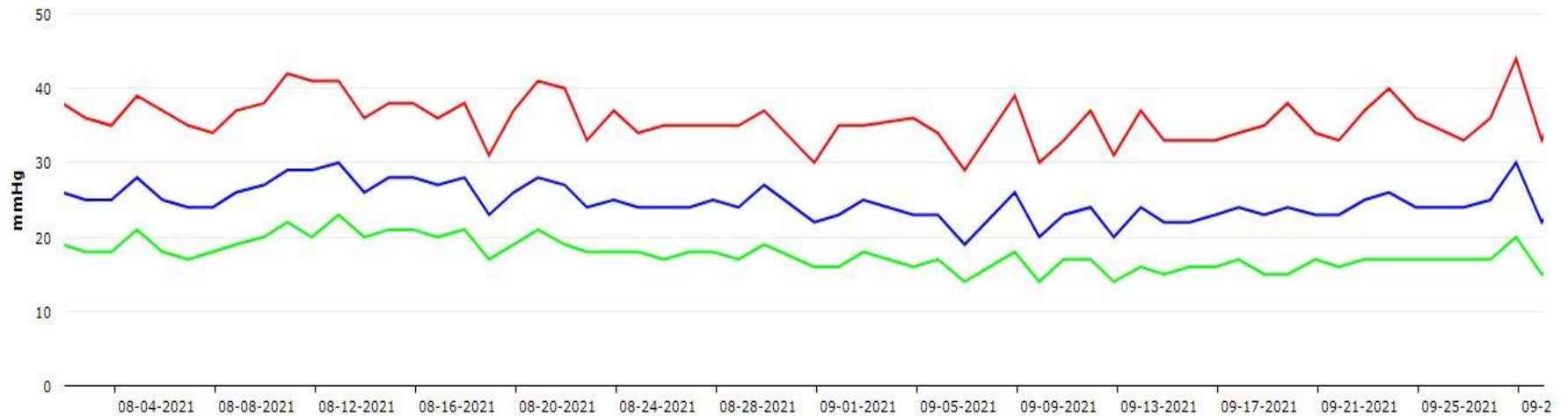
# Case 1

— PA Systolic

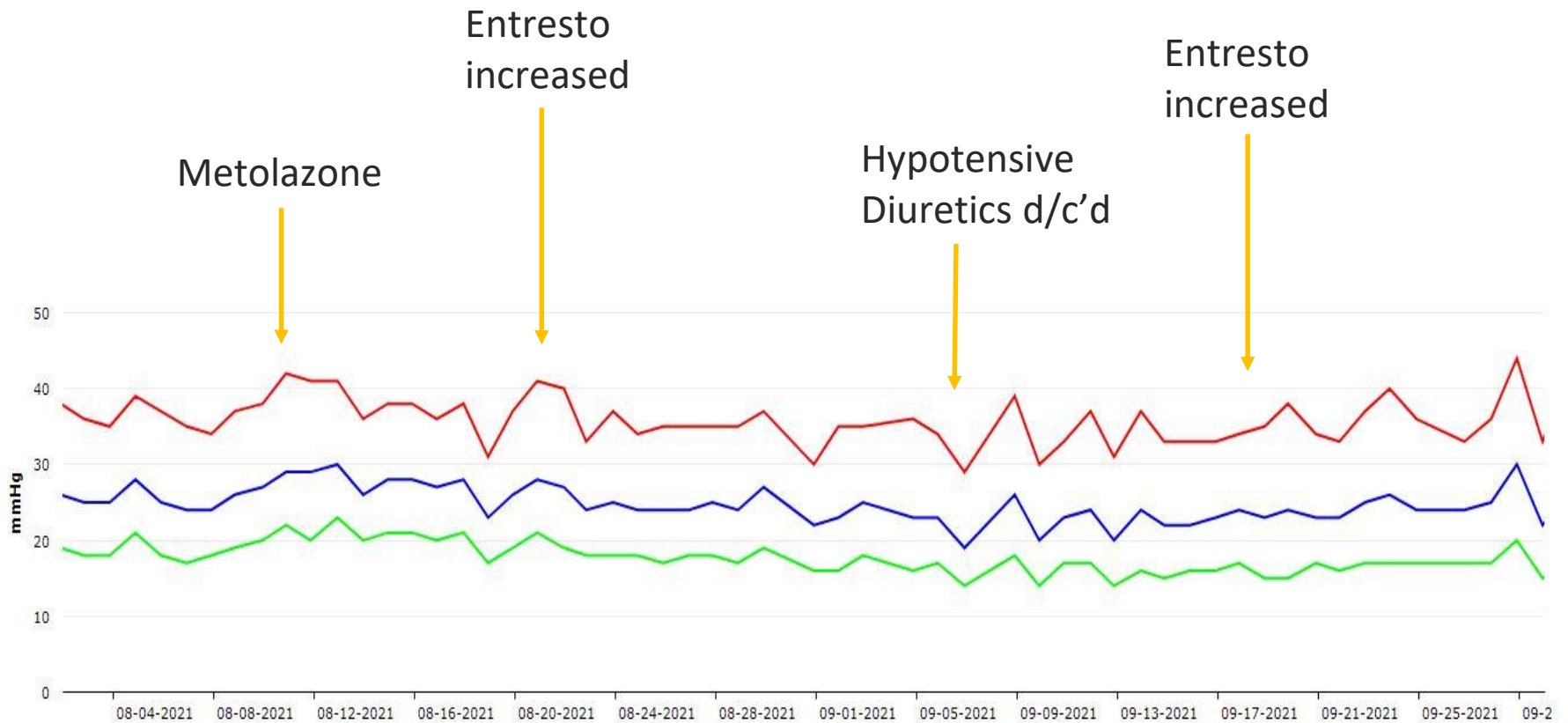
— PA Mean

— PA Diastolic

PA diastolic = Pulmonary capillary wedge pressure (PCWP)



# Case 1



# Case 1

- TTE: LVEF 40-45%
- On maximum doses of GDMT. Remains off diuretics.
- Remains sober

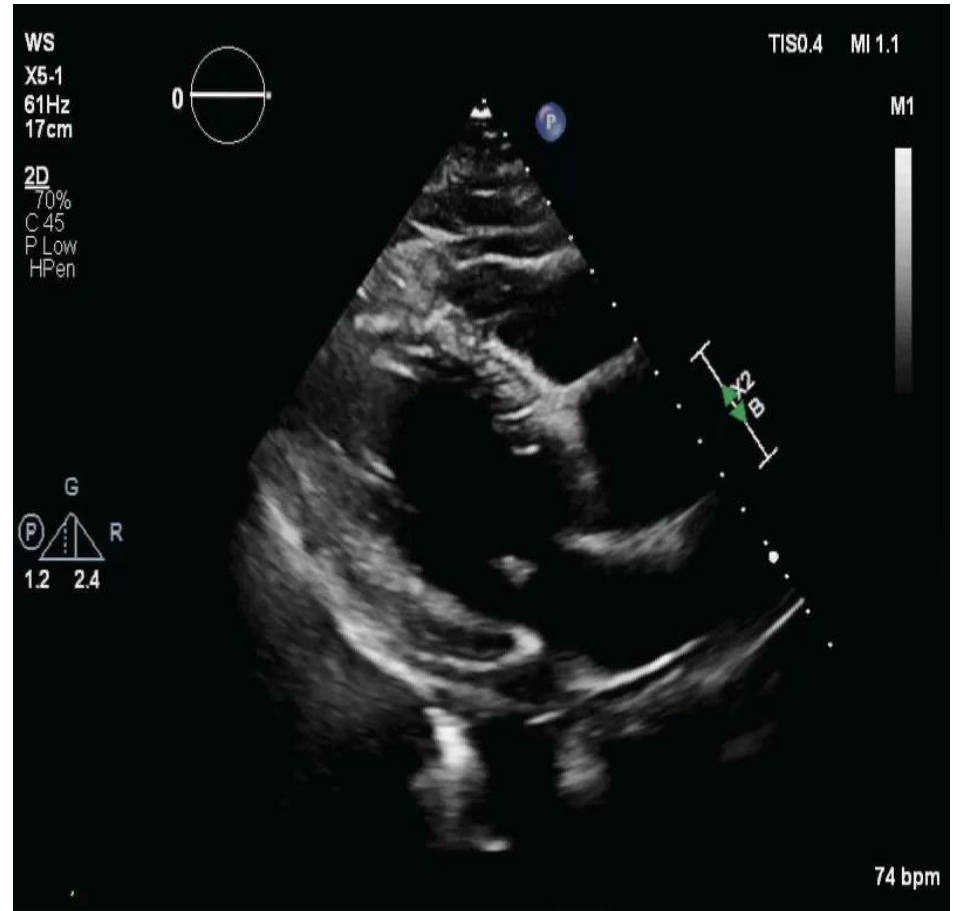


## Case 2

- 88yoM with PMH of HTN, COPD, DM, OSA, carpal tunnel, spinal stenosis s/p laminectomy who presents with LE edema
  - No shortness of breath, but mostly sedentary
  - Multiple US negative for DVT
- ROS: Positive for unintentional weight loss, frequent bowel movements
- FHx: No family history of cardiovascular disease
- PE: BP 133/70, BMI 32. Normal cardiac exam. Clear lungs. 1+ bilateral LE edema.
- Labs: Unremarkable

