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Neuro-ophthalmology Curriculum Standard

Purpose

The Neuro-ophthalmology Clinical Performance Standard covers the knowledge, process, skills and competencies required for the diagnosis and management of neuro-ophthalmic conditions.

Neuro-ophthalmology involves visual problems that are related to the nervous system. Neuro-ophthalmic disorders may be life threatening or sight threatening and require careful recognition and prompt management. A thorough knowledge of the diagnosis, examination, investigation and management of neuro-ophthalmic disorders is an essential skill for the ophthalmic trainee to acquire.

References

Neuro-ophthalmology Reading
In addition to the core texts, the following references are recommended:


Key Neuro-ophthalmology Randomised Clinical Trials

Additional Reading


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

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:

<table>
<thead>
<tr>
<th>Level</th>
<th>Description</th>
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<tbody>
<tr>
<td>***</td>
<td>Core knowledge of which trainees must be able to demonstrate understanding Skills and procedures that trainees must be able to perform autonomously</td>
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<tr>
<td>**</td>
<td>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</td>
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<tr>
<td>*</td>
<td>Knowledge, skills and procedures of which trainees must have some understanding</td>
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</tbody>
</table>
## Learning outcomes and performance criteria

### NO1 GENERAL MEDICAL AND OCULAR HISTORY RELEVANT TO NEURO-OPHTHALMIC CONDITIONS

This element covers the processes for observing, prompting and recording a general medical and ocular history in preparation for diagnosis and treatment of neuro-ophthalmic 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.

<table>
<thead>
<tr>
<th>LEARNING OUTCOMES</th>
<th>LEVEL OF MASTERY</th>
<th>PERFORMANCE CRITERIA</th>
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<tbody>
<tr>
<td>1.1 Take a thorough history of presenting symptoms, past medical history, ocular history and family history from a patient presenting with a neuro-ophthalmic condition</td>
<td>***</td>
<td>Identify and record features that have relevance to neuro-ophthalmic conditions in a patient who may present with the following symptoms:</td>
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<td>1.1.1 Visual loss</td>
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<td>1.1.2 Disorders of higher visual function including visual hallucinations</td>
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<td>1.1.3 Unexplained loss of vision</td>
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<td>1.1.4 Diplopia</td>
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<td>1.1.5 Proptosis or enophthalmos</td>
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<td>1.1.6 Ptosis</td>
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<td>1.1.7 Transient visual loss</td>
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<td></td>
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<td>1.1.8 Ansicoria</td>
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<td>1.1.9 Facial weakness or spasm</td>
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<td>1.1.10 Orbital pain or headache</td>
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<td>1.1.11 Nystagmus</td>
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</tbody>
</table>
NO2 PERFORM EYE EXAMINATIONS FOR NEURO-OPHTHALMIC CONDITIONS

This element covers the performance and interpretation of a range of eye examinations associated with neuro-ophthalmic conditions, as well as the demonstration of judgement 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. The trainee is also expected to have obtained and recorded a neuro-ophthalmic medical history.

<table>
<thead>
<tr>
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</table>
| 2.1 Undertake relevant neuro-ophthalmic examination based on presenting complaint | *** | 2.1.1 Perform, measure, record and interpret the results of the following examinations and note the relevance to the diagnosis of neuro-ophthalmic disease:  
- visual acuity (best corrected)  
- colour vision testing using Ishihara colour plates, HRR, or other office based techniques.  
- lid examination (identification, if present, of ptosis, retraction)  
- pupil examination – including pupil size in light and dark, shape, reaction, relative afferent pupillary defect and near response  
- ocular movements quantified with prisms  
- ascertain where there is deviation, whether it is comitant or incomitant  
- tests of muscle restriction, fatigability, saccadic, pursuit and nystagmoid movement including optokinetic nystagmus (OKN)  
- contrast sensitivity testing  
- photostress test  
- ocular motility with respect to specific nerve palsies – including patterns of lid abnormality  
- detect and quantify exophthalmos or enophthalmos  
- ascertain horizontal and vertical fusion range |
| 2.2 Perform slit lamp examination | *** | 2.2.1 Examine pupil, cornea, iris and lens and record and interpret observation of any disorders relevant to neuro-ophthalmic diagnosis |
| 2.3 Perform fundus examination | *** | 2.3.1 Examine fundus, record and interpret relevant retinal, choroidal, optic nerve and vitreal changes to aid neuro-ophthalmic diagnosis |
| 2.4 Perform visual field test to confrontation | *** | 2.4.1 Where relevant, perform and interpret results of confrontation visual field defect and be able to identify lesions that obey the vertical meridian |
| 2.5 Perform a neurological examination as relevant | *** | 2.5.1 Perform and interpret cranial nerve assessment |
| | | 2.5.2 Perform and interpret tests for tendon reflexes |
| | | 2.5.3 Perform and interpret basic office-based tests of vestibular and cerebellar function |
| 2.6 Undertake basic tests of higher cortical function | *** | 2.6.1 Where relevant, perform tests to identify alexia, agraphia, neglect |
| 2.7 Undertake appropriate examinations and tests to investigate functional visual loss | *** | 2.7.1 Where relevant, perform tests to identify functional visual loss |
NO3 SPECIAL NEURO-OPHTHALMIC TESTING

