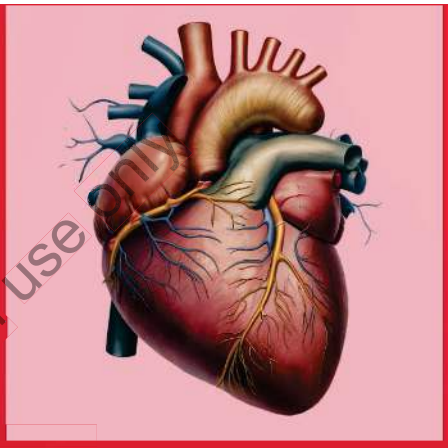
HEART SMARTS

The Cardiomyopathy Challenge



Learning Objectives

- Explain the physiology and types of cardiomyopathy on cardiovascular function
- Identify signs and symptoms of HCM, ATTR and Oncology treatment toxicity
- Discuss treatment and prevention strategies for worsening cardiomyopathy and heart failure

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QUICK 5 MIN REFRESHER

CARDIOMYOPATHY, AMYLOID HEART DISEASE, CANCER TREATMENT HEART DISEASE

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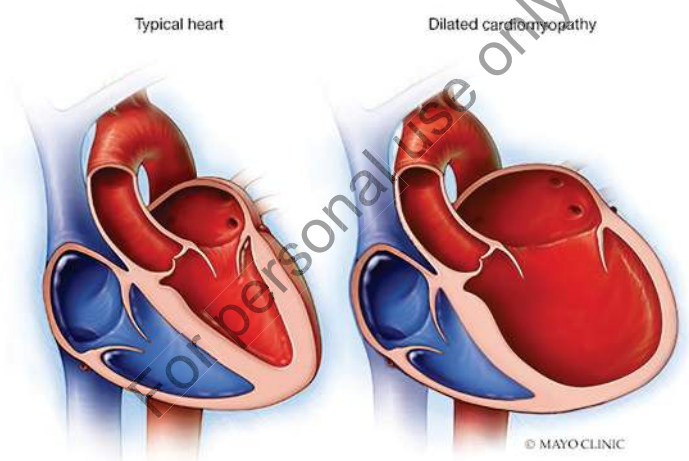
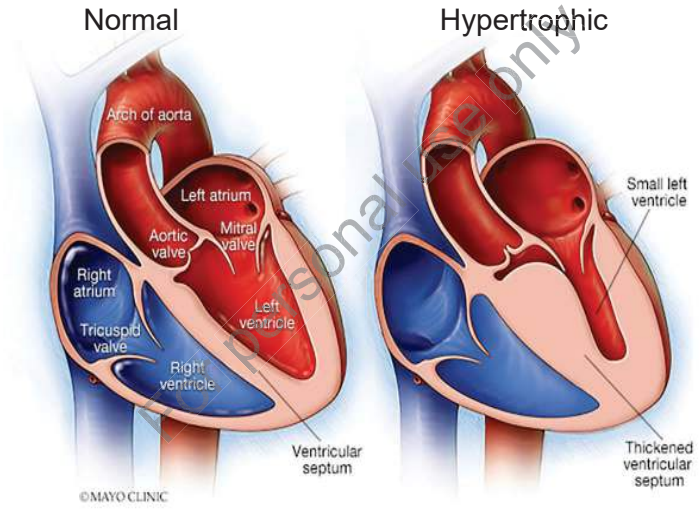
DILATED CARDIOMYOPATHY



