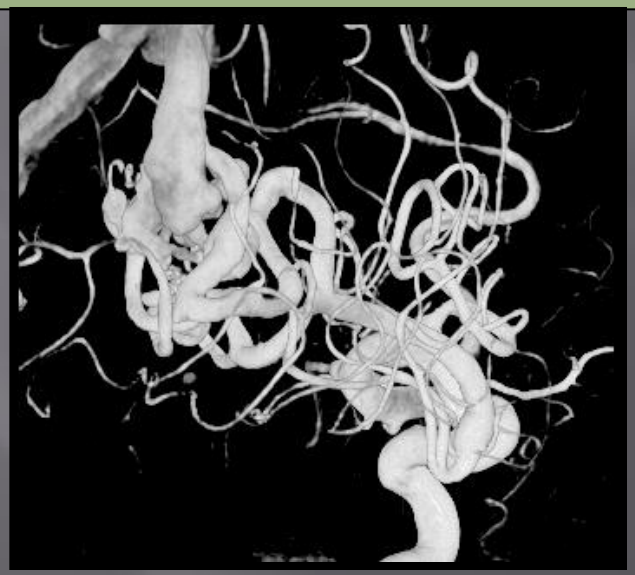
PEDIATRIC INTRACRANIAL HIGH FLOW ARTERIOVENOUS FISTULAE.

DIVISION OF INTERVENTIONAL NEURORADIOLOGY
DAVID GEFFEN SCHOOL OF MEDICINE
UNIVERSITY OF CALIFORNIA IN LOS ANGELES
XXV S.I.M.I.- 25th ANIVERSARIO.
4-6 DE JULIO 2016 HOTEL HILTON
BUENOS AIRES ARGENTINA.



Pial AVF:Incidence

Approximately 1.6% of all intracranial vascular malformations.

Approximately 30% of all pediatric intracranial vascular malfortmatitons.

INTRACRANIAL LOCALIZATION

VEIN OF GALEN A/V FISTULAE.

NON-GALENIC PIAL A/V FISTULAE
Cortical.
Deep.

DURAL A/V FISTULAE.

CLINICAL PRESENTATION

Clinical Presentation	Pathophysiology
Intracranial bruit	Incr. a/v shunt.
CHF	Incr. a/v shunt
Seizures	Venous hypert.
P.N.D.	Varix compress.
	.
Hemorrhage	Venous occluss. Hemorrh. Infarct.

Pial AVF: Pathophysiology

- **Hydrovenous Disorder**

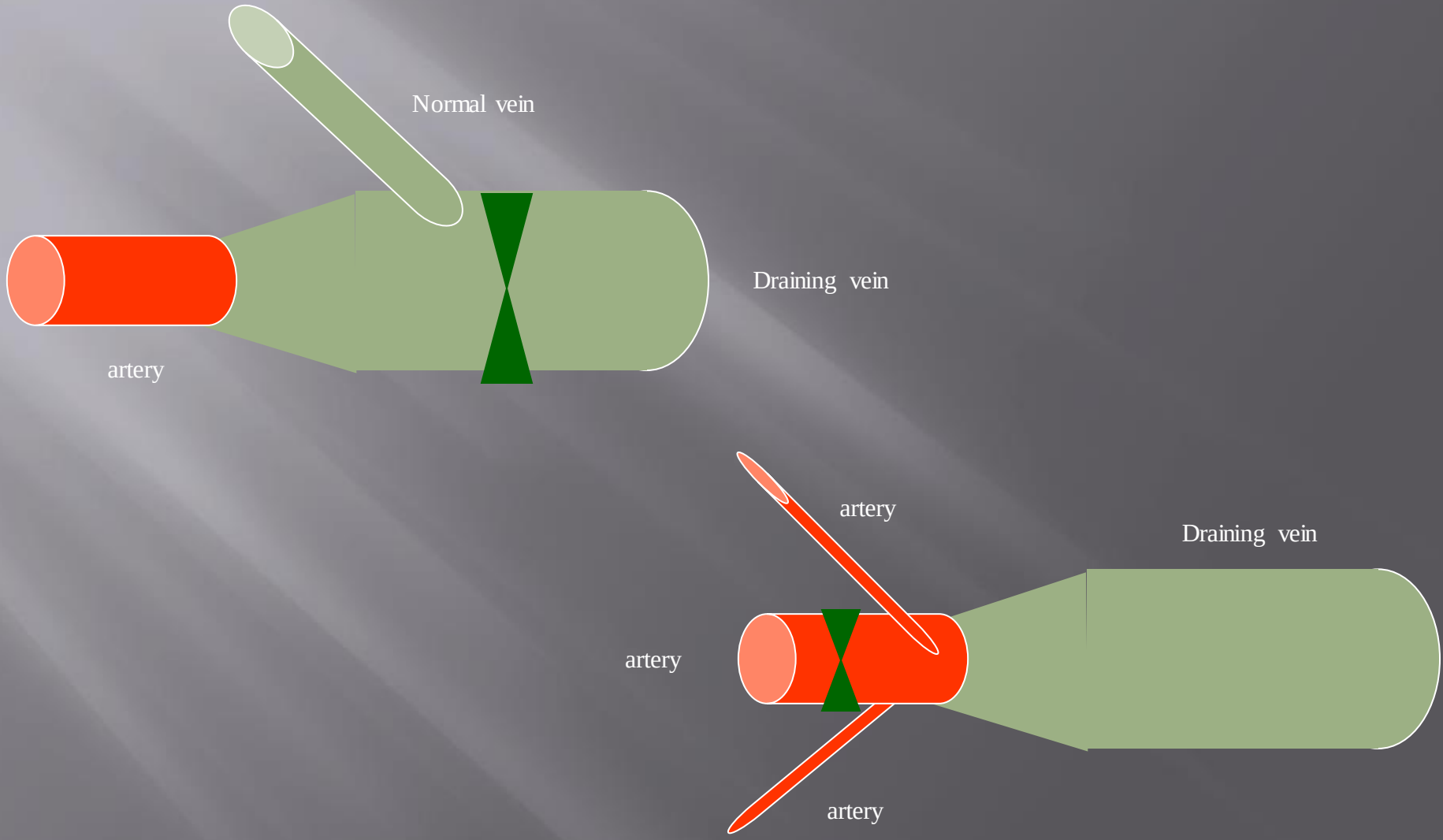
  Lasjaunias (1992,1994)

- **Most symptoms related to venous HYPERTENSION:**
  - **High flow leads to elevated venous/sinus pressure (outlet obstruction), then to impaired CSF dynamics :NON-COMMUNICATING HYDROCEPHALUS.**

THERAPEUTIC GOAL

- **ENDOVASCULAR OCCLUSION AT THE SITE OF THE ARTERIOVENOUS FISTULA.**
- **PROXIMAL OCCLUSION ELICITS EARLY RECANALIZATION.**
- **DISTAL VENOUS OCCLUSION ELICITS ACUTE VENOUS HYPERTENSION AND HEMORRHAGE.**

AVF: Occlusion Location



THERAPEUTIC APPROACHES

- **TRANSARTERIAL ++++**
- **TRANSVENOUS ++**
- **COMBINED ++**

EMBOLIC MATERIALS

- **DETACHABLE BALLOONS**
- **COILS AND MICROCOILS**
- **LIQUID EMBOLIC AGENT Acrylics.**
- **Onyx.**

TECHNICAL COMPLICATIONS

- OCCLUSION OF VENOUS OUTLET DISTAL TO A/V SHUNTING.
- DISTAL MIGRATION OF EMBOLIC MATERIAL WITH OCCLUSION OF DURAL SINUSES OR PULMONARY CIRCULATION.

PROCEDURAL MORBIDITY

- ICH
  - venous rupture.
  - arterial rupture.
  - venous infarct.
- Seizures
  - varix thrombus.
  - venous hypert.
  - venous infarction.
- PND
  - varix thrombosis.

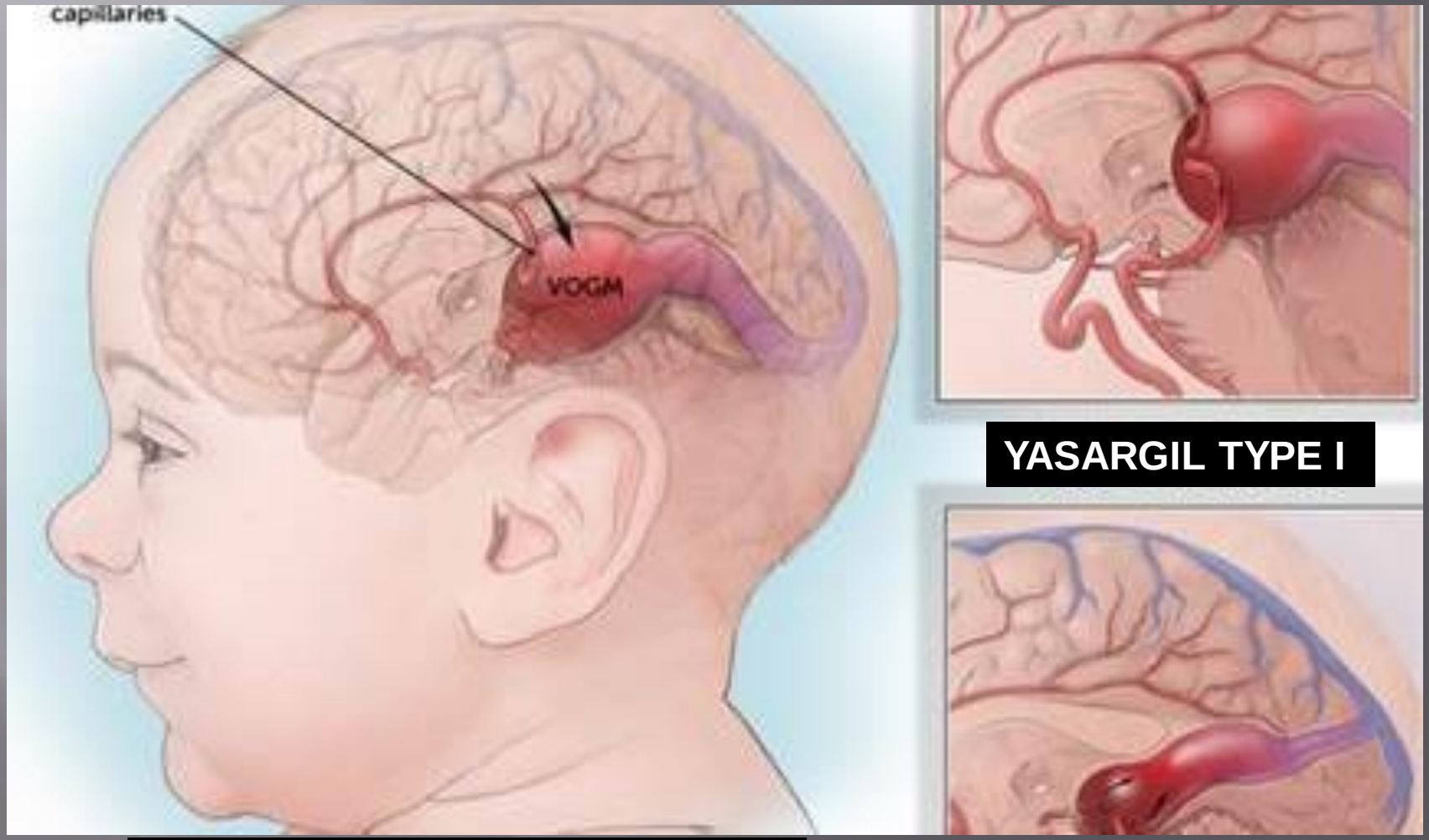
VEIN OF GALEN MALFORMATIONS.

- VOGMs are rare congenital vascular malformations resulting from the development of arteriovenous (AV) connections between primitive choroidal vessels (**limbic vascular system)** and the median prosencephalic vein of Markowski.

- The malformation develops between 6th and 11th week of fetal development.

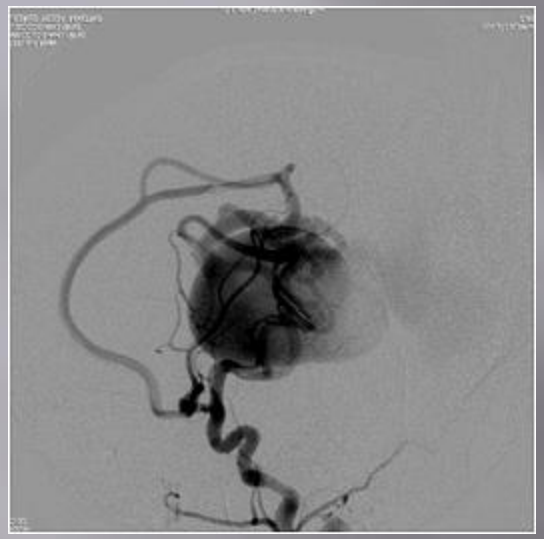
- IT comprises less than 1% of cerebral vascular malformations at any age, but in the pediatric vascular malformation group it might account up to 30%.
- The natural history of these lesions is mostly poor with a mortality of 42–91% if left untreated.

