

# Statement of Principles concerning RETINAL VASCULAR OCCLUSION (Reasonable Hypothesis) (No. 50 of 2020)

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

Dated 26 June 2020

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

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Professor Nicholas Saunders AO Chairperson

# Contents

	1	Name	3
	2	Commencement	3
	3	Authority	3
	4	Repeal	3
	5	Application	3
	6	Definitions	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	11
	11	Factors referring to an injury or disease covered by another Statement of Principles	11
Sched	Schedule 1 - Dictionary		
	1	Definitions	12

## 1 Name

This is the Statement of Principles concerning *retinal vascular occlusion* (*Reasonable Hypothesis*) (No. 50 of 2020).

### 2 Commencement

This instrument commences on 27 July 2020.

## 3 Authority

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

### 4 Repeal

The Statement of Principles concerning retinal vascular occlusive disease No. 83 of 2011 (Federal Register of Legislation No. F2011L01440) made under subsection 196B(2) of the VEA is repealed.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 retinal vascular occlusion and death from retinal vascular occlusion.

#### Meaning of retinal vascular occlusion

- (2) For the purposes of this Statement of Principles, retinal vascular occlusion:
  - (a) means obstruction to blood flow in either:
    - (i) the retinal artery or its branches; or
    - (ii) the retinal vein or its tributaries;

leading to visual impairment; and

- (b) excludes ischaemic optic neuropathy and ophthalmic artery occlusion.
- Note 1: Retinal vascular occlusion typically causes painless vision loss, ranging from mild to severe, and usually occurs suddenly. It may be accompanied by retinal ischaemia, macular oedema or retinal neovascularisation.

Note 2: Retinal vascular occlusion may manifest as transient monocular visual loss.

- (3) While retinal vascular occlusion attracts ICD-10-AM code H34, in applying this Statement of Principles the meaning of retinal vascular occlusion 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 retinal vascular occlusion

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

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 retinal vascular occlusion and death from retinal vascular occlusion 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 retinal vascular occlusion or death from retinal vascular occlusion with the circumstances of a person's relevant service:

 having smoked at least ten pack-years of tobacco products before the clinical onset of retinal vascular occlusion, and where smoking has permanently ceased, the clinical onset of retinal vascular occlusion has occurred within ten years of cessation;

Note: *pack-year of tobacco products* is defined in the Schedule 1 - Dictionary.

- (2) having hypertension before the clinical onset of retinal vascular occlusion;
- (3) having diabetes mellitus before the clinical onset of retinal vascular occlusion;

(4) being obese for at least five years within the 15 years before the clinical onset of retinal vascular occlusion;

Note: *being obese* is defined in the Schedule 1 - Dictionary.

(5) having dyslipidaemia before the clinical onset of retinal vascular occlusion;

Note: dyslipidaemia is defined in the Schedule 1 - Dictionary.

- (6) having hyperhomocysteinaemia before the clinical onset of retinal vascular occlusion;
- (7) having an arteritis from the specified list of systemic arteritides, at the time of the clinical onset of retinal vascular occlusion;

Note: specified list of systemic arteritides is defined in the Schedule 1 - Dictionary.

(8) having retinal vasculitis at the time of the clinical onset of retinal vascular occlusion;

Note: retinal vasculitis is defined in the Schedule 1 - Dictionary.

(9) having a non-inflammatory vasculopathy of the cerebral or retinal vessels from the specified list of vasculopathies at the time of the clinical onset of retinal vascular occlusion;

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

(10) having a disorder that is associated with a hypercoagulable state or hyperviscosity at the time of the clinical onset of retinal vascular occlusion;

Note: *disorder that is associated with a hypercoagulable state or hyperviscosity* is defined in the Schedule 1 - Dictionary.

- (11) having:
  - (a) a benign or malignant neoplasm; or
  - (b) a non-neoplastic mass lesion;

which compresses or infiltrates the retinal vessels of the affected eye at the time of the clinical onset of retinal vascular occlusion;

Note 1: Examples of a benign or malignant neoplasm include astrocytoma, optic nerve meningioma, leukaemia and lymphoma.

(12) having a vascular abnormality of the retinal vessels of the affected eye at the time of the clinical onset of retinal vascular occlusion;

Note: Examples of a vascular abnormality include arterio-venous malformation and prepapillary vascular loop.

