RANZCO

The Royal Australian
and New Zealand
College of Ophthalmologists

Vitreoretinal Curriculum Standard

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Purpose

The Vitreoretinal Clinical Performance Standard covers the specific knowledge, processes, skills and competencies required for the diagnosis and treatment of vitreoretinal disorders.

Retinal disease is the most frequent cause of irreversible blindness in Australia and many countries worldwide. Tremendous advances in new biologics, surgical techniques and imaging technology have enabled ophthalmologists to diagnose and effectively manage a growing number of retinal conditions.

Medical retina makes up a large proportion of the work of a general ophthalmologist. The range of new treatment options becoming available in the management of age-related macular degeneration, diabetic retinopathy and retinal vein occlusion reinforces the need for trainees to be exposed to the subtleties and complexities of retinal diagnostic challenges and their management.

References

In addition to the core texts, the following references are recommended:

Medical Retina Randomised Clinical Trials

1. AMD: ANCHOR, MARINA, PIER, CATT, VIEW, HORIZON/ SEVEN-UP
2. DR: DCCT, UKPDS, ETDRS, FIELD, ACCORD,
3. DME: ETDRS, DRCR.net, RESTORE, RISE/RIDE, BOLT, VIVID/VISTA
4. RVO: SCORE, CRUISE, BRAVO, GALILEO, COPERNICUS

Extended Vitreoretinal Reading

- Agarwal, A. & Gass, J.D.M. 2012, *Gass' atlas of macular diseases*, 5th edn, Elsevier Saunders, Edinburgh.
- Curtin, B.J. 1977, 'The posterior staphyloma of pathologic myopia', *Tr. Am. Ophth. Soc.*, vol. 75, pp. 67-86.<http://www.ncbi.nlm.nih.gov/pmc/articles/PMC1311542/>
- Edwards, A.O. 2008, 'Clinical features of the congenital vitreoretinopathies', *Eye,* vol. 22, pp.1233-1242.
- Holder, G.E., Celesia, G.G., Miyake, Y., Tobimatsu, S. & Weleber R.G. 2010, 'International federation of clinical neurophysiology: recommendations for visual system testing', *Clinical Neurophysiology*, vol. 121, pp. 1393–1409.
- Holz, F.G., Pauleikhoff, D., Spaide, R.F. & Bird, A.C. 2013, *Age-related macular degeneration*, 2nd edn., Springer, Berlin, Heidelberg.
- Kuhn, F. 2008, *Ocular traumatology*, Springer, Berlin.
- Lewis, H.2003, 'Peripheral retinal degeneration and the risk of retinal detachment', *Am. J. Ophthalmol.,* vol. 136, pp. 155-160.
- Macsai, M.S. (ed.) 2007, *Ophthalmic microsurgical suturing techniques,* Springer, Berlin.

- Mitry, D., Fleck, B.W., Wright, A.F., Campbell, H. & Charteris, D.G. 2010, 'Pathogenesis of rhegmatogenous retinal detachment: predisposing anatomy and cell biology', *Retina*, vol. 30, pp.1561-1572.

- Richards, A.J., Scott J.D, & Snead, M.P. 2002, 'Molecular genetics of rhegmatogenous retinal detachment', *Eye*, vol.16; pp. 388-392.

- Ryan, S.J. (ed.) 2013, *Retina*, 5th edn, Volume1 (*Retinal imaging and diagnostics; Basic science and translation to therapy*); Volume 2 (*Medical retina*); Volume 3 (*Tumors of the retina, choroid and vitreous*), Elsevier Saunders, London.
- Singh, A.D., Damato, B.E, Pe'er, J., Murphree, A.L., & Perry, J.D., 2007, *Clinical ophthalmic oncology*, Elsevier Saunders, Edinburgh .

- Spalton, D.J. et al. 2011, *Atlas of clinical ophthalmology*, 3rd edn, Elsevier/Mosby, Philadelphia, PA.

- Thompson J.A., Snead, M.P., Billington, B.M., Barrie, T., Thompson, T.R. & Sparrow, J.M. 2002, 'National audit of the outcome of primary surgery for rhegmatogenous retinal detachment. I. Sample and methods', Eye, vol. 16, pp. 766-70.

