



Australian Government
Repatriation Medical Authority

Statement of Principles concerning CARDIOMYOPATHY (No. 85 of 2015)

The Repatriation Medical Authority determines the following Statement of Principles.

Dated 19 June 2015

The Common Seal of the
Repatriation Medical Authority
was affixed to this instrument
at the direction of:

A handwritten signature in black ink, appearing to read 'Nicholas Saunders'.

Professor Nicholas Saunders AO
Chairperson

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1 Name

This is the Statement of Principles concerning cardiomyopathy (No. 85 of 2015).

2 Commencement

This instrument commences on **20 July 2015**.

3 Authority

This instrument is made under subsection 196B(2) of the *Veterans' Entitlements Act 1986*.

4 Revocation

The Statement of Principles concerning cardiomyopathy No. 23 of 2007, and the Statement of Principles concerning familial hypertrophic cardiomyopathy No. 35 of 2007, made under subsection 196B(2) of the VEA are revoked.

5 Application

This instrument applies to a claim to which section 120A of the VEA or section 338 of the *Military Rehabilitation and Compensation Act 2004* applies.

6 Definitions

The terms defined in the Schedule 1 - Dictionary have the meaning given when used in this instrument.

7 Kind of injury, disease or death to which this Statement of Principles relates

- (1) This Statement of Principles is about cardiomyopathy and death from cardiomyopathy.

*Meaning of **cardiomyopathy***

- (2) For the purposes of this Statement of Principles, cardiomyopathy:
- (a) means a disease of the heart muscle (myocardium) associated with mechanical or electrical cardiac dysfunction, in the absence of coronary artery disease, hypertension and valvular disease sufficient to cause the observed myocardial abnormality; and

- (b) includes familial hypertrophic cardiomyopathy and other forms of primary genetic cardiomyopathy, primary-mixed forms of dilated and restrictive cardiomyopathy, and takotsubo (stress) cardiomyopathy, but excludes pericardial disease or rejection of a heart transplant.

Note: *familial hypertrophic cardiomyopathy* and *takotsubo cardiomyopathy* are defined in the Schedule 1 - Dictionary.

- (3) While cardiomyopathy attracts ICD-10-AM code 099.4, 090.3, I42 or I43, in applying this Statement of Principles the meaning of cardiomyopathy is that given in subsection (2).
- (4) For subsection (3), a reference to an ICD-10-AM code is a reference to the code assigned to a particular kind of injury or disease in *The International Statistical Classification of Diseases and Related Health Problems, Tenth Revision, Australian Modification* (ICD-10-AM), Ninth Edition, effective date of 1 July 2015, copyrighted by the Independent Hospital Pricing Authority, ISBN 978-1-76007-020-5.

Death from cardiomyopathy

- (5) For the purposes of this Statement of Principles, cardiomyopathy, in relation to a person, includes death from a terminal event or condition that was contributed to by the person's cardiomyopathy.

Note: *terminal event* is defined in the Schedule 1 – Dictionary.

8 Basis for determining the factors

The Repatriation Medical Authority is of the view that there is sound medical-scientific evidence that indicates that cardiomyopathy and death from cardiomyopathy can be related to relevant service rendered by veterans, members of Peacekeeping Forces, or members of the Forces under the VEA, or members under the MRCA.

Note: *relevant service* is defined in the Schedule 1 – Dictionary.

9 Factors that must exist

At least one of the following factors must as a minimum exist before it can be said that a reasonable hypothesis has been raised connecting cardiomyopathy or death from cardiomyopathy with the circumstances of a person's relevant service:

- (1) being a prisoner of war of Japan before the clinical onset of cardiomyopathy;
- (2) for males only, drinking at least 125 kilograms of alcohol within a continuous five year period before the clinical onset of cardiomyopathy;

Note: *alcohol* is defined in the Schedule 1 - Dictionary.

- (3) for females only, drinking at least 75 kilograms of alcohol within a continuous five year period before the clinical onset of cardiomyopathy;

Note: *alcohol* is defined in the Schedule 1 - Dictionary.

