CRANIAL AVM:
CLASSIFICATION AND
MANAGEMENT STRATEGIES

Presented By: GOPAL SEDAIN
Definition

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

- Contrast with an AVF which has direct fistulous connection without intervening nidus
Epidemiology

- Determining true incidence difficult
- Incidence around 1 per 1 lakh per year in unselected population
- Point prevalence in adults around 18 per lakh
- Most reliable estimate would be detection rate for symptomatic lesions (0.14%-0.8% per year)
- Roughly one-tenth of frequency of aneurysms.
Epidemiology

- No sexual preference
- Mean age of presentation 30 to 40 years
- At least 15% remain asymptomatic, based on autopsy series
- Account for 3% stroke and 33% primary intra cerebral bleed in young adults
Angioarchitecture

Composed of:

a) Arterial feeder
b) Nidus
c) Draining vein
d) Intervening gliotic neural parenchyma
Arterial feeder (s)

- Single/multiple
- Pial /perforating /dural
- Direct feeders supply as terminal branch
- Indirect feeders supply *en passage*
The AVM nidus is a compact tangle of dysplastic, thin-walled vessels of varied length connecting feeding arteries to draining veins.

An AVM nidus can either be globular or conical in shape and may be compact or diffuse.

Within the nidus, arterial blood is shunted directly into draining veins without passage through a normal, high-resistance arteriolar–capillary network.
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
Hemodynamics

- The velocity of blood flow is considerably higher through AVMs than through normal brain parenchyma.
- As a result of the abnormal hemodynamic condition, feeding arteries and draining veins become progressively dilated and tortuous.
- AVMs could be compared to vascular “sponges”, which consume large volumes of blood, depriving the brain of normal circulation (steal phenomenon)
Etiology

- Initially thought to be congenital
- May be due to aberrant vasculogenesis and angiogenesis in 40-89mm length embryo
- Failure of the embryonic vascular plexus to fully differentiate and develop a mature capillary bed in the affected area
- Developmental theory has been challenged.
- New AVM in angiography negative patients have been found
- It is likely that a combination of congenital predisposition and extrinsic factors lead to their generation
AVM syndromes

- **Sturge-Weber syndrome** or encephalotrigeminal angiomatosis a cutaneous, facial angioma in the distribution of the trigeminal nerve, and an ipsilateral, parietal–occipital vascular malformation.

- **Rendu-Osler-Weber syndrome** (hereditary hemorrhagic telangiectasia) is an autosomal-dominant syndrome of multiple visceral, mucosal and cerebral vascular malformations.

- **Wyburn-Mason** (Bonnet-Blanc-Dechaume syndrome) or mesencephalo-oculo-facial angiomatosis corresponds to the association of unilateral retinal angiomatosis and a cutaneous hemangioma in an ipsilateral trigeminal distribution with an AVM located in the midbrain.
Natural history

- AVMs are dynamic, i.e., they undergo continuous subtle anatomic and hemodynamic changes.
- A cerebral AVM becomes clinically evident when the host’s capacity to effectively compensate has reached its threshold.
- Enlargement of brain AVMs is observed in young patients (under 30 years of age), and especially in childhood.
Natural history

• Spontaneous obliteration of cerebral AVMs is rare; only 50 cases have been reported in the literature.

• Several factors appear to be associated with spontaneous occlusion of cerebral AVM:
  - single draining vein (84% of cases of occlusion),
  - solitary arterial feeder (30%),
  - small size of the nidus (<3 cm in 50%)
Based on autopsy findings:

- **Cerebral hemisphere** – 60 to 70%
- **Cerebellum** - 11 to 18%
- **Brainstem** - 13 to 16%
- **Deep seated** - 8 to 9%
Presentation

1. Hemorrhage
2. Seizures
3. Headache
4. Neurological deficits
5. Asymptomatic
6. Pediatrics- hydrocephalus, heart failure
Hemorrhage

- Most common (32-82%)
- May be parenchymal, intraventricular or SAH
- Patient presents with sudden onset severe headache
- Vasospasm is rare (around 1-2%)
- Most AVMs are located subpially.
Factors Increasing the Risk of Bleeding

Nidus

82% **small** AVMs (less than 3 cm)
29% medium-sized AVMs (3–6 cm)
12% large AVM (greater than 6 cm)

Large AVMs can more often present in other ways than hemorrhage (seizures, deficits, headache) and may lead to an overestimate
Location

- Some authors suggest that AVMs in deep locations such as in the basal ganglia or in the periventricular or intraventricular space, have an increased risk of bleeding.

