Vascular malformation (VM) ABC

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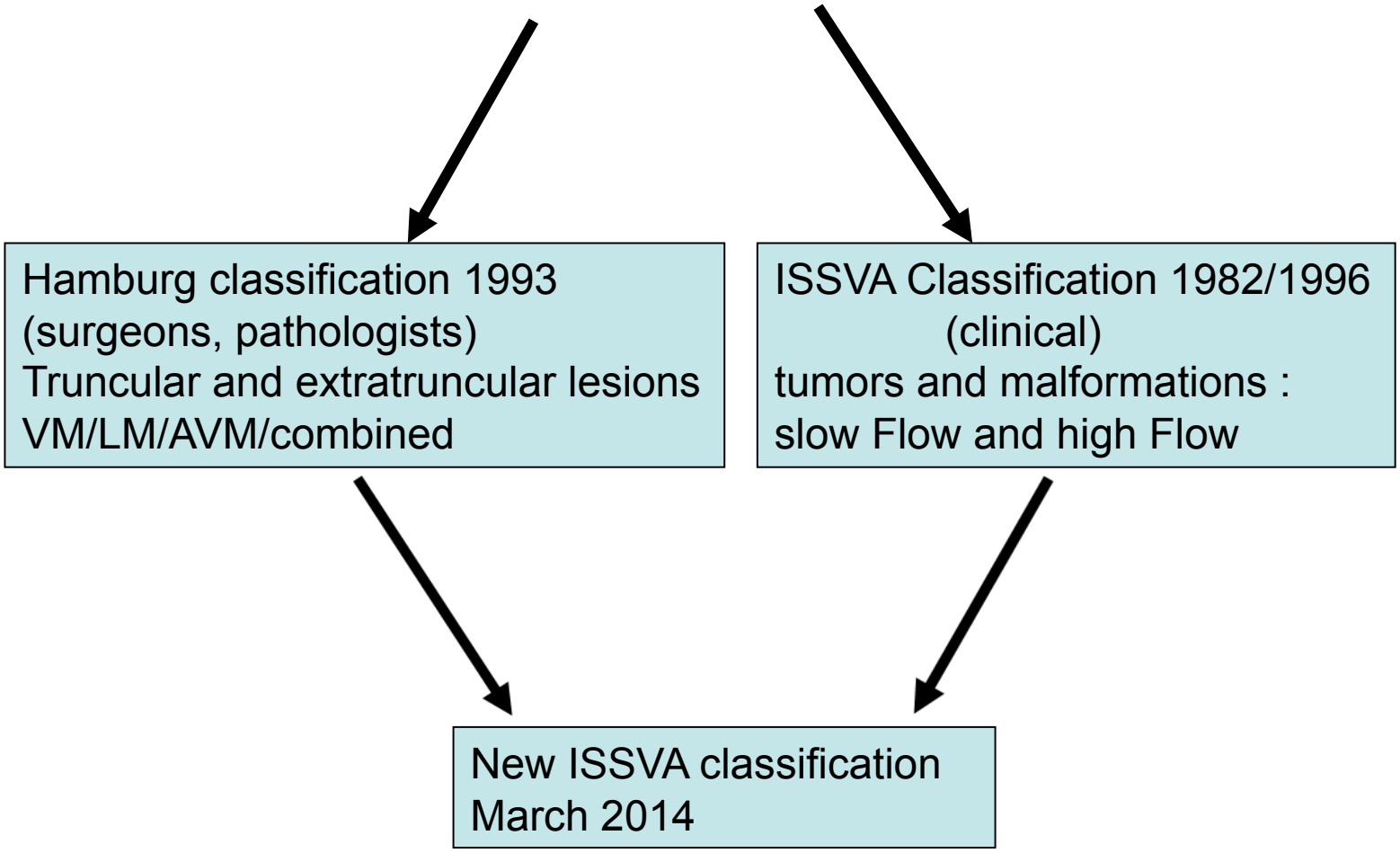
Disclosure

- Research grant
  - Siemens Medical
  - Object Research System
  - Bracco Diagnostic
  - Biotronik
  - TVA medical
- Consultant
  - Cook Medical

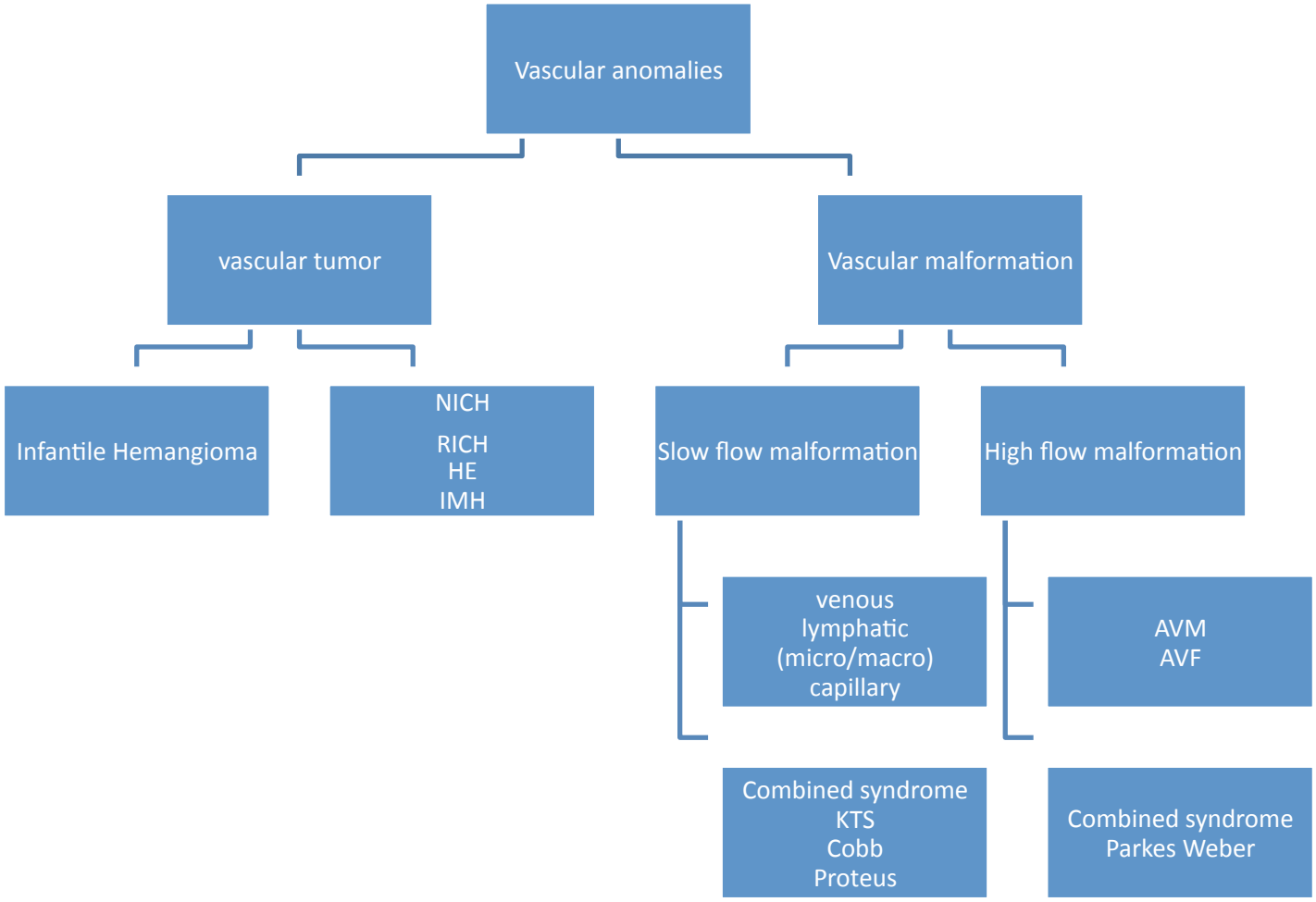
Basic principles

- Use an appropriate terminology
  - Mulliken classification + new ISSVA classification
- Always correlate imaging findings with clinical history and examination
  - Vascular anomalies clinic
- Multidisciplinary management
  - IR (diagnosis, intervention)
  - Plastic surgery, dermatology, ENT, internal medicine

CLASSIFICATIONS



Mulliken classification



Mulliken J. and Glowacki J.:Hemangiomas and vascular malformations in infants and children :a classification based on endothelial characteristics. *Plast Reconstr Surg 1982; 69 : 412-422*

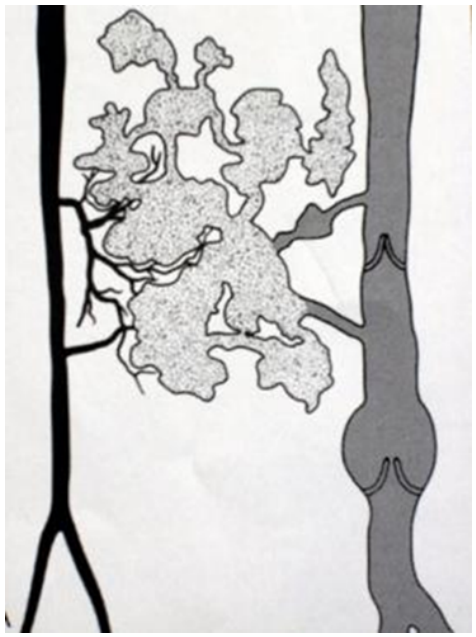
Hamburg Classification
of vascular malformations

