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AVM

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Arterio**V**enous **M**alfomations

Youmans and Winn Neurological Surgery, 8th edition, Jan 21, 2022

Cerebrovascular Malformations

- Capillary telangiectasia : incidental finding
- Venous angioma (Developmental venous anomaly) : M/C, incidental finding
- Cavernous malformation : 0.2 ~ 0.7 % / 90% asymptomatic
- AVM

- Direct fistulas, or Arteriovenous fistula

Cerebrovascular Malformations

- ~~Capillary telangiectasia : incidental finding~~
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- AVM

Patients with any type of vascular malformation of the brain may present with focal neurological deficit, seizure, or headache.

✓ due to hemorrhage or mechanical pressure, flow-related characteristics, or associated venous hypertension

Patients with DVAs and capillary telangiectasia have the lowest risk of bleeding, and if they do present with hemorrhage, an alternative source for such should be evaluated.

True arteriovenous malformations (AVMs)

- *Feeding arteries + Draining veins + a dysplastic vascular Nidus*
  - A conglomeration of numerous AV shunts w/o interposed brain tissue and no capillary bed

- Classically, congenital lesions
  - However, occurring as the result of upregulation or downregulation of multiple homeobox genes, which are involved in angiogenesis
- Predominantly sporadic
  - But can occur in certain hereditary diseases

The lack of a capillary bed

→ Low resistance + AV connections

→ High flow AV shunting

→ *Arterial dilation and Venous arterialization*

➔ Chronic high-flow shunt

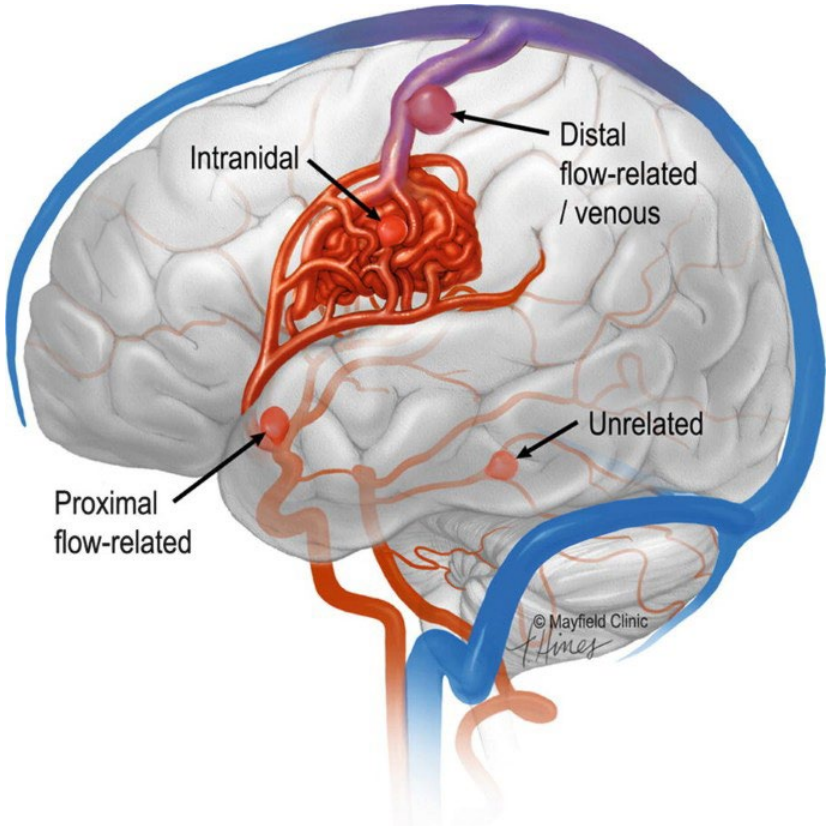
→ Dilation of the feeding arteries & thickening of the draining veins

→ The feeding arteries develop *smooth muscle hyperplasia*

associated with fibroblasts

→ Connective tissue elements known as *fibromuscular cushions*

- AVM associated aneurysms : 2.3% ~ 16.7%
  - The pathophysiology is not known definitively
  - Secondary to a high-flow vasculopathy



Arteriovenous Malformations: Presentation and Natural History, Andrew J. Ringer & Ryan Tackla,
Introduction to Vascular Neurosurgery pp 377–387

- The perinidal capillary network may be a cause of recurrence of surgically resected AVMs.
  - Dilated capillaries (10–25 times larger than normal capillaries) form a ring (1–7 mm) around the nidus.
- In contrast to CMs, intervening neural parenchyma may be present within the compact network of dysplastic vascular channels that forms the nidus.
  - Parenchymal elements tend to be gliotic, hemosiderin stained, and nonfunctional.

