### **Revocation and Determination**

of

# Statement of Principles

# PORPHYRIA CUTANEA TARDA

#### ICD-10-AM CODE: E80.1

## Veterans' Entitlements Act 1986

- 1. The Repatriation Medical Authority under subsection **196B(3)** of the *Veterans' Entitlements Act 1986* (the Act):
  - (a) revokes Instrument No.72 of 1994; and
  - (b) determines in its place the following Statement of Principles.

#### Kind of injury, disease or death

- 2. (a) This Statement of Principles is about porphyria cutanea tarda and death from porphyria cutanea tarda.
  - (b) For the purposes of this Statement of Principles, "**porphyria cutanea tarda**" means a disorder of porphyrin metabolism characterised by cutaneous photosensitivity, hyperpigmentation, hypertrichosis, deficiency of uroporphyrinogen decarboxylase and uroporphyria, attracting ICD-10-AM code E80.1.

#### **Basis for determining the factors**

**3.** On the sound medical-scientific evidence available, the Repatriation Medical Authority is of the view that it is more probable than not that **porphyria cutanea tarda and death from porphyria cutanea tarda** can be related to relevant service rendered by veterans or members of the Forces.

#### Factors that must be related to service

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

#### Factors

- 5. The factors that must exist before it can be said that, on the balance of probabilities, **porphyria cutanea tarda** or **death from porphyria cutanea tarda** is connected with the circumstances of a person's relevant service are:
  - (a) inhaling, ingesting or cutaneously absorbing a halogenated aromatic hydrocarbon within the one year immediately before the clinical onset of porphyria cutanea tarda; or
  - (b) suffering from alcoholic liver disease at the time of the clinical onset of porphyria cutanea tarda; or
  - (c) suffering from hepatitis at the time of the clinical onset of porphyria cutanea tarda; or
  - (d) suffering from cirrhosis of the liver before the clinical onset of porphyria cutanea tarda; or
  - (e) being infected with Human Immunodeficiency Virus (HIV) at the time of the clinical onset of porphyria cutanea tarda; or
  - (f) undergoing a course of oral oestrogen therapy for the 30 days immediately before the clinical onset of porphyria cutanea tarda; or
  - (g) suffering from hepatic haemosiderosis at the time of the clinical onset of porphyria cutanea tarda; or
  - (h) suffering from haemochromatosis at the time of the clinical onset of porphyria cutanea tarda; or
  - (j) undergoing haemodialysis for the one year immediately before the clinical onset of porphyria cutanea tarda; or
  - (k) suffering from a porphyrin-generating hepatocellular tumour at the time of the clinical onset of porphyria cutanea tarda; or

- (m) inhaling, ingesting or cutaneously absorbing a halogenated aromatic hydrocarbon within the one year immediately before the clinical worsening of porphyria cutanea tarda; or
- (n) suffering from alcoholic liver disease at the time of the clinical worsening of porphyria cutanea tarda; or
- (o) suffering from hepatitis at the time of the clinical worsening of porphyria cutanea tarda; or
- (p) suffering from cirrhosis of the liver before the clinical worsening of porphyria cutanea tarda; or
- (q) being infected with Human Immunodeficiency Virus (HIV) at the time of the clinical worsening of porphyria cutanea tarda; or
- (r) undergoing a course of oral oestrogen therapy for the 30 days immediately before the clinical worsening of porphyria cutanea tarda; or
- (s) suffering from hepatic haemosiderosis at the time of the clinical worsening of porphyria cutanea tarda; or
- (t) suffering from haemochromatosis at the time of the clinical worsening of porphyria cutanea tarda; or
- (u) exposing the affected area of skin to sunlight within the five days immediately before the clinical worsening of porphyria cutanea tarda; or
- (v) being treated with a drug or a class of drugs from the specified list, which cannot be ceased or substituted, at the time of the clinical worsening of porphyria cutanea tarda; or
- (w) inability to obtain appropriate clinical management for porphyria cutanea tarda.

#### Factors that apply only to material contribution or aggravation

6. Paragraphs 5(m) to 5(w) apply only to material contribution to, or aggravation of, porphyria cutanea tarda where the person's porphyria cutanea tarda was suffered or contracted before or during (but not arising out of) the person's relevant service; paragraph 8(1)(e), 9(1)(e) or 70(5)(d) of the Act refers.

#### **Inclusion of Statements of Principles**

7. 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.

#### **Other definitions**

8. For the purposes of this Statement of Principles:

**"being infected with Human Immunodeficiency Virus (HIV)"** means serological evidence of infection with Human Immunodeficiency Virus;

**"being treated with a drug or class of drugs from the specified list"** means therapeutic administration of one of the following drugs or class of drugs:

- (a) Barbiturates;
- (b) Busulfan;
- (c) Chloroquine;
- (d) Cyclophosphamide;
- (e) Dapsone;
- (f) Fluconazole;

- (g) Griseofulvin;
- (h) Hydroxychloroquine;
- (i) Iron supplements
- (j) Rifampicin;
- (k) Sulfadoxine-pyrimethamine;
- (l) Sulphonamides;

"cirrhosis of the liver" means a pathologically defined entity involving irreversible chronic injury of the hepatic parenchyma and includes extensive fibrosis in association with regenerative nodules;

"death from porphyria cutanea tarda" in relation to a person includes death from a terminal event or condition that was contributed to by the person's porphyria cutanea tarda;

"haemochromatosis" means a genetic disorder of iron storage in which an inappropriate increase in intestinal iron absorption results in the deposition of excessive quantities of iron in parenchymal cells, with eventual tissue damage and functional impairment of the organs involved, especially the liver, pancreas, heart and pituitary;

**"halogenated aromatic hydrocarbon"** means a chemical compound containing a benzene ring and multiple chlorine atoms. Halogenated aromatic hydrocarbon compounds include: dioxins, furans, polychlorinated biphenyls, hexachlorobenzene, DDT, chlordane, mirex, toxaphene or heptachlor;

**"hepatic haemosiderosis"** means the deposit of an abnormal quantity of haemosiderin in the liver, usually in Kupffer cells;

"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), Second Edition, effective date of 1 July 2000, copyrighted by the National Centre for Classification in Health, Sydney, NSW, and having ISBN 1 86487 271 3;

"porphyrin-generating hepatocellular tumour" means a tumour arising within the cells of the liver, with evidence of increased porphyrin production at the tumour site;

#### "relevant service" means:

- eligible war service (other than operational service); or (a)
- defence service (other than hazardous service); (b)

"terminal event" means the proximate or ultimate cause of death and includes:

- (a) pneumonia;
- respiratory failure; (b)
- cardiac arrest; (c)
- circulatory failure; or (d)
- cessation of brain function. (e)

#### Application

9. This Instrument applies to all matters to which section 120B of the Act applies.

Dated	this
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Nineteenth day of March

2001

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