- (4) having haematological or biochemical evidence of poisoning with cobalt at the time of the clinical onset of cardiomyopathy;
- (5) having a hypersensitivity reaction of the myocardium to a drug at the time of the clinical onset of cardiomyopathy;

Note: *hypersensitivity reaction of the myocardium to a drug* is defined in the Schedule 1 - Dictionary.

- (6) being treated with an anthracycline within the 25 years before the clinical onset of cardiomyopathy;

Note: *anthracycline* is defined in the Schedule 1 - Dictionary.

- (7) being treated for cancer with a chemotherapeutic agent from the specified list of chemotherapeutic agents, within the three months before the clinical onset of cardiomyopathy;

Note: *specified list of chemotherapeutic agents* is defined in the Schedule 1 - Dictionary.

- (8) receiving tacrolimus for organ transplantation within the three months before the clinical onset of cardiomyopathy;
- (9) being treated daily with chloroquine or hydroxychloroquine for at least the one year before the clinical onset of cardiomyopathy;

- (10) being treated with a drug or a drug from a class of drugs from Specified List of Drugs No. 1, within the two months before the clinical onset of cardiomyopathy;

Note: *Specified List of Drugs No. 1* is defined in the Schedule 1 - Dictionary.

- (11) using a drug from Specified List of Drugs No. 2 at the time of the clinical onset of cardiomyopathy;

Note: *Specified List of Drugs No. 2* is defined in the Schedule 1 - Dictionary.

- (12) having carbon monoxide poisoning within the 24 hours before the clinical onset of cardiomyopathy;
- (13) being infected with human immunodeficiency virus before the clinical onset of cardiomyopathy;
- (14) having infection-related myocarditis before the clinical onset of cardiomyopathy;
- (15) having Whipple's disease at the time of the clinical onset of cardiomyopathy;

Note: *Whipple's disease* is defined in the Schedule 1 - Dictionary.

- (16) having a disorder from the specified list of endocrine disorders at the time of the clinical onset of cardiomyopathy;

Note: *specified list of endocrine disorders* is defined in the Schedule 1 - Dictionary.

- (17) having acquired generalised lipodystrophy at the time of the clinical onset of cardiomyopathy;

Note: *lipodystrophy* is defined in the Schedule 1 - Dictionary.

- (18) having a catecholamine-secreting tumour at the time of the clinical onset of cardiomyopathy;

- (19) having giant cell myocarditis, or a disease from the specified list of inflammatory connective tissue diseases, involving the heart, at the time of the clinical onset of cardiomyopathy;

Note: *specified list of inflammatory connective tissue diseases* is defined in the Schedule 1 - Dictionary.

- (20) having a vasculitis from the specified list of systemic vasculitides, involving the heart, at the time of the clinical onset of cardiomyopathy;

Note: *specified list of systemic vasculitides* is defined in the Schedule 1 - Dictionary.

- (21) having coeliac disease at the time of the clinical onset of cardiomyopathy;

- (22) having an infiltrative disease affecting the myocardium at the time of the clinical onset of cardiomyopathy;

Note: *infiltrative disease* is defined in the Schedule 1 - Dictionary.

- (23) undergoing a course of therapeutic radiation for cancer, where the heart was in the field of radiation, before the clinical onset of cardiomyopathy;

- (24) having received a cumulative equivalent dose of at least 0.5 sievert of ionising radiation to the heart before the clinical onset of cardiomyopathy;

Note: *cumulative equivalent dose* is defined in the Schedule 1 - Dictionary.

- (25) having clinically apparent nutritional deficiency from the specified list of nutritional deficiencies at the time of the clinical onset of cardiomyopathy;

Note: *specified list of nutritional deficiencies* is defined in the Schedule 1 - Dictionary.

- (26) being peripartum at the time of the clinical onset of cardiomyopathy;

Note: *being peripartum* is defined in the Schedule 1 - Dictionary.

- (27) being obese at the time of the clinical onset of cardiomyopathy;

Note: *being obese* is defined in the Schedule 1 - Dictionary.

