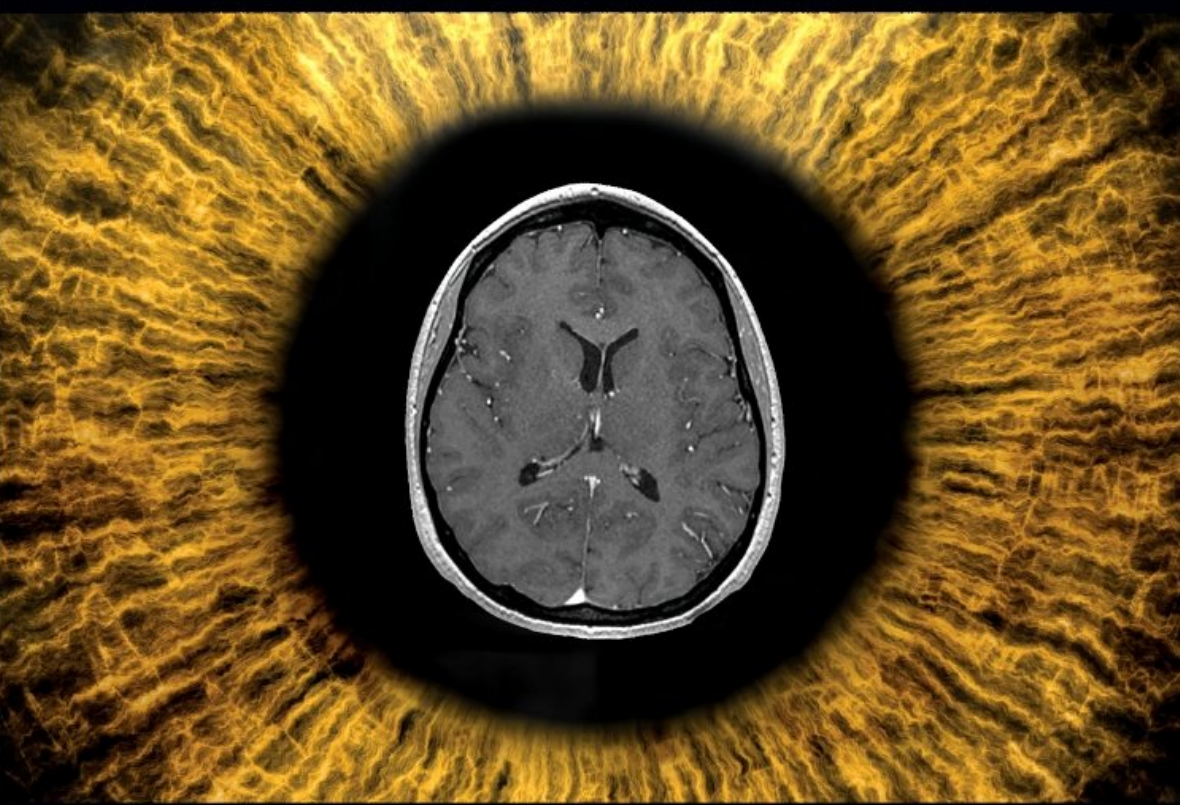
THE NEURO-OPHTHALMOLOGY SURVIVAL GUIDE

Second Edition



Anthony Pane
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Video contents

Index of key management flowcharts

Index of key clinical diagnostic criteria

Preface

Whether you are a busy optometrist who primarily performs refractions, an ophthalmologist who sees patients with cataracts or glaucoma, or a neurologist who sees a lot of patients with headache, you never know when a patient with a potentially vision- or life-threatening disorder will come to your clinic with a visual problem. Over the many years that we have practiced neuro-ophthalmology, we have encountered many patients who became permanently blind or neurologically impaired or who died because their otherwise skilled and well-meaning ophthalmologists, optometrists, or neurologists failed to **recognize** that they had a potentially devastating but treatable neuro-ophthalmic condition. This, despite the existence of many excellent and detailed neuro-ophthalmology texts. The problem is that none of these texts are written for the vast majority of practitioners who have no particular interest or expertise in neuro-ophthalmology. In addition, most of these texts are diagnosis-based and, therefore, only helpful once the diagnosis had been made. However, in our opinion, the three most difficult challenges for most practitioners are to recognize that their patient has a "neuro-ophth" problem in the first place, then to make the correct diagnosis, and, finally, to provide appropriate treatment in a timely fashion.

To address these challenges, we set out ten years ago to write a simple, practical clinical guide to benefit practitioners and their trainees. We made the guide symptom-based; i.e. listen to the patient's concern (e.g. "I see double"; "the vision in my left eye is slowly worsening"), turn to the appropriate chapter (e.g. Chapter 5: Double vision; Chapter 2: Blurred vision or field loss), and let the book guide you every step of the way to the correct diagnosis and treatment without presuming that you have any previous neuro-ophthalmic training.

In the ten years since the book's publication, there have been many advances in our ability to diagnosis and treat neuro-ophthalmic conditions. Accordingly, this second edition of

The Neuro-Ophthalmology Survival Guide provides an updated but still carefully structured approach. Specifically, it tells you what questions to ask, what to look for during the examination, what diagnostic tools might be useful to make the correct diagnosis, and, depending on the diagnosis, what the management options are, all using clear and simple bullet points and flowcharts. Also unique to this book are 60 videos showing various important eye movement and pupil abnormalities and some important examination techniques.

Neuro-ophthalmology is the "needle in the haystack" in the busy clinics of all practitioners who see patients with visual complaints. We hope this book helps you avoid sitting on too many of these needles.

AP
NRM
MB

CHAPTER 1

Staying out of trouble

CONTENTS

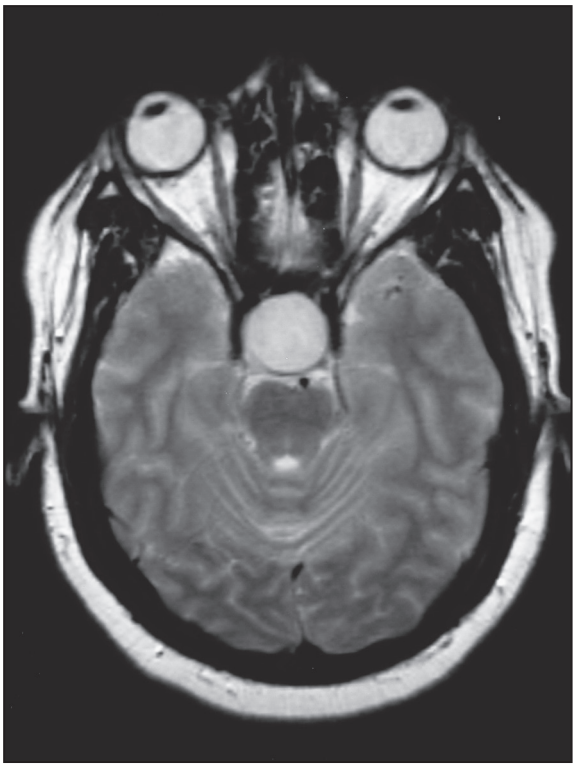
If you read only one chapter in this book, read this one. It covers the most common and most serious mistakes made by ophthalmologists and ophthalmic trainees when dealing with neuro-ophthalmic patients.

Twenty neuro "rules" to keep you out of trouble

The following 20 practice guidelines have a good chance of keeping your patients (and you) safe. Naturally, as with all "rules", there are rare exceptions to all of these, but they are still useful to keep in the back of your mind in the clinic or ophthalmic emergency department.

