



Australian Government
Repatriation Medical Authority

Statement of Principles
concerning
CREUTZFELDT-JAKOB DISEASE
(Reasonable Hypothesis)
(No. 80 of 2022)

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

Dated 23 August 2022

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

Professor Terence Campbell AM
Chairperson

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1 Name

This is the Statement of Principles concerning *Creutzfeldt-Jakob disease (Reasonable Hypothesis)* (No. 80 of 2022).

2 Commencement

This instrument commences on 19 September 2022.

3 Authority

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

4 Repeal

The Statement of Principles concerning Creutzfeldt-Jakob disease No. 76 of 2014 (Federal Register of Legislation No. F2014L01138) made under subsection 196B(2) of the VEA is repealed.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 Creutzfeldt-Jakob disease and death from Creutzfeldt-Jakob disease.

Meaning of Creutzfeldt-Jakob disease

- (2) For the purposes of this Statement of Principles, Creutzfeldt-Jakob disease:

- (a) means a spongiform encephalopathy characterised by an accumulation of abnormal prion protein in the brain; and
- (b) includes sporadic (classical) and variant Creutzfeldt-Jakob disease;

Note 1: Creutzfeldt-Jakob disease, also known as CJD, is characterised by rapidly progressive dementia, myoclonus, and motor disturbances.

Note 2: *variant Creutzfeldt-Jakob disease* is defined in the Schedule 1 – Dictionary.

- (3) While Creutzfeldt-Jakob disease attracts ICD-10-AM code A81.0, in applying this Statement of Principles the meaning of Creutzfeldt-Jakob disease 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 Creutzfeldt-Jakob disease

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

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 Creutzfeldt-Jakob disease and death from Creutzfeldt-Jakob disease 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 Creutzfeldt-Jakob disease or death from Creutzfeldt-Jakob disease with the circumstances of a person's relevant service:

- (1) receiving treatment involving a human tissue or product as specified at least 6 months before the clinical onset of Creutzfeldt-Jakob disease;

Note: *human tissue or product as specified* is defined in the Schedule 1 – Dictionary.

- (2) undergoing major surgery that requires general, spinal or epidural anaesthesia at least 6 months before the clinical onset of Creutzfeldt-Jakob disease;

Note: Major surgery that requires general, spinal or epidural anaesthesia includes neurosurgery, obstetric, gynaecological, or gastrointestinal surgical procedures. It does not include minor surgery such as needle aspiration or biopsy, superficial incision or endoscopy.

- (3) for variant Creutzfeldt-Jakob disease only:

- (a) receiving blood or blood products from a person infected with variant Creutzfeldt-Jakob disease at least 3 years before the clinical onset of Creutzfeldt-Jakob disease; or
- (b) consuming beef or a beef product from an area or country with evidence of bovine spongiform encephalopathy infection in cattle at the time of consumption, where the beef or beef product was consumed at least 3 years before the clinical onset of Creutzfeldt-Jakob disease; or
- (c) having percutaneous exposure to blood or tissue from an animal infected with bovine spongiform encephalopathy at least 3 years before the clinical onset of Creutzfeldt-Jakob disease.

Note 1: Consuming beef or beef product contaminated with bovine spongiform encephalopathy has occurred in countries without domestic bovine spongiform encephalopathy that have imported beef or beef product from a country with bovine spongiform encephalopathy.

Note 2: *variant Creutzfeldt-Jakob disease, blood products, beef product, and bovine spongiform encephalopathy* are defined in the Schedule 1 – Dictionary.

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.

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:

beef product means food prepared from or containing beef.

blood products means biopharmaceutical products made from human blood.

bovine spongiform encephalopathy, also known as ‘mad cow disease’, means a neurodegenerative disease of cattle caused by consumption of prion-contaminated meat or bone meal.

Creutzfeldt-Jakob disease—see subsection 7(2).

human tissue or product as specified means:

- (a) a corneal transplant;
- (b) a liver transplant;
- (c) an albumin infusion;
- (d) cadaveric human pituitary hormones; or
- (e) human dura mater as a graft or in radiographic embolisation procedures.

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.

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.

variant Creutzfeldt-Jakob disease means a form of Creutzfeldt-Jakob disease which is characterised by a younger age at onset, a more protracted clinical course, more sensory disturbances and more psychiatric symptoms than sporadic (classical) Creutzfeldt-Jakob disease, as well as a marked accumulation of abnormal prion protein and florid amyloid plaques in the brain tissue.

VEA means the Veterans' Entitlements Act 1986.