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Head Injury

18

Must know Must do

Must know

Differences between primary and secondary brain injury
Classification of head injuries
How to assess a patient with a head injury and to compute the Glasgow Coma Scale
Understand the pathophysiology of raised intracranial pressure and its management
Causes and treatment of intracranial haemorrhage
Classification of facial fractures

Must do

Follow a patient with a severe head injury, including stay in the intensive care unit
Follow a patient with a minor head injury
Examine computed tomographic scans showing the different types of intracranial haemorrhage
Talk to a professional involved in rehabilitation of a head injury patient

Introduction

More than 1 million patients present to UK emergency departments every year as a result of a head injury, which is thus one of the most common conditions seen in hospital. The causes of head injury are many and varied, although it is common to see 'head injury' as the only description of the incident in hospital notes. Every effort must be made to discover its underlying cause, paying particular attention to:

- the likely speed of impact;
- any events that may have led to the injury (e.g. epilepsy, subarachnoid haemorrhage, alcohol consumption);
- any events after its occurrence (e.g. vomiting, epilepsy, talking).

Pathology

The brain within the skull is liable to injury when deceleration occurs, i.e. when the neck flexes, extends or rotates. As the brain moves within the cranial cavity, it may strike sharp objects such as the sphenoid wing and the frontal and occipital poles. In addition, points where the brain is tethered, such as the foramen magnum and the cranial nerves, are also potential sites of injury. Shaking of the brain when the skull moves at high speed therefore results in haemorrhage in the subarachnoid space and at the frontal, temporal and occipital poles and in tearing of nerves and vessels. This damage can occur without the head being struck, for example in a high-speed car crash or in a fall from a height in which the body decelerates rapidly. It may also be associated with direct damage from a blow or a penetrating wound.

Primary damage

The impact from a direct blow to the head is absorbed by the scalp and the skull, which often fractures. The energy is then transmitted to the brain, damaging the tissue it strikes and causing brain movement within the skull. This damage occurring at the time of impact is called primary damage or concussion. Survival depends on the energy reaching the brain, which in turn depends on the velocity of impact (energy = $^1/_2 m \times v^2$, where m is mass and v is velocity). Therefore hitting a brick wall at 70 mph (110 kph) will be fatal but a kick to the head playing rugby will probably not. Similarly, a shot from a high-velocity rifle will prove fatal but a shot from an air rifle will probably not. The mechanism is the same, the degree is different. Severe primary damage is characterized by coma from impact. The ability to talk, even a few simple words, after injury indicates that, whatever else may happen, the primary injury was not severe and the injury is theoretically survivable.

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Secondary damage

Primary damage can be exacerbated by secondary damage (i.e. further insults to the damaged brain). These are important to understand as they can often be prevented and occasionally reversed, whereas nothing can be done about primary damage (except avoid the accident in the first place). The main secondary effects are respiratory complications, perfusion failure, intracranial haematoma, cerebral swelling, epilepsy, infection and hydrocephalus.

Respiratory complications

Hypoxia, hypercarbia or obstruction to breathing will have disastrous effects on a damaged brain and can worsen the clinical picture dramatically. Head-injured patients are especially prone to respiratory problems because of lack of central drive, airway obstruction, haemothorax or pneumothorax and/or aspiration pneumonia. It cannot be overemphasized that the most important aspect of care in the head-injured patient is care of the chest.

Perfusion failure

Perfusion failure will rapidly lead to cerebral ischaemia and a worsening of the clinical state. Head injury itself is rarely a cause of hypotension and other causes need to be investigated (e.g. ruptured spleen). Resuscitation must be rapid and patients with head injury accompanied by a systolic blood pressure of less than 60 mmHg for more than a few minutes rarely survive.

Intracranial haematomata

Intracerebral, subdural and extradural haematomata occur and can lead to deterioration following head injury. *Intracerebral* and *acute subdural* haematomata are usually associated with severe primary injury and carry a bad prognosis. In contrast, *subacute subdural* and *extradural* haematomata are often associated with little or no primary injury and bleed slowly over a few hours or days; removal before intracranial pressure (ICP) is excessive can lead to complete recovery.

Cerebral swelling

If you sprain your ankle, it swells; if you injure your brain, it also swells. Because the brain is encased in a rigid box, cerebral swelling itself can cause damage by increasing ICP to levels at which cerebral perfusion fails. This leads to ischaemia, which in turn leads to more brain swelling and a further increase in ICP. This vicious circle tends to be worse in those with severe primary damage.

Epilepsy

Fits are common in head injury and cause ischaemia while they are occurring. They must be stopped rapidly using intravenous diazepam in small doses, followed by phenytoin or valproate to prevent recurrence.

Infection

The development of meningitis or an abscess after injury can reverse a good recovery and must be watched for and treated vigorously.

Hydrocephalus

An absorptive hydrocephalus can occur during recovery and may slow or reverse recovery; however, it is relieved by shunting.

Head injury at a glance

Definitions

Head injury: the process whereby direct or decelerating trauma to the head results in skull and brain damage

Primary brain injury: damage that occurs to the brain immediately as the result of the trauma

Secondary brain injury: damage that develops later as a result of complications

Epidemiology

Head injury is very common: 1 million patients each year present to accident and emergency departments in the UK with head injury and about 5000 patients die each year following head injuries

Pathophysiology

Direct blow

- May cause damage to the brain at the site of the blow (*coup injury*) or to the side opposite the blow when the brain moves within the skull and hits the opposite wall (*contrecoup injury*).

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Rotation/deceleration
- Neck flexion, extension or rotation results in the brain striking bony points within the skull (e.g. the wing of the sphenoid bone)
- Severe rotation also causes shear injuries within the white matter of the brain and brainstem, causing axonal injury and intracerebral petechial haemorrhages

Crush
- Brain often remarkably spared direct injury unless severe (especially in children with elastic skulls)

Missiles
- Tend to cause loss of tissue with injury proportionate. Brain swelling less of a problem because the skull disruption automatically decompresses the brain
- Degree of primary brain injury is directly related to the amount of force applied to the head
- Secondary damage results from respiratory complications (*hypoxia*, *hypercarbia*, *airway obstruction*), hypovolaemic shock (head injury does not cause hypovolaemic shock: look for another cause), intracranial bleeding, cerebral oedema, epilepsy, infection, hydrocephalus

Clinical features
- History of direct trauma to head or deceleration
- Patient must be assessed fully for other injuries (full trauma survey)
- Level of consciousness determined by GCS: fully conscious, GCS = 15; deep coma, GCS = 3
- Pupillary inequalities or abnormal light reflex indicate intracranial haemorrhage
- Headache, nausea, vomiting, a falling pulse rate and rising blood pressure indicate cerebral oedema

Glasgow Coma Scale
Provides a simple method of monitoring global CNS function over a period rather than a precise index of brain injury at any one time

Investigations
- Skull X-ray: AP, lateral and Towne's views
- CT/MRI: show contusions, haematomata, hydrocephalus, cerebral oedema

Eye opening		Voice response		Best motor response	
Spontaneous	4	Alert and orientated	5	Obeys commands	6
To voice	3	Confused	4	Localizes pain	5
To pain	2	Inappropriate	3	Flexes to pain	4
No eye opening	1	Incomprehensible	2	Abnormal flexion to pain	3
		No voice response	1	Extends to pain	2
				No response to pain	1

Classification

There are four types of head injury.
- *Trivial*: little or no primary damage, no secondary effects.
- *Apparently trivial but potentially fatal*: little or no primary damage but underlying secondary effect (e.g. extradural haematoma).
- *Apparently hopeless but potentially salvageable*: moderate to severe but recoverable primary damage and avoidable or reversible secondary effects.
- *Hopeless*: overwhelming primary injury.

Management

The aims of management are to:
- recognize and treat those patients with apparently trivial but potentially fatal injuries at an early stage before brain damage occurs;
- recognize those with hopeless injuries and to withdraw treatment at an early stage;
- treat those with potentially salvageable severe injuries adequately and rapidly minimize secondary effects and allow complete or partial recovery.

Trivial head injury

If the patient is conscious, a history of the events leading up to the accident and subsequent to it is obtained. If the patient has sustained a period of unconsciousness, it is helpful to obtain corroboration of this from witnesses. Retrograde amnesia for events leading up to the accident is a significant factor in the history. The duration of unconsciousness and retrograde amnesia are indicative of the severity of the injury; if very transient and in the absence of other indications, they need not be a criterion for admission and further observation.

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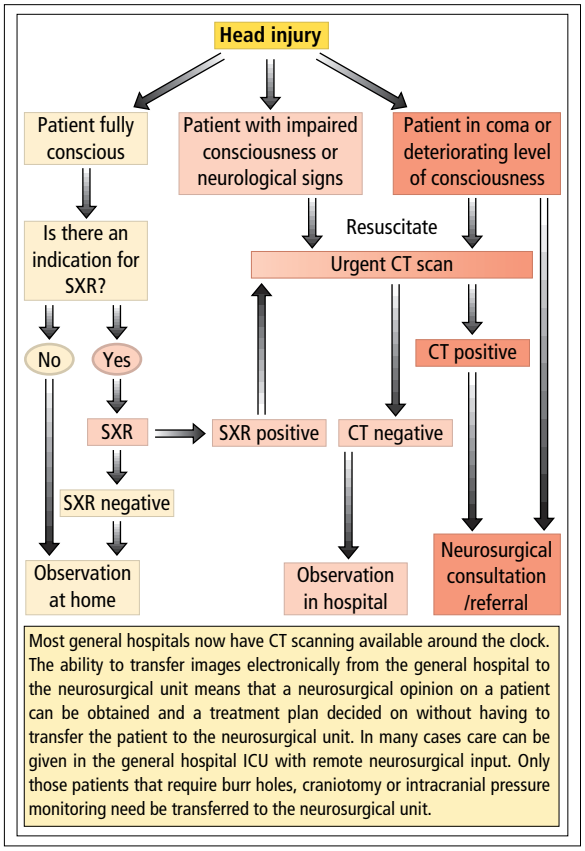

The questions to be asked include the following.