Patient presentation

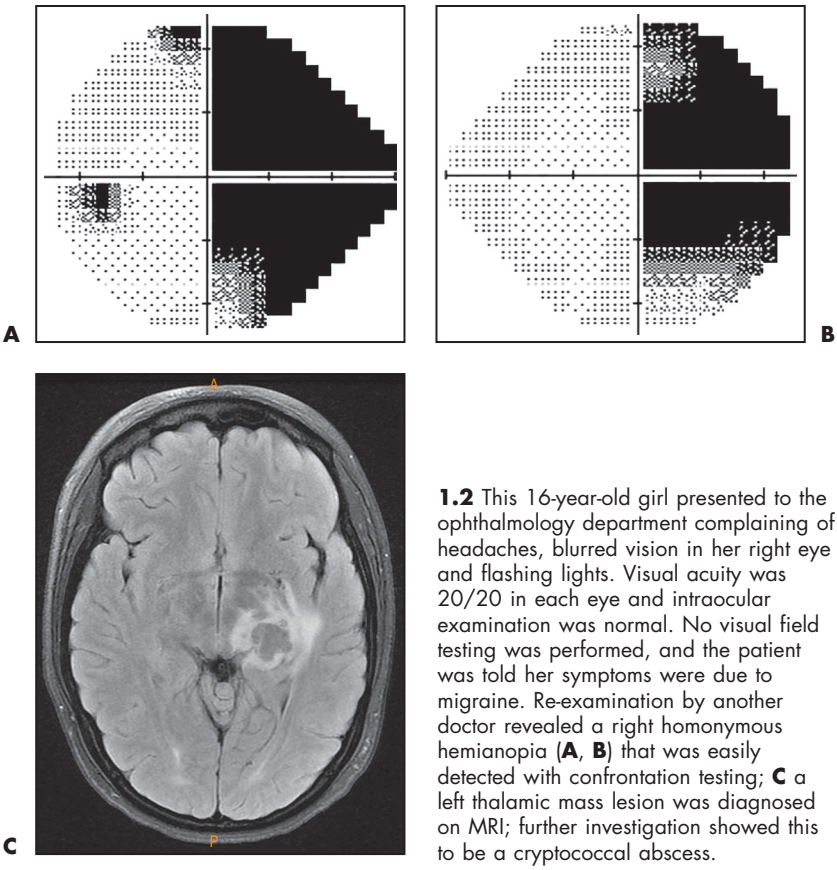
1. Beware the "silent" neuro-ophthalmic patient!
   - patients with optic nerve or brain tumors will sometimes be referred to you as "cataract", "glaucoma", "optic neuritis", "ischemic sixth nerve palsy", "senile ptosis" or other benign-sounding diagnoses (Fig. 1.1)



1.1 This 62-year-old patient was referred to an ophthalmologist "for treatment of cataracts". On initial examination visual acuity was right 20/40, left 20/60 despite only very early cataracts being present; color vision was not assessed, visual fields were not tested and no pupillary examination was performed. It was only when vision did not improve after bilateral cataract surgery that another cause was eventually suspected; this pituitary tumor was diagnosed as the real cause of the patient's blurred vision 2 years after first ophthalmic contact. Final visual acuity after tumor excision was only 20/120 in each eye; visual outcome could have been improved by earlier diagnosis of the tumor.

Examination

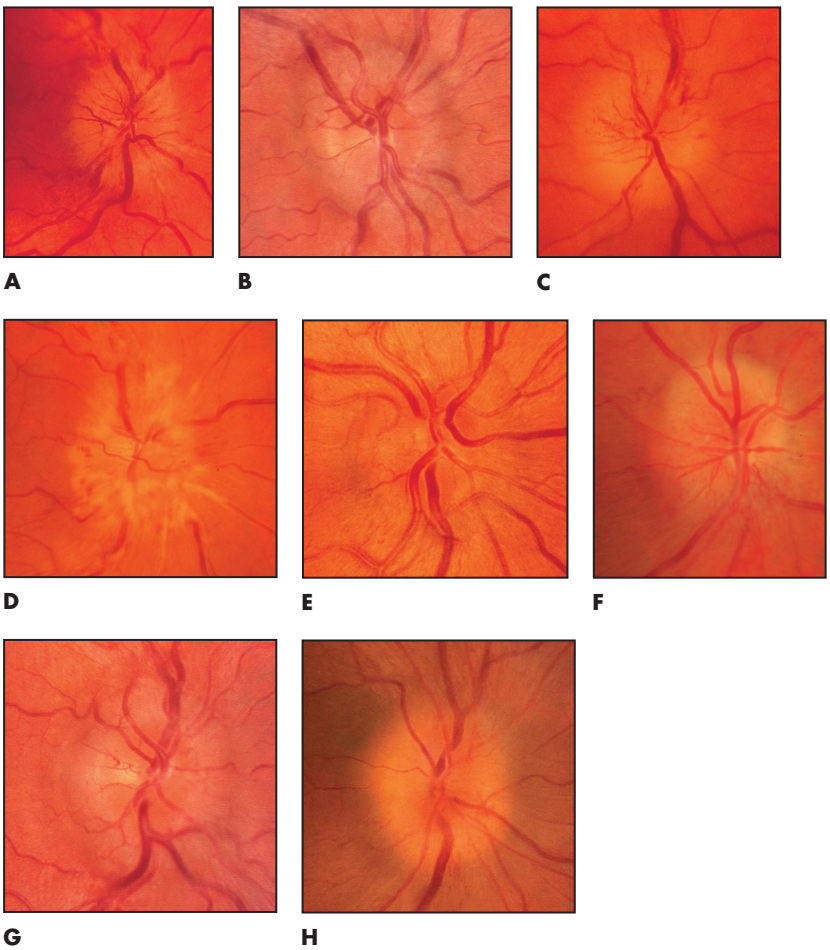
2. Every new eye patient complaining of blurred vision should have:
   - confrontation field testing (peripheral and central)
   - a "swinging torch test" for a relative afferent pupillary defect (RAPD) before dilation
   - perimetry if either of these is abnormal, the patient describes a field defect, or the degree of visual loss is not consistent with the ocular examination (Fig. 1.2)



1.2 This 16-year-old girl presented to the ophthalmology department complaining of headaches, blurred vision in her right eye and flashing lights. Visual acuity was 20/20 in each eye and intraocular examination was normal. No visual field testing was performed, and the patient was told her symptoms were due to migraine. Re-examination by another doctor revealed a right homonymous hemianopia (**A**, **B**) that was easily detected with confrontation testing; **C** a left thalamic mass lesion was diagnosed on MRI; further investigation showed this to be a cryptococcal abscess.

Blurred vision or field loss

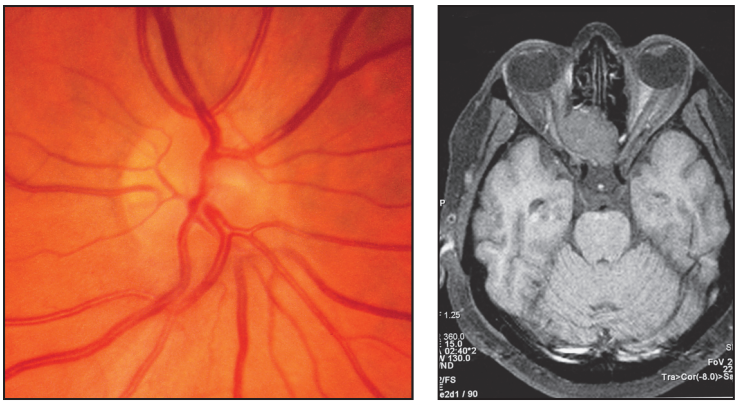
3. You can never diagnose the cause of optic nerve dysfunction just by looking at the disc (Fig. 1.3).



1.3 Hundreds of different diseases can present with a similar optic disc appearance. For example, these swollen discs have been caused by **A** sarcoid optic neuritis, **B** optic nerve infiltration by lymphoma, **C** non-arteritic AION, **D** papilledema, **E** Leber's hereditary optic neuropathy, **F** cat scratch disease, **G** idiopathic optic neuritis and **H** optic nerve sheath meningioma. In no case was the disc appearance diagnostic; diagnosis was made on careful history, other examination, perimetry and other investigations as suggested in the management flowchart on p. 31.