- Spontaneous obliteration does occur, but recurrence following confirmed obliteration is rare.

Etiology

- Sporadic AVMs : 0.04% to 0.52%
- Syndromic AVMs : 2% of cases
  - Hereditary hemorrhagic telangiectasia (HHT)
  - Cerebrofacial arteriovenous metameric syndromes (CAMSs)

- Apoplectic hemorrhage by rupture of nidal vessels or associated aneurysms

  or by venous outflow obstruction

  - Bleeding is typically from rupture of a draining vein, associated with dilation, kinking, and thrombosis, or from rupture of flow-related aneurysms, which are more prevalent than in adults.

- Large AVMs : an arterial steal phenomenon

  Older children : progressive neurological deterioration & chronic epilepsy

  - If sufficient AV shunting, neonates and infants may present with congestive cardiac failure.

Classification Criteria

- SPC class A is Spetzler-Martin grades I and II bAVM
- SPC class B is Spetzler-Martin grade III bAVM
- SPC class C is Spetzler-Martin grades IV and V bAVM

TABLE 452.1 Grading System of Spetzler and Martin for Arteriovenous Malformations[a]

Graded Feature	Points Assigned
AVM Size (Diameter)	
Small (<3 cm)	1
Medium (3–6 cm)	2
Large (>6 cm)	3
Eloquence of Adjacent Brain	
Noneloquent	0
Eloquent	1
Pattern of Venous Drainage	
Superficial only	0
Deep	1

[a]Grade = Size + Eloquence + Venous drainage.

Epidemiology

- 15 – 45 years with ICH ; 38% due to AVM
- Autopsy & MRI studies : 0.2%–1.0%
  - In autopsy studies, only 15% had symptoms
- Diagnosed in the 3~4th decade
- Common in men and women (M>F)

- Most are supratentorial and solitary.
- In the posterior fossa, the cerebellum is the most common site.

Clinical presentation

- Up to 40% of patients with symptoms unrelated to the AVM.
- Up to 50% of patients present to medical attention (a ruptured AVM with hemorrhage)
- ICH is the m/c, IVH, SAH, and rarely SDH

  including focal neurological deficit, headache, and seizure.

- The R2eD AVM score

  suggesting a 78% chance of hemorrhage

$$Y = -1.676 + 0.912\,(\text{Nonwhite}) + 0.516\,(\text{Deep Location}) + 0.486\,(\text{Small size}) + 0.547\,(\text{Deep drainage}) + 0.456\,(\text{Single feeder})$$

$$\frac{e^{y}}{1+e^{y}}.$$

- Hemorrhage at initial presentation often have significant morbidity and mortality (10%–40%).

- 15%–35% of patients first present with a seizure.
  - The mechanism :
    - Related an associated hemorrhage
    - Mass effect with cortical irritation
    - Flow characteristics leading to steal, ischemia, and neuronal damage
  - Risk factors : superficial or cortically based & a frontal, temporal, or parietal location, lack of prior hemorrhage, large nidus

- Fewer than 10% of patients present with neurological deficits without hemorrhage.
  - Steal phenomenon, microhemorrhages, mass effect from the AVM, or accompanying hydrocephalus.

- A large AVM may have ipsilateral headaches with a migraine-like quality.
  - Long-standing meningeal artery involvement and recruitment of blood supply
  - Venous outflow obstruction

Natural history

- The risk of hemorrhage : 2% – 4% per year
- The lifetime risk of AVM rupture: $1 - (\text{risk of no hemorrhage})^{\text{expected years of life}}$

  Simplified to lifetime risk = 105 − age

- The hemorrhage rate :

  <1% per year (asymptomatic) ~ >20% in first year (recently hemorrhage)

- The strongest and most consistent predictor of hemorrhage is prior hemorrhage.

- A high risk of rehemorrhage (20%–40%)
  - 1st yr : 15.4%
  - 4 yrs : 5.3%
  - After 5 yrs : 1.7%
  - Risk factors : age and sex, deep location with exclusive deep venous drainage, microhemorrhage, and large size ..
- These are not consistent findings in all studies.

