Intracranial Vascular Malformations

Last updated: January 16, 2021

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CLASSIFICATION according to predominant *VASCULATURE*:

1) **developmental venous anomaly (DVA), s. venous angioma** (*most common type*!) - anomalous *VEINS* without any direct feeding artery.
2) **capillary telangiectasia*** (*next most common type*!) - vessels morphologically resembling *CAPILLARIES* but slightly larger.
3) **arteriovenous malformation (AVM)**** (less common but *most clinically important type*!) - clusters of abnormal *ARTERIES* and *VEINS* without intervening capillaries.
4) **cavernous angioma (cavernoma)***
5) **direct (s. arteriovenous) fistula**** - acquired 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 **DVAs** 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 **NEUROIMAGING:** − not visible, (+) sometimes visible, + visible

Type	CT	MRI	Angio
AVM	+	+	+
Dural AV fistula	(+)	(+)	+
DVA	(+)	+	+
Telangiectasia	−	(+)	−

Type	CT	MRI	Angio
Cavernoma	(+)	+	–

ARTERIOVENOUS MALFORMATIONS (AVM)

PATHOLOGY, PATHOPHYSIOLOGY

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

- lesion is ***present from birth*** (vs. congenital aneurysms!).
- vascular conglomerate (numerous thin-walled, tortuous channels) is called **NIDUS**.
  - nidus commonly forms "pyramid / conus" 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. *or via caverns

Arteriovenous shunt is definitive characteristic!

- feeding *ARTERIES* may run serpentine course through sulci before entering nidus;
  - high flow subsequently leads to ARTERIAL DILATION
  - fibromuscular cushions - smooth muscle hyperplasia associated with fibroblasts and connective tissue elements
  - **arterial feeders**:
    a) entirely from **ICA branches** (*PURELY PIAL MALFORMATIONS*): MCA territory > ACA territory > PCA territory; occasionally (≈ 10%) recruit additional supply from ***meningeal arteries***.
    b) entirely from **ECA branches** (*PURELY DURAL MALFORMATIONS*).
  - **arterial structure is damaged** - duplication and fragmentation of internal elastic lamina, marked thickening or partial replacement of media by hyalinized connective tissue.
- draining *VEINS* often are dilated with thickened walls (due to high velocity blood flow through fistulae) - VENOUS ARTERIALIZATION;
  - AVMs of ACA and PCA may drain directly into vein of Galen, causing it to dilate in aneurysmal fashion. *see below (Vein of Galen Malformation) >>*

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

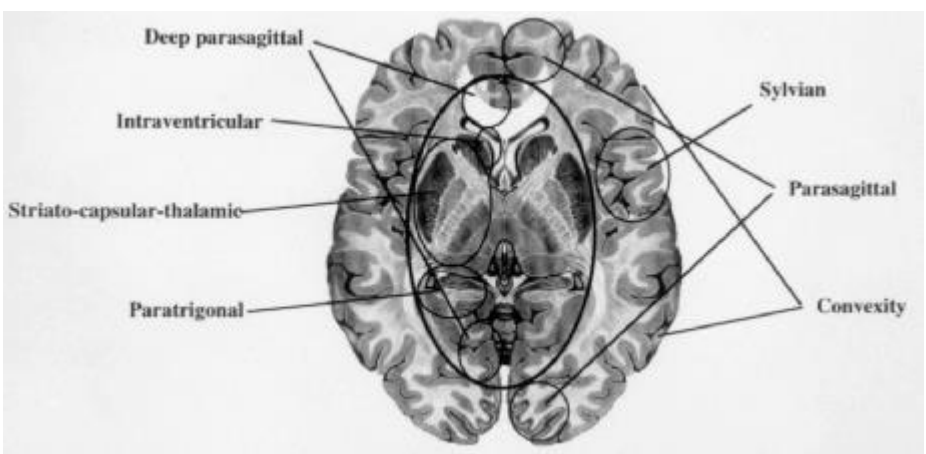
- involved ***vessels may enlarge*** with passage of time; some AVMs* decrease over time (spontaneous thrombosis?) up to total resolution (rarely).

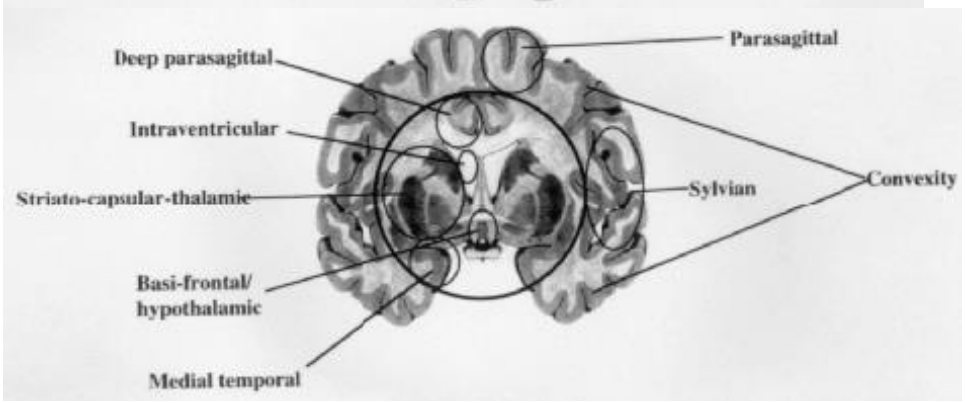
*esp. those around anterior fossa and chiasm

Microscopy: **entrapped** (between vessels) **brain tissue** is gliotic 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:





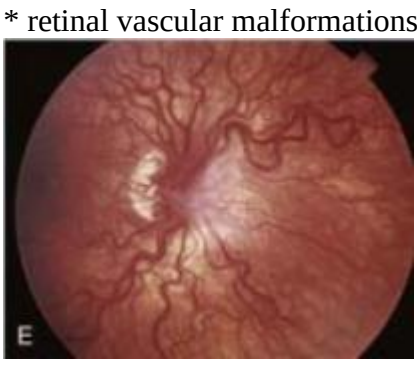
- AVMs occur in ***all parts of neuraxis*** (largest AVMs are most frequent in posterior half of hemispheres).
  - typically lie superficially (within brain substance or cerebral sulci).
  - wedge-shaped (apex directed toward ventricle).

- 2.3-16.7% 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 (s. Bonnet-Dechaume-Blanc 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

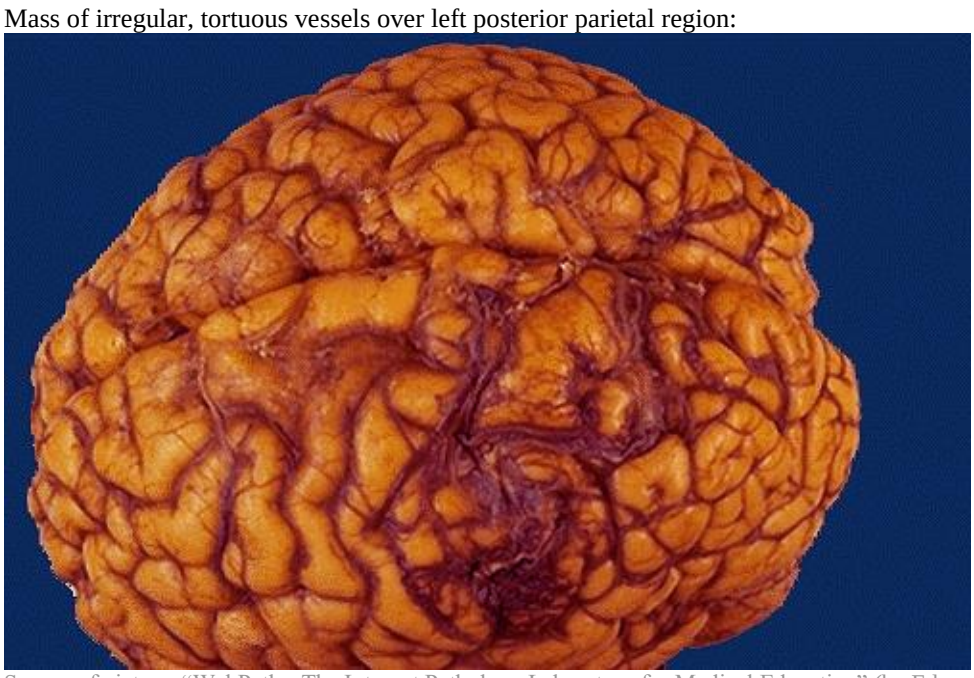
- flow shunted through AVM is *extremely pressure-dependent* (*no autoregulation*) and follows conditions described by **HAGEN-POISEUILLE equation** where flow (Q) is directly related to pressure difference (ΔP) and fourth power of radius (r) and is inversely related to tube length (L) and viscosity (n):

$$Q = \frac{\Delta P + \sim r^4}{8 \cdot L \cdot n}$$

- **bulk flow rates** (vary according to size and anatomy) are 150-900 ml/min (≈ 490 ml/min).

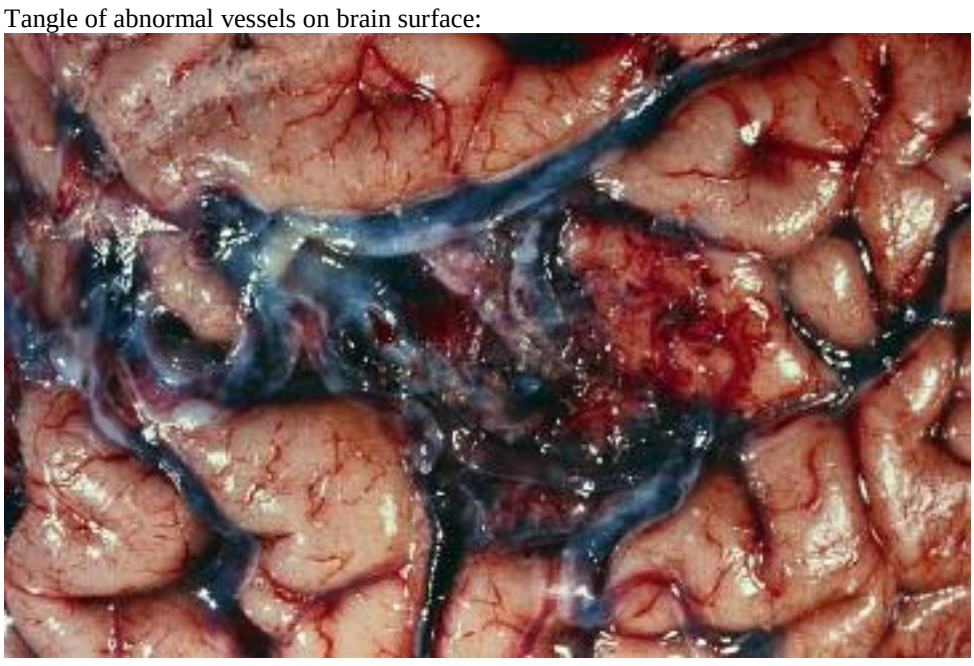
  Low resistance = High flow!
- AVM feeders have low intravascular pressure, high flow velocity, low peripheral stream resistance, and *very poor vasomotor reactivity* (e.g. relatively nonreactive to PCO_2 changes).
- **tissues adjacent to AVM** may be persistently *mildly hypoxic* (malformation may steal blood from adjacent healthy tissue).
- *as AVM is resected*, pressure within feeding arteries rises by ≈ 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:

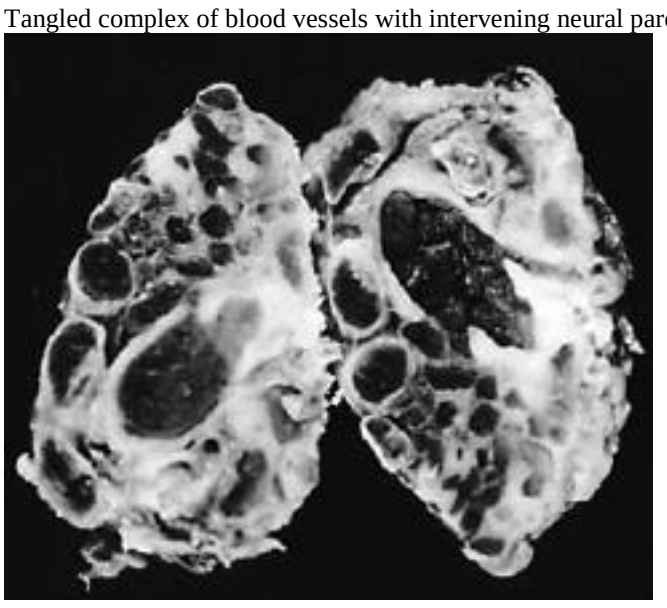


Source of picture: "WebPath - The Internet Pathology Laboratory for Medical Education" (by Edward C. Klatt, MD) >>

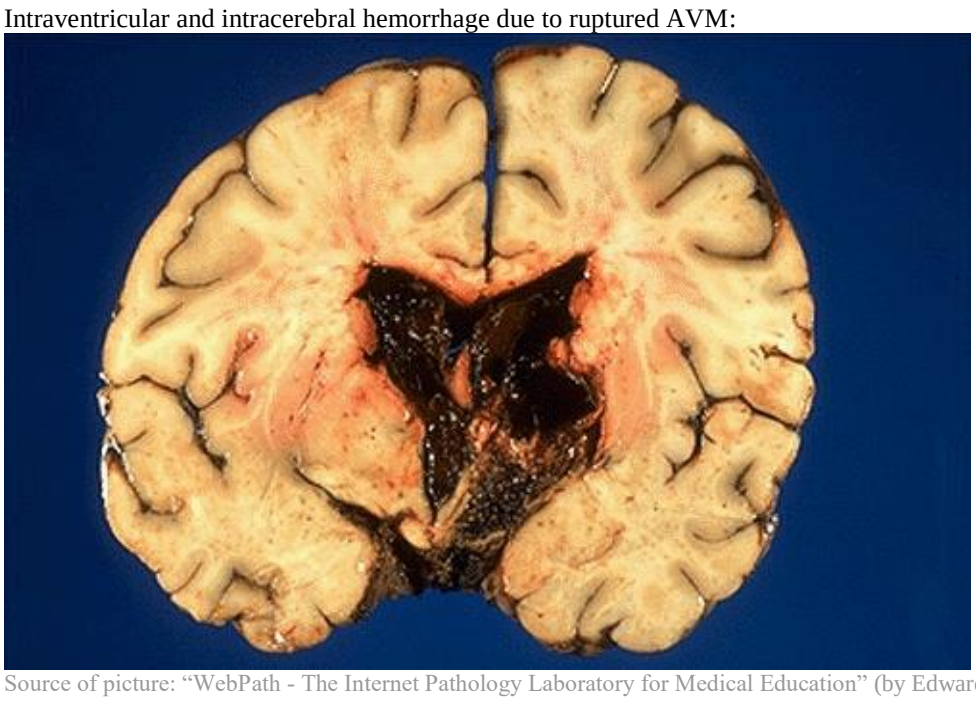
Tangle of abnormal vessels on brain surface:



Tangled complex of blood vessels with intervening neural parenchyma:



Intraventricular and intracerebral hemorrhage due to ruptured AVM:



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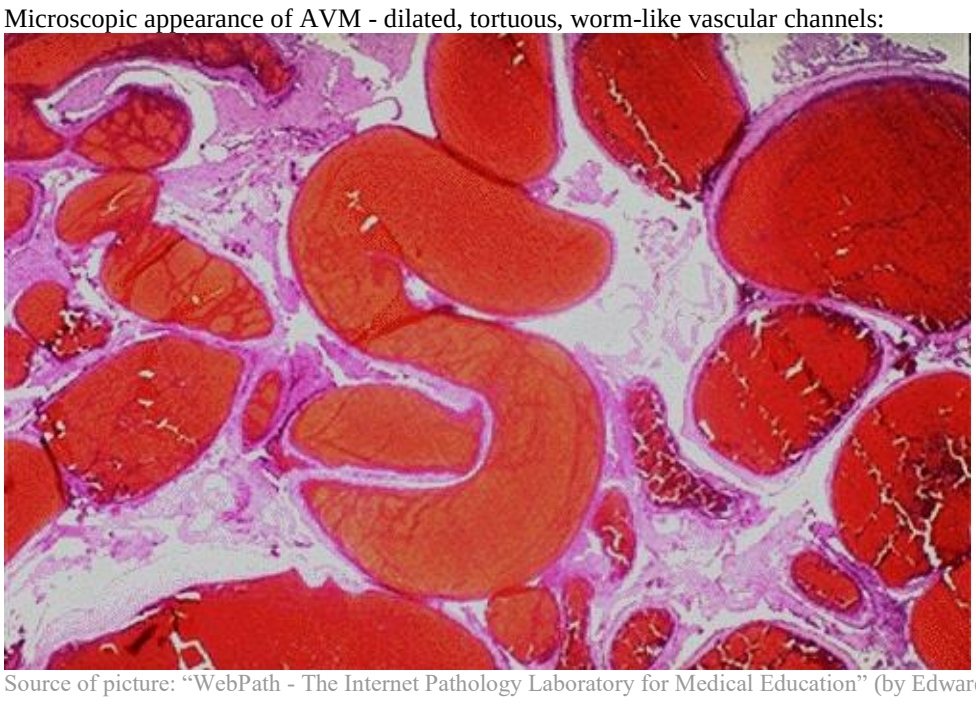
Microscopic appearance of AVM - dilated, tortuous, worm-like vascular channels:



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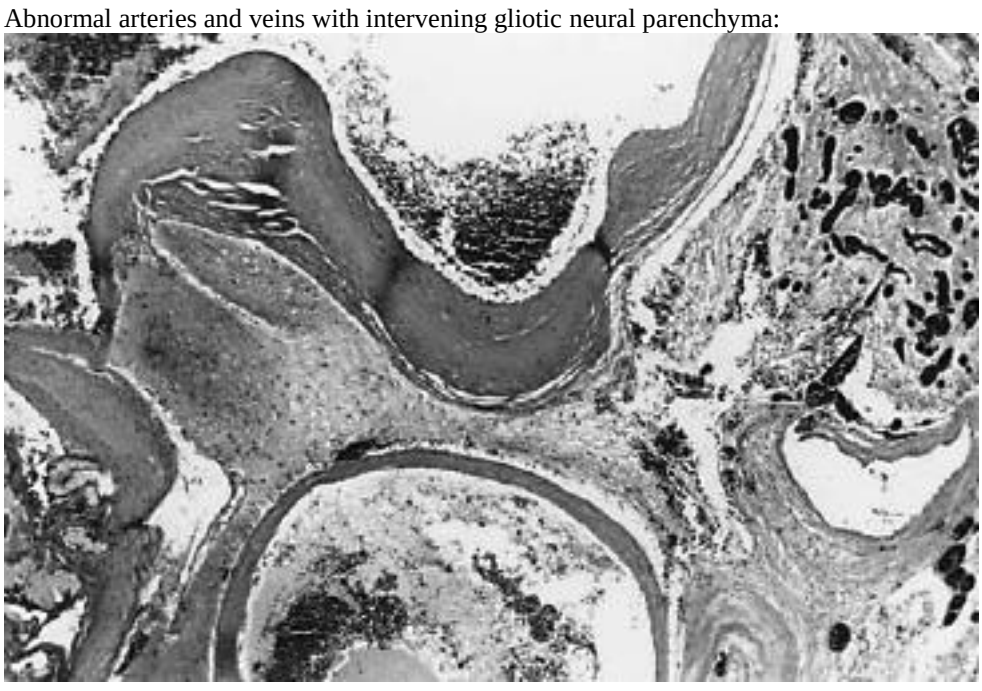
Abnormal arteries and veins with intervening gliotic neural parenchyma:



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 (CAMSs)

        CAMS-1 - prosencephalic AVMs affecting hypothalamus/hypophysis in association with facial AVM of nose.
        CAMS-2 - AVMs affecting lateral prosencephalon (occipital lobe, thalamus) in association with facial AVMs of maxilla.
        CAMS-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 ¼ patients present by age of 40)

- ***bruits*** (≈ 50%) may be audible either to patient or to examiner.
- scalp or face ***veins may be enlarged***.
- huge 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
B. Seizures
C. Progressive neurological deficit
D. Headache

A. Hemorrhage (presenting symptom in 38-70% patients) into:

a) **brain parenchyma** - most commonly!
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).

Mortality with each bleed is ≈ 15%

- 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) lobar 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.

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

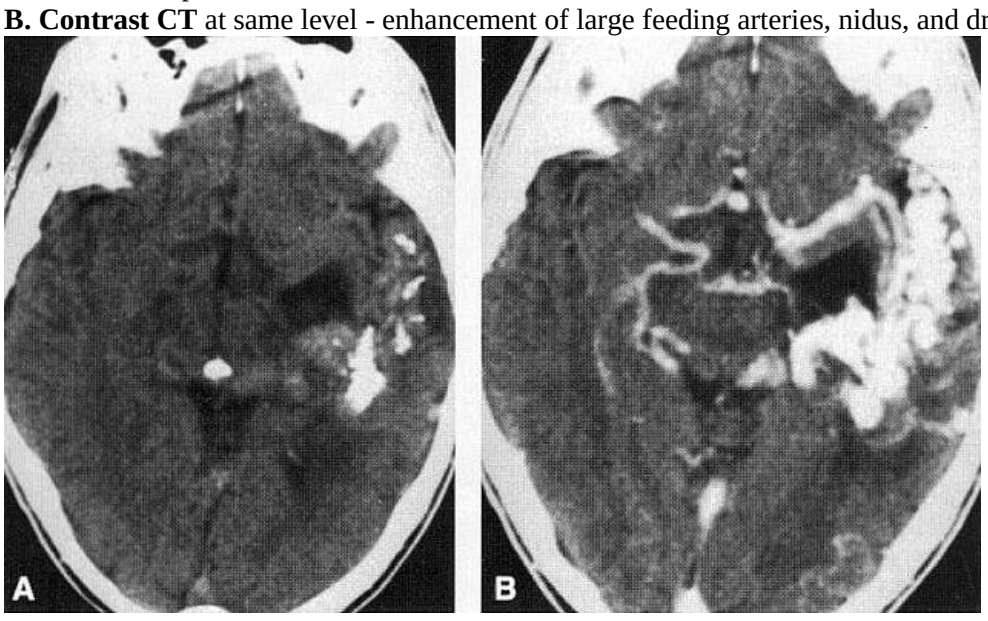
- AVM calcifications, increased vascular markings in overlying bone (calvarial vascular grooves and foramina).

CT

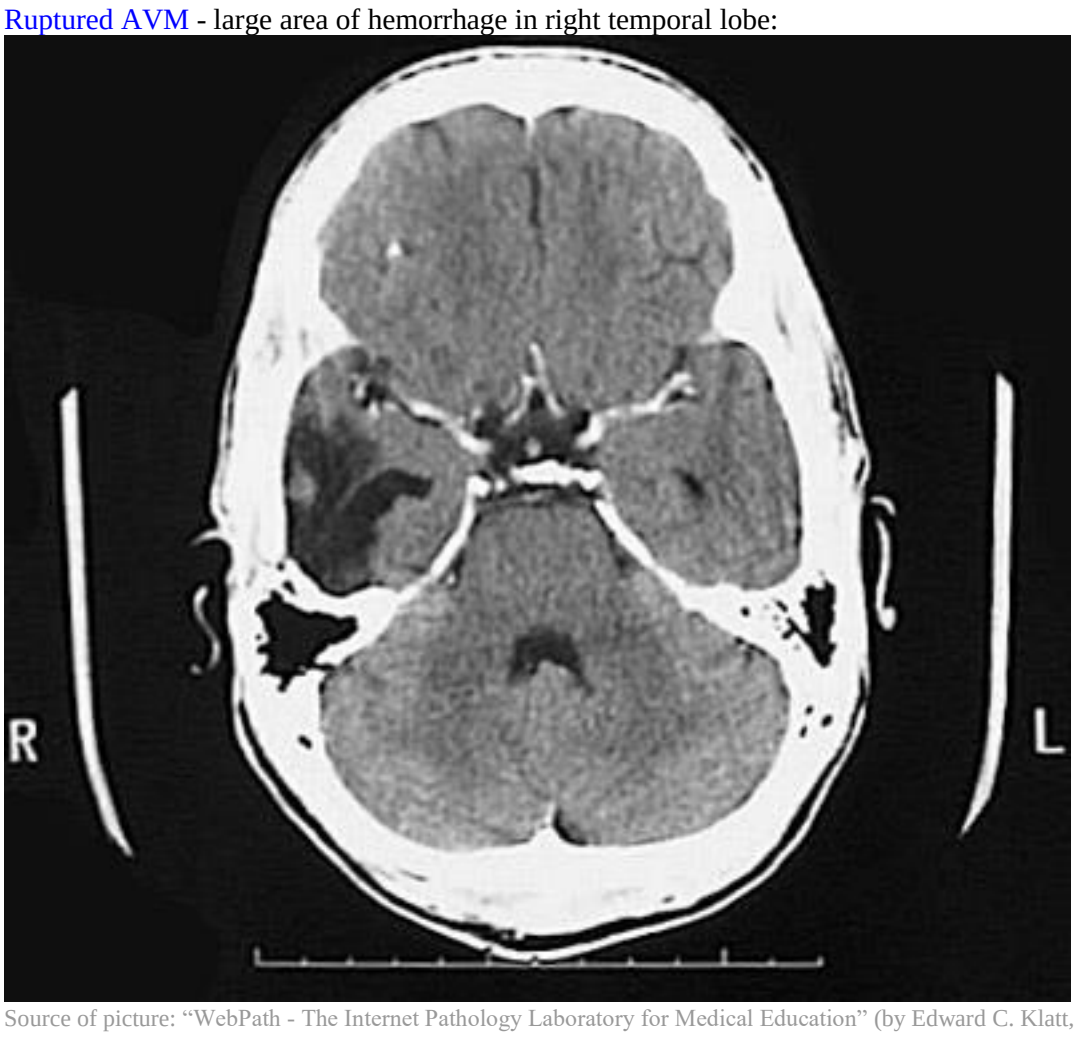
- can identify only large AVMs - serpiginous areas of high density.
- **contrast-enhanced CT** - striking enhancement* (classic pattern - irregular hyperdense central area from which extend multiple, well-defined serpentine structures of various sizes - dilated feeding arteries and draining veins).

  *due to increased blood pool within lesion + BBB impairment in adjacent neural parenchyma
- AVMs may be *surrounded by hypodense areas* of ***ischemic damage***.
- CT may show ***calcification***.

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



Ruptured AVM - large area of hemorrhage in right temporal lobe:



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)

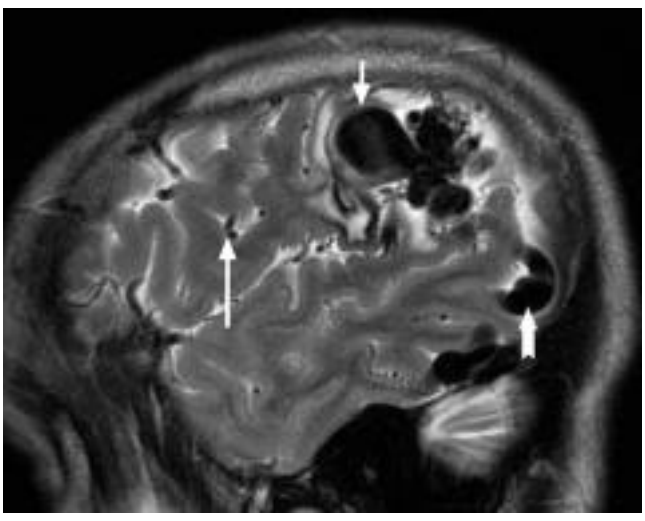
N.B. gadolinium does not facilitate detectability (vs. CT contrast).