(13) having migraine at the time of the clinical onset of retinal vascular occlusion;

Note 2: Examples of a non-neoplastic mass lesion include haematoma, sarcoidosis, neurofibromatosis and exophthalmos.

- (14) having infection with human immunodeficiency virus before the clinical onset of retinal vascular occlusion;
- (15) having an ocular or orbital infection involving the affected eye within the 30 days before the clinical onset of retinal vascular occlusion;

Note 1: Examples of an ocular or orbital infection include:

- (a) cat scratch disease;
- (b) herpes zoster ophthalmicus;
- (c) ocular tuberculosis; and
- (d) orbital abscess or cellulitis.

Note 2: This excludes isolated conjunctivitis or blepharitis of the affected eye.

(16) having direct external pressure to the eyeball of the affected eye for a continuous period of at least ten minutes, within the 24 hours before the clinical onset of retinal vascular occlusion;

Note: Examples of causes of direct external pressure to the eyeball include:

- (a) surgery in the prone position;
- (b) surgery involving the use of a device that compresses the eyes; and
- (c) positioning that places direct mechanical pressure on the eye.
- (17) having a blunt or penetrating injury involving the affected eye or the blood vessels supplying or draining the affected eye, including surgery or a therapeutic procedure, within the seven days before the clinical onset of retinal vascular occlusion;
  - Note 1: In the case of prolonged unconsciousness associated with trauma, the seven day period is taken to commence upon regaining consciousness.
  - Note 2: Examples of blunt or penetrating injury include:
    - (a) blunt trauma to the orbit;
    - (b) eyeball contusion; and
    - (c) intraocular foreign body.
  - Note 3: Examples of surgery or a therapeutic procedure include:
    - (a) cataract surgery;
    - (b) filler injection to the forehead, or the glabellar, nasal or nasolabial fold regions;
    - (c) intraocular or retrobulbar anaesthesia;
    - (d) intravitreal bevacizumab injection; and
    - (e) sclerotherapy of facial veins.
- (18) taking a drug from the specified list of drugs within the 72 hours before the clinical onset of retinal vascular occlusion;

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

- (19) taking a combined oral contraceptive pill for a continuous period of at least 21 days within the one year before the clinical onset of retinal vascular occlusion;
- (20) experiencing an acute hypotensive episode within the 24 hours before the clinical onset of retinal vascular occlusion;

Note: *acute hypotensive episode* is defined in the Schedule 1 - Dictionary.

Statement of Principles concerning Retinal Vascular Occlusion (Reasonable Hypothesis) (No. 50 of 2020) Veterans' Entitlements Act 1986

- (21) for retinal venous occlusion only:
  - (a) having open-angle glaucoma, angle-closure glaucoma or ocular hypertension, at the time of the clinical onset of retinal vascular occlusion;

Note: *ocular hypertension* is defined in the Schedule 1 - Dictionary.

(b) having chronic renal failure at the time of the clinical onset of retinal vascular occlusion;

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

(c) being severely dehydrated at the time of the clinical onset of retinal vascular occlusion; or

Note: *being severely dehydrated* is defined in the Schedule 1 - Dictionary.

- (d) having obstructive sleep apnoea at the time of the clinical onset of retinal vascular occlusion;
- (22) for retinal arterial occlusion only:
  - (a) having carotid artery disease at the time of the clinical onset of retinal vascular occlusion;
  - (b) having a cardiac disease with the potential to give rise to a retinal embolus at the time of the clinical onset of retinal vascular occlusion;

Note: Examples of a cardiac disease with the potential to give rise to a retinal embolus include:

- (a) acute myocardial infarction;
- (b) atrial fibrillation; and
- (c) mitral or aortic valve disease.
- (c) having a non-cardiac potential source of retinal embolus at the time of the clinical onset of retinal vascular occlusion; or

Note: Examples of a non-cardiac potential source of retinal embolus include:

- (a) aortic arch atherosclerosis;
- (b) atherosclerotic central retinal artery;
- (c) decompression sickness;
- (d) deep vein thrombosis in the presence of a potential route of paradoxical embolism;
- (e) pulmonary barotrauma; and
- (f) severe bone trauma.
- (d) undergoing a procedure to the heart or the carotid artery, or an intravascular procedure involving the vessels of the head or neck, within the seven days before the clinical onset of retinal vascular occlusion;

Note 1: Examples of a procedure to the heart include:

- (a) cardiac surgery or cardiac catheterisation;
- (b) coronary angiography; and
- (c) coronary artery bypass grafting.