- Does this patient need a skull radiograph?
- Do I need to admit this patient to hospital?
- Do I need to refer this patient to a neurosurgeon?

These questions are easily answered by following guidelines laid down by a group of neurosurgeons under the auspices of the Kings Fund and first published by the Royal College of Surgeons in 1986. They have recently been updated and published in the *British Journal of Neurosurgery* (see Evidence-based medicine). The guidelines are shown in Table 18.1. The risk of a significant intracranial haematoma following head injury is 1 in 6000 among alert patients with no skull fracture, 1 in 120 among alert patients with a fracture and 1 in 4 among drowsy patients with a fracture.

An admitted patient needs regular neurological observation. Patients who suffered no loss of consciousness or who had no more than retrograde amnesia or brief loss of consciousness may be allowed home if they remain stable for 24 h. Simple fractures are not an indication for continued hospital stay. Compound fractures, i.e. associated with open scalp wounds or basal fractures with otorrhoea or rhinorrhoea, will need a more prolonged hospital stay.

Table 18.1 Society of British Neurological Surgeons' guidelines for initial management of head injuries (1998).

Indications for skull radiography* after recent head injury

(A) Oriented patient

History of loss of consciousness or amnesia
Suspected penetrating injury
Cerebrospinal fluid or blood loss from the nose or ear
Scalp laceration (to bone or > 5 cm long), bruise or swelling
Violent mechanism of injury
Persistent headache or vomiting
In a child, fall from a significant height (which depends in part on the age of the child) and/or on to a hard surface; tense fontanelle, suspected non-accidental injury

(B) Patient with impaired consciousness or neurological signs

All patients, unless urgent CT is performed

Indications for admission to a general hospital

(A) Oriented patient

Skull fracture or suture diastasis
Persisting neurological symptoms or signs
Confusion or any other depression of the level of consciousness at the time of examination
Difficulty in assessing the patient (e.g. due to alcohol, young age, epilepsy, attempted suicide)
Inadequate social conditions or lack of responsible adult or relative
Coexistence of other medical conditions (e.g. coagulation disorders)

(B) All patients with impaired consciousness

Indications for urgent CT scanning and/or neurosurgical consultation

Coma, confusion or neurological signs persisting after resuscitation
Deteriorating consciousness or progressive neurological signs
Fractured skull in combination with:
  confusion or other impairment of consciousness
  epileptic seizure
  neurological symptoms or signs
Open injury with:
  depressed compound fracture of skull vault
  fractured base of skull
  penetrating injury
Unstable systemic state precluding transfer to neurosurgery
Diagnosis uncertain
Tense fontanelle or suture diastasis in a child

Indications for neurosurgical consultation after CT scanning

Abnormal CT scan

High- or mixed-density intracranial lesion
Midline shift
Obliteration of third ventricle
Relative dilatation of lateral ventricle(s)
Obliteration of basal cisterns
Intracranial air
Subarachnoid or intraventricular haemorrhage

CT scan normal but progress unsatisfactory

* Skull radiography is not necessary if CT is to be performed.

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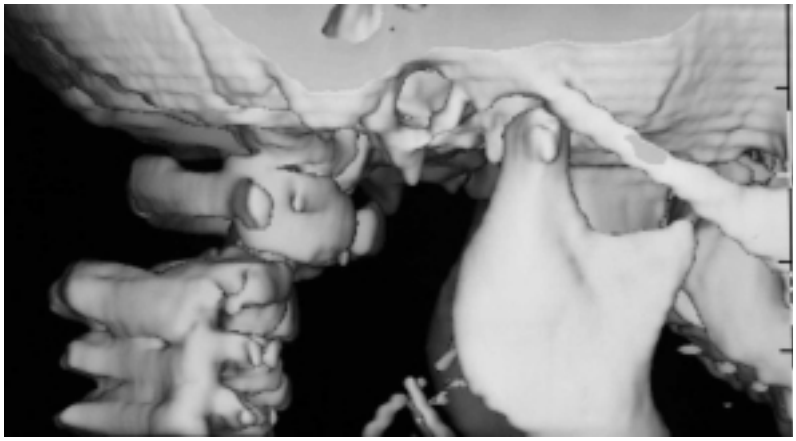

Figure 18.1 CT reconstruction of cervical spine C1/C2 fracture.

Traditionally prophylactic antibiotics have been given to these patients but are less commonly used nowadays. Persistent rhinorrhoea or otorrhoea requires surgical treatment to repair the skull base defect.

Following head injury, patients also need to be normally hydrated not fluid-restricted. An intravenous infusion should be used if the patient is too sleepy to drink or is vomiting.

The only other aspect of care is to deal carefully with any scalp wounds, remembering to:

- shave hair for at least 2 cm around the wound;
- remove foreign bodies;
- débride the skin edges;
- suture the scalp in a single layer, leaving stitches in for at least 7 days;
- use lidocaine with adrenaline to reduce bleeding.

Severe head injury

A person with a severe head injury will invariably arrive unconscious in the emergency department and the injury may be just one aspect of multiple trauma. It is important to stabilize the airway, breathing, circulation and exclude cervical fractures before concentrating on the head injury (Fig. 18.1). The management of head trauma is essentially the same whatever the cause and consists of intubation and ventilation, resuscitation, a thorough examination, imaging and decisions on treatment.

Intubation and ventilation

An unconscious patient with a head injury should be intubated to protect his or her airway. This usually involves sedation and paralysis and so ventilation will be required. There is no reason not to intubate. Traditional arguments against were that sedation prevented neurological monitoring of any deterioration and that intubation put at risk an unstable cervical spine. Nowadays, patients with severe head injuries undergo computed tomography (CT) so the first reason is less applicable and, although the cervical spine is also commonly involved in fatal head injuries, it is rarely injured in a survivable head injury. Having intubated and ventilated the patient, ensure the chest is moving properly, that there is no haemothorax or pneumothorax, and check blood gases to ensure adequate oxygenation.

Resuscitation

Insert one or more large-bore cannulae, a central venous pressure line if necessary, and a urinary catheter. Make sure that the patient's blood pressure and pulse are normal and that he or she has a good urine output. If there is bleeding, find its source and stop it. Stop any fits with intravenous diazepam and phenytoin.

Assess the level of consciousness

Once the airway has been established and cardiovascular stability achieved, the next priority is to assess the level of consciousness. Response to verbal and painful stimuli provides a simple evaluation, although more objective analysis can be obtained by using the Glasgow Coma Scale (GCS) (Table 18.2). Any depression of consciousness is an indication for admission for observation. Confusion may arise if the patient has been taking alcohol or other drugs but this increases rather than diminishes the necessity for careful observation.

Thorough examination

A full neurological and general examination is conducted to establish the presence or absence of focal neurological

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Table 18.2 Adult Glasgow Coma Scale.

	Score
Eye opening	
Spontaneous	4
To voice	3
To pain	2
No eye opening	1
Voice response	
Alert and orientated	5
Confused	4
Inappropriate	3
Incomprehensible	2
No voice response	1
Best motor response	
Obeys commands	6
Localizes pain	5
Flexes to pain	4
Abnormal flexion to pain	3
Extends to pain	2
No response to pain	1

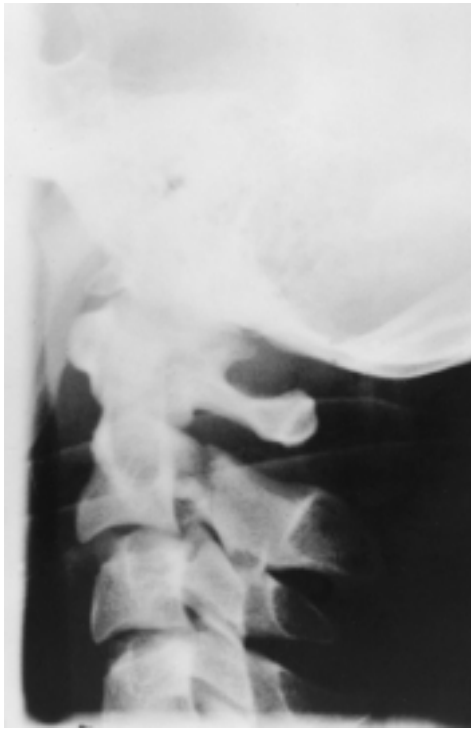

Figure 18.2 Radiograph of cervical spine showing fracture dislocation of the spine at C2/C3 level, so-called hangman's fracture.

signs. Pupillary inequalities or abnormal response to light are indicative of intracranial haemorrhage. A falling pulse rate and rising blood pressure are also indicative of increasing ICP. In the conscious patient, nausea and vomiting may suggest rising ICP due to bleeding. In the unconscious or semicomatose patient, vomiting may lead to aspiration into the air passage. Look at every part of the body. Pay particular attention to puncture wounds and the possibility of a spinal fracture.

Imaging

Accompanied by competent staff the patient should be imaged thoroughly. Skull (see Table 18.1), cervical spine (Fig. 18.2) and chest films are mandatory. If the patient has multiple injuries from a high-velocity injury, radiographs of the thoracic and lumbar spine, abdomen, pelvis and any suspected fractures should also be obtained. Nowadays, most multiple trauma patients will have CT of head, thorax and abdomen rather than radiography.

Treatment plan

There is no point in moving an unstable patient to another hospital for neurosurgery unless the instability is purely neurological. It is better to perform laparotomy or thoracotomy before transfer to stop bleeding and then to move the patient afterwards. Even if the patient is being transferred, spend a few minutes closing wounds and splinting fractures. Even if these procedures are crudely performed, it is better than not doing them at all, and they can be tidied up later.