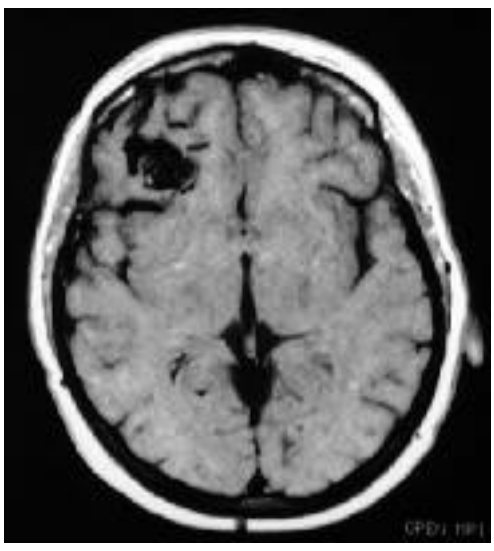
- irregular serpiginous or globoid masses with mixed signal anywhere within hemispheres or brain stem.
- large arteries and draining veins are particularly characteristic feature - shown as signal void rather than flow-related enhancement.

  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*.

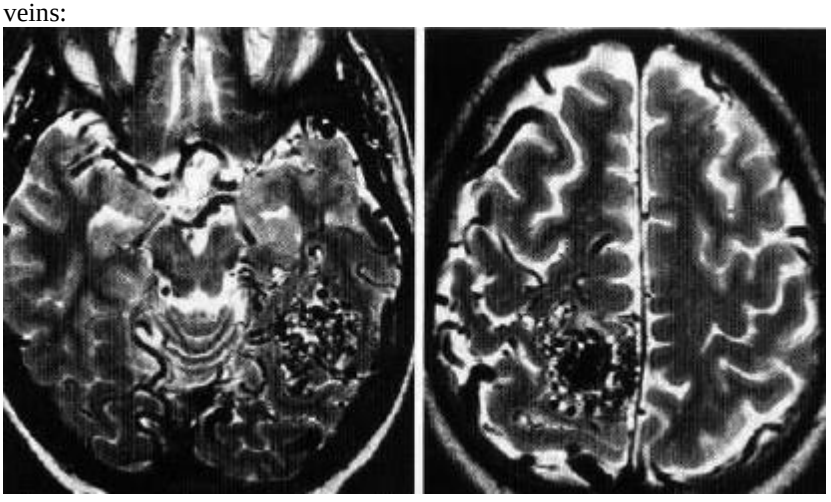
T2-MRI - parietal AVM; varices (*short arrow*), dilated arteries (*long arrow*) and draining veins (*notched arrow*):

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

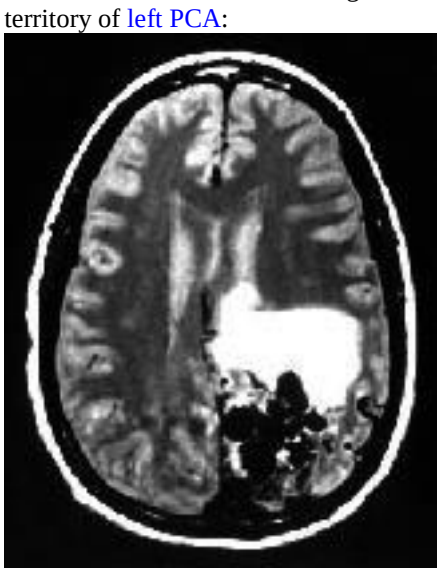




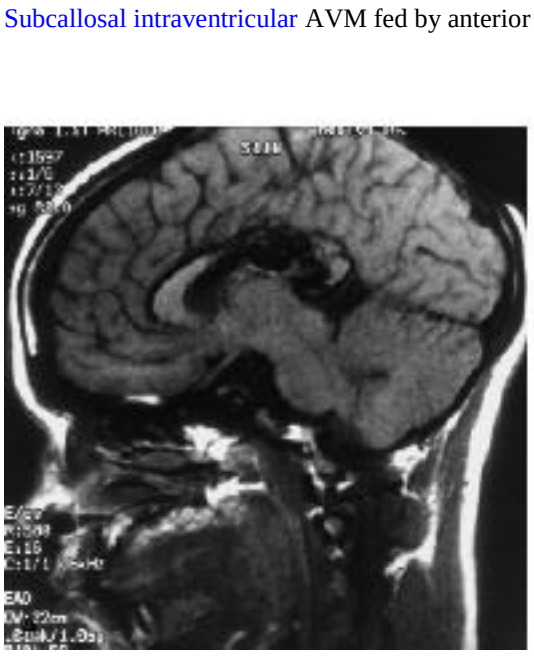
T2-MRI - extensive bilateral AVMs; multiple enlarged superficial drainage veins:

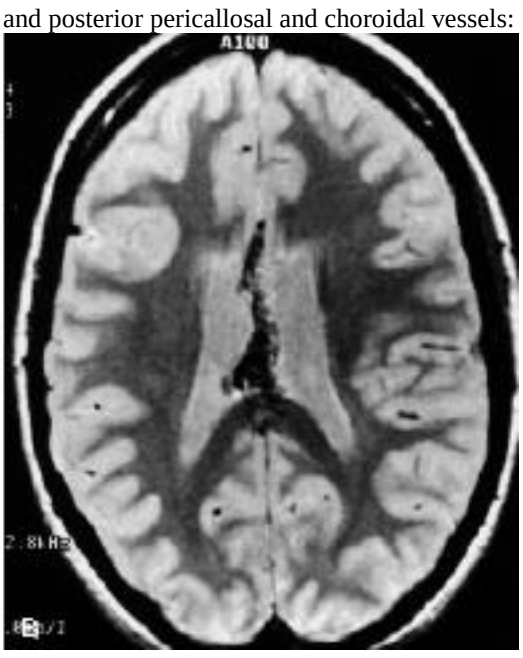


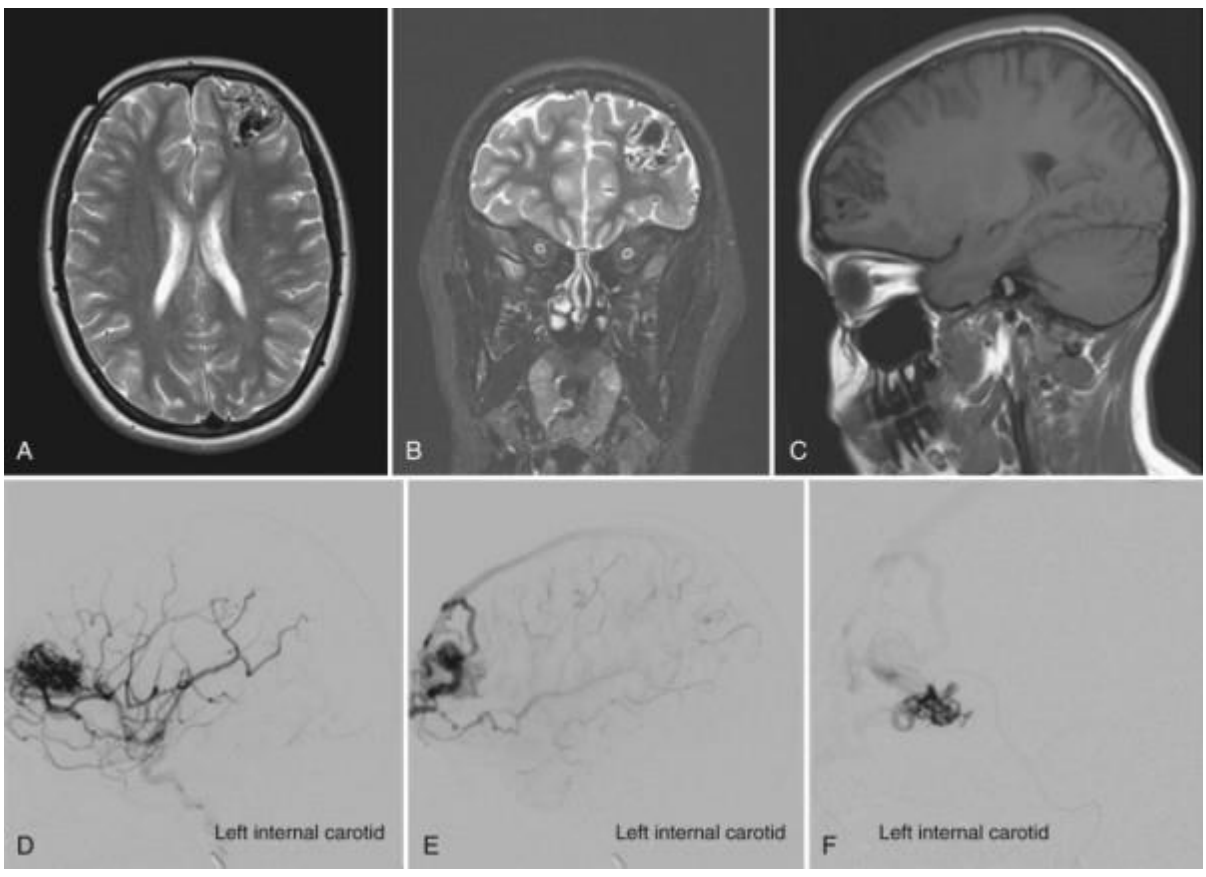
T2-MRI - AVM with hemorrhage in territory of left PCA:



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





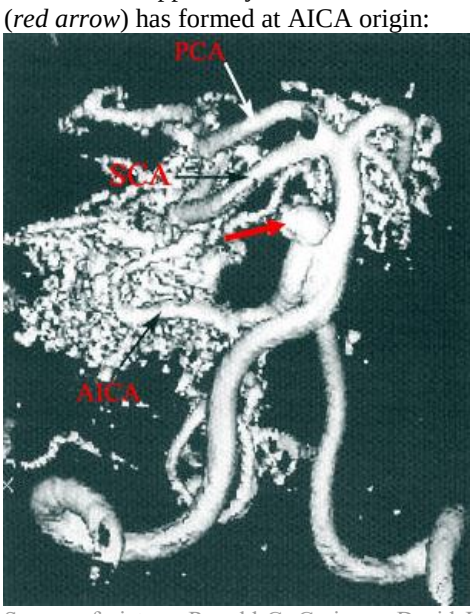


MRA

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

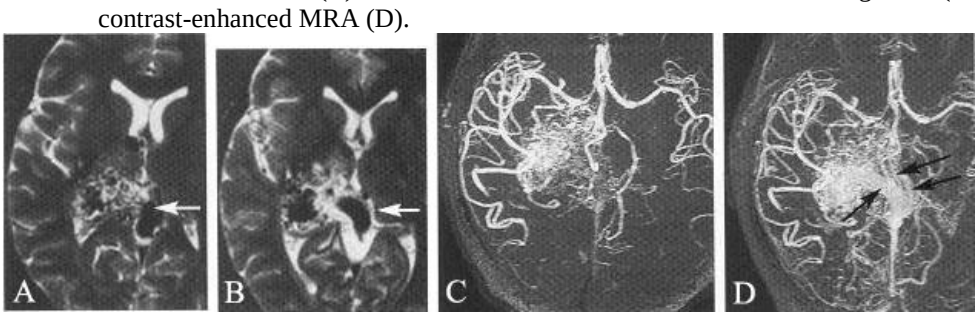
3D TOF MRA - hugely dilated left MCA feeders (*long arrow*), nidus (*short arrow*), varices (*arrowhead*) and superficial draining vein (*open arrow*):

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:

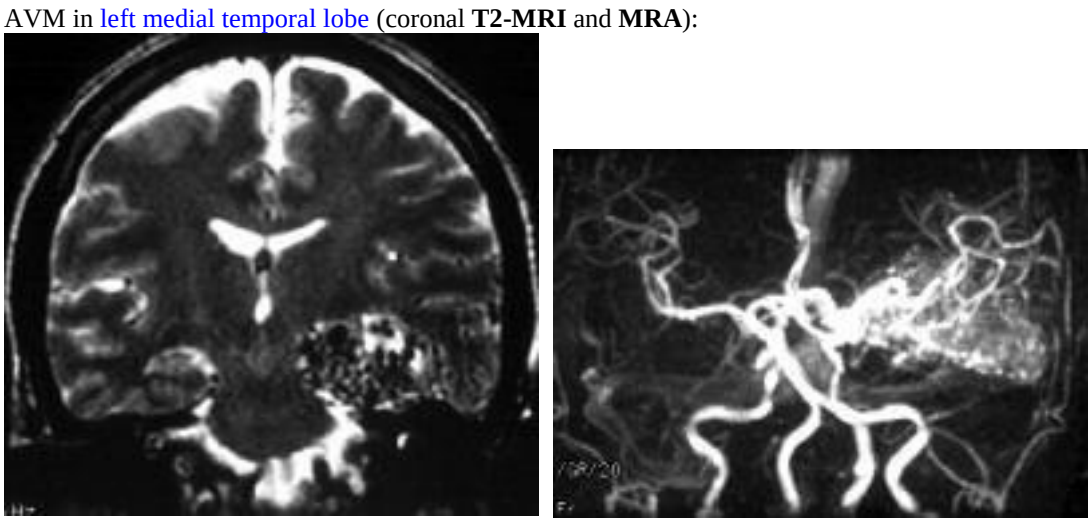


Source of picture: Ronald G. Grainger, David J. Allison "Grainger & Allison's Diagnostic Radiology: A Textbook of Medical Imaging", 4th ed. (2001); Churchill Livingstone, Inc.; ISBN-13: 978-0443064326 >>

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).



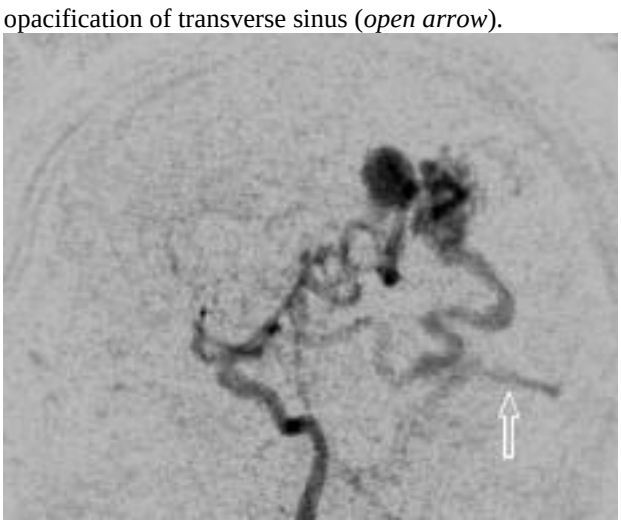
AVM in left medial temporal lobe (coronal **T2-MRI** and **MRA**):



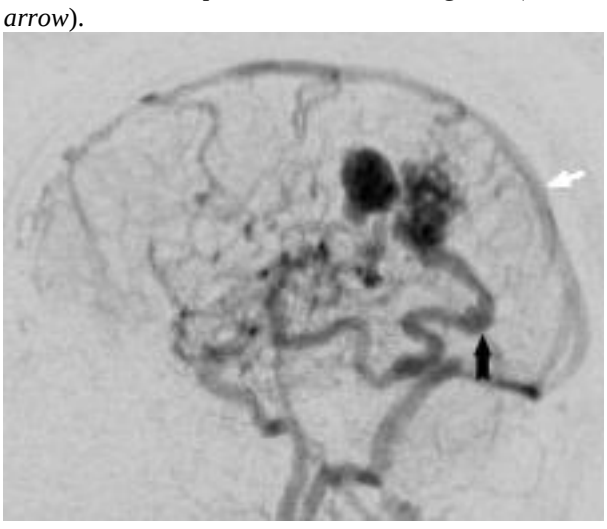
MR DSA (series of three frames in lateral projection acquired at 1-sec intervals during IV infusion of gadolinium):

1) feeding MCA branches (*long arrows*), nidus (*short arrow*), varices (*arrowhead*) and large superficial draining vein (*open arrow*) all apparent on first frame, indicating speed of shunting.

2) opacification of transverse sinus (*open arrow*).



3) opacification of superior sagittal sinus (*white arrow*); note small venous pouch on main draining vein (*black arrow*).



ANGIOGRAPHY

- required for DEFINITIVE diagnosis & TREATMENT planning (AVM morphology determines treatment algorithm!).

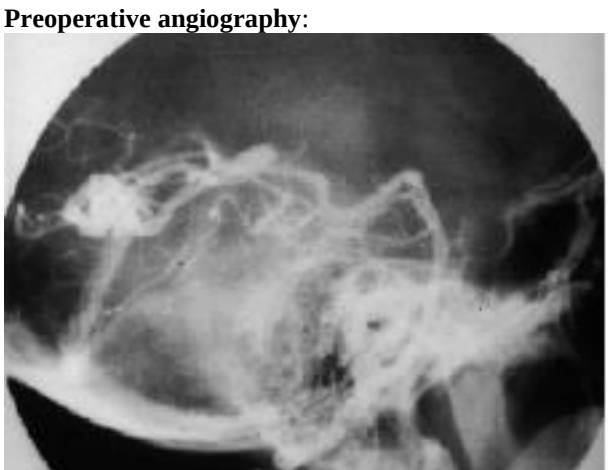
- dilated tortuous *feeding **arteries***; central *tangle of vessels*; rapid* *arterial-to-venous shunting* (early opacification of enlarged, tortuous *draining **veins***).
  **rapid sequence (high frame rate) filming* is essential otherwise (feeding vessels can be obscured by overlying veins in rapidly shunting lesions)!
- **four-vessel angiography** is indicated:
  - up to 10% AVMs are associated with saccular **aneurysms**.
  - extracranial or contralateral arteries occasionally supply intracranial AVMs.
- in case of hemorrhage, *hematoma may compress AVM* so completely that angiography cannot detect it (when AVM is suspected, angiography is best postponed until hematoma has resolved - after 6-8 weeks).
- aneurysms within nidus often show slow washout.

SUPERSELECTIVE angiography into AVM **feeding arteries**:

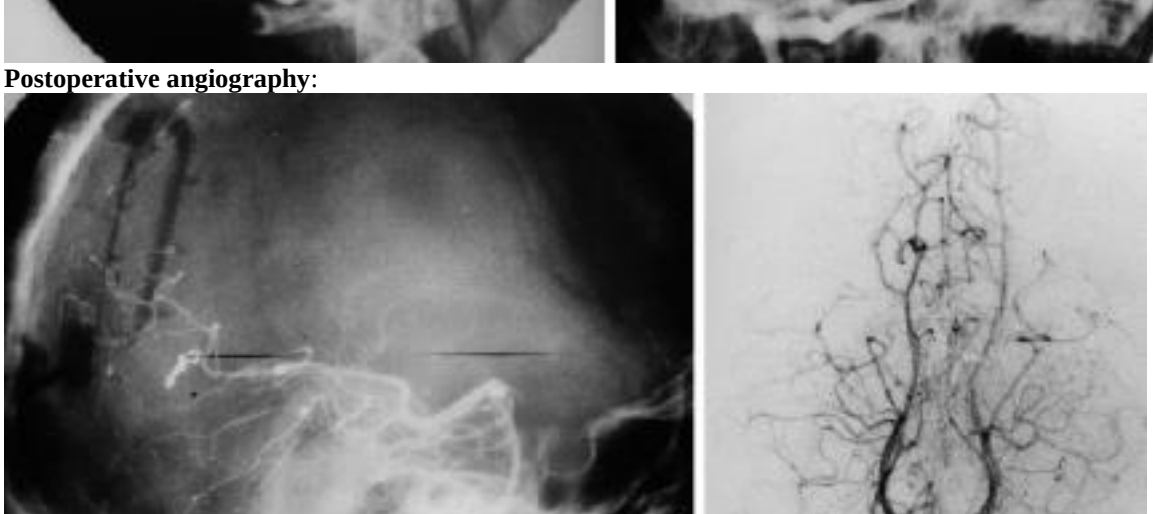
1) obtain ***pressure measurements*** (higher feeding pressures increase hemorrhage risk).
2) inject SODIUM AMYTAL (anesthetic agent) to produce temporary anesthesia of area perfused by artery - "***superselective Wada testing***" - language, memory, visual-spatial, sensory, motor function can be tested 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:

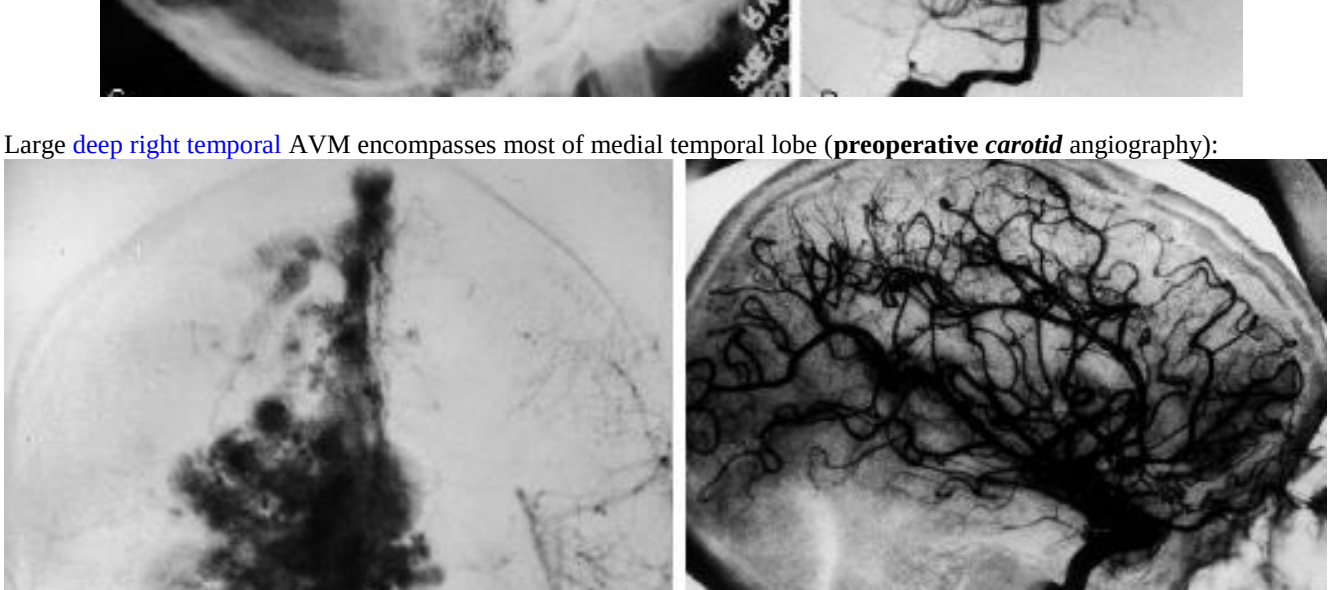
Preoperative angiography:



Postoperative angiography:

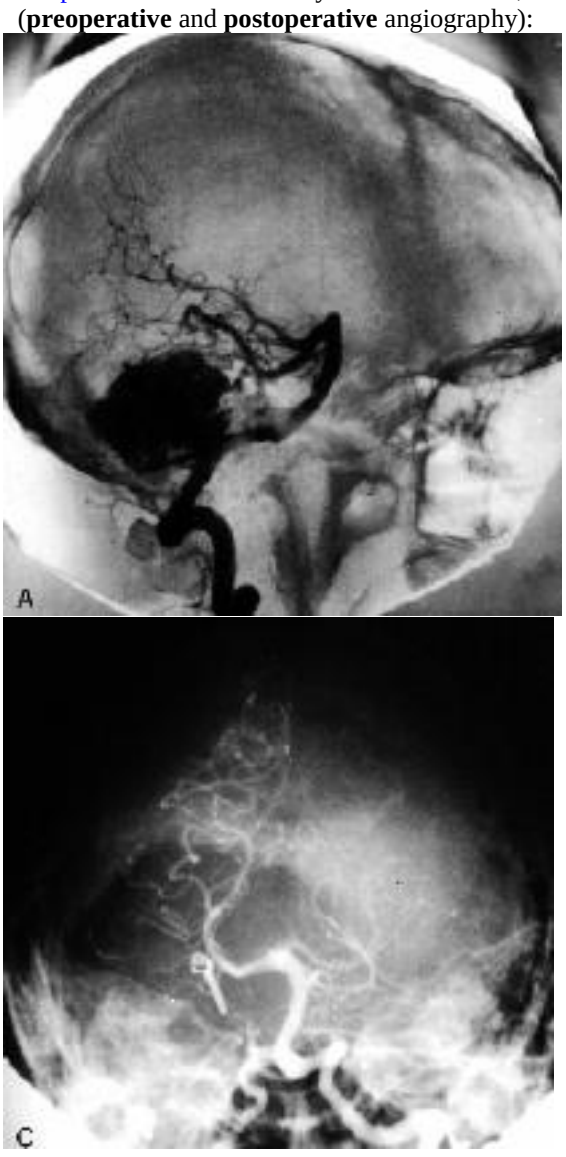


Large deep right temporal AVM encompasses most of medial temporal lobe (**preoperative *carotid*** angiography):

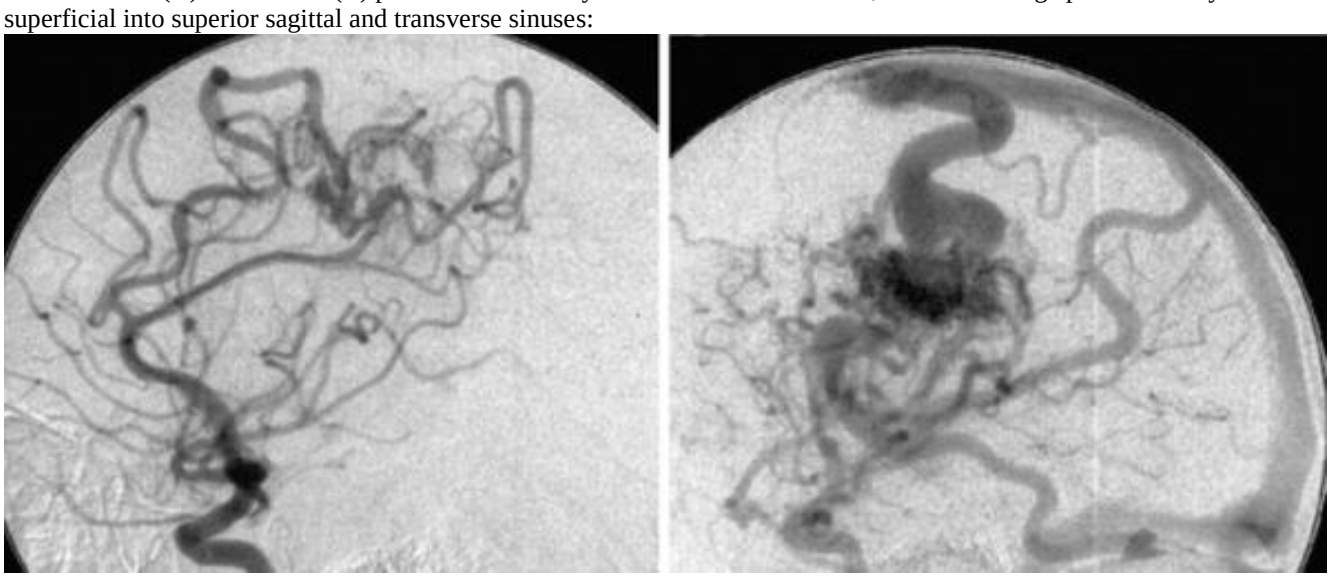


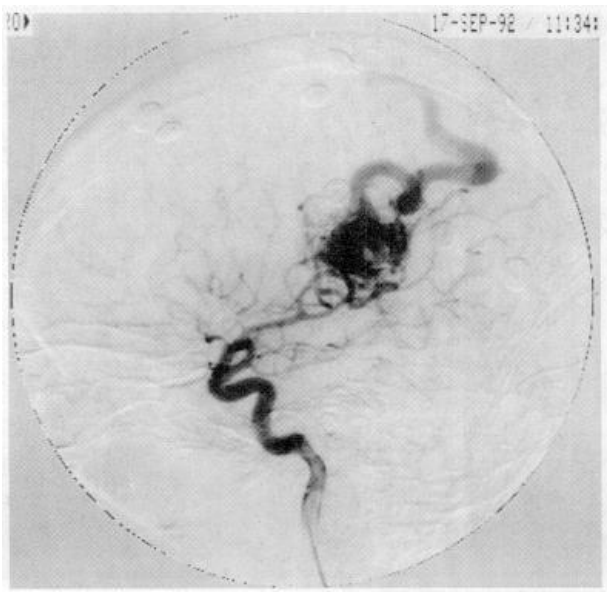
Angiogram (AP view) - AVM (3 cm in diameter) in deep MCA territory with deep draining vein (*arrow*):

