- Attributing the early headache of a space-occupying lesion to tension or hypertension

Seven masquerades checklist

Of the masquerades, depression and drugs are important causes of headache. Cervical dysfunction is certainly an important cause and tends to be ignored by some doctors. Australian figures are misleading because many of these patients tend to gravitate to alternative health professionals.

A UK study placed headache from cervical spondylosis on almost equal terms with migraine.[6]

The explanation for referral of pain from disorders of the upper cervical spine to the head and eye is that some afferent fibres from the upper three cervical nerve roots converge on cells in the posterior horn of the spinal cord (which can also be excited by trigeminal afferent fibres), thus conveying to the patient the impression of head pain through this shared pathway (see FIG. 45.1).

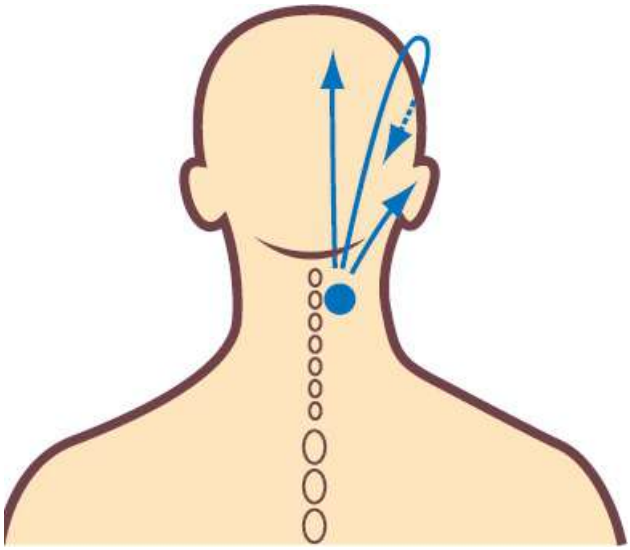

FIGURE 45.1 Typical headache referral patterns for dysfunction of the upper cervical spinal segments

Question the possibility of the effect of any drug the patient is taking: important examples are alcohol, analgesics (rebound), antihypertensives (several), caffeine, COCP, corticosteroids, NSAIDs (especially indomethacin), vasodilators (e.g. CCBs), nitrates, PDE inhibitors (e.g. sildenafil). Anaemia can cause headache, usually if the haemoglobin level falls below 100 g/L.[7] Hypo- and hyperthyroidism may also cause headache, and in diabetes hypoglycaemia is often responsible.

Psychogenic considerations

Headache, like tiredness, is one of those symptoms that may reflect a ‘hidden agenda’. Of course, the patient may be depressed (overt or masked) or may have a true anxiety state. The most characteristic feature of psychogenic headache is that the headache is present virtually every minute of the day for weeks or months on end. However, it is common for patients to not describe themselves as anxious, depressed or unduly stressed. For this reason a detailed history is important to identify lifestyle factors and historical events that can be associated with headache.

Some are fearful of their headache lest it represents a cerebral tumour, stroke or hypertension; they need appropriate reassurance.

Conversion reactions and other aspects of compensation rewards, especially following an accident (e.g. rear-end collision), may make the symptom of headache difficult to manage. Headache, like backache, is one of the prime symptoms perpetuated or exaggerated for secondary gain.

Severe headaches, especially simulated migraine, are common ‘tickets of entry’ for those seeking narcotics from empathic practitioners. Such patients require very skilled management.

Timelines for causes of headache/facial pain

Acute severe headache, e.g.:

- subarachnoid haemorrhage
- benign sex or exertional headache
- migraine/cluster headache
- reversible cerebral vasoconstriction

Subacute headache (recent onset, increasing):

- expanding intracranial lesion
- temporal arteritis

Recurrent episodes, e.g.:

- migraine/cluster headache
- benign sex or exertional headache
- neuralgias, e.g. trigeminal

Chronic headache, e.g.:

- tension-type headache

- chronic migraine/rebound headache
- cervicogenic/post-traumatic
- atypical facial pain

Diurnal patterns of pain

Plotting the fluctuation of headache during the day provides vital clues to the diagnosis (see FIG. 45.2). The person who wakes up with headache could have vascular headache (migraine), cervical spondylosis, depressive illness, hypertension or a space-occupying lesion. It is usual for migraine to last hours, not days, which is more characteristic of tension headache. The pain of frontal sinusitis follows a typical pattern, namely onset around 9 am, building to a maximum by about 1 pm, and then subsiding over the next few hours. In the absence of respiratory symptoms it is likely to be misdiagnosed as tension headache. The pain from combination headache tends to follow a most constant pattern throughout the day and does not usually interrupt sleep.

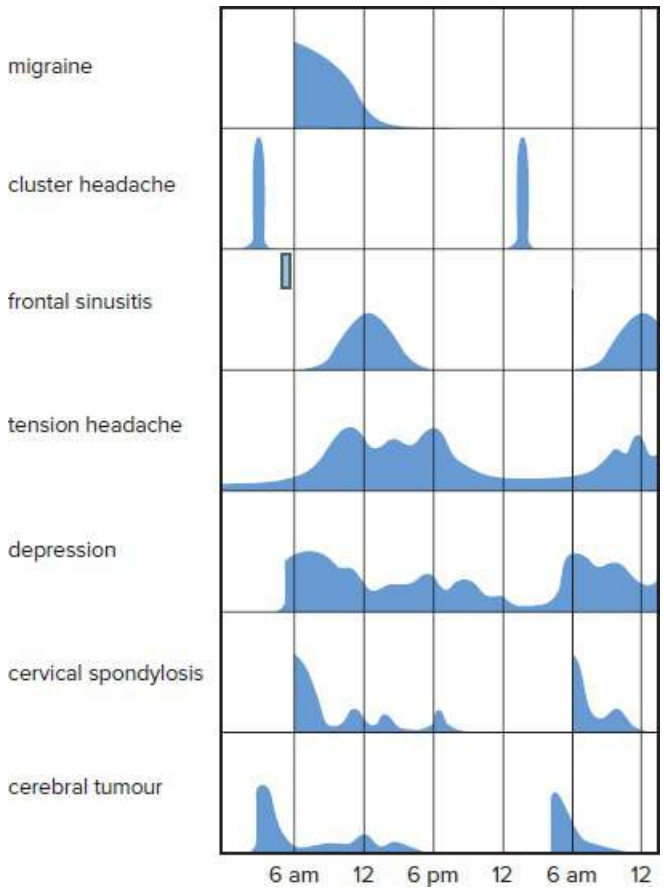

FIGURE 45.2 Typical diurnal patterns of various causes of headache; the relative intensity of pain is plotted on the vertical axis

The clinical approach

History

A full description of the pain including a pain analysis should be obtained using one of the mnemonics (see CHAPTER 82) with an emphasis on relieving factors and associations.

It is useful to get the patient to plot on a prepared grid the relative intensity of the pain and the times of day (and night) that the pain is present. The history, especially of the tempo of the condition, should help diagnose headaches secondary to specific pathology.

Key questions

- Can you describe your headaches?
- How often do you get them?
- Can you point to exactly where in the head you get them?
- Do you have any pain in the back of your head or neck?
- What time of the day do you get the pain?
- Do you notice any other symptoms when you have the headache?
- Do you feel nauseated and do you vomit?
- Do you experience any unusual sensations in your eyes, such as flashing lights?
- Do you get dizzy, weak or have any strange sensations?
- Does light hurt your eyes?
- Do you get any blurred vision?
- Do you notice watering or redness of one or both of your eyes?
- Do you get pain or tenderness on combing your hair?
- Are you under a lot of stress or tension?
- Does your nose run when you get the headache?
- What tablets do you take?
- Do you get a high temperature, sweats or shivers?

- Have you had a heavy cold recently?
- Have you ever had trouble with your sinuses?
- Have you had a knock on your head recently?
- What do you think causes the headaches?

Differences between the clinical features of migraine and tension headache are presented in TABLE 45.2 . 'Red flag' indicators of a serious cause of headache are outlined in the box.

Table 45.2 A comparison of typical clinical features of migraine and tension headache[5]

	Migraine	Tension headache
Family history	✓	
Onset before age 20	✓	
Prodromata	✓	
Bilateral		✓
Unilateral	✓	
Throbbing	✓	
Constant		✓
Less than one per week	✓	
Continuous daily		✓
Lasts less than 24 hours	✓	
Vomiting	✓	
Aggravated by the pill	✓	
Aggravated by alcohol	✓	
Relieved by alcohol		✓

Examination

For the physical examination it is appropriate to use the basic tools of trade, namely the thermometer, sphygmomanometer, pen torch, and diagnostic set, including the ophthalmoscope and the stethoscope. Inspect the head, temporal arteries and eyes. Areas to palpate include the temporal arteries, the facial and neck muscles, the cervical spine and sinuses, the teeth and

temporomandibular joints. Search especially for signs of meningeal irritation and papilloedema.

Red flag indicators for headache[3]

- Sudden onset, especially if no previous history
- Severe and debilitating pain
- Progressive
- Fever
- Vomiting
- Disturbed consciousness/confusion, drowsiness
- Personality change
- Worse with bending, posture, coughing or sneezing
- Maximum in morning
- Wakes patient at night
- Neurological and visual symptoms/signs
- Seizure
- Young, obese female with deteriorating vision (intracranial HTn)
- 'New' in elderly, especially >50 years
- Post-head injury
- Non-migraine headache in pregnancy or postpartum

A mental state examination is mandatory and includes looking for altered consciousness or cognition and assessment of mood, anxiety–tension–depression and any mental changes. Neurological examination includes assessment of visual fields and acuity, reactions of the pupils and eye movements in addition to sensation and motor power in the face and limbs and reflexes, including the plantar response. 'Red flag' pointers from physical examination are given in the box.

Red flag pointers: from physical examination

- Altered consciousness or cognition
- Meningism
- Abnormal vital signs: BP, temperature, respiration
- Focal neurological signs, including pupils, fundi, eye movement
- Tender, poorly pulsatile temporal arteries
- Papilloedema

Special signs

- *Upper cervical pain sign.* Palpate over the C2 and C3 areas of the cervical spine, especially two finger breadths out from the spinous process of C2. If this is very tender and even provokes the headache it indicates headache of cervical origin.
- *Ewing sign for frontal sinusitis.* Press your finger gently upwards and inwards against the orbital roof medial to the supra-orbital nerve. Pain on pressure is a positive finding and indicates frontal sinusitis.
- *The invisible pillow sign.* The patient lies on the examination table with head on a pillow. The examiner then supports the head with his or her hands as the pillow is removed. The patient is instructed to relax the neck muscles and the examiner removes the supporting hands. A positive test indicating tension from contracting neck muscles is when the patient's head does not readily change position. This is uncommon.

Investigations

Investigations can be selected from:

- haemoglobin: ?anaemia
- WCC: leucocytosis with bacterial infection
- ESR/CRP: ?temporal arteritis
- radiography:

    chest X-ray, if suspected intracerebral malignancy

    cervical spine

    sinus X-ray, if suspected sinusitis

    CT scan: detection of brain tumour (most effective), cerebrovascular accidents (valuable),

SAH, urgent non-enhanced cerebral CT scan for acute thunderclap headache[8]

radioisotope scan (technetium-99m) to localise specific tumours and haematoma

MRI: very effective for intracerebral pathology but expensive; produces better definition of intracerebral structures than CT scanning but not as sensitive for detecting bleeding; detects intracranial vasculitis in temporal arteries

lumbar puncture: diagnosis of meningitis, suspected SAH (only if CT scan normal)

Note: Dangerous if raised intracranial pressure.

Headache in children

Respiratory infections and febrile illnesses are common causes of headache in children but there are other causes that reflect the common causes in adults. Many childhood headaches are isolated but are chronic in a significant number. Migraine is relatively common before adolescence, while tension or muscle contraction headache is more common after adolescence.

Consider often overlooked causes such as hair traction, eye strain (measure and record vision) and hypoglycaemia. Children who have long periods without regular eating are prone to headache including exacerbations of migraine. They should not skip breakfast.[9]

Young children rarely experience sinus headache and this should not really be considered until the sinuses develop, around 5 years for the frontal sinuses.

Migraine affects 1% of children at age 7 and 5% or more at age 15, with girls developing it at a higher rate[2] with increasing age. There is a strong family history. The prognosis is good as the majority will have no migraines in the long term. The type is mainly common migraine with symptoms such as malaise or nausea: classic migraine with the typical aura is not a feature of childhood migraine. The rather dramatic vertebrobasilar migraine is frequent in adolescent girls and hemiplegia occurs in infants and children, especially with their first migraine attack.[10] Vomiting is not necessarily an associated symptom in children.

The possibility of cerebral space-occupying lesions requires due consideration, especially if the headaches are progressive. These are present typically in the morning and are associated with symptoms such as vomiting, dizziness, diplopia, ataxia, personality changes and deterioration of school performance. Symptoms that indicate a cerebral tumour or other serious problem are outlined in TABLE 45.3 .

Table 45.3 Pointers to serious causes of headache in children

Headache features

Persistent or recurrent
Present first thing in morning
Wakes child at night
No past history
No family history
Associated poor health
Associated neurological symptoms
Unilateral localisation

Source: Wright[2]

Neonates and children aged 6–12 months are at the greatest risk from meningitis and it is important to keep this in mind.

Management of the non-serious causes of headache includes reassurance (especially of parents), discouragement of excessive emphasis on the symptom and simple medications, such as paracetamol for the younger child and aspirin for the adolescent. Patients with undiagnosed and/or problematic headache should be referred.

Pharmacological treatment in children[10]

Tension headache and migraine:

paracetamol 20 mg/kg (o) statim then 15 mg/kg 4–6 hourly up to 90 mg/kg/day (max. 4 g daily)

or

ibuprofen 5–10 mg/kg (o) statim up to 40 mg/kg/day (not for children <6 months)

Headache in the elderly

A recent onset of headache in the elderly has to be treated with caution because it could herald a serious problem, such as a space-occupying lesion (e.g. neoplasm, subdural haematoma), TIA, trigeminal neuralgia or vertebrobasilar insufficiency. Cervical spondylosis is age-related and may be an important factor in the ageing patient. Age-related headaches are summarised in TABLE 45.4 .

Table 45.4 Age-related causes of headache

Children	Intercurrent infections

	Psychogenic Migraine Meningitis Post-traumatic
Adults, including middle age	Migraine Cluster headache Tension Cervical dysfunction Subarachnoid haemorrhage Combination
Elderly	Cervical dysfunction Cerebral tumour Temporal arteritis Neuralgias Paget disease Glaucoma Cervical spondylosis Subdural haemorrhage

Late-life migraine can be mistaken for cerebrovascular disease, especially in the presence of preceding neurological symptoms. It is the sequence of the visual and sensory symptoms with the spread from face to tongue to hand over some minutes, with clearing in one area as it appears that helps distinguish migraine from transient ischaemic attacks (TIAs). Although some patients experience headache with TIAs, it is not a distinguishing feature. Vomiting is suggestive of migraine rather than cerebrovascular disease.[11]

General headache disorders

Tension-type headache

Tension or muscle contraction headaches are typically a symmetrical (bilateral) tightness. They tend to last for hours and recur each day. They are often associated with cervical dysfunction and stress or tension, although the patient usually does not realise the headaches are associated with tension until it is pointed out. Seventy-five per cent of patients are females.[3]

IHS criteria for tension-type headache[12]

The International Headache Society (IHS3) criteria for episodic tension-type

headaches involve the following:

A The patient should have had at least 10 of these headaches.
B The headaches last from 30 minutes to 7 days.
C The headaches must have at least two of the following four:

1. non-pulsating quality
2. mild or moderate intensity
3. bilateral location
4. no aggravation with routine physical activity

D The headaches must have both of the following:

1. no nausea or vomiting
2. photophobia and phonophobia are absent, or one but not the other is present

E Not attributable to another disorder.

IHS = International Headache Society

Clinical features (tension headache)

Site:	frontal, over forehead and temples (see FIG. 45.3)
Radiation:	occiput
Quality:	dull ache, like a ‘tight pressure feeling’, ‘heavy weight on top of head’, ‘tight band around head’; may be tightness or vice-like feeling rather than pain
Frequency:	almost daily
Duration:	hours (can last days)
Onset:	after rising, gets worse during day
Aggravating factors:	stress, overwork with skipping meals
Relieving factors:	alcohol
Associated features:	lightheadedness, fatigue, neck ache or stiffness (occiput to shoulders), perfectionist personality, anxiety/depression
Physical examination:	muscle tension (e.g. frowning), scalp often tender to touch, ‘invisible pillow’ sign may be positive

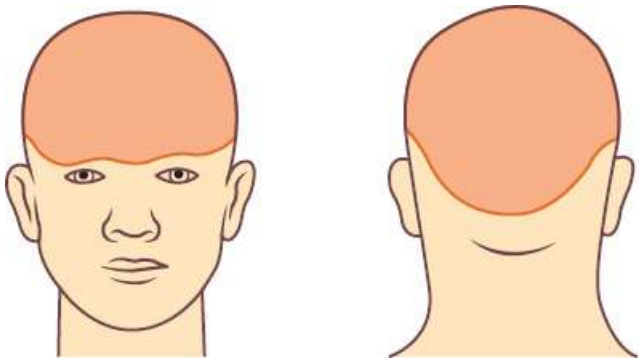

FIGURE 45.3 Typical distribution of pain in tension-type headache

Management[10]

- Careful patient education: explain that the scalp muscles get tight like the calf muscles when climbing up stairs.
- Counselling and relevant advice; CBT is as effective as any drug in all ages

  Learn to relax your mind and body.

  During an attack, relax by lying down in a hot bath and practise meditation.

  Be less of a perfectionist: do not be a slave to the clock.

  Don’t bottle things up, stop feeling guilty, approve of yourself, express yourself and your anger.
- Advise stress reduction, relaxation therapy and yoga or meditation classes. Page 544
- Advise and demonstrate massage of the affected area with a soothing analgesic rub.
- Medication—use mild non-opioid analgesics such as soluble aspirin, ibuprofen or paracetamol. Discourage stronger analgesics. Avoid tranquillisers and antidepressants if possible, but consider these drugs if symptoms warrant medication (e.g. amitriptyline 10 mg (o) nocte increasing to 75 mg or nortriptyline if necessary).

Special notes:

- The general aim is to direct patients to modify their lifestyle and avoid tranquillisers and analgesics.
- It is unusual to be awoken from sleep.

- Beware of depression.
- Consider muscle energy therapy and/or mobilisation of the neck followed by exercises if there is evidence of cervical dysfunction.
- Recommend a meditation program.

Migraine

Migraine, or the ‘sick headache’, is derived from the Greek word meaning ‘pain involving half the head’. It affects at least 1 person in 10, is more common in females (18% of women, 6% men) and peaks between 20 and 50 years. There are various types of migraine (see TABLE 45.5), with classic migraine (headache, vomiting and aura) and common migraine (without the aura) being the best known. The most common trigger factor is stress.[3] Also consider chocolate, cheese, alcohol, hangover and exercise.

Table 45.5 Types of vascular headache

Common migraine (aura is vague or absent)
Classic migraine
Complicated migraine
Aura without migraine (acephalgic migraine)
Unusual forms of migraine:
• hemiplegic
• basilar
• retinal
• migrainous (vestibular) vertigo
• migrainous stupor
• ophthalmoplegic
• migraine equivalents
• status migrainosus
Cluster headache
Chronic paroxysmal hemicrania
Menstrual migraine
Lower half headache
Benign exertional/sex headache (beware of SAH)
Miscellaneous (e.g. icepick pains, ‘ice-cream’ headache)

Source: Day[13]

DxT headache + vomiting + visual aura → migraine with aura (classic)

IHS3 criteria for common migraine[12]

The IHS3 criteria for migraine without aura involve the following checklist.

A The patient should have had at least five attacks fulfilling criteria B and D.

