VEIN OF GALEN MALFORMATION

Dr. Nguyen Ngoc Pi Doanh
Dr. Dang Ngoc Dung

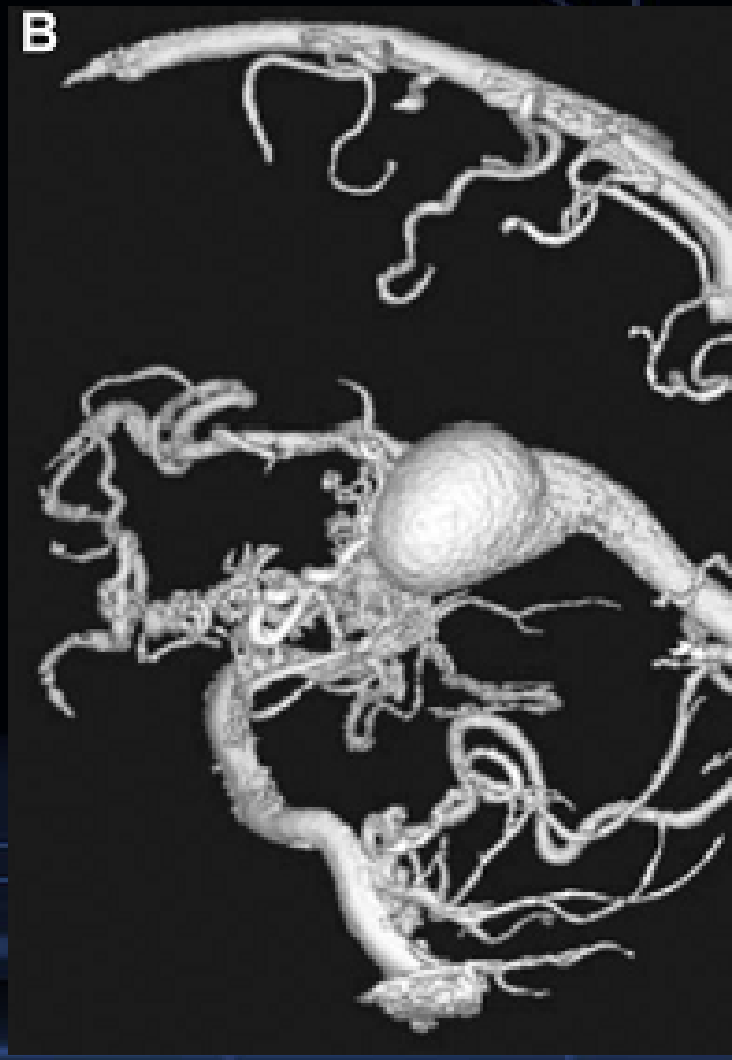
Contents

- Epidemiology
  - Anatomy
  - Pathology
- Classification
  - Symptoms
  - Diagnosis
  - Treatment
  - Outcome

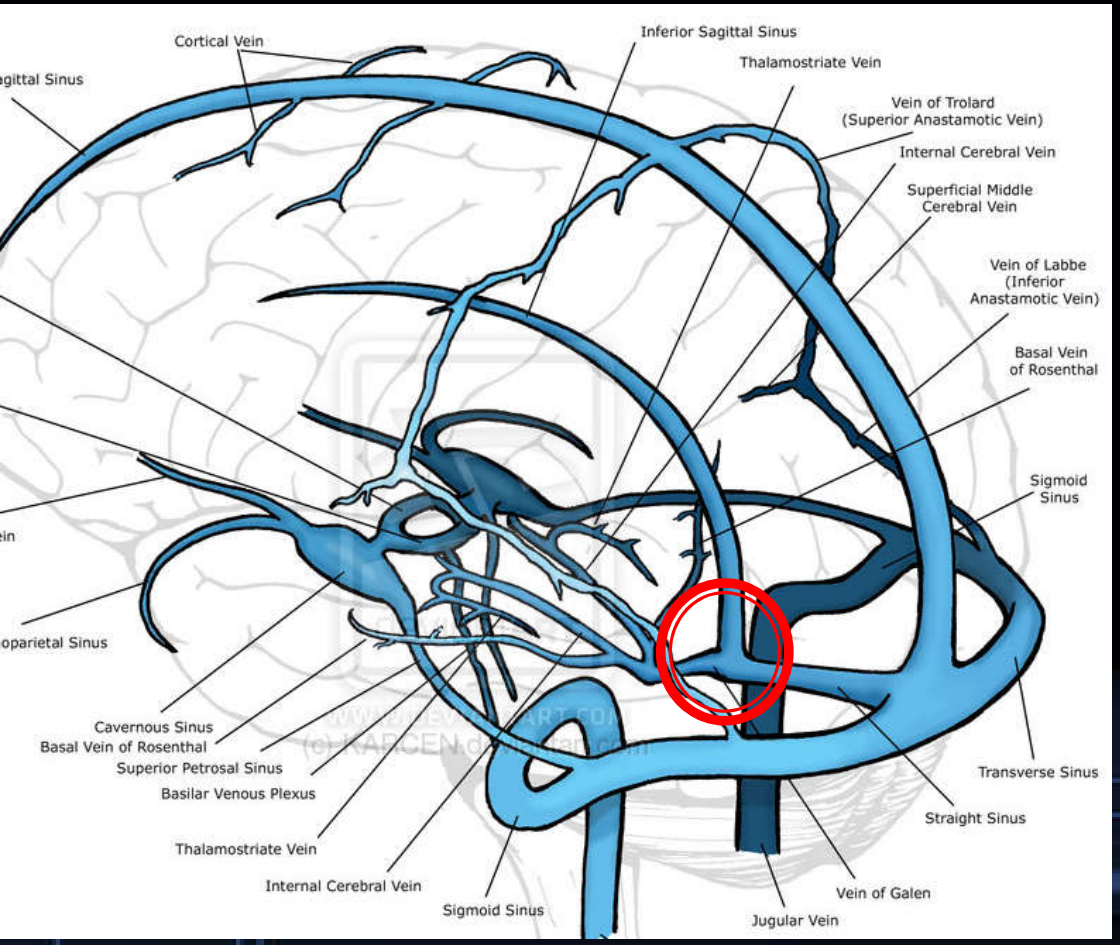
EPIDEMIOLOGY

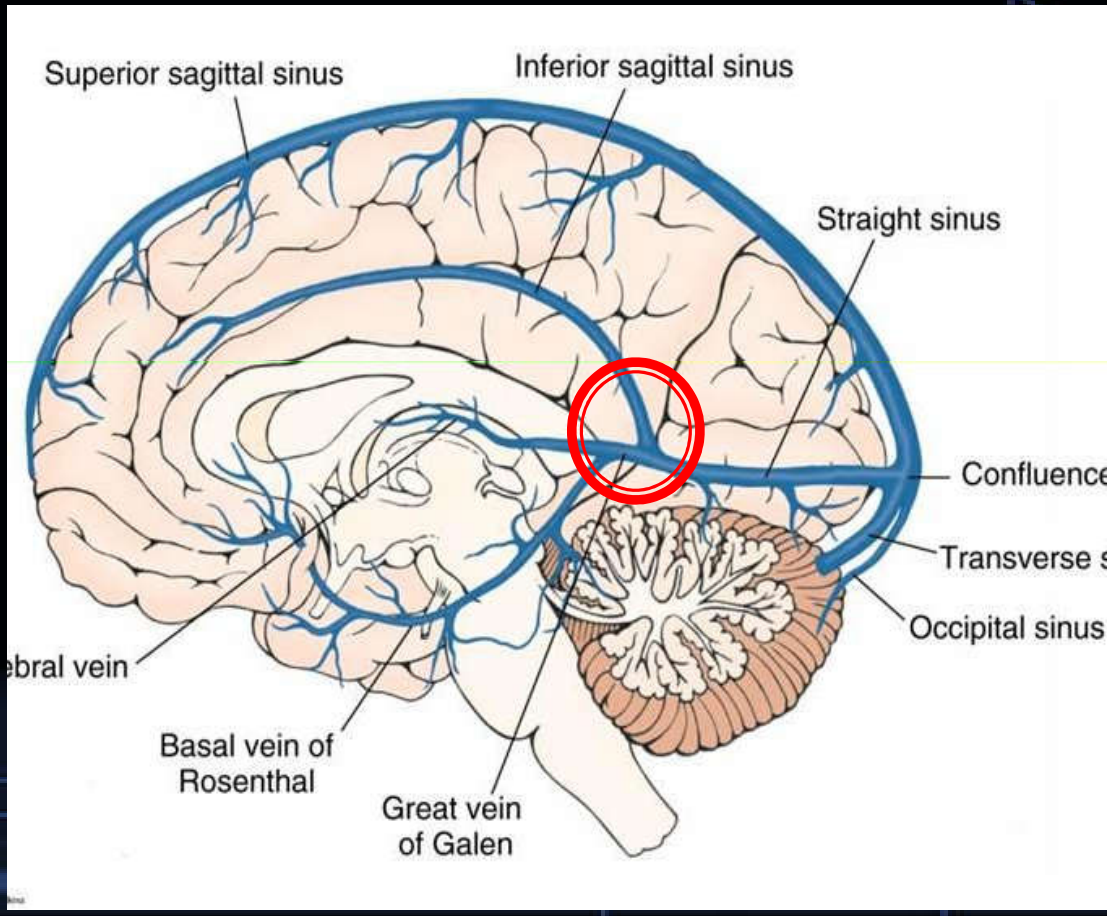
- Congenital malformation, *arteriovenous fistula*
- Weeks 6-11 of fetal development
- Incidence: unknown

