Cardiomyopathies

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Learning Objectives

- Define Heart Failure With Reduced Ejection Fraction (HFrEF)
- Review ambulatory treatment strategies for HFrEF
- Identify specific causes of HFrEF:
  - Ischemic Cardiomyopathy
  - Dilated Cardiomyopathy
- Identify cardiomyopathies with preserved EF:
  - Hypertrophic Cardiomyopathy
  - Infiltrative Cardiomyopathy

Diagnostics: History

- Clinical Syndrome
- Impaired filling of blood into the ventricle
- Impaired ejection of blood from the ventricle
- Diverse, complex

BOX 31.2 Framingham Heart Failure Criteria

Framingham Heart Failure Criteria[11]

Diagnosis of HF requires 2 major criteria OR 1 major and 2 minor criteria.

Major Criteria

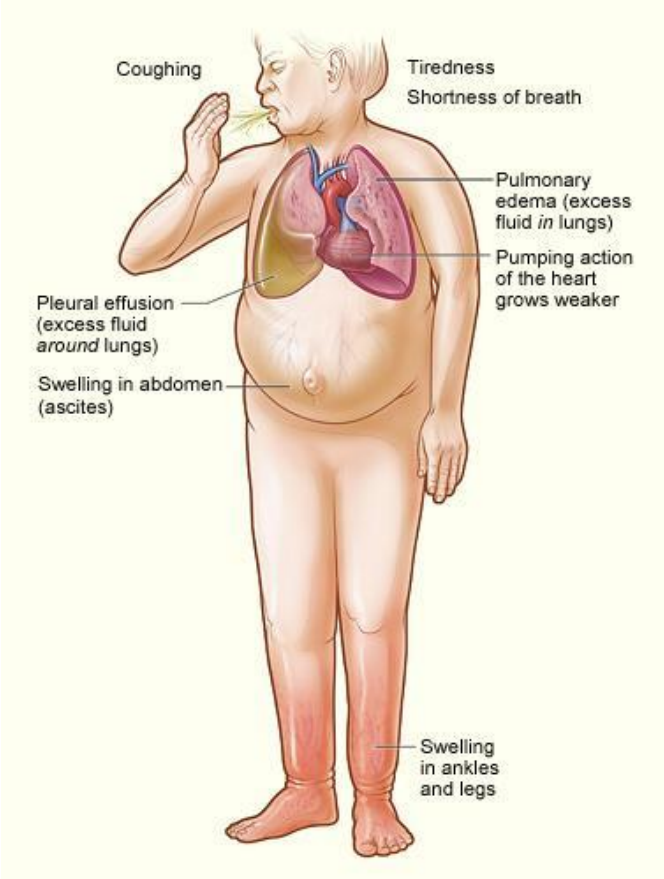
Acute pulmonary edema
Cardiomegaly
Hepatojugular reflux
Neck vein distention
Paroxysmal nocturnal dyspnea or orthopnea
Pulmonary rales
Third heart sound (S_3 gallop rhythm)
Weight loss >4.5 kg in 5 days in response to treatment

Minor Criteria

Ankle edema
Dyspnea on exertion
Hepatomegaly
Nocturnal cough
Pleural effusion
Tachycardia (HR >120)

Diagnostics: The Physical Exam

- Peripheral Edema:
  - Dependent
- Pulmonary Edema
  - Rales/Crackles
- Ascites
- Orthopnea
- Jugular Venous Distention

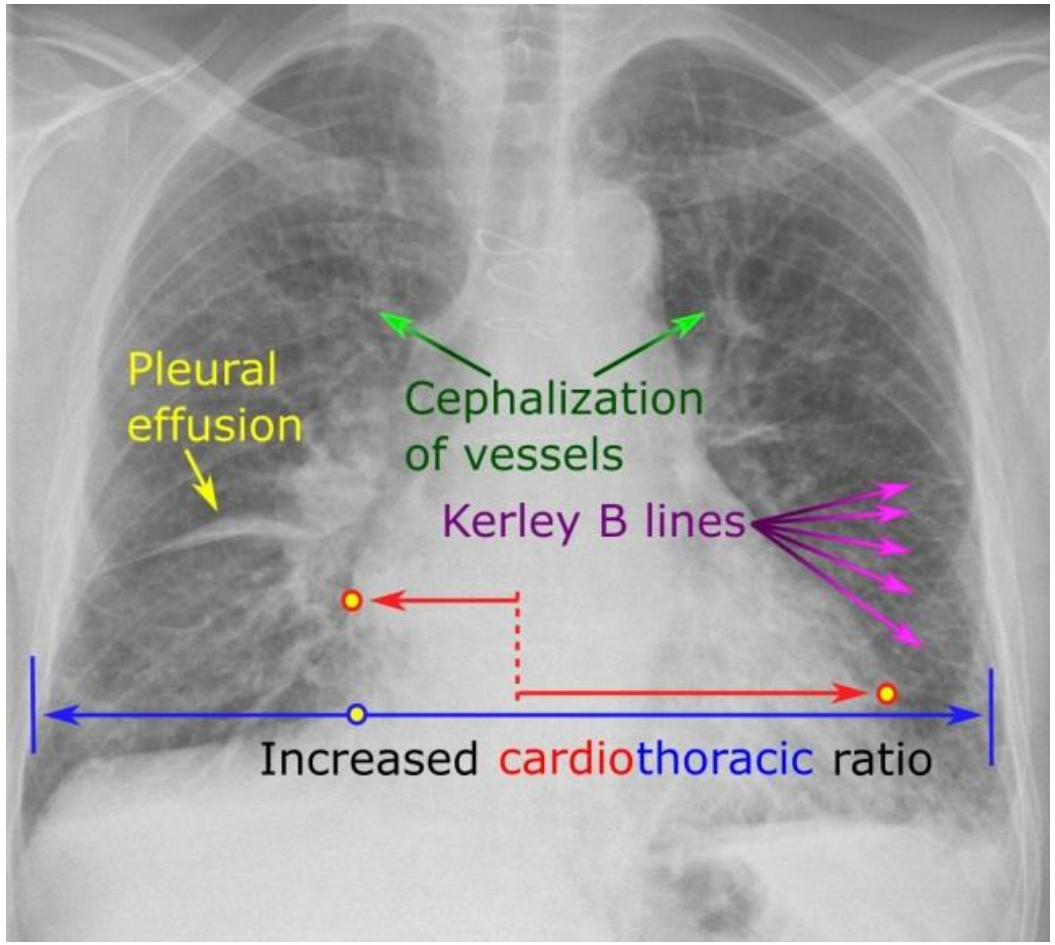


Diagnostics: The Physical Exam

	Sensitivity	Specificity
Physical examination		
Third heart sound (ventricular filling gallop)[36,41,43-45,48,53,56]	0.13	0.99
Abdominojugular reflux[31]	0.24	0.96
Jugular venous distension[36,41,43-45,48,53,56]	0.39	0.92
Rales[36,41,43-45,48,53,56]	0.60	0.78
Any murmur[36,44,48,53]	0.27	0.90
Lower extremity edema[41,43-45,53,56]	0.50	0.78
Valsalva maneuver[31]	0.73	0.65
Systolic blood pressure <100 mm Hg[48]	0.06	0.97
Fourth heart sound (atrial gallop)[36,48,53]	0.05	0.97
Systolic blood pressure ≥150 mm Hg[48]	0.28	0.73
Wheezing[36,44,45,48,53]	0.22	0.58
Ascites[48]	0.01	0.97

Abbreviations: CI, confidence interval; LR, likelihood ratio.
*LRs are not independent of each other and should not be multiplied in series when multiple findings are considered.

Diagnostics: Chest X-ray



https://glassboxmedicine.com/2019/03/23/radiology-abnormal-chest-x-rays-part-ii/

Next Steps

- Bedside history and exam are critical to identifying the presence of heart failure
- Limitation:
  - *Why* is the patient in heart failure?

Stages of Heart Failure

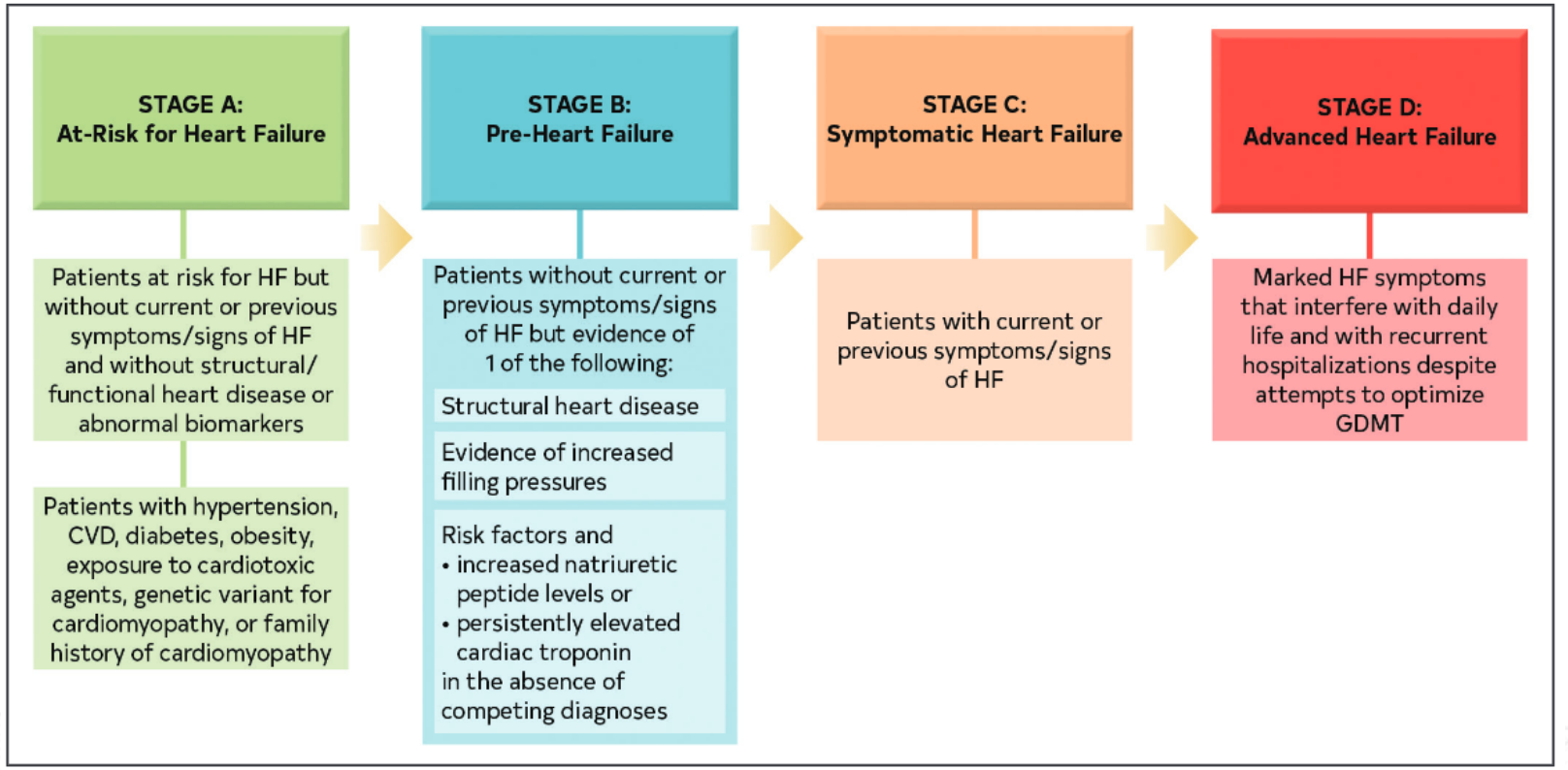


Figure 1. ACC/AHA Stages of HF.
The ACC/AHA stages of HF are shown. ACC indicates American College of Cardiology; AHA, American Heart Association; CVD, cardiovascular disease; GDMT, guideline-directed medical therapy; and HF, heart failure.

Stages of Heart Failure

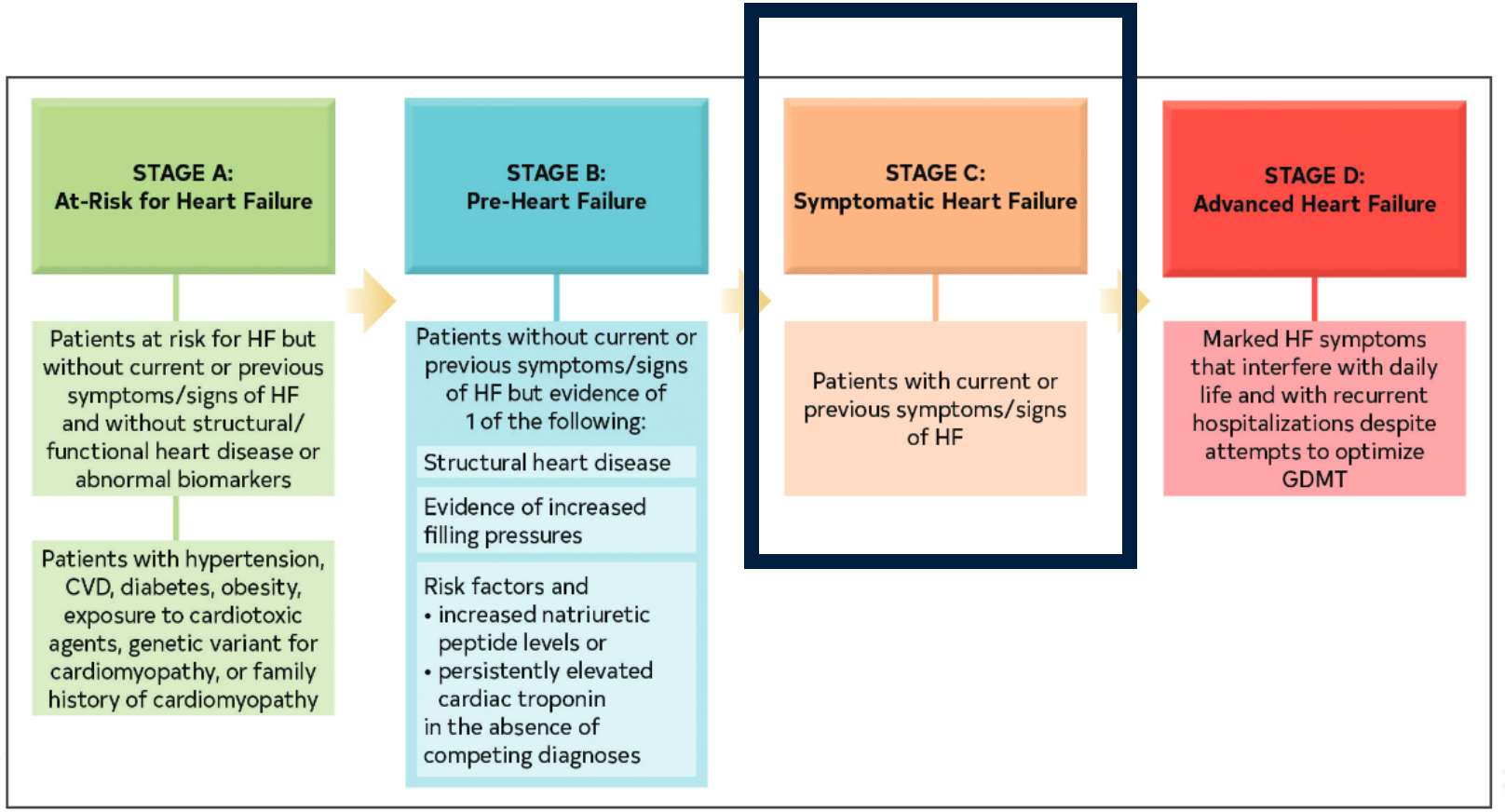


Figure 1. ACC/AHA Stages of HF.
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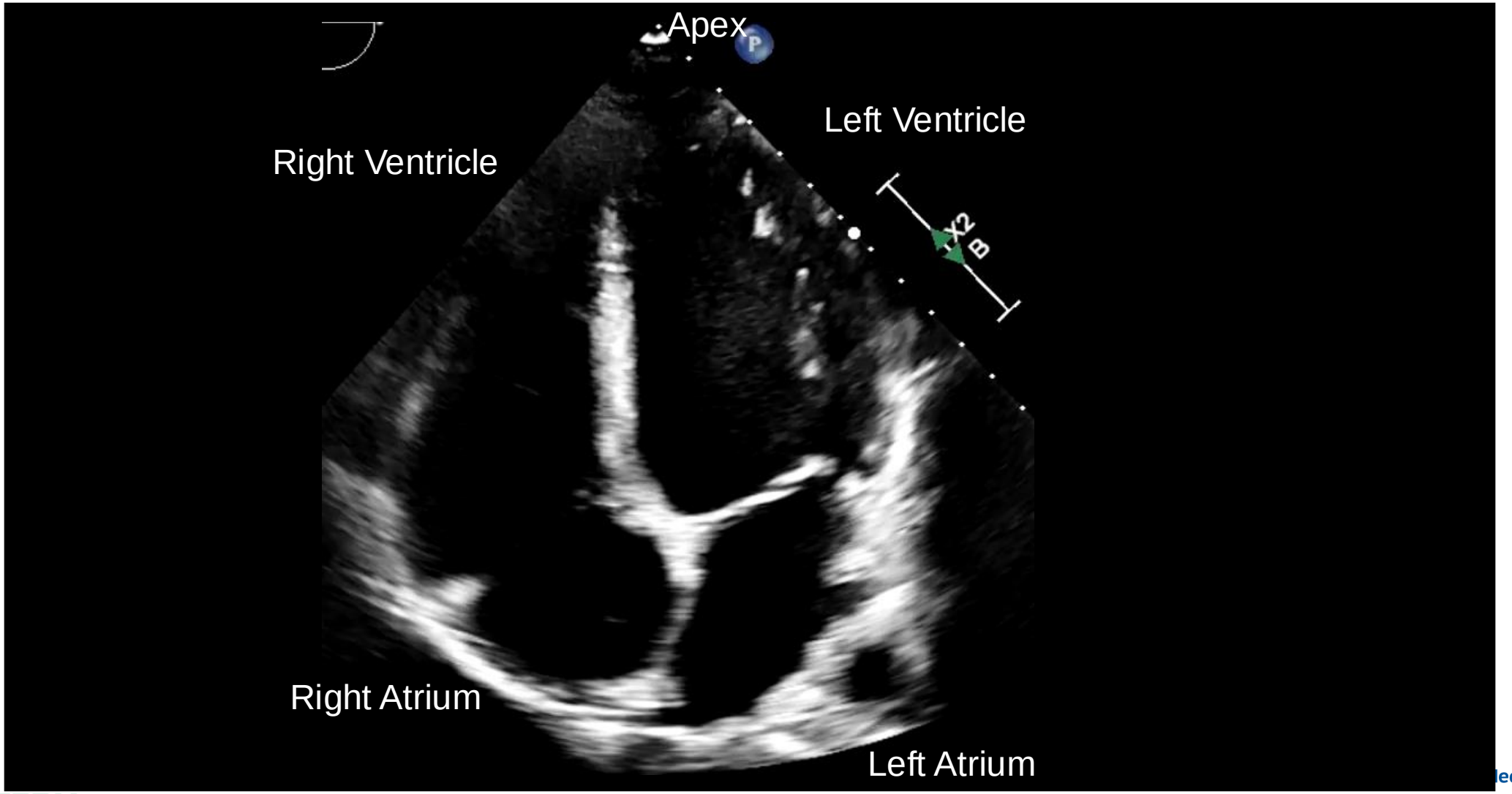
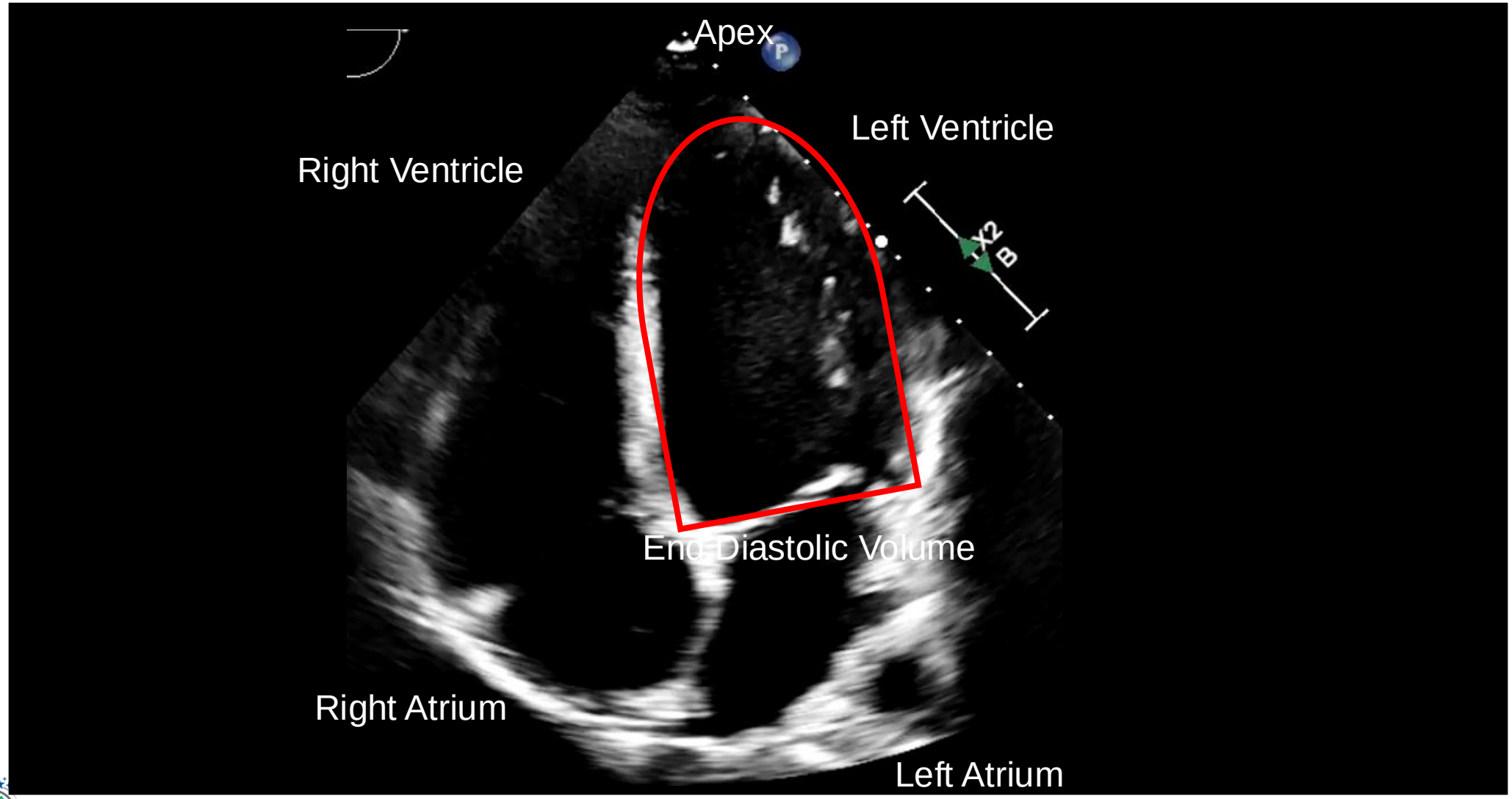
Identifying Specific Cardiomyopathies