## Case 2

- Severe concentric LVH
- LVEF 55-60%
- E/e' 10.1
- Grade 1 DD
- No significant valvular dysfunction
- PASP 36mmHg



# Heart Failure with Preserved Ejection Fraction

- H2FPEF Score:

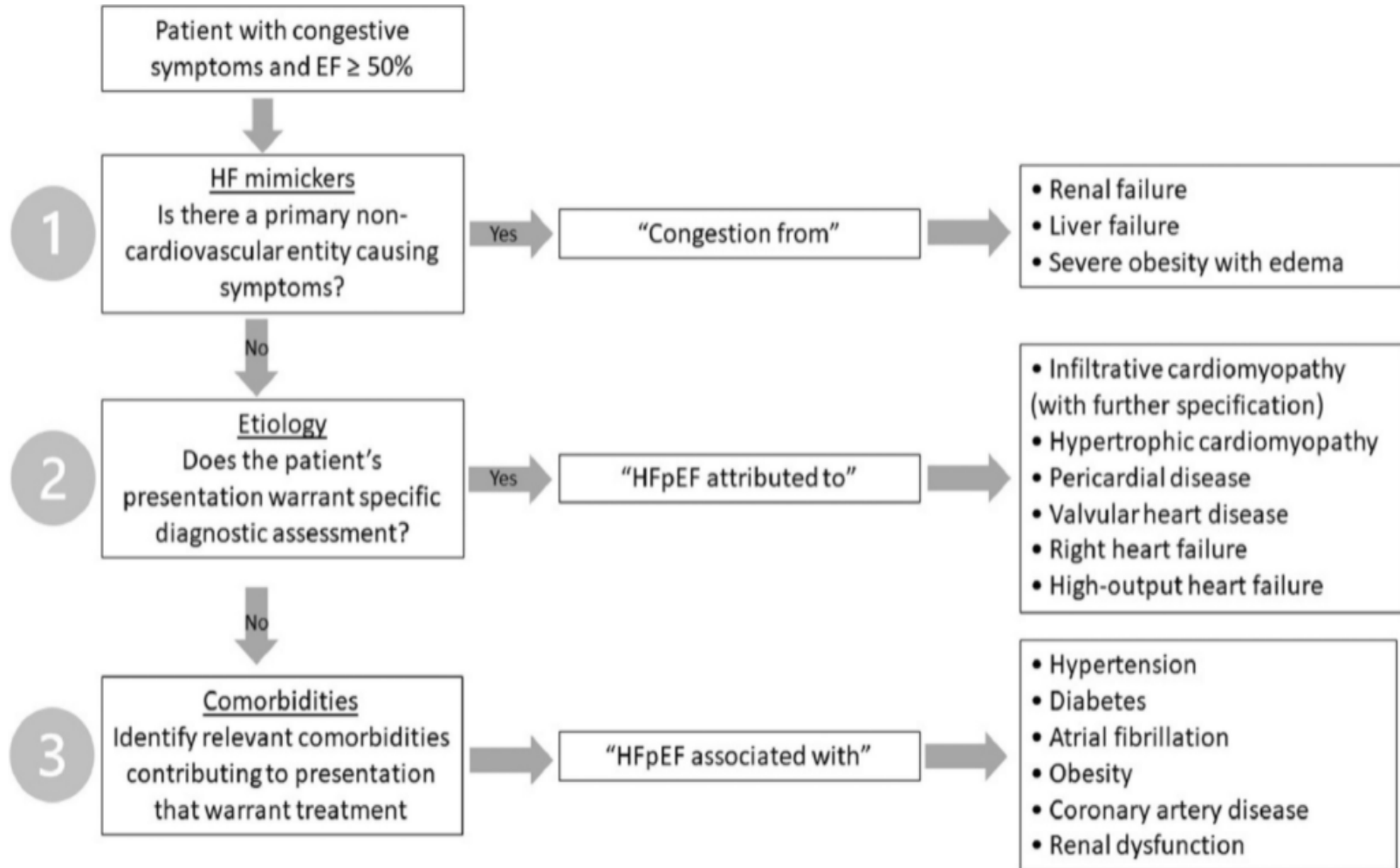
	Clinical Variable	Values	Points
H <sub>2</sub>	Heavy	Body mass index > 30 kg/m <sup>2</sup>	2
	Hypertensive	2 or more antihypertensive medicines	1
F	Atrial Fibrillation	Paroxysmal or Persistent	3
P	Pulmonary Hypertension	Doppler Echocardiographic estimated Pulmonary Artery Systolic Pressure > 35 mmHg	1
E	Elder	Age > 60 years	1
F	Filling Pressure	Doppler Echocardiographic E/e' > 9	1
H <sub>2</sub> FPEF score			Sum (0-9)
<div> <div>Total Points</div> <div> <div>0</div> <div>1</div> <div>2</div> <div>3</div> <div>4</div> <div>5</div> <div>6</div> <div>7</div> <div>8</div> <div>9</div> </div> </div> <div> <div>Probability of HFpEF</div> <div> <div>0.2</div> <div>0.3</div> <div>0.4</div> <div>0.5</div> <div>0.6</div> <div>0.7</div> <div>0.8</div> <div>0.9</div> <div>0.95</div> </div> </div>			

- Score of 0-1: Low risk
- Score of 6-9: High risk
- Score of 2-5: Intermediate risk

# Case 2

	Clinical Variable	Values	Points	
H <sub>2</sub>	Heavy	Body mass index > 30 kg/m <sup>2</sup>	2	←
	Hypertensive	2 or more antihypertensive medicines	1	←
F	Atrial Fibrillation	Paroxysmal or Persistent	3	
P	Pulmonary Hypertension	Doppler Echocardiographic estimated Pulmonary Artery Systolic Pressure > 35 mmHg	1	←
E	Elder	Age > 60 years	1	←
F	Filling Pressure	Doppler Echocardiographic E/e' > 9	1	←
H <sub>2</sub> FPEF score			Sum (0-9)	6
<div> <div>Total Points</div> <div> <div>0</div><div>1</div><div>2</div><div>3</div><div>4</div><div>5</div><div>6</div><div>7</div><div>8</div><div>9</div> </div> </div> <div> <div>Probability of HFpEF</div> <div> <div>0.2</div><div>0.3</div><div>0.4</div><div>0.5</div><div>0.6</div><div>0.7</div><div>0.8</div><div>0.9</div><div>0.95</div> </div> </div>				

# Focus on Etiology



## Case 2

- High probability of HFpEF based on H2FpF score
- Carpal tunnel, spinal stenosis
- GI symptoms
- Severe LVH



