# Intracranial Aneurysm (Cerebral Aneurysm) — Symptoms and Differential Diagnosis

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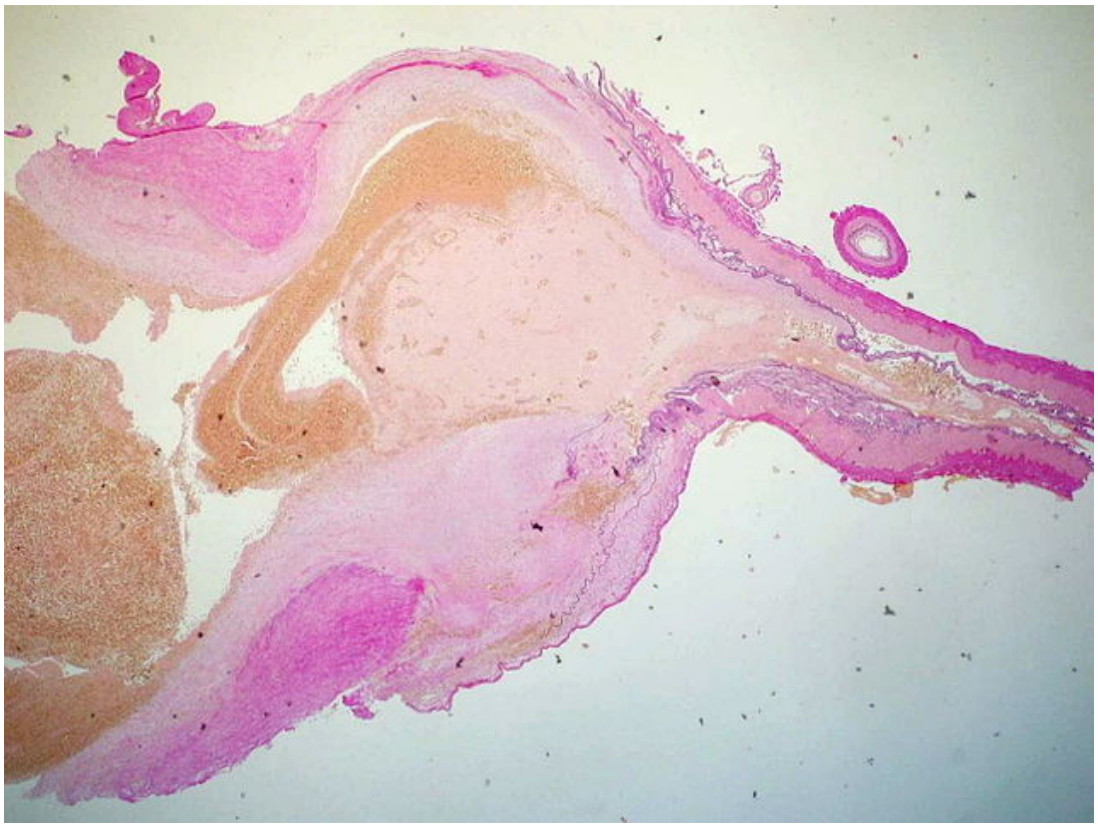
**This article looks at the basic science, diagnosis and treatment of ruptured and unruptured cerebral aneurysms. It provides a brief overview of cerebral aneurysm management.**



## Definition and Epidemiology of Intracranial Aneurysms

A cerebral aneurysm is an **abnormal dilation** of a local area of the artery wall in the central nervous system (CNS). The term is commonly used interchangeably with intracranial aneurysm. Most occur at junction points of the major arteries of the brain. This is usually around the **circle of willis**. They can either compress adjacent structures or rupture and cause hemorrhagic stroke.

## Prevalence of intracranial aneurysms

Studies done at autopsy show that 1 – 5% of the population suffer from unruptured

intracranial aneurysms.

The majority of aneurysms occur around the circle of Willis, which makes up the major blood supply to the **brainstem** and supplies most of the cortex.

