Subarachnoid Haemorrhage

Subarachnoid haemorrhage (SAH) is usually the result of bleeding from a berry aneurysm in the Circle of Willis. These are called berry aneurysms because of their shape. They were once thought to be mostly congenital but it is now thought that the aetiology may involve susceptibility of the elastic lamina, in some patients, to stressors such as hypertension and atherosclerosis.

Epidemiology[1, 2]

- SAH affects 7.9 people per 100,000 of the population per year and constitutes about 6% of first strokes.
- The relative risk for women is 1.3.
- SAH reported incidence declined by 40.6% in Europe between 1980 and 2010.
- This reduction may in part be attributed to a higher rate of CT scanning, excluding other haemorrhagic causes, although there is a clear correlation with the incidence of hypertension and smoking.
- Approximately 85% of patients bleed from intracranial arterial aneurysms, 10% from a non-aneurysmal peri-mesencephalic haemorrhage and 5% from other vascular abnormalities including arteriovenous malformation, vasculitis and abnormal blood vessels associated with tumour.
- SAH represents only 6% of cases of stroke but it is relatively far more important, as it tends to affect younger people, of whom about half die in that episode.
- The mean age is 50 years: most patients are under 60 years.
- Women have a higher risk than men; relative risk 1.6.
- Patients of Afro-Caribbean descent have a higher risk than white Europeans: relative risk 2.1:1
- Incidence is significantly higher in Finland and Japan.
- Spontaneous SAH is usually due to aneurysmal rupture but traumatic brain injury is a more common cause of blood in the subarachnoid space. Subarachnoid blood can be detected on CT scanning in as many as 60% of people with traumatic brain injury.

Risk factors

- Risk factors for spontaneous SAH are the same as for stroke in general: genetic factors operate in only a very small proportion of cases.
- The bigger the aneurysm, the more likely it is to bleed. However, as about 90% of aneurysms are small, the majority that bleed are less than 1 cm in diameter[3].
- Hypertension - the global SAH incidence declined with every millimetre of mercury decrease in systolic blood pressure by 7.1%[2].
- Smoking - the global incidence of SAH dropped by 2.4% for every 1% reduction in smoking prevalence[2].
- Cocaine use is a risk factor and is also associated with a worse outcome[4-5].
- Excessive alcohol intake roughly doubles the risk - its role has not been as well established as that of smoking, but drinking more than 37.5 units a week accounts for up to 1 in 5 cases of subarachnoid haemorrhage[6].
- Modifiable risk factors account for around two thirds of SAH[7].
- A combination of cigarette smoking and hypertension also increases the risk of a bleed from smaller aneurysms[8].
- Although no single gene has been isolated, genetic factors also play a part and account for around 10%[9].
- Linked genetic disorders include autosomal dominant adult polycystic disease, Ehlers-Danlos syndrome type IV and neurofibromatosis type 1. Berry aneurysms are found in 10% of patients with autosomal dominant adult polycystic kidney disease and represent 2% of cases of SAH.
- Marfan's syndrome has also been linked to SAH through the presence of berry aneurysms, although epidemiological studies have suggested that neurovascular complications are rare in this disorder[6, 11].
- Patients with a positive family history tend to have their first SAH at a younger age and are also more likely to have large and multiple aneurysms.
- First-degree relatives have a 3-7 times relative risk compared to the general population, but for second-degree relatives there is no increase in risk[1].
- Most patients with large or multiple aneurysms are sporadic rather than familial.

A note on berry aneurysms[12]

- Berry aneurysms are common with a prevalence of approximately 4%.
- Most berry aneurysms under 7 mm do not rupture, but they grow unpredictably.
- 85% occur in the Circle of Willis.
- Multiple aneurysms are seen in 30% of patients.
- Most are saccular aneurysms. These are not congenital but develop over the course of life, being rare in children[1].
- It is not clearly understood why some adults develop saccular aneurysms at arterial bifurcations in the Circle of Willis but most do not. There may be variation in the susceptibility of the elastic lamina of the arterial wall to the known stressors of hypertension and smoking.

