NOTES
CARDIOMYOPATHY

GENERALLY, WHAT IS IT?

PATHOLOGY & CAUSES

- Broad term, describes any issue resulting from disease of myocardium
- *Primary cardiomyopathy:* issue develops of its own accord
- *Secondary cardiomyopathy:* issue develops as compensation for another underlying disease

RISK FACTORS

- Positive family history

COMPLICATIONS

- Heart failure, arrhythmias, sudden cardiac death

SIGNS & SYMPTOMS

- Can be asymptomatic
- Heart failure signs, symptoms
- Heart murmurs

DIAGNOSIS

DIAGNOSTIC IMAGING

- Chest X-ray
- Echocardiogram/cardiac MRI

OTHER DIAGNOSTICS

- ECG

TREATMENT

MEDICATIONS

- See individual diseases

SURGERY

- Implantable cardioverter-defibrillator (ICD)
- Heart transplant

OTHER INTERVENTIONS

- Lifestyle changes

DILATED CARDIOMYOPATHY

osms.it/dilated-cm

PATHOLOGY & CAUSES

- Dilation of all four chambers of heart
- Most common type of cardiomyopathy
  - New sarcomeres added in series, creates larger chambers with relatively weak walls, less muscle for contraction → low systolic function
  - Chambers stretch → valves stretch → blood regurgitates back into atria

CAUSES

- Primary dilated cardiomyopathy most often idiopathic

Genetic mutations/conditions

- Duchenne muscular dystrophy (DMD), hemochromatosis

Myocarditis

- Can progress from myocarditis to dilated cardiomyopathy

Infection

- *Coxsackievirus B:* leads to myocarditis, heart muscle inflammation
- *Chagas disease:* protozoal infection

Linked to alcoholism

- Alcohol, metabolites have direct toxic effect on heart muscle

Linked to certain drugs

- *Chemotherapy:* doxorubicin, daunorubicin
- Cocaine

Wet beriberi

- *Beriberi:* illness caused by thiamine (vitamin B1) deficiency
- *Wet beriberi:* affects heart; ↓ thiamine levels impair myocardium energy production

Peripartum cardiomyopathy

- Can develop in third trimester of pregnancy/weeks after delivery
  - Related to pregnancy-associated hypertension
  - Half of individuals recover following pregnancy

Sarcoidosis

- Growth of granulomas in heart → dilation

RISK FACTORS

- Alcoholism, past family history of diseases implicated in DMD

COMPLICATIONS

- Systolic heart failure
  - *Valve regurgitation:* as chambers stretch, so do valves
- *Arrhythmias:* stretching muscle irritates conduction system

MNEMONIC: ID BIG MAPS

Causes of Dilated Cardiomyopathy

Idiopathic
Drugs/Doxorubicin (and cocaine)
Beriberi (wet)
Infection
Genetic
Myocarditis
Alcoholism
Peripartum cardiomyopathy
Sarcoidosis

SIGNS & SYMPTOMS

- Fatigue, dyspnea
- Lateral displaced point of maximum impulse (PMI)
- Chest pain on exertion
- Holosystolic murmur (mitral valve regurgitation during systole)
- S3 sound (blood rushing into, slamming into dilated ventricular wall during diastole)

DIAGNOSIS

DIAGNOSTIC IMAGING

X-ray

- Cardiomegaly, pulmonary edema, pleural effusion

OTHER DIAGNOSTICS

ECG

- Shows ventricular dilation, reduced ejection fraction

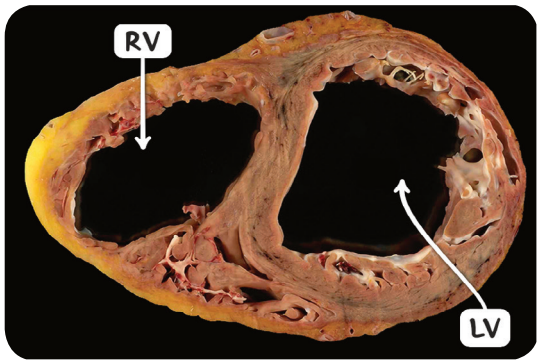

Figure 6.1 Gross pathology of dilated cardiomyopathy. Note the large ventricles and thin ventricular walls.