Around 40% of aneurysms arise at the junction between the anterior cerebral artery and the anterior communication artery. 20% occur at the junction of the posterior communicating artery and the middle cerebral artery; 34% occur at branches of the middle cerebral artery and 4% occur at the junction between the two posterior cerebral arteries and the basilar artery.

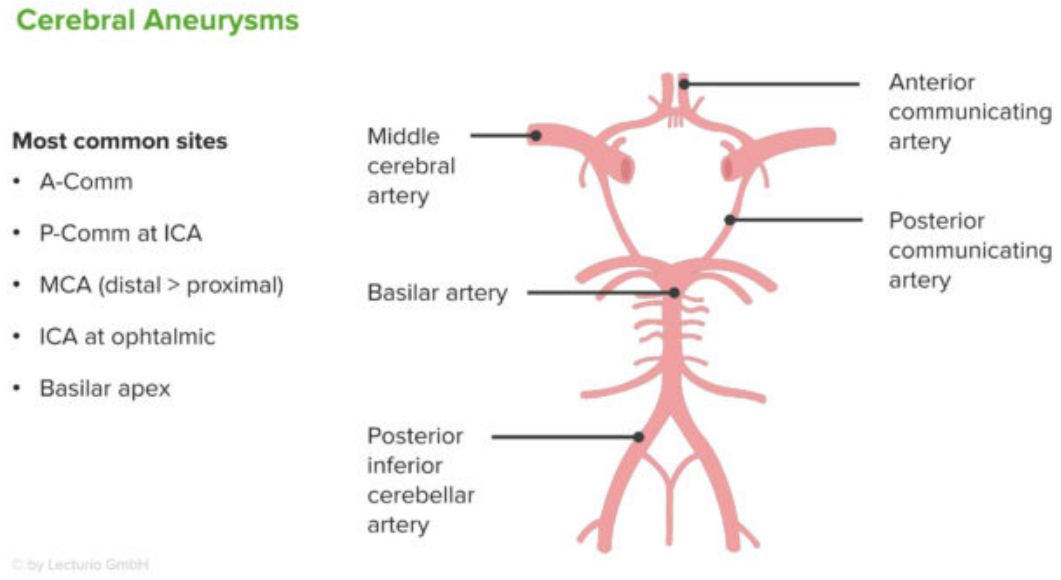
## Incidence of hemorrhagic stroke from intracranial aneurysms

Incidence of ruptured aneurysms is around 10 in 100,000 people per year. **Adults** and **women** are at an increased risk of developing cerebral aneurysms, as are those with a **family history** of them.

# Etiology of Intracranial Aneurysms

Cerebral aneurysms are an **acquired defect** typically resulting from **hemodynamic stressors**. However, they can arise as a result of **infections**, **tumors,** and **trauma**.

A **genetic component** is likely as a family history is a risk factor for the development of an intracranial aneurysm. Patients with **inherited connective tissue disorders** like polycystic kidney disease, neurofibromatosis type 1, Ehlers Danlos syndrome type IV and Marfan's syndrome have also been shown to have an increased risk of intracranial aneurysm development, and this should be taken into account during history in a patient with a suspected hemorrhagic stroke.




# Classification of Intracranial Aneurysms

Intracranial aneurysms are **classified by size and shape**. Their shape is split into three groups:

- **Saccular aneurysms**
    - Typically referred to as Berry aneurysms, these present as an outpouching of an artery and are named as such due to their resemblance with berries hanging off a branch.
- **Fusiform aneurysms**
    - A fusiform aneurysm causes widening of the entire blood vessel – rather than just one localized segment. These rupture less often.
- **Charcot Bouchard aneurysms (microaneurysms)**
    - These are associated with hypertension and form in small arteries, typically those in the lenticulostriate vessels of the basal ganglia. These commonly cause hemorrhage.

## Pathophysiology of Intracranial Aneurysms

Pathophysiologically, aneurysms form when there are **weaknesses in the walls of the arteries** supplying the CNS. However, it is less easy to ascertain how these form and the etiology of different classifications and sizes of aneurysms may be different.