1% all intracranial vascular malformations

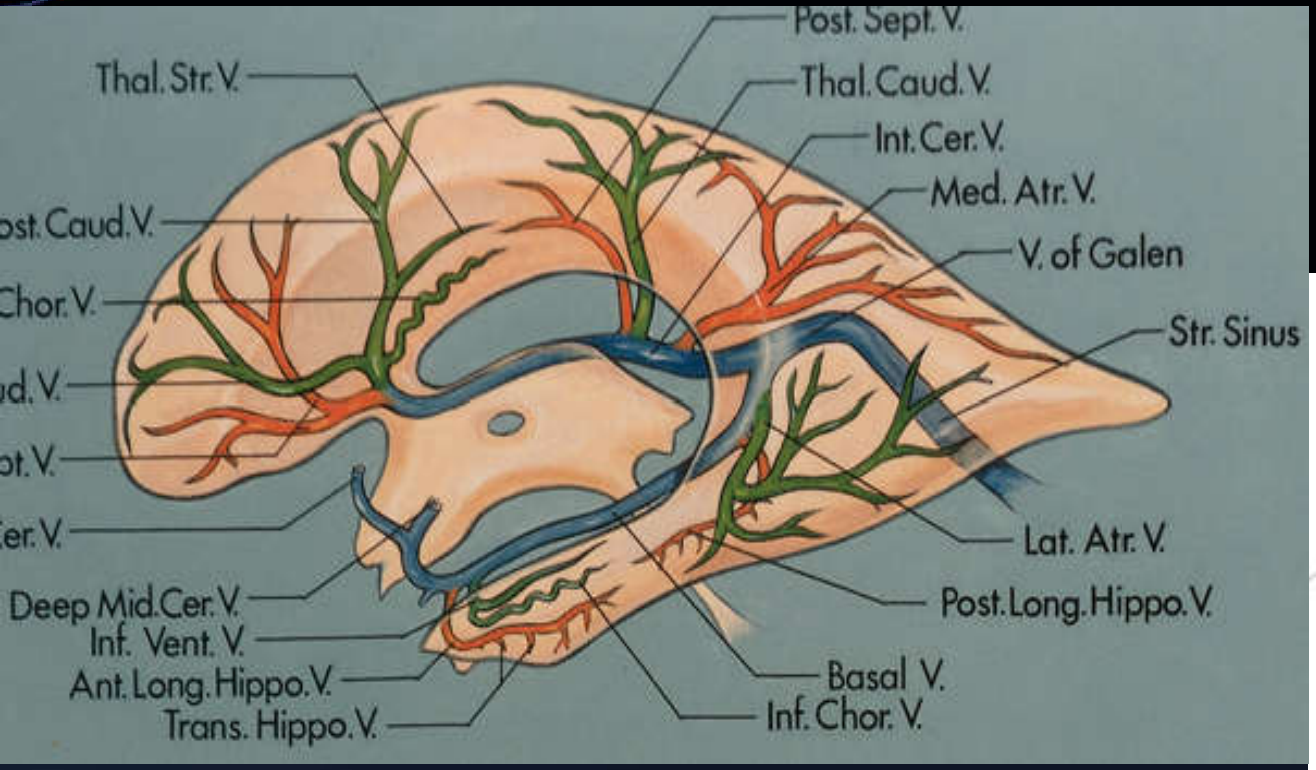


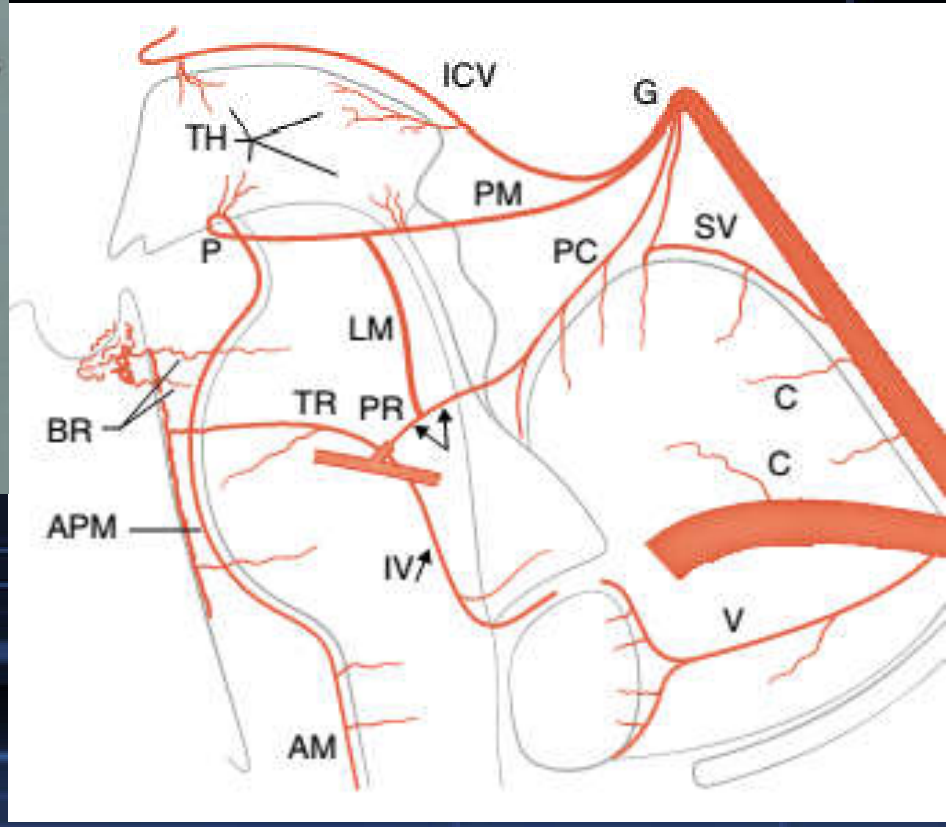
ANATOMY


Cortical Vein
Inferior Sagittal Sinus
Thalamostriate Vein
Vein of Trolard
(Superior Anastamotic Vein)
Internal Cerebral Vein
Superficial Middle
Cerebral Vein
Vein of Labbe
(Inferior
Anastamotic Vein)
Basal Vein
of Rosenthal
Sigmoid
Sinus
Transverse Sinus
Straight Sinus
Vein of Galen
Jugular Vein
Sigmoid Sinus
Internal Cerebral Vein
Thalamostriate Vein
Basilar Venous Plexus
Superior Petrosal Sinus
Basal Vein of Rosenthal
Cavernous Sinus


Superior sagittal sinus
Inferior sagittal sinus
Straight sinus
Confluence
Transverse
Occipital sinus
Great vein
of Galen
Basal vein of
Rosenthal

ANATOMY


Post. Sept. V.
Thal. Str. V.
Thal. Caud. V.
Int. Cer. V.
Med. Atr. V.
V. of Galen
Str. Sinus
Lat. Atr. V.
Post. Long. Hippo. V.
Basal V.
Inf. Chor. V.
Deep Mid. Cer. V.
Inf. Vent. V.
Ant. Long. Hippo. V.
Trans. Hippo. V.


