Figure 10.24 Paget's disease, showing bowing of the tibia

Examine for evidence of paraplegia, which is uncommon but can occur due to cord compression by bone or vascular shunting in the spinal cord. Cerebellar signs may rarely be present due to platybasia.

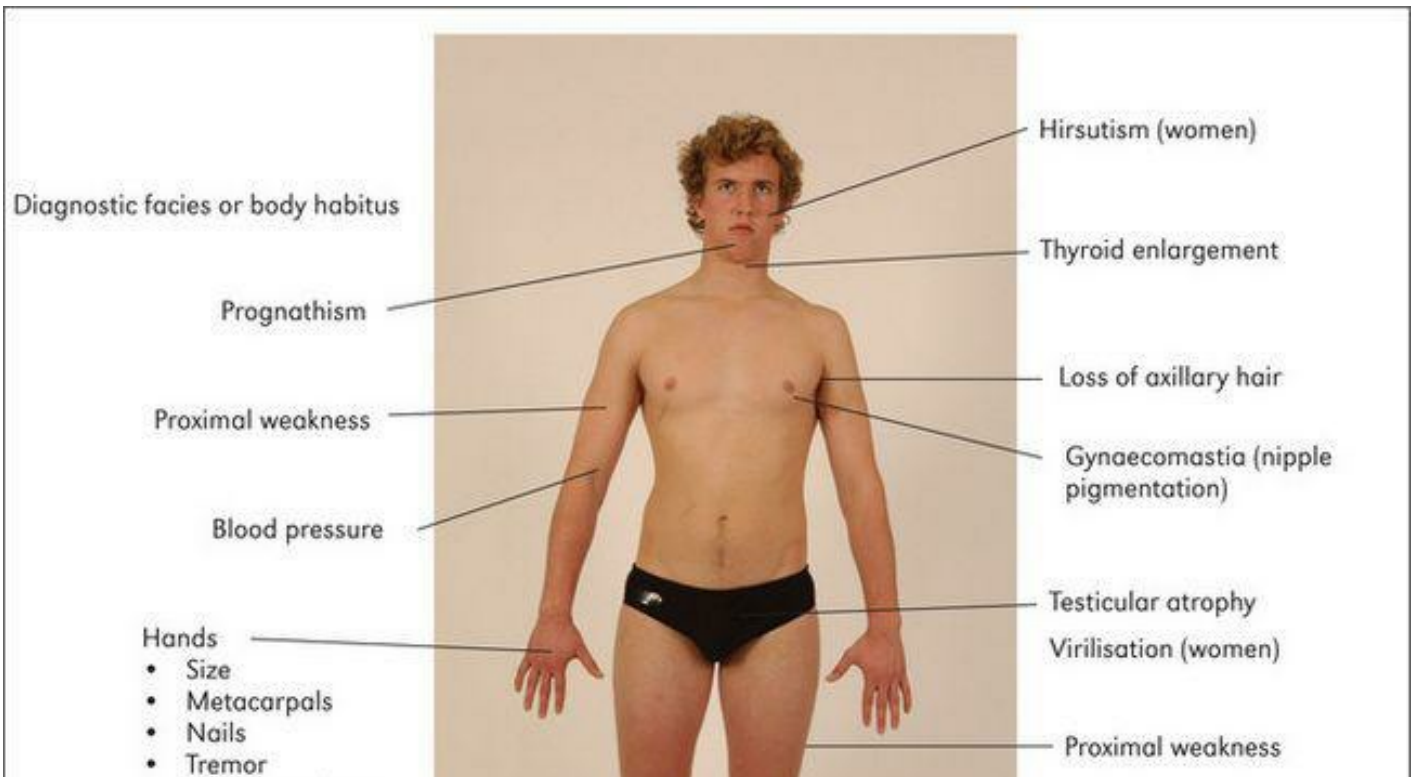
Urinalysis

Check for blood (there is an increased incidence of renal stones in Paget's disease).

Summary

The endocrine system: a suggested method of examination (Figure 10.25)

Inspect the patient for one of the diagnostic facies or body habituses. If the diagnosis is obvious, proceed with the specific examination outlined previously. If not, examine as follows.



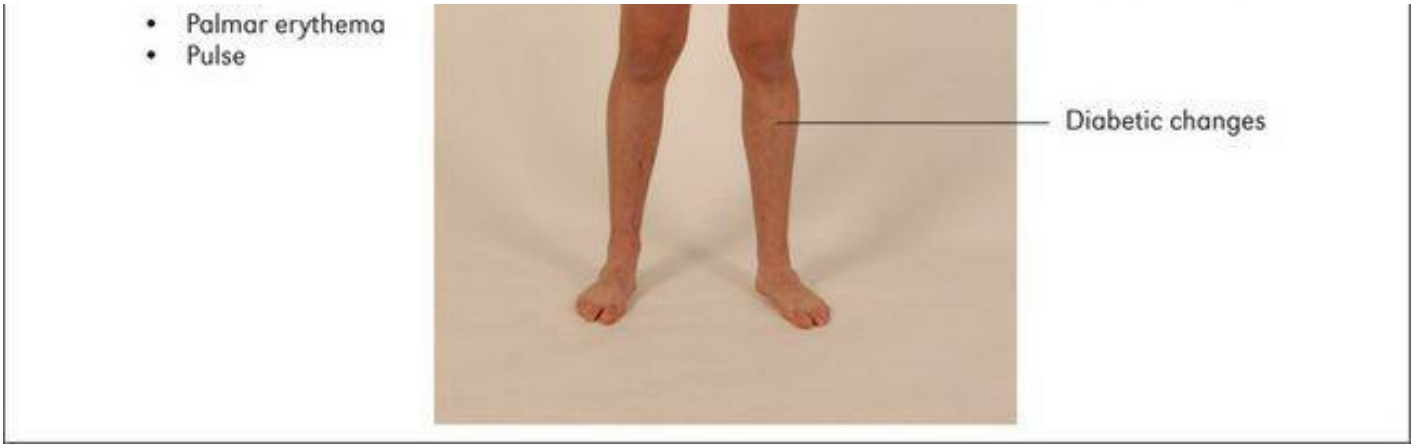


Figure 10.25 The endocrine examination

Pick up the **hands**. Look at the overall size (acromegaly), length of the metacarpals (pseudohypoparathyroidism and pseudopseudohypo-parathyroidism), for abnormalities of the nails (hyperthyroidism and hypothyroidism, and hypoparathyroidism), tremor, palmar erythema and sweating of the palms (hyperthyroidism).

Take the pulse (thyroid disease) and the blood pressure (hypertension in Cushing's syndrome, or postural hypotension in Addison's disease). Look for Trousseau's sign (tetany). Test for proximal muscle weakness (thyroid disease, Cushing's syndrome).

Go to the **axillae**. Look for loss of axillary hair (hypopituitarism), or acanthosis nigricans and skin tags (acromegaly).

Examine the **eyes** (hyperthyroidism) and the **fundi** (diabetes, acromegaly). Look at the **face** for hirsutism, or fine-wrinkled hairless skin (panhypopituitarism). Note any skin greasiness, acne or plethora (Cushing's syndrome).

Look at the **mouth** for protrusion of the chin and enlargement of the tongue (acromegaly) or buccal pigmentation (Addison's disease).

Examine the **neck** for thyroid enlargement. Note any neck webbing (Turner's syndrome). Palpate for supraclavicular fat pads (Cushing's syndrome).

Inspect the **chest** wall for hirsutism or loss of body hair, reduction in breast size in women (panhypopituitarism) or gynaecomastia in men. Look for nipple pigmentation (Addison's disease).

Examine the **abdomen** for hirsutism, central fat deposition, purple striae (Cushing's syndrome) and the **external genitalia** for virilisation or atrophy. Look at the **legs** for diabetic changes.

Measure the body **weight** and **height**, and examine the **urine**.

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Suggested reading

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[a] Hakaru Hashimoto (1881–1934), Japanese surgeon.

[b] Harry Fitch Klinefelter (b. 1912), Baltimore physician, described the condition when he was a medical student.

[c] The first person to distinguish an enlarged thyroid from cervical lymphadenopathy was the Roman medical writer Aulus Aurelius Cornelius Celsus (53 BC–7 AD). He is more famous for describing the four cardinal signs of inflammation: redness, swelling, heat and tenderness.

[d] Robert Graves (1796–1853), Dublin physician.

[e] Henry Plummer (1874–1936), physician at the Mayo Clinic, USA.

[f] John Dalrymple (1803–52), British ophthalmic surgeon.

[g] Friedrich von Graefe (1828–70), professor of ophthalmology in Berlin, described this in 1864. He was one of the most famous ophthalmologists of the 19th century; Horner was one of his pupils. He died of tuberculosis at the age of 42.

[h] Fritz de Quervain (1868–1940), professor of surgery, Berne, Switzerland.

[i] Franz Chvostek (1835–84), Viennese physician.

[j] Armand Trousseau (1801–1867), Parisian physician.

[k] The acral parts are the hands and feet.

[l] Acromegaly was first described by Pierre Marie in 1886 and was first called hyperpituitarism by Harvey Cushing in 1909.

[m] From the Latin—*habitus* = the state or condition of a thing.

[n] The enthusiastic student can calculate the central obesity index. This is the sum of three truncal circumferences (neck, chest and waist) divided by six peripheral ones (arms, thighs and legs on both sides). A normal index is less than 1.

[o] Warren Nelson (1906–64), American endocrinologist.

[p] Thomas Addison (1793–1860) described the disease in 1849. Addison, Bright and Hodgkin made up the famous trio of physicians at Guy's Hospital, London.

[q] This disease was called diabetes by ancient Greek and Roman physicians because the word diabetes means a siphon, referring to the large urine volume. Rather courageously, they distinguished diabetes mellitus from diabetes insipidus by the sweet taste of the urine: *mellitus*, 'sweet'; *insipidus*, 'tasteless'.

[r] Douglas Argyll Robertson (1837–1909), a Scottish ophthalmic surgeon and President of the Royal College of Surgeons, described these in 1869. The pupils are small, irregular and unequal, and react briskly to accommodation but not to light. Tertiary syphilis is another cause.

[s] Sir James Paget (1814–99), a surgeon at St Bartholomew's Hospital

Sir James Paget (1814–99), a surgeon at St Bartholomew's Hospital, London, was also Queen Victoria's doctor.

Chapter 11

The nervous system

> Who could have foretold, from the structure of the brain, that wine could derange its functions?
> *Hippocrates (460–375 BC)*

The neurological history

The neurological history begins in detail with the presenting problem or problems (Table 11.1). The patient should be allowed to describe the symptoms in his or her own words to begin with, and then the clinician needs to ask questions to clarify information and obtain more detail. It is particularly important to ascertain the *temporal course of the illness*, as this may give important information about the underlying aetiology.

TABLE 11.1 Neurological history

Presenting symptoms*
Headache, facial pain
Neck or back pain
Fits, faints or funny turns
Dizziness or vertigo
Disturbances of vision, hearing or smell
Disturbances of gait

Loss of or disturbed sensation, or weakness in limb(s)
Disturbances of sphincter control (bladder, bowels)
Involuntary movements or tremor
Speech and swallowing disturbance
Altered cognition
Risk factors for cerebrovascular disease
Hypertension
Smoking
Diabetes mellitus
Hyperlipidaemia
Atrial fibrillation, bacterial endocarditis, myocardial infarction (emboli)
Haematological disease
Family history of stroke

* Note particularly the temporal course of the illness, whether symptoms suggest focal or diffuse disease, and the likely level of involvement of the nervous system.

An acute onset of symptoms (within minutes to an hour) is suggestive of a vascular or convulsive problem (e.g. the explosive severe headache of subarachnoid haemorrhage or the rapid onset of a seizure).

For these episodes of sudden onset, a precipitating event (e.g. exercise) or warning (*aura*) may be present. The aura that precedes a seizure may be localising (e.g. auditory hallucinations, an unusual smell or taste, loss of speech, or motor changes) or non-localising (e.g. a feeling of apprehension). The occurrence of an aura followed by sudden unconsciousness is very suggestive of the diagnosis of a major seizure or complex partial seizure.

A *stroke* or *cerebrovascular accident* usually causes symptoms which appear over minutes or are present when the patient wakes from sleep. There is a focal problem with function of the brain. Patients may be unable to move

is a focal problem with function of the brain. Patients may be unable to move one side of the body (hemiplegia) or have difficulty with speech or swallowing, There may have been previous episodes. When there is resolution of the symptoms within 24 hours the episode is called a *transient ischaemic attack*—TIA). The rapid onset of focal symptoms almost always has a vascular cause—embolism, infarction or haemorrhage. If the patient can answer questions it is important to ask about the onset of the symptoms and about risk factors for stroke (*Questions box 11.1*).

Questions box 11.1

Questions to ask the (non-aphasic) patient with a possible stroke or transient ischaemic attack

1. What have you noticed has been wrong?
2. How quickly did it come on? How long ago?
3. Has it improved or gone away now?
4. Have you ever had a stroke before? How did that affect you?
5. Have you had high blood pressure or cholesterol (risk factors)?
6. Are you a diabetic (risk factor)?
7. Do you smoke (risk factor)?
8. Is there a history of strokes in the family?
9. Have you had palpitations or been told you have atrial fibrillation?
10. Have you been treated with blood-thinning drugs such as aspirin or warfarin?

The sudden onset of weakness on one side of the body followed by resolution and a severe headache is characteristic of hemiplegic migraine. Sudden resolution without headache suggests a transient ischaemic episode. The very gradual onset of muscle weakness suggests a muscle abnormality such as myopathy rather than a vascular event.

A subacute onset (hours to days) occurs with inflammatory disorders (e.g. meningitis, cerebral abscess or the Guillain-Barré[a] syndrome—acute inflammatory polyradiculoneuropathy).

A more chronic symptom course suggests that the underlying disorder may be related to either a tumour (weeks to months) or a degenerative process (months to years). Metabolic or toxic disorders may present with any of these time courses.