Frequently, patients who have suffered a serious head injury but do not have a neurosurgically remediable lesion (e.g. an extradural haematoma) are managed in the intensive care unit (ICU) of the admitting hospital. CT scanning is performed in the admitting hospital and the images are relayed electronically to the neurosurgical unit via an image link system. Based on the CT findings the neurosurgeons provide advice to the admitting hospital regarding treatment; these patients usually have some degree of cerebral oedema. Such a system reduces unnecessary transfers to the neurosurgical unit yet involves the neurosurgeons in the management of the patient's head injury. Modern minimally invasive ICP monitors (e.g. Codman, Micro-sensor, Cimino) are being increasingly used in district general hospitals without neurosurgical facilities by anaesthetists or other surgeons.

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Transferring a neurosurgical patient

Some patients will have to be transferred to a neurosurgical unit for treatment. Indications for transfer include contusions, haematomata or hydrocephalus seen on CT scanning. Unconscious patients do not travel well and care must be taken to ensure the safest journey. The patient must be stable before the journey starts because treatment of a deteriorating patient in a moving ambulance or helicopter is very difficult. The most experienced doctor available, preferably an anaesthetist, and a nurse should travel with the patient. Lines and tubes must be well secured before setting out and adequate supplies of paralysing agents, sedatives and fluids must be taken.

Neurosurgical management

Neurosurgical management will depend on the CT findings. If a haematoma or hydrocephalus is evident, treatment with craniotomy or drainage is necessary. If no clots are shown, or after surgery, a decision must be made whether to ventilate the patient. Views differ about who should be ventilated; in the author's practice, patients with respiratory problems or those at risk of developing them are ventilated to prevent hypoxia.

Ventilation is also used along with sedation, diuretics and intermittent boluses of mannitol (an osmotic diuretic) to control ICP. Moderate hyperventilation works by reducing Pco_2, which in turn reduces cerebral blood flow, although this mechanism tends to be effective for only 48–72 h. ICP monitoring is essential in these circumstances and is easily performed using a variety of methods. The most widely used technique is to insert a fine intraparenchymal wire sensor through a small twist-drill hole in the skull. The kits come prepacked with sensors and guarded drills and can be connected to the normal ICU monitoring systems (e.g. Codman, Microsensor, Cimino). Normal ICP is less than 10 mmHg. An ICP higher than 20 mmHg is worrying and needs to be treated; an ICP above 40 mmHg denotes severe problems and is rarely associated with a survivable injury. Note that these values apply to steady ICP levels. Coughing or chest physiotherapy may produce transient pressures of 60 mmHg or more but return to baseline when the stimulus stops.

The patient is ventilated with full nursing care (Table 18.3) for periods determined at the onset of ventilation (e.g. 48 or 72 h). After this period, if there are no problems and the ICP is low, sedation is stopped to allow the patient to waken. If the process runs smoothly, all well and good; however, if the patient becomes distressed or the ICP increases excessively, ventilation should be restarted for another set period. As the patient begins to wake, he or she may be extubated as soon as the airway is secure. As soon as recovery begins, an active programme of physiotherapy, speech therapy and occupational therapy should be started to maximize rehabilitation.

Table 18.3 Nursing the unconscious patient.

Elevate head 30°, turn patient frequently
Monitor arterial blood pressure, blood gases, ICP, serum sodium, glucose and osmolality
Endotracheal intubation, controlled ventilation
Control ICP: ventilation, intermittent boluses of mannitol
Enteral feeding, 2500 kcal/day (10 500 kJ/day)
Ulcer prophylaxis: proton pump inhibitors

ICP, intracranial pressure.

Special problems

Children

Infants have a tendency to deteriorate dramatically after relatively minor injuries, becoming comatose and floppy and then recovering again rapidly (infant concussion syndrome). They also have a tendency to fit and may develop status epilepticus after minor trauma. Skull fracture and intracranial haematomata are less common in children than in adults. Non-accidental injury should always be borne in mind, especially when a child presents with acute subdural haematomata.

Depressed fracture

In a depressed fracture, bone is driven inwards and may penetrate the dura. CT scanning will show whether the dura is lacerated. Scalp lacerations over a depressed fracture must be closed urgently and the patient given antibiotics. He or she may then be transferred at leisure to a neurosurgical unit where a decision can be made regarding wound exploration. If there is a suspicion that the dura is lacerated, the fracture is always explored, although the indication for exploring a slightly depressed fracture is usually only cosmetic (Fig. 18.3).

Missile injuries

While high-velocity injuries are usually fatal, low-velocity injuries can be survivable. The patient should be resuscitated and transferred to neurosurgical care where the wound will invariably be explored to remove bone, skin and hair fragments. However, the missile itself can often be left undisturbed as the heat involved usually sterilizes it.

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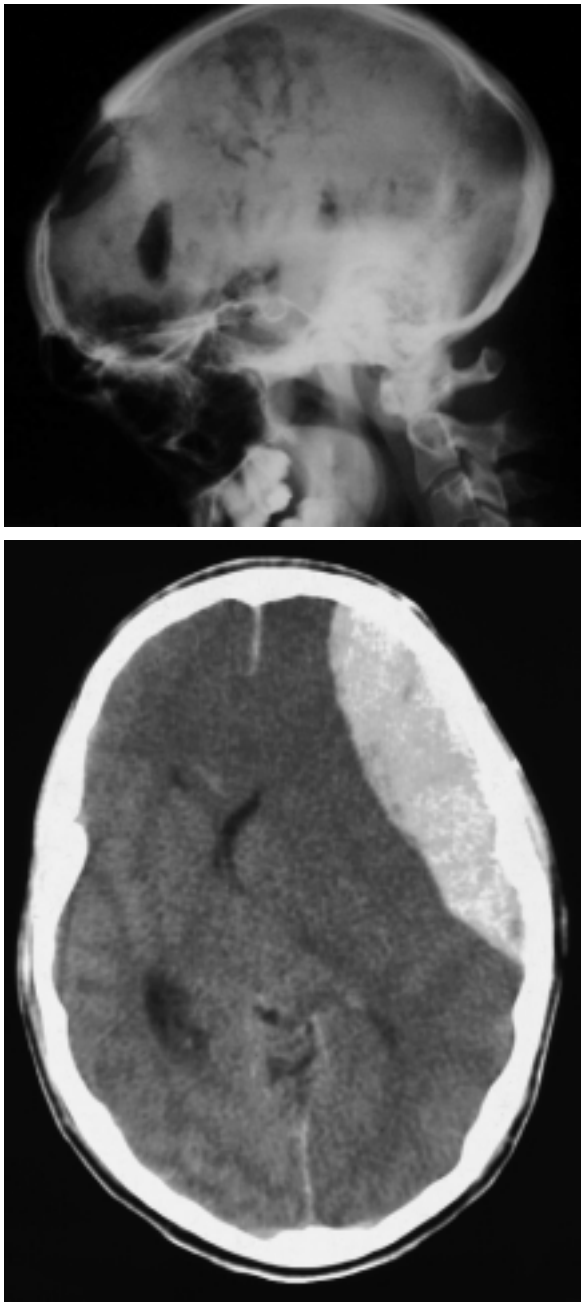

(a)

(b)

Figure 18.3 (a) Skull radiograph showing depressed fracture and air within the skull. (b) CT scan showing large extradural haematoma in the right frontal area with compression of the ventricles.

Basal fractures

Basal fractures may be accompanied by rhinorrhoea, periorbital haematoma and subconjunctival haemorrhage if the anterior cranial fossa is involved or otorrhoea if the middle cranial fossa is the site of injury. Subconjunctival haemorrhage, in which the blood tracks forwards from behind so that there is no posterior margin to the haematoma, is suggestive of anterior cranial fossa fracture. Direct injury to the eyeball produces a localized haematoma with visible margins. Periorbital bruising from direct injury is not confined by the orbicularis oculi, as is the case with intracranial (anterior fossa) injury.

Cerebrospinal fluid leak

Cerebrospinal fluid (CSF) leaks occur through the nose or ear, or silently down the back of the throat. Most clear up spontaneously within 2 weeks. Only a tiny percentage need surgical exploration to close the dural tear. Most neurosurgeons now advocate no treatment for CSF leaks, although traditionally broad-spectrum antibiotics (e.g. ampicillin and flucloxacillin) were given.

Cervical spine injury

Injury to the cervical spine may accompany a head injury and fractures and dislocations in this area are common after road traffic accidents and falls. If a patient complains of neck pain or weakness and/or numbness in the limbs or if there is loss of sphincter control, an injury to the cervical spine should be suspected. Precautions should be taken to prevent further damage. Extension and rotation of the neck should be prevented, preferably by halter traction. Skull callipers may be applied later. Sandbags may help prevent movement in the emergency situation. Collars should be maintained in position until radiography has been completed. Lateral radiographs of the cervical spine, including all seven cervical vertebrae, are done as soon as possible. Movement of the patient is especially hazardous: five individuals are needed to move the patient, keeping the head and neck immobile. Patients unconscious following a head injury should be assumed to have a cervical spine injury until proved otherwise by normal radiographs of the cervical spine (see Figs 18.1 & 18.2).

Other injuries

Even relatively minor head trauma can be complicated by injuries to the eyes, facial skeleton, ears and cranial nerves. The cooperation of ophthalmic, ear, nose and throat, plastic and faciomaxillary surgeons is therefore needed. More serious head injury may also be complicated by cervical

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spine and carotid artery trauma, which may not be apparent at first.

Specific head injuries

Skull fractures

Skull vault

Skull vault fractures, if linear and closed, do not of themselves require treatment and are more an indication of the severity of the trauma: attention is concentrated on the associated brain injury. Penetrating injuries may cause compound fractures, with pieces of bone and foreign bodies penetrating the meninges or the brain itself. These obviously require careful exploration and wound toilet. Depressed skull fractures may lacerate meninges or brain and pressure from a depressed fracture may cause traumatic epilepsy.

Base of skull

Basal skull fractures have already been mentioned. They are usually anterior or middle cranial fossa fractures and may communicate with the exterior, with the risk of infection. Antibiotics should be given. In anterior cranial fossa fractures, persistent rhinorrhoea through the cribriform plate of the ethmoid may need formal repair.