Presentation

The most characteristic feature is a sudden explosive headache. This may last a few seconds or even a fraction of a second. The patient may even look around and accuse someone of hitting them on the back of the head.
The headache is sometimes referred to as a thunderclap headache, although this is confusing as thunderclap headache is a nonspecific term. The term was introduced in 1986 by neurologists at the University of California, in a report of a woman aged 42 who had experienced several sudden headaches and was found to have an aneurysm that had not ruptured\textsuperscript{13}. There are other causes of thunderclap headaches, some of which are more common than SAH– including primary sexual headache.

- Sudden explosive headache may be the only symptom in a third of patients\textsuperscript{14}.
- Of patients who present with a sudden explosive headache as the only symptoms, around 10% have SAH\textsuperscript{15}.

SAH should be considered in any patient presenting with sudden-onset, severe and unusual headache with or without any associated alteration in consciousness.

It is important to note that the headache is not always severe but is usually, but not always, accompanied by nausea and vomiting\textsuperscript{16}.

- The lack of clinical features distinguishing reliably between SAH and more innocuous headache means that a brief hospital consultation is needed for ALL patients with an episode of severe headache that comes on in minutes. This approach serves the patient's best interests and is also cost-effective. The discomfort and cost of referring the 90% of patients with innocuous headache is outweighed by the enormous cost of overlooking a ruptured aneurysm\textsuperscript{1}.
- It is not unusual for SAH to be initially misdiagnosed as a migraine or tension headache, particularly in patients who lack other signs and symptoms. This can lead to a delay in obtaining a CT scan. In a 2004 study, this occurred in 12% of all cases and was more likely in people who had smaller haemorrhages and no impairment in their mental status. The delay in diagnosis led to a worse outcome\textsuperscript{17}.
- The headache is often diffuse. The dominant feature is the severity, often being described as the most severe ever experienced.
- It typically pulsates towards the occiput although this is not invariable.
- It often lasts a week or two.

Other presenting features

- It is difficult to suspect SAH without sudden headache, but with seizure or confusional state - there are many other more common causes of these presentations, but SAH should be on the list of differential diagnoses.
- Vomiting often occurs, although this does not distinguish it from other causes of headache.
- Seizures, occur in about 7%. When they do, they are highly suggestive of a haemorrhage.
- 1-2% of patients with SAH present with an acute confusional state\textsuperscript{1}.
- Neck stiffness and other signs of meningeal irritation may be present, although it usually presents around six hours after onset of SAH.
- Trauma may be confused with SAH if trauma has also occurred. Patients may also cause a motor vehicle accident as a result of SAH. SAH needs to be on the differential diagnostic list for patients with altered consciousness, headache or seizure after trauma, particularly if there is disproportionate headache or neck stiffness.
- SAH following head injury causes headache, decreased level of consciousness and hemiparesis. SAH is a frequent occurrence in traumatic brain injury, and carries a poor prognosis if it is associated with deterioration in the level of consciousness.

Warning symptoms and sentinel bleeds

There may be warning symptoms in the three weeks prior to SAH that represent small leaks. These are called sentinel bleeds or expansion of the aneurysm. These are usually headaches with the characteristics of SAH but which resolve by themselves without further symptoms. They are estimated to occur in 10-15% of patients.

- The most common symptoms are headache (48%), dizziness (10%), orbital pain (7%), diplopia (4%) and visual loss (4%).
- Signs may accompany these sentinel bleeds: sensory or motor disturbance (6%), seizures (4%), palsy (3%), bruits (3%) and dysphasia (2%).
- If a sentinel bleed is suspected, patients should be admitted urgently for investigations (treat as if an SAH has occurred).

