Statement of Principles concerning
PURE RED CELL APLASIA
(Reasonable Hypothesis)
(No. 60 of 2020)

The Repatriation Medical Authority determines the following Statement of Principles under subsection 196B(2) 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:

[Signature]

Professor Nicholas Saunders AO
Chairperson
Contents

1 Name.......................................................................................................................... 3
2 Commencement ............................................................................................................ 3
3 Authority...................................................................................................................... 3
4 Repeal ............................................................................................................................ 3
5 Application..................................................................................................................... 3
6 Schedules ..................................................................................................................... 3
7 Kind of injury, disease or death to which this Statement of Principles relates .......... 3
8 Basis for determining the factors ................................................................................. 4
9 Factors that must exist................................................................................................. 4
10 Relationship to service ............................................................................................... 5
11 Factors referring to an injury or disease covered by another Statement of Principles............................................................................................................. 5

Schedule 1 - Dictionary .................................................................................................. 7
1 Definitions..................................................................................................................... 7

Schedule 2 - Drugs ........................................................................................................... 9
1 Specified Drugs............................................................................................................ 9
1 Name
This is the Statement of Principles concerning *pure red cell aplasia (Reasonable Hypothesis)* (No. 60 of 2020).

2 Commencement
This instrument commences on 28 September 2020.

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

4 Repeal
The Statement of Principles concerning aplastic anaemia No. 50 of 2012 (Federal Register of Legislation No. F2012L01791) made under subsections 196B(2) and (8) 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 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).


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

The Repatriation Medical Authority is of the view that there is sound medical-scientific evidence that indicates that pure red cell aplasia and death from pure red cell aplasia 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 pure red cell aplasia or death from pure red cell aplasia 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 one year 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) taking a non-aspirin, nonsteroidal, anti-inflammatory drug on at least four days per week for a continuous period of at least four weeks, within the one year before the clinical onset of pure red cell aplasia;
(5) being exposed to benzene as specified on at least 30 days within the one year before the clinical onset of pure red cell aplasia;

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

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

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

(8) having an autoimmune disease from the specified list of autoimmune disease 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.

(9) having a haematological malignancy from the specified list of haematological malignancies within the six months before the clinical onset of pure red cell aplasia;

Note: specified list of haematological malignancies is defined in the Schedule 1 - Dictionary.

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

(11) having an infection with parvovirus B19 or acute infectious mononucleosis within the six months before the clinical onset of pure red cell aplasia;

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

(13) 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(12) and 9(13) 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(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:

- **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) 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 autoimmune diseases** means:
  - (a) ABO mismatched haematopoietic stem cell transplant;
  - (b) coeliac disease;
  - (c) eosinophilic fasciitis;
  - (d) graft versus host disease;
  - (e) hyperimmunoglobulinaemia;
  - (f) hypoinmunoglobulinaemia; or
  - (g) systemic lupus erythematosus.

- **specified list of haematological malignancies** means:
  - (a) chronic lymphocytic leukaemia/small lymphocytic lymphoma;
  - (b) Hodgkin's lymphoma;
  - (c) non-Hodgkin lymphoma; or
  - (d) T-cell large granular lymphocytic leukaemia.
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)

### Specified Drugs

<p>| | | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>alkylating agents (including temozolomide, busulfan, dacarbazine, cyclophosphamide, melphalan hydrochloride, nitrogen mustard)</td>
<td>2.</td>
</tr>
<tr>
<td>4.</td>
<td>azathioprine</td>
<td>5.</td>
</tr>
<tr>
<td>7.</td>
<td>chloramphenicol</td>
<td>8.</td>
</tr>
<tr>
<td>10.</td>
<td>clozapine</td>
<td>11.</td>
</tr>
<tr>
<td>13.</td>
<td>d-penicillamine</td>
<td>14.</td>
</tr>
<tr>
<td>16.</td>
<td>immune checkpoint inhibitors (including nivolumab, pembrolizumab)</td>
<td>17.</td>
</tr>
<tr>
<td>19.</td>
<td>isoniazid</td>
<td>20.</td>
</tr>
<tr>
<td>22.</td>
<td>leuproide</td>
<td>23.</td>
</tr>
<tr>
<td>25.</td>
<td>mycophenolate</td>
<td>26.</td>
</tr>
<tr>
<td>28.</td>
<td>procainamid</td>
<td>29.</td>
</tr>
<tr>
<td>31.</td>
<td>rifampicin</td>
<td>32.</td>
</tr>
<tr>
<td>34.</td>
<td>tacrolimus</td>
<td>35.</td>
</tr>
<tr>
<td>37.</td>
<td>zidovudine</td>
<td></td>
</tr>
</tbody>
</table>