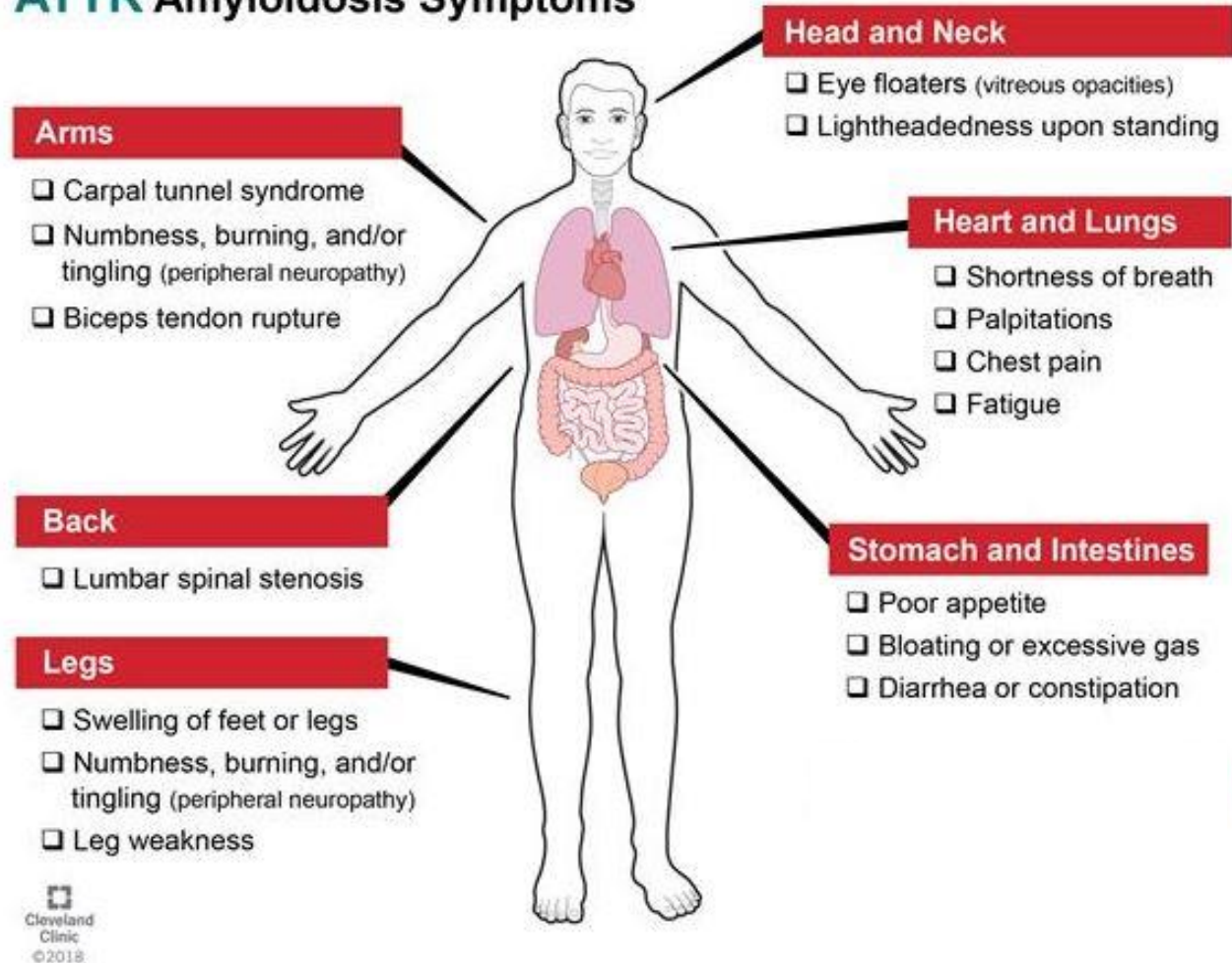
# Amyloid

- Caused by extracellular deposition of proteins in the myocardium
  - Proteins have an unstable structure that causes them to misfold, aggregate, and deposit as amyloid fibrils
- Types:
  - AL: misfolded immunoglobulin light chains from abnormal clonal proliferation of plasma cells
  - Transthyretin (ATTR): liver-synthesized protein
    - » Hereditary: autosomal dominant
    - » Wild-type: previously called senile cardiac amyloid
- Median survival after diagnosis in untreated patients is poor: 2.5 years for hATTR, 3.6 years for ATTRwt



# Amyloid

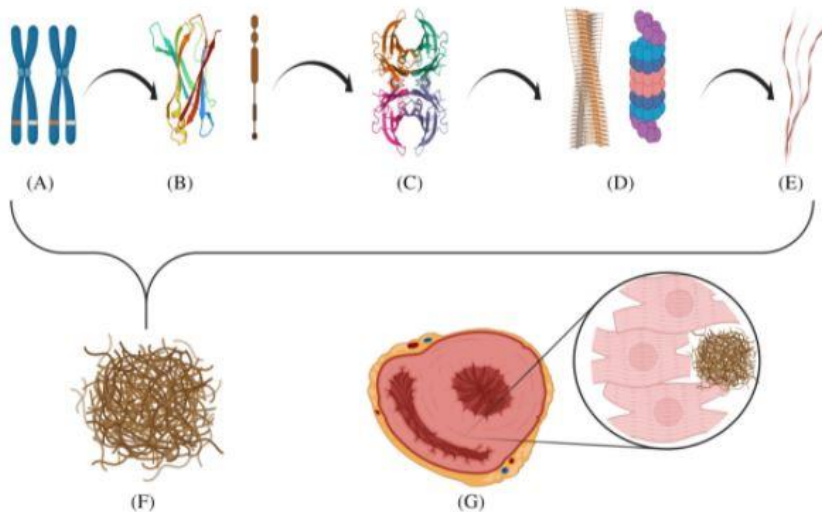
## ATTR Amyloidosis Symptoms





# Cardiac Amyloid

- Amyloid can deposit into any cardiac structure
- Restrictive cardiomyopathy
- Atrial fibrillation
- Aortic stenosis
- Hypotension



# Cardiac Amyloid: Clinical Clues

**Table 1. Clinical Clues From Routine Cardiac Evaluation That Should Prompt Additional Diagnostic Evaluation for ATTR-CM**

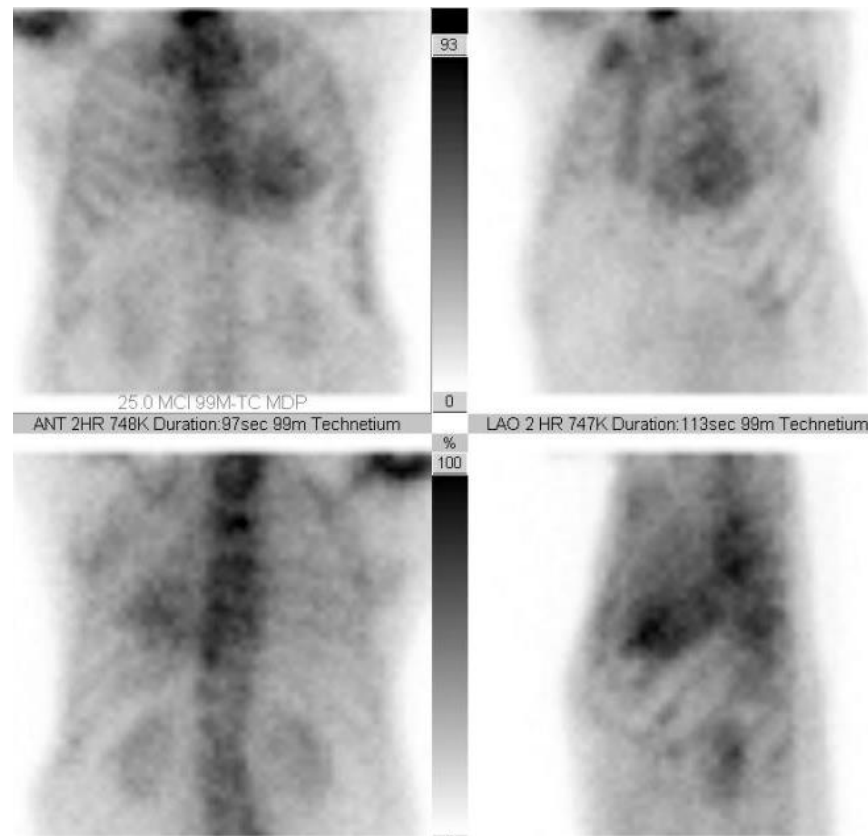
Traditional Cardiac Clues	Noncardiac Clues
Intolerance to antihypertensive or heart failure medications because of symptomatic hypotension or orthostasis	Neurological: sensorimotor polyneuropathy (paresthesias and weakness), autonomic dysfunction (orthostatic hypotension, postprandial diarrhea alternating with constipation, gastroparesis, urinary retention, and incontinence)
Persistent low-level elevation in serum troponin	Orthopedic: carpal tunnel syndrome, lumbar spinal stenosis, unprovoked biceps tendon rupture, hip and knee arthroplasty
Discordance between QRS voltage on an ECG and wall thickness on imaging	Black race
Unexplained atrioventricular block or prior pacemaker implantation	Family history of polyneuropathy
Unexplained LV wall thickening, right ventricular thickening, or atrial wall thickening	
Family history of cardiomyopathy	

# Cardiac Amyloid: Diagnosis

- Technitium pyrophosphate scintigraphy (PYP) scan
  - Nuclear imaging using a radiotracer that binds to TTR amyloid fibrils
  - Able to detect TTR fibrils prior to onset of cardiac hypertrophy and electrophysiologic changes
  - Sensitivity 91%

## Case 2

- PYP scan: Positive study with a visual grade of 3 and a heart to contralateral lung ratio of 1.54



