DGAFMS MEDICAL MEMORANDUM NO -175

CARDIOMYOPATHIES

1. The cardiomyopathies constitute a group of diseases in which the dominant feature is direct involvement of the heart muscle itself. They are not the result of pericardial, hypertensive, congenital, valvular, or ischemic diseases. Although the diagnosis of cardiomyopathy requires the exclusion of these etiological factors, the features of cardiomyopathy are often sufficiently distinctive—both clinically and hemodynamically—to allow a definitive diagnosis to be made.

CLASSIFICATION OF CARDIOMYOPATHIES

2. A variety of schemes have been proposed for classifying the cardiomyopathies. The most widely recognized classification is that by the World Health Organization (WHO) and the International Society and Federation of Cardiology (ISFC). In the WHO/ISFC classification, the cardiomyopathies are classified based on their predominant pathophysiological features; other diseases that affect the myocardium that are associated with a specific cardiac disorder or are part of a generalized systemic disorder are termed ***specific cardiomyopathies***.

3. Three basic types of functional impairment have been described: -

(a) **D*ilated*** (**DCM**, formerly called congestive), the most common form, accounting for 60 percent of all cardiomyopathies and characterized by ventricular dilatation, contractile dysfunction, and often symptoms of congestive heart failure.

(b) ***Hypertrophic*** **(HCM),** recognized by inappropriate left ventricular hypertrophy, often with asymmetrical involvement of the interventricular septum, with preserved or enhanced contractile function until late in the course.

(c) ***Restrictive*** **(RCM),** the least common form, marked by impaired diastolic filling and in some cases with endomyocardial scarring of the ventricle.

4. Two other forms of cardiomyopathy are recognized**, *Arrhythmogenic right ventricular cardiomyopathy*** and ***unclassified;*** the latter includes fibroelastosis, systolic dysfunction with minimal dilatation, and mitochondrial involvement.

5. Examples of ***specific cardiomyopathies*** include ischemic cardiomyopathy, valvular cardiomyopathy, hypertensive cardiomyopathy, and inflammatory cardiomyopathy (myocarditis with cardiac dysfunction). Most forms of specific cardiomyopathy are characterized by the DCM pattern.

6. The term ***ischemic cardiomyopathy*** has been used to describe the condition in which coronary artery disease causes multiple infarctions, diffuse fibrosis, and/or severe ischemia that

leads to left ventricular dilatation with congestive heart failure; it may or may not be associated with angina pectoris.

DILATED CARDIOMYOPATHY

7. **Dilated cardiomyopathy** is a syndrome characterized by cardiac enlargement and impaired systolic function of one or both ventricles.

8. **Clinical Manifestations.** Symptoms usually develop gradually in patients with DCM. Some patients are asymptomatic and yet have left ventricular dilatation for months or even years. This dilatation may be recognized clinically only later when symptoms develop or when routine chest X-ray reveals cardiomegaly.

9. The most striking **symptoms** of DCM are those of *left ventricular failure*. *Fatigue* and *weakness* are common. *Chest pain* occurs may suggest concomitant ischemic heart disease. Chest pain secondary to pulmonary embolism and abdominal pain secondary to congestive hepatomegaly are frequent late in the illness. *Right-sided heart failure* is a late sign and is associated with a poor prognosis.

10. A history of alcohol consumption must be taken because excessive alcohol consumption is a cause of DCM.

11. **Examination** usually reveals a variable degree of *cardiac enlargement* and findings of *congestive heart failure*. The systolic blood pressure is usually normal or low, and the pulse pressure is narrow. *Pulsus alternans* is common when severe left ventricular failure is present. *Cheyne-Stokes* breathing may be present and is associated with a poor prognosis. The *jugular veins* are distended with prominent *v* waves when right-sided heart failure appears. Grossly pulsatile jugular veins with prominent *v* waves indicate the presence of tricuspid valvular regurgitation. The *liver* may be engorged and pulsatile. *Peripheral edema* and *ascites* are present when right-sided heart failure is advanced.

12. The *precordium* usually reveals left and, occasionally, right ventricular impulses. The *apical impulse* is usually displaced laterally. The *second heart sound (S_2)* is usually normally split, although paradoxical splitting may be detected in the presence of left bundle branch block. If pulmonary hypertension is present, the pulmonary component of S_2 may be loud and the splitting may be narrow. *Presystolic gallop sounds (S_4)* are present and often precede the development of overt congestive heart failure. *Ventricular gallops (S_3)* are the rule once cardiac decompensation occurs.