Based on the history (and physical examination), a judgment is made as to whether the disease process is *localised or diffuse*, and which *levels of the nervous system* are involved (the nervous system may be thought of as having four different levels: the peripheral nervous system, the spinal cord, the posterior fossa, and the cerebral hemispheres). Consideration of the time course and the levels of involvement will usually lead to a logical differential diagnosis of the patient's symptoms. After detailed questions about the presenting problem, ask about previous neurological symptoms and about previous neurological diagnoses or investigations. The patient may know the results of CT or magnetic resonance imaging brain scans performed in the past. A thorough neurological history will include routine questions about possible neurological symptoms (*Questions box 11.2*). If the patient answers 'yes' to any of these, more-detailed questions about the nature of the problem and its time course are indicated.

Questions box 11.2

Questions to ask the patient with a possible neurological problem

1. Can you tell me what has been happening to you?
2. Are you right- or left-handed?
3. Have you had problems with headaches?
4. Have you been dizzy or had problems with your balance?
5. Have you noticed trouble with your speech?
6. Have you had problems with your vision?
7. Have you had weakness in an arm or leg?
8. Have you ever had a seizure or a blackout?
9. Have you ever had a head injury?

10. Have you had any back problems?

11. Have you had any scans of your brain or spinal cord?

12. What medications have you been taking?

13. Have you had high blood pressure?

14. Is there a history of neurological or muscle problems in the family?

15. Do you drink alcohol?

Headache and facial pain

Headache is a very common symptom (*Questions box 11.3*). It is important, as with any type of pain, to determine the character, severity, site, duration, frequency, radiation, aggravating and relieving factors and associated symptoms.[1,2] Unilateral headache that is preceded by flashing lights or zigzag lines and is associated with light hurting the eyes (photophobia) is likely to be a *migraine with an aura* ('classical migraine'); common migraine has no aura. Pain over one eye (or over the temple) lasting for minutes to hours, associated with lacrimation, rhinorrhoea and flushing of the forehead, and occurring in bouts that last several weeks a few times a year or less, is suggestive of *cluster headache.* This occurs predominantly in males and patients can't stay still. Headache over the occiput and associated with neck stiffness may be from *cervical spondylosis*. *Coital headache* occurs during intercourse close to orgasm.

Questions box 11.3

Questions to ask the patient with headache

1. What is it like, e.g. dull, sharp, throbbing or tight?

2. Where do you feel it—at the front or back, on one side or in the face?

3. How severe is it and how long does it last?

4. Has it begun very suddenly and severely?—Subarachnoid haemorrhage

5. Do you get any warning that it is about to start, e.g. flashing lights or zigzag lines in your vision?—Migraine

6. Is it associated with sensitivity to light (photophobia)?—Migraine

7. Do you feel drowsy or nauseated?—Raised intracranial pressure

8. Is the pain on one side over the temple and have you had any blurred vision?—Temporal arteritis

9. Is the pain worst over your cheek bones?—Sinusitis

10. Are the attacks likely to occur in clusters and associated with watering of one eye?—Cluster headache

11. Is there a prolonged feeling of tightness over the head but no other symptoms?—Tension headache

12. Did you drink large amounts of alcohol last night?—Hangover

A generalised headache that is worse in the morning and is associated with drowsiness or vomiting may reflect *raised intracranial pressure*, while generalised headache associated with photophobia and fever as well as with a stiff neck of more gradual onset may be due to *meningitis*. A persistent unilateral headache over the temporal area associated with tenderness over the temporal artery and blurring of vision suggests *temporal arteritis*.[3,4] This condition (Table 11.2) is often associated with jaw claudication, or jaw pain during eating, which can lead to considerable loss of weight. Headache with pain or fullness behind the eyes or over the cheeks or forehead occurs in *acute sinusitis*. The dramatic and usually instantaneous onset of severe headache that is initially localised but becomes generalised and is associated with neck stiffness may be due to a *subarachnoid haemorrhage*. Morning headaches worse with coughing, especially in an obese patient, may be due to *idiopathic intracranial hypertension*; visual loss may occur.

TABLE 11.2 Symptoms and signs of temporal arteritis

Symptoms	Frequency (%)	Positive LR	Negative LR
Headache often present for 2 or 3 months	77	1.5	0.82
Jaw claudication (pain in proximal jaw near TMJ after brief chewing of tough food)	51	4.2	0.72
Malaise	48	1.2	0.94
Polymyalgia rheumatica (shoulder girdle pain and stiffness)	34	0.97	0.99

Visual disturbance (often sudden monocular blindness)	29	0.85	1.2
Examination findings			
Fever	26	–	–
Tender temporal artery (palpated just anterior and superior to the tragus of the ear).	53	2.6	0.82
Enlarged temporal artery	–	4.3	0.67
There can be ischaemic changes of the tongue or scalp.			

TMJ = temporomandibular joint.
Likelihood ratios from McGee S, *Evidence-based physical diagnosis*, 2nd edn. St Louis: Saunders, 2007.

Finally, the most frequent type of headache is episodic or chronic *tension-type headache*; this is commonly bilateral, occurs over the frontal, occipital or temporal areas, and may be described as a sensation of tightness that lasts for hours and recurs often. There are usually no associated symptoms such as nausea, vomiting, weakness or paraesthesiae (tingling in the limbs), and the headache does not usually wake the patient at night from sleep.

Pain in the face can result from trigeminal neuralgia, temporomandibular arthritis, glaucoma, cluster headache, temporal arteritis, psychiatric disease, aneurysm of the internal carotid or posterior communicating artery, or the superior orbital fissure syndrome.

Faints and fits (see also page 41)

It is important to try to differentiate syncope (transient loss of consciousness) from *epilepsy* (*Questions box 11.4*). However, primary syncopal events can cause a few clonic jerks in a significant number of cases. Generalised tonic-clonic seizures (grand mal epilepsy) cause abrupt loss of consciousness, which may be preceded by an aura. Often the patient is incontinent of urine and faeces, and the tongue may be bitten. A witness may be able to describe the type of attack that occurred. It is important to try to determine whether any seizure is generalised or localised to one side of the body: a seizure affecting part of the body may indicate a focal lesion in the central nervous system, such as a tumour or abscess. If consciousness is impaired, these partial seizures are described as ‘complex’; if consciousness is unimpaired they are termed ‘simple’. Idiopathic absence seizures (‘petit mal’) occur in children. These are frequent brief episodes of loss of awareness often associated with staring. Major motor movements do not occur with this type of epilepsy.

Questions box 11.4

Questions to ask the patient with syncope or dizziness

1. Have you lost consciousness completely? How long for?

2. Do you black out or feel dizzy when you stand up quickly?—Postural hypotension

3. How often have episodes occurred?

4. Was the sensation more one of spinning?—Vertigo

5. Did the episode occur during heavy exercise or when you got up to pass urine at night? Exercise—suggests a left ventricular outflow tract obstruction such as aortic stenosis. Pass urine at night—micturition syncope

6. Have you injured yourself?

7. Do you get any warning?—A feeling of nausea and being in a stuffy room suggests a vasovagal episode; a strange smell or feeling of deja-vu suggests an aura and therefore a seizure

8. Have you passed urine during the episode?—Seizure

9. Have you bitten your tongue?—Seizure

10. Has anyone seen an episode and noticed jerking movements (tonic-clonic movements)?—Makes a seizure more likely but can also occur with cardiac syncope

11. Do you wake up feeling normal or drowsy? Normal—cardiac syncope. Drowsy—seizure

12. What medications are you taking—any antihypertensive medications, cardiac anti-arrhythmic drugs or anti-epileptic drugs?

Transient ischaemic attacks (TIAs) affecting the brainstem can occasionally cause blackouts. Use of the term 'drop attacks' means the patient falls but there is no loss of consciousness. In either case the patient falls to the ground without premonition and the attacks are of brief duration. *Hypoglycaemia* can lead to episodes of loss of consciousness. Patients with hypoglycaemia may also report sweating, weakness and confusion before losing consciousness. Bizarre attacks of loss of consciousness occur with *hysteria*.[b] During such attacks the patient may slump to the ground without sustaining any injury and there may be apparent fluctuations in the level of

consciousness for a prolonged period.

Dizziness

If a patient complains of dizziness, it is important to determine what is meant by this term. In true *vertigo*, there is actually a sense of motion, usually of the surroundings but also of the head itself (page 41).[5] When vertigo is severe it may not be possible for the patient to stand or walk, and associated symptoms of nausea, vomiting, pallor, sweating and headache may be present. Causes of vertigo include the 'peripheral vestibular lesions':

• benign positioning (positional) vertigo—recurrent brief episodes of vertigo precipitated by a change of head position, due to crystals in the saccule and utricle

• vestibular neuronitis—non-positional vertigo due to inflammation of the acoustic nerve with normal hearing, and

• acute labyrinthitis—associated with hearing loss.

Other causes of vertigo include:

• ototoxic drugs (e.g. aminoglycosides), associated with deafness or tinnitus;

• Ménière's disease,[c] which occurs in those over 50 years of age and presents with the triad of episodic vertigo and tinnitus (ringing in the ears) with progressive deafness;

• acoustic neuroma (where patients may also have deafness and tinnitus);

• central causes such as vertebrobasilar TIAs—these may be associated with diplopia (double vision; page 427), visual loss and ataxia; and

• rarely, internal auditory artery occlusion.

Visual disturbances and deafness

Problems with vision can include double vision (diplopia), blurred vision (amblyopia), light intolerance (photophobia) and visual loss. The causes of deafness are summarised on page 348.

Disturbances of gait

Disturbances of gait

Many neurological conditions can make walking difficult. These are described on page 376. Walking may also be abnormal when orthopaedic disease affects the lower limbs or spine. A bizarrely abnormal gait can sometimes be a sign of a hysterical reaction.

Disturbed sensation or weakness in the limbs

Pins and needles in the hands or feet may indicate nerve entrapment or a peripheral neuropathy (page 386) but can result from sensory pathway involvement at any level. The carpal tunnel syndrome is common; here there is median nerve entrapment, and patients experience pain and paraesthesiae in the hand and wrist. Sometimes pain may extend to the arm and even to the shoulder, but paraesthesiae are felt only in the fingers. These symptoms are usually worse at night and may be relieved by dangling the arm over the side of the bed or shaking the hand.

Nerve root, spinal cord and cerebral abnormalities can all cause disturbance of sensation and weakness.

Limb weakness can be caused by lesions at different levels in the motor system. There are a number of patterns of limb and muscle weakness:

- Upper motor neurone (UMN) weakness (page 383) is due to interruption of a neural pathway at a level above the anterior horn cell. The result is an increase in tone and peripheral reflexes. Interruption of this pathway has the greatest effect on the antigravity muscles and is called *pyramidal weakness*. There is little or no muscle wasting.

- Lower motor neurone (LMN) weakness (page 385) is due to a lesion that interrupts the reflex arc between the anterior horn cell and the muscle. There is a reduction in tone and reflexes, fasciculation (irregular contractions of small areas of muscle) may be seen and muscle wasting is prominent.

- Muscle disease causes weakness in a particular muscle or group of muscles. There is wasting, decreased tone, and the reflexes are reduced or absent.

- Disease at the neuromuscular junction (e.g. myasthenia gravis, page 394) causes generalised weakness, which worsens with repetition. The reflexes and tone are often normal.

- Non-organic weakness (e.g. due to hysteria) causes a non-anatomical

pattern of weakness in association with normal tone and power and, unless there has been prolonged disuse, normal muscle bulk.

Tremor and involuntary movements

Tremor is a rhythmical movement (Table 11.3). A slow tremor has, by definition, a rate between 3 Hz and 5 Hz. Rapid tremors are faster than 10 Hz. *Resting* tremors are present mostly during relaxation of the muscles, while *intention* tremors occur with deliberate movement and become more pronounced towards the end of the action. Tremors become worse with fatigue or anxiety. Shivering is a type of tremor brought on by cold. It is normal for there to be a fine tremor associated with holding a posture or performing a movement slowly. This is called a *physiological* tremor. It becomes more obvious with fright and fatigue. It is often increased by the beta-agonist drugs used to treat asthma or by caffeine. Thyrotoxicosis is a cause of exaggeration of physiological tremor. These movements are very fine and may be difficult to see unless looked for specifically. *Benign essential (familial)* tremor is an inherited disorder which causes tremor, but no other signs. The tremor is most easily seen when the patient's arms are stretched out; it can become worse during voluntary movements. It usually disappears when the muscles are at complete rest. Parkinson's disease[d] may present with a resting tremor (page 397). Intention (or target-seeking) tremor is due to cerebellar disease (page 398). *Chorea* involves involuntary jerky movements (page 399). Definitions of the terms used to describe movement disorders are shown in Table 11.4.

TABLE 11.3 Rates of tremors

Parkinson's disease	3 to 5 Hz
Essential/familial	4 to 7 Hz
Physiological	8 to 13 Hz

TABLE 11.4 Definitions of terms used to describe movement disorders

Akithesia	Motor restlessness; constant semi-purposeful movements of the arms and legs
Asterixis	Sudden loss of muscle tone during sustained contraction of an outstretched limb
Athetosis	Writhing, slow sinuous movements, especially of the hands and wrists
Chorea	Jerky small rapid movements, often disguised by the patient with a purposeful final movement: e.g. the jerky upward arm movement is transformed into a voluntary movement to scratch the head
Dyskinesia	Purposeless and continuous movements, often of the face and mouth; often a result of treatment with major tranquillisers for psychotic illness
Dystonia	Sustained contractions of groups of agonist and antagonist muscles, usually in flexion or extremes of extension; it results in bizarre postures
Hemiballism	An exaggerated form of chorea involving one side of the body; the proximal flexion movements

Hemiballismus	of the body: there are wild flinging movements which can injure the patient (or bystanders)
Myoclonic jerk	A brief muscle contraction which causes a sudden purposeless jerking of a limb
Myokymia	A repeated contraction of a small muscle group; often involves the orbicularis oculi muscles
Tic	A repetitive irresistible movement which is purposeful or semi-purposeful
Tremor	A rhythmical alternating movement

Speech and mental status

Speech may be disturbed by many different neurological diseases and is discussed on page 377. A number of different diseases can also result in delirium or dementia, as described on page 377 and in Chapter 12.