Deep cerebellar AVM fed by branches of SCA, AICA, and PICA (**preoperative** and **postoperative** angiography):

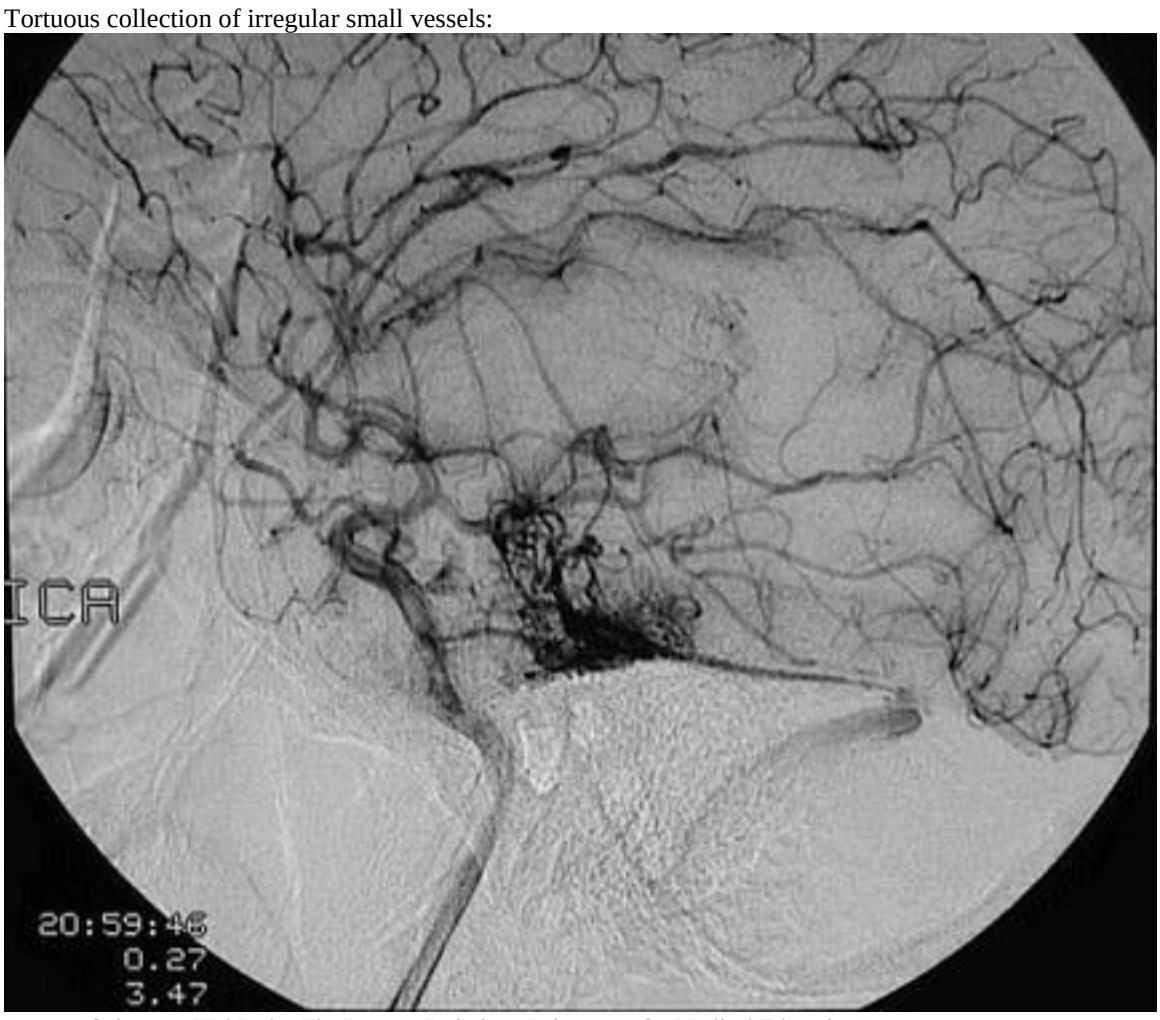


DSA - arterial (A) and venous (B) phase - AVM fed by ACA and MCA branches; venous drainage predominantly superficial into superior sagittal and transverse sinuses:



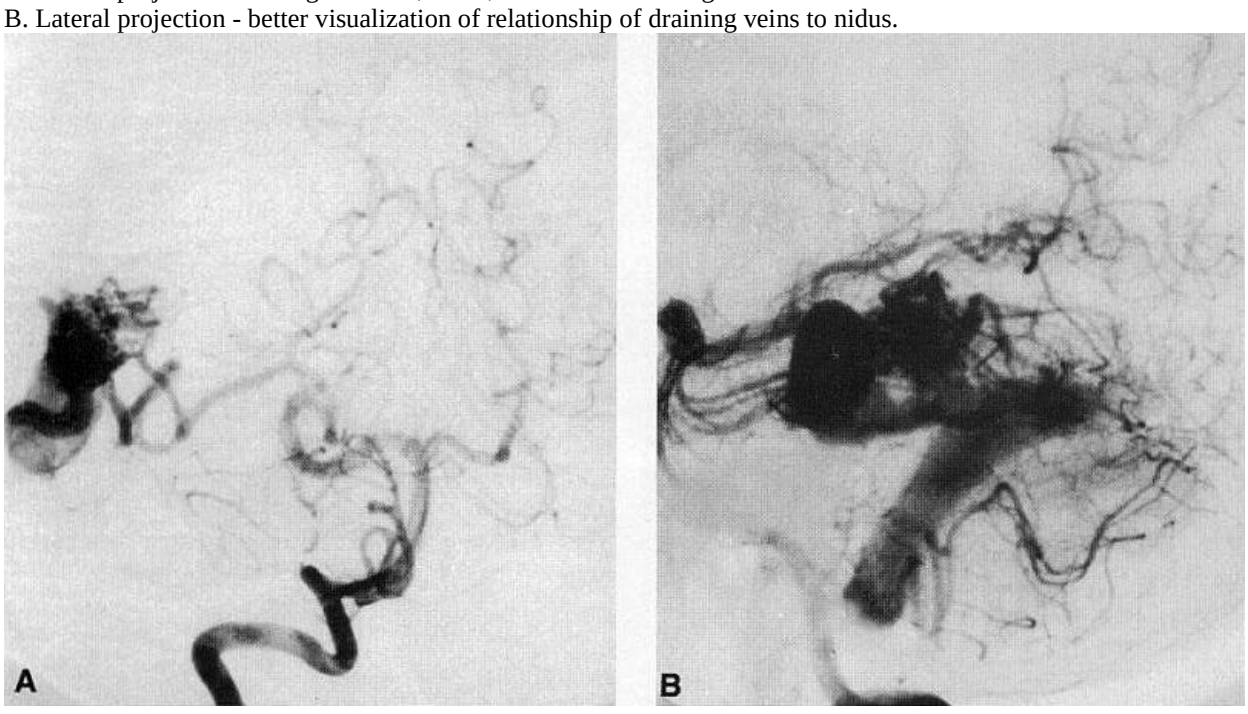


Tortuous collection of irregular small vessels:



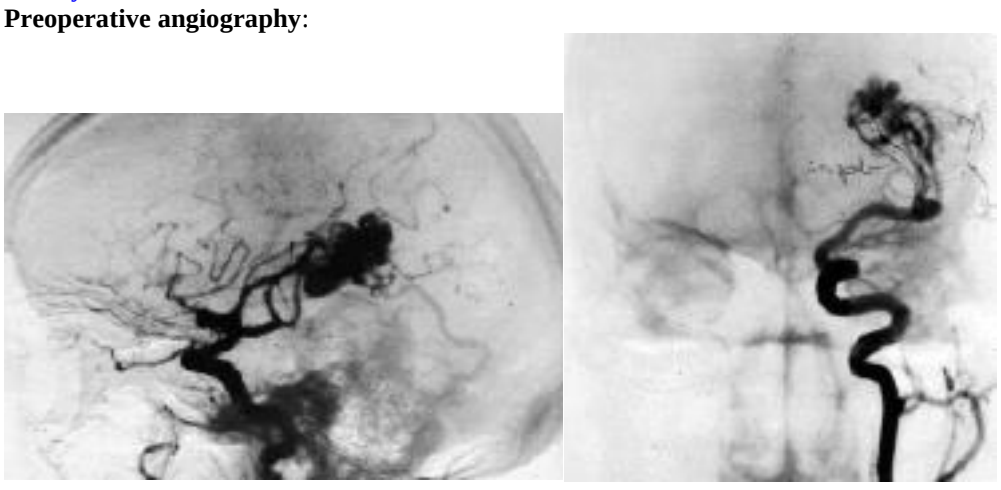
Source of picture: "WebPath - The Internet Pathology Laboratory for Medical Education" (by Edward C. Klatt, MD) >>

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.

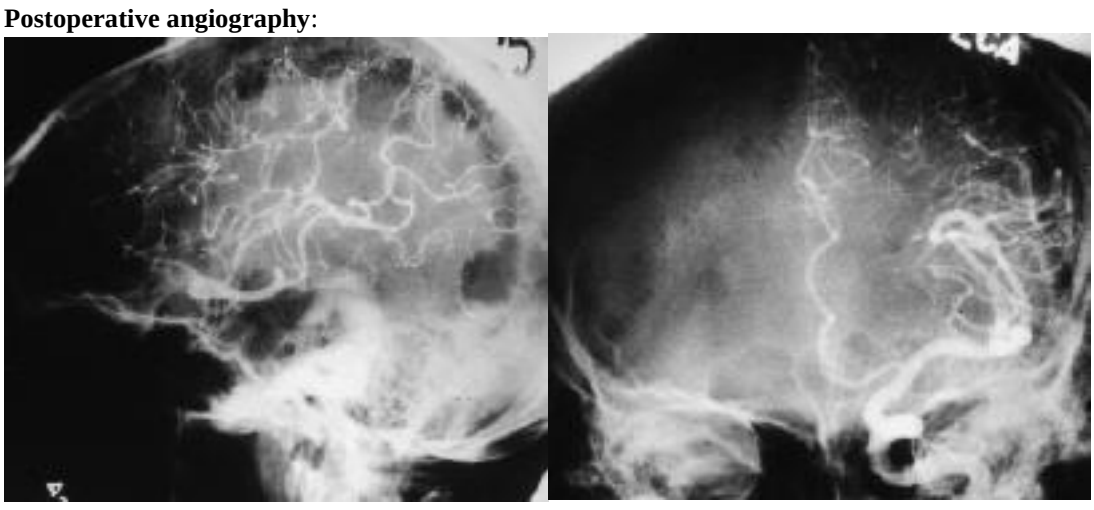


Right parasplenial AVM (**preoperative** and **postoperative angiography**):

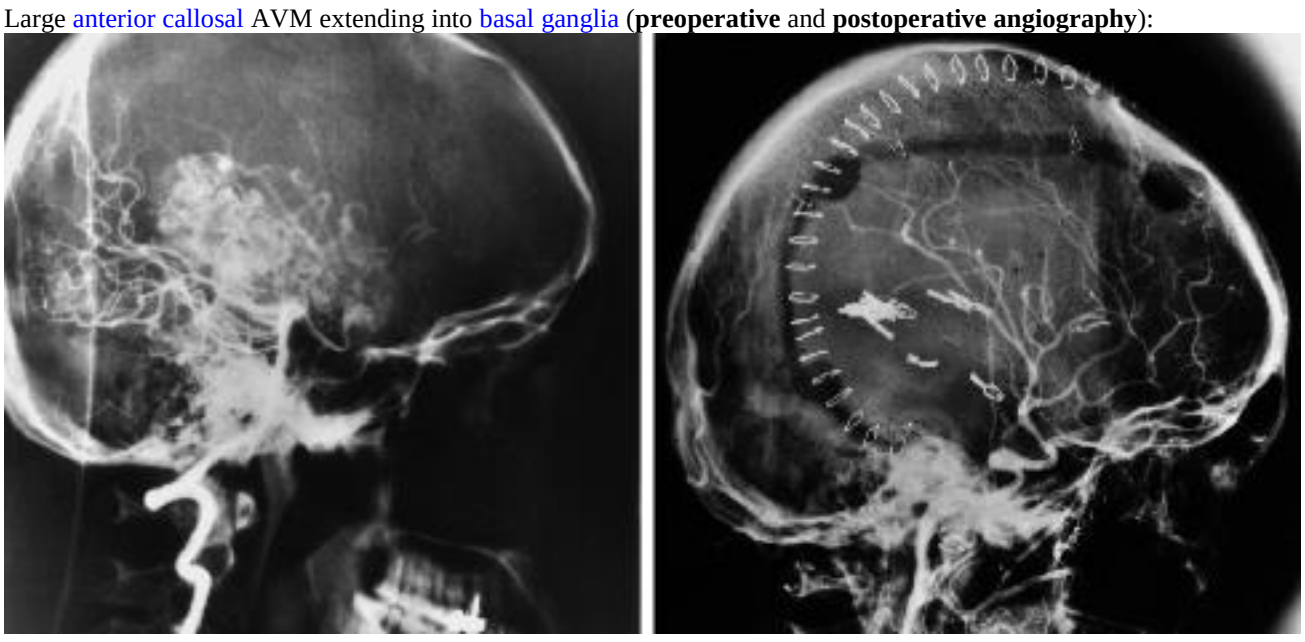
Left posterior sylvian AVM:
Preoperative angiography:



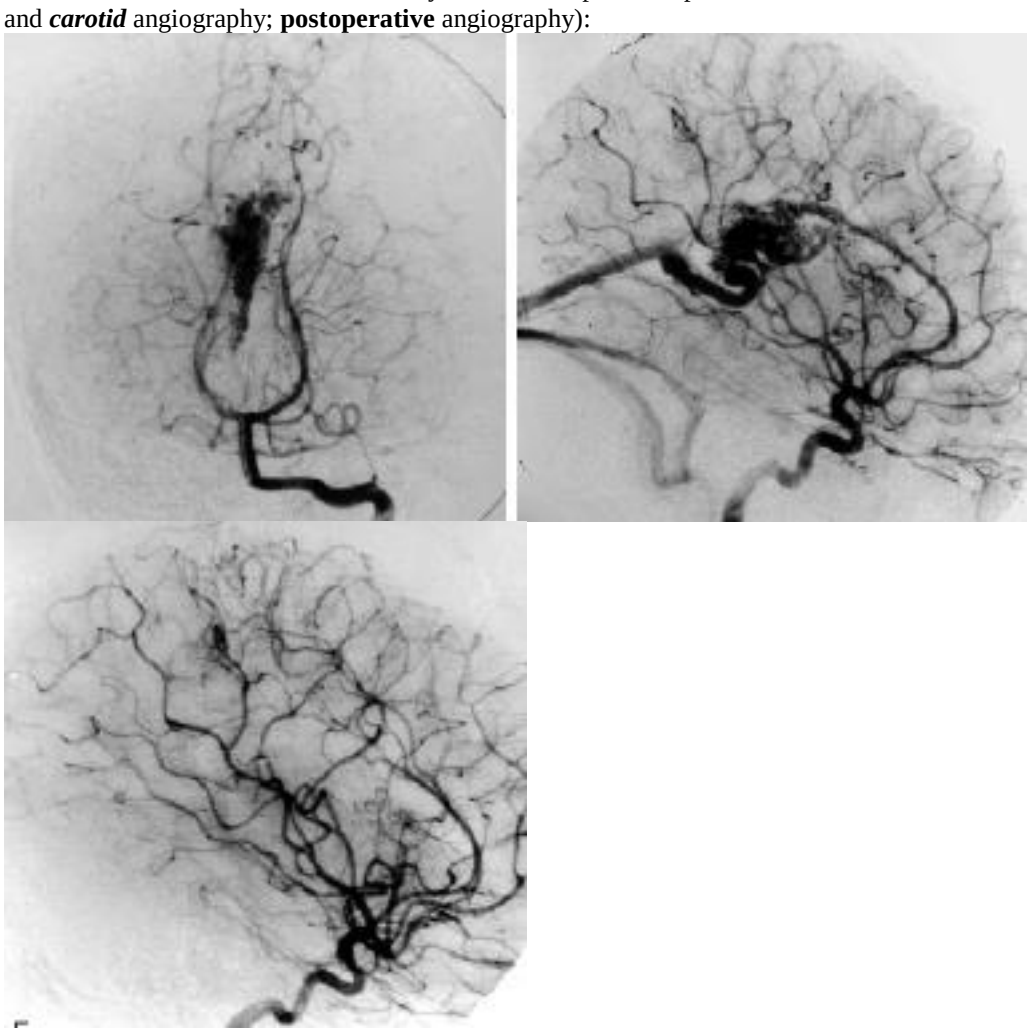
Postoperative angiography:



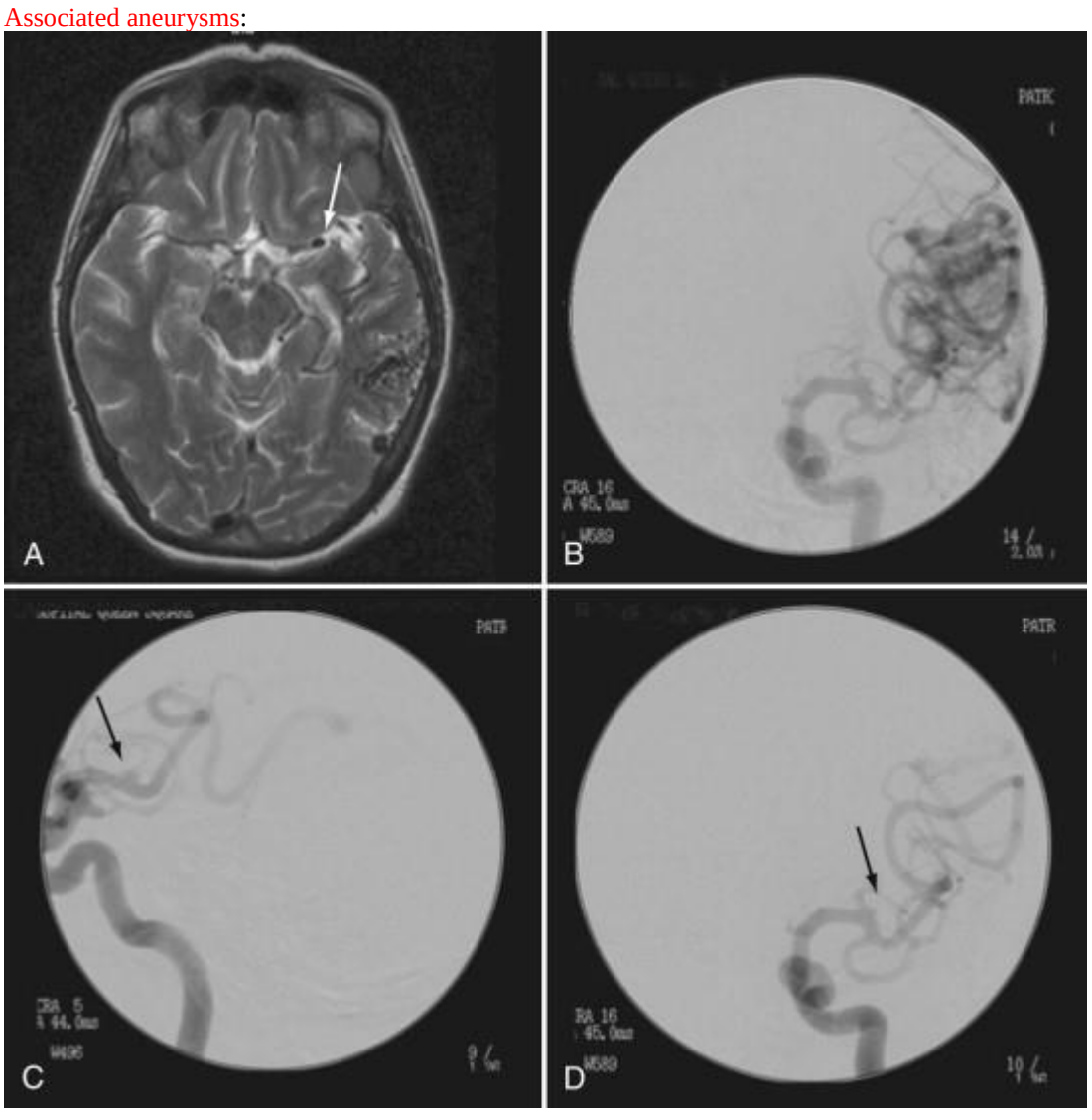
Large anterior callosal AVM extending into basal ganglia (**preoperative** and **postoperative angiography**):



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



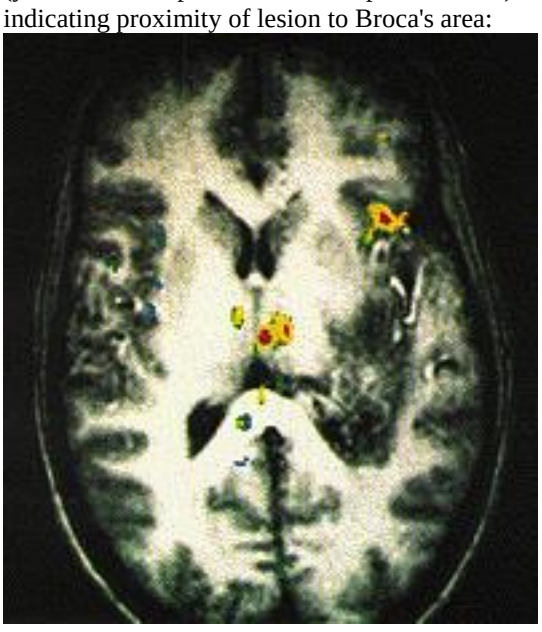
Associated aneurysms:



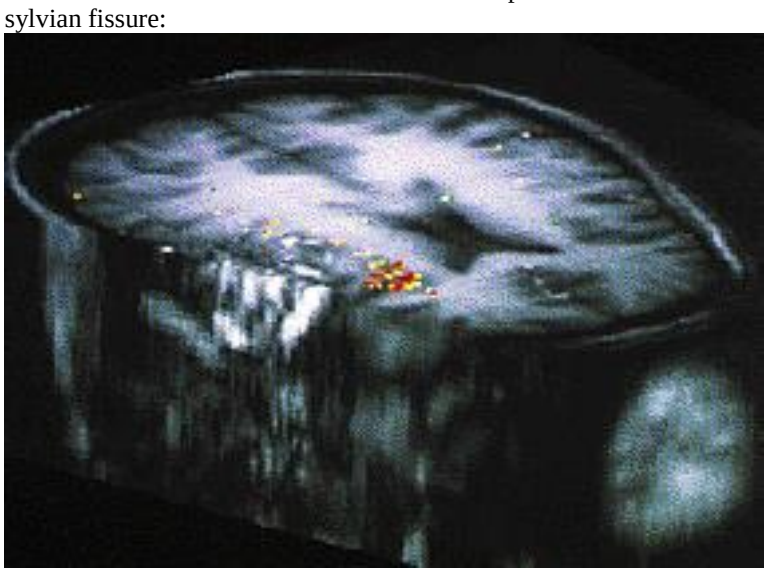
FUNCTIONAL MRI

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

fMRI - blood flow changes during silent speech (yellow and red pixels in frontal opercular area), indicating proximity of lesion to Broca's area:



3D MR reconstruction - relation of lesion to speech areas and sylvian fissure:



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 angiomas 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 (!) 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

     Giuseppe D'Aliberti "Intra-nidal and Flow Related Aneurysms in AVMs Pose Higher Risk of Bleeding Than Standard Saccular Aneurysms" World Neurosurgery - 2015 Feb

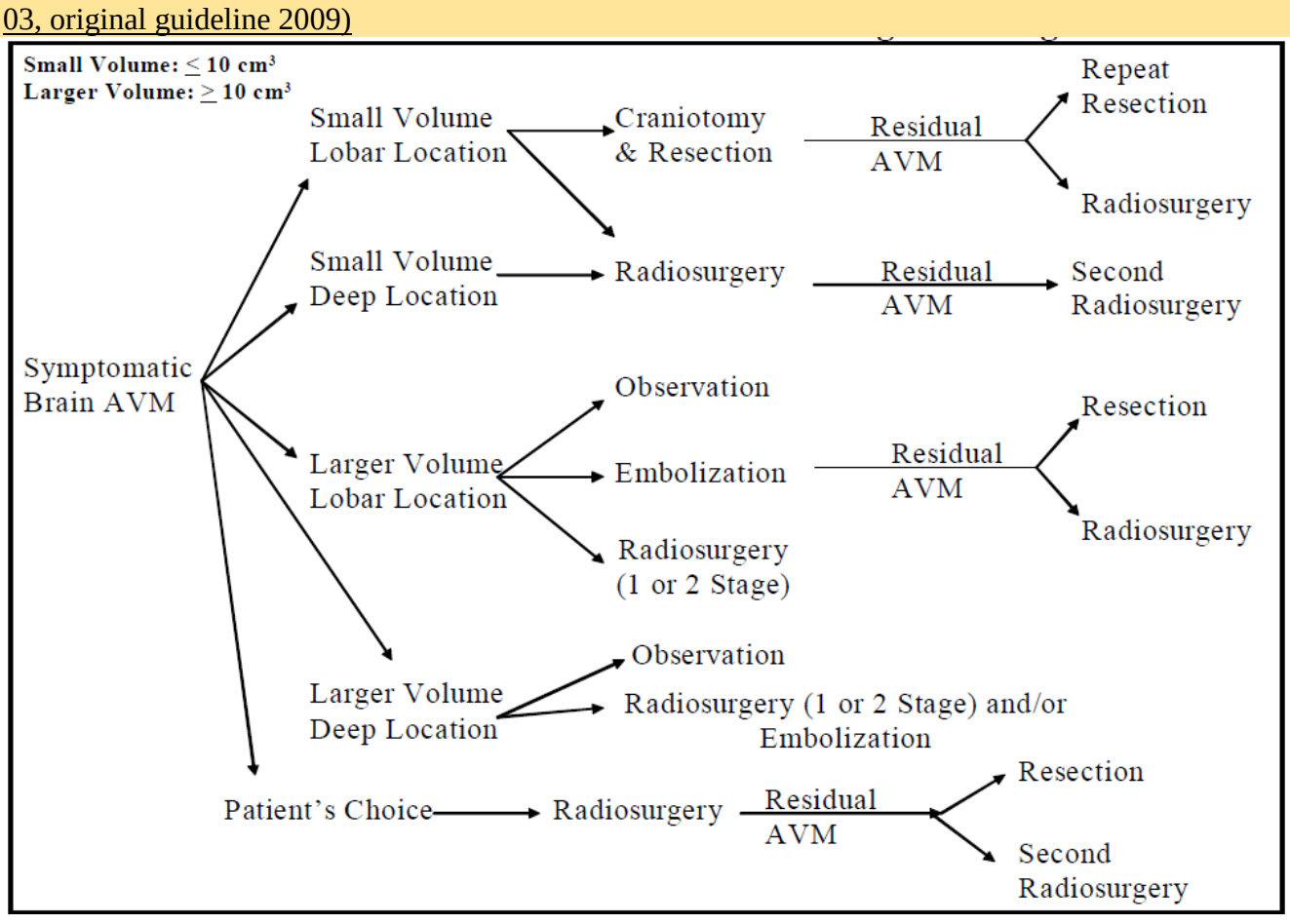
     AVMs that bleed often have intra-nidal aneurysms!!!
8) prior hemorrhage (rebleeding risk during 1st year 7-33%*, then 2.5% annually)
9) pregnancy *see below*

* i.e. much lower rate than in aneurysmal bleedings

Bleeding risk is not influenced by age!