Examination

- Conscious level: on admission to hospital two thirds have a depressed level of consciousness, of whom half are in coma. However, SAH patients can also walk into the surgery, complaining of sudden onset of headache.
- Neck stiffness may occur due to meningeal irritation by blood in the CSF, but it is not invariable.
- Ophthalmoscopy will show intracranial haemorrhages in around 15%, especially in those with a depressed level of consciousness.
- Isolated pupillary dilation with loss of light reflex may indicate brain herniation as a result of rising intracranial pressure.
- There may be focal neurological signs, suggestive of a stroke. Complete or partial palsy of the oculomotor nerve is well recognised, especially with rupture of aneurysms of the internal carotid artery at the origin of the posterior communicating artery.
- Intracranial haemorrhage may occur in response to the raised pressure and is more common in more severe SAH.
- Oculomotor nerve impairment may indicate bleeding from the posterior communicating artery.
- Hypertension is a risk factor for the condition but a marked rise in blood pressure may also occur as a sympathetic reflex following intracerebral haemorrhage. This sympathetic reflex can raise blood pressure to life-threatening levels, and surges of adrenaline (epinephrine) may contribute to associated cardiac arrhythmias which may both confuse the diagnosis and further threaten the patient.
- In 3% of cases cardiac arrest follows SAH.
- SAH in a person known to have seizures is suggestive of an arteriovenous malformation. New-onset seizures are more indicative of ruptured berry aneurysm.
Differential diagnosis

- Other causes of stroke.
- Meningitis (rarely features thunderclap headache).
- Trauma.
- Thunderclap headache of other aetiology.
- Primary sexual headache.
- Cerebral venous sinus thrombosis.
- Cervical artery dissection.
- Carotid artery dissection.
- Hypertensive emergency (severely raised blood pressure).
- Pituitary apoplexy (infarction or haemorrhage of the pituitary gland).

Investigations\cite{1, 18, 19}

The diagnosis of SAH cannot be made on clinical grounds alone.

CT scanning

- If SAH is suspected, CT scanning (without contrast) is the first line in investigation because of the characteristically hyperdense appearance of blood in the basal cisterns.\cite{6}
- CT without contrast will correctly identify 95-98% of cases, particularly if performed within 24 hours of onset. One study reported 100% sensitivity for CT performed within six hours of onset.\cite{20}
- The distribution of blood can give some indication of the location of the aneurysm.
- Every patient in whom SAH is suspected should have a CT scan at the earliest opportunity. This should be done immediately if the patient presents with sudden severe headache, and as soon as possible in all other cases.
- A false positive diagnosis of SAH on CT is possible in the presence of generalised brain oedema, which causes venous congestion in the subarachnoid space.

Angiography

- Cerebral panangiography continues to be the gold standard for detection, demonstration and localisation of ruptured aneurysms.\cite{6}
- After an SAH is confirmed, its origin needs to be determined. If the bleeding is likely to have originated from an aneurysm, the choice is between cerebral angiography (injecting radiocontrast through a catheter to the brain arteries) and CT angiography (visualising blood vessels with radiocontrast on a CT scan) to identify aneurysms. Catheter angiography also offers the possibility of coiling an aneurysm.
- Magnetic resonance angiography (MRA) is less invasive than cerebral panangiography. However, it may be less sensitive than conventional angiography.
- CT angiography is less time-consuming than MRA but its sensitivity decreases with aneurysm size.\cite{6}

Lumbar puncture

- CT is negative in 2% of patients with SAH.
- If the CT scan is negative but the history is suggestive, lumbar puncture should be undertaken, providing the scan shows no contra-indications. Around 3% of patients with a negative CT scan will prove, on lumbar puncture, to have had an SAH.\cite{21}
- Lumbar puncture to remove a CSF sample from the lumbar sac should ideally take place over 12 hours after the onset of the headache because if there are red cells in the CSF, sufficient lysis will have taken place during that time for bilirubin and oxyhaemoglobin to have formed.\cite{21}
- Spectrophotometry should be used to examine the CSF, to permit detection of small amounts of xanthochromia (yellow discolouration of the spinal fluid). The ‘three tube test’ (a decrease in red cells in consecutive tubes on visual inspection) is notoriously unreliable.