Past health

Inquire about a past history of meningitis or encephalitis, head or spinal injuries, a history of epilepsy or convulsions and any previous operations. Any past history of sexually transmitted disease (e.g. risk factors for HIV infection or syphilis) should be obtained. Ask about risk factors that may predispose to the development of cerebrovascular disease (Table 11.1). A previous diagnosis of peripheral vascular disease or of coronary artery disease indicates an increased risk of cerebrovascular disease. Chronic or paroxysmal atrial fibrillation is associated with a greatly increased risk of embolic stroke, especially for people over the age of 70.

Medication history

Previous and current medications may be the cause of certain neurological or apparently neurological syndromes (Table 11.5).

TABLE 11.5 Drugs and neurology

1 Anti-hypertensives

Therapeutic use: reduction of risk of stroke

Side-effects: postural dizziness, syncope, depression (methyldopa)

2 Anti-platelet drugs and anti-coagulants

Therapeutic use: reduction of risk of stroke

Side-effect: cerebral haemorrhage

3 Statins

Therapeutic use: reduction in stroke risk

Side-effect: myopathy

4 Major tranquillisers

Therapeutic use: treatment of psychoses

Side-effects: ataxia, sedation, Parkinsonian tremor

5 Other neurological symptoms associated with drugs

Headache: nitrates, sildenafil

Deafness: aminoglycoside antibiotics, aspirin, frusemide

Peripheral neuropathy: amiodarone, isoniazid, metronidazole

Non-Parkinsonian tremor: bronchodilators, amphetamines

Dysphagia: bisphosphonates

Confusion and loss of memory: minor tranquillisers

Seizures: lignocaine

Ask about treatment for neurological disorders with anticonvulsants, anti-Parkinsonian drugs, steroids, immunosuppressants, biological agents, anticoagulants, antiplatelet agents and for other drug treatment that may be associated with neurological problems; use of the contraceptive pill, antihypertensive agents or drugs for other disorders needs to be documented.

Social history

As smoking predisposes to cerebrovascular disease, the smoking history is relevant. It is useful to ask about occupation and exposure to toxins (e.g. heavy metals). Alcohol can also result in a number of neurological diseases (see Table 1.3, page 7).

Family history

Any history of neurological or mental disease should be documented. A number of important neurological conditions are inherited (Table 11.6).

TABLE 11.6 Inherited neurological conditions

X-linked	Colour blindness, Duchenne's and Becker's muscular dystrophy, Leber's* optic atrophy
Autosomal dominant	Huntington's chorea, tuberose sclerosis, dystrophia myotonica
Autosomal recessive	Wilson's disease, Refsum's† disease, Freiderich's ataxia, Tay-Sachs'‡ disease
Increased incidence in	Alzheimer's§ disease

families

* Theodor Karl von Leber (1840–1917). German ophthalmologist, professor of ophthalmology at Heidelberg. He began studying chemistry but was advised by Bunsen that there were too many chemists and so changed to studying medicine.

† Siguald Refsum (1907–91), Norwegian neurologist. He described this in 1945, calling it heredotaxia hemerlopica polyneuritiformis. This may be an argument for the use of eponymous names.

‡ Warren Tay (1843–1927), English ophthalmologist, described the ophthalmological abnormalities of the condition. Bernard Sachs (1858–1944), American neurologist and psychiatrist, described the neurological features in 1187. He studied in Germany and was a pupil of von Recklinghausen. The condition was originally called amaurotic family idiocy. This may be another reason to use eponymous names.

§ Alois Alzheimer (1864–1915), Bavarian neuropathologist, described the condition in 1906. His doctoral thesis was on the wax-producing glands of the ear.

The neurological examination

Examination anatomy

More than for any other system of the body, neurological diagnosis depends on localising the anatomical site of the lesion—in the brain, spinal cord or peripheral nerve. Figure 11.1 shows the gross anatomy of the brain and the major functional areas.

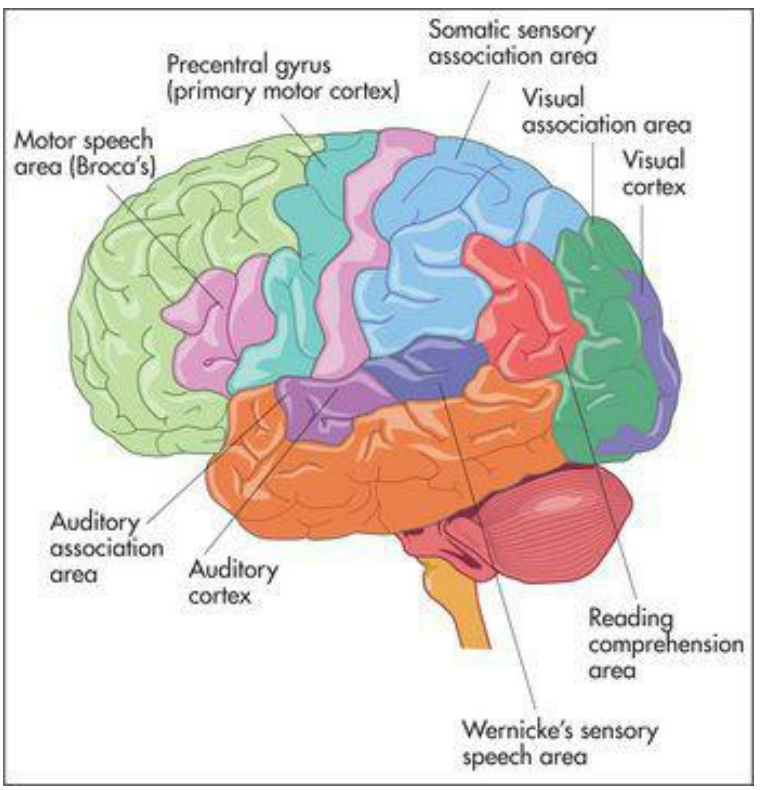


Figure 11.1 The functional areas of the brain

As a preliminary to the neurological assessment, the clinician should obtain some biographical information from the patient, including the age, place of birth, handedness, occupation and level of education.

The examination of the nervous system and the interpretation of findings require a lot of practice. In a viva voce examination, this system more than any other system requires a polished technique. The signs need to be elicited carefully because the precise anatomical localisation of any lesions can often be determined this way. It is important, therefore, to remember some elementary neuroanatomy.

Examination can be long and difficult and it is said to take much of a day if absolutely everything that can be done (including psychometric assessment) is done. This is obviously impractical, but a screening examination that will uncover most signs takes only a relatively short time.

In brief, the following aspects of the examination must be attended to:

1. General, including examination for neck stiffness, assessment of the higher centres, speech, and abnormal movements.

2. The cranial nerves II to XII.

3. The upper limbs. Motor system: inspection, tone, power, reflexes, coordination. Sensory system: pinprick sensation, proprioception, vibration sense, light touch.

4. The lower limbs: as for the upper limbs, but including assessment of walking (gait).

5. The skull and spine for local disease.

6. The carotid arteries for bruits.

General signs

Consciousness

Note the level of consciousness. If the patient is unconscious look for responses to various stimuli (page 401).

Neck stiffness

Any patient with an acute neurological illness, or who is febrile or has altered mental status **must** be assessed for signs of meningism.[6]

With the patient lying flat in bed, the examiner slips a hand under the occiput and gently flexes the neck passively (i.e. without assistance from the patient). The chin is brought up to approach the chest wall. Meningism may be caused by pyogenic or other infection of the meninges, or by blood in the subarachnoid space secondary to subarachnoid haemorrhage. There is resistance to neck flexion due to painful spasm of the extensor muscles of the neck. Other causes of resistance to neck flexion are characterised by an equal resistance to head rotation. They include: (i) cervical spondylosis; (ii) after cervical fusion; (iii) Parkinson's disease; and (iv) raised intracranial pressure, especially if there is impending tonsillar herniation. The *Brudzinski sign*[e] is spontaneous flexion of the hips during flexion of the neck by the examiner and indicates meningism.

Kernig's sign[f] should also be elicited if meningitis is suspected. Flex each hip in turn, then attempt to straighten the knee while keeping the hip flexed. This is greatly limited by spasm of the hamstrings (which in turn causes pain) when there is meningism due to an inflammatory exudate around the lumbar spinal roots.

Although the diagnostic value has been questioned (combined meningeal signs had a positive LR of 0.92 and a negative LR of 0.88),[6] we have found these signs useful clinically (and they have excellent specificity).

Handedness

Shake the patient's hand and ask if he or she is right- or left-handed. This is polite and allows the examiner to assess the likely dominant hemisphere. Ninety-four per cent of right-handed people and about 50% of left-handed people have a dominant left hemisphere. There is division of function between the two hemispheres, the most obvious distinction being that the dominant hemisphere controls language and mathematical functions.

Orientation

Test orientation in *person*, *place* and *time* by asking the patient his or her name, present location and the date (normal patients who have been in hospital for long periods often get the day wrong as one day seems very much like another in hospital). Disorientation is not a specific localising sign and may be acute and reversible (delirium) or chronic and irreversible (dementia). The mini-mental state examination (Table 12.7, page 420) is a

useful way to document the progress of a confusional state or dementia over time.

The cranial nerves[g]

Examination anatomy

The cranial nerves (Figure 11.2) arise as direct extensions of the brain (I and II) or from the brainstem (midbrain, pons and medulla)—Figures 11.10 (page 337), 11.13 (page 340) and 11.14 (page 341).

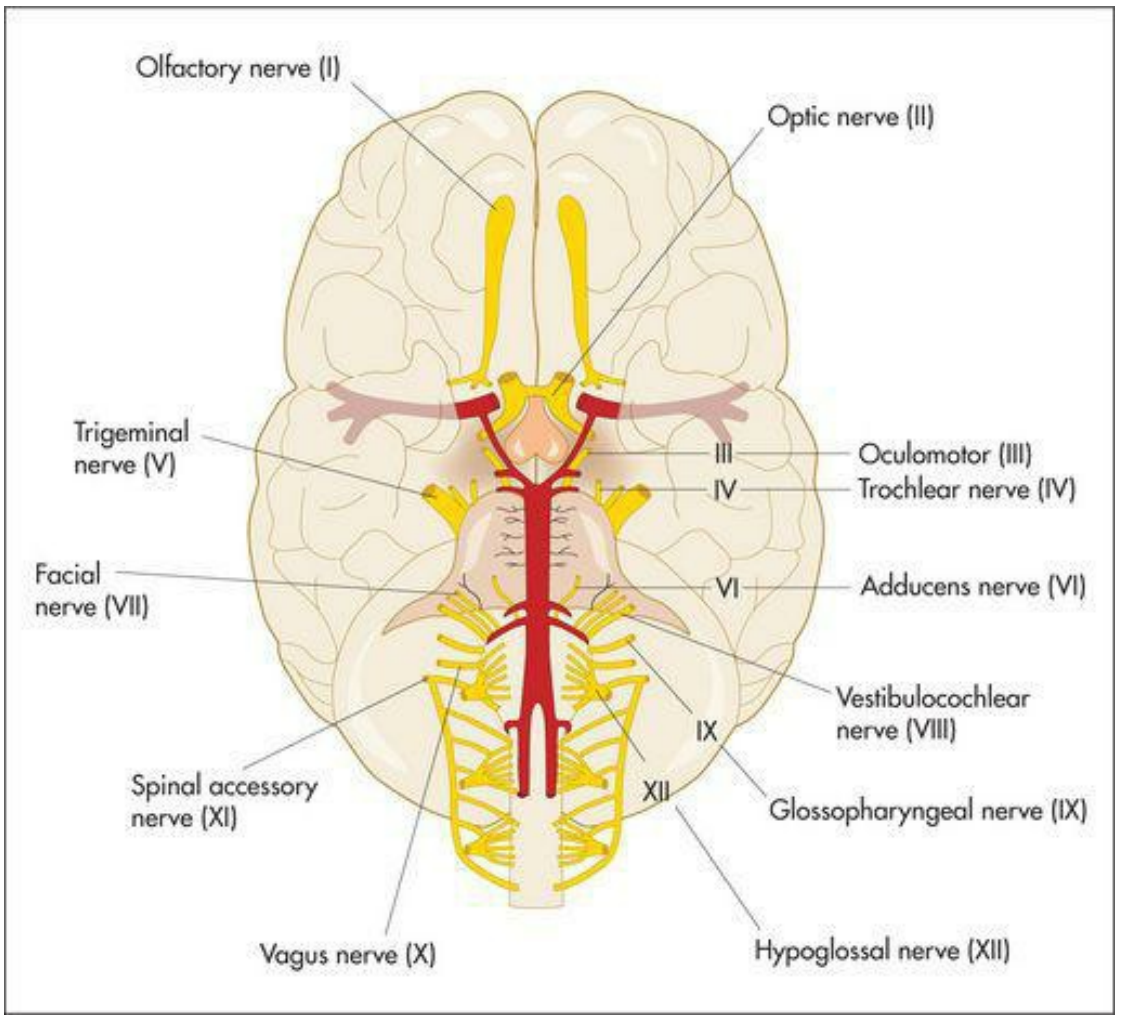


Figure 11.2 Cranial nerves

If possible, position the patient so that he or she is sitting over the edge of the bed. Look at the head, face and neck. If hydrocephalus has occurred in infancy—before closure of the cranial sutures—the head and face may

resemble an inverted triangle. Acromegaly (page 307), Paget's disease (page 320) or basilar invagination (page 321) may be obvious. A careful *general inspection* may reveal signs easily missed when each cranial nerve is examined separately. This is particularly true of ptosis (page 337), proptosis (page 303), pupillary inequality (page 336), skew deviation of the eyes and facial asymmetry. Inspect the whole scalp for craniotomy scars and the skin for neurofibromas (Figure 11.3). Look for skin lesions: for example, a capillary or cavernous haemangioma is seen on the face in the distribution of the trigeminal (V) nerve in the Sturge-Weber syndrome.[h] It is associated with an intracranial venous haemangioma of the leptomeninges and with seizures.