# Cardiac Amyloid: Treatment

## Suppression of TTR

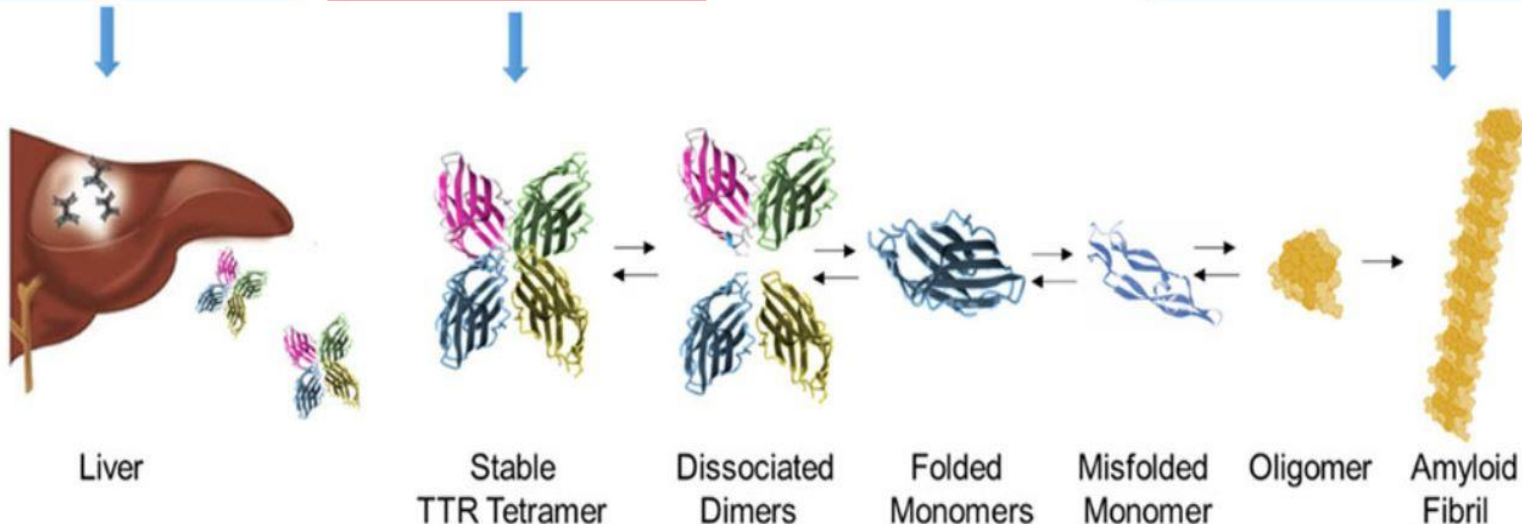
- Liver Transplantation
- TTR Gene silencers (Patisiran/Inotersen)

## TTR Stabilization

- Tafamidis
- Diflunisal
- Green Tea
- AG10

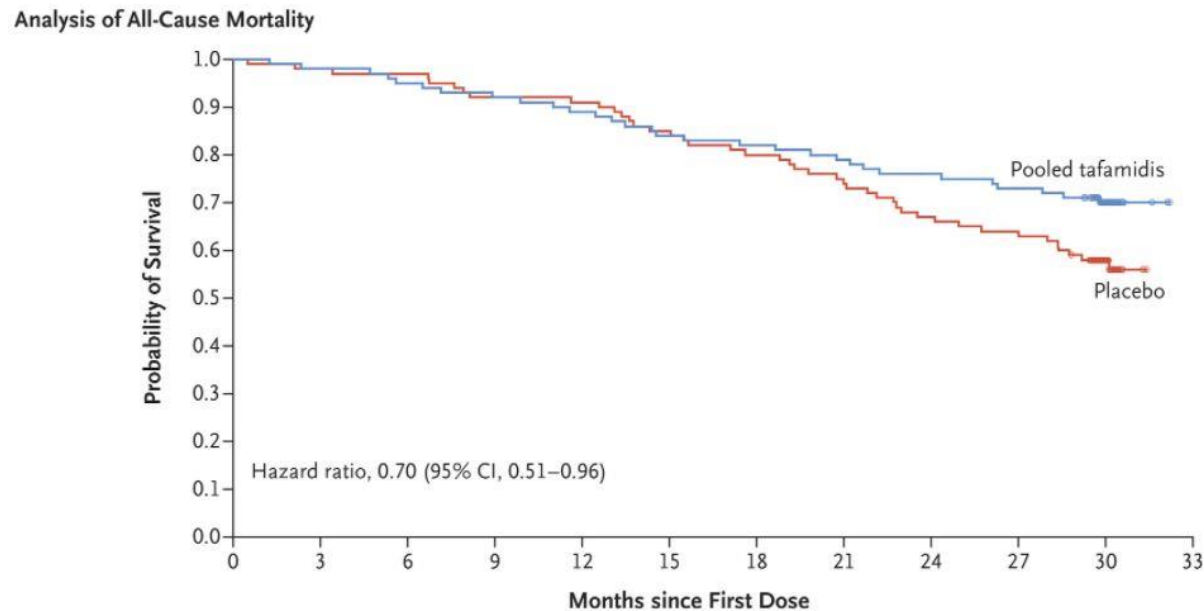
## TTR disruption/resorption

- Doxycycline/TUDCA
- Monoclonal antibodies



# Tafamadis: TTR stabilizer

- ATTR-ACT Trial
- 13.4% absolute reduction in overall mortality and 22% absolute reduction in hospitalization
- Improved QOL and functional status



## Case 3

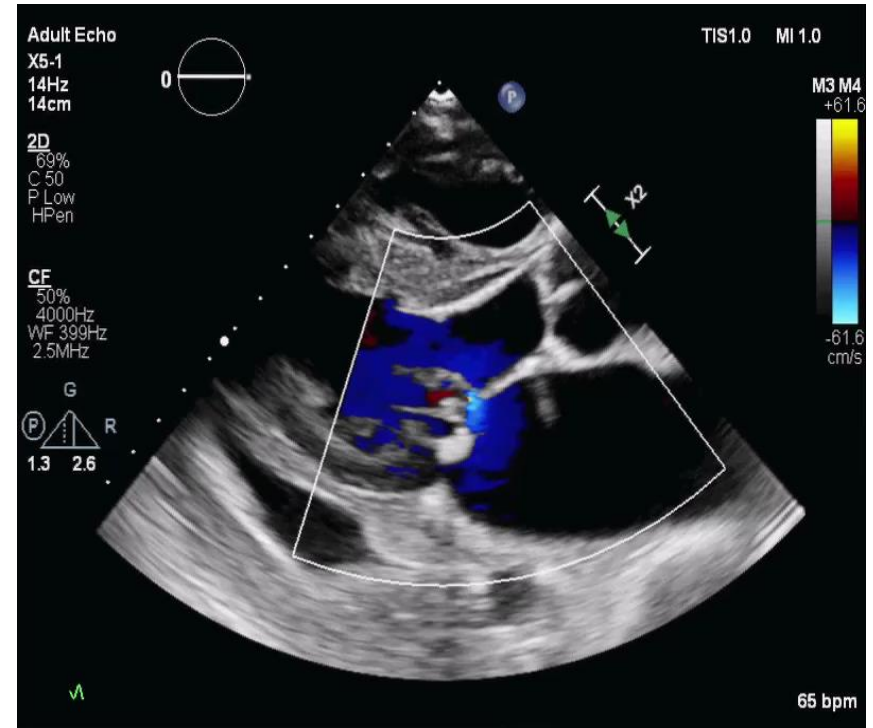
- 86yoM with HTN, HLD, hypothyroidism, and prostate cancer who presents to establish care after HF hospitalization
- Hospitalized 4 months prior with decompensated HF. Diagnosed with HFpEF
- Discharged on bumex 1mg BID, but self-discontinued due to dizziness
- Continues to have significant shortness of breath and edema

## Case 3

- Medications: Amlodipine, spironolactone, simvastatin, levothyroxine
- FHx: 1 daughter with HF, but does not know the specifics. No FHx of sudden cardiac death.
- PE: 147/71. BMI 28. 3/6 harsh systolic murmur at apex, increased with standing. 2+ lower extremity edema.



