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Heart muscle disease management in aircrew

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ABSTRACT

This manuscript focuses on the broad aviation medicine considerations that are required to optimally manage aircrew with suspected or confirmed heart muscle disease (both pilots and non-pilot aviation professionals). ECG abnormalities on aircrew periodic medical examination or presentation of a family member with a confirmed cardiomyopathy are the most common reason for investigation of heart muscle disease in aircrew. Holter monitoring and imaging, including cardiac MRI is recommended to confirm or exclude the presence of heart muscle disease and, if confirmed, management should be led by a subspecialist. Confirmed heart muscle disease often requires restriction to flying duties due to concerns regarding arrhythmia. Pericarditis and myocarditis usually require temporary restriction and return to flying duties is usually dependent on a lack of recurrent symptoms and acceptable imaging and electrophysiological investigations.

INTRODUCTION

The physiological demands of the aviation environment, with the potential for exposure to hypoxia, hypobaria, acceleration forces and positive pressure breathing, represent a unique challenge to the cardiovascular system. Cardiomyopathies represent a heterogeneous group of conditions affecting the heart muscle, with a variety of morphological and physiological characteristics. Their natural history is highly variable, both between conditions and between individuals with the same condition. The appropriate management of cardiomyopathies should involve a subspecialist in these conditions and/or inherited cardiac conditions (ICC) as the appropriate management of individuals is often nuanced and complex. As with all areas of aviation medicine, the impact and risk of both the condition and subsequent treatments on the ability of aircrew to undertake their duties must be fully considered.[i]

This manuscript uses the European Society of Cardiology 2008 classification as a basis for the categorisation of cardiomyopathies.[1] Their position statement defines cardiomyopathies as 'a myocardial disorder in which the heart muscle is structurally and functionally abnormal, in the absence of coronary artery disease (CAD), hypertension, valvular disease and congenital heart disease, sufficient to cause the observed myocardial abnormality'.[1] Myocarditis, pericarditis and athlete's heart are also included; however, 'ion channelopathies', although considered a cardiomyopathy by the American Heart Association,[2] are not, as they are discussed elsewhere.[3] The different groups of cardiomyopathies are considered in brief, although the rarest conditions are not considered due to the low likelihood of these being found in the aircrew population, and who would be considered for aircrew duties on a case-by-case basis.

Screening and investigation for cardiomyopathy in aircrew

Cardiomyopathies are most commonly discovered in aircrew due to abnormalities on the routine ECG undertaken in their periodic medical examinations or findings on physical examination. The ECG may manifest as T-wave inversion, frequent premature ventricular contractions (PVCs), or increased voltages. Rhythm and conduction disorders on ECG may warrant further assessment for a possible underlying cardiomyopathy.[4] The finding of left ventricular hypertrophy (LVH) by voltage criteria alone is seen more commonly in aircrew than the general population[5] and as an isolated finding has a low specificity, particularly in tall, thin, younger aircrew. In the absence of T-wave repolarisation abnormalities, LVH by voltage criteria alone should generally be considered a normal variant; however, height, weight and blood pressure should also be considered in making this determination. Aircrew

[i]Evidence-based cardiovascular risk assessment in aircrew poses significant challenges in the aviation environment as data to support decision making at the low level of tolerable risk in aviation is rarely available from the published literature. As a result, there are discrepancies between aviation authority's recommendations in different countries, and even between licensing organisations within single countries. The North Atlantic Treaty Organisation (NATO) HFM-251 Occupational Cardiology in Military Aircrew working group is constituted of full-time aviation medicine and aviation cardiology experts who advise both their militaries and civil aviation organisations including, but not limited to, the Federal Aviation Authority, Civil Aviation Authority, European Aviation Safety Agency and North American Space Agency. The recommendations of this group are based on a 3-year working group that considered best clinical cardiovascular practice guidelines within the context of aviation medicine and risk principles. This work was conducted independently of existing national and transnational regulators, both military and civilian, but considered all available policies, to determine best evidence-based practice in this field. The recommendations presented in this document, and associated manuscripts, is based on expert consensus opinion of the NATO group. This body of work has been produced to develop the evidence base for military aviation cardiology and to continue to update the relevant civilian aviation cardiology advice following the 1998 European Cardiology Society aviation cardiology meeting.

may also present for screening when a family member has been diagnosed with cardiomyopathy or has had a sudden cardiac death (SCD).