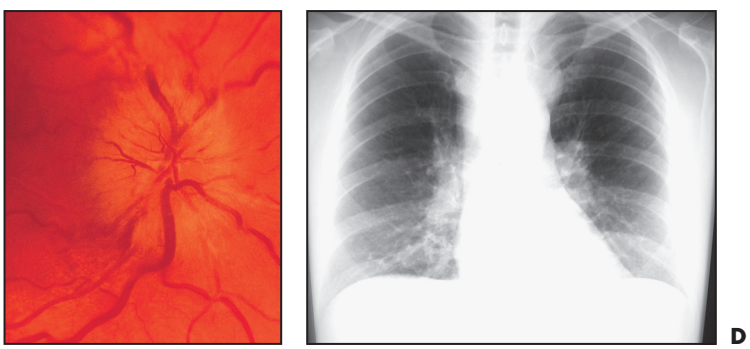
4. All patients with non-traumatic ACUTE optic nerve dysfunction who do not meet all the clinical diagnostic criteria for either:
   - typical optic neuritis (p. 35), or
   - anterior ischemic optic neuropathy (AION) (p. 36)

   require urgent referral to a neuro-ophthalmologist (or, if this is not possible, urgent investigation as suggested on p. 38) (Fig. 1.4).



A B

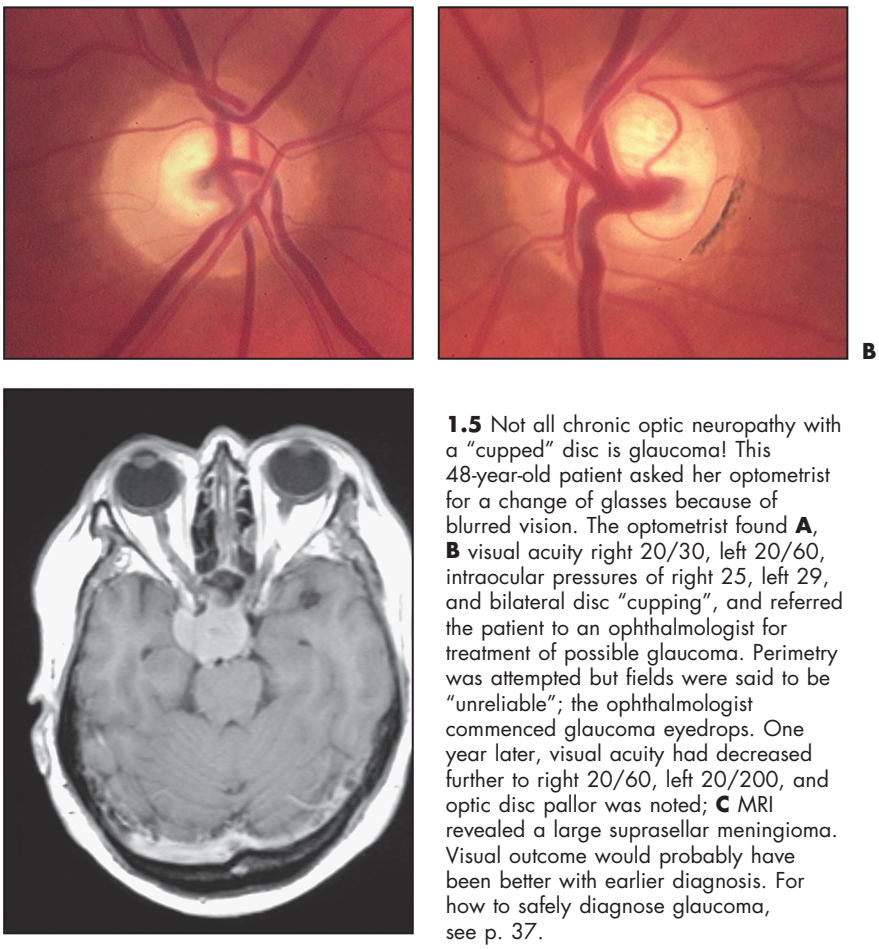
1.4 Not all acute optic neuropathies in young adults are optic neuritis! **A** This 32-year-old woman presented with progressive visual loss in the right eye over 3 weeks (right 20/60), pain behind the right eye, a right RAPD and normal optic discs. Her ophthalmologist diagnosed "retrobulbar optic neuritis" and reassured the patient that her vision would return spontaneously. Three months later, vision had worsened (20/200) and optic atrophy had developed; **B** MRI showed this large nasal tumor compressing the right optic nerve. Vision did not improve after removal of the tumor. Visual outcome would probably have been better with earlier diagnosis. For how to safely diagnose typical optic neuritis, see p. 35.



C D

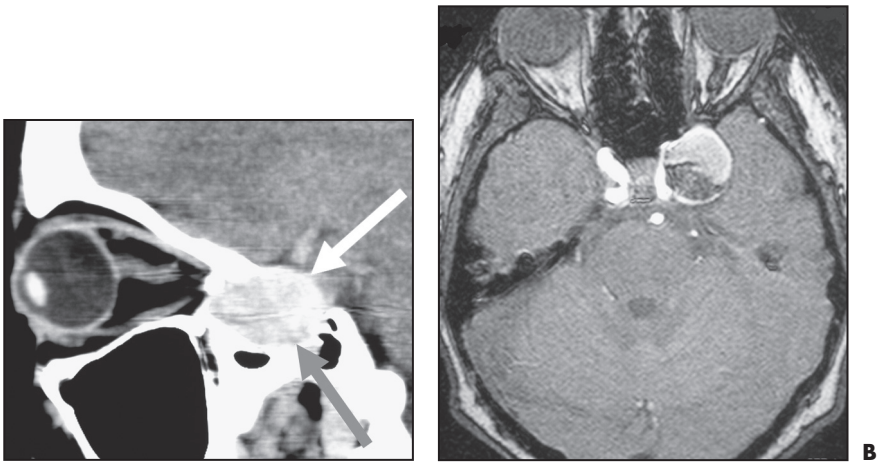
1.4 Not all acute optic neuropathies with a swollen disc are AION! This 55-year-old hypertensive man complained of progressive loss of vision in his right eye over 7 days. His ophthalmologist found **C** right visual acuity to be 20/40, a right RAPD and right optic disc swelling. A right inferior altitudinal scotoma was detected on perimetry. The ophthalmologist diagnosed "anterior ischemic optic neuropathy" and advised the patient that there was no treatment. Ten weeks later, right visual acuity (VA) had deteriorated to 20/400 and the right disc had become pale; further investigation revealed an increased serum angiotensin converting enzyme (ACE) and **D** hilar lymphadenopathy on chest x-ray. Biopsy of a lower eyelid conjunctival granuloma confirmed the diagnosis of sarcoidosis. Because diagnosis was delayed, right VA only returned to 20/80 with steroid treatment. For how to safely diagnose AION, see p. 36.

5. All patients with CHRONIC optic nerve dysfunction who do not meet all the clinical diagnostic criteria for glaucomatous optic neuropathy (p. 37) require referral to a neuro-ophthalmologist (or, if this is not possible, investigation as suggested on p. 38) (Fig. 1.5).



