Intracranial Vascular Malformations

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CLASSIFICATION according to predominant vasculature: 
1) developmental venous anomaly (DVA), s. venous angioma (most common type) - anomalous VENOUS without any direct feeding artery.
2) capillary telangiectasia* (next most common type!) - vessels morphologically resembling CAPILLARIES but slightly larger.
3) arteriovenous malformation (AVM)*** (less common but most clinically important type!) - clusters of abnormal ARTERIES and VENES without intervening capillaries.
4) cavernous angioma (cavernoma)*
5) direct (s. arteriovenous) fistula** - rapid lesions; no nidus:
a) dural arteriovenous fistula (DAVF) 
b) vein of Galen aneurysmal malformation 
c) carotid cavernous fistula
6) venous varix - no clinical significance.
* may represent extremes of one nosologic entity - capillary malformation
** malformations with arteriovenous shunt

AVMs and cavernomas are commonly encountered surgically, while DAVs and capillary telangiectasias are nearly exclusively seen incidentally at autopsy.

- transitional (mixed) types of malformations also occur.
- true malformations result from embryonic vascular network.
- some lesions may in some cases incorporate adjacent vessels (“cistern”).
- most significant manifestation - bleeding - most likely to occur in patients < 30 yrs.

Rapid guide to diagnosis by NEUROMAGING: – not visible, (+) sometimes visible, + visible

<table>
<thead>
<tr>
<th>Type</th>
<th>CT</th>
<th>MRI</th>
<th>Angiography</th>
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<tr>
<td>AVM</td>
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<td>+</td>
<td>+</td>
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<tr>
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ARTERIOVENOUS MALFORMATIONS (AVM)

PATHOLOGY, PATHOPHYSIOLOGY

AVM - congenital tangle of arteries and veins connected without intervening capillary bed (i.e. by one or more fistulae).

- lesion is present from birth (vs. congenital aneurysms).
- vascular conglomerate (numerous thin-walled, tortuous channels) is called nidus.
  - nidus commonly forms "pyramidal conus" in white matter with base covering part of cerebral surface, and apex directed toward lateral ventricle - bleeding can be subcortical, intracerebral, or intraventricular.
- nidus has no capillary bed (feeding arteries drain directly* to draining veins) - failure of normal capillary bed development: arteries and veins are congenitally normal.* or via caverns

**Arteriovenous shunt is definitive characteristic**

**feeding arteries**: may run serpentine course through sulci before entering nidus;
- high flow subsequently leads to arteriovenous dilatation
- fibromuscular cushions - smooth muscle hyperplasia associated with fibroblasts and connective tissue elements
- arterial feeders:
  - entirely from ICA branches (purely pial malformations): MCA territory > ACA territory > PCA territory, occasionally (< 10%) recruit additional supply from meningial arteries.
  - entirely from ECA branches (purely dural malformations).
- arterial structure is damaged - duplication and fragmentation of internal elastic lamina, marked thickening or partial replacement of media by hyalinized connective tissue.
- draining veins often are dilated with thickened walls (due to high velocity blood flow through fistulae) - venous arterIALIZation: AVMs of ACA and PCA may drain directly into vein of Galen, causing it to dilate in aneurysmal fashion. *see below (Vein of Galen Malformation) **+

Arterial supply and venous drainage may be any combination of superficial and deep vessels

- involved vessels may enlarge with passage of time; some AVMs* may decrease over time (spontaneous thrombosis)? up to total resolution (rarely). *esp. those around anterior fossa and chiasm

Microscopy: entrapped (between vessels) brain tissue is glastic and nonfunctional, often with evidence of past hemorrhage (calcification and hemosiderin deposition).

Nidus has no interposed normal brain tissue and no capillary bed

Anatomical classification of AVMs:

- AVMs occur in all parts of neuraxis (largest AVMs are most frequent in posterior half of hemispheres).
  - typically lie superficially (within brain substance or cerebral sulci).
  - wedge-shaped (apex directed toward ventricle).
- 2.3-16.7% patients with AVM develop aneurysm (high-flow vasculopathy):
  - type I - located proximally on ipsilateral major artery (most common*);
  - type IA - located proximally on contralateral major artery;
  - type II - located distally on superficially feeding artery;
  - type III - located proximally or distally on deep-feeding artery;
  - type IV - located on artery unrelated to AVM.

Types I-III (85%) are flow related aneurysms; intranidal aneurysms are rare (5.5%) *oral vascular malformations

Wyburn-Mason syndrome is, Bonnet-Dechaume-Blanc syndrome: AVM involving cerebral cortex, optic nerve, retina * facial nerve.

Cerebral Proliferative Angiopathy: type of proliferative or diffuse AVM without focal nidus; often seen in pediatric patients.

HEMODYNAMICS

- flow shunted through AVM is extremely pressure-dependent (no autoregulation) and follows conditions described by Hagen-Poiseuille equation where flow (Q) is directly related to pressure difference (DP) and fourth power of radius (r) and is inversely related to tube length (L) and viscosity (µ):
INTRACRANIAL VASCULAR MALFORMATIONS

- **bulk flow rates** (vary according to size and anatomy) are 150-900 ml/min (= 490 ml/min).
- **AVM feeders** have low intravascular pressure, high flow velocity, low peripheral stream resistance, and very poor vasomotor reactivity (e.g., relatively nonreactive to PCO₂ changes).
- Tissues adjacent to **AVM** may be persistently mildly hypoxic (malformation may steal blood from adjacent healthy tissue).
- As **AVM** is resected, pressure within feeding arteries rises by ≈ 60% to normal values, and normal CO₂ reactivity is immediately established in adjacent cerebral vessels.

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Mass of irregular, tortuous vessels over left posterior parietal region:

Source of picture: "WebPath - The Internet Pathology Laboratory for Medical Education" (by Edward C. Klatt, MD)>

Tangle of abnormal vessels on brain surface:

Source of picture: "WebPath - The Internet Pathology Laboratory for Medical Education" (by Edward C. Klatt, MD)>

Tangled complex of blood vessels with intervening neural parenchyma:

Source of picture: "WebPath - The Internet Pathology Laboratory for Medical Education" (by Edward C. Klatt, MD)>

Intraventricular and intracerebral hemorrhage due to ruptured AVM:

Source of picture: "WebPath - The Internet Pathology Laboratory for Medical Education" (by Edward C. Klatt, MD)>

Microscopic appearance of AVM - dilated, tortuous, worm-like vascular channels:

Source of picture: "WebPath - The Internet Pathology Laboratory for Medical Education" (by Edward C. Klatt, MD)>

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\[
\frac{\Delta P}{r^4} = \frac{Q}{8 \cdot L \cdot \eta}
\]
**AVMs produce**

Most manifest of intracranial aneurysms

1. Osler-Weber-Rendu syndrome (hereditary hemorrhagic telangiectasia) — although AVMs tend to be multiple but with more benign clinical course
2. Sturge-Weber disease
3. neurofibromatosis
4. von Hippel-Lindau syndrome.
5. cerebral/facial arteriovenous metaneprogenic syndromes (CAMs)

- CAMS I - proenephorrheic AVMs affecting hypothalamus/hypophysis in association with facial AVM of nose
- CAMS II - AVMs affecting lateral prosencephalon (occipital lobe, thalamus) in association with facial AVMs of maxilla.
- CAMS III - AVMs of rhomboencephalon (cerebellum, pons) in association with facial AVM of mandible.

- cerebral AVMs can be associated with pulmonary AVMs (but not with other organ AVMs)

**EPIDEMIOLOGY, ETIOLOGY**

Prevalence is not known, detection rate in general population = 0.04-0.52%, i.e. 1/5-1/7 incidence of intracranial aneurysms.