13. *Systolic murmurs* are common and are usually due to mitral or, less commonly, tricuspid valvular regurgitation. Gallop sounds and regurgitant murmurs can often be elicited or intensified by isometric handgrip exercise.

14. *Systemic emboli* resulting from dislodgement of intracardiac thrombi from the left atrium and ventricle and *pulmonary emboli* that originate in the venous system of the legs are common late complications.

15. **Noninvasive laboratory examination.** The *Chest X-Ray* usually reveals generalized cardiomegaly and pulmonary venous hypertension. Interstitial and alveolar edema are less common on initial presentation. Pleural effusions may be present. *HIV screening* should be done because this infection is an important and often unrecognized cause of congestive heart failure

16. **Electrocardiography.** The ECG often shows *sinus tachycardia* when heart failure is present. Various *atrial* and *ventricular tachyarrhythmias* may be seen. *Atrial fibrillation* is common. *Left atrial enlargement* is common. *Poor R wave progression* and *intraventricular conduction abnormalities*, especially left bundle branch block, are common. Anterior *Q waves* may be present when there is extensive left ventricular fibrosis even without evidence of coronary artery disease. *ST segment* and *T wave abnormalities* are common.

17. **24 Hour Holter Monitoring.** Half of monitored patients with DCM exhibit *nonsustained ventricular tachycardia*. Ventricular arrhythmias detected on ambulatory monitoring are a marker for the extent of myocardial damage in DCM and are associated with sudden death.

18. In occasional cases, recurrent and/or incessant supraventricular or ventricular tachyarrhythmias may be the cause of ventricular dysfunction (***Tachycardia induced cardiomyopathy***). Restoration of sinus rhythm or slowing of the heart rate may reverse the cardiomyopathy.

19. **Two-dimensional and Doppler Echocardiography** is useful in assessing the *degree of left ventricular dysfunction* and for *excluding concomitant valvular* or *pericardial disease*. It allows evaluation of the size of the ventricular cavity and thickness of the ventricular walls. A pericardial effusion may be present. Doppler studies are useful for determining the severity of *mitral and tricuspid regurgitation*.

20. **Dobutamine Stress Echocardiography** and **Thallium-201** imaging may be helpful in distinguishing left ventricular enlargement caused by DCM from that caused by coronary artery disease.

21. **Cardiac catheterization.** Certain patients with DCM require cardiac catheterization. It is indicated for those with chest pain and a *suspicion of ischemic disease* or in those thought to have a *treatable systemic disease* such as sarcoidosis or hemochromatosis, where myocardial biopsy is a part of the catheterization procedure. ***Coronary arteriography*** usually reveals normal vessels. This examination may be of particular value in excluding coronary artery disease in patients with abnormal Q waves on the ECG or regional left ventricular wall motion abnormalities on echocardiocardiography.

22. **Endomyocardial biopsy**. Using a flexible bioptome, tissue samples from the right ventricle (and left ventricle when required), maybe obtained with ease and safety. Biopsy may

be of *benefit in* detecting infiltrative disorders of the myocardium and in monitoring for anthracycline cardiotoxicity and cardiac transplant rejection.

23. **Management.** Because the cause of idiopathic DCM, by definition, is unknown, specific therapy is not possible. Treatment, therefore, is for heart failure. Physical, dietary, and pharmacological interventions may help to control symptoms. *Regular physical exercise (as tolerated)* increases exercise capacity. Only *cardiac transplantation* and *specific pharmacological therapy* (the vasodilators enalapril or hydralazine plus nitrates, the beta-adrenoceptor blocker carvedilol, and the aldosterone receptor blocker spironolactone) have been shown to prolong life. *Loop diuretics* (frusemide or torsemide) are needed when clinical LVF is present. *Digoxin* should be added more so when clinical heart failure and atrial fibrillation coexist.

24. **Beta-adrenergic blocker** therapy is now accepted as part of the *four-drug approach* (along with digoxin, vasodilators and diuretics) for all suitable patients with symptomatic congestive heart failure. Patients with advanced heart failure or in a decompensated state should not ordinarily be given a beta-adrenergic blocker as failure may worsen. *Carvedilol* substantially reduces mortality in DCM.

25. Calcium antagonists. Combining a calcium antagonist with traditional standard therapy (digoxin, diuretics, and vasodilator) does not appear to have substantial clinical benefit, nor does it reduce further the mortality in DCM.

