



Australian Government
Repatriation Medical Authority

Statement of Principles
concerning
PORPHYRIA CUTANEA TARDA
(Reasonable Hypothesis)
(No. 69 of 2021)

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

Dated 21 May 2021

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 *porphyria cutanea tarda* (*Reasonable Hypothesis*) (No. 69 of 2021).

2 Commencement

This instrument commences on 21 June 2021.

3 Authority

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

4 Repeal

The Statement of Principles concerning porphyria cutanea tarda No. 43 of 2012 (Federal Register of Legislation No. F2012L01357) made under subsection 196B(2) of the VEA is repealed.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 porphyria cutanea tarda and death from porphyria cutanea tarda.

Meaning of porphyria cutanea tarda

- (2) For the purposes of this Statement of Principles, porphyria cutanea tarda means a skin disorder resulting from decreased activity of the enzyme uroporphyrinogen decarboxylase in the liver and accumulation of porphyrins in the circulation.

Note: Typical clinical manifestations of porphyria cutanea tarda include blistering of the skin in sun-exposed areas, hyperpigmentation and facial hypertrichosis.

- (3) While porphyria cutanea tarda attracts ICD-10-AM code E80.1, in applying this Statement of Principles the meaning of porphyria cutanea tarda 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 porphyria cutanea tarda

- (5) For the purposes of this Statement of Principles, 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.

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 porphyria cutanea tarda and death from porphyria cutanea tarda 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 porphyria cutanea tarda or death from porphyria cutanea tarda with the circumstances of a person's relevant service:

- (1) for males, consuming a total of at least 100 kilograms of alcohol within the 10 years before the clinical onset of porphyria cutanea tarda;

Note: Alcohol consumption is calculated utilising the Australian Standard of 10 grams of alcohol per standard alcoholic drink.

- (2) for females, consuming a total of at least 50 kilograms of alcohol within the 10 years before the clinical onset of porphyria cutanea tarda;

Note: Alcohol consumption is calculated utilising the Australian Standard of 10 grams of alcohol per standard alcoholic drink.

- (3) having alcohol use disorder at the time of the clinical onset of porphyria cutanea tarda;

- (4) having cirrhosis of the liver or steatohepatitis at the time of the clinical onset of porphyria cutanea tarda;

- (5) having chronic renal failure at the time of the clinical onset of porphyria cutanea tarda;

Note: *chronic renal failure* is defined in the Schedule 1 - Dictionary.

- (6) having hepatic iron overload at the time of the clinical onset of porphyria cutanea tarda;
- Note: *iron overload* is defined in the Schedule 1 - Dictionary.
- (7) having a viral infection from the specified list of viral infections at the time of the clinical onset of porphyria cutanea tarda;
- Note: *specified list of viral infections* is defined in the Schedule 1 - Dictionary.
- (8) taking a drug from the specified list of drugs within the 30 days before the clinical onset of porphyria cutanea tarda;
- Note: *specified list of drugs* is defined in the Schedule 1 - Dictionary.
- (9) taking a drug which is associated in the individual with the clinical onset of porphyria cutanea tarda during drug therapy and either:
- (a) the improvement of porphyria cutanea tarda within 2 months of discontinuing or tapering drug therapy; or
 - (b) the redevelopment of porphyria cutanea tarda on rechallenge with the same drug; and
- where taking the drug continued for at least the 7 days before the clinical onset of porphyria cutanea tarda;
- (10) inhaling, ingesting or having cutaneous contact with a chemical agent contaminated by 2,3,7,8-tetrachlorodibenzo-para-dioxin (TCDD) within the 1 year before the clinical onset of porphyria cutanea tarda;
- Note: *inhaling, ingesting or having cutaneous contact with a chemical agent contaminated by 2,3,7,8-tetrachlorodibenzo-para-dioxin (TCDD)* is defined in the Schedule 1 - Dictionary.
- (11) ingesting food contaminated with hexachlorobenzene within the 1 year before the clinical onset of porphyria cutanea tarda;
- (12) having smoked tobacco products:
- (a) in an amount of at least 10 cigarettes per day or the equivalent thereof in other tobacco products; and
 - (b) for at least the 6 months before the clinical onset of porphyria cutanea tarda; and
- if smoking has ceased before the clinical onset of porphyria cutanea tarda, then that onset occurred within 30 days of cessation;
- Note: *cigarettes per day or the equivalent thereof in other tobacco products* is defined in the Schedule 1 - Dictionary.
- (13) for males, consuming a total of at least 100 kilograms of alcohol within the 10 years before the clinical worsening of porphyria cutanea tarda;
- Note: Alcohol consumption is calculated utilising the Australian Standard of 10 grams of alcohol per standard alcoholic drink.

- (14) for females, consuming a total of at least 50 kilograms of alcohol within the 10 years before the clinical worsening of porphyria cutanea tarda;

Note: Alcohol consumption is calculated utilising the Australian Standard of 10 grams of alcohol per standard alcoholic drink.