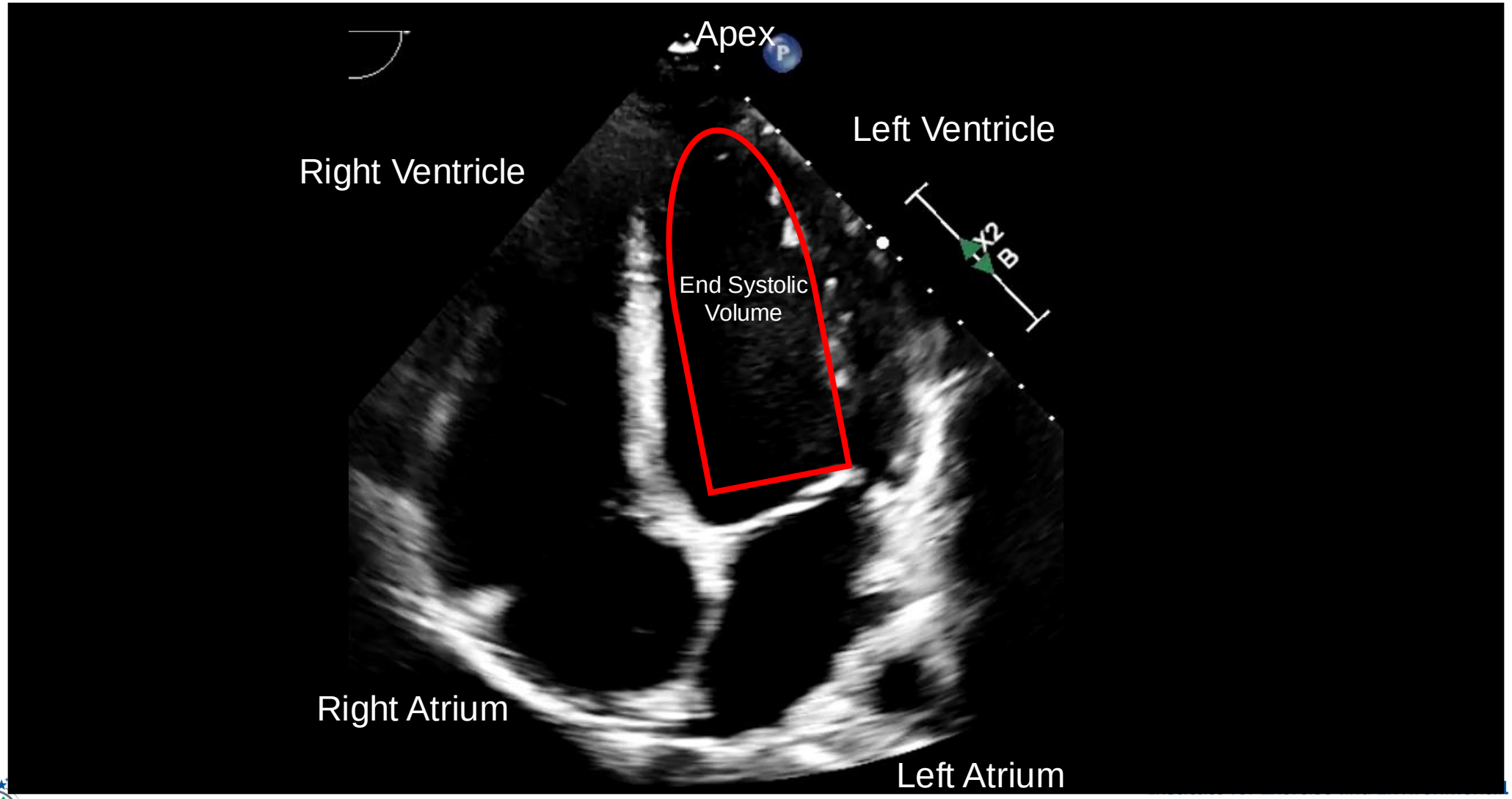
Cardiac anatomy and echocardiography



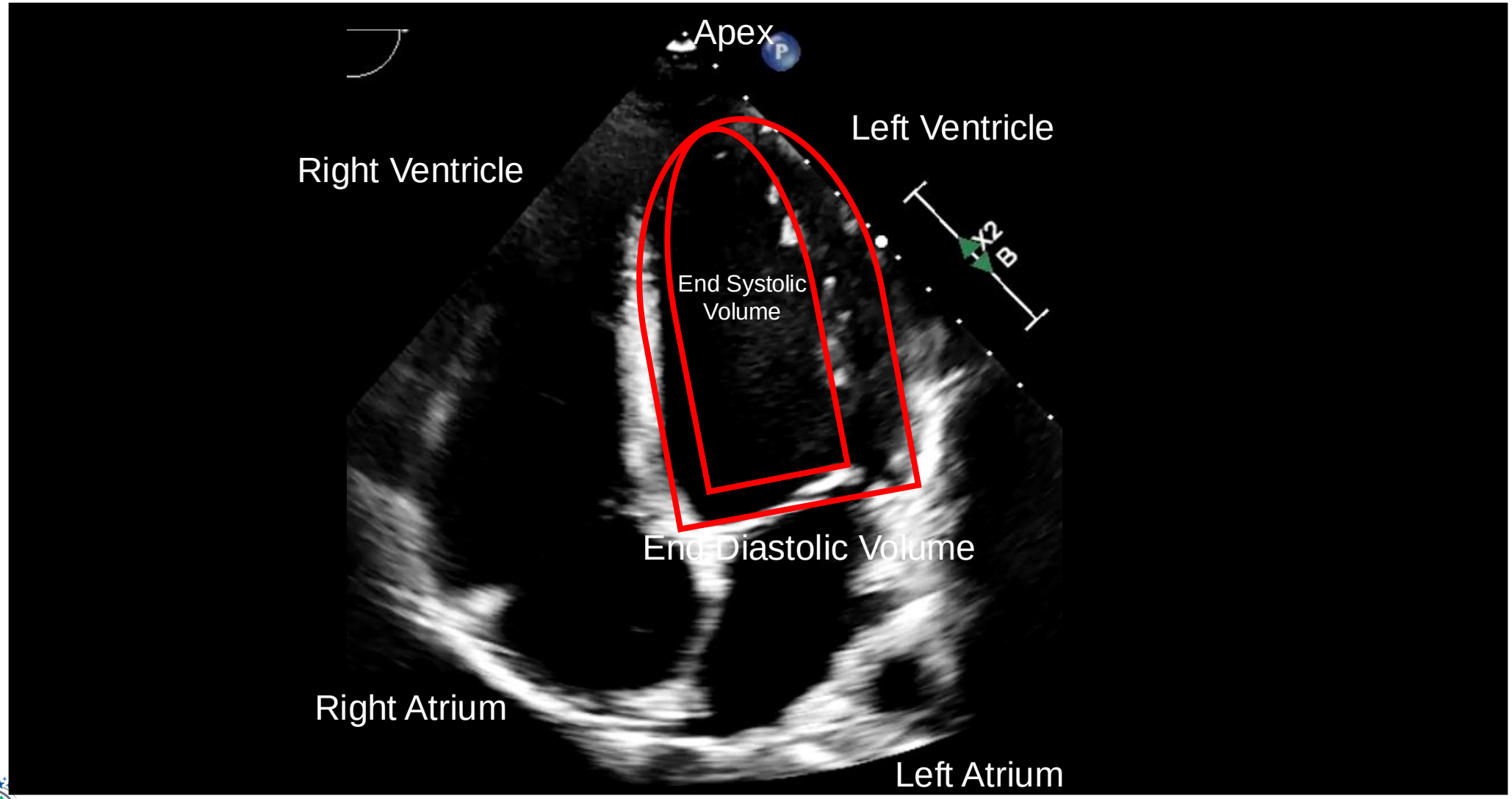
End-Diastolic Volume



End-systolic Volume

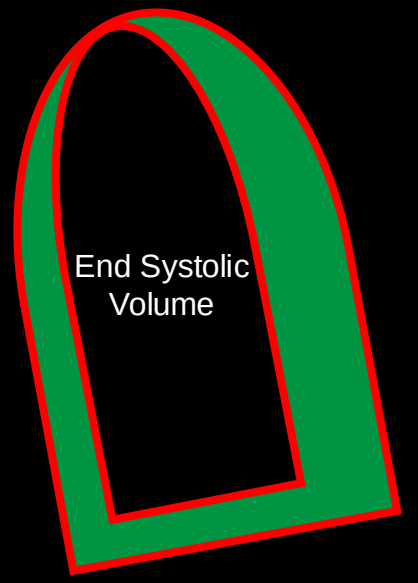


Left Ventricular Stroke Volume



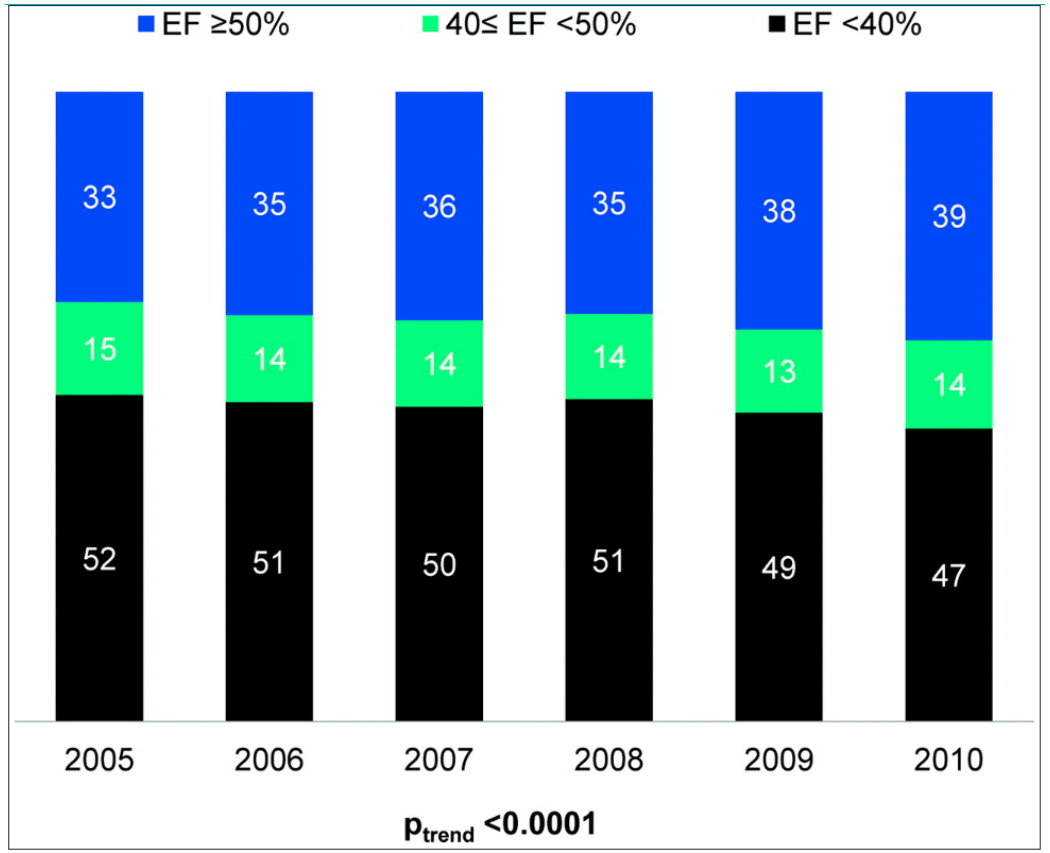
Left Ventricular Ejection Fraction

- Stroke Volume:
  - SV = EDV – ESV
- Ejection Fraction
  - EF = SV/EDV * 100
  - Normal is >52-55%
- Cardiac output:
  - SV x Heart Rate



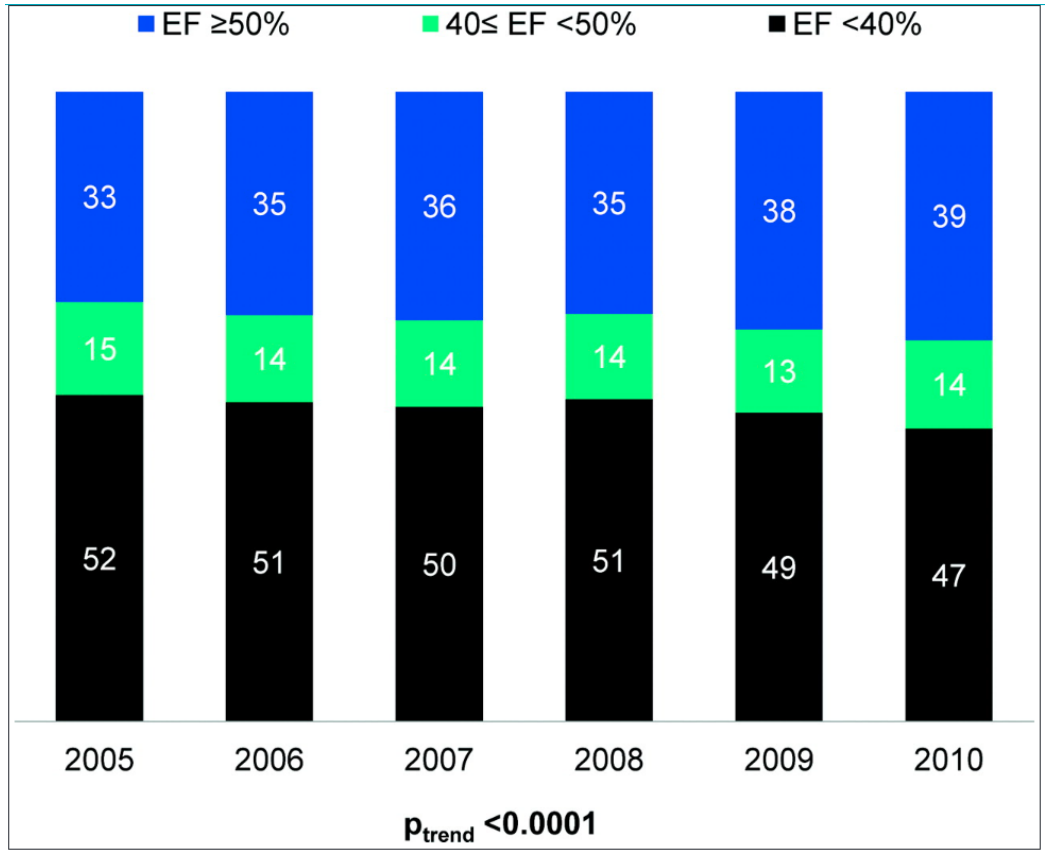
End Diastolic Volume

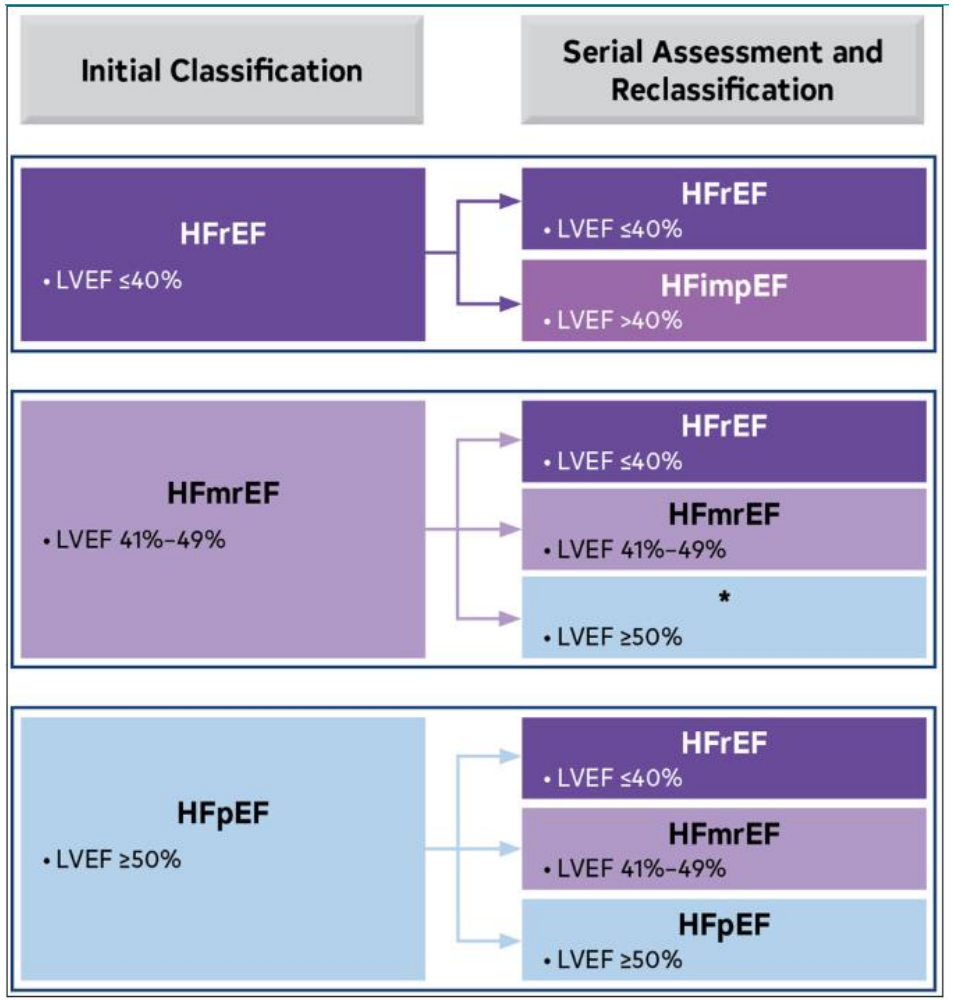
Role of Ejection Fraction



2022 AHA/ACC/HFSA HF Guidelines
Steinberg et al Circ 2012

Role of Ejection Fraction





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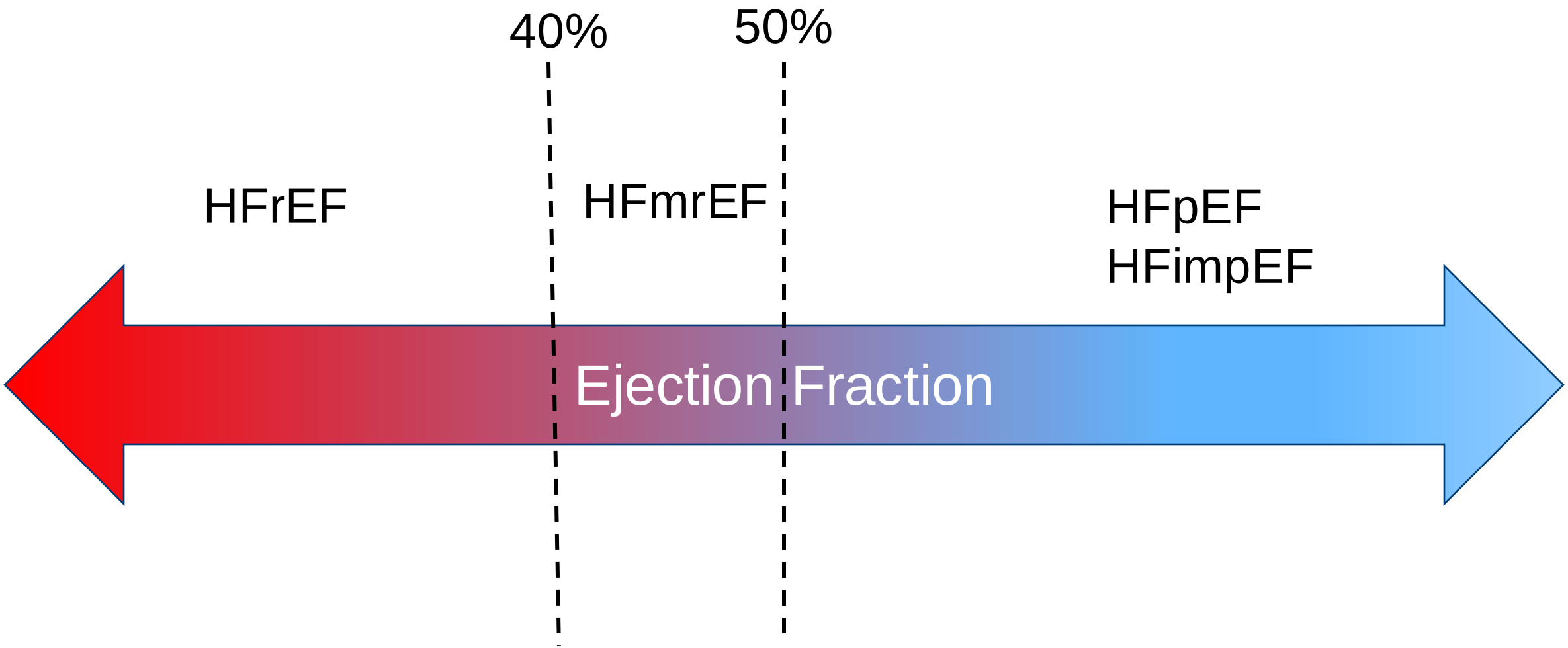

40%
50%
HFrEF
HFmrEF
HFpEF
HFimpEF
Ejection Fraction

Cardiomyopathies

Reduced LVEF

Systolic Dysfunction

- Dilated
  - Genetic
  - Toxin
  - Infectious
  - Peri-partum
  - Inflammatory
- Arrhythmogenic
- Ischemic

Preserved LVEF

Diastolic Dysfunction

- Infiltrative
  - Amyloidosis
  - Iron
- Genetic
  - Hypertrophic
  - Storage diseases
- Sarcoidosis
- Metabolic (“HFpEF”)

Dilated cardiomyopathy

Genetic

Cardiac phenotype:
- titin (TTN)
- lamin A/C (LMNA)
- myosin heavy chain (MYH7)
- troponin T (TNT2)
- Myosin binding protein C (MYBPC3) phospholamban (PLN)

Neuromuscular disorders:
- Duchenne muscular dystrophy
- Becker muscular dystrophy

Infection

Post-myocarditis:
- Enteroviruses (e.g., Coxsackieviruses),
- Parvovirus B19
- Adenoviruses,
- Herpes Viruses (Human Cytomegalovirus, Epstein-Barr virus, Human Herpesvirus 6),
- Echoviruses,
- Hepatitis C Virus,
- Influenza A virus

Other:
[e.g. HIV, Chagas]

Systemic immune-mediated disease

Autoimmune:
- Rheumatoid arthritis
- Coeliac disease
- Systemic lupus erythematosus
- Dermatomyositis, polymyositis
- Systemic sclerosis
- Primary biliary cirrhosis
- Vasculitis (some)
- Myasthenia gravis
- Pemphigus, pemphigoid

Autoinflammatory:
[e.g. Chron disease, ulcerative colitis, gout, reactive arthritis, rare monogenic disorders]

Toxic and overload

- Alcohol (ethanol)
- Cocaine
- Amphetamines
- Ecstasy
- Anabolic steroids
- Arsenic
- Cobalt
- Carbon-monoxide
- Lead
- Iron-overload/haemochromatosis
- Amyloidosis

Drugs

Chemotherapy:
[anthracycline, trastuzumab, antimetabolites, alkylating agents, monoclonal antibodies, tyrosine kinase inhibitors, immunomodulating agents]

Other:
[clozapine, olzapine, chlorpromazine, risperidone, lithium, trycyclic antidepressants, ephedrine
all-trans retinoic acid
phenothiazines]

Endocrine/ metabolic

- Diabetes mellitus,
- Acromegaly
- Pheochromocytoma
- Thyroid gland dysfunction

Peripartum

Idiopathic cardiomyopathy presenting with HFrEF towards the end of pregnancy, or in the months following delivery, where no other cause of HF is found

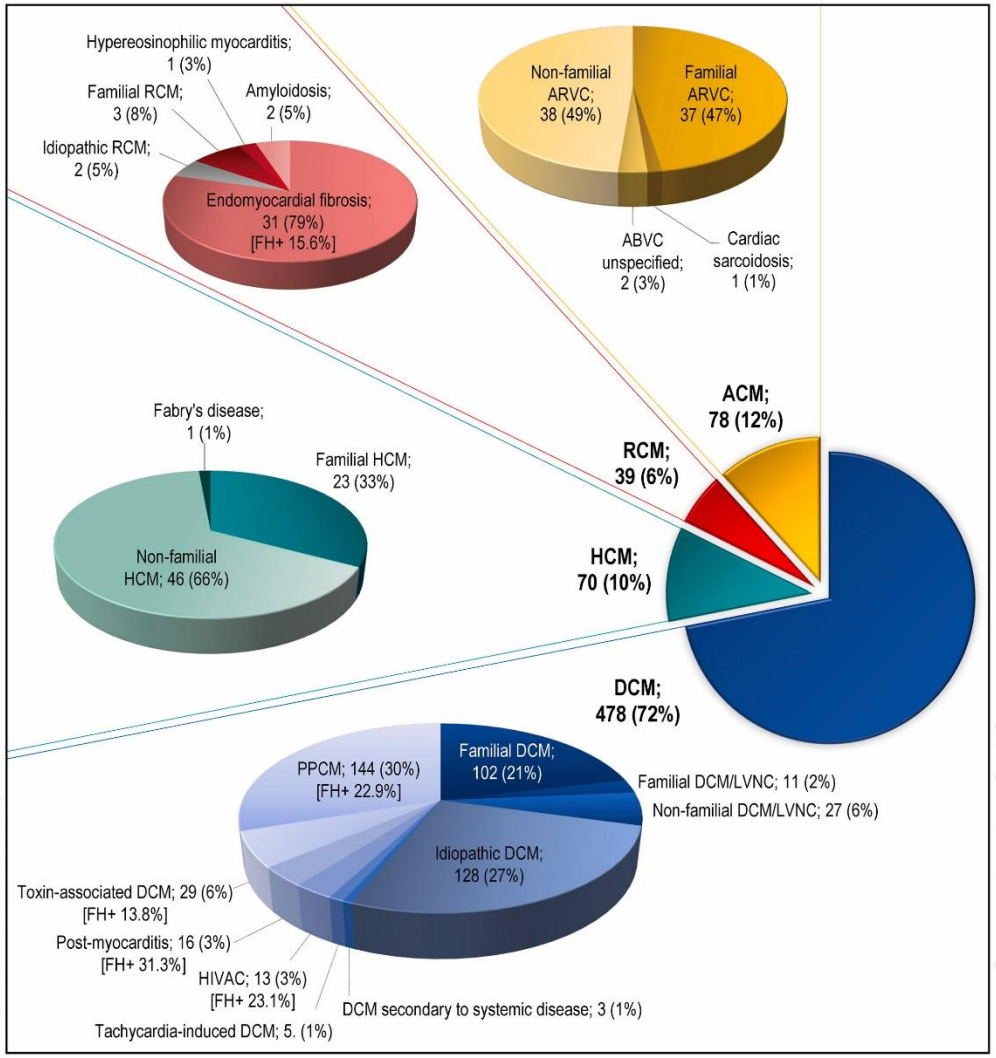
Preserved LVEF

Table 2 Potential specific aetiologies underlying heart failure with preserved ejection fraction-like syndromes in Step 4 (F_2)

Abnormalities of the myocardium		
Ischaemic		Myocardial post-infarction/scar[49] Myocardial stunning[50] Epicardial coronary artery disease[51] Microvascular and endothelial dysfunction[52,53–55]
Toxic	Recreational substance abuse	Such as alcohol,[56] cocaine,[57] and anabolic steroids[58]
	Heavy metals	Such as iron,[59] lead,[60] cadmium,[60] cobalt,[61] copper (M. Wilson)[62]
	Medications	Such as chloroquine,[63] ergotamine,[64] cytostatic drugs (e.g. anthracyclines),[64] immunomodulating drugs (e.g. interferons monoclonal antibodies such as trastuzumab, cetuximab)[64]
	Radiation	Mean cardiac radiation doses > 3 Gy[65,66]
Immune and inflammatory	Related to infection	Such as cardiotropic viruses,[67,68] HIV,[69–71] hepatitis,[72] helminths,[73] parasites (e.g. Chagas' disease[74])
	Not related to infection	Lymphocytic myocarditis,[75–79] autoimmune diseases (e.g. rheumatoid arthritis,[80] connective tissue disorders like scleroderma,[81] M. Raynaud,[55] systemic lupus erythematosus,[82] dermato/polymyositis,[83] and hypersensitivity and eosinophilic myocarditis[73,84–87]
Infiltrative	Related to malignancy	Direct infiltrations and metastases[88–90]
	Not related to malignancy	Amyloidosis,[19,91] sarcoidosis,[92,93] primarily and secondary haemochromatosis,[94–96] storage diseases[97] (e.g. Fabry disease,[98,99] Danon disease,[100–102] Pompe disease,[99,102] PRKAG2 deficiency,[99] Gaucher's disease[99]) [103,104,105,106]
Metabolic	Hormonal	Such as thyroid diseases,[107,108] parathyroid diseases,[109] acromegaly,[110] GH deficiency,[111] Cushing disease,[112] Conn's disease,[113] Addison disease,[114] phaeochromocytoma,[115] pathologies related to pregnancy and peripartum[116,117]
	Nutritional	Such as deficiencies in thiamine,[118] L-carnitine,[119] selenium,[120] (functional) iron,[121,122] complex malnutrition (e.g. AIDS, infections,[73] anorexia nervosa[73,123,124])
Genetic	Diverse forms	Such as HCM,[97,125,126] restrictive cardiomyopathies,[103,104,106] hypertrophic form of non-compaction cardiomyopathy,[127,128] early forms of muscular dystrophies (Duchenne/Becker disease[129]).
Endomyocardial		HES,[84] EMF,[71,127] endocardial fibroelastosis,[128] carcinoid,[130,131] endocardial calcification (Paget's disease[132])
Abnormalities of loading conditions		
Hypertension		Primary and secondary forms of hypertension[112,113,115,130,131]
Valvular and structural defects	Acquired	Heart valve diseases[133,134]
Valvular and structural defects	Congenital	Septal defects[132,135,136]
Pericardial and endomyocardial pathologies	Pericardial	Constrictive pericarditis and pericardial effusion[137,138]
	Endomyocardial	HES,[86] EMF,[73,139] endocardial fibroelastosis,[140] carcinoid,[141,142] endocardial calcification (Paget's disease[143])
High output states		Severe anaemia,[144] sepsis,[145] thyrotoxicosis,[105] arteriovenous fistula,[146] and pregnancy[147]
Volume overload		Renal failure and fluid overload[148,149,150]
Abnormalities of the cardiac rhythm		
Rhythm disorders		Atrial/ventricular arrhythmias, pacing, conduction disorders[38,151–153]

EMF, endomyocardial fibrosis; GH, growth hormone; HCM, hypertrophic cardiomyopathy; HES, hypereosinophilic syndrome (formerly known as Löffler's endocarditis); HIV/AIDS, human immunodeficiency virus/acquired immune deficiency; LV, left ventricular; PRKAG2, protein kinase AMP-activated non-catalytic subunit gamma 2.