First-line investigations

Echocardiography

Transthoracic echocardiography (TTE) is the main investigation in the initial assessment of heart muscle disease. It is a ubiquitous and cost-effective tool for both diagnosis and follow-up of most heart muscle diseases. Abnormal findings on TTE may be sufficient to make the diagnosis of cardiomyopathy and may be sufficient for initial risk stratification. In those with suspected myocarditis or pericarditis, TTE may be normal or demonstrate a pericardial effusion.

Table 1 First-line screening and investigation for cardiomyopathy in aircrew

The presence of isolated left ventricular hypertrophy by voltage criteria, in those with no additional ECG abnormalities, and normal blood pressure does not usually require additional investigation.	Strongly recommended
First-line investigation of aircrew suspected of having cardiomyopathy should include transthoracic echocardiography, ambulatory ECG monitoring and an exercise stress test.	Strongly recommended
Abnormal ECG findings and/or clinical findings suggestive of cardiomyopathy should warrant further assessment.	Strongly recommended
In aircrew where there is uncertainty with regard to the diagnosis of mild hypertrophic cardiomyopathy or 'athletic heart', cardiopulmonary exercise testing is recommended.	Recommended

Ambulatory ECG monitoring

The use of continuous ECG monitoring may be useful in assessment and risk stratification of cardiomyopathy in aircrew. Extended monitoring up to a week is preferred to increase the probability of capturing a relevant arrhythmia. In hypertrophic cardiomyopathy (HCM), the detection of atrial fibrillation may indicate the need for anti-arrhythmic medication and/or anticoagulation, and the presence of non-sustained/sustained ventricular tachycardia (VT) is a known risk factor for SCD. A high burden of PVCs detected on Holter monitoring (>2% on 24 hours Holter) may be suggestive of an underlying cardiomyopathy, or potentially the cause of a tachycardia-related cardiomyopathy, if long standing. Additionally, ECG abnormalities may form part of the criterion for diagnosis, such as in arrhythmogenic ventricular cardiomyopathy (AVC).[ii]

Exercise stress testing

Exercise stress testing (EST) may be used to assess exercise capacity in cardiomyopathy, and is of value in those with HCM, to assess dynamic changes in cardiac output. It must be undertaken under appropriate supervision, given the risk of clinically significant arrhythmia. Formal cardiopulmonary exercise testing (CPET), with formal measurement of maximal VO_2 may be useful in distinguishing milder phenotypes of HCM from 'athletic heart'. In the latter, peak oxygen consumption is usually supra-normal, whereas in those with HCM it is abnormal.

Second-line imaging investigations

Cardiac MRI

Cardiac MRI (CMR) may confirm a diagnosis of cardiomyopathy when echocardiography is doubtful or inconclusive (due to poor echo windows or borderline values). It is also useful for assessing apical segments of the heart, which may be subject to artefact on TTE. The ability to fully assess the right heart is particularly useful in cases of suspected AVC. A key aspect of CMR in the investigation of suspected cardiomyopathy is the use of gadolinium contrast agents to perform tissue characterisation. The specific appearances of late gadolinium enhancement (LGE) on CMR imaging may allow a cardiomyopathy to be diagnosed before any other imaging abnormalities are detectable.[4] Therefore, in those with a clinical suspicion of cardiomyopathy, CMR with gadolinium should be strongly considered, even if TTE is normal. The presence of LGE on CMR is associated with adverse outcomes[6 7] and may be useful in identifying those at highest risk.[8] Late gadolinium techniques may also allow for a distinction to be made between myocardial infarction and myocarditis. In aircrew, reaching the correct diagnosis is especially important in this regard, to ensure appropriate aeromedical and occupational disposal. Additionally, perfusion CMR may be used to rule out CAD as a cause of LV dysfunction.