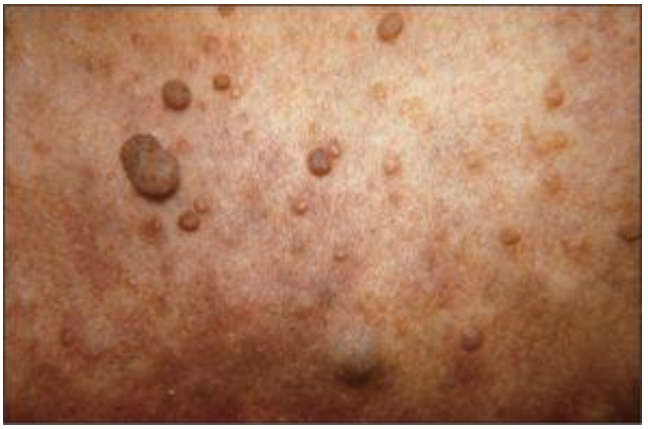


Figure 11.3 Subcutaneous neurofibromas in neurofibromatosis type I, associated with optic nerve and pontine gliomas (acoustic neuromas occur in type II)

The cranial nerves are usually tested in approximately the order of their number.[7]

The first (olfactory) nerve[i]

Examination anatomy

This is a purely sensory nerve, whose fibres arise in the mucous membrane of the nose and pass through the cribriform plate of the ethmoid bone to synapse in the olfactory bulb. From here the olfactory tract runs under the frontal lobe and terminates in the medial temporal lobe on the same side.

Examination of the nose and sense of smell

Note the external appearance of the nose. Look for rash or deformity. Then examine the nasal vestibule by elevating the tip of the nose (in adults a speculum is usually needed to give an adequate view).

The first nerve is not tested routinely. If the patient complains of loss of smell (anosmia) or there are other signs suggesting a frontal or temporal lobe lesion, then it should be examined. Anosmic patients sometimes complain of loss of taste rather than of smell because the sense of smell plays a large part in the appreciation of taste. Test each nostril separately with a series of bottles containing essences of familiar smells, such as coffee, vanilla and peppermint (this is traditional, but not very reliable). Pungent substances such as ammonia should not be used, first because they upset the patient and second because noxious stimuli of this sort are detected by sensory fibres of the fifth (trigeminal) nerve. An easy way to test smell is to use the isopropyl alcohol wipes present in most hospital clinics. These have a distinctive and non-pungent smell.

Examination of the nasal passages must be performed if anosmia is present. Polyps and mucosal thickening may be seen and may explain the findings.

Causes of anosmia

Most cases of anosmia are bilateral. Causes include: (i) upper respiratory tract infection (commonest); (ii) smoking and increasing age; (iii) ethmoid tumours; (iv) basal skull fracture or frontal fracture, or after pituitary surgery; (v) congenital—for example, Kallmann's syndrome (hypogonadotrophic hypogonadism); (vi) meningioma of the olfactory groove; and (vii) following meningitis. The main unilateral causes are head trauma without a fracture, or an early meningioma of the olfactory groove.[i]

The second (optic) nerve

Examination anatomy

The optic nerve is not really a nerve but an extension of fibres of the central nervous system that unites the retinas with the brain. It is purely sensory, contains about a million fibres and extends for about 5 cm (Figure 11.4), passing through the optic foramen close to the ophthalmic artery and joining the nerve from the other side at the base of the brain to form the optic chiasm. The spatial orientation of fibres from different parts of the fundus is

preserved so that fibres from the lower part of the retina are found in the inferior part of the chiasm, and vice versa. Fibres from the temporal visual fields (the nasal halves of the retinas) cross in the chiasm, whereas those from the nasal visual fields do not. Fibres for the light reflex from the optic chiasm finish in the superior colliculus, whence connections occur with both third nerve nuclei. The remainder of the fibres leaving the chiasm are concerned with vision, and travel in the optic tract to the lateral geniculate body. From here the fibres form the optic radiation and pass through the posterior part of the internal capsule, finishing in the visual cortex of the occipital lobe. In their course they splay out so that fibres serving the lower quadrants course through the parietal lobe, while those for the upper quadrants traverse the temporal lobe. The result of the decussation of fibres in the optic chiasm is that fibres from the left visual field terminate in the right occipital lobe, and vice versa.

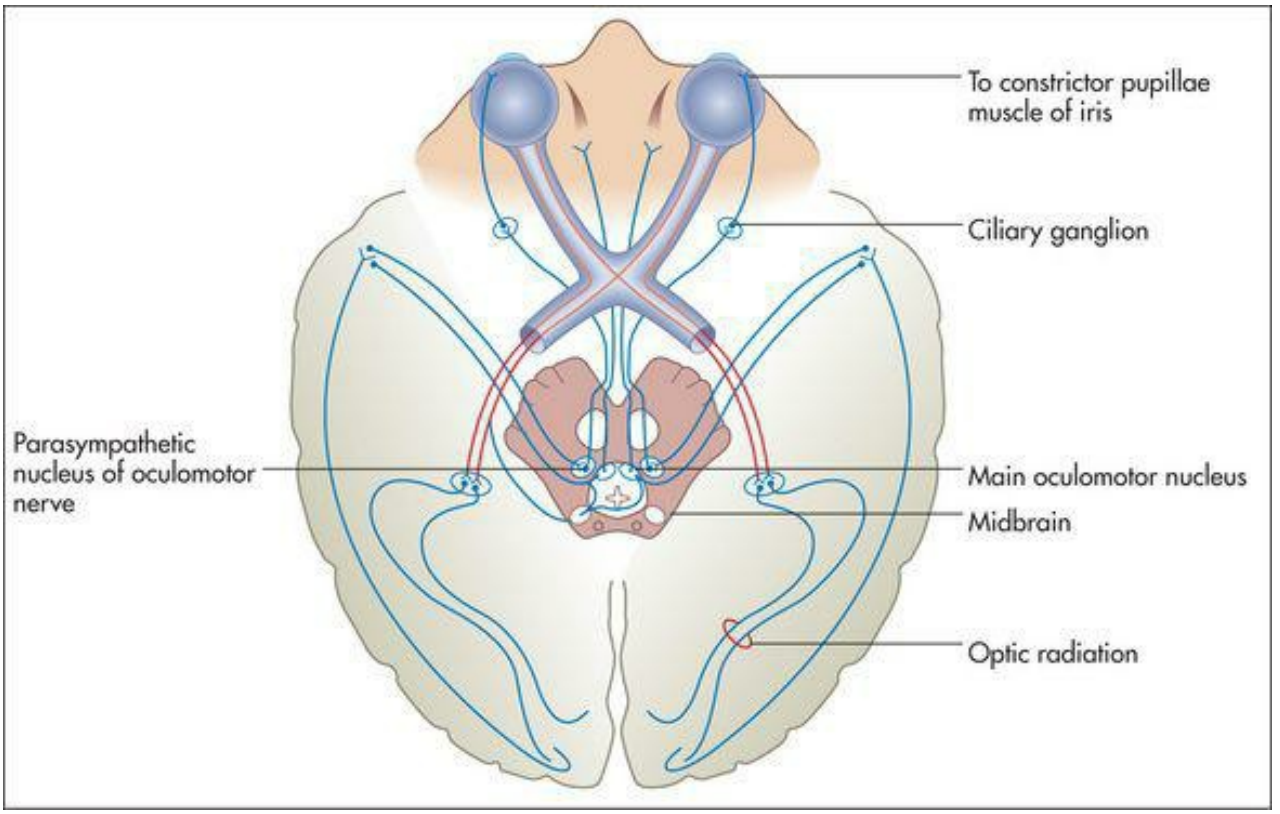


Figure 11.4 The optic pathways and visual reflexes

Adapted from Snell RS, Westmoreland BF, Clinical neuroanatomy for medical students, 4th edn. Boston: Little Brown, 1997.

History

The majority of visual symptoms involve reduction in visual acuity. These

are discussed in detail with the physical examination findings. Some patients notice a more specific change, which will help direct the examination. Ask about the time course of the visual disturbance and whether it seems to involve the vision of one eye or one visual field. Sudden loss of vision in one eye (often described as the awareness of a curtain being drawn across the eye) may be due to an embolus to the retina. These are called *negative* visual symptoms. There is usually, but not always, spontaneous return of vision. This is called *amaurosis fugax*. Migraine attacks may be preceded by subjective visual changes, including scintillating scotomas, photophobia, blurred vision or hemianopia. Visual hallucinations such as flashing lights and distortions of vision are called *positive* visual symptoms. They can occur in psychotic states or as the aura of an epileptic seizure.

More gradual loss of vision has many possible causes.

Examination

Assess visual acuity, visual fields and the fundi.

Visual acuity is tested with the patient wearing his or her spectacles, if used for reading or driving, as refractive errors are *not* considered to be cranial nerve abnormalities. Use a hand-held eye chart or a Snellen's chart[k] on the wall. Each eye is tested separately, while the other is covered by a small card.

Formal testing with a standard Snellen's chart requires the patient to be 6 metres from the chart. Unless a very large room is available, this is done using a mirror. Normal visual acuity is present when the line marked 6 can be read correctly with each eye (6/6 acuity). If poor visual acuity improves when the patient is asked to read the chart through a pin-hole, refractive error is likely to be the cause. A patient who is unable to read even the largest letter of the chart should be asked to count fingers held up in front of each eye in turn, and if this is not possible, then perception of hand movement is tested. Failing this, light perception only may be present.

Any abnormality of the lens, cornea, fundus or optic nerve pathway can cause reduction in visual acuity:

- *Causes of bilateral blindness of rapid onset* include bilateral occipital lobe infarction; bilateral occipital lobe trauma; bilateral optic nerve damage, as with methyl alcohol poisoning; and hysteria.

- *Sudden blindness in one eye* can be due to retinal artery or vein occlusion, temporal arteritis, non-arteritic ischaemic optic neuropathy and occasionally optic neuritis or migraine.

• *Bilateral blindness of gradual onset* may be caused by cataracts; acute glaucoma; macular degeneration; diabetic retinopathy (vitreous haemorrhages); bilateral optic nerve or chiasmal compression; and bilateral optic nerve damage—for example, tobacco *amblyopia* (blindness due to retinal disease).

Visual fields are examined by *confrontation* (Figure 11.5). Always remove a patient's spectacles first. The examiner's head should be level with the patient's head. Use a white- or red-tipped hat pin or pen. Test each eye separately. The examiner holds the pin at arm's length with the coloured head upwards. It should be positioned halfway between the patient and the examiner, and brought in from just outside the examiner's peripheral vision until the patient can see it. Make sure the patient is staring directly at the examiner's eye and explain that he or she is looking for the first sight of the pin out of the corner of the eye. When the right eye is being tested the patient should look straight into the examiner's left eye. The patient's head should be at arm's length and the eye not being tested should be covered. The pin should be brought into the visual field from the four main directions, diagonally towards the centre of the field of vision.

Figure 11.5 Visual field testing: 'Tell me when you first see the red pin come into view'

Next the blind spot can be mapped out by asking about disappearance of the pin around the centre of the field of vision of each eye. The pin is moved slowly across the field of vision. A large central scotoma will lead to its apparent temporary disappearance and then reappearance. Only a gross enlargement may be detectable.

If a patient has such poor acuity that a pin is difficult to see, the fields should be mapped with the fingers. The examiner's fingers can also be used to perform a quick screening test of the visual fields. Usually two fingers are

held up and brought into the centre of vision in the four quadrants. The examiner wriggles the fingers and asks the patient to say 'yes' when movement of the fingers is first seen. The following patterns of visual field loss may be detected (Figures 11.6 and 11.7):

• *Concentric diminution of the field (tunnel vision)* may be caused by glaucoma; retinal abnormalities such as chorioretinitis or retinitis pigmentosa; papilloedema; or acute ischaemia, as with migraine. Normally even a reduced field of vision widens as objects are moved further away. Tubular diminution of the visual fields suggests hysteria. There is always a small area close to the centre of the visual fields where there is no vision (the blind spot). This is the area where the optic disc is seen on fundoscopy and is the point where the optic nerve joins the retina. The blind spot enlarges with papilloedema.

• *Central scotomata, or loss of central (macular) vision,* may be due to demyelination of the optic nerve (multiple sclerosis causes unilateral or asymmetrical bilateral scotomata); toxic causes, such as methyl alcohol (symmetrical bilateral scotomata); nutritional causes, such as tobacco or alcohol amblyopia (symmetrical central or centrocecal scotomata); vascular lesions (unilateral); and gliomas of the optic nerve (unilateral).

• *Total unilateral visual loss* is due to a lesion of the optic nerve or to unilateral eye disease.

• *Bitemporal hemianopia* is due to a lesion that affects the centre of the optic chiasm, damaging fibres from the nasal halves of the retinas as they decussate. This will result in loss of both temporal halves of the visual fields. Causes include a pituitary tumour, a craniopharyngioma and a suprasellar meningioma.

• *Binasal hemianopia* is very rare and is due to bilateral lesions affecting the uncrossed optic fibres, such as atheroma of the internal carotid siphon.

• *Homonymous hemianopia* is due to a lesion that damages the optic tract or radiation, affecting the visual field on the right or left side. For example, left temporal and right nasal field loss will occur with a right-sided lesion. The exact nature of the defect depends on the site of interruption of the fibres. In the optic tract the defect is usually complete—there is no macular sparing. In the more posterior optic radiation the macular vision is usually spared if the cause is ischaemia, but not if a destructive process such as tumour or haemorrhage is responsible. The macular cortical area is thought to have some additional blood supply from the anterior and middle cerebral arteries.