TREATMENT

MEDICATIONS

- Angiotensin-converting-enzyme (ACE) inhibitor, angiotensin receptor blocker, beta blocker
  - Slows disease progression

SURGERY

- Heart transplant (extreme cases)

OTHER INTERVENTIONS

- *Left ventricular assist device (LVAD):* mechanical pump assists heart in delivering blood to body

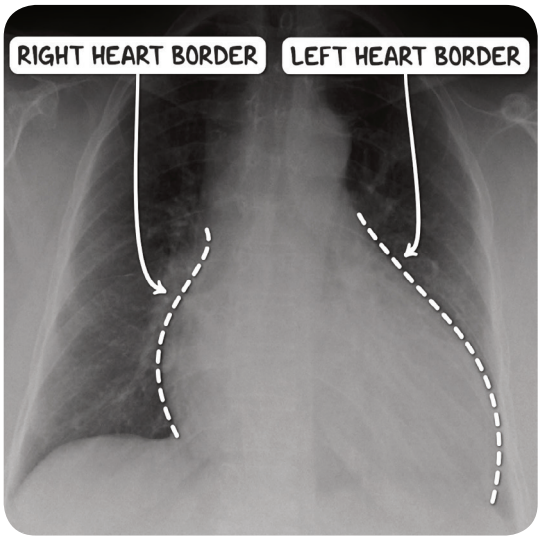

Figure 6.2 A chest radiograph demonstrating enlargement of the heart due to dilated cardiomyopathy. The heart occupies more than half the width of the chest.

HYPERTROPHIC CARDIOMYOPATHY (HCM)

osms.it/hypertrophic-cm

PATHOLOGY & CAUSES

- Myocardium becomes thick, heavy, hypercontractile
- Myocytes become disorganized, new sarcomeres added in parallel to existing ones
- Left ventricle most often affected
  - Muscle growth asymmetrical → interventricular septum grows larger relative to free wall
- Hypertrophy → walls taking up more space, ↓ blood fills ventricle
  - Walls become stiff, less compliant → less filling → low stroke volume → dysfunction in diastolic filling of left ventricle → diastolic heart failure
- *Arrhythmias:* larger muscles require more oxygen, coupled with heart having difficulty delivering blood to tissues → ischemia → arrhythmias

CAUSES

Genetic missense mutation, inherited as autosomal dominant trait

- Different genetic mutations affect different sarcomere proteins
- *Friedreich's ataxia:* autosomal recessive neurodegenerative disease

Hypertrophic obstructive cardiomyopathy (subtype)

- Interventricular septum growth blocks left ventricular outflow tract during systole → blood must flow quickly through small opening, ↓ pressure in this area (Venturi effect) → low pressure pulls anterior leaflet of mitral valve toward septum → further mitral valve obstruction towards septum → further obstruction overall

COMPLICATIONS

- Arrhythmias, sudden cardiac death

RISK FACTORS

- Positive family history of HCM/conditions known to be associated with HCM (e.g. Friedreich's ataxia)

SIGNS & SYMPTOMS

- Many individuals asymptomatic
- *Auscultation:* crescendo-decrescendo murmur
  - ↑ intensity with ↓ venous return (Valsalva, standing), ↓ in intensity with ↑ venous return (handgrip, squatting)
- Symptoms arise as complications arise
  - *Dyspnea:* left ventricle stiffening, atrium increasing back pressure into lungs → interstitial lung congestion
  - Fatigue
  - *Exertional chest pain:* ischemia
  - *Syncope with exertion:* brain receiving low oxygen
  - *Palpitations:* ischemia, arrhythmias
  - Sudden cardiac death
- *May exhibit bifid pulse:* two pulses felt
  - Mitral valve moves toward outflow tract → ↑ obstruction mid-systole

DIAGNOSIS

DIAGNOSTIC IMAGING

Echocardiography/cardiac MRI
- Enlarged heart chambers/↓ ejection fraction

Chest X-ray
- ↑ ratio of distance between heart, thoracic cage

LAB RESULTS

Genetic testing
- Cardiomegaly-implicated gene mutations

OTHER DIAGNOSTICS

ECG
- Detectable electrical changes, such as left ventricular hypertrophy

TREATMENT

MEDICATIONS

Beta blockers
- ↓ heart rate, contractile force

Calcium channel blockers
- If beta blockers not tolerated
- Slows down heart rate

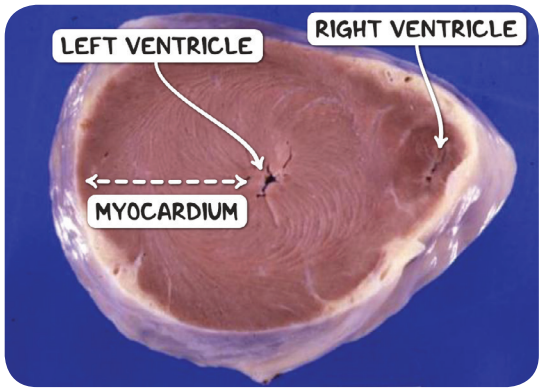

Figure 6.3 Gross pathology of hypertrophic cardiomyopathy. The myocardium has become so enlarged that both ventricles are almost entirely obliterated.

