



Australian Government
Repatriation Medical Authority

Statement of Principles
concerning
SICKLE-CELL DISORDER
(Balance of Probabilities)
(No. 41 of 2017)

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

Dated 30 June 2017

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 *sickle-cell disorder (Balance of Probabilities)* (No. 41 of 2017).

2 Commencement

This instrument commences on 31 July 2017.

3 Authority

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

4 Revocation

The Statement of Principles concerning sickle-cell disorder No. 44 of 2008 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 sickle-cell disorder and death from sickle-cell disorder.

Meaning of sickle-cell disorder

- (2) For the purposes of this Statement of Principles, sickle-cell disorder means:
- (a) a genetic disorder leading to the production of haemoglobin S (HbS), a defective form of haemoglobin; and includes:
 - (b) sickle-cell disease, in which there are two copies of the HbS gene (HbSS); and
 - (c) sickle-cell trait, in which there is one copy of the HbS gene and one normal gene (HbSA); and
 - (d) double heterozygous sickling disorders, in which there is one copy of the HbS gene plus one copy of another β -globin gene variant.

Note 1: Sickle-cell disorder may involve symptomatic episodes in which red blood cells become deformed, leading to vaso-occlusive, anaemic or infectious events.

Note 2: The diagnosis can also be made in asymptomatic individuals by genetic testing.

- (3) While sickle-cell disorder attracts ICD-10-AM code D57, in applying this Statement of Principles the meaning of sickle-cell disorder 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 sickle-cell disorder

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

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 sickle-cell disorder and death from sickle-cell disorder can be related to relevant service rendered by veterans 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 exist before it can be said that, on the balance of probabilities, sickle-cell disorder or death from sickle-cell disorder is connected with the circumstances of a person's relevant service:

- (1) experiencing a stimulus from Specified List 1 of stimuli within the 24 hours before the clinical worsening of sickle-cell disorder;

Note: **Specified List 1 of stimuli** and **clinical worsening of sickle-cell disorder** are defined in the Schedule 1 - Dictionary.

- (2) experiencing a stimulus from Specified List 2 of stimuli within the seven days before the clinical worsening of sickle-cell disorder;

Note: **Specified List 2 of stimuli** and **clinical worsening of sickle-cell disorder** are defined in the Schedule 1 - Dictionary.

- (3) smoking at least one pack-year of cigarettes, or the equivalent thereof in other tobacco products, before the clinical worsening of sickle-cell disorder, and where smoking has ceased, the clinical worsening of sickle-cell disorder has occurred within six months of cessation;

Note: *pack-year of cigarettes, or the equivalent thereof in other tobacco products* and *clinical worsening of sickle-cell disorder* are defined in the Schedule 1 - Dictionary.

- (4) being exposed to second-hand smoke for at least 1 000 hours before the clinical worsening of sickle-cell disorder, and where the last exposure to second-hand smoke occurred within the six months before the clinical worsening of sickle-cell disorder;

Note: *being exposed to second-hand smoke* and *clinical worsening of sickle-cell disorder* are defined in the Schedule 1 - Dictionary.

- (5) being treated with corticosteroids, other than inhaled or topical corticosteroids, within the 30 days before the clinical worsening of sickle-cell disorder;

Note: *clinical worsening of sickle-cell disorder* is defined in the Schedule 1 - Dictionary.

- (6) inability to obtain appropriate clinical management for sickle-cell disorder.

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(1) to 9(6) apply only to material contribution to, or aggravation of, sickle-cell disorder where the person's sickle-cell disorder 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:

a change in ambient temperature means a decrease in the mean daily temperature from one day to the next day of at least 5 degrees Celsius.

being exposed to second-hand smoke means being in an enclosed space and inhaling smoke from burning tobacco products or smoke that has been exhaled by a person who is smoking.

clinical worsening of sickle-cell disorder means permanent worsening of sickle-cell disorder as evidenced by:

- (a) a sickle-cell crisis or series of crises resulting in end-organ damage or death; or
- (b) an increase in the requirement for management of the manifestations or complications of sickle-cell disorder.

Note: ***sickle-cell crisis*** is also defined in the Schedule 1 - Dictionary.

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

pack-year of cigarettes, or the equivalent thereof in other tobacco products means a calculation of consumption where one pack-year of cigarettes equals 20 tailor-made cigarettes per day for a period of one calendar year, or 7300 cigarettes. One tailor-made cigarette approximates one gram of tobacco or one gram of cigar or pipe tobacco by weight. One pack-year of tailor-made cigarettes equates to 7.3 kilograms of smoking tobacco by weight. Tobacco products mean cigarettes, pipe tobacco or cigars, smoked alone or in any combination.

puerperal period means the 42 days following a birth.

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.

sickle-cell crisis means a vaso-occlusive, anaemic or infectious complication of sickle-cell disorder requiring hospitalisation or management by a medical practitioner.

sickle-cell disorder—see subsection 7(2).

Specified List 1 of stimuli means:

- (a) altitudes above 1 500 metres, including flying;
- (b) dehydration;
- (c) sleep apnoea or other causes of hypoxia; or
- (d) strenuous exercise.

Specified List 2 of stimuli means:

- (a) a change in ambient temperature which results in skin cooling;
- (b) fever;
- (c) granulocyte colony-stimulating factors;
- (d) immersion in cold water;
- (e) infection;
- (f) iodinated radiological contrast agents;
- (g) pregnancy or puerperal period;
- (h) scuba diving; or
- (i) surgery.

Note: *a change in ambient temperature* and *puerperal period* are also defined in the Schedule 1 - Dictionary.

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*.