VENOUS MALFORMATIONS

NEUROSURGERY LECTURE

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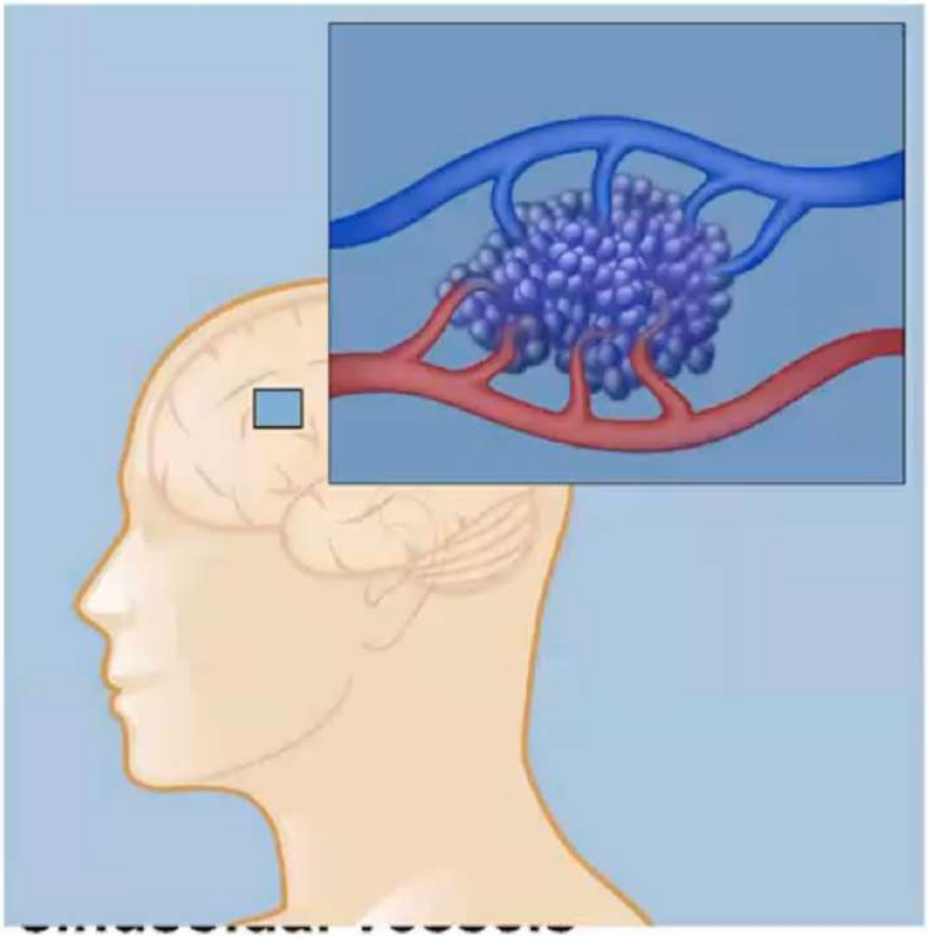
CAVERNOUS MALFORMATIONS

Cavernous angioma

Cavernoma

Cavernous hemangioma

Hemangioma



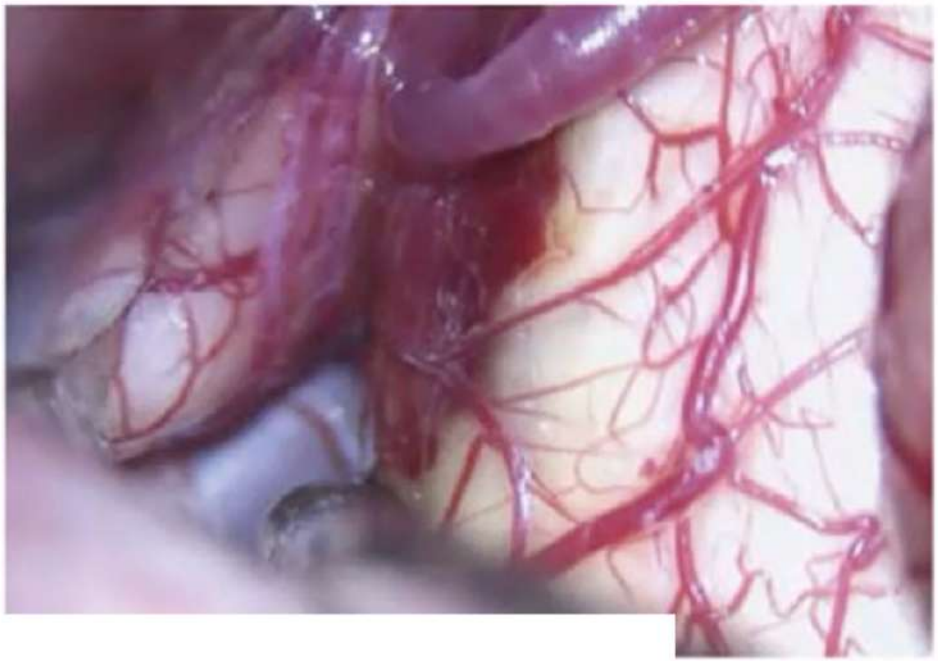
- Well circumscribed
- Benign vascular hamartoma
- Irregular thick and thin walled
- Lack intervening parenchyma, oarge feeders or large draining veins

- Usually 1-5 cm size
- Hemorrhage, calcify or thormbose
- Rerely in spinal cord

- Caverns are filled with blood
- Various stages of thrombus formation

- May arise de novo
- May grow, shrink or remain unchanged
- Capillary telangiectasia – adjacent and may be a precursor

- “hemorrhoid of the brain”
- Represent a mulberry



EPIDEMIOLOGY

- **5-13% of CVMs**
- **Occurrence**
  - 48-86% supratentorial
  - 4-35% brainstem
  - 5-10% basal ganglia
  - 23-50% multiple
- **Risk factor**
  - Radiotherapy – craniospinal
  - 42% spinal CM harbor >=1 intracranial CM
  - Hereditary – autosomal dominant, 3 genes

PRESENTATION

Seizures 50%

Hemorrhage 25%

Focal neurological deficit without hemorrhage 25%

Hydrocephalus or incidental 20-50%

SEIZURES

- 5 year risk of first time seizure – 6% among symptomatic CMs
- 4% among incidental CMs
- The iron in hemosiderin is epileptogenic

HEMORRHAGE

Defined as "symptomatic presence of blood outside the hemosiderin ring"

Risk of hemorrhage in cerebral CMs

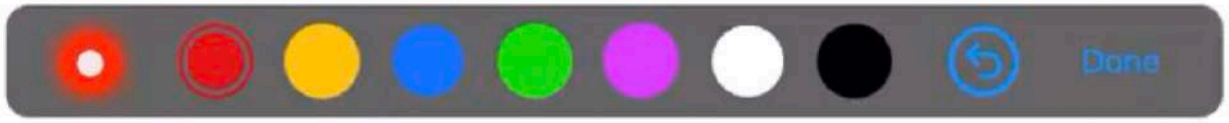
- 1st time hemorrhage – incidental CMs – very low (0.08%)
- CMs initially presenting with hemorrhage: ratio 5.6
- Brainstem CMs: hazard 4.4
- Annual risk of recurrent hemorrhage declines over time
- Higher rates in familial CMs (4.3-6%)

NO risk of hemorrhage with

- Female gender, CM size or CM multiplicity
- Pregnancy & parturition
- Platelet inhibitors, anticoagulants
- Physical activity

Spinal CMs

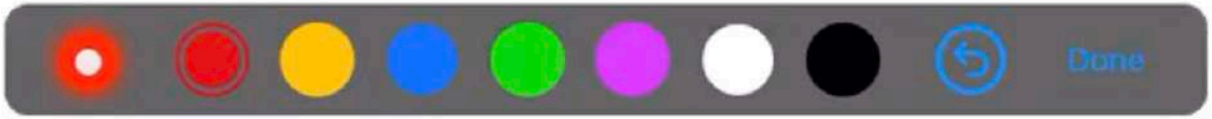
- Annual hemorrhage rate 2.1%
- 17% have cerebral CMs



IMAGING

Recommendations

- Brain MRI for diagnosis & follow up
- Include Gradient echo (GRE) or SWI images
- DSA not recommended unless AVM suspected
- Follow up imaging – to assess new or worsened symptoms and guide treatment