B The headaches last 4–72 hours.

C The headache must have at least two of the following:

1. unilateral location
2. pulsing quality
3. moderate or severe intensity, inhibiting or prohibiting daily activities
4. headache worsened by routine physical activity

D The headache must have at least two of the following:

1. nausea and/or vomiting
2. photophobia or phonophobia

E Not attributable to another disorder.

IHS3 criteria for migraine with typical aura (classic)[12]

A At least two attacks fulfilling criteria B and C.

B One or more of the following fully reversible aura symptoms: visual, sensory, speech and/or language, motor, brainstem, retinal.

C At least two of:

1. at least one aura symptom spreads gradually over at least 5 minutes
2. each aura symptom lasts 5–60 minutes
3. at least one symptom is unilateral
4. headache follows aura within 60 minutes

D Not attributable to another disorder including TIA.

Management

Patient education: provide explanation and reassurance, especially if bizarre visual and neurological symptoms are present. Patients should be reassured about the benign nature of their migraine. For each migraine sufferer, an individual treatment plan including a migraine action plan should be devised.

Counselling and advice

- Classical migraines with visual aura are a contraindication to prescribing the combined oral contraceptive.[14] Discuss other options.
- Tailor the advice to the individual patient.

- Avoid known trigger factors, especially tension, fatigue, hunger and constant physical and mental stress.
- Advise keeping a diary of foodstuffs or drinks that can be identified as trigger factors. Consider a low amine diet: eliminate chocolate, cheese, red wine, coffee, walnuts, tuna, Vegemite, spinach and liver.
- Practise a healthy lifestyle, relaxation programs, meditation techniques and biofeedback training.

Clinical features (classic migraine)

Site:	temporofrontal region (unilateral) (see FIG. 45.4); can be bilateral
Radiation:	retro-orbital and occipital
Quality:	intense and throbbing
Frequency:	1 or 2 per month
Duration:	4–72 hours (average 6–8 hours)
Onset:	paroxysmal, often wakes with it
Offset:	spontaneous (often after sleep)
Precipitating factors:	tension and stress (commonest); others in TABLE 45.6
Aggravating factors:	tension, activity
Relieving factors:	sleep, vomiting
Associated factors:	nausea, vomiting (90%), irritability, aura visual 25% (scintillation, scotoma, hemianopia, fortification)

	sensory (unilateral paraesthesia)
Other pointers:	abdominal pain in childhood; family history of migraine, asthma and eczema

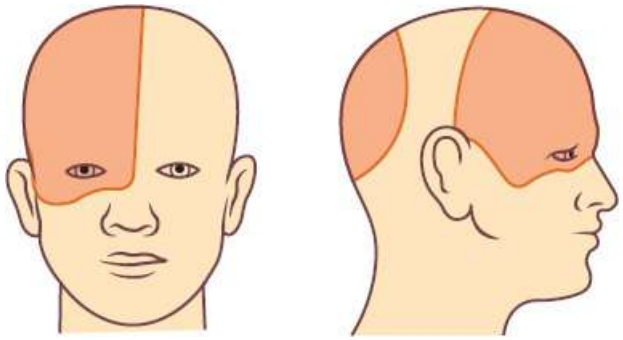

FIGURE 45.4 Typical distribution of pain in migraine (right side)

Table 45.6 Migrainous trigger factors

Exogenous
Foodstuffs—chocolate, oranges, tomatoes, citrus fruits, cheeses, gluten sensitivity (possible)
Alcohol—especially red wine
Drugs—vasodilators, oestrogens, monosodium glutamate, nitrites (‘hot dog’ headache), indomethacin, OCP, codeine
Glare or bright light* (32%)
Emotional stress* (63%)
Head trauma (often minor) (e.g. jarring—‘footballer’s migraine’)
Allergen
Climatic change
Excessive noise
Strong perfume
Endogenous
Tiredness, physical exhaustion, oversleeping
Lack of sleep
Stress, relaxation after stress—‘weekend migraine’
Exercise/physical stress
Hormonal changes

puberty
menstruation*
climacteric
pregnancy
Hunger
Familial tendency
?Personality factors

*most common

Treatment of the acute attack

- Commence treatment at earliest impending sign.
- Mild headaches may require no more than conventional treatment with ‘two aspirin (or paracetamol) and a good lie down in a quiet dark room’.[5,10]
- Rest in a quiet, darkened, cool room.
- Place cold packs on the forehead or neck.
- Drink copious water but avoid drinking coffee, tea or orange juice.
- Avoid moving around too much.
- Do not read or watch television.
- For patients who find relief from simply ‘sleeping off’ an attack, consider prescribing temazepam 10 mg or diazepam 10 mg in addition to the following measures.[3]
- For moderate attacks use oral ergotamine or sumatriptan and for severe attacks use injection therapy.
- Avoid pethidine and other opioids.

Medication (if necessary)[10,15,16]

First-line medication acute migraine:

- aspirin or paracetamol + anti-emetic: e.g. soluble aspirin 600–900 mg (o) *and* metoclopramide 10 mg (o)
- paracetamol or ibuprofen (for children)
- consider NSAIDs (e.g. ibuprofen, naproxen, diclofenac rapid)

- avoid opioids

If nausea and vomiting is a feature:

- metoclopramide 10–20 mg oral, IM or IV

  or

- prochlorperazine 12.5 mg IM or 12.5–25 mg rectally
- consider nasal sumatriptan

Alternatives (especially if above ineffective or severe):

Choose a triptan preparation—best at start of attack.

Triptans (effective in about 2 out of 3 patients)

- sumatriptan (a serotonin receptor agonist)[10] 50–100 mg (o) at the time of prodrome, repeat in 2 hours if necessary to maximum dose 300 mg/24 hours

  or

  nasal spray 10–20 mg per nostril (up to 40 mg/24 hours)

  or

  6 mg, SC injection, repeat in 1 or more hours to maximum dose 12 mg/24 hours

  or

- zolmitriptan 2.5–5 mg (o), repeat in 2 hours if necessary (max. 10 mg/24 hours)

  or

- naratriptan 2.5 mg (o), repeat in 4 hours (max. 5 mg/24 hours)

  or

- rizatriptan 10 mg wafer, repeat in >2 hours (max. 30 mg/24 hours)

  or

- eletriptan 40–80 mg (o) up to 160 mg/24 hours

Avoid triptans in patients with coronary artery disease, Prinzmetal angina, uncontrolled hypertension or during pregnancy. Do not use with ergotamine simultaneously and cease if chest pain develops, albeit transient in a young patient. Use with caution in patients taking SSRIs, MAOIs and lithium.

Treatment of the severe attack

(If other preparations ineffective.)

Practice tip for early migraine attack

A triptan + a NSAID, e.g. naproxen or ibuprofen[17]

Caution: Consider the possibility of underlying cerebral vascular malformation, SAH or opioid addiction.

- If at home:[13]

  sumatriptan 6 mg (SC)

- If in surgery or emergency room:

  metoclopramide 10 mg (IV) slowly over 2 minutes + oral analgesics

  or

  metoclopramide 10 mg (IV) + dihydroergotamine

  0.5 mg IV slowly

  or

  sumatriptan 6 mg (SC)

  or

  chlorpromazine 0.1 mg/kg IV infusion over 30 mins

Caution: Do not use ergotamine preparations if sumatriptan used in previous 6 hours, and do not use sumatriptan if ergotamine preparations used in previous 24 hours.

Practice tips for severe classic migraine:

- IV metoclopramide + 1 litre N saline IV in 30 minutes + oral aspirin or paracetamol
- Continue high fluid intake

Prophylaxis

Non-drug self-management with avoidance of any known trigger factors is the key. A 2016 Cochrane review of acupuncture found that a course of at least 6 treatments reduced the frequency of episodes compared to no acupuncture and placebo 'fake' acupuncture.[18]

Consider prophylactic therapy for frequent attacks that cause disruption to the patient's lifestyle and well-being, a rule of thumb being two or more severe migraine attacks per month; certainly consider it for weekly attacks and a poor response to therapy for the acute attack. Do not give ergotamine.[11]

The wide variety of drugs used reflects the lack of marked superiority of any one agent.[10] Weak-to-moderate evidence supports many of them (more so near the top of the list than the bottom) but comparisons are lacking. Individuals may need to trial a number of medications (one at a time, gradually increasing dose, for 8–12 weeks) before finding success.

- beta blockers—propranolol 20 mg (o) bd or tds (max. 160 mg/day)* (metoprolol, atenolol less evidence)
- tricyclic antidepressants—amitriptyline 10 mg (o) nocte (max. 75 mg)*; nortriptyline
- sodium valproate 200 mg (o) nocte (max. 500 mg bd)*
- pizotifen 0.5–2.0 mg at night (poorly tolerated)
- candesartan 4 mg (o) daily (max. 32 mg)
- calcium-channel blockers—verapamil SR 90 mg (o) daily (max. 240 mg); nifedipine
- topiramate
- cyproheptadine (ideal for children—seek specialist advice)
- clonidine
- NSAIDs—naproxen, indomethacin, ibuprofen

- MAOIs—phenelzine, moclobemide
- sumatriptan
- gabapentin
- botulinum toxin into muscles of the face, scalp or neck (only slightly more effective than placebo)

*first line

For consideration:

- melatonin

- anti-CGRP monoclonal antibodies

Menstrual migraine[10]

Naproxen 550 mg (o) bd, 48 hours before expected attack for 4–10 days, oestradiol gel 1.5 mg transdermally, once daily for 7 days or mefenamic acid.

Guidelines[10,15]

Select the initial drug according to the patient's medical profile:

- if low or normal weight—pizotifen
- if hypertensive—a beta blocker
- if depressed or anxious—amitriptyline
- if tension—a beta blocker
- if cervical spondylosis—naproxen
- food-sensitive migraine—pizotifen
- menstrual migraine—naproxen or mefenamic acid or ibuprofen or oestradiol transdermal gel

Commonly prescribed first-line drugs are propranolol or pizotifen:[11]

propranolol 40 mg (o) bd or tds (at first)

increasing to 320 mg daily (if necessary)

pizotifen 0.5–1 mg (o) nocte (at first) increasing to 3 mg a day (if necessary)

Each drug should be tried for 2 months before it is judged to be ineffective. Amitriptyline 50 mg nocte can be added to propranolol, pizotifen (beware of weight gain) or methysergide and may convert a relatively poor response to very good control.[3]

Cluster headache

Cluster headache is also known as migrainous neuralgia. It occurs in paroxysmal clusters of unilateral headache that typically occur nightly, usually in the early hours of the morning, although patients may have headaches that occur at other times. A hallmark is the pronounced cyclical nature of the attacks and at least five attacks. It occurs typically in males (6:1 ratio) and is rare in childhood. There are no visual disturbances or vomiting.

DxT retro-orbital headache + rhinorrhoea + lacrimation → cluster headache

Management[10,16]

Acute attack (brief treatment seldom effective):

- consider 100% oxygen 15 L/min for 15 min (usually good response)
- sumatriptan 6 mg SC injection (or 20 mg intranasal), rizatriptan 10 mg (o) or zolmitriptan 2.5 mg (o)

  or

- consider local anaesthetic—greater occipital nerve block

Avoid alcohol during cluster.

Prophylaxis (once a cluster starts)

Consider the following:

- for control of attack, naratriptan 2.5 mg (o) bd for 1 week
- methysergide 1 mg (o) once daily up to 3 mg bd
- prednisolone 50 mg/day for 10 days then taper over 3 weeks (as a bridging treatment)
- lithium 250 mg (o) bd
- verapamil SR 160 mg (o) daily up to 320 mg
- pizotifen
- indomethacin (helps confirm diagnosis)
- sodium valproate

Note: Some of the above can be used long term for frequent clusters. Prevention of further attacks is the main focus of cluster headache treatment.[5]

Clinical features

Site:	over or about one eye (see FIG. 45.5); always same side
Radiation:	frontal and temporal regions
Quality:	severe
Frequency:	one every other day and 8 per day for more than half the time
Duration:	15–180 minutes (average 30 minutes); the clusters last 4–6 weeks (can last months)

Onset:	suddenly during night (usually), same time about 2–3 hours after falling asleep; the ‘alarm clock’ headache (e.g. 2–4 am)
Offset:	spontaneous
Aggravating factors:	alcohol (during cluster)
Relieving factors:	drugs
Associated features:	family history; rhinorrhoea and/or congestion, ipsilateral nose; lacrimation; flushing and/or sweating of forehead and cheek; redness of ipsilateral eye; eyelid oedema; miosis and/or ptosis; sensation of fullness in ear; a sense of restlessness or agitation (see FIG. 45.6)

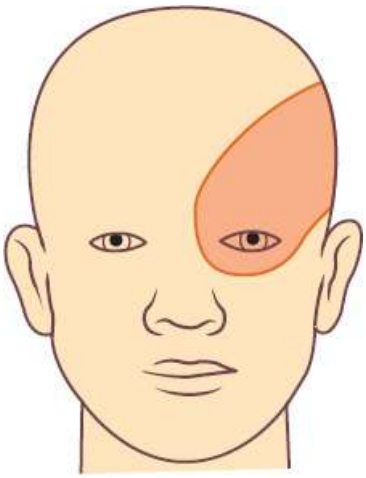

FIGURE 45.5 Typical distribution of pain in cluster headache

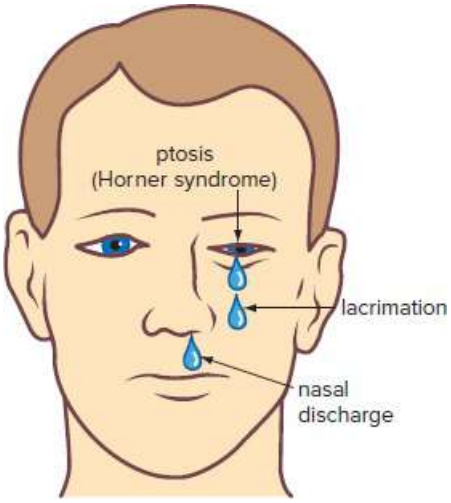

FIGURE 45.6 Features of an attack of cluster headache: ptosis, lacrimation and a discharge from the nostril on the side of pain

Cervicogenic headache

Headache from neck disorders (cervical dysfunction or spondylosis), referred to as cervicogenic, is far more common than realised and is very rewarding to treat by physical therapy, including mobilisation and manipulation and exercises in particular. See CHAPTERS 15 and 51 .

Headache can be caused by abnormalities in any structure innervated by the upper two cervical nerves C2, C3 (usually the C1–2, C2–3 facet joints). Pain from cervical structures can be referred retro-orbitally and over one-half of the head. The headache is often incorrectly diagnosed as migraine but clinical examination of the neck helps differentiation.[19] The neck may be responsible for so-called ‘tension’ headache but clinical differentiation can be more difficult.

Occipital neuralgia, which is uncommon, causes intermittent neuralgia or lancinating pain in the C2 distribution and may radiate to the front orbital area (see FIG. 45.7).

Clinical features

The pain is usually sited in the occipital region with possible radiation to the parietal region, vertex of skull and behind the eye (see FIG. 45.7). It is usually present on waking and settles during the day. There is usually a history of trauma including an MVA or blow to the head. Associated features include stiffness and grating of the neck. On examination there is usually tenderness to palpation over the C1, C2 and/or C3 levels of the cervical spine, especially on the side of the headache.

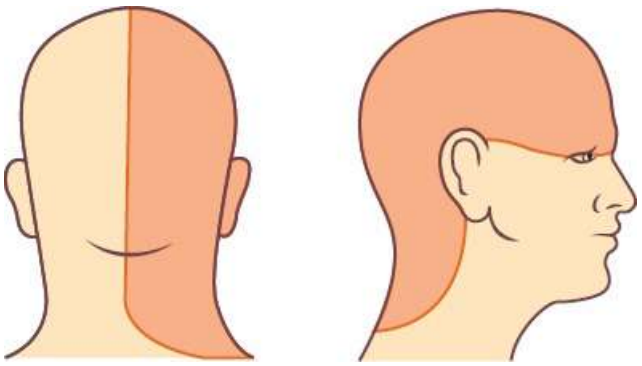

FIGURE 45.7 Typical distribution of pain in cervical dysfunction (right side)

Treatment

- Physiotherapy modalities: hydrotherapy, muscle energy therapy, mobilisation, manipulation (from experts) and neck exercises (very important)

- Supportive neck pillow
- Heat pack, especially for acute pain
- NSAIDs for cervical spondylosis
- For intractable cases consider mobilisation under general anaesthesia, injections of corticosteroids around, or surgical section of, the greater occipital nerve for occipital neuralgia.[19]

Combination headache

Combined (also known as mixed) headaches are common and often diagnosed as psychogenic headache or atypical migraine. They have a combination of various degrees of:

- tension and/or depression
- cervical dysfunction
- vasospasm (migraine)
- drugs: analgesics (rebound), alcohol, nicotine, caffeine, NSAIDs

The headache, which has many of the features of tension headache, is usually described as a heavy deep ache ‘as though my head is ready to burst’. It tends to be constant, being present throughout every waking moment. It tends to last for days (average 3–7) but can last for weeks or months. It is often related to stress and adverse working conditions, and sometimes follows an accident.

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Management

An important strategy is to evaluate each possible component of the headache as a stepwise trial by an elimination process:

- drug evaluation and modification
- cervical dysfunction—physical therapy if indicated
- depression
- tension and stress
- other psychogenic factors (e.g. conversion reaction)
- vasospasm

Treatment includes cognitive therapy, reassurance that the patient does not have a cerebral tumour and lifestyle modification. The most effective medication is amitriptyline or other

antidepressant. Propranolol and the anti-epileptics can be considered.

Temporal arteritis

TA is also known as giant cell arteritis or cranial arteritis. There is usually a persistent unilateral throbbing headache in the temporal region and scalp sensitivity with localised thickening, with or without loss of pulsation of the temporal artery. It is related to polymyalgia rheumatica—20% of sufferers will develop TA. See CHAPTER 21 .

TA is a type of collagen disease causing inflammation of extracranial vessels, especially the superficial temporal artery. It usually presents as a unilateral intermittent headache in a person over 50 years.

TA may also involve the intracranial vessels, especially the ophthalmic artery or posterior ciliary arteries, causing optic atrophy and blindness. Vision is impaired in about one-half of patients at some stage. Once the patient goes blind it is usually irreversible.

Age:	over 50 years (mean age 70 years)
Site:	forehead and temporal region (unilateral) (see FIG. 45.8)
Radiation:	down side of head towards occiput
Quality:	severe burning pain
Frequency:	daily, a constant ache
Duration:	usually constant (getting worse)
Onset:	non-specific, tends to be worse in morning
Offset:	nil
Aggravating factors:	stress and anxiety
Relieving factors:	nil
Associated features:	malaise, vague aches and pains in muscles (especially of neck), weight loss
Other pointers:	intermittent blurred vision
	tenderness on brushing hair
	jaw claudication on eating
	polymyalgia rheumatica
	hypertension
	abnormal emotional behaviour

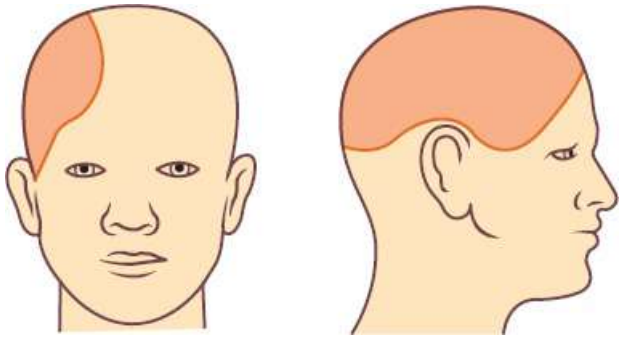

FIGURE 45.8 Typical distribution of pain in temporal arteritis (right side)

Diagnosis

Diagnosis is by biopsy and histological examination of the superficial temporal artery. The ESR is usually markedly elevated but may be normal. The biopsy may be normal as TA has a focal nature. MRI has a high sensitivity and specificity.