Predominant Type	Lesion Form	
	Truncular	**Extratruncular**
Arterial	Aplasia or Obstruction Dilatation	Infiltrative Limited
Venous	Aplasia or Obstruction Dilatation	Infiltrative Limited
Lymphatic	Aplasia or Obstruction Dilatation	Infiltrative Limited
Arteriovenous shunt	Deep Superficial	Infiltrative Limited
Combined / Mixed	Arterial and venous without shunt Haemolymphatic with or without shunt	Haemolymphatic infiltrative or limited

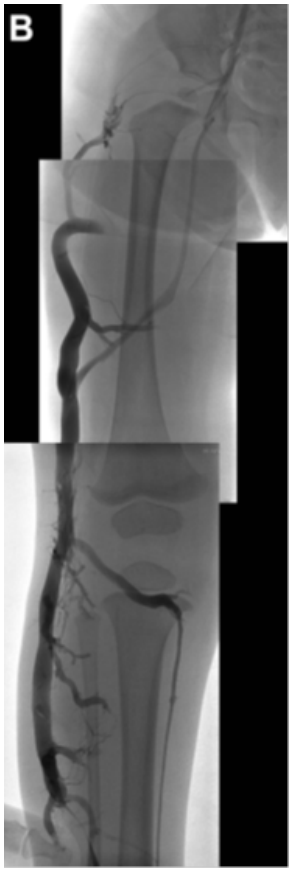
Belov S. Semin Vasc Surg 1993; 6:219

Extratruncal vs truncal

- Extratruncal
  - Defect occuring early in the embryogenesis
  - Mesenchymal cell memory (proliferation)
  - No or few connections with normal vascular system
- Truncal
  - Defect occuring later
  - Hypoplasia or dilatation of the vascular system



Extra truncal



Truncal

Lee-BB-JVD-2013

2014 ISSVA classification

Vascular anomalies				
Vascular tumors	**Vascular malformations**			
	Simple	**Combined °**	**of major named vessels**	**associated with other anomalies**
Benign Locally aggressive or borderline Malignant	Capillary malformations Lymphatic malformations Venous malformations Arteriovenous malformations* Arteriovenous fistula*	CVM, CLM LVM, CLVM CAVM* CLAVM* others	Details	Details

Anomalies of major named vessels

(aka "channel type" or "truncal" vascular malformations)

Affect

- lymphatics
- veins
- arteries

Anomalies of

- origin
- course
- number
- length
- diameter (aplasia, hypoplasia, stenosis, ectasia / aneurysm)
- valves
- communication (AVF)
- persistence (of embryonal vessel)

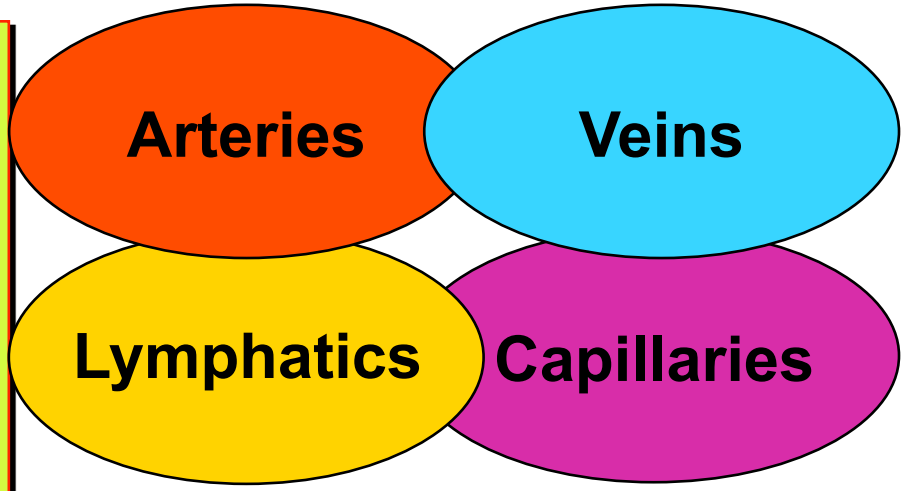
issva.org/classification

Vascular malformations associated with other anomalies		
Klippel-Trenaunay syndrome:	CM + VM +/- LM + limb overgrowth	
Parkes Weber syndrome:	CM + AVF + limb overgrowth	G
Servelle-Martorell syndrome:	limb VM + bone undergrowth	
Sturge-Weber syndrome:	facial + leptomeningeal CM + eye anomalies +/- bone and/or soft tissue overgrowth	G
Limb CM + congenital non-progressive limb hypertrophy		
Maffucci syndrome:	VM +/- spindle-cell hemangioma + enchondroma	
Macrocephaly - CM (M-CM / MCAP)		G
Microcephaly - CM (MICCAP)		G
CLOVES syndrome:	LM + VM + CM +/- AVM + lipomatous overgrowth	G
Proteus syndrome:	CM, VM and/or LM + asymmetrical somatic overgrowth	G
Bannayan-Riley-Ruvalcaba sd:	AVM + VM +macrocephaly, lipomatous overgrowth	G

Vascular malformations

no cellular proliferation

no regression

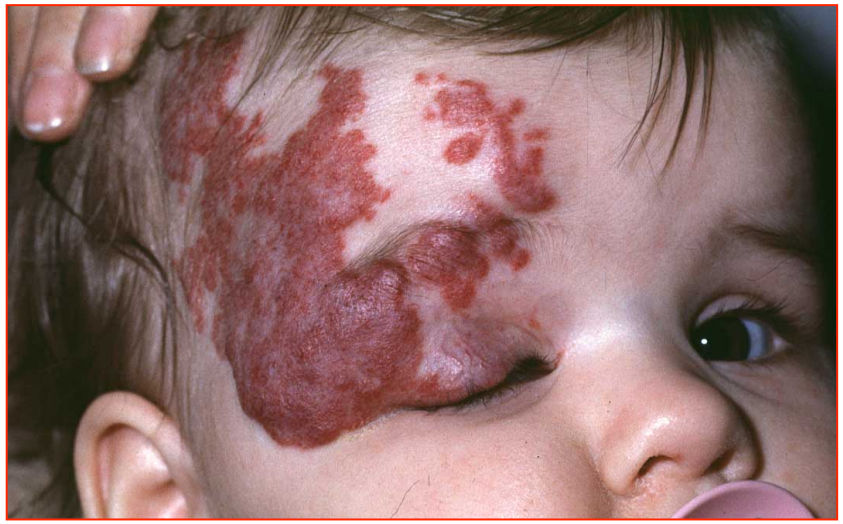


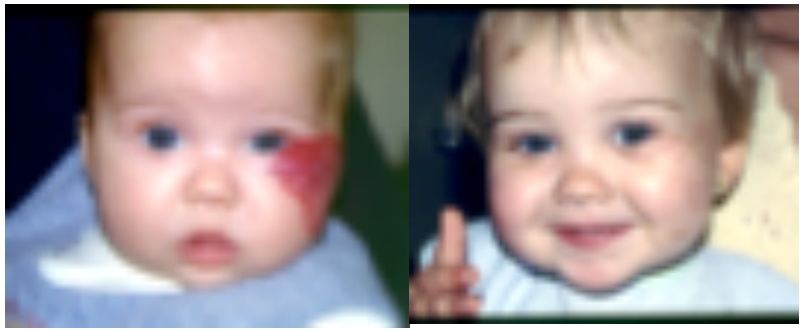
Investigation

- Clinical examination combined with DUS
  - Coloration
  - Softness
  - Thrill, expansion with valsalva
  - Vessel dilatation, venous reflux
  - Limb overgrowth
  - CM (angioma)
  - Tissular destruction

Infantile hemangioma

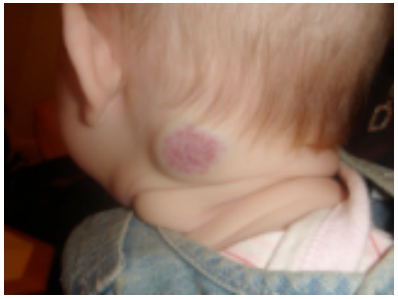
- Infantile hemangioma
  - Growth 0-1 year
  - Stabilization 1-2 year
  - Regression 2-5 year
  - Glut 1 +
  - Conservative management
  - Propanolol, interferon, vincristin for complicated cases

