Introduction
Brain and spinal cord vascular malformations can be separated into five main categories: 1) Arteriovenous malformation, 2) Capillary malformation, 3) Venous malformation, 4) Cavernous malformation (angioma) and 5) Arteriovenous fistula.

Arterial Venous Malformations

Overview & Epidemiology
Cerebral arteriovenous malformations (AVM) are congenitally formed complex lesions formed from abnormal development between the arterial and venous structures. They are characterized as aberrant connections between arteries and veins without an intervening capillary bed. Formation and maturation of AVMs lead to a state of high flow arterial to venous shunting via an intervening dysmorphic vascular nidus. Without the normal intervening capillary bed to act as a vascular buffer, arterial to venous shunting leads to the presenting symptoms such as headache, seizure, cerebral hemorrhage, cerebral ischemia, or progressive neurologic injury.

Natural History
The risk of spontaneous hemorrhage from a previously asymptomatic AVM is estimated to range from 1.5-4.0% annually (1,2). However, once the AVM has ruptured, the risk of recurrent hemorrhage is higher, estimated at 6% during the first year following rupture and then returns to an annual risk of 2-4%. Certain factors have been proposed to increase the likelihood of AVM hemorrhage such as: 1) size of the AVM nidus, 2) presence of a flow related aneurysm, 3) AVM location, 4) nidus characteristics, 5) venous draining pattern, and 6) draining vein stenosis. Of note, smaller sized (nidus) AVMs can have higher feeding artery pressures that may protect them from hemorrhage, but can predispose to cerebral steal syndromes and seizures.

Pathology
AVMs have three main structural components: 1) Feeding arteries, 2) core of dysplastic vessels (nidus), and 3) draining veins. The AVM nidus is a tangle of dysplastic vessels that replaces the normal capillary bed that buffers arteries and veins. Since the nidus is exposed to higher pressures than normal capillary architecture it is often the site of AVM hemorrhages. Other typical features is the absence of normal intervening brain tissue within the nidus and a gliotic rim (around the nidus) separating it from normal brain.
Clinical Presentation
Patients with AVM typically present in their middle years (20-50), with an unprovoked intracerebral hemorrhage (ICH); in the absence of a clear etiology such as hypertension, substance abuse, coagulopathy, AVM should be considered as a possible etiology. Intracranial hemorrhage is the most common presentation (50%) of patients with an AVM. Accompanying subarachnoid blood to a clot should raise suspicion of an underlying AVM. The remainder of patients present with seizures (25%), headaches and progressive neurologic decline (25%). The mortality rate of ruptured AVMs is 10-15% and are associated with a morbidity of 30%. In contrast, this is significantly lower than ruptured aneurysmal subarachnoid hemorrhage (aSAH), where the mortality and morbidity are 50% and 25% respectively. In addition, early rebleeding and arterial vasospasm are infrequent in AVM related hemorrhage despite the presence of subarachnoid blood, as compared with aneurysmal subarachnoid hemorrhage.

AVM Grading System
The risk of treating AVMs is in part determined by the its size, location, pattern of drainage, feeding arteries, amount of arterial cerebral steal from nearby normal brain, and the amount of overall blood flow through the AVM.

The most referenced and currently widely used grading system was reported by Spetzler and Martin in 1986 (4). They introduced a five tier grading system intended to predict the risk of surgical resection based on three variables: 1) AVM nidus size, 2) pattern of venous drainage, 3) location-eloquence of the adjacent brain. Size is scored by the greatest diameter of the AVM.

In general, the size of the nidus relates to the number and size of feeding arteries. Venous drainage pattern relates to the accessibility of surgery- deep being more difficult surgery with greater risk. Eloquence of adjacent brain includes motor and sensory cortex, language areas, visual areas, deeper structures such as hypothalamus, thalamus, brainstem, and cerebellar peduncles and nuclei. Higher grade AVMs, IV-V are reported to incur a 17-22% incidence of new neurologic deficits following surgery. Grades I-III are in general considered lower risk surgical candidates. In general surgical management for grade IV-V AVMs is reserved for patients with repeated intracerebral hemorrhage or experiencing progressive neurologic deterioration.
Surgical and Endovascular Treatment
The goal of AVM treatment is successful complete obliteration of the AVM to protect the patient from recurrent hemorrhage. Treatment must be individualized for each patient weighing the benefits and risks associated with surgical resection, endovascular therapy, radiotherapy, or a conservative watch and wait approach. Surgical treatments are preferred in patients in good neurologic condition with lower grade (I-III) Spetzler Martin AVMs. Higher grade patients (IV-V) or patients at high surgical risk or in poor neurologic condition may be candidates for endovascular treatment and or radiosurgical treatment. A detailed discussion of these is out of the scope of this review. See reference section for additional information.