- **both sexes** are affected equally.
- 3-20% of spondiac AVMs are diagnosed in children.
- no genetic, demographic, or environmental risk factors have been identified.
- familial cases are rare.
- in rare cases (2%), cerebral AVMs are associated with other inherited disorders:
  1) Osler-Weber-Rendu syndrome (hereditary hemorrhagic telangiectasia) — although AVMs tend to be multiple but with more benign clinical course
  2) Sturge-Weber disease
  3) neurofibromatosis
  4) von Hippel-Lindau syndrome.
  5) cerebral/facial arteriovenous metaneprogenic syndromes (CAMs)

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- CAMS III - AVMs of rhomboencephalon (cerebellum, pons) in association with facial AVM of mandible.

**CLINICAL FEATURES**

Only 12% AVMs become symptomatic!

Most manifest before 40 yrs! (vs. cerebral aneurysms - only 1/4 patients present by age of 40)

- **bruits** (> 50%) may be audible either to patient or to examiner.
- scalp or face veins may be enlarged.
- huge AVM (esp. if involve vein of Galen) may cause high-output congestive heart failure in newborns.
- **hydrocephalus** may result if vein of Galen enlarges as channel for AVM drainage.

**AVMs produce neurological dysfunction through 4 main mechanisms:**

A. **Hemorrhage** (presenting symptom in 38-70% patients) into:
   a) brain parenchyma - most common!
   b) intraventricular
c) subarachnoid space - less severe than with saccular aneurysms; blood tends to localize over cerebral convexities

- 2% of all hemorrhagic strokes.

N.B. AVMs are cause of hemorrhage in young adults! (peak in 2-4 decades)

N.B. AVMs are the most common cause of spontaneous brain hemorrhage in children (excluding neonatal period)

- **overall bleeding risk 2-4% per year**.
- **prognosis & recovery tendency to be better than in non-AMV-related (aneurysmal, hypertensive) hemorrhages**
  - vasospasm occurs only rarely (because less blood accumulates around large arteries at base of brain).
  - death occurs in 6-29% AVM hemorrhages (13-20% in rebleedings).

- **bleeding source**
  a) draining vein
  b) flow-related aneurysm

**Mortality with each bleed is > 15%**

B. **Seizures** unrelated to hemorrhage (presenting symptom in 15-46% patients).

- focal, may become secondarily generalized.
- **risk factors for seizures**:
  1) young age
  2) large AVM size
  3) lobes location (esp. temporal lobe) with feeders mainly from MCA.
- **secondary epileptogenesis and kindling** can persist after AVM removal (H: maintain anticonvulsants after treatment of AVM is accomplished).

C. **Progressive neurological deficit** (6-21%); - slowly progressive (over months + several years).

- **reflects AVM location**.
- **mechanisms**
  a) blood siphoning away from adjacent brain tissue (“steal phenomenon”).
  b) mass effect of enlarging AVM.
  c) venous hypertension in draining veins.
- detailed neuropsychological testing may disclose subtle right or left hemisphere dysfunction.
- history of subtle learning disorder is elicited in 66% adults with AVMs.
INTRACRANIAL VASCULAR MALFORMATIONS

D. Headache unrelated to hemorrhage (4-50% patients) - may be as typical migraine* or may be more generalized.
*typical migraine alternates from one side of head to other, whereas AVM headaches classically remain on same side.

E. If sufficient AV shunting is present, it may manifest as congestive cardiac failure in neonates and infants.

DIAGNOSIS

NEURAL RADIOGRAPHICS

• AVM calcifications, increased vascular markings in overlying bone (calvarial vascular grooves and foramina).

CT

• can identify only large AVMs - serpiginous areas of high density.
• contrast-enhanced CT - striking enhancement* (classic pattern - irregular hyperdense central area from which extend multiple, well-defined serpentine structures of various sizes - dilated feeding arteries and draining veins).
*due to increased blood pool within lesion + BBB impairment in adjacent neural parenchyma
• AVMs may be surrounded by hypodense areas of ischemic damage.
• CT may show calcification.

A. Noncontrast CT - areas of calcification and increased density in left temporal lobe; slight mass effect; dilated left temporal horn.
B. Contrast CT at same level - enhancement of large feeding arteries, nidus, and draining veins.

MRI

- essential for initial diagnosis* (also preferred screening procedure)
* N.B. gadolinium does not facilitate detectability (vs. CT contrast).
• irregular serpiginous or globoid masses with mixed signal anywhere within hemispheres or brain stem.
• large arteries and draining veins are particularly characteristic feature - shown as signal void rather than flow-related enhancement.
• Round, low-signal spots within / around mass are "flow voids" of feeding arteries, intranidal aneurysms, draining veins.
• if hemorrhage has occurred, mass of blood may obscure other diagnostic features (H. angiogram or follow-up MRI).
• low T1 signal of extracerebral hematoderin may be seen around or within AVM mass, indicating prior hemorrhage.
• there may be regional brain atrophy.

T2-MRI - parietal AVM; varices (short arrows), dilated arteries (long arrow) and draining veins (notched arrow):
**Intracranial Vascular Malformations**

**Vas30 (7)**

**T2-MRI** - extensive bilateral AVMs, multiple enlarged superficial drainage veins.

**T2-MRI** - AVM with hemorrhage in territory of left PCA.

Subcallosal intraventricular AVM fed by anterior and posterior pericallosal and choroidal vessels.

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**MRA**

- Can identify AVMs > 1 cm.
- Inadequate to delineate morphology of feeding arteries and draining veins; small aneurysms can be missed easily.

**3D TOF MRA** - hugely dilated left MCA feeders (long arrow), smaller (short arrow), varices (arrowhead) and superficial drainage vein (open arrow).

Surface-oded reconstruction of 3D TOF MRA - posterior fossa AVM supplied by SCA and AICA; flow-related aneurysm (red arrow) has formed at AICA origin.

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INTRACRANIAL VASCULAR MALFORMATIONS

AVM in left medial temporal lobe (coronal T2-MRI and MRA):

MR DSA (series of three frames in lateral projection acquired at 1-sec intervals during IV infusion of gadolinium):
1) feeding MCA branches (long arrows), nidus (short arrow), varices (arrowhead) and large superficial draining vein (open arrow) all apparent on first frame, indicating speed of shunting.
2) opacification of transverse sinus (open arrow).
3) opacification of superior sagittal sinus (white arrow); note small venous pouch on main draining vein (black arrow).

Angiography

- Required for definitive diagnosis & treatment planning (AVM morphology determines treatment algorithm):
  - Dilated tortuous feeding arteries; central tangle of vessels; rapid* arterial-to-venous shunting (early opacification of enlarged, tortuous draining veins).
  - Rapid sequence (high frame rate) filming is essential otherwise (feeding vessels can be obscured by overlying veins in rapidly shunting lesions!)

- Four-vessel angiography is indicated:
  - up to 10% AVMs are associated with saccular aneurysms.
  - Extracranial or contralateral arteries occasionally supply intracranial AVMs.
  - In case of hemorrhage, hematoma may compress AVM so completely that angiography cannot detect it (when AVM is suspected, angiography is best postponed until hematoma has resolved - after 6-8 weeks).
  - AVM nidus within nidus often show slow washout.

Superselective angiography into AVM feeding arteries:

1) Obtain pressure measurements (higher feeding pressures increase hemorrhage risk).
2) Inject SODIUM AMYTAL (anesthetic agent) to produce temporary anesthesia of area perfused by artery. "Superselective Wada testing" - language, memory, visual-spatial, sensory, motor function can be tested during 5 minutes to determine whether "eloquent" function originates in AVM region (risk for neurological deficits during embolization or surgery):
   a) Arteries directly feeding AVM
   b) "En passage" arteries - feed AVM but contain past AVM to feed normal brain tissue.
Intracranial Vascular Malformations

Paratrigonal AVM:

Preoperative angiography:

Postoperative angiography:

Large deep right temporal AVM encompasses most of medial temporal lobe (preoperative carotid angiography):

Angiogram (AP view) - AVM (3 cm in diameter) in deep MCA territory with deep draining vein (arrow): Deep cerebellar AVM fed by branches of SCA, AICA, and PICA (preoperative and postoperative angiography):

DSA - arterial (A) and venous (B) phase - AVM fed by ACA and MCA branches; venous drainage predominantly superficial into superior sagittal and transverse sinuses.
Intracranial Vascular Malformations

AVM in medial surface of left temporal lobe (left vertebral arteriogram):
A. Towne projection – enlarged feeder, nidus, and dilated draining vein are all seen.
B. Lateral projection – better visualization of relationship of draining veins to nidus.