Note: Consider it with any ‘new’ headache.

Treatment

TA is very responsive to corticosteroids; start treatment immediately to prevent permanent blindness. Initial medication is prednisolone 40–60 mg orally daily in two divided doses initially for a minimum of 4 weeks. Continue until symptoms resolve. Aspirin 100 mg daily helps prevent ischaemic events. Dose reduction and progress is monitored by the clinical state and ESR and CRP levels.[11] Concomitant use of H_2-receptor antagonists may be appropriate initially. Temporal arteritis may take 1–2 years to resolve.

Frontal sinusitis

The headache of frontal sinusitis can be a diagnostic problem especially in the absence of, or a lapse in time since, an obvious upper respiratory infection or vasomotor rhinitis. Some patients do not have a history of a preceding respiratory infection, nor any signs of nasal obstruction or fever. Contrary to popular belief, sinusitis is a relatively uncommon source of headache.

Clinical features

It presents typically as a frontal or retro-orbital headache (see FIG. 45.9). A characteristic is its diurnal variation, developing in the morning around 9 am, being most intense in the middle of the day, then subsiding to offset around 6 pm.

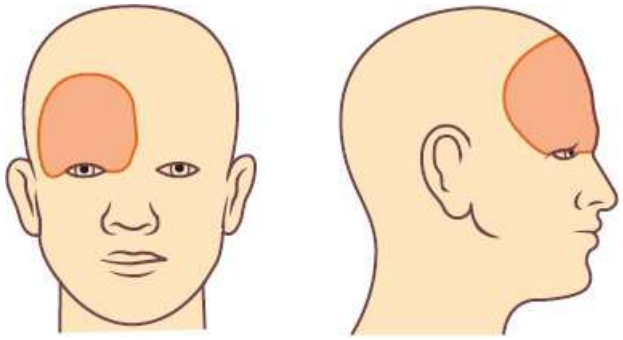

FIGURE 45.9 Typical distribution of pain of frontal sinusitis (right side)

Examination

There is tenderness over the frontal sinus and pain on percussion over the sinus. Ewing sign may be elicited. Fever and oedema of the upper eyelid may be present.

Management

Principles of treatment

- Drain the sinus conservatively using saline nasal irrigation or steam inhalations
- Use oral or intranasal antihistamine initially
- If no response, add in an intranasal corticosteroid
- Analgesics
- Refer to ENT if persistent and purulent nasal discharge—pus can spread. While waiting, a temporary trial of oral prednisolone 25 mg may be reasonable.

Complications

- Orbital cellulitis
- Subdural abscess
- Osteomyelitis
- Cavernous sinus thrombosis

Symptoms indicating spread of infection (requires antibiotics):

- increase in fever and chills

- vomiting
- oedema of the eyelids and forehead
- visual disturbances
- dulling of the sensorium
- convulsions

Raised intracranial pressure

Important causes of a space-occupying lesion include a cerebral tumour and subdural haematoma. Sometimes it is not possible to differentiate between a subdural and an extradural haematoma, although the latter classically follows an acute injury (see CHAPTER 64). Typical features are generalised headache, usually worse in the morning, aggravated by abrupt changes in intracranial pressure and later associated with vomiting and drowsiness. Headache is an uncommon presenting symptom of a cerebral tumour.

DxT drowsiness + vomiting + seizure → raised intracranial pressure

Site:	generalised, often occipital
Radiation:	retro-orbital
Quality:	dull, deep steady ache
Frequency:	daily
Duration:	may be hours in morning
Onset:	worse in mornings, usually intermittent, can awaken from sleep
Offset:	later in day (if at all)
Aggravating factors:	coughing, sneezing, straining at toilet
Relieving factors:	analgesics (e.g. aspirin), sitting, standing
Associated features:	vomiting (without preceding nausea); vertigo/dizziness; drowsiness; seizures; confusion (later); neurological signs (depending on side)

Examination

- Focal CNS signs

- Papilloedema (see FIG. 45.10) (but may be absent)

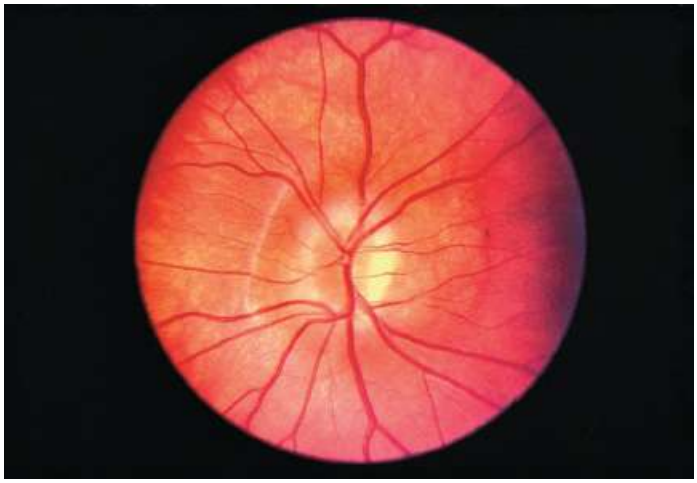

FIGURE 45.10 Papilloedema with swollen optic disc of the ocular fundus due to raised intracranial pressure

Intracerebral tumours

- Incidence is 5–10 per 100 000 population
- Two peaks of incidence: children <10 years[3] and 35–60 years
- Main types of tumour:

children: medulloblastoma, astrocytoma (posterior fossa), ependymoma, glioma (brain stem)

adults: cerebral glioma, meningioma, pituitary adenoma, cerebral metastases (e.g. lung)

Investigations

- CT scan and MRI

Subarachnoid haemorrhage

SAH is a life-threatening event that should not be overlooked at the primary care level. The incidence is 12 per 100 000 population per annum. About 40% of patients die before treatment, while about one-third have a good response to treatment.

Approximately 75% will present with an acute severe headache and the remainder with loss of consciousness.

Clinical features

- Sudden severe ‘thunderclap’ headache
- Occipital location
- Localised at first, then generalised
- Pain and stiffness of the neck follows
- Vomiting and loss of consciousness often follow
- Confusion or a lowered consciousness level
- ± Seizures
- Kernig sign positive (see CHAPTER 20)
- Neurological deficit may include: hemiplegia (if intracerebral bleed), third nerve palsy (partial or complete) (see FIG. 45.11)

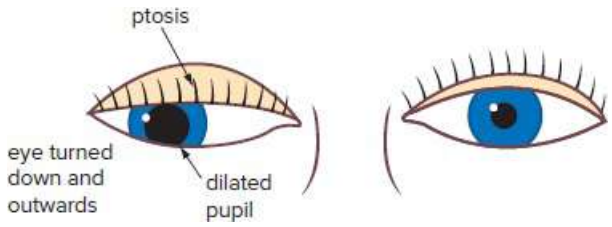

FIGURE 45.11 Third nerve palsy (right side)

About one-third of patients experience a ‘sentinel’ headache.

DxT occipital headache + vomiting + neck stiffness → SAH

Diagnosis

CT scanning is the investigation of choice and should be performed in the first few hours (blood has dispersed by 6 hours). Lumbar puncture is not necessary if the diagnosis can be made by CT, but is used if the CT scan is negative (usually 10–20% of cases). It may be falsely negative after 7 days. Homogenous blood staining of CSF and xanthochromia is a positive feature on lumbar puncture.

Special notes

- Less severe headaches can cause diagnostic difficulties.

- Consider an angioma rather than an aneurysm as the cause of SAH if previous episodes.

Management

Immediate referral for possible surgical intervention is required. If there is lingering doubt, provide ‘safety net’ instructions and review the patient within 12–24 hours.

Meningitis

The headache of meningitis is usually generalised and radiates to the neck. It is constant and severe and occasionally may begin abruptly. It is aggravated by flexion of the neck. Brudzinski sign (neck flexion) and Kernig sign are positive (see CHAPTER 20). Fever and neck stiffness are usually present. Urgent referral to hospital is necessary. If meningitis is suspected or if a child or adult has headache with fever and neck stiffness, antibiotics must not be given until a lumbar puncture has been performed.

Medication overuse (rebound) headache

Drug overuse can cause rebound headaches—even those drugs used to combat the symptom: analgesics, triptans and ergotamine, typically with regular use >15 days per month for 3 months. A long list of over-the-counter and prescription medications can cause rebound, such as aspirin, paracetamol, ibuprofen, opioids and caffeine. The headache is present on waking and typically persists throughout the day but fluctuates in intensity. It is a mild to moderate, dull, bilateral ache with a distribution similar to tension headache. Drug (or alcohol) rebound headaches should be suspected in any patient who complains of headache ‘all day, every day’. A careful drug history should be taken. Treatment includes gradual withdrawal of the drugs, bridging therapy with a short course of prednisolone or an NSAID such as naprosyn MR, and the substitution of anti-emetics and amitriptyline or beta blocker over about 14 days.

Chronic paroxysmal hemicrania

This is a rare headache syndrome that overlaps with cluster headache and facial pain. The unilateral pain, which can be excruciating, is located in the area of the temple, forehead, eye and upper face. It can radiate to the ear, neck and shoulder. It differs from cluster headaches in that the patients are invariably female, the paroxysms are short (average 20–30 minutes) and more frequent, with attacks occurring up to 14 times a day. The disorder resembles cluster headaches in nature and distribution and associated autonomic features, such as ipsilateral nasal stuffiness or rhinorrhoea, lacrimation, conjunctival injection and ptosis. The aetiology is unknown but the headache often responds dramatically to indomethacin (25 mg (o) tds).[11]

Post-traumatic headache

Following head trauma, this is a continuous, diffuse type of headache with associated psychological symptoms such as dizziness, irritability and depression. Long known to be a

potential sequelae to a single severe head injury, it is being increasingly recognised in the context of repetitive minor concussions, particularly in contact sports. Headaches can persist for 6–12 months and are best treated with aspirin or paracetamol. If unresponsive and persistent, amitriptyline or sodium valproate can be tried.[10]

Low CSF pressure headache[10]

The most common cause is a CSF fluid leak after dural puncture. The headache is usually present when standing or sitting and rapidly improves with lying flat. It can also be spontaneous or due to trauma. It can be severe with nausea and vomiting. In most, resolution occurs within 2–7 days. Treatment includes bed rest until resolution. If persistent, referral for an epidural blood patch is recommended.

Trigeminal neuralgia

The pain of trigeminal neuralgia comes in excruciating paroxysms, which last for seconds to minutes only and usually affect the face rather than the head (see CHAPTER 41). The lightning-like jabs of searing or burning pain usually last 1 to 2 minutes but can last as long as 15 minutes.

Icepick headache

Icepick headaches are similar sudden stabbing pains lasting a few seconds usually at the temple (often bilateral) and are more common in migraine sufferers. They can occur unpredictably 30 or more times a day. Treatment is with indomethacin 25 mg tds.[10]

Hypertension headache

It tends to occur only in severe hypertension such as malignant hypertension or hypertensive encephalopathy. The headache is typically occipital, throbbing and worse on waking in the morning.

The headache may be psychogenic in origin, developing after the diagnosis of hypertension is disclosed to the patient. Hypertension and headaches are two very common occurrences in general practice, so their association does not imply causation. However, the occasional patient has genuine headache related to milder hypertension and this serves as an accurate indicator of their blood pressure level.

Idiopathic intracranial hypertension (pseudotumour cerebri)

This is a rare but important sinister headache condition that typically occurs in young obese women mainly in the second to fifth decades but can occur at any age. Key features are headache, visual blurring and obscurations, nausea and papilloedema. It is considered to be due to a disturbance in the CSF circulation. The CT and MRI scans are normal but lumbar puncture

reveals increased CSF pressure 25 cm H_20 and normal CSF analysis.

It is sometimes linked to drugs, including tetracyclines (most common), nitrofurantoin, oral contraceptive pill, steroids and vitamin A preparations. The main concern is visual deficits from the high intracranial pressure. Urgent referral is essential. Medical treatment includes ceasing causative drugs (the key), weight reduction, corticosteroids and diuretics, usually acetazolamide. The treatment of choice to alleviate symptoms is repeated lumbar puncture. Surgery, which involves decompression of the optic nerves or lumbo-peritoneal shunting, is sometimes required for failed medical therapy.

DxT obese young patient + headache + visual obscurations + nausea → idiopathic intracranial hypertension

Headaches related to specific activities[20]

Sex headache

This can manifest as a dull or explosive headache, provoked by sexual arousal and activity, especially with orgasm. Some are clearly a form of exertional headache. Sometimes sex headache is mistaken for SAH, but if the severe headache coincided with orgasm, was not associated with vomiting or neck stiffness, and settled within hours, SAH is unlikely. Treatment is with prophylactic beta blockers or ergotamine 1 mg (o) 1–2 hours before sexual activity (may induce erectile dysfunction in males).

Cough and exertional headache

Some people experience a severe transient pain with factors such as coughing, sneezing, stooping, straining, lifting and various sporting activities. It is usually benign and examination is normal. A CT scan is indicated if there are focal signs or if the symptoms do not settle.

Treatment is indomethacin 25 mg (o) 2–3 times daily for cough headache and 1–2 hours before exertional activity.

Gravitational headache

Occipital headache, coming on when standing upright and relieved by lying down, is characteristic of a postlumbar puncture, an epidural block or low-pressure headache. It can last for several weeks after the procedure.

‘Ice-cream’ headache

Frontal or global headache can be provoked by the rapid ingestion of very cold food and drink. It is a form of vascular headache.

When to refer

- Thunderclap headache
- Evidence or suspicion of SAH or intracerebral haematoma
- Prolonged neurological symptoms
- Complicated migraine
- Uncertain diagnosis
- Positive neurological signs despite typical headaches
- Isolated aura as headache
- Needing acute treatment for >8–10 days a month

Practice tips

- Anyone >55 years presenting with unaccustomed headache has an organic disorder such as temporal arteritis (TA), intracerebral tumour or subdural haematoma until proved otherwise.
- The ESR is an excellent screening test to diagnose TA but occasionally can be normal in the presence of active TA.
- If a patient presents twice within 24 hours to the same practice or hospital with headache and vomiting, consider other causes apart from migraine before discharging the patient.[9]
- If migraine attacks are severe and unusual (e.g. always on the same side) consider the possibility of cerebral vascular malformation.
- CT scans and MRI have superseded other investigations in the diagnosis of cerebral tumours and intracranial haemorrhage but should be ordered sparingly and judiciously.
- If a headache is occipital in origin or accompanied by neck pain, consider the likely possibility of cervical dysfunction and refer to the appropriate therapist once the diagnosis is established.
- For recurrent migraine sufferers emphasise the importance of trigger factor avoidance and of taking aspirin and metoclopramide medication at the earliest warning of an attack.

- A severe headache of sudden onset is SAH until proved otherwise. It is overlooked sometimes.
- SAH is sometimes overlooked because it is not considered in the differential diagnosis. Suspect with very severe and protracted headache, drowsiness and neck stiffness.
- Medical evidence indicates that most headaches are related to fatigue, stress or migraine triggers and respond to application of heat or cold, exercise and common analgesics, including aspirin and ibuprofen.[21]
- A history of migraine with aura over the past five years is a contraindication to prescribing the combined contraceptive pill (venous thromboembolism risk).[14]
- The use of narcotics for migraine treatment (such as pethidine and codeine) is to be avoided whenever possible—the frequent use of ergotamine, analgesics or narcotics can transform episodic migraine into chronic daily headache.[4]
- Migraine, especially chronic and recurrent, is associated with an increased risk of anxiety and depressive disorders.

Patient education resources

Hand-out sheets from *Murtagh's Patient Education* 8th edition:

- Migraine
- Tension headache

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46 Hoarseness

Hoarseness results from imperfect phonation due to impairment of normal vocal cord mobility or vibration. It is an important symptom as it may signal a serious cause such as malignancy or a disease with potential for airway obstruction.[1]

RAYMOND L CARROLL 1996

Hoarseness (dysphonia) is defined as an altered voice due to a laryngeal disorder.[2] It is an important symptom of laryngeal disease presenting in general practice, and ranges from the very common, trivial, self-limiting condition of viral upper respiratory tract infection to a life-threatening disorder (see TABLE 46.1). It may be of sudden presentation lasting only a few days or develop gradually and persist for weeks or months. The cut-off point between acute and chronic hoarseness is three weeks' duration, by which time most self-limiting conditions have resolved. Hoarseness pertains to harsh, raspy, gravelly or rough tones of voice rather than pitch or volume. Rarely, hoarseness can be a functional or deliberate symptom referred to as 'hysterical aphonia'.[3] In this condition, people purposely hold the cords apart while speaking.

Table 46.1 Hoarseness: diagnostic strategy model

Probability diagnosis
Viral URTI: acute laryngitis
Non-specific irritative laryngitis (Reinke oedema)
Vocal abuse (shouting, screaming, etc.)
Nodules and polyps of cords
Presbyphonia in elderly: 'tired' voice
Acute tonsillitis
Serious disorders not to be missed
Cancer: larynx, lung, including recurrent laryngeal nerve palsy, oesophagus, thyroid
Imminent airway obstruction (e.g. acute epiglottitis, croup)
Other rare severe infections (e.g. TB, diphtheria)
Foreign body

Motor neurone disease

Aortic arch aneurysm

Myasthenia gravis

Pitfalls (often missed)

Toxic fumes

Vocal abuse

Benign tumours of vocal cords (e.g. polyps, ‘singer’s nodules’, papillomas)

Gastro-oesophageal reflux → pharyngolaryngitis

Goitre

Vocal cord palsy

Dystonia

Physical trauma (e.g. post-intubation), haematoma

Fungal oropharyngeal infections (e.g. *Candida* with steroid inhalation, immunocompromised)

Allergy (e.g. angioedema)

Leukoplakia

Vocal cord dysfunction

‘Floppy trachea’ syndrome

Systemic autoimmune disorders (e.g. SLE, Wegener granulomatosis, myaesthenia gravis)

Seven masquerades checklist

Drugs:

- antipsychotics
- anabolic steroids
- opium users

Smoking → non-specific laryngitis

Steroids → steroid inhaler laryngitis

Hypothyroidism, acromegaly

Is the patient trying to tell me something?

Functional aphonia

Functional stridor

Key facts and checkpoints

- In acute hoarseness, the diagnosis is usually obvious from the history alone. Examples include acute upper respiratory tract infection (URTI), vocal overuse or

regular steroid inhalation.

- Think ‘hypothyroidism’ if unusual hoarseness develops.
- Laryngeal cancer must be excluded if hoarseness persists for longer than 3 weeks in an adult. It can arise intrinsic or extrinsic to the vocal cords.
- Intermittent hoarseness is invariably secondary to a benign disorder. Constant or progressive hoarseness suggests malignancy.
- Non-malignant vocal cord lesions, which include polyps, vocal nodules, contact ulcers, granulomas, other benign tumours and leukoplakia, account for about half of all chronic voice disorders.
- In cases of chronic hoarseness the larynx must be visualised for diagnosis but the following are common:

  children—‘screamer’s nodules’

  adults—non-specific irritant laryngitis
- Acute laryngeal oedema may develop as a component of the life-threatening acute angioedemic allergic response.
- Elderly or debilitated patients may exhibit a shaky or soft ‘pseudohoarse’ voice due to a weakened respiratory effort. This is termed phonaesthenia or presbyphonia.
- Contact ulcers of the larynx occur on the posterior third of the vocal cords where the mucosa is thin. The resultant weak hoarse voice may be accompanied by painful phonation. The ulcers may develop into granulomas. Apart from intubation, the condition is usually found in forceful orators who misuse their larynx when attempting to lower the pitch of their voice.[3]

The clinical approach

History

Note the nature and duration of the voice change. Inquire about corticosteroid inhalations, excessive or unaccustomed voice straining (especially singing), recent surgery, possible reflux, smoking or exposure to environmental pollutants. Elicit associated respiratory or general symptoms such as cough and weight loss. Consider symptoms of hypothyroidism or Parkinson disease.