ICV
G
TH
PM
SV
P
PC
LM
TR
PR
C
C
BR
APM
IV
V
AM

PATHOLOGY

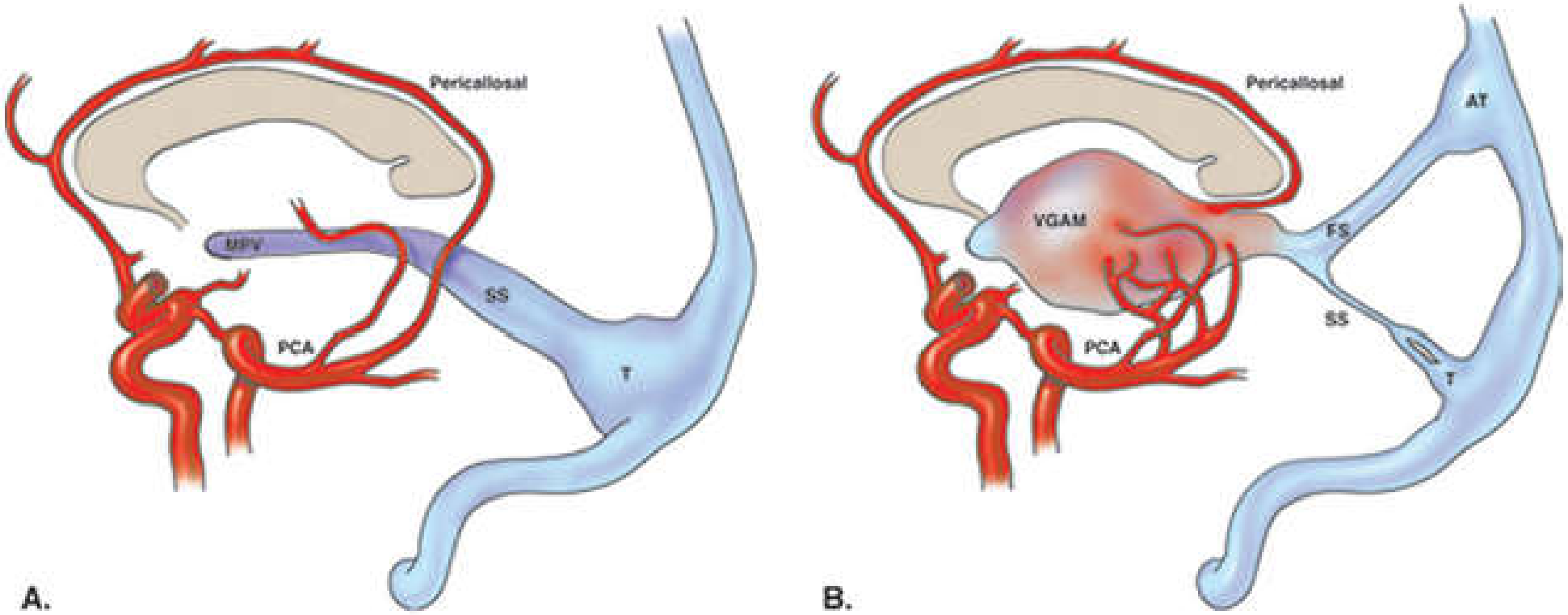


Fig. 2. Schematic illustration of the development of a VGAM and its tributaries. **A:** Note the normal anastomosis betwe the posterior branches of the pericallosal arteries and distal branches of the posterior cerebral arteries (PCA) during fe life. **B:** In VGAMs, those anastomoses degenerate and end up in the enlarged and malformed median prosencephalic vein Markowski (MPV), traditionally called a VGAM. Associated malformations are stenosis, fenestration, duplication, or absence straight sinus (SS), and decrease or absence of torcula (T). Sometimes there is an aberrant presence of falcine sinus (FS) a an accessory torcula (AT).

CLASSIFICATION

BLE 6: Angiographic comparison of all the existing and new classification systems*

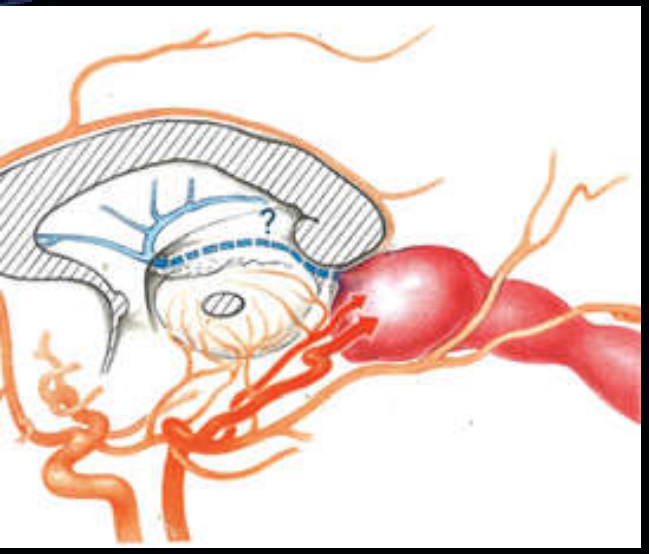
Litvak	Yaşargil	Lasjaunias	Proposed Classification
A	I	Type II (mural)	0
	II	Type I (choroidal)	1
	III		
B	IVA–B		excluded
C	IVC		

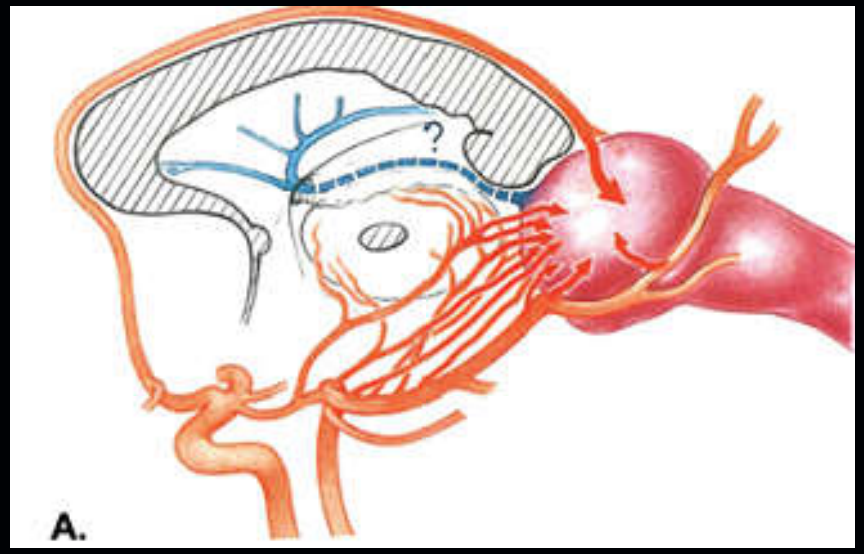
lote the exclusion of true AVM in the new proposed classification system. Note also that there is no perfect comparison a
e between the new proposed classification system and the 3 older classification systems, which do not include any cli
nptoms, age, treatment, and outcome. Only the angiographic component of our proposed classification system is shown in
e.

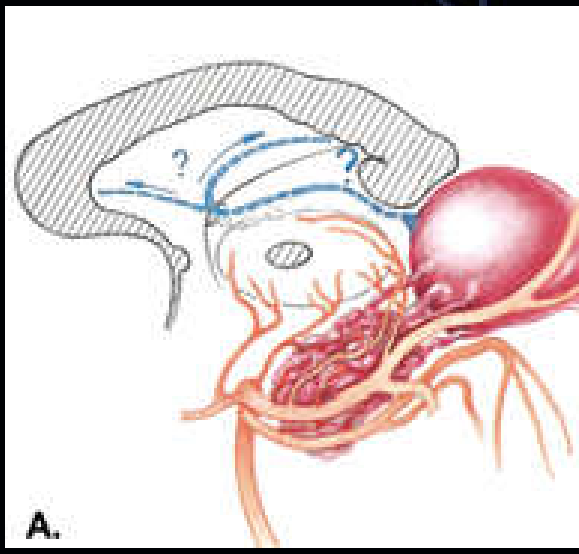
CLASSIFICATION (LASJAUNIAS)

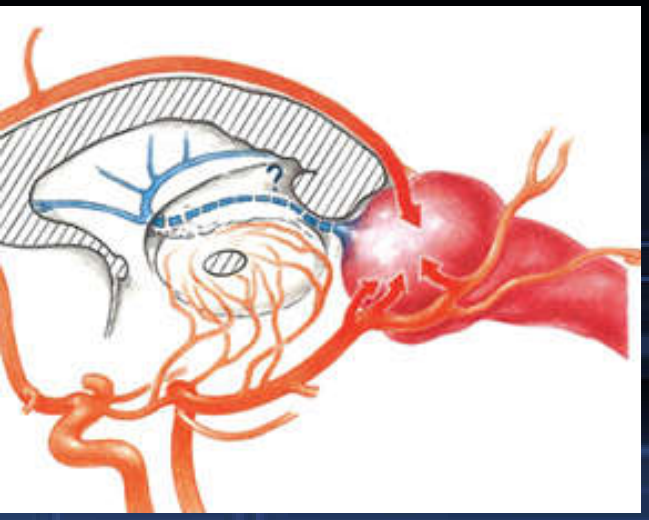
- ***Choroidal type***: Multiple high-flow fistulas
- ***Mural type***: One or few fistulas