Table 2 Cardiac MRI

In aircrew with clinical suspicion of cardiomyopathy, cardiac MRI (CMR) with gadolinium should be considered mandatory even if transthoracic echocardiography is normal.	Strongly recommended
In aircrew with significant levels of ventricular ectopy (>2% on 24 hours Holter), CMR with gadolinium is strongly recommendedto exclude an underlying cardiomyopathy, either as the aetiology of the ectopy, or as a result of long-standing ectopic disease (ectopy-related cardiomyopathy).	Strongly recommended

Coronary angiography

Coronary angiography, either invasive or CT based, may be used to exclude CAD as the aetiology of LV impairment.

Genetic testing

A variety of genetic abnormalities have been identified within the different cardiomyopathies, but there is significant genetic heterogeneity within each condition. Genetic testing may be undertaken with appropriate pretest counselling by a trained individual. Testing may also be used to assist in screening for cardiomyopathies but is often only possible if a genetic abnormality has been identified in the index patient. This may be particularly useful in aircrew, as it may provide a high degree of reassurance that they are unlikely to develop the disease and avoiding licensing restrictions.

[ii] Aircrew are defined somewhat differently in civil and military aviation. NATO and International Civilian Aviation Organisation delegates the definition of aircrew to national authorities. In the civilian sector, aircrew are often categorised as flight crew (pilots)/technical crew members and cabin crew, with separate regulation for air traffic controllers. The military define aircrew more broadly as 'persons having duties concerned with the flying or operation of the air system, or with passengers or cargo when in flight'. From a risk perspective, professional (commercial) pilots have a higher attributable risk than private pilots and non-pilot aircrew. Controllers are considered to have an attributable risk equivalent to professional pilots. From a cardiovascular perspective, aircrew whose flying role includes repetitive exposure to high acceleration forces (Gz) comprise a subgroup who, due to the unique physiological stressors of this flight environment, often require specific aeromedical recommendations. A more detailed description of aircrew is available in table 1 of the accompanying introductory paper on aviation cardiology (Nicol ED, *et al*. *Heart* 2018;**105**:s3–s8. doi:10.1136/heartjnl-2018-313019).

Endomyocardial biopsy

Endomyocardial biopsy may demonstrate typical histological appearances of HCM or AVC. However, it is invasive, associated with a risk of myocardial perforation and may result in false-negative results. It is rarely performed with advanced imaging techniques replacing its use.

Confirmed cardiomyopathies

General aeromedical concerns

The appropriate management of cardiomyopathies should involve a subspecialist in cardiomyopathy and/or ICC as the appropriate management of individuals is often nuanced and complex. The primary aeromedical concerns associated with cardiomyopathies are arrhythmias and LV dysfunction. Exposure to sustained acceleration (+G_z) is itself arrhythmogenic and may aggravate cardiomyopathy-associated arrhythmic activity resulting in a sudden decrease in cardiac output and G-induced loss of consciousness (G-LOC). Associated LV dysfunction may also compromise appropriate cardiac output augmentation in response to sustained acceleration (+G_z). Even in the absence of +G_z stress, arrhythmias may result in distracting symptoms which may compromise flight safety. Although in normally functioning hearts, repeated exposure to increased +G_z has not been shown to affect cardiac function,[9] it is not known whether exposure to +G_z, on a repeated basis, with associated catecholamine surges, might result in deterioration of cardiac function in aircrew with a cardiomyopathy.