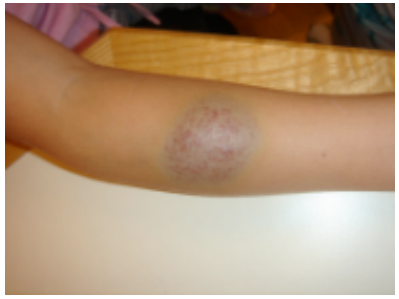




Congenital hemangioma

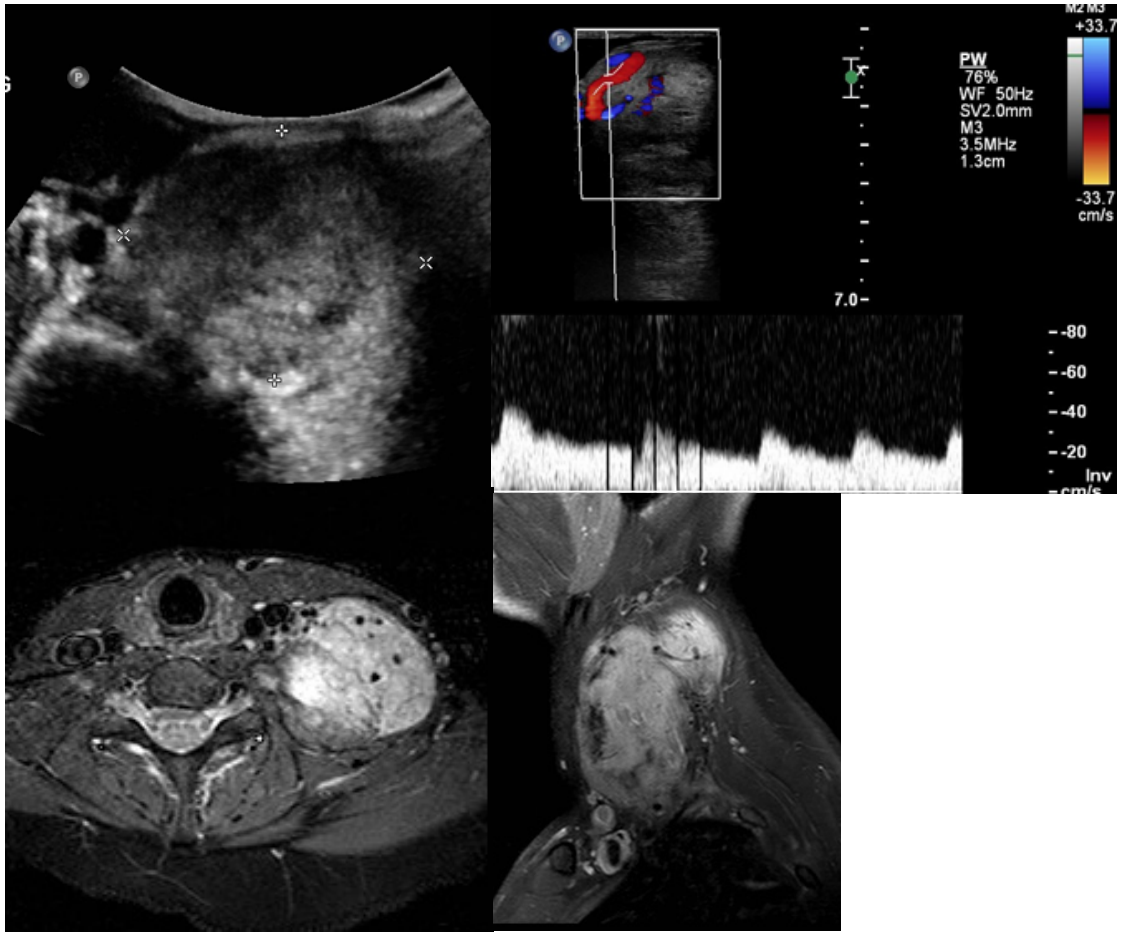
- RICH (rapidly involuted congenital hemangioma)
  - Completely grown at birth
  - Regression 12-14 months
  - Glut –
- NICH
  (non-involuted congenital hemangioma)
  - Completely grown at birth
  - No involution
  - Growth during teenage
  - Glut –
- Hemangioendothelioma
- Tufted angioma
- Intramuscular hemangioma ?





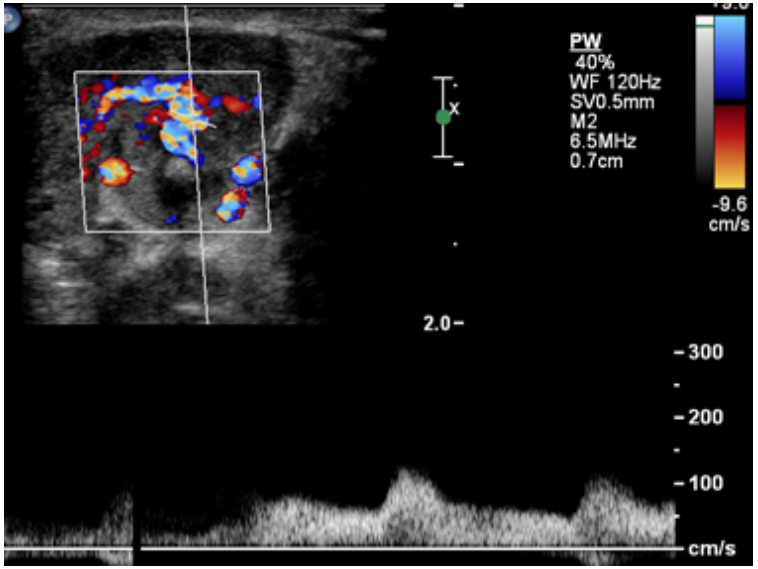
Intramuscular hemangioma = NICH ?

- Intra muscular vascular tumor
- Hypervascular
- No AV shunting
- Surgery

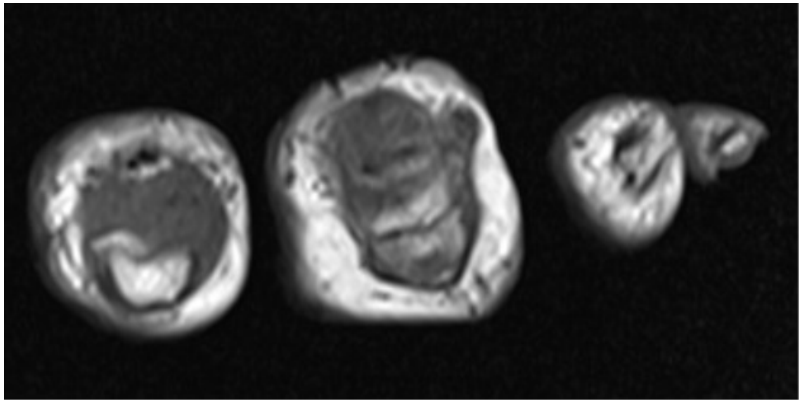


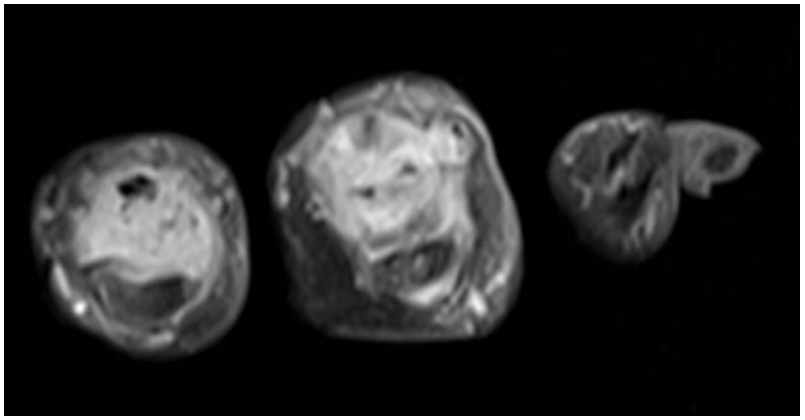
Hemangioendothelioma

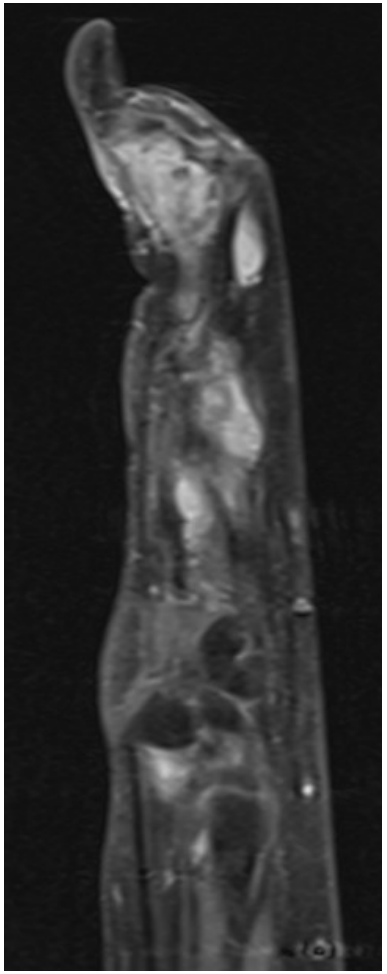
- F 46 ans
- Firm nodules since birth 1st 2nd, 3rd finger and wrist slowly growing