Disopyramide
- Can be used for its negative inotropic properties

Digoxin contraindicated
- ↑ force of contraction, can ↑ obstruction

SURGERY
- Implantable cardioverter-defibrillator

Surgical septal myectomy
- Involves removing portion of interventricular septum, ↓ obstruction

Septal ablation
- Chemical myomectomy to partially ablate septum

Heart transplant
- If unresponsive to all other forms of treatment

OTHER INTERVENTIONS

Lifestyle change
- Cessation of high-intensity athletics

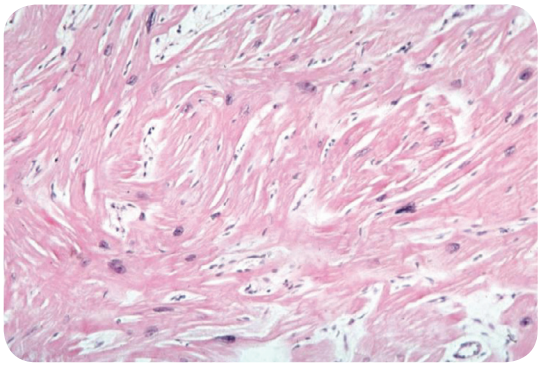

Figure 6.4 The histological appearance of the myocardium in a case of hypertrophic cardiomyopathy. There is complete myocyte disarray. The myocytes display bizarre forms with side to side branching and are arranged in a whorled configuration.

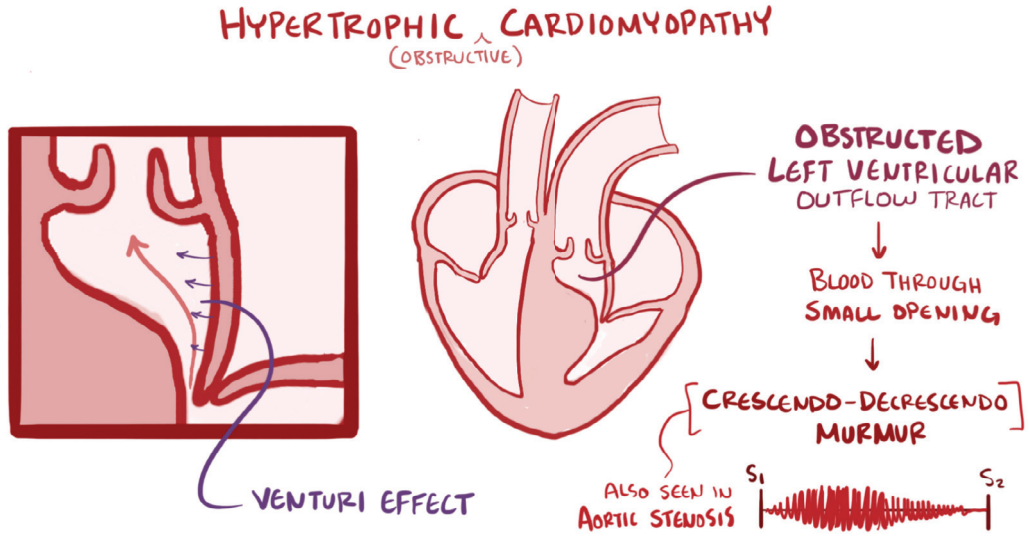

Figure 6.5 Illustration showing the Venturi effect: low blood pressure pulls the anterior leaflet of the mitral valve towards the septum, creating an obstruction. Blood can't get through the small opening, leading to a crescendo-descrescendo heart murmur.

RESTRICTIVE CARDIOMYOPATHY

osms.it/restrictive-cm

PATHOLOGY & CAUSES

- Cardiomyopathy: heart wall is rigid, has difficulty stretching, pumping
- Ventricles restrict filling, ↓ cardiac output

CAUSES

- Infiltrative diseases, storage diseases, endomyocardial diseases.

