



Australian Government
Repatriation Medical Authority

Statement of Principles concerning

PORPHYRIA CUTANEA TARDA

No. 43 of 2012

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 porphyria cutanea tarda No. 43 of 2012.

Determination

2. The Repatriation Medical Authority under subsection **196B(2)** and **(8)** of the *Veterans' Entitlements Act 1986* (the VEA):
 - (a) revokes Instrument No. 19 of 2001 concerning porphyria cutanea tarda; and
 - (b) determines in its place this Statement of Principles.

Kind of injury, disease or death

3.
 - (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 chronic disorder of porphyrin metabolism due to uroporphyrinogen decarboxylase deficiency, characterised by uroporphyrin, cutaneous photosensitivity, hyperpigmentation and facial hypertrichosis.
 - (c) Porphyria cutanea tarda attracts ICD-10-AM code E80.1.

- (d) In the application of this Statement of Principles, the definition of "**porphyria cutanea tarda**" is that given at paragraph 3(b) above.

Basis for determining the factors

4. 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 *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 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 is:
- (a) having exposure to a halogenated aromatic hydrocarbon as specified within the two years before the clinical onset of porphyria cutanea tarda; or
 - (b) having exposure as specified to a chemical agent contaminated with 2,3,7,8-tetrachlorodibenzo-para-dioxin (TCDD) within the two years before the clinical onset of porphyria cutanea tarda; or
 - (c) for males, consuming a total of 75 kilograms of alcohol within the five years before the clinical onset of porphyria cutanea tarda; or
 - (d) for females, consuming a total of 37 kilograms of alcohol within the five years before the clinical onset of porphyria cutanea tarda; or
 - (e) having a liver disease from the specified list at the time of the clinical onset of porphyria cutanea tarda; or
 - (f) being infected with human immunodeficiency virus before the clinical onset of porphyria cutanea tarda; or
 - (g) taking a course of oral oestrogen therapy for the 30 days before the clinical onset of porphyria cutanea tarda; or
 - (h) having hepatic iron overload at the time of the clinical onset of porphyria cutanea tarda; or
 - (i) undergoing haemodialysis or peritoneal dialysis for the six months before the clinical onset of porphyria cutanea tarda; or
 - (j) having a porphyrin-generating hepatocellular tumour at the time of the clinical onset of porphyria cutanea tarda; or

- (k) smoking at least ten cigarettes per day, or the equivalent thereof in other tobacco products, for the six months before the clinical onset of porphyria cutanea tarda, and where smoking has ceased, the clinical onset of porphyria cutanea tarda has occurred within one month of cessation; or
- (l) having the affected area of skin exposed to sunlight or ultraviolet light within the five days before the clinical onset of porphyria cutanea tarda; or
- (m) being treated with a drug or a drug from a class of drugs from the specified list, at the time of the clinical onset of porphyria cutanea tarda; or
- (n) being treated with a drug which is associated in the individual with:
 - (i) the development of porphyria cutanea tarda during drug therapy; and either
 - (ii) the improvement of porphyria cutanea tarda within two months of discontinuing or tapering drug therapy; or
 - (iii) the redevelopment of porphyria cutanea tarda on rechallenge with the same drug;

where treatment with the drug continued for at least the seven days before the clinical onset of porphyria cutanea tarda; or
- (o) having exposure to a halogenated aromatic hydrocarbon as specified within the two years before the clinical worsening of porphyria cutanea tarda; or
- (p) having exposure as specified to a chemical agent contaminated with 2,3,7,8-tetrachlorodibenzo-para-dioxin (TCDD) within the two years before the clinical worsening of porphyria cutanea tarda; or
- (q) for males, consuming a total of 75 kilograms of alcohol within the five years before the clinical worsening of porphyria cutanea tarda; or
- (r) for females, consuming a total of 37 kilograms of alcohol within the five years before the clinical worsening of porphyria cutanea tarda; or
- (s) having a liver disease from the specified list at the time of the clinical worsening of porphyria cutanea tarda; or
- (t) being infected with human immunodeficiency virus before the clinical worsening of porphyria cutanea tarda; or
- (u) taking a course of oral oestrogen therapy for the 30 days before the clinical worsening of porphyria cutanea tarda; or
- (v) having hepatic iron overload at the time of the clinical worsening of porphyria cutanea tarda; or
- (w) undergoing haemodialysis or peritoneal dialysis for the six months before the clinical worsening of porphyria cutanea tarda; or
- (x) having a porphyrin-generating hepatocellular tumour at the time of the clinical worsening of porphyria cutanea tarda; or