Treatments for cardiomyopathy are also of potential significance in the aviation context. The use of ACE inhibitors (ACEi), angiotensin receptor blockers (ARBs) and beta-blockers (βBs) are all indicated in the management of cardiomyopathies.[10–13] Treatment with βB may reduce an individual's G_z tolerance due to the negative inotropic and chronotropic effect of βB. Implantable cardioverter defibrillators (ICDs) may be indicated in those with cardiomyopathy and while they may prevent SCD, they may not prevent aeromedically significant arrhythmias, or syncope. An ICD discharge, either appropriate or inappropriate, may result in major distraction and/or incapacitation. In the military context, additional concerns regarding ICD include the increased risk of device-associated infection when operating in austere locations and the effect on the ICD from military equipment that emit electromagnetic radiation.

Dilated and hypertrophic cardiomyopathy

Both dilated cardiomyopathy (DCM) and HCM are seen in aircrew but are uncommon with a prevalence of 1 in 1000 and 1 in 2000, respectively.[14–18] The genetics of DCM and HCM are heterogenous and complex but may be useful for risk stratification. In HCM, certain patterns of disease are associated with specific mutations, and the presence of multiple mutations may result in a more extreme phenotype. A full genetic assessment may therefore be valuable. HCM is most frequently transmitted in an autosomal dominant pattern, but penetrance is incomplete, and is related to age.[19] DCM is familial in approximately half of cases[20]; however, genetic testing may not be possible unless a previous genetic mutation has been identified in the index case. Due to the variable disease progression, initial testing may be inconclusive, and therefore serial evaluation of aircrew with a first-degree relative diagnosed with DCM or HCM may be required over long periods of follow-up; it is recommended that screening continue at 5 yearly intervals in the general population up to the age of 50 years,[21] but in aircrew screening biannually should be strongly considered. The need for repeated investigation may be modified considering information from genetic screening when this is performed.

Table 3 Dilated and hypertrophic cardiomyopathy

Aircrew with confirmed cardiomyopathies should be managed in conjunction with a specialist cardiomyopathy or inherited cardiac conditions service to ensure appropriate specialist clinical management.	Strongly recommended
Aircrew with impaired LV function, documented arrhythmia, pharmacological treatment that may impair G_z tolerance, or ICD implantation shoud initially be made unfit to fly. . Return to limited flying duties may be considered on a case-by-case basis in non-pilot aircrew.	Not recommended
In aircrew with a first-degree relative with confirmed DCM or HCM, screening (with ECG and echocardiography) is recommended for both initial and relicensing.	Strongly recommended
Any aircrew with confirmed cardiomyopathy should be made unfit to fly ; return to limited flying duties may be possible but only in those with mild disease.	Not recommended
Pilot aircrew, or non-pilot aircrew with mission critical roles, who have confirmed mild DCM or HCM could be considered for multicrew and non-high-performance flying duties, if asymptomatic.	Consider
In those with a first-degree relative with HCM, this should include genetic testing, if a causal genetic mutation has been identified. If DCM, genetic testing should only be undertaken in accordance with guidelines and not performed routinely.	Strongly Recommended
If ECG and TTE is normal, interval screening for aircrew should be considered at a 2-yearly intervals.	Recommended

Dilated cardiomyopathy

It is important to note that the diagnosis of idiopathic DCM requires exclusion of alcohol, ischaemic and metabolic disorders. The pathophysiology of DCM is complex but is associated with cavity dilatation and systolic dysfunction of the left or both ventricles due to myocyte apoptosis. Wall stress, angiotensin II, catecholamines via the β-adrenergic system, reactive oxygen species, nitric oxide and inflammatory cytokines are all implicated in disease progression. The β-adrenergic surges associated with many forms of flying raises the possibility that in aircrew with DCM, exposure to high performance flying might hasten progression, or worsen the prognosis, of DCM.

