Intracranial Vascular Malformations

ARTERIOVENOUS MALFORMATIONS (AVM)

PATHOLOGY

- Hemodynamics
- Endothelial cell proliferation
- Hyperperfusion
- Hypervascularity
- Hemorrhage

CLINICAL FEATURES

- Headache
- Seizures
- Hemorrhage
- Hemiparesis
- Aphasia
- Visual symptoms

DIAGNOSIS

- Angiography
- CT
- MRI
- MRA
- DSA

TREATMENT

- Supportive treatment
- AVM treatment
- Endovascular embolization
- Surgical resection
- Stereotactic radiosurgery
- Tumoricidal radiosurgery
- Embolization
- Surgery
- Tumor debulking

OUTCOMES

- Recurrence
- Mortality

SPINAL VESSELS

- Embolization
- Surgery

Cavernous Angioma, s. Cavernoma, Cavernous Malformation

ETIOPATHOPHYSIOLOGY

- Hemodynamic
- Neuronal and astrocytic
- Perineuronal

CLINICAL FEATURES

- Headache
- Seizures
- Hemorrhage
- Hemiparesis
- Aphasia
- Visual symptoms

DIAGNOSIS

- CT
- MRI
- MRA
- T2-weighted imaging

TREATMENT

- Supportive treatment
- Radiosurgery
- Surgical resection
- Tumor debulking

BRAINSTEM Cavernomas

ETIOLOGIC FACTORS

- Trauma
- Radiation
- Infection

CAPILLARY TELEANGECTASIA

ETIOPATHOPHYSIOLOGY

- Hemodynamic
- Neuronal and astrocytic
- Perineuronal

CLINICAL FEATURES

- Headache
- Seizures
- Hemorrhage
- Hemiparesis
- Aphasia
- Visual symptoms

DIAGNOSIS

- CT
- MRI
- MRA
- DSA

TREATMENT

- Supportive treatment
- Radiosurgery
- Surgical resection
- Tumor debulking

Intracranial Vascular Malformations

M. Hochberg, et al.

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CLASSIFICATION according to predominant vascular feature:

1) developmental venous anomaly (DVA), s. venous angiomata (most common type*) - anomalous VENs without any direct feeding artery
2) capillary telangiectasia* (next most common type!) - vessels morphologically resembling capillaries but slightly larger.
3) arteriovenous malformation (AVM)** - vessels with abnormal arteries and veins without intervening capillaries.
4) cavernous angioma (cavernoma)*
5) direct (s. arteriovenous) fistula** - required 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 cautiously 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 increase in size by incorporating adjacent vessels ("recruitment").
- most significant manifestations - BLEEDING - most likely to occur in patients < 30 yrs.

Rapid guide to diagnosis by NEUROIMAGING:

- not visible, (+) sometimes visible, + visible

<table>
<thead>
<tr>
<th>Type</th>
<th>CT</th>
<th>MRI</th>
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Vas30 (1)
INTRACRANIAL VASCULAR MALFORMATIONS

Vas30 (2)
ARTERIOVENOUS MALFORMATIONS (AVM)

PATHOLOGY, PATHOPHYSIOLOGY

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

- lesion is present from birth (vs. congenital aneurysms)
- vascular conglomerate (numerous thin-walled, tortuous channels) is called nidus
  - nidus commonly forms "pyramidal/crown" in white matter with base covering part of cerebral surface, and apex directed toward lateral ventricle - bleeding can be subarachnoid, intraventricular, 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:
- high flow subsequently leads to arteriolar dilation
- fibromuscular cushions - smooth muscle hyperplasia associated with fibroblasts and connective tissue elements
- arterial feeders:
  - a) entirely from ICA branches (purely intracranial AVMs): ACA territory > ACA territory > PCA territory; occasionally (< 10%) recruit additional supply from meningeal arteries.
  - b) entirely from ICA branches (purely dural AVMs): arteriolar 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 fistulas) - venous arterialisation;
  - AVMs of ACA and PCA may drain directly into vein of Galen, causing it to dilate in aneurysmal fashion.