26. **Antiarrhythmics.** Although there is no definitive evidence that antiarrhythmic agents prolong life or prevent sudden death in DCM, it may be appropriate to use them in the treatment of symptomatic arrhythmias. Because of the adverse effects of most available agents, many of which depress myocardial contractility and have a proarrhythmic effect, *treatment should be individualized*, with both efficacy and toxicity carefully monitored. *Amiodarone* does not depress myocardial function and maybe considered for ventricular tachyarrhythmias requiring treatment.

27. Electrophysiological testing is of limited utility in DCM. The recording of late potentials by the signal-averaged ECG has appeared to be of benefit in assessing the risk of death in some studies. *Bundle branch reentrant VT* which maybe seen in patients suffering from DCM with intraventricular conduction defects maybe amenable to *Radiofrequency ablation.*

28. **The Implantable Cardioverter-Defibrillator (ICD)** may be considered in appropriate candidates with symptomatic ventricular tachyarrythmias , especially if not responding to optimum medical therapy.

29. **Cardiac resynchronization therapy (CRT) or Biventricular Pacing.** Many patients with advanced systolic heart failure exhibit significant inter- or intraventricular conduction delays (IVCD) that disturb the synchronous beating of the left and right ventricles so that they pump less efficiently (*ventricular dysynchrony).* This is easily recognized by a wide QRS complex with left bundle branch morphology on an ECG. Cardiac resynchronization therapy using *atrial-synchronized biventricular pacing* is indicated for the reduction of symptoms of moderate to severe heart failure (NYHA Function Class III or IV) in those patients who remain

symptomatic despite stable, optimal medical therapy, and have a left ventricular ejection fraction < 35% and a QRS duration >130 ms. In combination with optimal drug therapy, CRT has been shown to improve symptoms and increase exercise capacity in about 80 % of these patients More work is being done to define the exact duration of QRS and exact location for placement of left ventricular leads for optimum benefit.

30. **Combo Devices (combination of CRT and ICD)** may reduce mortality and can be considered in selected cases.In cases where the treating cardiologist , after a trial with optimal medical regime , considers implanting an ICD , CRT or Combo device , case summary with justification will be forwarded by him to Sr Advisor (Med & Cardio)/ Senior Consultant for sanction

31. **Anticoagulants** should be used in the presence of *atrial fibrillation*, if the patient has previously had a *stroke*, and when there is *visible thrombus* on echocardiography. Oral *Warfarin* or *Sintrom* is used to achieve a prolongation of the prothrombin time by 1.5 to 2 times control value or an international normalized ratio (INR) of 2.0 to 3.0.

32. **Surgical Treatment.** *Annuloplasty* or *replacement of regurgitant valves*, in some patients with DCM and prominent atrioventricular valvular regurgitation have shown some symptomatic improvement in some patients.

33. ***Cardiac transplantation*** may be an alternative to medical therapy, with a 5-year survival rate of about 75 percent in appropriately selected patients.

34. Dynamic cardiomyoplasty and partial ventriculotomy has been proposed as an additional surgical alternative to cardiac transplantation but the lack of a randomized control trial demonstrating efficacy has limited its use.

35. **Arrhythmogenic Right Ventricular Cardiomyopathy (ARVD) .** This unique cardiomyopathy is marked by myocardial cell loss with partial or total replacement of right ventricular muscle by adipose and fibrous tissue. ARVD is associated with *reentrant ventricular tachyarrhythmias of right ventricular origin* (a left bundle branch block configuration of the QRS complex) and the risk of *sudden death*. Noninvasive and invasive evaluation demonstrates a dilated, poorly contractile right ventricle, usually with a normal left ventricle. *Magnetic resonance imaging (MRI)* helps to identify these patients. Demonstration of fat in the right ventricular wall is a diagnostic finding. *Antiarrhythmic therapy*, especially with beta-adrenoceptor blockers, sotalol, or amiodarone, often is effective in controlling the arrhythmias. Catheter-based *radiofrequency ablation* has been successful in some patients unresponsive to or intolerant of antiarrhythmic drug therapy. Insertion of an *AICD* or *cardiac transplantation* is reserved for recalcitrant cases.

HYPERTROPHIC CARDIOMYOPATHY

36. The most characteristic **pathophysiological abnormalities** in HCM are

(a) ***Diastolic dysfunction*** characterized by abnormal stiffness of the left ventricle with resultant impaired ventricular filling, increased left ventricular end-diastolic pressure with resulting pulmonary congestion and dyspnea, despite typically hyperdynamic left ventricular systolic function.

(b) ***Dynamic pressure gradient*** across the left ventricular outflow tract in some patients with HCM is considered to be related to narrowing of an outflow tract by septal hypertrophy, possibly abnormal location of the mitral valve and by systolic anterior motion of mitral valve leaflets against the hypertrophied septum.