• *Homonymous quadrantanopia* is loss of the upper or lower homonymous

- *Homonymous quadrantanopia* is loss of the upper or lower homonymous quadrants of the visual fields. This may be due to temporal lobe lesions (e.g. vascular lesions or tumours), which cause upper quadrantanopia, or parietal lobe lesions (e.g. vascular lesions or tumours), which cause lower quadrantanopia.

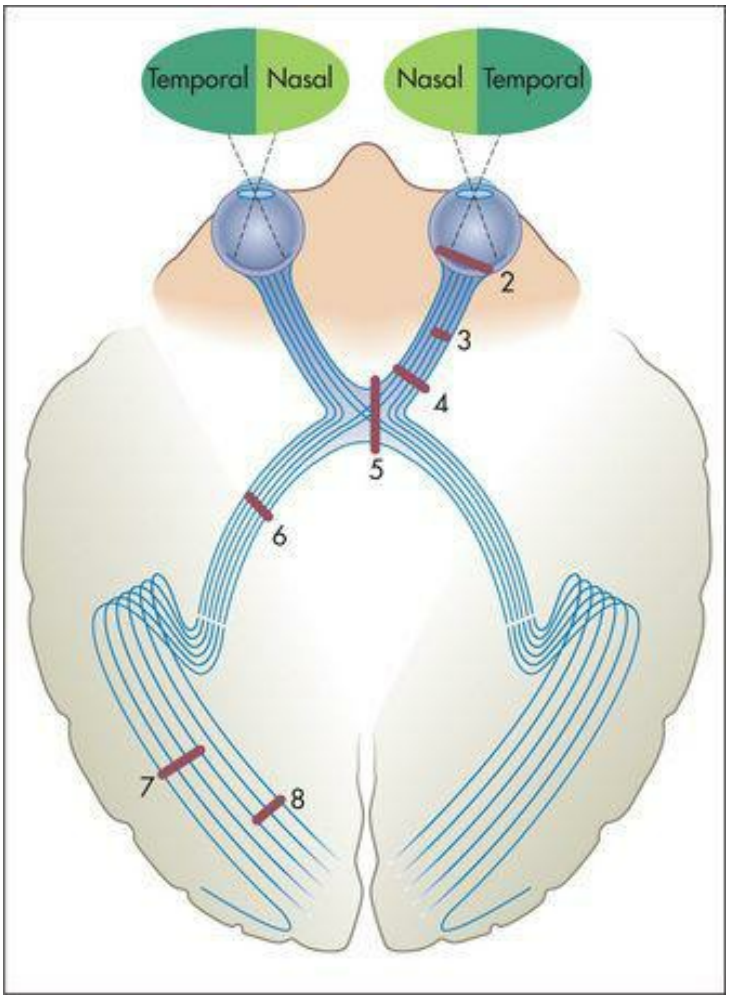


Figure 11.6 The visual fields and optic pathways Numbers indicate sites of lesions producing field defects shown in Figure 11.7.

Adapted from Snell RS, Westmoreland BF, Clinical neuroanatomy for medical students, 4th edn. Boston: Little Brown, 1997.

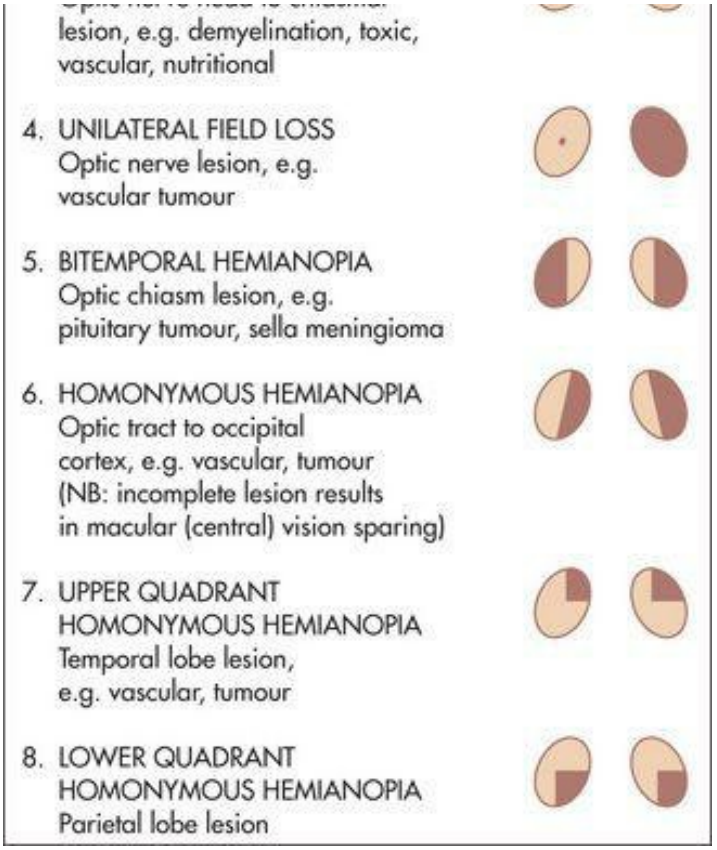


Figure 11.7 Visual field defects with lesions at various levels along the optic pathway, at sites indicated in Figure 11.6

Adapted from Bickerstaff ER, Spillane JA. Neurological examination in clinical practice, 5th edn. Oxford: Blackwell, 1989.

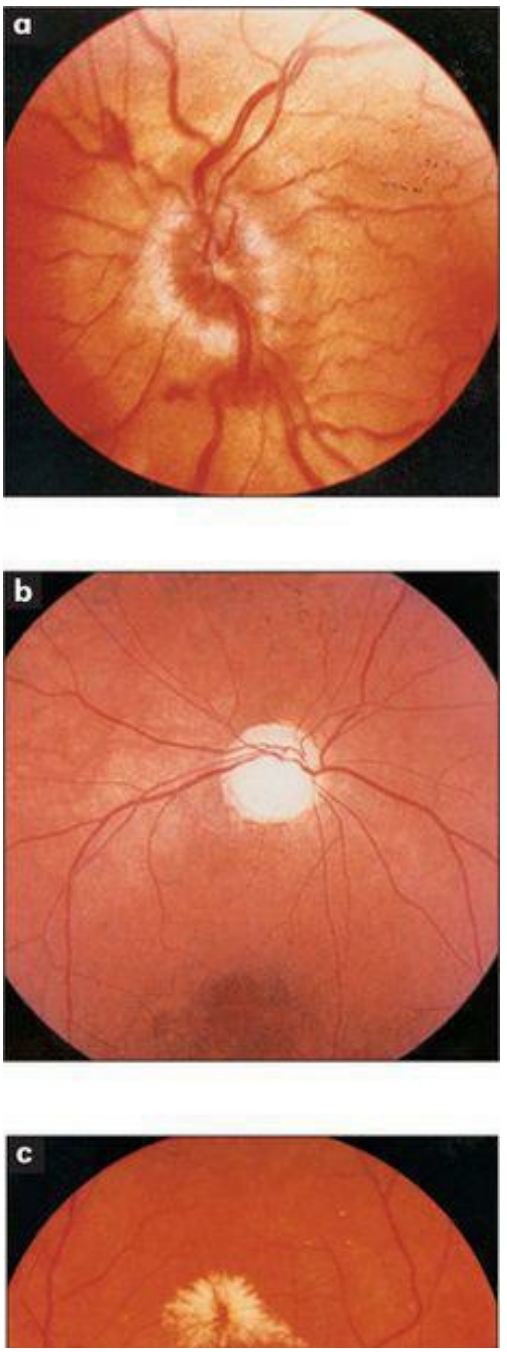
The presence of an abnormality has diagnostic value (positive LRs 4.2 to 6.8),[8] but absence is largely unhelpful.

Fundoscopy does not begin with the examination of the fundus, but rather with visualisation of the cornea with the ophthalmoscope. Use the right eye to look in the patient's right eye, and vice versa. This prevents contact between the noses of the patient and the examiner in the midline. Keep your head vertical so that the patient can fix with the other eye.

Begin with the ophthalmoscope on the +20 lens setting, with the patient gazing into the distance. This prevents reflex pupil contraction, which occurs if the patient attempts to accommodate. Look first at the cornea and iris, and then at the lens. Large corneal ulcers may be visible, as may undulation of the rim of the iris, which is due to previous lens extraction and is called iridodonesis.

By racking the ophthalmoscope down towards 0, the focus can be shifted towards the fundus. Opacities in the lens (cataracts) may prevent inspection of the fundus. When the retina is in focus, search first for the optic disc. This is done by following a large retinal vein back towards the disc. All these veins radiate from the optic disc.

The margins of the disc must be examined with care. The disc itself is usually a shallow cup with a clearly outlined rim. Loss of the normal depression of the optic disc will cause blurring at the margins and is called papilloedema (Figure 11.8a). It indicates raised intracranial pressure. If papilloedema is suspected, the retinal veins should be examined for spontaneous pulsations. When these are present raised intracranial pressure is excluded, but their absence does not prove the pressure is raised.[9] If the appearance of papilloedema is associated with demyelination in the anterior part of the optic nerve, it is called papillitis (Table 13.3, page 428). These two can be distinguished because papillitis causes visual loss but papilloedema does not.



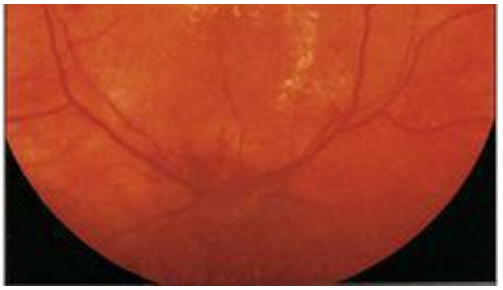


Figure 11.8 Fundoscopy in the neurological patient
(a) Papilloedema. (b) Optic atrophy. (c) Grade 4 hypertensive retinopathy, with papilloedema, a 'macular star' of hard exudates collecting around the fovea, and retinal oedema.

Next note the colour of the optic disc. Normally it is a rich yellow colour in contrast to the rest of the fundus which is a rich red colour. The fundus may be pigmented in some diseases and in patients with pigmented skin. When the optic disc has a pale insipid white colour, optic atrophy is usually present (Figure 11.8b).

Each of the four quadrants of the retina should be examined systematically for abnormalities. Look especially for diabetic and hypertensive changes (Figure 11.8c). Note haemorrhages or exudates.

The third (oculomotor), fourth (trochlear) and sixth (abducens) nerves—the ocular nerves

Examination anatomy

The size of the pupils depends on a balance of parasympathetic and sympathetic innervation. The parasympathetic innervation to the eyes is supplied by the Edinger-Westphal nucleus[1] of the third nerve (stimulation of these fibres causes pupillary constriction: *miosis*). The sympathetic innervation to the eye (stimulation causes pupillary dilatation: *mydriasis*) is as follows: fibres from the hypothalamus go to the ciliospinal centre in the spinal cord at C8, T1 and T2, synapse, and second-order neurones exit via the anterior ramus in the thoracic trunk and synapse in the superior cervical ganglion in the neck. Third-order neurones travel from here with the internal carotid artery to the eye. In addition, the pupillary reflexes (Figure 11.9) depend for their afferent limb on the optic nerve (Figure 11.4). Constriction of the pupil in response to light is relayed by the optic nerve and tract to the superior colliculus and then to the Edinger-Westphal nucleus of the third nerve in the midbrain. Efferent motor fibres from the oculomotor nucleus

(Figure 11.10) travel in the wall of the cavernous sinus, where they are in association with the fourth, ophthalmic division of the fifth, and the sixth cranial nerves (see Figure 10.10, page 308). These nerves leave the skull together through the superior orbital fissure. The iridoconstrictor fibres terminate in the ciliary ganglion, whence postganglionic fibres arise to innervate the iris. The rest of the third nerve supplies all the ocular muscles except the superior oblique (fourth nerve) and the lateral rectus (sixth nerve) muscles. The third nerve also supplies the levator palpebrae superioris, which elevates the eyelid (Figure 11.11).

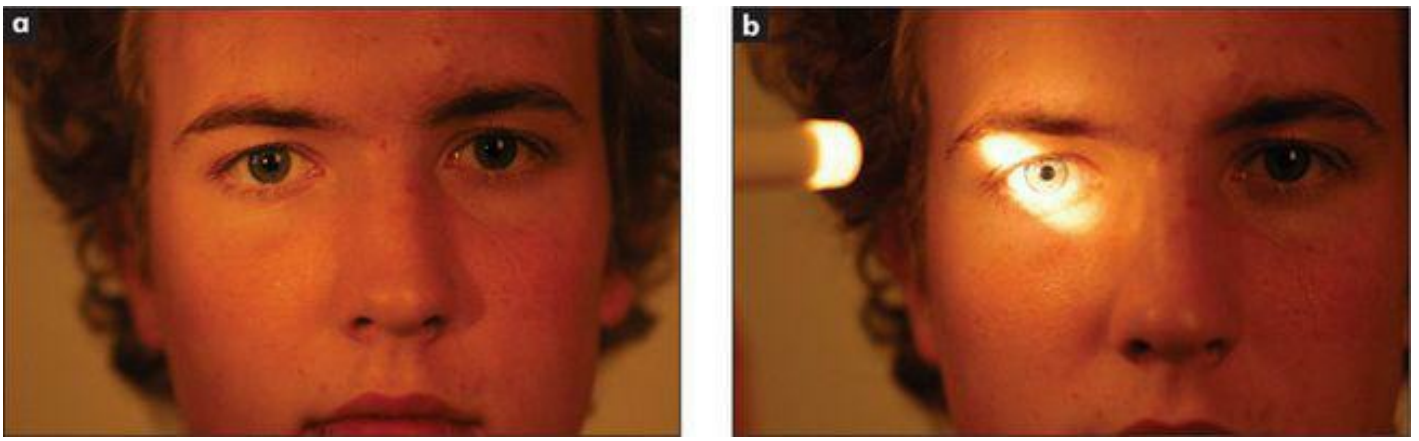


Figure 11.9 Cranial nerves II and III
(a) The pupils: inspect for size and symmetry. (b) Testing the pupillary reflex.