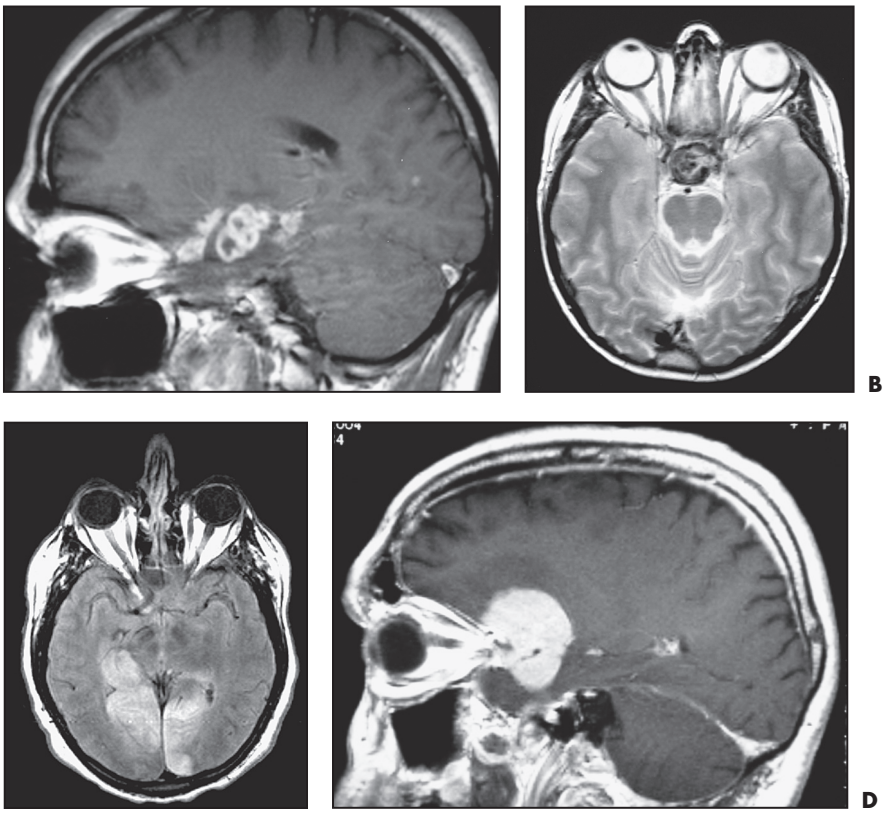
1.5 Not all chronic optic neuropathy with a "cupped" disc is glaucoma! This 48-year-old patient asked her optometrist for a change of glasses because of blurred vision. The optometrist found **A**, **B** visual acuity right 20/30, left 20/60, intraocular pressures of right 25, left 29, and bilateral disc "cupping", and referred the patient to an ophthalmologist for treatment of possible glaucoma. Perimetry was attempted but fields were said to be "unreliable"; the ophthalmologist commenced glaucoma eyedrops. One year later, visual acuity had decreased further to right 20/60, left 20/200, and optic disc pallor was noted; **C** MRI revealed a large suprasellar meningioma. Visual outcome would probably have been better with earlier diagnosis. For how to safely diagnose glaucoma, see p. 37.

6. Amblyopia is a specific diagnosis, with specific diagnostic features; never use a history of "lazy eye" as the explanation for worsening vision. Features of optic nerve disease should be absent and a demonstrable cause for the amblyopia should be present (Fig. 1.6).



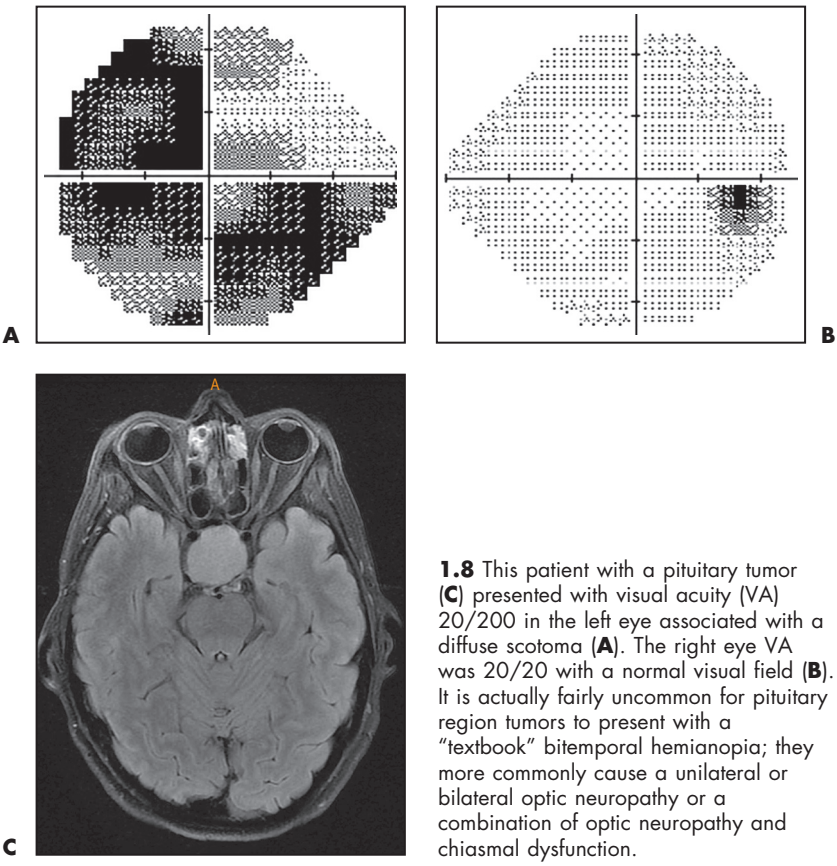
1.6 A 38-year-old woman presented to her ophthalmologist complaining of blurred vision in her left eye and the left eye "turning in". She said that the left eye had always been "a bit lazy" so the ophthalmologist recorded "left amblyopia" as the cause of the blurred vision without checking for an RAPD. The ophthalmologist diagnosed "decompensated congenital esotropia" and attributed the poor vision in the left eye (20/80) to "strabismic amblyopia". Eventually another ophthalmologist investigated the patient and found **A**, **B** a large internal carotid artery aneurysm causing compressive optic neuropathy and sixth nerve palsy. Unfortunately, by this time, the left eye was blind.

7. Whenever you look in an eye, think: is the level of vision explained by visible intraocular disease? If not, there could be disease behind the eye. Unexplained poor vision, optic atrophy, disc cupping or field loss always requires investigation (Fig. 1.7).



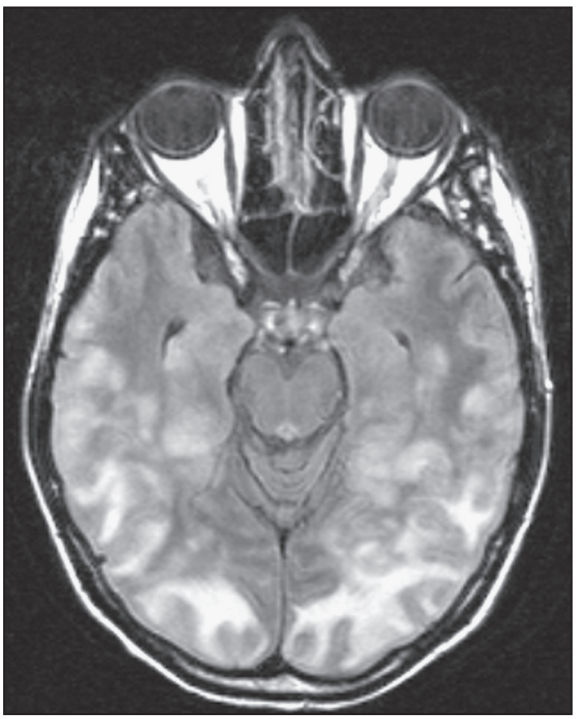
1.7 These patients were all initially referred to their local ophthalmologists "for treatment of cataracts": **A** tuberculous meningitis and optic neuritis; **B** craniopharyngioma; **C** bilateral occipital lobe stroke causing "cortical blindness"; **D** sphenoid wing meningioma. Suspect that the patient is not "just another cataract" if he or she has one or more of the following: visual acuity loss greater than that expected from the density of the cataracts; color vision loss; RAPD; or visual field loss (described by the patient or identified on confrontation testing). Optic disc pallor is helpful only when present (the discs may look entirely normal in early optic neuropathy); by the time pallor develops, irreversible damage has often occurred.