Risk stratification in DCM is challenging due to its heterogeneous nature. A confirmed diagnosis of DCM is likely to require restrictions to flying duties. The degree of LV dysfunction and dilatation should be considered, as these are associated with increased risk of malignant arrhythmia. There is some evidence that better outcomes are associated with improvement in LV function with medical therapy, younger age, briefer duration of symptoms, history of hypertension and, somewhat counterintuitively, a worse NYHA class at presentation.[22] However, aircrew with suspected or confirmed DCM require clinical follow-up over at least 2–3 years to establish whether this is the case in any individual. The use of ACEi, ARBs and βBs for the treatment of DCM is well established. ACEi and ARBs in themselves are compatible with continued flying; however, βBs may limit individuals G_z tolerance, requiring restriction from high-performance or aerobatic flight. Other medical therapy (eg, aldosterone antagonists, anti-arrhythmics) are likely to be used in those with greater degrees of LV impairment, or identified arrhythmias, and therefore limitations are likely to be due to the underlying cardiomyopathy, rather than the treatment itself.[3] Approximately half of patients with DCM will have an improvement in ejection fraction with medical treatment, but in a third of patients, inexorable progression occurs.

The onset of any symptoms in those with DCM is likely to reflect a level of risk of incapacitation, which is significantly >1% per annum, and therefore would be disqualifying regardless of medical therapy. Syncope is associated with a significant risk of SCD in DCM,[23] and any high-risk features by conventional measures should be a bar to flying.

Hypertrophic cardiomyopathy

In HCM, diastolic dysfunction, myocardial ischaemia, left ventricular outflow tract obstruction (LVOTO), abnormal vascular responses and arrhythmias are all important and must be carefully considered in aircrew. HCM is associated with significant risk of SCD, especially in those aged under 35 years,[24–26] those with heart failure[27] and atrial fibrillation.[28] Mortality rates in HCM are reported between 1% and 2% per annum, most commonly due to SCD, heart failure and thromboembolic events.[29] Mortality due to SCD is associated with severe LVH, family history of SCD, syncope and certain genetic phenotypes associated with poorer outcomes.[30] However, the variability of the disease, combined with its low prevalence, mean that few data are available from randomised trials. Most flight critical and mission critical aircrew will be barred from professional flight duties on the diagnosis of HCM due to their raised cardiovascular risk profile.

Around a third of patients with HCM are unable to significantly increase their systolic blood pressure (SBP), or drop their SBP on exercise. Abnormal blood pressure response on EST is a risk factor for SCD, particularly when seen in those under the age of 40 years.[31] In aircrew, a reduction in BP or inability to augment BP appropriately, when exposed to G_z may result in G-LOC with potentially catastrophic outcomes.

Ischaemia and fibrosis, as well as LVOTO, may act as a trigger for arrhythmia in HCM. Non-sustained VT and paroxysmal supraventricular arrhythmias (often asymptomatic) has been found in up to a fifth of patients with HCM.[32 33] Atrial fibrillation is the most common sustained arrhythmia in HCM and increases in frequency with age. Non-sustained VT during, or immediately following, exercise may indicate a high risk of SCD.[34] The onset of arrhythmia in flight may result in distraction or incapacitation and is a particularly important factor in aircrew. Anticoagulation and anti-arrhythmic therapy may both affect aeromedical disposition.

Medical therapy is usually indicated for symptoms, or in the presence of severe LVH. Transcoronary ablation of septal hypertrophy and surgical septal intervention are often considered in cases of LVOTO when symptoms are severe, and medical therapy provides inadequate relief. ICD implantation is strongly considered in those with severe LVH, and a history of VT (sustained or non-sustained), or syncope.[35] Although designed to produce symptomatic relief, and/or reduce risk of SCD, both medication and interventional procedures have the potential for side effects and complications. Both transcoronary ablation of septal hypertrophy and myectomy may result in myocardial scarring, which presents an arrhythmic substrate, with surgical myectomy associated with adverse remodelling with LV dilatation, and alcohol septal ablation associated with a possible risk of heart block requiring a permanent pacemaker; these sequelae may result in stringent restrictions to aircrew licensing. The indications for ICD implantation would usually have already resulted in withdrawal of flying privileges.