Heart Failure in South Africa and Mozambique



Sarah M. Kraus et al. *JACC Adv* 2024;

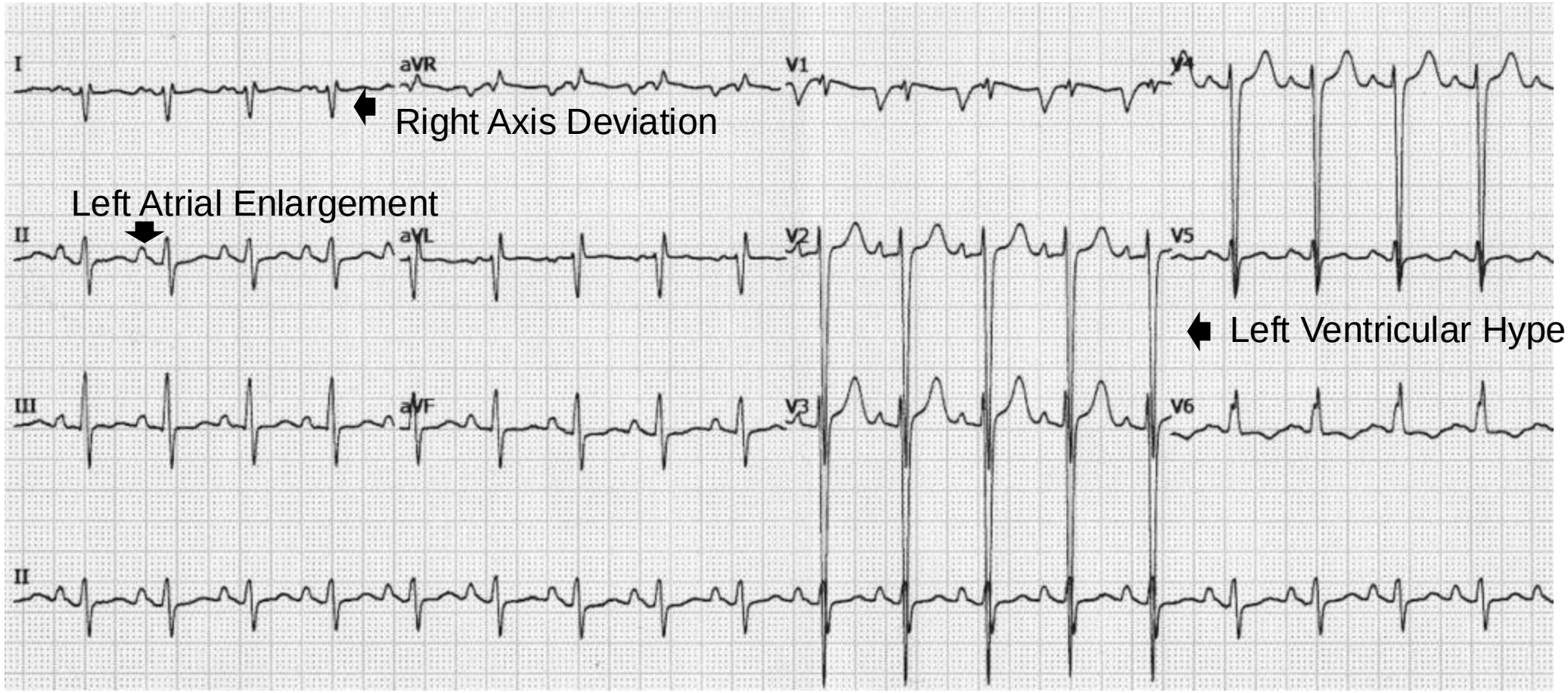
Cardiomyopathies for Today

1. Dilated
2. Hypertrophic
3. Arrhythmogenic
4. Restrictive
5. Ischemic

Dilated Cardiomyopathy

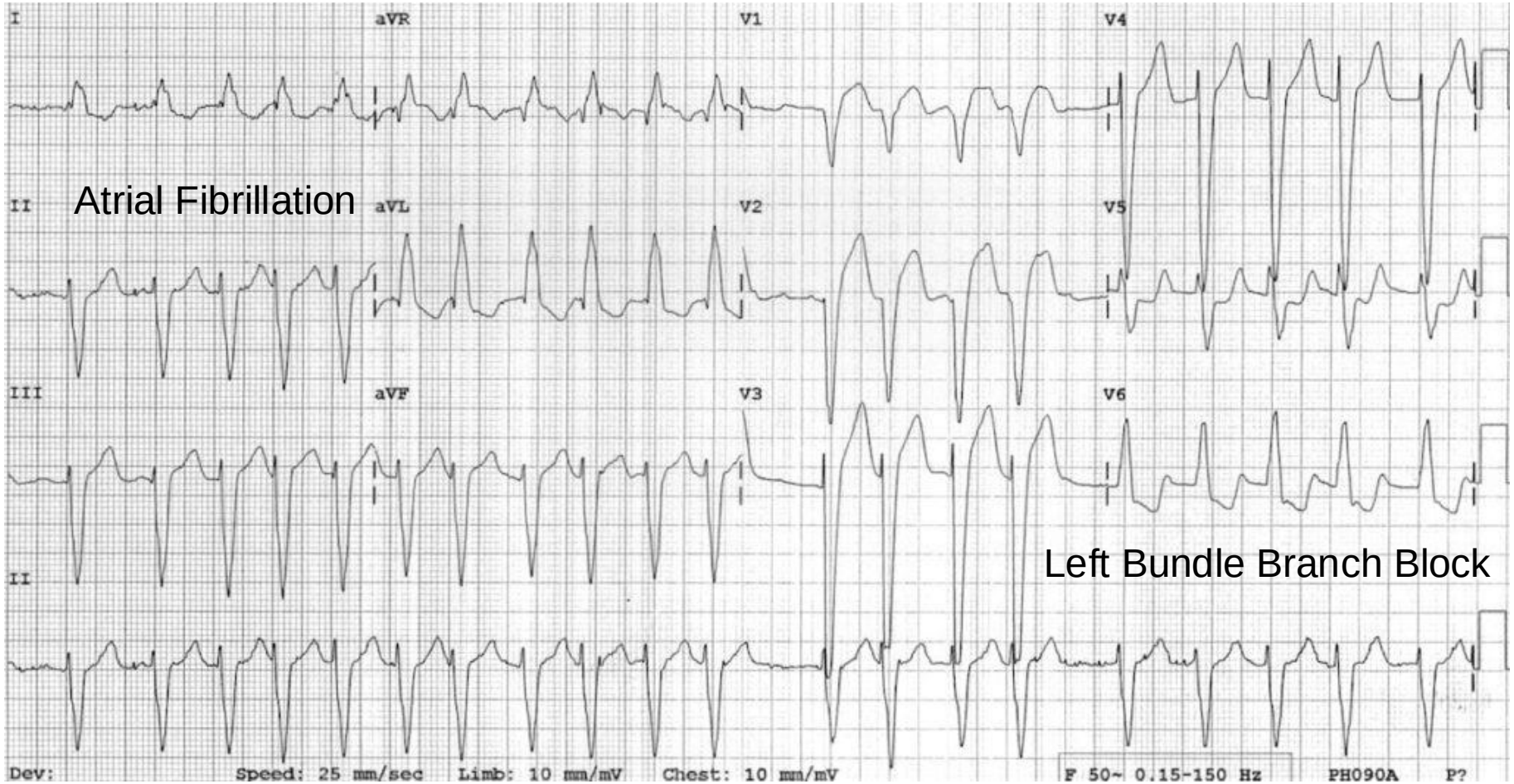
- Left ventricle is enlarged
- Systolic Dysfunction
  - LVEF < 50%
- Common in Africa:
  - 10-17% of all cardiac conditions
  - 17-48% of Heart Failure
- Male > Female
- 3rd and 4th decade of life
  - Can be any age

Dilated CM: ECGs



https://litfl.com/dilated-cardiomyopathy-dcm-ecg-library/

Dilated CM: ECGs



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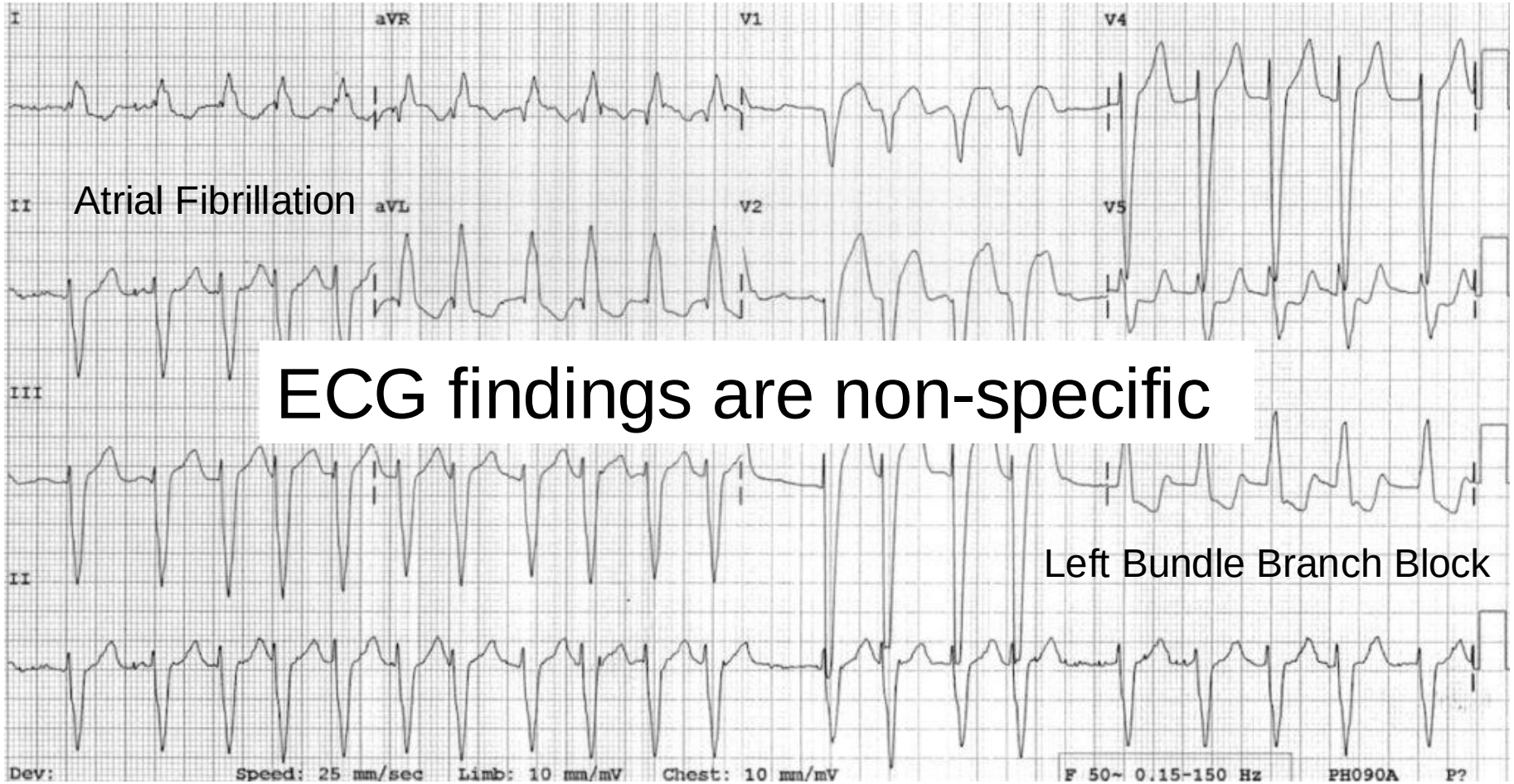
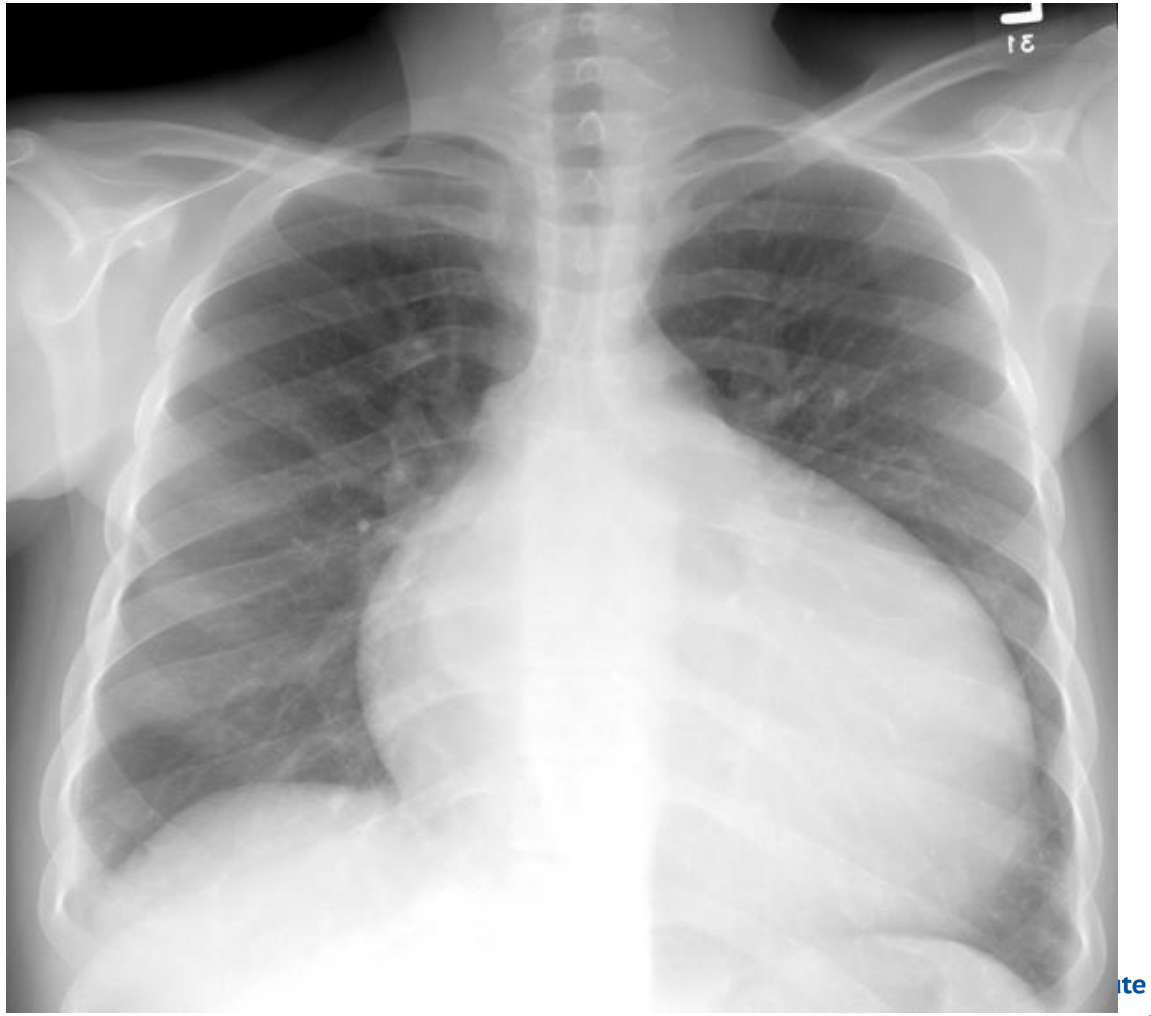
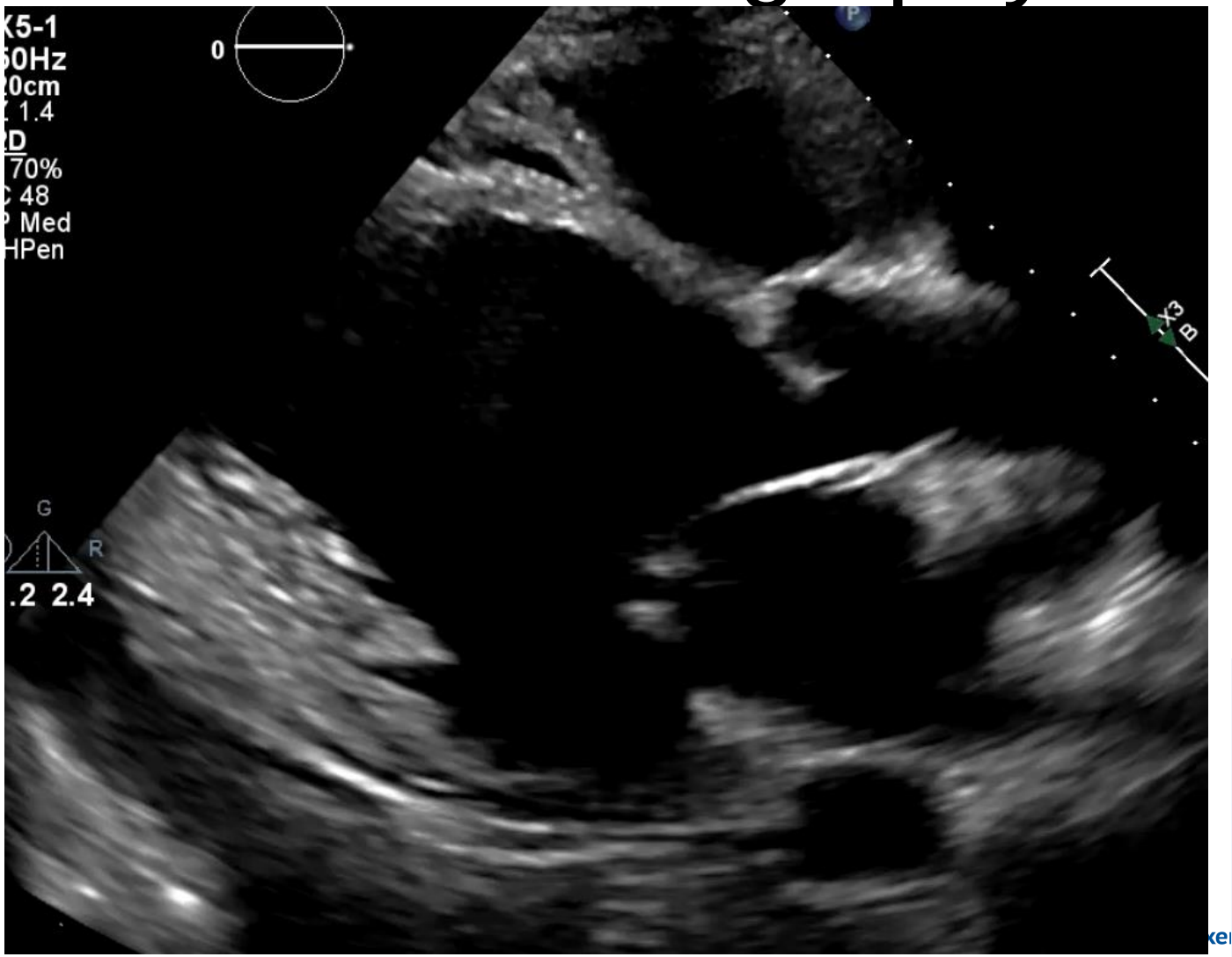


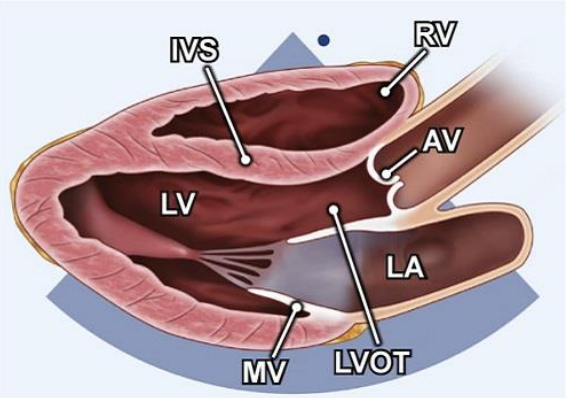
https://litfl.com/dilated-cardiomyopathy-dcm-ecg-library/

Dilated CM: X-ray



Dilated CM: Echocardiography





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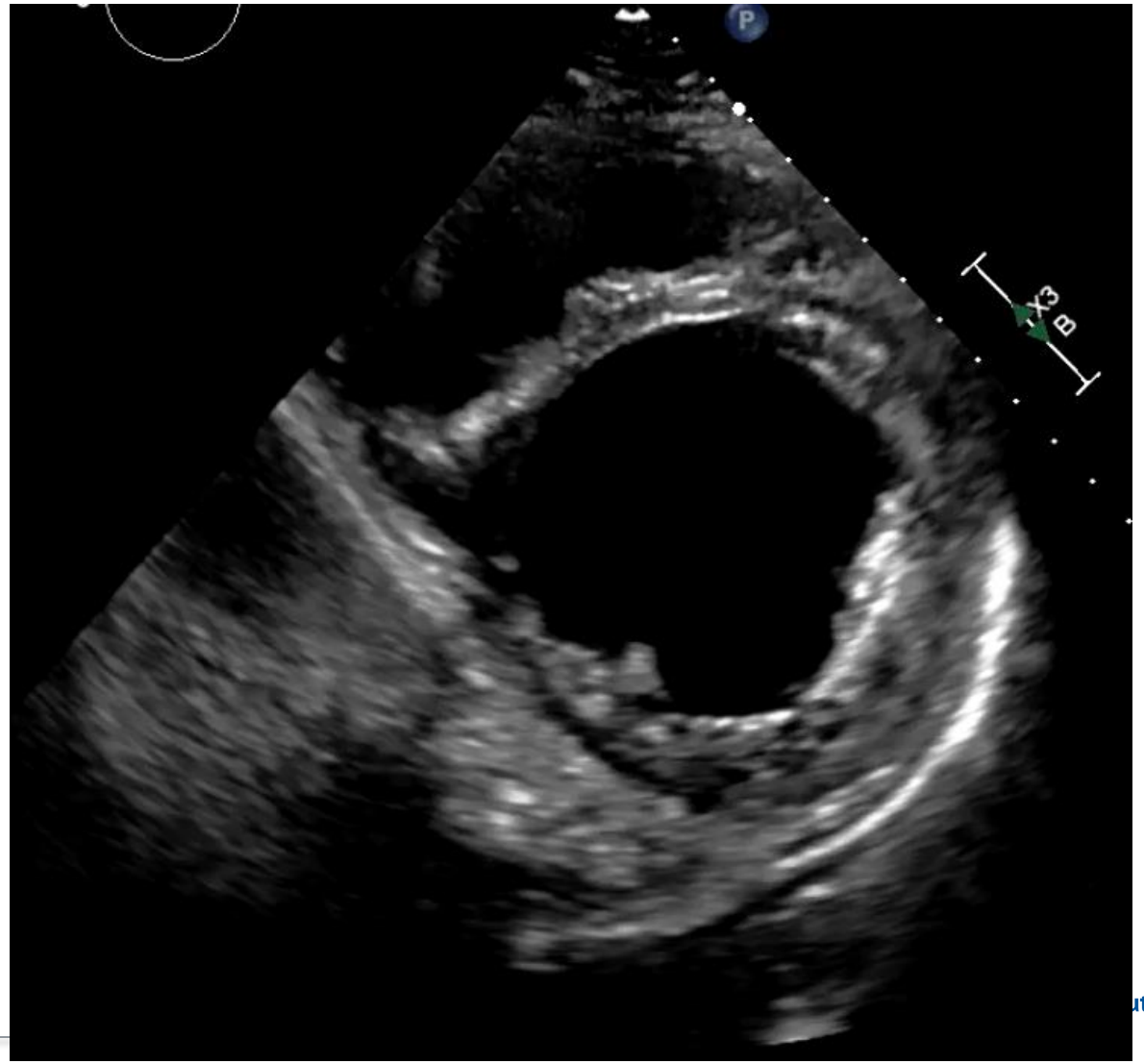