CT

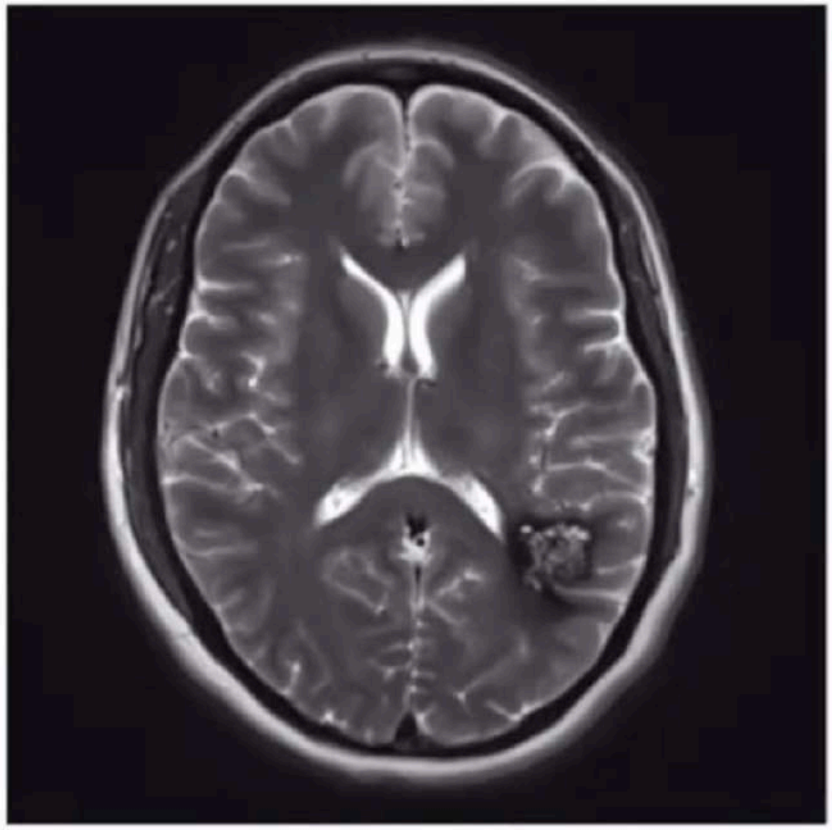
- Not sensitive

MRI

- Diagnostic test of choice

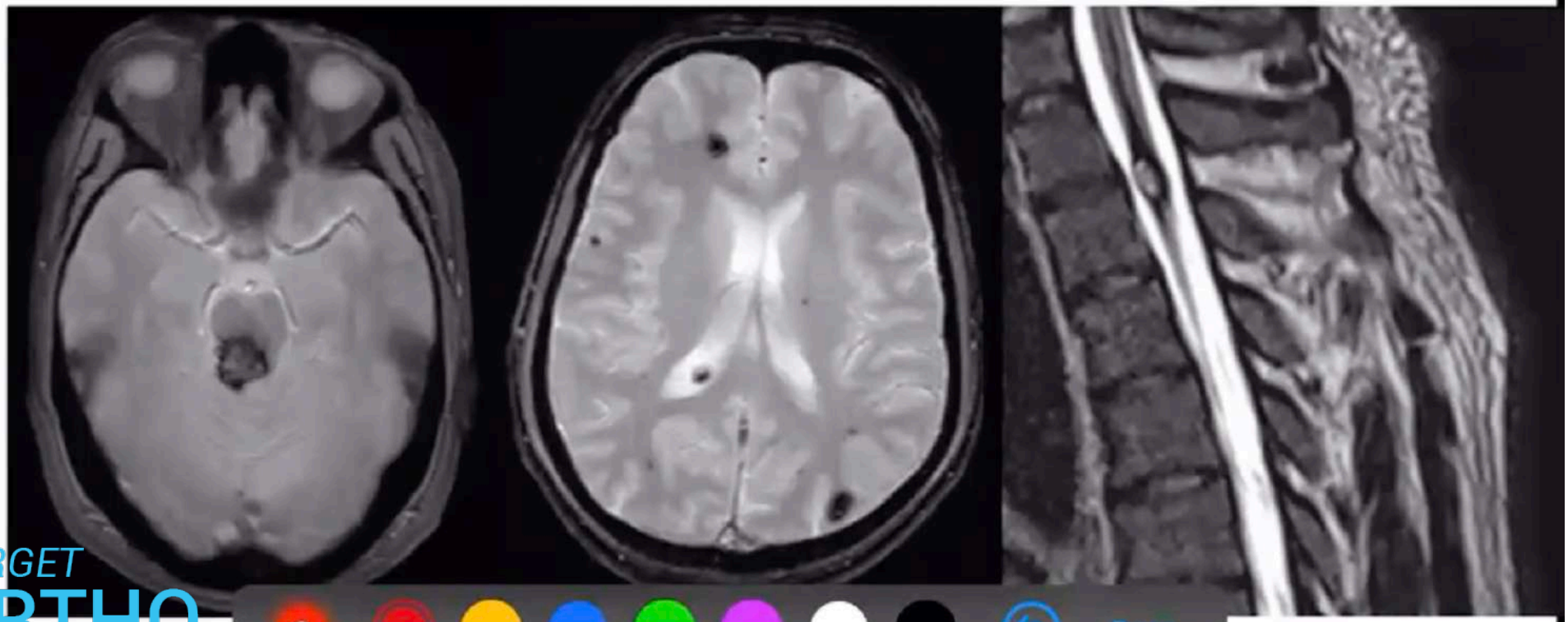
DSA

- Does not demonstrate lesion
- To rule out questionable diagnosis



Familial imaging

- 1st degree relatives with more than 1 member having CMs should have MRI screening



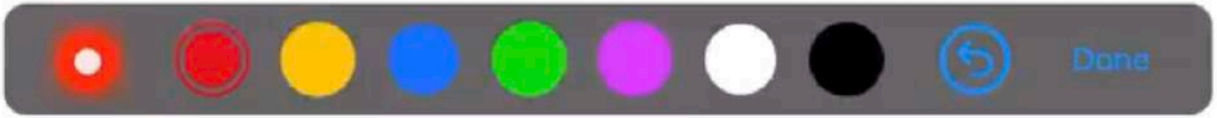
TREATMENT RECOMMENDATIONS

Incidental lesions

- Observation
- Serial imaging 2-3 years

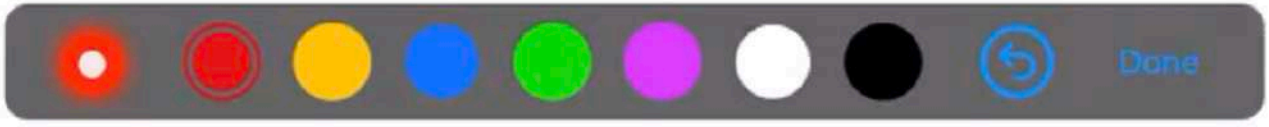
CM with ICH

- Follow management of ICH



Brainstem CMs

- **No surgery for CMs that have not bled**
- **Surgery**
  - For history of >2 prior hemorrhages
  - Pial/ependymal representation

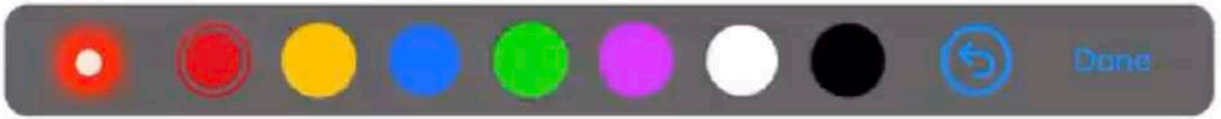


Spinal cord CMs

- Same as brainstem CMs

Cranial nerve CMs

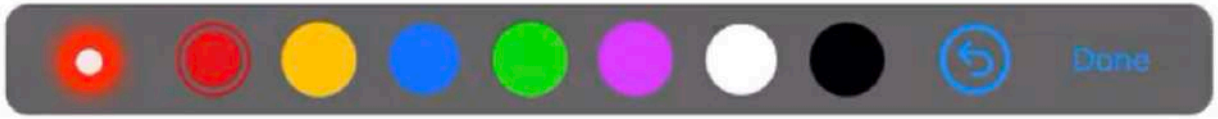
- Early surgical decompression



SURGERY

Indications

1. **Accessible lesions with**
    1. Focal deficit
    2. Symptomatic hemorrhage
    3. Seizures
2. **Less accessible lesions**
    1. Repeated hemorrhages with neorulogical deterioration



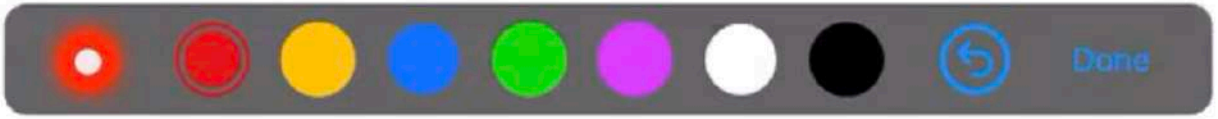
SRS

Controversial

Results comparable to natural history

Decrease in hemorrhage rates after 28 months

May promote development of new CMs in familial cases



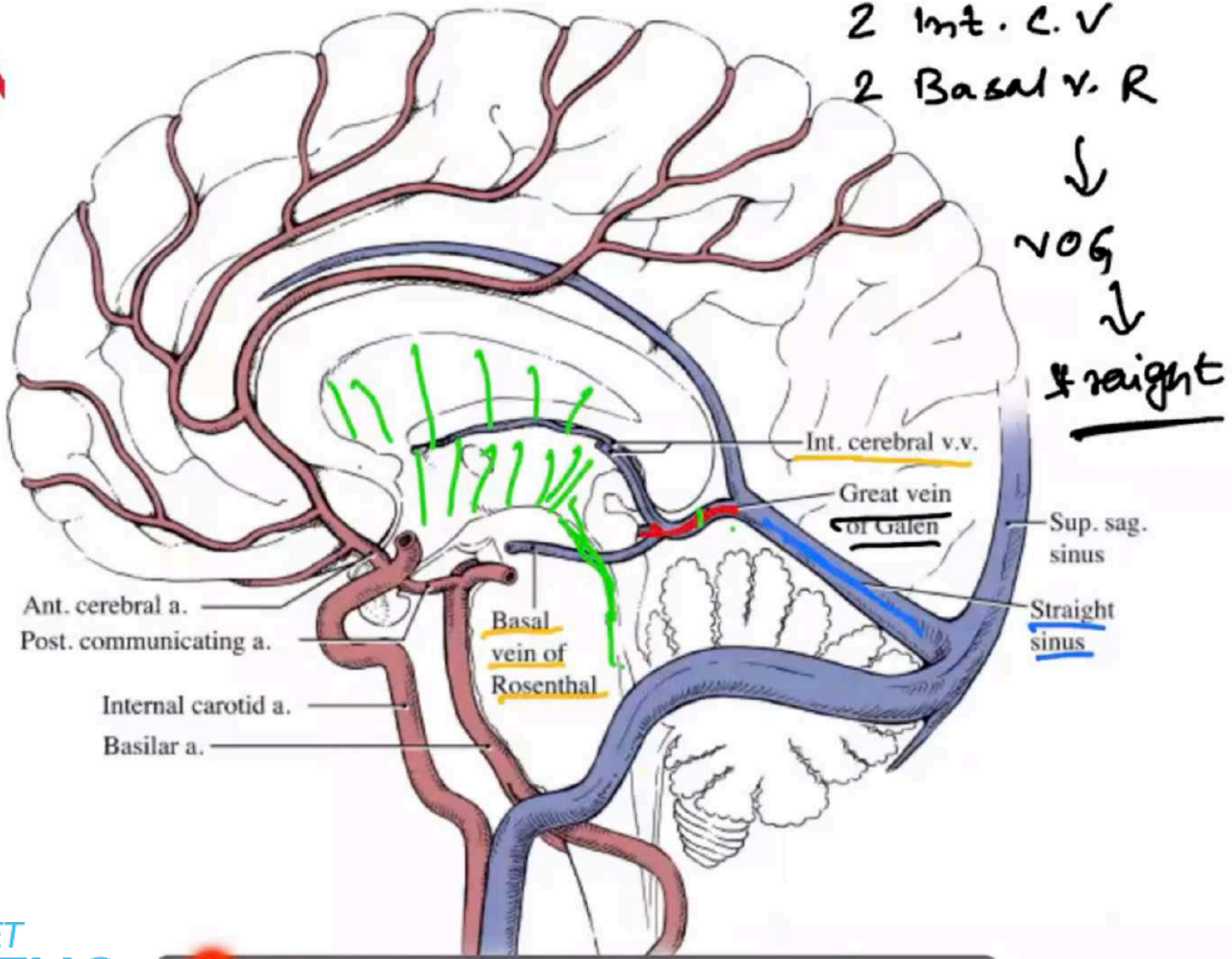
Practice guideline: Treatment recommendations for cavernous malformations