ECG

- An ECG may show changes. If these are wrongly interpreted as acute myocardial infarction and thrombolysis is given, the result will be disastrous.
- ECG changes are relatively common in SAH. They may include QT prolongation, Q waves, dysrhythmias and ST elevation.

Classification of subarachnoid haemorrhage

There are several grading scales for SAH, including the Glasgow Coma Scale (GCS).

Several different specialised scores are also used to evaluate SAH; in each, a higher number is associated with a worse outcome. These scales have been designed by retrospectively reviewing of patients against their outcomes. They include the Hunt and Hess score (1968), the Fisher Grading (based on CT appearance, 1980) and The World Federation of Neurosurgeons (WFNS) classification which uses GCS and focal neurological deficit.

A classification scheme has been suggested by Ogilvy and Carter (1998) to predict outcome and determine therapy. The system assigns one point for the presence or absence of each of five factors: age greater than 50; Hunt and Hess grade 4 or 5; Fisher scale 3 or 4; aneurysm size greater than 10 mm; and posterior circulation aneurysm 25 mm or more.\cite{22}
Management

Early management
Initial management of SAH aims to prevent further bleeding and to reduce the rate of secondary complications, such as cerebral ischaemia or hydrocephalus.

Re-bleeding is the most imminent danger; a first aim is therefore occlusion of the aneurysm. Endovascular obliteration by means of platinum spirals (coiling) is now the preferred mode of treatment, but some patients require a direct neurosurgical approach (clipping)\[3\].

Another complication is delayed cerebral ischaemia due to vasospasm; the risk is reduced with oral nimodipine and probably by maintaining circulatory volume. Hydrocephalus might cause gradual obtundation in the first few hours or days; it can be treated by lumbar puncture or ventricular drainage, dependent on the site of obstruction\[3\].

Supportive management
- Every patient should be referred to a specialist unit, usually neurosurgical, for investigation and, if appropriate, definitive treatment. This transfer should take place within 24 hours if appropriate.
- Patients should be under continuous observation in an intensive care unit, or in an intermediate care facility of a stroke or neurovascular unit\[6\].
- Staff should be experienced in assessing swallowing function to prevent pneumonia, a frequent complication after SAH and an independent risk factor for poor outcome\[6\].
- All necessary supportive care should be provided. This often includes intubation and ventilation in patients with depressed conscious level, together with nasogastric feeding. For most patients this will mean an intensive care bed.
- Analgesia and antiemetics are needed for conscious patients.

Prevention of vasospasm
- Vasospasm is a serious and common complication of SAH which can lead to ischaemic brain injury, and which can be fatal. It affects around a third of admitted patients.
- Calcium antagonists help to reduce vasospasm. Nimodipine 60 mg four-hourly is generally used, as it has been shown to improve outcomes\[23\].
- Nitroprusside (a potent vasodilator) and labetalol may be employed to treat hypertension; the level should be low enough to prevent re-bleeding whilst high enough to maintain cerebral perfusion.
- Statins have been suggested but meta-analyses have suggested that there is no benefit\[24\].
- Other novel treatments being tried include magnesium sulfate infusion and endothelial antagonists\[25\].
- Patients should not be given an antifibrinolytic agent or steroids.