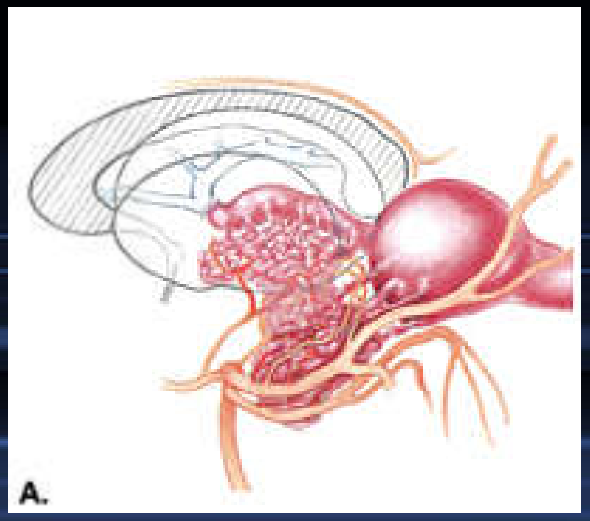
CLASSIFICATION
Yaşargil

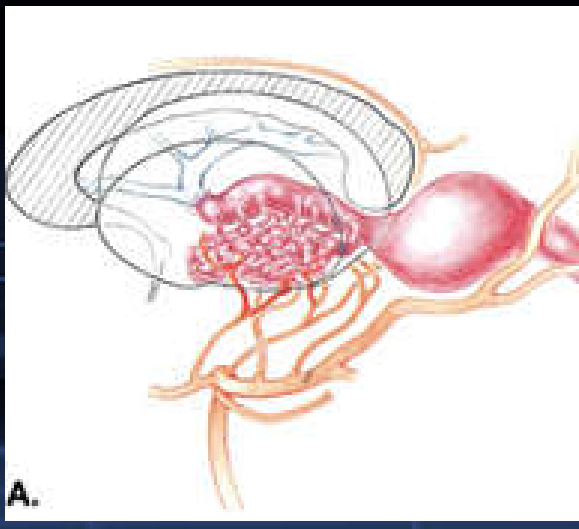




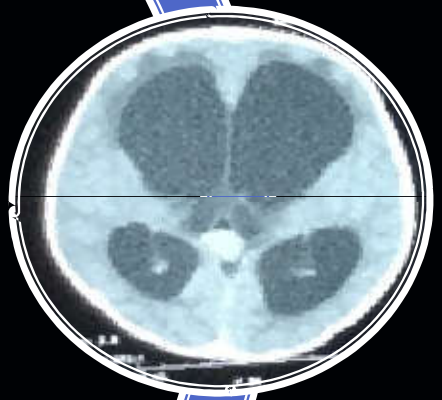




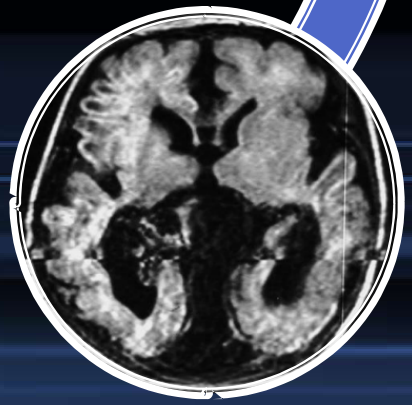




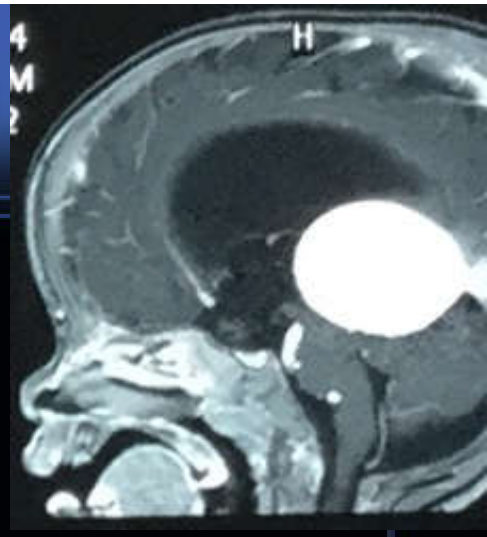
Heart failure

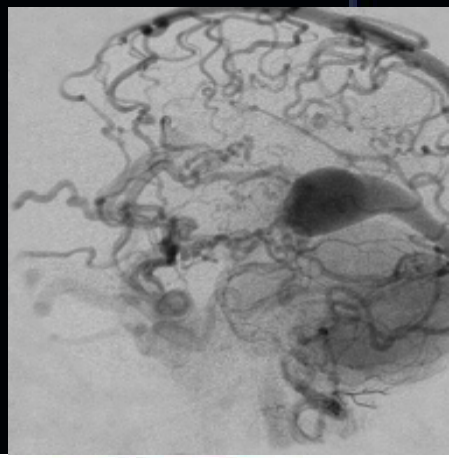


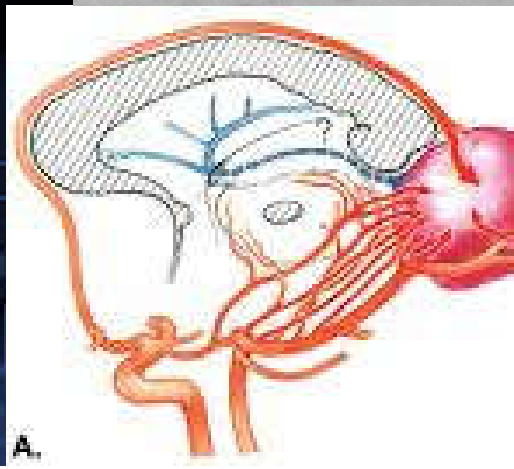
Hydro-cephalus



Seizure
Mental retardation







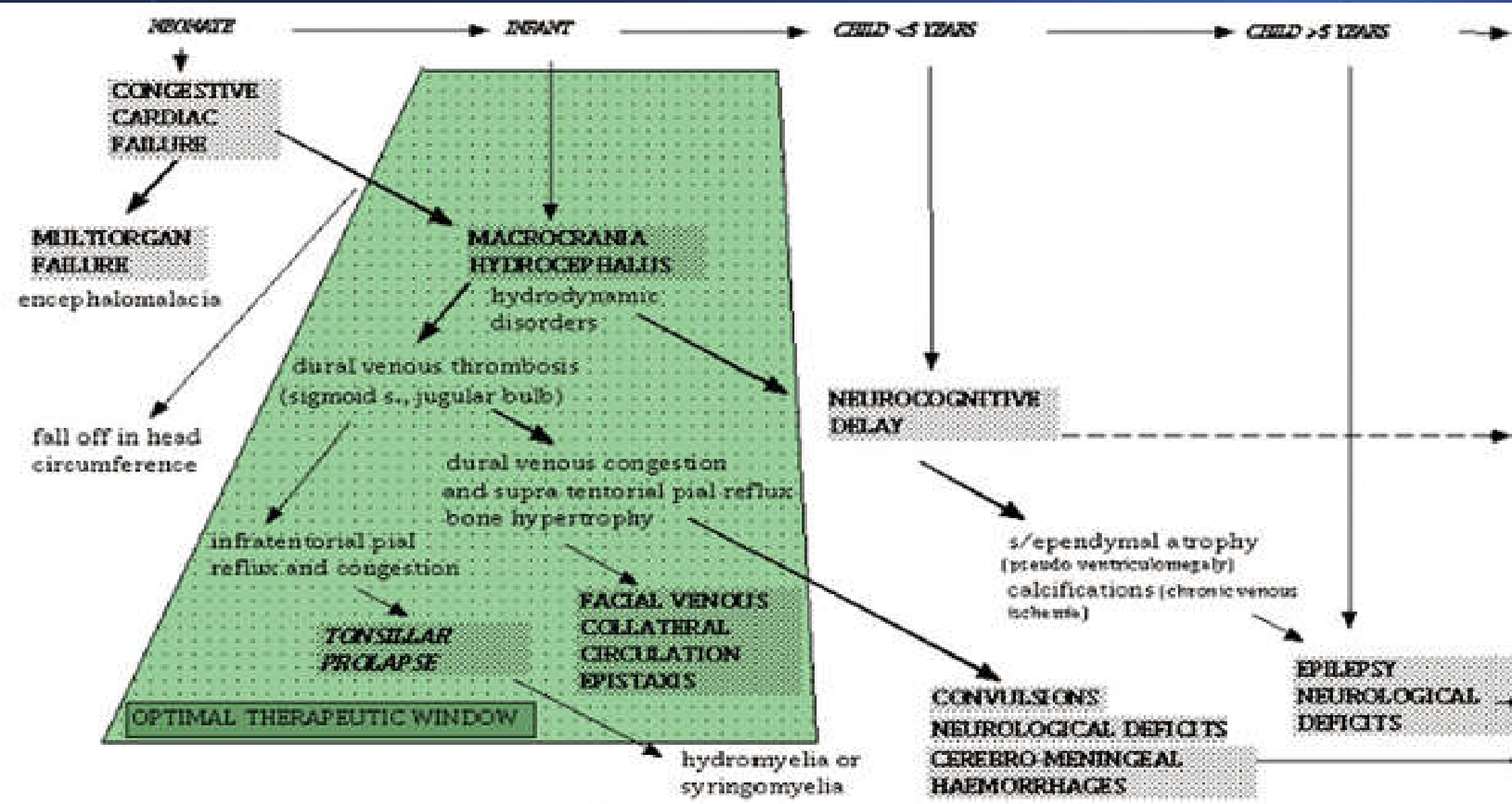
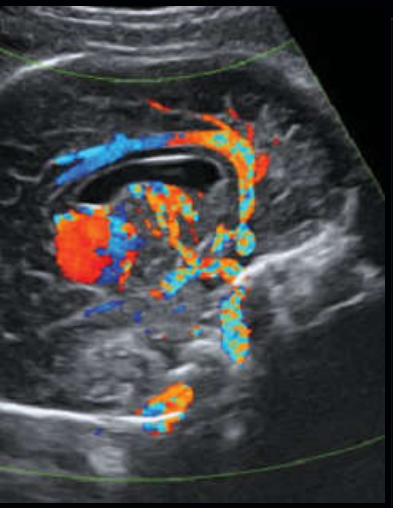
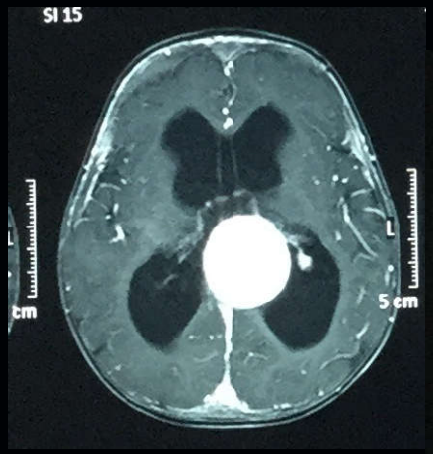
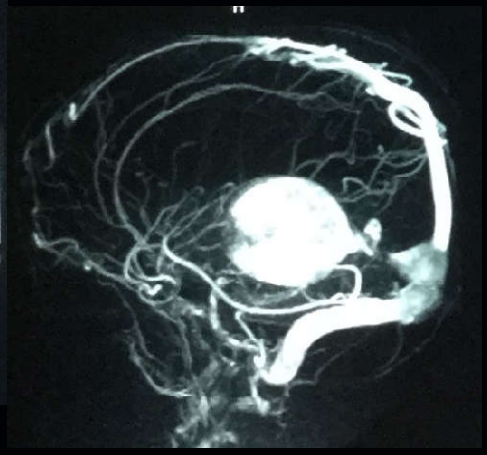