- Thompson J.A., Snead, M.P., Billington, B.M., Barrie, T., Thompson, T.R. & Sparrow, J.M. 2002, 'National audit of the outcome of primary surgery for rhegmatogenous retinal detachment. II. Clinical outcome', Eye, vol. 16, pp. 771-777.

- Yannuzzi, L.A. 2010, *The retinal atlas*, Saunders Elsevier, London.

It is recommended that reading be supplemented with appropriate articles from current and relevant peer-reviewed journals.

Best Practice Standards

RANZCO Clinical Guidelines for Performing Intravitreal Therapy
Accessed 25 March 2014, <http://www.ranzco.edu/index.php/about/policy-new>

RANZCO Fluorescein and Indocyanine Green Angiography Guidelines
Accessed 24 May 2014,
<http://www.ranzco.edu/images/documents/policies/Fluorescein_and_Indocyanine_Green_Angiography_Guidelines.pdf>

Level of Mastery

For each learning outcome, the level of mastery to be attained by the trainee at the end of training is indicated as follows:

***	Core knowledge of which trainees must be able to demonstrate understanding Skills and procedures that trainees must be able to perform autonomously
**	Knowledge of which trainees must have a good practical understanding Skills and procedures with which trainees should have assisted, and of which have good practical knowledge
*	Knowledge, skills and procedures of which trainees must have some understanding

Learning outcomes and performance criteria

VR1 GENERAL MEDICAL AND OCULAR HISTORY RELEVANT TO VITREORETINAL CONDITIONS		
This element covers the processes for observing, promoting and recording a general medical and ocular history in preparation for diagnosis and treatment of vitreoretinal conditions. *The trainee is expected to have obtained and recorded a general medical and ocular history (including family history) as outlined in the Ophthalmic Basic Competencies and Knowledge (OBCK) standard.*		
LEARNING OUTCOMES	**LEVEL OF MASTERY**	**PERFORMANCE CRITERIA**
1.1 Identify key features of symptoms that may assist in diagnosing vitreoretinal disease	***	1.1.1 Identification must include: • commencement/duration • fluctuation and severity • precipitating and exacerbating activities • recurrence 1.1.2 Ascertain symptoms of photopsia, floaters and field defects with respect to: • commencement / duration • distinguishing symptoms of field defects related to detachment, glaucoma and retina degeneration 1.1.3 Distinguish between different types of photopsia or scintillations related to vitreoretinal traction, migraine, ocular tumour and inflammatory chorioretinopathy

1.2 Identify general medical conditions including congenital/hereditary and acquired that may be associated with vitreoretinal disease	***	1.2.1 Ascertain relevant current and past history of illnesses, ocular history, surgical history, family history, diseases, allergies and medications / substances that may contribute to vitreoretinal conditions 1.2.2 Recognise principal risk factors for AMD and retinal vascular conditions: family history, hypertension, diabetes, lipid disorders, smoking, dietary factors 1.2.3 Identify and understand the association with vitreoretinal disease of the following: • congenital/hereditary conditions • acquired conditions: − degenerative − infective − tumours/malignancy − autoimmune and inflammatory − iatrogenic
	**	1.2.4 Identify patterns of family history for retinal detachment, retinoschisis or retinal dystrophy as having dominant, recessive, mitochondrial or X-linked inheritance 1.2.5 Elicit risk factors for progression to tractional detachment of ischaemic / proliferative retinopathy in diabetes mellitus, following radiation, carotid occlusion, and in retinopathy of prematurity
	*	1.2.6 Be familiar with the association between optic nerve head anomaly, coloboma and systemic diseases 1.2.7 Be familiar with systemic associations in inherited vitreoretinopathies

1.3 Ascertain relevant vitreoretinal surgical history	***	1.3.1 Ascertain previous retinal surgical history and outcomes 1.3.2 Ascertain relevant history related to previous complications related to anterior segment surgery (cataract, glaucoma etc.)
	**	1.3.3 Elicit specific surgical procedure history related to the different types of retinal detachment repair techniques, focusing on postoperative course and complications: • scleral buckle • pneumatic retinopexy • vitrectomy • cryo. versus laser retinopexy • gas, oil and heavy liquid tamponade
1.4 Identify vitreoretinal conditions arising from trauma	***	1.4.1 Ascertain history of trauma including: • nature of injury • closed or open globe • classification of trauma • presence of intraocular foreign body • trauma to surrounding tissue area • non-ocular injury 1.4.2 Identify features that may contribute to high risk of infection and morbidity
	**	1.4.3 Elicit symptoms of sympathetic ophthalmia from patients with history of trauma and multiple vitreoretinal procedures
	*	1.4.4 Systemic review to identify risk factors for Purtcher retinopathy, e.g. thoracic and cranial trauma

VR2 PERFORM EYE EXAMINATIONS FOR VITREORETINAL CONDITIONS

This element covers the performance and interpretation of a range of eye examinations associated with vitreoretinal conditions. It also covers the demonstration of judgment in selecting the appropriate examinations for particular patients.