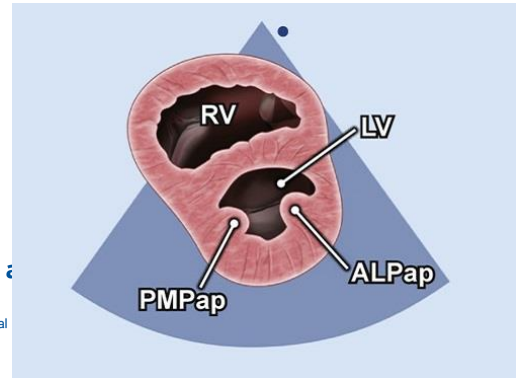

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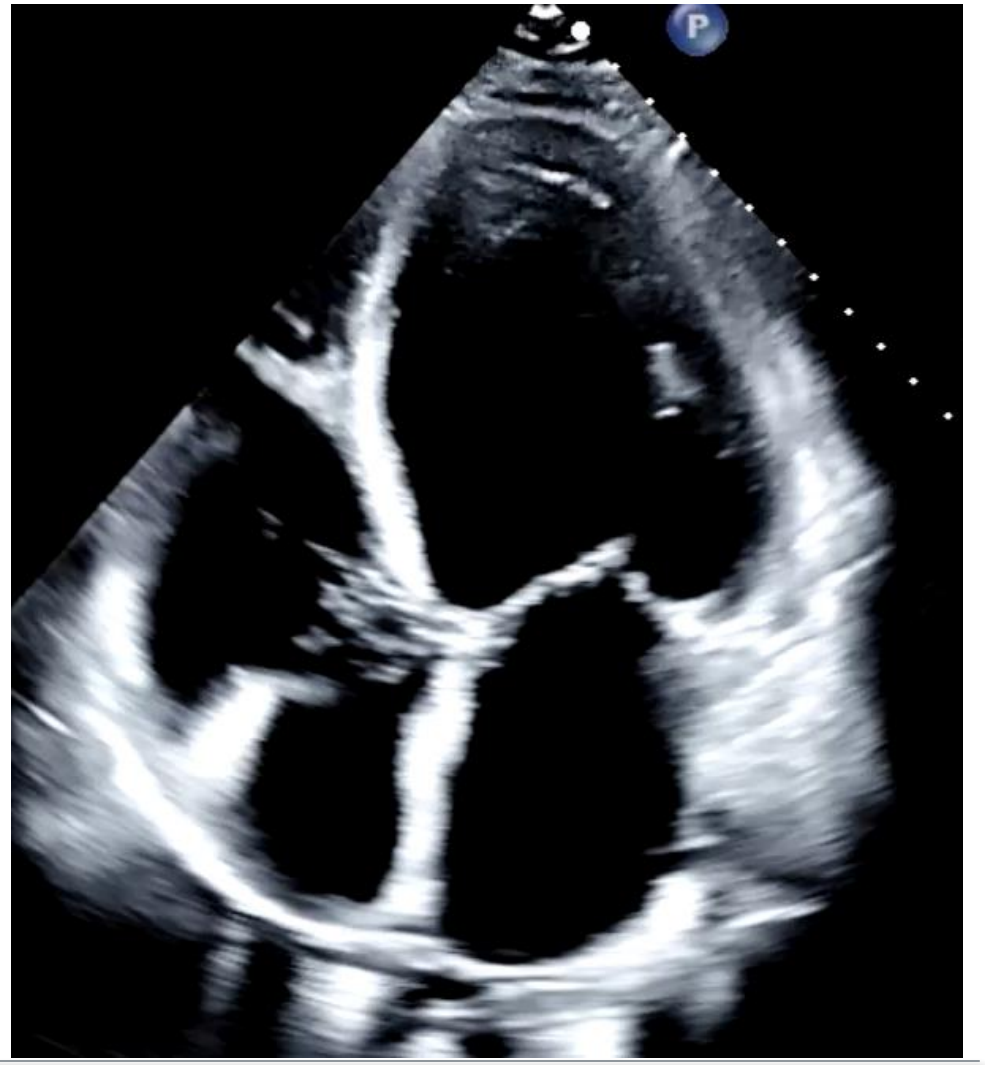
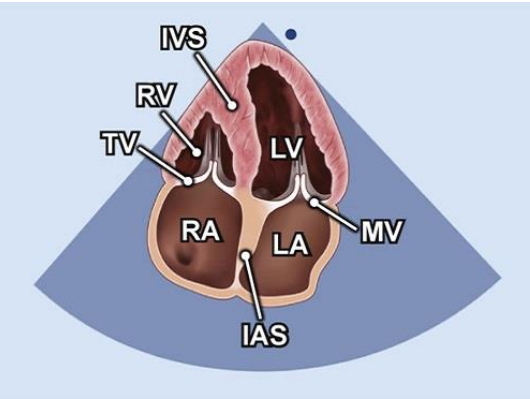
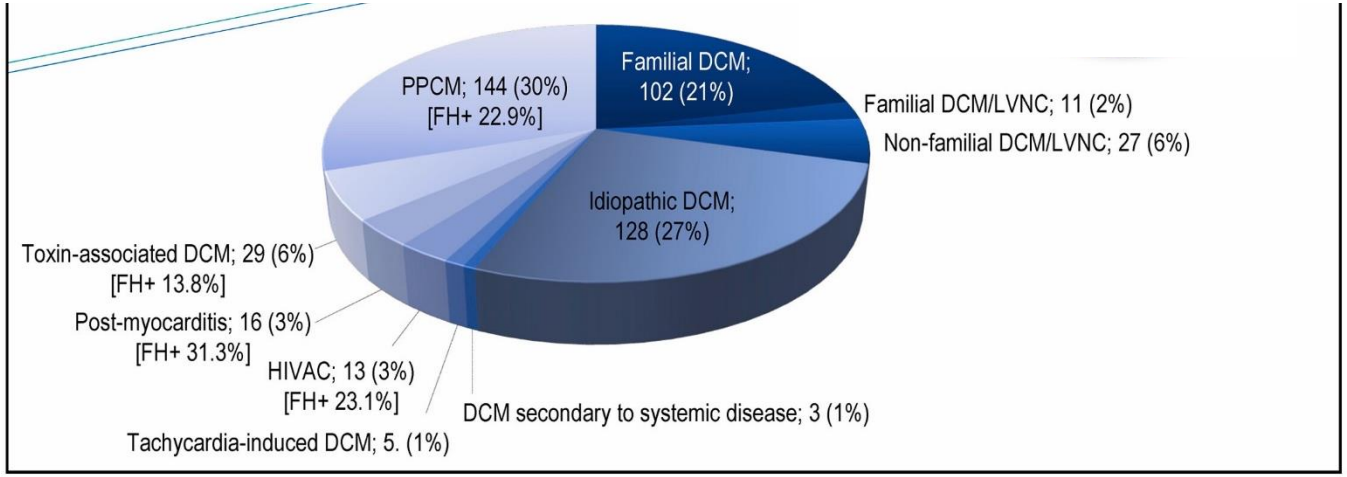
Dilated CM: Echocardiography



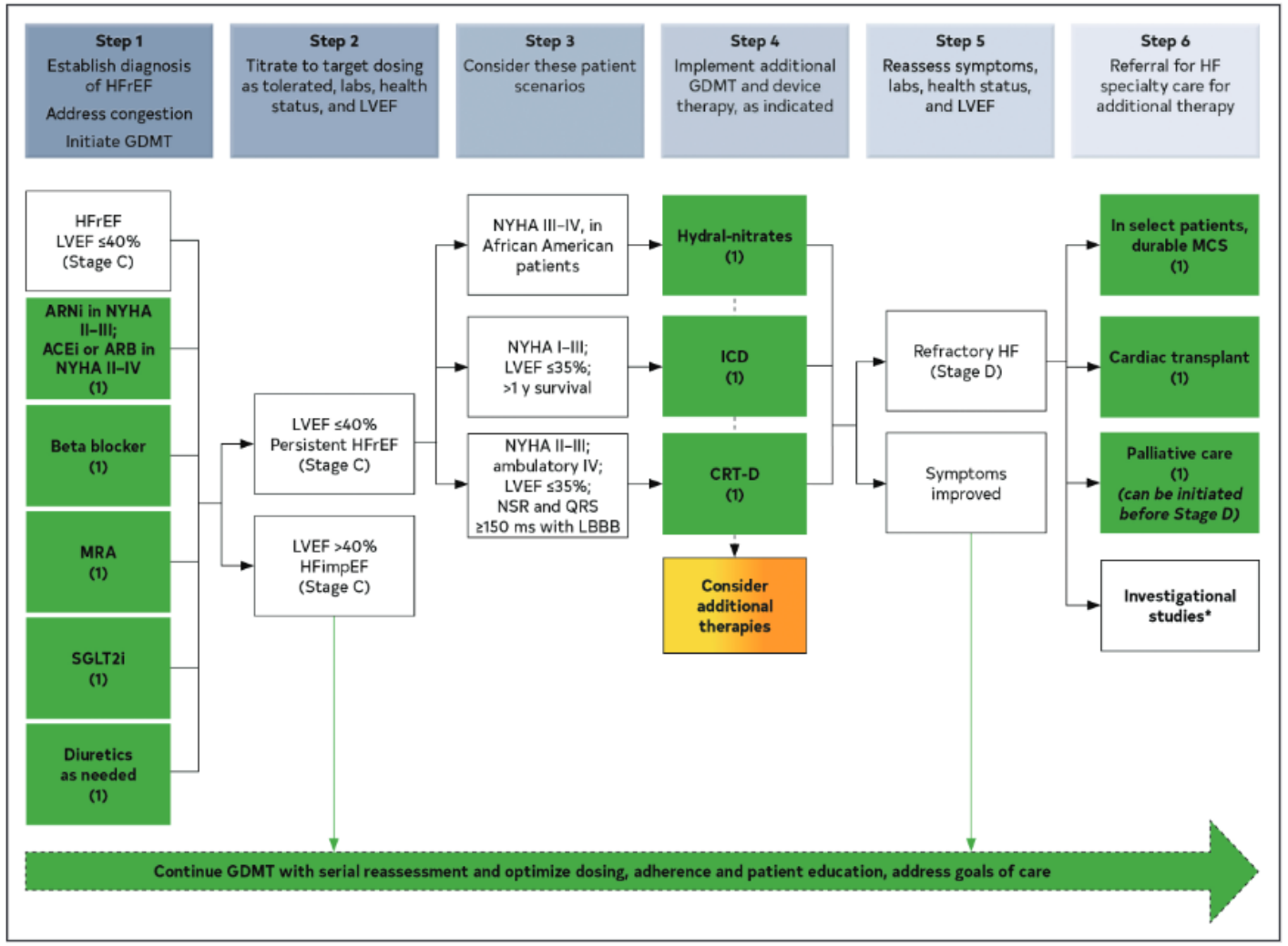


Dilated DM: Etiology

- Prevalence of etiologies of DM in Africa is a major unmet need
- Consider:
  - Peri-partum (30%)
    - Last month or within 5 months of pregnancy
  - Myocarditis
  - Toxin
    - Alcohol and Thiamine
  - “Burned out” HF
    - Hypertension
  - Genetic
- Idiopathic

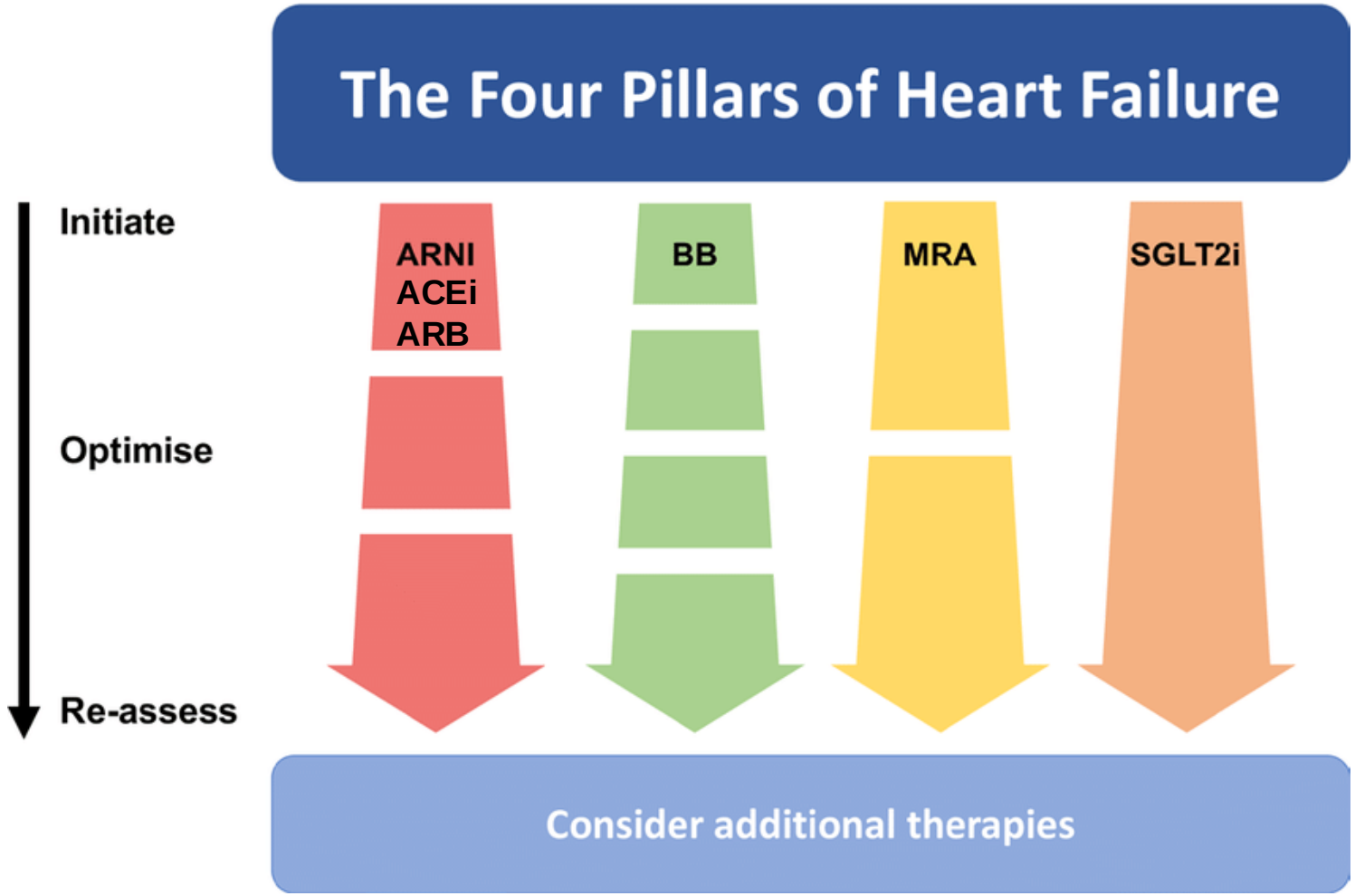


Management


Step 1
Establish diagnosis of HFrEF
Address congestion
Initiate GDMT
Step 2
Titrate to target dosing as tolerated, labs, health status, and LVEF
Step 3
Consider these patient scenarios
Step 4
Implement additional GDMT and device therapy, as indicated
Step 5
Reassess symptoms, labs, health status, and LVEF
Step 6
Referral for HF specialty care for additional therapy
HFrEF LVEF ≤40% (Stage C)
ARNi in NYHA II–III; ACEi or ARB in NYHA II–IV (1)
Beta blocker (1)
MRA (1)
SGLT2i (1)
Diuretics as needed (1)
LVEF ≤40% Persistent HFrEF (Stage C)
LVEF >40% HFimpEF (Stage C)
NYHA III–IV, in African American patients
NYHA I–III; LVEF ≤35%; >1 y survival
NYHA II–III; ambulatory IV; LVEF ≤35%; NSR and QRS ≥150 ms with LBBB
Hydral-nitrates (1)
ICD (1)
CRT-D (1)
Consider additional therapies
Refractory HF (Stage D)
Symptoms improved
In select patients, durable MCS (1)
Cardiac transplant (1)
Palliative care (1) (can be initiated before Stage D)
Investigational studies*
Continue GDMT with serial reassessment and optimize dosing, adherence and patient education, address goals of care

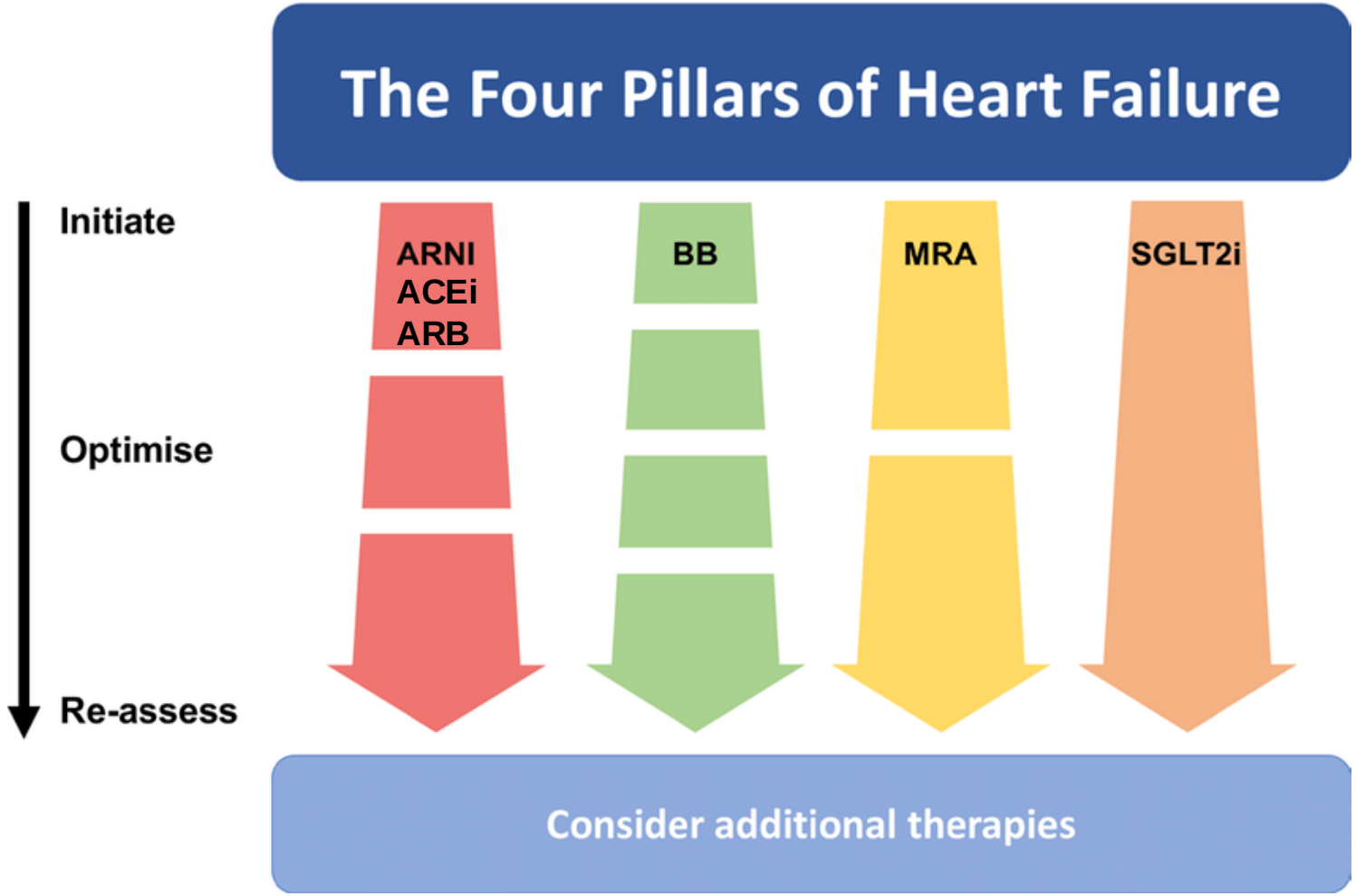
Medical Management

- Renin-Angiotension Aldosterone system:
  - Sacubitril/Valsartan
  - ACE Inhibitor
  - Angiotensin Receptor Blocker
- Start low and maximize dose
- Caution: Hypotension



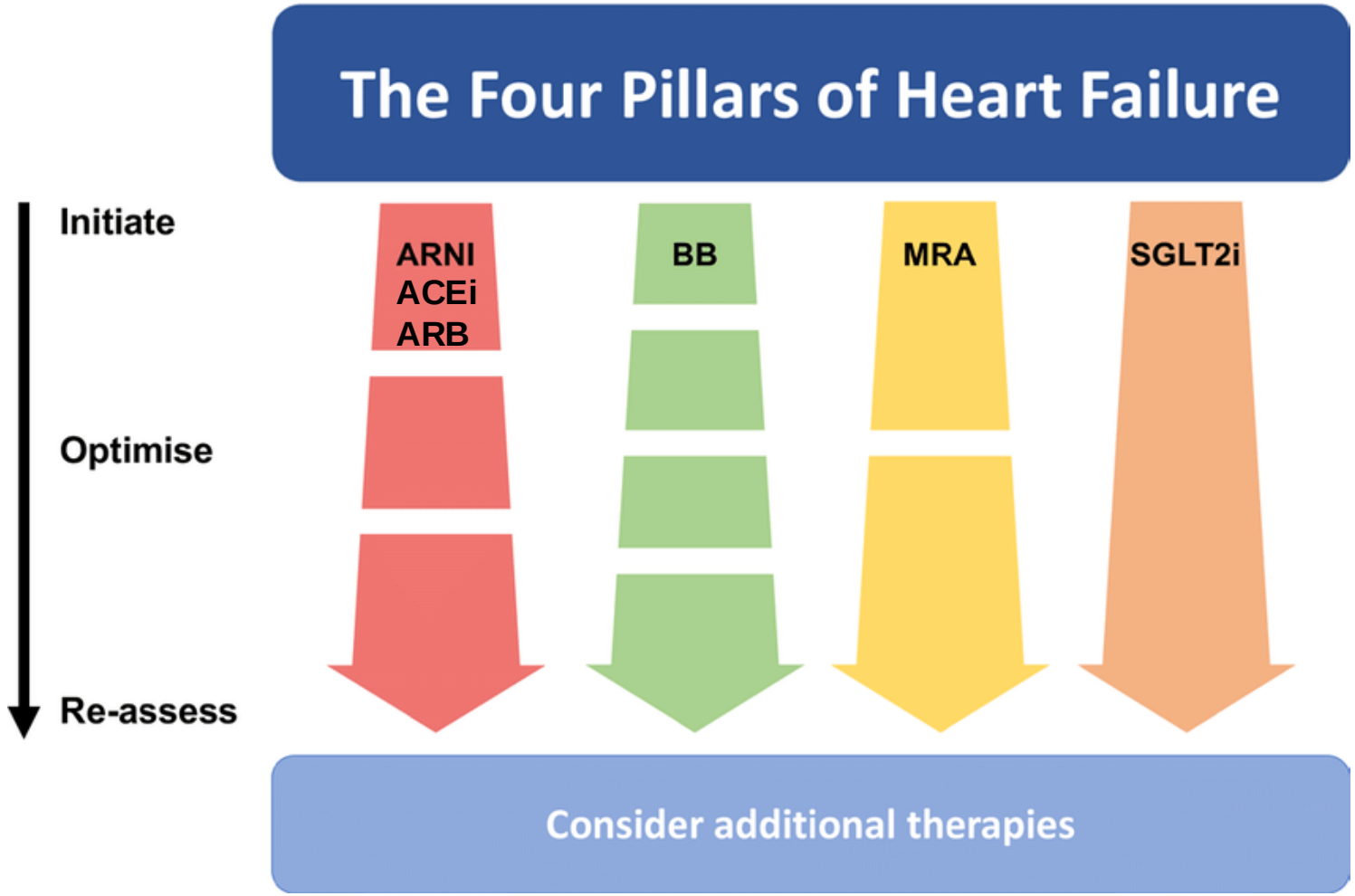
Medical Management

- Beta Blocker:
  - ↓ Mortality
  - Metoprolol
  - Carvedilol
    - Concurrent Hypertension
  - Bisoprolol
- Start low, titrate to maximally tolerated dose
- Caution: Acutely Decompensated