This element covers the performance and interpretation of a range of special neuro-ophthalmic tests associated with neuro-ophthalmic conditions. The trainee is required to demonstrate judgement in selecting the appropriate tests for particular patients.

<table>
<thead>
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</table>
| 3.1 Perform a range of specific tests to confirm diagnosis | 3.1.1 Perform, record and / or interpret the results of the following examinations and note the relevance to the diagnosis of neuro-ophthalmic disease:  
- automated threshold perimetry  
- Goldmann perimetry  
- Hess Screens testing  
- Ice and Rest tests  
- ocular ultra-sonography  
- optic disc morphology and nerve fibre layer abnormalities using contemporary investigative techniques, e.g. optical coherence tomography (OCT) | *** |
| | 3.1.2 Understand the performance and interpretation of the Tensilon test and:  
- recognise the parameters that are relevant to the diagnosis  
- identify the indications and contraindications for the test  
- be aware that necessary resuscitation equipment needs to be within easy reach | |
| | 3.1.3 Perform and interpret pharmacological pupil tests:  
- determine which pharmacological tests may be appropriate for investigation of anisocoria to identify the underlying cause | |
| | 3.1.4 Identify the indications for a fluorescein angiogram and interpret results | |
| | 3.1.5 Identify the indications for a temporal artery biopsy, perform surgery, use techniques that avoid possible complications and interpret results | |
| | 3.1.6 Identify optic nerve head drusen using ultrasound and/or autofluorescence | |
| 3.2 Use blood testing to confirm diagnosis | 3.2.1 Identify the indications for and interpret results from the following types of tests: |
|  | • haematology |
|  | • biochemistry |
|  | • microbiology |
|  | • immunology |

| 3.3 Use genetic testing to confirm diagnosis | 3.3.1 Identify the indications for and interpret results from genetic testing |
|  | 3.3.2 Seek appropriate informed consent from the patient for testing |
|  | 3.3.3 Provide or arrange genetic counselling |

| 3.4 Use radiologic testing to confirm diagnosis | 3.4.1 Identify the indications for and interpret results of: |
|  | • x-rays |
|  | • computed tomography (CT) scans |
|  | • magnetic resonance imaging (MRI) scans |
|  | • magnetic resonance arteriography (MRA) |
|  | • magnetic resonance venography (MRV) |
|  | • digital subtraction angiography |
|  | 3.4.2 Understand which is the most relevant imaging modality for diagnosis or exclusion of a condition |

| 3.5 Use ultrasonography testing of carotid and vertebral vasculature to confirm diagnosis | 3.5.1 Identify the indications for and interpretation of results of Doppler studies |