The trainee is expected to have performed preliminary eye examinations as outlined in the Ophthalmic Basic Competencies and Knowledge (OBCK) standard.

LEARNING OUTCOMES	LEVEL OF MASTERY	PERFORMANCE CRITERIA
2.1 Undertake an anterior segment examination including sclera and conjunctiva	***	2.1.1 Perform and interpret the results of these examinations and identify their relevance to the diagnosis of vitreoretinal conditions: • visual acuity (best corrected) • pupil responses • intraocular pressure • lens status and media clarity • grade anterior chamber cells and flare • identify keratic precipitates, rubeosis iridis
	**	2.1.2 Identify the following signs with specific relevance to surgical retina: • evidence of previous vitrectomy surgery • location and extrusion of scleral explant • abnormal lid movement and/or ocular motility due to scleral explant • peripheral iridotomy location and type of vitreous tamponade 2.1.3 Assess lens / capsule complex: • identify different types of cataract following vitreous surgery (lens touch, gas cataract, posterior subcapsular and nuclear sclerotic cataract) • assess location and fixation of intraocular lens implant (in-the-bag, sulcus fixation, sutured, iris fixation, optic captured and anterior chamber lens) • assess integrity of capsular support • assess stability of crystalline lens

	*	2.1.4 Identify and distinguish between different types of vitreous tamponade that may migrate into the anterior chamber 2.1.5 Assess anterior chamber and perform gonioscopy in postoperative ocular hypertension following vitreous surgery to distinguish between anterior chamber oil fill, silicone oil overfill, gas overfill (expansile concentration) and trabeculitis
2.2 Undertake a directed posterior segment examination	***	2.2.1 Use binocular indirect ophthalmoscopy to identify vitreoretinal structures and conditions 2.2.2 Assess vitreous status at slit lamp: • distinguish between attached and detached posterior cortical vitreous 2.2.3 Using suitable condensing lens (e.g. 78D), perform posterior pole examination to assess the optic nerve, macula, peripheral retina, noting abnormalities in the retina, macula, fovea, retinal vessels, pigment epithelium and choroid, and define conditions affecting these structures 2.2.4 Identify and distinguish between early and late signs of age-related macular degeneration (AMD), including neuro-sensory and retinal pigment epithelial (RPE) detachment, and presence of neovascular features (haemorrhage, exudate, fibrosis) 2.2.5 Identify and distinguish variants of AMD including polypoidal choroidal vasculopathy (PCV) and retinal angiomatous proliferation (RAP) 2.2.6 Identify epiretinal membranes, macular oedema, macular hole, vitreomacular traction, diabetic retinopathy, idiopathic central serous retinopathy, retinal venous occlusive disease, retinal emboli, retinal arteriolar occlusion, choroidal naevi, RPE hyperplasia, ocular ischaemia

		2.2.7 Identify abnormalities of ocular anatomy consistent with trauma
	**	2.2.8 Assess vitreous opacity: • grading of vitreous opacity • identify and distinguish between tobacco dust, vitreous haemorrhage, vitritis and vitreous seeding of tumours • identify patterns of vitreous abnormalities in vitreoretinopathies • distinguish between silicone oil, gas, and heavy liquid in vitreous cavity 2.2.9 Perform detailed peripheral retinal examination: • using indirect ophthalmoscopy and scleral indentation • using Goldmann 3 mirror lens • using wide field contact lens • visualise retinal breaks: horseshoe tears, atrophic and operculated holes, retinal dialysis, giant tears • assess adequacy of retinopexy • assess extent and adequacy of scleral indent following scleral buckling surgery 2.2.10 Recognise degenerative retinoschisis: • elicit clinical signs that distinguish between detachment and retinal schisis • distinguish between inner and outer leaf breaks • identify underlying retinal detachment arising from outer leaf break in retinoschisis 2.2.11 Recognise choroidal detachment: • distinguish between haemorrhagic and serous choroidal detachment, and simulating lesions 2.2.12 Distinguish between rhegmatogenous, exudative, and tractional retinal detachments

VR3 VITREORETINAL DIAGNOSIS AND INVESTIGATIONS

This element covers the performance and interpretation of a range of special vitreoretinal investigations associated with vitreoretinal conditions. Following examination, the provisional diagnosis and/or differential diagnosis is established. Further investigation may be required to establish the diagnosis.