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

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

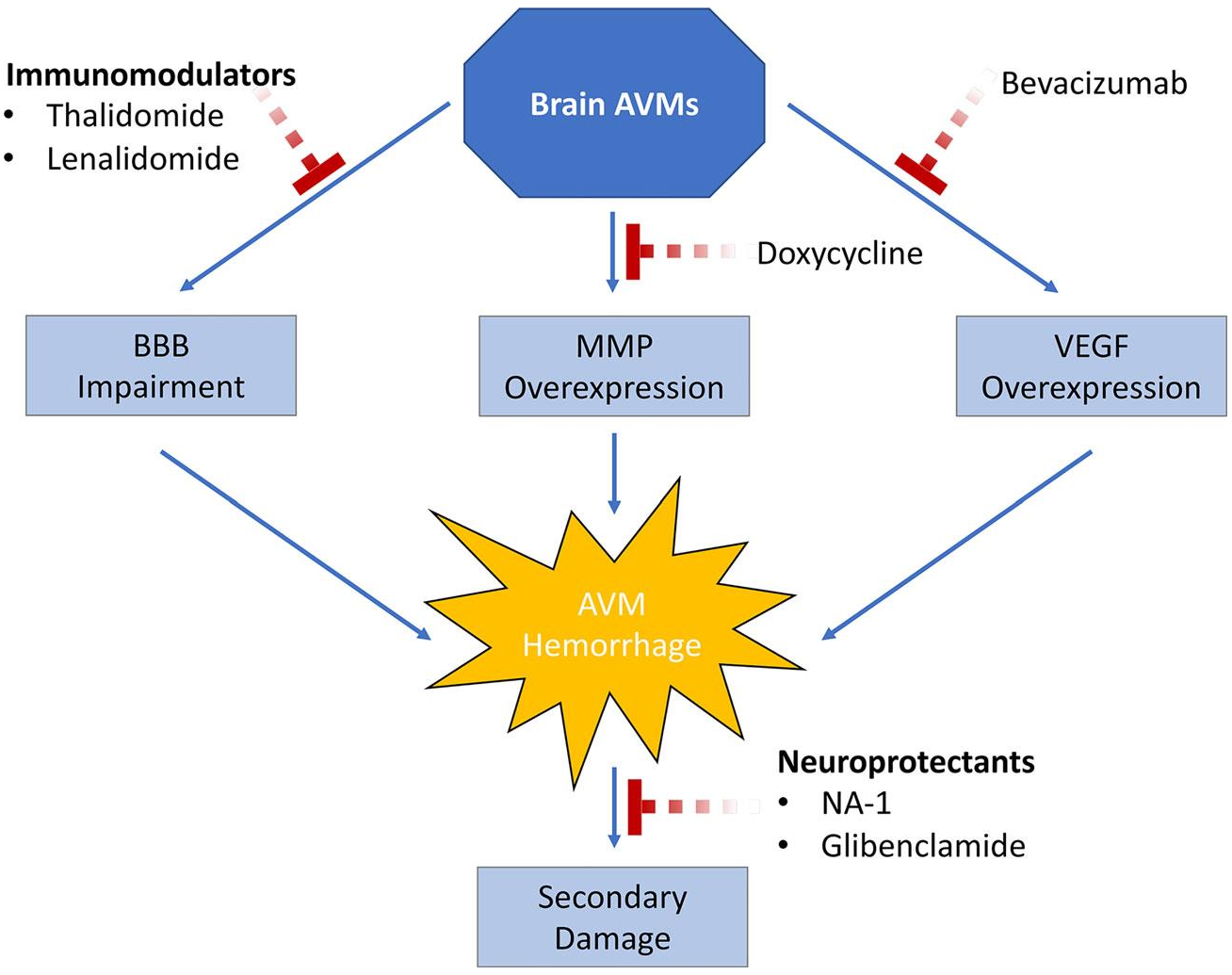


SUPPORTIVE treatment

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

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

PATHOPHYSIOLOGIC treatment



AVM treatment

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

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

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

SURGICAL TREATMENT RISK ESTIMATION

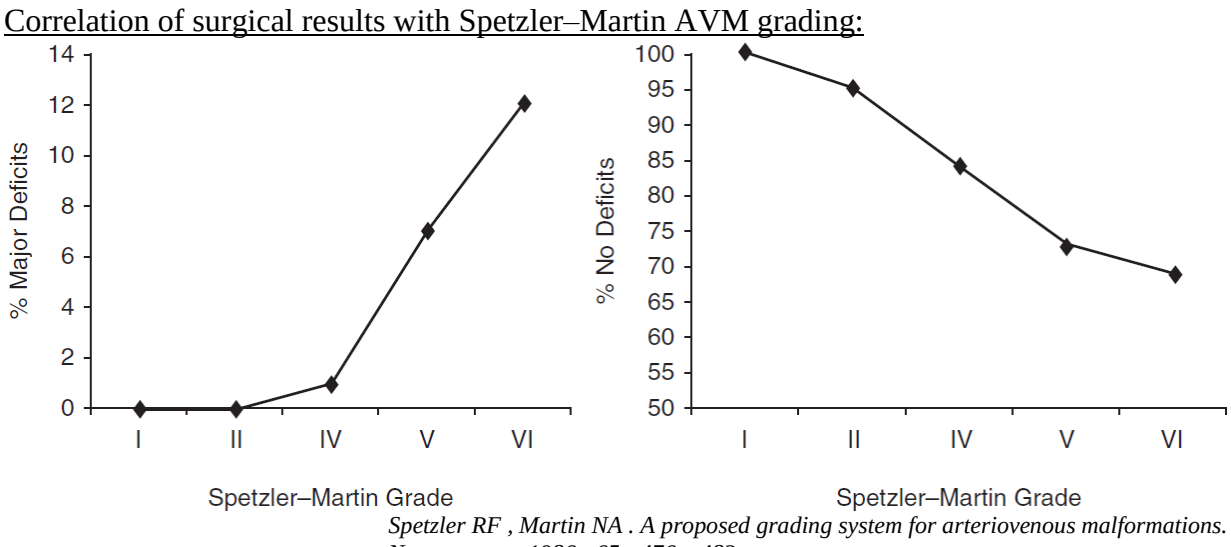
A. SPETZLER-MARTIN grading system – sum points from 3 factors:

I. AVM size (largest diameter of nidus on angiography):
1 point - AVMs < 3 cm
2 points - AVMs 3-6 cm
3 points - AVMs > 6 cm.

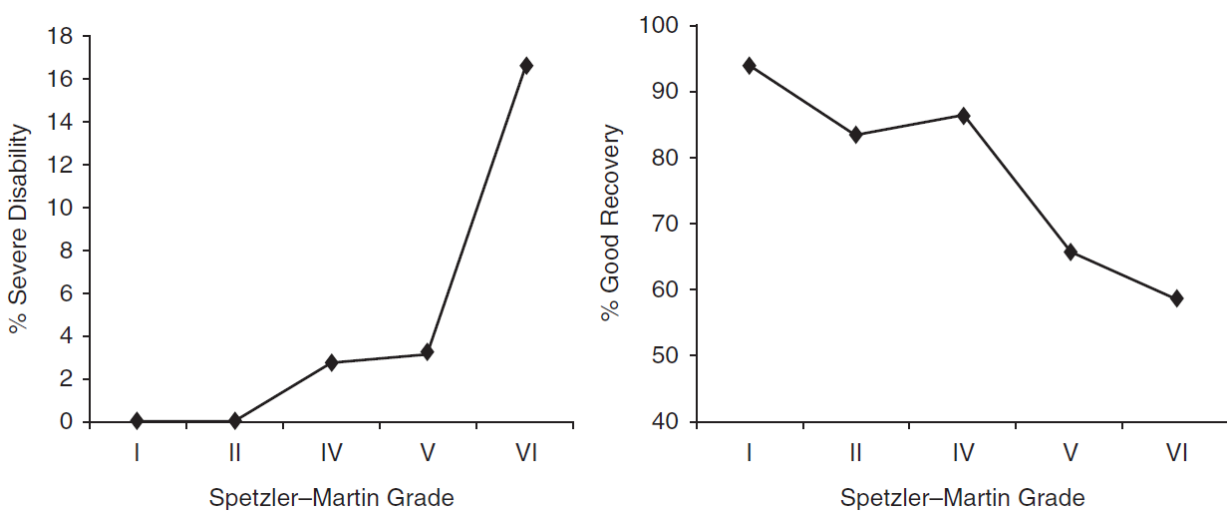
II. Eloquence of adjacent brain:
0 point – if AVM is located in *noneloquent area*.
1 point – if AVM is located in *functionally critical area* (e.g. language, motor, sensory, or visual cortex, thalamus, hypothalamus, internal capsule, brain stem, cerebellar peduncles, deep cerebellar nuclei).

III. Pattern of venous drainage:
0 point – if AVM has *superficial venous drainage* (all drainage is via cortical veins).
1 point – if AVM has component of *deep venous drainage*.

Correlation of surgical results with Spetzler–Martin AVM grading:



Spetzler RF , Martin NA . A proposed grading system for arteriovenous malformations. J Neurosurgery 1986 ; 65 : 476 – 483



Hamilton MG , Spetzler RF . The prospective application of a grading system for arteriovenous malformations. Neurosurgery 1994 ; 34 : 2 – 6

American Heart Association multidisciplinary MANAGEMENT GUIDELINES:

Spetzler-Martin grade I-II → **surgical extirpation**.

if AVM < 3 cm and surgery has increased risk → **radiosurgery**.

Spetzler-Martin grade III → **embolization** → **surgical extirpation**.
if high surgical risk, **embolization** → **radiosurgery**.

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

Spetzler-Ponce classes:
Class A - Spetzler-Martin grade I and II lesions
Class B - Spetzler-Martin grade III lesions

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

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

Spetzler-Martin Grading	Points	Supplementary Grading
Size, cm		Age, y
<3	1	<20
3-6	2	20-40
>6	3	>40
Venous drainage		Bleeding
Superficial	0	Yes
Deep	1	No
Eloquence		Compactness
No	0	Yes
Yes	1	No
Total	5	

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). *may be slowed by proximal balloon occlusion
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).

Approach:

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

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

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.
   - higher grade AVM (esp. supp-SM ≥ 3), higher role of embolization.
   - **definitive surgery** is performed ***within week*** of embolization (new feeding vessels and gliotic scar will make surgical procedures more difficult).

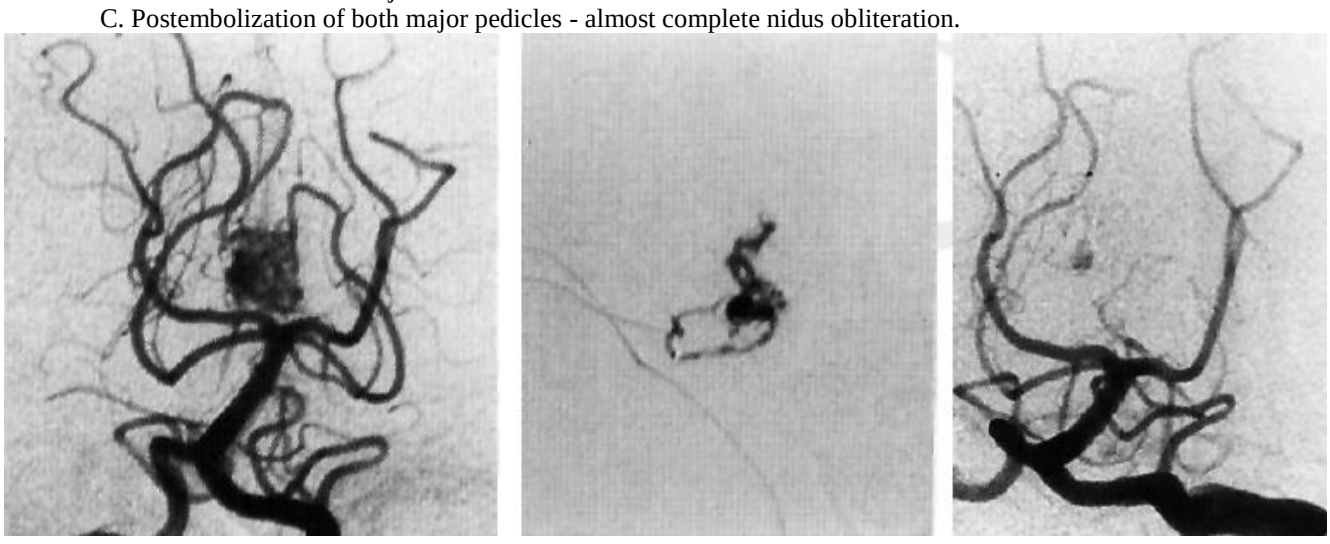
   N.B. embolization prior to radiotherapy reduced rate of total obliteration after SRS! – this statement has been challenged.

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:
a) inadvertent embolization of arteries supplying normal brain tissue;
   if vital 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 neurological picture
b) obliteration of venous outflow leading to intracerebral hemorrhages.

- MORBIDITY 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).
- ***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 **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 (tractional *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 extirpation with minimal deficits.
  – larger lesions are separated into compartments and isolated and collapsed as separate units.
- **aneurysms** are clipped surgically as well.
  When intra-nidal aneurysm is found in AVM-bleeding, it must be targeted for urgent therapy; i.e. presume that aneurysm (not AVM) is source bleeding - treat urgently to prevent rebleeding!
  – there is increase in resistance of vessels that have fed AVM after AVM is removed - if aneurysms are not treated before or at time AVM is treated, they may hemorrhage in postoperative period.
- measures to reduce blood loss:
  1) controlled hypotension during surgery (maintain MAP 40-60 mmHg)
  2) embolization of feeding vessels prior to surgery.

POSTOPERATIVE

- 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 negative postresection angiogram, have been reported).
- routine postoperative **seizure prophylaxis** (min. 3 months).

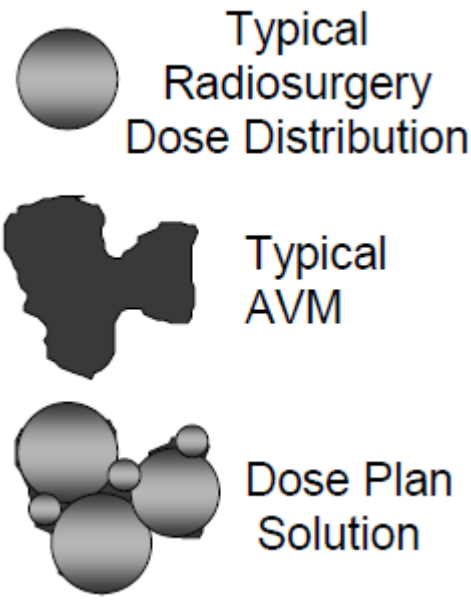
STEREOTACTIC RADIOSURGERY

- results using any of these techniques appear to be relatively similar:
  a) proton beam
  b) LINAC
  c) gamma knife
- noninvasive and can access all anatomic locations of brain (e.g. surgically inaccessible).
- ideal for ≤ 3 cm AVMs (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 cm^3), 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).



Stereotactic angiography must be performed with the angiographic index box affixed to the frame. Angiograms are taken with images obtained from the left or right, and front or back. If the AVM fills from more than one parent artery, then all arteries should be injected and imaged. The technician or nurse in attendance should make note (literally) of which side the x-ray source is on for the two projections. It is our practice to have the angiographer draw the nidus on the best images of the test.

Those images may be loaded into the system in several ways. Film may be scanned into Gamma Plan using the furnished transparency scanner. This method will be shows step-by-step. Alternatively, they may be transferred electronically to Gamma Plan using the institutions image network.

When imported, each image will show as an icon with a "?" superimposed on it. Clicking on "define" will allow you to register the image in stereotactic space. Two squares composed of dotted green lines will be superimposed on the image. Drag the fiducials on the square having 5 fiducials to the corresponding five "X"s or "+"s that appear on the image. Repeat with the other square with 4 fiducials. Enter the proper x-ray source and define. The error of the fifth marker should be < 0.5 mm. You will be asked to confirm that the coordinates of the index box is correct – these should be checked against those in the Gamma Knife operator's manual.

Once the appropriate AP (or PA) and lateral images are defined, bring them up in angio windows by dragging them (middle mouse button). Go to "Regions and Volumes", "Add" a new region then trace in the nidus. You may use one region for both are define a new region for each image. Make sure that "Project" is checked on the region. If so, the outer boundaries of the malformation (as determined from the angiogram) will project onto and volume image (e.g., MRI, MRA, CT, etc.). Use those lines to help you draw a volume for the nidus using MRI, MRA, CT, etc. Ultimately you will use that volume for planning and treatment, not the region on the angios. If everything is correct, your contour on any given image slice should touch all four walls of the projection on that slice at least once. If you are using multiple angios taken in the same direction, the nidus should border the outermost 4 projections. Also, the contours of the volume will be projected on the angiograms.

In reality, often there is not a perfect match between the angio and the image volume. Your job in any area is to decide which is right and that, ultimately, the volume is correct.

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.

	Author	N	Angiographic Obliteration	Permanent Deficit
Particle	Steinberg	86	39-94%	9.6%
	Colombo	153	80%	2.2%
LINAC	Englehart	212	72%	4.3%
	Friedman	158	69-89%	4.3%
	Pollock	315	66%	
Gamma Knife	Lunsford	227	58-100%	1.2%
	Yamamoto	121	75%	5%
	Karlsson	273	80%	
	Maruyama	500	81-91%	1.4%*

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)

3) undilated feeders (odds ratio = 0.36)
4) smaller AVM volume (odds ratio = 0.95)

SRS is treatment of choice for AVMs < 6 cm^3, even after bleeding!

Predictors of SRS failure:

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

Combination of embolization and SRS does not offer any advantages over SRS alone and may have significant disadvantages

- reduction in flow within the AVM does not improve SRS results; embolization can only be an effective adjunct to SRS if it results in permanent reduction of the nidus volume (*recanalization of embolized portions of the AVM that may have been outside the SRS target* results in persistent AV shunting and treatment failure).
- combination of embolization and radiosurgery *does not provide any additional protection against AVM hemorrhage* during the latency period.
- if embolization is used, the ***optimal time*** for SRS is not known, but generally waiting for a period of several weeks is beneficial to reduce the likelihood of vascular ischemic complications or residual cerebral edema sometimes associated with embolization followed by early radiosurgery.
- persistent out-of-field nidus (marginal failure) was identified in 18% of previously embolized vs. 5% of nonembolized patients (p = 0.006).

Friedman WA et al.: Analysis of factors predictive of success or complications in arteriovenous malformation radiosurgery. Neurosurgery 52:296-307; discussion 307- 308, 2003

- *avoid AVM embolization if planning radiosurgery*!

Kano H, Kondziolka D, Flickinger J, et al "Stereotactic radiosurgery for arteriovenous malformations after embolization: a case-control study: Clinical article"

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:

J Neurosurg. 2013 Oct;119(4):981-7. A practical grading scale for predicting outcome after radiosurgery for arteriovenous malformations: analysis of 1012 treated patients. Starke RM, Yen CP, Ding D, Sheehan JP.

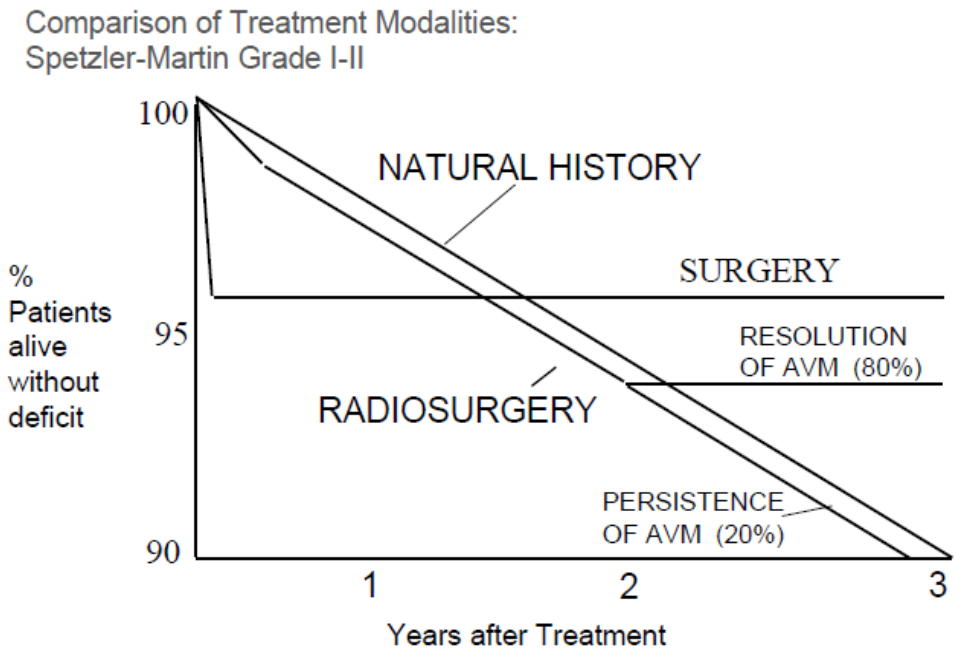
1) AVM **volume** of 2-4 cm^3 - 1 point; AVM volume > 4 cm^3 - 2 points
2) **eloquent** AVM location - 1 point
3) history of **hemorrhage** - 1 point

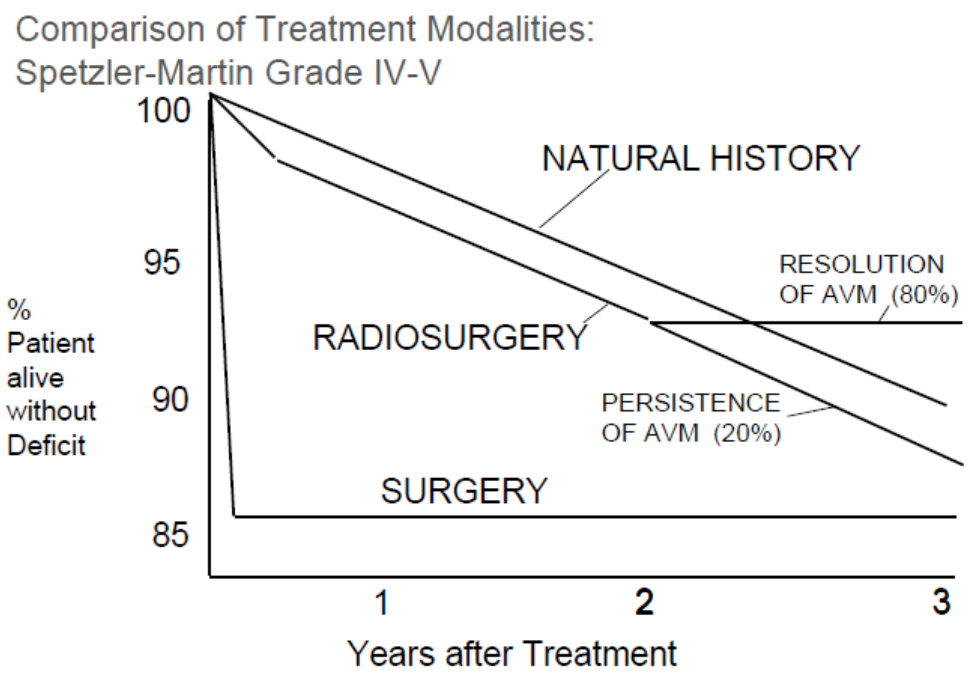
0-1 points – 80% of patients had a favorable outcome
2 points – 70%
3-4 points – 45%.

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





RECURRENCE

  - 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 unresected 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 8-fold: 8.1% per pregnancy (= 10.8% per year) vs. 1.1% baseline annual risk – data from Brigham and Women's Hospital and Harvard Medical School**.

**Risk of First Hemorrhage of Brain Arteriovenous Malformations During Pregnancy: A Systematic Review of the Literature. Christopher L Davidoff et al.Neurosurgery, Volume 85, Issue 5, November 2019, Pages E806–E814,*
***Blondel B, Schwab F, Ungar B, et al "Hemorrhage From Arteriovenous Malformations During Pregnancy"*

- 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, transsylvian-transinsular.
- approaches for **THALAMUS** – interhemispheric: transcallosal or trans-splenial.
- after microsurgical isolation of feeding perforating vessels, lateral ventricle is opened and lesion excised (incl. choroid plexus - carries deep arterial supply to nidus).
- surgical morbidity is formidable (hemiplegia, aphasia, 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. transtentorial route for lesions around tectum and superior cerebellum).
- **BRAINSTEM** AVMs:
  A) **microsurgical resection** - combined subtemporal-suboccipital-retrolabyrinthine-transtentorial 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 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) RIT1 (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

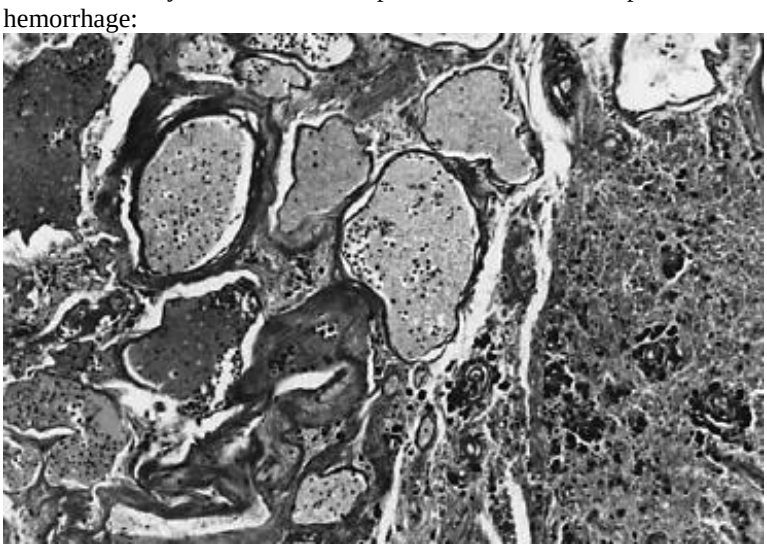
- hamartomatous enlarged sinusoidal capillaries, s. caverns (single layer of endothelium, thin collagenous wall, no smooth muscle, no elastic fibers).
- well-circumscribed, "mulberry" appearance, expand slowly.
- capillaries are immediately adjacent to each other.

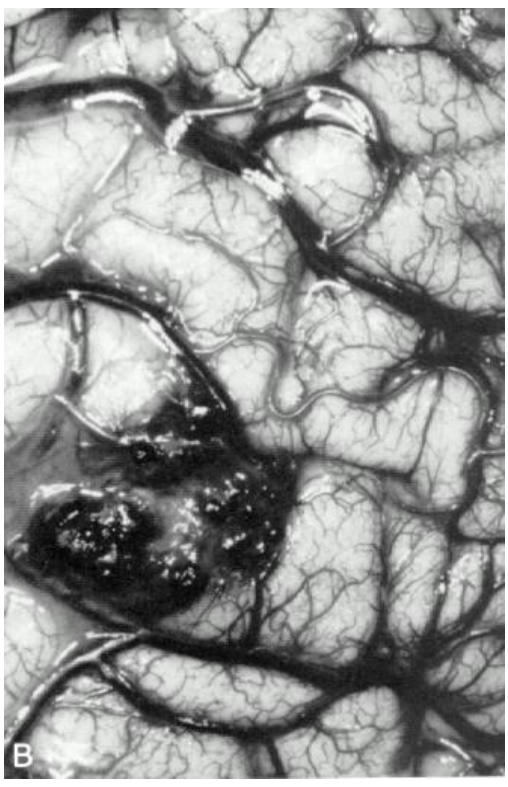
  No intervening neural tissue!!!

- not associated with enlarged feeding ARTERIES or draining VEINS.
- blood flow is low or even stagnated:
  - *difficult angiographic visualization*!
  - intra-lesional ***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 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.

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





Well circumscribed mulberry appearance:

CLINICAL FEATURES

≈ 40% of lesions are **ASYMPTOMATIC**.

10-25% lesions are **SYMPTOMATIC**:

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

*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 neurological deficits***.
- ***headaches*** are prominent wherever angiomas 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!