| 3.6 Use the results of lumbar puncture testing to confirm diagnosis | 3.6.1 Identify the indications for, the principles of conducting and interpretation of results from a lumbar puncture |
| 3.7 Understand the rationale behind and results of electrophysiological testing | ** | 3.7.1 Identify the indications for conducting the following testing of retinal and visual pathways and interpret the results thereof:  
- electro-oculogram (EOG)  
- electro-retinogram (ERG)  
- electro-myogram (EMG)  
- multifocal ERG  
- pattern ERG  
- visual evoked potential (VEP) |
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<tbody>
<tr>
<td>3.8 Use muscle biopsy testing to confirm diagnosis</td>
<td>*</td>
<td>3.8.1 Identify the indications for and interpretation of fibre patterns of muscle biopsies for neuro-ophthalmology</td>
</tr>
<tr>
<td>3.9 Use auto-immune testing to confirm diagnosis</td>
<td>*</td>
<td>3.9.1 Identify the indications for skin and buccal mucous membrane biopsies, the patterns of staining and significance</td>
</tr>
</tbody>
</table>
**NO4 IMPLEMENT A NEURO-OPHTHALMIC MANAGEMENT PLAN**

*This element covers the management of neuro-ophthalmic conditions using observation, medical therapies and surgery, including postoperative care.

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

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<thead>
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</table>
| 4.1 Determine and document in medical records a management plan for each individual patient | *** | 4.1.1 Integrate information from the history and examination to determine likely prognosis  
4.1.2 Maintain legible records in accepted format on the proposed management plan and the briefing of the patient  
4.1.3 Chose appropriate management strategies  
4.1.4 Establish initial management targets |
| 4.2 Educate the patient on the proposed management plan | *** | 4.2.1 Explain the nature of the patient’s neuro-ophthalmic condition  
4.2.2 Explain clearly the proposed management regimen and its potential consequences  
4.2.3 Obtain and record the patient’s informed consent, where necessary, to the management plan. |
| 4.3 Manage neuro-ophthalmic conditions using medical therapies | *** | 4.3.1 Identify the indications and contra-indications for pharmacological therapies  
4.3.2 Monitor the efficacy of the therapy, identify complications of the therapy and make necessary adjustments to the management regimen  
4.3.3 Consult with relevant medical specialties on management plan |
| 4.4 Understand the indications for the use of systemic steroids | 4.4.1 Clearly outline:  
- indications, dosage and management regimen of the use of systemic steroids in neuro-ophthalmic conditions  
- potential complications  
- appropriate management strategies to address complications (for example, bone-sparing treatments, treatment for potential gastric ulceration etc.) |
| 4.5 Manage neuro-ophthalmic conditions using surgery | 4.5.1 Perform temporal artery biopsy:  
- counsel the patient on surgery, potential outcomes and potential complications  
- understand the details of the surgical procedure and management of intraoperative complications  
- provide follow-up and continuing care |
| 4.6 Describe the rationale behind the management of neuro-ophthalmic conditions using radiotherapy | 4.6.1 Identify methods of delivery of radiation therapy for eye, neck and visual pathways  
4.6.2 Be familiar with the selected technique including effects and complications |
| 4.7 Provide psychological support for patient | 4.7.1 Counsel patient on condition, likely prognosis and progression  
4.7.2 Provide genetic counselling  
4.7.3 Provide emotional support  
4.7.4 Refer patient to relevant support services |
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 Genetics 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.

- Temporal arteritis
- Optic neuritis and demyelinating disease and its differentiation from atypical optic neuritis
- Myasthenia Gravis
- III Cranial Nerve Palsy