The trainee is required to demonstrate judgment in selecting the appropriate tests for particular patients.

LEARNING OUTCOMES	LEVEL OF MASTERY	PERFORMANCE CRITERIA
3.1 Undertake specific retinal investigations	***	3.1.1 Identify the indications and contra-indications for, and understand the adverse effects of the following retinal investigations. Perform or order tests, so as to provide the most useful diagnostic information, and interpret the results of: • optical coherence tomography (OCT) • retinal photography • fluorescein angiography (FA) • indocyanine green angiography (ICG) • fundus autofluorescence imaging (FAF) • B-scan ultrasound
	**	3.1.2 Use OCT imaging to assess maculae for presence of fluid (intraretinal, subretinal, sub-RPE), subretinal fibrosis, hard exudate, epiretinal membrane, thinning (atrophy), vitreo-macular attachment or traction, macular hole, subretinal lesions, and choroidal signs 3.1.4 Use OCT imaging to distinguish between retinal detachment and retinoschisis 3.1.5 Use autofluorescence imaging (FAF) to identify geographic atrophy and other FAF signs 3.1.6 Use B-scan ultrasound in patients with vitreous haemorrhage to, for example: • detect retinal tears • distinguish between vitreous detachment and retinal detachment • distinguish between serous and haemorrhagic choroidal detachment

	*	3.1.7 Understand indication and interpretation of fundus controlled perimetry (microperimetry) 3.1.8 Perform B-scan ultrasound to assist in differential diagnosis of conditions such as retinal detachment, choroidal osteoma, melanoma, metastasis, uveal lymphoma and uveal effusion
3.2 Use radiological testing to establish diagnosis	***	3.2.1 Identify the indications for use and interpret the results of: • X-rays • carotid duplex studies • computed tomography (CT) scans • magnetic resonance imaging (MRI)
	**	3.2.2 Demonstrate awareness of relevant neuroimaging for diagnosis of intracranial lesions associated with optic nerve head anomalies
3.3 Use electrophysiological testing to establish diagnosis	***	3.3.1 Identify the indications and limitations, and interpret the results of retinal and visual pathways tests using: • electro-oculogram (EOG) • electro-retinogram (ERG) – pattern ERG, full field (Ganzfeld) ERG, multi-focal ERG • visual evoked response (VER)
3.4 Use biopsy testing to establish diagnosis	*	3.4.1 Use vitreous biopsy to identify retinal lymphoma 3.4.2 Use fine needle biopsy to identify melanoma 3.4.3 Use scleral histology in uveal effusion syndrome to identify excess proteoglycan 3.4.4 Use aqueous sampling in selected cases to detect photoreceptor segments on electron microscopy

3.5 Use other systemic investigations to identify risk factors and/or establish diagnosis	***	3.5.1 Identify the indications for systemic investigations or screening to assist in identifying risk factors and co-morbidities for retinal vascular disease and retinal detachment
	*	3.5.2 Understand potential for gene testing to identify mutations in inherited vitreo/retinopathies 3.5.3 Describe systemic work-up in the process of confirming diagnosis of various inherited vitreo/retinopathies

VR4 IMPLEMENT A VITREORETINAL MANAGEMENT PLAN

This element covers the management of vitreoretinal conditions using observation, medical therapies and surgery including postoperative care.

The trainee must adhere to the standards of practice, particularly those regarding informed consent and clinical record-keeping, described in the Ophthalmic Basic Competencies and Knowledge (OBCK) standard.