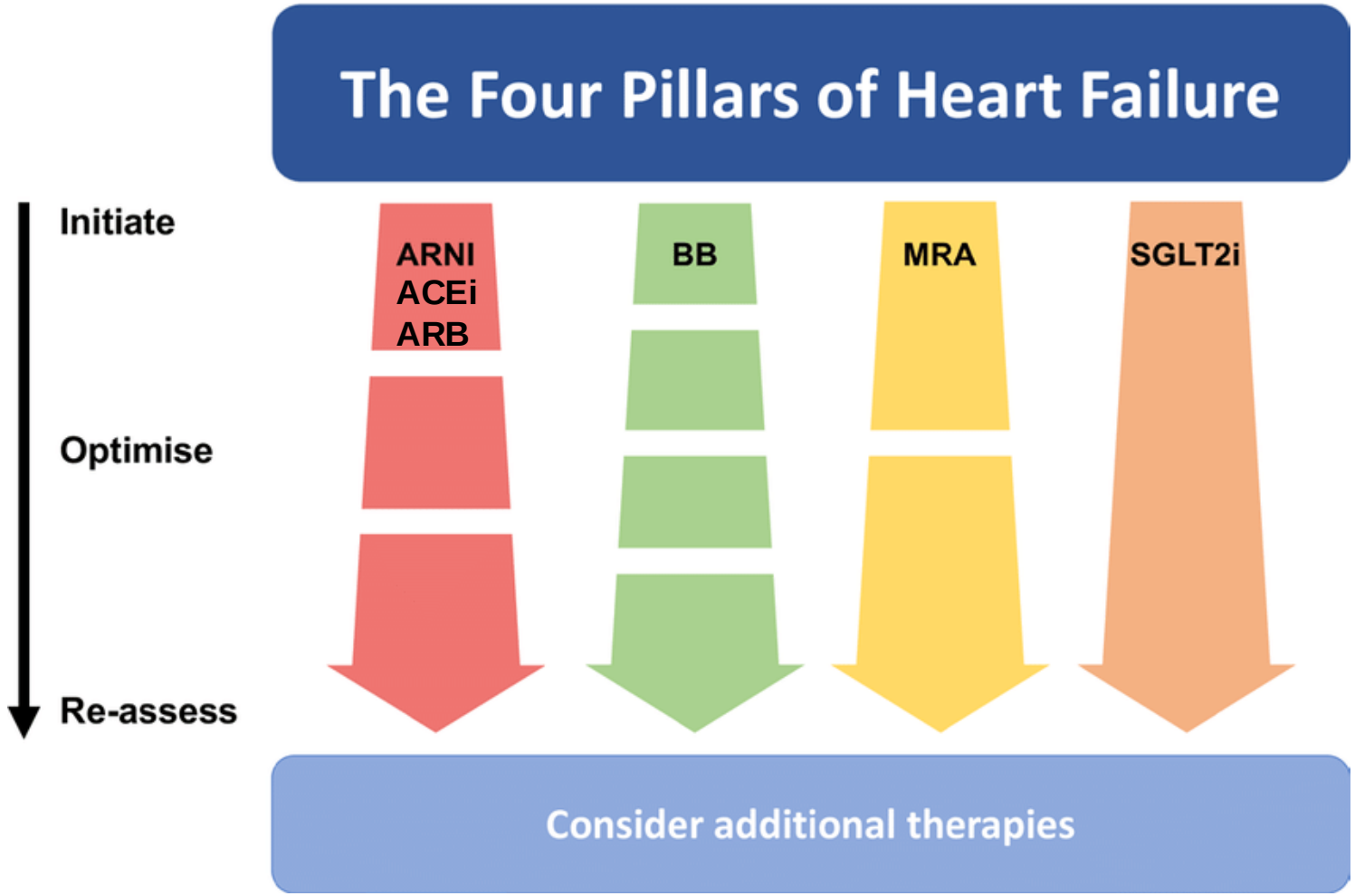
Medical Management

- Mineralocorticoid Receptor Antagonist
  - Spironolactone
  - Eplerenone
- 3rd agent
- Starting dose
  - 25mg daily
  - Increase to 50mg daily
- Caution: Kidney Disease, ↑K



Medical Management

- Sodium-Glucose Cotransporter-2 Inhibitors
  - Empagliflozin
  - Dapagliflozin
  - Others
- New Agent
- Fixed dose
- Caution: Patient Access, Type 1 Diabetes, Urinary Tract Infections



Medical Management

- ↓ vascular congestions and ↓ cardiac filling pressures
- ↓ Morbidity and symptoms
- Loop Diuretics
  - Forsemide
  - Toresmide
  - Bumetanide
- Combination Diuretic Therapy
  - Loop + Thiazide

Recommendations for Diuretics and Decongestion Strategies in Patients With HF
Referenced studies that support the recommendations are summarized in the Online Data Supplements.

COR	LOE	Recommendations
1	B-NR	1. In patients with HF who have fluid retention, diuretics are recommended to relieve congestion, improve symptoms, and prevent worsening HF.[1–5]
1	B-NR	2. For patients with HF and congestive symptoms, addition of a thiazide (eg, metolazone) to treatment with a loop diuretic should be reserved for patients who do not respond to moderate- or high-dose loop diuretics to minimize electrolyte abnormalities.[6]

Dilated CM: Drugs to Avoid

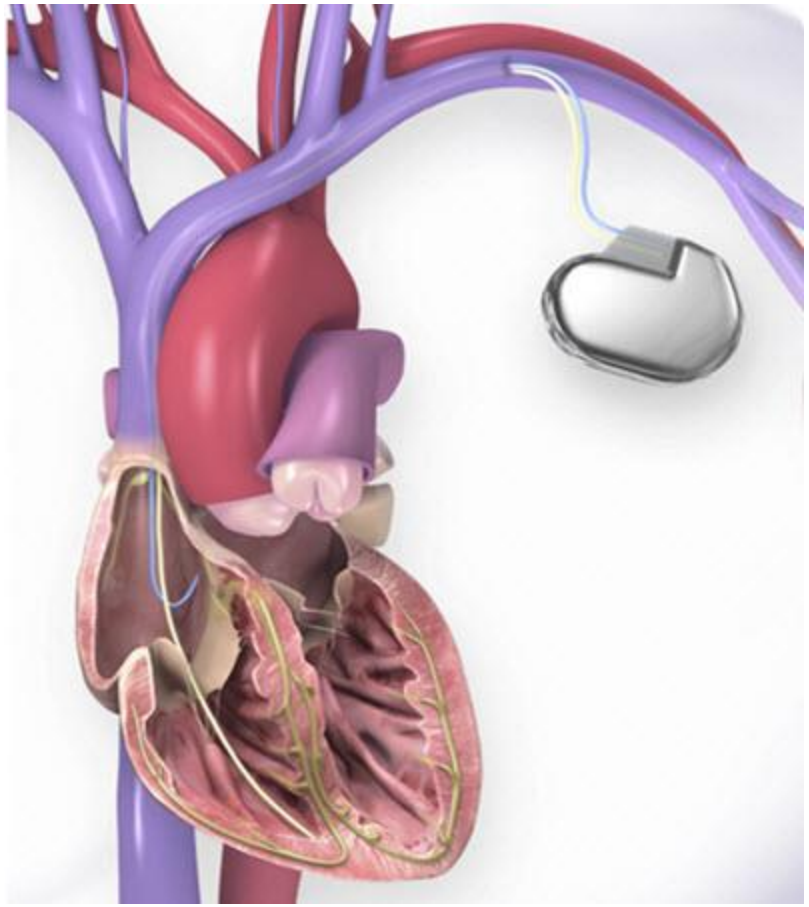
Recommendations for Drugs of Unproven Value or Drugs That May Worsen HF
Referenced studies that support the recommendations are summarized in the Online Data Supplements.

COR	LOE	Recommendations
3: No Benefit	A	1. In patients with HFrEF, dihydropyridine calcium channel-blocking drugs are not recommended treatment for HF.[1,2]
3: No Benefit	B-R	2. In patients with HFrEF, vitamins, nutritional supplements, and hormonal therapy are not recommended other than to correct specific deficiencies.[3–9]
3: Harm	A	3. In patients with HFrEF, nondihydropyridine calcium channel-blocking drugs are not recommended.[10–13]
3: Harm	A	4. In patients with HFrEF, class IC antiarrhythmic medications and dronedarone may increase the risk of mortality.[14–16]
3: Harm	A	5. In patients with HFrEF, thiazolidinediones increase the risk of worsening HF symptoms and hospitalizations.[17–21]
3: Harm	B-R	6. In patients with type 2 diabetes and high cardiovascular risk, the dipeptidyl peptidase-4 (DPP-4) inhibitors saxagliptin and alogliptin increase the risk of HF hospitalization and should be avoided in patients with HF.[22–24]
3: Harm	B-NR	7. In patients with HFrEF, NSAIDs worsen HF symptoms and should be avoided or withdrawn whenever possible.[25–28]

Implantable Cardiac Defibrialltor

Recommendations for ICDs and CRTs Referenced studies that support the recommendations are summarized in the Online Data Supplements.		
COR	**LOE**	**Recommendations**
1	A	1. In patients with nonischemic DCM or ischemic heart disease at least 40 days post-MI with LVEF ≤35% and NYHA class II or III symptoms on chronic GDMT, who have reasonable expectation of meaningful survival for >1 year, ICD therapy is recommended for primary prevention of SCD to reduce total mortality.[1–9]
Value Statement: High Value (A)		2. A transvenous ICD provides high economic value in the primary prevention of SCD particularly when the patient's risk of death caused by ventricular arrythmia is deemed high and the risk of nonarrhythmic death (either cardiac or noncardiac) is deemed low based on the patient's burden of comorbidities and functional status.[10–15]

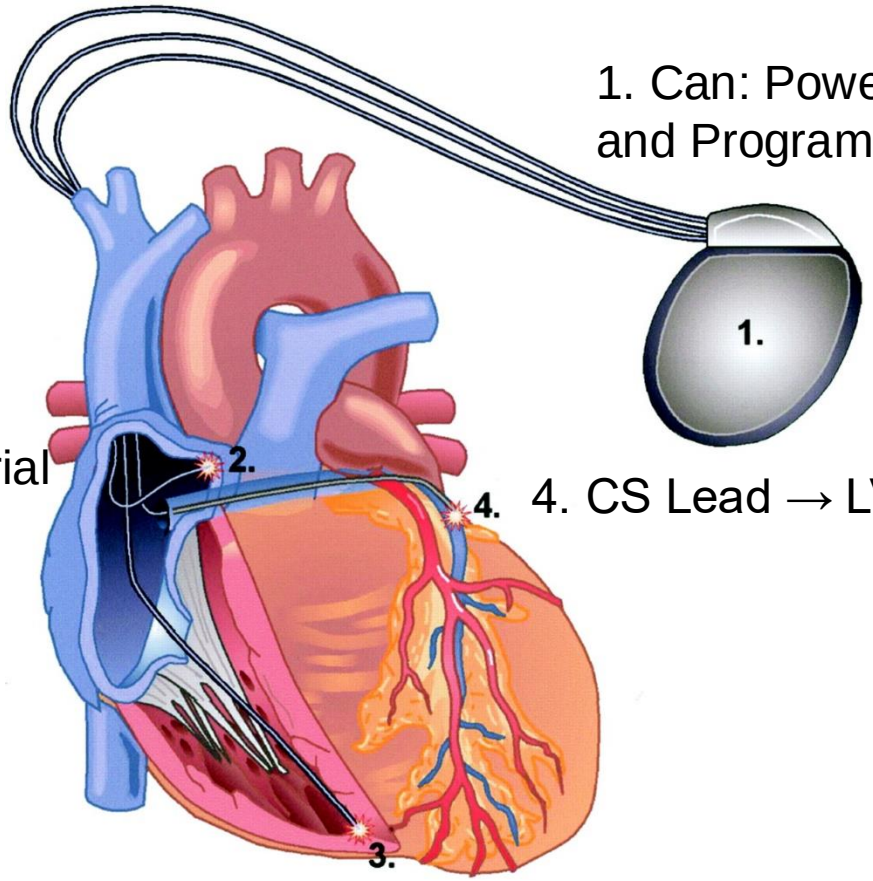
LVEF <35% despite GDMT



Cardiac Re-synchronization Therapy

1	B-R	4. For patients who have LVEF ≤35%, sinus rhythm, left bundle branch block (LBBB) with a QRS duration ≥150 ms, and NYHA class II, III, or ambulatory IV symptoms on GDMT, CRT is indicated to reduce total mortality, reduce hospitalizations, and improve symptoms and QOL.[16–21]
Value Statement: High Value (B-NR)		5. For patients who have LVEF ≤35%, sinus rhythm, LBBB with a QRS duration of ≥150 ms, and NYHA class II, III, or ambulatory IV symptoms on GDMT, CRT implantation provides high economic value.[22–27]

1. LBBB
2. LVEF <35%
3. Symptoms



Therapies Works in HFrEF

Table 15. Benefits of Evidence-Based Therapies for Patients With HFrEF[3–6,8,10–14,23,31–42]

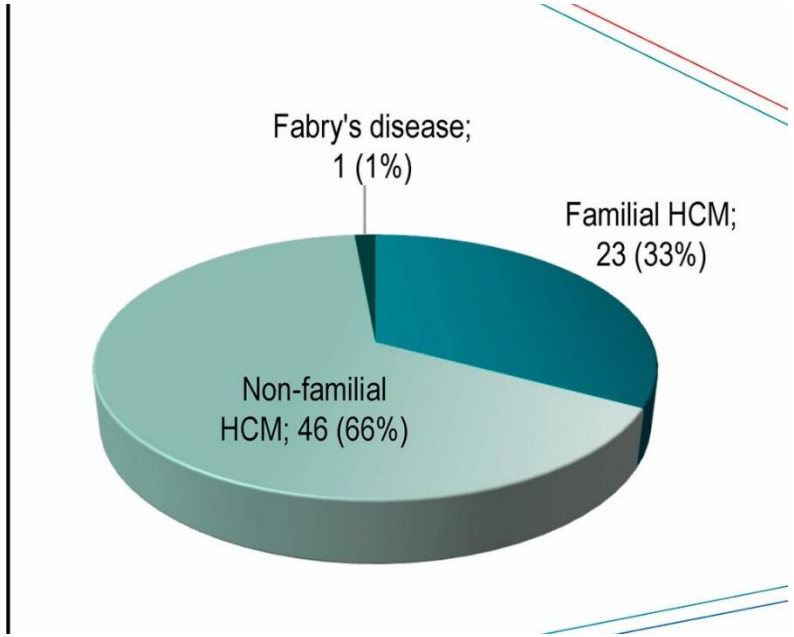
Evidence-Based Therapy	Relative Risk Reduction in All-Cause Mortality in Pivotal RCTs, %	NNT to Prevent All-Cause Mortality Over Time*	NNT for All-Cause Mortality (Standardized to 12 mo)	NNT for All- Cause Mortality (Standardized to 36 mo)
ACEi or ARB	17	22 over 42 mo	77	26
ARNi†	16	36 over 27 mo	80	27
Beta blocker	34	28 over 12 mo	28	9
Mineralocorticoid receptor antagonist	30	9 over 24 mo	18	6
SGLT2i	17	43 over 18 mo	63	22
Hydralazine or nitrate‡	43	25 over 10 mo	21	7
CRT	36	12 over 24 mo	24	8
ICD	23	14 over 60 mo	70	23

ACEi indicates angiotensin-converting enzyme inhibitor; ARB, angiotensin receptor blocker; ARNi, angiotensin receptor neprilysin inhibitor; CRT, cardiac resynchronization therapy; HFrEF, heart failure with reduced ejection fraction; ICD, implantable cardioverter-defibrillator; NNT, number needed to treat; RCT, randomized controlled trial; and SGLT2i, sodium-glucose cotransporter-2 inhibitor.

Hypertrophic Cardiomyopathy

Hypertrophic Cardiomyopathy

- Thickening of the heart muscle
  - Acquired
    - Hypertensive Heart Disease
    - Kidney Disease
    - Valve Disease
  - Genetic
- Common:
  - 34% of Cardiomyopathies in Ethiopia
  - 3rd most common CM in Ghana, S Africa or Mozambique



Acquired vs Famlial

Hypertensive Heart Disease

- No specific inheritance pattern
  - Can still “Run in families”

Hypertrophic Cardiomyopathy

- Autosomal Dominant
  - Genetic Disease of the Sarcomere
  - Many patients are first in their family (new mutations)

Acquired vs Famlial

Hypertensive Heart Disease

- No specific inheritance pattern
  - Can still “Run in families”
- Symmetric hypertrophy

Hypertrophic Cardiomyopathy

- Autosomal Dominant
  - Genetic Disease of the Sarcomere
  - Many patients are first in their family (new mutations)
- Asymmetric Hypertrophy

Acquired vs Famlial

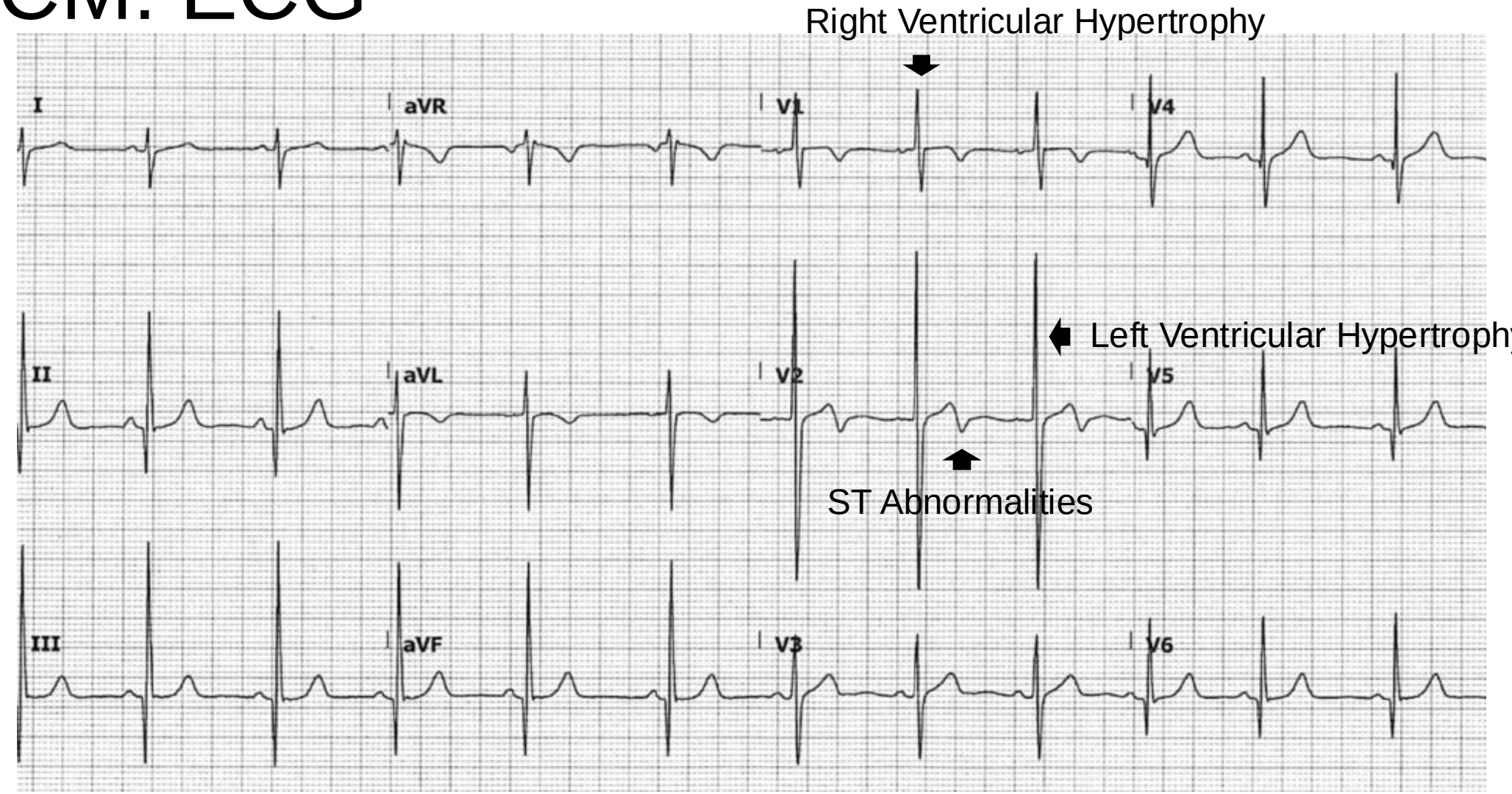
Hypertensive Heart Disease

- No specific inheritance pattern
  - Can still “Run in families”
- Symmetric hypertrophy
- Older patients
  - 6^{th}-7^{th} decade of life in Nigerian Cohort

Hypertrophic Cardiomyopathy

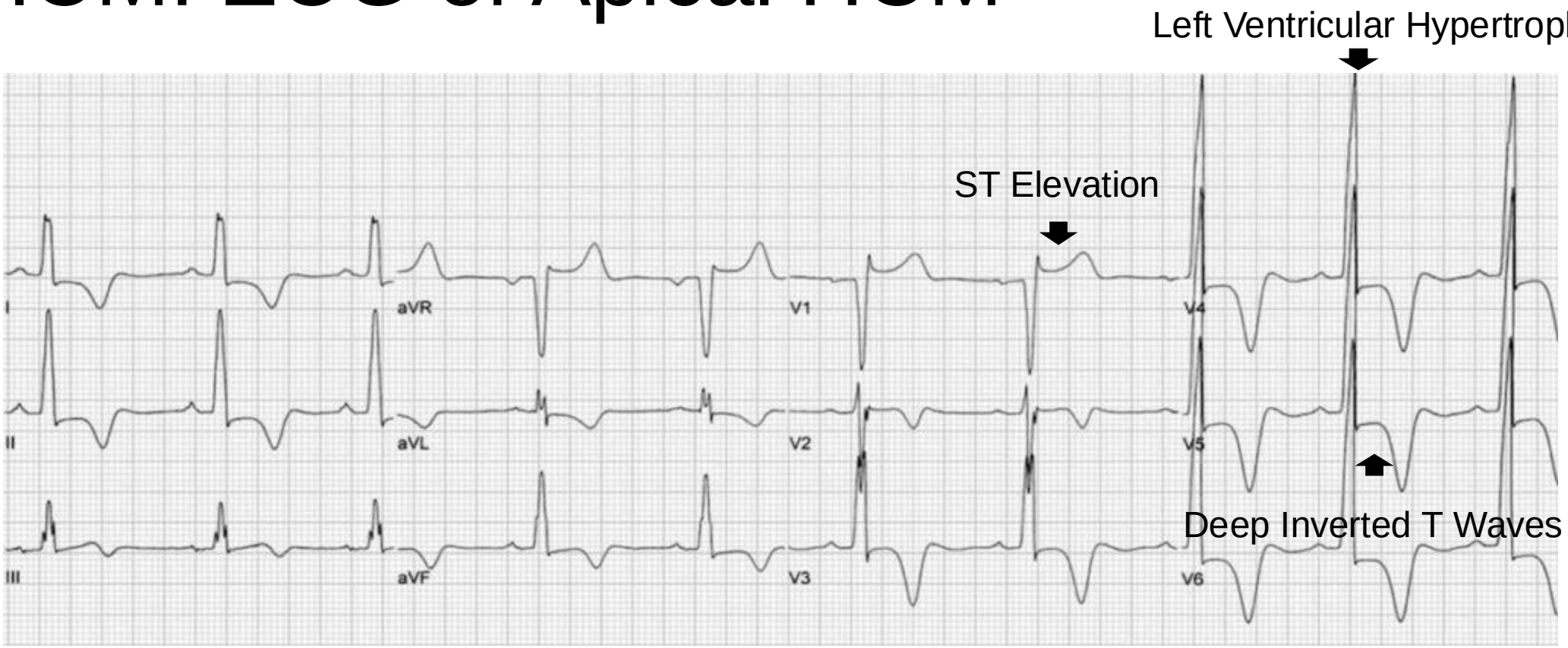
- Autosomal Dominant
  - Genetic Disease of the Sarcomere
  - Many patients are first in their family (new mutations)
- Asymmetric Hypertrophy
- Younger patients
  - 4-6^{th} decade of life in southern Africa