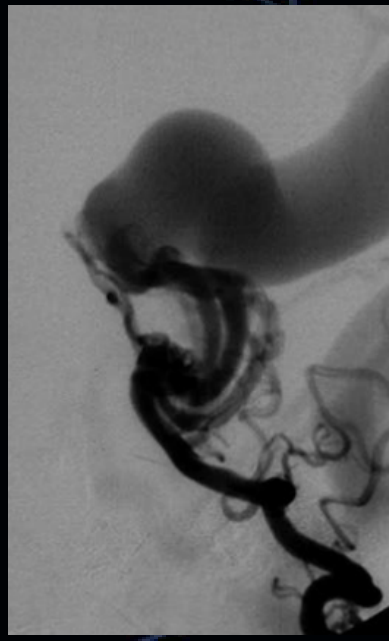
FIGURE 6. *Diagram illustrating the natural history of VGAMs over time. Note that the optimal therapeutic window is in infancy, typically at approximately 5 months of age* (from, Lasjaunias P: Vascular diseases in neonates, infants and children. Interventional Neuroradiology Management. Berlin, Springer, 1997 [15]).

DIAGNOSIS









rasound

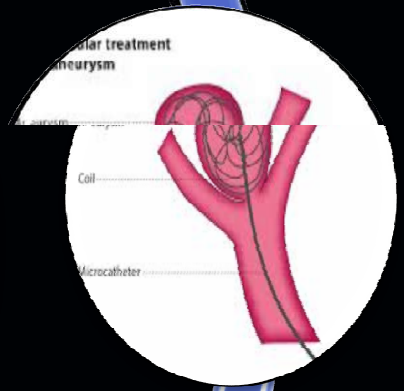
CT Scan

MRI / MRA

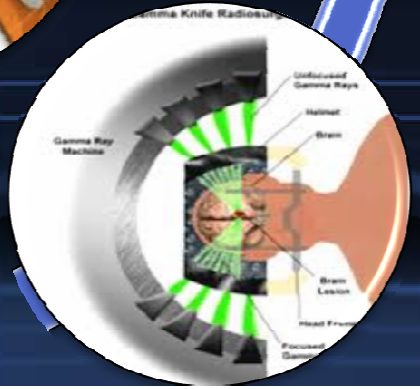
Angiograp

TREATMENT

Microsurgery



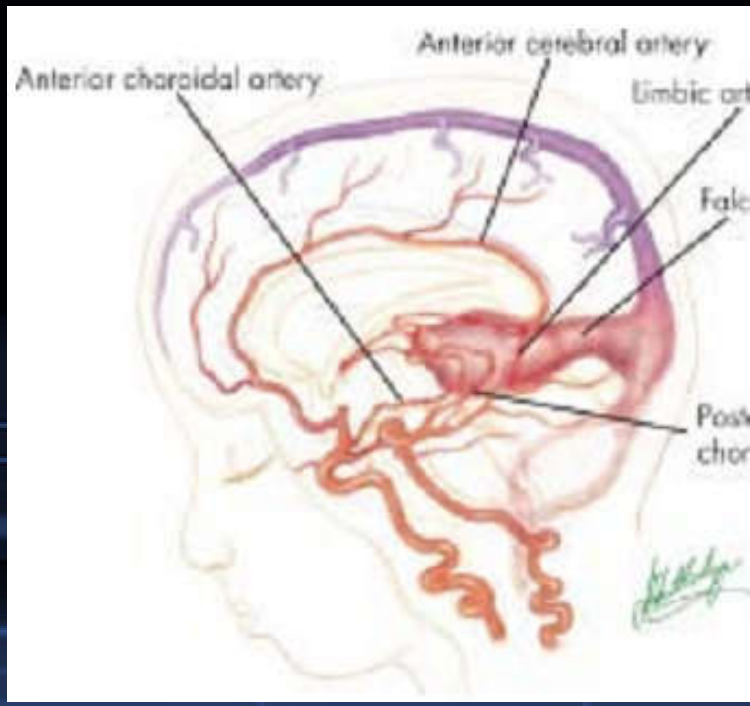
Endovascular treatment



Stereotactic Radiotherapy

GOAL

- Complete obliteration
- Hydrovenous equilibrium



Treatment - Surgery

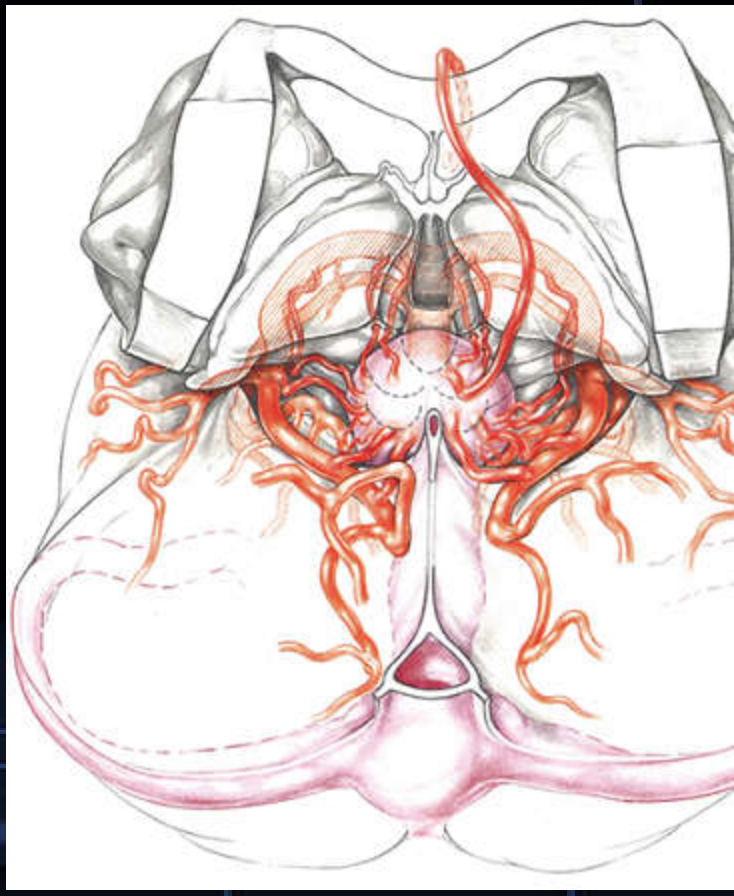
Johnston HI et al.– Neurosurgery 1987.