Facial fractures

Nasal bone fracture

Nasal bone fractures are common and are accompanied by swelling, deformity and bleeding from the nose. Reduction is carried out to correct the deformity, although this is often delayed to allow resolution of swelling before reassessment for deformity. If there is dislocation of the septum, reduction is necessary to relieve the obstructed nasal passage.

Fracture of the zygoma

Direct injury, such as a blow on the neck, may produce a depressed fracture of the zygoma. There are three common fracture sites: the arch, the region of the infraorbital foramen and the frontomalar suture. On examination, circumorbital ecchymosis is evident, with the bruising confined to an area within the orbital rim. Subconjunctival haemorrhage is also present without a posterior border. The cheek appears flattened and a bony step can be felt in the infraorbital rim, with an area of paraesthesia below that in the distribution of the infraorbital nerve. Diplopia may occur and the patient may have unilateral epistaxis. Movement of the mandible may be restricted. Diplopia is usually temporary due to bruising of the inferior rectus and inferior oblique muscles. However, if there is entrapment of the muscles in the fracture, the diplopia may persist and an inability to elevate the affected eye can be demonstrated. Blowout fractures of the orbit are due to direct trauma to the eye, with collapse of the orbital floor into the antrum. Entrapment of the inferior extraocular muscles may cause persistent diplopia. Zygomatic arch fracture should be elevated via a temporal incision within 10 days of the injury. An orbital blowout fracture requires elevation through the antrum or may need prosthetic replacement.

Fractures of the maxilla

Le Fort has separated fractures of the maxilla into three categories (Fig. 18.4).

Le Fort I
This fracture traverses the lower nasal septum and maxillary antrum, separating the dentoalveolar portion of the

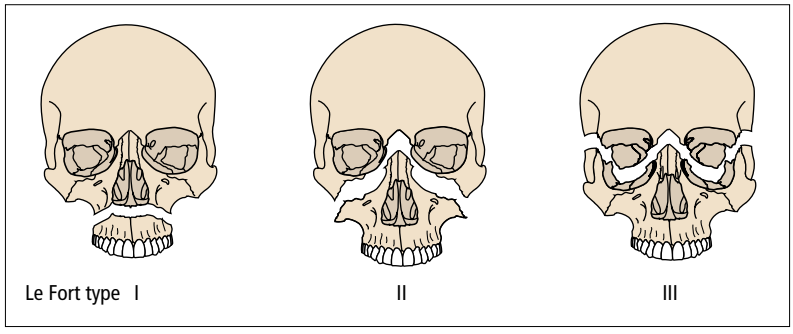

Figure 18.4 Le Fort classification of maxillary fractures.

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maxilla from the rest of the skull. The lower fragment is very mobile and dental occlusion is affected, giving an open bite.

Le Fort II
In this higher-level fracture, the line of the fracture extends through the nasal bones and medial portion of the orbit. Epistaxis and rhinorrhoea may occur. Periorbital bruising and subconjunctival haemorrhage are evident. Malocclusion is present with an anterior open bite.

Le Fort III
This is a very high fracture above the level of the zygomatic arch and including the whole maxilla, which is pushed downwards and backwards. There is extensive facial oedema and periorbital ecchymoses with subconjunctival haemorrhage. Involvement of the cribriform frontal sinuses in the line of the fracture gives rise to escape of CSF as rhinorrhoea.

Fractures of the mandible

These fractures often occur in two places, sometimes involving identical sites on both sides, sometimes involving quite different sites depending on the forces transmitted at the time of injury, e.g. a punch to the side of the jaw may cause a fracture through the premolar area on the side of the impact and a condylar fracture on the opposite side. The premolar area, angle of the jaw and the mandibular condyle are the common sites of fracture. The fracture is a compound fracture if it traverses a tooth socket, and the patient should accordingly be treated with antibiotics.

Clinically, a patient who sustains a mandibular fracture has a defective bite. Because of the upward pull of masseter and pterygoid muscles on the posterior fragment, posterior dental occlusion occurs prematurely and anterior dental occlusion cannot be achieved. The anterior fragment tends to be pulled downwards and backwards by the digastric muscles, thus further preventing occlusion. Damage to the inferior alveolar nerve may occur as it runs from mandibular to mental foramen, causing anaesthesia or paraesthesia in the area. A sublingual haematoma develops and a step may be felt in the line of the mandible.

Treatment is aimed at correcting the deranged dental occlusion and correcting the backward displacement of the horizontal ramus, which can obstruct the airway. As in any fracture, reduction with correct alignment of the fragments and immobilization achieves the desired result. Immobilization is achieved by fixing the upper and lower jaws together with cap splints or by direct wiring or plating of the fracture. Fixation for 4–6 weeks is usually required, except for unilateral condylar fractures where 10 days is deemed sufficient.

Intracranial haemorrhage

Extradural haemorrhage (Fig. 18.5 and Fig. 18.9)

An extradural haemorrhage is almost invariably caused by trauma, the only other cause being postoperative. Usually the patient has received a relatively minor blow that causes little primary brain damage but which fractures the skull. Beneath the fracture a dural vessel, most commonly the middle meningeal artery, is torn and the bleeding strips away the dura from the inner table of the skull. Bleeding is rapid and an enlarging haematoma collects between the skull and the dura mater and compresses the underlying brain. The signs of increased ICP quickly develop, with a rise in blood pressure, a fall in pulse rate, a dilated pupil and focal neurological signs, which include paresis or paralysis of the limbs on the opposite side and Jacksonian epilepsy. The patient gradually sinks into a coma and will die from raised ICP if no action is taken. Evacuation of the haematoma can only be carried out completely by craniotomy. Burr-holes placed over the clot to release some of it may be a life-saving procedure if there is not sufficient time for transfer to a neurosurgical unit, but if appropriate predictive care of a head injury has been carried out such heroics should not be neccessary. The possibility of an extradural haematoma should be considered in all those with a skull fracture (other than the fully alert patient) and a CT scan performed before further deterioration occurs.

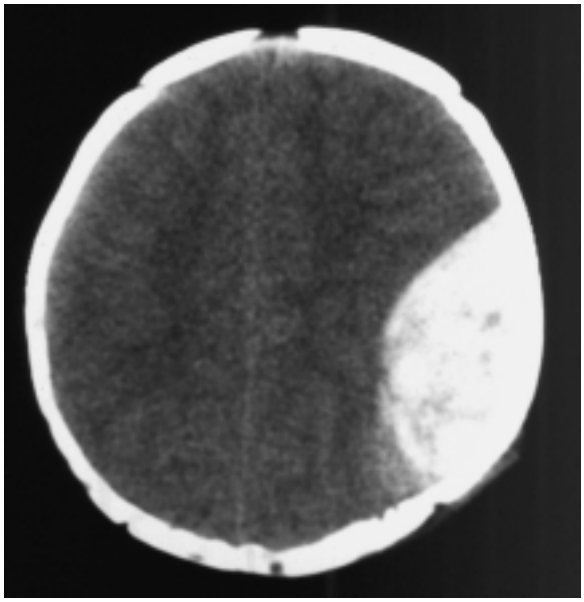

Figure 18.5 Extradural haematoma. CT scan showing large extradural haematoma in the left temporoparietal area with compression of the ventricles.

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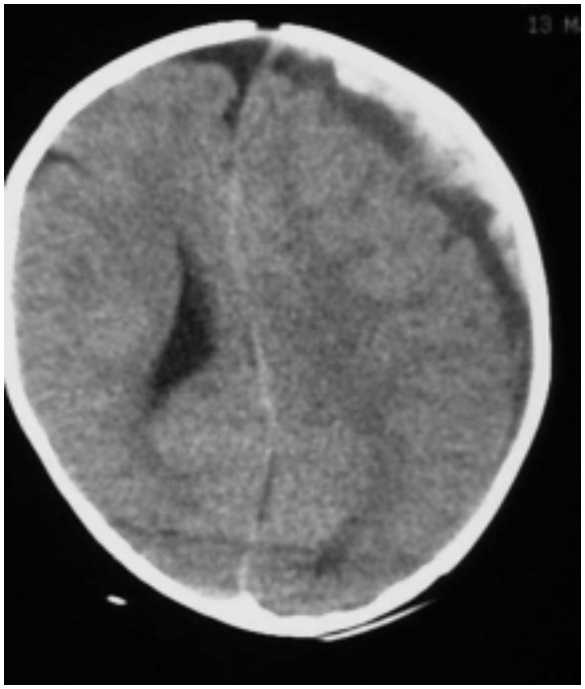

Figure 18.6 Acute subdural haematoma. CT scan showing large subdural haematoma in the left occipitoparietal area with compression of the left ventricles.

Acute subdural haematoma (Fig. 18.6 and Fig. 18.8)

Acute subdural haemorrhage carries a poor prognosis as it is usually associated with severe primary injury. This type of intracranial bleeding is more common than extradural haemorrhage and tends to occur in elderly patients who sustain a head injury because of the increased mobility of the brain within the skull cavity. It is accompanied by cerebral laceration or contusion and the patient tends to be in a confused or unconscious state from the time of the injury. The bleeding is the result of tearing of thin-walled veins traversing the space between the arachnoid and dura mater. The patient's neurological condition deteriorates progressively as the haematoma spreads. Even when the haematoma has been fully evacuated following craniotomy and incision of the dura mater, the patient may not fully recover because of the underlying brain injury or recovery may be slow.