(c) ***Myocardial ischemia*** is common and multifactorial in HCM. Causes include impaired vasodilator reserve, increased oxygen demand and elevated filling pressures with resultant subendocardial ischemia and occasionally compression of intramyocardial segments of the left anterior descending coronary artery (myocardial bridge).

37. **Clinical Manifestations.** The disease is identified most often in adults in their 30s and 40s. Because *syncope* and *sudden death* have been associated with competitive sports and severe exertion in patients with HCM, it is important to diagnose this condition so that these activities may be avoided.

38. The *clinical picture varies* considerably, ranging from the asymptomatic patient who has a slightly abnormal echocardiogram but no other overt manifestation of the disease to the patient with incapacitating symptoms. The majority are asymptomatic or only mildly symptomatic. The first clinical manifestation of the disease in such individuals may be sudden death.

39. The most common **symptoms** are *dyspnea* (up to 90 percent of symptomatic patients), *Angina pectoris* (75%), *fatigue, presyncope,* and *syncope.* Palpitations, paroxysmal nocturnal dyspnea, overt congestive heart failure, and dizziness are found less frequently. Severe congestive heart failure culminating in death may be seen. *Exertion* tends to worsen many of the symptoms. In children and adolescents presyncope and syncope identify patients at increased risk of sudden death.

40. **Physical examination.** Except for a left ventricular lift and a loud S_4, the physical examination *may be normal in asymptomatic patients without gradients*. Findings are usually prominent in patients with a left ventricular outflow tract pressure gradient.

41. The *apical precordial impulse* is often displaced laterally and is usually abnormally forceful and diffuse. A prominent *presystolic apical impulse* is often present. This may result in a *double apical impulse*. A more characteristic but less frequently recognized abnormality is a *triple apical beat*, the third impulse consisting of a late systolic bulge.

42. The *carotid pulse* typically rises briskly and then declines in midsystole followed by a secondary rise. The *jugular venous pulse* may demonstrate a prominent *a* wave.

43. **Auscultation**. The S_1 is normal and is often preceded by an S_4 that corresponds to the apical presystolic impulse. The S_2 usually is normally split. With severe outflow gradients, paradoxical splitting may be noted. *Systolic ejection sounds* may be found.

44. The auscultatory hallmark of HCM associated with an outflow gradient is a *systolic murmur* that is harsh and crescendo-decrescendo. It commences well after S_1 and is best heard between the apex and the left sternal border. In patients with large gradients, the murmur usually reflects both left ventricular outflow tract obstruction and mitral regurgitation. The murmur is often more holosystolic and blowing at the apex radiating to in the axilla (due to mitral regurgitation) and midsystolic and harsher along the lower sternal border (due to left ventricular outflow obstruction). The systolic murmur is *variable in intensity and duration*, and a variety of maneuvers may be used to augment or suppress it. The murmur of HCM characteristically increases with the strain phase of the Valsalva maneuver and during standing from a squatting position.

45. **Differentiation from Valvular Aortic Stenosis**. The *character of the carotid pulse* and *features of the murmur* are useful to differentiate HCM from valvular aortic stenosis. In aortic valvular stenosis, the carotid upstroke is slow and of low amplitude (pulsus parvus et tardus). With HCM the arterial upstroke is brisk. The murmur of HCM, as opposed to that of aortic stenosis increases with the Valsalva maneuver and during standing from a squatting position, and it decreases during passive leg elevation, and hand grip. The murmur radiates along the carotid arteries in valvular aortic stenosis.

46. **Electrocardiogram**. This is usually abnormal in HCM and invariably so in symptomatic patients with left ventricular outflow tract gradients. The most common abnormalities are *ST segment and T wave abnormalities*, followed by evidence of *left ventricular hypertrophy*, with QRS complexes that are tallest in the mid-precordial leads. Progressive ECG evidence of hypertrophy may develop over time. Giant negative T waves in the mid-precordial leads are characteristic of HCM involving the apex. Prominent *Q waves* are relatively common. The Q wave abnormalities often involve the inferior and/or precordial leads. Other ECG abnormalities may occur including usually *left-axis deviation* and *left atrial enlargement.*

47. **Ventricular arrhythmias** are common in patients with HCM. Runs of *nonsustained ventricular tachycardia* are found in about one fourth of patients. Treadmill testing may expose arrhythmias that are not present at rest.

48. **Continuous ambulatory monitoring** (Holter monitoring) is very useful in detecting repetitive *ventricular tachyarrhythmias*.