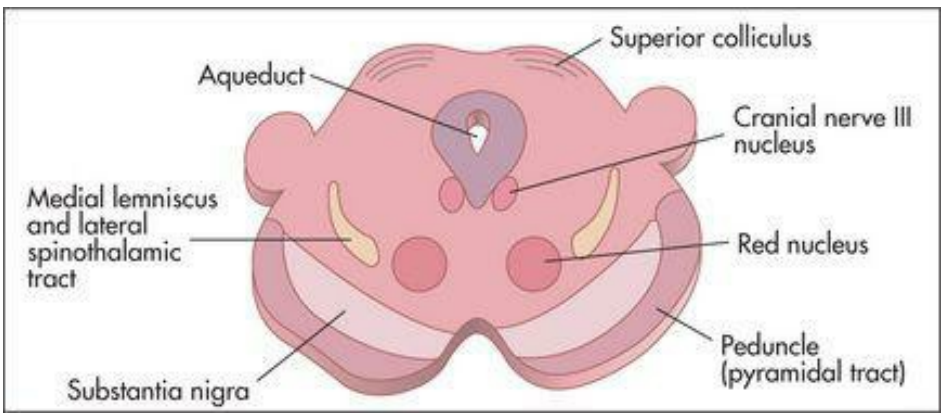
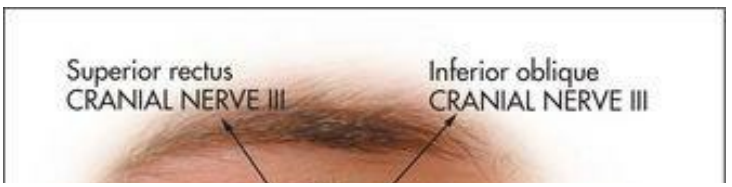


Figure 11.10 (left) Anatomy of the midbrain



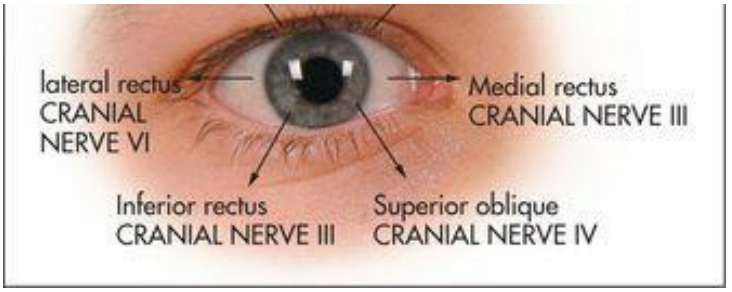


Figure 11.11 The eye muscles and nerve innervation

Examination

Assess the pupils and movements of the eye.

The pupils

With the patient looking at an object at an intermediate distance, examine the pupils for size, shape, equality and regularity. Slight differences in pupil size (up to 20%) may be normal.[m]

Look for *ptosis* (drooping) of one or both eyelids. Remember that *ectropion* or drooping of the lower lid is a common degenerative problem in old age but can also be caused by a seventh nerve palsy or facial scarring. There is often eye irritation and watering associated with it because of defective tear drainage.

Test the *light reflex*. Using a pocket torch, shine the light from the side (so the patient does not focus on the light and accommodate) into one of the pupils to assess its reaction to light. Inspect both pupils and repeat this procedure on the other side. Normally the pupil into which the light is shone constricts briskly—this is the *direct* response to light. Simultaneously, the other pupil constricts in the same way. This is called the *consensual* response to light.

Move the torch in an arc from pupil to pupil. If an eye has optic atrophy or severely reduced visual acuity from another cause, the affected pupil will dilate paradoxically after a short time when the torch is moved from the normal eye to the abnormal eye. This is called an *afferent pupillary defect* (or the Marcus Gunn pupillary sign[n]). It occurs because an eye with severely reduced acuity has reduced afferent impulses so that the light reflex is markedly decreased. When the light is shone from the normal eye to the abnormal one the pupil dilates, as reflex pupillary constriction in the abnormal eye is so reduced that relaxation after the consensual response dominates.

Now test *accommodation*. Ask the patient to look into the distance and

Now test *accommodation*. Ask the patient to look into the distance and then to focus his or her eyes on an object such as a finger or a white-tipped hat pin brought to a point about 30 cm in front of the nose. There is normally constriction of both pupils—the accommodation response. It depends on a pathway from the visual association cortex descending to the third nerve nucleus. Causes of an absent light reflex with an intact accommodation reflex include a midbrain lesion (e.g. the Argyll Robertson pupil of syphilis), a ciliary ganglion lesion (e.g. Adie's pupil[o]) or Parinaud's syndrome[p] (page 340). Failure of accommodation alone may occur occasionally with a midbrain lesion or with cortical blindness.

Eye movements

Here failure of eye movement, double vision (diplopia) and nystagmus are assessed.

Normally the eyes move in parallel except during convergence. When they move out of alignment the patient is said to have *strabismus* or a *squint.* This abnormality may be due to a cranial nerve palsy (III, IV or VI), and in these cases the angle of alignment changes depending on the direction of gaze (an *incomitant* squint). When the malalignment of the eye movement remains constant for any direction of gaze the squint is said to be *concomitant*. Concomitant squints are common in children and may be idiopathic or occasionally caused by an intracranial mass. Strabismus is associated with diplopia unless one of the images has been suppressed by the brain. This can happen quite quickly in children and may lead to severe visual loss in that eye—*amblyopia.*

Ask the patient to look at the invaluable hat pin. (The presence of these pins in the lapel of a well-cut white coat or expensive suit often indicates that the wearer is a neurologist.) Assess voluntary eye movements in both eyes first. Ask the patient to look laterally right and left, then up and down (Figure 11.12). Remember the lateral rectus (sixth nerve) only moves the eyes horizontally outwards, while the medial rectus (third nerve) only moves the eyes horizontally inwards. The remainder of the muscle movements are a little more complicated. When the eye is abducted, the elevator is the superior rectus (third nerve), while the depressor is the inferior rectus (third nerve). When the eye is adducted, the elevator is the inferior oblique (third nerve) while the depressor is the superior oblique (fourth nerve) (Figure 11.11). The practical upshot of all this is that the testing of pure movement (that is, one muscle only) for elevation and depression is performed first with the eye adducted and then with it abducted. Therefore, ask the patient to follow the moving hat pin, held by the examiner 30–40 cm from the patient and moved in an H pattern, with both eyes and to say if double images are

seen in any direction.

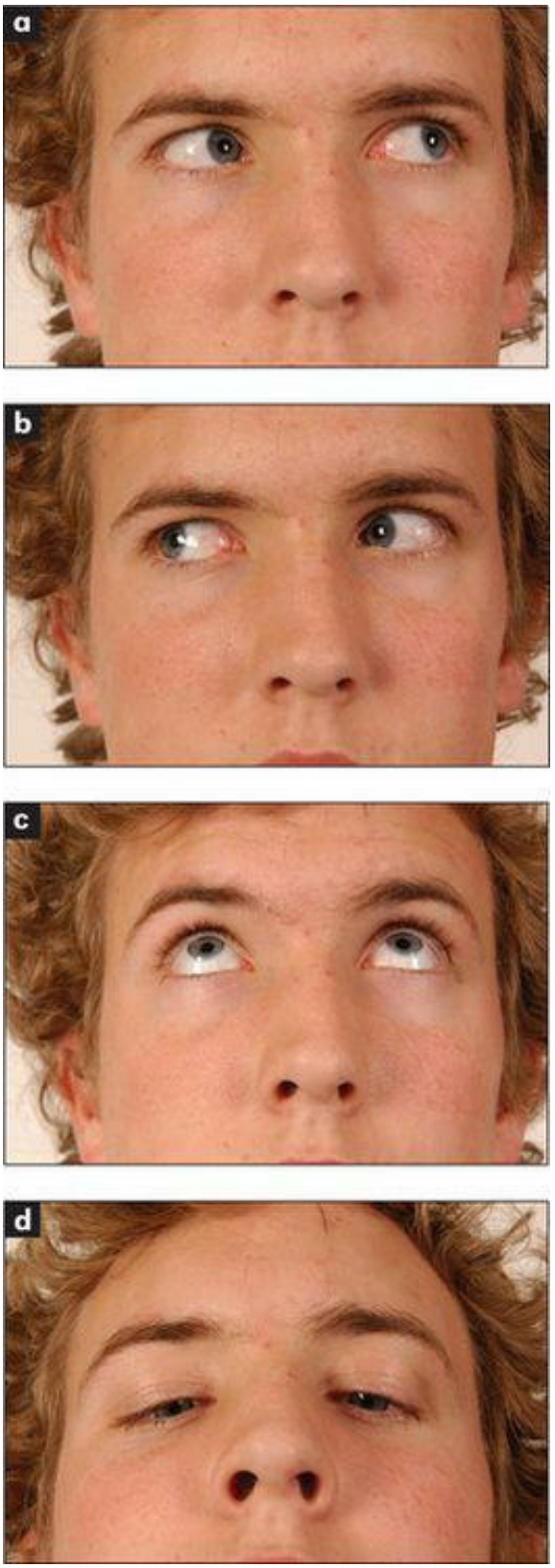


Figure 11.12 The cranial nerves III, IV and VI: voluntary eye movements
(a) 'Look to the left.' (b) 'Look to the right.' (c) 'Look up.' (d) 'Look down.'

Diplopia can be an early sign of ocular muscle weakness because the

Diplopia can be an early sign of ocular muscle weakness because the light falls on different parts of the corresponding retinas due to slight movement differences. If diplopia is present, further testing is necessary. The false image is usually paler, less distinct and always more peripheral than the real one. Ask the patient whether the two images lie side by side or one above the other. If they are side by side, only the lateral or medial recti can be responsible. If they lie one above the other, then either of the obliques or the superior or inferior recti may be involved. To decide which pair of muscles is responsible, ask in which direction there is maximum image separation. Separation is greatest in the direction in which the weak muscle has its purest action. At the point of maximum separation, cover one eye and find out which image disappears. Loss of the lateral image indicates that the covered eye is responsible. Diplopia that persists when one eye is covered can be due to astigmatism, a dislocated lens or hysteria.

Note failure of movement of either eye in any direction. This indicates ocular muscle involvement. If any abnormality is detected, then each eye must be tested separately. The other eye is covered with a card or with the examiner's hand. Abnormal eye movement may be due to III, IV or VI nerve palsy, or to an abnormality of conjugate gaze.

Features of a third nerve lesion

These are complete ptosis (partial ptosis may occur with an incomplete lesion); divergent strabismus (eye 'down and out'); and dilated pupil which is unreactive to direct light (the consensual reaction in the opposite normal eye is intact) and unreactive to accommodation. Always try to exclude a fourth (trochlear) nerve lesion when a third nerve lesion is present. One way to do this is by tilting the head to the same side as the lesion. The affected eye will intort if the fourth nerve is intact (remember SIN—the ***S****uperior* oblique ***IN****torts* the eye).

Aetiology of a third nerve palsy

Third nerve lesions are most commonly related to trauma or are idiopathic. Central causes include vascular lesions in the brainstem, tumours and rarely demyelination.

Peripheral causes include: (i) compressive lesions, such as an aneurysm (usually on the posterior communicating artery), tumour, basal meningitis, nasopharyngeal carcinoma or orbital lesions—for example, Tolosa-Hunt syndrome (superior orbital fissure syndrome—painful lesions of III, IV, VI and the first division of V); and (ii) ischaemia or infarction, as in arteritis,

diabetes mellitus and migraine.

Features of a fourth nerve lesion

Test this nerve by asking the patient to turn the eye in and then try to look down: a lesion results in paralysis of the superior oblique with weakness of downward (and outward) movement. The patient may walk around with his or her head tilted away from the lesion—that is, to the opposite shoulder (this allows the patient to maintain binocular vision).

An isolated fourth nerve palsy is rare and is usually idiopathic or related to trauma. It may occasionally occur with lesions of the cerebral peduncle.

Features of a sixth nerve lesion

These are failure of lateral movement, convergent strabismus and diplopia. These signs are maximal on looking to the affected side, and the images are horizontal and parallel to each other. The outermost image from the affected eye disappears on covering this eye (this image is usually also more blurred).

Aetiology of a sixth nerve palsy

Bilateral lesions may be due to trauma or Wernicke's encephalopathy (a syndrome of ophthalmoplegia, confusion and ataxia which is often associated with Korsakoff's psychosis due to thiamine deficiency). Mononeuritis multiplex and raised intracranial pressure are also causes of sixth nerve palsy.

Unilateral sixth nerve lesions are most commonly idiopathic or related to trauma. They may have a central (e.g. vascular lesion or tumour) or peripheral (e.g. raised intracranial pressure or diabetes mellitus) origin.

Abnormalities of conjugate gaze

Normal eye movements occur in an organised fashion so that the visual axes remain in the same plane throughout. There are centres for conjugate gaze in the frontal lobe for saccadic movements and in the occipital lobe for pursuit movements. Conjugate movement to the right is controlled from the left side of the brain. From these centres fibres travel to the region of the sixth nerve nucleus, from which area the medial longitudinal fasciculus coordinates movement with the contralateral third nerve (medial rectus) nucleus (Figure 11.13). A brainstem lesion causes ipsilateral paralysis of horizontal conjugate gaze, and a frontal lobe lesion causes contralateral paralysis of horizontal

gaze, and a frontal lobe lesion causes contralateral paralysis of horizontal conjugate gaze.

