CNS Arteriovenous Malformations: Classifications and Diagnosis

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Content layout

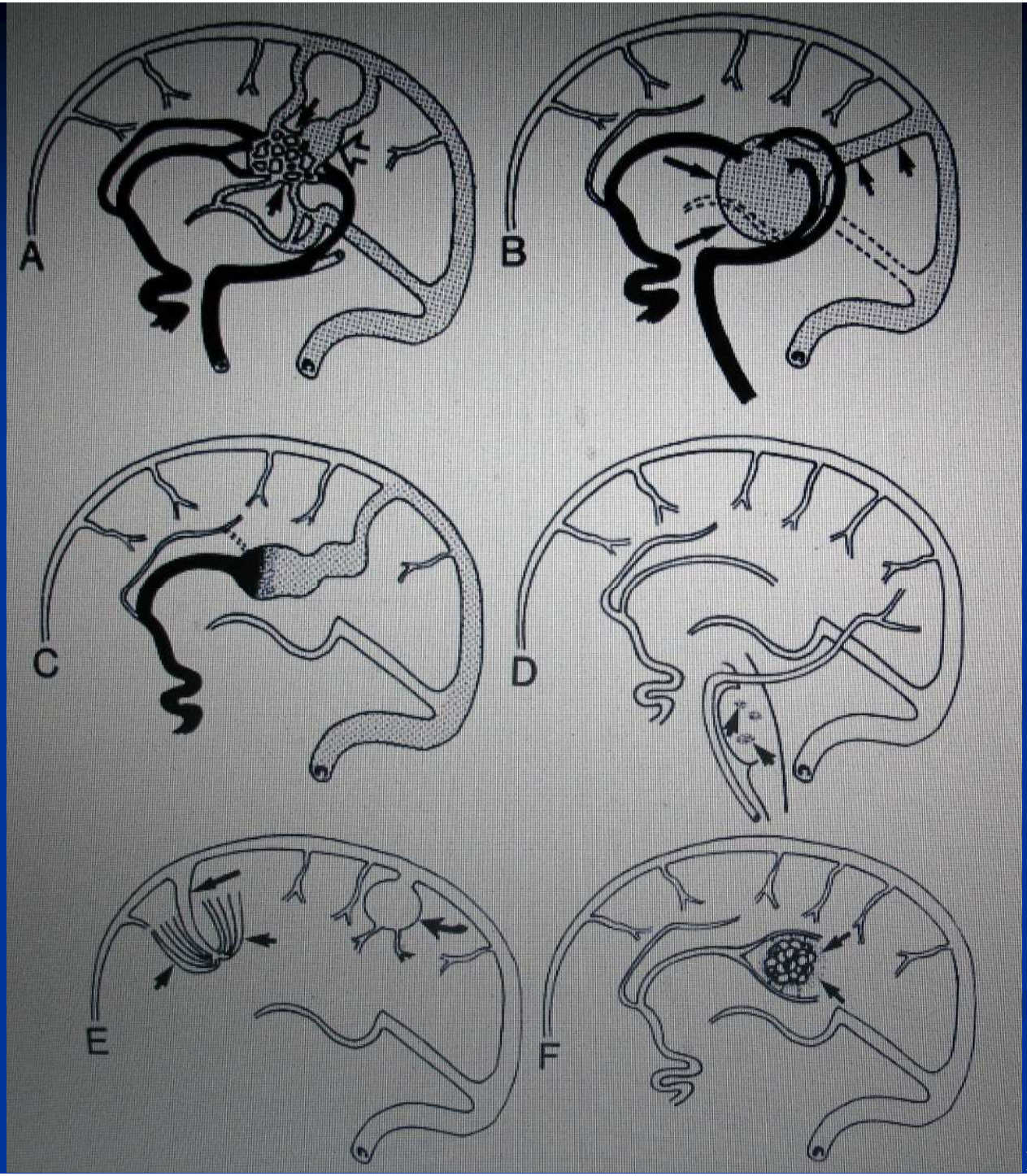
- Definition
- Epidemiology
- History
- Pathology
- Etiology
- Diagnosis
- Classification

Vascular malformation

- Mc Cormick (1966, modified 1978)
  - Arteriovenous malformation *(commonest in surgical series)*
  - Venous angioma *(commonest in autopsy series)*
  - Cavernous angioma
  - Capillary telangiectasia
  - Arteriovenous Fistula
    - Vein of Galen malformation
    - Dural AVM
    - Carotid cavernous fistula

Vascular malformation

- CVM with arteriovenous shunting
  - AVM
    - Plexiform nidus
    - Mixed (plexiform- fistulous) nidus
  - AVF
    - Single or multiple fistulae
    - Mono or multiple pedicular
    - CVM without AV shunt
- Capillary malformation
  - Venous malformation
  - Cavernous malformation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Definition

- Vascular abnormality constituted by a complex, tangled web of afferent arteries and draining veins linked by an abnormal dysplastic intervening capillary bed

History

1792	Hunter	Monograph on vascular malformation
1850s	Rokitansky	First described angiomas of intracranial cavity
1851	Virchow	Rudimentary classification of malformations
1895	D'Arcy Power/ Steinhill	Correlation of clinical features with location of AVM
1889	Pean	First attempt at surgical excision of vascular malformation
Late 1920s	Cushing/ Dandy	Described series of intracranial vascular malformations
1927	Egaz Moniz	Introduction of cerebral angiography
1936	Bergstrand	First angiographic diagnosis of cerebral AVM
1940s	Olivecrona	Series of angiographically documented AVM
1950s	Lussenhop	Attempted therapeutic embolisation of intracranial vessels
1960s	Yasargil/ Donaghy	Development of microvascular neurosurgery
1960s	Leksell	Clinical use of stereotactic focused beam radiation therapy
1990s		Elucidation of genomics of cavernous malformations

Epidemiology

- True incidence not known
- Unselected population: 1/ lakh / year
- Point prevalence in adults: 18/ lakh
- symptomatic lesions: 0.14- 0.8% per year
- 1/10th frequency of aneurysm

