

Statement of Principles
concerning

SICKLE-CELL DISORDER

No. 44 of 2008

for the purposes of the

Veterans' Entitlements Act 1986
and

Military Rehabilitation and Compensation Act 2004

Title

1. This Instrument may be cited as Statement of Principles concerning sickle-cell disorder No. 44 of 2008.

Determination

2. The Repatriation Medical Authority under subsection **196B(3)** and **(8)** of the *Veterans' Entitlements Act 1986* (the VEA):
 - (a) revokes Instrument No. 110 of 1995, as amended by Instrument No. 194 of 1995, concerning sickle-cell disease; and
 - (b) determines in their place this Statement of Principles.

Kind of injury, disease or death

3.
 - (a) This Statement of Principles is about **sickle-cell disorder** and **death from sickle-cell disorder**.
 - (b) For the purposes of this Statement of Principles, "**sickle-cell disorder**" means a genetic disorder leading to the production of haemoglobin S (HbS), a defective form of haemoglobin. It is characterised by episodes in which red blood cells become deformed, leading to vaso-occlusive, anaemic or infectious crises. This definition includes sickle-cell disease, in which there are two copies of the HbS gene (HbSS); sickle-cell trait, in which there is one copy of the HbS gene and one normal gene (HbSA); and

double heterozygous sickling disorders, in which there is one copy of the HbS gene plus one copy of another β -globin gene variant.

- (c) Sickle-cell disorder attracts ICD-10-AM code D57.
- (d) In the application of this Statement of Principles, the definition of "**sickle-cell disorder**" is that given at paragraph 3(b) above.

Basis for determining the factors

- 4. 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 *Military Rehabilitation and Compensation Act 2004* (the MRCA).

Factors that must be related to service

- 5. Subject to clause 7, at least one of the factors set out in clause 6 must be related to the relevant service rendered by the person.

Factors

- 6. The factor that 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 is:
 - (a) for a first sickle-cell crisis only, experiencing a specified stimulus within the 24 hours before the clinical onset of sickle-cell disorder; or
 - (b) experiencing a specified stimulus within the 24 hours before the clinical worsening of sickle-cell disorder; or
 - (c) inability to obtain appropriate clinical management for sickle-cell disorder.

Factors that apply only to material contribution or aggravation

- 7. Paragraph **6(b) to 6(c)** 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 not arising out of) the person's relevant service.

Inclusion of Statements of Principles

8. In this Statement of Principles if a relevant factor applies and that factor includes an injury or disease in respect of which there is a Statement of Principles then the factors in that last mentioned Statement of Principles apply in accordance with the terms of that Statement of Principles as in force from time to time.

Other definitions

9. For the purposes of this Statement of Principles:

"a specified stimulus" means:

- (a) a change in ambient temperature which results in skin cooling;
- (b) altitudes above 1500 metres, including flying;
- (c) dehydration;
- (d) fever;
- (e) immersion in cold water;
- (f) infection;
- (g) pregnancy;
- (h) scuba diving;
- (i) sleep apnoea or other causes of hypoxia; or
- (j) strenuous exercise;

"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 significant end-organ damage or death; or
- (b) a substantial increase in the requirement for hospitalisation for management of the manifestations or complications of sickle-cell disorder;

"death from 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;

"ICD-10-AM code" means a number assigned to a particular kind of injury or disease in The International Statistical Classification of Diseases and Related Health Problems, 10th Revision, Australian Modification (ICD-10-AM), Sixth Edition, effective date of 1 July 2008, copyrighted by the National Centre for Classification in Health, Sydney, NSW, and having ISBN 978 1 74210 016 6;

"relevant service" means:

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

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

"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.

Application

- 10.** This Instrument applies to all matters to which section 120B of the VEA or section 339 of the MRCA applies.

Date of effect

- 11.** This Instrument takes effect from 2 July 2008.

Dated this *nineteenth* day of *June* 2008

The Common Seal of the)
Repatriation Medical Authority)
was affixed to this instrument)
in the presence of:)

KEN DONALD
CHAIRPERSON