LEARNING OUTCOMES	LEVEL OF MASTERY	PERFORMANCE CRITERIA
4.1 Determine and document in medical records a management plan for each individual patient	***	4.1.1 Clearly identify and document any specific features, signs, and grading of macular or retinal diseases - e.g. degenerative retinal diseases including AMD (both early and late features, and types of CNV), the severity level for diabetic retinopathy (DR) by using ETDRS or AAO scales, and peripheral retinal pathologies 4.1.2 Clearly identify and document any specific features and signs of choroidal or RPE related pathologies - e.g. other secondary causes of choroidal neovascularisation including myopic retinal degeneration, angioid streaks, choroidal rupture, multifocal choroidopathy, birdshot choroidopathy, and other inflammatory conditions 4.1.3 Clearly identify and document any systemic conditions associated with the corresponding macular or retinal diseases - e.g. inflammatory (sarcoidosis) , infectious (TB), connective tissue disorders (pseudoxanthoma elasticum), inherited disorders (Usher syndrome) 4.1.4 Clearly identify and document the features, signs and sizes of any tumour in the posterior segment 4.1.5 Clearly identify and distinguish the differences between retinoschisis and retinal detachment, and document retinal detachment and grade any proliferative vitreoretinopathy on a retinal detachment chart

	**	4.1.6 Document the specific risks of primary detachment repair using vitrectomy technique, scleral buckle or pneumatic retinopexy - including postoperative endophthalmitis, failure of retinal reattachment, cataract, and glaucoma. Document the factors associated with poor visual outcome in these conditions 4.1.7 Formulate treatment plan and follow-up for both early and late-stage AMD, including role for dietary supplements and anti-VEGF therapy. Understand the need for long-term therapy for CNV, and the approach to monitoring using OCT and other investigations 4.1.8 Formulate treatment plan for different severity stages of diabetic retinopathy - e.g. non-proliferative DR versus proliferative DR, and centre- versus non-centre-involving diabetic macular oedema (DME) including indications for medical therapy (e.g. fenofibrate), focal, grid or panretinal laser (PRP) or anti-VEGF therapy 4.1.9 Formulate treatment plan for other vascular retinopathies including medical therapy, focal or peripheral laser, or anti-VEGF therapy 4.1.10 Formulate a surgical management plan for advanced DR and other proliferative retinopathies. Identify indications for vitrectomy – e.g. subhyaloid haemorrhage, non-clearing haemorrhage, recurrent haemorrhage, tractional detachment encroaching fovea

	*	4.1.11 Formulate a surgical management plan for rhegmatogenous retinal detachment including the reasoning behind the choices between scleral buckling, vitrectomy or pneumatic retinopathy 4.1.12 Discuss the rationale of various adjunctive techniques in repairing retinal detachment complicated by proliferative vitreoretinopathy – e.g. scleral buckle versus retinectomy versus using of vitreous substitute 4.1.13 Formulate a management plan for choroidal or retinal tumours (e.g. therapy for ocular melanoma, choroidal haemangioma, vasoproliferative tumour) 4.1.14 Discuss the indications and controversies in prophylaxis against retinal detachment in patients with high myopia, extensive lattice degeneration, and inherited vitreoretinopathy including those with collagen disorders
4.2 Educate the patient on the proposed management regimen	***	4.2.1 Explain the natural history, proposed management and potential outcomes for therapy of neovascular AMD, atrophic AMD and other degenerative retinal lesions 4.2.2 Explain the need for long-term anti-VEGF management of neovascular AMD, with appropriate monitoring using OCT and other assessments 4.2.3 Explain the likely natural history, proposed management regimen (laser or intravitreal therapy), alternative therapies and potential outcomes of the therapy for DR (DME and PDR) and other vascular retinopathies 4.2.4 Explain clearly the natural history, proposed management regimen, alternatives and the potential outcome with and without surgical repair in rhegmatogenous retinal detachment

		4.2.5 Explain the rationale and importance of posturing following vitreoretinal surgery 4.2.6 Explain the consequences of intraocular gas, heavy liquid and silicone oil tamponade including restriction on air travel and effects on vision 4.2.7 Advise patients presenting with symptomatic posterior vitreous detachment (with no retinal break on examination) of the symptoms and signs of retinal tears and detachment, and recommended course of action if these arise
	**	4.2.8 Educate the patient on the expected duration of tamponade with respect to the type of gas tamponade and concentration
4.3 Manage vitreoretinal surgical conditions using appropriate therapies	***	4.3.1 Determine the best treatment option in patients presenting with elevated intraocular pressure following vitreoretinal procedure
	**	4.3.2 Identify sympathetic ophthalmia in patients who have had ocular trauma and/or vitreoretinal procedures, and discuss treatment options
	*	4.3.3 Identify surgically induced necrotising scleritis and discuss differential diagnosis and treatment options