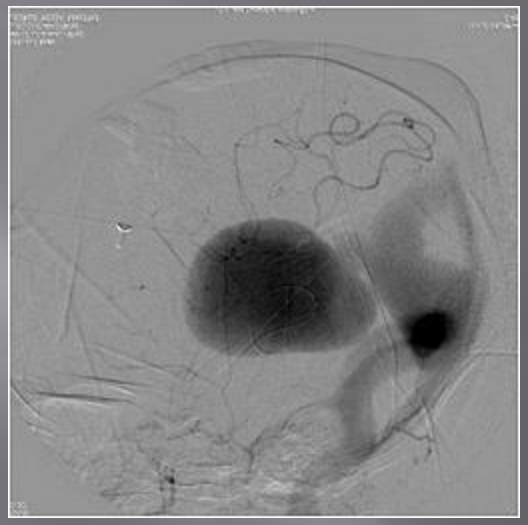
VEIN OF GALEN MALFORMATIONS (VOGM).

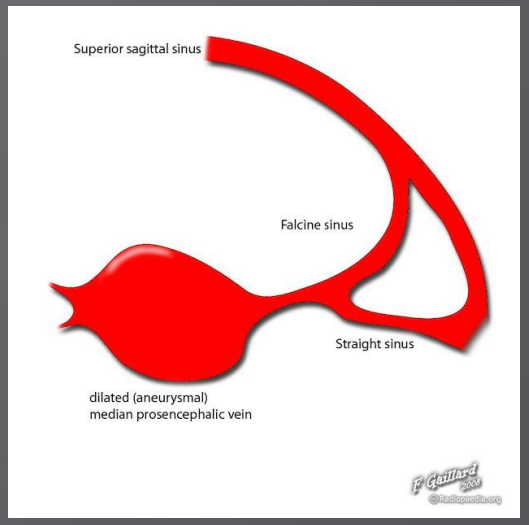


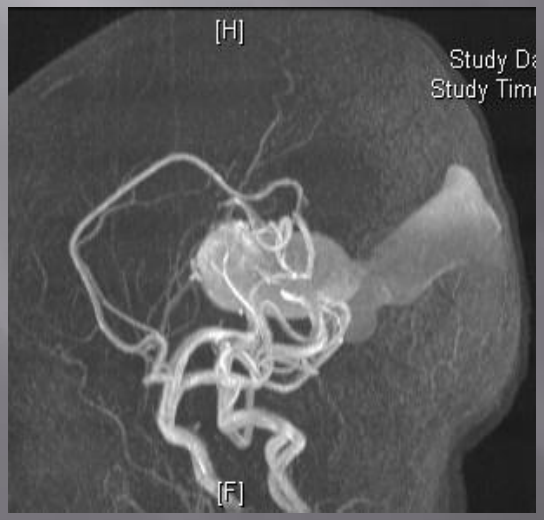
BOSTON CHILDREN'S HOSPITAL

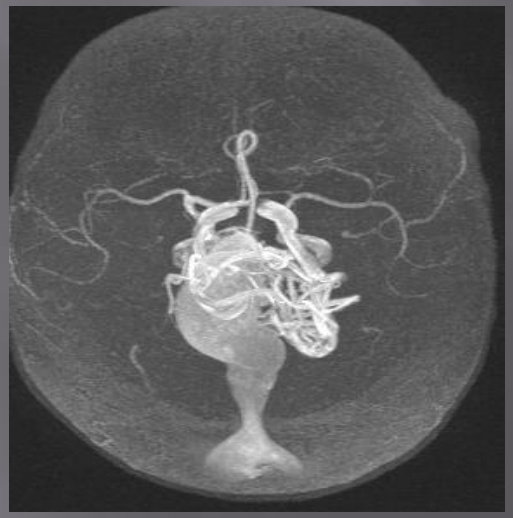
VEIN OF GALEN VASCULAR MALFORMATION YASARGYL TYPE I.

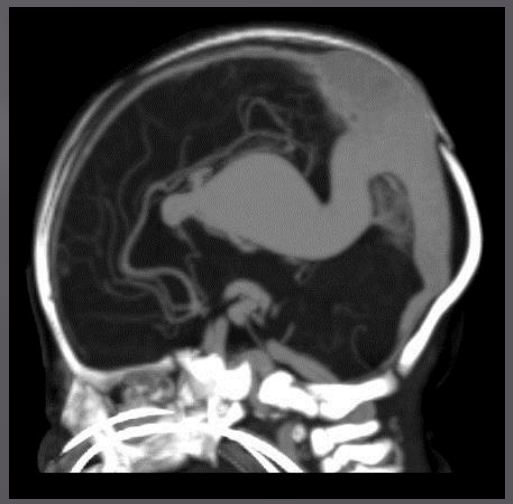




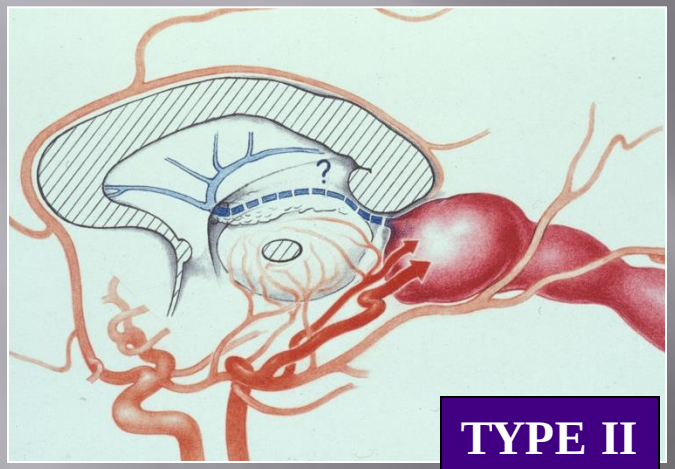


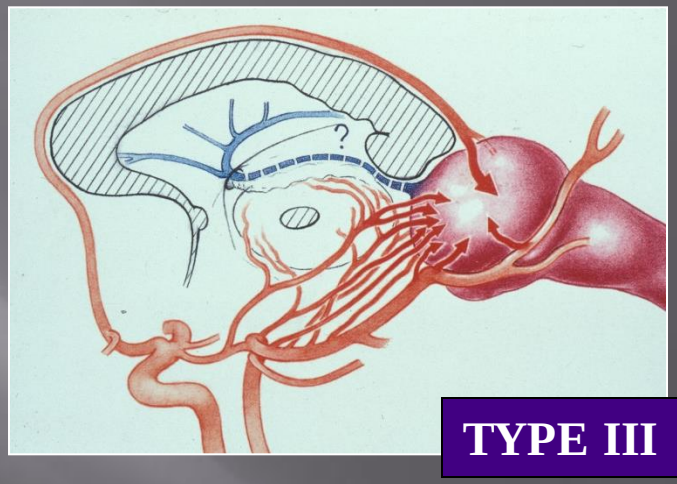


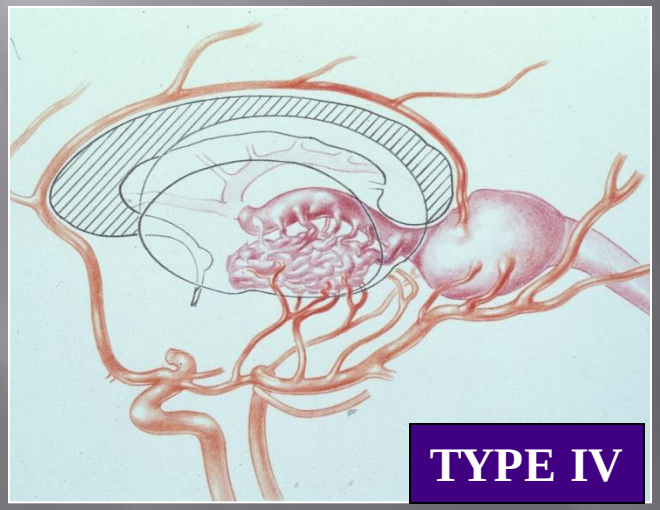


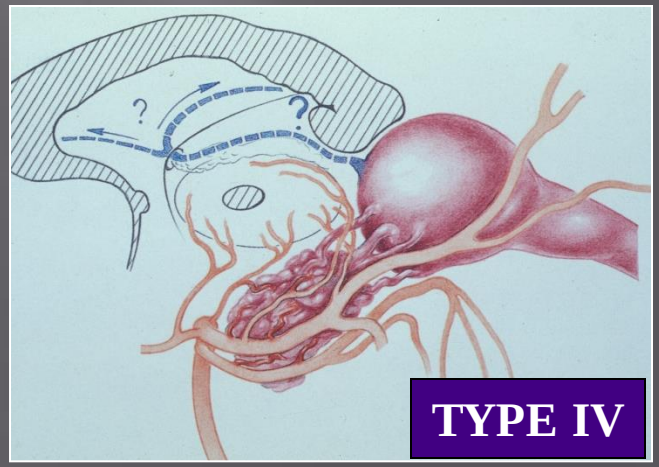


GALENIC MALFORMATIONS YASARGIL CLASSIFICATION







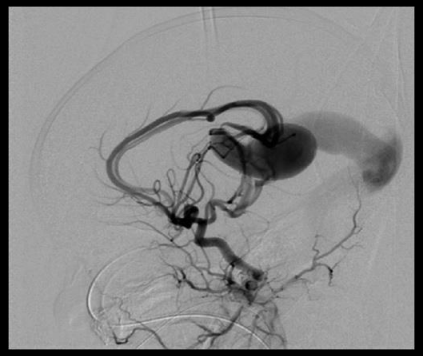


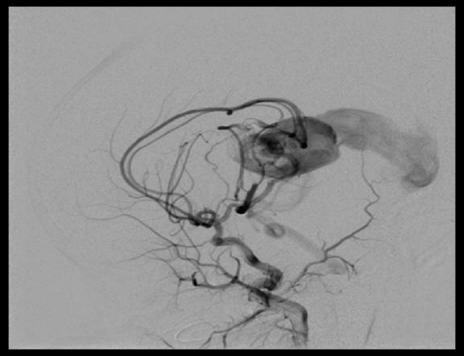
VEIN OF GALEN MALFORMATION

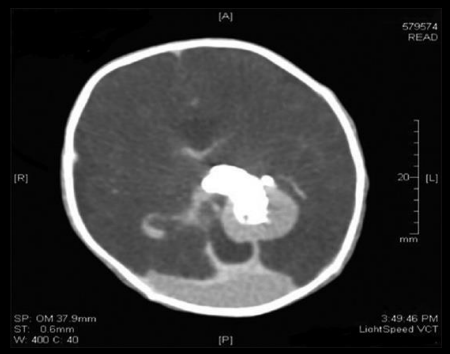
INTERDISCIPINARY TEAM	CLINICAL EVALUATION
Neonatal Intensive care. Cardiology. Neurology. Neuroanaesthesiology. Neurointervencionist. Pediatric neurosurgery. Medico-surgical Intensive Care Unit.	▣ Weight/ head circunference. ▣ Renal/liver functions. ▣ Encephalomalacia. ▣ Cardiac malformations. ▣ Brain MRI. ▣ Cerebral angio: **therapy.**

Ramakrishnan RM, Goraksha SU, Thakore BP, Monteiro JN, Butani MT. Anaesthetic management of vein of Galen malformation in a very low birth weight preterm baby for endovascular embolisation. J Neuroanaesthesiol Crit Care 2016;3:137-40

A preterm neonate delivered by caesarean section for foetal distress, with a very low birth weight of 1.75 kg, developed respiratory and severe cardiac failure soon after birth. On systemic examination, the child had a hyperdynamic precordium with a pansystolic murmur in the mitral area and hepatomegaly. The blood investigations revealed a deranged coagulation profile with a raised activated partial thromboplastin time - 51.9 seconds. Serum electrolytes were Na – 140 meq/l, K - 4.2 meq/l and the arterial blood gas showed PH - 7.3, PCO2 - 43.5, PO2- 70.5, HCO3- 21.8 on FiO2 0.4. Two dimensional echocardiography demonstrated severe pulmonary arterial hypertension with a ventricular septal defect with the right to left shunt.





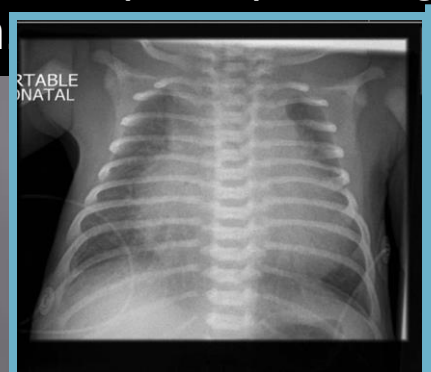


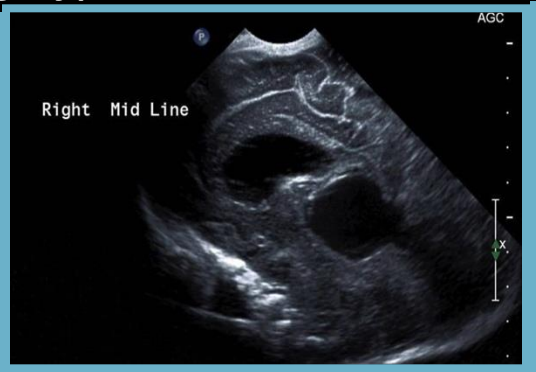
Vein of Galen malformation presenting as persistent pulmonary hypertension of newborn (PPHN).