- (28) having chronic renal failure at the time of the clinical onset of cardiomyopathy;

Note: *chronic renal failure* is defined in the Schedule 1 - Dictionary.

- (29) having cirrhosis of the liver before the clinical onset of cardiomyopathy;

- (30) being envenomated by a scorpion, a funnel web spider, a spider belonging to the genus *Latrodectus*, or a specified jellyfish, within the 24 hours before the clinical onset of cardiomyopathy;

Note: *specified jellyfish* is defined in the Schedule 1 - Dictionary.

- (31) for familial hypertrophic cardiomyopathy only, where the clinical onset is first manifest as heart failure, cardiac arrhythmia or sudden cardiac death, undertaking physical activity of greater than five METs at the time of the clinical onset of cardiomyopathy;

Note: *familial hypertrophic cardiomyopathy* and *MET* are defined in the Schedule 1 - Dictionary.

- (32) for takotsubo cardiomyopathy only:

- (a) experiencing a category 1A stressor within the 30 days before the clinical onset of cardiomyopathy;
- (b) experiencing a category 1B stressor within the 30 days before the clinical onset of cardiomyopathy;
- (c) having:
 - i) an injury or illness requiring admission to an intensive care unit or artificial ventilation;
 - ii) major trauma; or
 - iii) septicaemia;within the 30 days before the clinical onset of cardiomyopathy;
or
- (d) having a cerebrovascular accident or subarachnoid haemorrhage within the 30 days before the clinical onset of cardiomyopathy;

Note: *category 1A stressor*, *category 1B stressor*, *artificial ventilation* and *takotsubo cardiomyopathy* are defined in the Schedule 1 - Dictionary.

- (33) for males only, drinking at least 125 kilograms of alcohol within a continuous five year period before the clinical worsening of cardiomyopathy;

Note: *alcohol* is defined in the Schedule 1 - Dictionary.

- (34) for females only, drinking at least 75 kilograms of alcohol within a continuous five year period before the clinical worsening of cardiomyopathy;

Note: *alcohol* is defined in the Schedule 1 - Dictionary.

- (35) having haematological or biochemical evidence of poisoning with cobalt at the time of the clinical worsening of cardiomyopathy;

- (36) having a hypersensitivity reaction of the myocardium to a drug at the time of the clinical worsening of cardiomyopathy;

Note: *hypersensitivity reaction of the myocardium to a drug* is defined in the Schedule 1 - Dictionary.

- (37) being treated with an anthracycline within the 25 years before the clinical worsening of cardiomyopathy;

Note: *anthracycline* is defined in the Schedule 1 - Dictionary.

- (38) being treated for cancer with a chemotherapeutic agent from the specified list of chemotherapeutic agents, within the three months before the clinical worsening of cardiomyopathy;

Note: *specified list of chemotherapeutic agents* is defined in the Schedule 1 - Dictionary.

- (39) receiving tacrolimus for organ transplantation within the three months before the clinical worsening of cardiomyopathy;

- (40) being treated daily with chloroquine or hydroxychloroquine for at least the one year before the clinical worsening of cardiomyopathy;

- (41) being treated with a drug or a drug from a class of drugs from Specified List of Drugs No. 1, within the two months before the clinical worsening of cardiomyopathy;

Note: *Specified List of Drugs No. 1* is defined in the Schedule 1 - Dictionary.

- (42) using a drug from Specified List of Drugs No. 2 at the time of the clinical worsening of cardiomyopathy;

Note: *Specified List of Drugs No. 2* is defined in the Schedule 1 - Dictionary.

- (43) having carbon monoxide poisoning within the 24 hours before the clinical worsening of cardiomyopathy;

- (44) being infected with human immunodeficiency virus before the clinical worsening of cardiomyopathy;

- (45) having infection-related myocarditis before the clinical worsening of cardiomyopathy;

- (46) having Whipple's disease at the time of the clinical worsening of cardiomyopathy;

Note: *Whipple's disease* is defined in the Schedule 1 - Dictionary.