Athletic heart

High levels of athletic activity is associated with ECG changes due to increased vagal tone, abnormal cardiac chamber dimensions, increased LV mass and wall thickness.[36] Differentiating between healthy but enlarged hearts, and those with cardiomyopathy can be extremely challenging, however is critical in aircrew.

Some common findings such as sinus bradycardia, first-degree AV block, incomplete right bundle branch block (RBBB), early repolarisation and isolated QRS voltage criteria for LVH would be considered normal findings on an athlete's ECG. However, T-wave inversion, ST-depression, pathological Q waves, long-QT or short-QT interval and complete left, or right, bundle branch block would not be considered to be related to training.[37]

Although smaller degrees of increased LV thickness may be a physiological response to high-level intense exercise, this should not be ≥13 mm in male athletes[38] and should be lower in women. The use of Doppler echo techniques for assessing diastolic filling may also be helpful with diastolic function often impaired in HCM but is normal or enhanced in athletes.

Ventricular sizes may be increased in athletic heart, but overall systolic function should be within the normal range. Although ejection fraction may be at the lower end of the normal range, or even borderline, function should augment with exercise, unlike in significant pathological ventricular impairment. CPET to differentiate between cardiomyopathy and athletic adaptation can also be extremely useful. In athletes, the peak oxygen consumption may be supra-normal, whereas cardiomyopathy are associated with abnormal indices.

CMR may be useful in distinguishing between pathology, rather than physiological adaptation. The presence of characteristic patterns of LGE may provide additional evidence of an underlying cardiomyopathy. Distinguishing the appropriately adapted heart from the maladapted one can be extremely challenging. To appropriately assess aircrew, an integrated approach is required, including a full history and comprehensive investigation. Despite this, uncertainty may remain. In those with no significant clinical concerns in their history and no clear abnormalities on testing, flying can continue unrestricted. However, interval assessment should be undertaken at intervals of not more than 2 years.

Table 4 Athletic heart

Aircrew with suspicion of athletic heart may, in the absence of significant clinical concerns in their history and no clear abnormalities on testing, fly unrestricted.	Strongly recommended
In cases of uncertainty, cardiac MRI (CMR), cardiopulmonary exercise testing and specific Doppler analysis on transthoracic echocardiography is strongly recommended.	Strongly recommended
Repeat testing at intervals of no more than 2 years should be undertaken as per international guidelines.	Strongly recommended

Restrictive cardiomyopathies

Restricted cardiomyopathies (RCM) are usually classified into primary and secondary forms. Primary forms include Löffler's endocarditis, primary amyloid and endomyocardial fibrosis, and are usually associated with poor outcomes. Secondary forms of RCM are associated with infiltrative diseases (eg, amyloidosis, sarcoidosis), storage diseases (eg, haemochromatosis, Andersen-Fabry disease) or post irradiation (for those with radiation to the chest for malignancy). Impaired diastolic function, hypertrophy and restrictive filling patterns are associated with all forms, regardless of underlying aetiology. Concern from an aeromedical perspective is the potential for conduction defects in infiltrative and storage forms of the disease. Therefore, in aircrew the aetiology of any restrictive cardiomyopathy should be elucidated, as it impacts on aeromedical disposition, risk assessment, treatment and prognosis.

In aircrew with an established or potential diagnosis of sarcoidosis, cardiac involvement must be investigated by CMR with gadolinium. The finding of cardiac sarcoid is a bar to flying, due to the potential for bradyarrhythmia and heart block resulting from infiltration affecting the conducting system. Haemochromatosis may result in iron overload in the liver, pancreas, joints and heart if not identified and treated. Phlebotomy and iron-chelating agents mean that cardiac involvement is uncommon. The use of MRI to assess hepatic and cardiac iron loading is now standard practice. Any evidence of cardiac iron overload should result in restriction of flying privileges.