HCM: ECG



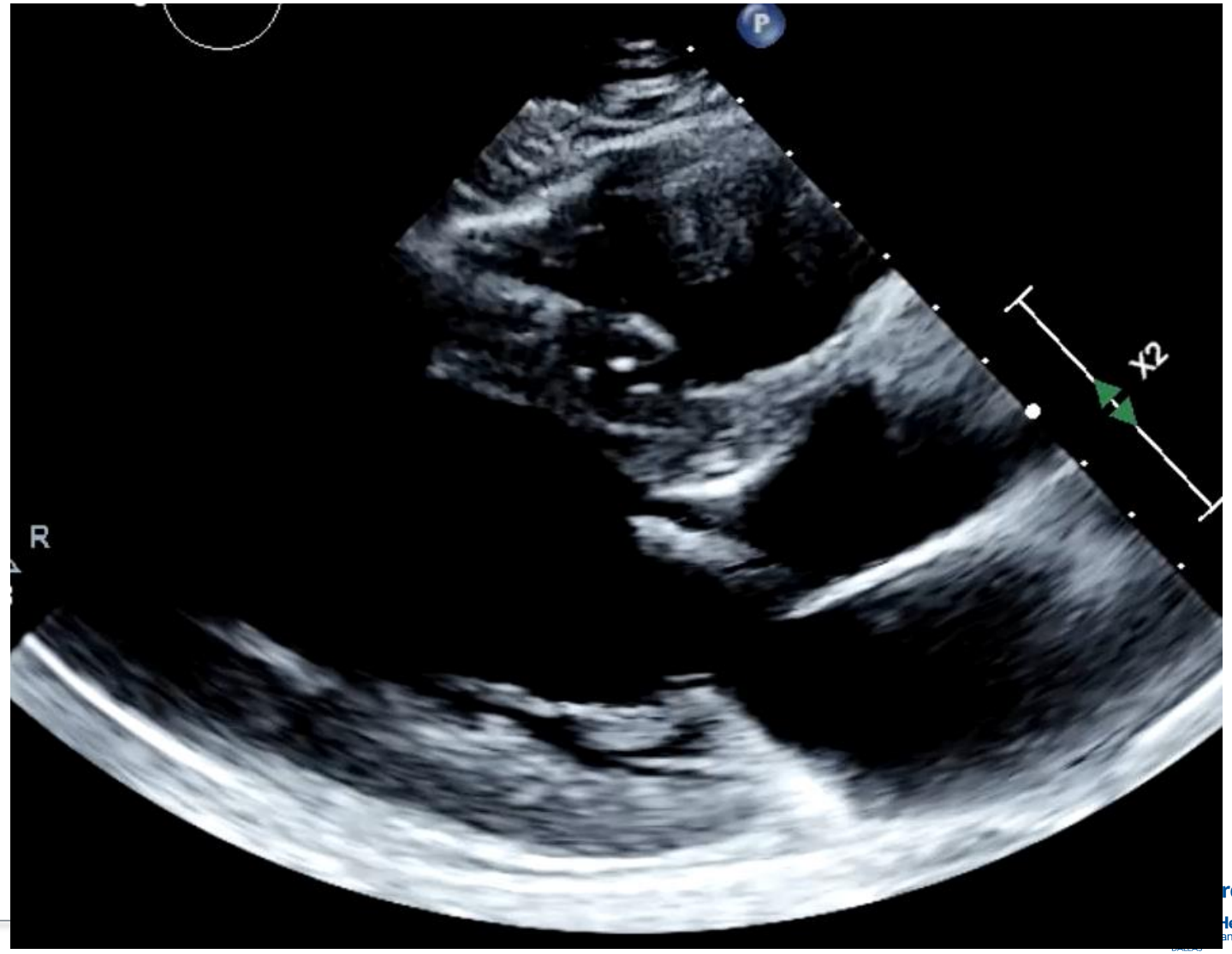
https://litfl.com/hypertrophic-cardiomyopathy-hcm-ecg-library/

HCM: ECG of Apical HCM

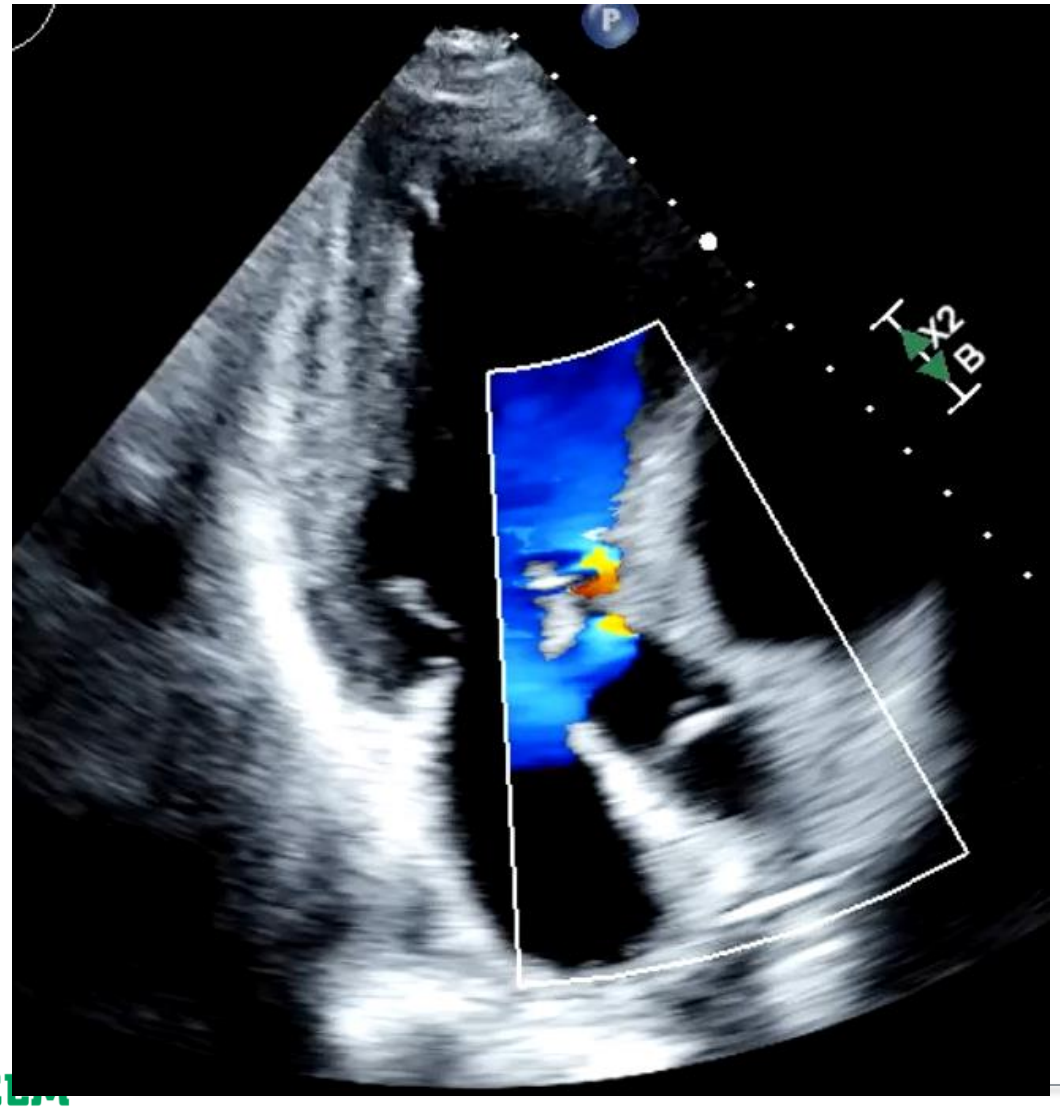


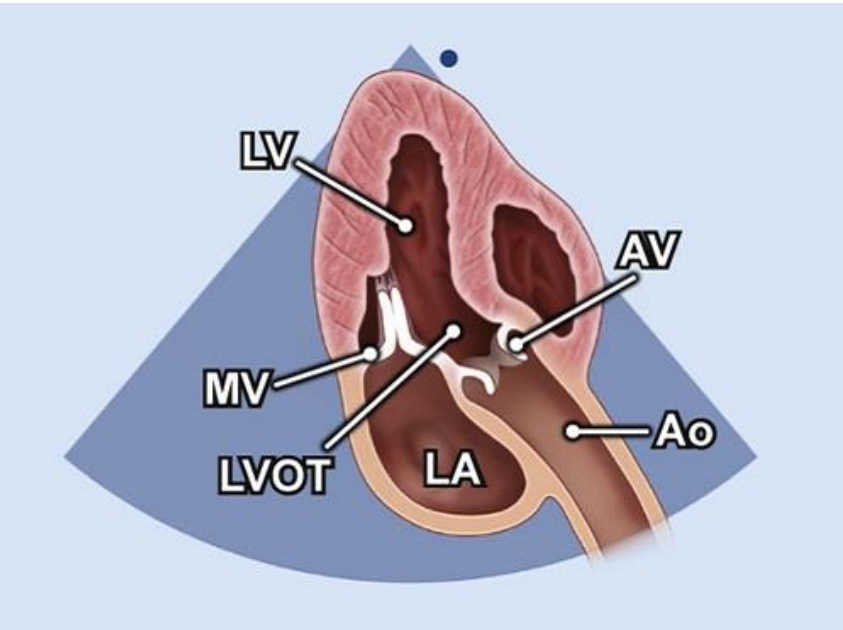
https://litfl.com/hypertrophic-cardiomyopathy-hcm-ecg-library/

HCM: Echocardiography

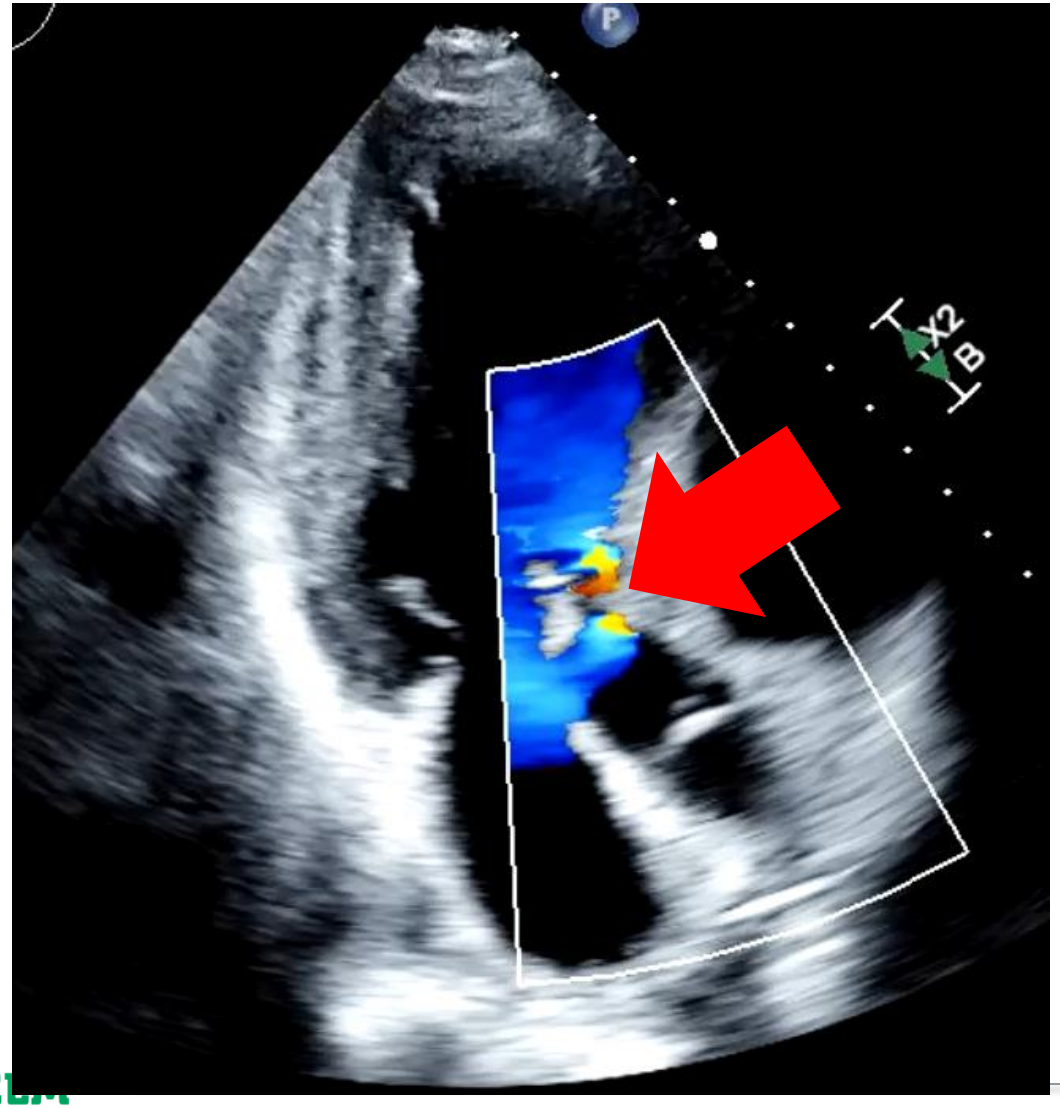


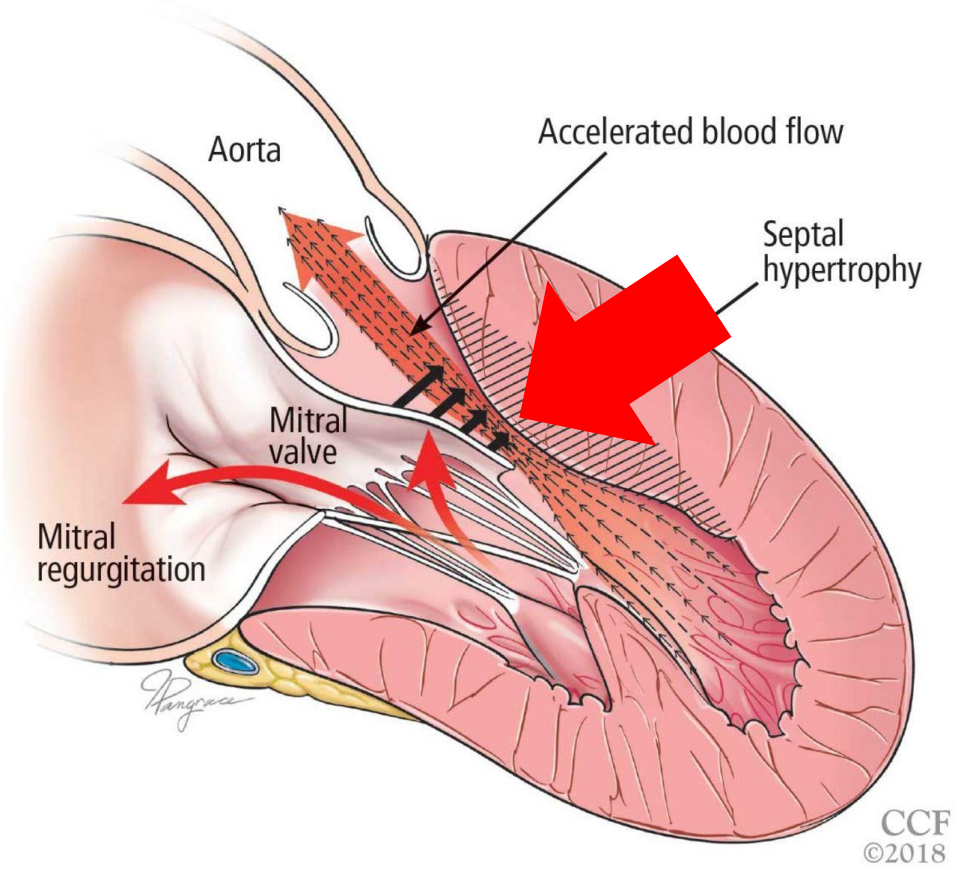
HCM: Echocardiography



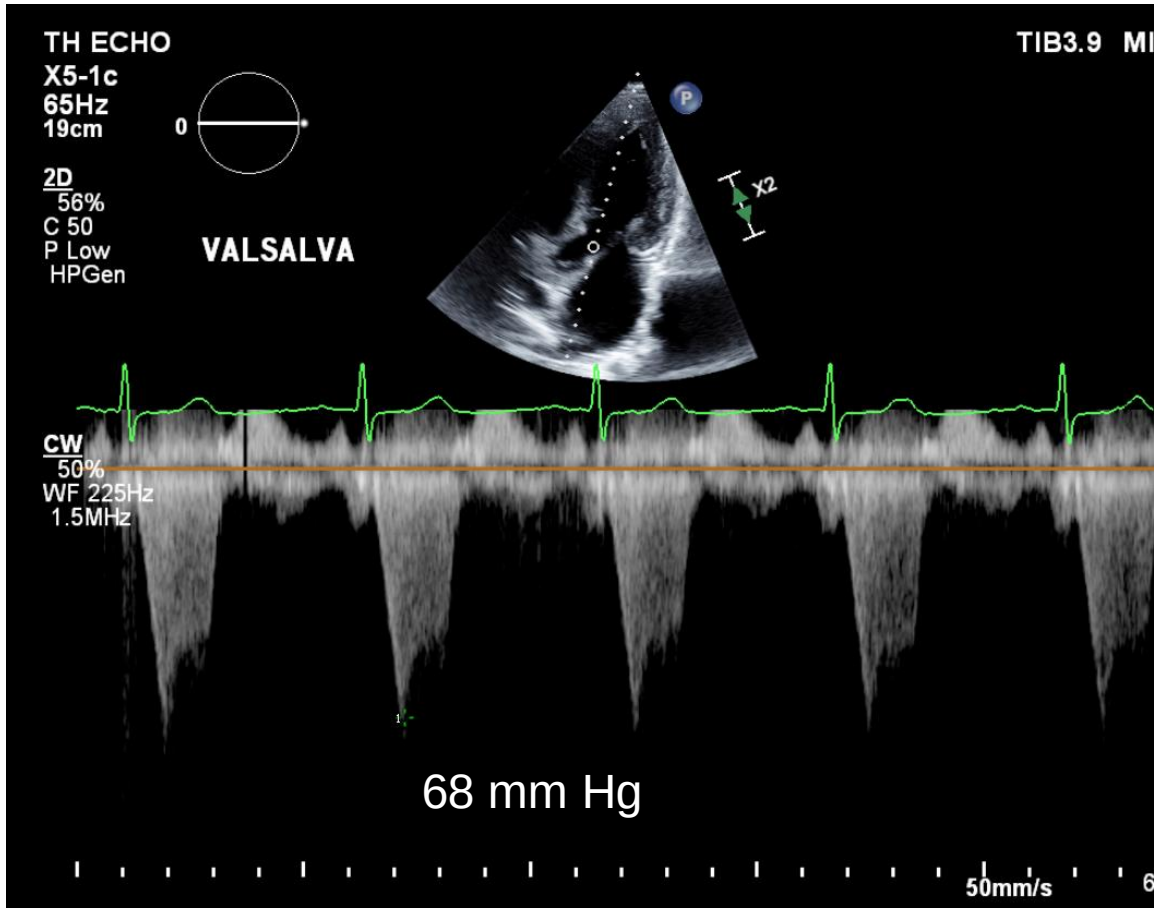


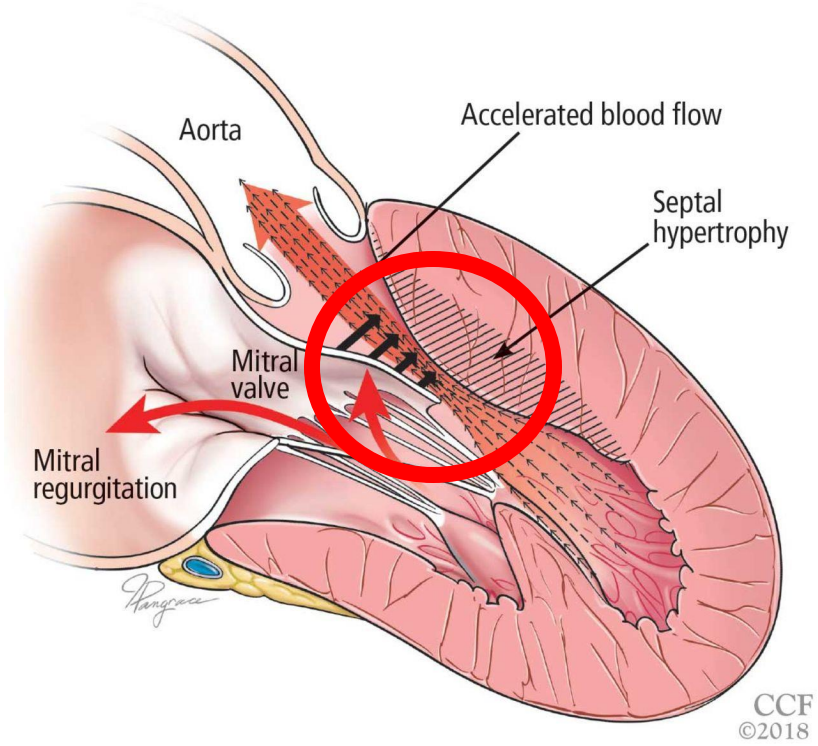
HCM: Echocardiography





HCM: Echocardiography





HCM: Management

Circulation

CLINICAL PRACTICE GUIDELINES

2024 AHA/ACC/AMSSM/HRS/PACES/SCMR Guideline for the Management of Hypertrophic Cardiomyopathy: A Report of the American Heart Association/American College of Cardiology Joint Committee on Clinical Practice Guidelines

Developed in Collaboration With and Endorsed by the American Medical Society for Sports Medicine, the Heart Rhythm Society, Pediatric & Congenital Electrophysiology Society, and the Society for Cardiovascular Magnetic Resonance

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HCM: Management

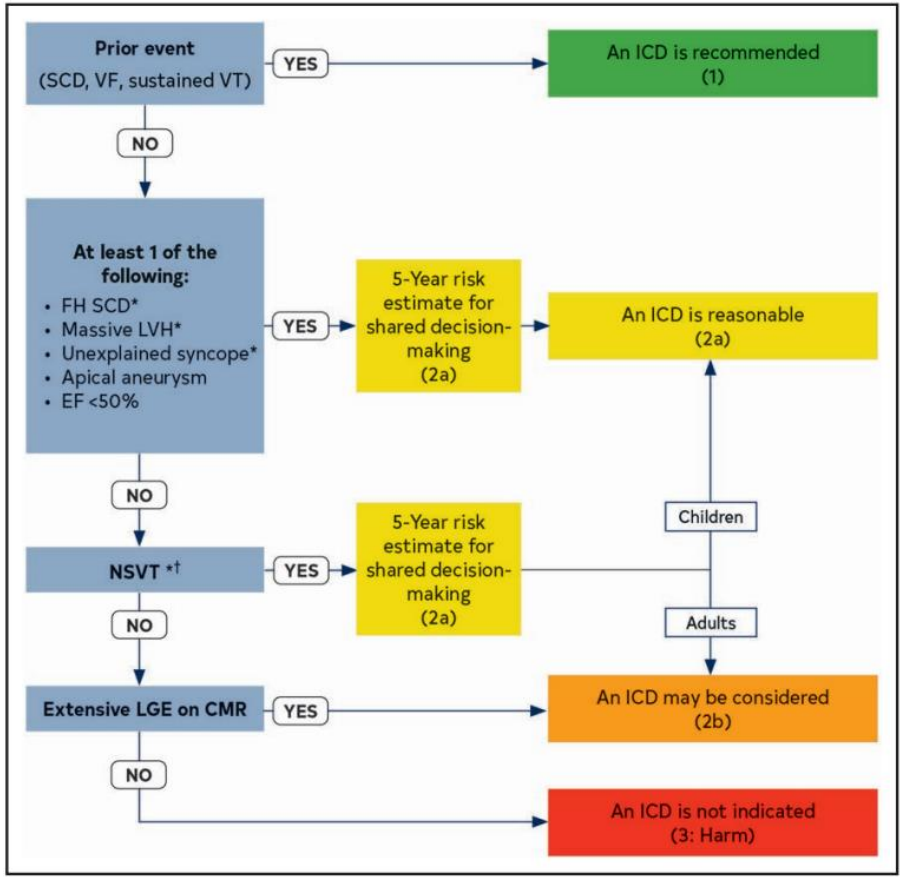
- Goal is to reduce obstruction
- Chronotropic Agents:
  - Beta blockers, Verapamil, Diltiazem
  - Lowers heart rate, increases EDV, decreases obstruction
- Septal Reduction Therapy
  - Surgical Myectomy
  - Alcohol Septal Ablation
  - Requires technical expertise
  - Patient access is a major problem

Recommendations for Pharmacological Management of Symptomatic Patients With Obstructive HCM
Referenced studies that support the recommendations are summarized in the Online Data Supplement.