Neuro-ophthalmology Topic List

- Visual loss
  - retinal disorders
    - big blind spot syndrome
    - acute macular neuro-retinopathy
    - multiple evanescent white dot syndrome (MEWDS)
    - birdshot choroidopathy
    - acute zonal occult outer retinopathy (AZOOR)
    - melanoma associated retinopathy (MAR)
    - cancer associated retinopathy (CAR)
    - occult maculopathy
    - cone dystrophy
    - vitamin A deficiency
    - retinitis pigmentosa
  - optic nerve conditions
    - retrobulbar optic neuritis
  - retrochiasmal conditions
  - cortical conditions
    - cortical blindness
• Conditions which present with sudden loss of vision
• Conditions which present with progressive loss of vision
• Unilateral swollen optic nerve
• Bilateral optic nerve swelling
• Pseudodisc swelling
• Non-metastatic manifestations of cancer
  – carcinoma associated
  – melanoma associated
  – optic neuropathy
• Phakomatoses
  – neurofibromatosis Type I and II
  – racemose haemangioma
  – tuberous sclerosis
  – Von-Hippel-Lindau
  – Kippel-Trelauny-Weber
• Retinal vascular
  – retinal vein and artery occlusion – branch, hemi and central
  – haematological disorders: hypertension, anaemia, subacute bacterial endocarditis (SBE)
  – Vogt-Koyanagi-Harada disease
  – migraine
  – neurological disease with retinovascular manifestations e.g. Susac syndrome
  – retinal vasculitides
• Sclera
  – anterior scleritis
  – posterior scleritis
• Pupil
  – anatomy
  – normal – testing
  – abnormal – testing
  – describe clinical setting and relevance of:
    • Horner syndrome
    • light-near dissociation
    • aberrant regeneration
    • Adie pupil
    • afferent pupil defect
    • mid-brain / Parinaud pupil
    • pharmacological blockade / tests
    • Raede paratrigeminal neuralgia
    • episodic dysfunction
    • traumatic pupillary abnormalities
• Orbit (orbital disease relevant to neuro- ophthalmology)
  – thyroid eye disease
    • discuss the pathogenesis and clinical manifestation
• describe the investigations and diagnosis (differential radiological findings)
• explain the immune function and its clinical relevance
• understand the principles of management:
  – systemic — explain the basis of treatment of thyrotoxicosis (medical, surgical and radiological treatments) and its effects on the eye
  – ophthalmic — describe the principles, order and timing of treatment

• Tumours
  – describe the modes of clinical presentation, treatments and complications of:
    • meningiomas
      – optic nerve sheath
      – spheroidal wing
      – optic disc swelling associated with meningiomas and optico-ciliary shunt vessels, differential diagnosis of shunt vessels
      – olfactory nerve
      – understand treatment options including modality of radiotherapy, dosage and complications of radiotherapy
    • lymphoma – relevant systemic and ophthalmic investigations from a suspected diagnosis of lymphoma
    • secondary tumours of the orbit – relevant systemic and ophthalmic investigations from a suspected diagnosis of a secondary tumour
      – vascular fistulas – clinical manifestations, the carotico-cavernous fistula, dural fistula
      – inflammation
        • non-infective: idiopathic orbital inflammation syndrome (Tolosa-Hunt)
        • infective: viral, bacterial and fungal infection or orbit and adnexal (including mucormycosis, sinusitis, orbital cellulitis and adnexal infection)

• Optic nerve
  – congenital abnormalities
  – true swelling versus pseudo-swelling — understand conditions mimicking optic disc swelling: optic nerve head drusen, hypermetropic discs
  – swollen disc:
    • hereditary
    • infiltrative
    • papilloedema (elevated intra-cranial pressure, idiopathic intra-cranial hypertension)
    • optic papillitis
    • granulomas on optic nerve — e.g. sarcoid, toxoplasmosis
    • infective — cat scratch
    • anterior ischaemic optic neuropathy: non-arteritic and arteritic
  – optic neuropathies
    • toxic
    • infiltrative
    • hereditary
    • radiation
    • ischaemic
    • compressive
    • traumatic
    • inflammatory
  – conditions mimicking optic disc swelling: optic nerve head drusen
• Ocular myasthenia
  – discuss the aetiology, clinical presentation and association with general myasthenia
  – describe clinical findings and specific clinical tests including ice test, Tensilon test, fatiguing, Cogan lid twitch, single muscle fibre electromyography (EMG)
  – discuss relevant systemic investigations and their significance
  – understand the principles of management of myasthenia gravis with appropriate referral to a neurologist for systemic disease management

• Mitochondrial diseases
  – describe acute and chronic presentations and their investigations, epidemiology, characteristic fundus findings
  – Leber hereditary optic neuropathy (LHON) common mutation
  – chronic progressive external ophthalmoplegia