Chronic subdural haematoma (Fig. 18.7)

Chronic subdural haemorrhage is seen in the elderly, alcoholics and infants. The most common cause in an infant is a non-accidental shaking injury and this possibility should always be considered. The condition develops over

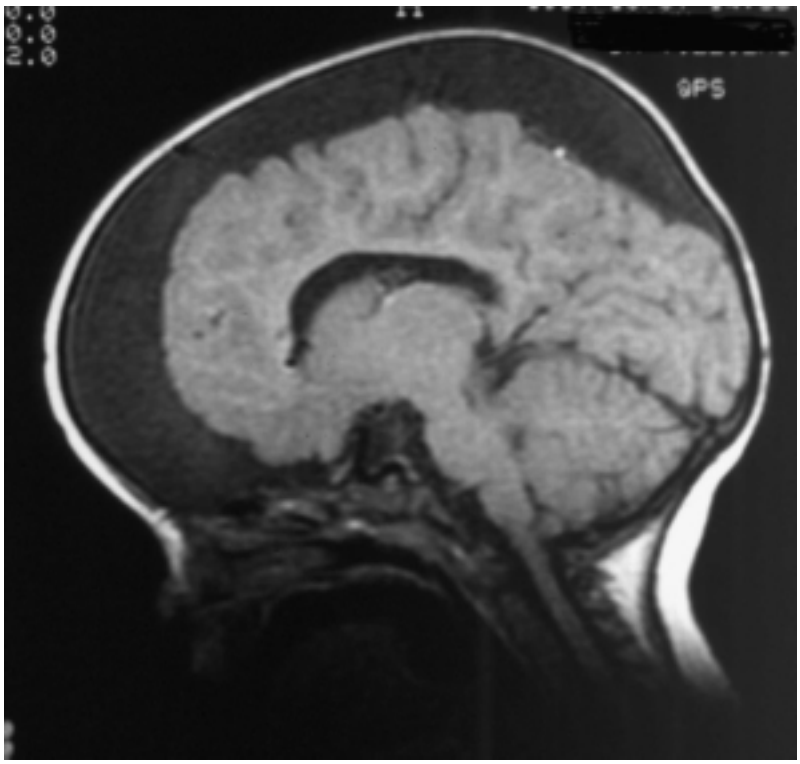

Figure 18.7 Chronic subdural haematoma. MRI scan showing large chronic subdural haematoma surrounding the cerebral cortex.

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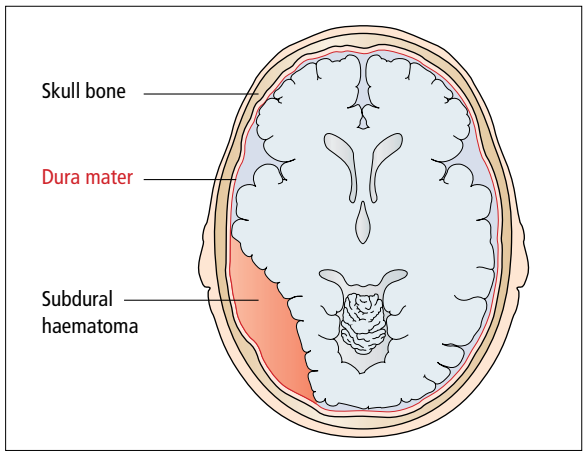

Figure 18.8 Subdural haematoma.

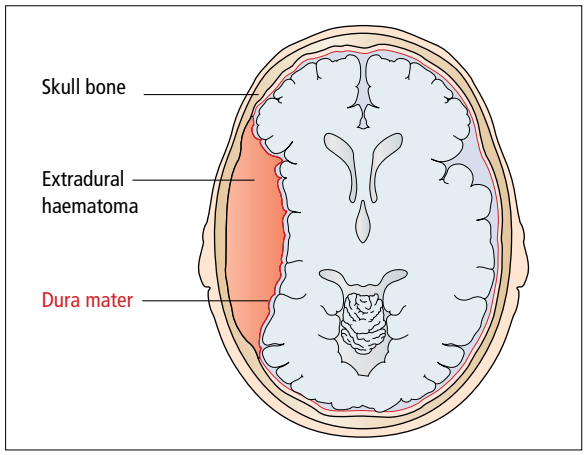

Figure 18.9 Extradural haematoma.

a period of days or weeks so that the initial head injury, often minor, which precipitated the bleeding is forgotten. A small subdural bleed, often from a torn cortical vein, gradually enlarges over several days or weeks. The haematoma gradually enlarges, not as a result of continued bleeding but because of absorption by osmosis of CSF across the semipermeable membrane of the arachnoid into the clot. As the haematoma increases in size and the pressure rises, the patient exhibits drowsiness and confusion. Headache is not uncommon. Hemiplegia may develop. Pupillary changes develop late and are an indication of imminent cone formation. The symptoms may wax and wane depending on fluctuations in brain volume, which are dependent on changes in blood gases.

Table 18.4 Outcome from head injury related to Glasgow Coma Scale (GCS) on admission.

GCS on admission (maximum score 15)	Mortality (%)
15	1
8–12	5
< 8	40

Identification depends on a high index of suspicion if symptoms such as headache or mental confusion persist after a head injury, even a minor one. Clinical diagnosis can be confirmed by CT. Treatment is by evacuation of the clot at operation. Burr-holes are made and the dura, often stained greenish-blue by blood pigments from the underlying haematoma, is incised. The haematoma is usually watery and after release of fluid the subdural space is copiously irrigated.

Intracerebral haemorrhage

A patient with intracerebral haemorrhage following head injury is likely to be semicomatose or comatose on admission. The neurological deficit depends on the location and extent of the intracerebral bleeding. Much of the brain damage is irreversible, although further damage from surrounding cerebral oedema and ICP can be minimized by ensuring adequate oxygenation. The period of unconsciousness may be very prolonged, requiring attention to nutritional requirements and other problems such as decubitus calculi, bladder function and bed sores.

Outcome from head injury

Mortality

Survival after head injury depends largely on the level of consciousness on arrival in hospital, which reflects the severity of the primary injury. Those with a GCS score of 15/15 should have a mortality of no more than 1%; those scoring 8–12/15 have a mortality of 5%; those in actual coma on arrival with a score of 8 or less have a mortality of 40% (Table 18.4).

Morbidity

Only 30% of those in coma on arrival (GCS score < 8) will make a full recovery, while 20% will have some disability

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Table 18.5 Professionals involved in rehabilitation following head injury.

Nurses: help patients with all aspects of general healthcare
Doctors: coordinate medical care and prescribe medication as indicated
Physiotherapist: helps patients recover physical ability and competence
Occupational therapist: helps patients develop independence in carrying out everyday tasks
Clinical psychologist: assesses and provides help with patients' mental skills (memory, emotional problems)
Speech and language therapist: helps patients communicate more effectively using both the spoken and written word
Social worker: helps patient and families receive the practical help needed (e.g. benefits, accommodation, transport, housing)

but will be able to work and care for themselves; 10% will require full care, being severely disabled or in a persistent vegetative state.

Postconcussion syndrome

Those recovering from even minor trauma may take a long time to recuperate, suffering from the postconcussion syndrome. This is characterized by headaches, difficulty concentrating, dizziness and depression, and usually disappears after 3–6 months. Nothing can be offered except reassurance.

Other long-term effects

Survivors of more severe injuries often fail to return to their preinjury state as a result of personality change, memory impairment and depression, as well as any residual disability. Of those with a closed head injury, 5% develop epilepsy that may prevent return to work. Families are under great strain in these circumstances, and may benefit from contact with self-help groups such as Headway (http://www.headway.org.uk).

Rehabilitation after head injury

Rehabilitation aims to achieve the optimum levels of physical, cognitive and social competence for the patient followed by integration into the most suitable environment. A whole host of professionals are involved in rehabilitation of a patient following serious head injury but the patients who do best are those with very good family support (Table 18.5). Often patients will be admitted to a rehabilitation centre for a period of intensive assessment and therapy. The greatest visible progress occurs in the first 6 months, after which improvement is often more subtle and less obvious. It is important to bear in mind that progress does not stop after 2 years but may continue for 5, 10 or even more years after a head injury. Rehabilitation has two stages, the first being the formal intervention to improve the individual, and the second when the family and carers work to maintain that improvement.

Spinal injury

Spinal injury is about one-tenth as common as head injury but the basic pathology is the same. Such an injury may be complete (rendering the patient functionless below the level of the lesion) or incomplete (some function is preserved). Secondary effects, including hypoxia and perfusion failure, can also occur. However, compression of the cord is less common than compression of the brain following head injury. The causes of cord compression are bone fragments, haematomata and acute disc prolapses.

Management

Three fundamental questions must be answered when managing a spinal injury.

- At what level is the injury?
- Is the cord lesion complete or incomplete?
- Is the bony architecture stable or unstable?

Flaccid paraplegia with no preservation of sensation usually indicates a complete lesion. The sacral dermatomes are the most resistant to injury and sacral sparing is a cause for *slight* optimism. If the lesion is incomplete, every effort must be made to prevent deterioration. Thus the spine must be held in a stable position, hypoxia prevented and hypotension corrected. Early investigation with magnetic resonance imaging (MRI) will exclude cord compression.

If the bony architecture is displaced or thought to be unstable, it should be reduced and stabilized. When the cervical spine is injured this can be done with traction only, but if the thoracic and lumbar spines are involved open reduction may be required. It is now common to perform an early operative fixation with bone grafts and metal for both incomplete and complete lesions. This allows early mobilization, which in turn reduces the risk of chest complications and deep vein thrombosis. At the earliest opportunity the patient should be transferred to a spinal injuries unit.