Examination

Palpate the neck for enlargement of the thyroid gland or cervical nodes. Perform a simple oropharyngeal examination except if epiglottitis is suspected. Check for signs of hypothyroidism, such as coarse dry hair and skin, slow pulse and mental slowing. With chest examination listen for stridor. Perform indirect laryngoscopy if skilled in the procedure. Note voice characteristics e.g.:[4]

- raspy—laryngopharyngeal reflux
- deep—hypothyroidism, Reinke oedema
- gravelly—vocal cord mass/nodule
- soft—Parkinson disease, vocal cord paralysis
- intermittent—functional dysphonia, vocal cord dysfunction
- strained—muscle tension dysphonia

Investigations

The following need to be considered:

- Thyroid function tests.
- Chest X-ray if it is possibly due to lung cancer with recurrent laryngeal nerve palsy.
- Indirect laryngoscopy (the gag reflex may preclude this).
- Direct laryngoscopy with a flexible fibre-optic endoscope with possible biopsy (the most sensitive investigation).
- The choice of imaging to detect suspected neoplasia or laryngeal trauma is a CT scan.

Red flags

- History of significant smoking
- Dysphagia/odynophagia/otalgia
- Neck mass
- Haemoptysis
- Stridor
- Constitutional, e.g. weight loss, fever

Management principles

Acute hoarseness

- Treat according to cause.
- Advise vocal rest or minimal usage at normal conversational level.
- Avoid irritants (e.g. dust, tobacco, alcohol).
- Consider inhalations and cough suppressants in cases of acute URTI and coughing paroxysms.

Chronic hoarseness

- Establish the diagnosis.
- Consider referral to ENT specialist.

Hoarseness in children

- It is worth bearing in mind that stridor in infants can be caused by a congenital abnormality of the larynx, including laryngomalacia (congenital laryngeal stridor), which is particularly noticeable when the child is asleep; laryngeal stenosis (congenital laryngeal narrowing); and laryngeal paralysis due to birth trauma of the vagus nerve. Vocal cord paralysis/palsy is the most common laryngeal abnormality in children (20% of cases) after laryngomalacia.[3]
- In children exclude the acute infections—laryngotracheobronchitis (croup), tonsillitis and epiglottitis.
- Persistent hoarseness in kinder/primary school-aged children is due commonly to vocal cord nodules related to vocal abuse, such as screaming and yelling, often due to noisy children's games.
- It is important to exclude a juvenile papilloma in a hoarse child.[5]

Specific conditions

Acute laryngitis

Most cases are caused by the respiratory viruses—rhinovirus, influenza, para-influenza, Coxsackie, adenovirus and respiratory syncytial virus—resulting in vocal cord oedema. Hoarseness is a useful feature to distinguish viral from bacterial upper respiratory infections,

although don’t discount group A *Streptococcus*. Short-term vocal abuse is also a factor. The main symptom is hoarseness, which usually persists for 3–14 days and leads to loss of voice. It is often self-limiting. Even speaking can be painful. Aggravating factors include smoking, excessive alcohol drinking and exposure to irritants and pollutants, air-conditioning systems and very cold weather.

Management

- Rest at home, including voice rest (the best treatment).
- Use the voice sparingly, avoid whispering.
- Use a warm sialagogue (e.g. hot lemon drinks).
- Drink ample fluids, especially water.
- Avoid smoking, passive smoke and alcohol.
- Have hot, steamy showers as humidity helps.
- Use steam inhalations (e.g. 5 minutes, 3 times a day).
- Use cough suppressants, especially mucolytic agents.
- Use simple analgesics, such as paracetamol or aspirin, for discomfort.
- Antibiotics are of no use unless there is evidence of bacterial infection (unusual). Corticosteroids are rarely indicated.

Chronic laryngitis: ‘barmaid syndrome’

This typically occurs in a heavy smoker who works in a heavy smoking environment, who is a heavy drinker and continually talks or sings. It is a combination of vocal abuse and chemical irritation. Hoarseness often comes and goes. Treatment involves modification of these factors and screening for vocal cord tumours.

Benign tumours of the vocal cords

These include nodules (most common) (see FIG. 46.1), polyps (second most common), cysts and papules. Vocal cord nodules, including ‘singer’s nodules’, may respond well to conservative measures such as voice rest and vocal therapy. If not, they can be removed by microlaryngeal surgery or laser therapy. Dependent polyps and papillomas are removed by microsurgery.

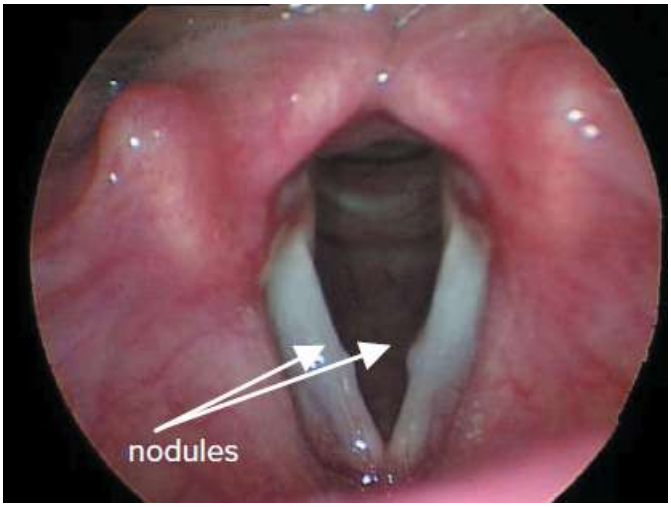

FIGURE 46.1 Vocal cord nodules

Pharyngolaryngitis

Also termed laryngopharingitis, it presents with a raspy voice, chronic throat clearing and GORD, e.g. heartburn. If due to laryngopharyngeal reflux, treat with an 8–12 week empirical course of proton-pump inhibitors as well as dietary and lifestyle modification.[2] It tends to be overdiagnosed, but should be referred to an otorhinolaryngologist if symptoms are persistent.[4]

Steroid inhaler induced laryngitis[4]

This common problem should be evident and readily treatable. Examine for oral thrush and, if present, treat with antifungal medication. Check inhaler/spacer technique and advise rinsing and gargling after use.

Laryngeal cancer

Squamous cell carcinoma usually occurs in people with a history of chronic laryngitis, smoking and alcohol use. Symptoms include hoarseness, stridor, haemoptysis and dysphagia. It may be preceded by leukoplakia, which is treated by vocal cord stripping under microsurgery. The diagnosis based on persistent hoarseness is made after fibre-optic laryngoscopy and biopsy by a specialist. The patient may present with an unexplained cervical lymph node. The condition is curable if detected early. Small local tumours can be treated by radiotherapy or laser therapy. Larger tumours usually require laryngectomy and perhaps dissection of the cervical lymph nodes (commando operation). Such radical surgery demands considerable patient support, including education about speech, eating and tracheostomy care.

Vocal cord dysfunction[6]

This condition is paradoxical vocal (or fold) adduction on inspiration and abduction on expiration, causing inspiratory airway obstruction and stridor. It tends to be misdiagnosed as asthma. Apart from dyspnoea, wheezing and stridor (usually inspiratory) symptoms may include intermittent hoarseness, chest and/or throat tightness, a noisy rasping sound and a choking or suffocating sensation. Patients may complain about a feeling 'like breathing through a straw'. Diagnosis is by observing inspiratory closure of the vocal cords with direct laryngoscopy. The mainstay of treatment is speech therapy.

Excessive dynamic airways collapse ('floppy trachea')[7]

Also known as adult tracheobronchomalacia, this is defined as pathological collapse and narrowing of the airway lumen by ≥50%. It is due to laxity of the posterior membrane into the airway lumen (in the presence of structurally intact cartilage) during forced expiration. Symptoms include breathing difficulty, coughing, difficulty clearing secretions, dyspnoea and stridor. Respiratory failure and death can occur. Diagnosis is with CT scanning and fibre-optic bronchoscopy. Treatment varies from conservative to surgery (minimal to radical). Refer to a respiratory physician.

When to refer[1]

- Acute cases that are unexplained, fail to respond by 3–4 weeks or recur; people >45 years.
- All chronic cases.
- Any case with stridor or non-tender cervical lymphadenopathy.
- Chronic hoarseness secondary to vocal abuse—refer for voice therapy.

Practice tips

- Consider intubation as a possible cause of transient hoarseness.
- Consider gastro-oesophageal reflux disease in the elderly but avoid such a diagnosis without specialist investigation for other causes.
- If stridor is present with acute hoarseness, the airway is compromised. Be on stand-by for possible emergency intervention.
- Prevention is the best treatment for laryngeal cancer (i.e. quit smoking).
- Recurrent laryngeal nerve palsy may be associated with cancer of the lung and mediastinum, or diabetes, or may be idiopathic.

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47 Jaundice

The disease is produced by black bile when it flows into the liver. The symptoms are these: 'an acute pain in the liver, also below the breast, a feeling of suffocation is strong during these days and becomes less strong later'. The liver is tender to palpation and the complexion of the patient is somewhat livid. These are symptoms that occur in the beginning but as the disease progresses, the fever diminishes in strength and the patient feels sated after ingesting a little amount of food. He must drink melikration [a mixture of water and honey].

HIPPOCRATES ON HEPATITIS

Jaundice is a yellow discolouration of the skin and mucosal surfaces caused by the accumulation of excessive bilirubin.[1] It is a cardinal symptom of hepatobiliary disease and haemolysis. Important common causes include gallstones, hepatitis A, hepatitis B, hepatitis C, drugs, alcohol and Gilbert syndrome. The commonest clinical encounter with jaundice is physiological jaundice in the newborn. As always, the history and examination are paramount, but investigations are essential to clinch the diagnosis of jaundice.

The three major categories of jaundice are (see FIG. 47.1):

- obstructive:
  - extrahepatic
  - intrahepatic
- hepatocellular
- haemolytic

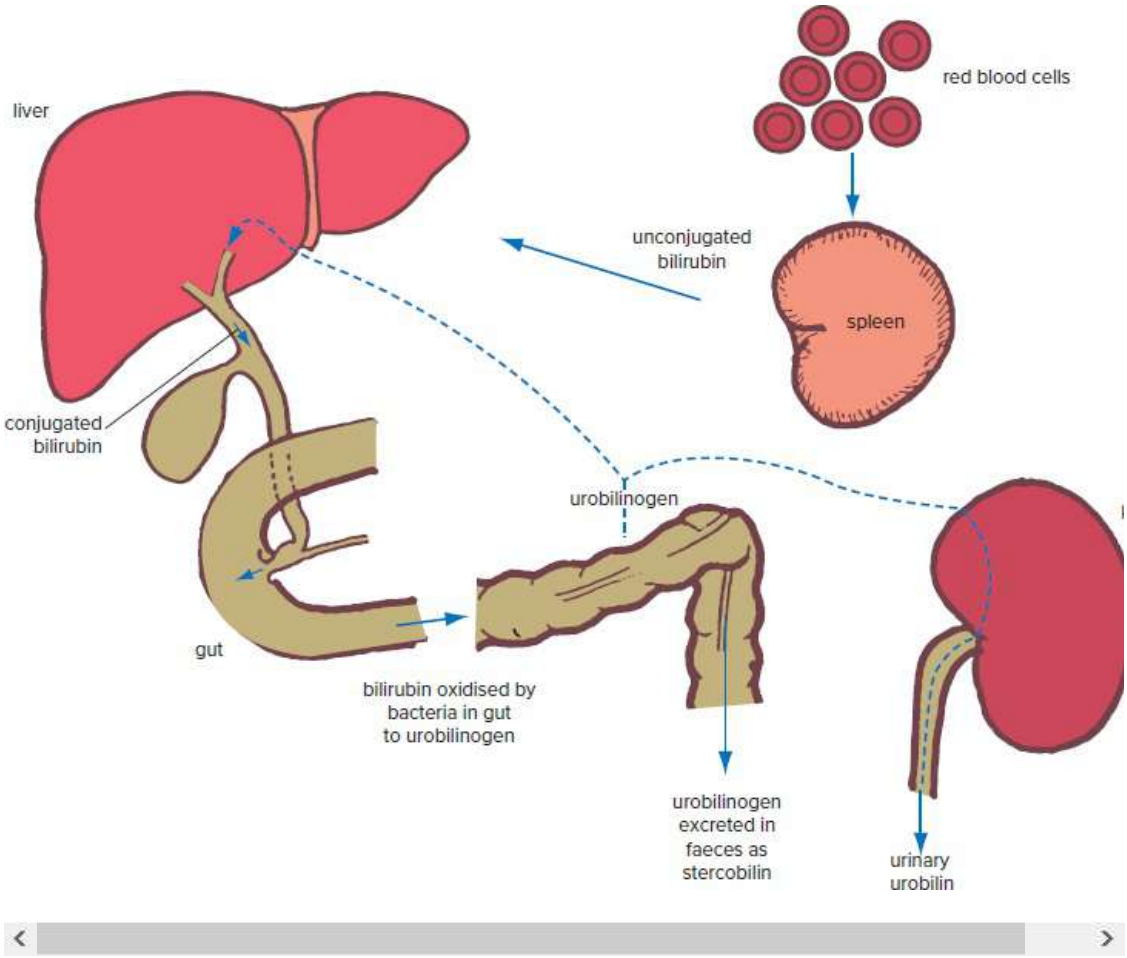


FIGURE 47.1 The jaundice pathway

Table 47.1 Abbreviations used in this chapter

Hepatitis A virus	HAV
Hepatitis A antibody	anti-HAV
Immunoglobulin M	IgM
Immunoglobulin G	IgG
Hepatitis B virus	HBV
Hepatitis B surface antigen	HBsAg
Hepatitis B surface antibody	anti-HBs
Hepatitis B core antibody	anti-HBc
Hepatitis B-e antigen	HBeAg
Hepatitis C virus	HCV

Hepatitis C virus antibody	anti-HCV
Hepatitis D (Delta) virus	HDV
Hepatitis E virus	HEV
Hepatitis F virus	HFV
Hepatitis G virus	HGV

Key facts and checkpoints

- Jaundice is defined as a serum bilirubin level exceeding 19 µmol/L.[2]
- Clinical jaundice manifests only when the bilirubin level exceeds 50 µmol/L.[1]
- However, jaundice is difficult to detect visually below 85 µmol/L if lighting is poor.
- Jaundice can be distinguished from yellow skin due to hypercarotenaemia (dietary excess of carrots, pumpkin, mangoes or pawpaw) and hypothyroidism because it involves the sclera.
- The most common causes of jaundice recorded in a general practice population are (in order) viral hepatitis, gallstones, pancreatic cancer, cirrhosis, pancreatitis and drugs.[3]
- Always take a full travel, drug and hepatitis contact history in any patient presenting with jaundice.
- Acute hepatitis is usually self-limiting in patients with hepatitis A and in adults with hepatitis B but progresses to chronic infections with hepatitis C (curable) and children with hepatitis B (containable).[4]
- A fatty liver (steatosis) can occur not only with alcohol excess but also with obesity, diabetes and starvation. There is usually no liver damage and thus no jaundice.
- Almost all patients with chronic hepatitis C will be cured with a course of oral direct-acting antiviral agents, but only if they are diagnosed, assessed, treated and monitored appropriately.

A diagnostic approach

A summary of the diagnostic strategy model is presented in TABLE 47.2 .

Table 47.2 Jaundice (adults): diagnostic strategy model

Probability diagnosis

Hepatitis A, B, C (mainly B, C)

Gallstones in common bile duct

Alcoholic hepatitis/cirrhosis

Serious disorders not to be missed

Malignancy:

- pancreas
- biliary tract
- hepatocellular (hepatoma)
- metastases

Severe infections:

- septicaemia
- ascending cholangitis
- fulminant hepatitis
- HIV/AIDS

Rarities:

- Wilson disease
- Reye syndrome
- acute fatty liver of pregnancy

Pitfalls (often missed)

Gallstones

Gilbert syndrome (↓ hepatic uptake)

Cardiac failure

Primary biliary cirrhosis

Autoimmune chronic active hepatitis

Primary sclerosing cholangitis

Chronic viral hepatitis

Haemochromatosis

Viral infections (e.g. CMV, EBV)

Leptospirosis

Seven masquerades checklist

Drugs (e.g. flucloxacillin)

Anaemia

Is the patient trying to tell me something?

Not usually applicable.

Probability diagnosis

The answer depends on the age and social grouping of the patient, especially if the patient indulges in risk-taking behaviour or has travelled overseas.

Viral hepatitis A, B or C account for the majority of cases of jaundice.

In the middle-aged and elderly group, a common cause is obstruction from gallstones or cancer. It is common for older people to have painless obstructive jaundice; bear in mind that the chances of malignancy increase with age.

Alcoholic liver disease is common and may present as chronic alcoholic cirrhosis with liver failure or as acute alcoholic hepatitis. It is worth emphasising that such patients can make a dramatic recovery when they cease drinking alcohol.

In family practice we encounter many cases of drug-induced jaundice, especially in the elderly. These drugs are outlined later in the chapter, under ‘Seven masquerades checklist’.

Serious disorders not to be missed

Malignancy must always be suspected, especially in the elderly patient and those with a history of chronic active hepatitis (e.g. post-hepatitis B or C infection). The former is more likely to have carcinoma of the head of the pancreas, and the latter, hepatocellular carcinoma (hepatoma).

Metastatic cancer must be kept in mind, especially in those with a history of surgery, such as large bowel cancer, melanoma and stomach cancer. An enlarged, knobbly, hard liver is a feature.

Hepatic failure can be associated with severe systemic infection (e.g. septicaemia and pneumonia), and after surgery in critically ill patients. The classic Charcot triad of upper abdominal pain, fever (and chills) and jaundice indicates ascending cholangitis until proved otherwise. Wilson syndrome, although rare, must be considered in all young people with acute hepatitis. A history of neurological symptoms, such as a tremor or a clumsy gait, and a family history is important. If Wilson disease is suspected, an ocular slit lamp examination, serum ceruloplasmin levels (low in 95% of patients) and a liver biopsy should be performed. Early diagnosis and treatment mean a better prognosis.

Reye syndrome is a rare and severe complication of influenza and some other viral diseases, especially in children when given aspirin. There is rapid development of hepatic failure and encephalopathy.

Pitfalls

Gallstones, especially in the absence of upper abdominal pain, can be overlooked, so this possibility should be kept in mind in the elderly.

Gilbert syndrome is worth considering, especially as it is the commonest form of unconjugated hyperbilirubinaemia. It affects at least 3% of the population and does not require treatment.

Cardiac failure can present as jaundice with widespread tenderness under the right costal margin. It can be insidious in onset or manifest with gross acute failure. It can be confused with acute cholecystitis. The biochemical abnormalities seen are very variable. Usually there is a moderate rise in bilirubin and alkaline phosphatase and sometimes, in acute failure, a marked elevation of transaminase may occur, suggesting some hepatocellular necrosis.

There are many other pitfalls for a family doctor, who may encounter the conditions very rarely, if at all. Such disorders include:

- inherited conjugated hyperbilirubinaemias (Dubin–Johnson and Rotor syndromes) caused by faulty excretion by liver cells
- haemochromatosis (associated pigmentation and diabetes)
- chronic active hepatitis
- primary biliary cirrhosis
- primary sclerosing cholangitis (associated with ulcerative colitis)

General pitfalls

- Excluding jaundice by examining the sclera in artificial light
- Not realising that the sclera in elderly patients often have an icteric appearance (without jaundice)
- Omitting to take a careful history, including illicit drugs
- Not referring for liver biopsy for all those with chronic hepatitis

Seven masquerades checklist

Of this group the haemolytic anaemias and drugs have to be considered.

