Intracranial Vascular Malformations

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Classification according to predominant vasculature:

1) developmental venous anomaly (DAV, s. venous angioma (most common type)) - anomalous veins without 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 veins 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 increase in size by incorporating adjacent vessels (recruitment).
- most significant manifestation - bleeding - most likely to occur in patients < 30 yrs.

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

<table>
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<th>Type</th>
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<th>MRA</th>
<th>Angio</th>
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<tr>
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<td>Cavernoma</td>
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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 "pyramid/corneum" in white matter with base covering part of cerebral surface, and apex directed toward lateral ventricle - bleeding can be subarachnoid, 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.

**ARTERIOVENOUS shunt is definitive characteristic!**

- feeding arteries may run serpentine course through sulci before entering nidus;
- high flow subsequently leads to ARTERY DILATION
  - fibromuscular channels - smooth muscle hyperplasia associated with fibrinoids and connective tissue elements
  - arterial feeders:
    - a) entirely from ICA branches (PURELY INTRACRANIAL AVMs): MCA territory > ACA territory > PCA territory; occasionally (< 10%) recruit additional supply from meningal arteries
    - b) entirely from ECA branches (PURELY DURAL AVMs)
- 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 ANEURYSMALIZATION;
- AVMs of ACA and PCA may drain directly into vein of Galen, causing it to dilate in aneurysmal fashion.

**ARTERIAL DILATION**

- 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 acute 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).

- 2.1-16.7% 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%)**

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

**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 (ΔP) and fourth power of radius (r) and is inversely related to tube length (L) and viscosity (η):

$$Q = \frac{\pi \times r^4 \times \Delta P}{8 \times \eta \times L}$$
**INTRA CRANIAL 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 PCO$_2$ changes).
- Tissues adjacent to AVM may be persistently mildly hypoxic (malformation may steal blood from adjacent healthy tissue).
- If AVM is corrected, pressure within feeding arteries rises by $\approx 60\%$ to normal values, and normal CO$_2$ reactivity is immediately established in adjacent cerebral vessels.

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

Tangle of abnormal vessels on brain surface:

Tangled complex of blood vessels with intervening neural parenchyma:

Intraventricular and intracerebral hemorrhage due to ruptured AVM:

Microscopic appearance of AVM – dilated, tortuous, worm-like vascular channels:
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 sporadic 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) cerebrofacial arteriovenous metameric syndromes (CAMSSs)

- **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 ⅓ patients present by age of 40)

- **bruits** (= 50%) may be audible either to patient or to examiner.
- **scap or face veins may be enlarged.**
- **large 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.

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 sacular 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 of 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).

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

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 to several years).

- reflects AVM location.
- **mechanism:**
  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**

**SKULL RADIOGRAPHS**
- 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 hypodense 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 + HBV 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.

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

**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 extracellular hemosiderin may be seen around or within AVM mass, indicating prior hemorrhage.
- there may be regional brain atrophy.

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

Vas30

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

Surface-based 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

Vas30 (7)

**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 arrows); varices (arrowhead) and large superficial draining vein (open arrow) all apparent on first frame, indicating speed of shunting.

2) early opacification of enlarged, tortuous draining veins.

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, lumbar or carotid AV fistulae is best evaluated by angiography.
  - Neuroanatomy cannot detect it (often 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 AMYTHAL (anesthetic agent) to produce temporary anesthesia of area perfused by artery - “superselective Wada testing” - language, memory, visual-spatial, sensory, motor functions can be tested during 5 minutes - to determine whether “eloquent” function originates in AVM region (risk for neurological deficits during embolization or surgery):

   - Arteries directly feeding AVM
   - “En passage” arteries - feed AVM but continue 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

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 parasplenic 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 anterior 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 (!) 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 kinking or stenosis or varix)
6. high pressure in feeding arteries (as measured during angiography)
7. aneurysms (10% patients);
   - 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
   - intra-arterial embolization or Gamma Knife intra-arterial embolization (AVMs that bleed often have intra-arterial 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!