1. **Level III**[70]: surgery is not recommended for asymptomatic CMs, especially those that are deep, or in eloquent areas or brainstem, or with multiple CMs
2. **Level II**[70]: consider surgery for solitary asymptomatic CMs if easily accessible and not in eloquent brain, to prevent future hemorrhage, because of psychological burden, expensive & time consuming follow-ups, to facilitate lifestyle or career decisions, or in patients who might need to be on anticoagulation
3. **Level II**[70]: consider early CM resection (≤ 6 weeks from hemorrhage) in patients with seizures, especially when medically refractory, if the CM is the likely cause of the seizure
4. **Level II**[70]: consider surgery in symptomatic easily accessible CMs (surgical morbidity & mortality is equivalent to living with the CM for 1–2 years after a first bleed)
5. **Level II**[70]: consider surgery after a second symptomatic bleed in a brainstem CM after reviewing the high risks of early post-op morbidity, mortality and impact on quality of life
6. **Level II**[70]: consider surgery in deep CMs if symptomatic or after prior hemorrhage (surgical morbidity & mortality is equivalent to living with the CM for 5–10 years after a first bleed)
7. **Level II**[70]: surgical indications are weaker after a single, disabling bleed from a brainstem CM
8. **Level II**[70]: consider radiosurgery in solitary CMs with previous symptomatic hemorrhage if the CM is located in eloquent areas that have an unacceptably high risk with surgery
9. **Level III**[70]: radiosurgery is not indicated for CMs that are asymptomatic, or surgically accessible, or are part of familial CMs because of the concern about precipitating formation of additional CMs

VEIN OF GALEN MALFORMATION

AN

2 Int. C. V
2 Basal v. R
↓
VOG
↓
Straight



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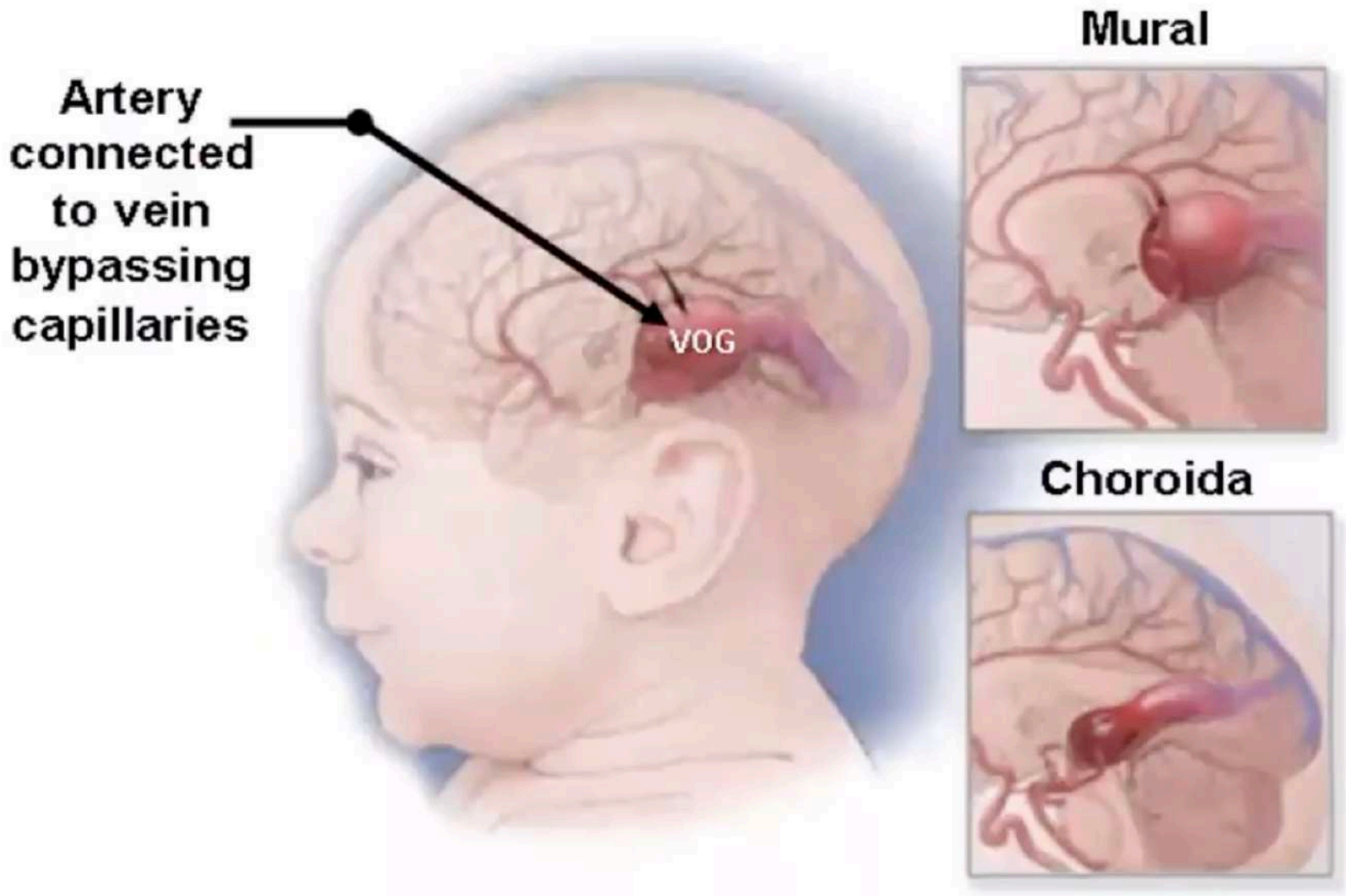
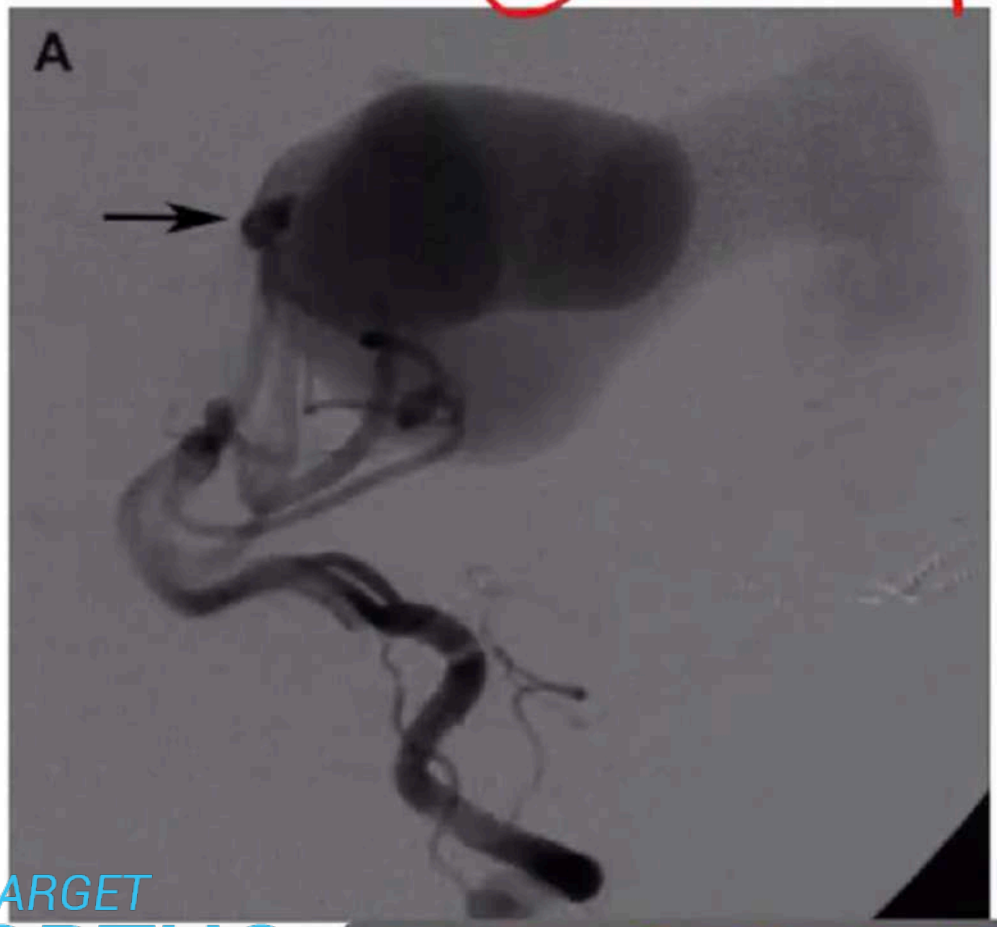
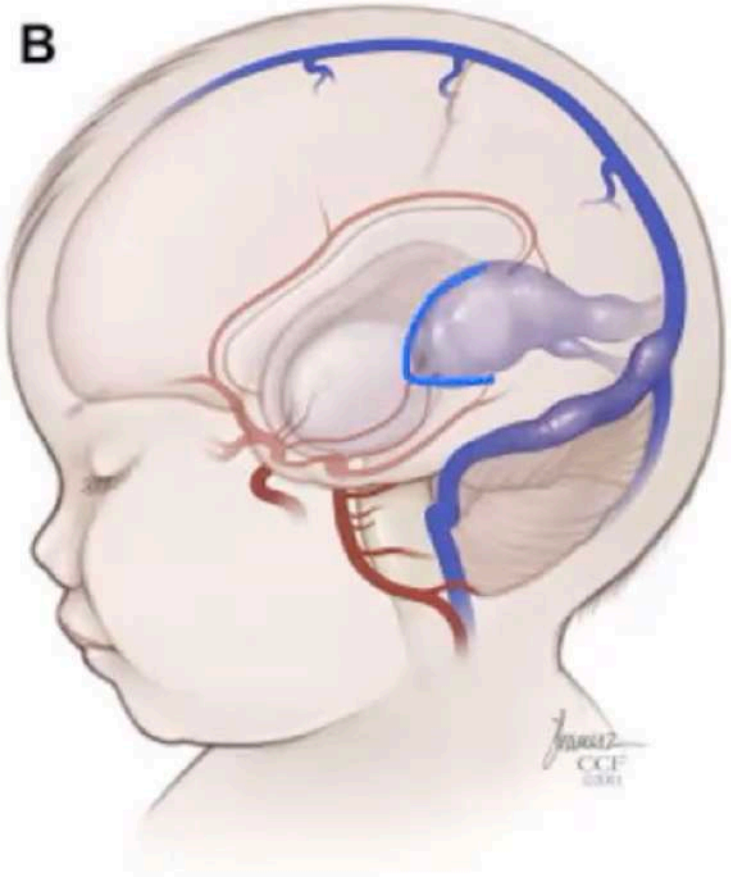


1. Callosomarginal / pericallosal A.
2. PCA
3. Choroidal A.
4. thalamoperforators





- **Congenital**
  - Develop before 3 month embryo stage
- **RASA 1 gene mutations**
- **Secondary**
  - Adjacent to deep AVMs or AVF
- **Feeders**
  - Medial & lateral choroidal
  - Circumferential, mesencephalic, anterior choroidal, pericallosal and meningeal arteries
- **Agenesis of straight sinus**