49. **Supraventricular tachycardia** is common. *Atrial fibrillation* maybe present and may result in clinical deterioration.

50. **Electrophysiological testing.** The role of electrophysiological studies in identifying HCM patients at increased risk of sudden death is believed to be of limited predictive value.

51. **Chest X-Ray**. The findings on X ray chest are variable; the *cardiac silhouette* may range from normal to markedly increased. Most cases of apparent "cardiomegaly" are the result

of left ventricular hypertrophy and/or left atrial enlargement. Left atrial enlargement is present when significant mitral regurgitation is present.

52. **Echocardiography.** It is useful in the study of patients with suspected HCM and also in the screening of relatives of HCM patients. It is of value in identifying and quantifying morphological features (i.e., distribution of septal hypertrophy), functional aspects (e.g., hypercontractile left ventricle), and hemodynamic findings (e.g., magnitude of outflow gradient).

53. There are three cardinal echocardiographic feature of HCM.

(a) The characteristic feature is ***asymmetrical hypertrophy of the septum (ASH)*** and anterolateral free wall. Maximal hypertrophy of the septum often occurs midway between the base and apex of the left ventricle. The septum is typically at least 15 mm in thickness (normal $\leq$ 11 mm). Diagnosis requires a ratio of septal wall thickness to left ventricular posterior wall thickness (in diastole) of $\geq$ 1.3.

(b) ***Left ventricular outflow tract obstruction*** is often found in HCM. When HCM is associated with a pressure gradient, there is abnormal systolic anterior motion of the anterior leaflet. The degree of mitral regurgitation often correlates with the severity of the outflow gradient.

(c) ***Abnormalities of diastolic function*** may be demonstrated by echocardiography and Doppler recordings in about 80 percent of patients.

54. **Radionuclide scanning**. Thallium-201 myocardial imaging with Single-Photon Emission Computed Tomography *[SPECT]*) permits direct determination of the relative thicknesses of the septum and free wall and is of value when the echocardiographic evaluation in a given patient with suspected HCM is unreliable. *Reversible thallium defects* indicative of ischemia, are common findings in HCM in the absence of obstructive coronary artery disease. Myocardial ischemia is an important factor and probably a mechanism of demise in younger patients.

55. **Cardiac catheterization**. Cardiac catheterization is not required for the diagnosis of HCM, because noninvasive evaluation is almost always enough. It is reserved for situations where *concomitant coronary artery disease* is suspected, or when *invasive modalities of therapy* (e.g., pacemaker, septal ablation, surgery) are being considered. The pressure gradient may be quite labile. A potent stimuli for enhancing the gradient is *postextrasystolic potentiation*. Patients may have *pulmonary hypertension* which is usually mild but may be moderate to severe.

56. **Apical HCM** is a less common variant with predominant involvement of the apex. Typical features include a characteristic spadelike configuration of the left ventricle during angiographic study giant negative T waves in the precordial ECG leads, the absence of an intraventricular pressure gradient, mild symptoms, and a generally benign course.

57. **Natural History.** The clinical course in HCM is *varied*. In many patients symptoms are absent or mild, remain stable. Clinical deterioration (aside from sudden death) usually is slow.

Although symptoms are usually unrelated to the severity or even the presence of a gradient. The percentage of severely symptomatic patients increases with age. The onset of *atrial fibrillation* may lead to an increase in symptoms. Progression of HCM to DCM) may occur.

58. **Sudden death.** Death is most often sudden in HCM and may occur in previously asymptomatic patients, in individuals who were unaware they had the disease, and in patients with an otherwise stable course. The features that identify ***high-risk patients*** include *young age* (< 30 years) at diagnosis, a *family history of HCM with sudden death*, an *abnormal blood pressure response to exercise* and *genetic abnormalities associated with increased prevalence of sudden death*. The presence or severity of an outflow tract gradient, the degree of functional limitation, and symptoms generally do not correlate with the risk of death.

59. **Management** of patients with HCM is aimed at relief of symptoms, prevention of complications, and reduction in the risk of death. Whether asymptomatic patients should receive drug therapy is not established.

60. **Strenuous exertion** should probably be *avoided in all patients with HCM* whether or not symptoms are prominent, especially if high-risk clinical characteristics are present. Almost half of deaths in HCM occur during or just after strenuous physical activity. Unsuspected HCM is the most common abnormality found at autopsy in young competitive athletes who die suddenly. Cardiovascular screening before participation in competitive sports appears to reduce the frequency of unexpected sudden death.