8. Compressive optic neuropathy from an orbital or brain tumor (including pituitary tumors) can present with:
   - ANY optic disc appearance (normal, swollen, pale or "cupped")—one or both eyes
   - ANY sort of field loss (including arcuate scotoma or altitudinal field defect)—one or both eyes (Fig. 1.8).



1.8 This patient with a pituitary tumor (**C**) presented with visual acuity (VA) 20/200 in the left eye associated with a diffuse scotoma (**A**). The right eye VA was 20/20 with a normal visual field (**B**). It is actually fairly uncommon for pituitary region tumors to present with a "textbook" bitemporal hemianopia; they more commonly cause a unilateral or bilateral optic neuropathy or a combination of optic neuropathy and chiasmal dysfunction.

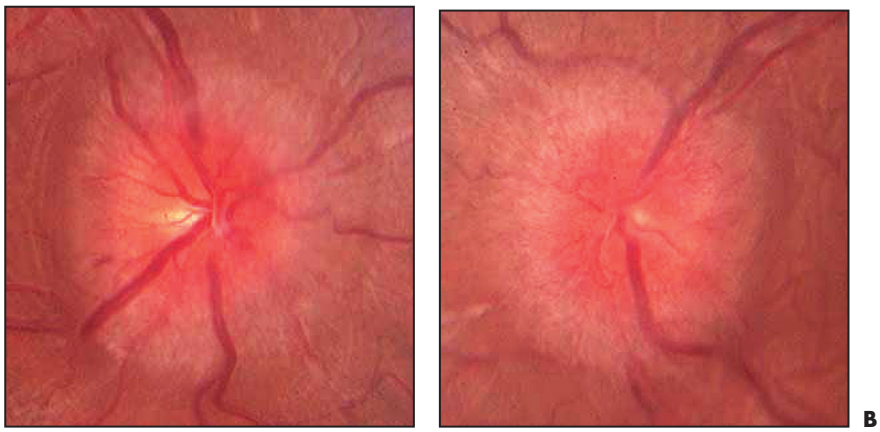
9. Just because both eyes look completely normal and there is no RAPD do not assume the patient has non-organic ("functional") visual loss; only diagnose this if you have thoroughly examined the patient, performed perimetry and have been able to "trick" the patient into demonstrating completely normal vision (see p. 34) (Fig. 1.9).



1.9 This 33-year-old man was thought by his ophthalmologist to have "functional visual loss". Despite complaining of poor vision in both eyes, he had normal pupil reactions and normal optic disc and retinal examination. The patient was referred to a neuro-ophthalmologist; MRI revealed posterior leukoencephalopathy affecting the optic radiations bilaterally.

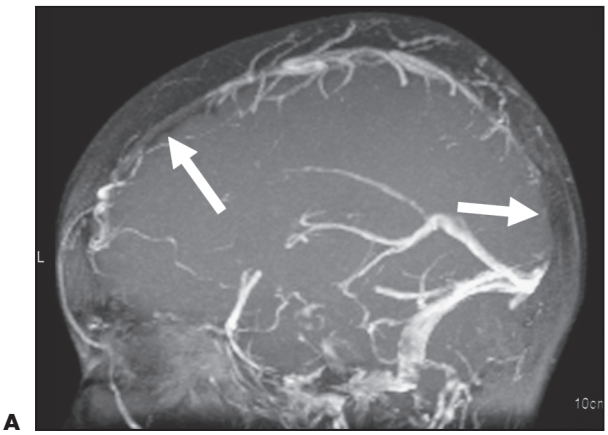
Bilateral disc swelling

10. Bilateral disc swelling could be papilledema (disc swelling due to raised intracranial pressure). The first investigation should be urgent (same-day) magnetic resonance imaging (MRI) plus magnetic resonance venography (MRV) to exclude a brain tumor or dural venous sinus thrombosis (see p. 120) (Fig. 1.10).

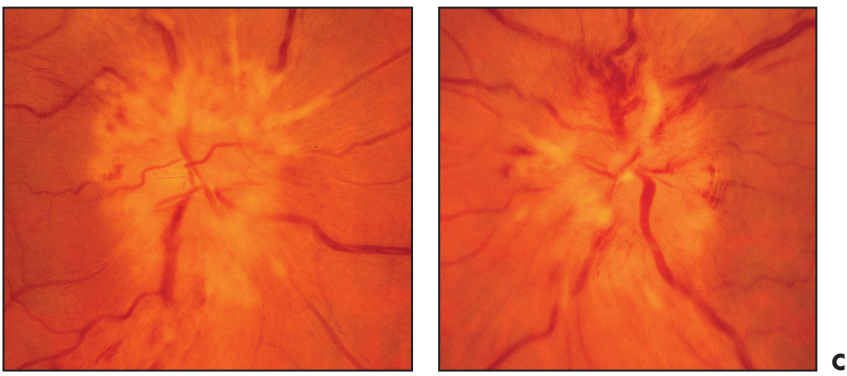


1.10 This 38-year-old man presented to an ophthalmic emergency department complaining of blurred vision and headaches. Examination revealed visual acuity 20/20 right and left and bilateral moderate disc swelling **A**, **B**. Because vision was good and there were "no other neurologic signs", the patient was allowed to go home and was scheduled for neuro-imaging as an outpatient. Two days later, while driving, the patient experienced a generalized seizure, resulting in severe injuries to himself and another driver. MRI revealed a brain tumor which was found at surgery to be an astrocytoma. For how to safely manage disc swelling with normal vision, see p. 110.

11. The diagnosis of primary pseudotumor cerebri (PTC), also called idiopathic intracranial hypertension (IIH), is based on specific diagnostic criteria (p. 113). MRI plus MRV and lumbar puncture must be performed in all cases of suspected IIH; not all women with disc swelling have IIH (Fig. 1.11).



A

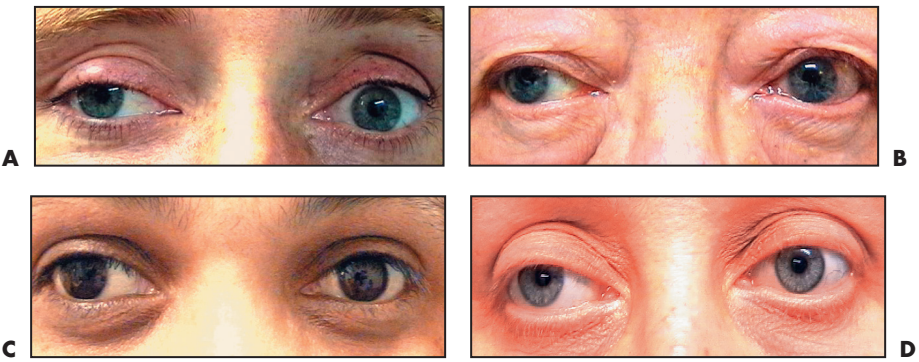


B C

1.11 Not all women with disc swelling have "IIH"! This 28-year-old, markedly obese woman presented to her ophthalmologist complaining of headaches and transient blurring of vision when she bent over or coughed. The ophthalmologist found visual acuities of 20/20 in both eyes and bilateral moderate optic disc swelling, diagnosed "benign intracranial hypertension" and requested a CT brain scan that was normal. The patient was treated with acetazolamide tablets and told to lose weight. One week later, the patient collapsed at home and was admitted to the neurologic intensive care unit; MRI plus MRV revealed a superior sagittal sinus thrombosis **A** and papilledema was noted to be very severe **B**, **C**. Despite intensive treatment, the patient suffered bilateral optic disc infarction secondary to the severe papilledema and final visual acuity was 20/200 in both eyes with bilateral postpapilledema optic atrophy. She was also left with permanent right-sided weakness due to secondary cerebral venous infarction. Earlier diagnosis might have resulted in a better visual and neurologic outcome. For how to safely manage disc swelling with normal vision, see p. 110.