CLASSIFICATION

Yasargil classification based on the location of fistula

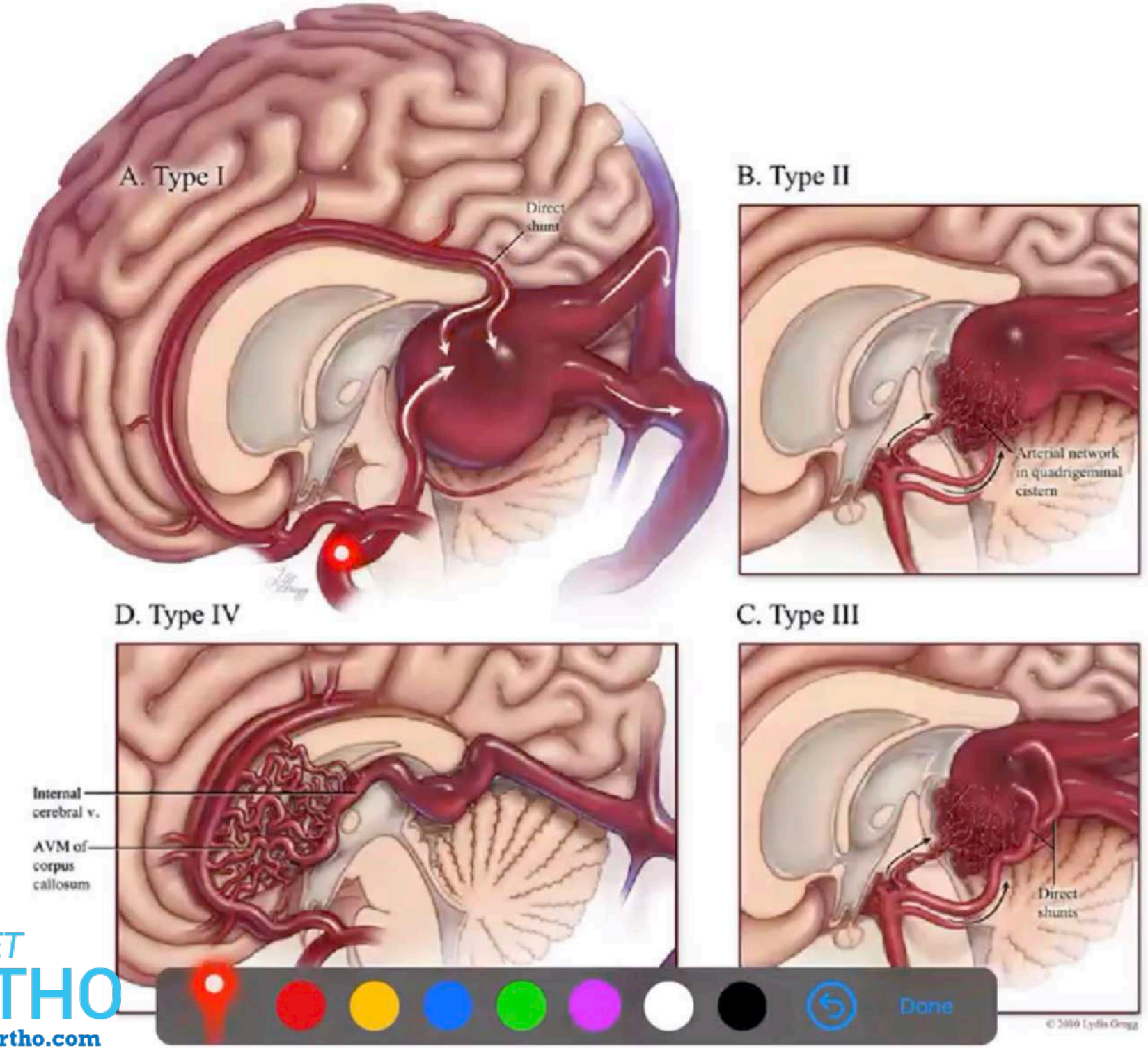
1. Pure internal fistulae: single or multiple
2. Fistulae between thalamoperforators and the VOG
3. Mixed form: most common
4. Pial AVMs

4 types

1: direct arterial feeder

2: perforator

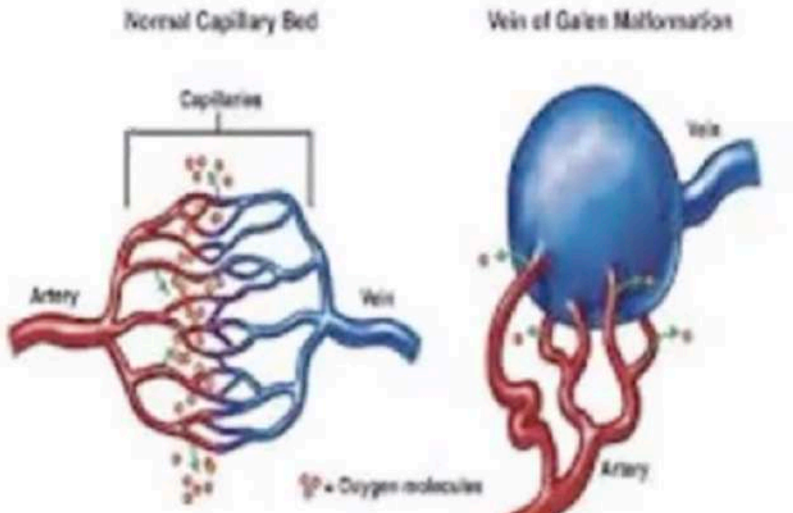
3: mixed



PRESENTATION

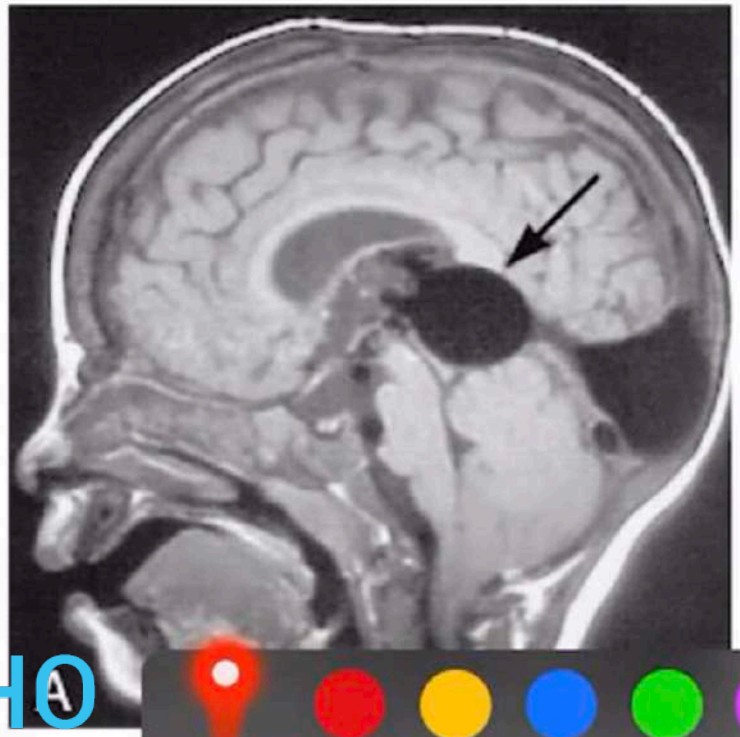
Neonates

- 40% patients diagnosed during neonatal period
- Congestive heart failure
- Cerebral artery "steal" → ischemia



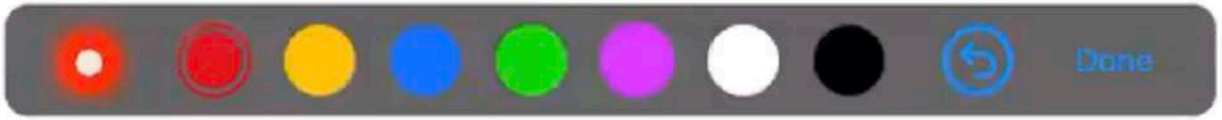
Infants

- Hydrocephalus & seizures
- Obstructive hydrocephalus vs impaired CSF absorption



Older children and adults

- low-flow fistula or Type IV malformation
- SAH, ICH, headaches
- Ocassional hydrocephalus