61. **Beta-adrenoceptor blockers**. These drugs are the mainstay of medical therapy of HCM. With their use, angina, dyspnea, and presyncope may all be improved. In patients with resting or provocable gradients beta-adrenoceptor blockade may prevent the increase in outflow obstruction that accompanies exertion. Beta-adrenoceptor blockade may prevent sudden death.

62. **Calcium antagonists**. These are an alternative to beta-adrenoceptor blockade in the management of HCM. Most of the experience has been with verapamil. Diltiazem maybe used. Amlodipine and other vasoselective calcium antagonists are of no use.

63. **Disopyramide**, an antiarrhythmic drug has produced symptomatic improvement and abolition of the pressure gradient in patients with HCM.

64. **Amiodarone** is effective in the treatment of both supraventricular and ventricular tachyarrhythmias in HCM. Limited and inconclusive data are available to show that amiodarone improves prognosis. Amiodarone may also improve symptoms and exercise capacity.

65. Digoxin should generally be avoided unless atrial fibrillation or systolic dysfunction develops. Cautious use of diuretics often helps reduce symptoms of pulmonary congestion, particularly when they are combined with beta-adrenergic blockers or calcium antagonists

66. *Atrial fibrillation* should usually be pharmacologically or electrically cardioverted.

67. **Devices**. Insertion of a dual-chamber DDD ***pacemaker*** may be useful in no more than 10 percent of patients with outflow gradient and severe symptoms, especially the elderly. The

long-term utility of pacing is not known at present, and a substantial placebo effect has been demonstrated. Hence, DDD pacing is not preferred as a primary treatment modality unless pacing is indicated per se.

68. In high-risk patients especially the minority of HCM patients with sustained monomorphic ventricular tachycardia or those with aborted sudden death, an ***AICD*** should be inserted.

69. **Percutaneous transluminal septal myocardial ablation (PTSMA)**. A number of patients with *severe obstruction with significant symptoms* have derived benefit from intentional infarction of a portion of the interventricular septum by the infusion of alcohol into a selectively catheterized septal artery, with reduction of the outflow gradient and improvement in symptoms.

70. **Surgical treatment**. Surgical procedures aimed at reducing the outflow gradient are most commonly used in the markedly symptomatic patient with a gradient at rest above 50 mm Hg who has not responded well to medical management. *Myectomy* is the most widely used operation for HCM and consists of excising a portion of the hypertrophied septum. Surgery results in long-term improvement in symptoms and exercise capacity in most patients. Myotomy-myectomy may be combined with *other necessary operative procedures* like coronary artery bypass grafting or plication of the anterior leaflet of the mitral valve and reconstruction of the submitral valvular apparatus.

RESTRICTIVE AND INFILTRATIVE CARDIOMYOPATHIES

71. The hallmark of the RCMs is ***abnormal diastolic function***; the ventricular walls are excessively rigid and impede ventricular filling. Systolic function is often unimpaired even in many cases with extensive infiltration of the myocardium.

72. A variety of specific pathological processes may result in restrictive cardiomyopathy, although the cause often remains unknown. Myocardial fibrosis, infiltration, or endomyocardial scarring is usually responsible. Myocardial involvement with amyloid is a common cause of RCM.

73. The clinical and hemodynamic features of restrictive heart disease *simulate those of chronic constrictive pericarditis* and differentiation of the two conditions is mandatory because of the potential for successful surgical treatment of constrictive pericarditis.

74. **Clinical manifestations.** *Exercise intolerance, weakness* and *dyspnea* are frequent. Exertional chest pain is usually absent but may be prominent in some patients. In advanced

cases, the *central venous pressure is elevated* with peripheral edema, enlarged liver, ascites, and anasarca. ***Physical examination*** may reveal *jugular venous distention* and *S3* or S_4, or *both*. An *inspiratory increase in venous pressure* may be seen.

75. **Laboratory studies.** *Echocardiography, cardiac catheterization, endomyocardial biopsy, CT* and *radionuclide angiography* may be particularly useful in differentiating the two diseases by demonstrating myocardial scarring or infiltration (biopsy) or thickening of the pericardium (CT and MRI). The *echocardiogram* may demonstrate thickening of the left ventricular wall and an increase of left ventricular mass in patients with infiltrative disease causing RCM. The pattern of filling of the left ventricle differs in the two conditions.

76. The **prognosis** in RCM is variable; usually it is one of relentless symptomatic progression and high mortality.

77. *No specific therapy* other than symptomatic is available excepting for the cardiomyopathy due to iron overload which is improved by removal of the iron and amyloidosis, in which some patients appear to benefit from alkylating-based chemotherapy.