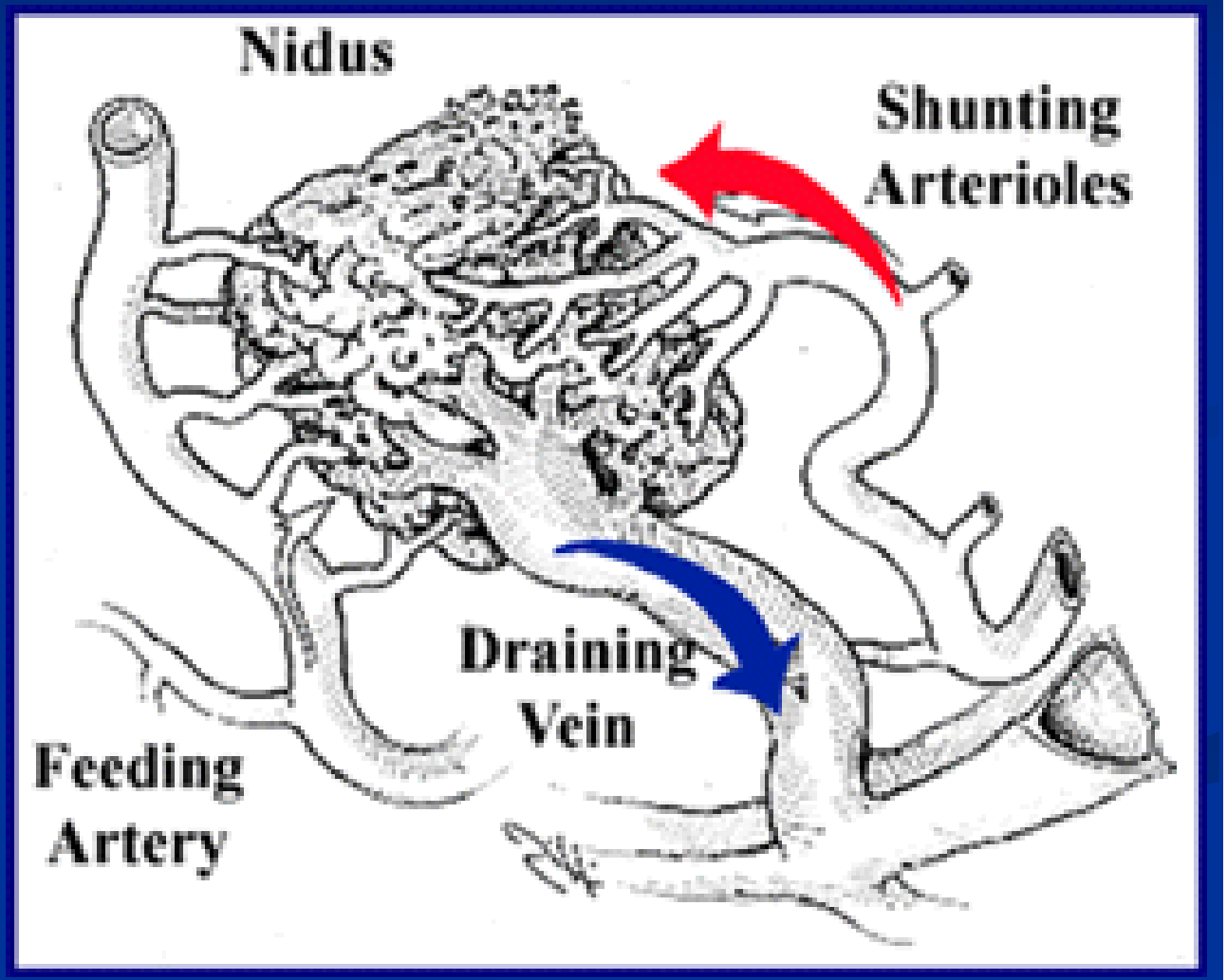
Epidemiology

- No gender preference
- Mean age at presentation: 30 to 40 years
- 15% asymptomatic (autopsy series)
- 3% stroke
- 33% primary intracerebral bleed in young adult

Pathology

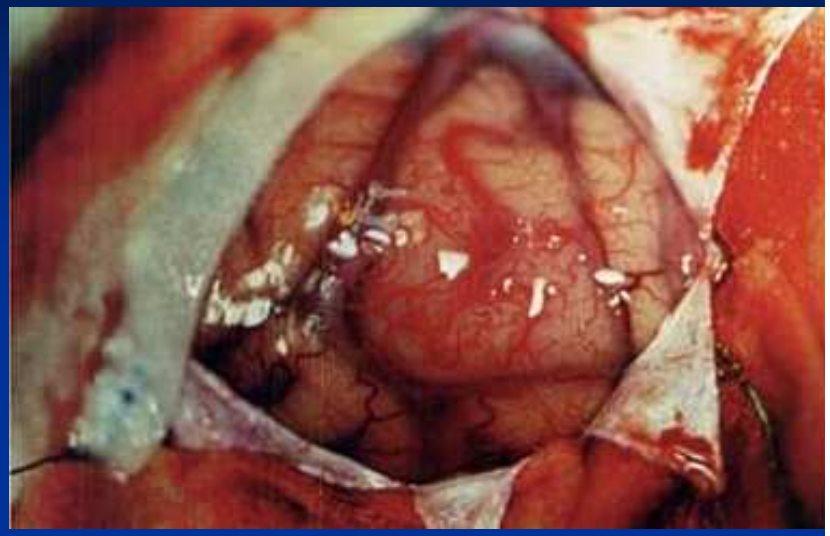
Angio-architecture

- Arterial feeder
- Nidus
- Draining vein
- Intervening gliotic neural parenchyma



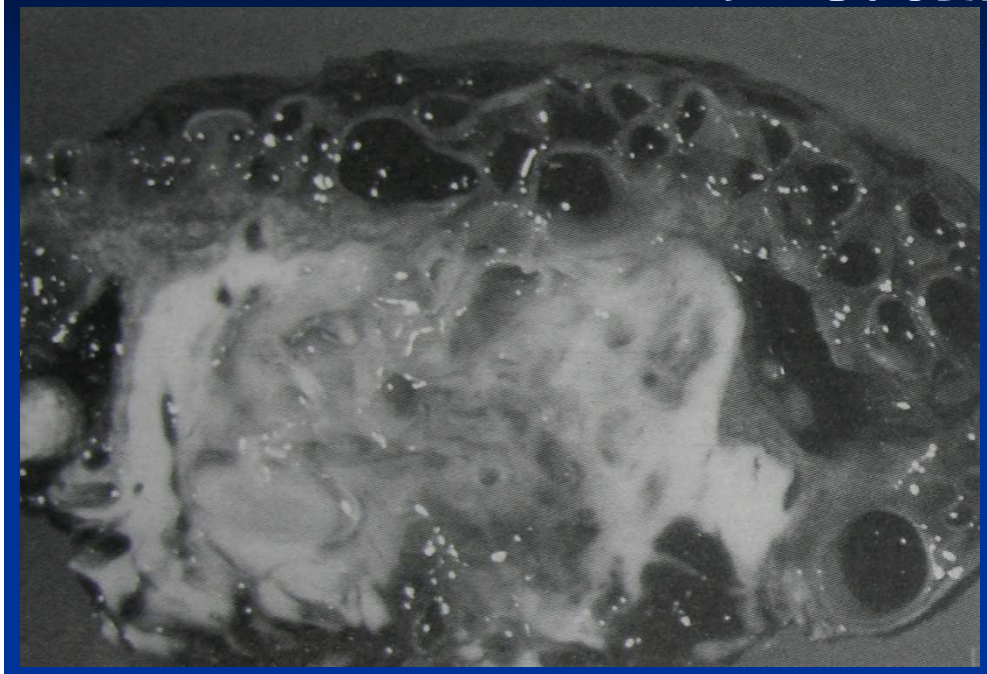
Pathology

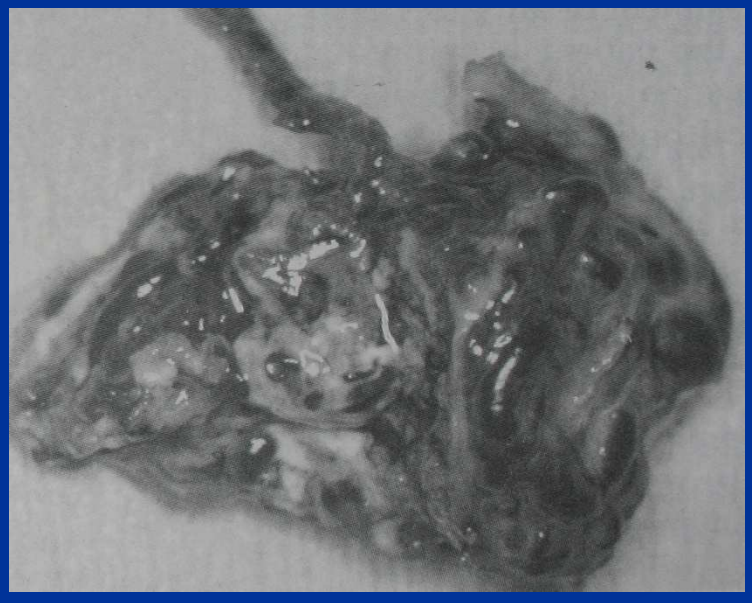
- Gross
  - leptomeninges- thick milky white
  - Gliotic wall: pseudocapsule
  - Arterialized cortical vein