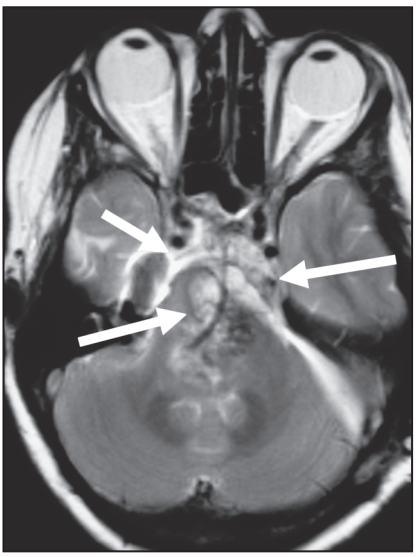
Double vision

12. Beware the "spot diagnosis" in acquired strabismus. Many diseases that cause motility disturbance can present with the same clinical picture (Fig. 1.12).

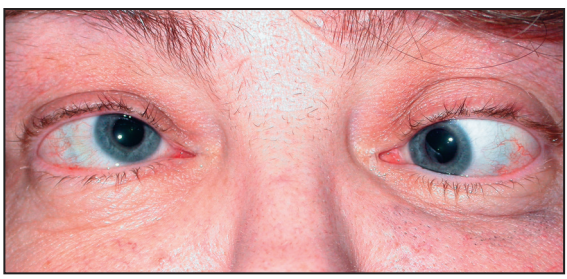


1.12 Right exotropia with decreased adduction of the right eye caused by: **A** partial third nerve palsy due to aneurysm; **B** myasthenia; **C** right lateral rectus myositis; and **D** internuclear ophthalmoplegia. Eye appearance and pursuit "eye movement" testing was not diagnostic in any case; diagnosis was made by a careful history, a detailed examination including saccade testing and other investigations. See p. 174 for a management flowchart for diplopia.

13. All cases of unexplained double vision (in which you are unable to make a definite diagnosis) require neuro-ophthalmic referral, urgently if of acute onset. There could be a serious underlying cause such as aneurysm, tumor or myasthenia (Fig. 1.13).



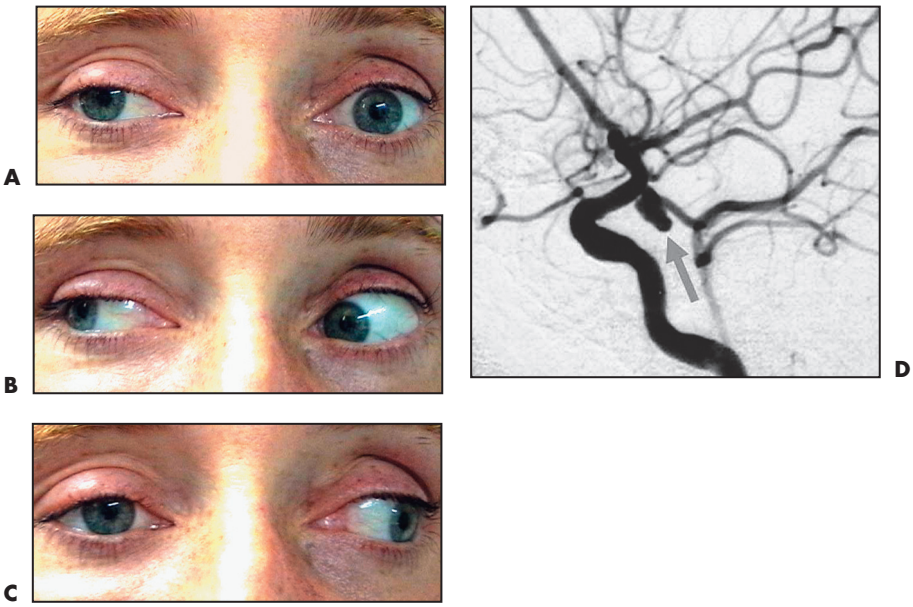
A



B

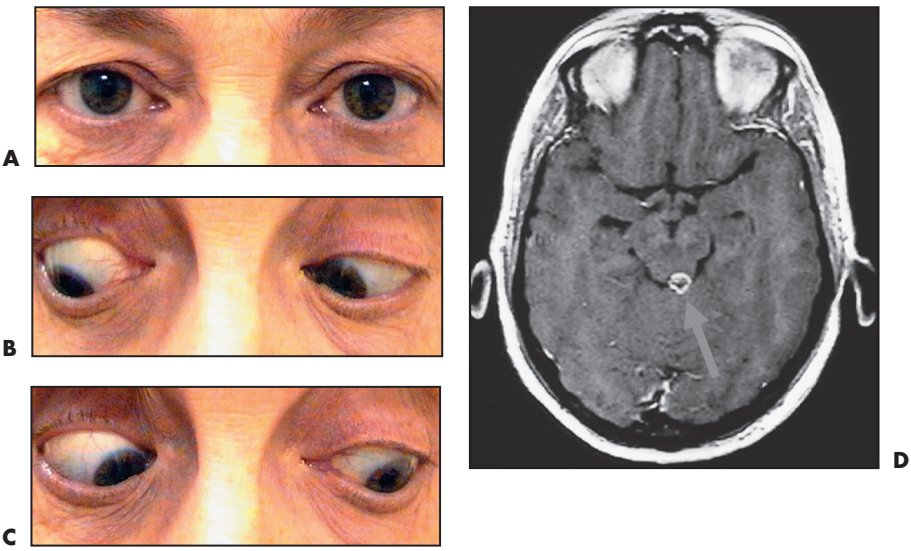
1.13 This 42-year-old woman initially presented to her local ophthalmologist complaining of gradual-onset horizontal double vision. At first examination, she had a small esotropia in primary position; however, eye movements were noted to be "full" (no restriction was seen in any direction). The ophthalmologist diagnosed "decompensated esophoria" and prescribed prism to be ground into the patient's spectacles. Although this relieved the diplopia, the amount of prism required gradually increased over the next 18 months until the spectacles were too heavy for the patient to wear comfortably; at this stage, it was noted that abduction was visibly limited in both eyes. MRI revealed a large clival tumor that on biopsy was found to be a chordoma **A**. The tumor had caused gradually progressive bilateral sixth nerve palsies that initially presented as a small comitant esotropia with no visible limitation of abduction. Further tumor spread eventually led to a vertical deviation being superimposed on the esotropia **B**, due to partial third nerve palsy.

14. All patients with partial or complete third nerve palsy require urgent MRI and magnetic resonance angiography (MRA) to exclude an aneurysm, except patients who meet all the clinical diagnostic criteria for ischemic third nerve palsy (p. 177) (Fig. 1.14).



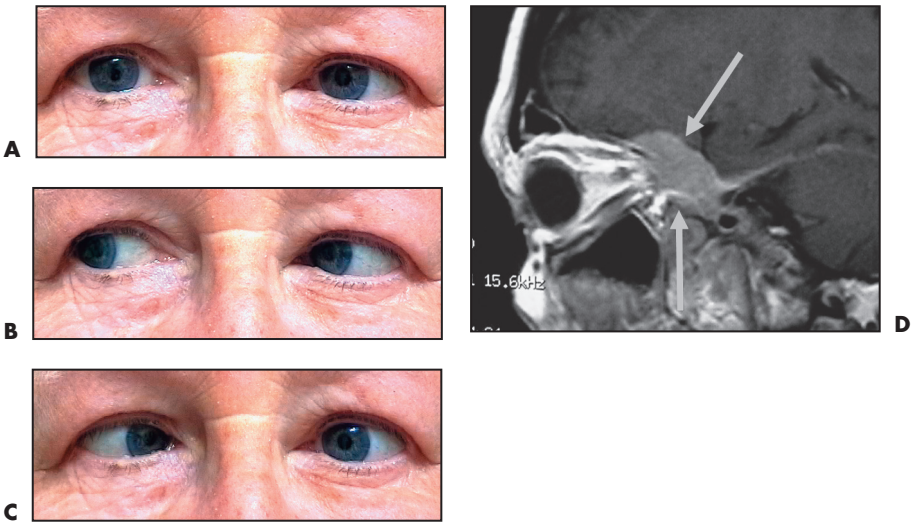
1.14 This 35-year-old diabetic woman presented with acute-onset horizontal diplopia. Her ophthalmologist noted a right exotropia, limitation of adduction of the right eye with normal elevation and depression, and a mild right ptosis **A**–**C**. The pupils were equal in size and both were briskly reactive to light. The ophthalmologist diagnosed a partial right third nerve palsy. He thought that this was "ischemic" in origin because the right pupil was "spared" and the patient was a diabetic. "To rule out any other cause", a CT scan of the brain was obtained that was normal; the patient was reassured the diplopia would resolve spontaneously and a return appointment was made for 3 months' time. The following day, the patient collapsed at home; an urgent MRI and MRA showed a right posterior communicating artery aneurysm that had ruptured, causing subarachnoid hemorrhage; this was confirmed on angiography **D**. The patient underwent urgent neurosurgical intervention and survived but was left with a dense left hemiplegia. Earlier diagnosis of the aneurysm before it ruptured could have prevented her serious permanent disability. For how to safely diagnose ischemic third nerve palsy, see p. 177.

