B.3. Diagnosis and Management of Headaches

a. Major Causes of Intracranial Hemorrhage.

i. Know the major causes of intracranial hemorrhage: Vasculopathy in the aged (hypertension and amyloidosis), aneurysm, vascular malformation, tumor, and coagulopathy.

aa. Hypertension.

Spontaneous intracerebral hemorrhage may occur with both acute and chronic hypertension. Acute hypertension sufficient to produce spontaneous intracerebral hemorrhage is sometimes seen with eclampsia and drug intoxication with sympathomimetic drugs. Chronic hypertension may lead to vascular changes of the basal perforating arteries including formation of Charcot-Bouchard aneurysms. The most common locations for these hemorrhages are (in decreasing order of frequency):

- Basal ganglia
- Thalamus
- Pons
- Cerebellum
- Cerebral white matter
- Brainstem

bb. Amyloid angiopathy.

This is characterized by a deposition of beta amyloid protein within the meningeal and cortical vessels. The clinical course is characterized by multiple lobar intracerebral hemorrhages. Risk factors include advanced age and Down's syndrome. Definitive diagnosis requires pathologic examination.

cc. Aneurysm.

Cerebral aneurysms result from weakening of the wall of major intracranial arteries. These aneurysms are generally saccular, though fusiform aneurysms also occur. Saccular aneurysms usually occur at branch points along the arterial tree. When a saccular aneurysm ruptures, it most commonly produces subarachnoid hemorrhage. Intracerebral, intraventricular, and subdural hemorrhage are also seen. Common sites of aneurysm rupture include (in decreasing order of frequency)

- Anterior communicating artery
- Posterior communicating artery
- Middle cerebral artery
- Basilar artery
- Vertebral artery

Multiple aneurysms are seen in approximately one-fourth of patients. Risk factors include hypertension, smoking, family history, and collagen vascular disease.

dd. Vascular malformation.

Vascular malformations include the high flow arteriovenous malformation, cavernous angioma, venous angioma, and capillary telangiectasias.

- The high flow arteriovenous malformation results from a direct communication between the arterial blood supply and draining veins without normal interposed capillaries. These are usually visible on arteriography. Saccular

aneurysm are frequently seen on feeding vessels. Intraparenchymal hemorrhage is most commonly seen, though subarachnoid and subdural hemorrhage may occur.

- Cavernous angiomas consist of irregularly formed vascular channels, without intervening brain parenchyma. Flow is usually low and these lesions, therefore, do not routinely manifest themselves on arteriography. Hemorrhage, when it occurs, is typically intraparenchymal. Family history is a risk factor.
- Venous angiomas result from a cluster of normal medullary veins draining into a central enlarged venous channel. The arterial system is uninvolved. Hemorrhage is infrequent and usually intraparenchymal. Many are associated with cavernous angiomas.
- Capillary telangiectasias are clusters of dilated capillaries with low flow. They are not visible on arteriography. They are sometimes associated with Osler-Weber-Rendu syndrome, Louis-Barr syndrome, Myburn-Mason syndrome, and Sturge-Weber syndrome.

ee. Tumor.

Both primary and metastatic tumors may hemorrhage. Of the malignant primary brain tumors, glioblastoma multiforme most commonly presents with hemorrhage. While lung carcinoma is the most common hemorrhagic cerebral metastasis, melanoma, choriocarcinoma, and renal cell carcinoma are the most likely metastases to present with cerebral hemorrhage. Most hemorrhages are intraparenchymal.

ff. Coagulopathy.

Even in the absence of underlying cerebral pathology, coagulopathy, either secondary to underlying medical disease or iatrogenically induced, increases the risk for intracranial hemorrhage. Particularly in elderly patients, subdural hematoma is the most common manifestation. Intraparenchymal hemorrhage may also occur, particularly in the setting of anticoagulation after cerebral infarction.

b. Recognize the Symptoms and Signs of Subarachnoid, Cerebral, and Cerebellar Hemorrhage.

i. Subarachnoid hemorrhage.