HYPERTROPHIC CARDIOMYOPATHY



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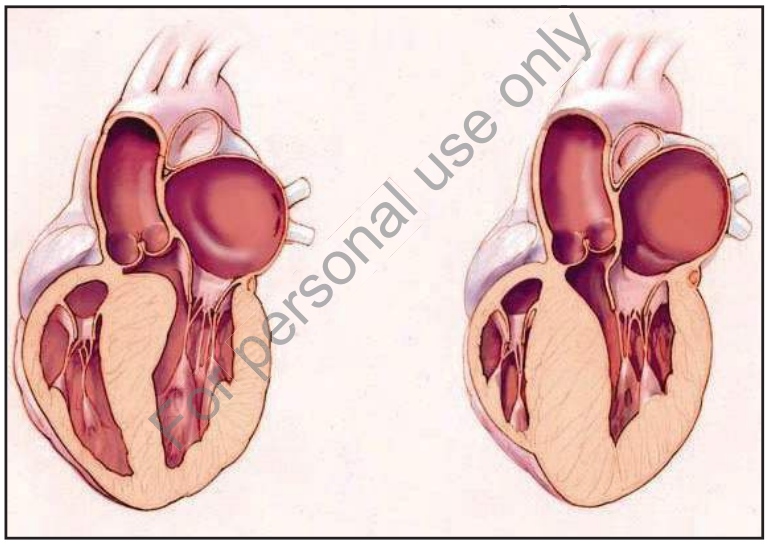
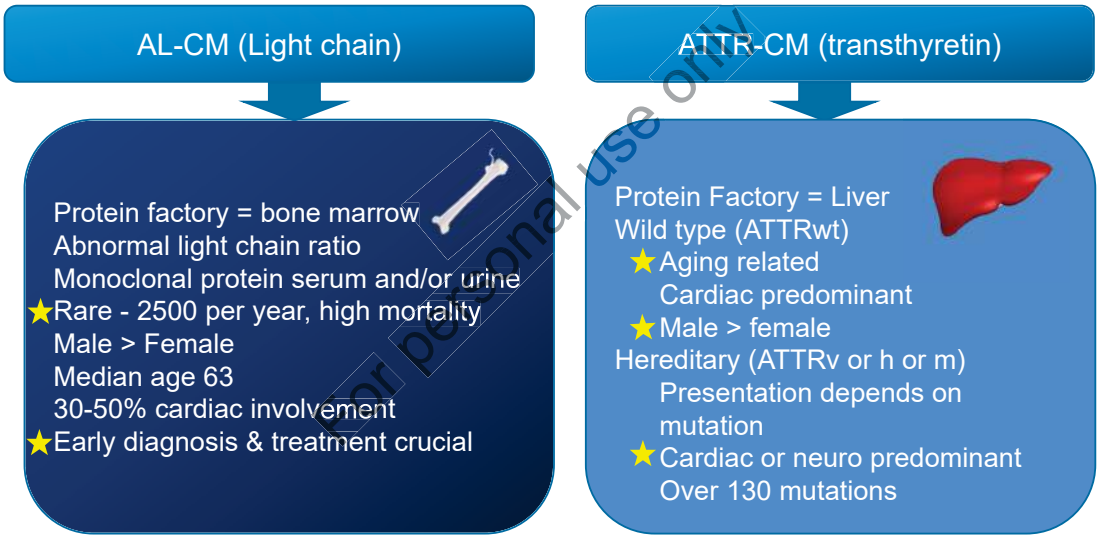


CARDIOMYOPATHIES

Dilated, hypertrophic, restrictive, toxin induced, amyloid

	Dilated	Hypertrophic	Restrictive
Pathophysiology	Fibrosis of myocardium and endocardium Dilated cardiomyopathy Thrombogenic	Hypertrophy of walls and septum (obstructed) Hypertrophy of walls except septum (non-obstructed)	Mimics constrictive pericarditis Restricted myocardial expansion & contraction Thrombogenic
Signs & symptoms	Fatigue, heart failure, weakness, pulm & systemic emboli Exam – S3, S4 cardiomegaly	Dyspnea, angina, fatigue, syncope, palpitations Exam – cardiomegaly, murmur that may inc w valsalva S4, SCD, HF	Dyspnea, fatigue activity intolerance Exam – S3, S4 heart block
Treatment	Diagnose & treat associated conditions CAD, HF, arrhythmias, toxins (radiation/chemo) Genetic testing, Advanced HF treatments, cardiac transplant	Symptomatic treatment, BB, CCB, myosin inhibitors, AAD, SCD risk assess, myectomy, ablation Genetic testing, ***dig, nitrates, vasodilators contraindicated in HOCM	Diagnose and treat associated conditions, HTN, arrhythmias, exercise recommendations
Surveillance	GDMT, clinical exam, surveillance, BP & symptom control important	Surveillance of symptoms, screening for SCD, screen first degree family members	Close follow up, GDMT, Monitor BP and assess symptoms, pulm edema

