

Myocardial Disease (Cardiomyopathy)

The questions signed by * are hard: read their explanation. ● Past Years ● TBL ● Others

This file belongs to the PULSE Cardiology question book: the explanations of the multiple choice answers and the model answers of the essay questions, in the words of the handout.

Answer Key | Multiple Choice Questions

Question no.	1	2	3	4	5	6	7	8	9	10	11	12
Answer	a	e	d	d	c	d	c	g	b	a	c	d

Question no.	13	14
Answer	d	c

► Second | Questions with a Note

Question no.	1
Answer	e

► Why this answer?

- 1** *Drugs to be avoided in HOCM are:*
(a) Vaso-Dilators / Diuretics / Digoxin
 HOCM is treated with avoidance of drugs that increase obstruction: +ve inotropic drugs (e.g. digoxin), and drugs that decrease ventricular volume as diuretics or afterload as vasodilators. B-blockers are part of the treatment.
- 2** *Infiltrative restrictive cardiomyopathy with increased LV wall thickness includes:*
(e) Glycogen storage diseases
 Glycogen storage disease fills the heart muscle cells, so the left ventricular wall becomes thick. Haemochromatosis (iron overload) is a myocardial cause of restrictive cardiomyopathy but usually leaves the wall thickness normal, and endomyocardial fibrosis, radiation and the idiopathic type stiffen the heart without thickening it.
- 3** *As regard treatment of restrictive cardiomyopathy which is wrong:*
(d) Digoxin is the preferred drug in RCM and AF
 Digoxin is not the preferred drug in restrictive cardiomyopathy, as toxicity occurs easily, especially in amyloid heart. Treatment is of heart failure, so diuretics, risk-factor control, anticoagulation in AF and SGLT2i, the only drug class with mortality benefit in HFpEF, are correct.
- 4** *The following is incorrect as regard HOCM*
(d) Autosomal recessive inherited disorder
 Hypertrophic cardiomyopathy is familial: autosomal dominant, not recessive. Sudden death is a complication, HOCM is a cause of cardiac syncope, and many patients have no symptoms early in life.

- 5** *The prognosis is poor for dilated cardiomyopathy patients with:*
(c) S3 Gallop
 A persistent S3 gallop reflects a high filling pressure and a severely failing ventricle, and it is a recognised sign of poor prognosis in dilated cardiomyopathy. A diffuse weak apex and soft heart sounds are usual findings of the disease and do not by themselves predict outcome. Functional MR and TR are also usual in dilated cardiomyopathy; marked MR adds risk, but the classic answer for poor prognosis is the S3 gallop.
- 6** *The least likely etiology of dilated cardiomyopathies is:*
(d) Hemochromatosis
 Viral infection and peripartum are listed causes of dilated cardiomyopathy, and dilated or ischemic cardiomyopathy causes left ventricular dilatation. Haemochromatosis is a cause of restrictive cardiomyopathy, so it is the least likely.
- 7** *The least likely diagnosis of restrictive cardiomyopathy is:*
(c) Systemic Lupus
 Amyloidosis, haemochromatosis and sarcoidosis are myocardial causes of restrictive cardiomyopathy. SLE is listed under collagen diseases causing dilated cardiomyopathy.
- 8** *Which of the following is the most probable diagnosis?*
(g) Obstructive cardiomyopathy
 Echo showing asymmetrical septal hypertrophy (ASH) is diagnostic of hypertrophic cardiomyopathy. Jerky pulse, giant A wave and a midsystolic murmur along the left sternal border also fit, with sudden death as a complication.
- 9** *The following is absent in patient with restrictive cardiomyopathy:*
(b) Left ventricular dilatation.
 Restrictive cardiomyopathy is a non-dilated, non-obstructive cardiomyopathy: the ventricles are stiff with a normal or small cavity, while both atria dilate. The wall can be thickened, for example in amyloidosis, so hypertrophy is not always absent. Left ventricular dilatation is the feature that is absent.
- 10** *Hypertrophic obstructive cardiomyopathy is the most common cause of sudden cardiac death in young. Its...*
(a) Autosomal dominant
 Hypertrophic cardiomyopathy is familial: autosomal dominant, thus screening of first degree relatives and genetic counselling are used.
- 11** *Which of the following drug/s should be avoided in patients with hypertrophic obstructive cardiomyopathy:*
(c) Amlodipine.
 Drugs that decrease afterload as vasodilators increase the obstruction and are avoided; amlodipine acts mainly as a vasodilator. B-blockers (metoprolol) and rate-slowing Ca-channel blockers like verapamil (diltiazem) are used in treatment.
- 12** *The following could be a reversible cause of dilated cardiomyopathy:*
(d) All could be reversible.
 In peripartum cardiomyopathy half of patients recover completely and most of the rest improve. Stress cardiomyopathy and tachycardia-induced cardiomyopathy also recover when the stress or the fast rate is controlled, so all could be reversible.
- 13** *Which of the following infectious agents have been associated with the development of inflammatory...*
(d) All of the above
 Diphtheritic and viral myocarditis are listed among specific fevers affecting the heart, and coxsackie B virus and Chagas disease (caused by *Trypanosoma cruzi*) are infective causes of dilated cardiomyopathy. So all are correct.
- 14** *Deficiency of which one of the following trace elements is implicated as a cause of cardiomyopathy?*
(c) Selenium
 Nutritional causes of dilated cardiomyopathy are Beri Beri and selenium deficiency [Keshan disease].