The most common symptom of subarachnoid hemorrhage is explosive onset of severe headache. The headache is described by the patient as "the worst headache of my life". The patient may complain of neck pain and stiffness, as well as photophobia. Low back pain may also be present. Nausea and vomiting may be present. There may be a history of transient loss of consciousness.

Signs of subarachnoid hemorrhage include nuchal rigidity, positive Kernig’s sign, and/or positive Brudzinski’s sign. A depressed level of consciousness may also be present. Ocular manifestations include hemorrhages and papilledema (secondary to elevated intracranial pressure). Focal neurologic deficit, particularly third nerve palsy ipsilateral to the site of aneurysm rupture, may be present and in some instances may help to localize the aneurysm.

ii. Intracerebral hemorrhage.

Headache may be present after intracerebral hemorrhage. Patients with nondominant hemorrhages may complain of weakness or numbness on their nondominant side.

Signs of intracerebral hemorrhage are typically those of hemispheric neurologic deficit. Patients with dominant-sided lesions are frequently found to have hemiparesis and hemianesthesia. Dysphasia is usually present, though the patient may present with frank aphasia. Patients with nondominant lesions typically have contralateral hemiparesis and hemianesthesia. While speech is frequently normal, neglect may be profound.

If the hemorrhage is large, the patient may present with signs of elevated intracranial pressure including depressed level of consciousness, decorticate or decerebrate posturing, or flaccid areflexia.

iii. Cerebellar Hemorrhage.

The awake patient with cerebellar hemorrhage may complain of severe headache, which may be localized to the suboccipital or upper cervical region.

Symptoms include those of cerebellar and lower brainstem dysfunction, as well as hydrocephalus, which is frequently present. Signs of cerebellar dysfunction include ipsilateral dysmetria, nystagmus when looking to the side of the lesion, and difficulties with speech. Lower brainstem dysfunction typically manifests as difficulty with swallowing. In the presence of hydrocephalus, signs of elevated intracranial pressure may ensue. These include a decreased level of consciousness, decorticate or decerebrate posturing, and flaccid areflexia.

c. Apply Diagnostic Tools in Evaluation of Acute Headache (CT, MRI, and Lumbar Puncture).

In the patient who presents with the "worst headache of my life" a subarachnoid hemorrhage should always be suspected. After initial resuscitation and complete neurologic examination, head CT without contrast is mandatory. In addition to verifying the presence or absence of subarachnoid blood, the CT scan should be carefully scrutinized for other intracranial pathology, including hydrocephalus.

If the CT scan is positive for subarachnoid hemorrhage, the patient should undergo cerebral arteriography. The arteriogram should be scrutinized for the location of the aneurysm, as well as for any associated lesions including additional unruptured aneurysms, arteriovenous malformations, and the presence or absence of vasospasm.

If the initial head CT is negative for subarachnoid hemorrhage, and the patient has a history that is highly suspicious for subarachnoid hemorrhage, a lumbar puncture should be performed. A small needle should be utilized and only a small volume of fluid should be withdrawn. Non-clotting blood, xanthochromia, or a red blood cell count greater than 100,000 which does not drop significantly from the first to the last tube are all highly suggestive of subarachnoid hemorrhage. If the lumbar puncture meets any of these criteria, the patient should undergo arteriography as described above.

If the cerebral arteriogram is negative, in the face of a CT scan indicative of subarachnoid hemorrhage or a positive lumbar puncture, the patient should be admitted to the hospital for observation. At this point, an MRI scan of the brain with and without contrast should be considered. MRI arteriography, if available, should be included. The purpose of this study is to rule out other structural causes

of subarachnoid hemorrhage including angiographically occult vascular malformations and tumor. If no bleeding source is identified with the MRI scan, most clinicians recommend repeating the arteriogram in 7 to 14 days. Occasionally, an intracranial aneurysm that was obscured by mass effect, thrombosis, or vasospasm, may be detected.

d. Understand the Natural History and Broad Treatment Strategies of Intracranial Aneurysms and Vascular Malformations.

i. Intracranial aneurysms - natural history.

aa. Ruptured intracranial aneurysms.

