MYOCARDIAL DISEASE

1. Restrictive CM causes delayed diastolic relaxation, decreased compliance, elevated filling pressures with nondilated ventricles.
2. Restrictive cardiomyopathy should be suspected with right-sided heart failure disproportionate to left-sided heart decompensation.
3. High BNP (>800 pg/mL) is highly sensitive but not specific for differentiating restrictive CM from constrictive pericarditis.
4. Low voltage on ECG + thick LV walls on echo, suggests amyloidosis (or other infiltrative process).
5. Abdominal fat pad, rectum, or gingiva biopsy can confirm diagnosis. Endomyocardial biopsy to prove definitive cardiac involvement.
6. Restrictive CM therapy focuses on lowering intracavitary diastolic pressures and controlling venous & systemic congestion and afib.
7. Bradycardia precipitates CHF & requires pacing in restrictive CM.
8. ICD should be considered in HCM with any major or > 1 minor risk factors. Absence of risk factors has 90% negative predictive value.
9. Myectomy or alcohol ablation should be considered with outflow obstruction, septum $\geq$ 18 mm, NYHA III - IV refractory to medication.
10. Strenuous exercise and competitive sports should be avoided in all patients with HCM.
11. Cardiac tumors are 20 x more likely to be mets or direct extension from noncardiac cause. Primary tumors are rare, 50% are myxomas.
12. Cardiac tumor symptoms include dyspnea, fatigue, chest pain, constitutional symptoms, embolic events, syncope, sudden death.
13. Pericardial tumors may cause tamponade or constriction.
    Endocardial tumors: valve dysfunction, emboli, cavity obliteration.
    Myocardial tumors: arrhythmias & conduction abnormalities.
14. Left atrial myxoma should be surgically removed even when the patient is asymptomatic to avoid systemic embolic events.
15. Myxomas may be recurrent; long-term surveillance is appropriate.

Risk Factors for Sudden Death in Patients with Hypertrophic Cardiomyopathy

Major Risk Factors
Prior cardiac arrest
Sustained ventricular tachycardia
Family history of sudden death (in a first-degree relative younger than 40 years)
Minor Risk Factors
Unexplained syncope (≥2 episodes within 1 year)
Left ventricular septal wall thickness >30 mm in diastole
Abnormal blood pressure on exercise stress testing (systolic decrease or increase of <20 mm Hg)
Nonsustained ventricular tachycardia
Left ventricular outflow obstruction
Microvascular disease
High-risk genetic defect

Characteristics of Selected Causes of Restrictive Cardiomyopathy

Type	Etiology	Notes
Noninfiltrative Myocardial Conditions		
Idiopathic	Unknown	Diagnosis of exclusion
Scleroderma	Patchy myocardial fibrosis often associated with contraction band necrosis	May result from recurrent vasospasm of small vessels
Infiltrative Myocardial Conditions		
Amyloidosis	Commonly associated with transthyretin gene mutation	Most common identifiable underlying cause of restrictive cardiomyopathy
Sarcoidosis	Noncaseating granulomas, inflammation, and fibrosis	Clinical manifestations uncommon but may include ventricular arrhythmias, conduction block, and sudden death
Hemochromatosis	Iron deposits may be associated with myocardial or endocardial fibrosis	Most often presents as dilated cardiomyopathy but may present as restrictive form
Myocardial Storage Conditions		
Fabry disease	X-linked deficiency of α-galactosidase causing accumulation of globotriaosylceramide	Has some features of hypertrophic cardiomyopathy
Endomyocardial Disorders		
Endomyocardial fibrosis	Unknown cause but may relate to nutritional deficiencies, eosinophilia, or genetics	Endocardial fibrosis of the right and left ventricular apices, occurring mainly in west and central Africa
Eosinophilic cardiomyopathy (Löffler endocarditis)	Hypereosinophilia, organ infiltration, and release of toxic mediators	Fibrosis of the endomyocardium
Toxic effect of anthracycline	Doxorubicin, daunorubicin, idarubicin, epirubicin, and mitoxantrone (an anthraquinone) are the most frequently implicated	Can cause dilated or restrictive disease; risk increases with concomitant irradiation
Radiation	Diffuse fibrosis in the interstitium of the myocardium	May occur years or decades after exposure

Treatment Approaches for Specific Causes of Restrictive Cardiomyopathy

Cause	Medical Therapy	Surgical Therapy
Amyloidosis		
Familial/mutant transthyretin	Stem cell transplant	Heart/liver transplant
AL (primary)	Chemotherapy	Heart transplant (controversial)
AA (secondary)	Specific for cause of inflammation or infection	
Granulomatous Disease		
Sarcoidosis	First-line therapy: corticosteroids Second line therapy: chloroquine, hydroxychloroquine, cyclosporine, methotrexate	
Hemochromatosis		
Hereditary	Phlebotomy	Heart transplant
Acquired	Iron chelation	
Endomyocardial Disease		
Endomyocardial fibrosis	Warfarin (for documented cardiac thrombus)	Endomyocardectomy (palliative)
Hypereosinophilic syndrome	First-line therapy: corticosteroids Second-line therapy: tyrosine kinase inhibitor, interferon, cyclosporine, chemotherapeutic drugs Warfarin (for documented cardiac thrombus)	Endomyocardectomy (palliative) Stem cell transplant (for treatment-resistant disease)
Storage Disease		
Fabry disease	α-Galactosidase A replacement	