Mortality: 38-91% - overall group

33-77% - operated group

Stanbridge Rde et al., 1983

- Mortality : 6 /8 pts



Treatment- Endovascular Therapy

thors & Year	Outcome
Quisling, 1986	authors used transtorcular embolization of the VGM using Gianturco coils in 3 pts; satisfactory outcome in 2 pts
n et al., 1993	improved embolization techniques decreased the mortality rate from 50% (w/ 22 neonates) in 1991 to 0% (11 neo 1993
et al., 2003	embolization in 27 pts resulted in 61% having no or mild developmental delay & a 15% mortality rate during hospi
al., 2006	embolization in 13 pts w/ VGM & 2 pts w/ VGAD resulted in complete obliteration in 66%, & a mortality rate of 20 15)
as et al., 2006	endovascular embolization in 233 pts resulted in angiographically confirmed 90–100% occlusion in 55% of pts; m rate 10.6%; 74% of survivors neurologically normal on follow-up

Evolution of treatment options for vein of Galen malformations (*A review*)

- 1983-2000: 265 pts

TABLE 2: Effectiveness of VGAM management (1983–2000)

Management	Total No. Pts	No. (%) w/ Outcome*		No. of Death
		Favorable	Unfavorable	
endovascular	200	144 (72.0)	56 (28.0)	30 (15.0)
GKS	9	8 (88.9)	1 (11.1)	0 (0)
microsurgery: craniotomy & clip occl of vessel	13	2 (15.4)	11 (84.6)	11 (84.6
no endovascular or microsurgical treatment	43	10 (23.3)	33 (76.7)	33 (76.7

J Neurosurg Pediatr. 2010 Nov;6(5):444-51. doi: 10.3171/2010.8.PEDS10231.

Evolution of treatment options for vein of Galen malformatic

Khullar D[1], Andeejani AM, Bulsara KR.

Evolution of treatment options for vein of Galen malformations (*A review*)

- 2001-2010: 350 pts

BLE 4: Effectiveness of endovascular treatment (2001–2010)

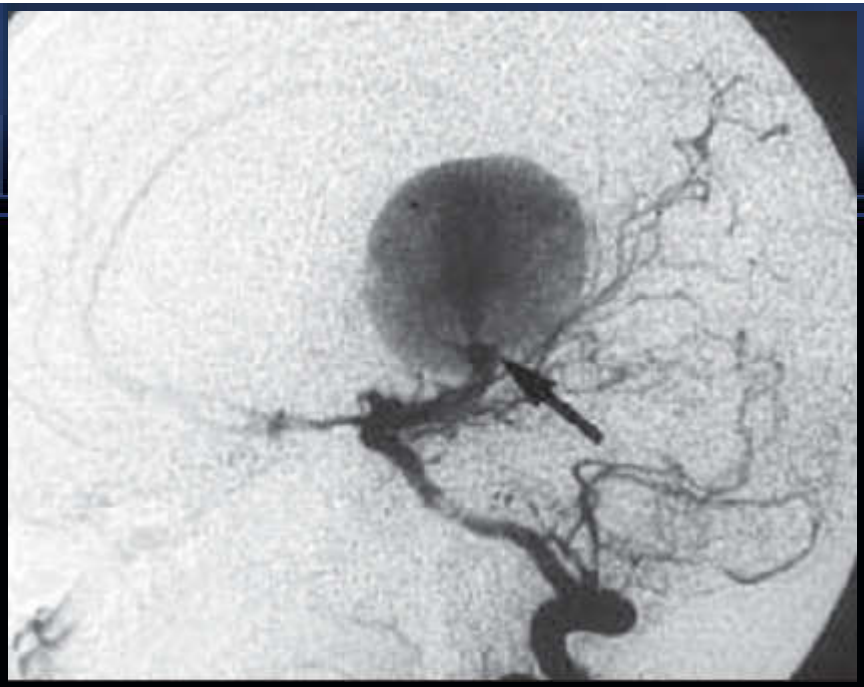
Management	Total No. of Pts	No. w/ Outcome (%)*		No. of Deaths
		Good	Fair	
endo embolization	337	205 (60.8)	79 (23.4)	53 (15.7)
no endo treatment	13	1 (7.7)	0	12 (92.3)

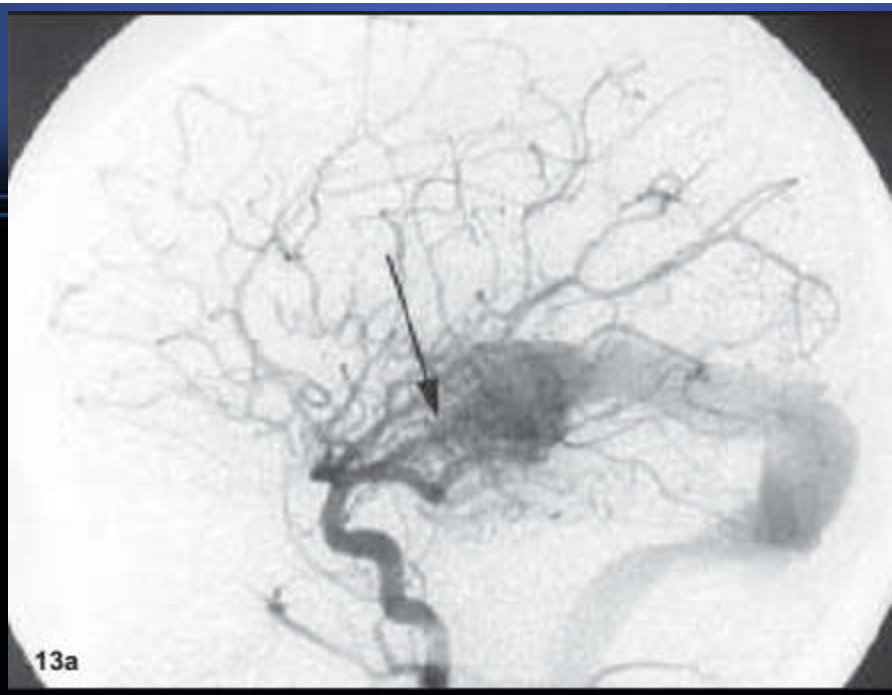
ood outcome is defined as normal or mild delay; fair is defined as moderate or severe delay.

J Neurosurg Pediatr. 2010 Nov;6(5):444-51. doi: 10.3171/2010.8.PEDS10231.

Evolution of treatment options for vein of Galen malformatic

Khullar D[1], Andeejani AM, Bulsara KR.




13a


13b

Timing of treatment

ABLE 7. Therapeutic results in the embolized group, 1981–2002[a]

	Neonates	Infants	Children	Total
rologically normal (BOS 3–5)	36.4% (4/11)	78.9% (112/142)	67.5% (27/40)	74% (143/
erate retardation (BOS 2)	54.5% (6/11)	11.3% (16/142)	20% (8/40)	15.6% (30/1
re retardation (BOS 1)	9.1% (1/11)	9.8% (14/142)	12.5% (5/40)	10.4% (20/1
h despite or because of embolization	52% (12/23)	7.2% (11/153)	0% (0/40)	10.6% (23/2

BOS, Bicêtre outcome score. Total of 216 patients, 193 surviving. Note that nearly 50% of neonates referred f anagement died. Many of these represent earlier cases that today would be scored below eight and, thus, would f to the nontreatment group.

Neurosurgery. 2006 Nov;59(5 Suppl 3):S184-94; discussion S3-13.

The management of vein of Galen aneurysmal malformati

Lasjaunias PL[1], Chng SM, Sachet M, Alvarez H, Rodesch G, Garcia-Monaco R.

Timing of treatment

TABLE 5: Age at presentation and clinical outcome for patients who underwent endovascular therapy (2001–2010)

Age at Presentation	Total	No. w/ Outcome (%)		
		Good	Fair	Died
neonates (<1 mo)	101	33 (32.7)	32 (31.7)	36 (35.6)
infants (≥1 mo to <2 yrs)	170	128 (75.3)	31 (18.2)	11 (6.5)
children or adults (≥2 yrs)	62	47 (75.8)	13 (21.0)	2 (3.2)
total	333	208 (62.5)	76 (22.8)	49 (14.7)

J Neurosurg Pediatr. 2010 Nov;6(5):444-51. doi: 10.3171/2010.8.PEDS10231.

Evolution of treatment options for vein of Galen malformatio

Khullar D[1], Andeejani AM, Bulsara KR.

EVALUATION

ABLE 4. Bicêtre neonatal evaluation score[a]

oints	Cardiac function	Cerebral function	Respiratory function	Hepatic function	Renal fun
5	Normal	Normal	Normal	—	—
4	Overload, no medical treatment	Subclinical, isolated EEG abnormalities	Tachypnea, finishes bottle	—	—
3	Failure; stable with medical treatment	Nonconvulsive intermittent neurologic signs	Tachypnea, does not finish bottle	No hepatomegaly, normal hepatic function	Normal
2	Failure; not stable with medical treatment	Isolated convulsion	Assisted ventilation, normal saturation $FIO_2 < 25\%$	Hepatomegaly, normal hepatic function	Transient a
1	Ventilation necessary	Seizures	Assisted ventilation, normal saturation $FIO_2 > 25\%$	Moderate or transient hepatic insufficiency	Unstable di with treatm
0	Resistant to medical therapy	Permanent neurological signs	Assisted ventilation, desaturation	Abnormal coagulation, elevated enzymes	Anuria

EEG, electroencephalogram; FIO_2, fractional inspired oxygen. Maximal score = 5 (cardiac) + 5 (cerebral) + 5 (respiratory) + 3 (hepatic) + 3 (renal) =

- ***<8: no treat***
- ***8-12: emergency endovascular intervention***
- ***>12: medical treatment → >5 mo***

Outcome and complications of endovascular embolization for vein of Galen malformations

- 34 studies
- Neonates: 44% - Infants: 41% - Children & Adult: 12%
- Complete : 57% - Partial : 43%
- Outcome: good: 68%- Poor: 31%
- Mortality: 10%- Complication: 37%
- Complication: Cerebral hemorrhage/ischemia, hydrocephalus, leg ischemia, vessel perforation

J Neurosurg. 2015 Oct;123(4):872-90. doi: 10.3171/2014.12.JNS141249. Epub 2015 Jul 31.