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Management of head injury at a glance

- Little can be done for primary brain injury apart from avoidance of head injury (seat-belts, helmets, don't drink and drive!)
- Prevention of secondary brain injury is the most important objective of head injury care

Trivial head injury

Patient is conscious, may be history of period of loss of consciousness. Retrograde amnesia for events prior to head injury is significant

Indications for skull X-ray

Oriented patient

- History of loss of consciousness or amnesia
- Suspected penetrating injury
- CSF or blood loss from the nose or ear
- Scalp laceration (to bone or > 5 cm long), bruise or swelling
- Violent mechanism of injury
- Persistent headache or vomiting
- In a child, fall from a significant height (depends in part on age of the child) and/or on to a hard surface; tense fontanelle, suspected non-accidental injury

Patient with impaired consciousness or neurological signs

- All patients, unless urgent CT is performed

Indications for admission

Oriented patient

- Skull fracture or suture diastasis
- Persisting neurological symptoms or signs
- Confusion or any other depression of the level of consciousness at the time of examination
- Difficulty in assessing the patient (e.g. due to alcohol, young age, epilepsy, attempted suicide)
- Inadequate social conditions or lack of responsible adult or relative
- Coexistence of other medical conditions (e.g. coagulation disorders)

All patients with impaired consciousness

Indications for urgent CT scanning ± neurosurgical consultation

- Coma, confusion or neurological signs persisting after resuscitation
- Deteriorating consciousness or progressive neurological signs
- Fractured skull in combination with:
  (a) Confusion or other impairment of consciousness
  (b) Epileptic seizure
  (c) Neurological symptoms or signs
- Open injury with:
  (a) Depressed compound fracture of skull vault
  (b) Fractured base of skull
  (c) Penetrating injury
- Unstable systemic state precluding transfer to neurosurgery
- Diagnosis uncertain
- Tense fontanelle or suture diastasis in a child

Indications for neurosurgical consultation after CT scanning

Abnormal CT scan

- High- or mixed-density intracranial lesion
- Midline shift
- Obliteration of third ventricle
- Relative dilatation of lateral ventricle(s)
- Obliteration of basal cisterns
- Intracranial air
- Subarachnoid or intraventricular haemorrhage

CT scan normal but progress unsatisfactory

Severe head injury

- Patient will arrive unconscious at hospital. Head injury may be part of a multiple trauma
- ABC: intubate and ventilate unconscious patients to protect airway and prevent secondary brain injury from hypoxia
- Resuscitate patient and look for other injuries, especially if the patient is in shock. Head injury may be accompanied by cervical spine injury and the neck must be protected by a cervical collar in these patients
- Treat life-threatening problems (e.g. ruptured spleen) and stabilize patient before transfer to neurosurgical unit. When transferring, ensure adequate medical supervision (anaesthetist + nurse) during transfer

Complications

Skull fractures

- Indicate severity of injury
- No specific treatment required unless compound, depressed or associated with chronic CSF loss (e.g. anterior cranial fossa basal skull fracture)

Raised ICP

- Trauma to the brain causes swelling but because the brain is encased in a rigid box (i.e. the skull) cerebral swelling results in raised ICP. If ICP rises above arterial blood pressure, cerebral perfusion fails. This leads to ischaemia, which in turn leads to more brain swelling and a further increase in ICP
- ICP tends to be worse in those with severe primary damage

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- Ventilation is used with sedation, diuretics and intermittent boluses of mannitol (osmotic diuretic) to control ICP. Moderate hyperventilation reduces P_{CO_2}, which in turn reduces cerebral blood flow but this mechanism tends to be effective for only 48–72 h
- Monitoring is essential in patients with raised ICP:
  (a) Normal ICP < 10 mmHg
  (b) ICP > 20 mmHg is worrying and needs to be treated
  (c) ICP > 40 mmHg is severe and rarely associated with a survivable injury

Intracranial haemorrhage

- Extradural haemorrhage: tear in middle meningeal artery. Haematoma between skull and dura. Often a 'lucid interval' before signs of raised ICP ensue (falling pulse, rising blood pressure, ipsilateral pupillary dilatation, contralateral paresis or paralysis). Treatment is by evacuation of haematoma via burr-holes
- Acute subdural haemorrhage: tearing of veins between arachnoid and dura mater. Usually seen in elderly. Progressive neurological deterioration. Treatment is by evacuation via craniotomy but even then recovery may be incomplete
- Chronic subdural haematoma: tear in cortical vein. Haematoma enlarges slowly by absorption of CSF. Often the precipitating injury is trivial. Drowsiness and confusion, headache, hemiplegia. Treatment is by evacuation of the clot
- Intracerebral haemorrhage: haemorrhage into brain substance causes irreversible damage. Efforts are made to avoid secondary injury by ensuring adequate oxygenation and nutrition

Prognosis

Related to level of consciousness on arrival in hospital

GCS on admission	Mortality
15	1%
8–12	5%
< 8	40%

Evidence-based medicine

Dickinson, K., Bunn, F., Wentz, R., Edwards, P. & Roberts, I. (2000) Size and quality of randomised controlled trials in head injury: review of published studies. *Br Med J* **320**, 1308–11.

Society of British Neurological Surgeons (1998) Guidelines for the initial management of head injuries. *Br J Neurosurg* **12**, 349–52.

http://www.braintrauma.org/guideems.ns American Brain Trauma Foundation website with useful evidence-based guidelines for management of head injury.

http://www.crash.ucl.ac.uk/ CRASH investigators website describing update on corticosteroids in head injury trial.

http://www.rcsed.ac.uk/Journal/vol146–3/463000s.htm Modern management of head injuries.

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Disorders of the Spleen

30

Must know Must do

Must know

Causes and clinical manifestations of hypersplenism
Causes and management of patients with splenomegaly
Adverse effects of splenectomy

Must do

Clerk and follow the management of a patient with idiopathic thrombocytopenic purpura
Learn to detect an enlarged spleen by palpation
Observe a patient undergoing laparoscopic splenectomy
Examine a blood film from a splenectomized patient

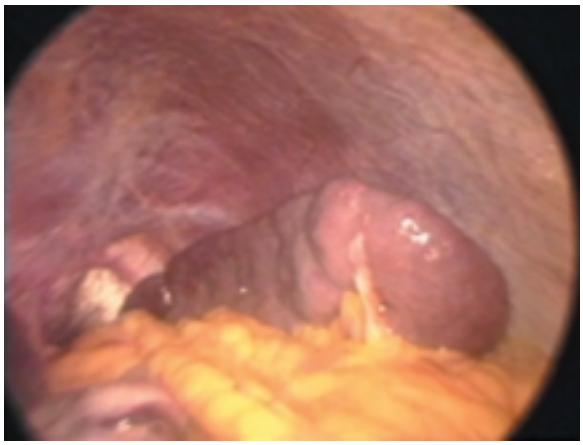

Figure 30.1 Appearance of normal spleen during laparoscopic antireflux surgery: the organ is well inside the lower ribcage.

Anatomy and physiology

The spleen is situated in the left upper quadrant tucked under and against the left dome of the diaphragm and overlain by the lower left 9th to 11th ribs (Fig. 30.1). It has to enlarge two to three times before becoming palpable. Accessory spleens (splenunculi) are present in 10% of adults and are most commonly situated near the hilum but may occur in other sites They are important in relation to splenectomy for haematological disorders, because if left behind they may hypertrophy and cause recurrence of the disease.

The spleen removes damaged or senescent red cells from the circulation (culling) and remodels the surface of maturing erythrocytes to achieve the ideal ratio between membrane surface area and cell volume. Hence target cells, which have excess membrane, appear in the peripheral blood after splenectomy. Intraerythrocytic inclusions are removed by the spleen, e.g. Howell–Jolly bodies (nuclear remnants), siderotic granules (haemosiderin aggregates) and Heinz bodies (aggregates of denatured haemoglobin). These appear in the peripheral blood after splenectomy. The spleen is very efficient in clearing particulate matter from the circulation. Following splenectomy, the primary antibody response is decreased and the secondary response is abnormal in that there is impaired switching from IgM to IgG antibody subtypes. The spleen also produces non-specific effectors of the immune response, e.g. tuftsin, which opsonizes particulate matter thereby facilitating phagocytosis. The spleen opsonizes pneumococci and is involved in the alternative pathway of complement activation. Loss of these immune functions accounts for the increased susceptibility to sepsis after splenectomy, especially in infants and children.

Hypersplenism

This is a syndrome of splenomegaly combined with destruction of formed blood elements leading to one or more of the following:

- anaemia;
- leukopenia $< 4–5 \times 10^9$/L;
- thrombocytopenia $< 100 \times 10^9$/L.

Primary hypersplenism is when the destruction of normal

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blood elements is caused by a primary lymphoreticular disorder, as distinct from *secondary hypersplenism* where the hyperactivity is associated with splenomegaly from other causes, e.g. liver disease. In both states, the bone marrow is unable to maintain normal numbers of circulating cells or platelets. Splenectomy is potentially curative of the cytopenias that occur in hypersplenism but there are other alternatives.

Hyposplenism

Hyposplenism is confirmed by the appearance of defective red cells in the peripheral circulation. The common causes of hyposplenism are summarized in Table 30.1. The most frequent cause is surgical splenectomy. Splenic hypoplasia forms part of the syndrome of Fanconi's anaemia (congenital hypoplastic anaemia). Acquired hyposplenism occurs in patients with coeliac disease and other gastrointestinal disorders, e.g. Crohn's disease, ulcerative colitis. Circulating autoantibodies and immune complexes in clinical autoimmune disorders, e.g. systemic lupus erythematosus (SLE), can cause a functional hypoplastic state secondary to Fc-receptor blockade. The hyposplenism of sickle cell anaemia is related to the extent of splenic infarction. Hyposplenism is also a feature of patients with full-blown human immunodeficiency virus (HIV) infection. These patients usually present with *Mycobacterium avium* complex infection, which often complicates HIV-related immune thrombocytopenic purpura (ITP).

Table 30.1 Causes of asplenism/hyposplenism.

Splenectomy
Splenic agenesis
Atrophy
Coeliac disease
Inflammatory bowel disease and collagenous colitis
Systemic amyloidosis
Old age
Dermatitis herpetiformis
Sickle cell anaemia
Systemic lupus erythematosus

Postsplenectomy sepsis

The risk of overwhelming sepsis is increased after splenectomy, with the greatest risk in infants and children up to 5 years old. Apart from age, the risk is also influenced by the nature of the disease necessitating splenectomy, with trauma having the lowest risk and thalassaemia the highest (Table 30.2). Some of the estimates of risk have been questioned because the majority of reported data are based on single case reports of pneumococcal infections with bacteraemia. Community-acquired pneumococcal pneumonia with bacteraemia is common in patients with normal splenic function and is seldom reported because of its established occurrence in susceptible groups.