Right parasplenic AVM (preoperative and postoperative angiograms):

Left posterior sylvian AVM:
Preoperative angiography:
INTRACRANIAL VASCULAR MALFORMATIONS

Postoperative angiography:

Large anterior callosal AVM extending into basal ganglia (preoperative and postoperative angiography):

Subcallosal intraventricular AVM fed by anterior and posterior pericallosal and choroidal vessels (preoperative vertebral and carotid angiography; postoperative angiography):

Associated aneurysms:

FUNCTIONAL MRI

- to map brain function (“eloquent” brain regions in and around AVM) during treatment planning.
**TREATMENT**

"It would be nothing less than foolhardy to attack one of the deep-seated racemose lesions…. The surgical history of most of the reported cases shows not only the futility of an operative attack upon one of these aneurysms but the extreme risk of serious cortical damage which it entails…. How many less successful attempts, made by surgeons less familiar with intracranial procedures, have gone unrecorded may be left to the imagination." — Harvey Cushing

Treatment planning depends on risk of hemorrhage!, seizures or headache may be treated conservatively.

<table>
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<th>Risk factors for hemorrhage:</th>
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<tr>
<td>1) male gender</td>
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<tr>
<td>2) small (!) AVM size (&gt; 2.5 cm)</td>
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<tr>
<td>3) deep location in basal ganglia or posterior fossa</td>
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<td>4) deep venous drainage</td>
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<td>5) single or only few draining veins (esp. with kinking or stenosis or varix)</td>
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<td>6) high pressure in feeding arteries (as measured during angiography)</td>
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<td>7) aneurysms (10% patients);</td>
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<td>- intranidal aneurysms have higher risk of rupture than those outside bounds of AVM (flow-related feeding artery aneurysms); in general, AVM-related aneurysms bleed more often than standard saccular aneurysms</td>
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<td>8) prior hemorrhage (rebleeding risk during 1st year 7-33%, then 2.5% annually)</td>
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<td>9) pregnancy see below</td>
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Bleeding risk is not influenced by age!

**ARUBA (A Randomized Trial of Unruptured Brain Arteriovenous Malformations) trial** - randomly assigned AVM patients to either a conservative medical management group or an intervention group (any sort - surgery, embolization, or Gamma Knife); trial was stopped by the National Institute of Neurological Disorders and Stroke because after a mean follow-up time of 33 months, the event rate (death or symptomatic stroke) was > 3 times higher among patients in intervention group than among those in conservative management group

**Radiosurgery Practice Guideline for Intracranial Arteriovenous Malformations** (Guideline Report #2, 03), original guideline 2009)

**SUPPORTIVE treatment**

1. Standard anticonvulsants for seizure control; seizures are well controlled with PHENYTOIN, CARBAMAZEPINE, VALPROIC ACID, LAMOTRIGINE.

2. Analgesics for headaches - either nonspecific or migraine-specific drugs.

**PATHOPHYSIOLOGIC treatment**
AVM treatment
- indicated for patients with high risk of hemorrhage
  a) all patients with AVM-related hemorrhage
  b) younger patient with unruptured AVM and ≥ 1 high-risk features for AVM rupture.

If annual rebleeding rate 1-2% is maintained for life, young individual faces 50-60% chance of incapacitating / fatal rebleeding during normal lifespan.

Attempts should be made to completely eradicate lesion:
1. Endovascular embolization
2. Microsurgical resection
3. Radiosurgery.

SURGICAL TREATMENT RISK ESTIMATION
A. SPETZLER–MARTIN grading system – sum points from 3 factors:
I. AVM size (largest diameter of indus on angiography):
   1 point - AVMs < 3 cm
   2 points - AVMs 3-6 cm
   3 points - AVMs > 6 cm.
II. Eloquence of adjacent brain:
   0 point – if AVM is located in noneloquent area.
   1 point – if AVM is located in functionally critical area (e.g. language, motor, sensory, or visual cortex, thalamus, hypothalamus, internal capsule, brain stem, cerebellar peduncles, deep cerebellar nuclei).
III. Pattern of venous drainage:
   0 point – if AVM has superficial venous drainage (all drainage is via cortical veins).
   1 point – if AVM has component of deep venous drainage.

Correlation of surgical results with Spetzler–Martin AVM grading:

American Heart Association multidisciplinary MANAGEMENT GUIDELINES
Spetzler-Martin grade I-II → surgical extirpation.
if AVM < 3 cm and surgery has increased risk → radiosurgery.
Spetzler-Martin grade III → embolization → surgical extirpation.
if high surgical risk, embolization → radiosurgery.
Spetzler-Martin grade IV-V (not amenable to surgical treatment alone - high procedural risk) → combination of embolization, radiosurgery and/or surgery.

Spetzler-Ponce classes:
Class A - Spetzler-Martin grade I and II lesions
Class B - Spetzler-Martin grade III lesions
B. Supplemental Spetzler-Martin Grading System (SM-Supp, s. Lawton-Young grading system) - ABCs of AVMs: patient Age, Bleeding or hemorrhagic presentation, and AVM Compactness

**Endovascular Embolization**

**Thrombosing agents:**
1. Quick-acting acrylate glue (N-butyl cyanoacrylate [NBCA]) – glue dilution degree governs speed of polymerization (to suit rapidity of AV shunting* and distance to be crossed by embolic agent).
2. Onyx® liquid embolic system – only approved for patients who will undergo surgical removal of AVM
   - catheter entrapment (catheter stuck in implanted Onyx material) has been reported.
3. Cordan® Trufill®-NBCA – indicated for embolization of cerebral AVM when presurgical devascularization is desired.
4. Finely graded particles (e.g. polyvinyl alcohol ± microfibrillar collagen)
5. Thrombus-inducing coils
6. Detachable balloons.

**Principal uses:**
- try to occlude nids, not just arteries (in time, collateral supply can develop to left intact nids).
- do not occlude veins!
- do not embolize > 1/3 during one session.
- do not occlude > 1/3 during one session.
- always try to embolize deep parts first (may be fed by PCA branches vs. the rest of AVM fed by MCA).

**Approach:**

**Transarterial – classical:**
- plug-and-push technique: create plug (e.g. balloon) around the catheter (to prevent embolization material reflux) and push glue; may cause catheter gluing in situ.
- direct access embolization – for distal vessels, for tortuous vessels: microcatheter inserted directly into feeding artery intra-operatively.

**Transvenous – make sure to embolize entire nidus or else will cause bleeding.**

**TABLE 1. Comparison of the Spetzler-Martin and Supplementary Grading Systems**

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<th>Spetzler-Martin Grading</th>
<th>Points</th>
<th>Supplementary Grading</th>
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<td>Size, cm</td>
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</tr>
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</table>

Sum of two scores (SM-Supp) ≤ 6 – acceptably low surgical risks (0%-24%).
Sum of two scores (SM-Supp) > 6 – significant increase in surgical risk (39%-63%).

**Vermian AVM embolization (left posterior view):**

**Vermian AVM embolization (left posterior view):**

- Pre-embolization - vermian AVM supplied by vermian branches of SCA.
- Microcatheter NBCA injection into AVM nidus.
- Postembolization of both major pedicles - almost complete nidus obliteration.

**Surgical Resection**
INTRAOPERATIVE

- most effective with easily accessible smaller lesions (Stryker-Martin grades 1-3, 4).
- Electrocortical stimulation and surface EEG recording while local anesthetic is useful in delineating specific cortical function and seizure focus localization.

INTRAOPERATIVE ANGIOGRAPHY is gold standard (may be supplemented by BEG)*. ANGIOGRAPHY helps to delineate arterial feeders and draining veins. It can be safely isolated and ligated.