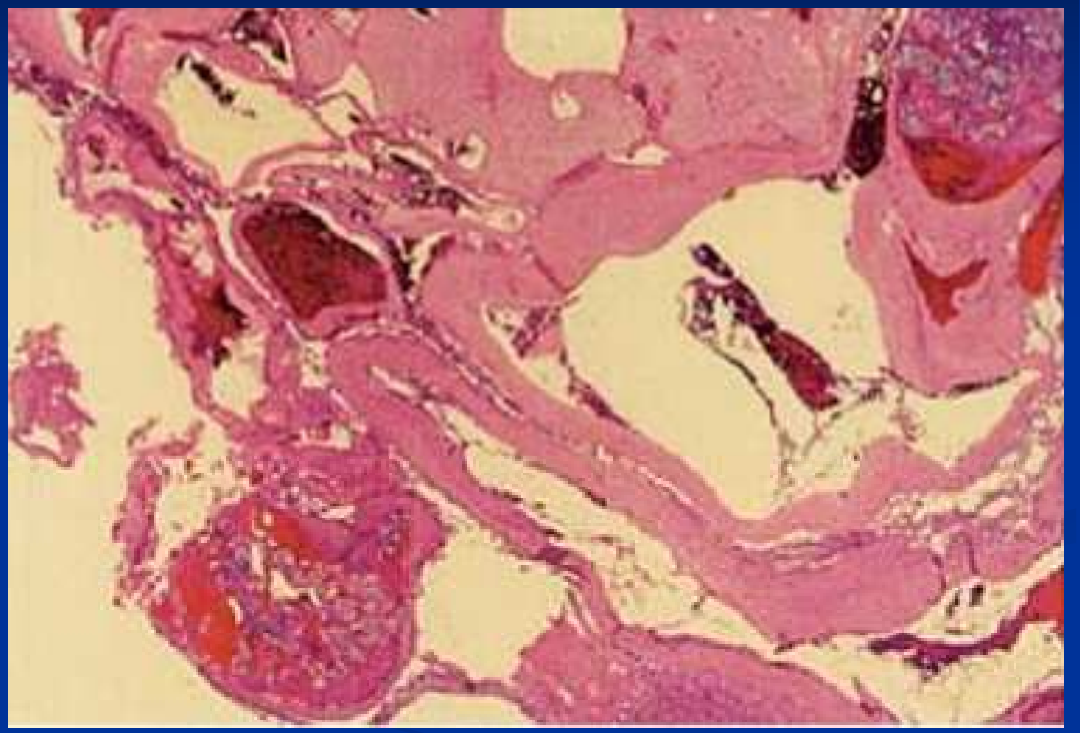


- Microscopic:
  - Arterial Media (Smooth muscle): hyaline degenerate- collagenous
  - Arterialised draining veins: smooth muscle & intima projects into lumen
  - Intervening gliotic parenchyma with hemosiderin laden microphages

Pathology







Multiple irregular thick and thin walled vessel

Pathology

Cavernous malformations	AVM	Venous malformations	Capillary telangiectases
Mulberry-like assembly of thin walled vascular sinusoids	Tangled, serpiginous mass of abnormally dilated vessels corresponding to arteries and veins	Single dilated or conglomerates of varicose veins drained by a single large vein	Dilated capillaries of differing calibre
Low flow dynamics	High flow dynamics		Occasionally petechial haemorrhage appearance
Single layer of endothelium, collagenous adventitia	Arteries with muscular and elastic laminae	Venous walls mostly normal or thickened by muscular hyperplasia and hyalinisation	Walls consist of basement membrane and endothelium
No smooth muscle, no elastic fibers	Veins "arterialised", thickened collagenous walls with increased cellularity due to proliferation of fibroblasts		No smooth muscle, no elastic fibres
No intervening brain parenchyma	Intervening gliomatous brain parenchyma within the interstices of the nidus	Normal intervening brain parenchyma	Normal intervening brain parenchyma
Peripheral rim of haemosiderin deposits	Haemosiderin pigmentation often present	No haemosiderin pigmentation	No haemosiderin pigmentation
Gliomatous reaction in surrounding parenchyma	Gliomatous reaction	No gliomatous reaction	Rarely gliomatous reaction
Hyalinisation, thrombosis, calcification, cholesterol crystals	Foci of calcification possible	Thrombosis possible	

Dr Amit Thapa; 5/9/2009

Pathology

- Arterial feeder
  - Single or multiple
  - Pial or perforating or dural

Terminal	May supply normal brain proximally Eventually end directly within nidus
Pseudo-terminal	Supply Normal brain distal to their supply to AVM nidus
Indirect	Terminate in AVM nidus, but arise typically at right angles form larger arteries that feed normal brain. “en passage” as too small to catheterize

Pathology

- Nidus or epicenter
  - Compact tangle of dysplastic, thin walled vessels of varied length connecting feeding arteries to draining veins
  - Arterial blood shunted directly into draining veins without passage through a normal high resistance arteriolar capillary network
  - Shape: Globular or conical
  - Compact or diffuse

Pathology

AVM associated Aneurysms

- About 2.3%-16.7% of time

Type I: unrelated dysplastic /incidental

Type II: flow related on proximal vessel

Type III: on distal small feeding vessel

Type IV: intra- nidal aneurysm

Pathology

- Haemodynamics
  - Blood flow velocity higher through AVM
  - Feeding artery and draining vein: progressively dilate and become tortuous
  - **Steal phenomenon:**
    - AVM like vascular sponges
    - deprives brain of normal circulation

Pathology

- Etiology
  - Congenital: aberrant vasculogenesis\ angiogenesis (embryo 40 to 89 mm length)
  - Developmental theory: failure of embryonic vascular plexus to fully differentiate and develop into a mature capillary bed in affected area
  - Fails to explain new AVM in angio negative patients
  - Combination of congenital predisposition and extrinsic factors

Pathology

AVM syndromes

- ***Sturge–Weber syn /*** encephalotrigeminal angiomatosis:
  facial angioma (CN V distribution) + I/L PO AVM
- ***Rendu–Osler–Weber syndrome*** (HHT): AD
  multiple visceral + mucosal + cerebral AVM
- ***Wyburn-Mason*** (*Bonnet-Blanc- Dechaume syndrome)* or mesencephalo-oculo-facial angiomatosis:
  U/L retinal angiomatosis
  cutaneous hemangioma (I/L CN V distribution)
  midbrain AVM