Drug-related jaundice

Drug-induced jaundice is common and many drugs are implicated. The patterns of drug-related liver damage include cholestasis, necrosis (‘hepatitis’), granulomas, chronic active hepatitis, cirrhosis, hepatocellular tumours and veno-occlusive disease.[4,5] Some drugs, such as methyldopa, can initiate haemolysis.

The important drugs to consider are presented in TABLE 47.3 . Antibiotics, especially flucloxacillin, amoxicillin + clavulanate and erythromycin, are commonly implicated.

Table 47.3 Drugs that can cause jaundice

Haemolysis

Methyldopa

Hepatocellular damage

Dose-dependent:

- paracetamol (can cause acute hepatic necrosis)
- salicylates
- tetracycline

Dose-independent:

- anaesthetics (e.g. halothane)
- antidepressants (e.g. MAOIs, duloxetine)
- anti-epileptics (e.g. phenytoin, sodium valproate, carbamazepine)
- antibiotics (a long list, e.g. penicillins, sulfonamides)
- antimalarials (e.g. Fansidar)
- antiretrovirals (e.g. efavirenz, nevirapine)
- antituberculosis (e.g. isoniazid)
- anti-inflammatories (e.g. NSAIDs, various)
- carbon tetrachloride
- cardiovascular (e.g. amiodarone, methyldopa, hydralazine, perhexiline)
- statins (e.g. simvastatin)

Cholestasis

Anti-thyroid drugs

Chlorpromazine

Erythromycin estolate

Penicillins, esp. flucloxacillin

Gold salts

Oral contraceptives/oestrogens

Synthetic anabolic steroids (e.g. methyltestosterone)

Hypoglycaemic drugs (e.g. chlorpropamide)

Amitriptyline

Others

Allopurinol

Cimetidine (aggravated by alcohol)

DMARDs (e.g. methotrexate, azathioprine, ipilimumab)
Immunomodulators (e.g. interferon, TNF-alpha I)
Etretinate
Nitrofurantoin
Illicit drugs, e.g. MDMA/ecstasy, cocaine
Vitamin A (mega dosage)
Various complementary medicines (e.g. herbal agents)

Haemolysis

The person may present with the symptoms of underlying anaemia and jaundice with no noticeable change in the appearance of the urine and stool. The degree of haemolysis may vary from the lemon-yellow tinge of pernicious anaemia in an elderly patient to a severe haemolytic crisis precipitated by drugs or broad beans (favism) in a person with an inherited red cell deficiency of glucose-6-phosphate dehydrogenase (G6PD). More common causes include the hereditary haemolytic anaemias, such as congenital spherocytosis and thalassaemia major. Acquired causes include incompatible blood transfusions, malignancies (such as lymphoma), severe sepsis and some drugs.

Splenomegaly occurs in most people with haemolytic anaemia, and decreased red cell survival can be measured.

Psychogenic considerations

This is not really applicable for an organic problem such as jaundice. Nevertheless, the cause may be related to lifestyle factors that the patient may be reluctant to reveal, such as sexual orientation or intravenous drug abuse. The GP's skills at empathetic questioning become paramount.

Red flag pointers for jaundice

- Unexplained weight loss
- Progressive jaundice including painless jaundice
- Oedema
- Cerebral dysfunction (e.g. confusion, somnolence)

The clinical approach

History

The history should include questioning about the following:

- any episodes of jaundice
- change in colour of faeces and urine
- anorexia, sore throat, weight loss, pruritus
- abdominal pain
- residence and members of household
- contact with others with hepatitis or jaundice
- recent overseas travel
- exposure to blood or blood products
- needle-stick injuries or exposure to needles, such as acupuncture, tattooing and intravenous drugs
- dietary history—shellfish, drinking water
- sexual history and orientation
- drug history, including alcohol, paracetamol
- recent medical history, including surgery

- family history—family contacts who have had jaundice, haemolytic disease and other genetic liver diseases
- ethnic history—liable to haemolytic disease, contact with hepatitis B
- occupational history—exposure to hazards

Significance of various symptoms

- Pain in the right hypochondrium:

    gallstones

    acute hepatitis (a constant ache)

    cholecystitis

- Anorexia, dark urine, fever:

viral hepatitis probable

alcoholic liver disease possible

drug-induced hepatitis possible

- Pruritus:

  cholestasis probable

  possible with all liver diseases

- Arthralgia, rash:

  viral hepatitis

  autoimmune hepatitis

Examination

The abdominal examination is very important. The liver should be palpated carefully for enlargement, consistency and tenderness under the right costal margin. Search for enlargement of the gall bladder and the spleen. The gall bladder lies in the transpyloric line. A palpable gall bladder indicates extrahepatic biliary obstruction, and splenomegaly may indicate haemolytic anaemia, portal hypertension or viral hepatitis. Test for ascites.

Skin excoriation may indicate pruritus, which is associated with cholestatic jaundice. Look for evidence of chronic liver disease, such as palmar erythema, easy bruising, spider naevi and muscle wasting, and testicular atrophy and gynaecomastia. Test for hepatic flap (asterixis) and fetor, which indicate liver failure. Search for lymphadenopathy, which may be indicative of malignancy.

The examination should include dipstick urine testing for bilirubin and urobilinogen.

A summary of the possible findings is presented in FIGURE 47.2 .

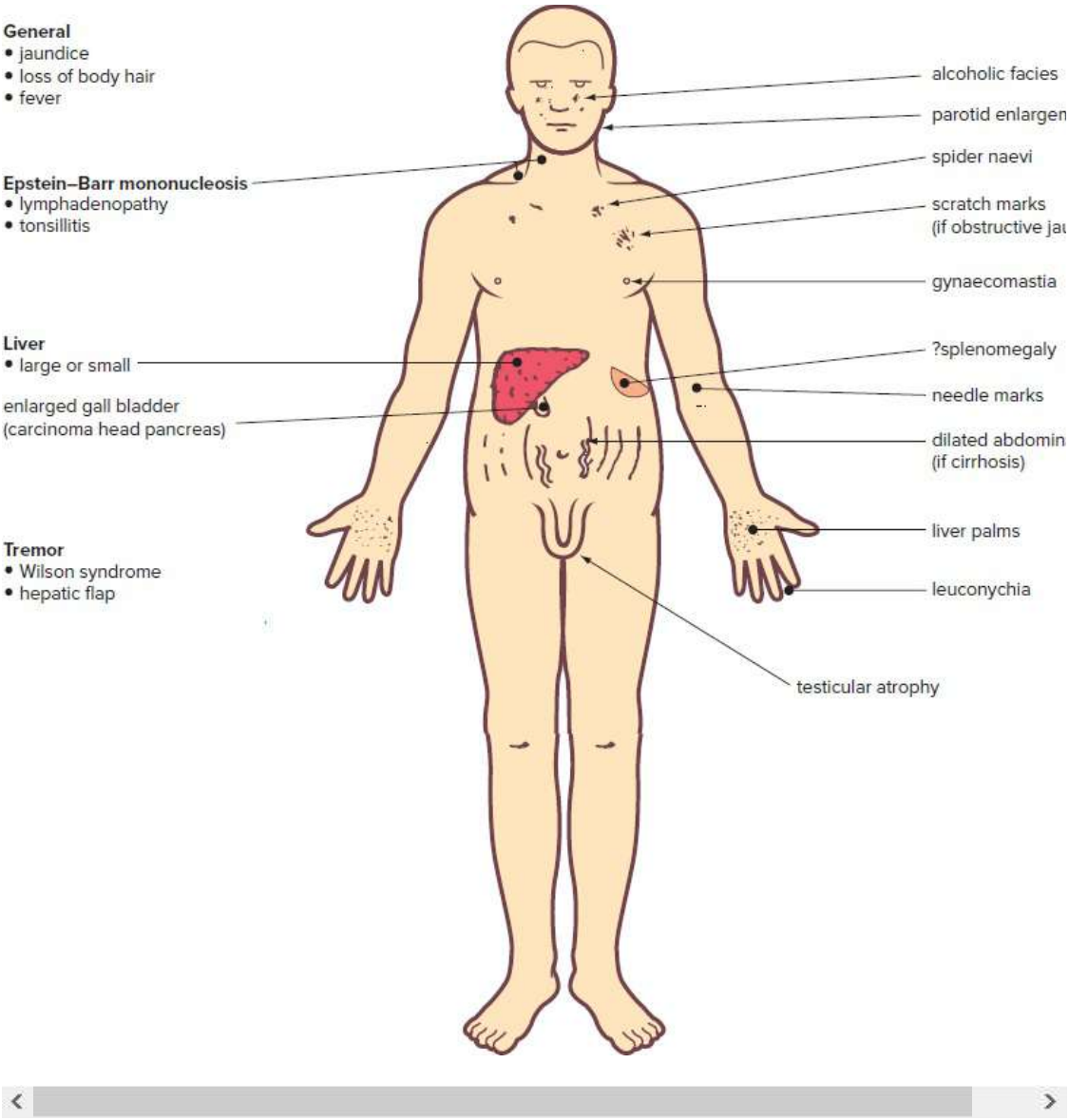


FIGURE 47.2 Possible findings on examining the jaundiced patient

Investigations

The main investigations are FBE, LFTs and the standard viral serology for the infective causes, particularly hepatitis A, B and C virus (also EBV and CMV).

A summary of the general findings for liver function tests is shown in TABLE 47.4 . Consideration should be given to ordering fractionalisation of bilirubin to determine whether it is conjugated or unconjugated (important in diagnosis of Gilbert syndrome).

Table 47.4 Characteristic liver function tests for selected types of liver disease

Liver function tests (serological)	Hepatocellular (viral) hepatitis	Haemolytic jaundice	Obstruction	Gilbert syndrome	Liv me
Bilirubin	↑ to ↑ ↑ ↑	↑ unconjugated	↑ to ↑ ↑ ↑	↑ up to 50 unconjugated	↑ t
Alkaline phosphatase	↑ <2N	N	↑ ↑ ↑ >2N	N	↑ ↑
Alanine transferase (ALT)	↑ ↑ ↑ >5N	N	N or ↑	N	↑
Gamma-glutamyl transferase	N or ↑	N	↑ ↑	N	↑
Albumin	N or ↓	N	N	N	N t
Globulin	N or ↑	N	N	N	N

N: is within normal limits

Diagnostic markers for hepatitis

- Hepatitis A: IgM antibody (anti-HAV)
- Hepatitis B: surface antigen (HBsAg)
- Hepatitis C: HCV antibody (anti-HCV)

Hepatobiliary imaging

Tests to identify causes such as malignancy or gallstones are now sophisticated and should be chosen with care.

- X-ray: a plain abdominal X-ray shows gallstones that are opaque (15–20%)
- Transabdominal ultrasound (US): the most useful investigation for detecting gallstones and dilatation of the common bile duct; also detects liver metastases and other diffuse liver diseases. The rapid increase in the diagnostic label of ‘fatty liver’ parallels the increased use of high resolution ultrasound.
- HIDA scintiscan: useful in diagnosis of acute cholecystitis

- CT scan: for diagnosis of enlargement of the head of the pancreas and other pathology; indicated if US unsatisfactory

- PTC: percutaneous transhepatic cholangiography: shows imaging of biliary tree
- ERCP: endoscopic retrograde cholangiopancreatography; PTC and ERCP (best) determine the cause of the obstruction and relieves it by sphincterotomy and removal of CBD stones
- MRCP: magnetic resonance cholangiopancreatography provides non-invasive planning for obstructive jaundice
- Liver isotopic scan: useful for liver cirrhosis, especially of the left lobe
- Fibroscan (transient elastography) to measure liver fibrosis

Specific tests

Some specific tests include:

- autoantibodies for autoimmune chronic active hepatitis and primary biliary cirrhosis
- carcinoembryonic antigen to detect liver secondaries, especially colorectal
- serum iron studies, especially transferrin saturation—elevated in haemochromatosis
- alpha-fetoprotein—elevated in hepatocellular carcinoma; mild elevation with acute or chronic liver disease (e.g. cirrhosis)
- serum ceruloplasmin level—low in Wilson disease
- liver biopsy
- EBV/cytomegalovirus serology (consider if hepatitis serology negative)

Jaundice in children

Jaundice in the infant

Jaundice in the newborn is clinically apparent in 50% of term babies and more than 80% of preterm.[6] Icterus is therefore common and almost invariably physiologically benign. However, it is a cause for concern as there are many other causes and investigation is needed to determine whether the bilirubin is conjugated (always pathological) or unconjugated. If conjugated, consider the serious biliary atresia (stools are white); also a cyst obstructing the bile duct or neonatal hepatitis. Prompt referral is essential.

Jaundice occurring in the first 24 hours after birth is not due to immature liver function but is pathological and usually due to haemolysis consequent on blood group incompatibility. In primigravidas it is usually due to ABO incompatibility. Rhesus factor is the most severe form of alloimmune haemolytic disease of the newborn. At birth, it presents with hydrops, anaemia,

jaundice and hepatosplenomegaly. Prevention is with injections of anti-D immunoglobulin for Rh negative women at 28 and 34–36 weeks.

Causes of pathological jaundice are presented in TABLE 47.5 Such causes demand referral.

Table 47.5 Pathological neonatal jaundice: diagnostic strategy model (brief)

Probability diagnosis
Physiological: day 2–5
ABO incompatibility: first 24 hours
Breast milk jaundice: late first week
Serious disorders not to be missed
ABOI: first 24 hours
Biliary atresia (conjugated)
Haemolysis: ABOI, G6PD, Rhesus incompatibility
Sepsis
Seven masquerades checklist
Drugs
Hypothyroidism
Hereditary spherocytosis, other inherited disorders
Polycythaemia

Bilirubin encephalopathy

Unconjugated bilirubin can be regarded as a neurological poison. With increasing serum levels an encephalopathy (which may be transient) can develop, but if persistent can lead to the irreversible brain damage known as kernicterus. The level of bilirubin causing kernicterus is totally unpredictable, but a guideline as a cause for concern in babies with Rh disease is a serum unconjugated bilirubin of 340 µmol/L (20 mg/dL).

Guidelines for treatment for hyperbilirubinaemia (at 24–36 hours)—an example:

- >285 µmol/L—phototherapy
- >360 µmol/L—consider exchange transfusion

An example of a normogram is presented in FIGURE 47.3 .

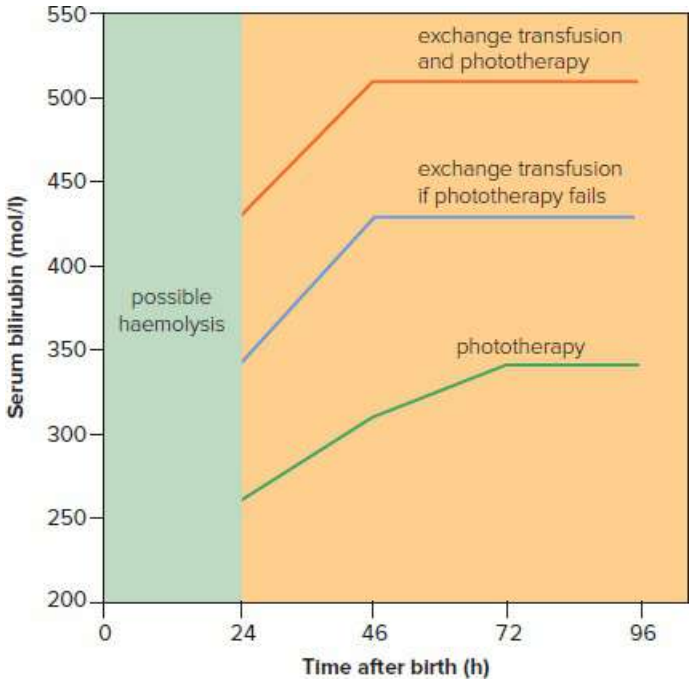


FIGURE 47.3 Typical normogram for decision making in healthy infants with jaundice

Physiological jaundice

This mild form of jaundice, which is very common in infants, is really a diagnosis of exclusion. In a term infant the serum bilirubin rises quickly after birth to reach a maximum by day 3–5, then declines rapidly over the next 2–3 days before fading more slowly for the next 1–2 weeks. Management includes phototherapy.

ABO blood group incompatibility

This is antibody-mediated haemolysis (Coomb test positive):

- Mother is O
- Child is A or B

Jaundice develops within first 24 hours.

Treatment

- Perform a direct Coomb test on infant.
- Phototherapy is required immediately.

- These children require follow-up developmental assessment including audiometry.

Breast milk jaundice

If the secondary causes of prolonged jaundice are excluded, the baby is well and breastfeeding, the likely cause of unconjugated elevated bilirubin is breast milk jaundice. It occurs in 2–4% of breastfed infants. It usually begins late in the first week and peaks at 2–3 weeks. Diagnosis can be confirmed by suspending (not stopping) breastfeeding for 24–48 hours, although some doctors recommend against this. The serum bilirubin falls and then breastfeeding can continue. The mother, who can express milk for this short period, must be reassured that there is nothing wrong with the milk and advised to resume.

Jaundice in older children

Viral infection is the commonest cause of jaundice in the older child, especially hepatitis A and hepatitis B. It is uncommon for viral hepatitis to become chronic in childhood.

Jaundice in the elderly

If an elderly person presents with jaundice the usual causes and investigations have to be considered. Obstructive jaundice is the commonest form of jaundice in the elderly and may be caused by gallstones blocking the common bile duct (may be painless) and carcinoma of the head of the pancreas, the biliary tract itself, the stomach or multiple secondaries for other sites. While it is not uncommon for a gallstone to produce marked obstructive jaundice and yet be painless, it is appropriate to adhere to the old adage that painless obstructive jaundice is due to neoplasm—particularly if the gall bladder is palpable (Courvoisier's law).

Alcoholic liver disease, although most frequently affecting those aged 40 to 60 years, can present for the first time over age 60 years. The commonest cause of hepatocellular jaundice in the elderly is probably alcoholic cirrhosis; hepatitis A is still relatively uncommon in old persons.

Drugs do not cause jaundice in the elderly as frequently as they once did, particularly as phenothiazines, especially chlorpromazine, are not prescribed as often as previously. However, drugs should be considered as a potential cause and a careful check of the drug history is important.

Infective causes of jaundice

A generation ago hepatitis A (infectious hepatitis or yellow jaundice) was the commonest recognised form of viral hepatitis, presenting usually with an abrupt onset of fever, anorexia, nausea and vomiting. It usually occurred in epidemics and hence was common in overcrowded institutions and camps. Now hepatitis B and C are the most commonly reported types of viral hepatitis with an onset that is more insidious and with a longer incubation period.[4,7] Symptoms include malaise, anorexia, nausea and polyarthritis. Acute hepatitis C is often subclinical.

The various forms of hepatitis are summarised in TABLE 47.6 . All forms of hepatitis are common in developing countries and travellers are at risk of contracting these infections: hepatitis A and E from faeco-oral transmission; and hepatitis B, C, D and G from intravenous drugs and bodily fluids (from sexual transmission, in particular, for hepatitis B).

Table 47.6 Characteristic profiles of viral hepatitis A–E

Characteristic	Hepatitis A	Hepatitis B	Hepatitis C	Hepatitis D	Hepa
Pseudonyms	Infectious hepatitis	Serum hepatitis	Parenterally transmitted non-A, non-B	Delta hepatitis	Enter transı non-A
Agent (virus)	27 nm RNA	42 nm DNA	50 nm RNA	35 nm RNA	30 nn
Transmission	Faecal–oral Contaminated water/food	Blood and other body fluids Mother to child	Infected blood ?Other body fluids	Blood and other body fluids	Faec Cont wate
Incubation period	15–45 days	40–180 days	14–180 days	30–50 days	15–4
Severity of acute illness	Mild to moderate; often subclinical—no jaundice	Mild to severe; jaundice common; arthralgia and rash common	Mild to moderate; often subclinical	Moderate to severe; high mortality; usually jaundice	Mild t mode often subcl
Chronic liver disease	No	Yes 10–15%	Yes 65–80%	Yes Potentially worst	No
Carrier state	No	Yes	Yes	Yes	No
Risk in travellers	Yes, applies to all A–E: East and South-East Asia, Asian subcontinen India), South Pacific Islands (e.g. Fiji), sub-Saharan Africa, Mexico, R other developing countries. A and E with poor sanitation; B, C, D also drug use; B, D sexual contact.				