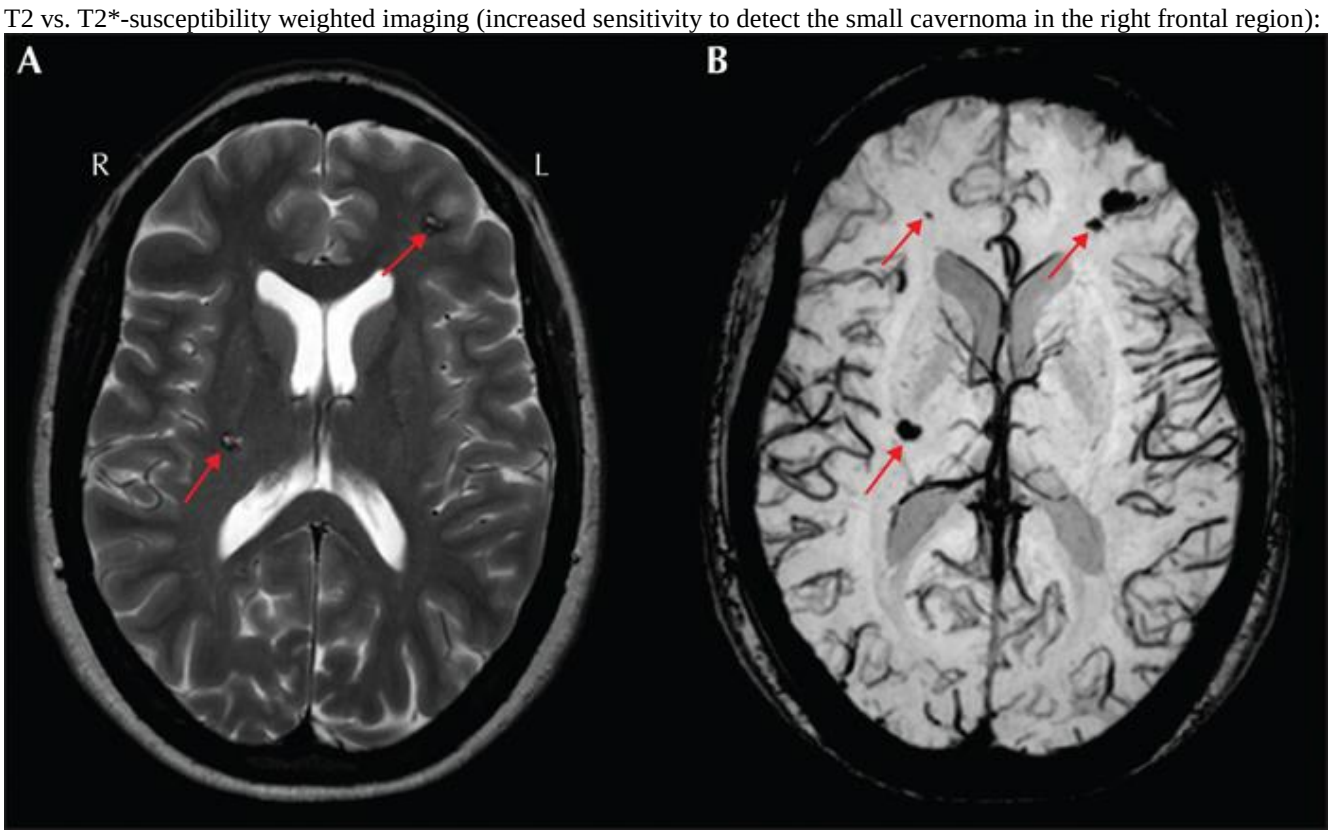
MRI

- most-sensitive neuroimaging - appearance is sufficiently characteristic:

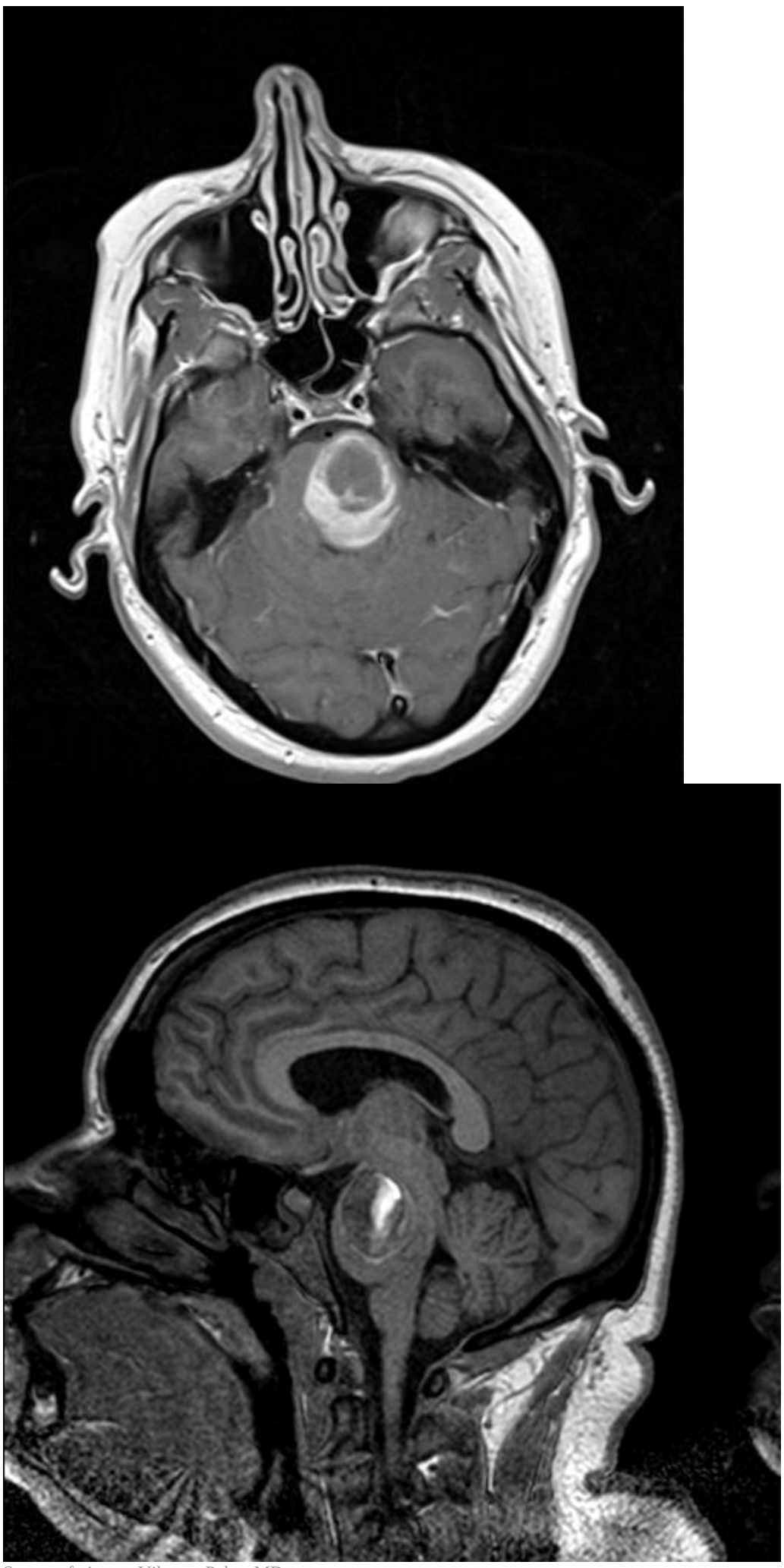
Well-defined ***reticulated - multilobular*** *core of mixed-signal intensity* ("**POPCORN**"), typically surrounded by dark ***hemosiderin*** *ring* on T2.
T2*-susceptibility weighted imaging which shows increased sensitivity.

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

T2 vs. T2*-susceptibility weighted imaging (increased sensitivity to detect the small cavernoma in the right frontal region):

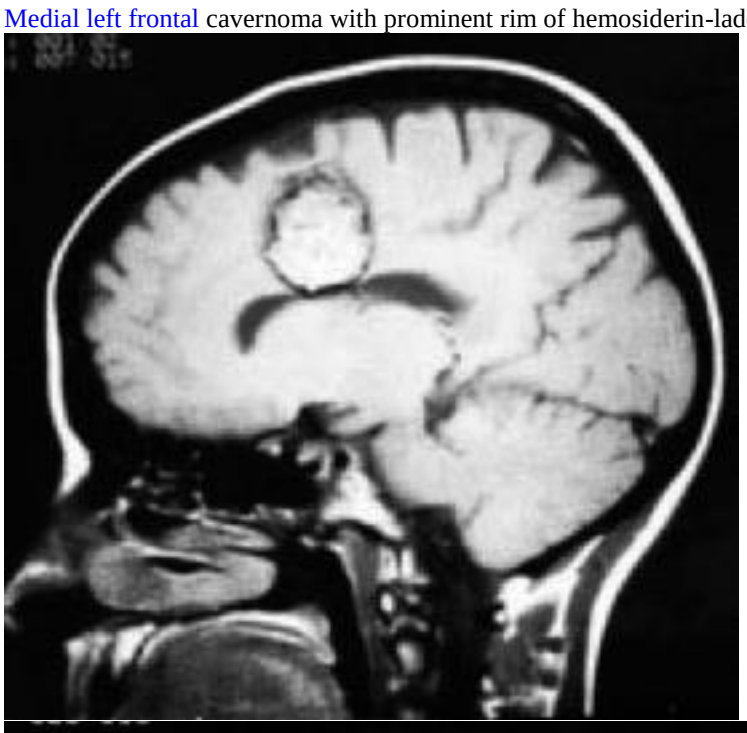


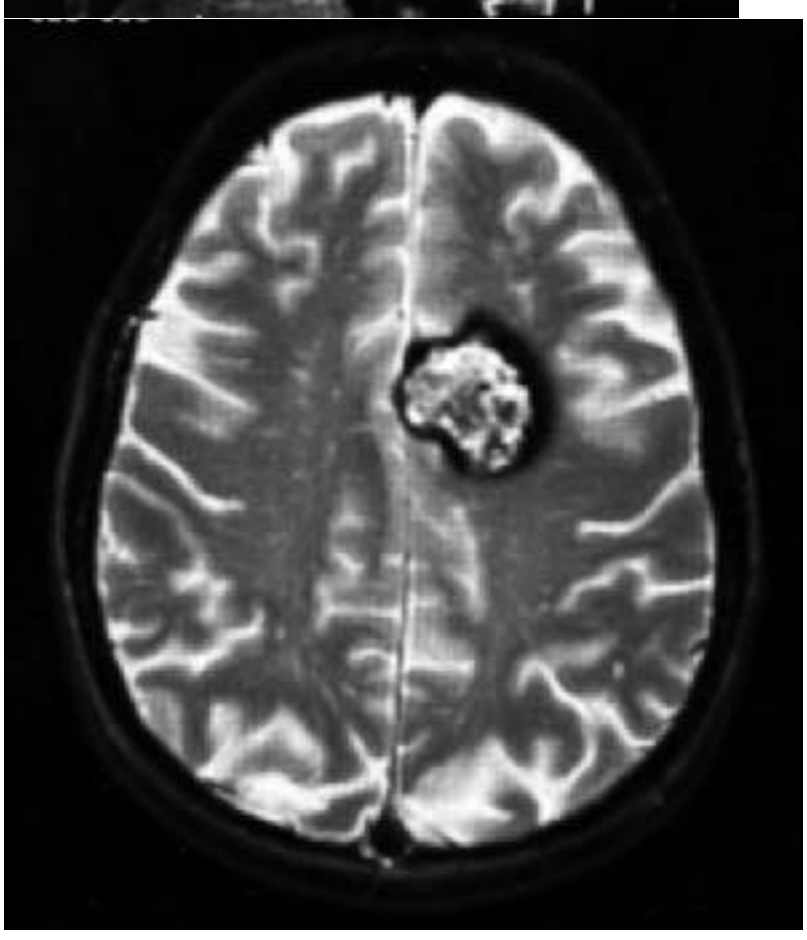
Midbrain cavernoma:



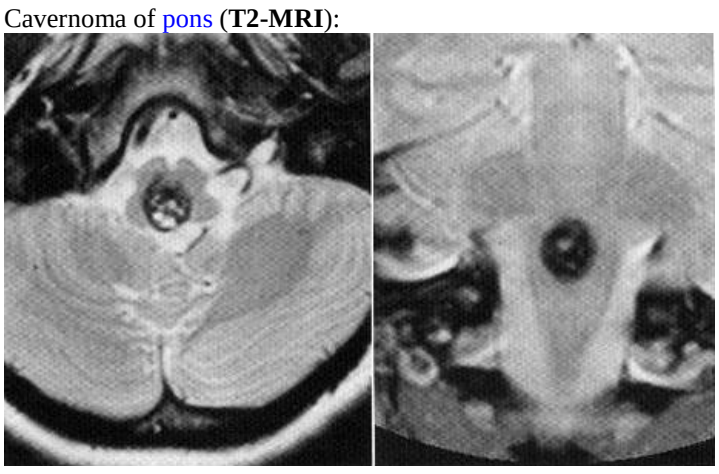
Source of picture: Viktoras Palys, MD >>

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

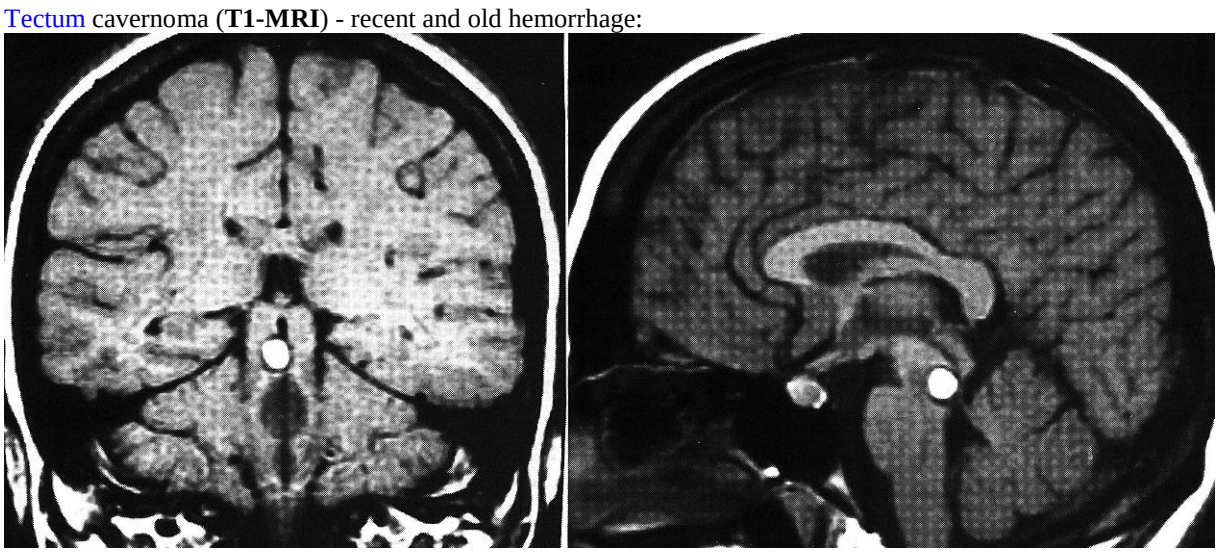




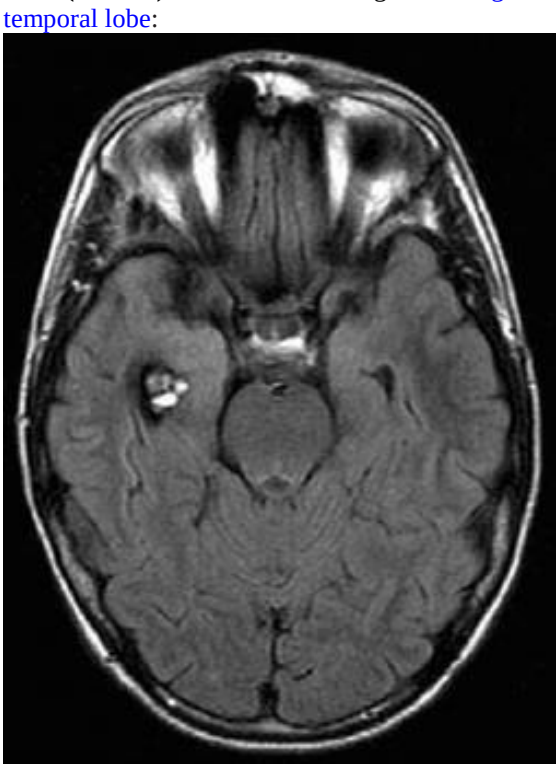
Cavernoma of pons (**T2-MRI**):



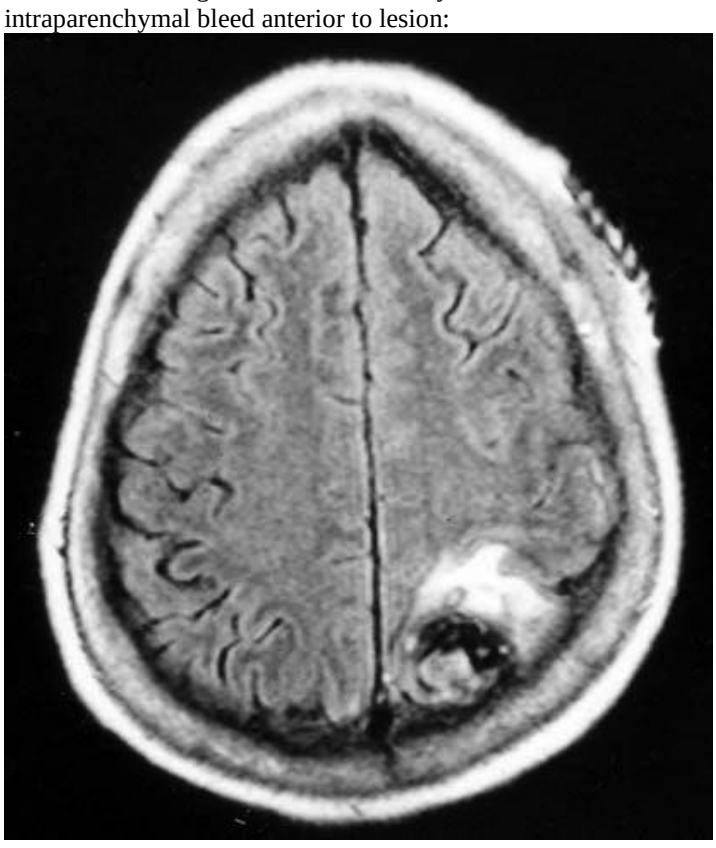
Tectum cavernoma (**T1-MRI**) - recent and old hemorrhage:



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

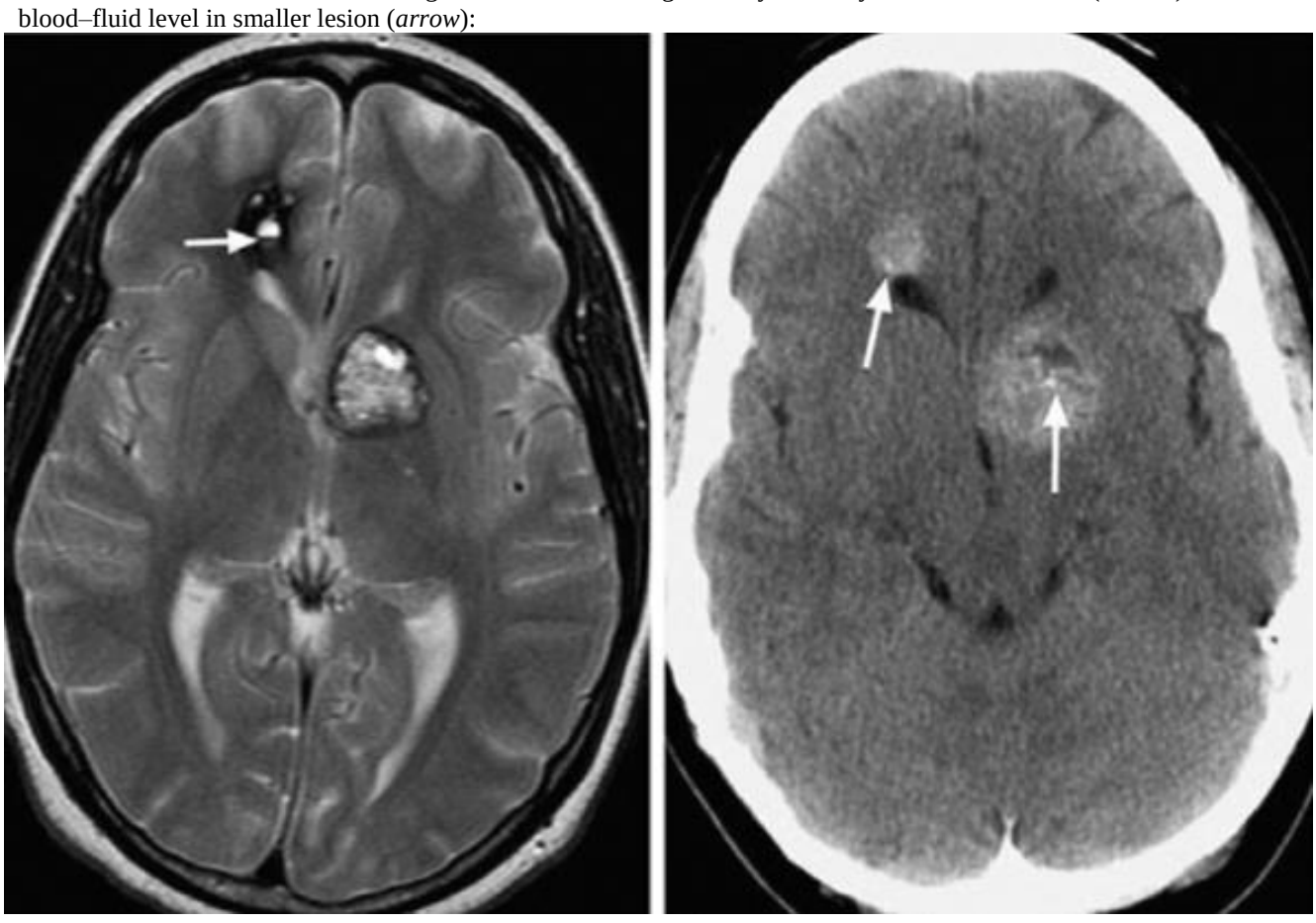


T1-MRI - left-sided parietal 2-cm cavernous malformation with areas of hemorrhage of various chronicity + acute intraparenchymal bleed anterior to lesion:

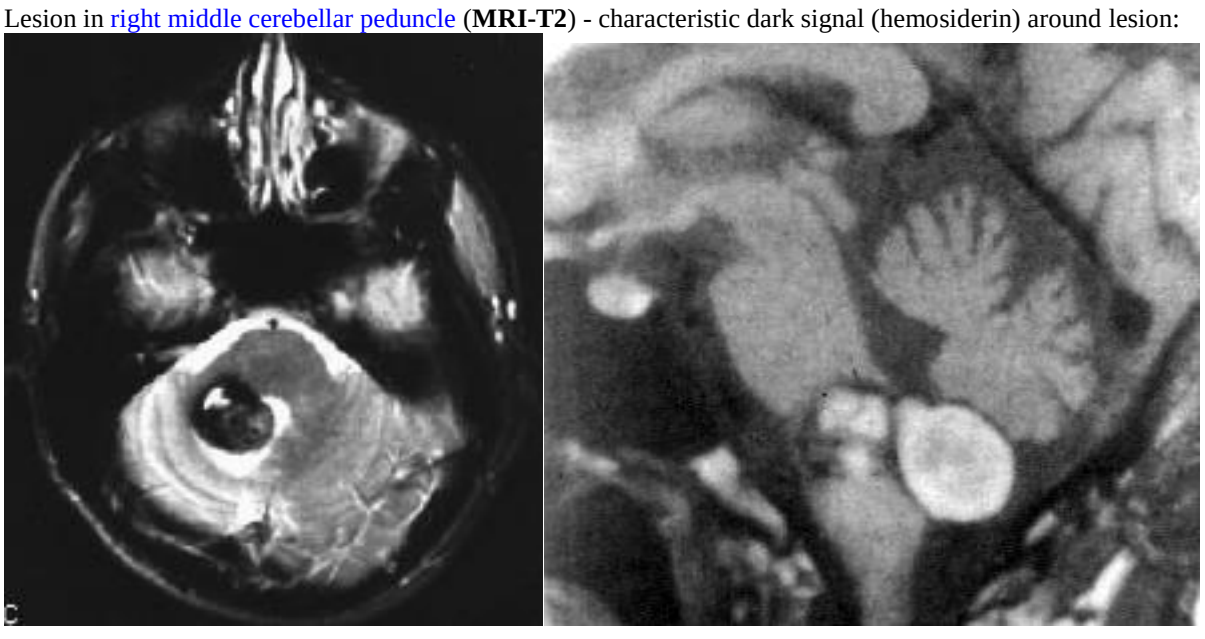


T2-MRI: high signal due to MetHb, low signal rim of hemosiderin indicates old hemorrhage; note blood–fluid level in smaller lesion (*arrow*):

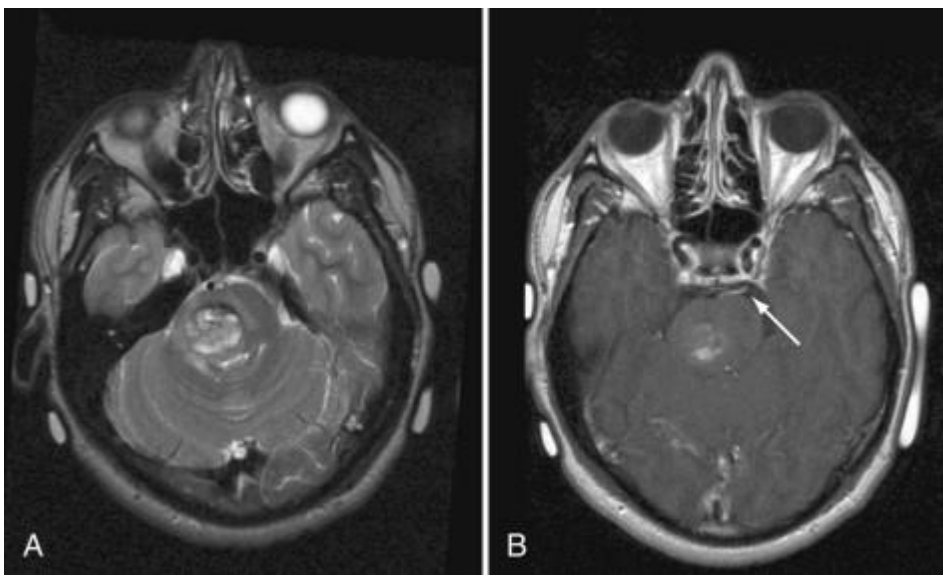
Unenhanced CT of same patients - lesions predominantly of high density with tiny foci of calcification (*arrows*):



