



Australian Government
Repatriation Medical Authority

Statement of Principles
concerning
SYSTEMIC SCLEROSIS
(Balance of Probabilities)
(No. 26 of 2018)

The Repatriation Medical Authority determines the following Statement of Principles under subsection 196B(3) of the *Veterans' Entitlements Act 1986*.

Dated 2 March 2018

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 'N. Saunders'.

Professor Nicholas Saunders AO
Chairperson

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1 Name

This is the Statement of Principles concerning *systemic sclerosis (Balance of Probabilities)* (No. 26 of 2018).

2 Commencement

This instrument commences on 2 April 2018.

3 Authority

This instrument is made under subsection 196B(3) of the *Veterans' Entitlements Act 1986*.

4 Revocation

The Statement of Principles concerning systemic sclerosis No. 65 of 2009 made under subsection 196B(3) of the VEA is revoked.

5 Application

This instrument applies to a claim to which section 120B of the VEA or section 339 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 systemic sclerosis and death from systemic sclerosis.

Meaning of systemic sclerosis

- (2) For the purposes of this Statement of Principles, systemic sclerosis (also known as scleroderma):
- (a) means an autoimmune, fibrosing connective tissue disorder characterised by deposition of collagen in skin and internal organs, and microvascular lesions, especially in the skin, lungs and kidneys; and
 - (b) excludes localised sclerosis, graft versus host disease with dermal fibrosis, and scleroderma-like conditions (for example, scleromyxoedema and eosinophilic fasciitis).

Note 1: The three main subsets of systemic sclerosis are limited cutaneous systemic sclerosis (previously referred to as CREST syndrome, comprising calcinosis cutis, Raynaud phenomenon, oesophageal dysmotility, sclerodactyly, telangiectasias), diffuse cutaneous systemic sclerosis and systemic sclerosis sine scleroderma.

Note 2: The diagnosis of systemic sclerosis is based on clinical findings, although histopathologic confirmation is sometimes needed to rule out other diseases. Serological markers for systemic sclerosis include elevated levels of anticentromere antibodies and anti-Scl-70 antibodies.

- (3) While systemic sclerosis attracts ICD-10-AM code M34, in applying this Statement of Principles the meaning of systemic sclerosis 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)*, Tenth Edition, effective date of 1 July 2017, copyrighted by the Independent Hospital Pricing Authority, ISBN 978-1-76007-296-4.

Death from systemic sclerosis

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

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

8 Basis for determining the factors

On the sound medical-scientific evidence available, the Repatriation Medical Authority is of the view that it is more probable than not that systemic sclerosis and death from systemic sclerosis can be related to relevant service rendered by veterans or members of the Forces under the VEA, or members under the MRCA.

Note: *MRCA*, *relevant service* and *VEA* are defined in the Schedule 1 – Dictionary.

9 Factors that must exist

At least one of the following factors must exist before it can be said that, on the balance of probabilities, systemic sclerosis or death from systemic sclerosis is connected with the circumstances of a person's relevant service:

- (1) inhaling respirable crystalline silica dust, at the time material containing crystalline silica was being:
 - (a) produced;
 - (b) excavated;
 - (c) drilled, cut or ground; or
 - (d) used in construction, manufacturing, cleaning or blasting,for a cumulative period of at least 5 000 hours before the clinical onset of systemic sclerosis;
- (2) being treated with bleomycin at the time of the clinical onset of systemic sclerosis;

- (3) inhaling, ingesting or having cutaneous contact with trichloroethylene for a cumulative period of at least 5 000 hours within a period of ten consecutive years, before the clinical onset of systemic sclerosis;
 - (4) inhaling respirable crystalline silica dust, at the time material containing crystalline silica was being:
 - (a) produced;
 - (b) excavated;
 - (c) drilled, cut or ground; or
 - (d) used in construction, manufacturing, cleaning or blasting,
 for a cumulative period of at least 5 000 hours before the clinical worsening of systemic sclerosis;
 - (5) being treated with bleomycin at the time of the clinical worsening of systemic sclerosis;
 - (6) inhaling, ingesting or having cutaneous contact with trichloroethylene for a cumulative period of at least 5 000 hours within a period of ten consecutive years, before the clinical worsening of systemic sclerosis;
 - (7) for clinical worsening of systemic sclerosis manifesting as scleroderma renal crisis only, being treated with 15 milligrams or more per day of prednisone, or equivalent oral glucocorticoid therapy, for a period of at least four weeks within the six months before the clinical worsening of systemic sclerosis;
- Note: *equivalent oral glucocorticoid therapy* and *scleroderma renal crisis* are defined in the Schedule 1 - Dictionary.
- (8) inability to obtain appropriate clinical management for systemic sclerosis.

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(4) to 9(8) apply only to material contribution to, or aggravation of, systemic sclerosis where the person's systemic sclerosis 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(3) 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:

equivalent oral glucocorticoid therapy means a glucocorticoid in the following table, at the doses specified in the table, or a therapeutically equivalent dose of another glucocorticoid:

Glucocorticoid	Minimum average rate (milligrams/day)
cortisone	75
hydrocortisone	60
prednisone	15
prednisolone	15

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

relevant service means:

- (a) eligible war service (other than operational service) under the VEA;
- (b) defence service (other than hazardous service and British nuclear test defence service) under the VEA; or
- (c) peacetime service under the MRCA.

Note: **MRCA** and **VEA** are also defined in the Schedule 1 - Dictionary.

scleroderma renal crisis means a disorder characterised by the abrupt onset of marked hypertension, rapidly progressive renal failure due to thrombotic microangiopathy vasospasm, and tissue ischaemia. Up to 10 percent of patients with scleroderma renal crisis are normotensive, although their blood pressure may be increased from baseline values.

systemic sclerosis—see subsection 7(2).

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

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

VEA means the *Veterans' Entitlements Act 1986*.