Statement of Principles concerning Retinal Vascular Occlusion (Reasonable Hypothesis) (No. 50 of 2020) Veterans' Entitlements Act 1986 Note 2: Examples of a procedure to the carotid artery include:

- (a) carotid angioplasty;
- (b) carotid endarterectomy; and
- (c) carotid stenting.

Note 3: Examples of an intravascular procedure involving the vessels of the head or neck include:

- (a) cosmetic vein sclerotherapy to the orbital adnexa, face, nose or sinuses;
- (b) transcatheter thyroid artery embolisation; and
- (c) transfemoral cerebral angiography.
- (23) having smoked at least ten pack-years of tobacco products before the clinical worsening of retinal vascular occlusion, and where smoking has permanently ceased, the clinical worsening of retinal vascular occlusion has occurred within ten years of cessation;

Note: pack-year of tobacco products is defined in the Schedule 1 - Dictionary.

- (24) having hypertension before the clinical worsening of retinal vascular occlusion;
- (25) having diabetes mellitus before the clinical worsening of retinal vascular occlusion;
- (26) being obese for at least five years within the 15 years before the clinical worsening of retinal vascular occlusion;

Note: *being obese* is defined in the Schedule 1 - Dictionary.

(27) having dyslipidaemia before the clinical worsening of retinal vascular occlusion;

Note: dyslipidaemia is defined in the Schedule 1 - Dictionary.

- (28) having hyperhomocysteinaemia before the clinical worsening of retinal vascular occlusion;
- (29) having an arteritis from the specified list of systemic arteritides, at the time of the clinical worsening of retinal vascular occlusion;

Note: specified list of systemic arteritides is defined in the Schedule 1 - Dictionary.

(30) having retinal vasculitis at the time of the clinical worsening of retinal vascular occlusion;

Note: retinal vasculitis is defined in the Schedule 1 - Dictionary.

(31) having a non-inflammatory vasculopathy of the cerebral or retinal vessels from the specified list of vasculopathies at the time of the clinical worsening of retinal vascular occlusion;

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

(32) having a disorder that is associated with a hypercoagulable state or hyperviscosity at the time of the clinical worsening of retinal vascular occlusion;

Note: *disorder that is associated with a hypercoagulable state or hyperviscosity* is defined in the Schedule 1 - Dictionary.

#### (33) having:

- (a) a benign or malignant neoplasm; or
- (b) a non-neoplastic mass lesion;

which compresses or infiltrates the retinal vessels of the affected eye at the time of the clinical worsening of retinal vascular occlusion;

Note 1: Examples of a benign or malignant neoplasm include astrocytoma, optic nerve meningioma, leukaemia and lymphoma.

Note 2: Examples of a non-neoplastic mass lesion include haematoma, sarcoidosis, neurofibromatosis and exophthalmos.

(34) having a vascular abnormality of the retinal vessels of the affected eye at the time of the clinical worsening of retinal vascular occlusion;

Note: Examples of a vascular abnormality include arterio-venous malformation and prepapillary vascular loop.

- (35) having migraine at the time of the clinical worsening of retinal vascular occlusion;
- (36) having infection with human immunodeficiency virus before the clinical worsening of retinal vascular occlusion;
- (37) having an ocular or orbital infection involving the affected eye within the 30 days before the clinical worsening of retinal vascular occlusion;

Note 1: Examples of an ocular or orbital infection include:

- (a) cat scratch disease;
- (b) herpes zoster ophthalmicus;
- (c) ocular tuberculosis; and
- (d) orbital abscess or cellulitis.

Note 2: This excludes isolated conjunctivitis or blepharitis of the affected eye.

(38) having direct external pressure to the eyeball of the affected eye for a continuous period of at least ten minutes, within the 24 hours before the clinical worsening of retinal vascular occlusion;

Note: Examples of causes of direct external pressure to the eyeball include:

- (a) surgery in the prone position;
- (b) surgery involving the use of a device that compresses the eyes; and
- (c) positioning that places direct mechanical pressure on the eye.
- (39) having a blunt or penetrating injury involving the affected eye or the blood vessels supplying or draining the affected eye, including surgery or a therapeutic procedure, within the seven days before the clinical worsening of retinal vascular occlusion;
  - Note 1: In the case of prolonged unconsciousness associated with trauma, the seven day period is taken to commence upon regaining consciousness.
  - Note 2: Examples of blunt or penetrating injury include:
    - (a) blunt trauma to the orbit;
    - (b) eyeball contusion; and
    - (c) intraocular foreign body.