In the patient who survives their initial aneurysm rupture, the most serious complication is rebleeding. The rate of rebleeding is approximately 4% in the first 24 hours and decreases to 1 to 2% per day for the first two weeks. After the first two weeks, the rate of rebleeding drops to approximately 3% per year. A second potential serious complication of aneurysmal subarachnoid hemorrhage is a vasospasm. Vasospasm is defined as delayed narrowing of large and medium size arteries at the base of the brain. If severe, this may lead to cerebral infarction. The risk of symptomatic cerebral vasospasm is maximum between 4 and 11 days of subarachnoid hemorrhage. The incidence of vasospasm, detected by angiography, is approximately 70%. The incidence of symptomatic vasospasm (resulting in neurologic deficit) is approximately 25 to 30%. The major risk factor for the development of vasospasm is the amount and location of subarachnoid blood visualized on the CT scan. This is graded using the Fisher system.

Group	Blood on CT Scan
1	No blood detected
2	Diffuse or vertical layers < 1mm thick
3	Localized clot and/or vertical layer >= 1mm thick
4	Intracerebral or intraventricular clot with diffuse or no SAH

bb. Unruptured intracranial aneurysm - natural history.

Occasionally aneurysms are discovered prior to rupture. Again, the primary risk is rupture. The available information suggests that the annual rate of rupture for an unruptured intracranial aneurysm is between 1% and 3% per year. The risk of rupture is affected by several factors. Larger aneurysms, particularly those greater than or equal to 10 mm in diameter, have a higher rate of rupture. Aneurysms located in the posterior communicating artery, vertebrobasilar/posterior cerebral artery, and in the basilar tip are more likely to rupture.

cc. Treatment strategies for intracranial aneurysms.

The goal of intracranial aneurysm treatment is to prevent bleeding or rebleeding. Additionally, in the patient with a ruptured intracranial aneurysm, treatment is also instituted to lessen the risk of symptomatic vasospasm.

In patients with subarachnoid hemorrhage due to aneurysm rupture, outcome is related to the modified Hunt-Hess grade. Higher-grade patients have a statistically poorer outcome.

Grade	Description
0	Unruptured aneurysm
1	Asymptomatic, or mild headache and slight nuchal rigidity

1a No acute meningeal/brain reaction, but with fixed neurologic deficit
2 Cranial nerve palsy (e.g. III, IV), moderate to severe headache, nuchal rigidity
3 Mild focal deficit, lethargy, or confusion
4 Stupor, moderate to severe hemiparesis, early decerebrate rigidity
5 Deep coma, decerebrate rigidity, moribund appearance
*Add one grade for serious systemic disease (e.g. HTN, DM, severe atherosclerosis, COPD), or severe vasospasm on arteriography

The patient with aneurysmal subarachnoid hemorrhage should be admitted to the neurological intensive care unit. Adequate oxygenation and respiration should be assured. The patient should be maintained normotensive. Hypertension should not be treated unless the systolic blood pressure rises above 160 mm of mercury. Hydrocephalus, which is present in at least 15% of patients with subarachnoid hemorrhage, should be treated with cautious external ventricular drainage if symptomatic. Routine use of external ventricular drainage is controversial. Cerebral selective calcium channel blockers lessen the risk of symptomatic vasospasm and are started at this time. Use of anti-fibrinolytic agents and corticosteroids may be considered.

Treatment of the ruptured intracranial aneurysm should at this point be entertained. Direct surgical exclusion of the aneurysm from the circulation (craniotomy and clipping) remains the "gold standard". Endovascular obliteration of the aneurysm through the use of detachable coils may be considered in certain situations (difficult or "unclippable" aneurysms, medically unstable patient). The long-term results of this modality of treatment are unknown and currently under study.