- Can be partly explained as they are unlikely to have other presentation.
Other factors

- *Deep Venous Drainage*
- *Venous Stenosis*
- *Single draining vein*
- *Feeding Artery Pressures*
- Arterial stenosis and arterial angioectasia decrease bleeding risk
Annual rate of bleeding

• For 1\textsuperscript{st} hemorrhage is 2-4% per year
• Recurrent hemorrhage-6 to 18% in 1\textsuperscript{st} year declines to pre hemorrhage rates over 5 years
• Life time risk of hemorrhage = 1- (risk of no hemorrhage) expected years of remaining life
• E.g. if expected years of living is 40yrs, AVM bleed risk is 3% per year, i.e
  1-(0.97)^{40}= 70%
  Simple equation given by Brown et al is risk = 105-age in years
Seizures

2nd most common (15-35%) clinical presentation of a cerebral AVM

Nearly three-quarters of patients experience a seizure at some point during their life.

AVM-related seizures may be caused by:

a) overt intracranial hemorrhage
b) from hemosiderin deposition following recurrent micro-hemorrhages
c) secondary to venous hypertension
d) ischemia following steal

In majority of cases, seizures are partial or complex partial and controlled on medications
Headaches

- **Chronic** headache is the initial symptom in 7%–48%.

- Migraine-like headaches commonly are associated with occipital lobe AVMs fed by the posterior cerebral artery,
Neurological deficits

• Focal neurologic deficits such as hemiparesis, aphasia without hemorrhage are the initial symptom in 1%–40% of patients

• May be due to: vascular steal, mass effect, venous hypertension or peri focal edema
Types and size of AVM

- Cryptic (absent on angio/surgery/pathology)
- Occult (absent on angio /surgery/seen on pathology examination)
- Micro-AVM (<1 cm)
- Small (<2.5 cm)
- Moderate (2.5-5 cm)
- Large (>5 cm)
Imaging

Imaging has several roles and goals:

1. To establish the diagnosis of brain AVM in various clinical situations
2. To make a pre therapeutic evaluation of the AVM to help in decision-making
3. To treat the AVM as a sole therapy or in association with surgery or radio surgery
4. To perform post-therapeutic evaluation
CT Scan

- CT is usually the first imaging modality used to rule out hemorrhage.
- AVM is suspected in young patients, if hematoma has lobar topography and if hyperdense serpigenous structures (calcifications) are seen.
- Parenchymatous calcifications are observed in 20% of cases, related to intravascular thrombosis or evolution of an old hematoma.
MRI

- MRI is currently used in case of unruptured AVM or to find the underlying lesion in case of lobar hematoma days or weeks after the bleeding.
- On T1- and T2-weighted images, circulating vessels have no signal because of the flow void phenomenon. On T1-weighted images with gadolinium, vessels are enhanced.
- The size and the anatomic location of the nidus are precisely delineated by MRI.
MRI

- Magnetic resonance angiograms (MRAs) can reveal the anatomy of the AVM non invasively.
- Functional MRI can assist in treatment planning by defining the functionality of adjacent brain.
Angiogram

• Selective angiography is still always necessary to make a decision regarding the treatment.

• The exact and therapeutically relevant anatomic and functional information still has to be obtained by angiography.
IADSA
Classification

A) Luessenhop-gennarelli (1977)
B) Spetzler- Martin (1986)
C) Garretson
D) Nataf
E) Vienna classification
Spetzler- Martin grading

Size of nidus

Small (<3 cm) 1
Medium (3–6 cm) 2
Large (>6 cm) 3

Cortical Eloquence (sensorimotor, language, visual, thalamus, hypothalamus, internal capsule, brain stem, cerebellar peduncles, and deep cerebellar nuclei)

Yes 1
No 0

Deep venous drainage

Yes 1
No 0

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

Spetzler- Martin grading

- The Spetzler and Martin (1986) classification was established to grade AVMs according to their degree of surgical difficulty and the risk of surgical morbidity and mortality.
- There are 5 grades:
  - Low grade AVM: Grade I, II, III
  - High grade AVM: Grade IV, Grade V
  - Inoperable lesions: Grade VI
**Drawbacks of S-M grading**

- Definition of eloquence
- Lack of consideration for nidus compactness
- Not useful for posterior fossa AVM
- Only applicable for surgical outcome
- Grade III in homogeneity
Nataf grading

Five angiographic parameters were considered to be determinants of the bleeding risk, leading to a four-grade classification.

