Australian Government
Repatriation Medical Authority

Statement of Principles
concerning
PURE RED CELL APLASIA
(Balance of Probabilities)
(No. 61 of 2020)

The Repatriation Medical Authority determines the following Statement of Principles under subsection 196B(3) of the *Veterans' Entitlements Act 1986*.

Dated 28 August 2020

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

Professor Nicholas Saunders AO
Chairperson

Contents

1 Name

This is the Statement of Principles concerning *pure red cell aplasia (Balance of Probabilities)* (No. 61 of 2020).

2 Commencement

This instrument commences on 28 September 2020.

3 Authority

This instrument is made under subsection 196B(3) of the *Veterans' Entitlements Act 1986*.

4 Repeal

The Statement of Principles concerning aplastic anaemia No. 51 of 2012 (Federal Register of Legislation No. F2012L01793) made under subsections 196B(3) and (8) of the VEA is repealed.

5 Application

This instrument applies to a claim to which section 120B of the VEA or section 339 of the *Military Rehabilitation and Compensation Act 2004* applies.

6 Schedules

Any item in a Schedule to this Instrument has effect according to its terms.

7 Kind of injury, disease or death to which this Statement of Principles relates

(1) This Statement of Principles is about pure red cell aplasia and death from pure red cell aplasia.

*Meaning of **pure red cell aplasia***

(2) For the purposes of this Statement of Principles, pure red cell aplasia:

(a) means complete or nearly complete cessation of red cell production in the bone marrow without effects on other haematopoietic cells and characterised by anaemia, reticulocytopaenia and absent or rare erythroid precursor cells in the bone marrow; and

(b) excludes:

(i) congenital Diamond-Blackfan anaemia;
(ii) myelodysplastic syndrome; and
(iii) paroxysmal nocturnal haemoglobinuria.

(3) While pure red cell aplasia attracts ICD-10-AM code D60, in applying this Statement of Principles the meaning of pure red cell aplasia 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 ***pure red cell aplasia***

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

Note: ***terminal event*** is defined in the Schedule 1 - Dictionary.

8 Basis for determining the factors

On the sound medical-scientific evidence available, the Repatriation Medical Authority is of the view that it is more probable than not that pure red cell aplasia and death from pure red cell aplasia can be related to relevant service rendered by veterans 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 exist before it can be said that, on the balance of probabilities, pure red cell aplasia or death from pure red cell aplasia is connected with the circumstances of a person's relevant service:

(1) being pregnant at the time of the clinical onset of pure red cell aplasia;

(2) being treated with a drug specified in the Schedule 2 - Drugs of this Instrument within the six months before the clinical onset of pure red cell aplasia;

(3) being treated with a drug which is associated in the individual with:

  (a) the development of pure red cell aplasia within six months of drug therapy; and
  (b) the improvement of pure red cell aplasia within six months of discontinuing or tapering drug therapy;

(4) being exposed to benzene as specified on at least 45 days within the six months before the clinical onset of pure red cell aplasia;

Note: ***being exposed to benzene as specified*** is defined in the Schedule 1 - Dictionary.

(5) having acute hepatitis within the one year before the clinical onset of pure red cell aplasia;

(6) having a liver transplant within the three months before the clinical onset of pure red cell aplasia;

(7) having an autoimmune disease from the specified list of autoimmune diseases within the two years before the clinical onset of pure red cell aplasia;

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

(8) having chronic lymphocytic leukaemia/small lymphocytic lymphoma or T-cell large granular lymphocytic leukaemia within the six months before the clinical onset of pure red cell aplasia;

(9) having a thymoma or thymic carcinoma before the clinical onset of pure red cell aplasia;

(10) having an infection with parvovirus B19 within the six months before the clinical onset of pure red cell aplasia;

(11) being pregnant at the time of the clinical worsening of pure red cell aplasia;

(12) inability to obtain appropriate clinical management for pure red cell aplasia.

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(11) and 9(12) apply only to material contribution to, or aggravation of, pure red cell aplasia where the person's pure red cell aplasia 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(3) 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:

8-hour time-weighted average (TWA) means the averaging of different exposure levels to benzene during an average exposure period equivalent to eight hours.

being exposed to benzene as specified means:

(a) having cutaneous contact with liquids containing benzene greater than 1% by volume; or
(b) ingesting liquids containing benzene greater than 1% by volume; or
(c) inhaling benzene vapour where such exposure occurs at an ambient 8-hour time-weighted average (TWA) benzene concentration exceeding five parts per million.

Note: ***8-hour time-weighted average (TWA)*** is defined in the Schedule 1 - Dictionary.

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

pure red cell aplasia—see subsection 7(2).

relevant service means:

(a) eligible war service (other than operational service) under the VEA;
(b) defence service (other than hazardous service and British nuclear test defence service) under the VEA; or
(c) peacetime service under the MRCA.

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

specified list of autoimmune diseases means:

(a) ABO mismatched haematopoietic stem cell transplant;
(b) eosinophilic fasciitis;
(c) graft versus host disease;
(d) hyperimmunoglobulinaemia;
(e) hypoimmunoglobulinaemia; or
(f) systemic lupus erythematosus.

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*.

Schedule 2 - Drugs

Note: See Section 6, Subsection 9(2)

1 Specified Drugs

1. alkylating agents (including temozolomide, busulfan, dacarbazine, cyclophosphamide, melphalan hydrochloride, nitrogen mustard)	2. allopurinol	3. antimetabolite agents (including 6-mercaptopurine, fludarabine, fluouracil, methotrexate, pemetrexed)
4. azathioprine	5. carbamazepine	6. carbonic anhydrase inhibitors (including acetazolamide, methazolamide)
7. chloramphenicol	8. dapsone	9. d-penicillamine
10. gold	11. hydantoins	12. immune checkpoint inhibitors (including nivolumab, pembrolizumab)
13. isoniazid	14. lamivudine	15. linezolid
16. mycophenolate	17. phenytoin	18. procainamide
19. recombinant erythropoietin	20. sulphonamide antibiotics (including trimethoprim, sulfamethoxazole) and drugs containing sulphonamide antibiotics (including sulfasalazine)	21. sulphonylureas (including chlorpropamide, tolbutamide)
22. valproic acid	23. zidovudine	