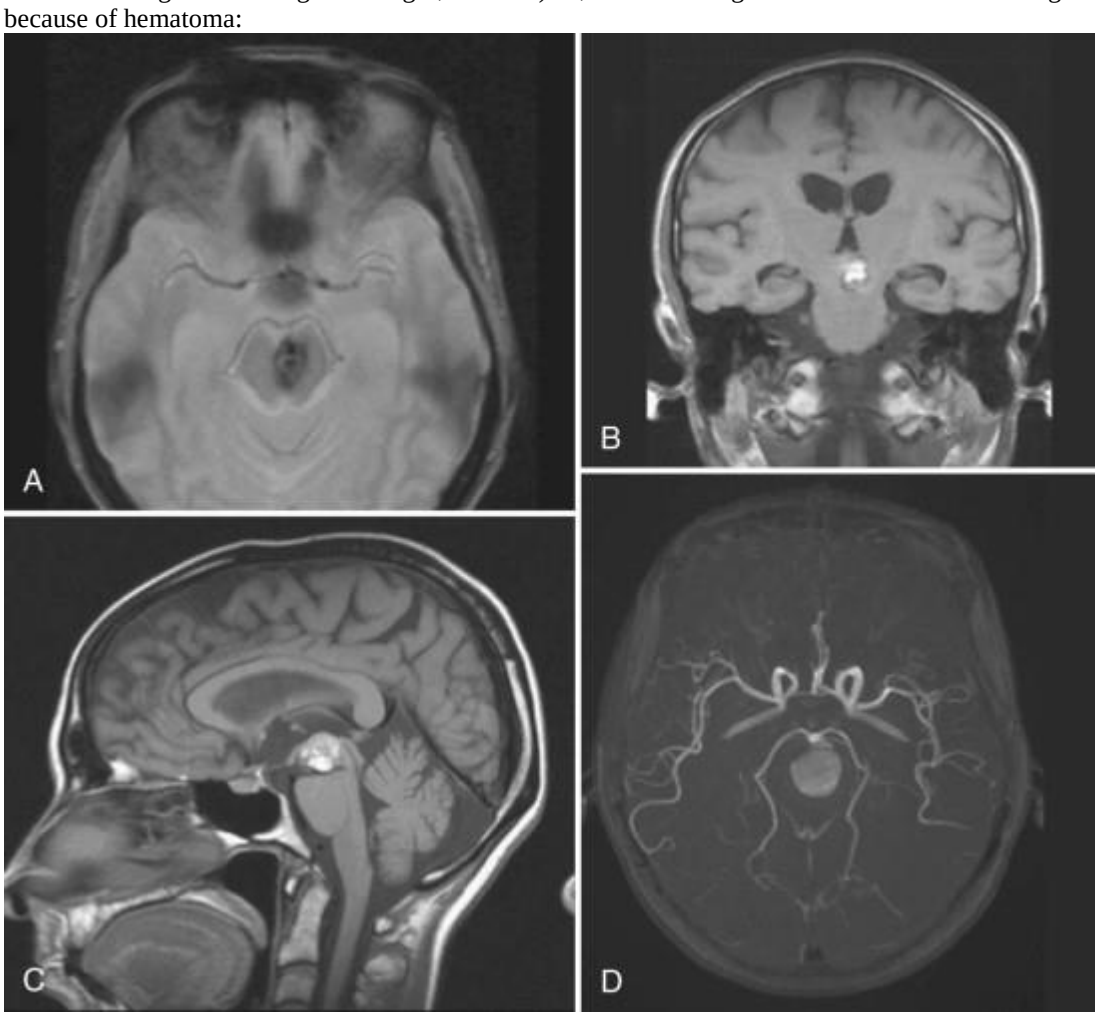
Lesion in right middle cerebellar peduncle (**MRI-T2**) - characteristic dark signal (hemosiderin) around lesion:



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 caverrnoma, 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:



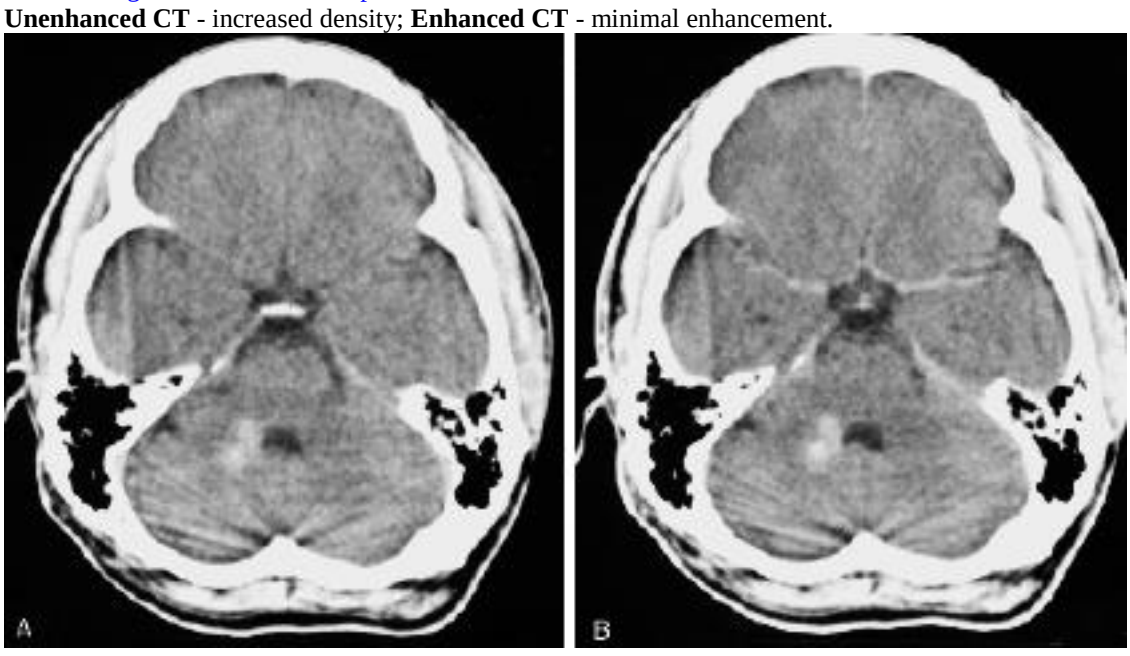
CT

- homogeneous* *focal hyperdensity* ± *calcifications* (≈ 30%).

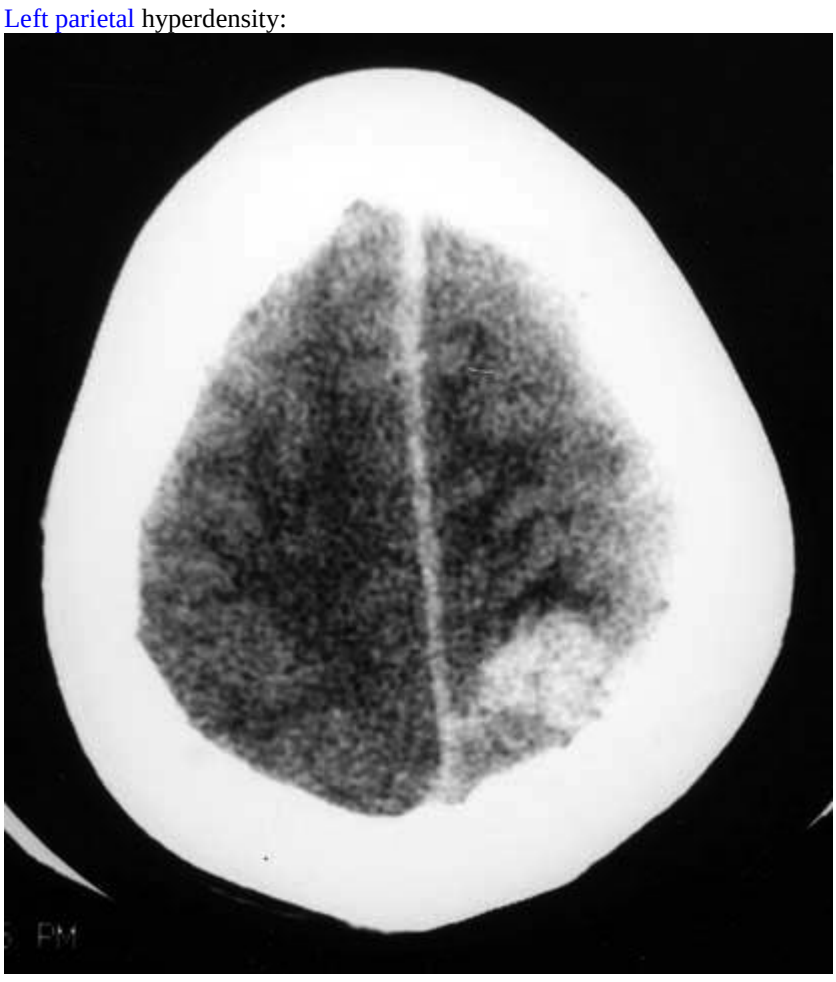
*unless thrombosed, or after hemorrhage

- no edema or mass effect.
- IV contrast may show only ***faint patchy enhancement***.

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



Left parietal hyperdensity:



TREATMENT

Indications:

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!

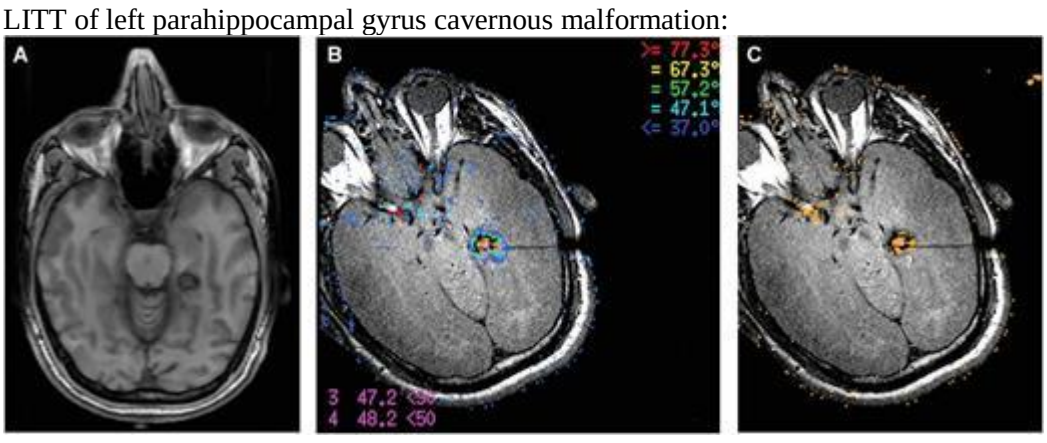
RADIOSURGERY

- 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 and, thus, unlikely to provide direct hemostasis.

LITT of left parahippocampal gyrus cavernous malformation:



Case series

McCracken "Magnetic Resonance Thermometry-Guided Stereotactic Laser Ablation of Cavernous Malformations in Drug-Resistant Epilepsy: Imaging and Clinical Results" Operative Neurosurgery, March 2016 - Volume 12 - Issue 1 - p 39–48

- 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.
- reimaging (6-21 months) indicated lesion diminution with surrounding liquefactive necrosis.

Case series

Willie JT, Malcolm JG, Stern MA, et al. Safety and effectiveness of stereotactic laser ablation for epileptogenic cerebral cavernous malformations. Epilepsia. 2019;60:220– 232. 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 became so following open resection.

  LITT presents no barrier to subsequent intracranial monitoring or open resection.
- delayed imaging validated CCM involution (median 83% volume reduction) and ablation of surrounding cortex.
- histopathologic examination of one previously ablated CCM following open surgery confirmed obliteration.
- SLA caused no detectable hemorrhages.
- 2 symptomatic neurologic deficits (visual and motor) were predictable, and neither was permanently disabling.

BRAINSTEM CAVERNOMAS

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

Garcia, Roxanna M "Brainstem Cavernous Malformations: Surgical Results in 104 Patients and a Proposed Grading System to Predict Neurological Outcomes" Neurosurgery: March 2015 - Volume 76 - Issue 3 - p 265–278

CAPILLARY TELANGIECTASIA

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

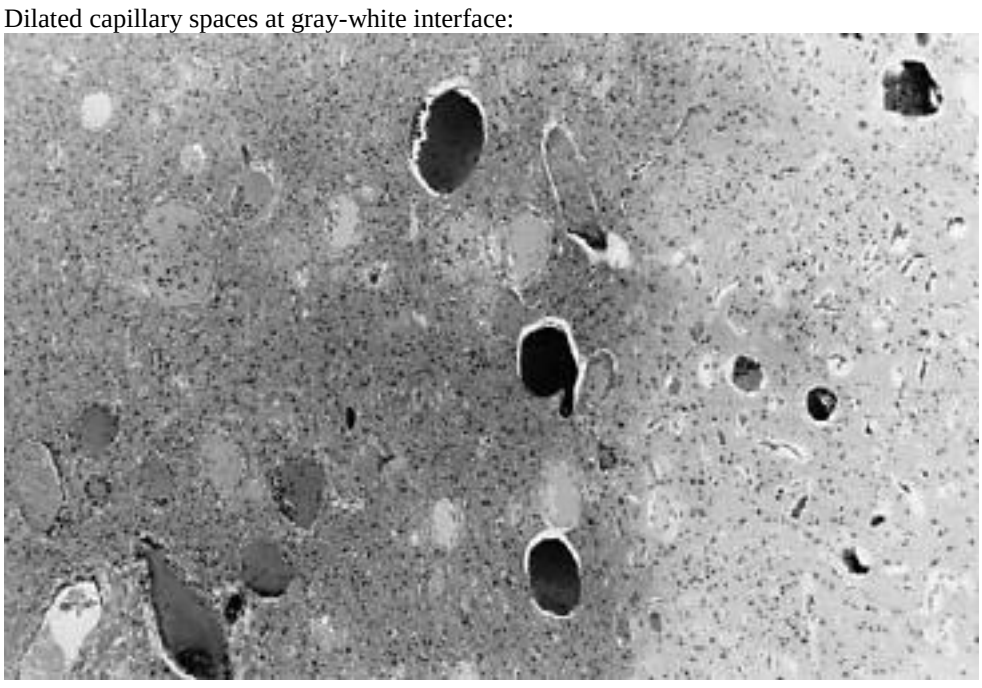
PATHOLOGY

- 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).

  The only feature that differentiates CAPILLARY TELANGIECTASIAS from CAVERNOUS ANGIOMAS is *presence of brain parenchyma between vascular channels*
- no evidence of associated hemorrhage.
- grossly - tiny lesions having appearance of **punctate hemorrhages**.
- usually deep within brain (particularly in brain stem).

Dilated capillary spaces at gray-white interface:



CLINICAL FEATURES

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

DIAGNOSIS

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

TREATMENT

No treatment is indicated.

DEVELOPMENTAL VENOUS ANOMALY (DVA), s. Venous Angioma, Venous Malformation

- *most common type* of intracranial vascular malformation! (PREVALENCE ≈ 2.6%).

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).

*vs. **VARIX** - single dilated vein.

- **postcapillary** structure (no ARTERIAL or CAPILLARY abnormalities).
- veins receive drainage from adjacent healthy tissues (neural parenchyma in and around angioma is histologically normal).

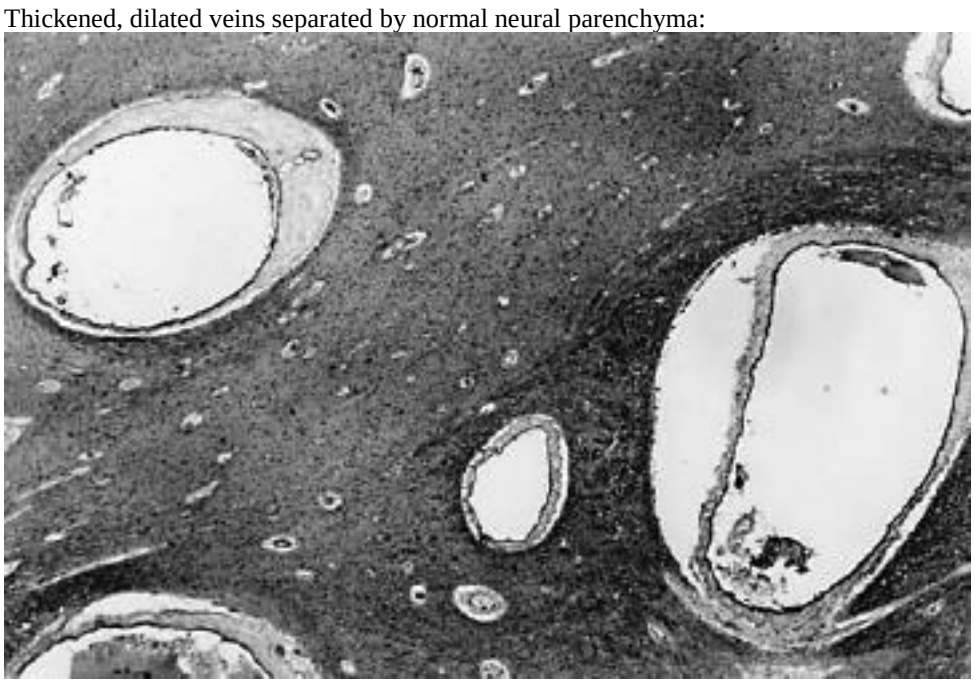
Veins separated by normal brain tissue

- 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! – 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

Thickened, dilated veins separated by normal neural parenchyma:



CLINICAL FEATURES

Generally, ***completely asymptomatic***!

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

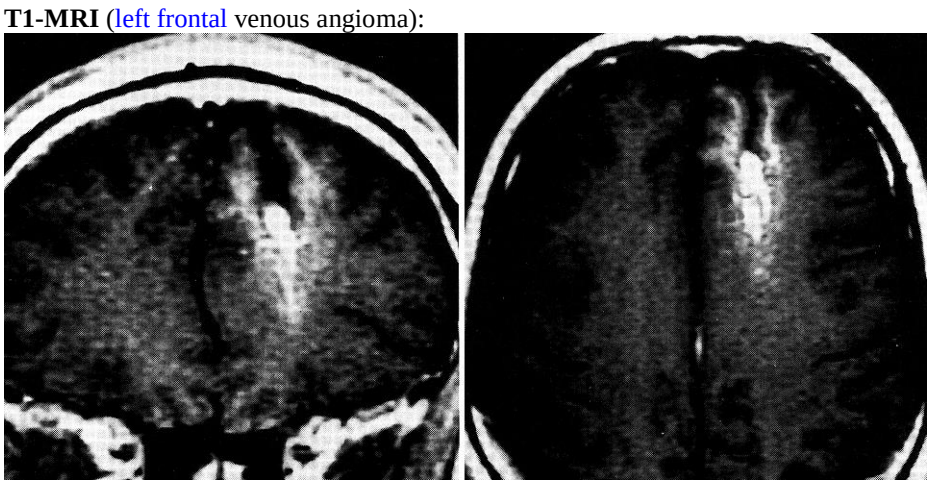
DIAGNOSIS

CT / MRI

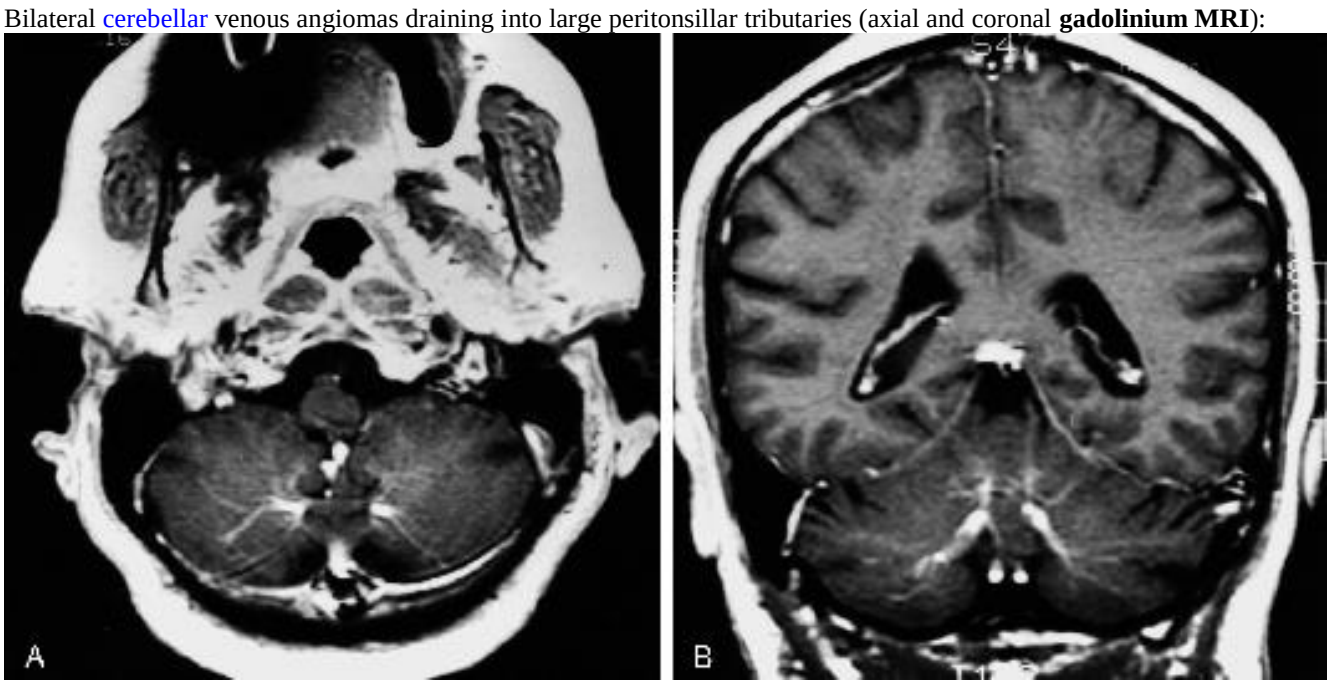
- tubular curvilinear structure ("spokes of wheel"; **MRI** may have sufficient resolution to reveal "caput medusae").
- **CT** may reveal enhancing area (linear, tubular, spotty, or nodular).

Postcontrast CT - two tubular enhancing structures that extend from ventricular margin to brain surface through normal brain tissue; superficially these became continuous with surface veins, which drained into superior sagittal sinus:

T1-MRI (left frontal venous angioma):



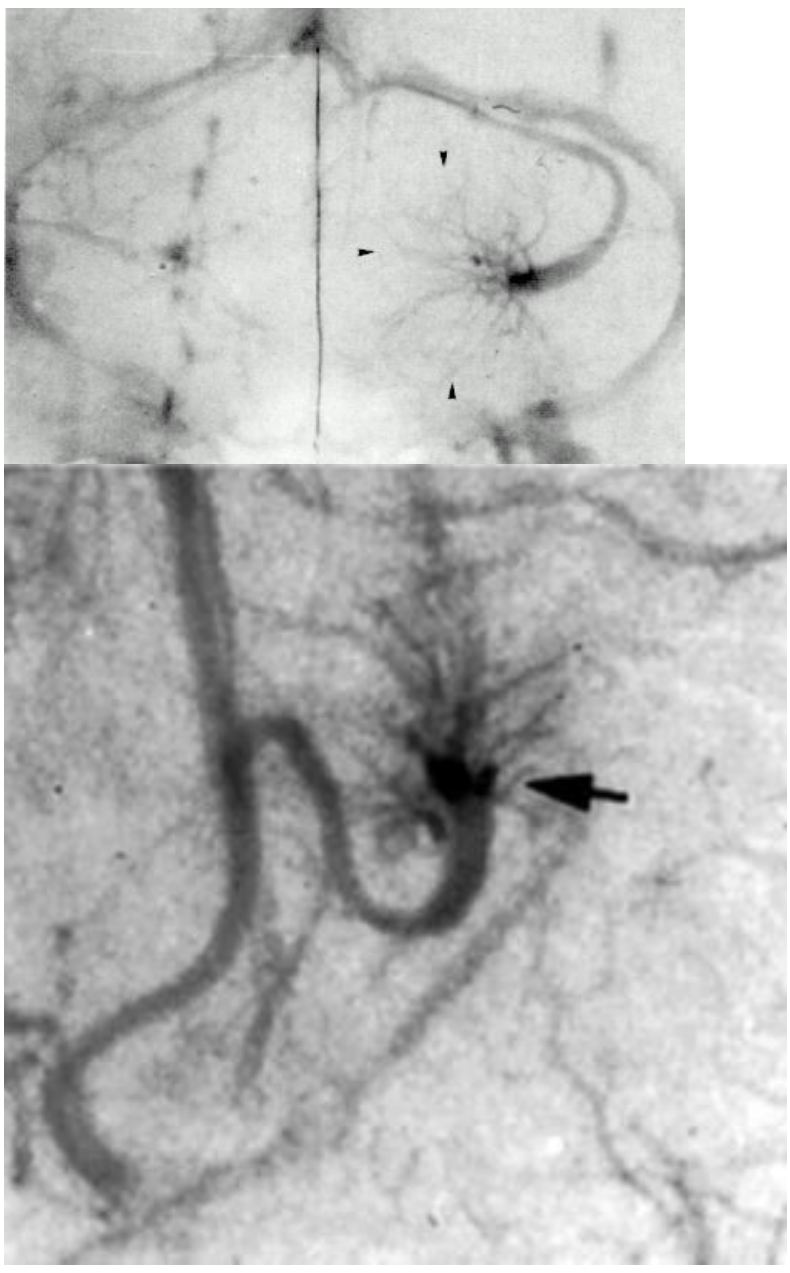
Bilateral cerebellar venous angiomas draining into large peritonsillar tributaries (axial and coronal **gadolinium MRI**):

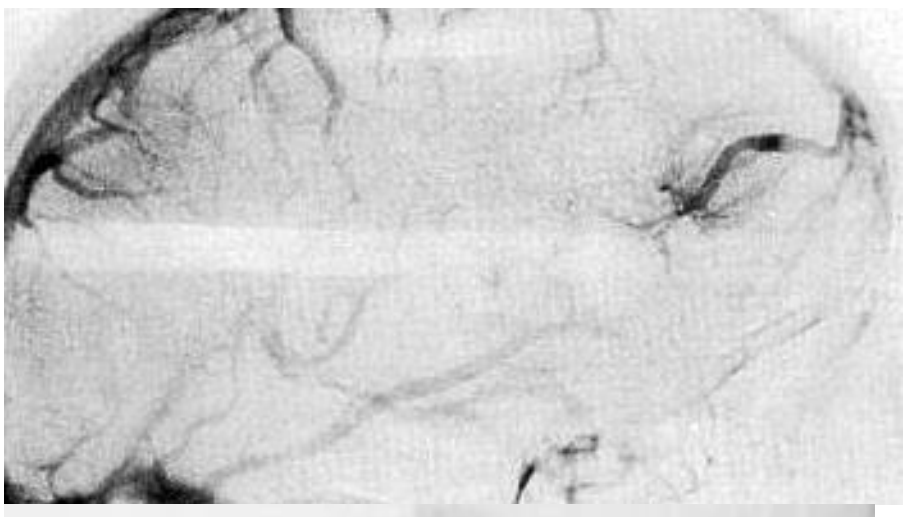


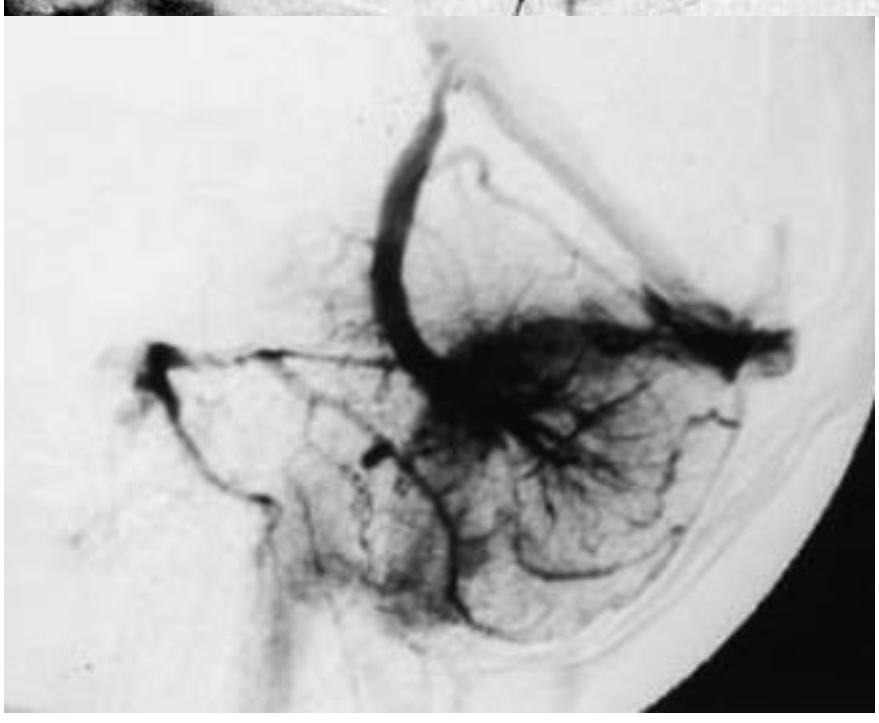
ANGIOGRAPHY

- "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.







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** see p. TrH9 >>

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

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

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

Carotid-Cavernous Fistula → see p. TrH9 >>

ETIOPATHOPHYSIOLOGY

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

Etiology – ACQUIRED*: *therefore, term "MALFORMATION" is not correct

a) ***traumatic*** tear in branch of middle meningeal, intraorbital or even occipital, artery (drainage into venous sinus develops later).
b) dural sinus ***thrombosis*** → attempted recanalization → opening of embryonic AV communications → fistula creation.