Streptococcus pneumoniae is responsible for 60% of septic episodes in asplenic patients, although infections by other encapsulated bacteria (e.g. *Haemophilus influenzae*, *Haemophilus pertussis*, *Neisseria meningitidis*) are also common, as are infections by Gram-negative bacteria. The syndrome of overwhelming postsplenectomy infection (OPSI) begins insidiously with a non-specific viral-like illness with rapid progression to a fulminant infection with high fever, rigors, vomiting, dehydration, hypotension and coma, unless halted by effective resuscitation and aggressive antibiotic therapy. The mortality rate of OPSI exceeds 50%. Treatment is with intravenous broad-spectrum antibiotics effective against encapsulated cocci in the first instance until blood culture results become available. Intravenous colloids are used to correct the hypovolaemia using central venous pressure as a guide to therapy.

The prevention of OPSI is based on vaccination, administration of oral penicillin and patient education. Prophylactic antibiotic therapy is recommended (together with vaccination) in children and some would extend it indefinitely to adults. Vaccination is carried out at least 10–14 days prior to splenectomy for maximum effective immunization and includes polyvalent pneumococcal vaccine and *Haemophilus* vaccines. Immunization after

Indication	Incidence of sepsis (%)
Trauma	1.4
Immune (idiopathic) thrombocytopenia	2.0
Incidental (iatrogenic injury)	2.1
Congenital spherocytosis	3.5
Acquired haemolytic anaemia	7.5
Portal hypertension	8.2
Reticulosis/lymphomas	11.5
Thalassaemia	24.8

Table 30.2 Risk of postsplenectomy sepsis with indication for splenectomy. Modified from Singer, D.B. (1973) Postsplenectomy sepsis. *Perspect Pediatr Pathol* **1**, 285–311.

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splenectomy (trauma cases) is much less effective but is advisable. All patients should carry a 'splenectomy card'.

Splenic infarction

Infarction of the spleen has a variable clinical presentation, ranging from patients with acute life-threatening complications (splenic rupture, splenic abscess) to others where symptoms are minor or absent. There is a high risk of infarction in patients with sickle cell disease and those with splenomegaly due to chronic myeloid leukaemia and myelosclerosis. Splenic infarction may also be due to thromboembolic disorders (atrial fibrillation), diabetic microvascular disease and acute torsion of an *ectopic (wandering) spleen*. A variety of other disorders may be complicated by the development of splenic infarction, e.g. falciparum malaria, acquired immunodeficiency syndrome (AIDS), severe necrotizing pancreatitis. The splenic infarction in patients with AIDS is due to arterial thrombosis of the coeliac trunk and is associated with thrombocytopenia and a coagulopathy.

The age range of splenic infarction varies widely. The most common symptoms are acute upper left quadrant abdominal pain, fever, chills and malaise. However one-third of patients are asymptomatic. Physical examination reveals tenderness and guarding maximal in the left upper abdomen. Splenic infarction results in a capsular inflammatory reaction causing irritation of the left diaphragm, with left basal pleurisy/effusion and left shoulder pain (Kerr's sign). The diagnosis is confirmed by abdominal computed tomography (CT). Initially, management is conservative with analgesia and antibiotics. Surgery is indicated if the diagnosis is in doubt or for complications (splenic abscess, bleeding from splenic rupture) when splenectomy is indicated.

Splenomegaly

There are several causes of splenomegaly (Table 30.3) and the relative incidence varies in different parts of the world. In the West the distribution is as follows:

- hepatic diseases, most commonly cirrhosis;
- haematological disease;
- infectious diseases (16%, increasingly AIDS);
- inflammatory non-infectious disease;
- primary splenic disease;
- others (3%, e.g. congestive heart failure, endocarditis).

The spleen may enlarge transiently in a variety of acute bacterial and viral infections, chronic infections and in subacute bacterial endocarditis. Parasitic infections such as malaria cause massive congestive splenomegaly and these very large spleens can rupture. AIDS now accounts for 60% of splenomegaly caused by infectious disease. Portal hypertension causes mild to moderate splenomegaly unless it is part of Budd–Chiari syndrome or follows splenic vein thrombosis. Splenomegaly accompanies both hereditary and acquired red cell defects. The increasing splenic size predisposes these patients to increased destruction of the abnormal red cells. Splenomegaly regularly accompanies myeloproliferative disorders, e.g. myelosclerosis, leukaemia or lymphoma.

Clinical features

When palpable, the spleen is at least two to three times normal size. Pressure symptoms of splenomegaly are present when the spleen becomes massively enlarged and include chronic dragging abdominal pain or pain when lying on the side, abdominal discomfort and early satiety. In addition patients may complain of attacks of colicky left upper quadrant pain. Physical examination shows a left upper quadrant mass that disappears beneath the costal margin such that it is impossible to reach its superior limit. The enlarged spleen lies against the abdominal wall and is dull to percussion. Capsular inflammation of the spleen may produce a rub heard with the stethoscope. Physical examination must include a careful search for lymphadenopathy and stigmata of chronic liver disease. The cause of splenic enlargement is identified by history, physical examination and a few key tests. Haematological causes are fully characterized by a peripheral blood smear and a bone marrow biopsy. Serological testing identifies most infectious cases, e.g. AIDS, mononucleosis (positive Paul–Bunnell test, rising anti-Epstein–Barr virus titre). Patients with a history of travel or those living in endemic areas should have blood smears examined for malaria or a marrow biopsy examined for Leishman–Donovan bodies.

The size, shape and consistency of the spleen is accurately visualized by either CT or ultrasonography. The splenic blood vessels can be imaged using either duplex ultrasound or high-dose contrast helical CT. Splenic ultrasound is used for:

- detection of accessory spleens;
- confirmation of splenomegaly but not its cause;
- differentiation of solid from cystic intrasplenic focal masses;
- detection of calcification, wall thickening, internal debris, and gas within cystic-type lesions;
- detection of splenic cavernous haemangiomas;
- diagnosis of splenic infarction;
- diagnosis of splenic trauma and monitoring of patients with splenic injuries managed conservatively.

An accurate assessment of the spleen's function can be obtained by injection of labelled platelets, cells (red, white) or carrier molecules and radiotracer studies. The splenic uptake rate of ^{99m}Tc-labelled sulphur colloid or Tc-labelled tin colloid provides a sensitive and quantitative assessment of splenic function.

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Table 30.3 Disorders producing splenomegaly.

Infections
Acute
Hepatitis
Mononucleosis
Salmonellosis
Toxoplasmosis
Cytomegalovirus
Abscess

Subacute
AIDS
Bacterial endocarditis
Tuberculosis
Brucellosis
Malaria

Chronic
Fungal disease
Syphilis
Bacterial endocarditis

Congestive
Intrahepatic portal hypertension
Cirrhosis
Wilson's disease
Haemochromatosis
Congenital hepatic fibrosis

Prehepatic portal hypertension
Portal vein thrombosis

Posthepatic portal hypertension
Budd–Chiari syndrome
Congestive cardiac failure

Segmental (left-sided portal hypertension)
Splenic vein occlusion

Haematological
Haemolytic disorders
Hereditary cell membrane defects
Autoimmune haemolytic states (warm antibodies)
Thalassaemia
Sickle cell disease
Haemoglobin C disease

Myeloproliferative disorders
Myeloid metaplasia
Polycythaemia vera
Essential thrombocythaemia

Miscellaneous
Primary splenic hyperplasia
Megaloblastic anaemia
Iron deficiency

Malignant
Haematological malignancies
Acute or chronic leukaemias
Leukaemic reticuloendotheliosis
Malignant lymphomas
Malignant histiocytosis
Myelomatosis

Primary intrinsic malignancies
Lymphosarcoma
Plasmacytoma
Fibrosarcoma
Angiosarcoma

Intrinsic secondary malignancies
Carcinoma
Melanoma

Benign
Haemangioma
Lymphangioma

Inflammatory or granulomatous
Felty's syndrome
Systemic lupus erythematosus
Rheumatic fever
Serum sickness
Sarcoidosis

Storage disease
Gaucher's disease
Wilson's disease
Niemann–Pick syndrome
Histiocytosis X
Hurler's syndrome
Tangier disease

Miscellaneous
Cysts
Parasitic
Pseudocysts
Congenital
Traumatic

Others
Hyperthyroidism
Osler–Weber–Rendu syndrome
Splenic mastocytosis
Albers–Schönberg disease

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Table 30.4 Indications for splenectomy.

Definite
Non-salvageable spleen injury
En-bloc resection of adjacent neoplasms (usually proximal gastric cancer)
Neoplasms of the spleen (usually lymphomas)
Splenic abscess
Echinococcal cysts
Bleeding gastric varices due to sinistral portal hypertension (splenic vein thrombosis)
Desirable (selective)
Hereditary spherocytosis
Immune (idiopathic) thrombocytopenic purpura
AIDS-related thrombocytopenic purpura
Autoimmune haemolytic anaemia
Sickling syndromes (sickle cell disease and sickle β-thalassaemia)
Debatable
Non-parasitic splenic cysts
Thalassaemia syndromes
Lymphoma with specific cytopenia or pancytopenia
Thrombotic thrombocytopenic purpura
Myeloproliferative disorders

The indications for splenectomy are shown in Table 30.4. All patients undergoing elective splenectomy should be immunized against *Streptococcus pneumoniae* and *Haemophilus* spp. and this should be carried out 2 weeks before surgery. If the platelet count is low, platelet transfusion to a level greater than 60×10^9/L is indicated both prior to and for the first few days following operation. If the thrombocytopenia is caused by immune disease (e.g. ITP), then preoperative platelet transfusions are less useful, whereas human IgG increases the platelet count. Because of its advantages (fewer perioperative complications, reduced morbidity, shorter hospital stay), laparoscopic splenectomy is now preferable to open operation for some but not all disorders requiring splenectomy. It is the procedure of choice for benign disease especially when the spleen is not very enlarged, e.g. ITP, AIDS-related thrombocytopenic purpura and acquired haemolytic anaemia (Fig. 30.2).