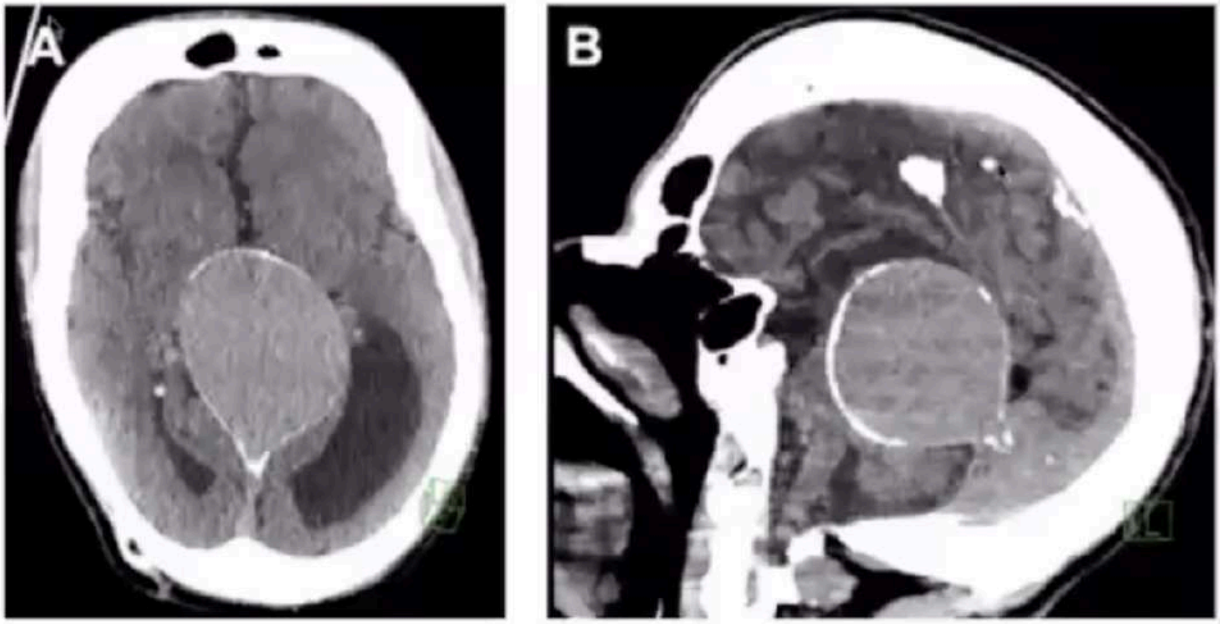
IMAGING

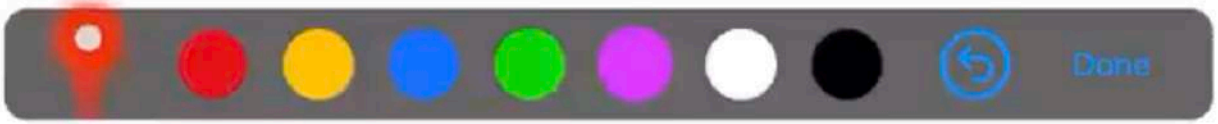
CT

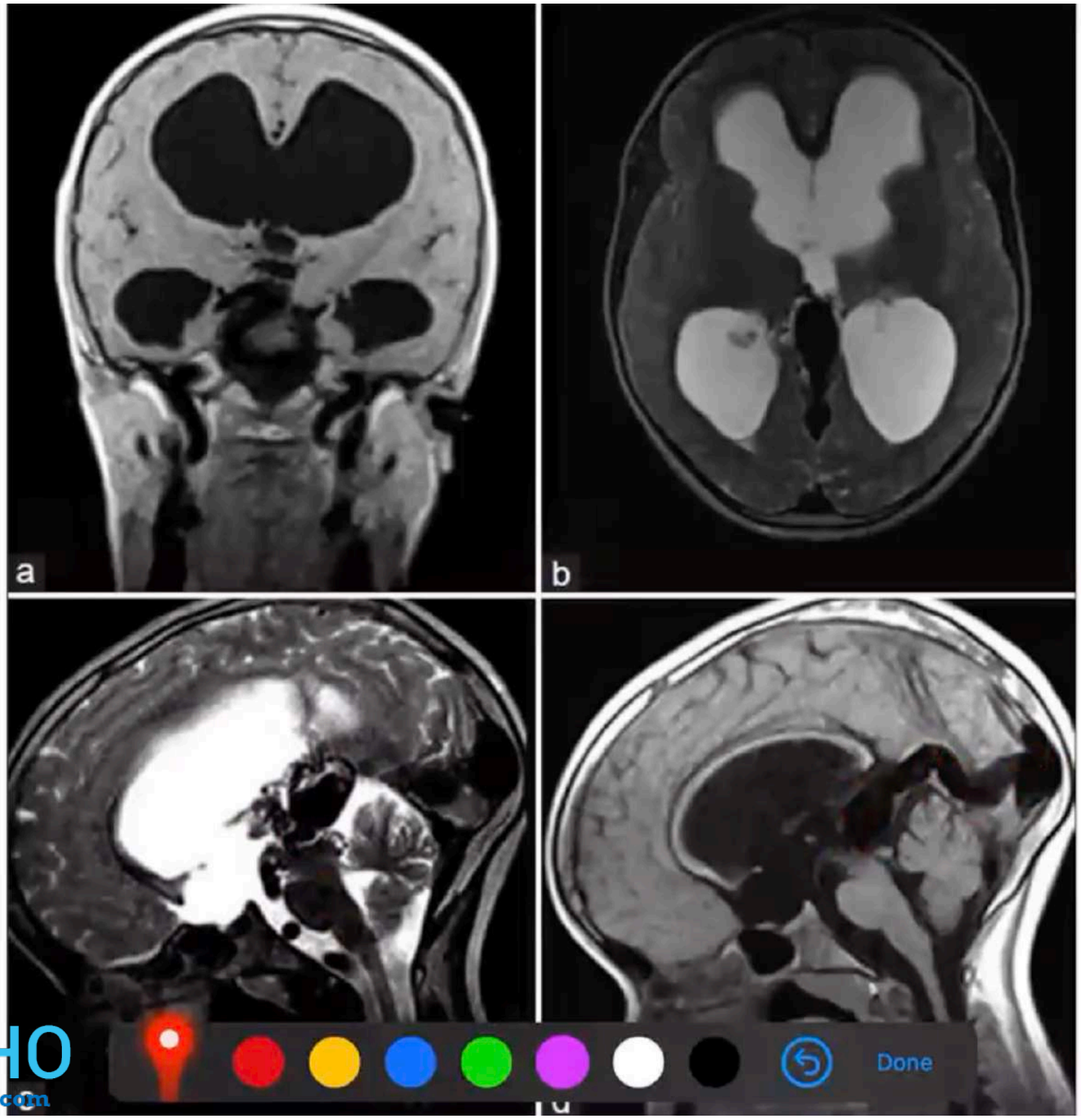
- Calficifation, low-density cystic spaces

MRI

- More sensitive
- Gives details about ischemic changes







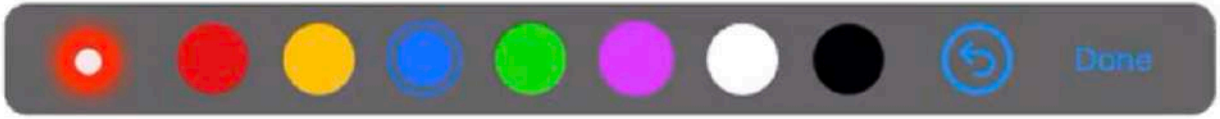
DSA

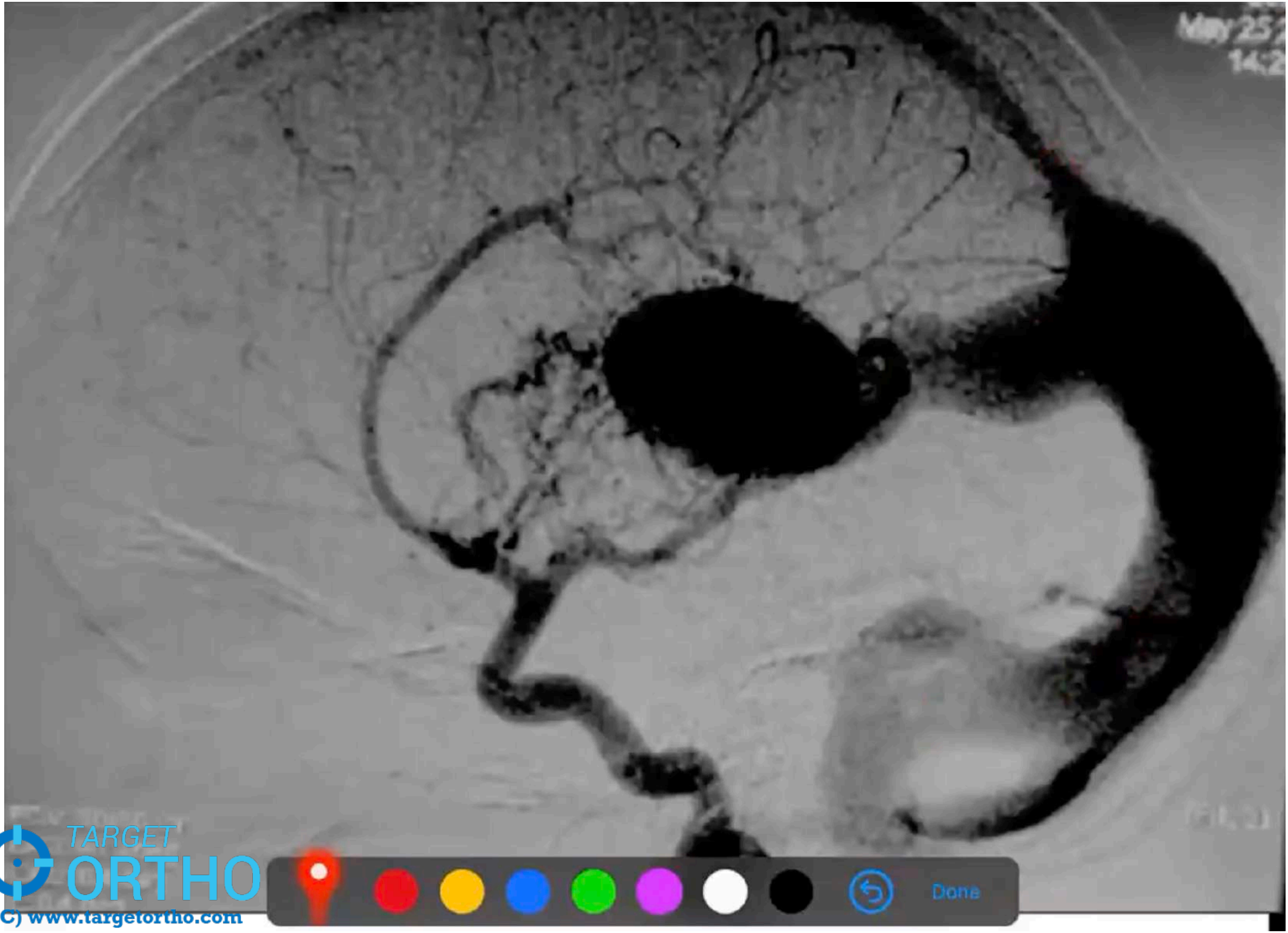
- Diagnostic
- Vascular anatomy
- Flow pattern