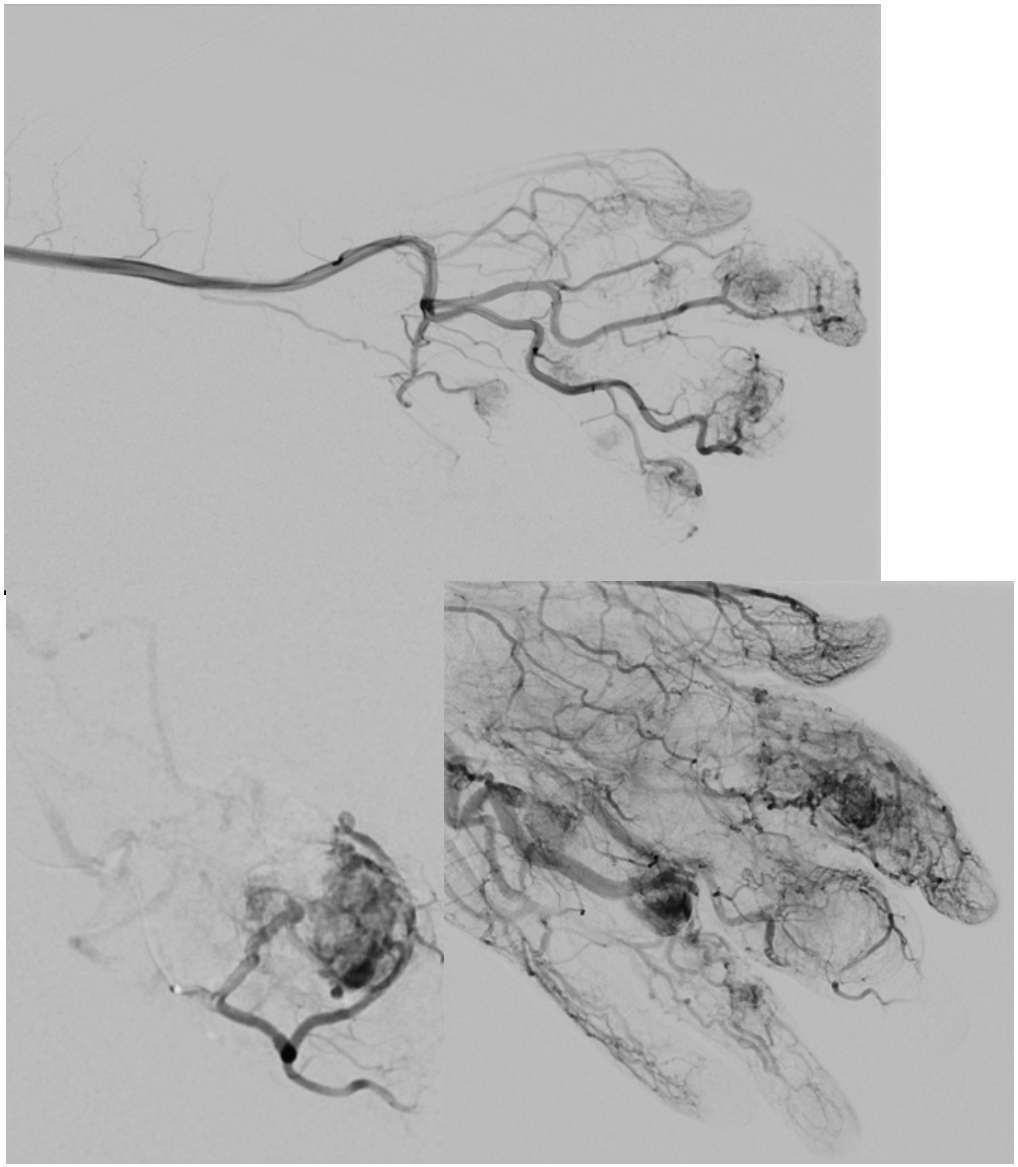
Hemangioendothelioma







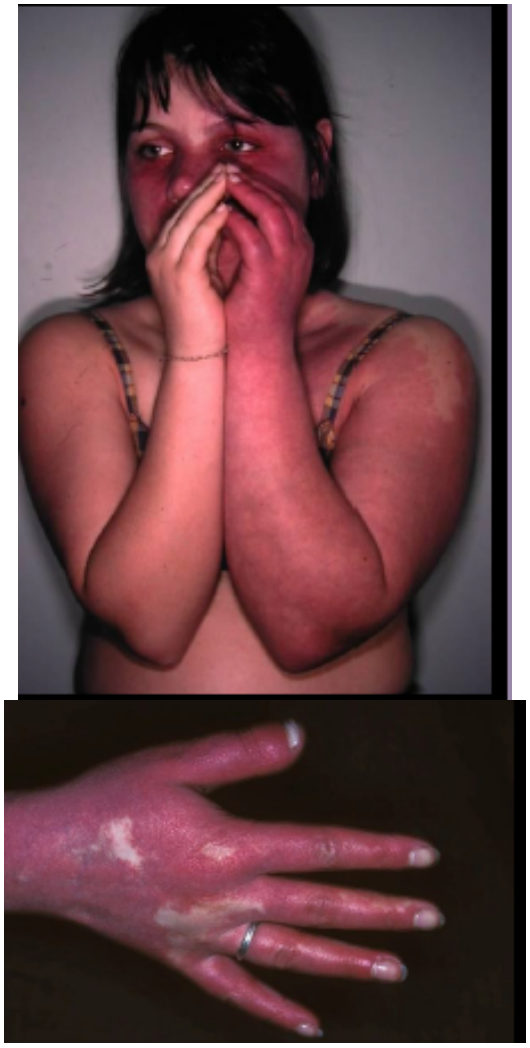
hemangioendothelioma



- Embolization
  - Particle
  - Onyx
- Surgery
- Vascular tumors in an adult patient & growth = biopsy

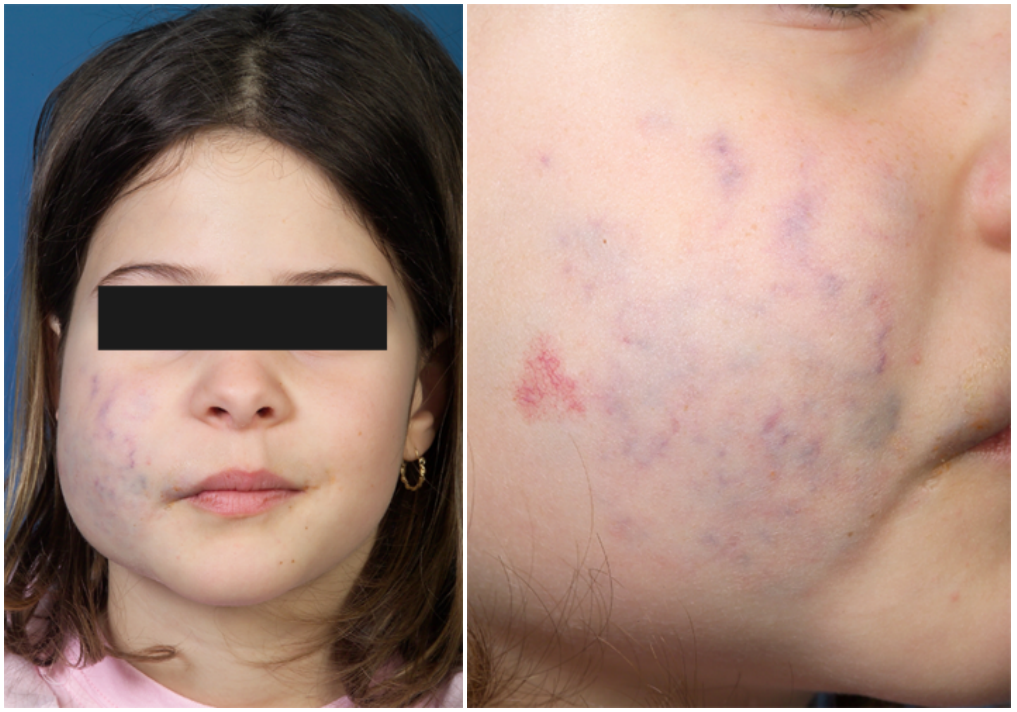
Capillary malformation

- Port wine stain
- No pulsation
- No mass
- Rule out AVM
- Associated syndrome
  - Struge Weber
  - KT
  - PKWS



Venous malformation

- Low flow
- Most frequent
  - Head and neck 40%
  - Body 20%
  - Limbs 40%
- Expansion
  - Valsalva
  - Dependent position
- Bluish coloration



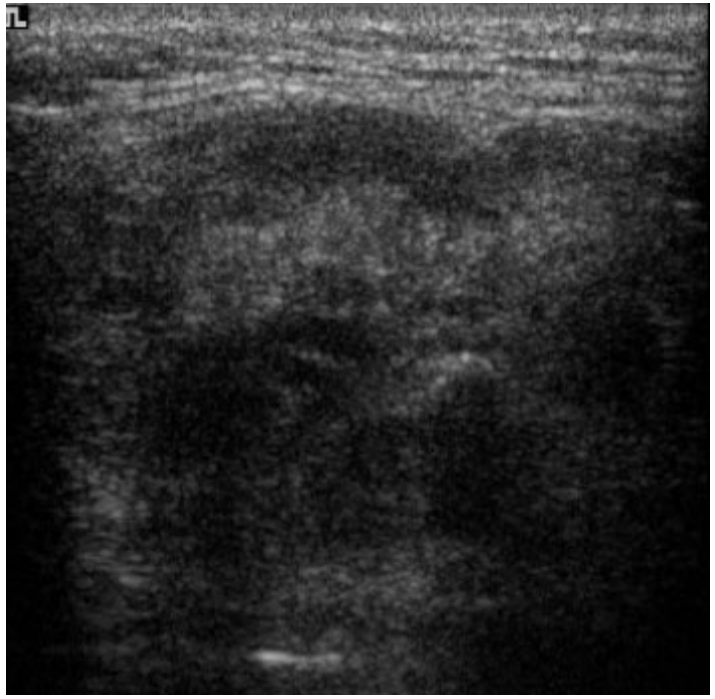
Investigation

- Coagulation for large VM
  - 88% have a localized intravascular coagulopathy (LIC)[1]
- Doppler ultrasound
- MRI if a treatment is considered or diagnostic unclear
- TDM not contributive
- Angiography not necessary +++