**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

Surgical treatment risk is estimated by:

- Spezelt-Martin grading system — sum points from 5 factors:
  I. AVM size (largest diameter of nidus on angiography):
     a) 1 point - AVMs < 3 cm
     b) 2 points - AVMs 3-6 cm
     c) 3 points - AVMs > 6 cm.
  II. Elocuence of adjacent brain:
     a) 0 point — AVM is located in noneloquent area.
     b) 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:
     a) 0 point — if AVM has superficial venous drainage (all drainage is via cortical veins).
     b) 1 point — if AVM has component of deep venous drainage.

American Heart Association multidisciplinary MANAGEMENT GUIDELINES:

- Spezelt-Martin grade I-II → surgical extirpation.
- Spezelt-Martin grade III-IV → embolization, surgical extirpation.

Surgical treatment risk, embolization, radiosurgery

Spezelt-Martin grade IV-V (not amenable to surgical treatment alone - high procedural risk) → combination of embolization, radiosurgery and/or surgery.

B. Supplemented Spezelt-Martin Grading System (SM-Supp) — ABCs of AVMs: patient Age, Bleeding or hemorrhagic presentation, and AVM Compactness.
**TABLE 1. Comparison of the Spetzler-Martin and Supplementary Grading Systems**

<table>
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<th>Spetzler-Martin Grading</th>
<th>Points</th>
<th>Supplementary Grading</th>
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<tr>
<td>Size, cm</td>
<td>Age, y</td>
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<td>&lt; 3</td>
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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**

**Thrombogenic agents:**
1. quick-acting acrylate glue (M-butyl cyanoacrylate [NBCA]) – glue dilution speed 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
6. detachable balloons
   - try to occlude 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).

**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.
   - definitive surgery is performed within week of embolization (new feeding vessels and gliotic scar will make surgical procedures more difficult).
   - N.B. embolization prior to radiotherapy reduced rate of total obliteration after SRS.*
B. Palliative / partial 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 hypertension (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 Amytal may be injected → patient evaluated for alterations in EEG and neuro-visual picture.
- bolization of venous outflow leading to intracerebral hemorrhages.

- MORTALITY 9-22%; MORTALITY 0-9%.

**Vermian AVM embolization** (left vertebral angiogram):
A. Pre-embolization – vermian AVM supplied by vermian branches of SCA.
B. Microcatheter NBCA injection 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).
- electrophoretical 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 ICG* ANGIOGRAPHY) – helps to delineate arterial feeders and draining veins – can be safely isolated and ligated
- indocyanine green
- preserve “en passage” arteries!
- two strategies of approach:
  a) peripheral isolation of feeding arteries with marginal resection of nidus (trabecular en bloc method).
  b) central retrograde isolation of major draining vein as guide to nidus (merit of collapsing lesion from within).
Predictors of SRS failure

Positive predictors of obliteration

- cortical veins drain arterialized blood radically from lesion; major draining vein is often in center and is initially identified and localized (match with angiogram) → follow into nidus
- shunting venous and arteriolar calibers are coarcted and cut along central vein (venous radicals: 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. γ-therapy: 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 difficulties, you have to proceed with resection (cannot do only partial resection)
- giotic interface at periphery of nidus allows for plane of dissection to be developed - surgical excision with minimal deficits.
- 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 bleeding loss:
  1. controlled hypotension during surgery (maintain MAP 40-60 mmHg)
  2. embolization of feeding vessels prior to surgery.

Postoperative

- outcome correlates with score on Spetzler-Martini scale.
- AVM removal → progresses to progressive neurological improvement.
- surgical morbidity 8.6%, mortality 3.3%.
- complications:
  1. perfusion-breakthrough (Spetzler theory) - profuse edema and generalized hemorrhage from AVM bed (increased flow to previously underperfused vessels with lost autoregulation - weeks to months are required for brain to adapt to blood flow changes).
  2. postoperative embolization, staged resections for large high-flow AVMs, postoperative hypotension.
  3. postoperative bleeding / venous infarction due to occlusion of venous drainage (Yasargil theory).
  4. damage to adjacent neural tissue.