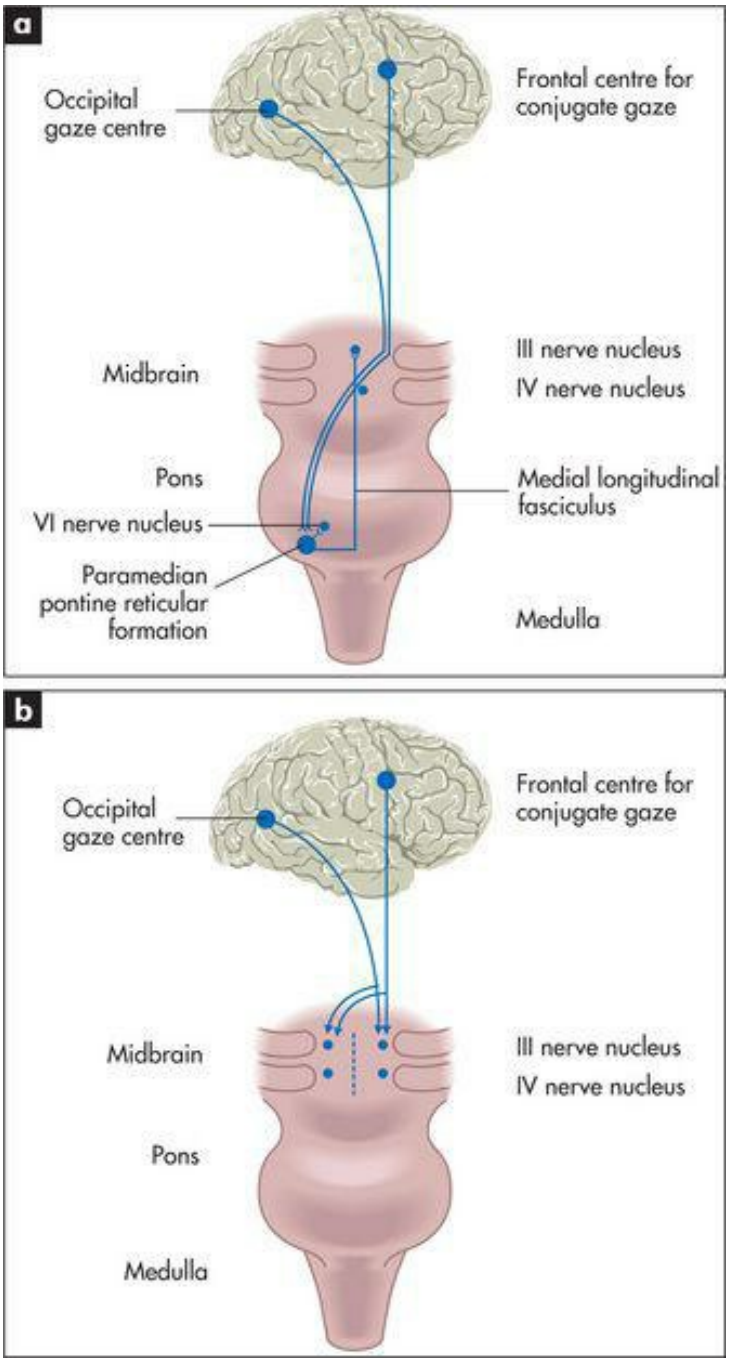


Figure 11.13 Horizontal (a) and vertical (b) eye movements

Adapted from Lance JW, McLeod JG. A physiological approach to clinical neurology, 3rd edn. London: Butterworths, 1981.

There are a number of possible causes for deviation of the eyes to one side. For example, *deviation of the eyes to the left* can result from: (i) a destructive lesion (usually vascular or neoplastic), which involves the pathways between the *left* frontal lobes and the oculomotor nuclei; (ii) a destructive lesion of the *right* side of the brainstem; or (iii) an irritative lesion

destructive lesion of the *right* side of the brainstem, or (iii) an irritative lesion, such as an epileptic focus, of the *right* frontal lobe, which stimulates deviation of the eyes to the left.

Supranuclear palsy is loss of vertical or horizontal gaze or both (Figure 11.13). The clinical features that distinguish this from third, fourth and sixth nerve palsies include: (i) both eyes are affected; (ii) pupils may be fixed and are often unequal; (iii) there is usually no diplopia; and (iv) the reflex eye movements—for example, on flexing and extending the neck—are usually intact.

• *Progressive supranuclear palsy* (or Steele Richardson Olszewski syndrome[9]): here there is loss of vertical and later of horizontal gaze, which is associated with extrapyramidal signs, neck rigidity and dementia. Reflex eye movements on neck flexion and extension are preserved until late in the course of the disease.

• *Parinaud's syndrome* is loss of vertical gaze often associated with nystagmus on attempted convergence (see below). There are pseudo-Argyll Robertson pupils. The causes of Parinaud's syndrome include a pinealoma, multiple sclerosis and vascular lesions.

• Involuntary upward deviation of the eyes (*oculogyric crises*) occurs with post-encephalitic Parkinson's disease and may be seen in patients sensitive to phenothiazine derivatives or in patients on levodopa therapy.

One-and-a-half syndrome is rare but important to recognise. These patients have a horizontal gaze palsy when looking to one side (the 'one') plus impaired adduction on looking to the other side (the 'and-a-half'). Other features often include turning out (exotropia) of the eye opposite the side of the lesion (paralytic pontine exotropia). One-and-a-half syndrome can be caused by a stroke (infarct), plaque of multiple sclerosis or tumour in the dorsal pons.

Nystagmus

The eyes are normally maintained at rest in the midline by the balance of tone between opposing ocular muscles. Disturbance of this tone, which depends on impulses from the retina, the muscles of the eyes themselves and various vestibular and central connections, allows the eyes to drift in one direction. This drift is corrected by a quick movement (saccadic) back to the original position. When these movements occur repeatedly nystagmus is said to be present. The direction of the nystagmus is defined as that of the fast (correcting) movement, although it is the slow drift that is abnormal. Nystagmus from any cause tends to be accentuated by gaze in a direction

Nystagmus from any cause tends to be accentuated by gaze in a direction away from the midline. In many instances nystagmus is not present when the eyes are at rest, and is only detected when the eyes are deviated (gaze-evoked nystagmus). At the extremes of gaze, fine nystagmus is normal (physiological). Therefore test for nystagmus by asking the patient to follow your pin out to 30 degrees from the central gaze position.

Nystagmus may be jerky or pendular.

Jerky horizontal nystagmus may be due to (i) a vestibular lesion (acute lesions cause nystagmus away from the side of the lesion while chronic lesions cause nystagmus to the side of the lesion); (ii) a cerebellar lesion (unilateral diseases cause nystagmus to the side of the lesion); (iii) toxic causes, such as phenytoin and alcohol (may also cause vertical nystagmus but less often); and (iv) *internuclear ophthalmoplegia*. Internuclear ophthalmoplegia is present when there is nystagmus in the abducting eye and failure of adduction of the other (affected) side. This is due to a lesion of the medial longitudinal fasciculus. The most common cause in young adults with bilateral involvement is multiple sclerosis; in the elderly, vascular disease is an important cause.

Jerky vertical nystagmus may be due to a brainstem lesion. (Vertical nystagmus means nystagmus where the oscillations are in a vertical direction.) Upbeat nystagmus suggests a lesion in the midbrain or floor of the fourth ventricle, while downbeat nystagmus suggests a foramen magnum lesion. Phenytoin or alcohol can also cause this abnormality.

With **pendular nystagmus** the nystagmus phases are equal in duration. Its cause may be retinal (decreased macular vision, e.g. albinism) or congenital. This condition is thought to occur as a result of poor vision or increased sensitivity to light. It develops in childhood and occurs as the patient performs searching movements in an attempt to fixate or improve the visual impulses.

A summary of how to approach the medical eye examination is provided on in Chapter 13.

The fifth (trigeminal) nerve

Examination anatomy

This nerve contains both sensory and motor fibres. Its motor nucleus and its sensory nucleus for touch lie in the pons (Figure 11.14), its proprioceptive nucleus lies in the midbrain, while its nucleus serving pain and temperature sensation descends through the medulla to reach the upper cervical cord. It is the largest of the cranial nerves.

the largest of the cranial nerves.

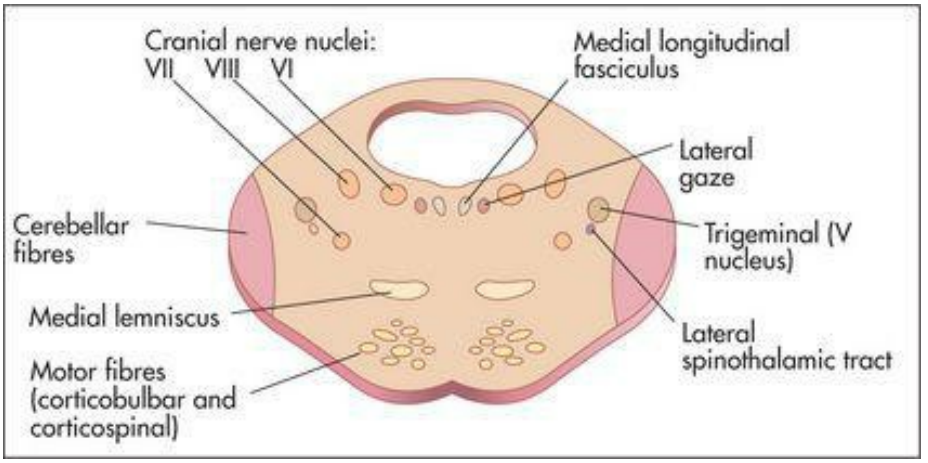


Figure 11.14 Anatomy of the pons

The nerve itself leaves the pons from the cerebellopontine angle and runs over the temporal lobe in the middle cranial fossa. At the petrous temporal bone the nerve forms the trigeminal (Gasserian[1]) ganglion and from here the three sensory divisions arise. The first (ophthalmic) division runs in the cavernous sinus with the third nerve and emerges from the superior orbital fissure to supply the skin of the forehead, the cornea and conjunctiva. The second (maxillary) division emerges from the infraorbital foramen and supplies skin in the middle of the face and the mucous membranes of the upper part of the mouth, palate and nasopharynx. The third and largest (mandibular) division runs with the motor part of the nerve, leaving the skull through the foramen ovale to supply the skin of the lower jaw and mucous membranes of the lower part of the mouth (Figures 11.15 and 11.16).

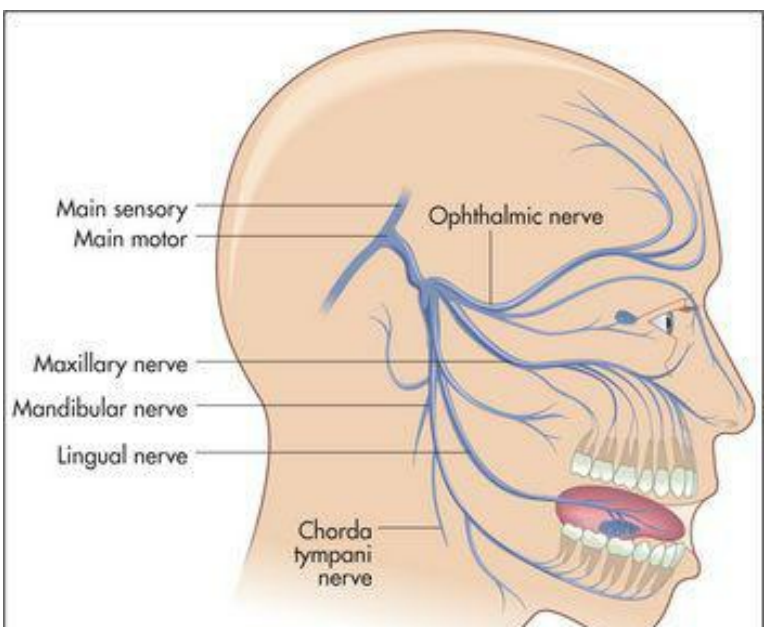


Figure 11.15 The trigeminal nerve (cranial nerve V)

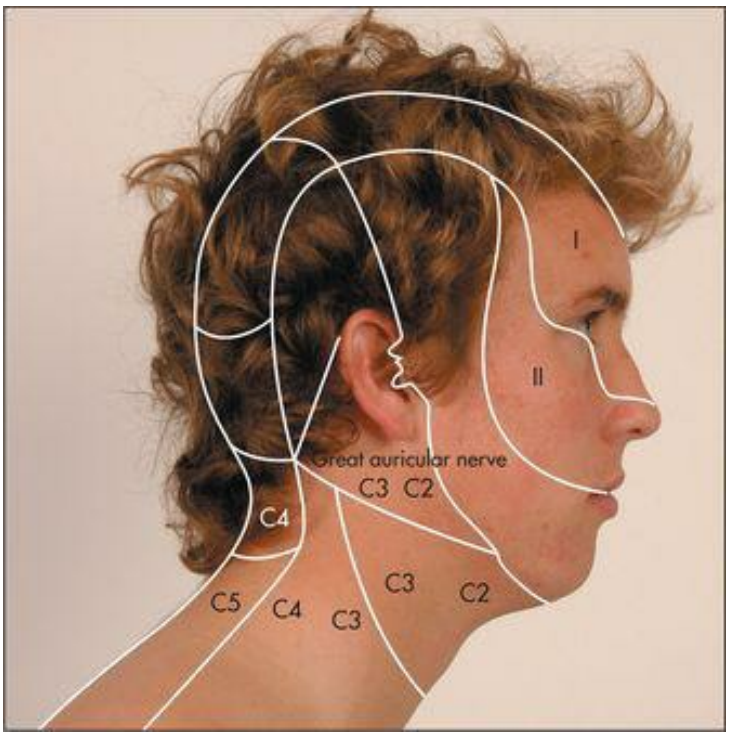


Figure 11.16 Dermatomes of the head and neck

Pain and temperature fibres from the face run from the pons through the medulla as low as the upper cervical cord, terminating in the spinal tract nucleus as they descend. The second-order neurones arise in this nucleus and ascend again as the ventral trigeminothalamic tract. Touch and proprioceptive fibres terminate in the pontine or main sensory and mesencephalic nuclei, respectively, to form the dorsal and ventral mesencephalic tracts. Because of this segregation in the brainstem, lesions of the medulla or upper spinal cord can cause a *dissociated sensory loss of the face*—loss of pain and temperature sensation, but retention of touch and proprioception.