Pathology

- Natural history
  - Dynamic: continuous subtle anatomic/ hemodynamic changes
  - Clinically evident: host capacity to compensate saturates
  - Enlargement of brain AVM: < 30 yrs (especially childhood)
  - Spontaneous obliteration: rare (only 50 cases)
    - 84% single draining vein
    - 30% solitary arterial feeders
    - 50% Small nidus (<3cm)

Pathology

- Distribution
  - Based on autopsy findings

Cerebral hemisphere	60- 70%
Cerebellum	11- 18%
Brain stem	13- 16%
Deep seated	8- 9%

Presentation

1.Hemorrhage

2.Seizures

3.Headache

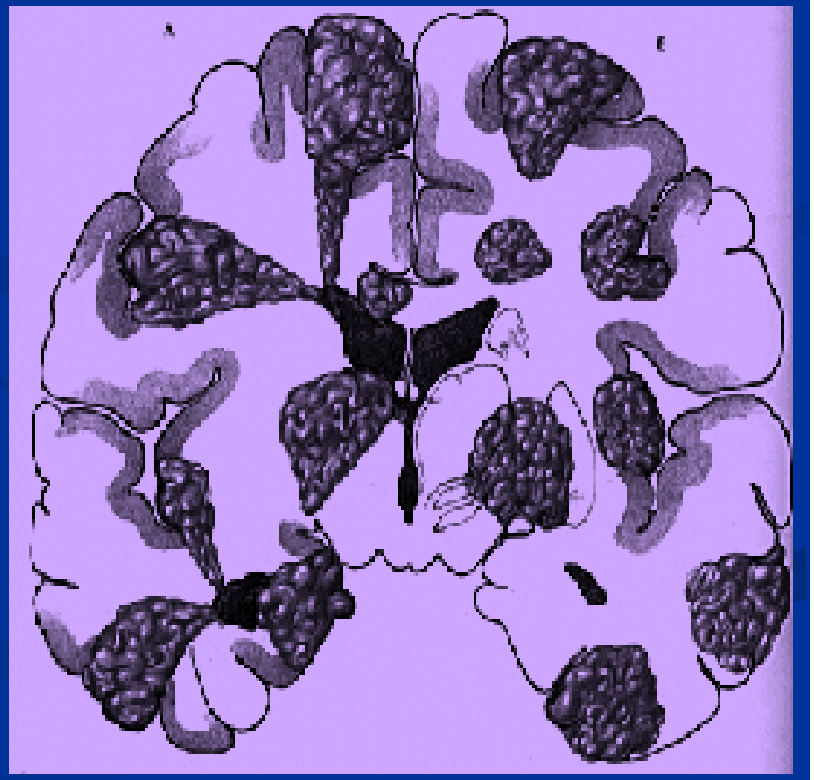
4.Neurological deficits

5.Asymptomatic

6.Pediatrics- hydrocephalus, heart failure

Presentation

- Hemorrhage
  - Commonest (32- 82%). Peak age: 15- 20 years
  - Parenchymal / Intraventricular/ SAH
  - Sudden onset headache
  - Vasospasm rare (1-2%)
  - Subpial location commonest
  - 10 % mortality
  - 30- 50% morbidity



Presentation

- Hemorrhage: Risk factors
  - Size of Nidus:

Small	< 3 cm	82%
Medium	3- 6 cm	29%
Large	> 6 cm	12%

  - Location: Deeper (basal ganglia, periventricular, IV, PF)
  - Intranidal aneurysm
  - Venous stasis
  - Draining vein: Single (89%), deep (94%), obstruction (94%)
  - Arterial stenosis/ angioectasia (decrease bleeding risk)

Presentation

- Hemorrhage: *Annual rate*
- First bleed: 2 to 4%/ year
- Recurrent bleed:

6% *Graf (JNS 1983; 58: 331)* to 18%/ 1st year *Fults (Neurosurgery 1984; 15: 658)*

Constant: 4%/ yr *Ondra (JNS 1966; 25: 467)*

- Calculating risk of bleed
  - Life time risk

    = 1- (risk of no hemorrhage) expected years of remaining life
  - Brown *et al*:

    risk= 105- age (in years)

Presentation

Unruptured cerebral AVM

Citation	No. of Cases	Bleeding (%)	Morbidity (%)	Mortality (%)	Average Follow-up (Years)
Perret and Nishioka, 1966 (97)	77	30	—	—	>20
Kelly et al., 1969 (49)	26	—	34.5	11.5	12.5
Forster et al., 1972 (27)	46	25	20	17	15
Graf et al., 1983 (31)	71	39	—	—	20
Fults and Kelly (28)	26	27	19	12	8.7
Brown et al., 1988 (11)	168	18	5	4	8.2
Ondra et al., 1990 (91)	46	4.2/yr	0.7/yr	0.9/yr	>20

Current scientific data do not tell us conclusively whether there is less chance of brain injury when an unbled AVM is eliminated or is left alone.

ARUBA trial – still undergoing

Presentation

- Seizures
  - Second most common (15- 35%)
  - ¾ patients during life
  - Cause
    - Overt bleed
    - Recurrent micro- hemorrhage (hemosiderin)
    - Secondary to venous hypertension
    - Ischemia following steal phenomenon
  - Majority: partial or complex partial
  - Controlled on medications

Presentation

- Headache
  - Chronic headache (initial symptom: 7 to 48%)
  - Occipital lobe AVM (fed by PCA)

    cause migraine like headache

Presentation

- Neurological Deficits
  - Focal (hemiparesis, aphasia)

    Without bleed (initial symptoms: 1 to 40%)
  - Cause:
    - Vascular steal
    - Mass effect
    - Venous hypertension
    - Peri-focal edema