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

Involved vessels may enlarge with passage of time; some AVMs* decrease over time (spontaneous thrombosis?) up to total resolution (rarely).

* esp. those around anterior fossa and chiasm

Microscopy:
- entrapped (between vessels) brain tissue is plastic and nonfunctional, often with evidence of prior 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% 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-II (85%) are flow related aneurysms; intranidal aneurysms are rare (5.5%)

Wyburn-Mason syndrome (or, Bonnet-Dechaume-Blanche syndrome) – AVM involving cerebral cortex, optic nerve, retina* + facial nevus.

*retinal vascular malformations

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 (∆P) and fourth power of radius (r) and is inversely related to tube length (L) and viscosity (n):
INTRACRANIAL VASCULAR MALFORMATIONS

**INTRACRANIAL VASCULAR MALFORMATIONS**

**Vas30**

- **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.

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)
Altered arteries and veins with intervening glotic neural plexuses

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 saccular aneurysms 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 perinatal 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 metanephrine syndromes (CAMs).
- CMS-I - proencephalic AVMs affecting hypothalamus/hypophysis in association with facial AVM of nose.
- CMS-2 - AVMs affecting lateral preencephalon (occipital lobe, thalamus) in association with facial AVMs of maxilla.
- CMS-3 - AVMs of rhombencephalon (cerebellum, pons) in association with facial AVMs of mandible.
- cerebral AVMs can be associated with pulmonary AVMs! (but not with other organ AVMs)

CLINICAL FEATURES

Only 12% AVMs become symptomatic!
Most manifest before 40 yrs! (vs. cerebral aneurysms - only ¾ patients present by age of 40)
- x-rays (> 50%) may be audible either to patient or to examiner.
- scalp or face veins may be enlarged.
- huge AVMs (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.

AVM producing neurological dysfunction through 4 main mechanisms:
A. Hemorrhage
B. Seizures
C. Progressive neurological deficit
D. Headache

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, risk factors for hemorrhage - see below >>
- prognosis & recovery tends to be better than in non- AVM-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).

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) lobate 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

NUCLEUS RADIOPHAGIS

- 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; dented 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: angio or follow-up MRI).
- low T1 signal of extracerebral hematoidin 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):

T1-MRI - small subcortical AVM in right frontal lobe.

Source of images: WebPath. The Internet Pathology Laboratory for Medical Education, by Edward C. Klatt, MD.>>
INTRACRANIAL VASCULAR MALFORMATIONS

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.

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 arrows), middle cerebral arteries, various (arrowheads) and superficial draining vein (open arrow).

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

INTRACRANIAL VASCULAR MALFORMATIONS

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

T2-weighted FSE images (A) and (B); 3D TOF MRA before (C) and after intravenous contrast (D):

- AVM nidus is of mixed signal intensity on (A) and (B); low signal areas are due to flow void in intranidal vessels; venous drainage is predominantly deep (arrow).
- Nonenhanced MRA (C) shows arterial structures and nidus, but draining veins (arrows) are only visible on contrast-enhanced MRA (D).

MR DSA (series of three frames in lateral projection acquired at 1-second 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).

- 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).

- aneurysms 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 continue past AVM to feed normal brain tissue.
Paratrigonal AVM: Preoperative angiography.

Postoperative angiography:

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

Deep cerebellar AVM fed by branches of SCA, AICA, and PICA (preoperative and postoperative 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

Tortuous collection of irregular small vessels.

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 parasplenial AVM (preoperative and postoperative angiography):

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.