- (47) having a disorder from the specified list of endocrine disorders at the time of the clinical worsening of cardiomyopathy;

Note: *specified list of endocrine disorders* is defined in the Schedule 1 - Dictionary.

- (48) having acquired generalised lipodystrophy at the time of the clinical worsening of cardiomyopathy;

Note: *lipodystrophy* is defined in the Schedule 1 - Dictionary.

(49) having a catecholamine-secreting tumour at the time of the clinical worsening of cardiomyopathy;

(50) having giant cell myocarditis, or a disease from the specified list of inflammatory connective tissue diseases, involving the heart, at the time of the clinical worsening of cardiomyopathy;

Note: *specified list of inflammatory connective tissue diseases* is defined in the Schedule 1 - Dictionary.

(51) having a vasculitis from the specified list of systematic vasculitides, involving the heart, at the time of the clinical worsening of cardiomyopathy;

Note: *specified list of systemic vasculitides* is defined in the Schedule 1 - Dictionary.

(52) having coeliac disease at the time of the clinical worsening of cardiomyopathy;

(53) having an infiltrative disease affecting the myocardium at the time of the clinical worsening of cardiomyopathy;

Note: *infiltrative disease* is defined in the Schedule 1 - Dictionary.

(54) undergoing a course of therapeutic radiation for cancer, where the heart was in the field of radiation, before the clinical worsening of cardiomyopathy;

(55) having received a cumulative equivalent dose of at least 0.5 sievert of ionising radiation to the heart before the clinical worsening of cardiomyopathy;

Note: *cumulative equivalent dose* is defined in the Schedule 1 - Dictionary.

(56) having clinically apparent nutritional deficiency from the specified list of nutritional deficiencies at the time of the clinical worsening of cardiomyopathy;

Note: *specified list of nutritional deficiencies* is defined in the Schedule 1 - Dictionary.

(57) being peripartum at the time of the clinical worsening of cardiomyopathy;

Note: *being peripartum* is defined in the Schedule 1 - Dictionary.

(58) having chronic renal failure at the time of the clinical worsening of cardiomyopathy;

Note: *chronic renal failure* is defined in the Schedule 1 - Dictionary.

(59) having cirrhosis of the liver before the clinical worsening of cardiomyopathy;

- (60) being envenomated by a scorpion, a funnel web spider, a spider belonging to the genus *Latrodectus*, or a specified jellyfish, within the 24 hours before the clinical worsening of cardiomyopathy;

Note: *specified jellyfish* is defined in the Schedule 1 - Dictionary.

- (61) undertaking physical activity of greater than five METs at the time of the clinical worsening of cardiomyopathy;

Note: *MET* is defined in the Schedule 1 - Dictionary.

- (62) for takotsubo cardiomyopathy only:

- (a) experiencing a category 1A stressor within the 30 days before the clinical worsening of cardiomyopathy;
- (b) experiencing a category 1B stressor within the 30 days before the clinical worsening of cardiomyopathy;
- (c) having:
 - i) an injury or illness requiring admission to an intensive care unit or artificial ventilation;
 - ii) major trauma; or
 - iii) septicaemia;within the 30 days before the clinical worsening of cardiomyopathy; or
- (d) having a cerebrovascular accident or subarachnoid haemorrhage within the 30 days before the clinical worsening of cardiomyopathy;

Note: *category 1A stressor*, *category 1B stressor*, *artificial ventilation* and *takotsubo cardiomyopathy* are defined in the Schedule 1 - Dictionary.

- (63) inability to obtain appropriate clinical management for cardiomyopathy.

10 Relationship to service

- (1) The existence in a person of any factor referred to in section 9 must be related to the relevant service rendered by the person.
- (2) The factors set out in subsections 9(33) to 9(63) apply only to material contribution to, or aggravation of, cardiomyopathy where the person's cardiomyopathy was suffered or contracted before or during (but did not arise out of) the person's relevant service.

11 Factors referring to an injury or disease covered by another Statement of Principles

In this Statement of Principles:

- (1) if a factor referred to in section 9 applies in relation to a person; and
- (2) that factor refers to an injury or disease in respect of which a Statement of Principles has been determined under subsection 196B(2) of the VEA;

then the factors in that Statement of Principles apply in accordance with the terms of that Statement of Principles as in force from time to time.