Treatment of unruptured aneurysms takes into account both the risk of rupture, as well as age of the patient, severity and progression of symptoms, and treatment alternatives. Older patients have a statistically shorter life span and therefore lower cumulative risk of rupture. Recent recommendations suggest that symptomatic intradural aneurysms of all sizes should be considered for treatment. Unruptured aneurysms of all sizes in patients with subarachnoid hemorrhage due to another treated aneurysm should be considered for treatment. Treatment for unruptured small (less than 10 mm) aneurysms is not generally recommended except in special circumstances. These include aneurysms near the 10 mm size, those with daughter sac formation, and in patients with family history of aneurysm or aneurysmal subarachnoid hemorrhage. Asymptomatic aneurysms greater than or equal to 10 mm in diameter warrant strong consideration for treatment.

ii. Arteriovenous Malformations - Natural History

aa. Risks.

The primary risk of arteriovenous malformations is rupture. The rate of rupture has been estimated to be approximately 4% per year for symptomatic arteriovenous malformations and 2% per year for asymptomatic arteriovenous malformations. The rate of arteriovenous malformation rupture appears to be higher in small malformations than in large malformations. The second risk of arteriovenous malformation is seizures. Nearly 60% of patients with an arteriovenous

malformation have a history of seizures. Larger malformations are associated with a greater risk of preoperative seizures.

bb. Treatment strategies for arteriovenous malformations.

Treatment for arteriovenous malformations is recommended when the risk of subsequent hemorrhage is greater than the risk of treatment. The risk of treatment may be estimated using the Spetzler-Martin classification:

Graded Feature	Points Assigned
Size of arteriovenous malformation	
Small (<3cm)	1
Medium (3-6cm)	2
Large (>6cm)	3
Eloquence of adjacent brain	
Noneloquent	0
Eloquent	1
Pattern of venous drainage	
Superficial only	0
Deep	1

Grade = [size] + [eloquence] + [venous drainage]

There are several treatment options for arteriovenous malformations. These include craniotomy and direct surgical excision, embolization using interventional radiologic techniques, and Gamma Knife radiosurgery. Because the risk of treatment rises with increasing Spetzler-Martin grade, the following recommendations have been made:

- Grade I or II arteriovenous malformations should be treated.
- Grade III lesions should be treated if symptomatic.
- Grade IV or V arteriovenous malformations should be treated only when significant or repetitive intracerebral hemorrhage has occurred or the patient is experiencing progressive neurologic disability.
- The treatment modality of choice remains controversial. Surgical removal remains the "gold standard". With all treatment modalities, the goal remains complete removal or obliteration of the arteriovenous malformation. This goal should be pursued until it has been achieved.

e. Differentiate the Symptomatology of Migraine, Cluster, and Tension Headache, and Sinusitis Headache.

i. Migraine headache. Features of classical migraine headache include:

- Onset in childhood, adolescence or early adulthood.
- Positive family history.
- More common in females.
- Usually unilateral, throbbing pain.
- Nausea and vomiting common.
- Scintillating scotomata.
- Duration: Several hours.

ii. Cluster headache.

- Much more common in men.
- Pain severe and retro-orbital or temporal, usually unilateral.
- Pain at night.
- Accompanied by lacrimation, rhinorrhea, periorbital edema.
- Duration of pain 20 to 60 minutes and may recur several times within a 24-hour period.
- Pain-free period of months to years is typical.

iii. Tension headache.

- Onset in adolescence or young adulthood.
- Non-familial.
- Pain is typically bilateral, bifrontal, bitemporal, or suboccipital.
- Pain described as throbbing or a sensation of "pressure".
- Nausea and vomiting rare, no lacrimation or rhinorrhea.
- More frequent in afternoon or evening.
- Duration hours to days.
- Precipitated by stress, depression, or anxiety.

iv. Sinusitis.

- History of allergic background or frequent prior sinusitis.
- Unilateral or bilateral.
- Localized pain and tenderness over affected sinuses common.
- Associated conditions include nasal obstruction, fevers, and chills. No history of scintillating scotomata, nausea, or vomiting.
- Headache persists until sinusitis is cleared.