

Statement of Principles concerning KAPOSI SARCOMA (Reasonable Hypothesis) (No. 1 of 2020)

The Repatriation Medical Authority determines the following Statement of Principles under subsection 196B(2) of the *Veterans' Entitlements Act 1986*.

Dated 20 December 2019

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

Professor Nicholas Saunders AO

Chairperson

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1 Name

This is the Statement of Principles concerning *Kaposi sarcoma (Reasonable Hypothesis)* (No. 1 of 2020).

2 Commencement

This instrument commences on 27 January 2020.

3 Authority

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

4 Repeal

The Statement of Principles concerning Kaposi's sarcoma No. 9 of 2011 (Federal Register of Legislation No. F2010L03266) made under subsection 196B(2) of the VEA is repealed.

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 Kaposi sarcoma and death from Kaposi sarcoma.

Meaning of Kaposi sarcoma

- (2) For the purposes of this Statement of Principles, Kaposi sarcoma:
 - (a) means a multicentric vascular neoplasm arising from endothelial cells and characterised by the proliferation of spindle cells, formation of new blood vessels and inflammation; and
 - (b) excludes soft tissue sarcoma.

Note: Typical clinical manifestations are coloured patches, plaques or nodules on the skin, although lesions may also involve the mucous membranes, lymph nodes and viscera.

(3) While Kaposi sarcoma attracts ICD-10-AM code C46, in applying this Statement of Principles the meaning of Kaposi sarcoma 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 Kaposi sarcoma

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

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 Kaposi sarcoma and death from Kaposi sarcoma 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: 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 as a minimum exist before it can be said that a reasonable hypothesis has been raised connecting Kaposi sarcoma or death from Kaposi sarcoma with the circumstances of a person's relevant service:

- (1) having infection with human herpesvirus type 8 before the clinical onset of Kaposi sarcoma;
- (2) having infection with human immunodeficiency virus before the clinical onset of Kaposi sarcoma;
- (3) having non-Hodgkin lymphoma or chronic lymphocytic leukaemia/small lymphocytic lymphoma before the clinical onset of Kaposi sarcoma;
- (4) having chronic renal failure at least six months before the clinical onset of Kaposi sarcoma;
 - Note: *chronic renal failure* is defined in the Schedule 1 Dictionary.
- (5) having an autoimmune disease from the specified list of autoimmune diseases at least six months before the clinical onset of Kaposi sarcoma;

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

- (6) having diabetes mellitus for at least six months before the clinical onset of Kaposi sarcoma;
- (7) having lymphoedema involving the affected site at least five years before the clinical onset of Kaposi sarcoma;
- (8) undergoing solid organ, stem cell or bone marrow transplantation before the clinical onset of Kaposi sarcoma;
- (9) being treated with an immunosuppressive drug:
 - (a) for a continuous period of at least three months before the clinical onset of Kaposi sarcoma; and
 - (b) where that exposure has ceased, the clinical onset of Kaposi sarcoma has occurred within ten years of cessation;

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

- (10) being treated with an angiotensin converting enzyme inhibitor:
 - (a) for at least three months before the clinical onset of Kaposi sarcoma; and
 - (b) where that exposure has ceased, the clinical onset of Kaposi sarcoma has occurred within ten years of cessation;
- (11) inability to obtain appropriate clinical management for Kaposi sarcoma.

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 factor set out in subsection 9(11) applies only to material contribution to, or aggravation of, Kaposi sarcoma where the person's Kaposi sarcoma 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:

chronic renal failure means:

- (a) having a glomerular filtration rate of less than 15 mL/min/1.73 m² for a period of at least three months; or
- (b) a need for renal replacement therapy (dialysis or transplantation) for treatment of complications of decreased glomerular filtration rate which would otherwise increase the risk of morbidity and mortality; or
- (c) undergoing chronic dialysis.

immunosuppressive drug means a drug or an agent which results in substantial suppression of immune responses.

Note: Examples of an immunosuppressive drug include, but are not limited to:

- (a) chemotherapeutic agents used for the treatment of cancer;
- (b) corticosteroids, other than inhaled or topical corticosteroids;
- (c) drugs used to prevent transplant rejection; and
- (d) tumour necrosis factor-α inhibitors.

Kaposi sarcoma—see subsection 7(2).

MRCA 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.

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

specified list of autoimmune diseases means:

- (a) Addison disease;
- (b) autoimmune haemolytic anaemia;
- (c) Behcet disease;
- (d) granulomatosis with polyangiitis (Wegener granulomatosis);
- (e) myasthenia gravis;
- (f) pemphigoid;
- (g) pemphigus;
- (h) polyarteritis nodosa;
- (i) polymyalgia rheumatica;
- (j) psoriasis;
- (k) rheumatoid arthritis;
- (1) systemic lupus erythematosus;
- (m) systemic or localised scleroderma; or

(n) ulcerative colitis.

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.