**Risk factors for hemorrhage:**

1. male gender
2. small (i) AVM size (< 2.5 cm)
3. deep location in basal ganglia or posterior fossa
4. deep venous drainage
5. single or only few draining veins (esp. with aneurysm or varix)
6. high pressure in feeding arteries (as measured during angiography)
7. aneurysms (10% patients);
   - intracranial 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
8. prior hemorrhage (rebleeding risk during 1st year 7-33%*, then 2.5% annually)
9. pregnancy see below

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 medical/surgical 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

- **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
INTRANASCAL VASCULAR MALFORMATIONS

Surgical treatment risk is estimated by:

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. Eloquent 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:

- 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
- Class C - Spetzler-Martin grade IV and V lesions.

B. Supplemented Spetzler-Martin Grading System (SM-Supp, s. Lawton-Young grading system)
- ABCs of AVMs: patient Age, Bleeding or hemorrhagic presentation, and AVM Compactness

* May be slowed by proximal balloon occlusion

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

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<thead>
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<th>Spetzler-Martin Grading</th>
<th>Supplementary Grading</th>
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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%).

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) Codman Trufill n-BCA 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.

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.
Vermian AVM: Transvenous Approach:

- try to resect nidus; not just arteries (in time, collateral supply can develop to left intact nidus).
- do not occlude veins!
- do not embolize > 1/3 during one session.
- deepest part of AVM is most difficult to control if starts bleeding during the surgery, therefore, always try to embolize deep part 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; 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.

Principal uses:

A. Pre-operative measure - serial embolizations reduce AVM size (and Spetzler-Martin grade) → safer microsurgery / radiosurgery;
- embolization alone is usually not sufficient to completely obliterate AVM.
- defining surgery is performed within week of embolization (new feeding vessels and glotic scar will make surgical procedures more difficult).
- N.B. embolization prior to radiotherapy reduced rate of total obliteration after SRS!

B. Palliative / parent embolization:

a) to produce neurological relief – in Spetzler-Martin grade IV-V with venous outflow obstruction (to reduce arterial inflow to control edema) or true steal phenomenon (to block high-velocity blood shunting from high-pressure arterial system into venous system).

b) to produce headache relief – by reducing venous hypertention (eliminating ECA supply is often effective).

c) to reduce hemorrhage risk – by targeting specific components (e.g., aneurysm).

Complications - persistent neurological deficits:

- inadvertent embolization of arteries supplying normal brain tissue;
- if total portions of brain are thought to be irrigated by same vessels supplying malformation, sodium Arsenyl may be injected → patient evaluated for alterations in EEG and neurological picture
- b) obliteration of venous outflow leading to intracerebral hemorages.

MORTALITY 9-22%, MORTALITY 0.9%.

Vermian AVM embolization (left vertebral angiogram):

A. Pre-embolization: supply by vermian branches of SCA.
B. Microcatheter NBCA injected into AVM nidus.
C. Postembolization of both major pedicles – almost complete nidus obliteration.

Surgical Resection - excision without injury to adjacent brain tissue - mainstay of definitive treatment.

Intraoperative:

- most effective with easily accessible smaller lesions (Spetzler-Martin grades 1-3, ± 4).
- electrocortical stimulation and surface EEG recording under local anesthesia are useful in delineating specific cortical function and seizure focus localization.

Intraoperative Angiography is gold standard (may be supplemented by ECG+ ANGIOGRAPHY) – helps to delineate arterial feeders and draining veins – can be safely isolated and ligated.

- "nudocyanine green preserve "en passage" arteries!
- two strategies of approach:
  a) peripheral isolation of feeding arteries with marginal resection of nidus (tactical en bloc method).
  b) central retrograde 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 center and is initially identified and localized (match with angiogram) → follow into nidus;
- shunting venules and arterioles are cauterized and cut along central vein (venules are thin-walled and tear easily) - occlude 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.

nidus is totally resected.

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 excision with minimal deficits.
- larger lesions are separated into compartments and isolated and collapsed as separate units.

aneurysms are clipped surgically as well.

When intranidal aneurysm is found in AVM bleeding, it must be targeted for urgent therapy; i.e. pressure 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 hypertension during surgery (maintain MAP 40-60 mmHg)
- 2) embolization of feeding vessels prior to surgery.