Antigens	HAV Ag	HBsAg, HBcAg, HBeAg	HCV Ag	HDV Ag	HEV
Diagnosis—antibodies	anti-HAV	HBsAg anti-HBc anti-HBs	anti-HCV	anti-HDV	anti-H
Vaccine	Hepatitis A vaccine	Hepatitis B vaccine	None	Hepatitis B vaccine	None
Curable	No progression	No	Yes	No	Varia

Evidence points to more viruses causing non-ABC hepatitis.[8] Hepatitis F virus has been claimed to be transmitted enterically while the newly designated hepatitis G virus (HGV) is transmitted parenterally. It does not appear to cause a severe illness in recipients. It can be predicted that the hepatitis alphabet will continue to expand.

In hepatitis A, liver damage is directly due to the virus, but in hepatitis B and C it is due to an immunologic reaction to the virus.

Other infections that can present with jaundice as part of a systemic disease are malaria, Epstein–Barr mononucleosis, cytomegalovirus, Q fever, toxoplasmosis, syphilis, leptospirosis and, rarely, measles, varicella, yellow fever, rubella, herpes simplex, dengue fever, Lassa fever and Marburg and Ebola virus.

Hepatitis A

Hepatitis A is becoming relatively less prevalent in developed countries. It is enterically transmitted and arises from the ingestion of contaminated food, such as shellfish, or water. There is no carrier state and it does not cause chronic liver disease. Hepatitis A most often causes a subclinical or self-limited clinical illness.

Clinical features

Pre-icteric (prodromal) phase:

- anorexia, nausea ± vomiting
- malaise
- headache
- distaste for cigarettes in smokers

- mild fever
- ± diarrhoea

- ± upper abdominal discomfort

Icteric phase (many patients do not develop jaundice):

- dark urine
- pale stools
- hepatomegaly
- splenomegaly (palpable in 10%)

Recovery usually in 1–10 weeks (average 6 weeks).

Fulminant hepatitis with liver coma and death may occur but is rare.

Investigations

LFTs and viral markers confirm the diagnosis. The antibodies to HAV are IgM, which indicates active infection, and IgG antibodies, which means past infection and lifelong immunity and which is common in the general population. Ultrasound is useful to exclude bile duct obstruction, especially in an older patient.

Outcome and treatment

Hepatitis A has an excellent prognosis with most patients making a complete recovery, and patients should be reassured. The mortality is less than 0.5%. Admission to hospital is not usually necessary. There is no specific treatment, so management is as follows.

- Provide appropriate reassurance and patient education.
- Rest as appropriate.
- Follow a fat-free diet.
- Avoid alcohol, smoking and hepatotoxic drugs (until recovery).
- Advise on hygiene at home to prevent spread to close contacts and family members. Hep A can also be spread sexually and by IV drug use.
- Wash hands carefully after using the toilet and disinfect them with antiseptic.

- Do not handle food for others with your fingers.
- Do not share cutlery and crockery during meals.

- Do not use tea-towels to dry dishes.

Prevention

Simple health measures such as good sanitation, effective garbage disposal and handwashing are probably responsible for the major decrease in the disease. Immune serum globulin (0.03–0.06 mL/kg IM) confers satisfactory passive immunity for close contacts (within 2 weeks of contact) and for travellers to endemic areas for up to 3 months. An active vaccine consisting of a two-dose primary course is the best means of prevention.

Hepatitis B

Hepatitis B has protean clinical manifestations. Transmission is by blood spread, percutaneous, sexual transmission, perinatal spread or by close prolonged family contact. Infection may be subclinical or self-limited acute hepatitis. Fulminant hepatitis is rare. Five per cent of subjects go on to become chronic carriers of the virus. Most are ‘healthy carriers’ but some may develop chronic active hepatitis, cirrhosis and hepatoma. The serology of hepatitis B involves antibody responses to the four main antigens of the virus (core, DNA polymerase, protein X and surface antigens). Passive and active vaccines are available, and should be used freely in groups at risk, including babies of infected mothers. High-risk groups are presented in TABLE 47.7 . The clinical features are the same as those found in hepatitis A infection but may be less abrupt in onset but more severe in the long term.[7] A serum sickness-like immunological syndrome may be seen with transient rashes (e.g. urticaria or a maculopapular rash), and polyarthritis affecting small joints in up to 25% of cases in the prodromal period.

Table 47.7 Higher-risk groups for contracting hepatitis B (vaccination advisable)[7]

Babies born to hepatitis B positive (carrier) mothers
Migrants from high prevalence regions, e.g. Africa, Asia
Garbage collectors
Health care workers
Household contacts of hepatitis B carriers
Institutionalised people with intellectual disability
Users of intravenous drugs
People on kidney dialysis
Men who have sex with men
Prisoners
Recipients of blood or blood products (prior to testing)

Sex industry workers

Sexual partners of hepatitis B carriers (especially acute HBV)

Travellers to endemic areas

Investigations[5,9,10]

The main viral investigation for HBV is HBsAg (surface antigen), which is searched for routinely. If detected, indicating hepatitis B positive or carrier, a full viral profile is then formed. HBeAg is a soluble protein from the pre-core and core. Antibodies develop to both HBsAg and HBeAg.

HBsAg may disappear or persist. Its presence indicates a current or chronic infection as well as a carrier state (see FIG. 47.4).

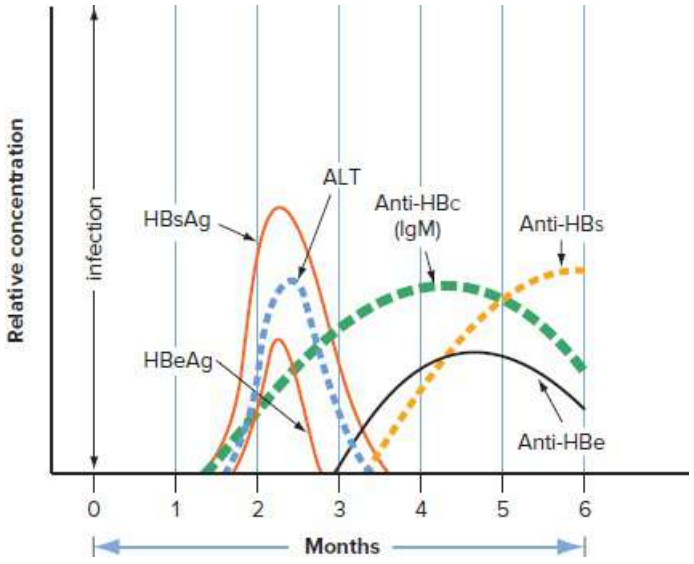


FIGURE 47.4 Time course of acute hepatitis B infection

Chronic hepatitis B (carriage) is defined as the presence of HBsAg for at least 6 months. The typical natural history model of chronic hepatitis B infection is outlined in FIGURE 47.5 .

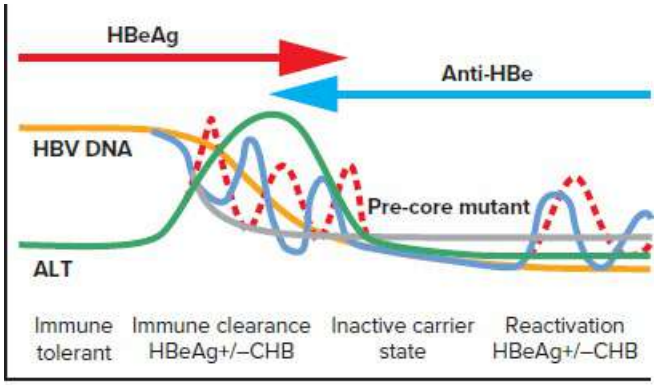
The revised four phases of the chronic infection are:

1. Immune tolerant—HBeAg +ve; normal LFTs
2. Immune clearance—HBeAg +ve or −ve chronic hepatitis; abnormal LFTs
3. Immune control—inactive carrier (residual) state; HBeAg −ve; normal LFTs
4. Immune escape (reactivation)—HBeAg −ve; anti-HBe +ve; abnormal LFTs

Practice tip

Phase 2: Spontaneous seroconversion <30 years has a favourable long-term prognosis, but if prolonged, hepatic fibrosis and cirrhosis may develop. These complications and hepatocellular carcinoma are a high risk in phase 4.

Furthermore, liver histological activity is classified as mild; moderate to severe; nil or minimum or moderate to severe active (see FIG. 47.5).[11]



Liver histological activity			
Mild	Moderate/Severe	None/Mild	Moderate/Severe

FIGURE 47.5 Typical disease models of chronic hepatitis B (CHB) infection

Source: Reproduced with permission from Hepatitis B Foundation. Diagnosed With Chronic Hepatitis B? What Phase – HBeAg-Positive Chronic Hepatitis/Immune Reactive/Immune Clearance? 2013. https://www.hepb.org/blog/diagnosed-with-chronic-hepatitis-b-what-phase-immune-clearance/

Serological patterns

Acute hepatitis B

HBsAg +ve, anti-HBcIgM +ve, anti-HBs −ve

Chronic hepatitis B

HBsAg +ve, anti-HBcIgG +ve, anti-HBs −ve

Resolved hepatitis B

HBsAg −ve, anti-HBcIgG +ve, anti-HBs +ve

Serology guidelines

HBsAg = acute or persistent infection; carrier

anti-HBs = past infection and immunity

HBeAg = highly infectious; high viral replication

HBV DNA = circulating and replicating virus

anti-HBc IgM = recent and continuing infection

anti-HBc IgG = past infection

anti-HBe = seroconversion

Monitoring and outcome[9, 10]

The possible course of events is shown in FIGURE 47.6 . The majority recover completely with the outcome depending on several factors, including the virulence of the virus and the immune state and age of the patient. Some will develop chronic hepatitis, some will develop a fulminant course, and others will become asymptomatic carriers and present a health risk to others.

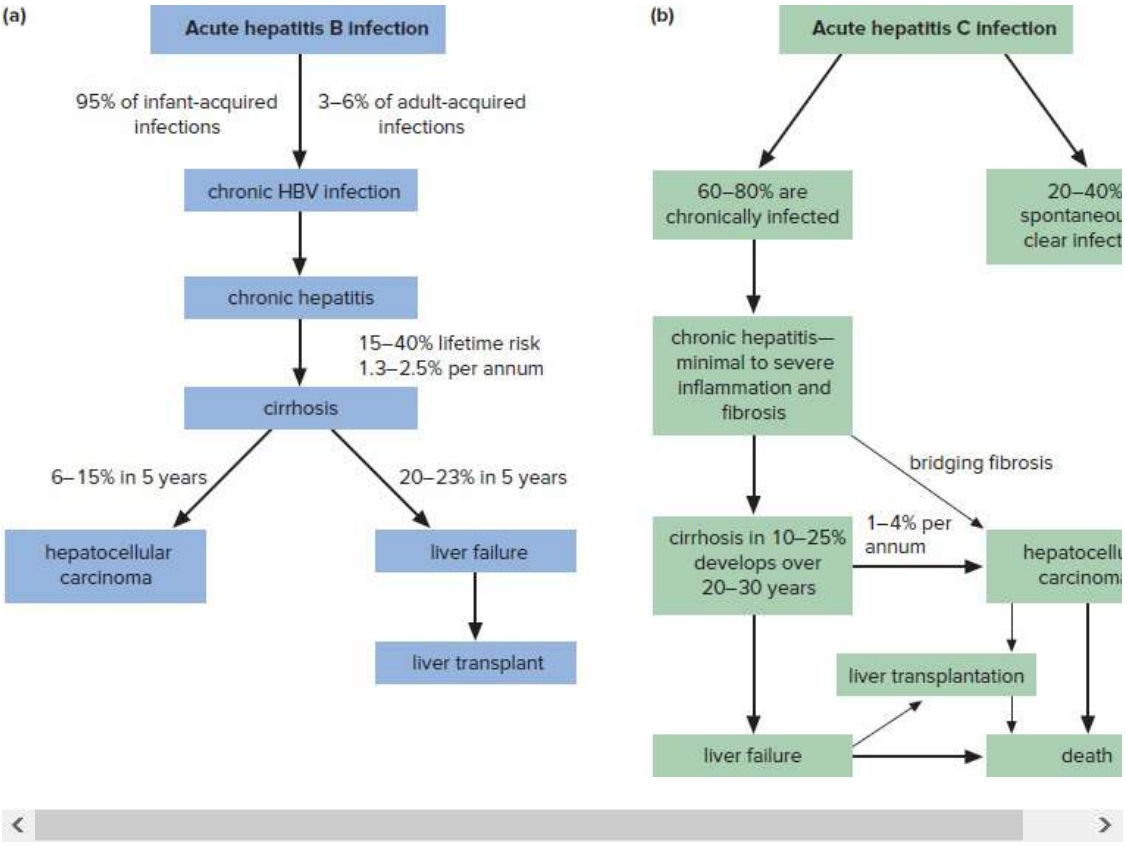


FIGURE 47.6 Natural history of **(a)** hepatitis B infection; **(b)** hepatitis C infection

Source: Reproduced with permission from W Sievert, B Katz, Department of Gastroenterology, Monash Medical Centre.

Monitor progress with 6–12 monthly LFTs, HBeAg and HBV DNA.

- Negative HBsAg and HBV DNA (with anti-HBe) = resolving, with anti-HBs = full recovery.
- Positive HBsAg and HBV DNA = replicating and infective—refer.
- Monitor LFTs every 6 months. Refer if ALT elevated.

Treatment[5]

The aim of treatment is prolonged suppression. There is no specific treatment initially—appropriate reassurance and patient education are necessary. Advise avoidance of alcohol. Avoid certain drugs, e.g. sedatives, NSAIDs, OCP, until recovery (normal LFTs). Advise about prevention of transmission, especially safe sex and sharing needles. Refer to a liver transplantation centre if encephalopathy or severe coagulopathy develop. Treatment of chronic hepatitis B infection (abnormal LFTs) is with the immunomodulatory and antiviral agents—peginterferon alpha-2a and entecavir, tenofovir or other agents. Long term, this is expensive but it achieves permanent remission in 25% of patients, and temporary remission in a further 25%.[7]

Liver transplantation has been performed, but is often followed by recurrence of hepatitis B in the grafted liver. Follow up with regular LFTs and alpha-fetoprotein screening. It is appropriate to refer any HBsAg positive patient with an abnormal ALT and/or signs of chronic liver disease to a specialist since the evaluation of chronic hepatitis B can be complex.[4]

Prevention

Active immunisation using a course of three hepatitis B vaccinations has been a major breakthrough in the management of this serious illness. If there is a negative antibody response after 3 months, revaccinate with a double dose. If the response is positive, consider a test in 5 years with a view to a booster injection.

For non-immune patients at risk (e.g. after a needle-stick injury), hepatitis B immunoglobin (HBIg), which contains a high level of HBV surface antibody, is appropriate.

Prenatal screening of pregnant women and appropriate use of HBIg and HB vaccine is useful in preventing perinatal vertical transmission of HBV.

Hepatitis C[5,9,12,13]

Hepatitis C virus is responsible for most cases of viral hepatitis in Australia. It is primarily contracted from intravenous drug use or tattooing. It does not seem to be spread very readily by sexual contact although there is a small risk. It is also not readily spread perinatally.

Clinical symptoms of hepatitis C are usually minimal (often asymptomatic), and the diagnosis is often made after LFTs are found to be abnormal. An important feature is that there are at least six major genotypes of HCV and treatment decisions are based on the genotype; thus, patients with acute hepatitis C should have HCV genotype testing (genotype 1 to 6).

Hepatitis C infection may be self-limiting, but more commonly (in about 70% of cases) without treatment it causes a slow, relentless progression to chronic hepatitis, cirrhosis (20%) and also hepatoma.[7] See FIGURE 47.7 . The severity of hepatic fibrosis can be assessed by liver biopsy or, preferably, by a non-invasive device called a fibroscan that assesses ‘hardness or stiffness’ of the liver via the technique of transient elastography. A raised ALT level that is tested three times over the next 6 months implies disease activity. HCV RNA (a PCR test) is present when the ALT becomes abnormal while the anti-HCV rises more slowly and may not be detectable for several weeks. If the PCR test is negative, the hepatitis C infection has resolved.

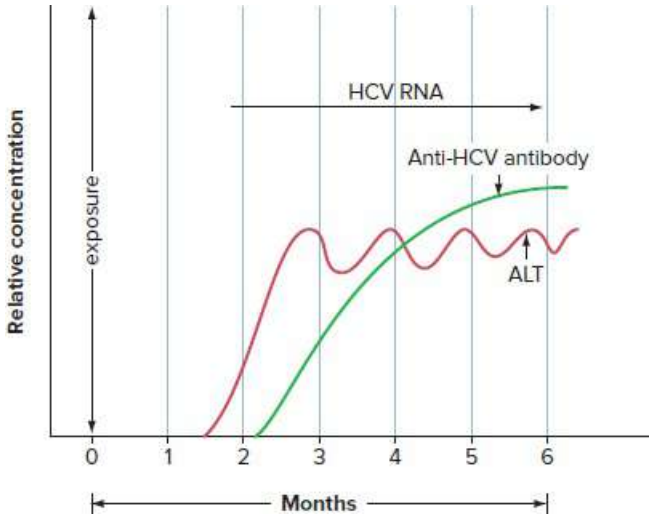


FIGURE 47.7 Time course of active hepatitis C infection

Diagnosis and progress

This is by serology:

- HCV Ab (anti-HCV) +ve = exposure (current or past)
- HCV-RNA +ve = chronic viraemia

  –ve = spontaneous clearance
- CD_4/HCV = viral load
- ALTs on LFTs indicate disease activity (tested 3 times over next 6 months)

  ALT persistently normal = good prognosis

  ALT ↑ ↑ = requires referral for treatment
- If PCR +ve + significant viral load + ALT ↑ perform HCV genotype—determines treatment
- PCR –ve, ALT –ve = infection clear

Treatment[5,13]

General strategies to prevent progression include cease or minimise alcohol and cannabis use, quit smoking, lose weight and vaccinate against hepatitis A and B. The widespread use of the direct-acting antivirals (DAAs) taken daily by mouth (usually for 8 or 12 weeks) has dramatically improved the prognosis, and a cure can now be expected in more than 90%. Side

effects of fatigue, headache, nausea and insomnia are uncommon and typically mild, not necessitating treatment cessation.[14] The determination of genotype and viral load, as well as hepatic status, will identify those most likely to respond to therapy.

Examples of DAAs are:

- protease inhibitors, e.g. simeprevir
- nucleotide polymerase inhibitors, e.g. sofosbuvir
- non-nucleotide inhibitors, e.g. dasabuvir
- NS5A inhibitors, e.g. daclatasvir, ledipasvir

These agents are given in combination according to the genotype. Treatment can be managed within general practice, in collaboration with a specialist gastroenterologist.[15]

The recommended steps for pretreatment assessment are:[15,16]

1. Confirm the diagnosis of chronic HCV infection.
2. Test for hepatitis C virus genotype and viral load.
3. Document the HCV treatment history.
4. Evaluate comorbidity and liver status, especially cirrhosis (refer if present), FBE, APRI, LFTs, blood glucose, creatinine U and E, HIV, HBV.
5. Discuss contraception and pregnancy (if applicable).
6. Consider concomitant medication.
7. Assess adherence to treatment.
8. Select treatment regimen (8 or 12 weeks) and review potential drug interactions.
9. Consult with a specialist.
10. Treat and monitor.