- routine postoperative 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
- nontargetive and can access all anatomic locations of brain (e.g. surgically inaccessible).
- ideal for lesions < 3 cm. AVM must be resected for AVM. Ideally small deep-seated, large lesions are separated into compartments and isolated and collapsed as separate units.
- single fraction tissue-destructive dose (16-25 Gy) is given.
- radiotherapy induces subendothelial collagen deposition → narrowed lumen of vessels → thrombosis over 1-3 years (risk of hemorrhage remains during this “latency period”!!)

Only 7.8% of AVMs obliterate within 18 mos post SRS:
- complications (clinical worsening attributable to SRS is seen in 3.8%)
- 1) white matter edema.
  N.B. if you see edema around VM, 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.
- 4) late effect - accelerated atherosclerosis in surrounding blood vessels.
- if aneurysm is identified in AVM selected for SRS, additional endovascular or surgical strategies could be considered to reduce risk of bleeding during latency interval).

- results: obliteration rate after SRS ≥ 65-69%, after multiple RS ≥ 75%.

Follow up MRI every 6 mos, when AVM is gone, confirmatory angiography

Positive predictors of obliteration:

1) higher marginal dose (odds ratio = 1.16).
2) compact nidus (odds ratio = 3.16)
3) undilated feeders (odds ratio = 0.36).
4) smaller AVM volume (odds ratio = 0.95).
5) SRS is treatment of choice for AVMs < 6 cm3, even after embolization!

Predictors of SRS failure:

1) prior bleed
2) prior embolization
- avoid AVM embolization if planning radiosurgery!

N.B. Martin scale
- 1 to 2: TemoAVM (Dr. Sheehan: SRS can be used for any AVM, ideally small deep-seated, > 3 cm AVMs must be treated in stages).
- 3) damage to adjacent neural tissue.

- routine postoperative angiography (AVM reappearance; years after negative postresection angiogram, have been reported)
- routine postoperative seizure prophylaxis (min. 3 months).

The Virginia Radiosurgery AVM Scale.

**RESIDUES**

- 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 uncontrolled but embolized portions of the AVM — follow-up angiography at 1 to 3 years appears to be warranted.

**PERINATAL 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 vs. 1.1% baseline annual risk — data from Brigham and Women's Hospital and Harvard Medical School*. 
  

- intracranial hemorrhage during pregnancy is due in AVM in 20-48% cases.

- once hemorrhage occurs during pregnancy there is 25% chance of recurrent hemorrhage.

**SPECIAL SITUATIONS**

**STRIATUM - POSTERIOR FOSSA**

- 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, transsylvian-transanular.

- approaches for THALAMUS — interhemispheric: transcallosal or trans-sphenial.

- 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 rebleed 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. transventricular route for lesions around tectum and superior cerebellum).

- BRAINSTEM AVMs: microsurgical resection, radiosurgery, combined subtemporal-suboccipital-retro labyrinthine-transventricular approach.

**HEMORRHAGE**

- risk of immediate rebleeding is relatively low - treatment of AVM is delayed (4 to 6 weeks) to allow time for:
  1) hemorraghe 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 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)

- 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.
INTRACRANIAL VASCULAR MALFORMATIONS

Vas30

Not associated with enlarged feeding arteries or draining veins.
- Blood flow is low or even stagnant.
- Intrallesional thrombosis, calcification and recanalization are typical.
- Range from soft to hard.
- 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.

CLAIRINAL 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; 40% 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* (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 angiomas 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
- High-sensitivity 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:

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 patients - 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

A. Left parietal hyperdensity:

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 Cavernous Malformations: Surgical Results in 104 Patients and a Proposed Grading System to Predict Neurological Outcome* Neurosurgery: March 2015 - Volume 76 - Issue 3 - p 265–278

CAPILLARY TELANGIECTASIA

- microscopic nests of dilated capillary vessels (saccular or fusiform dilations) with normal brain tissue in between.
- related pathologically to CAVERNOUS ANGIOMAS (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).
- no evidence of associated hemorrhage.
- grossly - tiny lesions having appearance of punctate hemorrhages.
- usually deep within brain (particularly in brain stem).

