



**Australian Government**  
**Repatriation Medical Authority**

**Statement of Principles**  
**concerning**  
**SICKLE-CELL DISORDER**  
**(Reasonable Hypothesis)**  
**(No. 40 of 2017)**

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The Repatriation Medical Authority determines the following Statement of Principles under subsection 196B(2) 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 (Reasonable Hypothesis)* (No. 40 of 2017).

**2 Commencement**

This instrument commences on 31 July 2017.

**3 Authority**

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

**4 Revocation**

The Statement of Principles concerning sickle-cell disorder No. 43 of 2008 made under subsection 196B(2) of the VEA is 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 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  $\beta$ -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**

The Repatriation Medical Authority is of the view that there is sound medical-scientific evidence that indicates that sickle-cell disorder and death from sickle-cell disorder 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 sickle-cell disorder or death from sickle-cell disorder 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) having symptomatic asthma within the one year before the clinical worsening of sickle-cell disorder;

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

- (7) 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(7) 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(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:

***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) 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.

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*.