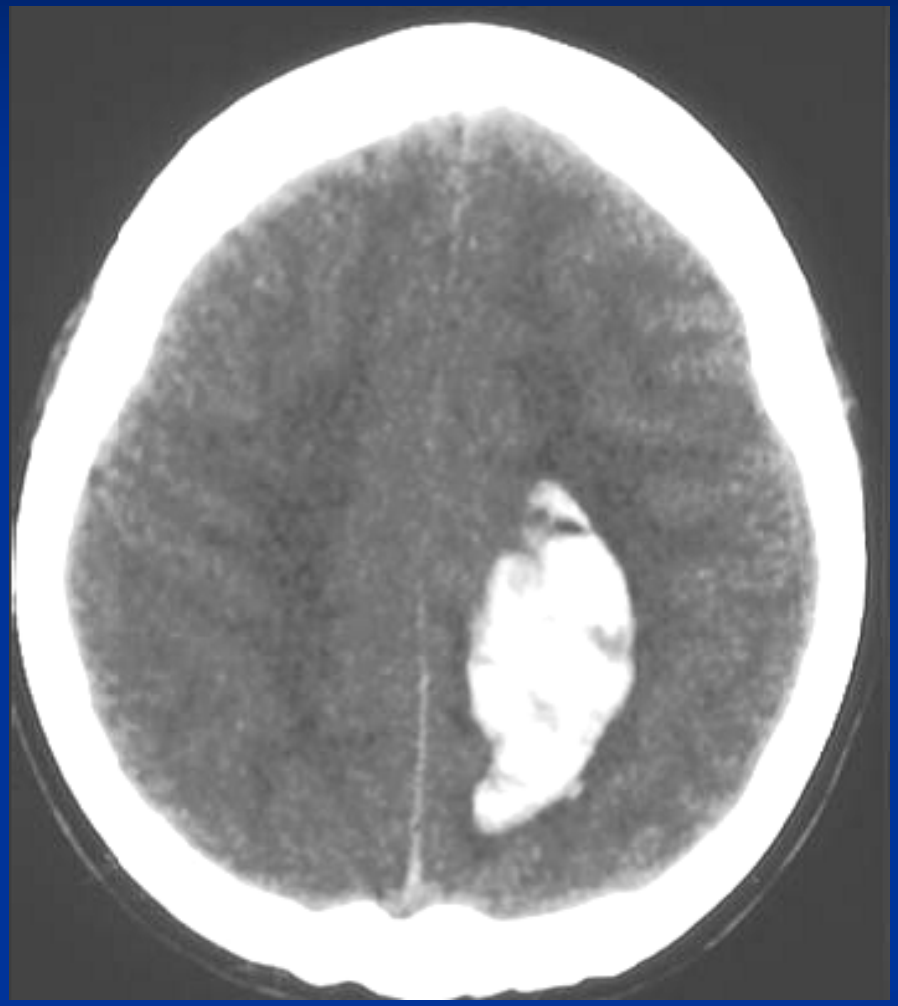
Investigations

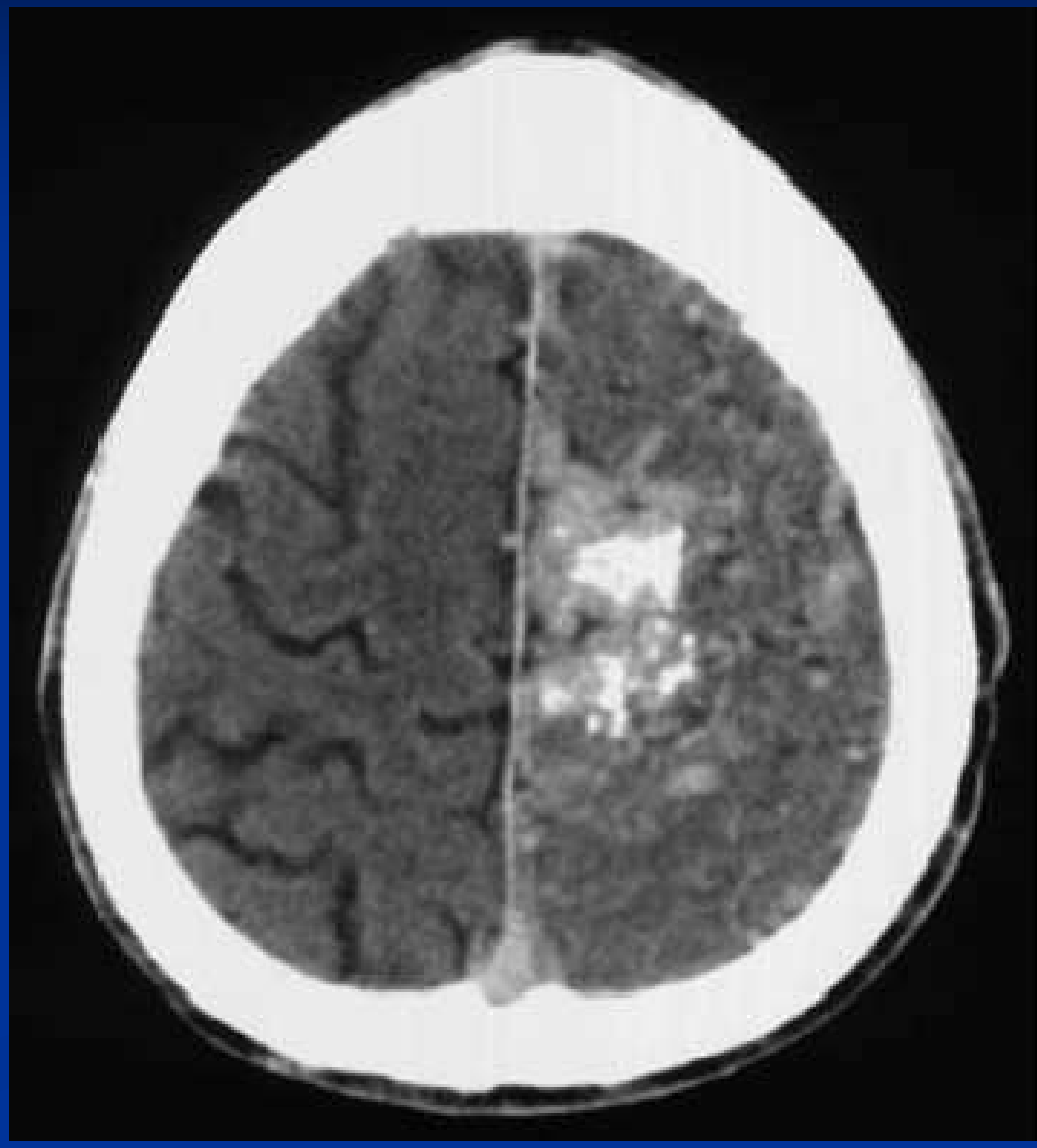
- Imaging
  - Establishes diagnosis in various clinical scenario
  - Pre treatment evaluation (decision making)
  - Treatment
  - Post therapeutic evaluation

Investigations

- Imaging (CT Scan)
  - First modality to rule out bleed
  - Suspect AVM if
    - Young patient
    - Lobar location
    - Hyperdense serpiginous structures (calcifications)
    - Nidus sparing sign : hypo-density within hematoma
    - CECT: serpiginous enhancing with early draining vein
  - Parenchymatous calcifications: 20%
    - Related: intravascular thrombosis or old hematoma

Imaging (CT Scan)