Statement of Principles concerning Retinal Vascular Occlusion (Reasonable Hypothesis) (No. 50 of 2020) Veterans' Entitlements Act 1986

- Note 3: Examples of surgery or a therapeutic procedure include:
  - (a) cataract surgery;
  - (b) filler injection to the forehead, or the glabellar, nasal or nasolabial fold regions;
  - (c) intraocular or retrobulbar anaesthesia;
  - (d) intravitreal bevacizumab injection; and
  - (e) sclerotherapy of facial veins.
- (40) taking a drug from the specified list of drugs within the 72 hours before the clinical worsening of retinal vascular occlusion;

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

- (41) taking a combined oral contraceptive pill for a continuous period of at least 21 days within the one year before the clinical worsening of retinal vascular occlusion;
- (42) experiencing an acute hypotensive episode within the 24 hours before the clinical worsening of retinal vascular occlusion;

Note: *acute hypotensive episode* is defined in the Schedule 1 - Dictionary.

- (43) for retinal venous occlusion only:
  - (a) having open-angle glaucoma, angle-closure glaucoma or ocular hypertension, at the time of the clinical worsening of retinal vascular occlusion;

Note: ocular hypertension is defined in the Schedule 1 - Dictionary.

(b) having chronic renal failure at the time of the clinical worsening of retinal vascular occlusion;

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

(c) being severely dehydrated at the time of the clinical worsening of retinal vascular occlusion; or

Note: *being severely dehydrated* is defined in the Schedule 1 - Dictionary.

- (d) having obstructive sleep apnoea at the time of the clinical worsening of retinal vascular occlusion;
- (44) for retinal arterial occlusion only:
  - (a) having carotid artery disease at the time of the clinical worsening of retinal vascular occlusion;
  - (b) having a cardiac disease with the potential to give rise to a retinal embolus at the time of the clinical worsening of retinal vascular occlusion;

Note: Examples of a cardiac disease with the potential to give rise to a retinal embolus include:

- (a) acute myocardial infarction;
- (b) atrial fibrillation; and
- (c) mitral or aortic valve disease.
- (c) having a non-cardiac potential source of retinal embolus at the time of the clinical worsening of retinal vascular occlusion; or

Note: Examples of a non-cardiac potential source of retinal embolus include:

- (a) a ortic arch atherosclerosis;
- (b) atherosclerotic central retinal artery;
- (c) decompression sickness;
- (d) deep vein thrombosis in the presence of a potential route of paradoxical embolism;
- (e) pulmonary barotrauma; and
- (f) severe bone trauma.
- (d) undergoing a procedure to the heart or the carotid artery, or an intravascular procedure involving the vessels of the head or neck, within the seven days before the clinical worsening of retinal vascular occlusion;

Note 1: Examples of a procedure to the heart include:

- (a) cardiac surgery or cardiac catheterisation;
- (b) coronary angiography; and
- (c) coronary artery bypass grafting.

Note 2: Examples of a procedure to the carotid artery include:

- (a) carotid angioplasty;
  - (b) carotid endarterectomy; and
  - (c) carotid stenting.

Note 3: Examples of an intravascular procedure involving the vessels of the head or neck include:

- (a) cosmetic vein sclerotherapy to the orbital adnexa, face, nose or sinuses;
- (b) transcatheter thyroid artery embolisation; and
- (c) transfemoral cerebral angiography.
- (45) inability to obtain appropriate clinical management for retinal vascular occlusion.

#### **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(23) to 9(45) apply only to material contribution to, or aggravation of, retinal vascular occlusion where the person's retinal vascular occlusion 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:

*acute hypotensive episode* means a sudden drop in blood pressure of a sufficient degree to cause cerebral hypoperfusion.

being obese means having a Body Mass Index (BMI) of 30 or greater.

Note: **BMI** is also defined in the Schedule 1 - Dictionary.