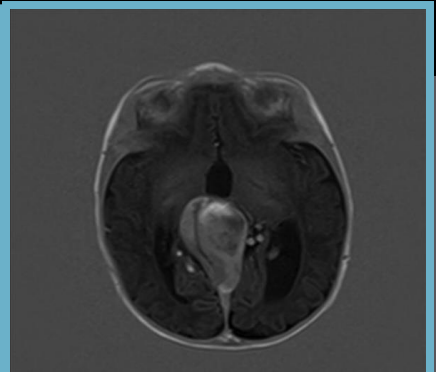
Tiwary S, Geethanath RM, Abu-Harb M.

BMJ Case Rep. 2013 Sep 26;2013. pii: bcr2013200425. doi: 10.1136/bcr-2013-200425.
PMID: 24072831

Term baby boy delivered by elective caesarean section at a local district hospital. Birth weight was 3650 g (75th centile) and head circumference 37.5 cm (91st centile). The pregnancy was uncomplicated with normal antenatal anomaly scan at 20 weeks of gestation. This baby was born in good condition and did not need any resuscitation at birth. He was discharged home at 24 h of age. The baby presented to the accident and emergency on day 4 with feeding difficulties, breathlessness and lethargy. On examination, the baby was noted to be tachycardic, breathless with marked chest recessions, an overactive precordium, a loud systolic murmur over tricuspid area, normal preductal oxygen saturation 95%, low postductal oxygen saturation (85%) in air and cardiomegaly on the chest X-ray. The rest of the clinical examination was essentially normal (no hepatomegaly) and there was no evidence of dysmorphism



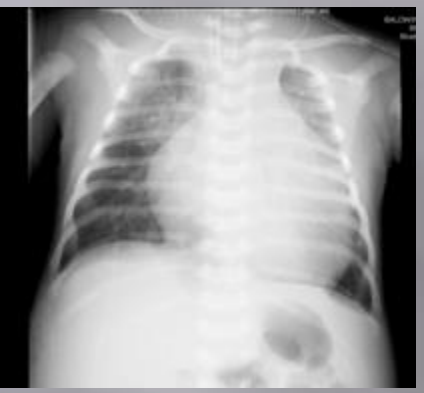
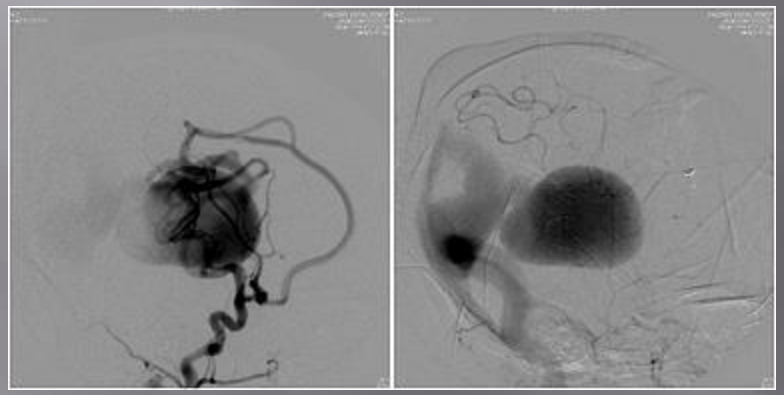
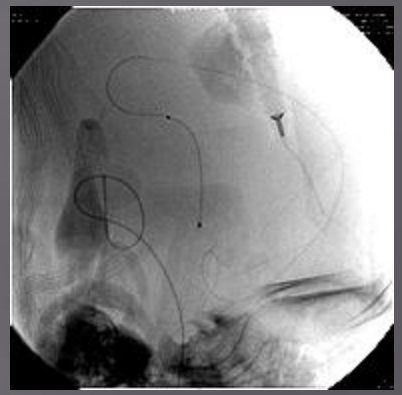
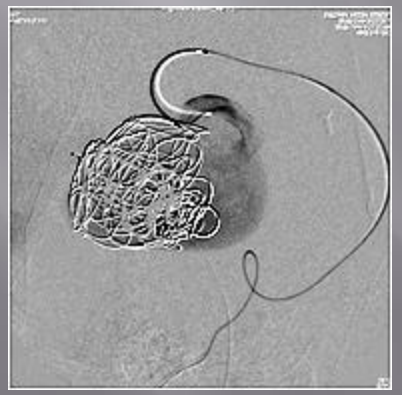
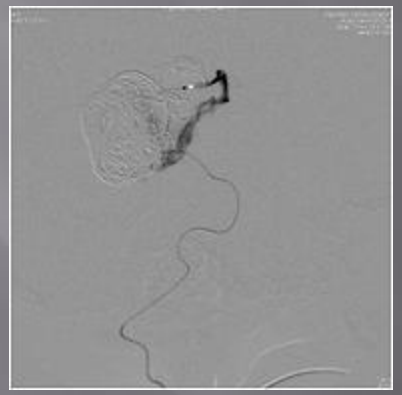
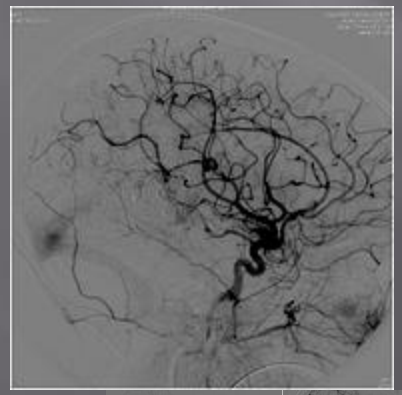
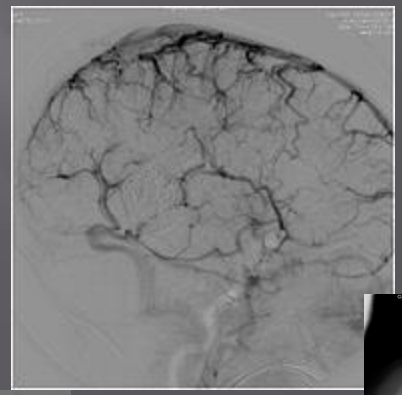
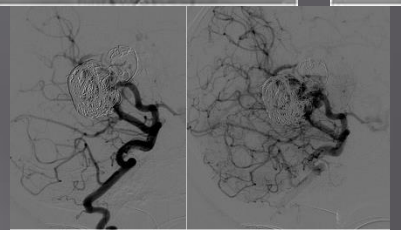
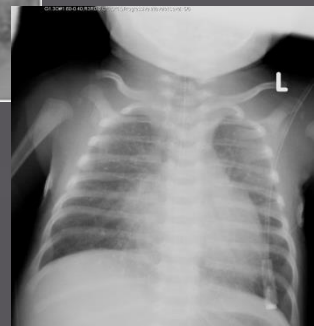




YASARGIL TYPE I

3M OLD MODERATE CHF

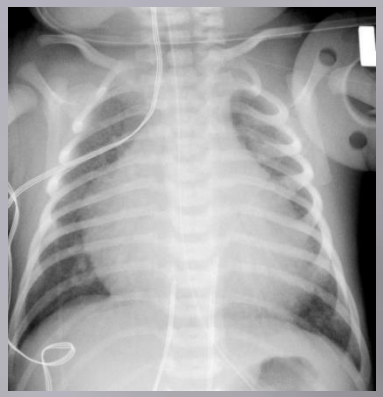
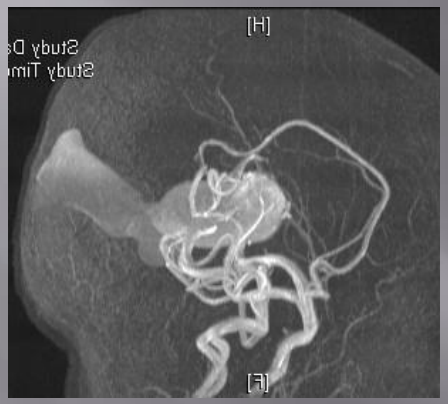
T.A. APPROACH COILS + ONYX

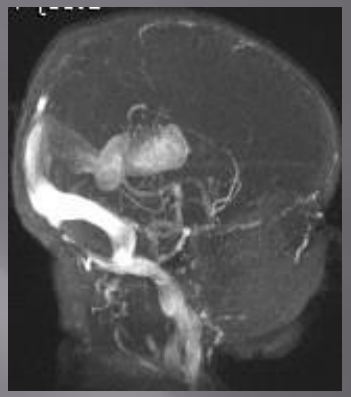
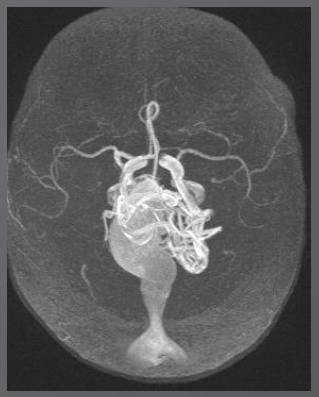
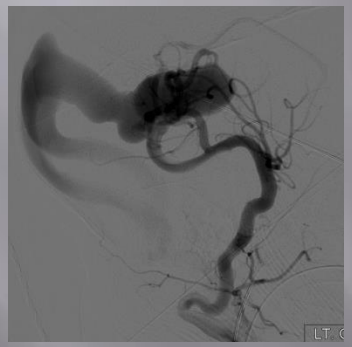
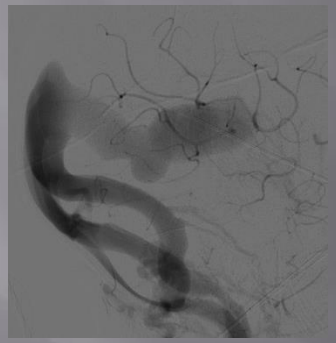
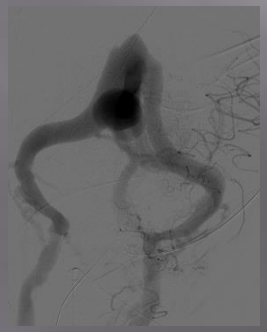
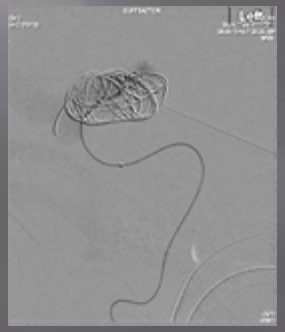
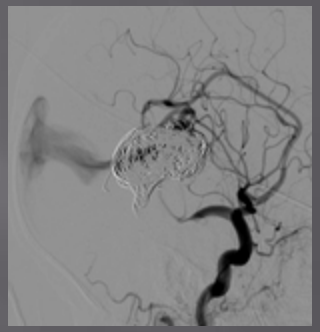
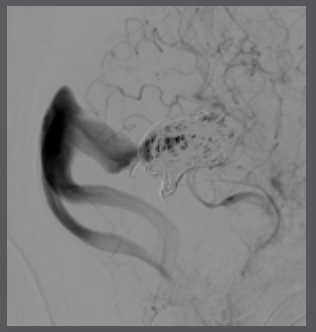
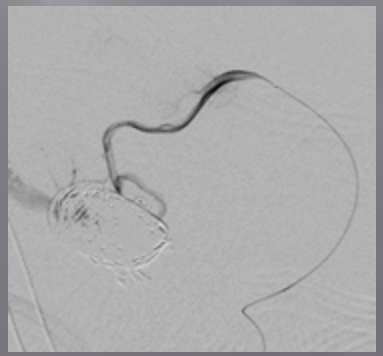
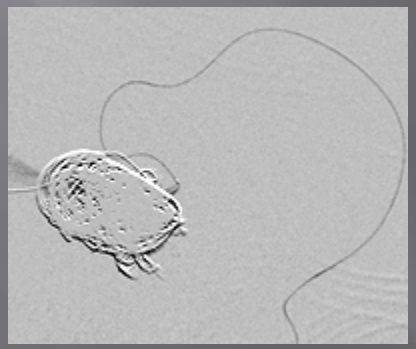
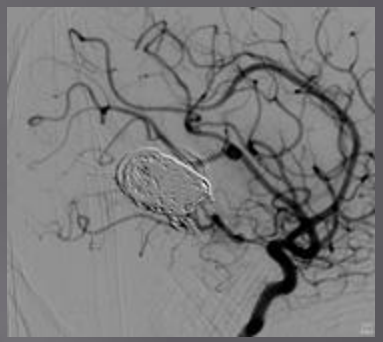
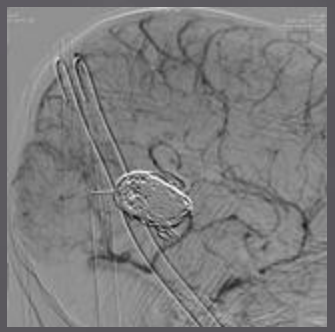
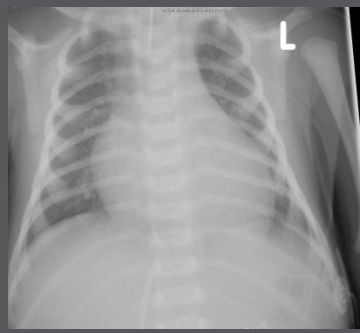










YASARGYL TYPE I.

EMBOLIZATION X 2 I/A COILS + ONYX.

.COMPLETE AT 3 MONTHS .