4.4 Use laser or intravitreal injection procedures to manage vitreoretinal conditions	***	4.4.1 Demonstrate safe use of laser techniques in vitreoretinal management 4.4.2 Perform laser treatment to manage diabetic retinopathy, including focal and grid laser for threatened or actual DME, and panretinal laser for PDR 4.4.3 Perform intravitreal injection of different agents (steroids, anti-VEGF therapies) to manage neovascular AMD, DME or other vascular retinopathies 4.4.4 Recognise symptoms and signs suggesting endophthalmitis following intravitreal therapy, and perform tap and inject procedures to manage suspected endophthalmitis 4.4.5 Understand the role of vitrectomy surgery in endophthalmitis
	**	4.4.6 Using contact lens or indirect-laser delivery in performing laser treatment to seal retinal breaks
	*	4.4.7 Perform or observe use of endolaser treatment during vitreous surgery
4.5 Participate in the surgical management of vitreoretinal conditions	***	4.5.1 Counsel patient on potential for endophthalmitis after intravitreal therapy, warning of specific symptoms and need for urgent review 4.5.2 Counsel patient on procedures, potential outcomes and potential complications of retinal detachment repair 4.5.3 Participate in vitrectomy and buckling procedures, and identify and describe common intraoperative complications 4.5.4 Identify postoperative complications following retinal detachment procedures

	**	4.5.5 Manage cataract following vitrectomy surgery 4.5.6 Assist with removal of silicone oil from posterior chamber with or without cataract extraction 4.5.7 List the surgical steps in retinal detachment repair using scleral buckling or pars plana vitrectomy 4.5.8 Perform cryopexy to seal retinal break/s with or without intravitreal gas injection 4.5.9 Assist with removal of segmental scleral buckle in buckle extrusion
4.6 Provide psychological support for patient	***	4.6.1 Counsel patient on diagnosis and prognosis of retinal detachment 4.6.2 Provide support for patients with low vision due to retinal diseases through referral to low vision services 4.6.3 Refer patients to relevant support services, including genetic counselling
	*	4.6.4 Interpret the results of genetic testing for inherited retinal disease, and advise the patient 4.6.5 Be aware of and able to discuss experimental treatments such as stem cell therapy or bionic eye with the patient

Context

In order to fulfil the clinical performance standards, the trainee must apply the knowledge and skills described in the:

Ophthalmic Sciences (Anatomy, Clinical Ophthalmic and Emergency Medicine, Optics, Physiology, Clinical Genetic and Microbiology and Evidence-based Ophthalmic Practice);
Ophthalmic Basic Competencies and Knowledge (OBCK); and,
Basics of Ophthalmic Surgery (BOS) curriculum standards.

Clinical practice

The following list is provided to identify the conditions, their causes and sequelae, and the treatment approaches that may be encountered by the trainee in clinical practice. The list is not exhaustive; it is intended as a guide for the use of the trainee when planning his or her learning.

Conditions deserving special emphasis

These conditions are of particular importance because of their prevalence and impact on society. It is expected that the trainee will have a very detailed knowledge of these conditions.

- Medical retinal conditions
  - diabetic retinopathy
  - choroidal neovascularisation
  - other common retinal vascular diseases (branch/ central retinal vein/ artery occlusion)
  - atrophic retinal or macular disease

- Surgical retinal conditions
  - retinal breaks and retinal detachment
  - retained lens fragments and dislocated intraocular lens
  - endophthalmitis
  - posterior segment trauma

The aspects of these conditions that should be covered include:

- epidemiology and public health significance;
- clinical presentation including ocular and non-ocular presentation;
- association with systemic conditions;
- differential diagnosis;
- pathology;
- a detailed account of treatment modalities with reference to applicable clinical trials;
- counselling of patient and family;
- long-term prognosis; and
- long-term management and support.