Outcome and complications of endovascular embolization for vein of Galen malformations: a systematic review and meta-analysis.

Yan J[1], Wen J[2], Gopaul R[1], Zhang CY[1], Xiao SW[1].

Full te

Save i

Outcome

TABLE 7. Therapeutic results in the embolized group, 1981–2002[a]

	Neonates	Infants	Children	Total
urologically normal (BOS 3–5)	36.4% (4/11)	78.9% (112/142)	67.5% (27/40)	74% (143/
derate retardation (BOS 2)	54.5% (6/11)	11.3% (16/142)	20% (8/40)	15.6% (30/1
vere retardation (BOS 1)	9.1% (1/11)	9.8% (14/142)	12.5% (5/40)	10.4% (20/1
ath despite or because of embolization	52% (12/23)	7.2% (11/153)	0% (0/40)	10.6% (23/2

[a] BOS, Bicêtre outcome score. Total of 216 patients, 193 surviving. Note that nearly 50% of neonates referred f management died. Many of these represent earlier cases that today would be scored below eight and, thus, would fa into the nontreatment group.

Neurosurgery. 2006 Nov;59(5 Suppl 3):S184-94; discussion S3-13.

The management of vein of Galen aneurysmal malformation

Lasjaunias PL[1], Chng SM, Sachet M, Alvarez H, Rodesch G, Garcia-Monaco R.