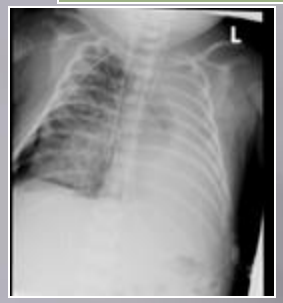



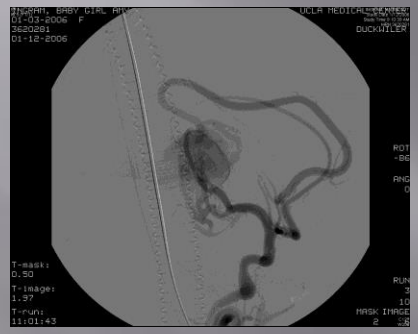














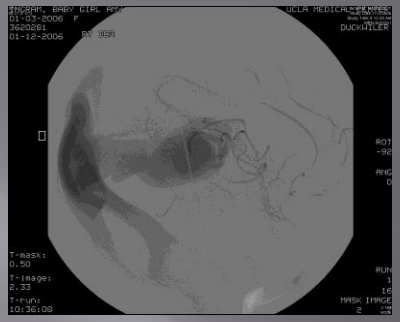
YASARGYL TYPE III

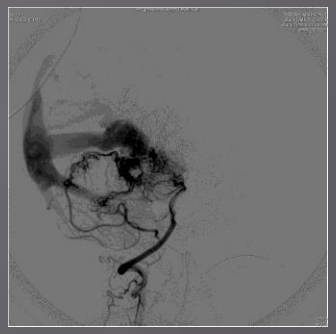
SEVERE BI-VENTRICULAR FAILURE 2 DAYS OLD