• Myotonic dystrophy

• Eye movement disorders
  – III nerve
    • clinical presentation including signs of aberrant regeneration
    • develop appropriate differentials of the cause of the III nerve palsy based on clinical presentation and age e.g. aneurysm, microvascular, other causes
    • perform appropriate investigation(s)
  – IV nerve
    • recognise clinical presentation of IV nerve palsy.
    • determine the differentials (e.g. congenital versus acquired) based on the clinical findings
    • perform appropriate investigation and discuss appropriate management options.
  – V nerve
    • recognise clinical presentations of V nerve dysfunction, e.g. trigeminal neuralgia, orbital apex syndromes, cavernous sinus lesions
    • manage clinical complications of V nerve dysfunction, e.g. neusotrophic keratitis
  – VI nerve
    • recognise clinical presentations of VI nerve palsy.
    • able to generate appropriate differential e.g. trauma, tumour, microvascular
    • aware of conditions that may present with VI nerve palsy as a false localising sign
  – VII nerve
    • upper motor neuron versus lower motor neuron
    • blepharospasm
    • hemifacial spasm
    • botulinum toxin treatment including pharmacology and treatment effect
    • meiges syndrome
    – combined cranial neuropathies
      • recognise the clinical features of combined cranial neuropathies
      • localization of pathology and discuss differentials of the cause

• Abnormal vertical dissociation and deviation

• Supranuclear gaze disorders
  – progressive supranuclear palsy: Steele-Richardson syndrome
  – Parkinsonism
• Nystagmus – methods for diagnosing, clinical types and localising types
  – upbeat
  – downbeat
  – horizontal
  – rotary
  – convergence / retraction
  – opsoclonus
  – periodic alternating
  – latent
  – congenital
  – pendular
  – monocular
  – seesaw

• Optic chiasm
  – pituitary tumour
    • describe the generalised clinical manifestation (e.g. acromegaly, Cushing, Addison)
    • explain the spectrum of visual field and central visual loss
    • recognise urgency and presentation of pituitary apoplexy
    • impact of modality of treatment on visual function
    • understand indications of neuro-surgery and endocrine treatments
  – craniopharyngiomas (particularly in children)
  – meningiomas (including those occurring more posteriorly impinging on visual pathways and eye movement)

• Optic tract and radiation
  – describe the visual field defects – congruous vs. non-congruous, macula sparing vs. macular involving
  – anatomical characterisation and localization on the visual pathway

• Lateral geniculate nucleus related visual field defects

• Higher disorders of visual function
  – visual hallucination (pathological, non-pathological) – differential diagnosis of formed and unformed visual hallucinations and their clinical significance
  – ocular manifestations of dementia

• Transient visual loss
  – embolic
  – amaurosis fugax: clinical manifestations and association with vascular disease; appropriate investigations and treatment
  – ocular ischaemic syndrome
  – hypoperfusion
  – transient obscuration associated with disc swelling
  – migraine
  – ocular surface disorders
  – in association with optic nerve tumour
  – in association with glaucoma

• Functional visual loss
Neuro-ophthalmology Curriculum Standard

- paediatric – including strategies for detection
  - adult

- Headaches
  - sinus
  - neck disease
  - migraine
    - ophthalmoplegic migraine
    - retinal migraine
    - common migraine
    - classic migraine
    - variants – cluster headache
    - acephalgic migraine
  - occipital stroke
  - carotid artery and vertebral artery dissection
  - tumours
  - tension
  - cluster
  - aesthenopic

- Idiopathic intra-cranial hypertension / pseudotumour cerebri
  - discuss the clinical manifestations
  - outline the investigations and differential diagnosis (including venous sinus thrombosis)
  - discuss management options

- Other systemic disease with neuro-ophthalmic manifestations
  - infective – include human immunodeficiency virus (HIV), tuberculosis (TB), syphilis, Lyme disease
  - non-infective – include Behçet disease
  - functional visual loss

- Combined cranial neuropathies
  - recognise the clinical features of combined cranial neuropathies
  - localisation of pathology and discuss differentials of the cause
  - superior orbital fissure syndromes
  - cavernous sinus disease

- Orbit (orbital disease relevant to neuro-ophthalmology)
  - thyroid eye disease
  - discuss the pathogenesis and clinical manifestation
  - describe the investigations and diagnosis (differential radiological findings)
  - explain the immune function and its clinical relevance
  - understand the principles of management
  - systemic - explain the basis of treatment of thyrotoxicosis (medical, surgical and radiological treatments) and its effects on the eye
  - ophthalmic - describe the principles, order and timing of treatment

- Orbital apex syndromes