Pre-operative and Post-operative Evaluation and Management
In general a complete four vessel cerebral angiogram and brain imaging (CT or MRI) is required prior to surgical or endovascular planning. CT is useful for identifying hemorrhage and the need for urgent clot evacuation and intracranial pressure management. MRI can assist in identifying the spatial relationships between the AVM and surrounding brain tissue.

Intra-operative and Post operative management
Barbiturates may be used early in surgery to reduce cerebral blood flow. This can be neuroprotective against brain ischemia during AVM resection. Intra-operative EEG can be helpful to confirm EEG-Burst suppression pattern and help gauge barbiturate dosing and management. Barbiturates may also minimize or blunt rapid post resection hemodynamic blood flow fluctuations. The disadvantages of barbiturate use is the long half-life which may mask post-operative neurologic examination changes from operative related brain hemorrhage or other complications. A post-operative head CT should be considered in patients who are slow to wake up or are noted to have new neurologic deficits.

Following AVM resection, either intensive care or a monitored setting is important for close neurologic and hemodynamic assessments (5). Blood pressure and fluid management in the early post-operative period is important to minimize additional cerebral blood flow fluctuations. Excessive hypertension or hypotension may have deleterious effects. Strict or predetermined blood pressure guidelines do not exist and management should be tailored to the individual patient.

A short course of steroids may be considered to treat operative related cerebral edema. Some patients are at increased risk for post operative seizures due to irritation of surrounding cerebral cortex and or from hemodynamic fluctuations. Infrequently, seizures may indicate mass effect on cortical brain areas or a new postoperative hemorrhage. Even despite prophylactic anticonvulsant therapy, patients can develop post operative seizures. Ongoing seizures can aggravate cerebral edema and alter local cerebral blood flow in the micro-circulation of the AVM resection bed. Higher than usual doses of anticonvulsants and or multiple agents may be required for seizure control.
REFERENCES


Cavernous Malformations (angiomas)

Cavernous malformations (CM), also referred to as cryptogenic or angiographically occult malformations, occur throughout the brain and spinal cord. They often appear as well defined round or irregularly shaped hemorrhagic masses. Unlike AVMs they do not have a readily identifiable feeding artery or draining vein. The consist of dysmorphic irregular vascular masses without intervening normal brain parenchyma. Cavernous malformations can enlarge over time due to a life cycle of recurrent hemorrhages leading to hemosiderin deposition, and gradual thrombosis.

Incidence, Pathological Features, Etiology, Location

Autopsy and brain imaging studies demonstrate a population prevalence of 0.47%-0.53%(1). A familial entity of multiple cavernous malformations is now well established (2). A familial form with an autosomal dominant inheritance pattern with incomplete expression has been reported (2). Moreover, Dubovsky et al (3) established a gene responsible for familial cavernous malformations mapping to chromosome 7q. Other occurrences of presumably acquired causes of cavernous malformation formation have been reported following cranial radiation and brain biopsy. Venous malformations, such as venous angiomas, are not frequently associated with cavernous malformations.

Cavernous malformations occur throughout the nervous system; cerebral hemispheres, both cortical and deep structures, brain stem, cerebellum, dura, intraventricular system, spinal cord, and rarely within spinal and cranial nerves. Rarely, CMs can be found in extracerebral locations such as within the dura and without attachment in the subarachnoid space.
The use of modern MRI imaging has greatly increased the identification of cavernous malformations throughout the nervous system. The cerebral hemispheres are the most common site of cavernous malformations. Although they distributed throughout the CNS, the cerebral hemispheres are the most common location, most located in the cortical or subcortical regions and often around the rolandic fissure. Temporal lobe is also a common location, often precipitating seizures as the presenting symptom. Other common locations include the cerebellum, pons, midbrain, and cerebellar peduncles.