Many hypotheses have been suggested for the **link between smoking and saccular aneurysms**. It has been suggested that inhibition of proteases (by smoking) causes the loss of connective tissues in the arterial walls. The pressure at these points along the arteries causes outpouching and development of aneurysms.

The obvious **progression** of a cerebral aneurysm is a **rupture**.

## Symptoms of Intracranial Aneurysms

Many patients will have **no symptoms** associated with an unruptured aneurysm. However, an outpouching of the vessel can press on surrounding structures and cause various symptoms. Patients can experience:

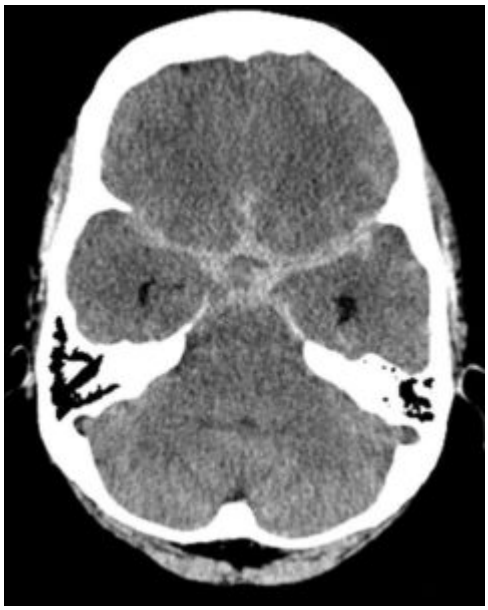


Image: “Subarachnoid hemorrhage in CT. One can see the blood hyperattenuating in the basal cisterns.” by Hellerhoff. License: CC BY-SA 3.0

- Loss of vision

- Double vision
- Pain around the eye
- Numbness/weakness of one side of the face
- Difficulty speaking
- Headaches
- Loss of balance (vertigo)

Aneurysms can rupture, typically causing a **subarachnoid hemorrhage**. This is often described in patients as “the worst headache of their life” and its onset will be sudden. Patients will often also experience:

- Photophobia
- Loss of consciousness
- A third nerve cranial palsy (this causes deviation of the eye out and downwards, as the third cranial nerve controls a number of muscles in the eye. Patients can also present with full ptosis. This occurs when a posterior communicating artery aneurysm compresses the ipsilateral third cranial nerve).
- Nausea.
- Vomiting.
- Unilateral or bilateral sixth cranial nerve palsies (this presents as double due to loss of lateral rectus. It indicates a raised intracranial pressure often caused by a bleed).
- Focal neurological deficits.

# Diagnosis of Intracranial Aneurysms

## Diagnosis of an unruptured cerebral aneurysm

Diagnosis of an unruptured cerebral aneurysm may be difficult as patients may present with vague and unspecific symptoms. A full history should be taken whereby risk factors for cerebral aneurysms should be obtained. Key risk factors are:

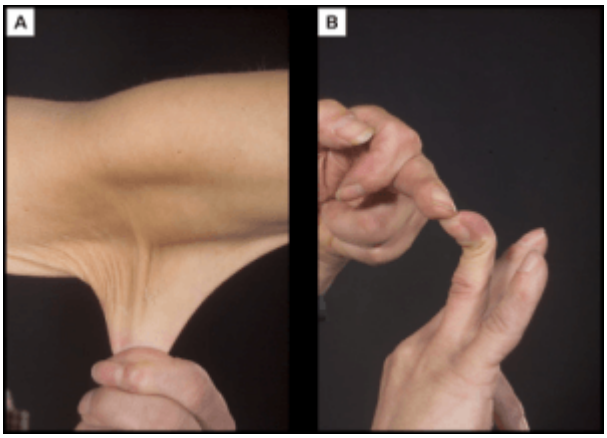



Image: “(A) Prominent hyperextensible skin (B) hypermobile joints in Ehlers Danlos syndrome.” by Openi. License: CC BY 2.0

- Smoking
- Alcohol
- Previous diagnosis of a subarachnoid hemorrhage
- A family history of subarachnoid hemorrhage

- Heritable connective tissue disease Marfan's, Ehlers Danlos syndrome, pseudoxanthoma elasticum and neurofibromatosis type 1.