Vas30 [14]
POSTOPERATIVE
- outcome correlates with score on Spetzler-Martin scale.
- AVM removal → better tissue perfusion → progressive neurological improvement.
- surgical morbidity 8.6%, mortality 3.3%.
- complications:
  1) “normal perfusion pressure breakthrough (NPPB)” (theory described by Spetzler 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 blood 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 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
- noninvasive and can access all anatomic locations of brain (e.g. surgically inaccessible).
- ideal for 3 cm AVM (Dr. 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 subendothelial 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:
- stereotactic volumetric axial plane imaging (MRI or CT) supplemented by conventional or digital subtraction angiography* is usually necessary for complete conformal dose planning. *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 methylprednisolone at the conclusion of the SRS procedure.
- if aneurysm is identified in AVM 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 > 15 cc) – 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 recreated 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 it 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).
INTRACRANIAL 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.  

<table>
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<tr>
<th>Author</th>
<th>N</th>
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<td>153</td>
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<td>227</td>
<td>56-100%</td>
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<td>Maruyama</td>
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<td>81-91%</td>
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</tr>
</tbody>
</table>

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 (recurrence 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. 3% of nonembolized patients (p = 0.006).

Dr. Sheehan

Prior embolization reduces rate of total obliteration after SRS (but risks of hemorrhage during latency period are not affected by prior embolization).

Indication (for AVM 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 angiogram 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
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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) preoperative 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 (i.e. 10% per year) vs. 1.1% baseline annual risk – data from Brigham and Women’s Hospital and Harvard Medical School**

- **Hemorrhage From Arteriovenous Malformations During Pregnancy. Christopher L Davidoff et al. Neurosurgery, Volume 85, Issue 1, January 2019, Pages 25% – 10.8% per year) vs. 1.1% baseline risk parameters.

- 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: transfrontal-transventricular, transcallosal-transventricular, transvesslans-transventricular.
- approaches for THALAMUS – interhemispheric: transcallosal or trans-splenial.
- after microsurgical isolation of feeding perforating vessels, lateral ventricle is opened and lesion excised (incl. choroidal plexus - carries deep arterial supply to nidus).
- surgical morbidity is formidable (hemiplegia, aphasia, hemianopsia, 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 ceded 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 n = 20%)

- CEREBELLUM AVMs should be surgically removed (natural course is treacherous).
- approach is tailored to specific location (e.g. transcisternal route for lesions around tentorium and superior cerebellum).

- BRAINSTEM AVMs:
  a) microsurgical resection - combined subtemporal-suboccipital-retro labyrinthine-transcisternal approach.
  b) radiosurgery (rate of obliteration at 2, 3, 5, 7, and 10 yr after SRS - 24.5%, 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 hemotoma requires emergent evacuation surgery – decompress hemotoma while avoiding AVM.
- attempting to more completely remove hemotoma can result in bleeding.
  - in case of significant brain swelling, adding a dural 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) RITI (CCM1) b) MGC4607 (CCM2) c) PDCD10 (CCM3) d) 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**

- hamartomatous enlarged sinusoidal capillaries, s. cavern (sangle 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.

---

**No invironing neural tissue**

- not associated with enlarged feeding arteries or draining veins.
- base flow is low or even stagnant.
- intra-lesional thrombosis, calcification and recanalization are typical.
- range from soft to hard.

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INTRACRANIAL VASCULAR MALFORMATIONS

Adjacent neural tissue may be affected – gliotic (form capsule), small subclinical hemorrhages (perileisional hemosiderin may incite epileptogenic focus).

30% cases have associated DVAs.

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.

Adjacent neural tissue may be affected – gliotic (form capsule), small subclinical hemorrhages (perileisional hemosiderin may incite epileptogenic focus).

30% cases have associated DVAs.

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.

- Clinical Features

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.

*less than with AVMs or dural AV fistulae.

4. Mass effect: headache (6-52%)

Supratentorial lesions are frequently associated with seizures while infratentorial lesions are likely to be associated with focal neurologic deficits.