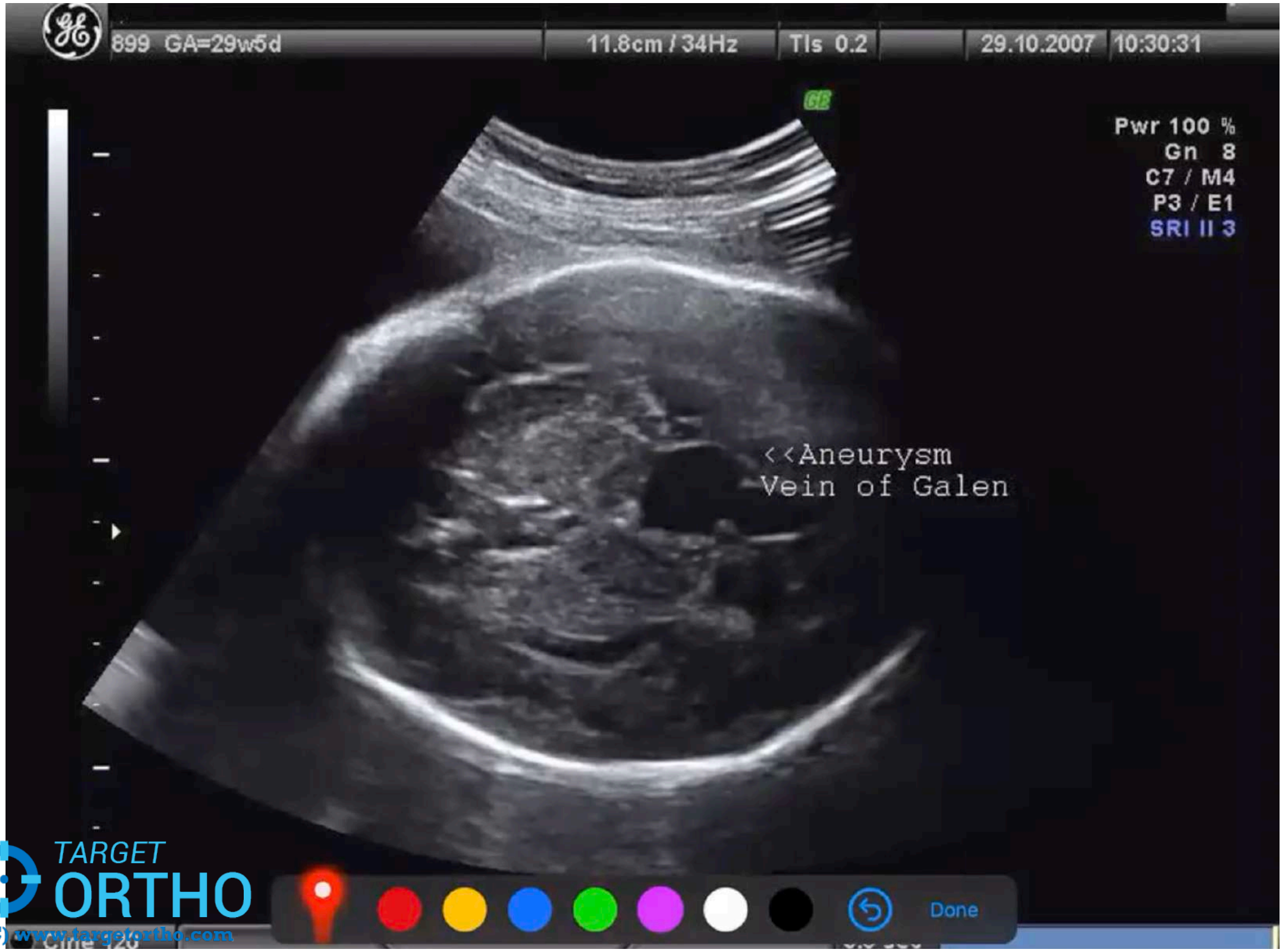
Prenatal USG

VOGM

- Characteristic midline tubular anechoid structure (comet tail or keyhole sign)
- Extracardiac left to right shunt







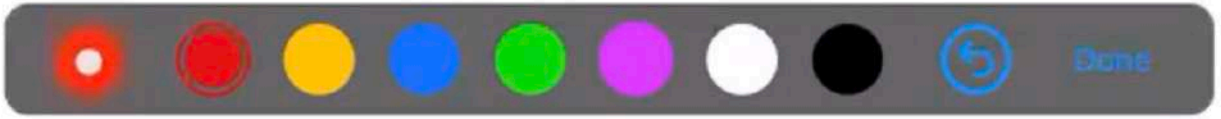
TREATMENT

Untreated neonates → 100% mortality

With treatment, mortality in neonates → 79% and others 39%

Goal:

To restore normal growth conditions than a normal morphological appearance



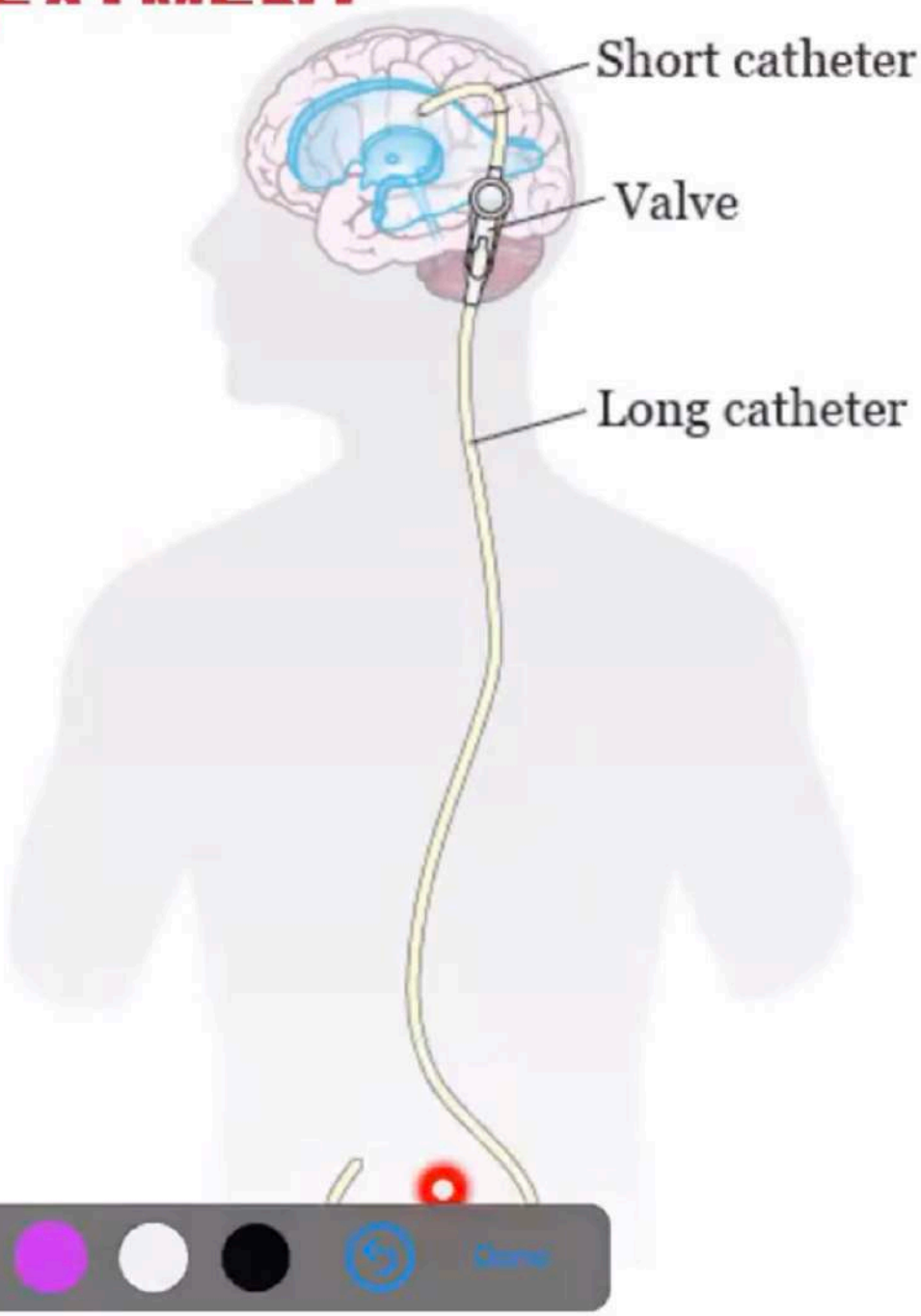
MEDICAL TREATMENT

Newborn

- Medical stabilisation
- CHF, pulmonary HTN and cardiomegaly management
- Intervention delayed until infant is 6 months old

HYDROCEPHALUS TREATMENT

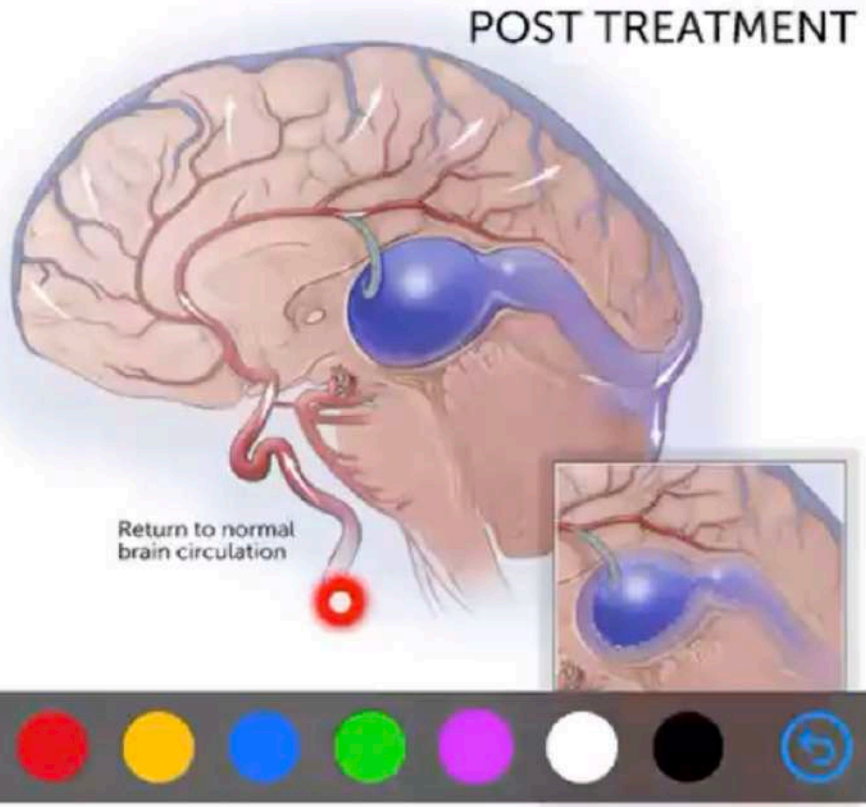
The CSF pressure is low, hence results of most shunt procedures in infants and neonates are disappointing



ENDOVASCULAR TREATMENT

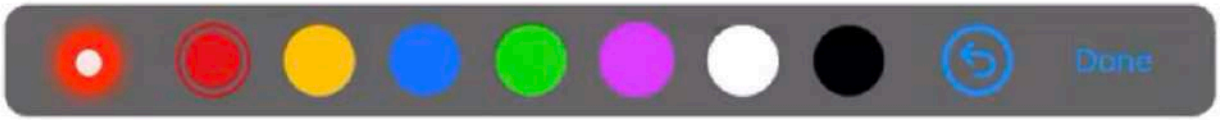
Transarterial or transvenous approach

- TA approach for Type I-III malformations
- TV approach for failed TA approach
- TV approach for type IV malformation