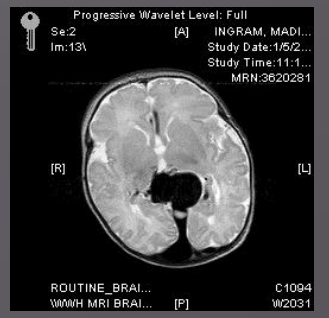
TRANSARTERIAL + TRANSVENOUS

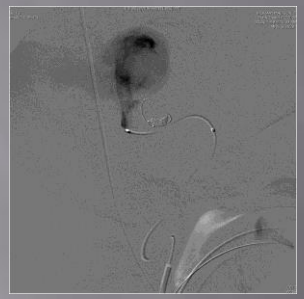


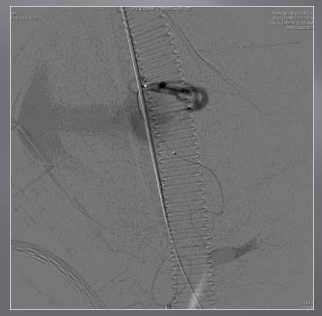


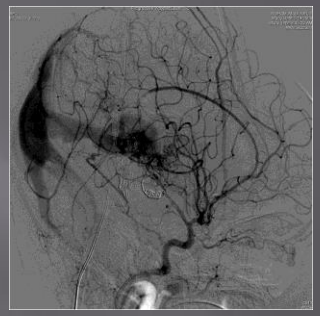


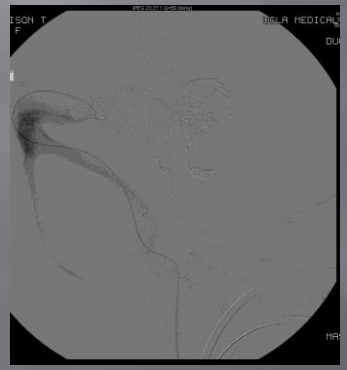


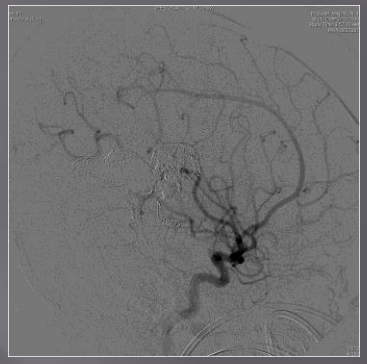


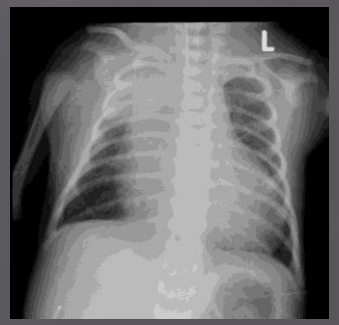






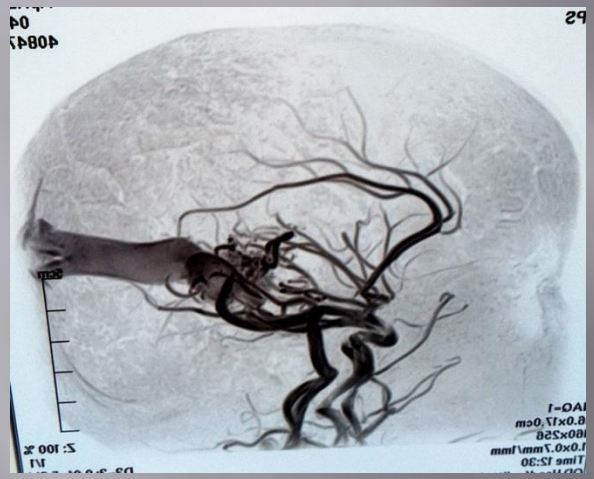


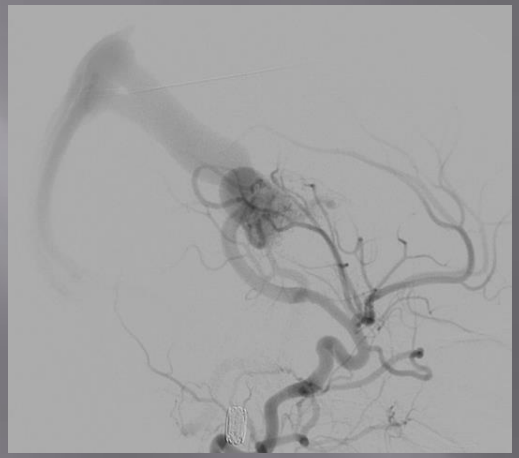


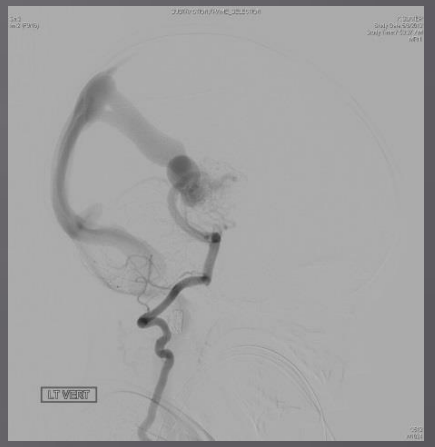


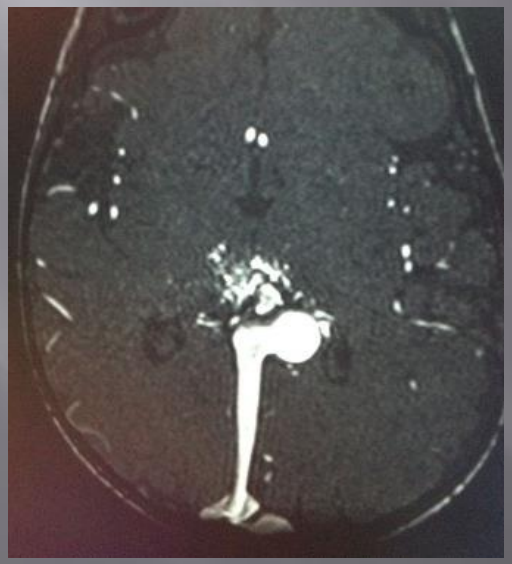
YASARGYL TYPE III

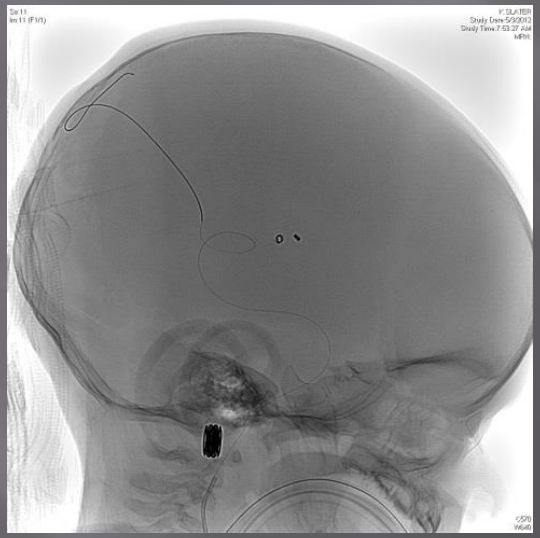
3M. MILD CHF . TA

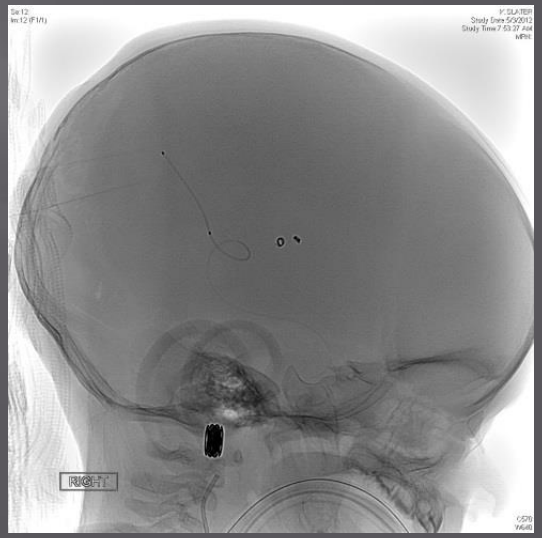




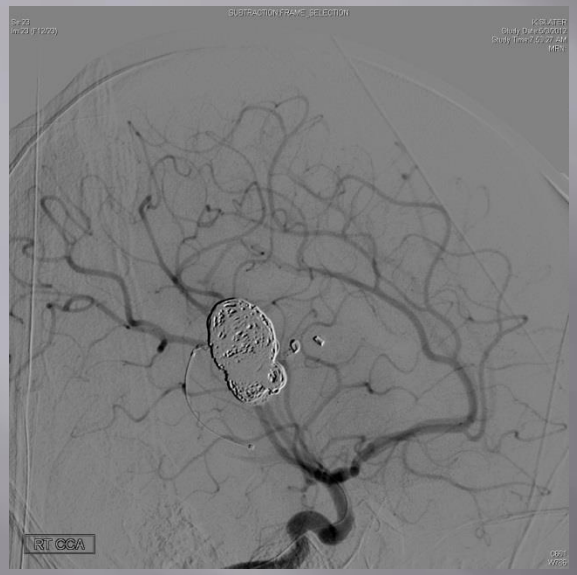


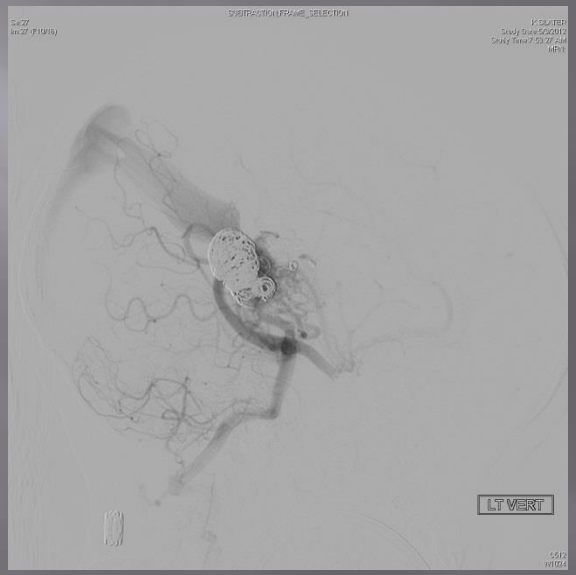


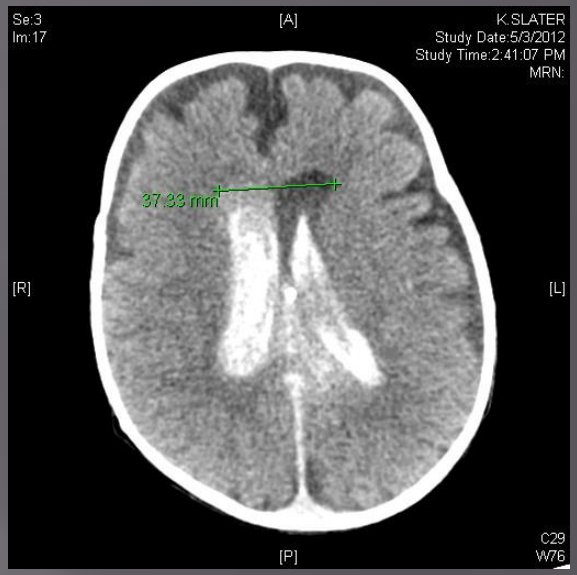


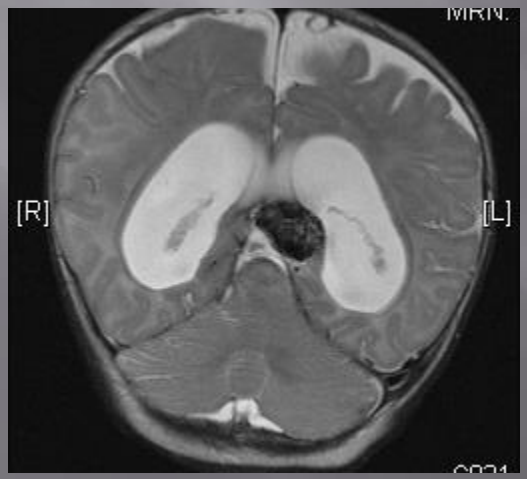


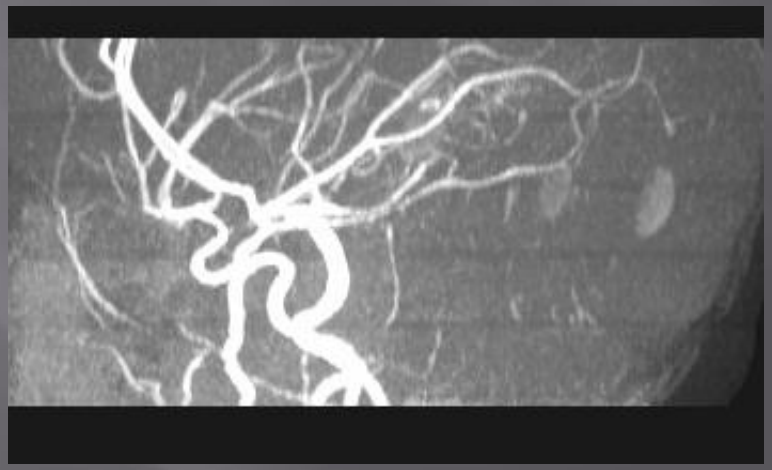
IV AND THALAMIC BLEED
6 MONTHS COMPLETE RECOVERY







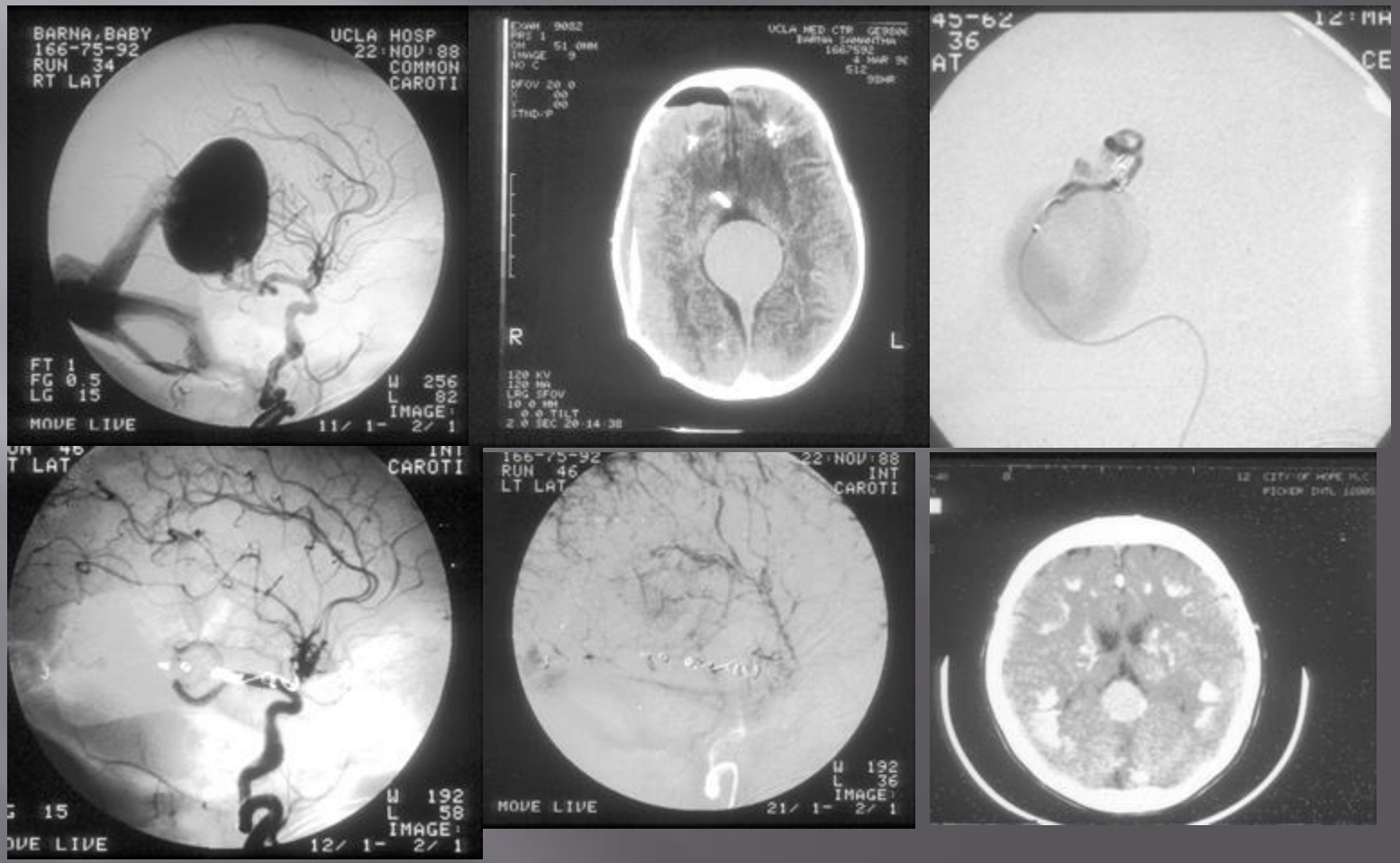




YASARGIL TYPE I

VENOUS HYPERTENSION /CALCIFICATIONS

STATUS EPILEPTICUS



CLINICAL COMPLICATONS

GALENIC		**112 pts.**
Neonates/Infants (72 pts)	13/72	**18.5%**
DEATH	**7/72**	**9,72%**
Children/adults (40 pts)	3/40	**7.5%**
DEATH	**0/40**	**0%**
NON-GALENIC (53 pts)	**4/53**	**7%**
DEATH	**0/53**	**0%**

OUTCOME AND COMPLICATIONS OF ENDOVASCULAR EMBOLIZATION FOR VEIN OF GALEN MALFORMATIONS: A SYSTEMATIC REVIEW AND META-ANALYSIS

Jun Yan, MD , PhD,1 Jing Wen, MD , PhD,2 Roodrajeetsing Gopaul, MD ,1 Chao-Yuan Zhang, MD ,1 and

Shao-wen Xiao, MD 1

Departments of 1Neurosurgery and 2Rheumatism, The First Affiliated Hospital of Guangxi Medical University, Nanning,

Guangxi, China

J Neurosurg Volume 123 • October 2015

Literature research using PuB Med datasabe.

Thirty four studied with **667 pts** treated using endovascular embolization.

Evaluation of efficacy and safety of treatment.

- Neonates 44%
- Infants 41%
- Children/adults 12%

- Complete occlusion 57%
- Partial occlusion 43%

- Good outcome 68%
- Poor outcome 31%

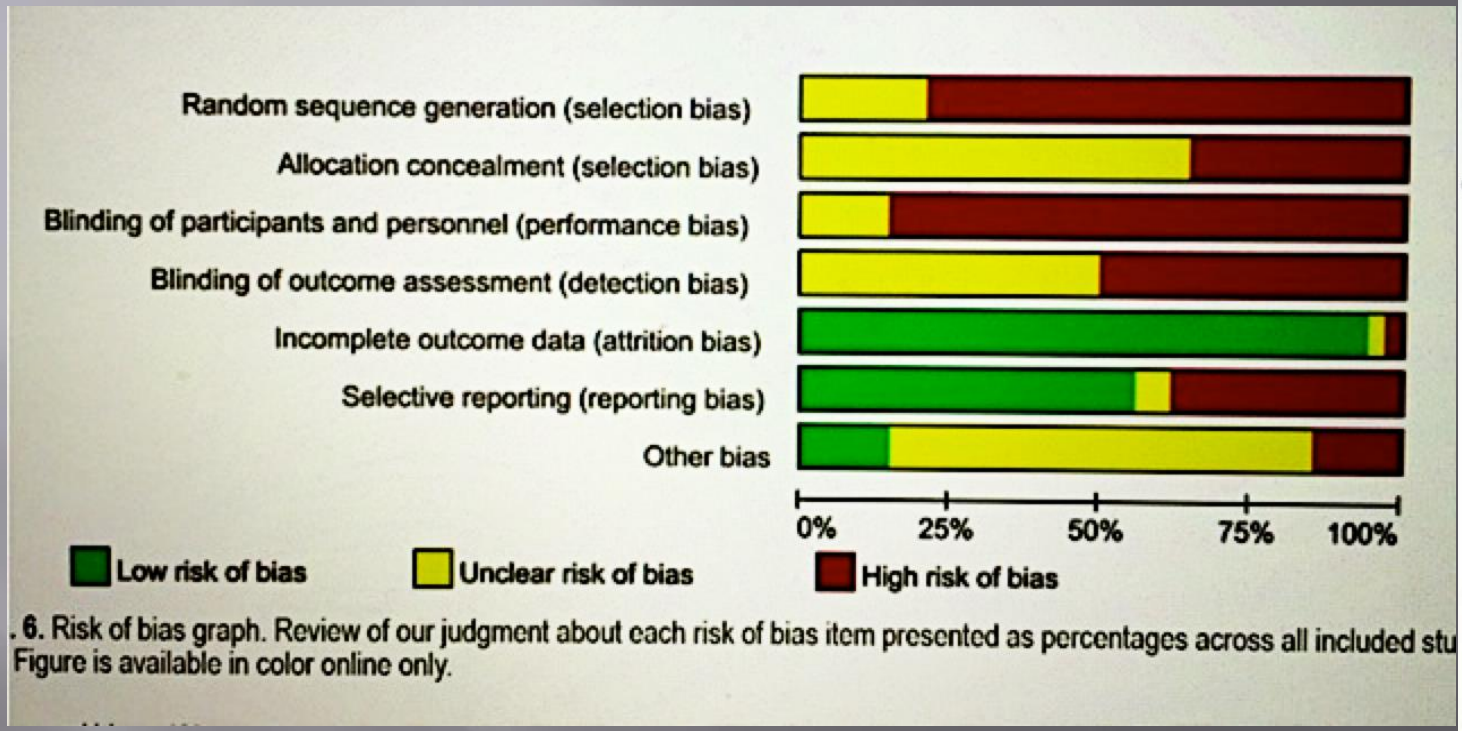
- Neonates <1 month
- Infants >1 month<2 yrs.
- Children >2 yrs.

- Complications 10%
- Death 10%

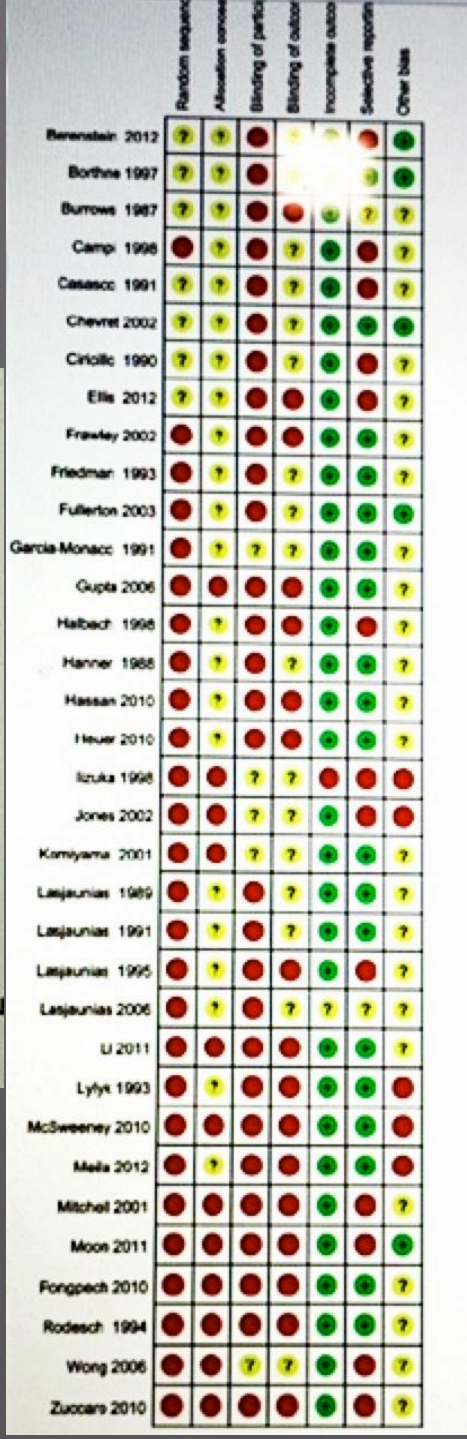
Complications: cerebral hemorrhage, cerebral ischemia, hydrocephalus, leg ischemia, and vessel perforation.

- Patients with VGM total occlusion
- treated in **1 stage** had highest
- complications (cerebral hemorrhage [32%] and venous thrombosis [27%]). Patients with VGM occlusion post **2 or 3 stages** had fewer complications (cerebral hemorrhage [21%] and venous
- thrombosis [24%]). Patients treated **more than 3 times** had the complications (cerebral hemorrhage [18%] and venous thrombosis [24%]).