The motor part of the nerve supplies the muscles of mastication.

History

Pain in the distribution of part of the trigeminal nerve is common. *Tic douloureux* (*trigeminal neuralgia*) is a sudden severe shooting pain in one of the divisions of the nerve. It is more common in elderly people. The occurrence in a young woman suggests multiple sclerosis. The pain is brief

but very distressing. It may be precipitated by an activity such as eating or brushing the teeth. The pain is caused by a pontine lesion or by compression of the trigeminal nerve by a vascular abnormality. Pain due to sinusitis, dental abscess, malignant disease of the sinuses and herpes zoster may be felt in a trigeminal nerve distribution. Muscle weakness of the trigeminal nerve may lead the patient to complain of difficulty eating or talking.

Examination

Test the *corneal reflex.* Lightly touch the cornea (*not* the conjunctiva) with a wisp of cottonwool brought to the eye from the side. Reflex blinking of *both* eyes is a normal response. Ask the patient whether he or she feels the touch of the cottonwool. The sensory component of the reflex is mediated by the ophthalmic division of the fifth nerve, while the reflex blink (motor) results from facial nerve innervation of the orbicularis oculi muscles. Absence of corneal sensation is associated with corneal ulceration.

Note: If blinking occurs only with the contralateral eye this indicates an ipsilateral seventh nerve palsy. The patient will then still feel the touch of the cottonwool on the cornea.

Test *facial sensation* in the three divisions of the nerve, comparing each side with the other (Figure 11.17). Test first with the sharp end of a new neurological pin for pain sensation (never use an old pin in these days of hepatitis B, HIV etc).[10] The pin is applied lightly to the skin and the patient is asked whether it feels sharp or dull. Some examiners ask patients to shut their eyes. Loss of pain sensation will result in the pinprick feeling dull. An area of dull sensation should be mapped by testing pinprick sensation progressively: testing should go *from the dull to the sharp area.* Test also above the forehead progressively back over the top of the head. If the ophthalmic division is affected sensation will return when the C2 dermatome is reached (Figure 11.16). It is important to exercise caution: too sharp a pin will leave a little trail of bloody spots, which is embarrassing. Temperature is not tested routinely unless syringobulbia is suspected, as temperature loss usually accompanies loss of pain sensation.

Figure 11.17 Facial sensation V, maxillary division: 'Does this feel sharp or blunt?'—test all three divisions on each side

The patient keeps the eyes closed and a new piece of cottonwool is used to test light touch in the same way. The patient should be instructed to say 'yes' each time the touch of the cottonwool is felt (do *not* stroke the skin). Proprioception loss is not routinely tested on the face (and indeed it would be rather a difficult thing to do!).

Now examine the *motor division* of the nerve. Begin by inspecting for wasting of the temporal and masseter muscles. Ask the patient then to clench the teeth and palpate for contraction of the masseter above the mandible (Figure 11.18). The strength of these muscles can be tested by asking the patient to bite forcefully onto a wooden tongue depressor with the molar teeth. The depth of the teeth marks on each side give an indication of the relative strengths of the muscles. The examiner can attempt to withdraw the tongue depressor as the patient bites it. A bite of normal strength will prevent this. Then get the patient to open the mouth (pterygoid muscles) and hold it open while the examiner attempts to force it shut. A unilateral lesion of the motor division causes the jaw to deviate towards the weak (affected) side.

Figure 11.18 Cranial nerve V (motor): 'Clench your jaw'—feel the masseter muscles

Test the *jaw jerk* or *masseter reflex*. The patient lets the mouth fall open slightly and the examiner's finger is placed on the tip of the jaw and

tapped lightly with a tendon hammer (Figure 11.19). Normally there is a slight closure of the mouth or no reaction at all. In an upper motor neurone lesion above the pons the jaw jerk is greatly exaggerated. This is commonly seen in pseudobulbar palsy).[s]

Figure 11.19 Cranial nerve V: the jaw jerk

Causes of a fifth nerve palsy

Central (pons, medulla and upper cervical cord) causes include a vascular lesion, tumour or syringobulbia.

Peripheral (middle fossa) causes include an aneurysm, tumour (secondary or primary) or chronic meningitis.

Trigeminal ganglion (petrous temporal bone) causes include a trigeminal neuroma, meningioma, or fracture of the middle fossa.

Cavernous sinus causes involve the ophthalmic division only and are usually associated with third, fourth and sixth nerve palsies. They include aneurysm, tumour or thrombosis.

Remember, if there is total loss of sensation in all three divisions of the nerve, this suggests that the level of the lesion is at the ganglion or the sensory root—for example, an acoustic neuroma (Figure 11.20). If there is total sensory loss in one division only, this suggests a postganglionic lesion. The ophthalmic division is most commonly affected because it runs in the cavernous sinus and through the orbital fissure, where it is vulnerable to a number of different insults.

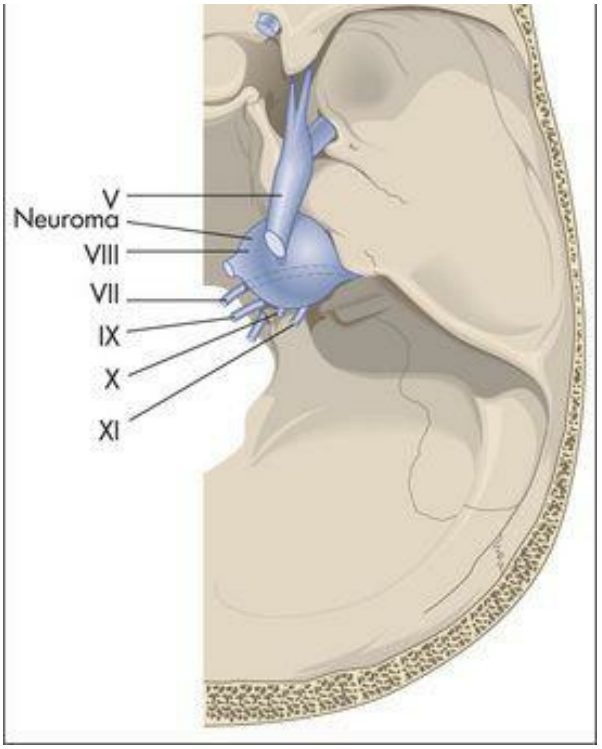


Figure 11.20 Cerebellopontine angle tumour
A neuroma arising from the acoustic (VIII) nerve compresses adjacent structures, including the trigeminal (V) and facial (VII) nerves and the brainstem and cerebellum (removed to permit the cranial nerves to be seen).

Adapted from Simon RP, Aminoff MJ, Greenberg DA, Clinical neurology 1989. Appleton & Lange, 1989.

If there is dissociated sensory loss (loss of pain, but preservation of touch sensation) this suggests a brainstem or upper cord lesion, such as syringobulbia, foramen magnum tumour, or infarction in the territory of the posterior inferior cerebellar artery. If touch sensation is lost but pain sensation is preserved, this is usually due to an abnormality of the pontine nuclei, such as a vascular lesion or tumour. Motor loss can also be central or peripheral.

Irritative motor changes

Convulsive seizures that involve the precentral gyrus can include clenching of the jaw and biting of the tongue. Parkinson's disease and essential tremor can cause a rhythmic tremor of the lips or jaw. *Trismus* is a forceful clenching of the jaw that can occur in tetanus and encephalitis. The patient may be unable to open the mouth. Repetitive chewing and yawning movements can occur as an effect of antipsychotic drugs (*tardive orofacial dyskinesias*).

The seventh (facial) nerve

Examination anatomy

The seventh nerve nucleus lies in the pons next to the sixth cranial nerve nucleus (Figure 11.14). The nerve (Figure 11.21) leaves the pons with the eighth nerve through the cerebellopontine angle. After entering the facial canal it enlarges to become the geniculate ganglion. The branch that supplies the stapedius muscle is given off from within the facial canal. The chorda tympani (containing taste fibres from the anterior two-thirds of the tongue) joins the nerve in the facial canal. The seventh nerve leaves the skull via the stylomastoid foramen. It then passes through the middle of the parotid gland and supplies the muscles of facial expression. The frontalis muscle receives upper motor innervation bilaterally, the other muscles receive innervation from the contralateral cortex.

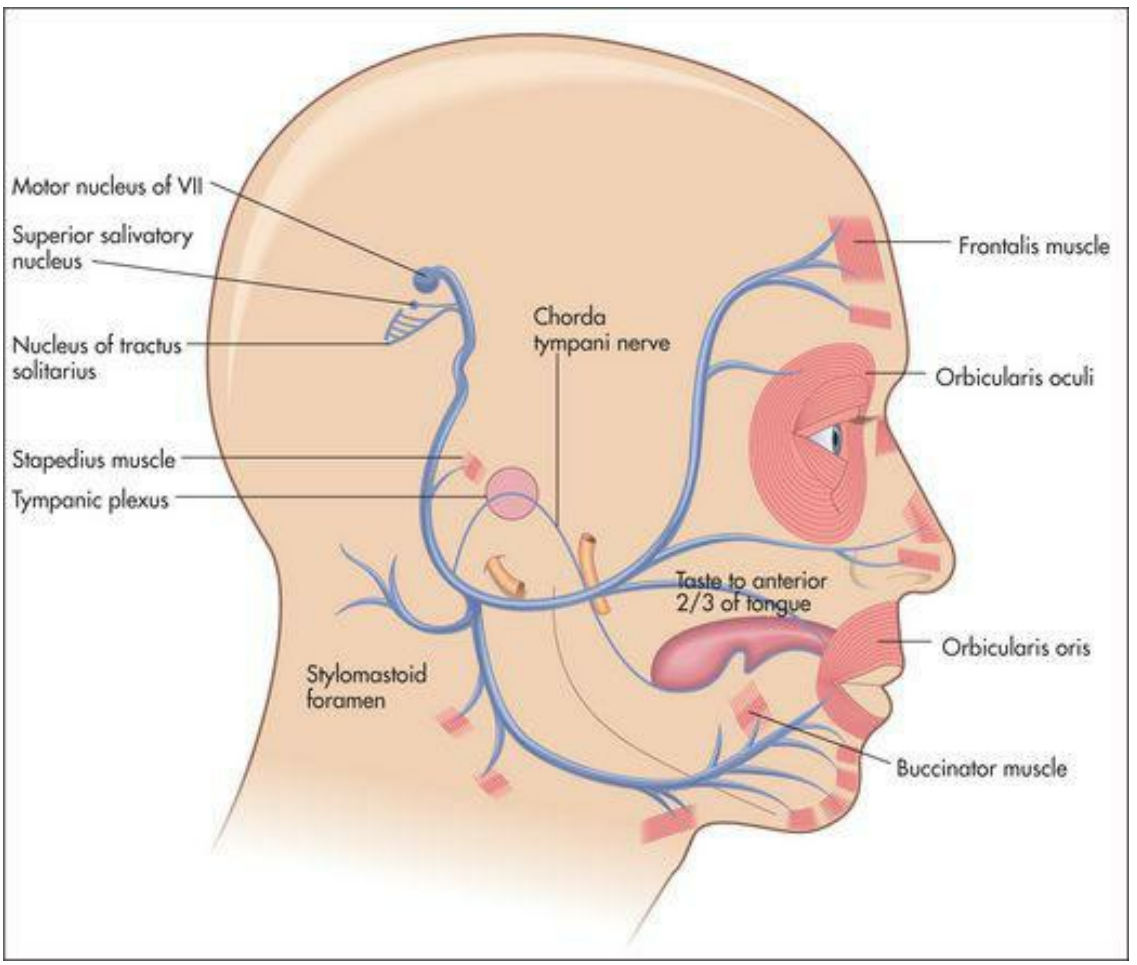


Figure 11.21 The facial nerve (cranial nerve VII)
Note: The branches of the facial nerve: 'Two zebras bit my car'—***t****emporal*, ***z****ygotic*, ***b****uccal*, ***m****andibular*, ***c****ervical*.

History

The patient may have noticed the onset of difficulty with speaking and keeping liquids in the mouth or may have noticed facial asymmetry in the mirror. He or she may be aware of dryness of the eyes (decreased lacrimation) or the mouth (decreased salivary production). Paralysis of the stapedius muscle can cause *hyperacusis* or intolerance of loud or high-pitched sounds. Normal contraction of the stapedius muscle occurs in response to loud noises such as popular music and dampens movement of the ossicles.

Examination

Inspect for *facial asymmetry*, as a seventh nerve palsy can cause unilateral drooping of the corner of the mouth, and smoothing of the wrinkled forehead and the nasolabial fold (Figure 11.22). However, with bilateral facial nerve palsies symmetry can be maintained.

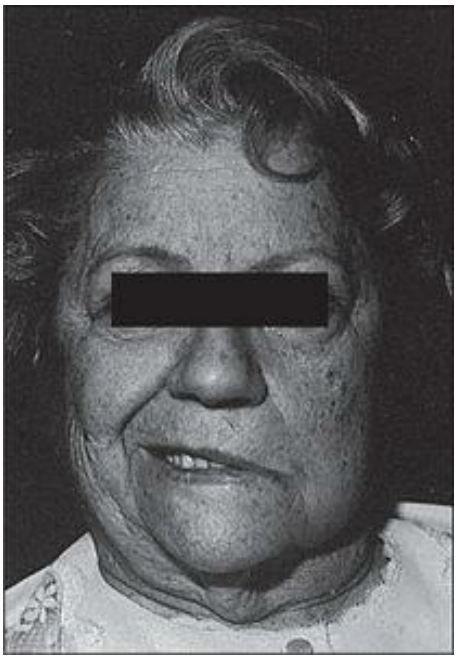


Figure 11.22 Left upper motor neurone facial weakness, showing drooping of the corner of the mouth, flattened nasolabial fold, and sparing of the forehead; the lesion is in the right side of the brain

Test the *muscle power*. Ask the patient to look up so as to wrinkle the forehead (Figure 11.23). Look for loss of wrinkling and feel the muscle