Headaches are prominent wherever angiomata are located.

Symptomatic lesions are likely to remain symptomatic or progress.

DIAGNOSIS

Angiography

- demonstrates 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.

- Blood products of various ages in center.

- Mass effect only seen if bleeding has occurred.

- Local edema may be present.

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

Cavernoma of pons (T2-MRI):

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

MRI (FLAIR) - cavernous hemangioma in right temporal lobe:

MRI (T1) - left-sided parietal 2-cm cavernous malformation with areas of hemorrhage of various chronicity + acute intra-parenchymal blood anterior to lesion:

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

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 (arrow).

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.

*unless thrombosed, or after hemorrhage
INTRACRANIAL VASCULAR MALFORMATIONS

Lesion in right middle cerebellar peduncle:
Unenhanced CT – increased density: Enhanced CT – minimal enhancement

Left parietal hyperdensity:

TREATMENT

Indications:
1) intractable (!) seizures; some experts think that 3 seizures under adequate AED is enough to warrant surgery (due to esp. high risk of kindling with repeated seizures in cavernomas; thus, some experts say it is enough to fail just one AED to qualify for surgery).
2) symptomatic increase in lesion size (thus, newly diagnosed cavernomas should be followed yearly with MRI, esp. if near eloquent areas).
3) gross (!) hemorrhage or ≥ 2 rebleeds.

SURGERY

- ictal ECoG‐guided resection including surrounding cortex* is considered the gold standard (70–80% 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!

RADIOSURGERY

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

LASER ABLATION

- 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 electrocautery, thus, unlikely to provide direct hemostasis.

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 1 outcome), with follow-up 12-28 months.
- reimagining (6-21 months) indicated lesion diminution with surrounding liquefactive necrosis.

Case series

  https://doi.org/10.1111/epi.14634
- 19 patients with epilepsy, Emory. Visualase system.
  - most received IV dex.
  - magnetic susceptibility of sequestered blood products within CCMs 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 before 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.

N.B. cavernoma itself is not epileptogenic (it contains no nervous tissue) but adjacent hemosiderin-lade cortex is!
INTRACRANIAL VASCULAR MALFORMATIONS

2 symptomatic neurologic deficits (visual and motor) were predictable, and neither was permanently disabling.

BRAINSTEM Cavernomas

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

CAPILLARY TELANGIECTASIA

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

PATHOLOGY

- microscopic nests 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.
- intercalated among healthy brain parenchyma, not associated with gliosis (vs. cavernomas – no intervening neural tissue, surrounding tissue is gliotic).
- related pathologically to CAVERNOMAS (extremes of one nosologic entity – CAPILLARY MALFORMATION).
- no evidence of associated hemorrhage.
- grossly - tiny lesions having appearance of punctate hemorrhages.
- usually deep within brain (particularly in brain stem).

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%).

DVA – dilated communication between deep and superficial venous systems; no AV shunting?

ETIOPATHOPHYSIOLOGY

- intrauterine ischemic event during formation of medullary veins → collateral venous drainage:

PATHOLOGY

- enlarged collection of dilated veins (architecture essentially normal, except for size).
- postcapillary structure (no ARTERIAL or CAPILLARY abnormalities).
- veins receive drainage from adjacent healthy tissues (neural parenchyma in and around angioma is histologically normal).
- veins separated by normal brain tissue.
- 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?
- no mass effect!
- most frequent in white matter (cerebral hemispheres > cerebellum), usually close to brain's surface.
**INTRACRANIAL VASCULAR MALFORMATIONS**

- **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 become continuous with surface veins, which drained into superior sagittal sinus.