Grade I: no risk factor
   Ia : with venous recruitment
   Ib : without venous recruitment

Grade II: venous stenosis or venous reflux

Grade III: deep venous drainage only

Grade IV: intra- or juxta nidal aneurysm

In the series mentioned, there were 13% of hemorrhages in grade Ia, 38% in grade Ib, 48% in grade II and 90% in grade III and IV.
# Vienna classification

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<tr>
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<th>Gr 0</th>
<th>Gr I</th>
<th>Gr II</th>
<th>Gr III</th>
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<tbody>
<tr>
<td>Feeders</td>
<td></td>
<td>Pial</td>
<td>Pial+perf.</td>
<td>Perf.</td>
</tr>
<tr>
<td>No.of feeders</td>
<td>1–2</td>
<td>&gt;2</td>
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<tr>
<td>Nidus (cm)</td>
<td>&lt;2</td>
<td>2–4</td>
<td>&gt; 4</td>
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The algorithm is similar to that of Spetzler and Martin for use in arteriovenous malformation surgery, allowing the comparison of surgical and endovascular feasibility.
Management

- AVM
  - microsurgery
  - embolization
  - radiosurgery
Microsurgery

• Microsurgical AVM excision is the most effective treatment
• Not every AVM is amenable to or best treated with surgery
• Factors associated with increased surgical risk are large size, deep venous drainage, deep location, diffuse nidus, feeders from deep perforating system.
Microsurgical principles

- Surgical AVM excision should be an elective procedure, even in patients with ruptured AVMs.
- Pre-op steroids, anticonvulsants and antibiotics need to be given.
- Wide craniotomy and dural opening: When the nidus is below the surface, an arterialized draining vein (red vein) can be followed to the AVM.
- Localization may be assisted with ultrasound or frameless stereotaxy.
• Careful dural opening
• Circumferential nidus dissection layer by layer
• At least one major draining vein should be preserved till end of dissection
• At end of dissection this vein becomes bluish proving that feeders have been eliminated
• Complete AVM excision should be documented by post op angiogram.
Surgical complications

- Intra op rupture
- Post retraction edema
- New onset seizures
- Retrograde thrombosis of feeding vessels
- Resection of eloquent brain
- Normal perfusion pressure breakthrough
Microsurgical outcome

- Risk of surgery is quite well estimated by the Spetzler-Martin grading system, with a favorable outcome in:
  - 92%–100% grade I
  - 95% grade II
  - 88% grade III
  - 73% grade IV
  - 57% grade V

(Spetzler and Martin 1986; Heros et al. 1990)
• A meta-analysis reviewed all series of more than 50 patients published since 1990 (25 series, 2452 patients) (Castel and Kantor 2000).
• Global mortality varied from 0% to 15% (mean 3.3%)
• Postoperative global morbidity was 1.5%–18.7% (mean 8.6%)
• Small superficial AVMs may be operated on with a very low morbidity (1.5%–9.7%) (Pik and Margan 2000; Schaller et al. 1998; Sisit et al. 1993).
• In contrast, morbidity of deep-seated lesions is much higher, at 9% in 22 patients (Lawton et al. 1995), 17% in 18 patients (De Oliveira et al. 1997), 20% in 22 patients (Sasaki et al. 1998), and 25% in 16 patients (U et al. 1992)
Embolization

- First described in 1960 by Luessenhope and Spence
- A rapidly evolving technique
- Embolic agents (silk, NBCA, histoacryl with lipiodol, onyx) and delivery systems (balloons with calibrated leaks, flow-guided micro catheters, over-the-wire micro catheters) have undergone drastic modifications
- Initially used as an adjunct to microsurgery or radio surgery
- In some cases can be a sole treatment modality
Aims of embolization

• Curative embolization
• Palliative embolization
• Partial (targeted embolization)
• Pre-op embolization
• Pre-radio surgery embolization
Principles of embolization

- Pre procedure steroids and anti epileptics
- Generally under GA
- Intra nidal catheter placement
- Embolic agent dilution
- Injection speed
- Stopping the injection
ONYX