Cure is defined as undetectable plasma HCV RNA at least 12 weeks after cessation of treatment.

Note: DAAs are implicated with reactivation of HBV.

Those at increased risk of having hepatitis B and C

- Blood transfusion recipients (prior to HBV and HCV testing)
- Intravenous drug users (past or present)

- MSM who have practised unsafe sex
- Kidney dialysis patients
- Sex industry workers
- Those with abnormal LFTs with no obvious cause
- Those with tattoos/body piercing

Prevention of transmission of hepatitis B and C viruses

Advice to those who are positive for HBV and HCV:

- Do not donate blood or any body organs or tissues.
- Do not share needles.
- Advise health care workers, including your dentist.
- Do not share intimate equipment such as toothbrushes, razors, nail files and nail scissors.
- Wipe up blood spills in the home with household bleach.
- Cover up cuts or wounds with an adequate dressing.
- Dispose of bloodstained tissues, sanitary napkins and other dressings safely.
- Use safe sex practices such as condoms.
- Avoid tattooing.

Hepatitis D

Hepatitis D is a small defective virus that lacks a surface coat. The coat is provided by the hepatitis B virus, and so hepatitis D infection occurs only in patients with concomitant hepatitis B.

It is usually spread parenterally and if chronic is usually associated with progressive disease with a poor prognosis. Treatment with peginterferon for 48 months has a variable success rate. Antibodies to the delta virus, both anti-HDV and anti-HDV IgM (indicating a recent infection) as well as HDV Ag can be measured.[17] Referral to a specialist is recommended.

Markers of cirrhosis in infective hepatitis:

- increased INR
- thrombocytopenia

- hypoalbuminaemia
- AST/ALT ratio >1

Hepatitis E

Hepatitis E is an enterically transmitted virus that occurs in outbreaks in certain countries with a poor water supply, such as some Asian subcontinent countries. Epidemiologically, HEV behaves like HAV, with well-documented water-borne epidemics in areas of poor sanitation. There is a high case fatality rate in endemic areas (10–20%) and in pregnant females.

Hepatitis F

Researchers claim to have identified HGF virus, which is spread enterically.[18] Treatment can be comfortably managed within general practice.

Hepatitis G

HGV has been identified as a transfusion-spread virus. It has subsequently been found to be prevalent among Queensland blood donors.[10,19]

Cholestatic jaundice

Cholestasis refers to the syndrome of biliary obstructive jaundice whereby there is obstruction to the flow of bile from the hepatocyte to the duodenum, thus causing bilirubin to accumulate in the blood. It is classified into two main groups:

- intrahepatic cholestasis—at the hepatocyte or intrahepatic biliary tree level
- extrahepatic cholestasis—obstruction in the large bile ducts by stones or bile sludge

The significant causes are listed in TABLE 47.8 .

Table 47.8 Significant causes of cholestasis in adults

Intrahepatic
Alcoholic hepatitis/cirrhosis
Drugs
Primary biliary cirrhosis
Viral hepatitis

Extrahepatic

Cancer of bile ducts

Cancer of pancreas

Other cancer: primary or secondary spread

Cholangitis

Primary sclerosing cholangitis (?autoimmune)

IgG_4-related disease

Common bile duct gallstones

Pancreatitis

Postsurgical biliary stricture or oedema

Symptoms

- Jaundice (greenish tinge)
- Dark urine and pale stools
- Pruritus—worse on palms and soles
- Pain varies from nil to severe

Gallstones and jaundice

Gallstones can be found in the following (see FIG. 47.8):

- gall bladder (asymptomatic up to 75%)—the majority remain here
- neck of gall bladder (biliary ‘colic’ or acute cholecystitis)
- cystic duct (biliary ‘colic’ or acute cholecystitis)
- common bile duct—may cause severe biliary ‘colic’, cholestatic jaundice or cholangitis

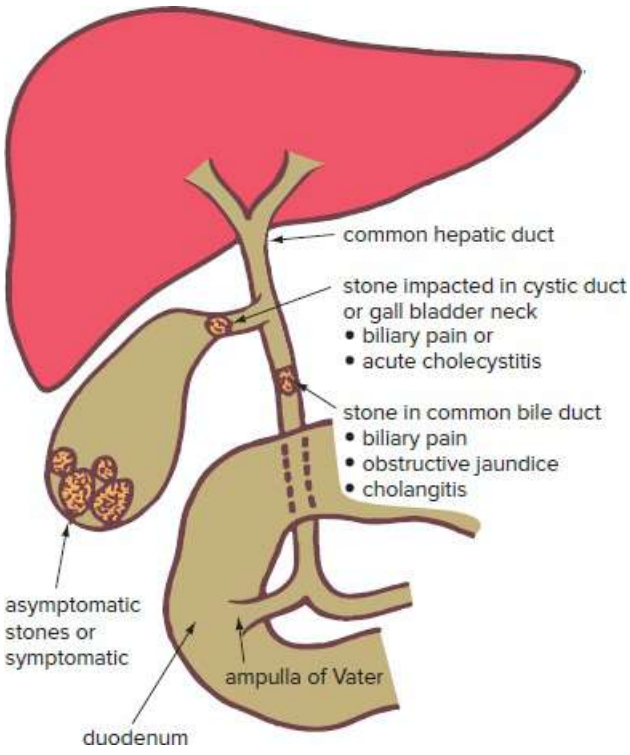


FIGURE 47.8 Clinical presentation of gallstones

Acute cholecystitis is accompanied by mild jaundice in 25% of cases, due to accompanying common duct stones.[17]

Common bile duct stones may be asymptomatic or may present with any one or all of the triad of abdominal pain, jaundice and fever. The jaundice varies, depending on the amount of obstruction. The liver is moderately enlarged if the obstruction lasts for more than a few hours.

The investigations of choice for cholestatic jaundice are ultrasound and ERCP.

Acalculous cholecystitis

- Acute—in very ill patients often with diabetes.
- Chronic—associated with polyps, sludge or inflammation.

Acute cholangitis

This is due to bacterial infection of the bile ducts secondary to abnormalities of the bile duct, especially gallstones in the common duct. Other causes are neoplasms and biliary strictures.

Charcot triad (present in 70%) is shown in the diagnosis box.

DxT fever (often with rigor) + upper abdominal pain + jaundice → acute cholangitis

Older patients can present with circulatory collapse and Gram-negative septicaemia. Urgent referral is necessary.

Carcinoma of head of the pancreas

Pancreatic cancer is the fourth commonest cause of cancer death in the UK and US.[17]

Clinical features

- M > F
- Mainly >60 years of age
- Obstructive jaundice
- Pain (over 75%)—epigastric and back
- Enlarged gall bladder (50–75%)

Possible features

- Weight loss, malaise, diarrhoea
- Migratory thrombophlebitis
- Palpable hard, fixed mass
- Metastases (e.g. left supraclavicular gland of Virchow—Troisier sign)
- Occult blood in stool
- Glycosuria

Diagnosis

- Scanning by ultrasound or CT scan may show mass
- ERCP

DxT jaundice + constitutional symptoms (malaise, anorexia, weight loss) + epigastric pain (radiating to back) → pancreatic cancer

Prognosis

Prognosis is poor: 5-year survival rates vary from 5–11%.

Cirrhosis of the liver

Cirrhosis is accompanied by jaundice as a late and serious manifestation with the exception of primary biliary cirrhosis, where jaundice appears before advanced liver failure. The development of jaundice usually indicates that there is minimal hepatic reserve and is therefore found in conjunction with other signs of liver failure (see FIG. 47.9).

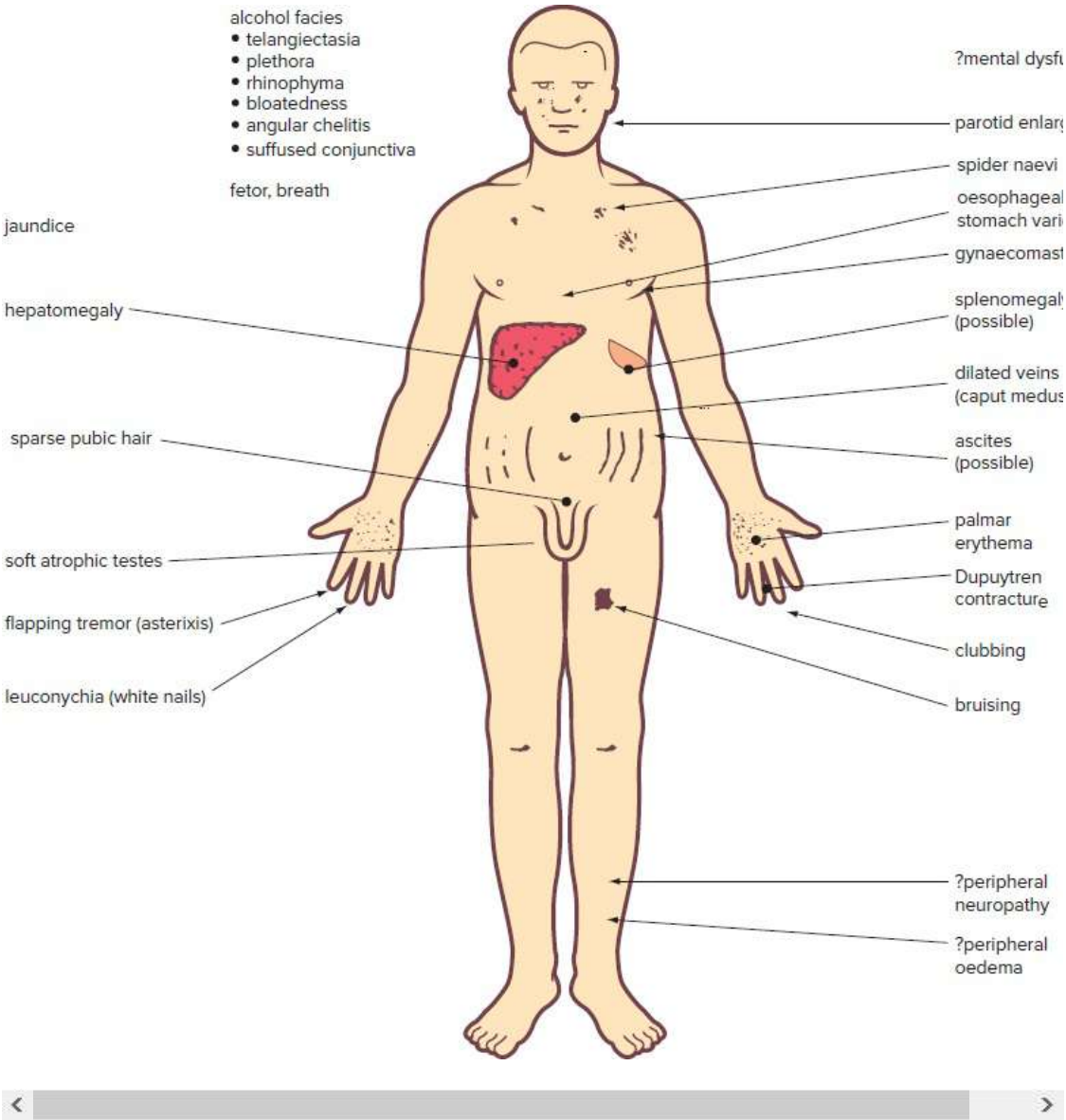


FIGURE 47.9 Possible features of chronic alcoholic liver disease

Causes

Common:

- alcohol excess
- chronic viral hepatitis (esp. HBV, HCV)

Others:

- autoimmune chronic active hepatitis
- primary biliary cirrhosis (autoimmune)
- haemochromatosis
- Wilson disease
- drugs (e.g. methotrexate)
- cryptogenic (no cause found)

Clinical features

- Anorexia, nausea ± vomiting
- Swelling of legs
- Abdominal distension
- Bleeding tendency
- Drowsiness, confusion or coma (if liver failure)

Signs

- Spider naevi (distribution of superior vena cava)
- Palmar erythema of hands
- Peripheral oedema and ascites
- Jaundice (obstructive or hepatocellular)
- Enlarged tender liver (small liver in long-term cirrhosis)
- Ascites
- Gynaecomastia

- ± Splenomegaly (portal hypertension)

Investigations

- FBE
- Fibroscan
- ± Biopsy

Complications

- Ascites
- Portal hypertension and GIT haemorrhage
- Portosystemic encephalopathy
- Hepatoma
- Kidney failure

Autoimmune chronic active hepatitis (ACAH)[5]

Also termed idiopathic ACAH, this usually affects young females (10–40 years) who present insidiously with progressive fatigue, anorexia and jaundice. Diagnosis is made by abnormal LFTs (which should raise suspicion), positive smooth muscle antibodies, a variety of other autoantibodies and a typical liver biopsy. If untreated, most patients die within 3–5 years. Treatment is with prednisolone orally, monitored according to serum alanine aminotransferase levels, and supplemented with azathioprine or mercaptopurine. About 80% respond, while 20% develop chronic liver disease. Specialist referral is advisable.

Primary sclerosing cholangitis[5]

This uncommon inflammatory disorder of the biliary tract presents with progressive jaundice and other features of cholestasis such as pruritus. It is often associated with ulcerative colitis. Diagnosis is based on characteristic cholangiopancreatography findings. There is no specific therapy, but refer for possible ERCP. Ursodeoxycholic acid may benefit some patients. Patients have an increased risk of colorectal cancer.

Primary biliary cirrhosis[5]

This is an uncommon cause of chronic liver disease that often presents with pruritis, malaise and an obstructive pattern of liver biochemistry. Found mainly in women. Treatment is with ursodeoxycholic acid orally.

Alcoholic liver disease

The main effects of alcohol excess on the liver are:

- acute alcoholic liver disease
- fatty liver
- alcoholic hepatitis (progresses to cirrhosis if alcohol consumption continues)
- alcoholic cirrhosis

If diagnosed, patients are advised to stop drinking alcohol for life except for fatty liver when small amounts can be drunk later.

Fatty liver

Alcohol can cause hepatic steatosis (fatty liver), which is almost universal in obese alcoholics. Non-alcoholic causes include obesity, diabetes mellitus, hypertriglyceridaemia and corticosteroids. A significant number with this very common condition (one in five Australians) will develop cirrhosis. Fatty liver is usually asymptomatic but some complain of malaise and tiredness. Serology is unhelpful. Diagnosis is by liver biopsy and perhaps CT scan. The treatment is weight loss through diet, which improves liver function and reduces fatty deposits.

Haemochromatosis

See CHAPTER 23 .

Special patient groups

The returned overseas traveller

The overseas traveller presenting with jaundice may have been infected by any one of the viruses —hepatitis A, B, C, D or E. All are prevalent in developing countries, especially in south-eastern and eastern Asia, some Pacific islands and Africa.

Other causes to consider are malaria, ascending cholangitis and drug-induced hepatic damage due to, for example, the antimalarials, including mefloquine (Lariam) and Fansidar. Refer to CHAPTER 129 .

Jaundice during pregnancy

Important hepatic disorders in pregnancy leading to jaundice are intrahepatic cholestasis of pregnancy, acute fatty liver of pregnancy and severe pre-eclampsia. Delivery is advisable at 37–38 weeks.

Postoperative jaundice

There are many possible causes of postoperative jaundice either in the immediate or the long-term postoperative phase. Hypoxia associated with shock in a severely ill patient or in a patient with cardiopulmonary disease may lead to transient abnormalities in liver function. Other causes include:

- post-transfusion hepatitis
- coincident viral hepatitis
- drugs, including anaesthetics
- transfusion overload (haemolysis)
- sepsis
- unmasked chronic liver disease and biliary tract disease
- cholestasis: post major abdominal surgery

Neonates of HBeAg positive mothers

These neonates should have the following (see CHAPTER 101):

- hepatitis B immunoglobulin IM within 24 hours of birth
- hepatitis B vaccine at birth, 1 month and 6 months

The vaccine may fail because some infants can be infected in utero.

When to refer

- All patients with fulminant hepatitis
- All patients with chronic liver disease or cirrhosis
- Painless obstructive jaundice
- Evidence of malignancy
- Symptomatic gallstones
- Acute fatty liver of pregnancy (very urgent)
- Suspected rare conditions (e.g. Wilson syndrome)

Practice tips

- All drugs should be suspected as potential hepatotoxins.
- With hepatitis A, the presence of IgM antibodies reflects recent infection, and IgG antibodies indicate past infection and lifelong immunity.
- There is no chronic carrier state of hepatitis A and E.
- All patients with jaundice should be tested for hepatitis B surface antigen (HBsAg).
- Hepatitis B infection is usually benign and short-lived, but it can be fatal if chronic hepatitis develops, which may lead later to cirrhosis and hepatocellular carcinoma.
- Up to 5% of patients with hepatitis B will become chronic carriers (especially users of IV drugs).
- Such carriers are identified by persistent titres of HBsAg and possibly HBeAg, the latter indicating the presence of the whole virus, and active replication and high infectivity.
- A raised gamma-glutamyl transferase accompanied by a raised MCV is a good screening test for alcohol abuse.
- A systolic murmur may be heard over the liver in alcoholic hepatitis and hepatoma.
- A distaste for smoking (with jaundice) suggests acute viral hepatitis.
- Life expectancy is short with ascites.

Patient education resources

Hand-out sheets from *Murtagh's Patient Education* 8th edition:

- Fatty liver
- Gallstones
- Hepatitis A
- Hepatitis B
- Hepatitis C

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48 Nasal disorders

The face of Mrs Gamp—the nose in particular—was somewhat red and swollen, and it was difficult to enjoy her society without becoming conscious of the smell of spirits.

CHARLES DICKENS (1812–1870), *MARTIN CHUZZLEWIT*

Disorders of the nose, which include the everyday problems of rhinitis, postnasal drip, epistaxis, folliculitis and disorders of smell, are very common in everyday general practice.

The main functions of the nose are:

- airflow
- filtration—of dust, organisms and other air-borne particles
- olfaction (smell)
- self-cleansing and moisturising of the mucous membrane
- humidification and warming of air in its passage to the lungs
- vocal resonance

The main symptoms of nasal disorders are discharge, blockage, sneezing, anosmia, itching, postnasal drip, bleeding and snoring (see TABLE 48.1).[1]

Table 48.1 Typical symptoms for nasal disorders[2]

Foreign body	Unilateral discharge, unilateral blockage
Acute sinusitis	Facial pain, toothache, nasal discharge, postnasal drip
Allergic rhinitis	Sneezing, rhinorrhoea, itch, eye irritation
Infective rhinitis	Blockage, purulent discharge, postnasal drip
Deviated septum	Blockage, postnasal drip

Nasal polyps	Blockage, reduced sense of smell
Nasal tumour	Blockage, unilateral discharge, epistaxis
Adenoidal hypertrophy	Bilateral blockage, snoring, halitosis
Nasal vestibulitis	Local pain, crusting, malodour

Nasal discharge is a common and important symptom to evaluate. The characteristics of nasal discharge are summarised in TABLE 48.2 .

Table 48.2 Characteristics of nasal discharge

Nature of discharge	Think of
Blood	Neoplasia, trauma, bleeding disorder, rhinitis, infection, hypertension
Mucopurulent	Bacterial rhinitis, foreign body
Serosanguineous	Neoplasia, foreign body
Watery/mucoid	Viral rhinitis, allergic rhinitis, vasomotor rhinitis, CSF

A major presenting problem is nasal obstruction with the complaint of a blocked or ‘stuffy’ nose. In those without a current URTI, common causes are physiological (the nasal cycle), rhinosinusitis (allergic or non-allergic), polyps, adenoid hypertrophy and mechanical such as septal deformity.