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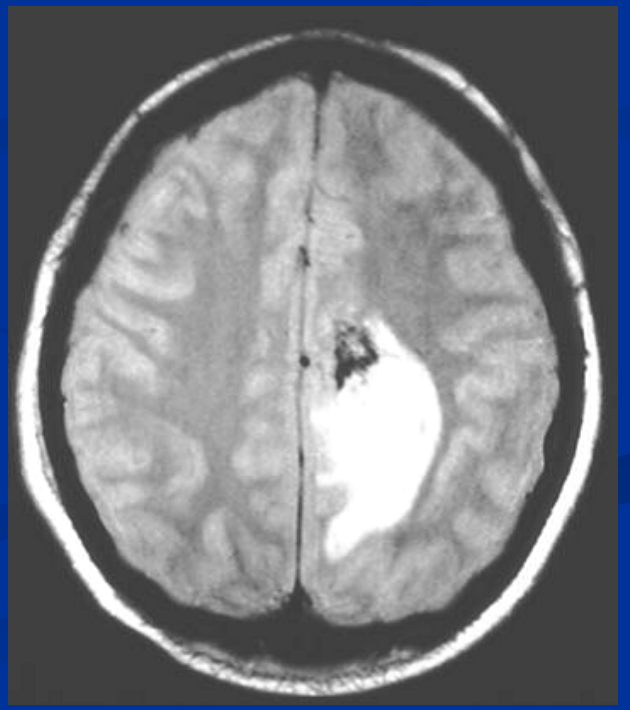
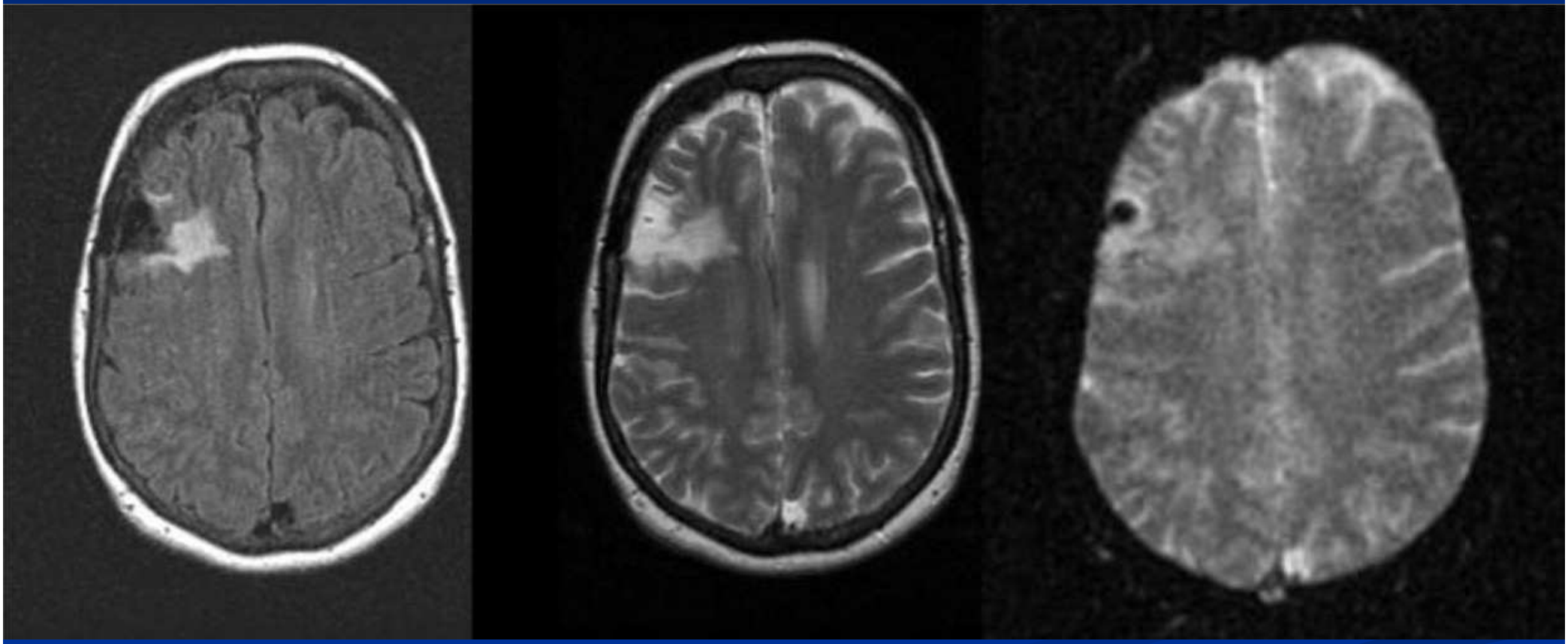
Investigations

- Imaging (MRI)
  - Unruptured AVM/ cause of lobar bleed
  - T1 / T2WI: flow void phenomenon
  - Gad T1WI: enhanced circulating vessels
  - Precise delineation of nidus
  - MRA (TOF): non invasive
  - Functional MRI: assist treatment



Investigations



Investigations

- Imaging (MRI)
  - D/D
    - Tumor with bleed: significant edema around lesion
    - AVM: complete ring of low density around lesion (hemosiderin)

Investigations

- Abnormal vessels foci + AV shunting – no D/D
- Parenchymal hematoma

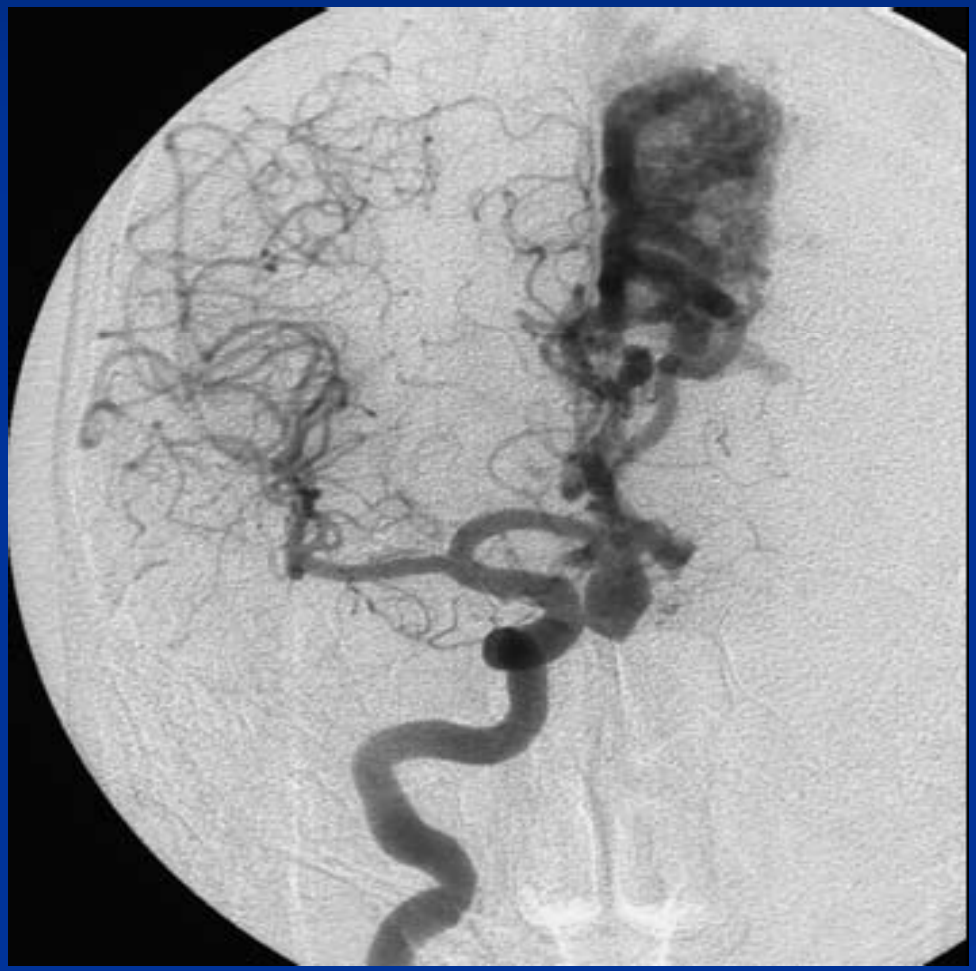
Hypertensive hemorrhage	H/O HTN, typical location
Cavernous malformation	Typical MRI, angiographically occult
Underlying neoplasm	Other brain lesions, suspicious enhancement
Amyloid angiopathy	Elderly, foci of hemosiderin T2WI or GE
Trauma	History, scalp injury
Dural sinus thrombosis	Occluded venous sinus
Mycotic aneurysm rupture	SAH, endocarditis