COR	LOE	Recommendations
1	B-NR	1. In patients with obstructive HCM and symptoms* attributable to LVOTO, nonvasodilating beta blockers, titrated to effectiveness or maximally tolerated doses, are recommended.[1–3]
1	B-R	3. For patients with obstructive HCM who have persistent symptoms* attributable to LVOTO despite beta blockers or nondihydropyridine calcium channel blockers, adding a myosin inhibitor (adult patients only), or disopyramide (in combination with an atrioventricular nodal blocking agent), or SRT performed at experienced centers,§ is recommended.[7–14]

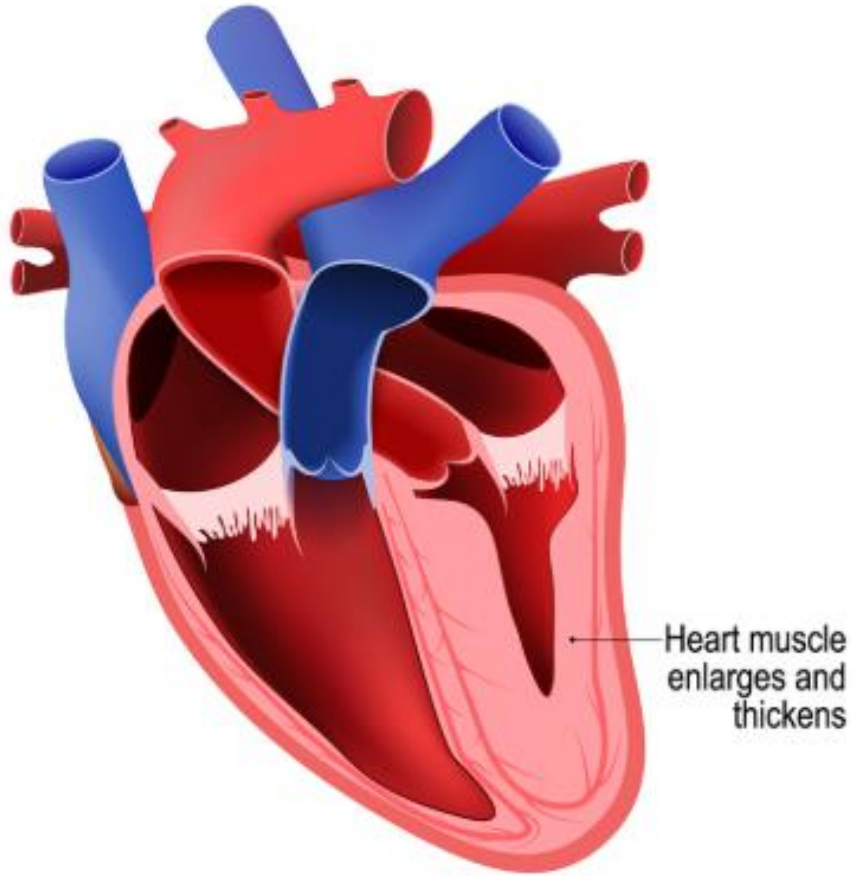
HCM: Risk of Sudden Cardiac Death

- Some patients with HCM are higher risk for sudden cardiac death
  - Ventricular Tachycardia and ventricular fibrillation
- No medical treatment
- ICD is indicated for high risk patients
- NOT for hypertensive heart disease with LVEF >35%



Hypertensive Heart Disease

- If LVEF >50%:
  - Treat blood pressure
  - Diuretics as needed
  - SGLT-2i
  - Exercise
- If LVEF <50%L
  - Treat with HFrEF GDMT

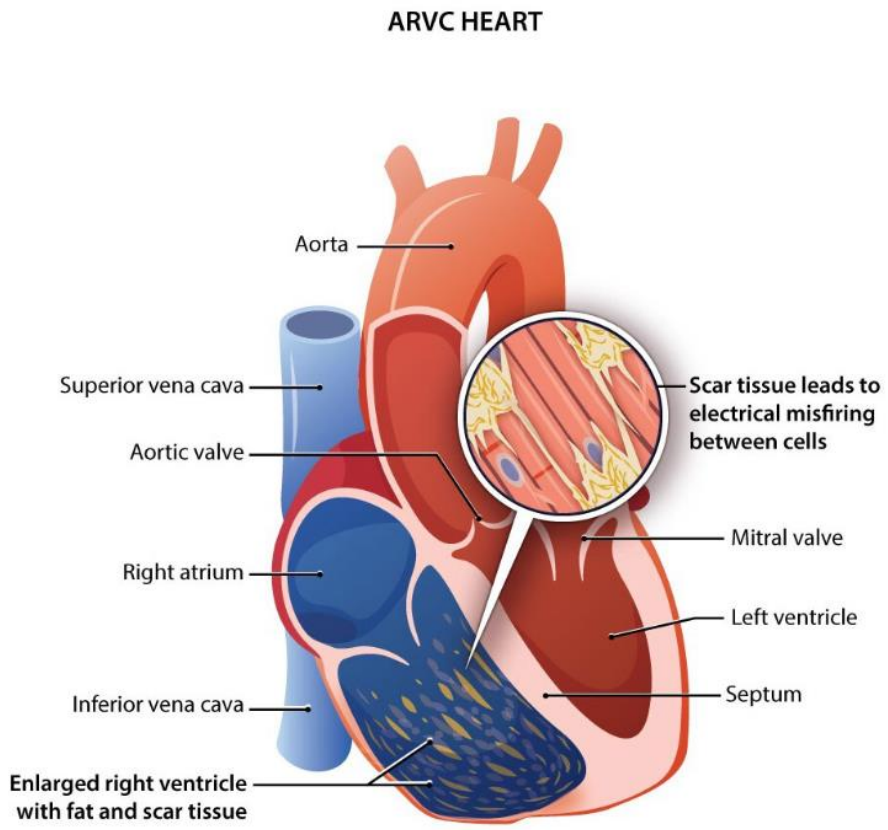


Quick Hitters

Arrhythmogenic, Restrictive, and Ischemic

ARCV: Arrhythmogenic Right Ventricular Cardiomyopathy

- 2nd most common CM in some parts of Africa
- Genetic condition
  - 25% PKP2
- Male > Female
- 3rd decade of life
- 5 year mortality is 10% in South African registry

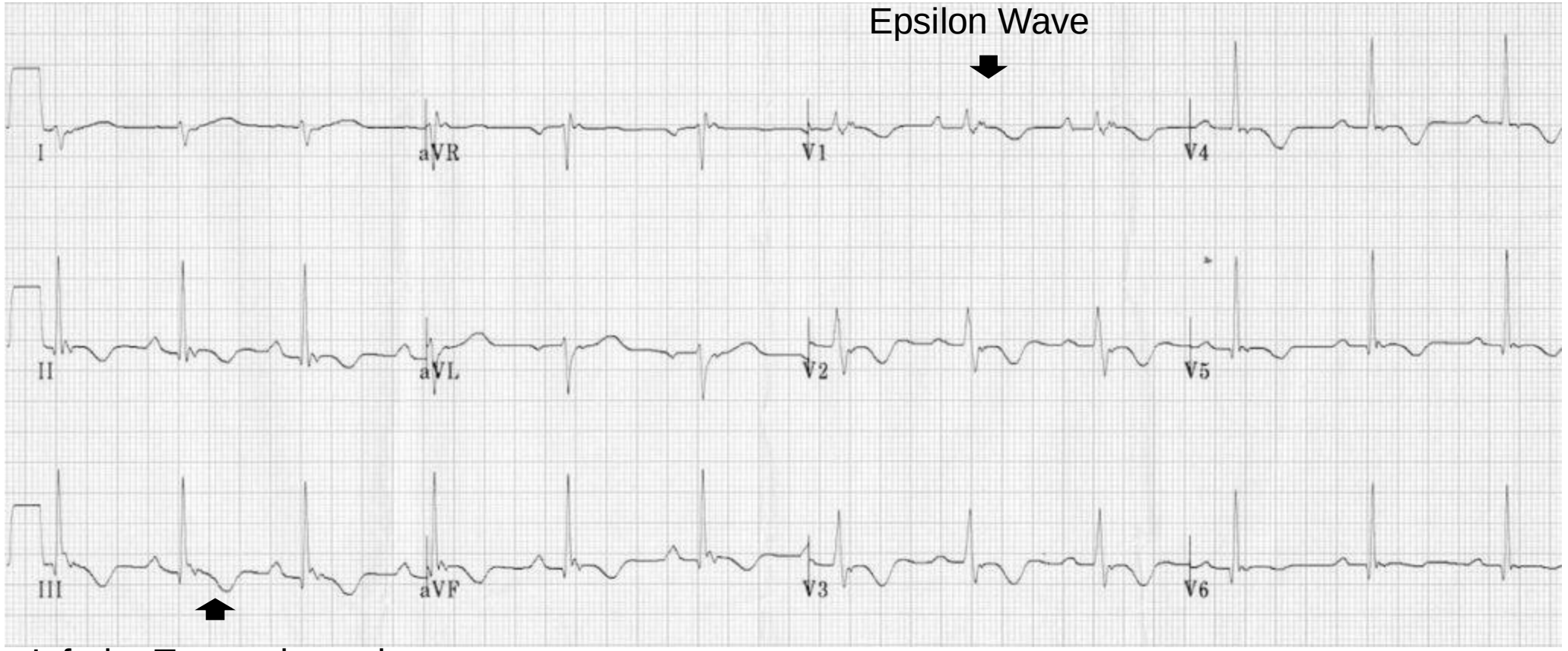


ARCV: Symptoms

- Asymptomatic in early stages
- Syncope
- Palpitations
- Shortness of breath
- Fatigue

- Diagnosed:
  - ECG
  - Echo
  - Genetic Testing
  - Family History

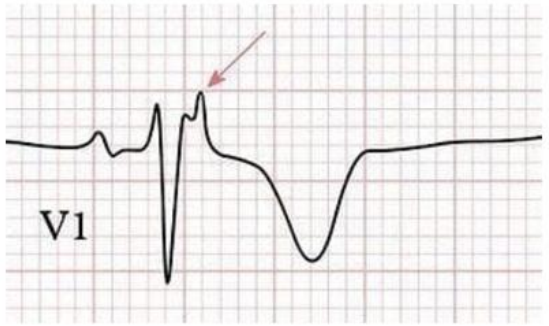
ARCV: ECG



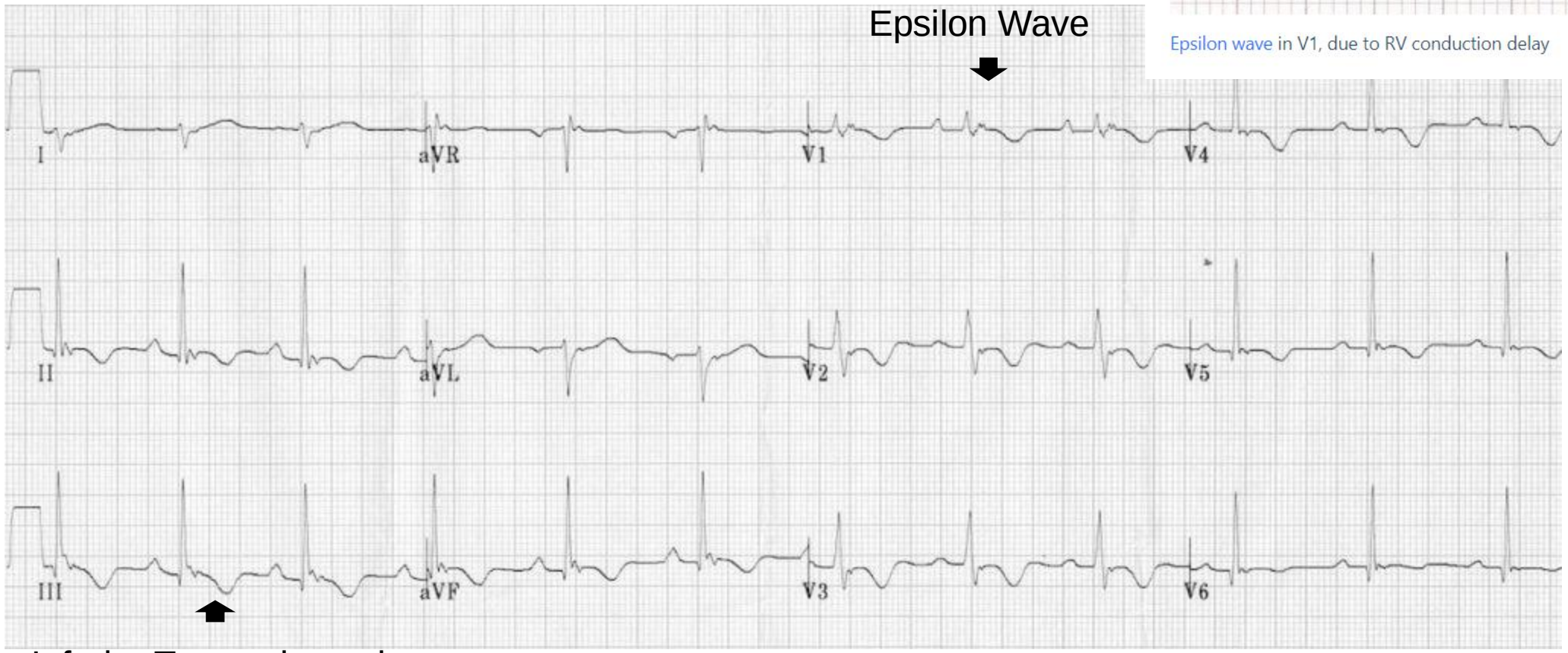
Inferior T wave inversion

https://litfl.com/arrhythmogenic-right-ventricular-dysplasia-arvd/

ARCV: ECG



Epsilon wave in V1, due to RV conduction delay



Inferior T wave inversion

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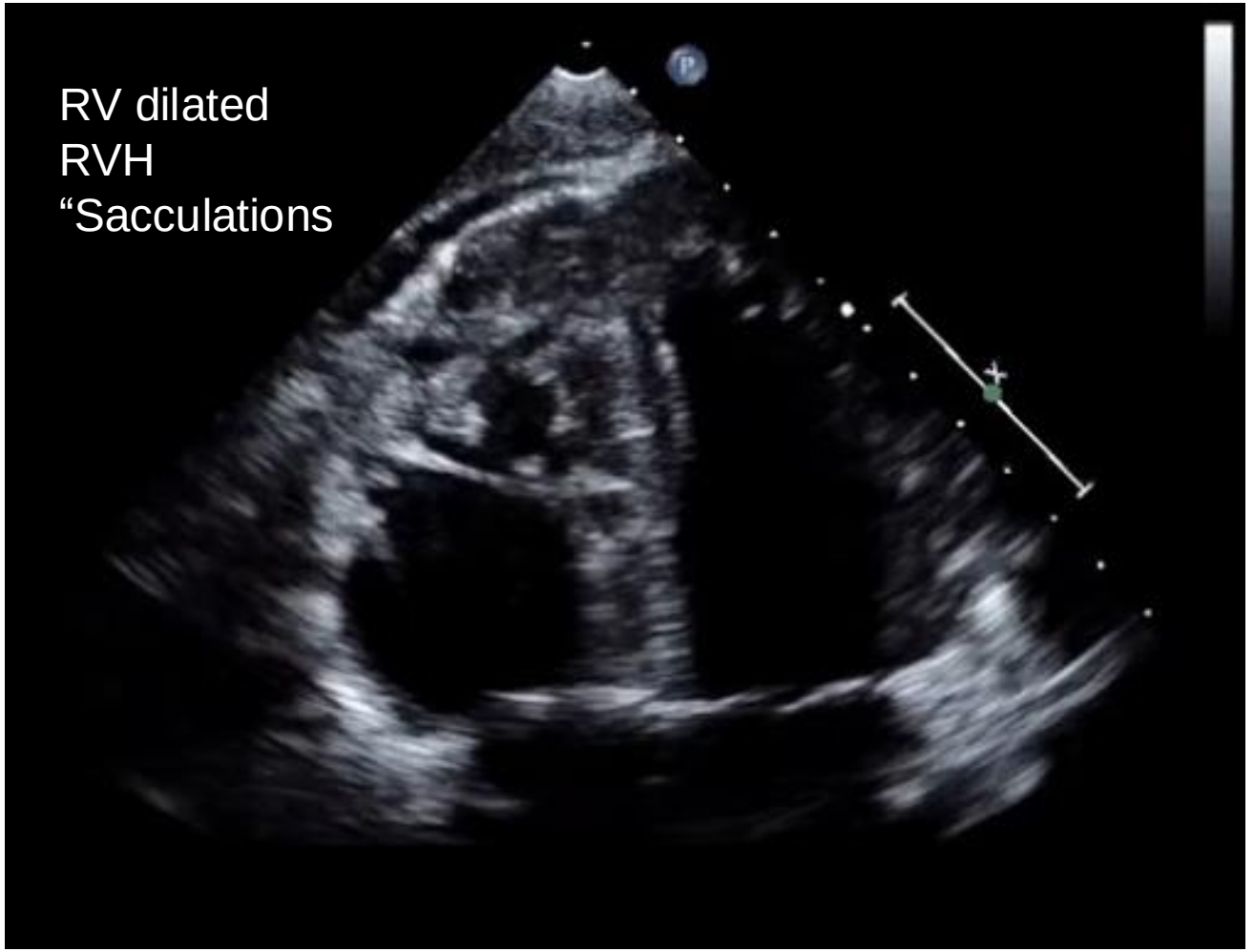
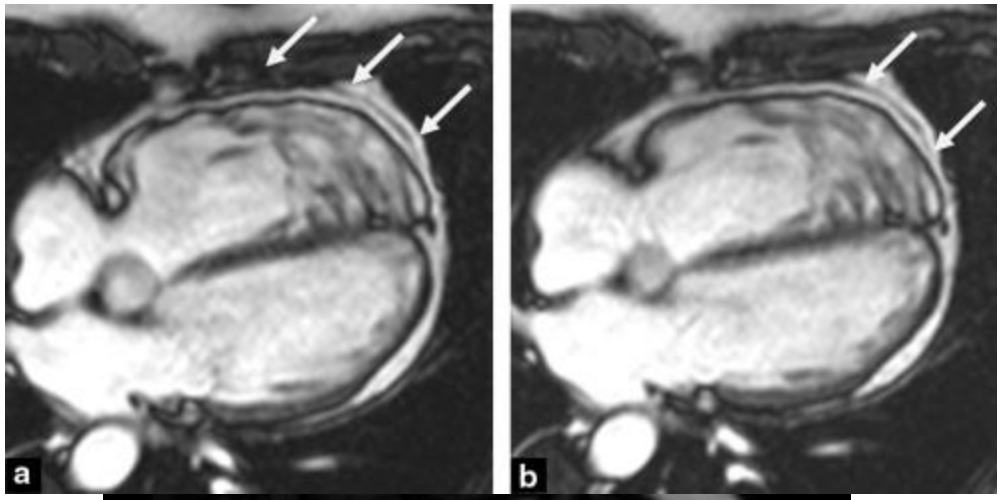
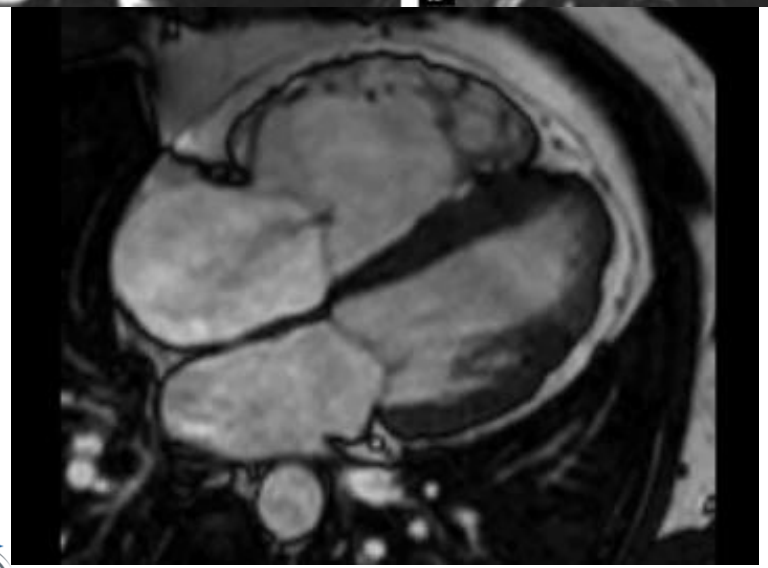
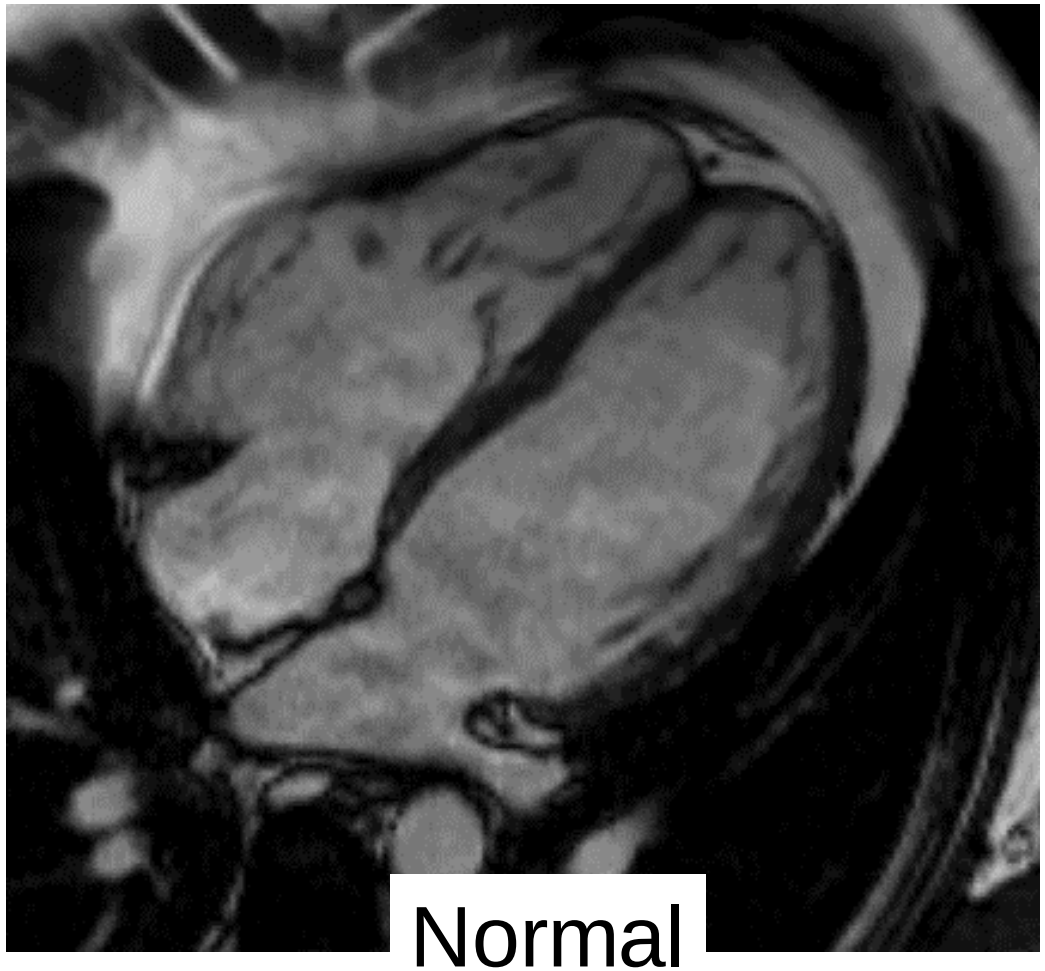
ARCV: Echocardiogram



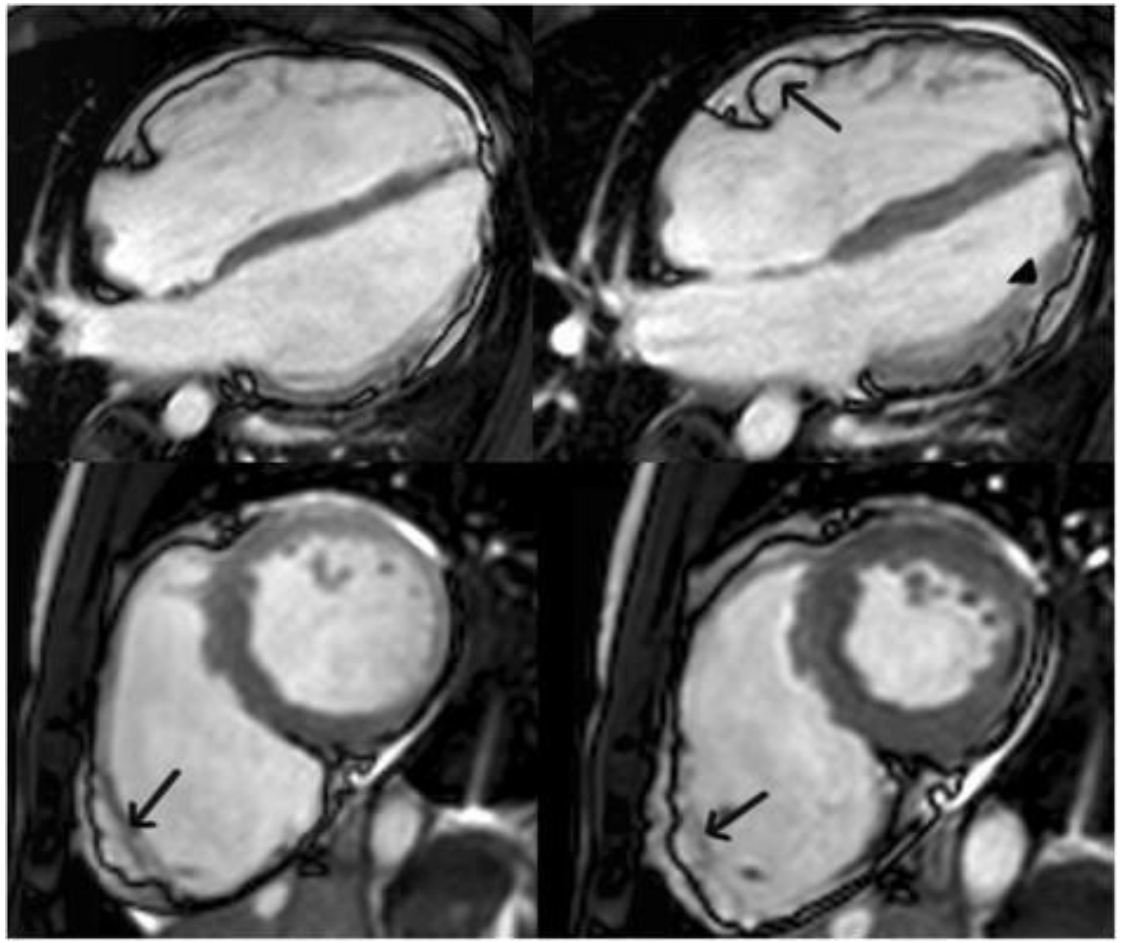
ARCV: MRI is best modality







ARCV: MRI is best modality



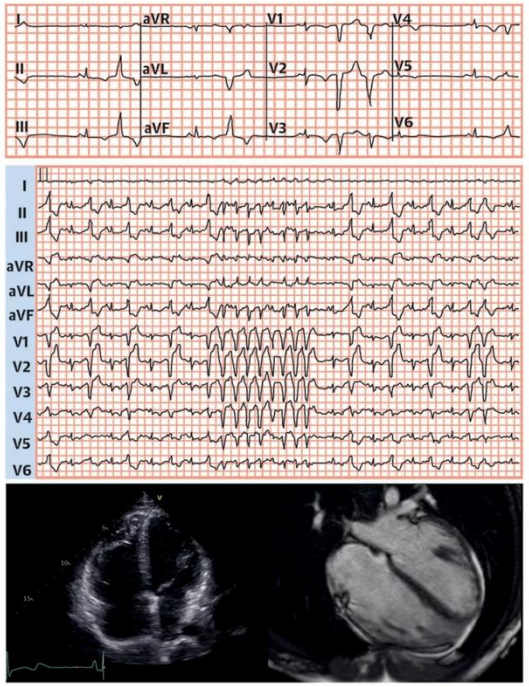
ARCV: Management

CENTRAL ILLUSTRATION: Clinical Approach to Arrhythmogenic Right Ventricular Cardiomyopathy