TYPES OF CARDIAC AMYLOID



WHEN TO SUSPECT?
DIAGNOSTICS SUGGESTIVE OF AMYLOID

Echocardiogram	Lab	EKG	Cardiac MRI
Concentric LV thickness ≥ 12 mm	Persistent mild elevation of troponin	Atrioventricular block	Marked extracellular volume expansion
EF normal or low		Low voltage	Abnormal nulling
LV not dilated			Diffuse late gadolinium enhancement
Increased RV thickness			
Strain < -18% (apical sparing pattern)			
Pericardial effusion			

Courtesy Janell Frantz Grazzini NP, Mayo Clinic

What about cardiomyopathies associated with Chemo/Radiation?

Cardiac surveillance following anthracycline-based chemo

Screen before and throughout treatment, followed by a posttreatment echo 6 to 12 months in the presence of 1 or more CV risk factors:

- Cumulative doxorubicin dose of ≥250 mg/m^2 (lifetime)
- Smoking, HTN, HLD, DM, Obesity during or after completion of therapy
- Age (≥65 years) at treatment
- Baseline (EF) <54% or history of CVD

•Cardiology rec if: post tx EF <53%, EF reduc of >10% pre/post tx, abn strain, cardiac symptoms

•Patients with signs and/or symptoms of cardiovascular disease should have an echocardiogram regardless of history of receipt of anthracycline, and should be referred to cardiology

Cardiac surveillance recs post mediastinal radiotherapy

•Stress echocardiogram 10 years after completion of radiotherapy due to the risk of late onset coronary artery disease

ATTRWT AMYLOID CLINICAL CONSIDERATIONS

- Males > 60 yrs (91% male)
- HFpEF in absence of HTN
- Bilateral carpal tunnel
- Newly dx HCM over age 60
- Low flow aortic stenosis
- Atrial fibrillation
- Angina despite normal coronary angiogram
- Repeated episodes embolic strokes
- Low QRS voltage or pseudo infarct on EKG
- Spinal stenosis
- Bicep tendon rupture
- Right sided HF
- Intractable pleural effusions
- Pericardial effusion
- Orthostatic hypotension
- Intolerant to standard HF meds
- Symptomatic hypotension in previous HTN
- PPM for AV block or bradycardia
- Any heart block

ESC HF, Vol 6, Issue 6, 1128-1139. 2019

AL CARDIAC AMYLOID CLINICAL FEATURES/RED FLAGS

- Heart failure
  - Cardiac involvement single most adverse prognostic factor
- Multi-organ involvement common
- Fatigue
- Weight loss +/- diarrhea
- Hepatomegaly
- Nephrotic syndrome
- Peripheral neuropathy
- Autonomic neuropathy


Periorbital purpura


macroglossia

Hamed, R et al. Blood Cancer Journal 11, 97 (2021)

Family Screening and Genetic Testing for Genetic Cardiomyopathy

Importance of Family Screening:

- **Risk Assessment:** Identifies at-risk family members who may inherit genetic cardiomyopathy.
- **Early Detection:** Facilitates early diagnosis and management, improving outcomes for affected individuals.
- **Informed Decision-Making:** Empowers families with knowledge about their genetic risks and options.

Genetic Testing Overview:

- **Types of Tests:**
  - **Single Gene Testing:** Focuses on specific genes known to be associated with cardiomyopathy (e.g., MYH7, MYBPC3).
  - **Panel Testing:** Analyzes multiple genes simultaneously to identify variants linked to cardiomyopathy.
  - **Whole Exome/Genome Sequencing:** Comprehensive analysis of all coding regions or entire genome for rare or unknown mutations.

Process of Genetic Testing:

1. **Referral to Genetic Counselor:** Essential for discussing testing options, implications, and emotional support.
2. **Informed Consent:** Patients and families must understand the purpose, risks, and benefits of testing.
3. **Sample Collection:** Blood or saliva samples are collected for analysis.
4. **Result Interpretation:** Genetic counselors and healthcare providers explain results and implications for family members.