- (15) having alcohol use disorder at the time of the clinical worsening of porphyria cutanea tarda;
- (16) having cirrhosis of the liver or steatohepatitis at the time of the clinical worsening of porphyria cutanea tarda;
- (17) having chronic renal failure at the time of the clinical worsening of porphyria cutanea tarda;

Note: *chronic renal failure* is defined in the Schedule 1 - Dictionary.

- (18) having hepatic iron overload at the time of the clinical worsening of porphyria cutanea tarda;

Note: *iron overload* is defined in the Schedule 1 - Dictionary.

- (19) having a viral infection from the specified list of viral infections at the time of the clinical worsening of porphyria cutanea tarda;

Note: *specified list of viral infections* is defined in the Schedule 1 - Dictionary.

- (20) taking a drug from the specified list of drugs within the 30 days before the clinical worsening of porphyria cutanea tarda;

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

- (21) taking a drug which is associated in the individual with:

- (a) the clinical worsening of porphyria cutanea tarda during drug therapy; and
- (b) the improvement of porphyria cutanea tarda within 2 months of discontinuing or tapering drug therapy; and

where taking the drug continued for at least the 7 days before the clinical worsening of porphyria cutanea tarda;

- (22) inhaling, ingesting or having cutaneous contact with a chemical agent contaminated by 2,3,7,8-tetrachlorodibenzo-para-dioxin (TCDD) within the 1 year before the clinical worsening of porphyria cutanea tarda;

Note: *inhaling, ingesting or having cutaneous contact with a chemical agent contaminated by 2,3,7,8-tetrachlorodibenzo-para-dioxin (TCDD)* is defined in the Schedule 1 - Dictionary.

- (23) ingesting food contaminated with hexachlorobenzene within the 1 year before the clinical worsening of porphyria cutanea tarda;

- (24) having smoked tobacco products:
- (a) in an amount of at least 10 cigarettes per day or the equivalent thereof in other tobacco products; and
 - (b) for least 6 months before the clinical worsening of porphyria cutanea tarda; and

if smoking has ceased before the clinical worsening of porphyria cutanea tarda, then that worsening occurred within 30 days of cessation;

Note: *cigarettes per day or the equivalent thereof in other tobacco products* is defined in the Schedule 1 - Dictionary.

- (25) inability to obtain appropriate clinical management for porphyria cutanea tarda.

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(13) to 9(25) 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 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 3 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.

cigarettes per day or the equivalent thereof in other tobacco products means:

- (a) cigarettes, pipe tobacco or cigars, alone or in any combination; and
- (b) 1 gram of cigar, pipe or other smoking tobacco (including roll your own smoking tobacco) is equivalent to one tailor made cigarette.

inhaling, ingesting or having cutaneous contact with a chemical agent contaminated by 2,3,7,8-tetrachlorodibenzo-para-dioxin (TCDD) means:

- (a) decanting or spraying;
- (b) cleaning or maintaining equipment used to apply;
- (c) being sprayed with;
- (d) handling or sawing timber treated with;
- (e) being in an environment shrouded in dust from timber treated with; or
- (f) using cutting oils contaminated with;

one of the following chemicals:

- (i) 2,4,5-trichlorophenoxyacetic acid;
- (ii) 2,4,5-trichlorophenoxypropionic acid;
- (iii) 2,4,5-trichlorophenol;
- (iv) 2-(2,4,5-trichlorophenoxy)-ethyl 2,2-dichloropropionate;
- (v) o,o-dimethyl-o-(2,4,5-trichlorophenyl)-phosphorothioate;
- (vi) pentachlorophenol;
- (vii) 2,3,4,6-tetrachlorophenol;
- (viii) 2,4,6-trichlorophenol;
- (ix) 1,3,5-trichloro-2-(4-nitrophenoxy)benzene;
- (x) 2,4-dichloro-1-(4-nitrophenoxy)benzene; or
- (xi) 2,4-dichloro-1-(3-methoxy-4-nitrophenoxy)-benzene.

iron overload means an accumulation of excess iron in tissues and organs which has been confirmed by elevated ferritin or transferrin saturation levels.

Note: Common causes of hepatic iron overload are haemochromatosis and blood transfusions.

MRCAs means the *Military Rehabilitation and Compensation Act 2004*.

porphyria cutanea tarda—see subsection 7(2).

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 drugs means:

- (a) carbamazepine;
- (b) chloroquine;
- (c) cyclophosphamide;
- (d) highly active antiretroviral therapy;
- (e) hydroxychloroquine;
- (f) imatinib;
- (g) interferons;
- (h) iron supplements;
- (i) olmesartan;
- (j) oral estrogens;
- (k) phenobarbital;
- (l) phenytoin;
- (m) ribavirin;
- (n) rifampicin;
- (o) sulfonyleureas; or
- (p) tamoxifen.

specified list of viral infections means:

- (a) hepatitis B virus;
- (b) hepatitis C virus; or
- (c) human immunodeficiency virus.

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*.