Table 5 Restrictive cardiomyopathy

In aircrew with proven or probable sarcoid disease, assessment with CMR (plus gadolinium) is recommended to confirm/exclude cardiac involvement. If cardiac involvement is confirmed aircrew should be considered unfit.	Strongly recommended
In aircrew with haemochromatosis, CMR should be strongly considered to assess for cardiac iron overload. If confirmed aircrew should be considered unfit.	Recommended
Aircrew with primary restrictive cardiomyopathy, confirmed cardiac sarcoid or cardiac haemochromatosis are not recommended for aircrew duties.	Not recommended

Arrhythmogenic ventricular cardiomyopathy

The most common clinical presentation of arrhythmogenic ventricular cardiomyopathy (AVC) is with exercise-triggered, symptomatic VT. Although the right ventricle is classically affected in AVC, a significant proportion of cases involve the LV or both ventricles.[39] Localised or diffuse atrophy of the myocardium occurs, with fibrous or adipose tissue infiltration seen on histology. AVC is associated with ventricular dysfunction, and malignant ventricular arrhythmias which may result in SCD. It is recognised as being one of the leading causes of SCD in those aged ≤35 years and may be responsible for up to 1 in 10 cases of SCD in those aged ≤65 years.[40 41] The incidence of sudden death in AVC due to ventricular arrhythmias is thought to be 1%–2% per annum.[39 42] Therefore, a diagnosis of AVC in aircrew is considered disqualifying. Due to the potential for familial transmission, aircrew with a first-degree relative who have a diagnosis of AVC should be fully investigated for the condition.

Table 6 Arrhythmogenic ventricular cardiomyopathy

Aircrew with suspicion of arrhythmogenic ventricular cardiomyopathy (AVC), or a first-degree relative with AVC, should be grounded while fully investigated.	Recommended
A diagnosis of AVC should be considered disqualifying for aircrew duties in both applicants and trained aircrew due to the potential of malignant arrhythmias and sudden cardiac death.	Not recommended

Myocarditis

In the Western world, myocarditis is most commonly associated with cardiotropic viruses. In other regions, Chagas disease, Borrelia infection and diphtheria may be the underlying cause. Myocarditis may present acutely, with typical chest pain symptoms or with more severe symptoms of heart failure and arrhythmia. In these cases, diagnosis is based on serum troponin measurements, TTE and CMR, and will also often involve coronary angiography to rule out CAD. It may be missed due to its subtle symptoms or be detected on CMR as an incidental finding following an asymptomatic episode. Progression from myocarditis to DCM occurs in approximately a fifth of those affected. SCD is a well-recognised association with acute myocarditis, most commonly in younger patients and in association with strenuous physical exertion, with the highest risk being in the 6 months following diagnosis.

The clinical presentation of myocarditis and acute coronary syndromes maybe similar; however, the occupational ramifications of these two diagnoses in aircrew differ substantially and mandate optimal assessment to discriminate between them. The use of CMR to look for myocarditis, either using T2-weighted sequences to look for oedema, or using LGE to look for fibrosis, is strongly encouraged. There are some data to suggest that the presence of LGE in myocarditis is associated with a worse prognosis,[43] which further supports its use in aircrew. CMR imaging may be useful for the follow-up of aircrew with myocarditis, to assess LV function and fibrosis burden.

Although full recovery from myocarditis is thought to occur in approximately 80% of those with myocarditis, there are no clinical measures that have proven useful in predicting outcomes in these patients. Even those with fulminant disease, with rapid onset of symptoms and haemodynamic compromise may have an excellent outcome.[44] General features of postviral syndromes and reduced exercise capacity may persist for many months following an episode of myocarditis, and this should also be borne in mind when considering returning aircrew to flying duties. Therefore, a cautious approach, with initial restriction to flying duties and close follow-up over a period of time is required in aircrew.