Considerations for Nursing Care:

- **Education:** Provide information about the benefits and limitations of genetic testing.
- **Support:** Offer emotional support to families navigating the testing process.
- **Follow-Up:** Assist in coordinating care for further evaluation or management based on test results.

Resources

GCAC[Cardiomyopathy Awareness 2023 - Global Heart Hub]

Novel Therapies for Genetic Cardiomyopathy

Overview of Emerging Treatments:

1. **Cardiac Myosin Inhibitors:**
   - **Mechanism:** These agents target the cardiac myosin protein, reducing its interaction with actin, which can help decrease cardiac contractility and improve heart function in patients with hypertrophic cardiomyopathy (HCM).
   - **Examples:**
     - **Mavacamten:** Approved for HCM, it has shown to improve symptoms and exercise capacity.
     - **Aficamten:** Currently in clinical trials, showing promise in managing HCM.
2. **Gene Therapy:**
   - **Approach:** Aims to correct or replace defective genes responsible for cardiomyopathy. This can potentially halt disease progression or reverse cardiac dysfunction.
   - **Current Developments:**
     - **AAV-based therapies:** Utilizing adeno-associated viruses to deliver therapeutic genes directly to cardiac tissue.

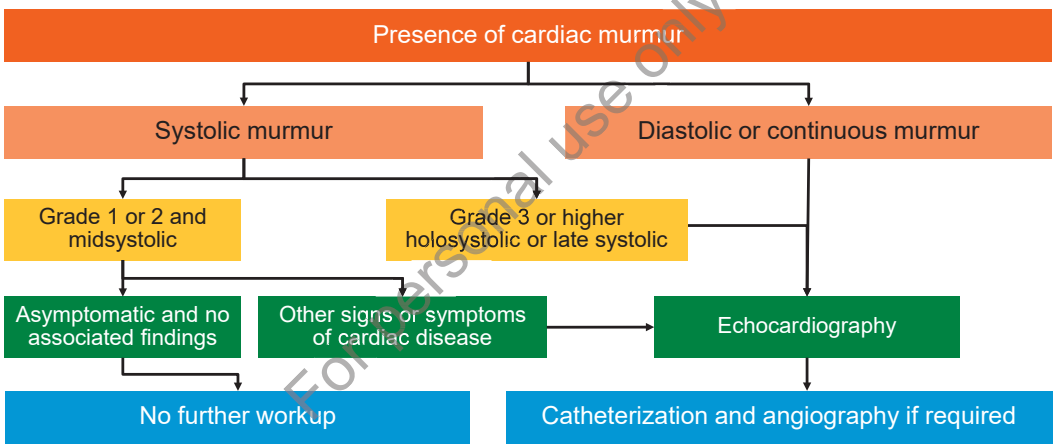
Implications for Nursing Care:

- Stay informed about these therapies to educate patients and families.
- Monitor for potential side effects and efficacy of new treatments.
- Support patients in understanding the implications of genetic testing and therapy options.

Resources

Gene Therapy in Cardiovascular Disease: Recent Advances and Future Directions in Science: A Science Advisory From the American Heart Association
Guide to Cardiovascular Genomics - Professional Heart Daily | American Heart Association
Lifelong Learning - Professional Heart Daily | American Heart Association

WHEN TO GET AN ECHO/WHEN NOT TO



Redrawn from: Bonow et al: JACC 32:1486, 1998

RESOURCES

- 2020 ACC/AHA guideline for the management of patients with valvular heart disease: A report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. Journal of the American College of Cardiology, 2021
- Mayo Clinic policy library: Transfemoral transcatheter aortic valve replacement (tfTAVR) guideline Mayo Clinic
- Transcatheter aortic-valve replacement with a balloon-expandable valve in low-risk patients The New England Journal of Medicine, 2019
- 2014 AHA/ACC Guideline for the Management of Patients With Valvular Heart Disease: Executive Summary: A Report of the American College of Cardiology/American Heart
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Prevention and Monitoring of Cardiac Dysfunction in Survivors of Adult Cancers: American Society of Clinical Oncology Clinical Practice Guideline

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Publication: Journal of Clinical Oncology • Volume 35, Number 8 • https://doi.org/10.1200/JCO.2016.70.5400