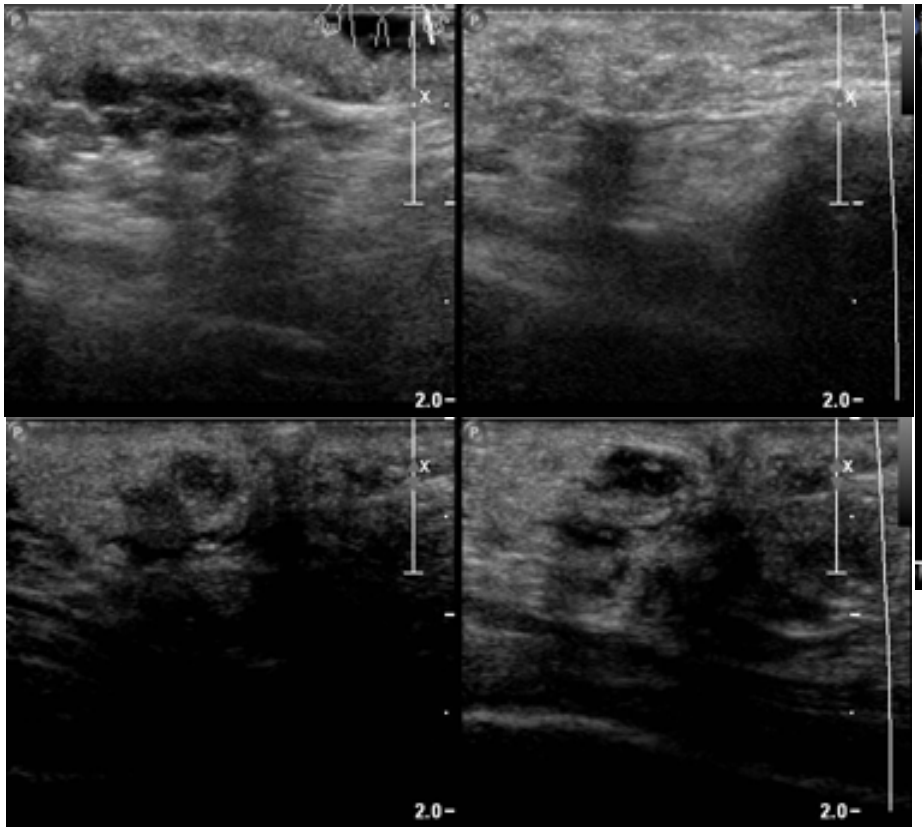
1. Enjolras O, J Am Acad Dermatol 1997;36:219–25.

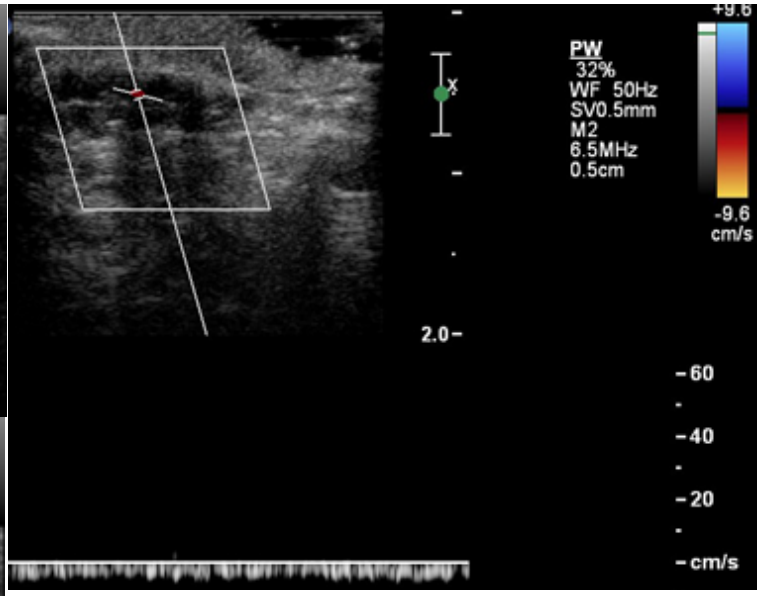
Venous malformation

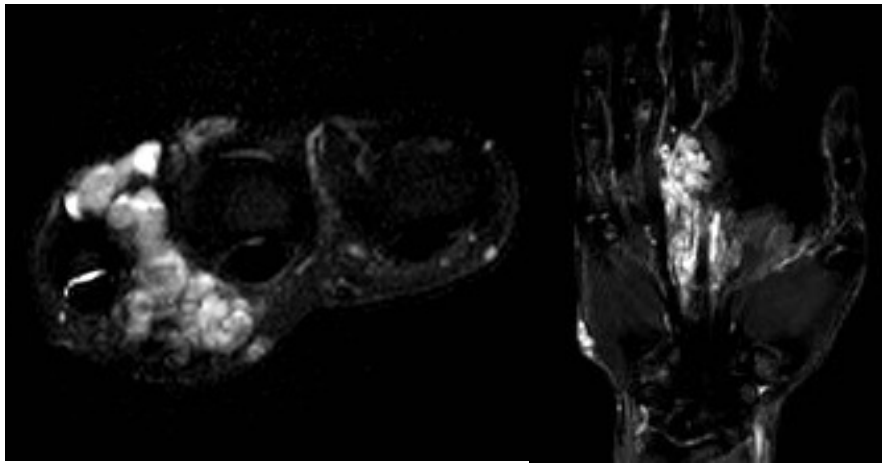
- Doppler ultrasound
  - Low flow
  - Hypoechoic
  - Compressible venous dilatation
  - Phlebolitis
  - Evaluate feasibility of needle guidance for sclerotherapy