Table 7 Myocarditis

Aircrew with suspected myocarditis should be grounded and assessed with transthoracic echocardiography to determine LV function and/or pericardial effusion.	Strongly recommended
If confirmed myocarditis, aircrew should be grounded for 6 months initially.	Strongly recommended
CMR is strongly recommended to distinguish myocarditis from an acute coronary event and to determine future prognosis.	Strongly recommended
Prior to return to flight, full assessment with 24 hours Holter, echocardiography and exercise stress testing is strongly recommended.	Strongly recommended
In those with acceptable findings, unrestricted flying is possible. Restrictions are required if LV dysfunction or aeromedically significant arrhythmia is detected.	Strongly recommended
In mild myocarditis, restricted aircrew duties may be possible after 3 months when first-line investigations and CMR show normal results. Unrestricted aircrew duties may be considered after 6 months.	Consider

Pericarditis

Acute pericarditis may present with severe pain, which may be have an acute onset. Any associated pericardial effusion may result in haemodynamic compromise. In high-income countries, a viral aetiology is the most common cause,[45] although in countries where tuberculosis is endemic it is most commonly the cause of pericarditis. Although pericarditis involves primarily the pericardium, the inflammatory process may also involve the myocardium (myopericarditis). The resulting myocarditis is usually mild.

Diagnosis of acute pericarditis is based on pericarditic chest pain, pericardial rubs, new widespread ST-elevation or PR depression on ECG, and a new or worsening pericardial effusion.[46] Elevated inflammatory markers, or imaging evidence

of pericardial inflammation may also provide support to the diagnosis. The presence of a pericardial effusion may result in haemodynamic compromise, which may have a more profound effect with exposure to G_z in particular. Therefore, aircrew with suspected or known pericarditis must undergo TTE to look for pericardial effusion. The use of aspirin or non-steroidal anti-inflammatory drugs is recommended for the treatment of pericarditis until symptoms fully resolve, along with colchicine which is continued for 6–12 weeks.[46] Corticosteroids are associated with an increased risk of recurrence and use as a first-line treatment is not recommended. Aircrew are likely to be off medication by the time a return to flying is being considered, although if they remain on treatment, consideration should be given to a further period of grounding to ensure that symptoms have settled.

A period of reduced physical activity is recommended following an episode of pericarditis[47 48] and aircrew should not fly during this period. Recurrence occurs in approximately 15%–30% of cases of acute pericarditis, although this may be halved if colchicine is used.[49] In order to look for evidence of recurrence, and to avoid increased activity in the short-term following an attack of pericarditis, aircrew should be grounded for a minimum period of 3 months.

Table 8 Pericarditis

Aircrew with suspected pericarditis should be grounded and assessed with transthoracic echocardiography to determine left ventricular function and/or pericardial effusion.	Strongly recommended
Aircrew with a confirmed diagnosis of pericarditis must be grounded for 3 months initially. For idiopathic or viral aetiologies, treatment with aspirin/non-steroidal anti-inflammatory drugs and colchicine (which should be continued for at least 6 weeks) is strongly recommended.	Strongly recommended
Before returning to aircrew duties, a full assessment with first-line investigations must be performed. In those with acceptable findings, unrestricted aircrew duties are possible. For aircrew in whom chest pain is precipitated or aeromedically significant arrhythmia is detected, aircrew restrictions or continued grounding is required.	Strongly recommended
In mild myopericarditis, return to restricted flying is possible after 3 months, provided the first-line investigations show satisfactory results. Return to unrestricted flying maybe considered after 6 months.	Consider

CONCLUSION

Heart muscle diseases are a heterogeneous group of pathologies, with highly variable natural history and presentation. The risk of arrhythmia is significant in aircrew. All confirmed diagnoses of cardiomyopathy, myocarditis or pericarditis will result in initial restriction to flying privileges, with grounding being a common outcome in cardiomyopathy. However, in those with equivocal or borderline diagnoses, continued flying may be possible, potentially with restrictions and subject to ongoing close follow-up and acceptable periodic investigations.

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