**T1-MRI (left frontal venous angioma)**

Bilateral cerebellar venous angiomas 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**

**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 pathophysio logically related to Cavernous 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

- 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.
INTRACRANIAL VASCULAR MALFORMATIONS

**Etiology – acquired**: "Therefore, term “malformation” is not correct

- Traumatic tear in branch of middle meningeal, intraorbital or even occipital, artery (drainage into venous sinus develops later).
- 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. Proptosis (lesions shunting into cavernous sinus).
3. Cranial nerve involvement (III, 7, 8, and 12 most common).
4. CNS manifestations (headache, seizures, motor weakness, brain stem and cerebellar syndromes, neuropsychiatric syndromes); mechanisms:
   - Intracranial venous hypertension
   - Decreased CSF absorption
   - Venous sinus thrombosis
   - Intracranial hemorrhage (subdural, SAH, ICH) - only lesions which reflux into cortical veins! In general, AVFs do not bleed!!!
   - Steal phenomenon → neurologic deficits
     - Some cases present as PSEUDOTUMOR with papilledema and headache only.

- Spontaneous thrombosis with symptom remission can occur.

### DIAGNOSIS

**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**

*Case courtesy of Dr Ian Bickle, Radiopaedia.org, rID: 31702*
INTRACRANIAL VASCULAR MALFORMATIONS

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

W 1855: L 2303

Case courtesy of APFOS (Anat Frank Gaillard, Radiopaedia.org, rID: 9971)

CLASSIFICATIONS

COGNARD classification
Type I: confined to sinus wall with normal anterograde flow
Type II A: confined to sinus with reflux (retrograde) into sinus but not cortical veins
Type II B: drains into sinus with reflux (retrograde) into cortical veins (10-20% hemorrhage)
Type III: drains 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 sinus.
Type Ia – supplied by single meningeal artery
Type Ib – supplied by multiple meningeal arteries
Type II: drains into sinus with both anterograde and retrograde drainage (via subarachnoid veins)
Type III: drains retrograde into subarachnoid veins (no sinus drainage)

Annual bleeding rates:

<table>
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<tr>
<th>Bleeding rates</th>
<th>Bleeding rates if has venous ectasia</th>
<th>Rebleeding rates after hemorrhage</th>
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<tbody>
<tr>
<td>I 0%</td>
<td></td>
<td>0%</td>
</tr>
<tr>
<td>II 6%</td>
<td>46%</td>
<td>46%</td>
</tr>
<tr>
<td>III 10%</td>
<td>21%</td>
<td>46%</td>
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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 DAVFs spontaneously convert into aggressive DAVF types; therefore, Borden type I need observation.
- anterior cranial fossa DAVFs always have retrograde cortical drainage (and are always treated) because there is no venous sinus in the proximity of the fistula to route the venous drainage away from cortical veins.

DIJNDJAN 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.
(Dijndjian and Merland 1978).

TREATMENT

AVFs do not bleed!
Major indication for treatment – retrograde cortical drainage!

A. OBSERVATION (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 Labbe),
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 disconnected.
B. Vascular compressions – initial treatment for lesion is \textit{transverse} of \textit{sigmoid sinus} - manual compression of \textit{rectal artery} (behind mastoid) for up to 30 min \textit{--} diminished inflow can induce thrombosis; contraindication to procedure – cortical venous drainage.

C. Endovascular embolization \textit{treatment} of choice:

- \textit{A} \textit{P}VA \textit{particles} - low morbidity rate but high recanalization chances.
- \textit{b} \textit{liquid materials} - high cure rates but high morbidity rate (great risk of stroke).

- \textit{Onyx-18} (ethylenevinylalcohol copolymer)
- \textit{N}BCA (N-butyryl-cyancrylate)
- \textit{c} \textit{coils} (e.g. into sinus \textit{-->} laceration for life)

- \textbf{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!

\textbf{approaches:}

- \textit{transarterial} - superselective catheterization of arterial feeding vessels that can be occluded, but the fistula itself is rarely obliterated.
  - \textit{not curative} because many smaller feeders cannot be embolized. 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.

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

- \textit{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 sinus dural fistulas.

- \textit{feasible} only if venous phase angiography has documented the absence of venous drainage of normal brain by the involved sinus.