# Case 3

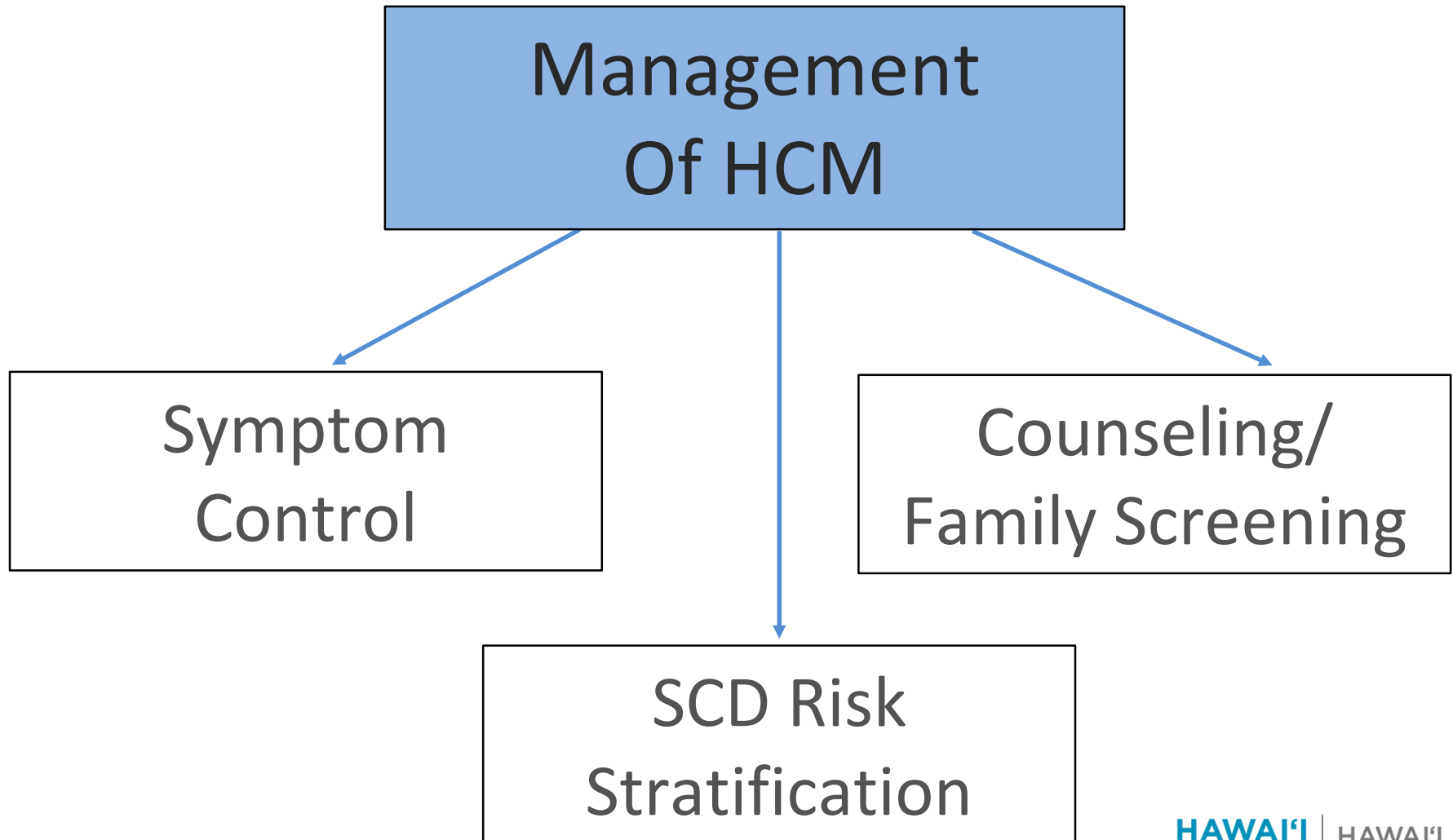


- LVEF 60-65%, severe septal hypertrophy, LVOT 86mmHg at rest, increased to 213mmHg with Valsalva. + SAM with severe MR.

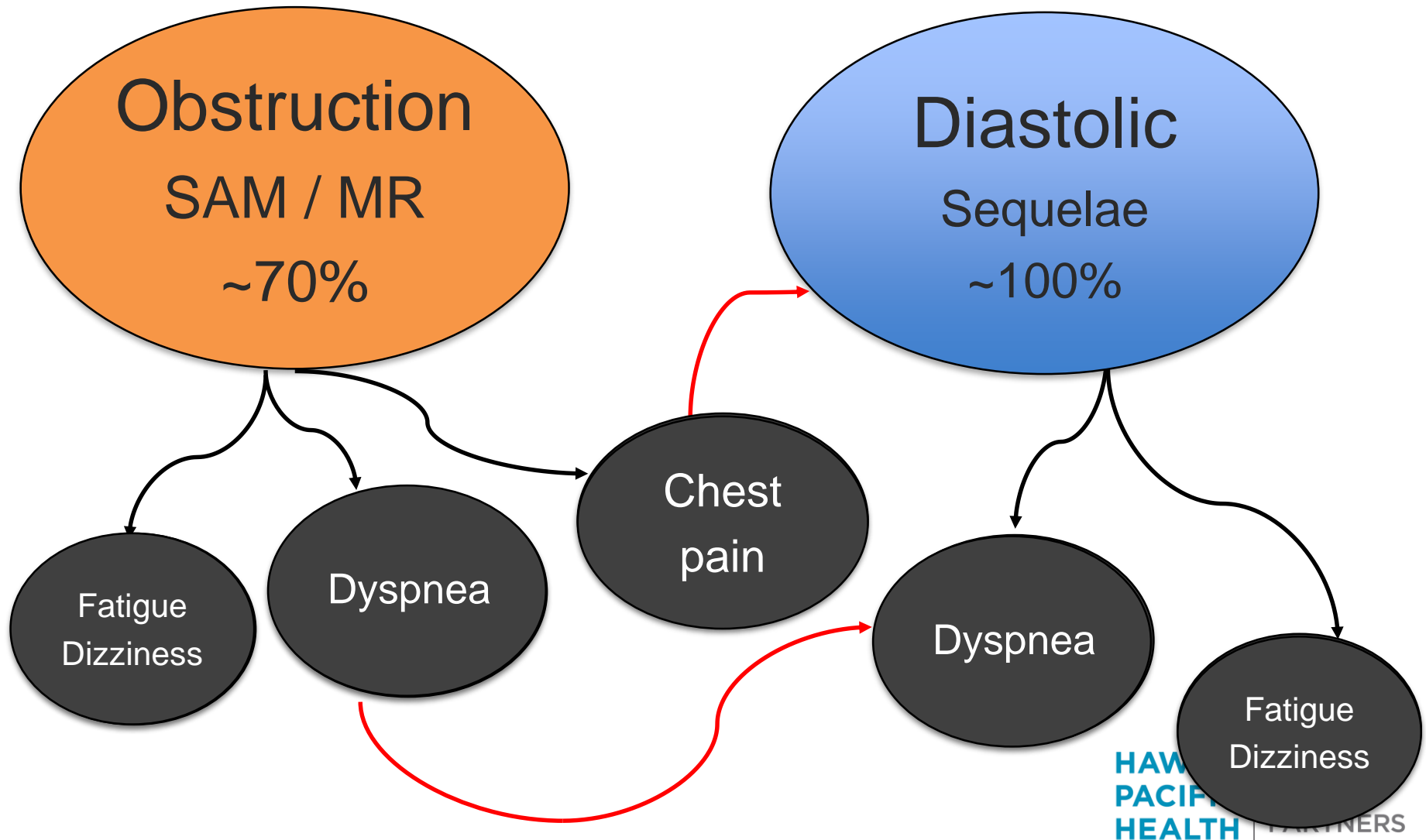
# Hypertrophic cardiomyopathy

- Most common genetic cardiomyopathy
  - Prevalence of 1:500
- Mutations are transmitted in an autosomal dominant manner and exhibit variable and age-related penetrance
- Natural history ranges from a benign course with minimal symptoms to progressive disease and the development of HF
- Increased risk of sudden cardiac death