- Single dose (40 mg) of indocyanine green preserves "en passage" arteries!
- Two strategies of approach:
  a) Peripheral isolation of feeding arteries with marginal resection of nidus (trabecular en bloc method).
  b) Central resectional isolation of major draining vein as guide to nidus (merit of collapsing lesion from within).
  - Cortical veins drain arterialized blood radially from lesion; major draining vein is often in cortex and is initially identified and localized (match with angiogram) ➔ follow into nidus.
  - Shunting vessels and arterioles are cauterized and cut along central vein (venous radicals are then "shocked" by multiple bipolar coagulations under saline irrigation along with topical hemostatic-cottonoid pledget tamponade).

N.B. premature occlusion of venous drainage from incompletely resected nidus results in severe hemorrhage.

- N.B. surgical occlusion of arterial feeders alone does not have lasting value - collateral feeding arteries quickly enlarge and malformation persists!
- Even small residual pieces of nidus can lead to catastrophic hemorrhage!
- N.B. awake craniotomy is not feasible - even if you encounter deficits, you have to proceed with resection (cannot do only partial resection)

- Gliotic interface at periphery of nidus allows for plane of dissection to be developed - surgical extirpation with minimal deficits.
- Larger lesions are separated into compartments and isolated and collapsed as separate units.
- Aneurysms are clipped surgically as well.
- When intra-nidal aneurysm is found in AVM, bleeding, it must be targeted for urgent therapy; i.e. presume that aneurysm (not AVM) is source bleeding - treat urgently to prevent rebleeding!
- There is increase in resistance of vessels that have fed AVM after AVM is removed - if aneurysms are not treated before or at time AVM is treated, they may hemorrhage in postoperative period.

- Measures to reduce blood loss:
  1) controlled hypotension during surgery (maintain MAP 40-60 mmHg)
  2) embolization of feeding vessels prior to surgery.

POSTOPERATIVE

- uncomplcates with correlates on Spetzler-Martin scale.
- M1 resection ➔ better tissue perfusion ➔ progressive neurological improvement.
- Surgical morbidity 8.6%; mortality 3.3%.
- Complications:
  1) "Normal perfusion pressure breakthrough" (NPPB) (theory described by Stryker in 1978) - profuse edema and generalized hemorrhage from resected AVM bed (increased flow to previously underperfused vessels with lost autoregulation - weeks to months are required for brain to adapt to flow changes).
- H: preoperative embolization, staged resections for large high-flow AVMs, postoperative hypotension.
- 2) postoperative bleeding / venous infarction due to occlusion of venous drainage (Yasargil theory).
- 3) damage to adjacent neural tissue.
- routine POSTSURGICAL ANGIOGRAPHY (AVM reappearance, years after negative postresection angiogram, have been reported).
- routine postoperative seizure prophylaxis (min. 3 months).

STEREOTACTIC RADIOSURGERY

- results using any of these techniques appear to be relatively similar:
  a) proton beam
  b) LINAC
  c) gamma knife
- noninvavve and can access all anatomic locations of brain (e.g. surgically inaccessible).
- ideal for small AVMs (O. Sheehan: SRS can be used for any AVM, ideally small deep-seated: > 3 cm AVMs must be treated in stages).
- mechanism of action: radiotherapy induces subependymal collagen deposition ➔ narrowed lumen of vessels ➔ thrombosis over 1-3 years (risk of hemorrhage remains during this "latency period"!!)

- Contraindications: small volume (< 3 cm³), lobar location AVMs that can be easily removed or resected without permanent neurological deficits.

Methodology:
- stereoelectro volumetric axial plane imaging (MRI or CT) supplemented by conventional or digital subtraction angiography* is usually necessary for complete conformal dose planning. Isotopes are not absolutely necessary but serves as a reality check.
- single session tissue-destructive dose (16-25 Gy at the margin) is given.
- single dose (40 mg) of methyleneblueindole at the conclusion of the SRS procedure.
- If aneurysm is identified in nidus selected for SRS, additional endovascular or surgical strategies should be considered (to reduce risk of bleeding during latency interval).
- If AVM is large (total treatment volume > 5 cm³) – use volume staging with margin dose at a minimum of 16 Gy; AVM is divided into approximately equal volumes on MRI (medial or lateral, superior or inferior components) using certain identified landmarks such as major vessel blood supply, the ventricles or other anatomic structures such as the internal capsule, each stage is defined at the first procedure, and then re惦ated at subsequent stages using internal anatomic landmarks; second stage SRS is performed 3-6 months after the first procedure.
- SRS after surgery for hemorrhage: safe interval between surgery and SRS is not known, but is reasonable to perform SRS once the patient has achieved a stable neurological recovery or plateau (generally within 2-3 months after the intracranial hemorrhage or prior surgery).
INTRACRA NIAL VASCULAR MALFORMATIONS

Complications (clinical worsening attributable to SRS is seen in 3.8%; estimated risk of permanent new neurological deficits related to radiation is 3-5%):

1) white matter edema. N.B. if you see edema around AVM, it is likely from radiation damage (ask patient about previous treatments!)
2) radiation-induced necrosis.
3) seizure frequency may increase in first weeks after radiosurgery. Use perioperative AED in lobar AVMs!
4) late effect - accelerated atherosclerosis in surrounding blood vessels, cyst formation (4.7%).

Results: obliteration rate after single SRS is 50-95%; after multiple SRS - 75%.
process is cumulative, with earliest obliterations noted within 2-3 months, 50% within 1 year, 80% within 2 years and 90% within 3 years.

Follow up after SRS: MRI at 6 month intervals for the first 3 years (gradual obliteration; MRI has 96% accuracy for obliteration detection), at 3-year mark:

a) complete closure of the AVM nidus → confirmatory angiogram (if MRI before 3 years suggests obliteration, angiography is generally delayed until 3 full years have elapsed).
b) AVM nidus not obliterated (on MRI or angiogram at 3 years) → repeat SRS (or other strategy).
   - post-radiosurgery MRI changes (new areas of high T2 signal in brain surrounding the irradiated AVM nidus) develop in approximately 30% of patients 1-24 months after SRS.
   - during latent period, risk of bleeding may be increased or decreased (published results vary); risk of hemorrhage is further reduced, although not eliminated, after obliteration (estimated lifetime risk of a bleed is < 1%).

Positive predictors of obliteration:

1) higher marginal dose (odds ratio = 1.16).
2) compact nidus (odds ratio = 3.16).
Predictors of SRS failure:

1. prior bleed
2. lower marginal dose
3. sex (slightly worse in women)
4. prior embolization

Combination of embolization and SRS does not offer any advantages over SRS alone and may have significant disadvantages
- reduction in flow within the AVM does not improve SRS results; embolization can only be an effective adjunct to SRS if it results in permanent reduction of the nidus volume (recanalization of embolized portions of the AVM that may have been outside the SRS target results in persistent AV shunting and treatment failure).
- combination of embolization and radiosurgery does not provide any additional protection against AVM hemorrhage during the latency period.
- if embolization is used, the optimal time for SRS is not known, but generally waiting for a period of several weeks is beneficial to reduce the likelihood of vascular ischemic complications or residual cerebral edema sometimes associated with embolization followed by early radiosurgery.
- persistent out-of-field nidus (marginal failure) was identified in 18% of previously embolized vs. 5% of nonembolized patients (p = 0.0006).
- avoid AVM embolization if planning radiosurgery


Prior embolization reduces rate of total obliteration after SRS (but risks of hemorrhage during latency period are not affected by prior embolization).
- indication (for AV embolization before radiosurgery - palliative (e.g. patient has neurodeficits and you want decrease AVM flow instead of waiting 3 years for SRS effect)
- embolization harms:
  1. Embolization material gives radio artefacts - difficult to target SRS.
  2. Embolization material "breaks" one big nidus into several smaller - SRS difficult to target.
- Dr. Sheehan: embolization just delays obliteration (Onyx does not have this adverse effect).
5) larger AVM volume
6) eloquent location

The Virginia Radiosurgery AVM Scale

1) AVM volume of 2-4 cm³ - 1 point; AVM volume > 4 cm³ - 2 points
2) eloquent AVM location - 1 point
3) history of hemorrhage - 1 point

0-1 points - 80% of patients had a favorable outcome
2 points - 70%
3-4 points - 45%

Repeat SRS for AVM
- if residual nidus volume is < 3 mL then no angiogram is needed for the GK procedure as MRI spatial resolution should suffice; if > 3 mL then an angiogram is performed with the angiographic fiducial box the day the stereotactic frame is placed and the targeting MRI is obtained - in this case the spatial resolution of the nidus in the angiogram is superior to that of the MRI (i.e. the nidus will be overdrawn by using MRI only).
- no firm guideline how much time between GK procedures (more time between the 2 procedures the better - less probability of running into toxicity issues).