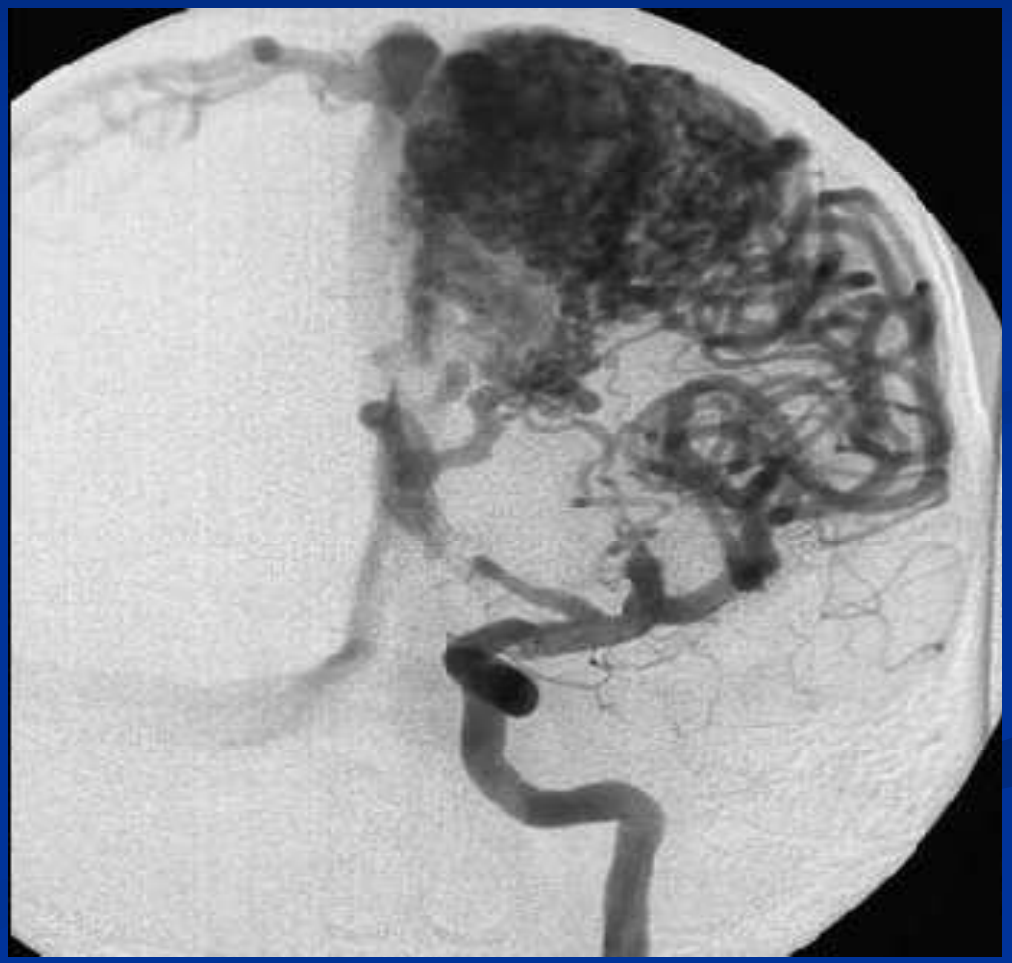
Investigations

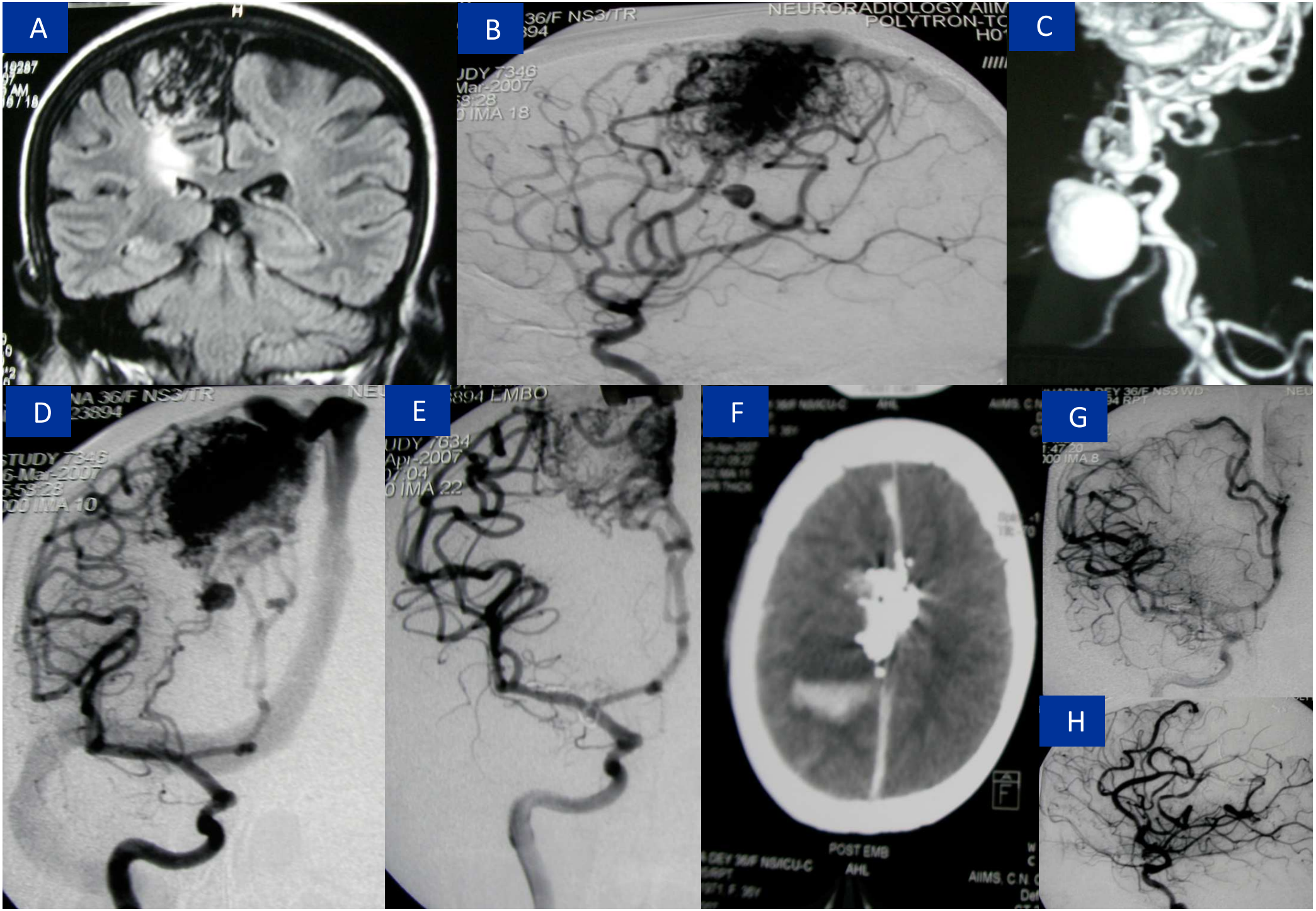
- Imaging (IADSA): **gold standard**
  - 4 vessel angiography still necessary
  - Exact and therapeutically relevant
  - Anatomic and functional information
    - Tangle of vessels
    - Large feeding artery
    - Large draining veins