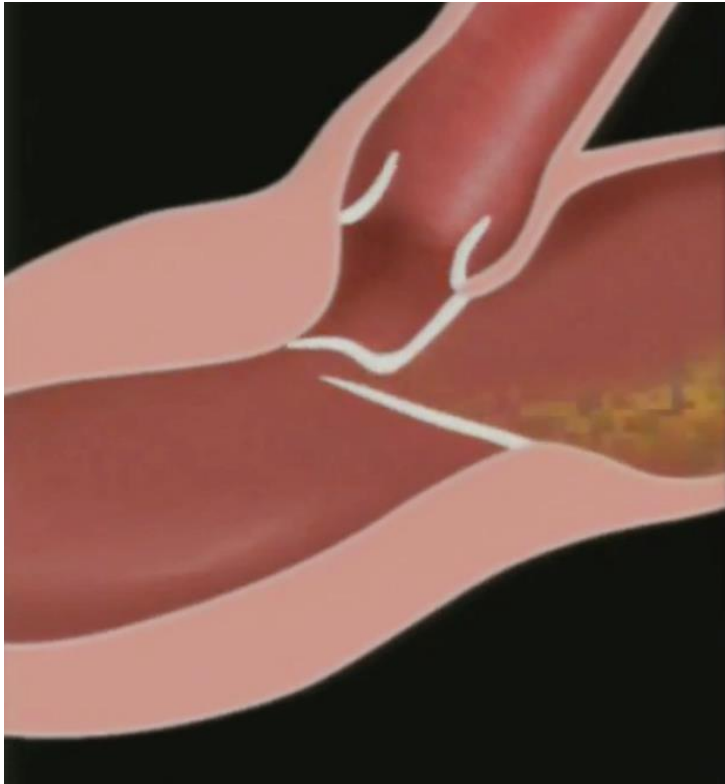
# Hypertrophic cardiomyopathy



# Symptoms of Hypertrophic Cardiomyopathy

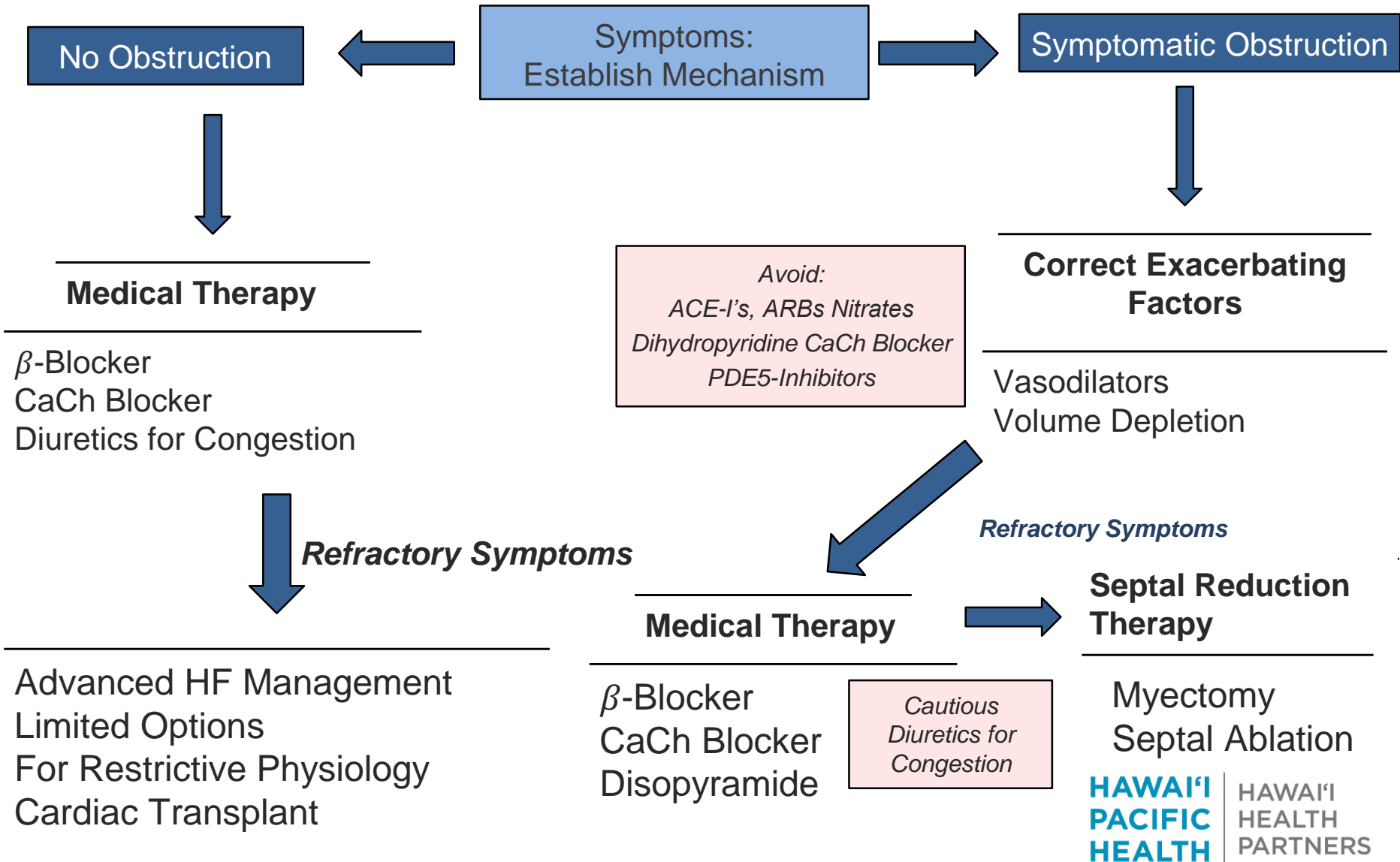


# Symptom Management: LVOT Obstruction

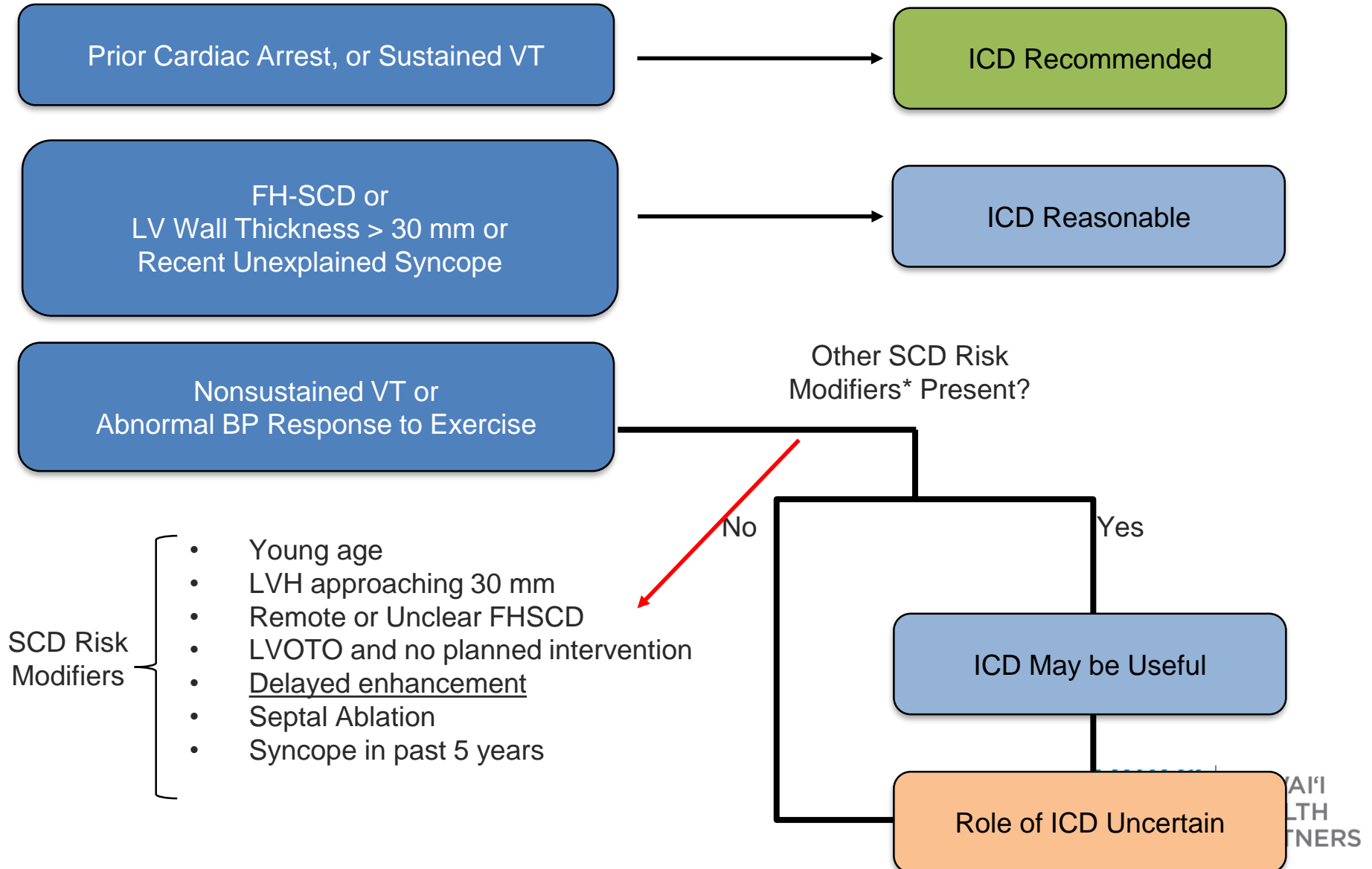


- Obstruction worsens with:
  - More contractility  
Give negative inotropes
  - Decreased afterload  
Withdraw afterload reducing agents
  - Decreased preload  
Avoid volume depletion
- All of these occur with exercise

# Approach to Therapy



# Indications for ICD



# Family Screening

Screen in adolescence or at any consideration of competitive athletics every 12-18 months

Adults (18-20yrs) every 5 years

Genetic testing possible

- Genetic testing starts with the proband
- If the mutation is identified, confirmatory testing is done in family members
- 35% likelihood that a pathogenic mutation will be identified (higher with positive family history)
- Does not influence treatment strategies and does not identify high-risk patients who may benefit from ICD



## Case 3

- Amlodipine discontinued
- Started on metoprolol XL and diltiazem
- Remained on spironolactone
- Shortness of breath and edema improved
- Repeat echocardiogram: No significant LVOT gradient, no SAM
- Genetic testing pending
- No high risk features to warrant an ICD