OUTCOMES
AVM recurrences (4% in one series) in the operated adult population may have a multifactorial origin: risk factors:

1) deep venous drainage
2) diffuse nidus
3) prooperative embolization - may also be a contributing factor with the potential for recurrence of unselected but embolized portions of the AVM - follow-up angiography at 1 to 3 years appears to be warranted.

- perinidal capillary network may be cause of recurrence of surgically resected AVMs.
- dilated capillaries (10 to 25 times larger than normal capillaries) form a ring (1-7 mm) around nidus - connected to nidus / feeding arteries / draining veins, and to surrounding normal brain vessels. 

### SPECIAL SITUATIONS

#### PREGNANCY

- conflicting data:
  a) pregnancy does not increase risk of hemorrhage if AVM has not previously hemorrhaged; risk doubles if AVM has previously hemorrhaged.
  b) pregnancy increases risk 5-fold; 8.1% per pregnancy (= 10.8% per year) vs. 1.1% baseline annual risk – data from Brigham and Women’s Hospital and Harvard Medical School**.


- intracranial hemorrhage during pregnancy is due to AVM in 20-48% cases.
- once hemorrhage occurs during pregnancy there is 25% chance of recurrent hemorrhage.
- surgical management should be based on neurosurgical principles; majority of AVM hemorrhages can be managed nonoperatively until delivery.
- AVM does not preclude normal vaginal delivery (i.e. method of delivery should be determined by obstetrical principles).

#### STRIATUM - THALAMUS

- 8-18% of all AVMs.
- usually drain via thalamostriate vein and basal vein of Rosenthal into galenic system.
- approaches for STRIATUM: transfornamental, transcapsular-transventricular, transylvian-transversal.
- approaches for THALAMUS – interhemispheric: transcapsular or trans-splenial.
- after microsurgical isolation of feeding perforating vessels, lateral ventricle is opened and lesion excised (incl. choroid plexus - carries deep arterial supply to nidus).
- surgical morbidity is formidable (hemiplegia, aphasia, hemianopia, memory impairment, and hydrocephalus), but these deficits also are part of natural course.
- profound neurological deficits can resolve (owing to rich collateral supply that naturally accompanies AVMs).

#### POSTERIOR FOSSA

- 5-7% of all AVMs.
- tend to be eclabbed frequently.
- large arterial feeders also supply cerebellum and brainstem - branches can be surgically occluded only if they clearly enter nidus.
- best handled surgically, although posterior fossa AVMs are listed in nonoperated group in many series (combined operative morbidity and mortality is ≥ 20%).
- CEREBELLUM AVMs should be surgically removed (natural course is treacherous).
  - approach is tailored to specific location (e.g. transfornamental route for lesions around tectum and superior cerebellum).

#### BRAINSTEM AVM

- microsurgical resection - combined subtemporal-suboccipital-retrolabyrinthine-transcranial approach.
- radiosurgery (rate of obliteration at 1 to 3 years appears to be 43.3%, 62.3%, 73%, and 81.8% respectively)

### HEMORRHAGE

- risk of immediate rebleeding is relatively low - treatment of AVM is delayed (4 to 6 weeks) to allow time for 1) hemorrhage to resolve and edema to subside – better brain tolerance for retraction
  2) AVM to stabilize its architecture for treatment planning.
- life-threatening hematoma requires urgent evacuation surgery - decompress hematoma while avoiding AVM.
- attempting to more completely remove hematoma can result in bleeding.
- in case of significant brain swelling, adding a dorsal patch and leaving large bone flap out is extremely helpful.

### CAVERNOUS ANGIOMA, s. Cavernoma, Cavernous Malformation

- sporadic; at least 6% are familial (> 50% such patients have multiple lesions); responsible genes: a) RIT1 (CCM1) b) MGC4607(CCM2) c) PDCD10 (CCM3)
- potential existence of CCM4 increased incidence and multiplicity among Mexican-American families.
- occur in 0.1-0.8% of general population.
- 10-15% of all CNS vascular malformations (second most common type after developmental venous anomalies).

### ETIOPATHOPHYSIOLOGY

- hamartomatic enlarged sinusoidal capillaries, s. cavern (single layer of endothelium, thin collagenous wall, no smooth muscle, no elastic fibers).
- well-circumscribed, “mulberry” appearance, expand slowly.
- capillaries are immediately adjacent to each other.

- not associated with enlarged feeding arteries or draining veins.
- not known in low or even standard.
- intra-lesional thrombosis, calcification and recanalization are typical.
- range from soft to hard.

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*No invoving neural tissue!!!

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INTRACRANIAL VASCULAR MALFORMATIONS

- adjacent neural tissue may be affected – gliotic (form capsule), small subclinical hemorrhages (perilesional hemosiderin may incite epileptogenic focus).
- 30% cases have associated DVA/s.
- usually located within brain parenchyma (can occur anywhere in CNS) but rarely may be located within dura.
- natural history - dynamic lesions - can grow and regress.

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- 30% cases have associated DVA/s.
- usually located within brain parenchyma (can occur anywhere in CNS) but rarely may be located within dura.
- natural history - dynamic lesions - can grow and regress.

Back-to-back hyalinized vascular spaces with evidence of prior hemorrhage:

Well-circumscribed mulberry appearance:

CLINICAL FEATURES

- 40% of lesions are ASYMPTOMATIC.
- 10-25% lesions are SYMPTOMATIC:
  1. Seizures (23-70%, esp. supratentorial) - result from surrounding hemosiderin deposits, cerebral gliosis, and cortical irritation (cavernoma itself is not epileptogenic as contains no brain tissue); risk of second seizure within 5 years is > 90%; 40-50% develop medically refractory epilepsy.
  2. Focal neurologic deficits (20-45%) - lesions in or close to cerebral cortex.
  3. Hemorrhage (9-56%); 0.1-1.0% annual rate* or 15.8% / 5-yr symptomatic hemorrhage risk (rate increases dramatically if lesion enlargement within one year is documented; other risk factors - previous bleed, infratentorial location); may be severe enough to result in mortality or long-term disability.
  4. Mass effect: headache (6-52%)

- supratentorial lesions are frequently associated with seizures while infratentorial lesions are likely to be associated with focal neurological deficits.
- headaches are prominent wherever angiomomas are located.
- symptomatic lesions are likely to remain symptomatic or progress.

DIAGNOSIS

ANGIOGRAPHY
- demonstrate no vascular abnormalities!!!

Cavernomas are “angiographically occult” or “cryptic” vascular malformations!

MRI
- Most-sensitive neuroimaging - appearance is sufficiently characteristic:
  - Well-defined reticulated - multilobular core of mixed-signal intensity (“POPCORN”), typically surrounded by dark hemosiderin ring on T2.
  - T2*-susceptibility weighted imaging which shows increased sensitivity.

- blood products of various ages in center.
- mass effect only seen if bleeding has occurred.
- local edema may be present.

A

Well-defined reticulated - multilobular core of mixed-signal intensity (“POPCORN”), typically surrounded by dark hemosiderin ring on T2.

B

T2*-susceptibility weighted imaging which shows increased sensitivity.

Midbrain cavernoma:
Medial left frontal cavernoma with prominent rim of hemosiderin-laden macrophages and no associated edema.