Prevention of re-bleeding
- Re-bleeding is common, with a cumulative incidence by six months of 50%. If a ruptured aneurysm is left untreated, about one third of the patients who recover from the initial haemorrhage die because of recurrent bleeding within six months after SAH\[6\].
- When it will occur and in which patients are difficult to predict. It is prognostically very grim. Interventions are aimed at reducing this risk early.
- There are two options: neurosurgical clipping and endovascular coiling. Clipping requires a craniotomy followed by the placement of clips around the neck of the aneurysm. Coiling is performed through femoral catheterisation with platinum coils that obliterate the aneurysm by causing a blood clot to form in it\[5, 26\].
- Until coiling or clipping, systolic blood pressure should be kept below 180 mm Hg\[6\].
- Clipping was first introduced in 1938 by a Baltimore neurosurgeon, Walter Dandy, and endovascular coil embolisation in 1991 by Italian neurosurgeon, Guido Guglielmi.
- A meta-analysis of studies of early (within 72 hours) versus late clipping showed significantly better outcomes in patients with a good clinical/neurological condition on admission, and a trend towards better outcomes in patients with worse clinical condition\[27\].
- The decision between the two is made by the operating team, depending on the location and accessibility of the aneurysm, its size and the condition of the patient. Aneurysms of the middle cerebral artery circulation are hard to reach with angiography and tend to be amenable to clipping. Those of the basilar and posterior cerebral circulation are harder to reach surgically and are more accessible for endovascular management.
- One study comparing clipping and coiling showed better outcomes for the latter, but was criticised for having a high proportion of patients in good clinical condition after SAH\[28\].
- Patients with large haematomas causing focal neurological symptoms or depressed consciousness may benefit from very urgent evacuation of the clot, with obliteration of the aneurysm at the same time. In other patients, stabilisation may take initial priority.
- Insertion of an endovascular platinum coil is increasingly being used for posterior circulation aneurysms, as evidence indicates that this has less associated mortality than clipping\[29\].
- The use of stents to facilitate coil insertion is also being explored.

Prevention of seizures
- About 7% of patients have seizures at onset and another 10% have seizures in the first few weeks but their impact on prognosis is unclear.
- The use of anti-epilepsy medication for the prevention of seizure is controversial. It has been associated with worse outcomes, although it is unclear whether this is due to selection bias, as patients with seizures have worse outcomes\[29, 30, 31\]. It is not recommended in the European Stroke Organisation guidelines\[8\].
Prognosis

- Cumulative case fatality rates over time after SAH are as follows: day 1: 25-30%; week 1: 40-45%; first month: 50-60%; sixth month: 55-60%; year 1: 65% and year 5 after SAH: 65–70%. Approximately 12% of patients die before reaching medical attention[6].
- Up to 60% of deaths occur in the first month. 10% die immediately without any warning symptoms. Re-bleeding, a major complication, carries a mortality rate of 51-80%.
- Delayed cerebral ischaemia due to vasospasm affects 20%.
- Long-term prognosis is related to initial presentation.
- The variables most closely related to outcome are neurologic condition of the patient on admission, age (younger patients do better) and the amount of subarachnoid blood on the initial CT scan. Of these, the initial level of consciousness is by far the most important.
- Improvement tends to occur between 4 and 18 months after SAH but even those who have independent living often have some cognitive impairment or sequelae.
- 46% of people who have had an SAH have cognitive impairment that affects their quality of life.
- Over 60% report frequent headaches.
- More than a quarter of people with a previous SAH may develop some degree of hypopituitarism.
- In a survey of 610 patients who were interviewed a mean of 8.9 years after SAH, there was marked morbidity[33]:
  - Of employed patients, 26% stopped working altogether and 24% modified or reduced their work commitment.
  - On average, patients returned to work 9.4 months after discharge. Related problems caused divorce in 7%.
  - There were changes in personality in 59%, with the most common being increased irritability (37%) or emotionality (29%).

Complications[1]