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

CLINICAL FEATURES

Many are **asymptomatic**!

Clinical presentation depends on **location** and **venous drainage pattern**:

1. **Bruit**, **pulsatile tinnitus** (lesions shunting into *transverse* or *sigmoid sinus*).
2. **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.

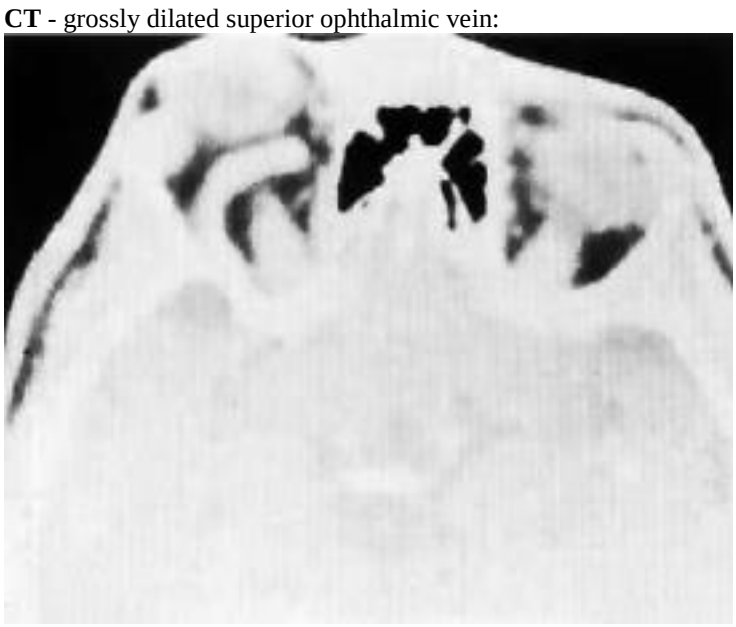
DIAGNOSIS

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

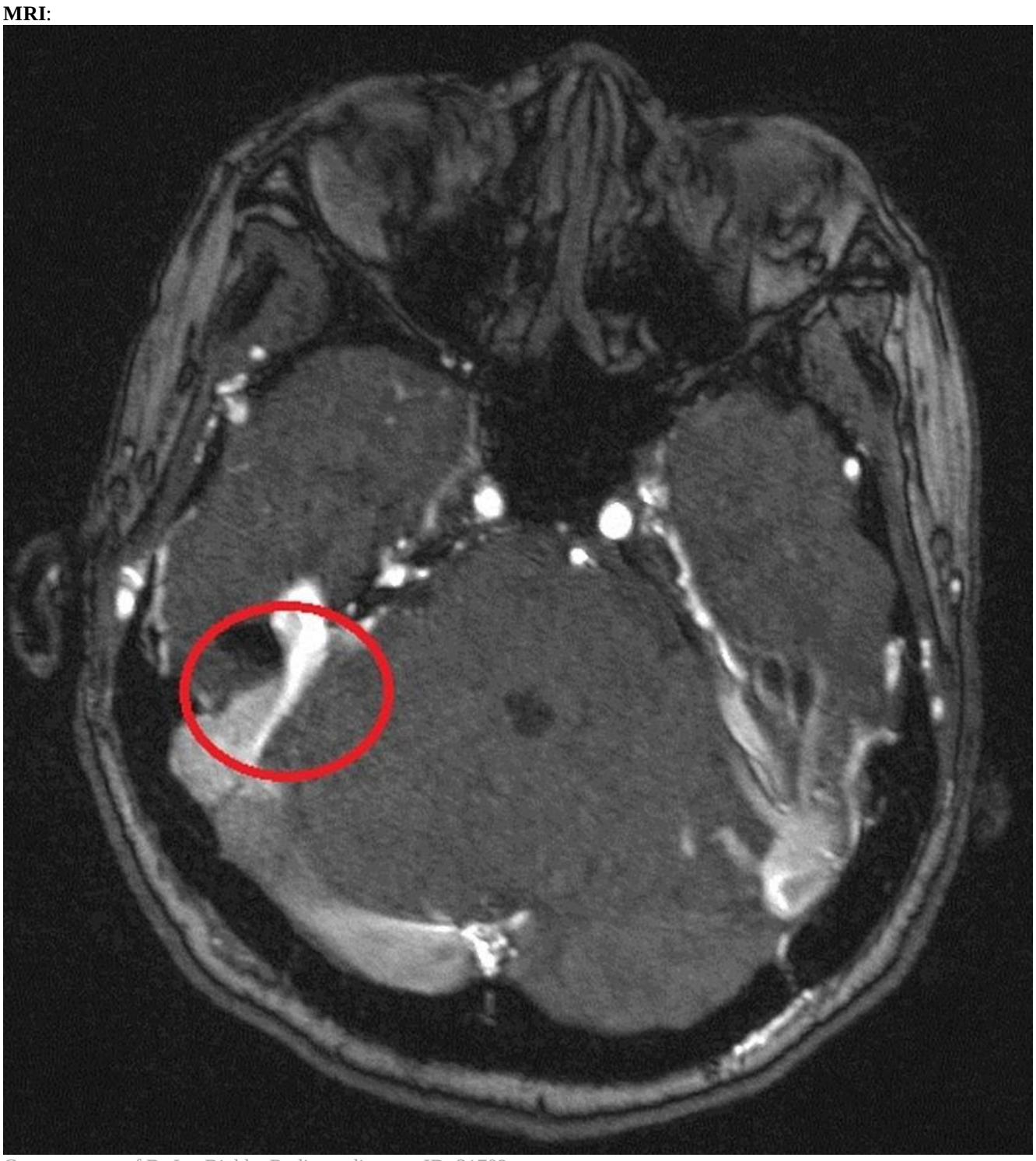
Normally dural arterial branches are not seen angiographically; but DAVFs are well visualized!

- 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:

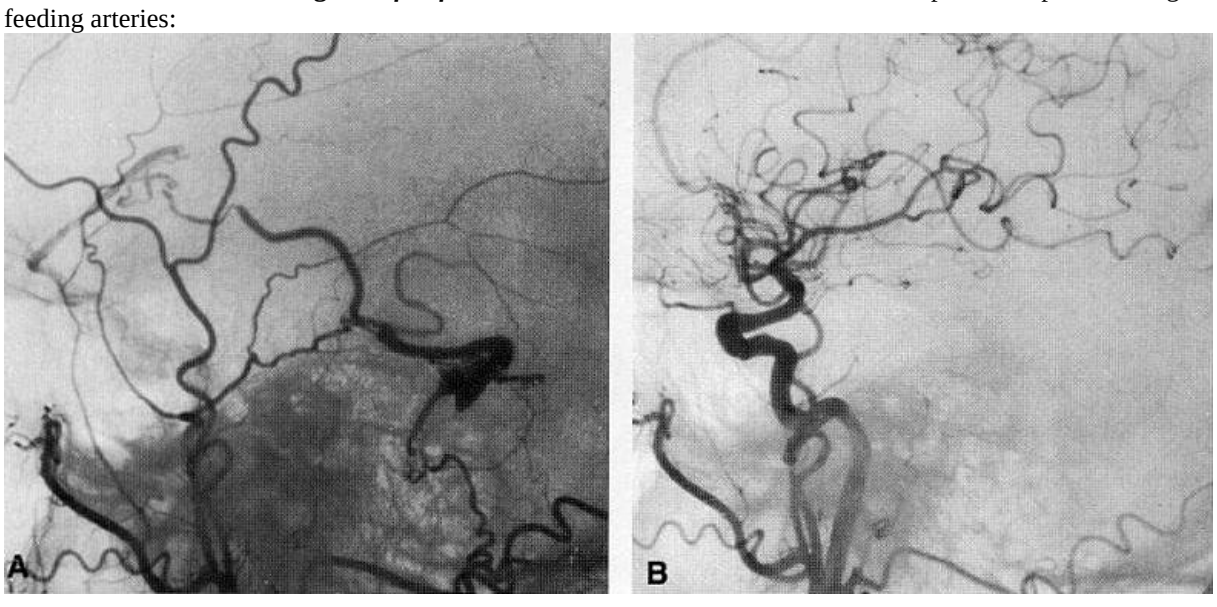


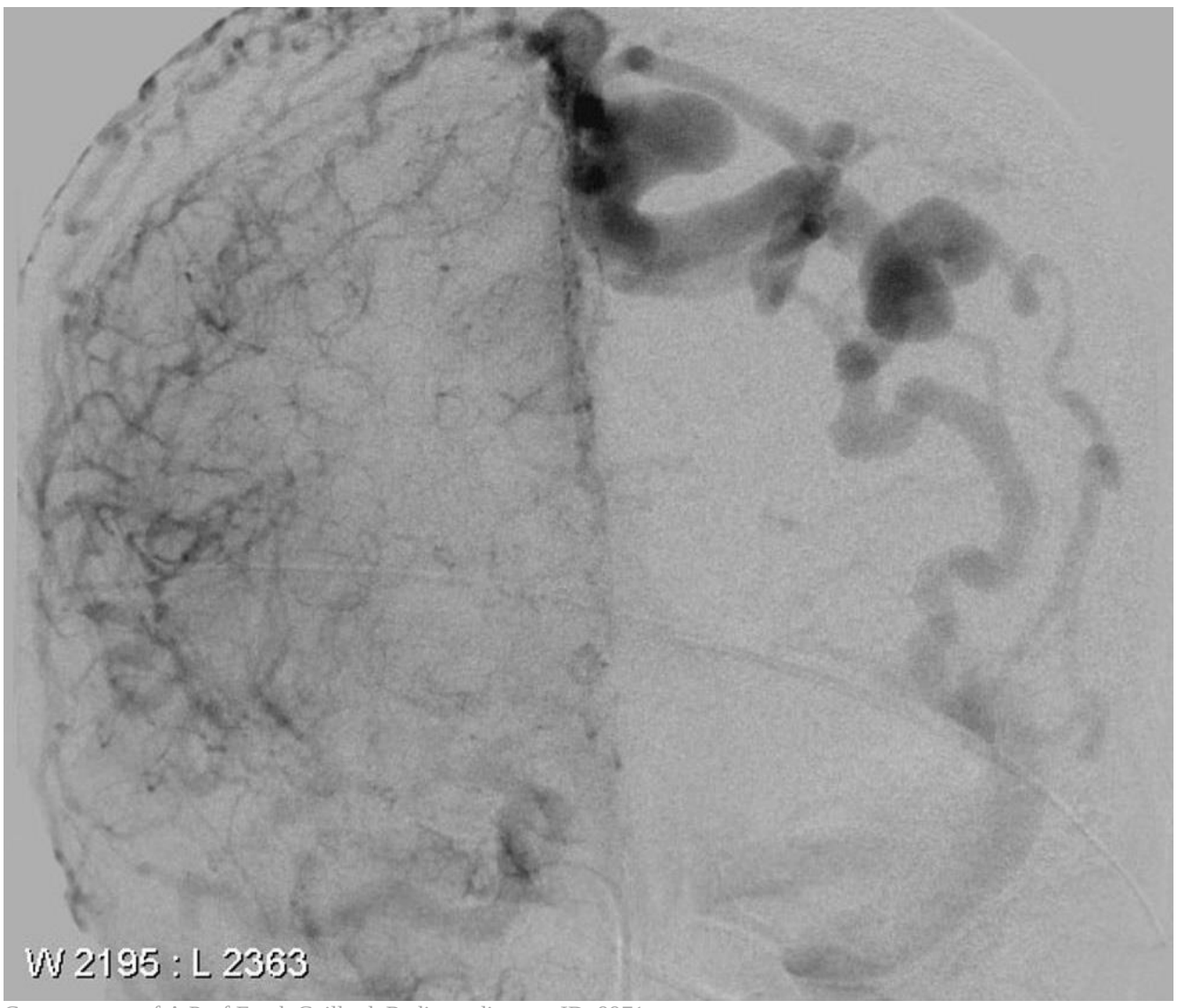
MRI:



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

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





Case courtesy of A.Prof Frank Gaillard, Radiopaedia.org, rID: 9971

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:

Borden	Bleeding rates	Bleeding rates if has venous ectasia	Rebleeding rates after hemorrhage
I	0%		
II	6%		46%
III	10%	21%	46%

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 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.

DJINDJAN 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. (Djindjan 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 Labbé), only arterial feeder disconnection should be done without disturbing the venous aspect of the lesion. If there is no evidence of brain dependence on cortical reflux veins, a classic cortical venous reflux (CVR) disconnection can be done. In patients with neurologic deficits that may be due to venous congestion, only arterial feeders are disconnected.

B. Vascular COMPRESSION – initial treatment for lesion in **TRANSVERSE** or **SIGMOID sinus** - manual compression of occipital artery (behind mastoid) for up to 30 min → diminished inflow can induce thrombosis; contraindication to procedure - cortical venous drainage.

C. Endovascular EMBOLIZATION (treatment of choice):
   a) **PVA particles** - low morbidity rate but high recanalization chances.
   b) **liquid materials** - high cure rates but high morbidity rate (great risk of stroke).
      1) **Onyx-18** (ethylenevinylalcohol copolymer)
      2) **NBCA** (N-butyl-cyanoacrylate)
   c) **coils** (e.g. into sinus → heparin 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*** **-** superselective 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 sinus, these lesions often are hard to cure.
  - very effective adjunct before surgical treatment of DAVF because it can significantly reduce procedural blood loss.

b) ***transvenous*** - often necessitates sacrifice of venous sinus segment at zone of AV shunting; indications - multiple sites of shunting, multiple sources of arterial supply, inaccessible arterial sources; particularly useful in treatment of cavernous 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 Borden III lesions*, which do not drain through a venous sinus but directly in cortical veins.

D. SURGERY:

a) **excision** (traditional surgical treatment of DAVFs) – complete excision of the fistula and the surrounding dura - approach involves the disconnection of all feeding arteries and arterialized leptomeningeal veins and excision of the draining 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 DVAFs 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 patiently until the bleeding stops generally works well.

Suzuki procedure – resecting transverse sinus due to multiple dAVFs.

E. RADIOSURGERY; results in obliteration of DAVFs between one and three years after treatment; disadvantage – delayed fistula closure (not suitable for aggressive DAVFs with annual bleeding risk up to 15%).
- embolization is performed after SRS to avoid the pitfall of having embolization temporarily obscure portions of the nidus that would then not be targeted during the SRS.

2. VEIN OF GALEN Malformation / Ectasia / Aneurysm

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

PATHOLOGY, CLASSIFICATIONS

YASARGIL classification according to ***arterial feeders*** *(Yasargil, M. G.: Microneurosurgery. Vol IIIB. Stuttgart, Thieme, 1988)*:

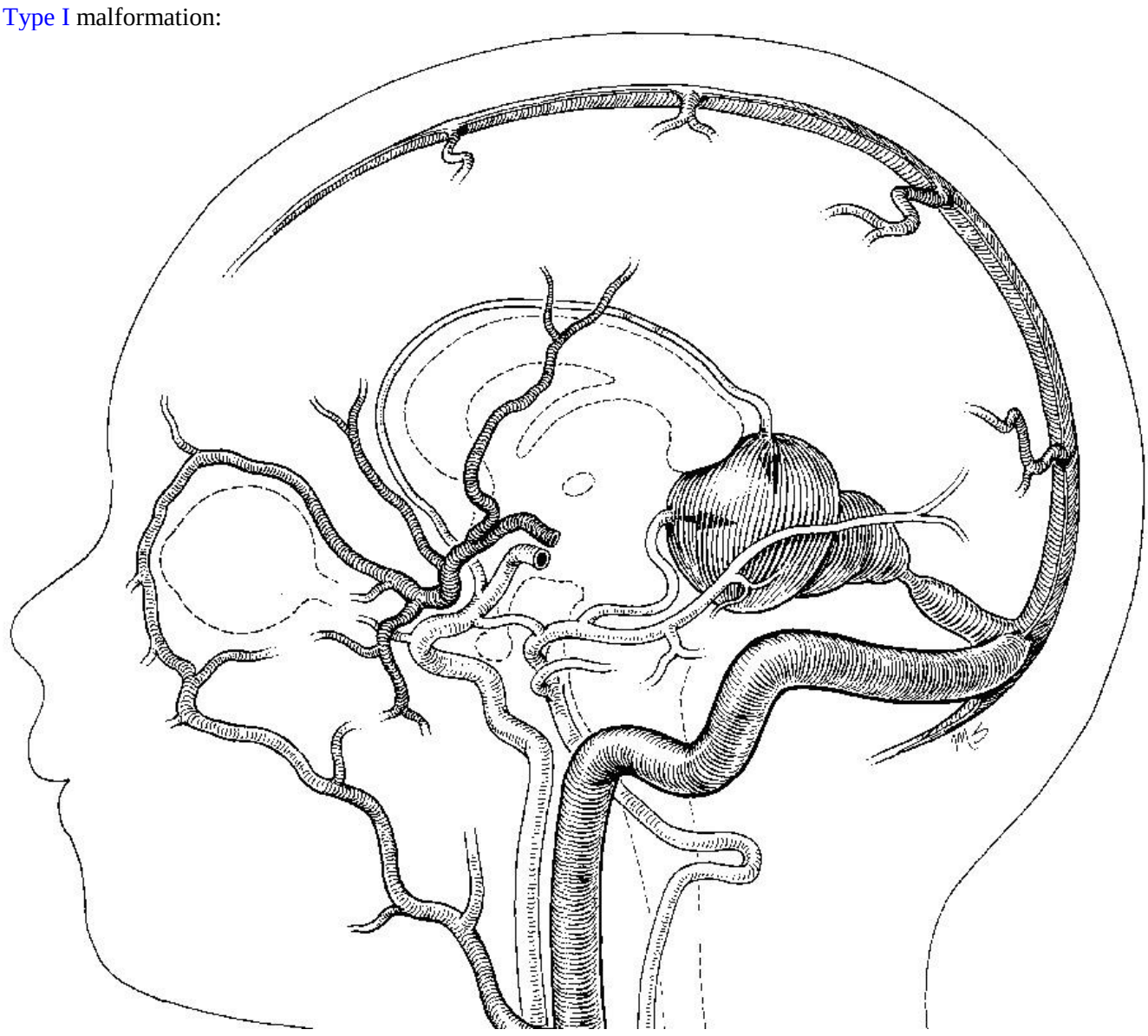
type I - fistulae located in varix wall arise from feeders from ***ACA*** (***pericallosal***) and / or PCA (***posterior choroidal***) arteries (i.e. pericallosal and choroidal arteries are sole supply to varix).

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).

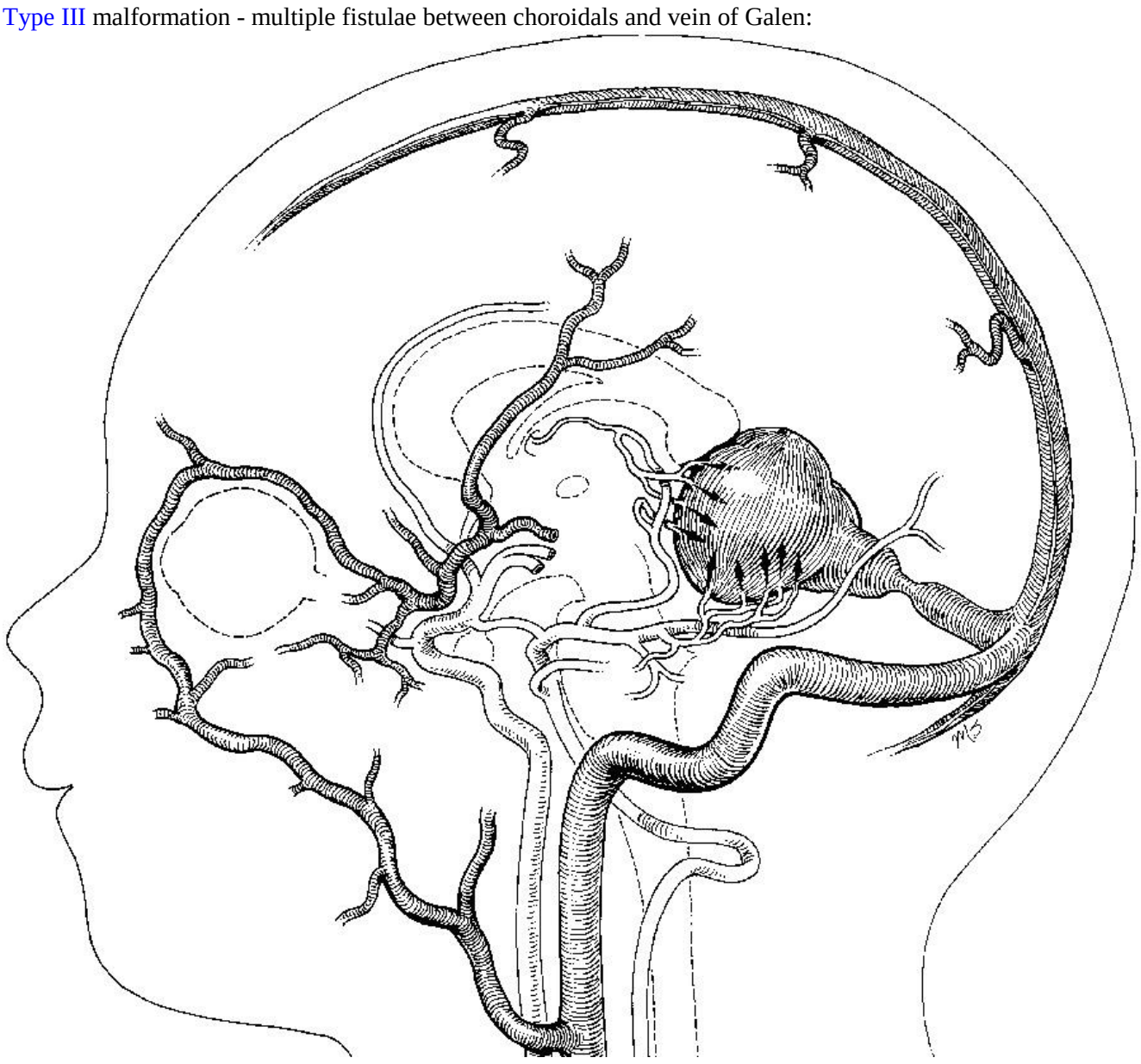
type III (most common type) - ***combination*** of types I and II.

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

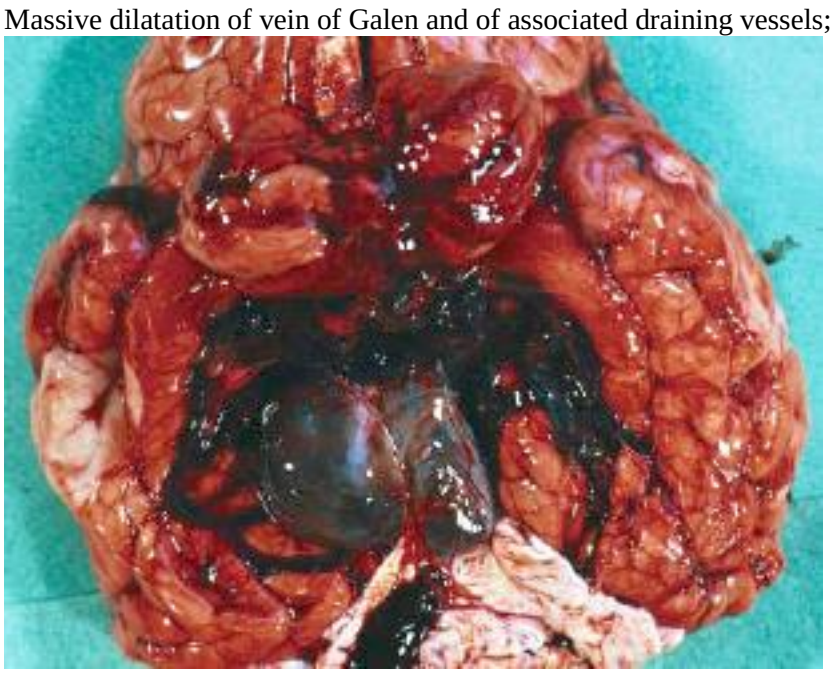
Type I malformation:



Type III 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).

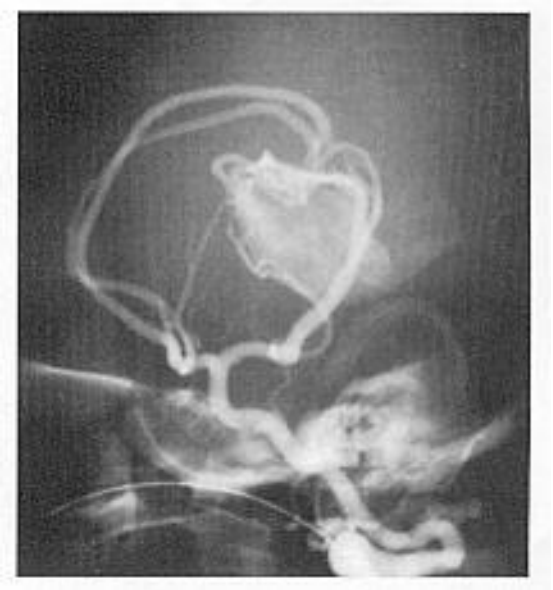
- tends to be progressive even in its most benign forms (frequently fatal!).

DIAGNOSIS

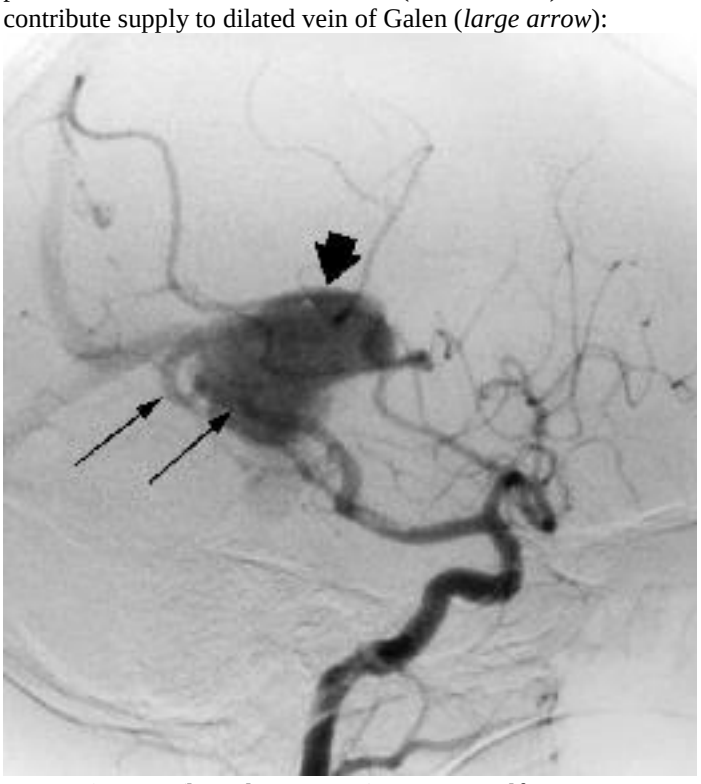
Doppler imaging in infants - detection of ectatic vein (hypervascular midline structure with demonstrable pulsations) → **MRI** / **angiography**.

- ***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).