Other weaker risk factors can include:

- Hypertension
- Head trauma
- Infection (intracranially)
- Tumor
- Drug abuse

Common symptoms were described above. The patient will typically give a **history of a headache**. They may have had **seizures** or have some **nuchal rigidity**. A **decreased level of consciousness** can be found in some patients.

**The examination** is typically **unremarkable**. In patients with strong risk factors and unexplained symptomatology, it may be prudent to order a number of diagnostic investigations. Investigations that may aid diagnosis are:

- **CT head** scan can show blood in the subarachnoid space from leaking or a ruptured aneurysm. If the aneurysm has calcified, it may also be seen.
- **MRI** can show unruptured aneurysms.
- **Lumbar puncture** can show elevated red blood cell count with xanthochromia
- **CT angiography** to locate aneurysm location and size
- One can also use **magnetic resonance angiography**

## Diagnosis of a ruptured cerebral aneurysm

Patients whose aneurysm has ruptured will have a different presentation from those who have unruptured aneurysms. If a patient is suffering from a **sudden onset severe headache**, a subarachnoid hemorrhage (SAH) should be ruled out. Typically, these patients will also suffer from **vomiting** and **loss of consciousness** – **coma** and **death** can occur quickly and so suspected SAH is considered a **medical emergency**.

Other symptoms will include **neck stiffness** (nuchal rigidity) and **Kernig's sign**.

**CT head** scan is your first port of call with any patient with severe onset headache. SAH is usually seen on a CT and has a sensitivity of 95% within 24 hours. If a CT confirms a subarachnoid hemorrhage, then lumbar puncture is not necessary, although in some cases where CT is inconclusive this may be indicated.

On **lumbar puncture,** you typically find **xanthochromic CSF** (yellowing of the CSF). **Visual inspection of the CSF** is usually sufficient for diagnosis.

## Differential diagnosis

**Differential Diagnosis for an unruptured cerebral aneurysm**

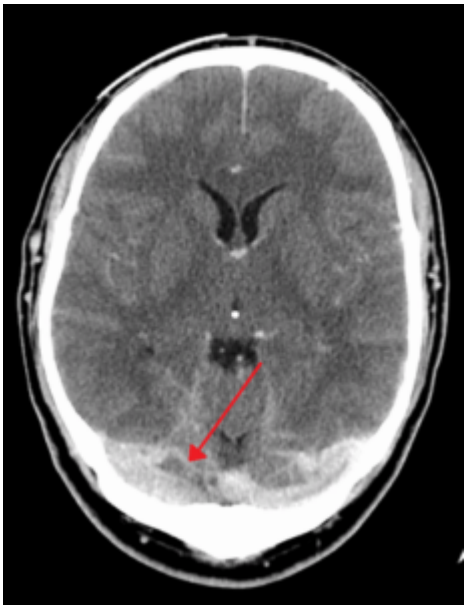


Image: “A dural venous sinus thrombosis of the transverse sinus. Greater on the right than left.” by James Heilman, MD – Own work. License: CC BY-SA 4.0

- Arteriovenous malformation
    - Usually has a subacute presentation
    - Can also present with seizures
    - A cerebral angiography or MRI
- Hypertensive intracerebral hemorrhage
    - This typically occurs in patients above 55 years old. They will have a history of **hypertension**. Hemorrhage causes focal neurological signs.
    - A CT head scan should be ordered to rule out a bleed
- Cerebral venous sinus thrombosis
    - This typically presents in middle-aged women. They will have **papilloedema** on examination and describe a subacute history (hours to days).
- Traumatic **subarachnoid hemorrhage**
    - Usually seen in older patients who had a fall. A CT head scan can rule this out.
- Hemorrhagic tumor
    - The patient usually has a history of some malignancy.