Source of picture: Viktoras Palys, MD.
Intracranial Vascular Malformations

Cavernoma of pons (T2-MRI):

Tectum cavernoma (T1-MRI) - recent and old hemorrhage:

MRI (FLAIR): cavernous hemangoma in right temporal lobe.

T2-MRI: high signal due to MetHb, low signal rim of hemosiderin indicates old hemorrhage, note blood-fluid level in smaller lesion (arrows).

Unenhanced CT of same patient - lesions predominantly of high density with tiny foci of calcification (arrows).

Lesion in right middle cerebellar peduncle (MRI-T2) - characteristic dark signal (hemosiderin) around lesion:

Hemorrhage into pontine cavernoma, hemorrhagic products are surrounded by partial hemosiderin rim, and associated developmental venous anomaly is also noted (arrows).
INTRACRANIAL VASCULAR MALFORMATIONS

Vas30 (22)

45-year-old woman with progressive neurological deterioration - blood products of various stages within midbrain cavernoma, including deoxyhemoglobin or hemosiderin (gradient echo, A), and methemoglobin (increased signal on the coronal and sagittal T1-weighted images, B and C). D, A time-of-flight MRA shows T1 shortening within cavernoma because of hematoma:

CT:
- homogeneous* focal hyperdensity ± calcifications (∼30%)
- no edema or mass effect
- IV contrast may show only faint patchy enhancement.

Lesion in right middle cerebellar peduncle:
Unenhanced CT - increased density; Enhanced CT - minimal enhancement.

Left parietal hyperdensity:

TREATMENT

Indications:
SURGERY

- icctal ECoG-guided resection including surrounding cortex* is considered the gold standard (70% seizure freedom)
- "additional resection of perilesional hemosiderin deposits and gliosis provides higher rate of seizure control (vs. pure lesionectomy) but it is problematic in eloquent areas (patient outcome consists of both – seizure freedom and no postop deficits)
- N.B. cavernoma itself is not epileptogenic (it contains no nervous tissue) but adjacent hemosiderin-lade cortex is!

CUTAEROGRAPHY

- advocated for deep-seated lesions not easily accessible by conventional surgery; delayed and variable rates of seizure freedom (25-64% of patients)

ENDOSCOPY

- currently used for seizure control only; up to 80% seizure freedom; immediate therapeutic effects without collateral damage from approach, hemorrhage, or clinical side effects relatable to edema.
- tissue temperatures achieved during LITT are well below those achieved by direct current without collateral damage from approach, hemorrhage.
- 82% of patients with epilepsy; 64% achieved Engel class I outcomes with follow-up 12-28 months; re-imaging (6-21 months) indicated lesion diminution with surrounding liquefactive necrosis.

LITT of left parahippocampal gyrus cavernous malformation:

Case series


- 5 patients with epilepsy; Visualase system
- no acute hemorrhage
- no adverse events or neurological deficits.
- 4 of 5 (80%) patients achieved freedom from disabling seizures (Engel class I outcome), with follow-up 12-28 months;
- re-imaging (6-21 months) indicated lesion diminution with surrounding liquefactive necrosis.

Case series


- 19 patients with epilepsy; Erbitu; Visualase system.
- most received IV dex.
- magnetic susceptibility of quested blood products within CCM can compromise MR thermography within the boundaries of these lesions, but perilesional cortex is imaged with relative ease.
- 82% (>12 months of follow-up) achieved Engel class I outcomes (59% were Engel class IA) – same as with open resection; 2 patients who were not seizure-free became so following open resection.
- LITT presents no barrier to subsequent intracranial monitoring or open resection.
- delayed imaging validated CCM involution (median 83% volume reduction) and ablation of surrounding cortex.
- histopathologic examination of one previously ablated CCM following open surgery confirmed obliteration.
- SLA caused no detectable hemorrhages.
- 2 symptomatic neurologic deficits (visual and motor) were predictable, and neither was permanently disabling.

BRAINSTEM Cavernomas

Once considered inoperable lesions, brainstem cavernomas are now surgically curable with acceptable operative morbidity. Recommending surgery is facilitated by grading system designed specifically for brainstem cavernomas:

Garcia, Roxanna M "Brainstem Cavernomas: Surgical Results in 14 Patients and a Proposed Grading System to Predict Neurological Outcome." Neurosurgery: March 2015 - Volume 76 - Issue 3 - p. 265-278

- prevalence = 0.4%.
- may accompany Sturge-Weber syndrome or Rendu-Osler disease.

CAPILLARY TELANGIECTASIA

- microscopic tracts of dilated capillary vessels (saccular or fusiform dilations) with normal brain tissue in between.
- related pathologically to Cavernomas (extremes of one nosologic entity - CAPILLARY MALFORMATION). 
- vessels lack muscular and elastic components.
- interconnected among highly brain parenchyma, not associated with glialosis (vs. cavernomas – no intervening neural tissue, surrounding tissue is glotic).
- no evidence of associated hemorrhage.
- grossly - tiny lesions having appearance of punctate hemorrhages.
- usually deep within brain (particularly in brain stem).

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PATHOLOGY

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- usually deep within brain (particularly in brain stem).

The only feature that differentiates CAPILLARY TELANGIECTASIA FROM CAVENOMS ANOMDAS is presence of brain parenchyma between vascular channels.
INTRACRANIAL VASCULAR MALFORMATIONS

### Dilated capillary spaces at gray-white interface

**CLINICAL FEATURES**
- almost always are **clinically silent**
- clinically significant hemorrhage very rare.

**DIAGNOSIS**
- not detectable radiographically by MRI, CT, angiography!!
- found incidentally on autopsy.
- occasionally visible on T2-MRI - as tiny area of intensity change (represents previous subclinical hemorrhage).

**TREATMENT**
No treatment is indicated.

**DEVELOPMENTAL VENOUS ANOMALY (DVA), s. Venous Angioma, Venous Malformation**
- most common type of intracranial vascular malformation! (prevalence ≈ 2.6%).

**ETIOPATHOPHYSIOLOGY**
- intrauterine ischemic event during formation of medullary veins → collateral venous drainage.

**PATHOLOGY**
- enlarged collection of dilated veins (architecture essentially normal, except for size).
  - vs. VARIX - single dilated vein.
- postcapillary structure (no ARTERIAL or CAPILLARY abnormalities).
- veins receive drainage from adjacent healthy tissues (neural parenchyma in and around angioma is histologically normal).
- radial arrangement - all veins converge on enlarged central venous trunk (this trunk drains into healthy superficial [cortical] or deep [subependymal] venous systems).
- No interruption of physiologic drainage! – venous angioma is anomaly (or even normal variant) rather than pathological structure.
- no mass effect!
- most frequent in white matter (cerebral hemispheres > cerebellum), usually close to brain's surface.
- walls of veins are thickened and hyalinized and usually lack elastic tissue and smooth muscle.

**CLINICAL FEATURES**
Generally, completely asymptomatic!
- some patients may present with headache, hemorrhage, seizure (esp. frontal lobe lesions), focal neurologic deficit (esp. posterior fossa lesions).
- if DVA bleeds (ICH) – can be associated with cavernoma.

**DIAGNOSIS**
- CT/MRI – tubular curvilinear structure (“spokes of wheel”); MRI may have sufficient resolution to reveal “caput medusae”.
- CT may reveal enhancing area (linear, tubular, spotty, or nodular).
Postcontrast CT - two tubular enhancing structures that extend from ventricular margin to brain surface through normal brain tissue; superficially these became continuous with surface veins, which drained into superior sagittal sinus.

T1-MRI (left frontal venous angioma):

Bilateral cerebellar venous angioma draining into large peritonsillar tributaries (axial and coronal gadolinium MRI):

**ANGIOGRAPHY**

- “Hydra” or “caput medusae” appearance (smaller radial veins converging on central draining venous trunk) – confirms diagnosis (but MRI appearance is sufficiently characteristic to forgo angiography!).
  - Angiography is used if AV shunting is suspected; DVA is best seen in late venous phase, i.e. no AV shunting.
  - Trace lesion from its nidus to either ventricular or subarachnoid surface.