Assessment

Evaluation
- History and pedigree
- ECG, SAECG, Holter
- Echo, MRI
- Genetic testing

Diagnosis
- 2 major TFC
- 1 major + 2 minor TFC
- 4 minor TFC



Risk Stratification

Established Markers
- Cardiac arrest
- VT
- Age
- Male sex
- Cardiogenic syncope
- Extent of TWI
- PVC count
- Reduced LV/RV

Treatment

Conservative
- Exercise restriction

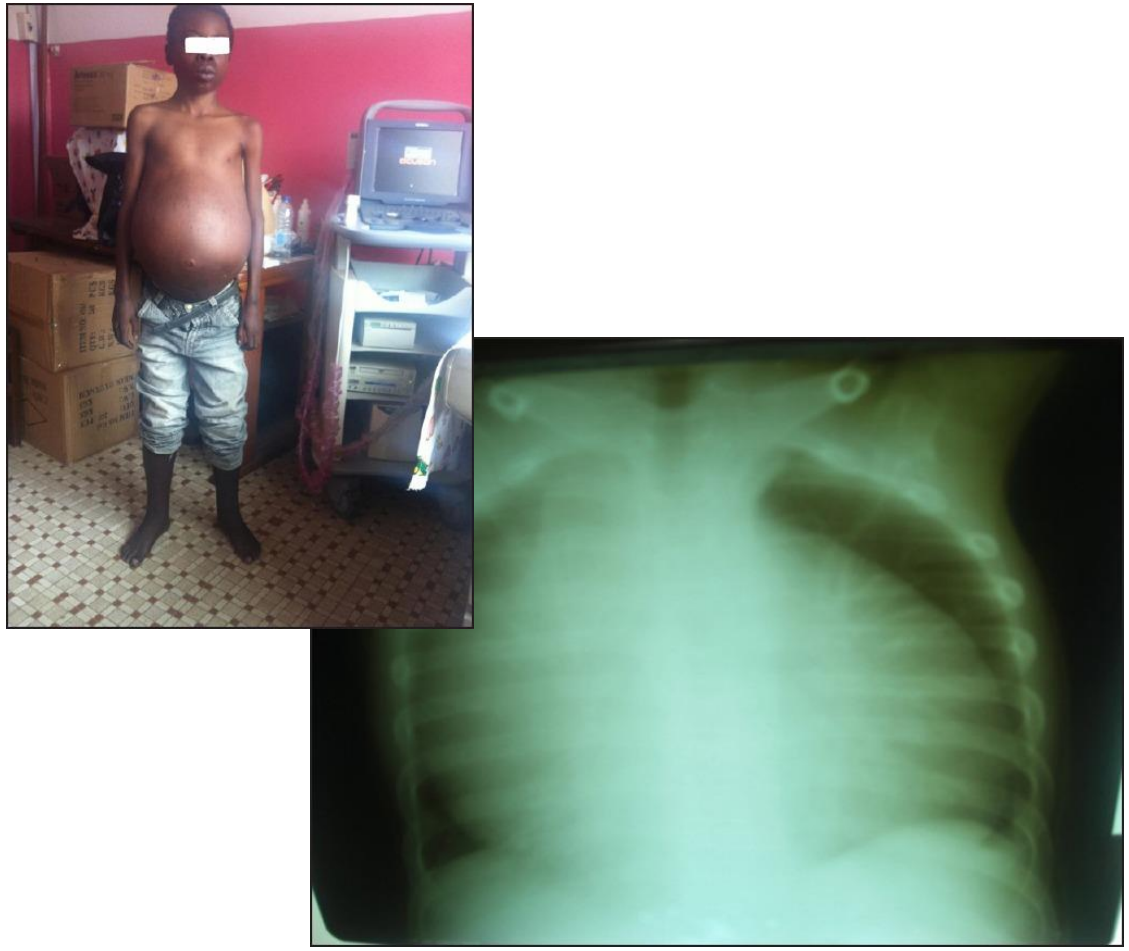
Pharmacological
- β-blockers
- Sotalol, flecainide, amiodarone

Interventional
- ICD
- Ablation

Krahn AD, et al. J Am Coll Cardiol EP. 2022;8(4):533-553.

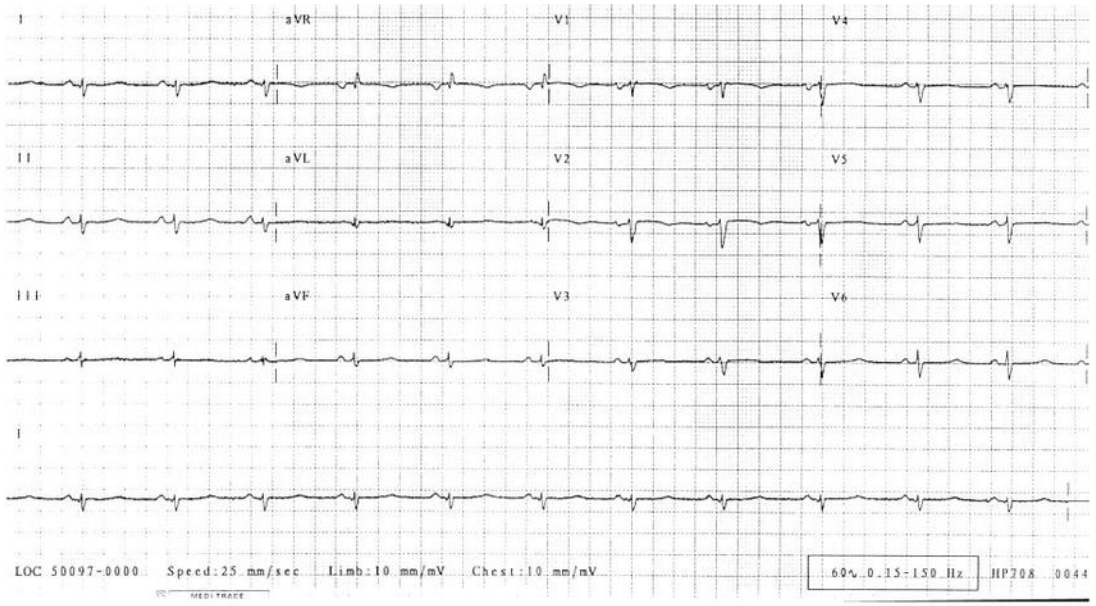
Restrictive Cardiomyopathies

- Endomyocardial fibrosis is very common
  - 2nd most common CM in central Africa (20%)
- Davie's Disease
- Response to treatment is poor
  - Mortality in high: 75% at 2 years
- Cause remains elusive



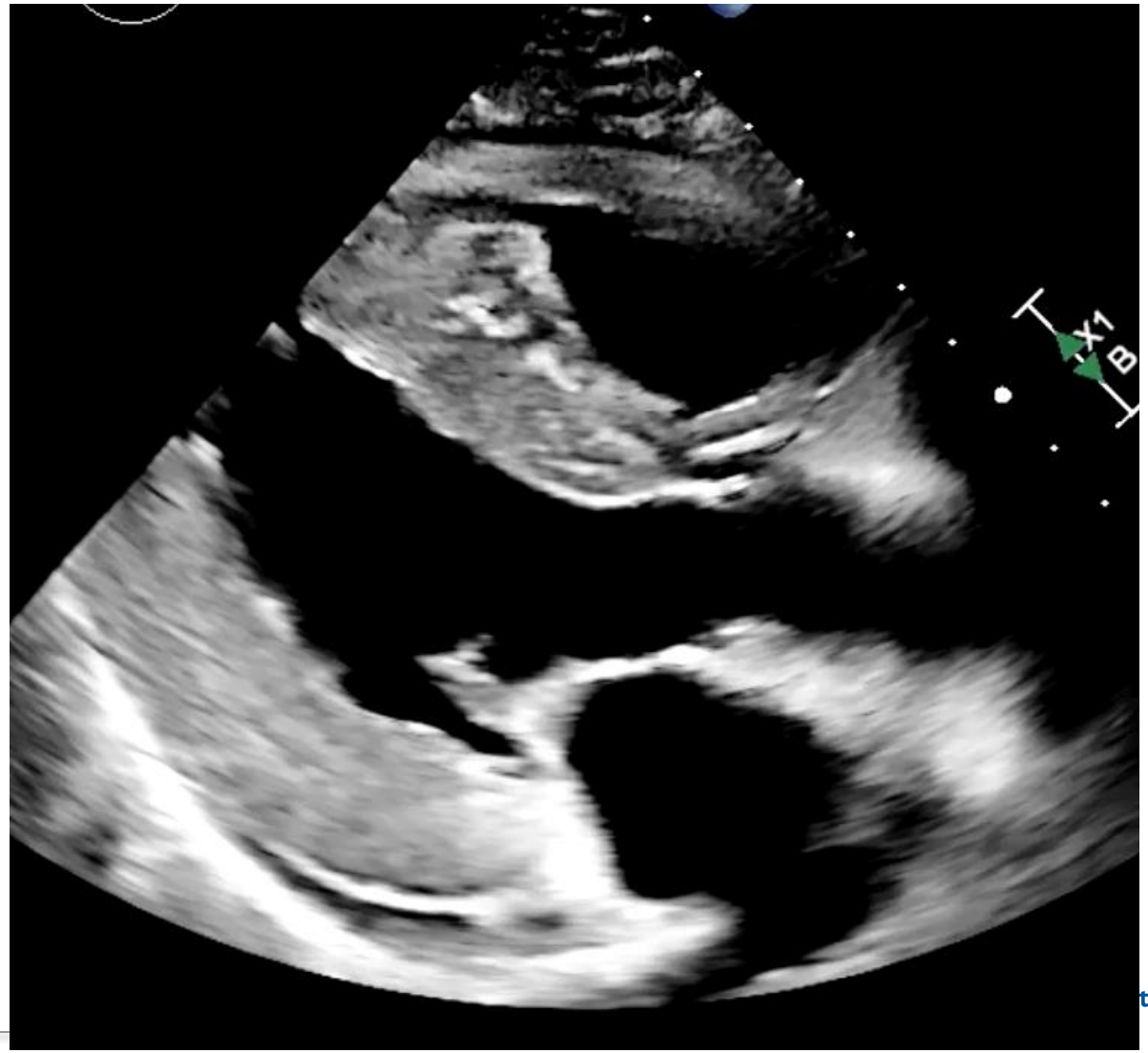
Infiltrative Cardiomyopathies: Amyloid

- Amyloidosis:
  - Deposition of protein in the myocardium
  - Restrictive cardiomyopathy
  - Can progress to dilated
- Hereditary Type:
  - aTTR is found in Americans of African descent
  - Prevalence in Africa is unknown



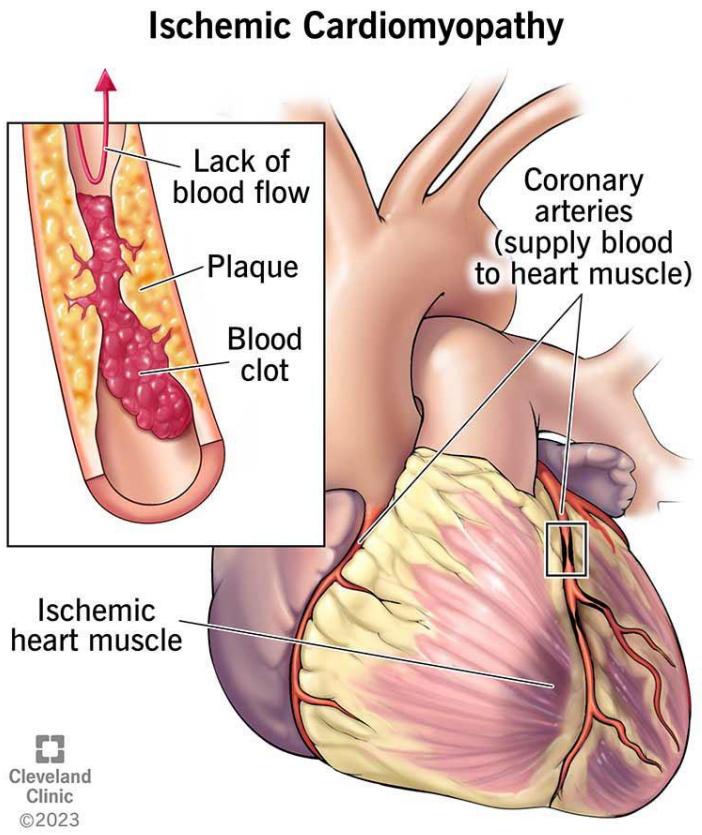
ECG with low voltage

Infiltrative Cardiomyopathies: Amyloid

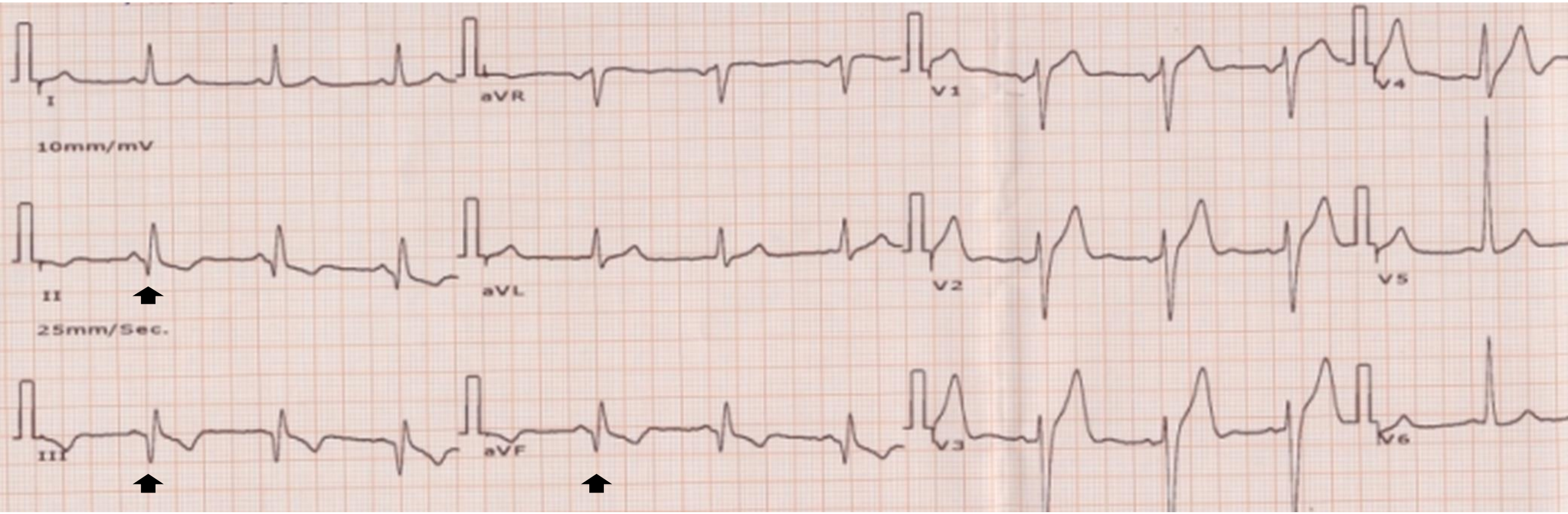


Ischemic Cardiomyopathy

- Sequalae of myocardial infarction or long-standing coronary artery disease
- Most common type in the United States
- Remains uncommon in Africa
  - Risk Factors are rising:
  - Diabetes, obesity, hypertension, smoking

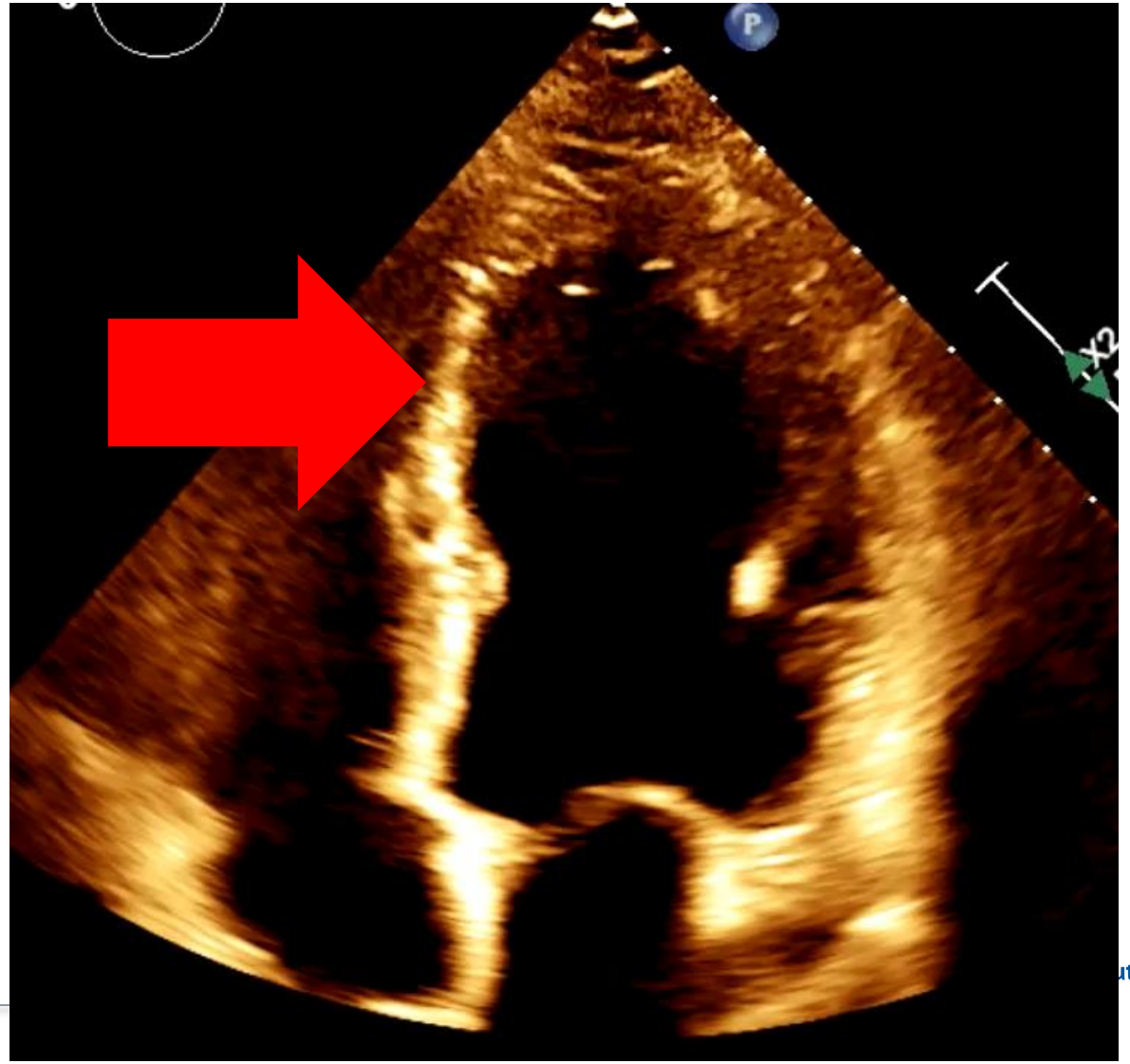


Ischemic Cardiomyopathy

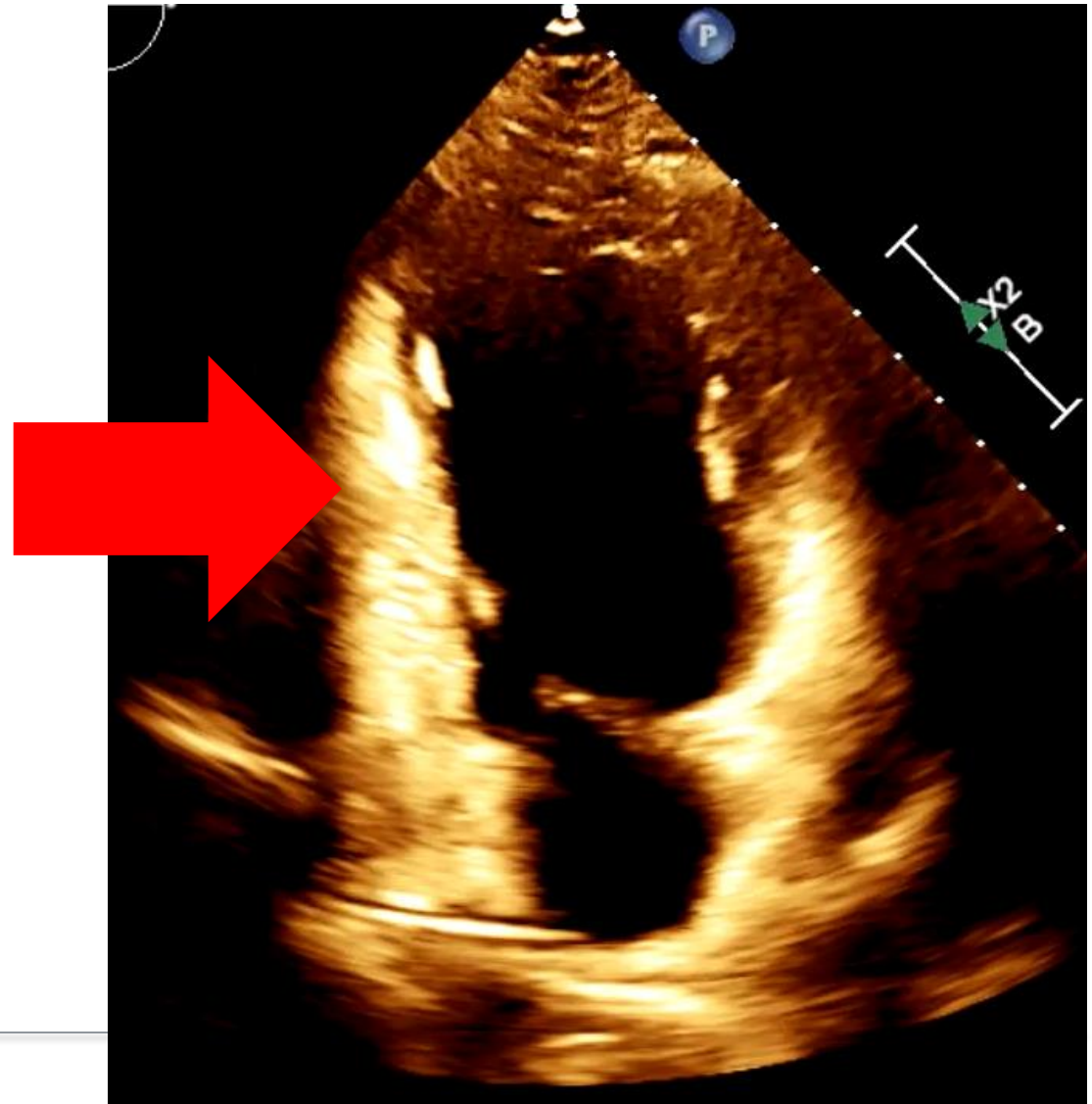


Inferior Q waves, old myocardial infarction

Ischemic Cardiomyopathy



Ischemic Cardiomyopathy



ICM: Management

- Treat with GDMT for HFrEF if LVEF is <50%
- Consider ICD or CRT if indicated
- In US, recommend coronary evaluation of all patients where ICM is possible
  - Coronary Angiogram or CT coronary angiogram

6. HF is often caused by coronary atherosclerosis,[79] and evaluation for ischemic heart disease can help in determining the presence of significant coronary artery disease (CAD). Noninvasive stress imaging with echocardiography or nuclear scintigraphy can be helpful in identifying patients likely to have obstructive CAD.[24,25] Invasive or computed tomography coronary angiography can detect and characterize extent of CAD.[26,27]

Conclusions

1. Cardiomyopathies are common and complex heart conditions in Africa
2. Physical exam, history and bedside tests are effective at diagnosing syndrome of heart failure
3. Cardiac Imaging is needed to characterize the cardiomyopathy
4. Etiologies of cardiomyopathies are diverse
5. Different cardiomyopathies have distinct treatment pathways