15. Patients with fourth nerve palsy require an MRI to exclude a brain tumor or vascular malformation, except patients who meet all the clinical diagnostic criteria for either:
    - congenital fourth nerve palsy (p. 180), or
    - ischemic fourth nerve palsy (p. 178) (Fig. 1.15)



1.15 This 56-year-old hypertensive woman presented with a 2-week history of vertical and torsional diplopia. She was initially diagnosed with a "right ischemic fourth nerve palsy" and reassured that the diplopia would resolve spontaneously. However, closer examination revealed 12 degrees of excyclotorsion on double Maddox rod testing, a right hypertropia in left gaze that changed to a left hypertropia in right gaze, and mild bilateral superior oblique underaction (decreased depression of each eye in adduction) **A**–**C**. A diagnosis of bilateral fourth nerve palsy was made. MRI with contrast revealed a small enhancing lesion in the collicular area **D**, biopsy of which diagnosed a glioma. For how to safely diagnose ischemic or congenital fourth nerve palsy, see pp. 178 and 180.

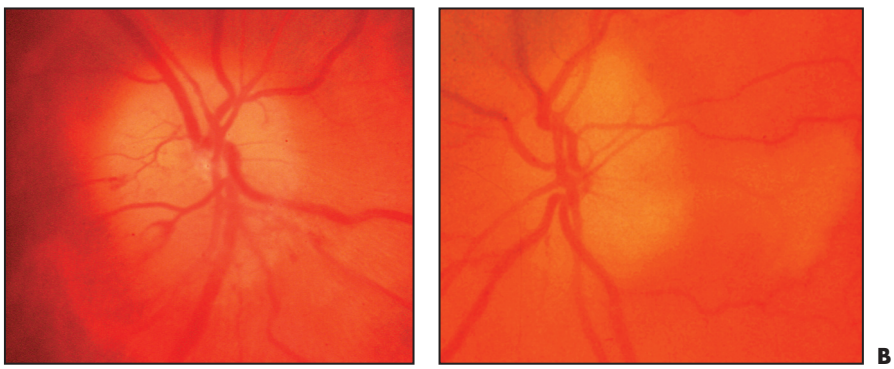
16. All patients with sixth nerve palsy require an MRI to exclude a brain tumor, except patients who meet all the clinical diagnostic criteria for ischemic sixth nerve palsy (p. 181) (Fig. 1.16).



1.16 This 62-year-old hypertensive diabetic woman presented with horizontal diplopia that had gradually worsened over the previous 2 weeks. Her ophthalmologist found a left esotropia with reduced abduction of the left eye and diagnosed an "ischemic" left sixth nerve palsy. However, over the next 6 months, the diplopia did not resolve and in fact worsened; the patient was eventually examined by a neuro-ophthalmologist who found reduced left corneal and supraorbital sensation in addition to a marked esotropia with left abduction weakness consistent with a left sixth nerve palsy **A**–**C**. MRI revealed a large left sphenoid wing meningioma that had invaded the cavernous sinus **D**. The patient was angry that she had been initially observed instead of investigated. For how to safely diagnose ischemic sixth nerve palsy, see p. 181.

Giant cell arteritis (GCA)

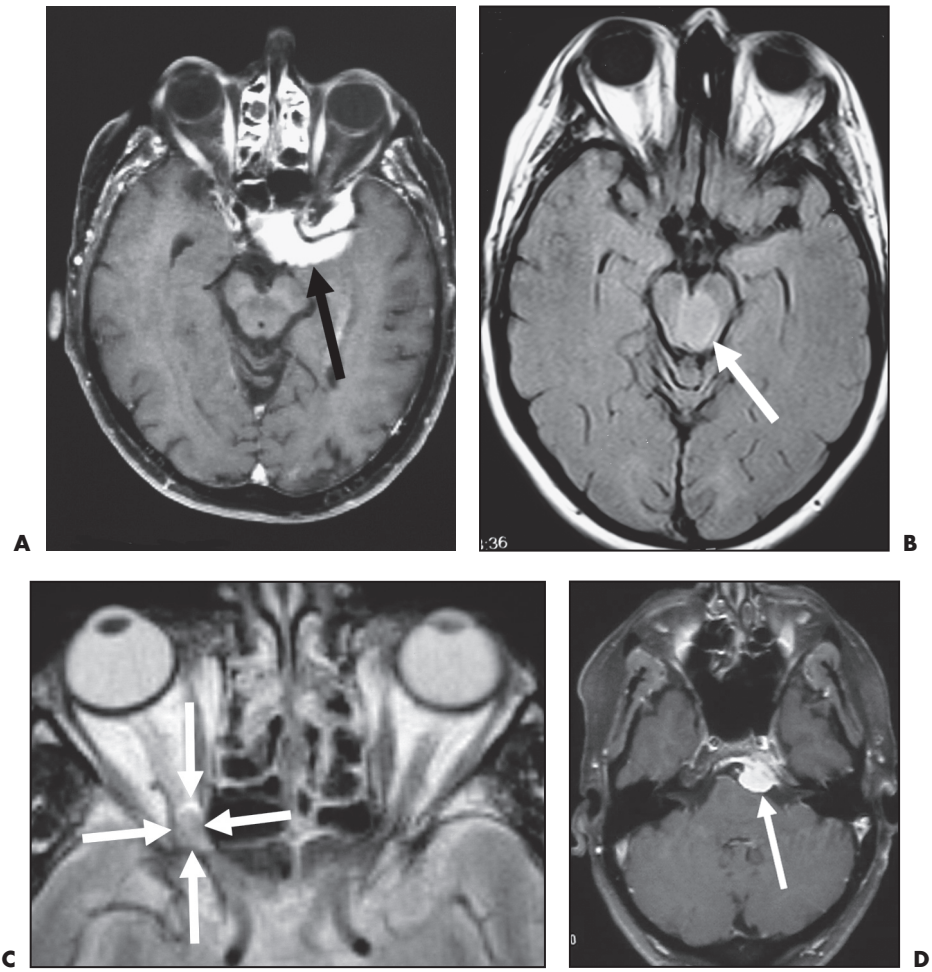
17. Don't miss GCA (Fig. 1.17): think of it in every patient over 50 with:
    - transient visual loss (including "amaurosis fugax")
    - sudden, persistent visual loss
      - with optic disc swelling (e.g. AION)
      - with evidence of a central retinal artery occlusion
      - with a normal fundus but other evidence of an optic neuropathy (retrobulbar ischemic optic neuropathy)
    - transient double vision (even if your examination is normal)
    - persisting double vision (including third, fourth or sixth nerve palsies)



1.17 A 68-year-old woman was referred to her local ophthalmologist with vague visual complaints of 2 weeks' duration. She complained of "seeing double" for minutes at a time and of loss of vision in her right eye lasting minutes and then resolving. The ophthalmologist found ocular motility and intraocular examination to be entirely normal and reassured the patient that there was "nothing wrong with the eyes". Three days later, the patient suddenly went blind in the right eye, followed an hour later by loss of vision in the left eye, due to AION **A**, **B**. Specific questioning revealed a 3-month history of a new type of headache, fatigue, myalgias and scalp pain when she tried to brush her hair. Despite urgent treatment with intravenous and then oral steroids, the final visual acuity was right no perception of light, left light perception. Temporal artery biopsy confirmed GCA.