Compressibility & Doppler

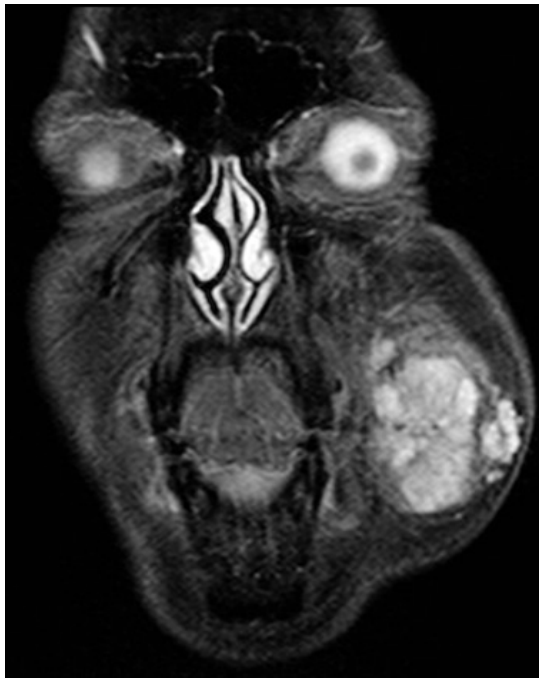


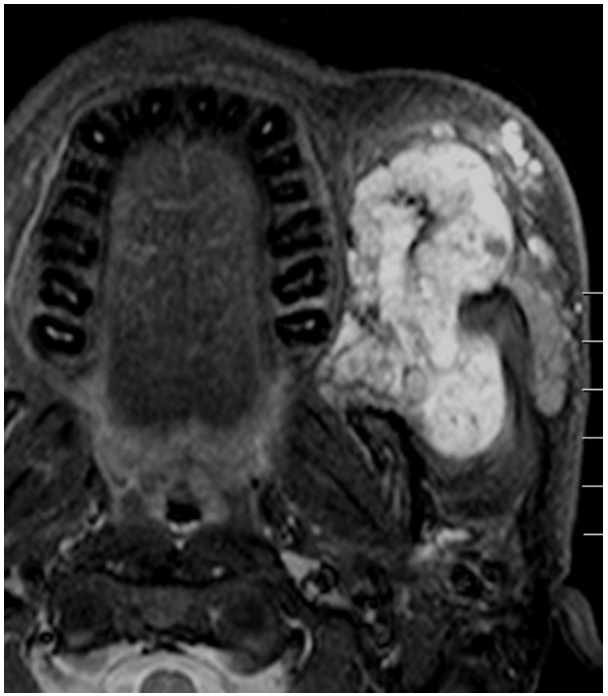


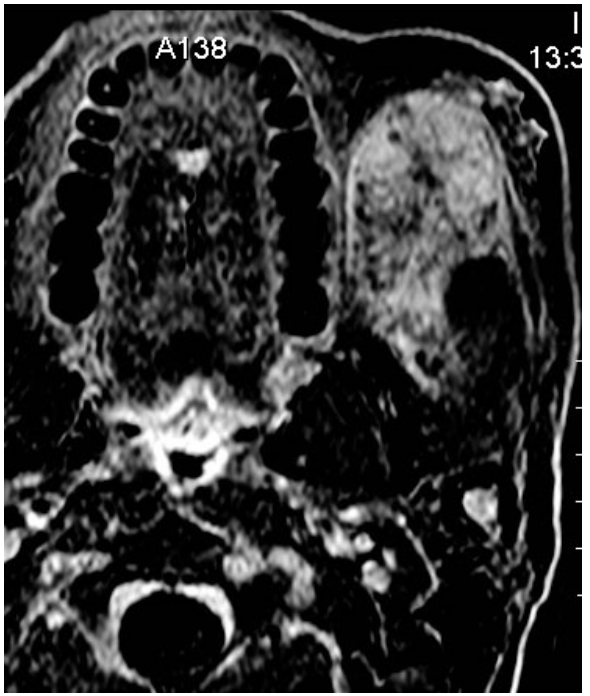


VM & MRI

- Best examination for extension
- T2 (STIR), T1 and T1 fat sat post gado





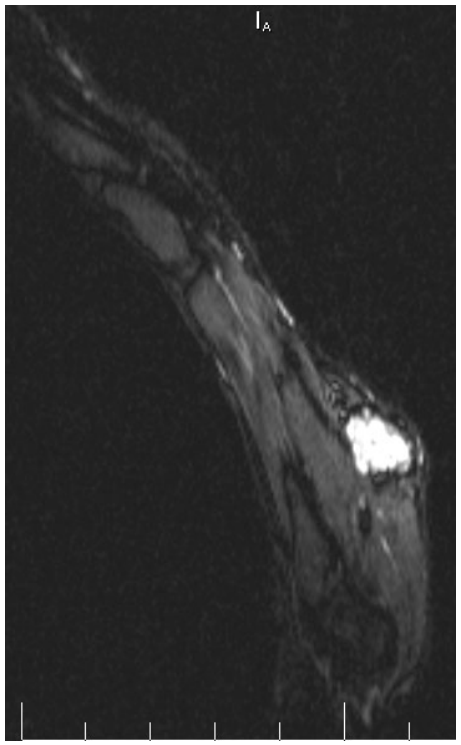


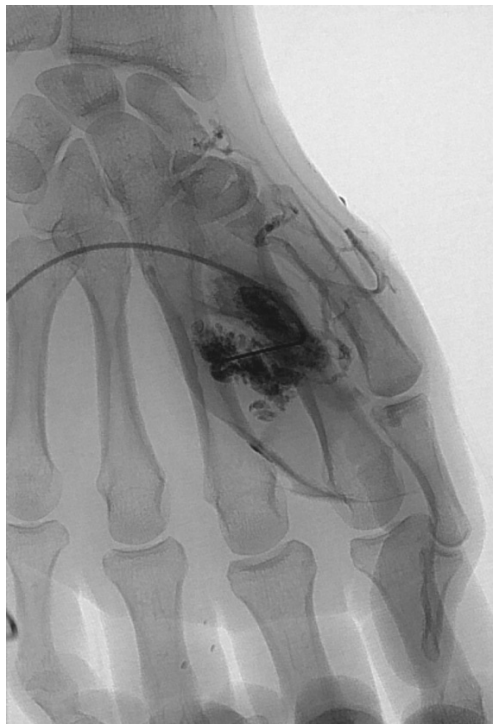
Invasive treatment

- Sclerotherapy
  - Failure of conservative treatment
    - Pain
    - Aesthetic
    - Bleeding
    - Oropharyngeal compression
- Rarely surgery
  - Intramuscular
  - FAVA

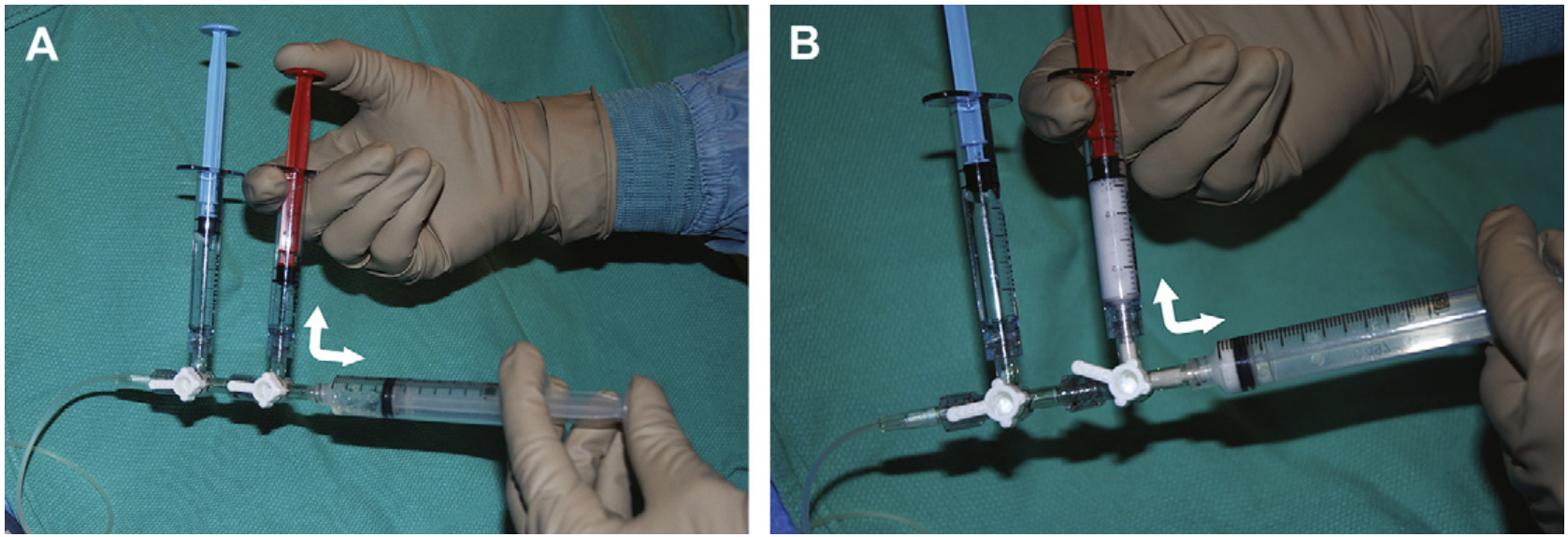
Sclerotherapy

- Ethanol
- STS 3% (foam air + lipiodol)
- Fluorosocopic ans ultrasound guidance
- Session 6-8 weekss





