Given the complexity and quantity of clinical knowledge required to correctly identify and treat ocular disease, a quick reference text with high quality color images represents an invaluable resource to the busy clinician. Despite the availability of extensive resources online to clinicians, accessing these resources can be time consuming and often requires filtering through unnecessary information. In the exam room, facing a patient with an unfamiliar presentation or complicated medical problem, this series will be an invaluable resource.

This handy pocket sized reference series puts the knowledge of world-renowned experts at your fingertips. The standardized format provides the key element of each disease entity as your first encounter. The additional information on the clinical presentation, ancillary testing, differential diagnosis and treatment, including the prognosis, allows the clinician to instantly diagnose and treat the most common diseases seen in a busy practice. Inclusion of classical clinical color photos provides additional assurance in securing an accurate diagnosis and initiating management.

Regardless of the area of the world in which the clinician practices, these handy references guides will provide the necessary resources to both diagnose and treat a wide variety of ophthalmic diseases in all ophthalmologic specialties. The clinician who does not have easy access to sub-specialists in Anterior Segment, Glaucoma, Pediatric Ophthalmology, Strabismus, Neuro-ophthalmology, Retina, Oculoplastic and Reconstructive Surgery, and Uveitis will find these texts provide an excellent substitute. World-wide recognized experts equip the clinician with the elements needed to accurately diagnose treat and manage these complicated diseases, with confidence aided by the excellent color photos and knowledge of the prognosis.

The field of knowledge continues to expand for both the clinician in training and in practice. As a result we find it a challenge to stay up to date in the diagnosis and management of every disease entity that we face in a busy clinical practice. This series is written by an international group of experts who provide a clear, structured format with excellent photos.

It is our hope that with the aid of these six volumes, the clinician will be better equipped to diagnose and treat the diseases that affect their patients, and improve their lives.

Marian S. Macsai and Jay S. Duker
If your patient has a visual problem or funny eye movements and you cannot quite figure out what is going on, chances are the problem falls within the domain of neuro-ophthalmology.

This is neuro-ophthalmology in a shot glass - short but potent. It is all here in concentrated form - the manifestations, the disease mechanisms, the pitfalls, the practical guidelines, and the pictures.

The text is bulleted for easy grasp. The fundus photographs are my best-in-show from material extending back over 40 years of clinical practice. The brain imaging illustrations are prepared to highlight the lesions. Eye movement and alignment abnormalities are a challenge to illustrate without videos; I have chosen to present schematic illustrations because still photographs so rarely tell the story properly.

I hope that you will find this book useful and that it will make you wonder at the marvels of the nervous system.

Jonathan D. Trobe
Section 1

Transient Visual Loss

Transient Monocular Visual Loss (Amaurosis Fugax)  2
Transient Binocular Visual Loss  4
Migraine with Typical Visual Aura  6
Retinal Migraine (Presumed Retinal Vasospasm)  8
Transcranial Magnetic Stimulation

Key Facts
- Abrupt visual loss affecting one eye that lasts <60 min
- Sometimes associated with scintillations (photopsias, positive visual phenomena)
- Caused by reduced perfusion of eye (ocular transient ischemic attack, TIA)
- **Common causes:**
  - cervical carotid stenosis
  - systemic hypotension
  - idiopathic (possible retinal artery vasospasm)
  - impending retinal or optic nerve infarction
  - papilledema
- Evidence that carotid endarterectomy benefits patients suffering only ocular TIA is weak

Clinical Findings
- Eye examination is usually normal but may show intra-arterial retinal platelet–fibrin–cholesterol (Hollenhorst) plaque, optic disc edema, or venous stasis retinopathy

Ancillary Testing
- Carotid ultrasound, CT angiography, or magnetic resonance angiography to rule out stenosis, dissection, and dysplasia
- Blood pressure (including orthostatic) testing to rule out hypertension or hypotension
- Electrocardiography to rule out atrial fibrillation
- Cardiac echography to rule out cardioembolic source
- **Blood tests to rule out hypercoagulable state:**
  - complete blood count
  - serum protein electrophoresis
  - prothrombin and partial thromboplastin times
  - antiphospholipid antibodies
  - antithrombin-3
  - factor V Leiden
  - prothrombin gene mutation
  - homocysteine
  - sickle hemoglobin
  - serum viscosity

Differential Diagnosis
- Embolism from cervical carotid artery, aortic arch, or cardiac valve or wall
- Systemic hypertension or hypotension
- Hypercoagulable state
- Impending retinal vascular occlusion
- Ischemic oculopathy
- Retinal vasospasm (see Retinal migraine)
- Papilledema