*being severely dehydrated* means having clinical dysfunction due to inadequate body fluids, in which the loss of body fluids, mostly water, exceeds intake. Symptoms and signs of severe dehydration include limited or absent urine output, dizziness or lightheadedness, and orthostatic hypotension.

Note: Examples of causes of dehydration include strenuous physical activity, vomiting, diarrhoea and febrile illness.

*BMI* means  $W/H^2$  where:

- (a) W is the person's weight in kilograms; and
- (b) H is the person's height in metres.

## chronic renal failure means:

- (a) having a glomerular filtration rate of less than 15 mL/min/1.73 m<sup>2</sup> for a period of at least three 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.

*disorder that is associated with a hypercoagulable state or hyperviscosity* means an acquired defect in blood components that alters blood viscosity or disrupts the coagulation cascade, characterised by an increased predisposition to form blood clots.

Note: Examples of a disorder that is associated with a hypercoagulable state or hyperviscosity include:

- (a) antiphospholipid antibody syndrome;
- (b) lymphoplasmacytic leukaemia;
- (c) lymphoplasmacytic lymphoma (Waldenstrom macroglobulinaemia);
- (d) myeloma;
- (e) myeloproliferative neoplasm;
- (f) nephrotic syndrome;
- (g) paroxysmal nocturnal haemoglobinuria; and
- (h) polycythaemia.

*dyslipidaemia* means persistently abnormal blood lipid levels, diagnosed by a medical practitioner and evidenced by:

- (a) a total serum cholesterol level greater than 5.5 mmol/L; or
- (b) a serum low density lipoprotein level greater than 4.0 mmol/L; or
- (c) a serum high density lipoprotein level less than 1.0 mmol/L; or

- (d) a serum triglyceride level greater than or equal to 2.0 mmol/L; or
- (e) the regular administration of drug therapy to normalise blood lipid levels.

MRCA means the Military Rehabilitation and Compensation Act 2004.

*ocular hypertension* means intra-ocular pressure greater than 21 mm Hg, without evidence of optic atrophy or visual field loss.

#### pack-year of tobacco products means:

- (a) 20 cigarettes per day for a period of one calendar year; or
- (b) 7,300 cigarettes in a period of one calendar year; or
- (c) 7,300 grams of smoking tobacco by weight, either in cigarettes, pipe tobacco or cigars, or a combination of same, in a period of one calendar year.

#### 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.

#### retinal vascular occlusion—see subsection 7(2).

*retinal vasculitis* means inflammation of the retinal artery or its branches, or the retinal vein or its tributaries. Retinal vasculitis can be due to autoimmune disease, infection, drugs, or may be idiopathic. It can be confined to the retinal vessels, or may be an element of a systemic vasculitis.

Note 1: Autoimmune diseases that can cause retinal vasculitis include:

- (a) Behcet disease;
- (b) eosinophilic granulomatosis with polyangiitis (EGPA);
- (c) granulomatosis with polyangiitis (Wegener granulomatosis);
- (d) microscopic polyangiitis;
- (e) polyarteritis nodosa;
- (f) Susac syndrome;
- (g) systemic lupus erythematosus; and(h) systemic sclerosis (scleroderma).

Note 2: Infections that can cause retinal vasculitis include:

- (a) cytomegalovirus;
- (b) herpes simplex virus;
- (c) human immunodeficiency virus;
- (d) syphilis;
- (e) toxoplasmosis;
- (f) tuberculosis;
- (g) varicella zoster virus; and
- (h) viral uveitis.

Note 3: Drug-induced causes of retinal vasculitis include intravitreal vancomycin and intraocular gentamycin.

#### specified list of drugs means:

- (a) amphetamines and amphetamine-like compounds;
- (b) cannabis;

- (c) intranasal cocaine;
- (d) intravenous heroin contaminated with talc;
- (e) phosphodiesterase type 5 (PDE5) inhibitors;
- (f) testosterone supplementation; or
- (g) tranexamic acid.

# specified list of systemic arteritides means:

- (a) giant cell (temporal) arteritis;
- (b) polyarteritis nodosa;
- (c) Takayasu arteritis; or
- (d) thromboangiitis obliterans (Buerger's disease).

# specified list of vasculopathies means:

- (a) fibromuscular dysplasia;
- (b) Moyamoya disease; or
- (c) Sneddon syndrome.

*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.