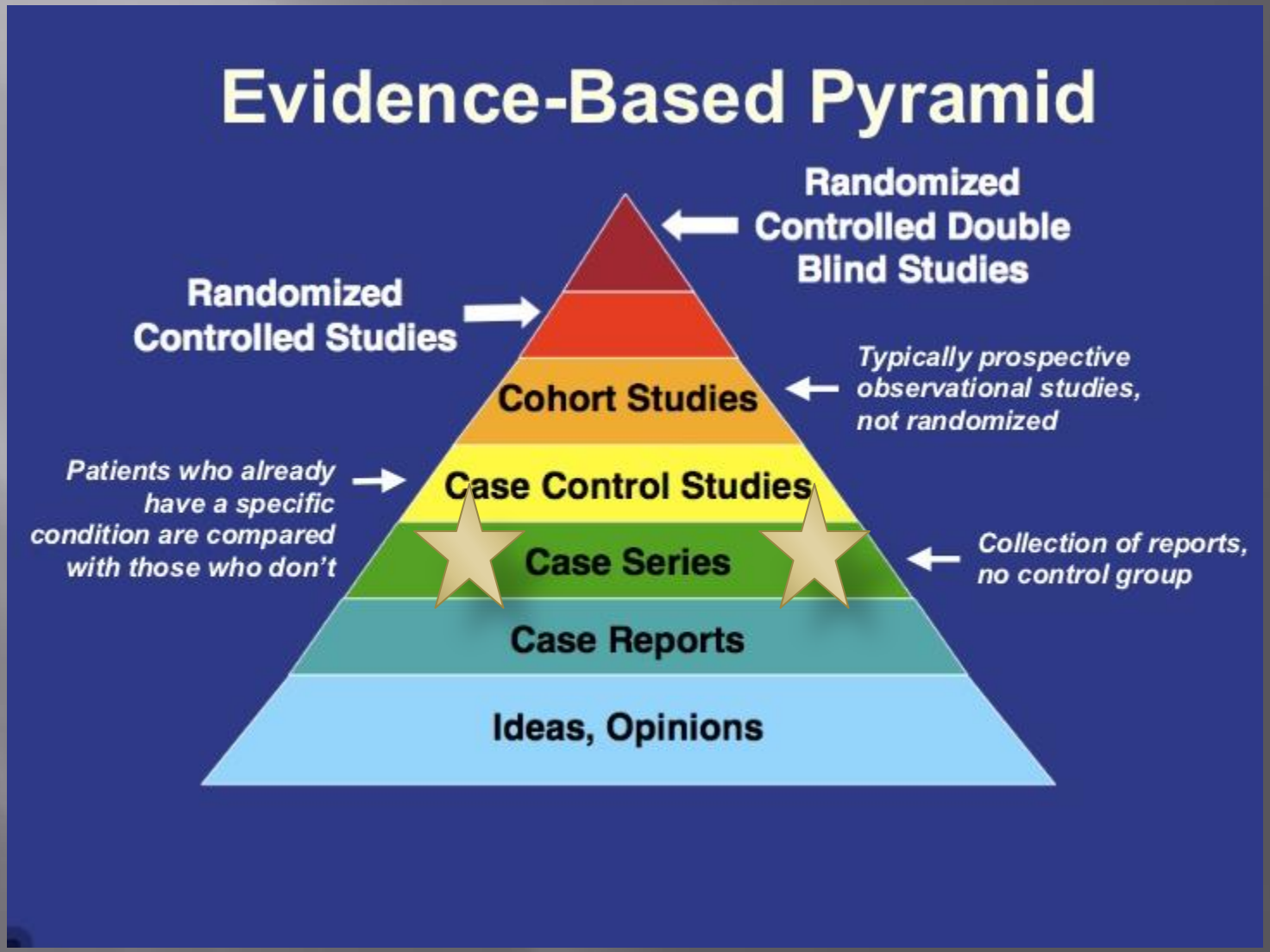
Cochrane Collaboration's risk of bias tool.



. 6. Risk of bias graph. Review of our judgment about each risk of bias item presented as percentages across all included stu
Figure is available in color online only.



EVIDENCE BASED MEDICINE.



CENTRE FOR EVIDENCE-BASED MEDICINE.

Level	Type of evidence
1A	Systematic review (with homogeneity) of RCTs
1B	Individual RCT (with narrow confidence intervals)
1C	All or none study
2A	Systematic review (with homogeneity) of cohort studies
2B	Individual Cohort study (including low quality RCT, e.g. <80% follow-up)
2C	"Outcomes" research; Ecological studies
3A	Systematic review (with homogeneity) of case-control studies
3B	Individual Case-control study
4	Case series (and poor quality cohort and case-control study
5	Expert opinion without explicit critical appraisal or based on physiology bench research or "first principles"

CONCLUSIONS

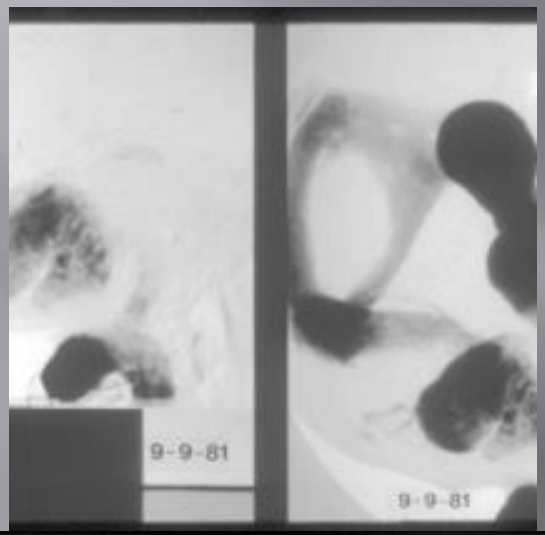
- Tailor treatment to clinical syndrome.
- Aggressive treatment in CHF, hydrovenous disorder and hemorrhage.
- Consider anticoagulation in cases of postembolization severe venous stagnation.
- Staging may be a valid alternative in multipedicular high flow a/v fistulae.

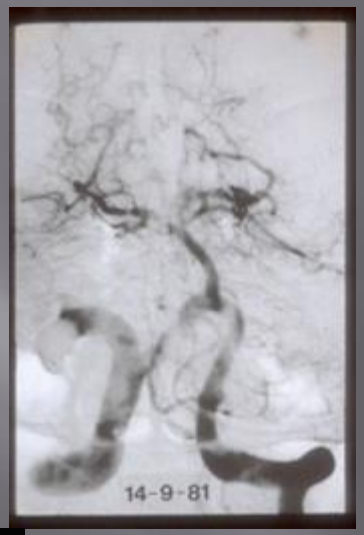
PIAL HIGH FLOW ARTERIOVENOUS FISTULAE

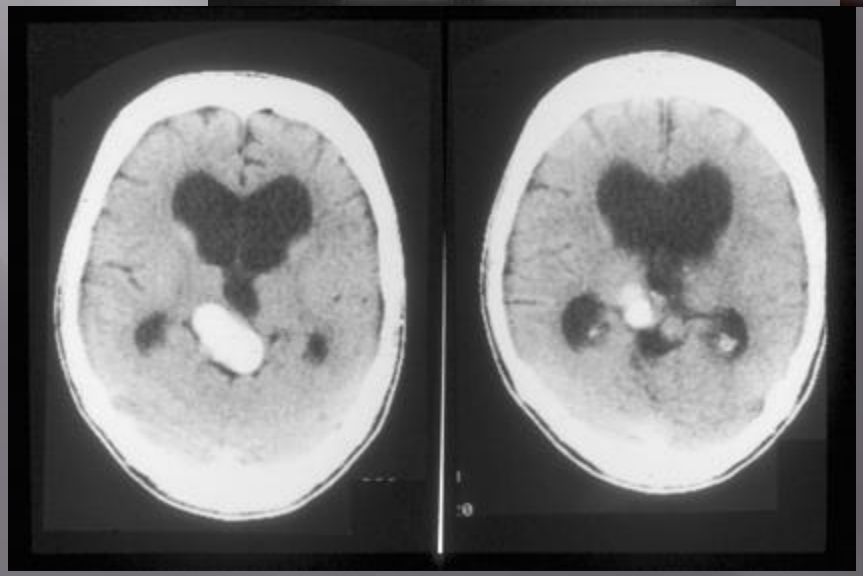
NON-GALENIC ARTERIOVENOUS

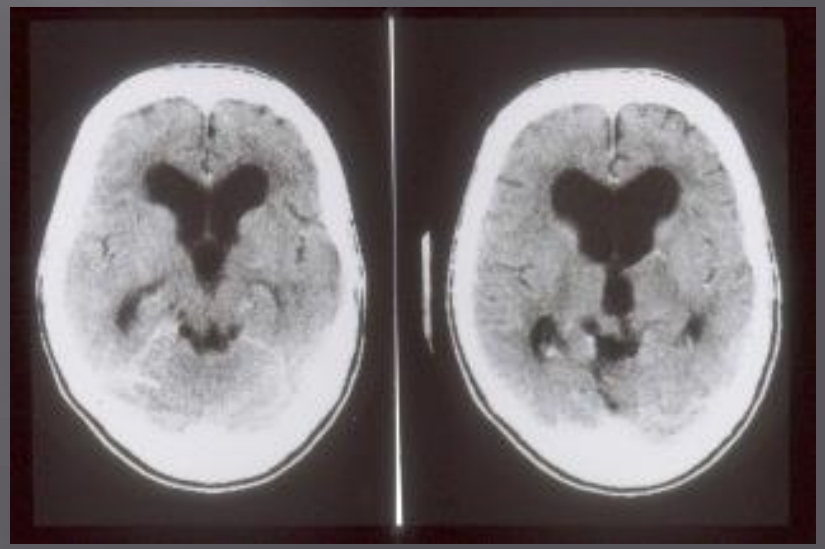
FISTULAE.