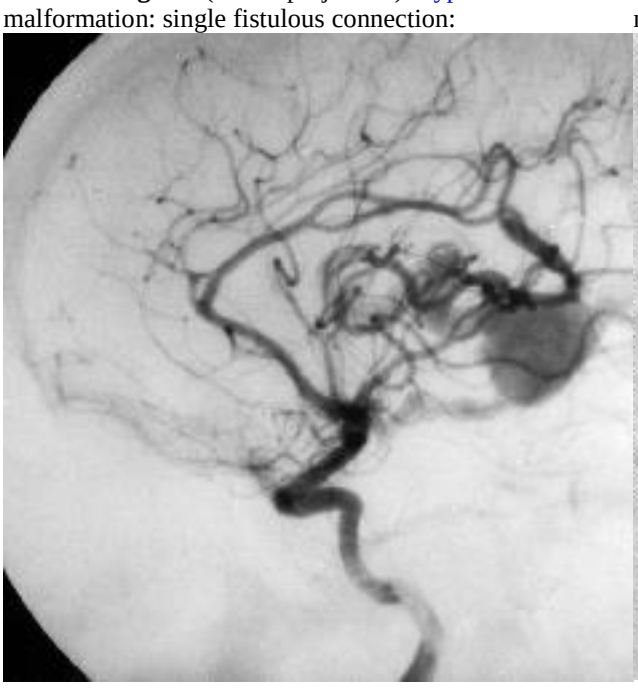
ICA arteriogram (lateral projection) - type I malformation: enlarged ACA and posterior choroidal artery entering nidus of vein of Galen malformation:



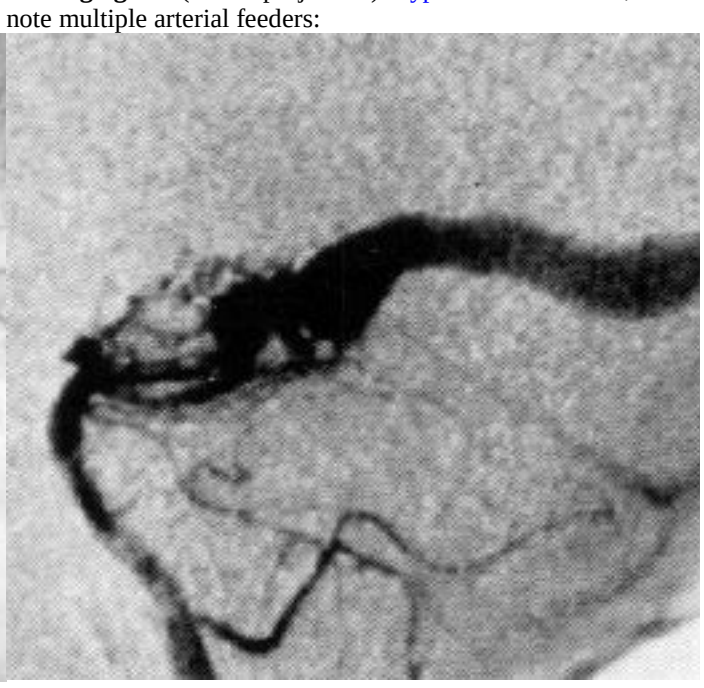
ICA angiogram (lateral projection) - type I malformation: posterior choroidal branches of PCA (*small arrows*) contribute supply to dilated vein of Galen (*large arrow*):



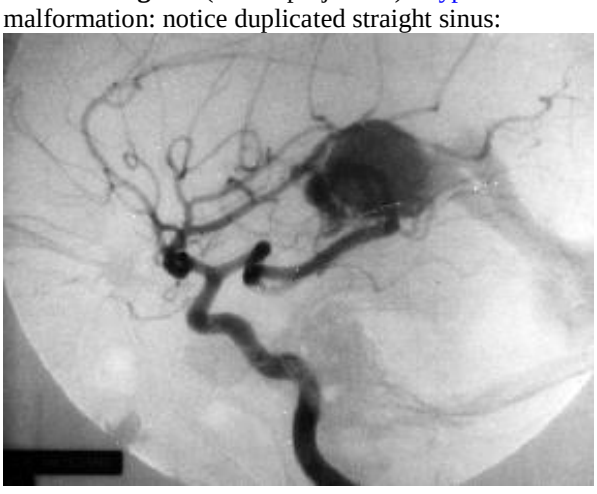
ICA arteriogram (lateral projection) - type I malformation: single fistulous connection:



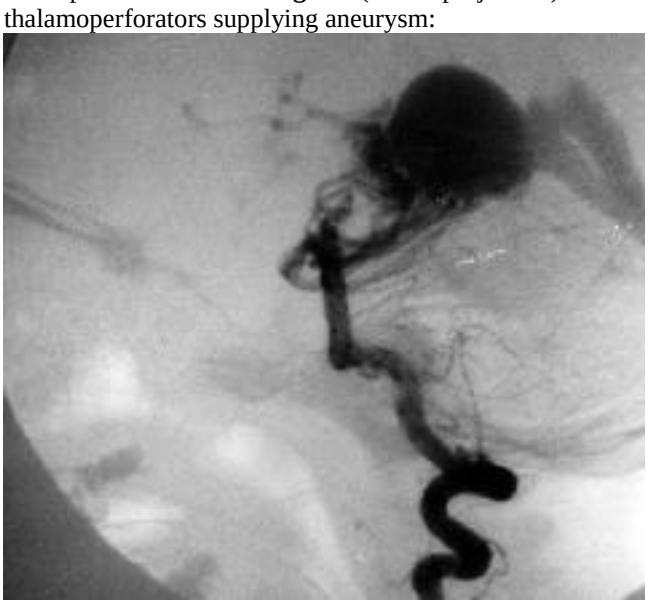
VA angiogram (lateral projection) - type II malformation; note multiple arterial feeders:



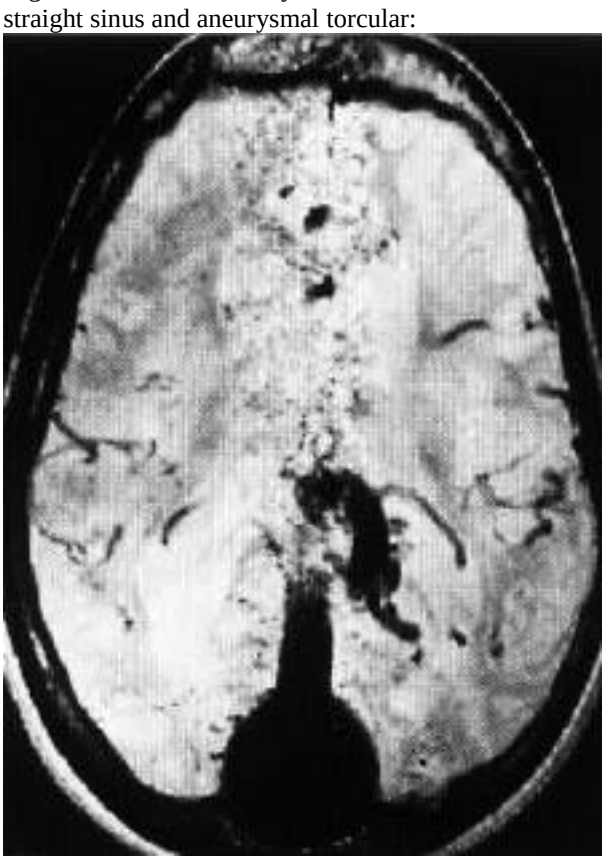
ICA arteriogram (lateral projection) – type III malformation: notice duplicated straight sinus:



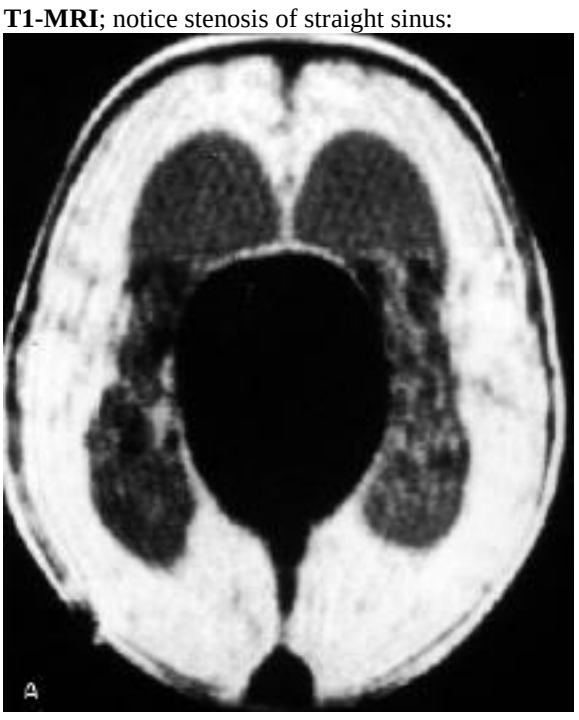
Same patient – **VA arteriogram** (lateral projection): thalamoperforators supplying aneurysm:



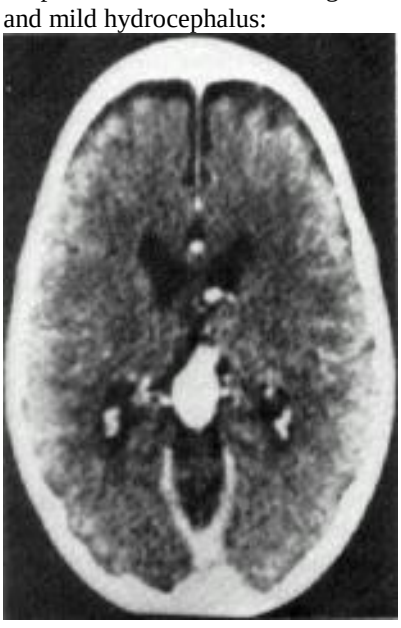
T1-MRI – type IV malformation: large complex midline angioma drains into aneurysmal vein of Galen, note dilated straight sinus and aneurysmal torcular:



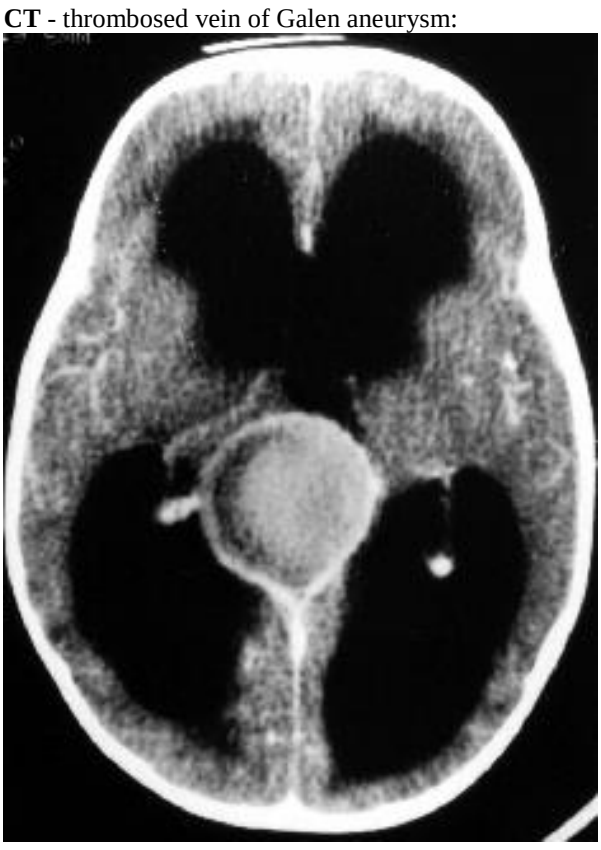
T1-MRI; notice stenosis of straight sinus:



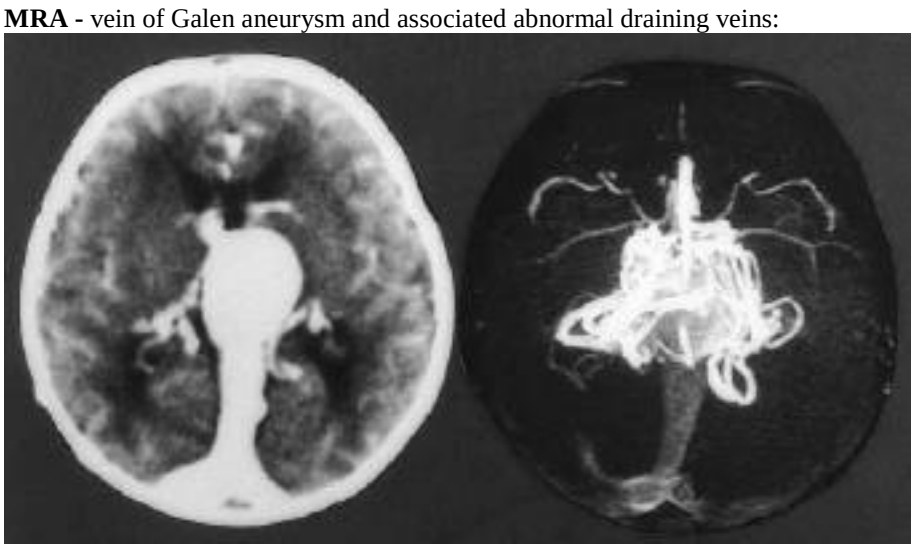
CT - aneurysmal dilatation of Galen vein owing to large, deep malformation; note enlargement of draining sinuses and mild hydrocephalus:



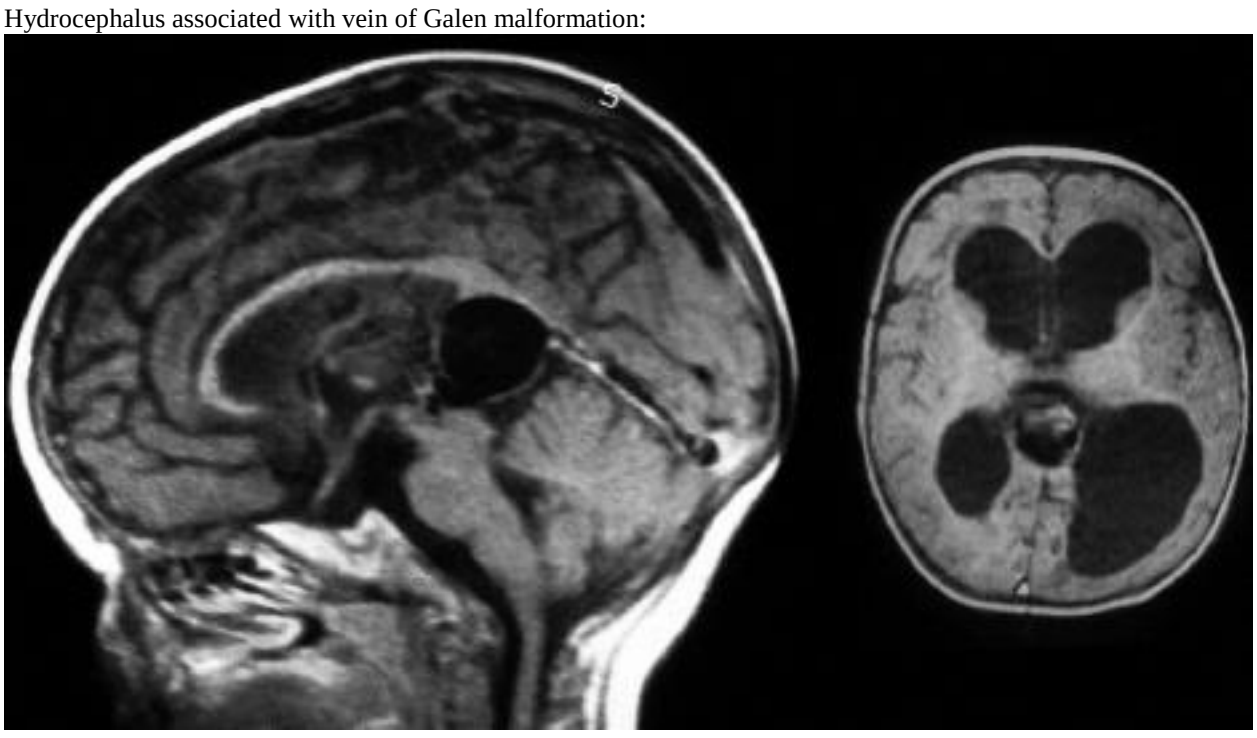
CT - thrombosed vein of Galen aneurysm:



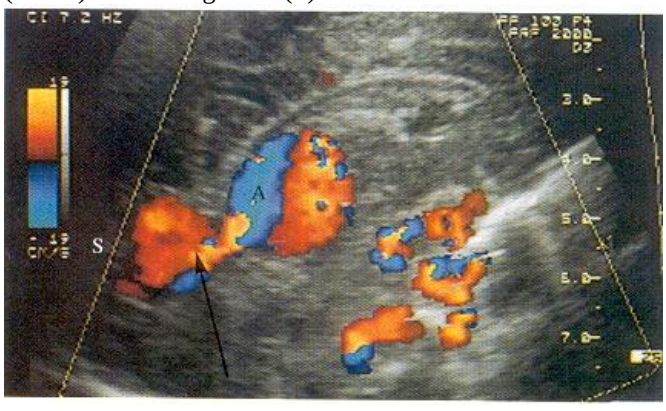
MRA - vein of Galen aneurysm and associated abnormal draining veins:



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):



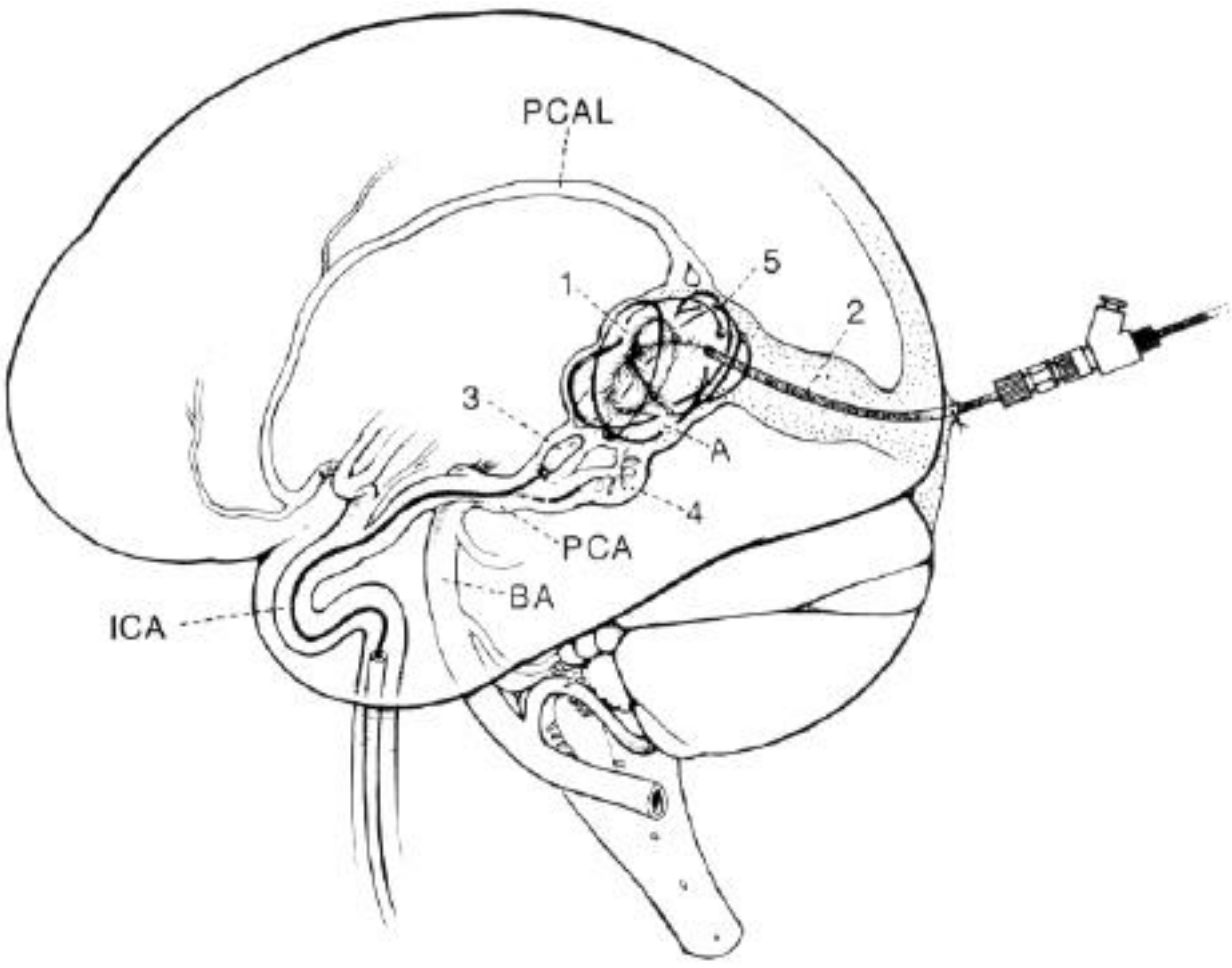
TREATMENT

- 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 (transtorcular 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):



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

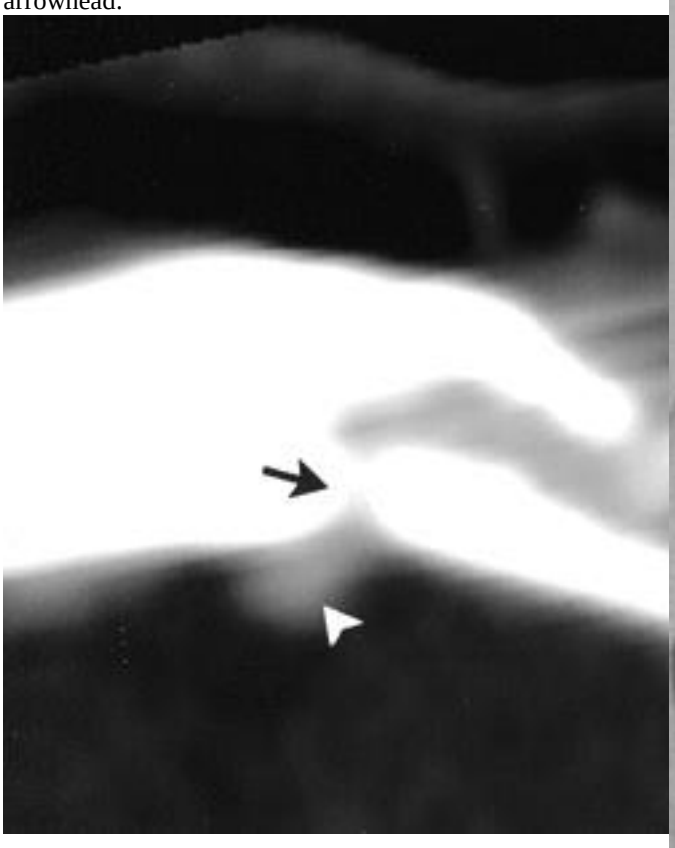
- high risk of hemorrhage! - treat in ***graded***, ***multisession fashion*** (2-4 treatments during first several days of life).
- preoperative correction of cardiac failure is critical.
  Cardiac failure unresponsive *to medical management* is indication for urgent embolization in neonatal period.
- 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

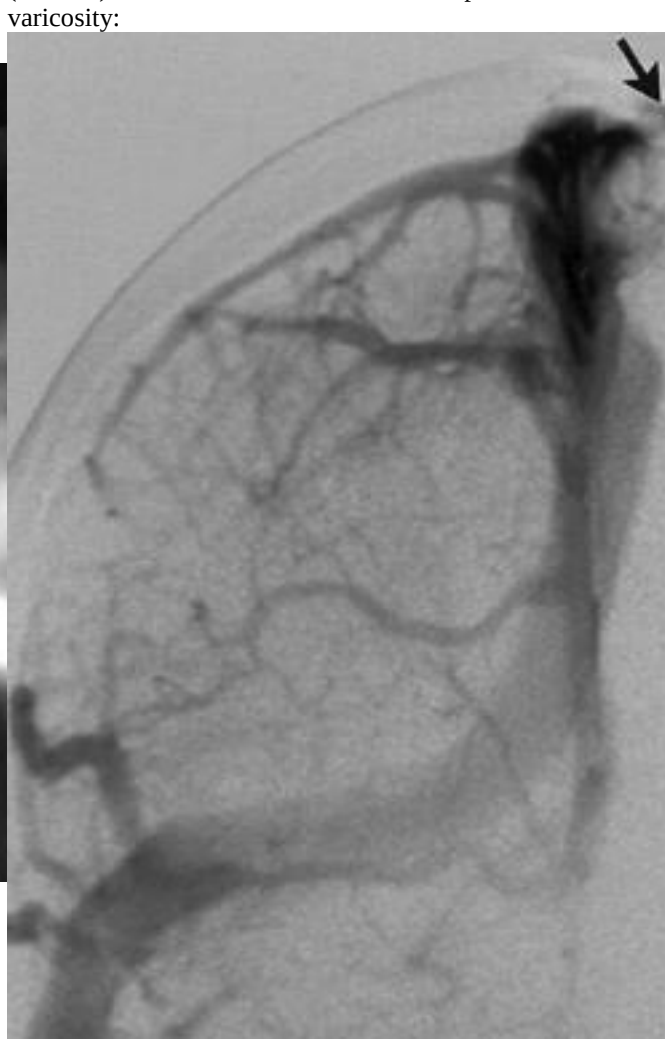
- thin-walled vascular spaces interconnected by numerous anastomoses that protrude from skull and communicate with superior sagittal sinus.

- EXTERNAL PROTUBERANCE - 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.

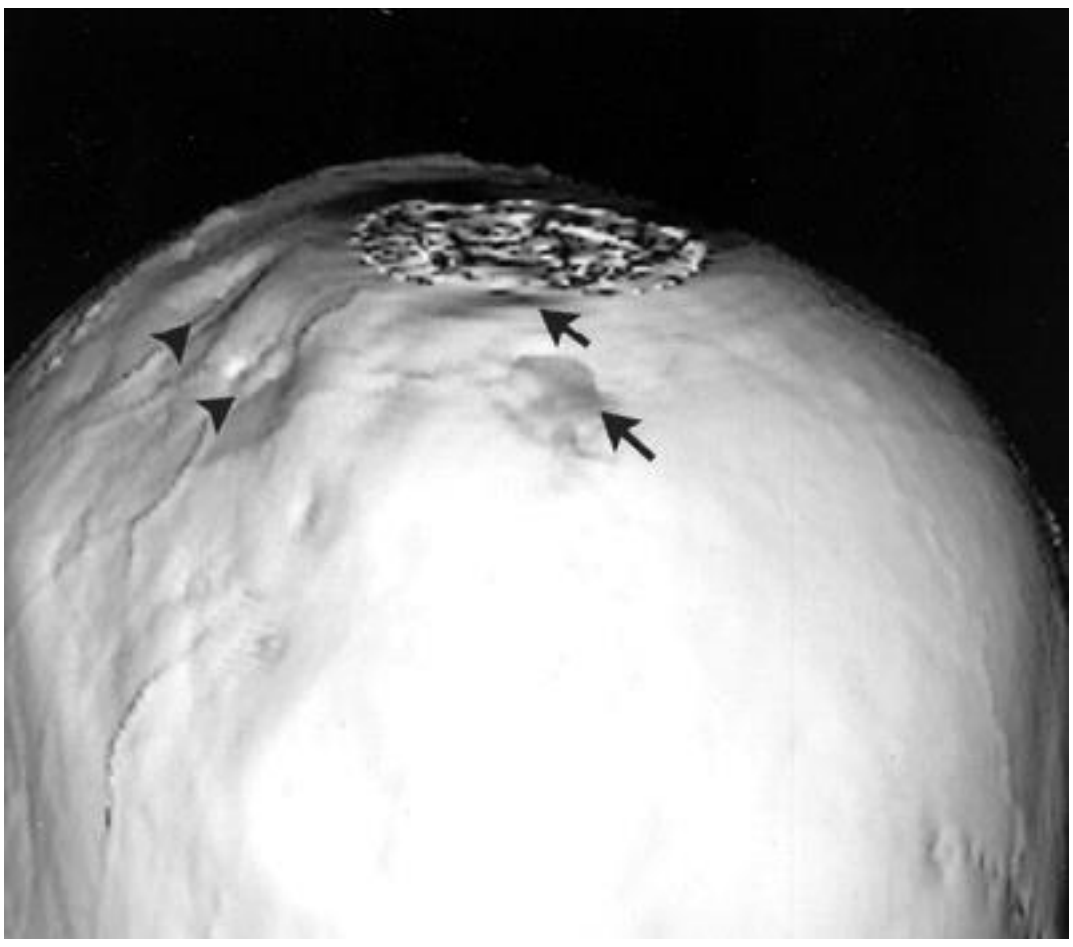
Reformatted oblique coronal **CT** through filling varicosity (*asterisk*) - defects within both inner and outer tables (*arrows*) of calvaria and filling of diploe with venous blood; connecting intracranial vein is denoted by white arrowhead:



Selective right **ICA angiogram**, AP view (venous phase) - venous connections (*arrow*) 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*):



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