78. **Cardiac Amyloidosis.** Involvement of the heart is a common finding in *immunocyte dyscrasia, familial amyloidosis* and *senile amyloidosis.* In *secondary amyloidosis*, clinically significant cardiac involvement is uncommon. Cardiac amyloidosis occurs more commonly in men than in women, and it is rare before the age of 30 years.

79. Involvement of the cardiovascular system by amyloidosis occurs in four general forms:-

(a) *RCM.* Right-sided findings dominate the clinical presentation.

(b) *Congestive heart failure* due to systolic dysfunction. The course of this form is often one of relentless progression, usually poorly responsive to treatment.

(c) *Orthostatic hypotension* most likely due to amyloid infiltration of the autonomic nervous system, of blood vessels and maybe deposition in the heart and adrenals.

(d) An *abnormality of cardiac impulse formation* and *conduction* is the least common mode of presentation. *Sudden death* is relatively common and may be preceded by episodes of *syncope*.

80. **Diagnosis**. *Biopsy techniques* permit the diagnosis is now made before death in the majority of patients. An *abdominal fat aspirate*, biopsy of rectum, gingiva, bone marrow, liver, kidney and other tissues have been used. *Endomyocardial biopsy* of the right or left ventricles may be helpful if the abdominal fat aspirate is negative.

81. **Endomyocardial Fibrosis.** It is characterized by fibrous endocardial lesions of the inflow of the *right* or *left ventricle* or *both* and often involves the atrioventricular valves, resulting in regurgitation. Right atrial *thrombi* occur commonly. Left ventricular involvement is similar. Endocardial calcific deposits may occur. The epicardial coronary arteries are free of obstructive lesions.

82. The disease is equally frequent in both genders and is common in young adults It is commoner in South India.

83. A pericardial effusion, which may be quite large, may be present.

84. **Clinical Manifestations.** Because EMF may involve both ventricles and either ventricle selectively, symptoms vary. *Left-sided involvement* results in symptoms of pulmonary congestion, whereas predominant *right-sided disease* may present features of an RCM. There is often regurgitation of one or both atrioventricular valves.

85. The onset of the disease is usually insidious. EMF is usually relentlessly progressive. Death is due to progressive myocardial failure, often associated with pulmonary congestion, infection, or infarction, or sudden, unexpected cardiovascular collapse due to arrhythmia. Survival appears to be unrelated to the site of predominant involvement (right or left ventricle), although patients presenting in advanced right-sided failure have a worse prognosis than other patients.

86. **Diagnosis.** This is based on the presence in an individual of the typical clinical and laboratory features, particularly echocardiographic and angiographic, from the appropriate geographical area.

87. **Management.** The *medical treatment* of EMF is often difficult and not particularly effective. In patients with advanced disease, the outlook is poor, with a 35 to 50 percent 2-year mortality. Better survival may be seen in less symptomatic patients who have milder forms of the disease. *Digitalis glycosides* may be helpful in controlling the ventricular rate in patients with atrial fibrillation, but the response of congestive symptoms is disappointing. Diuretics are not particularly helpful in the treatment of ascites. Once endomyocardial disease has reached the fibrotic stage, *surgery* offers the possibility of symptomatic improvement and is the treatment of choice. Operative mortality has been high and long-term results suggest that surgery is at best palliative.

DISPOSAL

88. All cases of suspected cardiomyopathy should be referred for evaluation by a cardiologist at the nearest cardiology centre. The cardiologist shall evaluate the patient clinically and by using appropriate non-invasive investigative modalities and decide on the course of therapy. The cardiologist will, based on the clinical and functional status and long term prognosis, decide the appropriate medical classification. A cardiologist would periodically review these patients. The frequency of review would be specified and endorsed at time of giving an opinion based on the clinical status and the requirement of monitoring and delivering further therapy.

89. Some cases would be required to be referred to a centre with facilities of a cardiac catheterization laboratory or advanced radiology modalities and/or cardiothoracic surgery for further evaluation and therapy. Examples are:

(a) Where concomitant coronary artery disease is suspected.

(b) When invasive modalities of therapy e.g., pacemaker, septal ablation, AICD implantation, radiofrequency ablation, cardiac resynchronization therapy or surgery etc are being considered.

(c) In those thought to have infiltrative disorders of the myocardium where endomyocardial biopsy would assist in the diagnosis.

(d) Where cardiac catheterization, endomyocardial biopsy, CT and radionuclide angiography are required to differentiate restrictive heart disease from chronic constrictive pericarditis (because of the potential for successful surgical treatment of constriction).