POSTERIOR FOSSA A/F FISTULA BALLOON OCCLUSION 1981

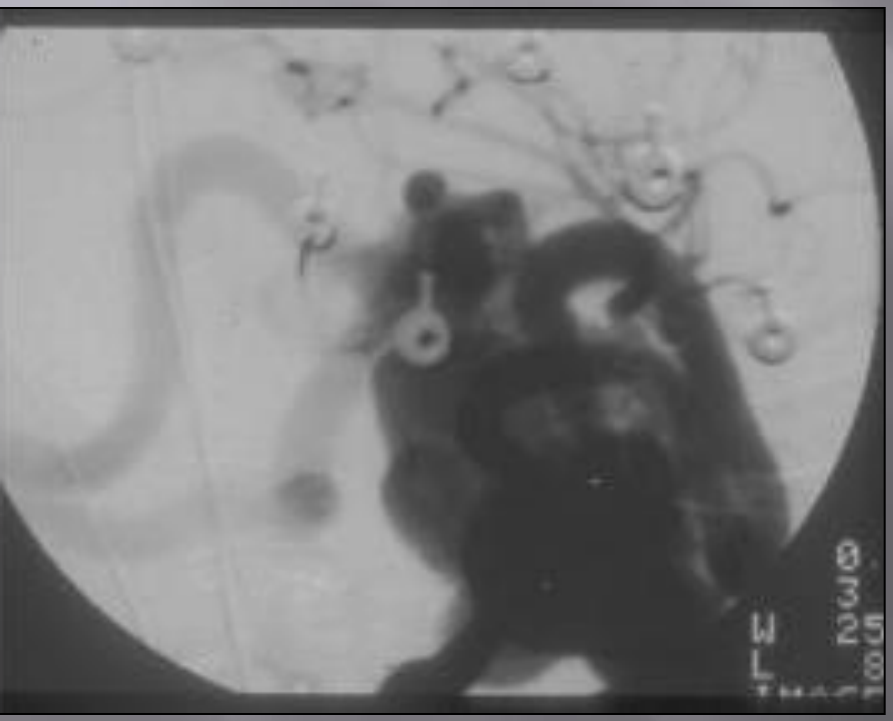


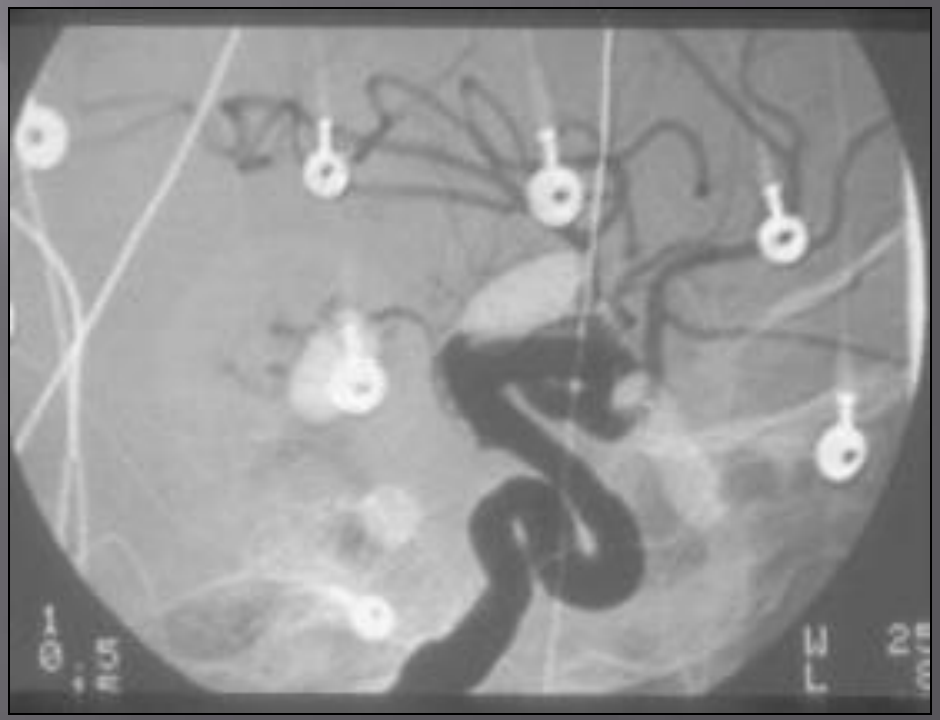




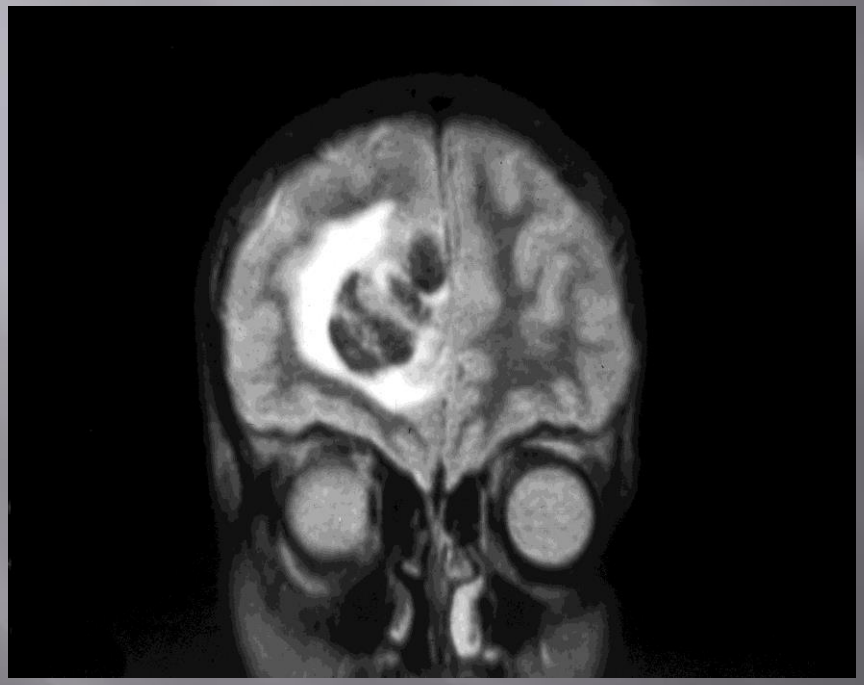


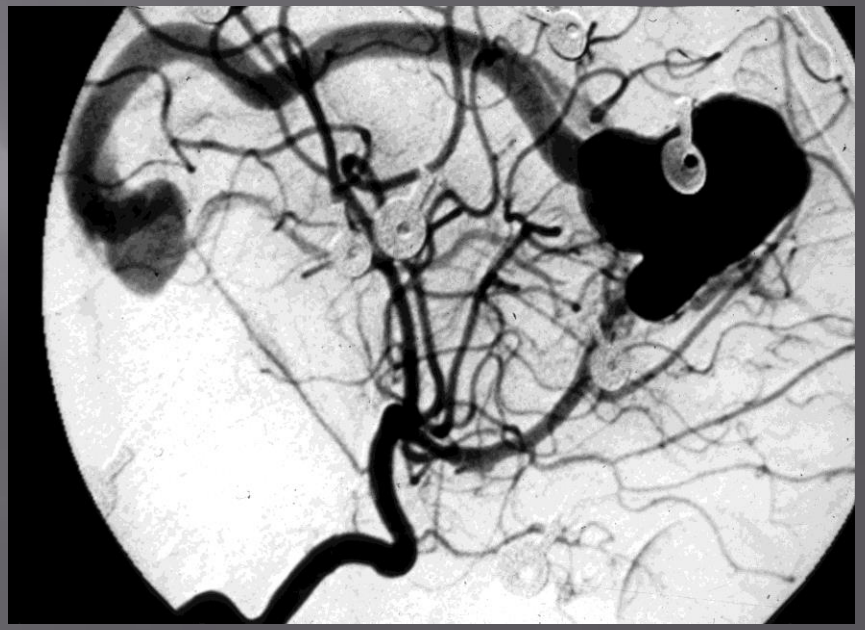
ANTERIOR TEMPORAL ARTERIOVENOUS FISTULA 1982



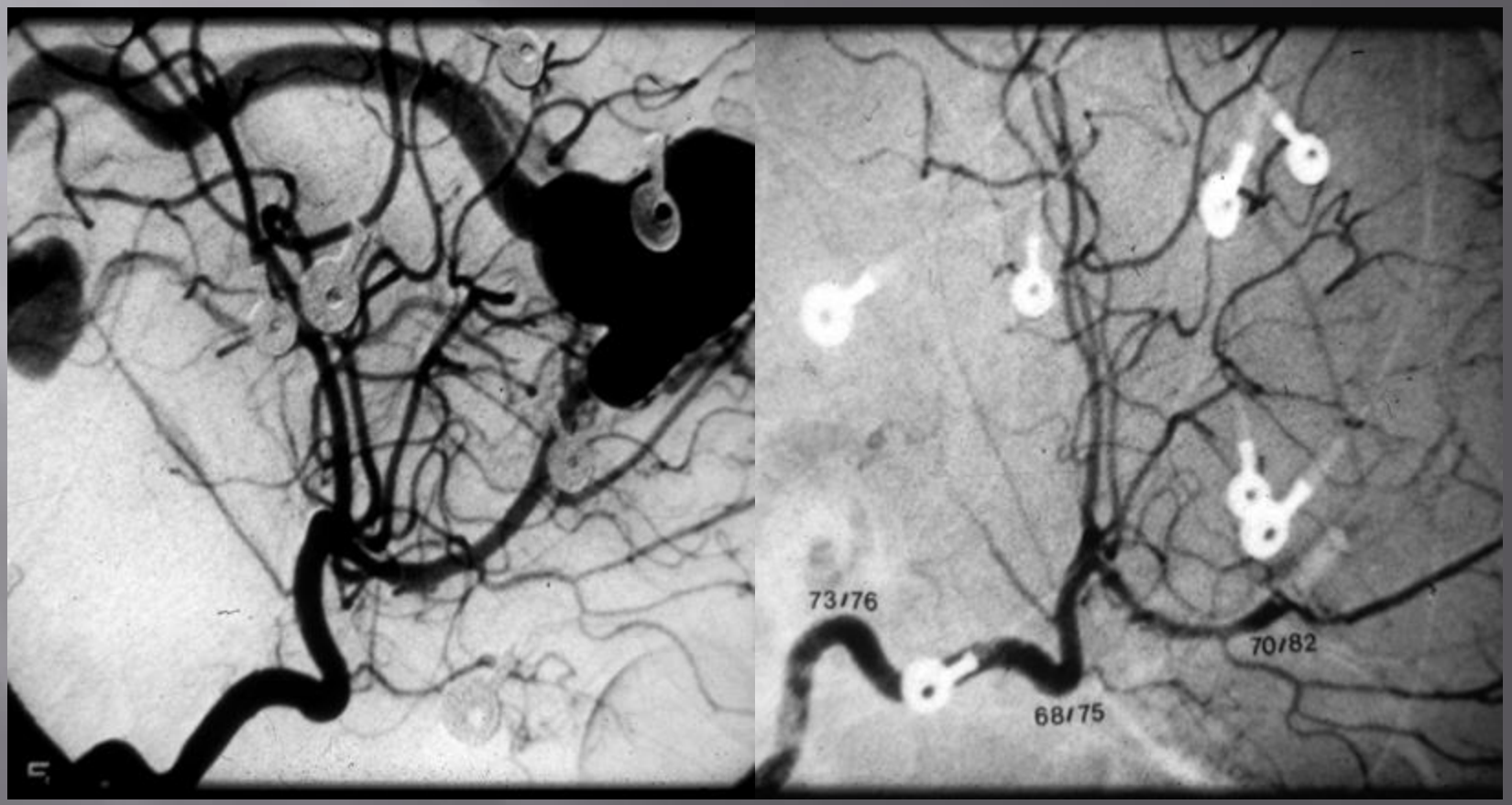


RUPTURED ANTERIOR FRONTAL CONGENITAL A/V FISTULA 1985

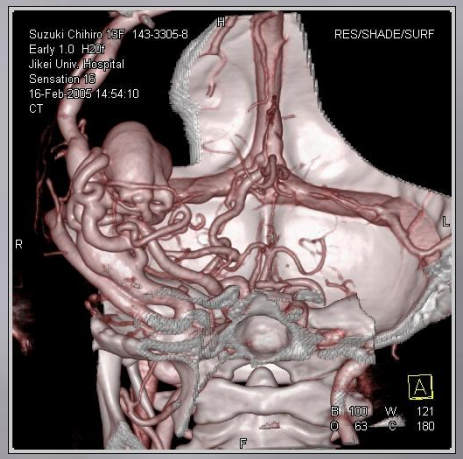


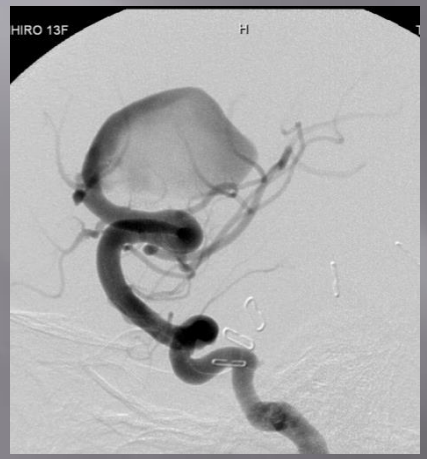


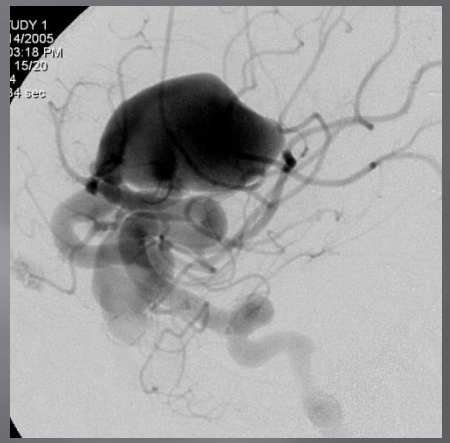
RUPTURED ANTERIOR FRONTAL CONGENITAL A/V FISTULA

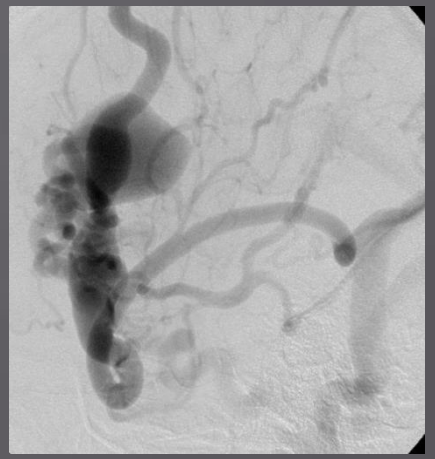


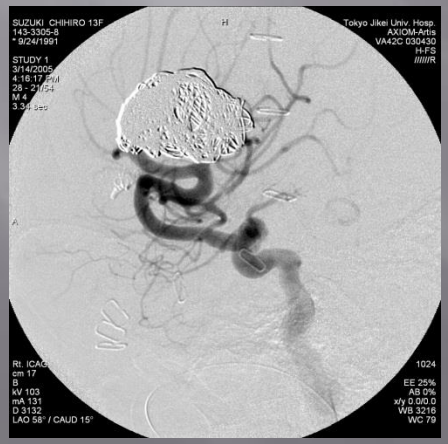
RIGHT FRONTAL PIAL ARTERIOVENOUS FISTULA. TA COIL EMBOLIZATION

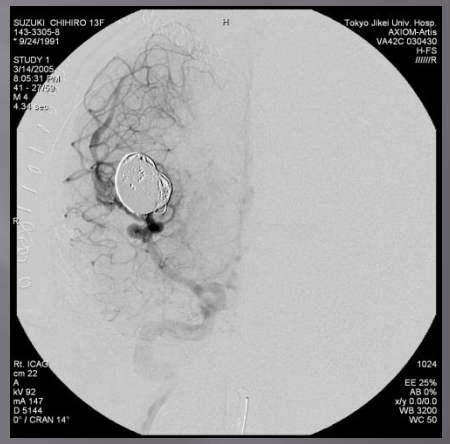


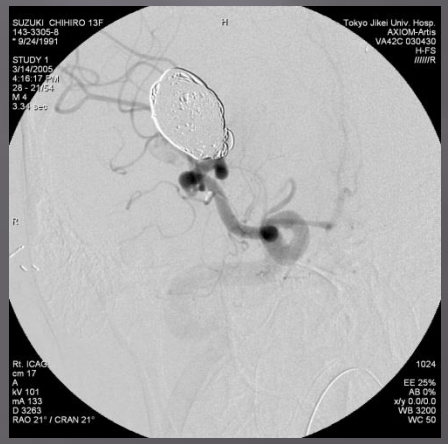




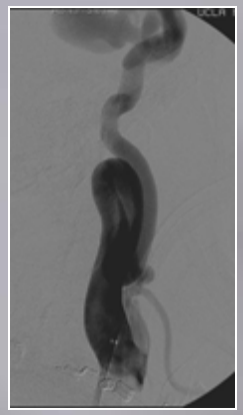