**Natural History**
Unfortunately, there is limited data on the long term risk of hemorrhage from an incidental CM or a recurrent hemorrhage (from a CM). Annual hemorrhage from previously asymptomatic lesions is estimated at 0.25-1.1%\(^1\)\(^4\). Robinson et al\(^1\) suggest a higher rate of hemorrhage for re-bleeding of 25\%. The highest morbidity and mortality is associated with brainstem lesions. In general, the overall literature suggest that the denovo bleeding rate for asymptomatic patients with CMs is 0.6\% and 4.5\% in symptomatic patients\(^5\).

**Imaging**
For acute hemorrhage CT imaging is the study of choice, although less sensitive for subacute and chronic brain hemorrhage. Magnetic resonance imaging (MRI) is the study of choice for imaging CMs due its sensitivity for subacute and chronic hemorrhages. MRI T-2 weighted sequences typically reveal a well circumscribed hemorrhagic lesion with a central area of heterogeneous signal intensity. This appearance represents blood products from recurrent micro-bleeding, often asymptomatic, of the CM. The mixed signal is described as “popcorn kernel” from a black/white appearing hemorrhagic mass surrounded by a dark hemosiderin ring representing old hemorrhage. MRI imaging is often highly suggestive of CMs and best seen on hemosiderin sensitive
MRI sequences such as gradient echo. Other differential pathologies to consider are hemorrhagic metastases such as choriocarcinoma and melanoma and low grade glial tumors. Due to a lack of feeding arteries or clear draining vessels, conventional cerebral angiography may be normal or reveal a faint venous blush. When CMs are associated with venous angiomas angiography can be suggestive.

**Clinical Presentations and Features:**
Similar to AVMs, the most common clinical presentations of patients with CMs are seizures (especially in supratentorial hemispheric location), acute hemorrhage, and progressive neurologic deficits. Although CMs occurs without a gender bias, women tend to present more commonly with acute hemorrhage and men present more commonly with seizures. Brainstem and infratentorial locations can also present with acute hemorrhage, although not uncommonly with progressive brainstem neurologic deficits such as cranial nerve palsies, ataxia, and weakness/sensory loss.

Factors associated with a higher re-hemorrhage rate are: younger patients, woman, multiple re-hemorrhages, associated venous angioma, deep location, familial occurrence, infratentorial location, and after incomplete prior resection.

**Management:**
Conservative management, a “watch and wait approach”, is a reasonable strategy for patients with minimal or no neurologic symptoms. This is especially the case in patients with multiple cavernous malformations, such as in the familial category. The indications for surgical management are patients with recurrent hemorrhage, refractory seizures, and progressive neurologic deterioration. Stereotaxic radiosurgery has been used to treat patients with either surgically inaccessible or high risk surgical candidates. Unfortunately, the benefits of radiosurgery are difficult to assess given the lack of long term studies and an incomplete understanding of the natural history of these malformations.

**REFERENCES**

Venous Angiomas

Overview:
Venous Angiomas are the most common type of vascular malformations in the brain; arterial venous malformations, cavernous malformations, capillary telangiectases, venous angioma. Venous angioma are typically congenital anomalies rather than true vascular malformations such as Arteriovenous malformations or Cavernous malformations. They are found to occur throughout the brain and consist of a radial array of small medullary veins which converge into a larger venous vessel. The can drain either superficially to the cortical surface or deeper into a dural based sinus. In comparison to AVMs and CMs, they are rarely associated with repeated micro-bleeding an hemosiderin staining on MRI. Venous angioma can be solitary or multiple, and have been associated with cavernous malformations. In general venous angiomas provide venous drainage from normal brain tissue.

Natural History and Clinical Presentation:
Similar to other vascular malformation of the nervous system, venous angiomas can present with brain hemorrhage, seizures, and progressive neurologic deficits. However, more often they present as asymptomatic as incidental finding on a brain image. In general, venous angiomas have a fairly benign natural course. The estimated risk of hemorrhage is less estimated at less than 1% for asymptomatic cases.

Clinical Management:
In the majority of cases, the literature and experience suggests that venous angiomas should be managed conservatively. Currently there is no established role for radiosurgery or endovascular intervention. For venous angiomas associated with symptomatic cavernous malformations it is recommended to resect the cavernous malformation with attention to preserve the venous angioma. Other medical management strategies such as seizure management is crucial.