- Incomplete treatment of the nidus does not alter the natural history.

- Hemorrhage risk in patients with hereditary hemorrhagic telangiectasia may be lower.

Therapeutic Decision Making

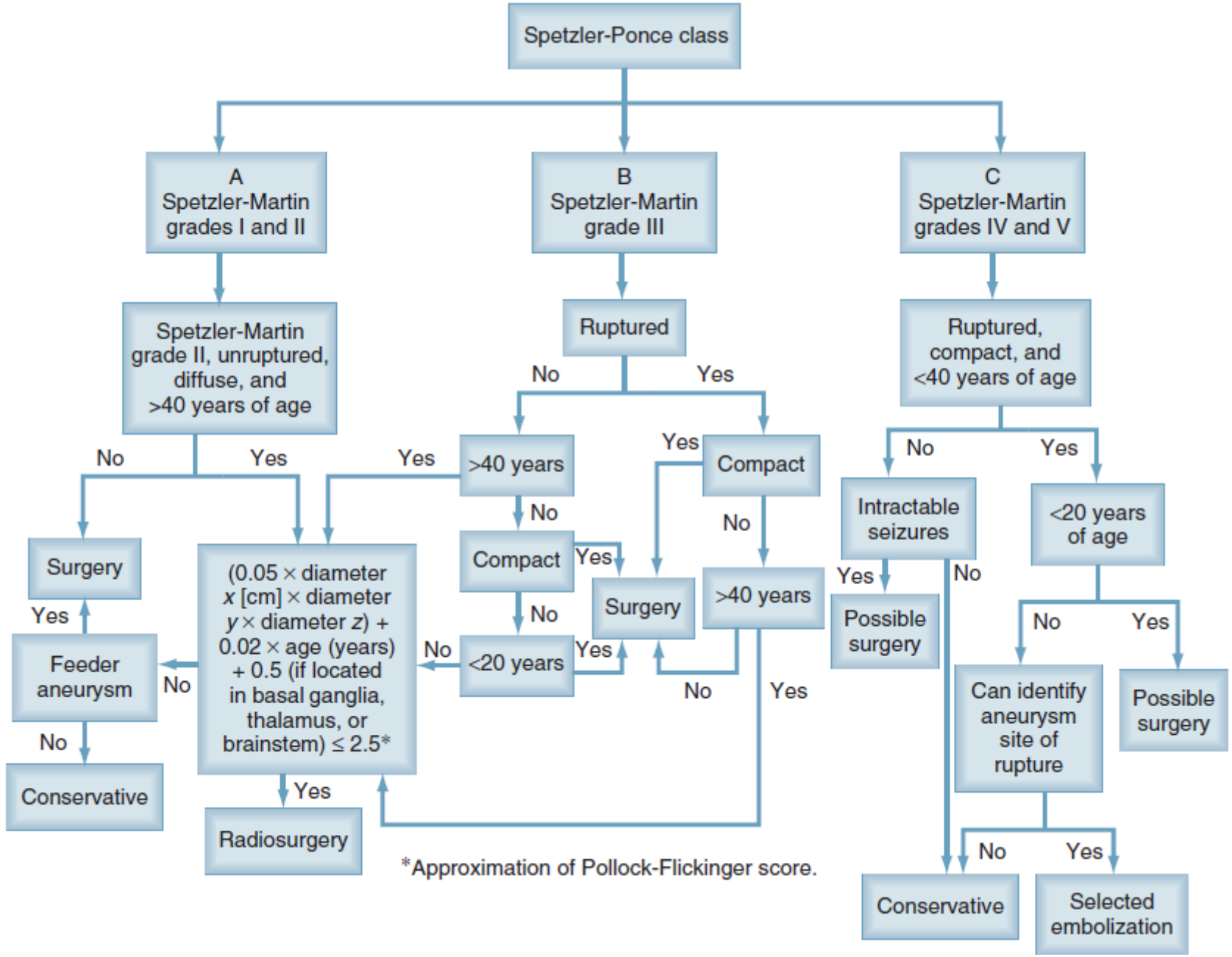
- A 10- year risk for first hemorrhage : 15% ~ 25%
- A 10-year risk for ruptured bAVM : 20% ~ 35%

- The consequence of each future hemorrhage includes a 40% risk of a permanent neurological deficit or death.