Vitreoretinal Topic List

Pathology, aetiology, genetics, epidemiology, clinical manifestations, systemic manifestations, diagnostic criteria, and natural history of:

Medical Retinal Disease

(i) Congenital / hereditary

For the following congenital/inherited conditions, recognise clinical manifestations and any systemic associations; identify pattern of inheritance, and management principles

- Rod cone dystrophies
  - retinitis pigmentosa (RP) and variants
  - RP and deafness syndromes (Usher syndrome)
  - RP and systemic syndromes (Refsum, abetalipoproteinaemia, Kearns-Sayre syndromes; mitochondrial encephalomyopathy latic acidosis and stroke (MELAS); Bardet Biedel syndrome,
  - Leber congenital amaurosis

- Cone and cone rod dystrophy

- Stationary rod or cone dysfunction
  - congenital stationary night blindness
  - achromatopsia

- Macular dystrophies
  - Best disease
  - Stargardt disease / fundus flavimaculatus
  - pattern dystrophy of the RPE
  - adult vitelliform macular dystrophy
  - maternally inherited diabetes and deafness (MIDD)

- Retinal dysplasia
  - Norrie disease

- Vitreoretinopathies
  - congenital x-linked retinoschisis
  - Goldmann Favre / enhanced S-cone syndrome

- Choroidal dystrophies
  - choroideraemia
  - gyrate atrophy

- Developmental
  - ocular coloboma syndromes
  - persistent hyperplastic primary vitreous

- Systemic
  - albinism (oculocutaneous and ocular)
  - phakomatoses
  - pseudoxanthoma elasticum – angioid streaks
  - metabolic/storage diseases

- Combined hamartoma of retina and RPE

- Familial exudative vitreoretinopathy

- Retinopathy associated with TORCH syndromes (toxoplasma gondii, rubella virus, cytomegalovirus, and herpes simplex virus)

(ii) <u>Acquired</u>

- Infections
  - toxoplasmosis
  - toxocariasis
  - endophthalmitis: postoperative, traumatic, endogenous
  - viral retinitis syphilis
  - ocular histoplasmosis
  - tuberculosis
  - nematode infestation
  - onchocerciasis

- Inflammation
  - multifocal choroiditis +/- panuveitis
  - punctate inner choroidopathy (PIC)
  - multiple evanescent white dot syndrome (MEWDS)
  - acute zonal outer occult retinopathy (AZOOR)
  - acute macular neuroretinopathy (AMN)
  - acute posterior multifocal placoid pigment epitheliopathy (APMPPE)
  - pars planitis
  - sympathetic ophthalmia
  - Behçets syndrome
  - Vogt-Koyanagi-Harada syndrome
  - sarcoidosis
  - serpiginous choroidopathy
  - birdshot choroidopathy
  - uveal effusion syndrome
  - cancer associated retinopathies

- Benign neoplasm
  - choroidal naevus
  - choroidal haemangioma – diffuse and circumscribed
  - retinal capillary haemangioma
  - retinal cavernous haemangioma
  - retinal vasoproliferative tumour (reactive glioangiosis or pseudoangiomatous retinal gliosis)
  - melanocytoma
  - astrocytoma
  - sclerochoroidal calcification
  - choroidal osteoma
  - congenital hypertrophy of the RPE
  - arteriovenous malformation

- Malignant neoplasm
  - retinoblastoma/retinocytoma
  - uveal melanoma
  - choroidal metastasis
  - intraocular lymphoma
  - leukaemia

- Degenerative disease
  - age-related macular degeneration (AMD), including early and late signs
  - idiopathic polypoidal choroidal vasculopathy
  - retinal angiomatous proliferation
  - presumed ocular histoplasmosis syndrome (POHS)

  - pathologic myopia
  - epiretinal membrane (macular pucker)
  - choroidal folds
  - posterior staphyloma
  - maculopathy related to tilted disc syndrome

- Retinal toxicity of systemic and locally administered agents
  - chloroquine and hydroxychloroquine
  - phenothiazines
  - tamoxifen
  - aminoglycosides
  - other

- Retinal vascular disease
  - diabetic retinopathy
  - central retinal vein occlusion
  - branch retinal vein occlusion
  - central retinal artery occlusion
  - branch retinal artery occlusion
  - retinal emboli
  - hypertensive retinopathy and choroidopathy
  - retinal arterial macroaneurysm
  - ocular ischaemic syndrome
  - Coats disease / retinal telangiectasia
  - macular telangiectasia
  - retinopathy of prematurity
  - radiation retinopathy
  - retinal vasculitis
  - Eales disease
  - drug abuse
  - sickle cell retinopathy
  - familial exudative vitreoretinopathy