Schedule 1 - Dictionary

Note: See Section 6

1 Definitions

In this instrument:

alcohol is measured by the alcohol consumption calculations utilising the Australian Standard of ten grams of alcohol per standard alcoholic drink.

anthracycline means a class of cell-cycle non-specific drugs used in cancer chemotherapy derived from *Streptomyces peucetius* var. *caesius*, and includes daunorubicin (daunomycin), doxorubicin (adriamycin), epirubicin, idarubicin, valrubicin and mitoxantrone.

artificial ventilation means a method to assist or replace spontaneous breathing, which may involve a mechanically-controlled ventilator, manually-assisted bag ventilation of an intubated patient, or expired air resuscitation.

being obese means having a Body Mass Index (BMI) of 30 or greater.

The $BMI = W/H^2$ and where:

W is the person's weight in kilograms; and

H is the person's height in metres.

being peripartum means being in the last trimester of pregnancy or being within the six months immediately postpartum.

cardiomyopathy—see subsection 7(2).

category 1A stressor means one of the following severe traumatic events:

- (a) experiencing a life-threatening event;
- (b) being subject to a serious physical attack or assault including rape and sexual molestation; or
- (c) being threatened with a weapon, being held captive, being kidnapped, or being tortured.

category 1B stressor means one of the following severe traumatic events:

- (a) being an eyewitness to a person being killed or critically injured;
- (b) viewing corpses or critically injured casualties as an eyewitness;
- (c) being an eyewitness to atrocities inflicted on another person or persons;
- (d) killing or maiming a person; or
- (e) being an eyewitness to or participating in, the clearance of critically injured casualties.

Note: **eyewitness** is also defined in the Schedule 1 - Dictionary.

chronic renal failure means having a glomerular filtration rate of less than 60 mL/min/1.73 m² for a period of at least three months, or the presence of irreversible kidney damage.

cumulative equivalent dose means the total dose of ionising radiation received by the particular organ or tissue. The formula used to calculate the cumulative equivalent dose allows doses from multiple types of ionising radiation to be combined, by accounting for their differing biological effect. The unit of equivalent dose is the sievert. For the purposes of this Statement of Principles, the calculation of cumulative equivalent dose excludes doses received from normal background radiation, but includes therapeutic radiation, diagnostic radiation, cosmic radiation at high altitude, radiation from occupation-related sources and radiation from nuclear explosions or accidents.

eyewitness means a person who observes an incident first hand and can give direct evidence of it. This excludes a person exposed only to media coverage of the incident.

familial hypertrophic cardiomyopathy means a genetic disease of cardiac muscle caused by a variety of mutations in genes encoding sarcomeric proteins. It is characterised by left ventricular hypertrophy and diastolic dysfunction, and may cause obstruction to left ventricular outflow and symptoms and signs of heart failure.

hypersensitivity reaction of the myocardium to a drug means eosinophilic myocarditis due to an allergic reaction, as indicated by:

- (a) skin rash, fever, peripheral eosinophilia, ECG changes, tachycardia and elevated cardiac enzymes; or
- (b) endomyocardial biopsy showing infiltration into the myocardium by eosinophils, multinucleated giant cells and leukocytes, occurring during drug therapy.

infiltrative disease means:

- (a) a primary or metastatic neoplasm;
- (b) amyloidosis;
- (c) iron overload; or
- (d) sarcoidosis.

Note: ***iron overload*** is also defined in the Schedule 1 - Dictionary.

iron overload means an accumulation of excess iron in tissues and organs which has been confirmed by elevated ferritin or transferrin saturation levels. Causes include haemochromatosis and blood transfusions.

lipodystrophy means a condition due to defective metabolism of fat, resulting in the absence of subcutaneous fat.

MET means a unit of measurement of the level of physical exertion. 1 MET = 3.5 ml of oxygen/kg of body weight per minute, or 1.0 kcal/kg of body weight per hour, or resting metabolic rate.