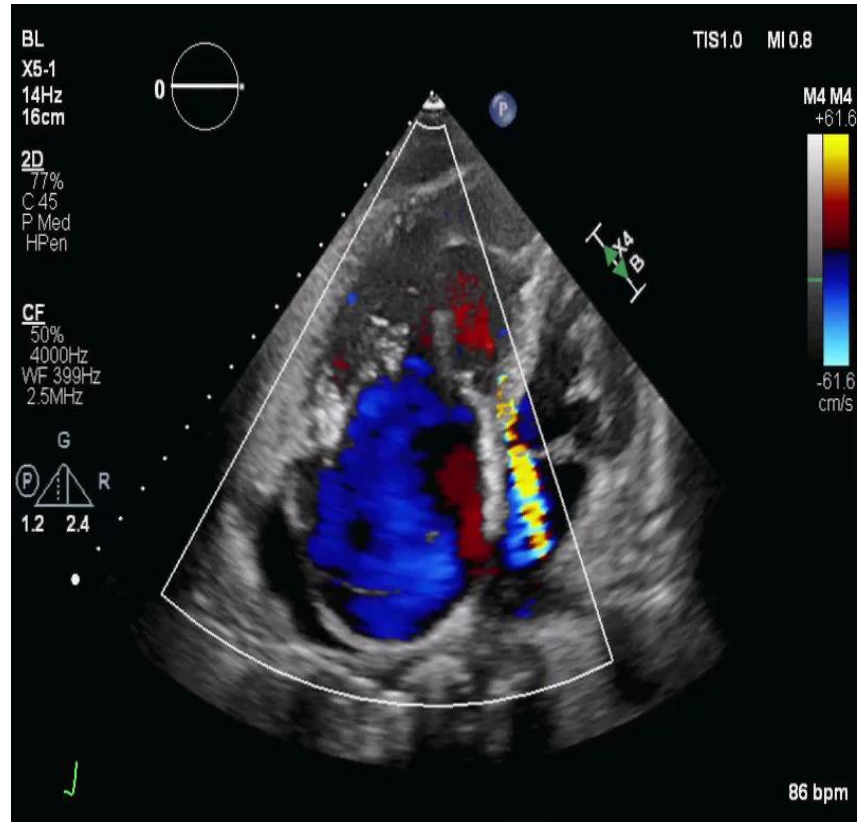
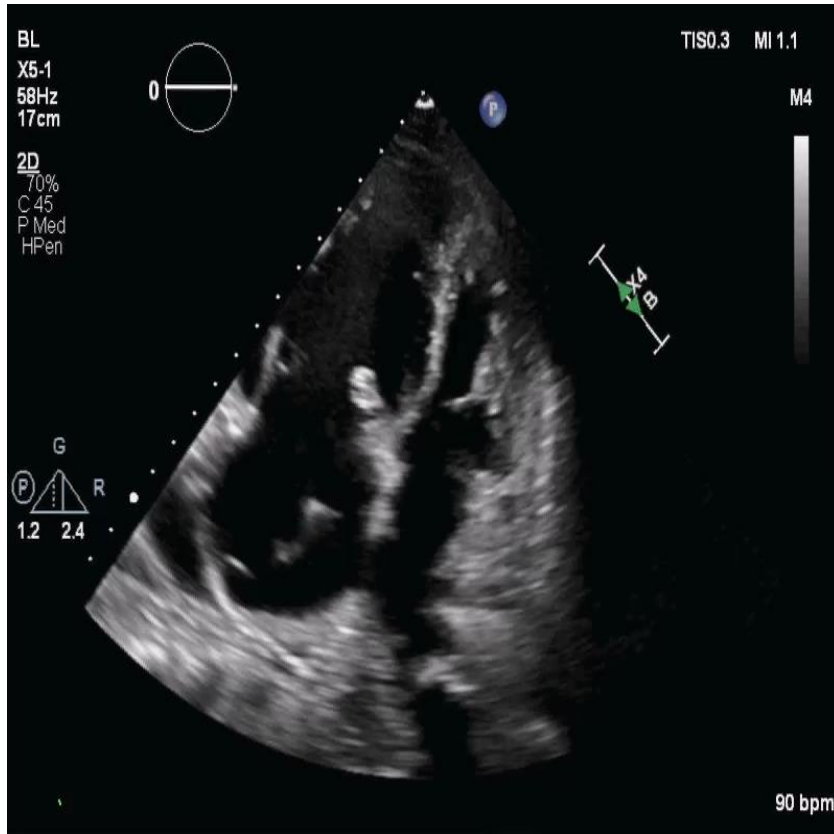
## Case 4

- 65yoF with history of HTN and joint pain who presents to the ED with shortness of breath and LE edema
  - Diagnosed with COVID 1 month ago. Mild symptoms, no hospitalization.
  - Saw rheumatology 3 months prior for elevated ANA (>2560). Labs pending.
- Medications: Amlodipine, HCTZ
- FHx: No significant cardiovascular disease
- SHx: Non-smoker. No EtOH use.

## Case 4

- Exam: BP 110/70, HR 120s, SpO2 88%. Systolic murmur at LLSB. LE warm with 2+ pitting edema.
- Notable labs: Cr 0.9, NTproBNP 2447, troponin 67
- ECG: Sinus tachycardia, RAD, RV hypertrophy
- CTA chest: Negative for PE.
  - Findings suggestive of right heart enlargement and contrast reflux into the hepatic veins
  - Small bilateral pleural effusions

# Case 4



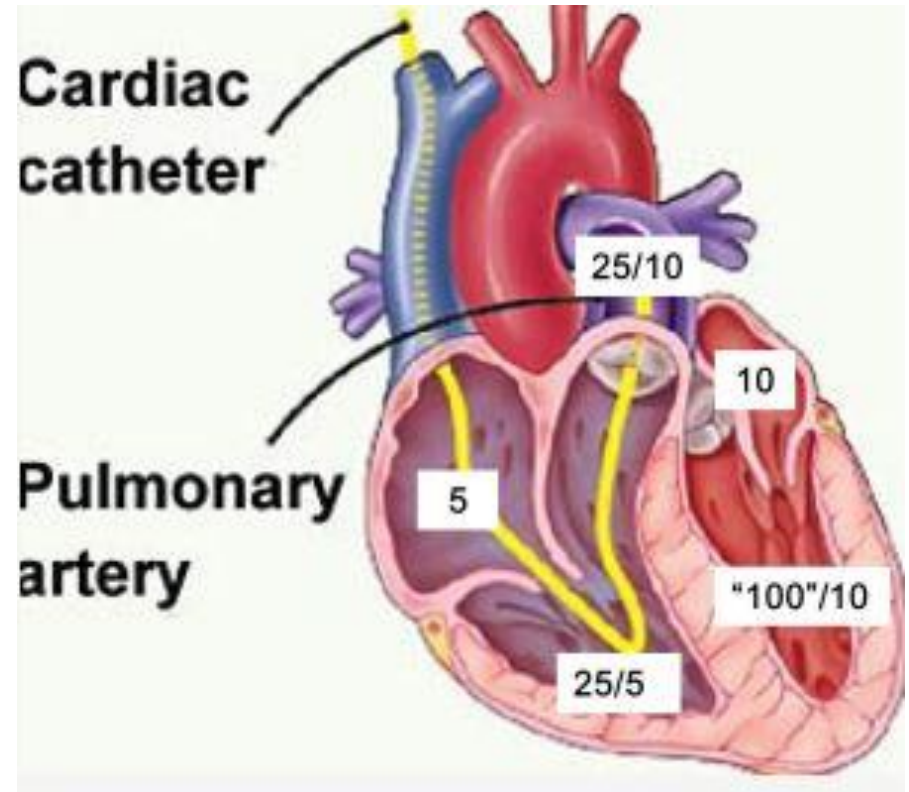
- Severe right-sided chamber enlargement. Severe TR. Severe pulmonary HTN, PASP 64mmHg. Small pericardial effusion.

## Case 4

- Diuresed with Lasix IV with significant urine output
- Cr increased, up to 1.7
- More cool on exam
- Started on milrinone 0.25mcg/kg/min with improvement in perfusion markers
- TEE: mild tricuspid annular dilation, structurally normal tricuspid valve leaflets

