

Disclaimer: This Clinical Practice Guideline ('CPG') was written for use in The Royal Victorian Eye and Ear Hospital Emergency Department. It should be used under the guidance of an Ophthalmology or ENT registrar. If clinical advice is required, please contact the Eye and Ear Admitting Officer for assistance: EYE: +61 3 9929 8033; ENT: +61 3 9929 8032. Links to internal Eye and Ear documents cannot be accessed from the website CPG.

Description:

Occlusion of the central retinal vein at the level of the lamina cribrosa. CRVO may be either ischaemic or non-ischaemic which will have implications in treatment and prognosis.

Red Flags:

- Differential diagnosis:
 - Diabetic retinopathy: retinal haemorrhages and microaneurysms, usually bilateral.
 - Ocular ischaemic syndrome: mid-peripheral retinal haemorrhages with dilated veins, which are not tortuous.
 - Hypertensive retinopathy (bilateral retinopathy)
 - Blood dyscrasia (often bilateral)
- Beware '90 day' glaucoma: neovascular glaucoma occurs in ~50% of ischaemic CRVO after 3 months
- CRVO in a 'young' patient (<55 years old) – systemic risk factors should be investigated by GP or physician

How to Assess:

History:

- Progressive, painless, decrease in vision.
 - Loss of vision at presentation tends to be more severe in ischaemic CRVO or in the presence of macular oedema.
- History of systemic hypertension, cardiovascular disease, diabetes mellitus, hyperlipidaemia, smoking or hypercoagulability (e.g. factor V Leiden mutation), glaucoma and ocular hypertension.
 - Other associations: oral contraceptive pill, sleep apnoea, myeloproliferative disorders and other inflammatory disorders associated with retinal vasculitis.

Examination:

- Pupils: test for relative afferent pupillary defect (RAPD) (in ischaemic CRVO)
- Intraocular pressure (IOP)
- Gonioscopy: for neovascularisation of iris and angle; narrow angles
- Dilated retinal examination:
 - Diffuse retinal haemorrhages, cotton wool spots and exudates with dilated and tortuous retinal veins in all four quadrants.
 - Macular oedema
 - Neovascularisation at the disc (NVD), or 'elsewhere' (NVE)
 - Signs of 'old' CRVO: disc collaterals, pigmentary changes in the macula
 - Closing venous pressure – this is a specific sign for CRVO. Visualise the veins exiting from the disc margin, while applying digital pressure on the globe. Normally, venules collapse with mild pressure. However, in CRVO, venules collapse at the same time OR after the arterioles.
- Clinical signs suggestive of ischaemic CRVO include: poor presenting visual acuity, RAPD, significant retinal haemorrhages, cotton wool spots and presence of neovascularisation.

Investigations

- Macular OCT to assess for presence and extent of cystoid macular oedema (CMO) – if in hours (not mandatory)
- Colour fundus photographs to document retinal findings
- Fundus fluorescein angiogram is important to assess for areas of retinal ischaemia and macular involvement. This can be arranged via Medical Retinal Clinic/private specialist.
- Systemic investigations - Hypertension and hyperlipidaemia are primary risk factors, even in the younger age group
 - Consider ordering: FBE, UE, lipid profile, HbA1c, BSL level, ESR, CRP
 - Do NOT order thrombophilia screens from ED, unless discussed with haematologist or medical retina fellow
 - Blood pressure measurements: Perform repeated blood pressure measurements during presentation in ED to ensure fluctuations are not missed. Consider requesting GP's to perform a 24hr BP monitor to ensure things

Acute Management:

- Measure blood pressure
- Treat elevated intraocular pressure

Follow up:

- Discuss with medical retina fellow regarding timing of fluorescein angiogram and follow-up:
 1.
 - A. Non ischaemic (not only eye) – Medical Retinal Clinic 4 weeks
 - B. Non ischaemic and only eye – Medical Retinal Clinic 1 week
 2.
 - A. Ischaemic with rubeosis – add on urgently to next Medical Retinal Clinic for consideration of intravitreal Avastin®
 - B. Ischaemic without rubeosis – Medical Retinal Clinic 2 weeks
 3. Rubeosis and high IOP – also contact GIRU fellow regarding consideration of cyclodiode/further management
 4. Intravitreal anti-VEGF injection for macular oedema. Level 1 evidence for anti-VEGF therapy for cystoid macular oedema (CMO) in non-ischaemic CRVO. Private specialists can now access Lucentis® and Eylea® via PBS for this indication.
 5. Due to limited capacity in RVEEH intravitreal injection clinics, consider referral to local ophthalmologist for follow - up/anti-VEGF therapy.
- Patients with neovascular glaucoma should be referred to the Glaucoma Clinic
- Patients with neovascularisation require urgent referral to Medical Retinal Clinic for consideration of pan-retinal photocoagulation treatment +/- intravitreal anti-VEGF injections
- GP or physician referral to further investigate and manage risk factors: hypertension, cardiovascular disease, diabetes mellitus, hyperlipidaemia or hypercoagulability. This is the only way to reduce the risk of involvement of the contralateral eye.

Evidence Table

Author(s)	Title	Source	Level of Evidence (I – VII)
Coscas G, Loewenstein A, Augustin A, et al.	Management of retinal vein occlusion--consensus document.	International journal of ophthalmology 2011;226:4-28.	IV-II
La Spina C, De Benedetto U, Parodi MB, Coscas G, Bandello F.	Practical management of retinal vein occlusions.	Ophthalmology and therapy 2012;1:3.	IV-II
Gerstenblith AT, Rabinowitz	The Wills eye manual : office and emergency room diagnosis and treatment of eye disease, 6th ed.	Wolters Kluwer/Lippincott Williams & Wilkins, 2012	VII
Sobha Sivaprasad, Winfried Amoaku, Tom Williamson, Paul Dodson, James Talks, Katherine Talks, Khanchan Bhan and Philip Hykin	The Royal College of Ophthalmologists Guidelines on retinal vein occlusions: executive summary.	Eye 29, 1633–1638 (2015). https://doi.org/10.1038/eye.2015.164	

The Hierarchy of Evidence

The Hierarchy of evidence is based on summaries from the National Health and Medical Research Council (2009), the Oxford Centre for Evidence-based Medicine Levels of Evidence (2011) and Melynck and Fineout-Overholt (2011).

- I) Evidence obtained from a systematic review of all relevant randomised control trials.
- II) Evidence obtained from at least one well designed randomised control trial.
- III) Evidence obtained from well-designed controlled trials without randomisation.
- IV) Evidence obtained from well-designed cohort studies, case control studies, interrupted time series with a control group, historically controlled studies, interrupted time series without a control group or with case series.
- V) Evidence obtained from systematic reviews of descriptive and qualitative studies.
- VI) Evidence obtained from single descriptive and qualitative studies.
- VII) Expert opinion from clinician, authorities and/or reports of expert committees or based on physiology.

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