► Second | Questions with a Note: why this answer?

- 1 Which of the following is the most probable diagnosis?

(e) Dilated cardiomyopathy

A woman with a past mastectomy has had breast cancer, and adriamycin is a toxic cause of dilated cardiomyopathy. The displaced apex fits biventricular enlargement, the irregular pulse fits AF, and the pansystolic murmurs fit functional MR and TR.

Model Answers | Essay Questions

- 1 Enumerate causes and categories of dilated cardiomyopathy.

Aetiology of dilated cardiomyopathy

- ① Idiopathic.
- ② **Infective:**
 - Viral esp. coxsackie B virus.
 - Chagas disease.
- ③ **Collagen diseases** e.g. SLE.
- ④ **Endocrinal:**
 - Myxoedema.
 - D.M.
 - Acromegaly.
- ⑤ **Peripartum cardiomyopathy:**
 - In the last trimester or within **6 months** of delivery.
 - Half of patients recover completely & most of the rest improving.
 - Avoid further pregnancy to decrease risk of recurrence.
 - ACE inhibitors are contraindicated in pregnancy.
- ⑥ **Toxic:**
 - Alcohol.
 - Adriamycin.
- ⑦ **Neurologic:**
 - Duchenne's myopathy.
 - Myotonia dystrophica.
 - Friedreich's ataxia.
- ⑧ **Familial.**
- ⑨ **Nutritional:** Beri Beri, selenium deficiency [Keshan disease].

- 2 Enumerate the investigations of dilated cardiomyopathy.

Investigation	Findings
Chest x-ray	Cardiac enlargement involving all the chambers; pulmonary congestion.
ECG	Low voltage; depressed ST segment; flat or inverted T-wave; may be A.F.
Echocardiography and Doppler U/S	Dilated ventricles with no hypertrophy; decreased contractility; MR and TR.

3 Demonstrate investigations of hypertrophic cardiomyopathy.

Investigation	Findings
Chest x-ray	Left ventricular hypertrophy; pulmonary congestion.
ECG	Left ventricular enlargement; right ventricular hypertrophy if there is associated R.V. outflow tract obstruction.
Echocardiography and Doppler U/S	Hypertrophy of the left ventricle with the septal wall thickness exceeds the free wall thickness (asymmetrical septal hypertrophy, ASH): diagnostic .
Microscopic examination	Myocardial fibre disarray "malalignment" [whorls of cells].

4 What is the management?

Treatment

- ① Treatment of H.F. with **avoidance of drugs that increase obstruction**, i.e. +ve inotropic drugs and drugs that decrease ventricular volume as diuretics or afterload as vasodilators.
- ② **B-blockers**: e.g. propranolol decrease tachycardia: "better filling of left ventricle" and prevent syncopal attacks.
- ③ **Ca-channel blockers** e.g. verapamil improve diastolic filling.
- ④ **Surgical treatment**: resection of the septum (myotomy or myomectomy) or laser myoplasty.

Preventive measures

Familial: autosomal dominant, thus screening of first degree relatives and genetic counselling are used as preventive measures.