- Intracranial Vascular Malformations

Via30 (25)
INTRACRANIAL VASCULAR MALFORMATIONS

INTRACRANIAL VASCULAR MALFORMATIONS

Vas30 (26)

TREATMENT

Angioma may be part of established venous drainage for adjacent healthy neural tissue - avoid excision or ablation - can lead to venous infarction.

- venous angioma is pathophysiologically related to CAVEROUS ANGIOMAS - in case of hemorrhage, investigate for adjacent cavernous angioma - if it is found, resect clot and cavernous angioma, but do not resect venous angioma!!

Direct (s. Arteriovenous) Fistula

- acquired lesions:
  a) dural arteriovenous fistula (DAVF)
  b) vein of Galen aneurysmal malformation
c) carotid cavernous fistula (see p. TrH9)

- single or multiple dilated arterioles that connect directly to vein without nidus.
- high-flow, high-pressure lesions.
- low incidence of hemorrhage (except some dural AVFs).

1. DURAL ARTERIOVENOUS Fistula (DAVF) / Shunt / Malformation

- women > men.
- rare (10-15% of all intracranial VMs).
- most patients > 60 yrs.

ETIOPATHOPHYSIOLOGY

- Direct AV shunt located within dura (e.g. dural sinus wall) between meningeal arterial branches and DURAL VENOUS SINUSES.

Etiology – ACQUIRED:

*-therefore, term “MALFORMATION” is not correct

a) traumatic tear in branch of middle meningeal, intracranial or even occipital, artery (drainage into venous sinus develops later).

b) dural sinus thrombosis → attempted recanalization → opening of embryonic AV communications → fistula creation.

- arterial supply - meningeal (dural) branches of ICA/ ECA/ vertebral artery.
- venous drainage - into nearest sinus (occasionally to adjacent cortical veins).
- posterior fossa > above tentorium.

CLINICAL FEATURES

Many are asymptomatic!

Clinical presentation depends on location and venous drainage pattern:

1. Bruit, pulsatile tinnitus (lesions shunting into transverse or sigmoid sinus).
2. Pulsations (lesions shunting into cavernous sinus).
3. Cranial nerve involvement (3, 7, 8, and 12 most common).
4. CNS manifestations (headache, seizures, motor weakness, brain stem and cerebellar syndromes, neuropsychiatric syndromes); mechanisms:
   a) intracranial venous hypertension
   b) decreased CSF absorption
   c) venous sinus thrombosis
   d) intracranial hemorrhage (subdural, SAH, ICH) – only lesions which reflux into cortical veins! In general, AVFs do not bleed!!
   e) steal phenomenon → neurologic deficits – some cases present as PSEUDOTUMOR with papilledema and headache only.

- spontaneous thrombosis with symptom remission can occur.

DIAGNOSIS
INTRACRANIAL VASCULAR MALFORMATIONS

Disease of flow – diagnosis and classification requires detailed catheter angiography (incl. ICA and ECA);

Normally dural arterial branches are not seen angiographically, but DAVFs are well visualized!

- cannot be consistently diagnosed with CT or MRI (may only show enlarged dural sinuses or cortical veins; also complications - hemorrhage and infarction).
- MRA / CTA may show abnormal vessels but catheter angiography is still required to make definitive diagnosis.

CT - grossly dilated superior ophthalmic vein

MRI - grossly dilated superior ophthalmic vein

Case courtesy of Dr Ian Bickle, Radiopaedia.org, rID: 31702

A. External carotid arteriogram – early opacification of sigmoid sinus and adjacent veins.
B. Common carotid arteriogram after fistula embolization – obliterated AV shunts, preserved proximal segments of feeding arteries.
**CLASSIFICATIONS**

**Cognard classification**

- **Type I**: confined to sinus wall with normal antegrade flow
- **Type IA**: confined to sinus with reflux (retrograde) into sinuses but not cortical veins
- **Type IB**: drains into sinuses with reflux (retrograde) into cortical veins (10-20% hemorrhage)
- **Type II A+B**: drains into sinuses with reflux (retrograde) into both sinuses and cortical veins
- **Type III**: drains directly into cortical veins (not into sinus) (40% hemorrhage)
- **Type IV**: drains directly into cortical veins (not into sinus) with venous ectasia (65% hemorrhage)
- **Type V**: spinal perimedullary venous drainage (associated with progressive myelopathy)

**Borden classification**

- **Type I**: drains anterograde into sinuses. Do not bleed!
  - **Type Ia**: supplied by single meningeal artery
  - **Type Ib**: supplied by multiple meningeal arteries
- **Type II**: drains into sinuses with both anterograde and retrograde drainage (via subarachnoid veins).
- **Type III**: drains retrograde into subarachnoid veins (no sinus drainage).

**Annual bleeding rates:**

<table>
<thead>
<tr>
<th>Border</th>
<th>Bleeding rate</th>
<th>Bleeding rate if has venous ectasia</th>
<th>Rebleeding rates after hemorrhage</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>0%</td>
<td></td>
<td>46%</td>
</tr>
<tr>
<td>II</td>
<td>6%</td>
<td></td>
<td>46%</td>
</tr>
<tr>
<td>III</td>
<td>10%</td>
<td>21%</td>
<td>46%</td>
</tr>
</tbody>
</table>

N.B. DAVF types with retrograde drainage into cortical veins (Borden type II-III) are called aggressive DAVF – due to high risk (>15%/yr) of complications (mostly hemorrhagic);
- 2-3% benign DAVF spontaneously convert into aggressive DAVF types; therefore, Borden type I is used observation.
- anterior cranial fossa DAVFs always have retrograde cortical drainage (and are always treated) because there is no venous sinuses in the proximity of the fistula to route the venous drainage away from cortical veins.

**Djindjian and Merland classification**

A more aggressive natural history and more severe clinical presentation are associated with retrograde venous drainage and venous drainage into leptomeningeal veins.
(Djindjian and Merland 1978).

**TREATMENT**

**AVFs do not bleed!**

- Major indication for treatment – retrograde cortical drainage!

**A. Observations**

- (for Borden I, Cognard I-IIA), any symptom change → MRA / CTA – if suspicious → angiography.

**TREATMENT**

In the event the brain uses cortical reflux veins for drainage (especially if there is reflux in the vein of Labbé), only arterial feeder disconnection should be done without disturbing the venous aspect of the lesion. If there is no evidence of brain dependence on cortical reflux veins, a classic cortical venous reflux (CVR) disconnection can be done. In patients with neurologic deficits that may be due to venous congestion, only arterial feeders are treated. In patients with neurologic deficits that may be due to venous congestion, only arterial feeders are treated.

**B. Vascular compression**

- initial treatment for lesion in transverse or sigmoid sinus – manual compression of occipital artery (behind mastoid for up to 30 min) – diminished inflow can induce thrombosis; contraindication to procedure – cortical venous drainage.

**C. Endovascular embolization**

- treatment of choice:
  - a) PVA particles - low morbidity rate but high recanalization chances.
  - b) Liquid materials - high cure rates but high morbidity rate (great risk of stroke).
  - c) coils (e.g. into sinus → hepatic for life)
  - goal – occlusion of AV shunting site or vein with preservation of venous sinus patency.
  - Care should be taken not to redirect venous flow toward veins that drain brain by occlusion of alternative outflow pathways!
  - approaches:
    - transarterial - selective catheterization of arterial feeding vessels that can be occluded, but the fistula itself is rarely obliterated;
      - not curative because many smaller feeders cannot be emobolized. If residual flow is present in the fistula, further feeders are likely to be recruited leading to recurrence.
Some arteries are too microscopic to be occluded, and because of small arterioles in the dura and wall of sinuses, these lesions often are hard to cure. 

- very effective adjunct before surgical treatment of DAVF because it can significantly reduce procedural blood loss.

b) transvenous - often necessitates sacrifice of venous sinus segment at zone of AV shunting; indications - multiple sites of shunting, multiple sources of arterial supply, inaccessible arterial sources; particularly useful in treatment of cavernous sinuses dural fistulas.