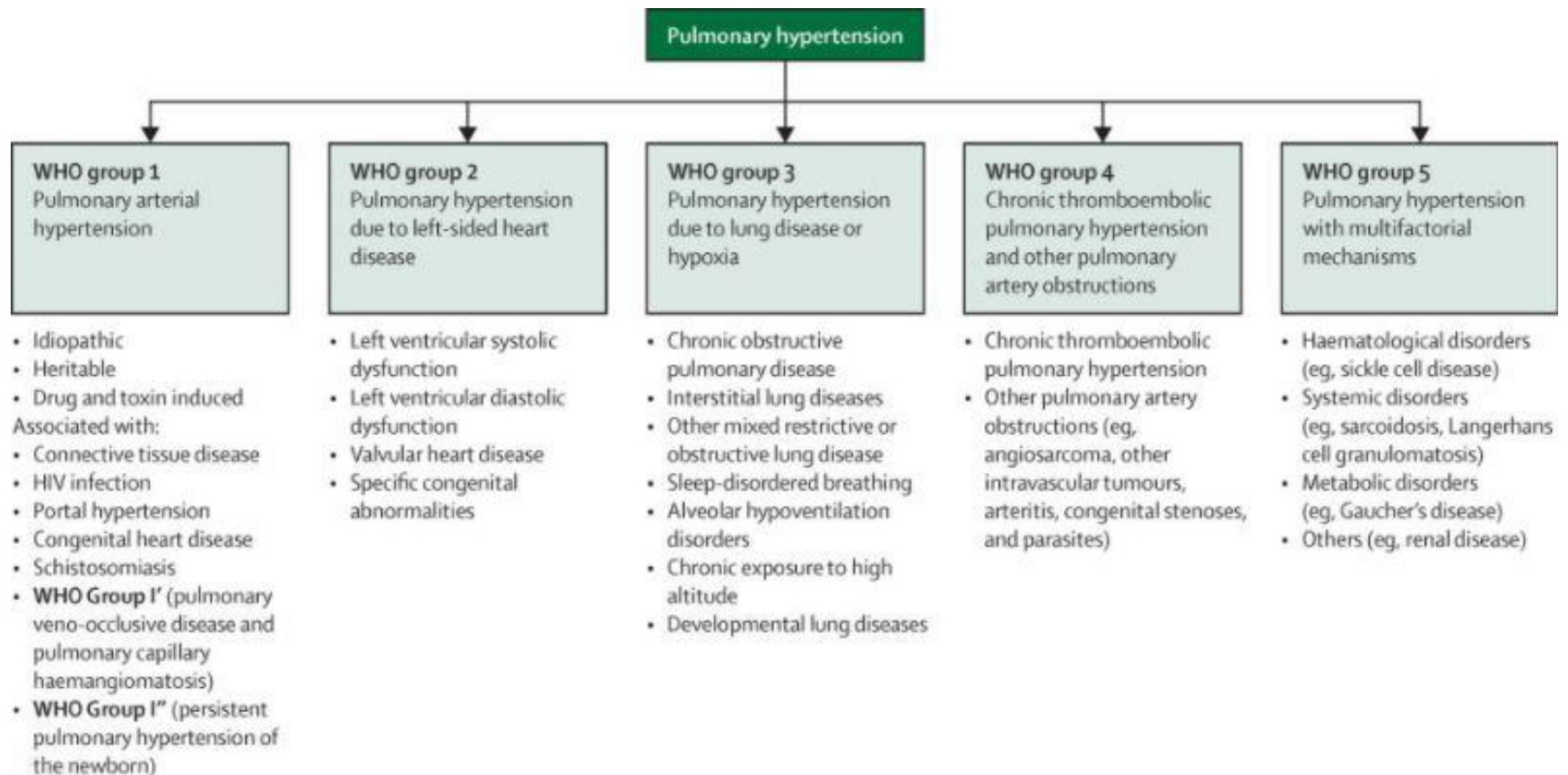
# Case 4

- LHC: Negative for obstructive CAD
- RHC:
  - RA 34
  - RV 83/23
  - PA 91/43/59
  - PCWP 14
  - CO/CI 2.4/1.7
  - PVR 12.8
  - No evidence of intracardiac shunt
  - No changes with iNO

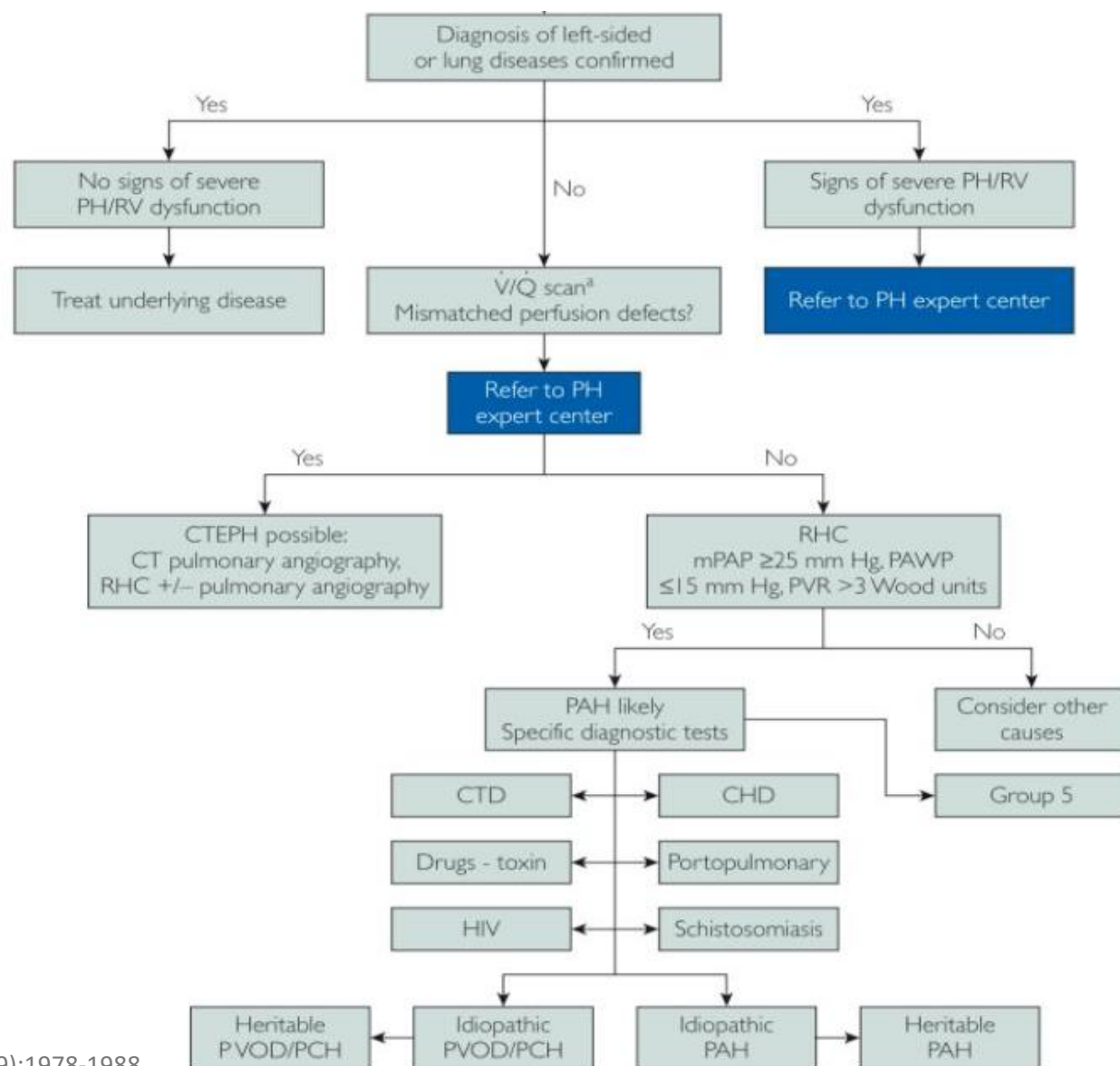


# Pulmonary Hypertension: Definition

- Mean PAP >20mmHg with PVR >3 WU



# Pulmonary Hypertension: Evaluation





# Right Heart Failure

- Increased RV afterload eventually leads to right-sided heart failure
  - Characterized by venous congestion, underfilling of the LV, and cardiogenic shock
- RHF is the leading cause of death in pulmonary hypertension
  - 5-year survival of 57%

# Right Heart Failure: Chronic Management

- Decongestion with diuretics or renal replacement therapy
  - Increases RV stroke volume and cardiac output
  - Improves LV filling
- Afterload reduction with pulmonary vasodilators
  - PDE-5 inhibitors
  - Endothelin-receptor antagonists
  - Soluble guanylate cyclase inhibitors
  - Prostacycline inhibitors
  - Benefit primarily seen in WHO group 1 and 4
- Maintain systemic blood pressure
  - RV highly susceptible to subendocardial ischemia

# Case 4

- Diuresed aggressively
- VQ Scan negative for CTEPH
- Rheumatology labs: Positive Sm/RNP, SSA
- Diagnosed with mixed connective tissue disease
- Started on Plaquenil
- Established care with pulmonary hypertension specialist
  - Started on dual therapy
  - Remains on Lasix 20mg PO

# Take Home Points

- GDMT with quadruple therapy significantly reduces morbidity and mortality
- Consider CardioMEMS in patients with NYHA class III symptoms and frequent hospitalizations for HF
- Look for clinical clues that may suggest other causes of HFpEF
- Treatment of hypertrophic cardiomyopathy includes symptom management, SCD risk stratification, and familial screening
- Pulmonary arterial hypertension is a progressive condition with high mortality if left untreated. Survival improves with early diagnosis and timely therapy.

# Q&A

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# Thank you!

- A recording of the meeting will be available afterwards
- Unanswered question?
  - Contact us at [info@hawaiihealthpartners.org](mailto:info@hawaiihealthpartners.org)

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