The only feature that differentiates CAPILLARY TELANGIECTASIA from CAVERNOUS ANGIOMAS is presence of brain parenchyma between vascular channels

PATHOLOGY

- microscopic nests of dilated capillary vessels (saccular or fusiform dilations) with normal brain tissue in between.
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- usually deep within brain (particularly in brain stem).

TREATMENT

Indications:

1) intractable (!) seizures; some experts think that 3 seizures under adequate AED is enough to warrant surgery (due to risk of kindling with repeated seizures)
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

Modalities

A) surgical removal - additional resection of perilesional hemosiderin deposits and gliosis provides higher rate of seizure control vs. pure lesionectomy
- N.B. cavernoma itself is not epileptogenic but adjacent hemosiderin-laden cortex is!

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

C) 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

- no acute hemorrhage following Visualase fiber placement.
- 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.
- reimaging (6-21 months) indicated lesion diminution with surrounding liquefactive necrosis.

CAPILLARY TELANGIECTASIA

- prevalence ≈ 0.4%
- may accompany Sturge-Weber syndrome or Rendu-Osler disease.
INTRACRANIAL VASCULAR MALFORMATIONS

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).
  - 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
- 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.

CT/MRI (left frontal venous angioma):

Bilateral cerebellar venous angiomata 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 that 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 pathophysiologically 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.

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 (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);

- 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

CLASSIFICATIONS

Cognard classification
Type I: confined to sinus wall with normal anterograde flow
Type IIA: confined to sinus with reflux (retrograde) into sinus but not cortical veins
Type IIB: drains into sinus with reflux (retrograde) into cortical veins (10-20% hemorrhage)
Type II A+B: drains into sinus with reflux (retrograde) into both sinus 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 sinus. Do not bleed!
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>
<thead>
<tr>
<th>Borden</th>
<th>Bleeding rates</th>
<th>Bleeding rate if has venous ectasia</th>
<th>Rebleeding rates after hemorrhage</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>1%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>II</td>
<td>6%</td>
<td>40%</td>
<td>21% and 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-IIID) 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 DVAFs 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.

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

B. Vascular compression – initial treatment for lesion in TRANSVERSE or SIGMOID sinuses - manual compression of opthalmic vein (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) PTA particles - low morbidity rate but high recanalization chances. b) liquid materials - high cure rates but high morbidity rate (great risk of stroke). 1) Onyx-18 (ethylenevinylalcohol copolymer) 2) NBCA (N-butyryl-cyanoacrylate) 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: a) transarterial - supraselective catheterization of arterial feeding vessels that can be occluded, but the fistula itself is rarely obliterated: 
  - 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: 
  - very effective adj 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 sinus dural fistulas: 
  - feasible only if venous phase angiography has documented the absence of venous drainage of normal brain by the involved sinus. 
  - impossible in Border 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 sinus, when not used by brain, together with pathologic dura (if brain uses the sinus, the sinus is skeletonized and left patent). b) direct surgical exposure, catheterization, and packing of the involved sinus 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 (indocyanine green) angiography 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 patient until the bleeding stops generally works well. Suzuki procedure – resecting transverse sinus due to multiple DAVFs. E. Radiosurgery: disadvantage – delayed fistula closure (not suitable for aggressive DAVFs with annual bleeding risk up to 15%).

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) - aneurysmal enlargement of vein of Galen is only secondary manifestation of AVM!
INTRACRANIAL VASCULAR MALFORMATIONS

Type I malformation:

Type II malformation - multiple fistulae between choroidals 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).

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

ICA arteriogram (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.

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

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

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

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

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 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**

- Gianturco coil
- Short angiography catheter
- Detachable balloon
- Microcoil
- 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).
- Prophylactic 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 PROTRUSION** - 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) - defects within both inner and outer tables (arrows) of calvaria and filling of diploë 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