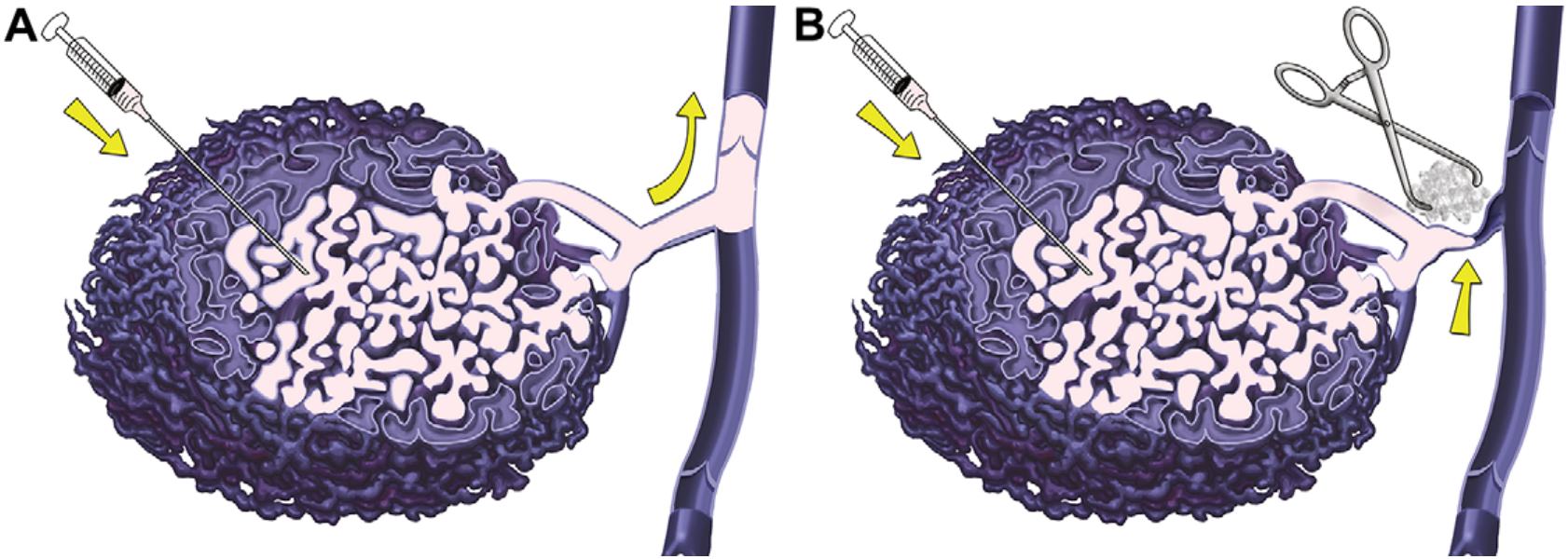
Foam-STS



Efficacy foam-STS > STS liquide

Yamaki T et al. *J Vasc Surg*. 2008;47:578-584

Foam STS

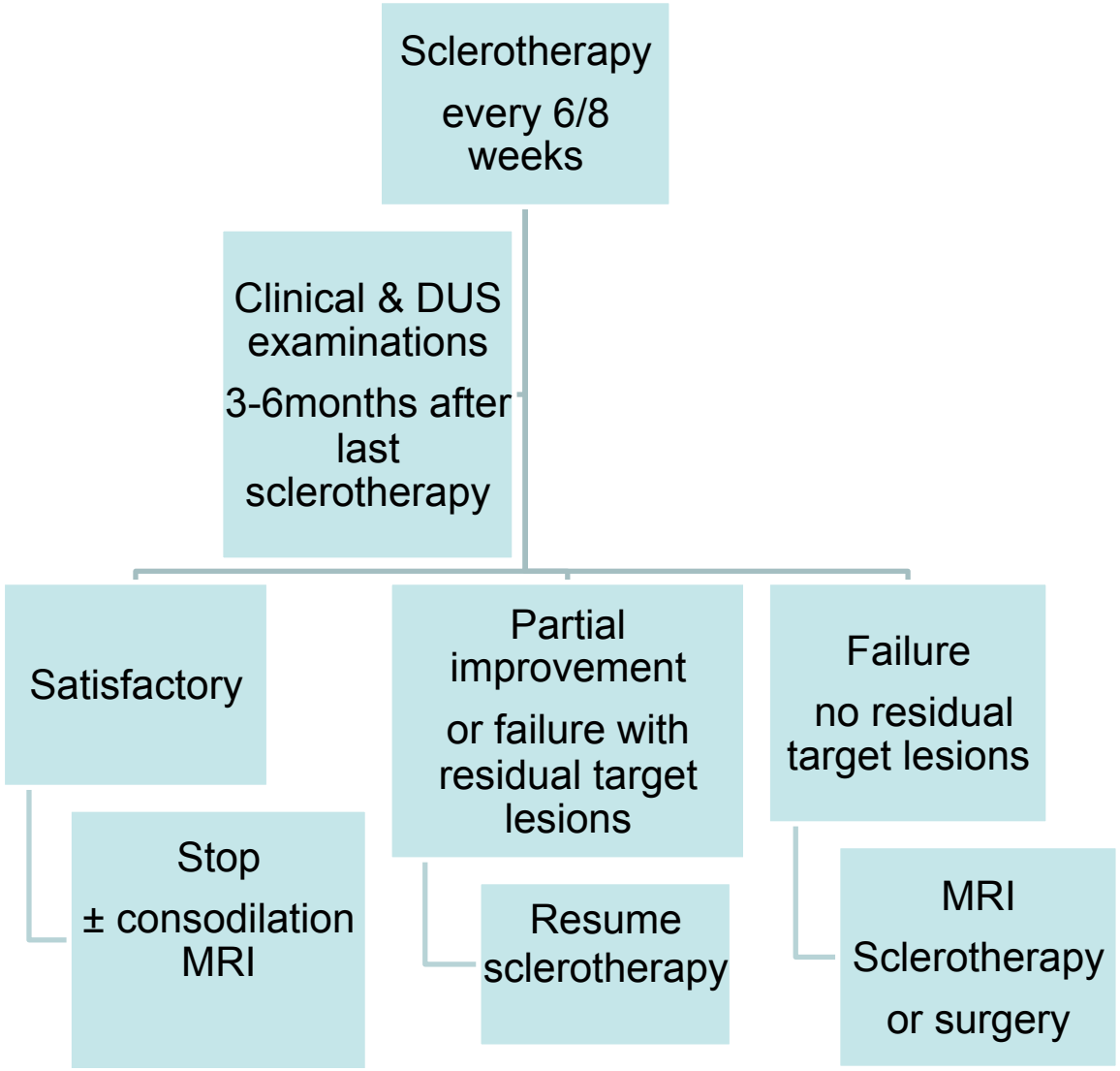


Legiehn et al Radiol Clin North Am 2008

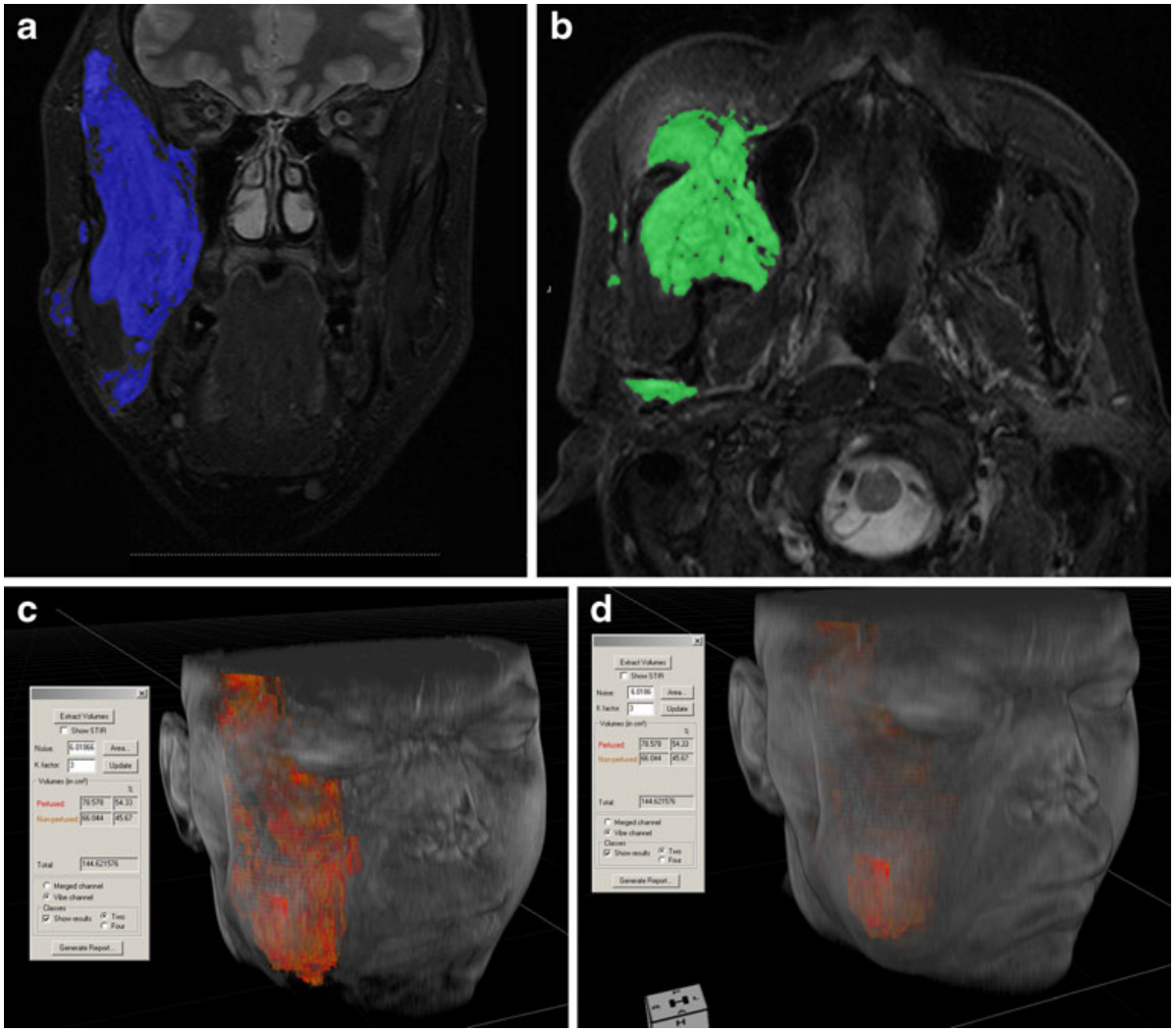
Post intervention

- NSAID/corticoid
- Pain medication
- LMWH if suspicion of foam migration in central venous & truncal lesions

VM follow-up


Sclerotherapy
every 6/8 weeks
Clinical & DUS examinations
3-6months after last sclerotherapy
Satisfactory
Stop
± consodilation MRI
Partial improvement
or failure with residual target lesions
Resume sclerotherapy
Failure
no residual target lesions
MRI
Sclerotherapy
or surgery