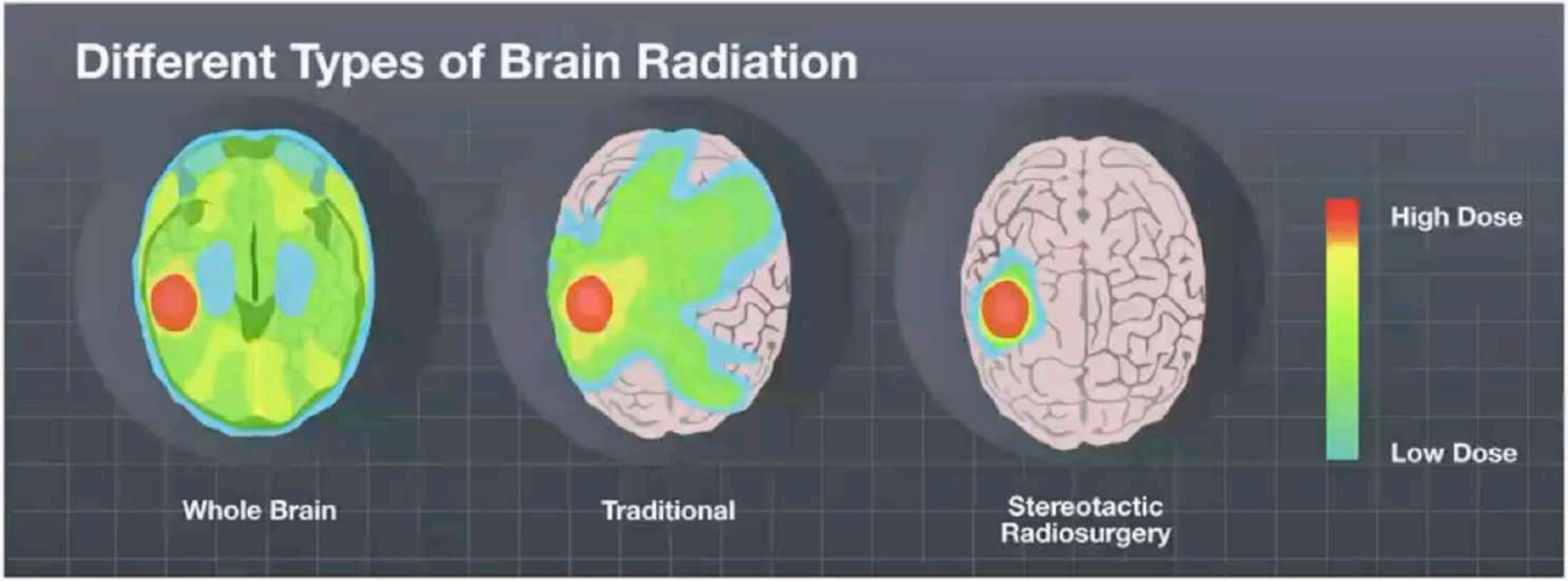


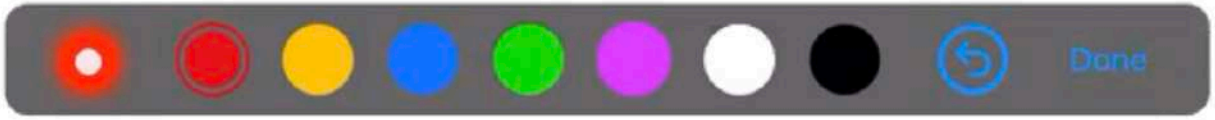
SURGERY

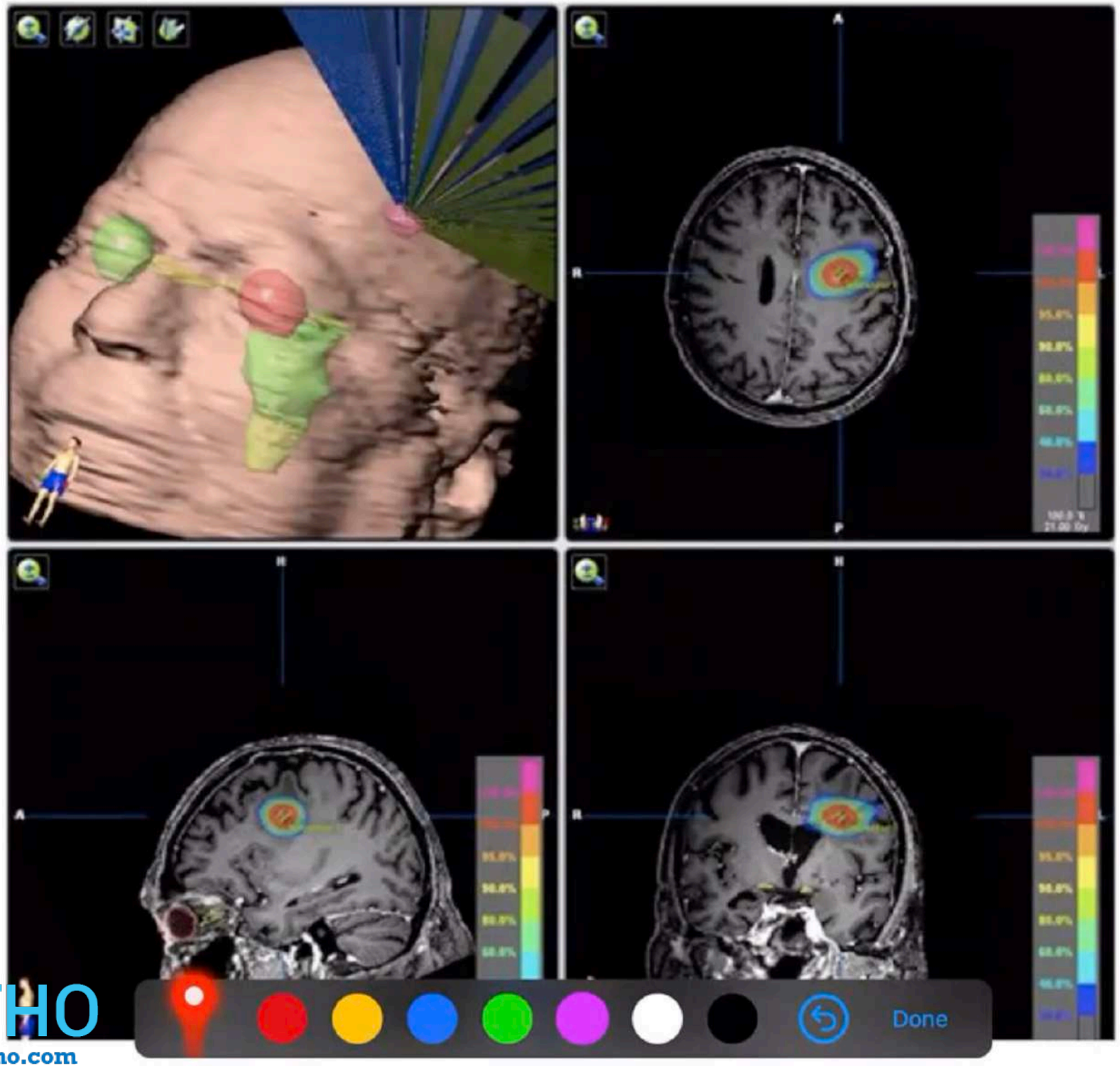
- Definitive management of hydrocephalus
- Medical condition and nutritional status are optimised
- Pre-operative embolisation of accessible feeders



RADIOSURGERY







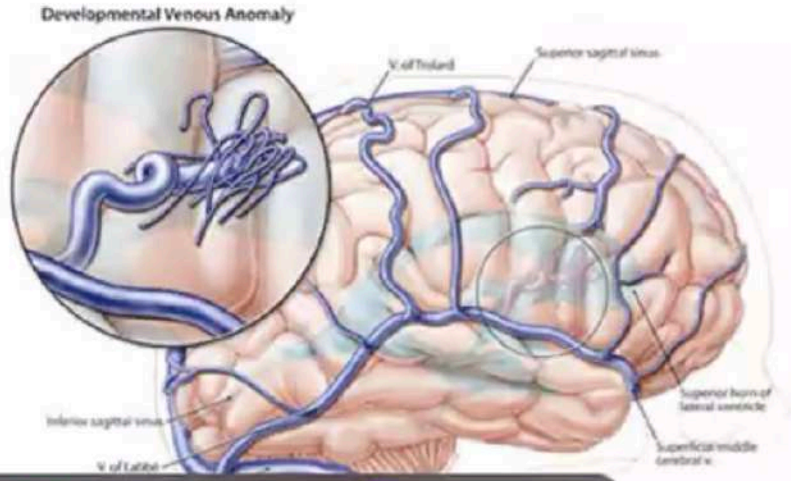
VENOUS ANGIOMA

Developmental venous anomaly (DVA

Venous malformation

Venous angioma

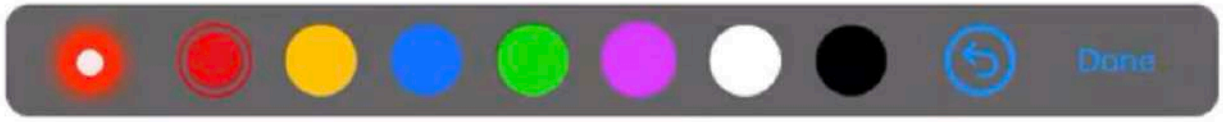
- Tuft of medullary veins
- Converge into an enlarged central trunk
- Intervening brain parenchyma



- Mostly in region of MCA or vein of Galen
- Drain into superficial or deep venous system
- May have an associated cavernous malformation
- Non hereditary
- Low flow
- Low pressure

PRESENTATION

- Mostly clinically silent
- Rarely seizures
- Less freq hemorrhage
- Venous infarcts may be there