Investigations

- Imaging (IADSA)



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POST EMB
AHL

AOVM

- Angiographically occult AVM
  - Truly occult: cavernoma, telangiectasia, thrombosed AVM
  - Transiently occult: destruction/ compression- hematoma / edema
  - Micro AVM
  - Intra-luminal thrombosis- stagnation / turbulence
  - Changes in blood vessels- fibrosis, spasm, dysplasia
  - Variable filling of AVM during angiography

Investigations

- CT Angiography- 3 D format, low acquisition time, cheaper
- Transcranial Doppler- neonate/ paed
- SPECT- hypoperfusion
- Functional PET- hypoperfusion

Investigations

- Complete blood count (CBC) with platelets:
  infection

  to identify hemorrhagic risk and complications.
- Prothrombin time (PT)/activated partial thromboplastin time (aPTT)
- Serum chemistries (electrolytes and osmolarity)
- Toxicology screen and serum alcohol level
- Screening for hematologic, infectious, and vasculitic etiologies

Classification

- Age and neurological status
- Size
- Location
- Configuration of nidus
- Arterial feeders: number, size, type, source
- Venous drainage
- Hemodynamic: shunt flow, steal,

Classification

- Types (presentation)
  - Cryptic (angio, surgery, pathology: absent)
  - Occult (angio & surgery: absent, pathology: seen)

- Types (AVM size)
  - Micro : < 1 cm
  - Small: < 2.5 cm
  - Moderate: 2.5 to 5 cm
  - Large : > 5 cm

Classification

- Types (location)
  - Dural
  - Parenchymal
    - Pial
    - Subcortical
    - Paraventricular
    - combined

Classification

- Luessenhop and Gennarelli (1977)
- Spetzler and Martin Grading (1986)
- Yasargil classification (1988)
- Valavanis classification (1996)
- Garretson
- Nataf Grading
- Vienna classification

Classification

- Spetzler Martin Grading system (1986)

Journal of neurosurgery 65:476,1986

Character		Points
Nidus (size in cm)	Small (<3 cm)	1
	Medium (3-6 cm)	2
	Large (> 6 cm)	3
Eloquent cortex	Yes	1
	No	0
Deep venous drainage	Yes	1
	No	0

AVM grade = sum (size + eloquence + deep component)

Classification

- Spetzler Martin Grading system
  - AVM grade:

    Surgical difficulty

    Risk of surgical morbidity and mortality

  5 grades
  - Low grade AVM: Grade I, II, III
  - High grade AVM: Grade IV, V
  - Inoperable: Grade VI

Classification

- Spetzler Martin Grading system

Risk of surgery *Spetzler and Martin 1986; Heros et al 1990*

Grade	Minor Deficits	Major Deficits	Favorable outcome
I	0	0	92- 100%
II	5%	0	95%
III	12%	4%	88%
IV	20%	7%	73%
V	19%	12%	57%

Classification

- Spetzler Martin Grading system (Drawbacks)
  - Definition of eloquence
  - Nidus compactness (Needs consideration)
  - Posterior fossa AVM: not useful
  - Difficulty in comparing modality other than surgery (Only applicable for surgical outcome)
  - No homogeneity in Grade III AVM

Classification

Shi-Chen Scale (1986)

J Neurosurg. 1986; 65: 484-489

Parameter	Finding	Grade
size in cm (diameter)	< 2.5 cm	1
	2.5 - 5.0 cm	2
	5.01 - 7.5 cm	3
	> 7.5 cm	4
location and depth	superficial and non-crucial area	1
	superficial and in functional area	2
	deep not involving a vital structure	3
	deep involving a vital structure of the brain (brainstem, diencephalon, etc.)	4
arterial blood supply	single superficial branch of the middle and/or anterior cerebral artery	1
	multiple superficial branches of the middle and/or anterior cerebral artery	2
	(branches of the posterior cerebral artery) or (deep branches of the middle cerebral artery) or (deep branches of the anterior cerebral artery) or (branches of the vertebral artery)	3
	(vertebrobasilar artery) or (main branches from all 3 cerebral arteries)	4
venous drainage	single, emptying into superficial dural sinuses	1
	multiple, all drainage into superficial dural sinuses	2
	(deep cerebral vein emptying into vein of Galen or straight sinus) or (superficial venous drainage)	3
	deep cerebral vein with huge dilatation or aneurysm-like structures	4

Classification

- **Shi-Chen Scale**

Pattern of 4 Grades	**Final Grade**	Operative Morbidity
all Grade 1	I	0%
3 Grade 1, 1 Grade 2	I-II	0%
2 Grade 1, 2 Grade 2	II	0%
1 Grade 3, 3 Grades 1 and/or 2	II-III	17%
2 Grade 3, 2 Grades 1 and/or 2	III	20%
<1 Grade 4, 3 others>	III-IV	80%
<2 or more Grade 4, others>	IV	

Classification

- Yasargil (1988)
  - Ask- Upmark concept (1938)- evolution of brain and its vascular system
- 2 groups
  - Convexity : supra and infratentorial
  - Deep central: supra and infratentorial
- Disadv:
  - Feeding arteries did not correlate with location so did not work for embolization

Classification

- Valavanis (1996)
  - Convexity or cortical AVM
    - Sulcal AVM
    - Gyral AVM
    - Mixed or sulco- gyral AVM
    - Diffuse cortical AVM
    - Subcortical AVM
  - Deep or central AVM
    - Subarachnoid AVM
    - Parenchymal AVM
    - Plexal or Intraventricular AVM
    - Mixed deep AVM

Classification

- Nataf Grading
  - 5 angiographic parameters: risk of bleeding

Grade	Description	Bleed
I	(No risk factor)	
I a	Venous recruitment	13%
I b	No venous recruitment	38%
II	Venous stenosis or venous reflux	48%
III	Deep venous drainage only	90%
IV	Intra or juxta nidal aneurysm	

Classification

- Vienna Classification
  - Similar to Spetzler Martin,
  - However useful in comparison/ endovascular feasibility

Grade	Feeders	Number of Feeders	Nidus size (cm)
0		1-2	<2
I	Pial	>2	2- 4
II	Pial + Perforator		>4
III	Perforator		

AIIMS series

- 1/1/2000 to 31/7/2008
- 868 patients evaluated for suspected AVM
- 790 had intracranial AVM
- 111 were surgically managed
- Presentations
  - Headache 70%
  - Loss of consciousness 12%
  - Seizure 32%
  - GCS $\leq$ 8: 12%

AIIMS series

Location	Frontal Temporal Parietal Cerebellar Brain stem	31% 14% 9% 13% 1%
Spetzler Martin Grade	I II III a III b IV V	14% 40% 9% 19% 16% 2%
Size of AVM	<3cm 3-6cm >6cm	46% 47% 7%
Location of bleed-	Lobar Intraventricular bleed SAH	55% 10% 7%
Associated findings on IADSA	Aneurysm Cavernoma	12% 1%

THANK YOU