Red flag pointers for nasal disorders

- Unilateral nasal ‘polyp’
- Unilateral bloodstained discharge
- Toddler with offensive nasal discharge, esp. unilateral
- Post-traumatic periseptal swelling
- Rhinitis medicamentosa
- Chronic sinusitis + LRTI = ?Wegener granulomatosis

Disorders of smell

The basic sense of smell is detected in the olfactory region by the olfactory nerve (cranial nerve I) while irritant sensors in the nose, mediated by the maxillary branch of the trigeminal nerve (cranial nerve V), detect some noxious odours.

The disorders can be classified as:[3]

- anosmia—no smell
- hyposmia—reduced smell
- hyperosmia—increased sensitivity to odours
- dysosmia—distortion of smell perception

    cacosmia—normal odours seem foul or unpleasant

    parosmia—a perverse sense of smell

Disorders of smell can be caused by conductive or sensorineural disturbances, part of normal ageing or considered as idiopathic (see TABLE 48.3). Conductive disorders present as anosmia or hyposmia, while sensorineural disorders can present with all of the above disorders.[3] Most cases of idiopathic anosmia are considered to be viral neuropathies and may last from a few days to several months. Head trauma, which can cause conductive or sensorineural disturbances, is considered to be caused either from a fracture of the skull involving the cribriform plate or, more commonly, by posterior head trauma. Some patients will never recover their sense of smell. There is no effective treatment. Those with anosmia lack flavour discrimination and often have accompanying loss of sense of taste. They are also vulnerable to an unawareness of smoke, gas, dangerous chemicals and unhealthy food.

Table 48.3 Causes of reduced sense of smell

Conductive defects
Head trauma
Nasal polyps
Septal deviation
Rhinitis and sinusitis
Rare (not to be missed)
Nasal tumour
Wegener granulomatosis
Central/sensorineural defects
Ageing

Chemicals (e.g. benzene, chlorine, formaldehyde, cement dust)
Cigarette and other smoking/inhalation
Drugs
Endocrine disorders (e.g. diabetes, hypothyroidism)
Frontal lobe tumour
Parkinson disease
Head trauma
Kallmann syndrome (anosmia + hypogonadism)
Nutritional deficiencies
Viral infections

The clinical approach

- History: head injury or surgery, recent URTI, drugs, occupation including chemical exposure
- Physical examination, including inspection via a Thudicum nasal speculum
- Sniff test—qualitative and quantitative odours (e.g. coffee, cloves, lemon, peppermint, water placebo). Ammonia (for irritant sensation)
- Investigations (e.g. CT scan for sinus disease, nasal polyps)

Treatment of anosmia[2,4]

- Explanation and reassurance
- Education about smoke detectors, caution about chemicals including gas, excessive perfume, food safety including milk and meat contamination
- Consider dietary supplement (weak supportive evidence) with daily zinc sulphate, vitamin A and thiamine

  For chronic anosmia following an URTI: prescribe a nasal decongestant such as Spray-Tish Menthol for 5–7 days

Rhinitis

Rhinitis is inflammation of the nose causing sneezing, nasal discharge or blockage for more than an hour during the day. Rhinitis is subdivided into various types:

- According to time span:

  seasonal rhinitis: occurs only during a limited period, usually springtime

  perennial rhinitis: present throughout the year

- According to pathophysiology:

  allergic rhinitis: an IgE-mediated atopic disorder

  vasomotor rhinitis: due to parasympathetic overactivity

Both allergic and vasomotor rhinitis have a strong association with asthma.

The classification can be summarised as:

- seasonal allergic rhinoconjunctivitis = hay fever
- perennial rhinitis

  allergic (usually due to house dust mites)

  non-allergic = vasomotor: eosinophilic, non-eosinophilic

Note: Allergic rhinitis (hay fever) is presented in detail in CHAPTER 72 .

Clinical features

- Nasal symptoms:

  sneezing

  nasal obstruction and congestion

  hypersecretion—watery rhinorrhoea, postnasal drip

  reduced sense of smell

  itching nose (usually allergic)

- Throat symptoms:

  dry and sore throat

  itching throat

- Irritated eyes (allergic)
- Abnormal nasal mucous membrane—pale, boggy, mucoid discharge. A transverse nasal

crease indicates nasal allergy, especially in a child.

Allergens

- Pollens from trees (spring) and grass (in summer)
- Moulds
- House dust mites (perennial rhinitis)
- Hair, fur, feathers (from cats, dogs, horses, birds)
- Some foods (e.g. cow's milk, eggs, peanuts, peanut butter)

Diagnosis

Allergic rhinitis—nasal allergy:

- detection of allergen-specific IgE antibodies (not specific)
- RAST test or skin testing for specific allergens (can get false negatives)

Vasomotor rhinitis—a diagnosis of exclusion.

Other causes of rhinitis

- Chronic infection (viral, bacterial, fungal)
- Rhinitis of pregnancy
- Rhinitis medicamentosa—following overuse of OTC decongestant nasal drops or oxymetazoline sprays
- Drug-induced rhinitis:

    various antihypertensives

    aspirin

    phenothiazines

    oral contraceptives

    cocaine, marijuana

- Chemical or environmental irritants (vasomotor rhinitis):

    smoke and other noxious fumes

paints and sprays

cosmetics

Factors aggravating rhinitis (vasomotor)

- Emotional upsets
- Fatigue
- Alcohol
- Chilly, damp weather
- Air-conditioning
- Sudden changes in temperature and humidity

Rhinosinusitis

Acute sinusitis

Acute sinusitis is acute inflammation in the mucous membranes of the paranasal sinuses. About 5% of URTIs are complicated by an acute sinusitis,[4] which is mainly viral initially, while secondary bacterial infection can follow. Any factor that narrows the sinus openings into the nasal cavity (the ostia) will predispose to acute sinusitis.

The two prime clinical presentations are:

1. an URTI persisting for longer than 10 days
2. an URTI that is unusually severe with pyrexia and a purulent nasal discharge

Refer to CHAPTER 41 for features of acute maxillary sinusitis. Note that antibiotics are usually not helpful.

Chronic sinusitis

Chronic sinusitis is the most common complication of acute sinusitis. In chronic sinusitis, the symptoms and signs of inflammation persist for more than 8–12 weeks and are more likely to be associated with factors that impair drainage via the osteomeatal complex, including nasal polyps.

Treatment[5]

- Intranasal irrigation with saline

- Trial antihistamine nasal spray
- Add in (or switch to, first or second line) intranasal corticosteroid spray
- While awaiting surgical options, pain may be relieved by oral prednisolone

If the above therapies are ineffective, a mechanical saline sinus irrigation procedure to remove stagnant mucus is beneficial.[6]

Refer:

- for surgical drainage if there is no response to the above regimen
- those with orbital or facial cellulitis (refer urgently)

Nasal polyps

Nasal polyps are round, soft, pale, pedunculated outgrowths arising from the nasal or sinus mucosa. They are basically prolapsed, congested, oedematous mucosa, described by some as ‘bags of water’ (see FIG 48.1). They occur in patients with all types of rhinitis, but especially in allergic rhinitis (see FIG. 48.2). Polyps usually arise from the middle meatus and turbinates.

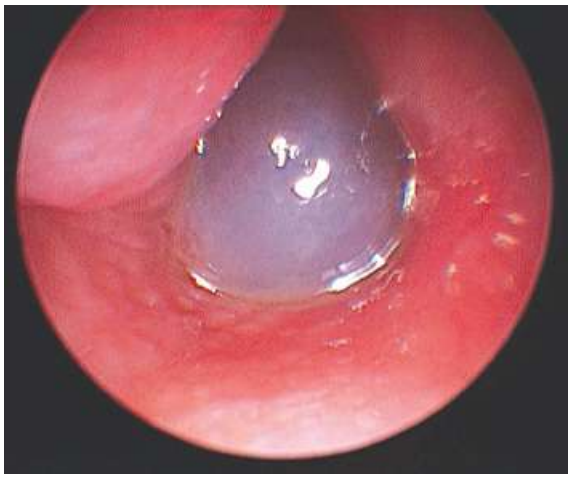

FIGURE 48.1 Nasal polyp in right nasal cavity in a patient with inflamed mucosa from allergic rhinitis

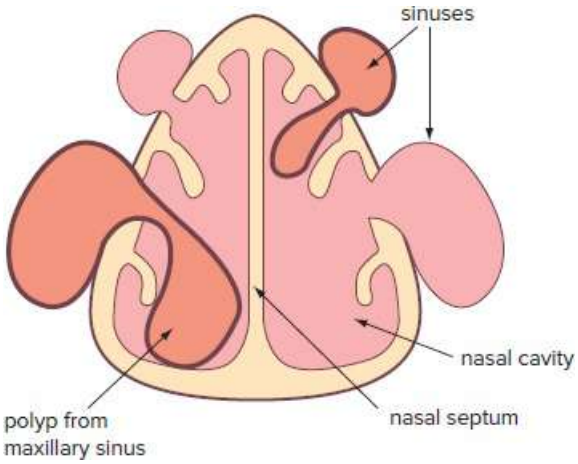

FIGURE 48.2 Transverse cross-section of nose, demonstrating origin of nasal polyps

Symptoms include nasal obstruction, watery discharge, postnasal drip and loss of smell.

Note:

- Nasal polyps may be associated with asthma and aspirin sensitivity.
- Cystic fibrosis should be considered in any child with nasal polyps.
- A polyp that does not have the typical smooth, pale appearance may be malignant.
- A unilateral ‘polyp’ may be a neoplasm.
- If there is a purulent discharge, swab and give antibiotics.

Treatment

The initial treatment should be medical.[7] A medical ‘polypectomy’ can be achieved with oral steroids, for example, prednisolone 50 mg daily for 7 days. Supplement this with a corticosteroid spray (betamethasone, fluticasone, budesonide, mometasone) starting simultaneously and continuing for at least 3 months.[8] Give antibiotics for any purulent nasal discharge.

Simple polyps can be readily snared and removed, but referral to a specialist surgeon is advisable for surgical intervention since the aim is to remove the polyp with the mucosa of the sinuses (often ethmoidal cells) from which it arises. This complex procedure reduces the incidence of recurrence.

Epistaxis[9]

This common emergency should, in some instances, be treated as a life-threatening problem. The

common situation is intermittent anterior bleeding from Kiesselbach plexus located in Little area, seen in children and the young adult (90% of episodes), while posterior epistaxis (10%) is more common in the older hypertensive patient. It has a strong association with higher URTI (rhinitis, sinusitis), hot dry climates and trauma. Neoplasms should be kept in mind. Bleeding often occurs at night due to vascular vasodilation. Causes of epistaxis are presented in the diagnostic strategy model in TABLE 48.4 . The secret of good management is to have the right equipment, good lighting and effective local anaesthesia.

Table 48.4 Epistaxis: diagnostic strategy model

Probability diagnosis

Idiopathic: spontaneous from Little area

URTI: common cold, influenza, sinusitis

Rhinitis

Vestibulitis

Trauma (incl. nose picking, nose injury)

Drugs (e.g. anticoagulants, aspirin)

Serious disorders not to be missed

Vascular:

- hypertension and arteriosclerosis

Infection:

- systemic febrile illness (e.g. malaria)
- HIV/AIDS

Cancer/neoplasia:

- tumours of nose/sinuses/nasopharynx
- intracranial tumours
- leukaemia

Other:

- thrombocytopenia
- coagulopathy (e.g. haemophilia, liver disease)

Pitfalls (often missed)

Exposure to toxic agents

Vitamin deficiencies: C and K

Septal granulomas and perforations

Foreign bodies (esp. in children)

Cocaine abuse

Rarities:

- hereditary haemorrhagic telangiectasia

Diagnostic tips

- Recent onset of persistent bleeding in elderly points to carcinoma.
- Severe epistaxis is often caused by liver disease coagulopathy.
- Difficult-to-control posterior bleeding is a feature of the hypertensive elderly.

Ideal equipment

Head light, Thudicum nasal speculum, Tilley nasal packing forceps, suction cannula and tubing, Co-Phenylcaine forte spray ± 5% cocaine solution.

Tamponade options (for difficult bleeding)

Merocel expandable or dental pack, Kaltostat, BIPP (bismuth iodoform paraffin paste) with ribbon gauze, dental pack, Foley catheter (no. 12, 14 or 16) with a 30 mL balloon and self-sealing rubber stopper, anterior/posterior balloon, Epistat catheter with or without Kaltostat. Or instead, many retrieval services stock a (rather expensive) Rapid Rhino nasal device for posterior or uncontrolled anterior bleeds.

Treatment

First line is to clear clots—blow the nose and then apply 5–6 sprays of a decongestant nasal spray, e.g. Drixine.

Simple tamponade:

- Pinch ‘soft’ part of nose below the nasal septum between thumb and finger for 5 or up to 20 minutes
- Apply ice packs to bridge of nose
- Another simple method is to insert a cotton wool ball soaked in lignocaine with adrenaline or other decongestant

Simple cautery of Little area (see FIG. 48.3) (under local anaesthetic, e.g. Co-Phenylcaine forte nasal spray ± 5% cocaine solution):

- Use one of three methods: electrocautery, trichloracetic acid or silver nitrate stick (preferred). Beware of silver nitrate stains. Apply petroleum jelly to cauterised area.

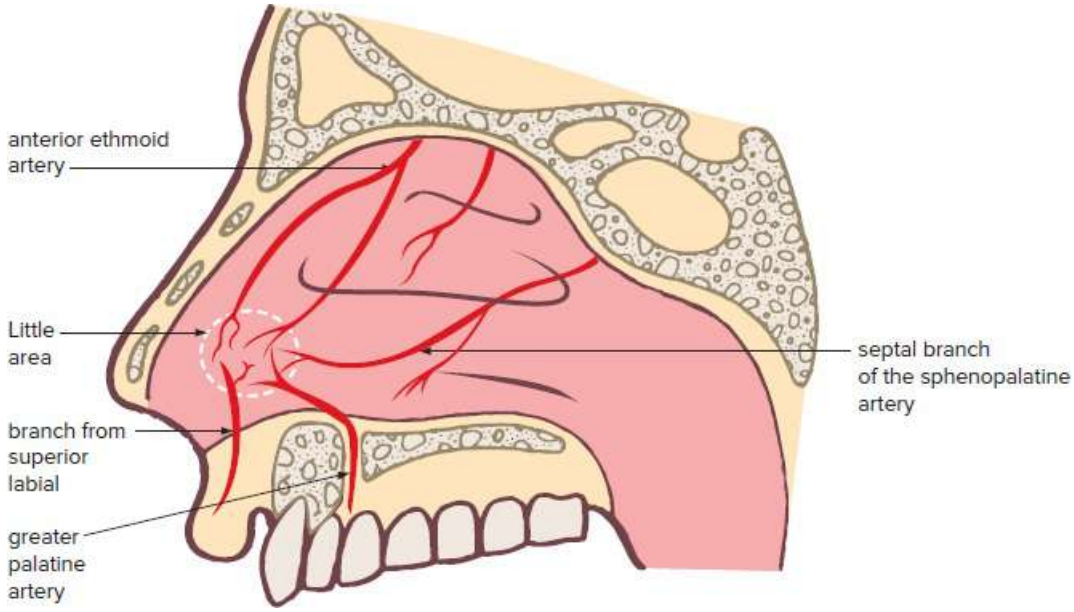

FIGURE 48.3 Little area on the nasal septum where several blood vessels anastomose. Bleeding is common here, especially in young people.

Persistent anterior bleed

Merocel (surgical sponge) nasal tampon or Kaltostat pack.

Tranexamic acid can be effective, given orally or intravenously. Two RCTs have also demonstrated the effectiveness of topical application, which is of even lower risk and gives more immediate results.[10] Pour an ampoule (if none avail, dissolve capsule contents) of tranexamic acid onto ribbon gauze and insert nasally.

‘Trick of the trade’ for intermittent minor anterior epistaxis:

topical antibiotic (e.g. Aureomycin ointment) bd or tds for 10 days

or (better option)

Nasalate nasal cream tds for 7–10 days

or

Rectinol ointment or Vaseline

Avoid digital trauma and nose blowing.

Severe posterior epistaxis

Use a Foley catheter or an Epistat catheter plus anterior pack or the Rapid Rhino nasal pack.

Nasal vestibulitis[11]

Infection of the nasal vestibule can cause a tender, irritating, crusty problem. First-line simple treatment is the daily application of sesame seed oil (e.g. Fess, Flo, Nozoil) or a thin smear of petroleum gel (Vaseline). Low-grade infections and *folliculitis*, which are evident on inspection, cause localised pain, crusts and bleeding, especially if picked from habit. Treatment is with bacitracin or preferably mupirocin (intranasal) ointment topically for 5–7 days; apply with cotton bud.

Furunculosis of the nasal vestibule is usually due to *Staphylococcus aureus.* It starts as a small superficial abscess in the skin or the mucous membrane and may develop into a spreading cellulitis of the tip of the nose. The affected area becomes tender, red and swollen. It is best treated by avoiding touching it, hot soaks and systemic antibiotics such as dicloxacillin or as determined by culture from swabs of the vestibule.

Tip: Staphylococcus aureus colonises the nose of 20–30% of the population.[8] Carriers are prone to transmit nosocomial infection and have an increased risk of serious infections in the presence of serious medical disorders. Treatment includes strict hygiene and eradication with an agent such as mupirocin ointment (match-head size) 2–3 times daily for 5–7 days (max. 10 days).[6]

Fissure: Painful fissures often develop at the mucocutaneous junction. They may become crusted and chronic. Fissures can be treated by keeping the area moist with petroleum jelly (Vaseline) or saline gel, using hot compresses and the use of an antibiotic or antiseptic ointment if necessary.

Offensive smell from nose

This may be caused by vestibulitis but ensure no foreign body is present.

Treatment

- Take nasal swab for culture.

  consider mupirocin 2% nasal ointment, instilled 2–3 times a day for 10 days

  or

  Kenacomb ointment, instil 2–3 times a day

Rhinophyma

This disfiguring swelling of the nose is due to hypertrophy of the nasal sebaceous glands. There is no causal association with alcohol; the visible facial vasodilatory effects of acute intoxication

may perhaps have supported this belief. Rhinophyma is almost exclusive to men over the age of 40 years, often associated with rosacea.

Treatment

- Good control of rosacea may reduce the risk (see CHAPTER 113).
- If surgical correction is warranted, refer to a specialist.
- Carbon dioxide laser therapy is the treatment of choice.
- Shave excision is another effective therapy.

Nasal septal deviation

This causes blockage as a solitary symptom. Mild septal deviation tends to cause alternating blockage while severe deviation causes persistent blockage on one side.

The septum can be divided into anterior and posterior segments. The anterior portion is necessary to support the cartilaginous pyramid of the nose whereas the posterior portion has no supporting role and can be removed without disturbing the support of the nose. The classic submucous resection operation is therefore suitable for posterior septal deviations. Repair of anterior septal deviations is more complex.

Nasal cosmetic surgery

Rhinoplasty is undertaken to improve the function of an obstructed nasal airway or for cosmetic reasons. In counselling for rhinoplasty it is important to undertake careful planning with realistic anticipated outcomes. The GP should provide non-judgmental support for the patient's decision on cosmetic surgery before referral to an expert in rhinoplasty. Each case has to be assessed individually and the surgery tailored to the deformity. Surgical attention to the airway is important, otherwise the nose may become partially obstructed and stuffy after cosmetic surgery alone.

Septal perforation

A hole in the nasal septum is commonly caused by chronic infection, including tuberculosis, repeated trauma such as vigorous nose 'picking' or following nasal surgery. Rarely, it is encountered in syphilis, Wegener granulomatosis (CHAPTER 21) or overzealous medical cautery. It is a known occupational hazard, particularly among chrome workers, and is seen in drug users who sniff cocaine. In about 5–10% of cases, perforation is a result of malignant disease.[4] The condition may be asymptomatic depending on the cause, but there is often an irritating nasal crust and a whistling sound on nasal inspiration. It can be demonstrated by looking in one nares while a light is shone in the opposite one. The cartilaginous part is usually involved. The perforation can be closed using a silicone septal button.