- Other
  - optic disc pit maculopathy
  - central serous chorioretinopathy
  - idiopathic uveal effusion syndrome
  - angioid streaks (non PXE)
  - cancer associated retinopathies
  - choroidal rupture
  - cystoid macular oedema
  - Valsalva retinopathy
  - Terson syndrome
  - Purtscher retinopathy

Surgical Retinal Disease

- Posterior vitreous detachment

- Vitreous haemorrhage

- Systemic and inherited diseases associated with rhegmatogenous retinal detachment
  - collagen type II, V, XI, XVIII, versican and fibrillin
  - Stickler syndrome
  - Knobloch syndrome

  - Marshall syndrome
  - Ehlers-Danlos syndrome
  - Wagner syndrome
  - Marfan syndrome
  - snowflake vitreoretinal degeneration (SVD)
  - autosomal dominant vitreoretinochoroidpathy (ADVIRC)

- Pre-detachment retinal and disc lesions and vitreoretinal degeneration
  - horse shoe tear
  - giant retinal tear
  - retinal dialysis
  - atrophic round holes
  - myopic macular hole
  - lattice degeneration
  - degenerative retinoschisis (see below)
  - vitrectomy related: entry site breaks trauma related: chorioretinitis sclopetaria
  - optic disc pit
  - morning glory anomaly
  - posterior segment coloboma

- Retinal detachment
  - rhegmatogenous
  - tractional breaks secondary to PVD
  - break without PVD: round hole, dialysis and myopic macular hole
  - related to disc pit, morning glory anomaly and coloboma
  - tractional
    - diabetic tractional detachment
    - sickle cell retinopathy
    - retinopathy of prematurity
  - exudative or serous
    - Coats disease
    - uveal effusion syndrome
    - ocular melanoma

- Degenerative retinoschisis
  - reticular retinoschisis
  - typical retinoschisis
  - inner leaf break
  - outer leaf break
  - schisis-detachment

- Choroidal detachment (serous and haemorrhagic)
  - hypotony induced choroidal effusion
  - uveal effusion syndrome
  - intraoperative choroidal haemorrhage
  - spontaneous choroidal haemorrhage

- Macular disorders
  - full-thickness macular hole
  - epiretinal membrane, pseudohole
  - vitreomacular traction syndrome
  - lamellar hole
  - macular micro hole
  - myopic foveoschisis with or without foveal detachment / macular hole

  - submacular haemorrhage
- Endophthalmitis
  - postoperative endophthalmitis
  - post-injection endophthalmitis
  - endogenous endophthalmitis (fungal, syphilitic and bacterial)
  - panophthalmitis

Trauma

- Management of different types of trauma
  - closed globe – contusion or lamellar laceration
  - open globe – rupture or laceration including penetrating, perforating, intraocular foreign body
  - principle of primary traumatic repair (to close the globe) and secondary repair (e.g. vitrectomy for vitreous haemorrhage)
  - management of closed globe injury (e.g. hyphema, retinal/choroidal contusion)
  - exploratory surgery (in severe globe rupture) and eye removal in trauma
  - management of traumatic cataract during globe repair
  - basic principles of anterior and pars plana vitrectomy in trauma

- Surgical planning
  - appropriate imaging and interpretation in order to assess the extent of the injury and to plan for surgical repair
  - preoperative management plan for traumatic case (e.g. anaesthetic consideration, antimicrobial/antibiotic cover, tetanus toxoid)
  - options for repair of different corneal and scleral wounds (e.g. stellate, shelved and non-shelved)
  - management of tissue loss (e.g. iris preservation, management of prolapsed choroid and retina)
  - use of tissue glue (e.g. cyanoacrylate)

Anterior segment related vitreoretinal conditions

- Retained lens fragment

- Dislocated intraocular lens

- Aqueous misdirection, malignant glaucoma

Other vitreous conditions or therapy associations

- Intravitreous drug delivery systems
  - ganciclovir, steroid or other implants

- Complications from gas and air
  - glaucoma
  - IOL dislocation

- Complications from silicone oil, heavy liquid and heavy oil tamponade
  - glaucoma
  - anterior chamber migration
  - subretinal heavy liquid and silicone oil

- Scleral buckles related conditions

- extrusion of scleral explant
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- Miragel expansion
- anterior segment ischaemia
- diplopia and muscle trauma from scleral explant
- scleral explant infection
- surgically induced necrotising scleritis