Neuro-imaging

18. MRI is far superior to computed tomography (CT) in detecting most of the causes of neuro-ophthalmic disease and is the investigation of choice. A normal brain CT does NOT exclude serious disease (Fig. 1.18).
    - you need to know what sort of MRI to order, and in some cases you need to push to have it done urgently
    - MRI should always be requested "with contrast" as many orbital and brain lesions are difficult to detect unless contrast injection is given



1.18 These tumors were all missed on brain CT scan but detected with contrast-enhanced MRI. **A** Sphenoid wing meningioma. **B** Pontine glioma. **C** Orbital apex meningioma. **D** Clival meningioma.

Ophthalmic emergencies

19. The prime ophthalmic emergencies, in order of importance and urgency, are:

Life-threatening:

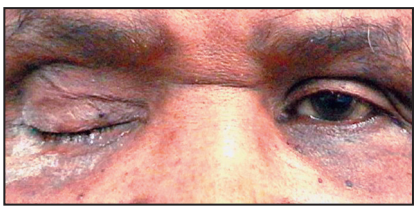
- double vision due to possible partial or complete third nerve palsy in which an aneurysm cannot be clinically excluded
- bilateral optic disc swelling due to brain tumor or dural venous sinus thrombosis
- acute unilateral or bilateral ophthalmoplegia from acute myasthenia, pituitary apoplexy (Fig. 1.19), cavernous sinus thrombosis, carotid-cavernous fistula or orbital cellulitis
- ptosis or diplopia with dyspnea, dysphagia or severe systemic weakness due to severe myasthenia
- ptosis and ipsilateral small but reactive pupil from Horner syndrome related to an acute internal carotid artery dissection

Bilateral sight-threatening:

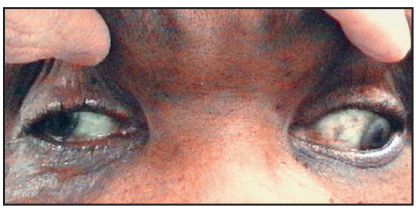
- acute loss of vision from GCA, pituitary apoplexy or neuromyelitis optica

Unilateral sight-threatening:

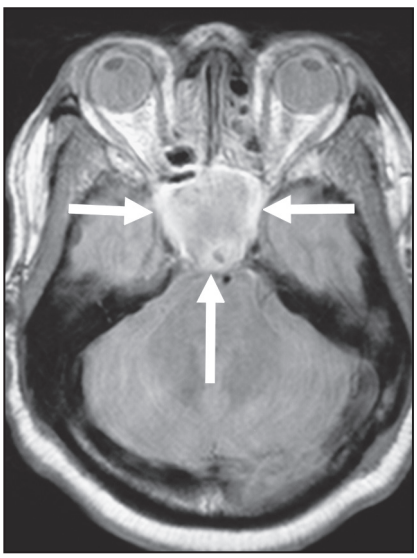
- acute glaucoma, endophthalmitis, penetrating eye injury, sphenoid sinus mucocele



A



B



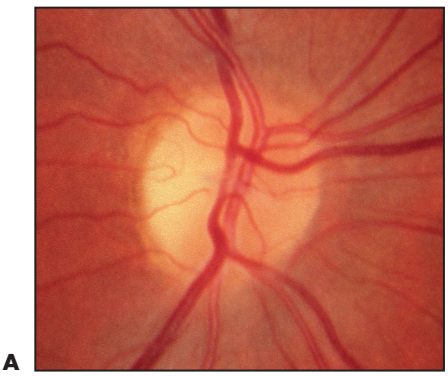
C

1.19 This 42-year-old man presented to the ophthalmic emergency department complaining of the sudden onset of blurred vision, diplopia, right ptosis and headache of 3 hours' duration **A**, **B**. The ophthalmology resident was informed but was too busy with other patients to see him immediately. Six hours later, the patient collapsed in the waiting room and was admitted via the general emergency department. Urgent MRI revealed pituitary apoplexy (sudden infarction of a previously undiagnosed pituitary tumor) **C**. Urgent medical therapy and neurosurgical treatment saved the patient's life. In retrospect, this patient's case was much more urgent than the retinal detachments and penetrating eye injury with which the resident had been occupied.

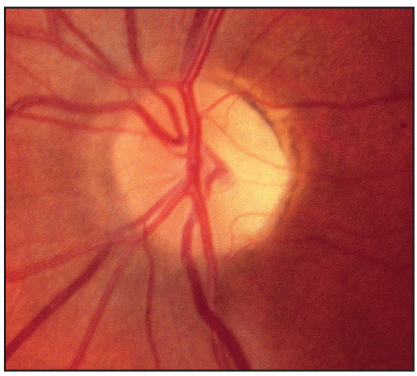
Three common mistakes

20. Three common mistakes made by ophthalmologists that lead to permanent blindness, disability or death of patients with neuro-ophthalmic disease are:
    - not suspecting the possibility of serious orbital or brain disease as the cause of a patient's "eye" complaints
    - not performing a thorough history and examination
    - not referring to a neuro-ophthalmologist early and urgently when necessary

These three mistakes cause much more harm than not knowing all the details of rare neuro-ophthalmic diseases (Fig. 1.20).



A



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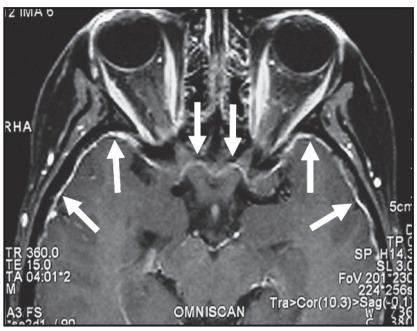

C

1.20 This 33-year-old woman was referred to her local ophthalmologist complaining of blurred vision in both eyes for the past 2 months. Examination revealed visual acuity of 20/30 in each eye, reduced color vision in both eyes and temporal pallor of both optic discs **A**, **B**. The ophthalmologist requested an MRI (but did not specifically ask for contrast enhancement) and ordered the following blood tests: vitamin B12, folate, erythrocyte sedimentation rate (ESR), c-reactive protein (CRP) and complete blood count. The MRI was said to be normal, as were the blood tests, except for the B12 level that was slightly low; the patient was told she had "nutritional deficiency optic neuropathy" and was given multivitamins and B12 injections. Despite this treatment, her vision worsened to 20/200 in both eyes over the next 12 months. At this stage the patient was advised that she might have "optic neuritis" and was given a 3-day course of intravenous methylprednisolone without effect. A blood test was then performed for Leber hereditary optic neuropathy (LHON), which was negative. Eighteen months after first seeing the ophthalmologist, the patient's visual acuity had decreased to "light perception" in each eye and she was finally referred to a neuro-ophthalmologist. **C** A contrast-enhanced MRI revealed diffuse meningeal enhancement (arrows) suggestive of a chronic granulomatous meningitis; and serum angiotensin converting enzyme (ACE), CT of the mediastinum and lumbar puncture resulted in a diagnosis of chronic sarcoid meningitis and optic neuritis. Because of the severe optic atrophy, vision did not improve despite immunosuppressive treatment. If, at the start, the patient had been promptly referred to a neuro-ophthalmologist or investigated thoroughly, rather than investigated inadequately and "piecemeal" over a long period, it is likely that she would have retained good vision in both eyes.