DGAFMS MEDICAL MEMORANDUM NO 174

CARDIAC MURMURS

(Issued in 2009)

(This supersedes DGAFMS Medical Memorandum No 138 of 1993)

Distribution

(a) One copy per medical officer
(b) One copy per medical unit
(c) One copy per Medical Branches of Formation Headquarters.

Issued under the authority of

THE DIRECTOR GENERAL ARMED FORCES MEDICAL SERVICES

(Distributed by the Controller of Publications, Civil Lines Delhi – 54)

Case File No – PC/39869/DGAFMS/DG-3C

DGAFMS MEDICAL MEMORANDUM NO 175

CARDIOMYOPATHIES

(Issued in 2009)

Distribution

(a) One copy per medical officer
(b) One copy per medical unit
(c) One copy per Medical Branches of Formation Headquarters.

Issued under the authority of

THE DIRECTOR GENERAL ARMED FORCES MEDICAL SERVICES

(Distributed by the Controller of Publications, Civil Lines Delhi – 54)

Case File No – PC/39870/DGAFMS/DG-3C

DGAFMS & SR COL COMDT AWARD

CHIEF OF ARMY STAFF AWARD

CHIEF OF NAVAL STAFF AWARD

CHIEF OF AIR STAFF AWARD

LATE LT GEN RS HOON, PVSM, AVSM AWARD

LATE MAJ GEN NG LATELY MEMORIAL PRIZE AND EASSY COMPETITION AWARD

SCIENTIFIC SESSION (MEDICINE & ALLIED SUBJECTS INCL PATHOLOGY, PREVENTIVE AND SOCIAL MEDICINE, HOSPITAL ADMINISTRATION AND NURSING)

SCIENTIFIC SESSION (SURGERY & ALLIED SUBJECTS INCLUDING DENTAL SURGERY AND NURSING)

CME (MEDICINE & ALLIED SUBJECTS)

CME (SURGERY & ALLIED SUBJECTS)

CME (PATHOLOGY INCL MICROBIOLOGY, TRANSFUSION MEDICINE, FORENSIC MEDICINE AND BIOCHEMISTRY)

CME (PREVENTIVE AND SOCIAL MEDICINE)

CME (HOSPITAL ADMINISTRATION, ANATOMY, PHYSIOLOGY AND PHARMACOLOGY)

CME (DENTAL SURGERY

Tele : 23094808

REGISTERED / SDS

Office of the DGAFMS/DG-3C
Ministry of Defence
PIN – 908713
C/O 56 APO

PC-39869/DGAFMS/DG-3C

Jun 2009

Lt Col Sunil Sofat
Cl Spl (Cardio)
AH (R&R)
Delhi Cantt – 10

PRINTING OF DGAFMS MEDICAL MEMORANDUM NO 174 (CARDIAC MURMURS)

1. Second Proof of the draft publication in respect of DGAFMS Medical Memorandum No 174 Titled "Cardiac Murmurs" received from the Printing Press Faridabad is being forwarded herewith for your proof reading the document and checking of any errors.

2. It is requested that the same duly corrected be returned to this Dte Gen by 20 Jun 2009.

(Smita Shahi)
Lt Col
Jt Dir AFMS (MR)
for DGAFMS

Encls : As above

Internal :-

DGAFMS/DG (Hindi) - Hindi version of Second proof of the draft publication in Respect of above DGAFMS Medical Memorandum is forwarded herewith for your checking the correctness of the document. You are requested to return the same duly corrected by 20 Jun 2009

Tele : 23094808

REGISTERED / SDS

Office of the DGAFMS/DG-3C
Ministry of Defence
PIN – 908713
C/O 56 APO

PC-39869/DGAFMS/DG-3C

Jun 2009

Wg Cdr Hemant Madan
Cl Spl (Cardio)
AH (R&R)
Delhi Cantt – 10

PRINTING OF DGAFMS MEDICAL MEMORANDUM NO 175 (CARDIAOMYPATHIES)

1. Second Proof of the draft publication in respect of DGAFMS Medical Memorandum No 175 Titled "Cardiomypathies" received from the Printing Press Faridabad is being forwarded herewith for your proof reading the document and checking of any errors.

2. It is requested that the same duly corrected be returned to this Dte Gen by 20 Jun 2009.

(Smita Shahi)
Lt Col
Jt Dir AFMS (MR)
for DGAFMS

Encls : As above

Internal :-

DGAFMS/DG (Hindi) - Hindi version of Second proof of the draft publication in Respect of above DGAFMS Medical Memorandum is forwarded herewith for your checking the correctness of the document. You are requested to return the same duly corrected by 20 Jun 2009