Ectopic (wandering) spleen

Ectopic (wandering) spleen is rare and occurs more commonly in women (7 : 1). It is due to lax attachments of the spleen to the retroperitoneum and long splenic vessels, such that the spleen 'wanders' the quadrants of the abdomen (Fig. 30.3). It may present acutely with abdominal pain due to torsion that may progress to infarction,

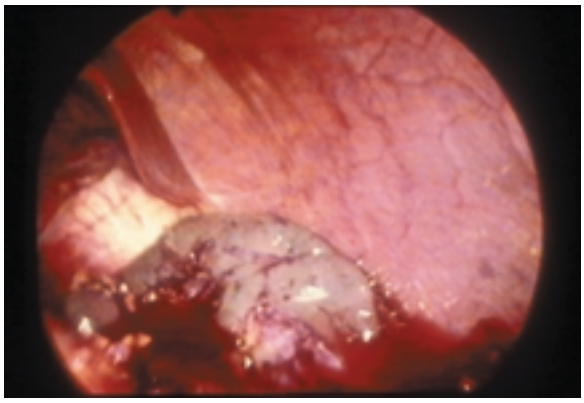

Figure 30.2 Laparoscopic splenectomy: detached spleen prior to removal.

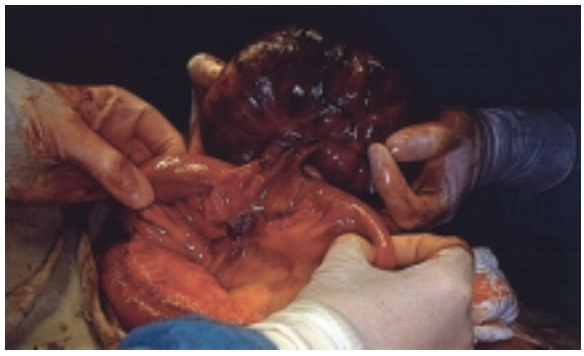

Figure 30.3 Emergency splenectomy for necrotic wandering spleen following torsion of the splenic vessels. Courtesy of Professor R.J.C. Steele, University of Dundee.

with hypersplenism (due to congestion), or simply with an abdominal mass with or without associated pain. The diagnosis is confirmed by CT or duplex ultrasonography. Treatment of ectopic spleen is by splenopexy (viable spleen) or splenectomy when the spleen is infarcted.

Splenic cysts

Splenic cysts are primary or secondary. Secondary cysts are more common and develop after splenic injuries, hence the name 'traumatic splenic pseudocysts' (no lining epithelium). Splenic traumatic pseudocysts may be totally asymptomatic but have a tendency to enlarge (Fig. 30.4). The time interval between initial injury and presentation or diagnosis is extremely variable. Some traumatic pseudocysts remain asymptomatic and are discovered accidentally during investigation by ultrasound. Others develop abdominal pain and a palpable mass. Acute presentation with rupture is well documented. Surgical

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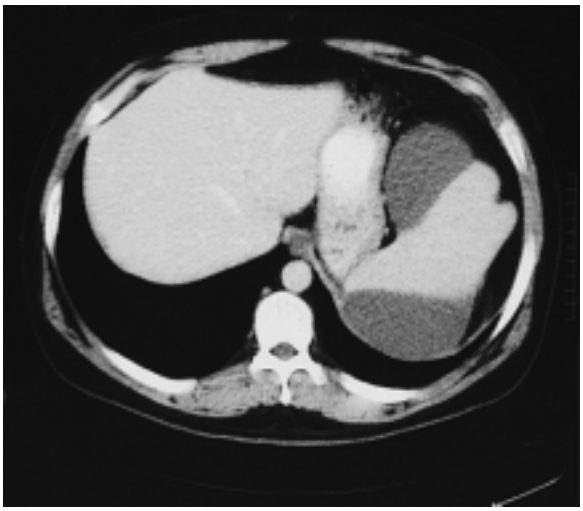

Figure 30.4 Computed tomography showing traumatic cysts at the poles of the spleen following blunt traumatic injury. The lesions were treated laparoscopically with splenic preservation.

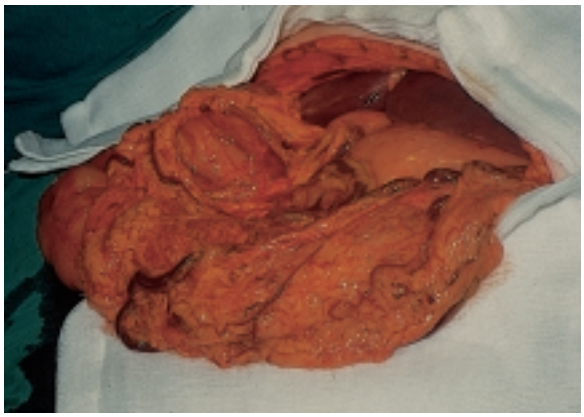

Figure 30.5 Sectorial (sinistral, left-sided) portal hypertension following splenic vein thrombosis in a patient with chronic pancreatitis. Note engorged and prominent left gastroepiploic veins. The patient presented with bleeding gastric varices.

treatment is only necessary for large symptomatic cysts after confirmation of the diagnosis by ultrasound or CT. Spleen-preserving excision is possible unless the cyst is very large or presents acutely with rupture and bleeding.

Primary splenic cysts have an epithelial lining and can be:

- epithelial;
- dermoid;
- lymphangiomatous;
- mucinous cystic lesions;
- parasitic (hydatid).

Splenic tumours

Apart from lymphomas, primary and secondary tumours of the spleen are both rare. Vascular tumours include primary angiosarcoma, haemangioma and haemangioendotheliomas. Secondary tumour deposits are very rare, with a reported frequency of 2–5%. Cutaneous melanoma can metastasize to the spleen, and direct involvement from pancreatic and retroperitoneal sarcomas can occur.

Splenic vein thrombosis

Most commonly, this occurs as a complication of acute or chronic pancreatitis. It can also be caused by pancreatic cancer. Isolated splenic vein thrombosis results in splenomegaly and sectorial or left-sided portal venous hypertension. The condition is characterized by varices involving the short gastric and gastroepiploic veins. The most common presentation is with bleeding gastric varices in patients with normal or good liver function (Fig. 30.5). Treatment is by splenectomy.

Disorders of the spleen at a glance

Functions of the spleen

- Removes damaged and senescent red blood cells from circulation
- Remodels the surface of maturing red blood cells
- Removes intraerythrocytic inclusions:
  (a) Howell–Jolly bodies (nuclear remnants)
  (b) Siderotic granules (haemosiderin aggregrates)
  (c) Heinz bodies (denatured haemoglobin)
- Removes particulate matter from the circulation
- Produces non-specific effectors of immune response, e.g. tuftsin
- Opsonizes pneumococci
- Involves an alternative pathway of complement activation

Hypersplenism

- Syndrome of splenomegaly and destruction of formed elements of blood resulting in:

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(a) Anaemia
(b) Leukopenia
(c) Thrombocytopenia
- Primary hypersplenism: caused by primary lymphoreticular disorder
- Secondary hypersplenism: results from hyperactivity associated with splenomegaly due to some other cause, e.g. liver disease

Hyposplenism
- Caused by failure or absence of the spleen
- Characterized by appearance of defective red blood cells and increase in platelets in the circulation

Causes
- Splenectomy
- Splenic agenesis
- Atrophy caused by:
  (a) Coeliac disease
  (b) Inflammatory bowel disease
  (c) Systemic amyloidosis
  (d) Old age
  (e) Dermatitis herpetiformis
  (f) Sickle cell anaemia
  (g) SLE

Overwhelming postsplenectomy infection
- After splenectomy there is an increased risk of overwhelming sepsis
- Greatest risk in infants and children up to 5 years of age
- Risk depends on reason for splenectomy:
  (a) Least after trauma and ITP
  (b) Greatest after lymphomas and thalassaemia
- Usually caused by encapsulated bacteria: *Streptococcus pneumoniae* (60%), *Haemophilus influenzae*, *Neisseria meningitidis*
- Mortality rate for OPSI > 50%
- Prevention by vaccination (preferably 10–14 days before splenectomy), oral penicillin (probably indefinitely) and patient education (carry splenectomy card)

Splenic infarction
Causes
- Sickle cell disease
- Splenomegaly secondary to chronic myeloid leukaemia and myelosclerosis
- Thromboembolism
- Diabetic microvascular disease
- Acute torsion of ectopic spleen
- Complication of falciparum malaria, AIDS, necrotizing pancreatitis

Clinical features and diagnosis
- Asymptomatic in 30%
- Acute left upper quadrant abdominal pain, fever, chills and malaise
- Tenderness and guarding in left upper abdomen
- Left basal effusion and left shoulder pain
- Diagnosis: CT

Management
- Conservative: analgesia and antibiotics
- Splenectomy for complications only: abscess, rupture

Splenomegaly
- Spleen has to be enlarged two to three times to be clinically palpable

Disease groups that result in splenomegaly
- Hepatic disease: portal hypertension
- Haematological disease: haemolytic disorders, myeloproliferative disease
- Infectious disease: hepatitis, mononucleosis, AIDS, malaria, bacterial endocarditis
- Malignant disease: leukaemias, lymphomas, myelomatosis, lymphosarcoma
- Granulomatous disease: SLE, sarcoidosis, rheumatic fever, Felty's syndrome

Ectopic (wandering) spleen
- Rare phenomenon due to long attachments of the spleen to the retroperitoneum
- Torsion may occur leading to splenic infarction

Splenic cysts
Primary
Primary splenic cysts are rare and classified as:
- Epithelial
- Dermoid
- Lymphangiomatous
- Mucunious cystic lesions
- Hydatid

Secondary
- Secondary (pseudocysts) are seen more often than primary but are not common
- Develop after splenic injury

Evidence-based medicine

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