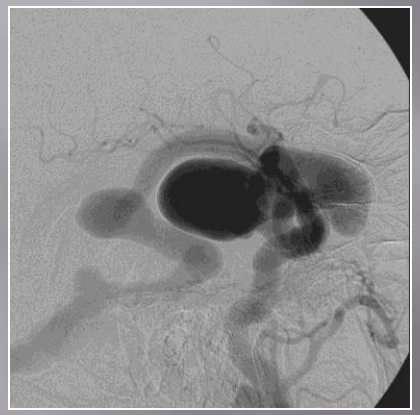
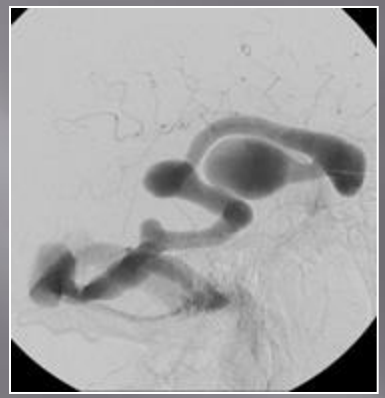
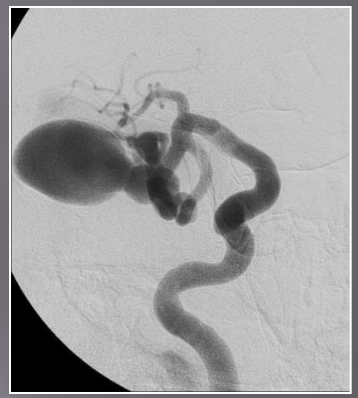
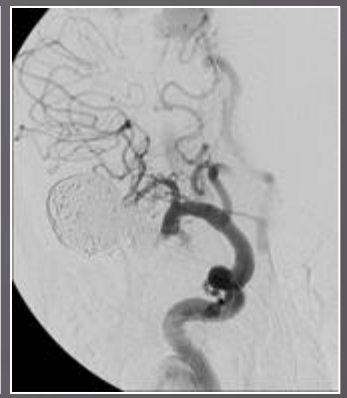
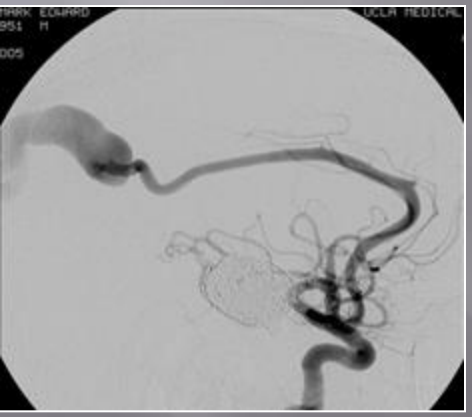
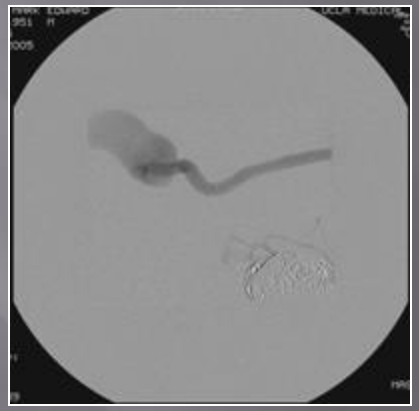
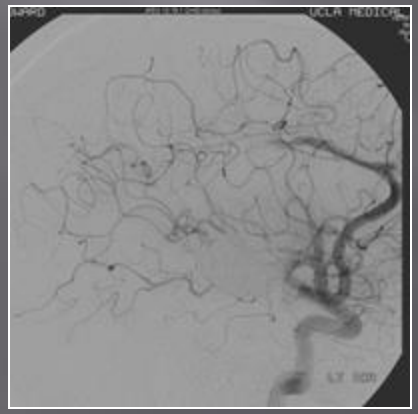
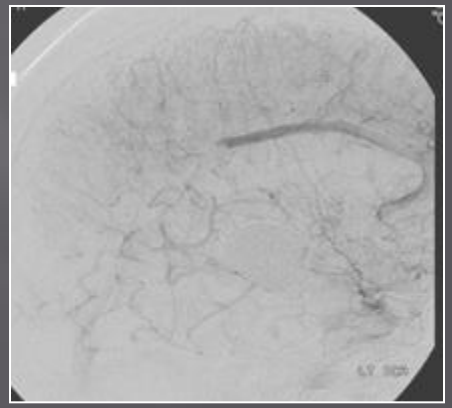




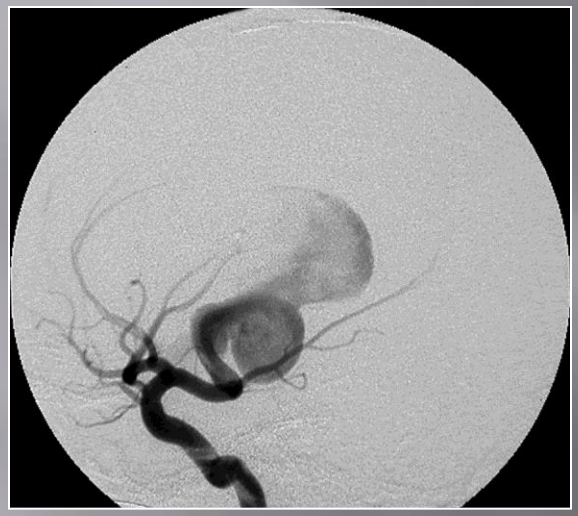


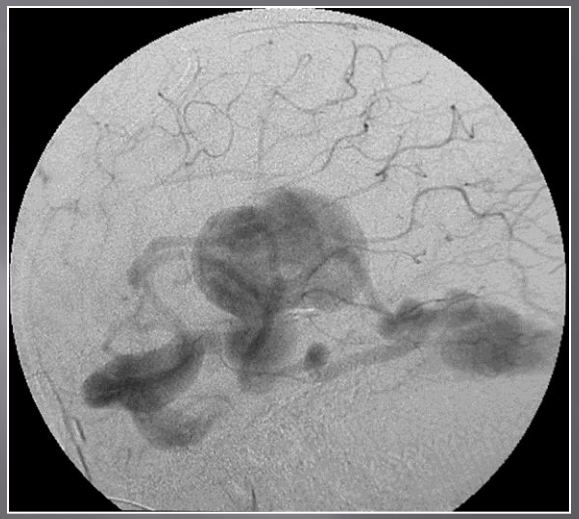


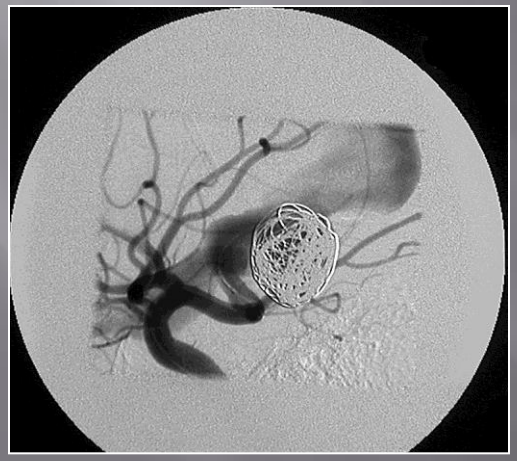
HIGH FLOW TEMPORAL AND OCCIPITAL AV FISTULAE. TRANSARTERIAL EMBOLIZATION.

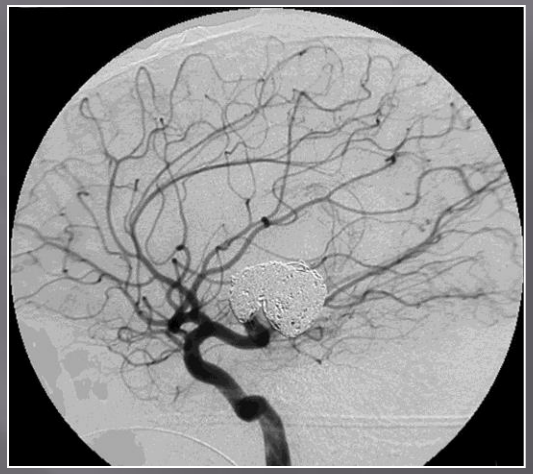










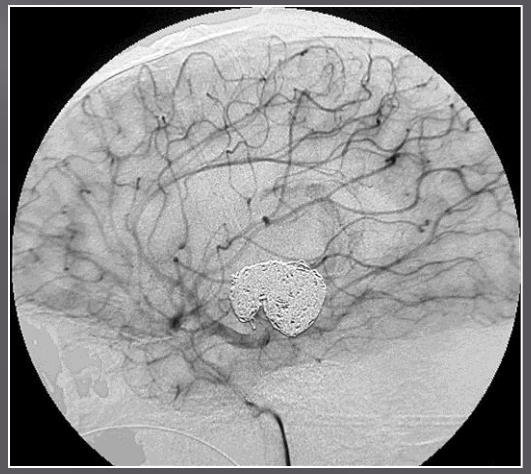
GIANT TEMPORAL A/V FISTULA SEIZURES COILS + ONYX



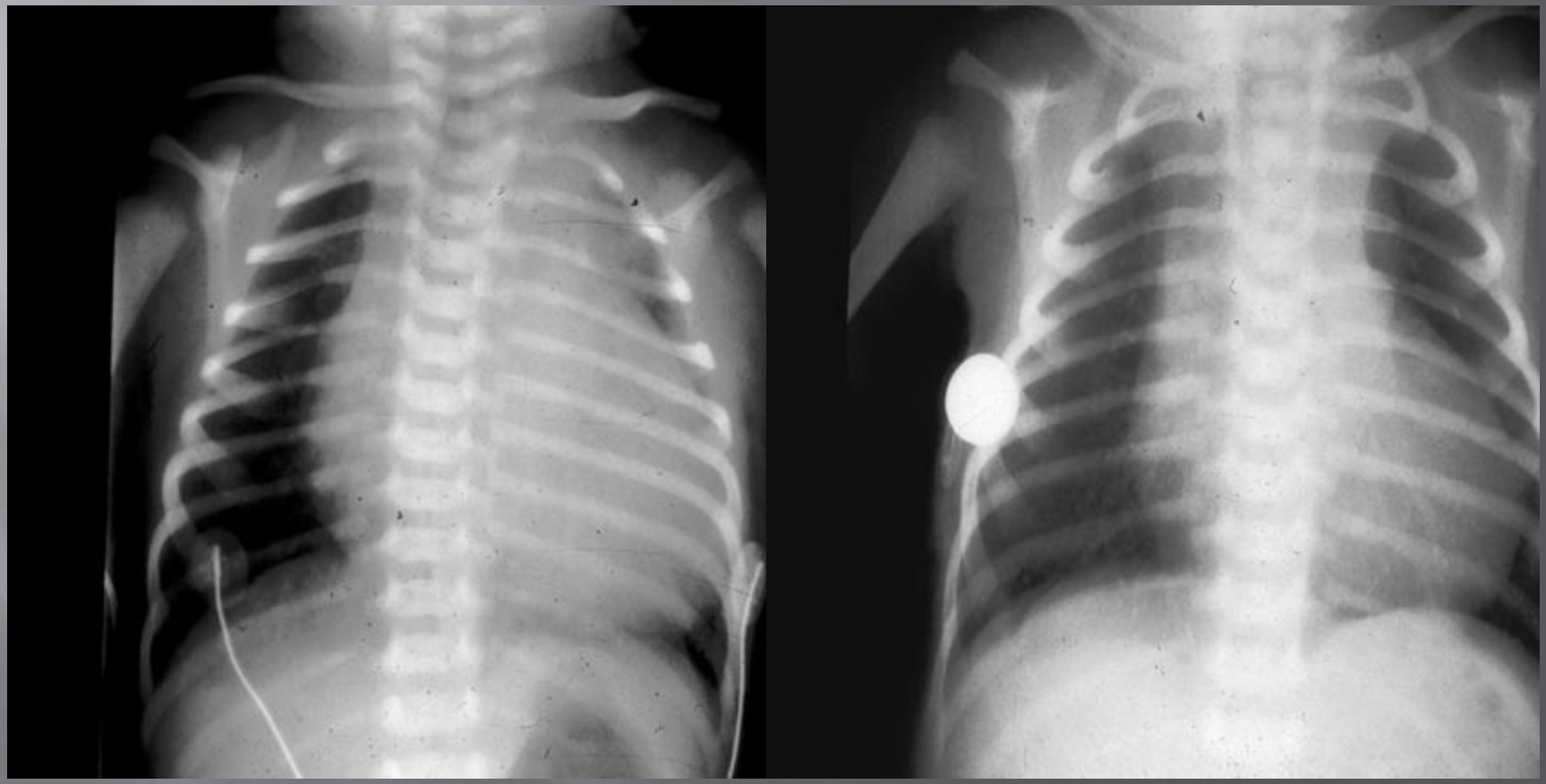








BASILAR DIENCEPHALIC A/V FISTULA 3 YRS CHF.



DELAYED VENOUS THROMBOSIS:
HEMORRHAGIC VENOUS INFARCTION