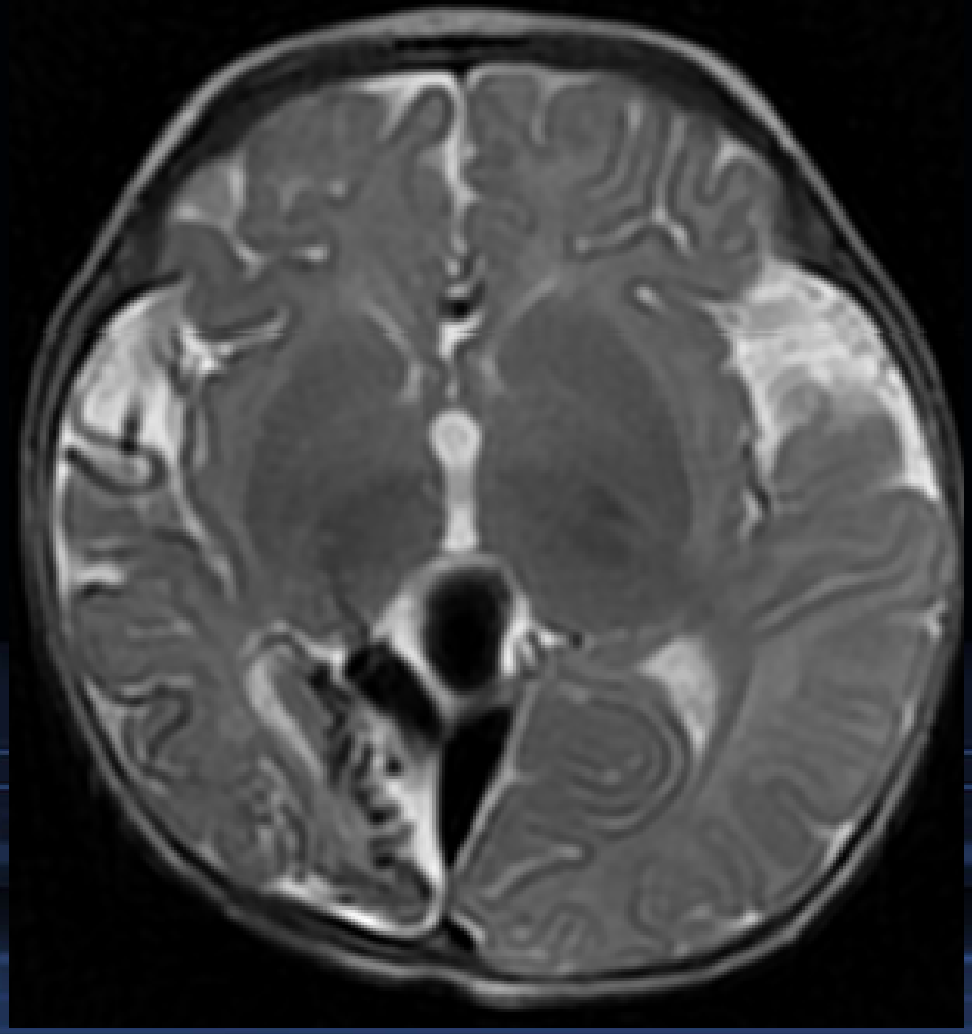
Case Report

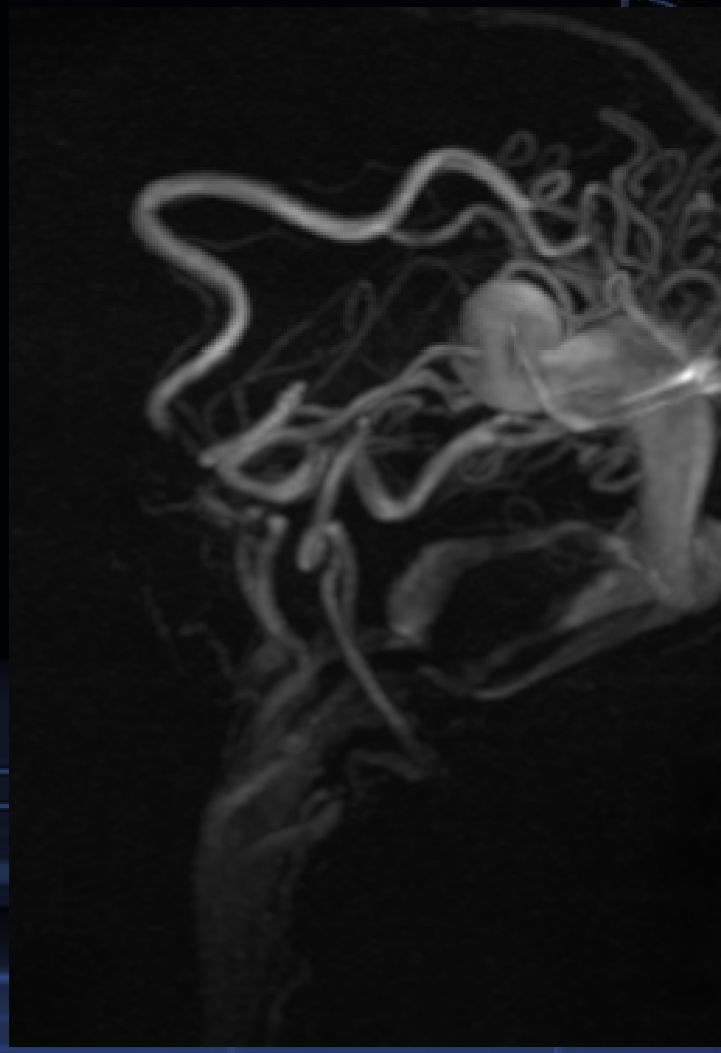
oy, 3 mo

lild heart
ailure

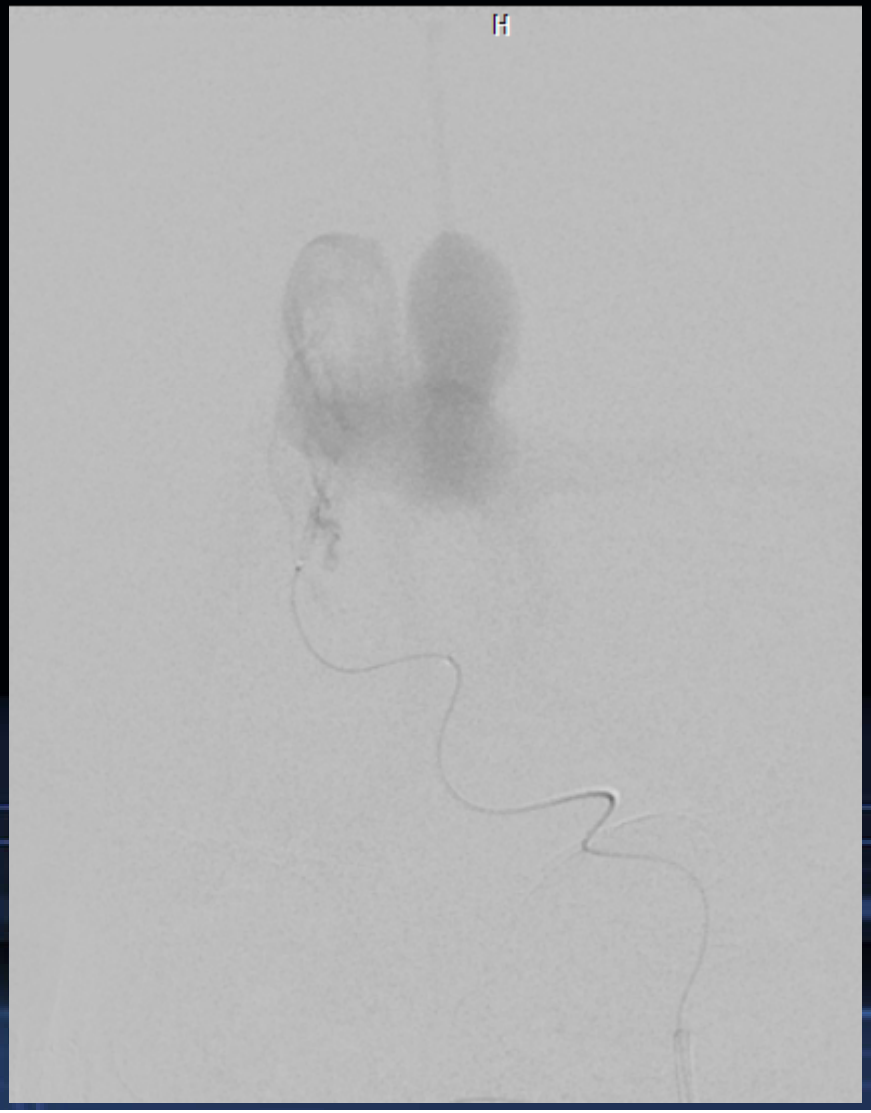
AP: 30mmHg

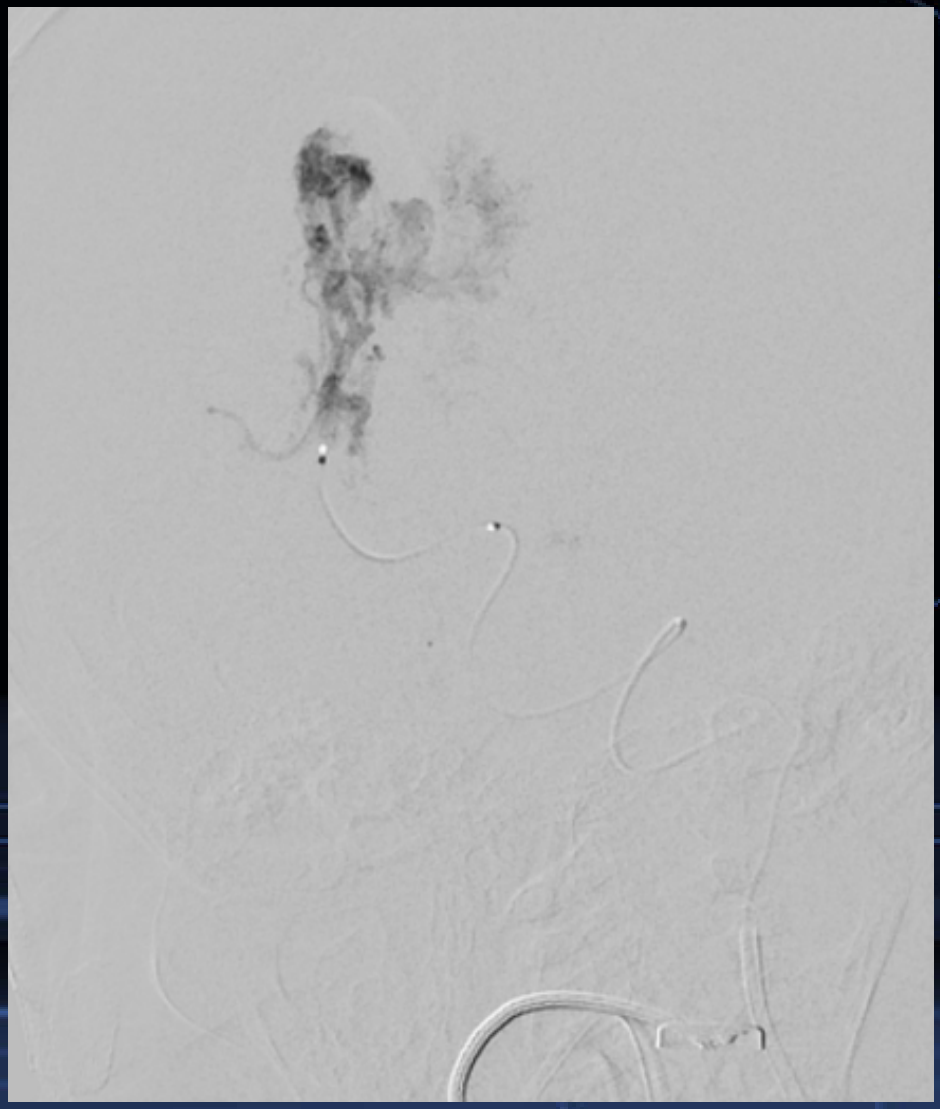
lurmur on the
ead



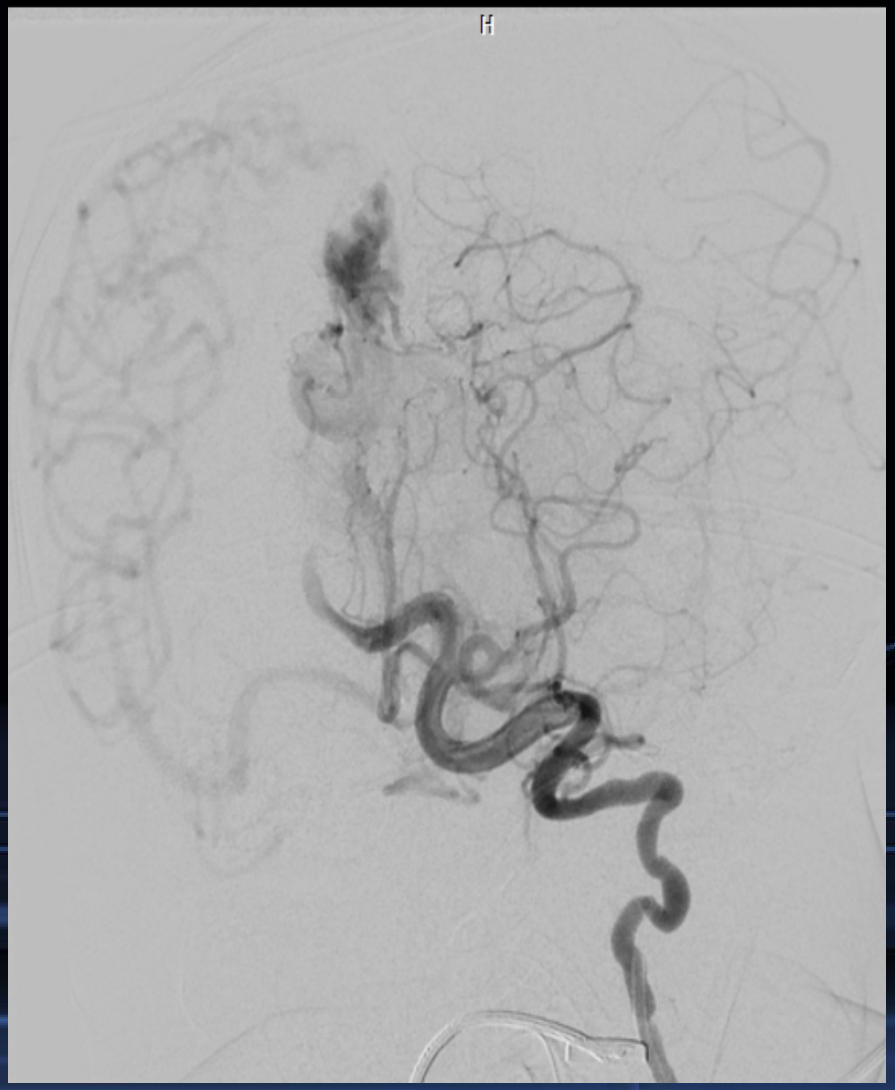


Case Report





Case Report



CONCLUSIONS

- A congenital AVF.
- Untreated → poor outcome
- Endovascular therapy is effective, acceptable mortality rate, complications and good clinical outcome

Thank you for your attention