- Innovative and promising material
- Non-adhesive and available in ready to use vials
- Composition: DMSO (di methyl sulfoxide) EVOH (ethylene vinyl alcohol co polymer) and tantalum.
- Can be used only after intranidal catheter placement
- It is easily injected, non adhesive, prolonged injection times are common.
Complications

- Incomplete embolization
- Intra cranial hemorrhage
- Ischemia
- Gluing of micro catheter
- Perfusion pressure breakthrough
Normal perfusion pressure breakthrough

- Post op /post embo swelling or hemorrhage
- ? Loss of auto regulation
- <5% incidence
- Mx – prevent post procedure hypertension

Embolization outcome

- Complete obliteration by embo alone 10-40% of AVMs.
- Multiple sessions are required most of the times
- Gobin et al (1996) 11% obliteration rate
- Bernstein et al (1990) 17% OR
Radiosurgery

- Harvey Cushing first advocated x-ray therapy for cerebral AVM.
- Currently GK (cobalt-60), Linac or proton beams used.
- In terms of radiobiology, AVM are late responding target within late responding normal tissue.
- Following bleed, part of nidus may be hidden or compressed by clots hence it is rational to wait till resolution or first evacuate it.
Radiosurgery

Radiation dose causes endothelial damage, smooth muscle cell proliferation, progressive sclerosis and subsequent thrombosis of nidal channels over time.

The success of stereotactic radiosurgery depends on AVM size and the radiation dose delivered.
Factors influencing obliteration rate

- AVM volume
- Target determination (complete nidus)
- Angio architecture and hemodynamics
- AVM location (hemispheric better)
- Radiation dosage
**Drawbacks of radiosurgery**

- Risk of bleed during latency
- Unknown long term perspective
- Unknown individual radio sensitivity
- Neurologic deficits related to radiosurgery
Radiosurgery outcome

• A series of 705 patients treated by radiosurgery alone or in combination with embolization or surgery. The overall complete obliteration rate (OR) was 55%. The OR was correlated to size:
  
<table>
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<tr>
<th>OR</th>
<th>nidus size</th>
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<tr>
<td>77%</td>
<td>&lt;15 mm,</td>
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<td>62%</td>
<td>15 mm - 25 mm</td>
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<td>44%</td>
<td>&gt;25 mm</td>
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• Mortality was 1.6%, due mainly to recurrent bleeding, which occurred in 6.5%

• Rate of recurrent bleeding was 2.98%/year/patient

• Neurological deficits related to radiosurgery and not related to hemorrhage were observed in 5.37% of the cases and were permanent in 1.46%

Radiosurgery outcome

- Peripheral or marginal dose is most significant factor
- Probability of obliteration = 35.69 x ln (marginal dose) - 39.66
- Marginal dose (Gy) OR
  - 6  24%
  - 12 49%
  - 20 67%
  - 28 79%

( Karlsson et al 1999)
Tools for treatment of brain arterio venous malformations: pros and cons

<table>
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<tr>
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<th>Embolization</th>
<th>Surgery</th>
<th>Radiosurgery</th>
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<tr>
<td>1. Low procedural invasivity</td>
<td>2</td>
<td>1</td>
<td>3</td>
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<tr>
<td>2. Occlusion capacity</td>
<td>1–2</td>
<td>3</td>
<td>2</td>
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<td>3. Speed of efficacy</td>
<td>2–3</td>
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<td>4. Long-term reliability</td>
<td>2–3</td>
<td>3</td>
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<tr>
<td>5. Independent from size</td>
<td>3</td>
<td>2</td>
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<tr>
<td>6. Independent from brain</td>
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<tr>
<td>functionality</td>
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<td>7. Independent from angio-architecture</td>
<td>1</td>
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<td>3</td>
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<td>8. Independent from flow</td>
<td>1</td>
<td>3</td>
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1, no/low; 2, medium; 3, yes/high.
# General recommendation

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<tr>
<th>SM grade</th>
<th>Deep perf. vessel</th>
<th>size</th>
<th>1&lt;sup&gt;st&lt;/sup&gt; choice</th>
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<td>3</td>
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<td>3</td>
<td>Present</td>
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<td>Rx+Ex</td>
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Thanks for Your Attention & Have a Good Day!!!