MRCIA means the *Military Rehabilitation and Compensation Act 2004*.

relevant service means:

- (a) operational service under the VEA;
- (b) peacekeeping service under the VEA;
- (c) hazardous service under the VEA;
- (d) British nuclear test defence service under the VEA;
- (e) warlike service under the MRCA; or
- (f) non-warlike service under the MRCA.

specified jellyfish means a carybdeid ('box') jellyfish or *Pelagia noctiluca*.

specified list of chemotherapeutic agents means:

- (a) 5-fluorouracil (non-topical);
- (b) alemtuzumab;
- (c) bevacizumab;
- (d) cisplatin;
- (e) cladribine;
- (f) cyclophosphamide;
- (g) dasatinib;
- (h) denileukin difitox;
- (i) gemcitabine;
- (j) ifosfamide;
- (k) imantinib;
- (l) interferon alpha;
- (m) interleukin-2;
- (n) lapatinib;
- (o) lenalidomide;
- (p) mitomycin C;
- (q) paclitaxel;
- (r) pentostatin;
- (s) sorafenib;
- (t) sunitinib; or
- (u) trastuzumab.

Specified List of Drugs No. 1 means:

- (a) amphotericin B;
- (b) anagrelide;
- (c) antiretroviral agents (zidovudine, didanosine, zalcitabine);
- (d) clozapine;
- (e) denileukin difitox;
- (f) exogenous catecholamine;
- (g) isotretinoin or tretinoin;
- (h) itraconazole;
- (i) lithium;
- (j) mesalamine;
- (k) methylphenidate; or
- (l) olanzapine.

Specified List of Drugs No. 2 means:

- (a) amphetamines or amphetamine derivatives, including 3, 4-methylenedioxymethamphetamine (Ecstasy);
- (b) androgenic-anabolic steroids; or
- (c) cocaine.

specified list of endocrine disorders means:

- (a) acromegaly;
- (b) adrenal insufficiency;
- (c) Cushing's syndrome;
- (d) diabetes mellitus;
- (e) Graves' disease;
- (f) hyperthyroidism or thyrotoxicosis;
- (g) hypoparathyroidism; or
- (h) hypothyroidism or Hashimoto's thyroiditis.

specified list of inflammatory connective tissue diseases means:

- (a) ankylosing spondylitis;
- (b) dermatomyositis;
- (c) inclusion body myositis;
- (d) polymyositis;
- (e) rheumatoid arthritis;
- (f) scleroderma (progressive systemic sclerosis);
- (g) Sjogren's syndrome; or
- (h) systemic lupus erythematosus.

specified list of nutritional deficiencies means:

- (a) beriberi (thiamine deficiency);
- (b) carnitine deficiency;
- (c) Keshan disease (selenium deficiency);
- (d) pellagra (niacin deficiency); or
- (e) scurvy (Vitamin C deficiency).

specified list of systemic vasculitides means:

- (a) Behcet's disease;
- (b) eosinophilic granulomatosis with polyangiitis (Churg Straus syndrome);
- (c) giant cell (temporal) arteritis;
- (d) granulomatosis with polyangiitis (Wegener's granulomatosis);
- (e) mucocutaneous lymph node syndrome (Kawasaki disease);
- (f) microscopic polyangiitis;
- (g) polyarteritis nodosa; or
- (h) Takayasu's arteritis.

takotsubo cardiomyopathy, also known as stress cardiomyopathy, means a type of acute non-ischaemic cardiomyopathy in which there is a sudden temporary weakening of the myocardium, with ballooning of the left ventricular apex and a hypercontractile left ventricular base.

terminal event means the proximate or ultimate cause of death and includes:

- (a) pneumonia;
- (b) respiratory failure;
- (c) cardiac arrest;
- (d) circulatory failure; or
- (e) cessation of brain function.

VEA means the *Veterans' Entitlements Act 1986*.

Whipple's disease means a systemic infectious disease caused by the bacterium *Tropheryma whipplei*, which primarily causes malabsorption, but may also affect any part of the body.