**Differential Diagnosis for subarachnoid hemorrhage**

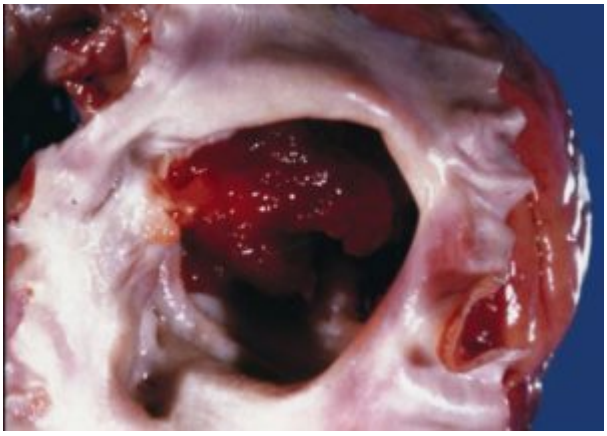


Image: “A gelatinous tumor is attached by a narrow pedicle to the atrial septum. The myxoma has an irregular surface and nearly fills the left atrium.” by The Armed Forces Institute of Pathology (AFIP) – PEIR Digital Library (Pathology image database). Image# 410012. License: Public Domain

- Arterial dissection
    - Patients will not describe as excruciating pain. Pain is often localized behind the eye or to the neck.
- Non-aneurysmal perimesencephalic SAH
    - Very difficult to differentiate from a SAH.
    - No aneurysms are found with angiography.
- Cerebral arteriovenous malformation
    - Can be seen with cerebral angiography (or CT).
- Vasculitis
    - A headache is usually less severe. The neurological symptoms are recurrent.
- Saccular aneurysms of spinal arteries.
- Cardiac myxoma
- Septic aneurysm
- Pituitary apoplexy

# Therapy of Intracranial Aneurysms

## Management of unruptured aneurysms

Patients can either be treated **conservatively** and be **observed**, or **surgery** can be performed. Treatment is either by “**clipping**” or **endovascular obliteration**. This decision is made on a case by case basis.

When making the decision, a number of things should be considered. These include the **patient’s age** (older patients with less life expectancy are more likely to be given observation), the **size and location of the aneurysm** (some aneurysms are very unlikely to rupture e.g. cavernous carotid artery aneurysm and observation may trump treatment due to the risk associated with clipping, etc).

The **risks of treatment** should also be considered: rupturing the aneurysm during surgery can lead to hemorrhage and ultimately death.

## Management of ruptured aneurysms

Patients with a ruptured aneurysm are considered a **medical emergency**. **Hunt and Hess grading** should be done as this impacts treatment severity and swiftness. Hunt and Hess's grades are used to assess how severe a subarachnoid hemorrhage is and are as follows:

1. Asymptomatic, the patient has a mild headache with some nuchal rigidity.
2. Moderate to a severe headache with nuchal rigidity.
3. Drowsiness, mild focal neurological deficit, patients are often confused.
4. Patient is in a stupid coma, decerebrate posture.

Give **calcium channel blockers** (to reduce vasospasm) and maintain a systolic blood pressure press 135 mmHg. **Clipping** or endovascular therapy should then be considered for any unruptured aneurysms. **Endovascular therapy** is preferable.

# Complications of Intracranial Aneurysms

Patients with an unruptured cerebral aneurysm are at significant risk of **rupture** and **death**. Patients can also develop **hyponatremia**, experience **seizures** and **hydrocephalus**. In the longer term, aneurysms may recur.

# Prevention of Intracranial Aneurysms

Prevention is done via **screening for unruptured aneurysms**. **Magnetic resonance angiography (MRA)** is indicated in patients with two+ family members who have been diagnosed with cerebral aneurysms. Screening is also performed for patients with **heritable connective tissue disorders** like Ehlers Danlos syndrome. Patients who have had a **previous history of aneurysms** should also be screening periodically (every few years).

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Notes