- (y) smoking at least ten cigarettes per day, or the equivalent thereof in other tobacco products, for the six months before the clinical worsening of porphyria cutanea tarda, and where smoking has ceased, the clinical worsening of porphyria cutanea tarda has occurred within one month of cessation; or
- (z) having the affected area of skin exposed to sunlight or ultraviolet light within the five days before the clinical worsening of porphyria cutanea tarda; or
- (aa) being treated with a drug or a drug from a class of drugs from the specified list, at the time of the clinical worsening of porphyria cutanea tarda; or
- (bb) being treated with a drug which is associated in the individual with:
 - (i) the worsening of porphyria cutanea tarda during drug therapy; and either
 - (ii) the improvement of porphyria cutanea tarda within two months of discontinuing or tapering drug therapy; or
 - (iii) the worsening of porphyria cutanea tarda on rechallenge with the same drug;

where treatment with the drug continued for at least the seven days before the clinical worsening of porphyria cutanea tarda; or
- (cc) inability to obtain appropriate clinical management for porphyria cutanea tarda.

Factors that apply only to material contribution or aggravation

7. Paragraphs **6(o) to 6(cc)** 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.

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 drug or a drug from a class of drugs from the specified list" means:
 - (a) alpha-methyldopa;
 - (b) amphetamines;
 - (c) barbiturates;
 - (d) busulfan;
 - (e) carbamazepine;

- (f) chloroquine;
- (g) colchicine;
- (h) cyclophosphamide;
- (i) dapsone;
- (j) diclofenac;
- (k) ergot derivatives;
- (l) erythromycin;
- (m) fluconazole;
- (n) frusemide;
- (o) griseofulvin;
- (p) hydantoins;
- (q) hydralazine;
- (r) hydroxychloroquine;
- (s) imatinib mesylate;
- (t) interferon alpha;
- (u) iron supplements;
- (v) lignocaine;
- (w) metronidazole;
- (x) nalidixic acid;
- (y) phenylbutazone;
- (z) phenytoin
- (aa) ribavirin;
- (bb) rifampicin;
- (cc) sulfadoxine-pyrimethamine;
- (dd) sulphonamides;
- (ee) tamoxifen;
- (ff) tetracyclines;
- (gg) theophylline; or
- (hh) valproic acid;

"a halogenated aromatic hydrocarbon from the specified list" means:

- (a) cacodylic acid;
- (b) hexachlorobenzene; or
- (c) picloram;

"a liver disease from the specified list" means:

- (a) cirrhosis of the liver;
- (b) steatohepatitis; or
- (c) viral hepatitis;

"a specified chemical agent" means one of the following chemicals:

- (a) 2,4,5-trichlorophenoxyacetic acid;
- (b) 2,4,5-trichlorophenoxypropionic acid;
- (c) 2,4,5-trichlorophenol;
- (d) 2-(2,4,5-trichlorophenoxy)-ethyl 2,2-dichloropropionate;
- (e) o,o-dimethyl-o-(2,4,5-trichlorophenyl)-phosphorothioate;
- (f) 2,3,4,6-tetrachlorophenol;
- (g) 2,4,6-trichlorophenol;

- (h) 1,3,4-trichloro-2-(4-nitrophenoxy)benzene;
- (i) 2,4-dichloro-1-(4-nitrophenoxy)benzene; or
- (j) 2,4-dichloro-1-(3-methoxy-4-nitrophenoxy)-benzene;

"alcohol" is measured by the alcohol consumption calculations utilising the Australian Standard of 10 grams of alcohol per standard alcoholic drink;

"cigarettes per day or the equivalent thereof in other tobacco products" means either cigarettes, pipe tobacco or cigars, alone or in any combination where one tailor made cigarette approximates one gram of tobacco; or one gram of cigar, pipe or other smoking tobacco;

"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;

"having exposure as specified to a chemical agent contaminated with 2,3,7,8-tetrachlorodibenzo-para-dioxin (TCDD)" means:

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

"having exposure to a halogenated aromatic hydrocarbon as specified" means:

- (a) being sprayed with a halogenated aromatic hydrocarbon from the specified list;
- (b) cleaning or maintaining equipment used to apply a halogenated aromatic hydrocarbon from the specified list;
- (c) decanting or spraying a halogenated aromatic hydrocarbon from the specified list; or
- (d) ingesting food contaminated with a halogenated aromatic hydrocarbon from the specified list;

"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), Seventh Edition, effective date of 1 July 2010, copyrighted by the National Centre for Classification in Health, Sydney, NSW, and having ISBN 978 1 74210 154 5;

"iron overload" means an accumulation of excess iron in tissues and organs which has been confirmed by elevated ferritin or transferrin saturation levels. Causes include haemochromatosis or blood transfusions;

"porphyrin-generating hepatocellular tumour" means a tumour arising from hepatocellular tissue, with evidence of increased porphyrin production at the tumour site;

"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;

"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 120A of the VEA or section 338 of the MRCA applies.

Date of effect

- 11.** This Instrument takes effect from 11 July 2012.

Dated this *twenty-first* day of *June* 2012

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

KEN DONALD
CHAIRPERSON