- Surgical excision of SPC class A bAVM : 5 yrs in ruptured / 9 yrs in unruptured
- Surgical excision of SPC class B bAVM : 17 yrs in ruptured / 23 yrs in unruptured

- SPC class A bAVM : surgery (unless old, unruptured, and diffuse)
- SPC class B bAVM : determined by following the Lawton-Young grading system
- SPC class C bAVM : conservative management (unless young and a tight nidus)

- A Lawton-Young grade of 7 or greater is more likely to lead a permanent deficit or death from surgery in comparison to conservative treatment for at least the first 40 years after diagnosis.


Spetzler-Ponce class
A
Spetzler-Martin grades I and II
B
Spetzler-Martin grade III
C
Spetzler-Martin grades IV and V
Spetzler-Martin grade II, unruptured, diffuse, and >40 years of age
Ruptured
Ruptured, compact, and <40 years of age
No
Yes
Surgery
Feeder aneurysm
Conservative
(0.05 × diameter x [cm] × diameter y × diameter z) + 0.02 × age (years) + 0.5 (if located in basal ganglia, thalamus, or brainstem) ≤ 2.5*
Radiosurgery
>40 years
Compact
<20 years
Surgery
>40 years
Intractable seizures
Possible surgery
<20 years of age
Can identify aneurysm site of rupture
Possible surgery
Conservative
Selected embolization
*Approximation of Pollock-Flickinger score.

Endovascular Management

- Nidus size <3 cm, noneloquent, fewer, larger, and less tortuous feeding arteries
  - More than half of all lesions may be completely obliterated.
- The high reported complication rates
- New endovascular techniques have the potential for cure a safe and feasible option.
  - Multiplug intranidal flow control and transvenous embolization
  - Improved devices and novel embolic agents
- Preoperative embolization

  to reduce the risk for complications and to allow safe resection
- Palliative embolization in selected cases

  intractable headaches or progressive neurological deficits

  related to arterial steal or venous hypertension

Microsurgery

- Grade I and II AVMs : surgical excision

  Provided that they are relatively young and healthy
- Grade III AVMs : not straightforward

  Although many can be surgically resected with acceptable morbidity

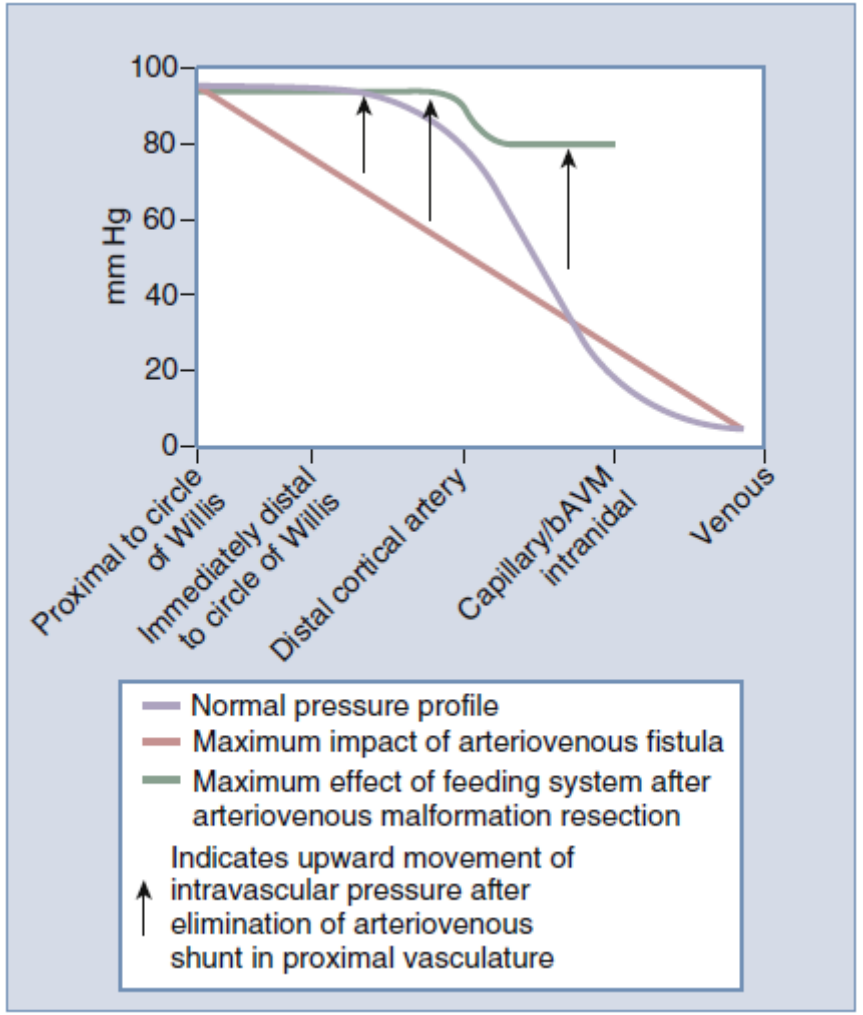
- Grade IV and V lesions : should be treated conservatively