Treatment
- Direct at underlying condition
- Endarterectomy often advocated for >70% ipsilateral cervical carotid stenosis, but evidence of benefit for purely ocular TIA is weak
- Reduce modifiable arteriosclerotic risk factors (diabetes, hypertension, dyslipidemia, lack of exercise, obesity, cigarette smoking)
- Correct very high blood pressure but avoid excessive blood pressure lowering (may lead to perfusion failure and stroke of eyes or brain)
- Aspirin 81 mg/day for underlying arteriosclerosis

Prognosis
- Depends on underlying condition
- *Amaurosis fugax* is an old term that is out of favor because it does not specify whether transient visual loss is monocular or binocular
Fig. 1.1 Hollenhorst plaque. The refractile yellow dot (arrow) is an impacted platelet–fibrin embolus that traveled from the ipsilateral common carotid artery bifurcation in the neck. It produced transient visual loss by causing ischemia to the retina. Caution: most patients with transient monocular visual loss have no abnormalities on fundus examination.

Fig. 1.2 Cervical carotid stenosis. The critical narrowing of the proximal internal carotid artery (arrow) is the result of arteriosclerosis. This lesion probably gave rise to the Hollenhorst plaque in Fig. 1.1.
Transient Binocular Visual Loss

Key Facts
- Abrupt visual loss affecting both eyes and lasting <60 min
- Caused by dysfunction of visual cortex or optic nerves in both eyes
- Common causes:
  - migraine
  - visual cortex transient ischemic attack (TIA)
  - visual cortex seizure
  - papilledema

Clinical Findings
- **Migraine:** scintillations (often with zigzag border) that march across one hemifield and last 20–30 min; headache may follow
- **TIA:** homonymous or total visual loss lasting minutes that may have scintillations; headache is rare
- **Focal seizure:** stationary, often colored visual scintillations lasting seconds to minutes; persistent homonymous hemianopia sometimes
- **Papilledema:** blackouts of vision lasting seconds, often precipitated by upright posture

Ancillary Testing
- **If suspecting TIA:**
  - echocardiography or vascular imaging may show source of emboli
  - cardiac telemetry may show atrial fibrillation
  - blood pressure testing may show hypertension or hypotension
- **If suspecting focal seizure:**
  - brain imaging may show mass, encephalomalacia, enhancing meninges
  - electroencephalography may show epileptic focus

Treatment
- **Migraine:** usually no treatment unless headaches are frequent and debilitating
  - If so, treat with standard prophylactic agents (beta blockers, calcium channel blockers, valproic acid, topiramate, aspirin)
- **Visual cortex TIA:**
  - warfarin if cardiogenic emboli, otherwise aspirin 81 mg/day or clopidogrel 75 mg/day
  - correct discretionary arteriosclerotic risk factors
- **Visual cortex seizure:** treat underlying lesion if possible, anticonvulsants
- **Papilledema:** treat underlying cause

Prognosis
- Depends on underlying condition
Fig. 1.3 Epileptogenic occipital mass. Postcontrast axial T1 MRI shows left occipital mass (arrow) that proved on resection to be metastatic breast cancer. It provoked seizures manifested by transient binocular sparkling (scintillations) in the right hemifield and a partial right homonymous hemianopia.
Migraine with Typical Visual Aura

Key Facts
- Episodic neuronal discharge starting in brainstem and ending in visual cortex
- Usually no underlying disease (primary headache disorder)
- Typical visual aura (hallucination) is a bright (scintillating) scotoma with zigzag (fortification) border that traverses the hemifield in 20–30 min
- Headache, photophobia, or sonophobia may follow aura and last ≤48 h
- Visual aura may occur without headache (acephalgic migraine, dissociated migraine), especially in older adults
- First attack usually occurs before age 30 but may occur much later

Clinical Findings
- Visual fields may show homonymous hemianopia during the aura
- No ophthalmic abnormalities after aura has dissipated
- Diagnosis based entirely on history

Ancillary Testing
- Brain imaging necessary only if visual fields are abnormal, patient has other neurologic manifestations, or symptoms are atypical of migraine

Differential Diagnosis
- Visual cortex transient ischemic attack
- Visual cortex seizure
- Papilledema
- Migrainous event secondary to underlying cerebral pathology (mitochondrial disorders, lupus erythematosus, occipital lobe arteriovenous malformations, and cerebral autosomal dominant arteriopathy with strokes and leukoencephalopathy [CADASIL])

Treatment
- Eliminate precipitating factors if possible
- Prescribe migraine prophylactic agents (beta blockers, calcium channel blockers, topiramate, valproic acid, aspirin) if attacks are frequent and debilitating
- Prescribe migraine abortive agents (triptans) if non-visual symptoms are debilitating
- Advise patient to discontinue smoking and contraceptive medication because they increase risk of stroke

Prognosis
- Abortive agents have no effect on visual aura but usually attenuate non-visual symptoms
- Prophylactic agents usually reduce attack frequency and intensity
- Risk of stroke is small but not negligible if patient is a smoker or uses oral contraceptive medication