- \textit{impossible in Borden III lesions}, which do not drain through a venous sinus but directly in cortical veins.

D. \textbf{SURGERY:}

- \textit{sclerosis} (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 sinus, when not used by brain, together with pathologic dura (if brain uses the sinus, the sinus is skeletonized and left patent).

- \textit{direct surgical exposure}, catheterization, and \textit{packing} of the involved sinus with coils or other thrombogenic material (e.g., Gelfoam, silk sutures)

- \textit{selective disconnection} of the arterialized leptomeningeal veins – simpler, less invasive, and less morbid option of selectively eliminating cortical venous drainage (to convert DAVFs to 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 safe 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.

- \textit{preoperative embolization} and \textit{reduction} of arterial feeders is a useful adjunct to surgery.

- \textit{intraoperative ICG (indocyanine green) angiography} in gold standard.

- \textit{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.

\textbf{Suzuki procedure} – resecting transverse sinus due to multiple DAVFs.

e. \textbf{RADIOSURGERY:} results in obliteration of DAVFs between one and three years after treatment; 
\textbf{disadvantage} – delayed fistula closure (not suitable for aggressive DAVFs with annual bleeding risk up to 15%).

- \textbf{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.

\section*{2. VEIN OF GALLEN Malformation / Ectasia / Aneurysm}

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

\section*{PATHOLOGY, CLASSIFICATIONS}


\begin{itemize}
  \item \textbf{type I} - fistulae located in varix; wall arise from feeders from ACA (pericalloital) and/or PCA (periorbit chorial) arteries (i.e. pericalloital and chorial arteries are sole supply to varix).
  \item \textbf{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).
  \item \textbf{type III} (most common type) - \textit{combination} of types I and II.
  \item \textbf{type IV} - \textit{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!
\end{itemize}
**INTRACRANIAL VASCULAR MALFORMATIONS**

**Type I malformation:**

Type I malformation - multiple fistulae between choroisals 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**
INTRACRANIAL VASCULAR MALFORMATIONS

Doppler imaging in infants - detection of ectatic vein (hypervascular midline structure with demonstrable pulsations) \(\rightarrow\) 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 prenat al cardiomegaly).

- **ICA arteriogram (lateral projection)** - type I malformation: enlarged ACA and posterior choroidal artery entering midline vessel with Galen malformation.
- **VA angiogram (lateral projection)** - type II malformation: posterior choroidal branches of PCA (small arrows) contribute supply to dilated vein of Galen (large arrow).
- **ICA arteriogram (lateral projection)** - type III malformation: single fistulous connection.
- **VA angiogram (lateral projection)** - type IV malformation: note duplicated straight sinus.
- **ICA arteriogram (lateral projection)** - type III malformation: notice duplicated straight sinus.
- **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.

Note: Contrast enhancement is not necessary if CT or MRI is used. However, it may be necessary in cases of cardiac or other organ failure to minimize the risk of contrast agent toxicity. Transvaginal ultrasound can be helpful in prenatal diagnosis, especially in cases with prenatal cardiomegaly. The images illustrate different types of intracranial vascular malformations, with corresponding diagnosis and treatment options.
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 para-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 (transocular 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

**INTRACRANIAL VASCULAR MALFORMATIONS**

Vas30 (33)

1. Gianturco coil
2. Short angiography catheter
3. Detachable balloon
4. Microcoil
5. Wire basket within vein of Galen aneurysm.

- **high risk of hemorrhage!** - treat in **graded, multisession 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.
  - **EXTERNAL PROTRUBANCES** - at any portion of **SKULL MIDLINE** (most often in midportion of forehead);
    - Appears early in life.
    - Soft and compressible, increases in size when venous pressure in head is raised (by coughing, straining, lowering head).
    - May enlarge slowly over years.
  - **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) - defect 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 ▶

Viktor’s Notes™ for the Neurosurgery Resident
Please visit website at www.NeurosurgeryResident.net