  Surgical resection is associated with serious morbidity

  The likelihood of complete obliteration with multimodal therapy is low

Arterio-capillary-venous hypertensive syndrome

- Brain hemorrhage during and after resection
- The underlying mechanism : intravascular pressure ↑
  - Venous outflow occlusion
  - A failure of autoregulation
    (normal perfusion pressure breakthrough)
  - A rise in pressure within proximal arteries with insufficient integrity because of chronically low pressures
- Remodeling of the arteries over time will return the intravascular pressures to normal.
  - Shear stress ↓ → Endothelial NO synthase ↓
  → Pulsation ↑ → Endothelin release ↑



- A feeding artery : < ½ of the internal carotid in the vicinity of the Pcom & < 3 cm
  Remodeling is complete within 7 days following surgery in the majority of cases.
- For other combinations, the majority have not fully remodeled within 7 days of surgery.
- During this period,
  the patient is vulnerable to both hemorrhage and rupture of thin-walled vessels
- It is important to be sure that the nidus has been completely excised.

- For larger than 3 cm maximum diameter and with large feeding arteries, this may require active blood pressure reduction to levels known to be safe for normal brain (i.e., a perfusion pressure above 50 mm Hg) but significantly lower than normal.

- CTA or DSA demonstrating remodeling can assist in deciding when an aggressive hypotensive regimen can be eased.

- Vasospasm can occur with devastating consequences.
  - For a therapeutic hypotensive regimen, prophylaxis against vasospasm is reasonable to attempt

- It is possible to mistake the remodeled arteries for arteries in vasospasm, and attempted angioplasty may be fraught with danger.

- The author manages large bAVMs in the ICU with intravenous CCB (with nimodipine and/or magnesium);

  also useful in inducing the desired effect of supplementing BP control to reduce the risk of arterio-capillary venous hypertensive syndrome,

  with a possible protective effect against the development of vasospasm

Focused Irradiation

- A consideration
  - Radiation-related complications
  - Time delay between treatment and cure
  - The likelihood of cure

- Small-volume bAVMs
  - 5yr S-M grade I and II : ≤ 2cm, 87% and > 2cm, 67% / 5yr S-M grade III : 70%

- The risk of hemorrhage remains unchanged until complete occlusion has occurred

- Pollock and Flickinger predicted radiosurgery obliteration without deficit :

  = 0.1 × volume (mL) +0.02 × age (years) +0.5*

  (*if located in basal ganglia, thalamus, or brainstem)