Amyloidosis

- Amyloids are misfolded proteins → insoluble → deposit in tissues, organs → organs less compliant
- Familial amyloid cardiomyopathy
- Mutant transthyretin (TTR) protein; misfolded deposits preferentially in heart tissue
- Senile cardiac amyloidosis; TTR protein/ wild type TTR deposits in heart over time

Sarcoidosis

- Immune cell collections form granulomas in heart tissue

Endocardial fibroelastosis

- Fibrosis develops in endocardium (inner lining of heart) and subendocardium (layer underneath endocardium)

Loffler syndrome

- Eosinophils accumulate in lung tissue
- *Loeffler endocarditis/Loeffler endomyocarditis:* eosinophils also accumulate in endocardial layer of heart tissue → inflammation, endocardial fibrosis → restrict heart tissue

Hemochromatosis

- Iron deposits in heart tissue, contributes to restricted tissue

Other causes

- Heart tissue radiation
  - Radiation generates reactive oxygen species → inflammation over time → myocardial fibrosis → tissue stiff,

restrictive

COMPLICATIONS

- Can → diastolic heart failure
  - Stiff ventricles → cannot stretch → less filling → low cardiac output → heart failure

MNEMONIC: LASHER

Causes of Restrictive cardiomyopathy

Loffler syndrome
Amyloidosis
Sarcoidosis
Hemochromatosis
Endocardial fibroelastosis
post-**R**adiation

SIGNS & SYMPTOMS

- *Auscultation:* stiff ventricle → S4 heart sound
- *Presents as congestive heart failure:* dyspnea; paroxysmal nocturnal dyspnea; orthopnea; crackles; intraalveolar hemorrhage; fatigue; inability to exercise; appetite loss; abdomen swelling; swelling of feet, ankles; uneven/rapid pulse; chest pain; low urine output; nocturia

DIAGNOSIS

OTHER DIAGNOSTICS

ECG

- *Low-amplitude signals:* peak to nadir measurement of QRS complex being < 5mm (limb leads)/< 10mm (precordial leads). Low voltage produced due to loss of viable myocardium
- *Small QRS complexes:* QRS complexes represent ventricular contraction, restricted tissue → weaker contraction

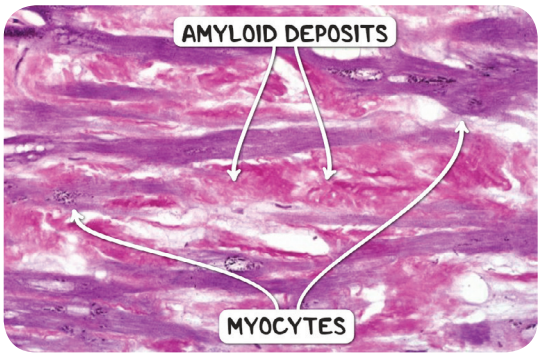

Figure 6.6 Histological appearance of the myocardium in a case of cardiac amyloidosis; a cause of restrictive cardiomyopathy. Cardiac myocytes (dark purple) are surrounded by amyloid deposits (light pink).

TREATMENT

MEDICATIONS

Loop diuretics

- ↓ systemic, pulmonary congestion

Beta-blocker, calcium channel blocker, angiotensin converting enzyme inhibitors

- Slows heart rate, ↑ ventricular-filling time

SURGERY

- Heart transplant

CARDIOMYOPATHY TYPES

	DILATED	HYPERTROPHIED	RESTRICTED
ETIOLOGY	Genetic, infectious, alcoholic, peripartum	Genetic	Infiltrative and storage diseases; endomyocardial disease
PATHOLOGY	Large chambers, relatively weak wall	Asymmetric myocardial growth; markedly thick walls, overgrowth of septum	Rigid, noncompliant heart wall resists filing
VENTRICULAR DYSFUNCTION	Impaired systolic contraction	Impaired diastolic relaxation; hyperdynamic systolic function	Impaired diastolic relaxation; normal systolic function
HEART SOUNDS	S3	S4	S4
VALVULAR INCOMPETENCE	AV valves; especially mitral	Mitral valves	AV valves
ECG	Atrial fibrillation, ventricular tachycardia	Atrial fibrillation; ventricular fibrillation	Low amplitude signals; small QRS signals; AV block
CXR	Enlarged cardiac silhouette (ballooned); pulmonary venous congestion	Normal to enlarged cardiac silhouette	Usually normal cardiac silhouette
ECHOCARDIOGRAM	Decreased LV wall thickness; LV ejection fraction decreased	Hypertrophied LV; LV outflow obstruction; mitral regurgitation; LV ejection fraction normal to increased	Enlarged right atrium; LV ejection fraction decreased; infiltrative disorders cause "speckled" appearance
CLINICAL PEARL	Increased risk of thromboembolic events	May lead to sudden death in young athletes	JVD may manifest as Kussmaul's sign