- feasible only if venous phase angiography has documented the absence of venous drainage of normal brain by the involved sinuses.
- impossible in Borden III lesions, which do not drain through a venous sinus but directly in cortical veins.

D. SURGERY:

a) excision (traditional surgical treatment of DAVFs) – complete excision of the fistula and the surrounding dura - approach involves the disconnection of all feeding arteries and arterialized leptomeningeal veins and excision of the draining sinuses, when not used by brain, together with pathologic dura (if brain uses the sinuses, the sinus is skeletonized and left patent).

b) direct surgical exposure, catheterization, and packing of the involved sinuses with coils or other thrombogenic material (e.g., Gelfoam, silk sutures)

c) selective disconnection of the arterialized leptomeningeal veins – simpler, less invasive, and less morbid option of selectively eliminating cortical venous drainage to convert DAVFs into benign lesions and eliminate the risk of bleeding and neurologic deficit; safe only when the brain does not use the reflux veins for its own drainage.

- even if it is judged from angiography that CVR veins do not drain normal brain, it is safer to apply a clip on major draining veins before disconnecting them. The brain should be observed for any swelling resulting from impaired venous drainage for a few minutes before coagulating and anatomically dividing the vein.
- because several veins can contribute to the fistula, some of them being smaller and more difficult to identify than the major draining vein, it cannot be stressed enough how important it is to ensure all arterialized veins are identified and disconnected.

- preoperative embolization and reduction of arterial feeders is a useful adjunct to surgery.
- intraoperative ICG (indoxyline green) angangiography is gold standard.
- if profuse venous bleeding is encountered from a tear at the junction of a vein and its draining sinus, compression and holding patiently until the bleeding stops generally works well.

Suetsugu procedure – resecting transverse sinus due to multiple DAVFs.

E. RADIOTHERAPY: results in obliteration of DAVFs between one and three years after treatment; drawback – delayed fistula closure (not suitable for aggressive DAVFs with annual bleeding risk up to 15%)

- embolization is performed after SRS to avoid the pitfall of having embolization temporarily obscure portions of the nidus that would then not be targeted during the SRS.

2. VEIN OF GALEN Malformation / Ectasia / Aneurysm

- specific form of congenital AVM - abnormal vessels drain directly into vein of Galen without interconnected capillary system.

PATHOLOGY, CLASSIFICATIONS


type I - fistulae located in varix wall arise from feeders from ACA (pericallosal) and/or PCA (posterior choroidal) arteries (i.e. pericallosal and choroidal arteries are sole supply to varix).

type II - fistulae located in wall of varix arising from feeders from trans-mesencephalic and trans-diencephalic arteries (i.e. fistulae are purely from arteries traveling through mesencephalon and diencephalon into varix).

type III (most common type) - combination of types I and II.

type IV - separate diencephalic / mesencephalic AVMs draining into enlarged, but otherwise normal, vein of Galen (i.e. no direct fistula to vein of Galen itself) - anomalous enlargement of vein of Galen is only secondary manifestation of AVM!

Type I malformation
**Intracranial Vascular Malformations**

**Type III malformation - multiple fistulae between choroidal and vein of Galen**

Massive dilatation of vein of Galen and of associated draining vessels; mural thrombi formed on their walls.

**CLINICAL FEATURES**

Usually manifests during early childhood:

1. **Tremendous A-V shunting:**
   1) neonatal progressive high-output cardiac failure (becomes apparent 1-2 hours after birth - babies born with very hyperactive precordia).
   2) pan-cardiac cycle *bruit* involving chest, neck, and head.
2. **Obstructive hydrocephalus (80%)** - due to:
   a) venous hypertension
   b) obstruction of CSF pathways (e.g. midbrain compression).

In older children: headache, seizures, SAH, progressive neurological deficits (due to cerebral ischemia).

- tends to be progressive even in its most benign forms (frequently fatal!).

**DIAGNOSIS**

*Doppler imaging* in infants - detection of ectatic vein (hypervascular midline structure with demonstrable pulsations) → *MRI / angiography.*

- *contrast enhancement is not necessary* if CT or MRI is used.

  N.B. minimally toxic effects of contrast agent may prove significant in newborn baby with cardiac or other organ failure (minimal allowance of contrast agent should be saved for any possible therapeutic intervention!)

- *transvaginal ultrasound* is very helpful in prenatal diagnosis (in fetuses with prenatal cardiomegaly).
Intracranial Vascular Malformations

ICA arteriogram (lateral projection) - type I malformation: enlarged ACA and posterior choroidal artery entering nidal of vein of Galen malformation.

ICA angiogram (lateral projection) - type I malformation: posterior choroidal branches of PCA (small arrows) contribute supply to dilated vein of Galen (large arrow).

ICA arteriogram (lateral projection) - type I malformation: single fistulous connection.

VA angiogram (lateral projection) - type II malformation: note multiple arterial feeders.

ICA arteriogram (lateral projection) - type III malformation: notice duplicated straight sinus.

Same patient – VA arteriogram (lateral projection): thalamoperforators supplying aneurysm.

T1-MRI – type IV malformation: large complex midline angioma drains into aneurysmal vein of Galen, note dilated straight sinus and aneurysmal torcular.

T1-MRI notice stenosis of straight sinus.
INTRACRANIAL VASCULAR MALFORMATIONS

CT - aneurysmal dilatation of Galen vein owing to large, deep malformation; note enlargement of draining sinuses and mild hydrocephalus.

CT - thrombosed vein of Galen aneurysm.

MRA - vein of Galen aneurysm and associated abnormal draining veins.

Hydrocephalus associated with vein of Galen malformation.

Doppler (sagittal section): rounded midline vascular structure (A), with swirling flow (red and blue), proximal stenosis (arrow) on draining sinus (S).

TREATMENT

- aimed at AV shunt reduction.

A. **Open SURGICAL occlusion** of fistula (via supratentorial parieto-occipital approach).

B. **Percutaneous EMBOLIZATION** (treatment of choice!): using wire coils (Gianturco coils) via tethering plunger system that allows precise positioning of coils.
   a) transvenous approach (transorbital or transfemoral).
   b) transarterial approach
      - all depositions of wires must be extremely careful - ventral part of malformation is paper-thin!
      - various interventional treatment modalities (PCAL, pericallosal artery; A, aneurysm).
INTRACRANIAL VASCULAR MALFORMATIONS

Vas30

- Gianturco coil
- Short angiography catheter
- Detachable balloon
- Microcoil
- Wire basket within vein of Galen aneurysm.

- high risk of hemorrhage! - treat in graded, multisection fashion (2-4 treatments during first several days of life).
- preoperative correction of cardiac failure is critical. Cardiac failure unresponsive to medical management is indication for urgent embolization in neonatal period.
- complete anatomical occlusion may not always be achieved, but cardiac failure can be rapidly reversed by reducing shunt flow (further staged treatment can be performed after child maturation).
- hydrocephalus needs ventriculoperitoneal shunt early in therapy (be careful - subependymal veins are dilated as result of abnormal flow patterns around vein of Galen complex).

SINUS PERICRANII

- thin-walled vascular spaces interconnected by numerous anastomoses that protrude from skull and communicate with superior sagittal sinus

- no symptoms (except for external swelling, occasional pulsating tinnitus, ICP↑).

- radiograph - bone defect, through which lesion communicates with longitudinal sinus.

Reformatted oblique coronal CT through filling varicosity (asterisk) - defects within both inner and outer tables (arrows) of calvaria and filling of diploe with venous blood; connecting intracranial vein is denoted by white arrowhead:

Selective right ICA angiogram, AP view (venous phase) - venous connections (arrows) to pericranial varicosity (asterisk); flow is both into and out of this pericranial varicosity:

Volume-rendered CTA of calvaria viewed from behind - multiple calvarial depressions (arrowheads) underlie slow-filling varicosities; varicosities with more direct connections to dural sinuses already show enhancement (arrows).
INTRACRANIAL VASCULAR MALFORMATIONS

BIBLIOGRAPHY for ch. “Neurovascular Disorders” — follow this LINK >>