  - FU of 70 months, the obliteration rate without deficit

    90% for scores ≤1

    70% for scores >1 and ≤1.5

    60% for scores >1.5 and ≤2

    less than 50% for scores >2

  - mRS score decline in the 95% CI range

    0%–10% for scores ≤1

    10%–20% for scores >1 and ≤1.5

    15%–30% for scores >1.5 and ≤2

    25%–50% for scores >2

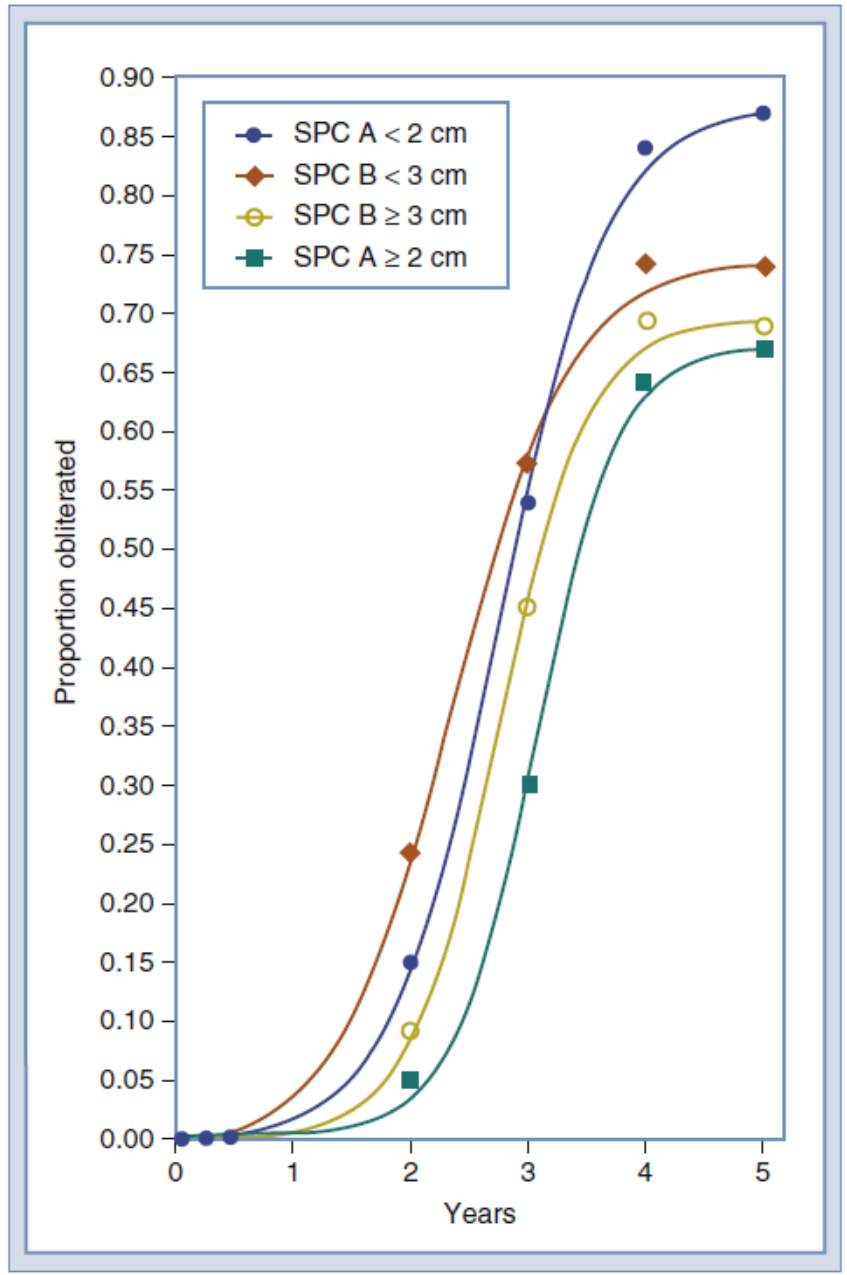


Figure 452.3. Obliteration rate estimation for Spetzler-Ponce classification *(SPC)* classes A and B arteriovenous malformations of the brain *(bAVMs)* based on data from Kano and colleagues.[32,44] Sigmoid curve of best fit for the estimated obliteration

- The best obliteration rate is in small bAVMs (<3 cm) with no prior embolization.

- Repeat focused irradiation can be performed and has a reported obliteration rate of 35% and 68% at 3 and 4 years, respectively, after the repeat treatment.
- Planned staged volumetric reduction has been reported for a small number of cases with an obliteration rate of no better than 20% at 4 years and with a similar percentage of patients dying of hemorrhage.

- With such results, embarking on treatment for large bAVMs with focused irradiation should be done only after consideration of alternative management pathways.

- Over a mean of 7 years, VRAS score,

  FO following radiosurgery in 81%, 75%, 66%, 47%, and 41% for 1 - 5.
- The Spetzler-Martin grade

  FO was observed in 75%, 69%, 61%, and 37% of patients for I - IV.

Management of Grade IV and V AVM

- A formidable challenge for neurosurgeons.
- An annual rupture rate : 2% ~ 4% for unruptured AVMs

  4% ~ 18% for previously ruptured AVMs
- Morbidity and mortality rates following AVM rupture are 50% and 10%, respectively.
  - ✓ Historically, surgical management of these lesions has been associated with high rates of perioperative morbidity and mortality.
  - ✓ Endovascular embolization and stereotactic radiosurgery are generally not effective when used as a single modality for high-grade AVMs.
  - ✓ Multimodal, staged therapy consisting of preoperative embolization and/or stereotactic radiosurgery can achieve high obliteration rates with acceptable morbidity and mortality in select patients.