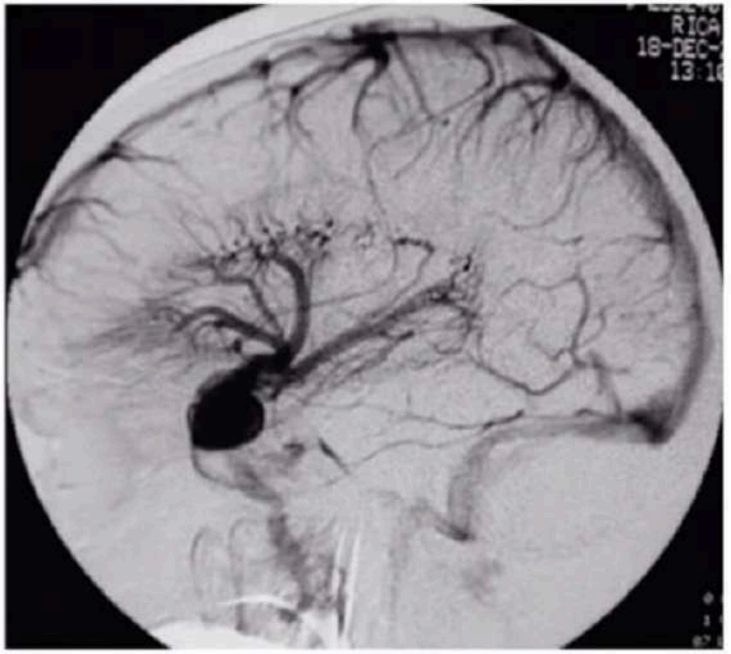
IMAGING

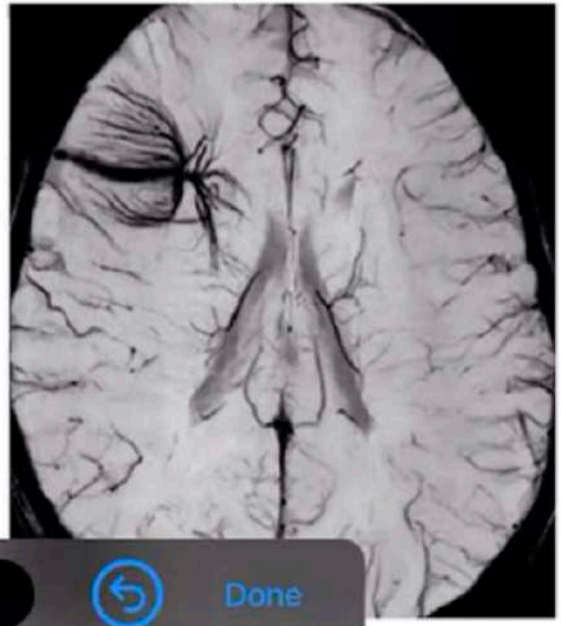
MRI

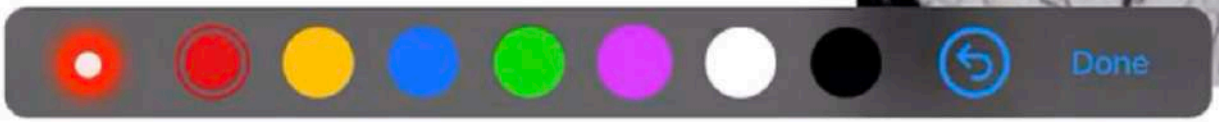
- T2 hyperintense on FLAIR

DSA

- Distinct caput medusae
- Long draining vein
- Arterial phase – no AV shunting





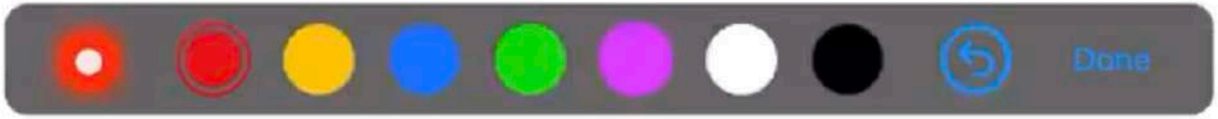


TREATMENT

- Left untreated
- Surgery for cavernoma, the angioma left alone

Surgery indications

- Documented bleeding
- Intractable seizures



AOVM

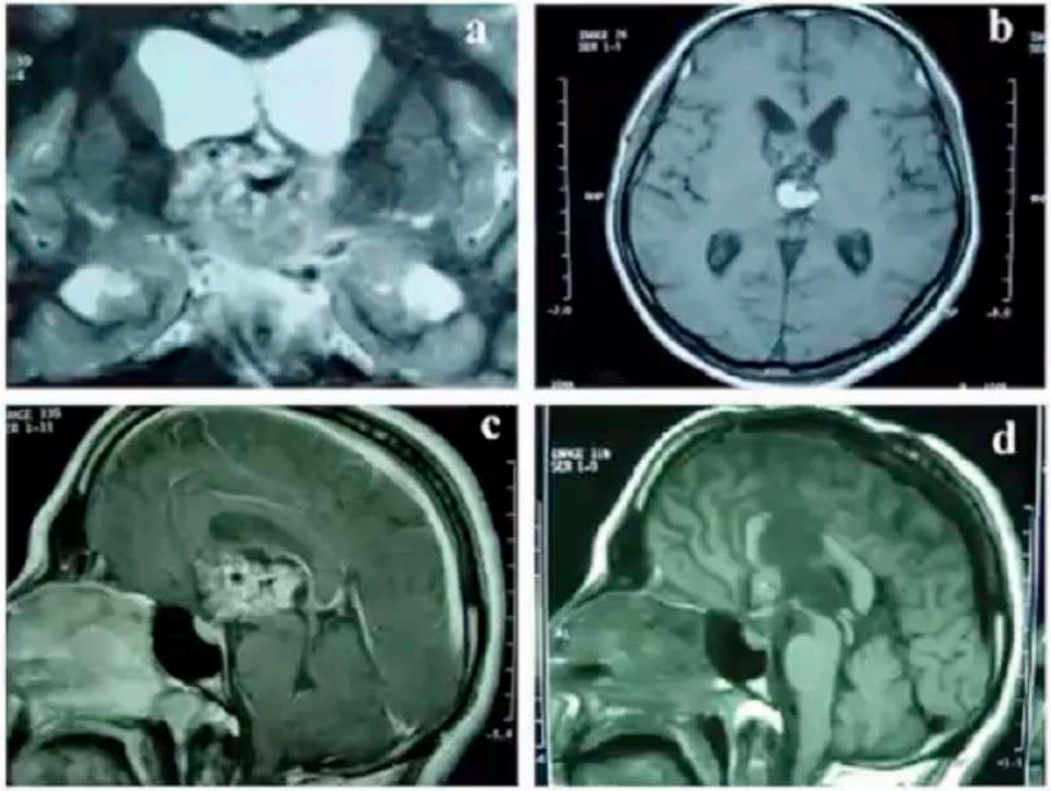
ANGIOGRAPHICALLY OCCULT VASCULAR

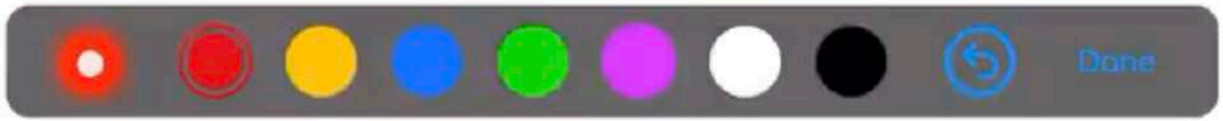
- **Malformations not demonstrable on a good quality angiogram**
    - Despite magnification, antiotomography, rapid angiograms or delayed films

Reasons

- **Lesion may have hemorrhaged**
- **Sluggish flow**
- **Small size of abnormal vessels**

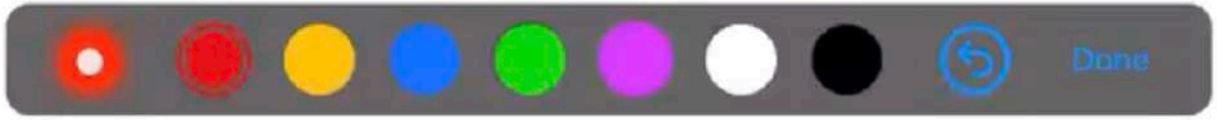
- Represent 10% of all CVMs
- Average age at diagnosis – 28 years



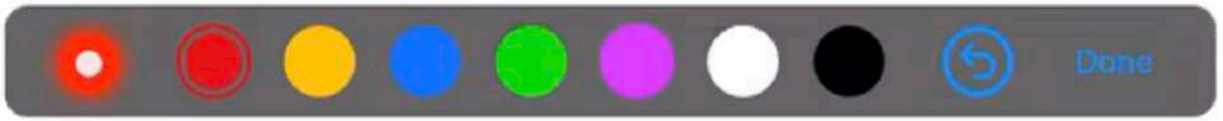


PRESENTATION

- Seizures
- Headache
- Spontaneous ICH
- Incidental

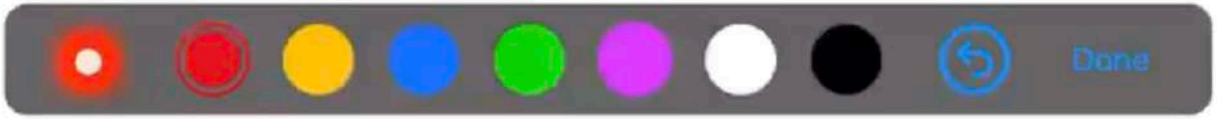


CAPILLARY TELANGIECTASIA



Aka hereditary hemorrhagic telangiectasia

- Slightly enlarged capillaries
- Low flow
- Solitary or part of syndrome
  - Osler-Weber-Rendu
  - Louis Barr (ataxia telangiectasia)
  - Mybum – Mason
  - Sturge-Weber



IMAGING

CT

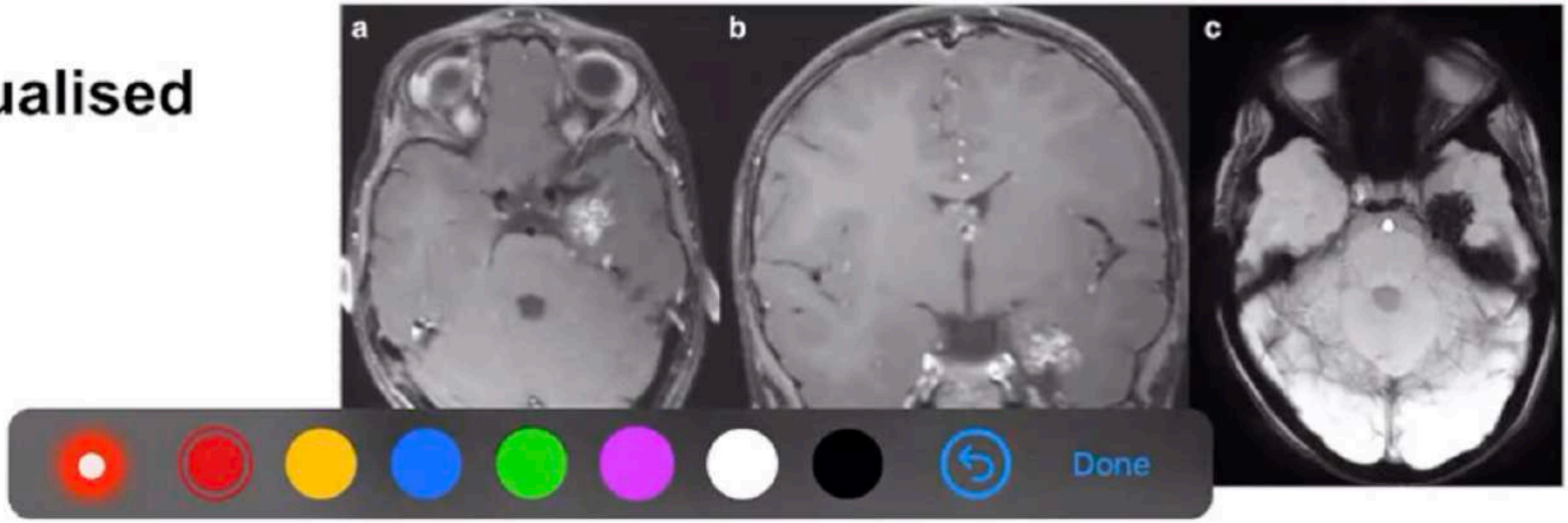
- High density lesion, homogenous, well demarcated

MRI

- Previous hemorrhage
- Hemosiderin ring

DSA

- Not visualised



TREATMENT

Surgery

- Evacuation of hematoma or diagnosis
- Recurrent hemorrhages
- Intractable seizures

SUMMARY

Capillary telangiectasia — DSA ✗, Rx ✗

Venous angioma — Rx

AOVM — DSA ✗, Rx

Cavernous malformations — Rx

VOGM