- Not all patients can be saved. The overall death rate is still around 50%, including those who die pre-hospital. Irreversible brain damage can occur immediately after aneurysm rupture. This may occur through bleeding, through hydrocephalus, through brain stem dysfunction or through prolonged cerebral ischaemia at the time of the bleed. This latter condition is thought to occur as a result of the pressure in the CSF spaces being elevated to the level of that in the arteries, for as long as a few minutes.
- In about 3%, cardiac arrest occurs at the onset but, of those successfully resuscitated, about half can leave hospital for an independent existence.
- The risk of later cerebral ischaemia is reduced with oral nimodipine and probably by maintaining circulatory volume. The peak time for occurrence is 5-14 days after the haemorrhage.
- Hydrocephalus might cause gradual reduction in conscious level in the first 24 hours or more. If untreated the outcome is poor. It can be treated by lumbar puncture or ventricular drainage, combined with fibrinolysis through the drain site, depending on the site of obstruction.
- Abnormalities of biochemistry are common and need appropriate management in intensive care.
- 5% or more of patients develop epilepsy after discharge.
- In the first few hours after admission for the initial haemorrhage, up to 15% of patients have a sudden episode of clinical deterioration that suggests re-bleeding.
- Intra-parenchymal haematomas occur in up to 30% of patients with ruptured aneurysms, and have a worse outcome than SAH alone.
- A sudden deterioration in level of consciousness within the first few hours suggests further bleeding and occurs in around 15%. It carries a mortality rate of 51-80%.
- At present it is virtually impossible to prevent this from happening, although medical or surgical intervention can prevent recurrent haemorrhages occurring.
- In patients who survive the first day, the risk of re-bleeding is evenly distributed over the next four weeks. The total risk of re-bleeding without medical or surgical intervention in the four weeks after the first day is estimated at 35-40%.
- Between four weeks and six months after the haemorrhage, the risk of re-bleeding gradually decreases from the initial level of 1-2% a day to a constant level of ~3% a year.
- Treatment with antifibrinolytic drugs does appear to reduce the re-bleed rate, but does not improve the overall outcome.

Further management

- All surviving patients should be advised on secondary prevention, especially on treatment for hypertension and the need to stop smoking.
- 10-15% of patients with SAH do not survive as far as hospital.
- Any patient with residual impairment after investigation and treatment should be referred to an appropriate specialist rehabilitation service.
- Patients with a strong family history (one other affected first-degree relative and/or with a history of polycystic kidney disease) should be advised that their immediate family may be at greater risk of SAH, and a referral made to a neurovascular specialist for up-to-date information and advice.

Ventricular drainage

- Hydrocephalus may complicate SAH both in the short and in the long term. Relief of hydrocephalus can cause an enormous improvement in a patient’s clinical status.
- Half of patients will improve spontaneously and there is a risk of re-bleeding and infection. Therefore some neurosurgeons prefer not to use a ventricular drain immediately[8].
- Surgical intervention may be contra-indicated in patients who fail to respond to medically stabilising measures, in those over the age of 80, in those with other medical problems or where the aneurysm is large or complicated.
- All patients should be monitored for the development of treatable complications, especially hydrocephalus, cerebral ischaemia, electrolyte imbalance and hypotension.
Patients with SAH had a statistically significant higher mean depression score than the control population. Approximately 10% of the patients had a hospital anxiety and depression (HAD) score in the range of a probable depressive or anxious state. Only 25% reported a complete recovery without psychosocial or neurological problems. Patients with traumatic brain injury with SAH have as much as twice the risk of dying as those who do not, and a higher risk of severe disability and persistent vegetative state. However, more than 90% of people with traumatic subarachnoid bleeding and a GCS over 12 have a good outcome.

Prevention

As hypertension, smoking and excessive alcohol consumption are risk factors, individuals need to address such issues. The question of management of known aneurysms falls into three groups.

Patients found to have incidental aneurysms during other investigations

The management of incidental findings will depend upon many aspects. The risk of rupture increases with age but so does morbidity of the procedure and younger people stand to gain more years. It is a difficult decision and family history and patient wishes must be considered.

Patients with SAH who have one unruptured aneurysm or more

- Those who have already had an SAH are usually offered treatment, although if it is small or difficult to reach, it may be left alone.
- One study found that predictive factors for rupture of an aneurysm were width:height ratio, maximum diameter of neck (≤3 mm) and family history of cerebrovascular disease.

Screening for aneurysms in patients who survive an SAH episode and in first-degree relatives of patients with SAH

The results of screening first-degree relatives of patients for aneurysms are not promising and few apparently dangerous lesions are found. Even routine screening of people with adult polycystic kidney disease is not recommended.

Further reading & references


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