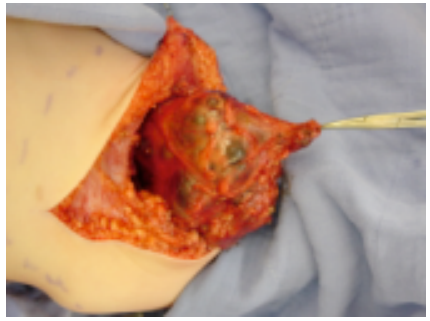
3D modeling

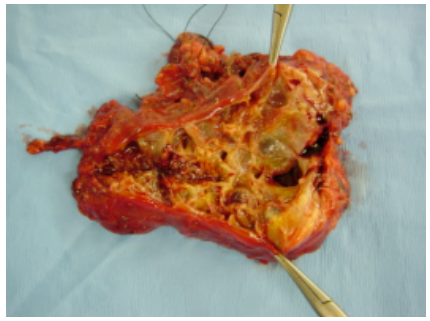


Caty V et al. Europ Radiol 2013

Lymphatic malformation

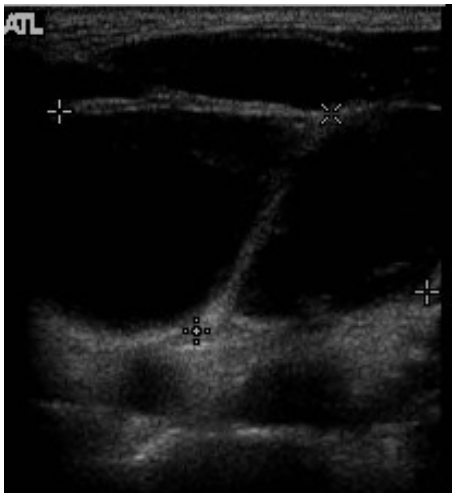
- Cystic cavity lined by an endothelial layer filled by a lymphatic fluid
  - ML macrocystic (> $2cm^3$)
  - ML microcystic (< $2cm^3$)
  - Mixed lesion (micro-macro)
  - Mixed lesion lymphatic and venous
- Present at birth
- Growth childhood-teenage

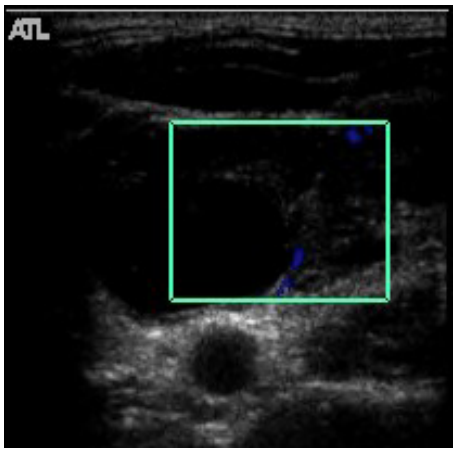




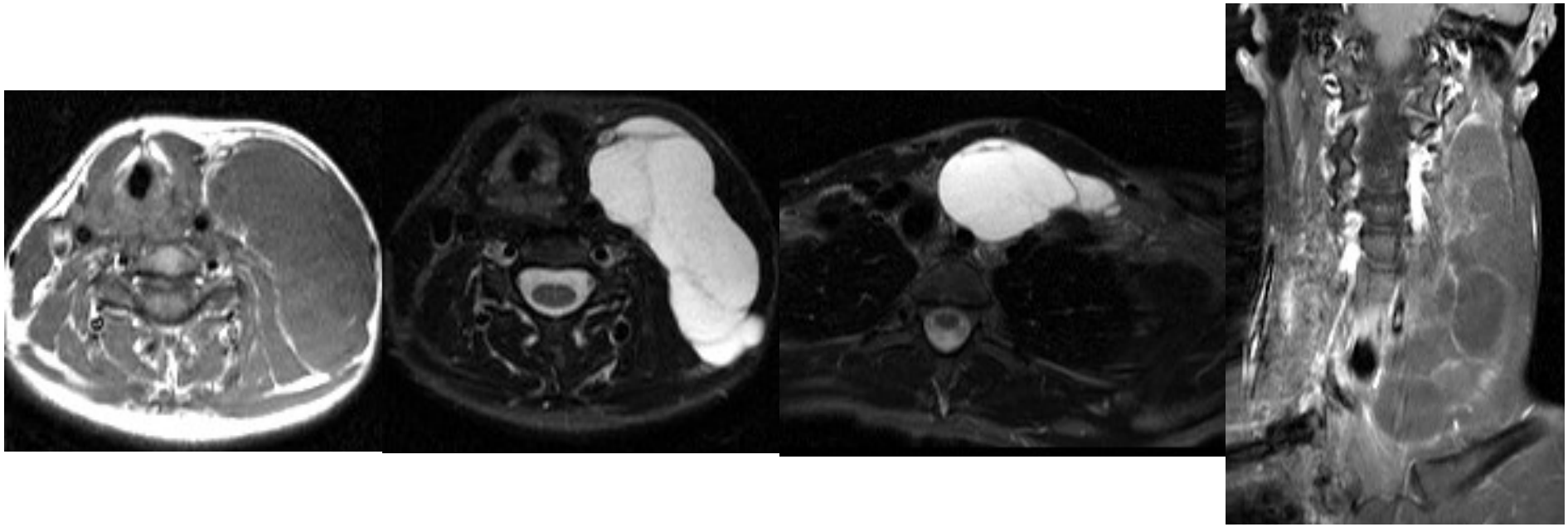
Lymphatic malformation

- Doppler ultrasound
  - Macrocyst with septation
  - Doppler
    - No flow in the cyst
    - High resistance flow in septa
  - Microcystic lesions infiltration of soft tissue by hyperechoic or hetereogeneous mass

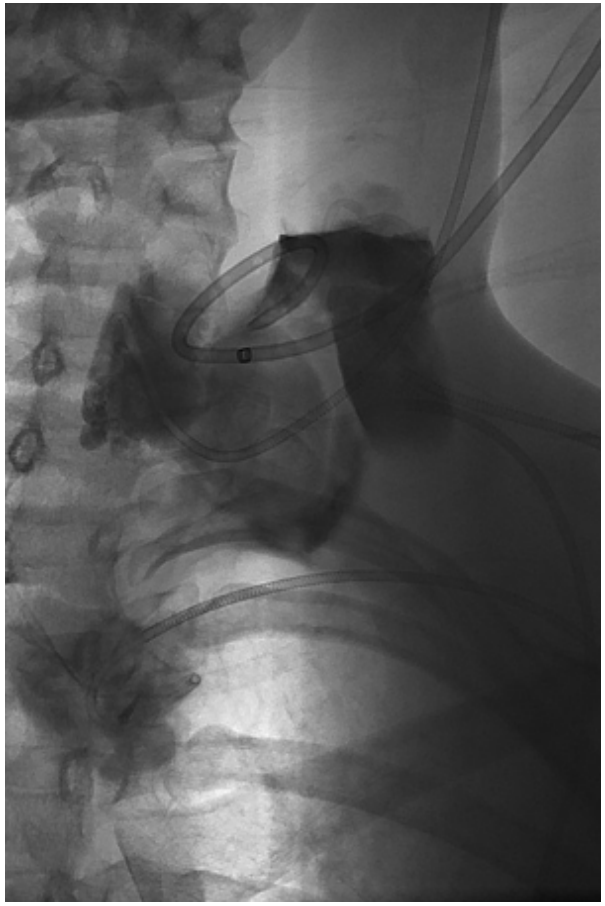


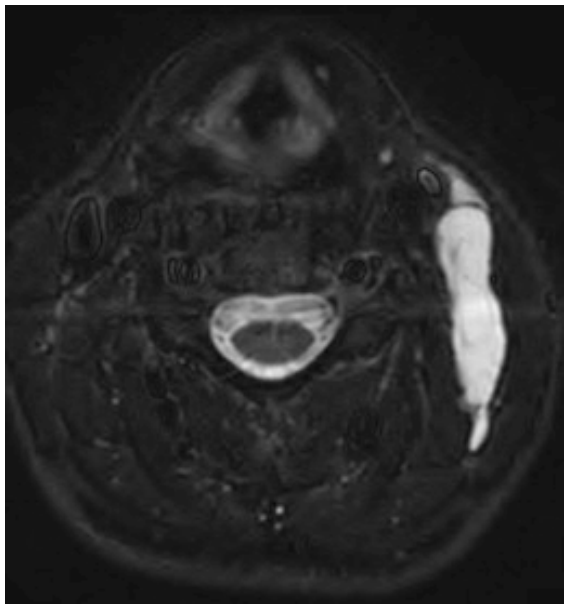


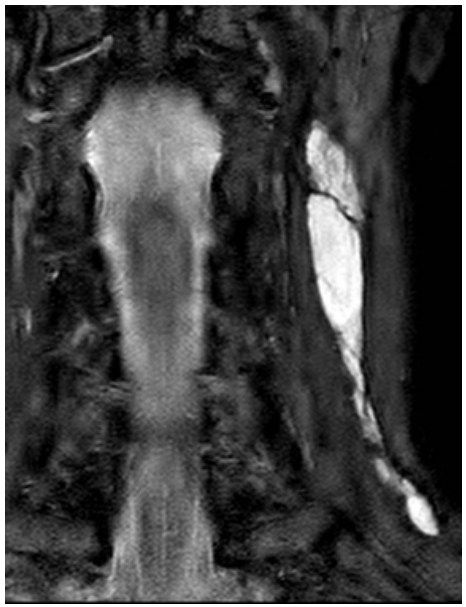
MRI & LM



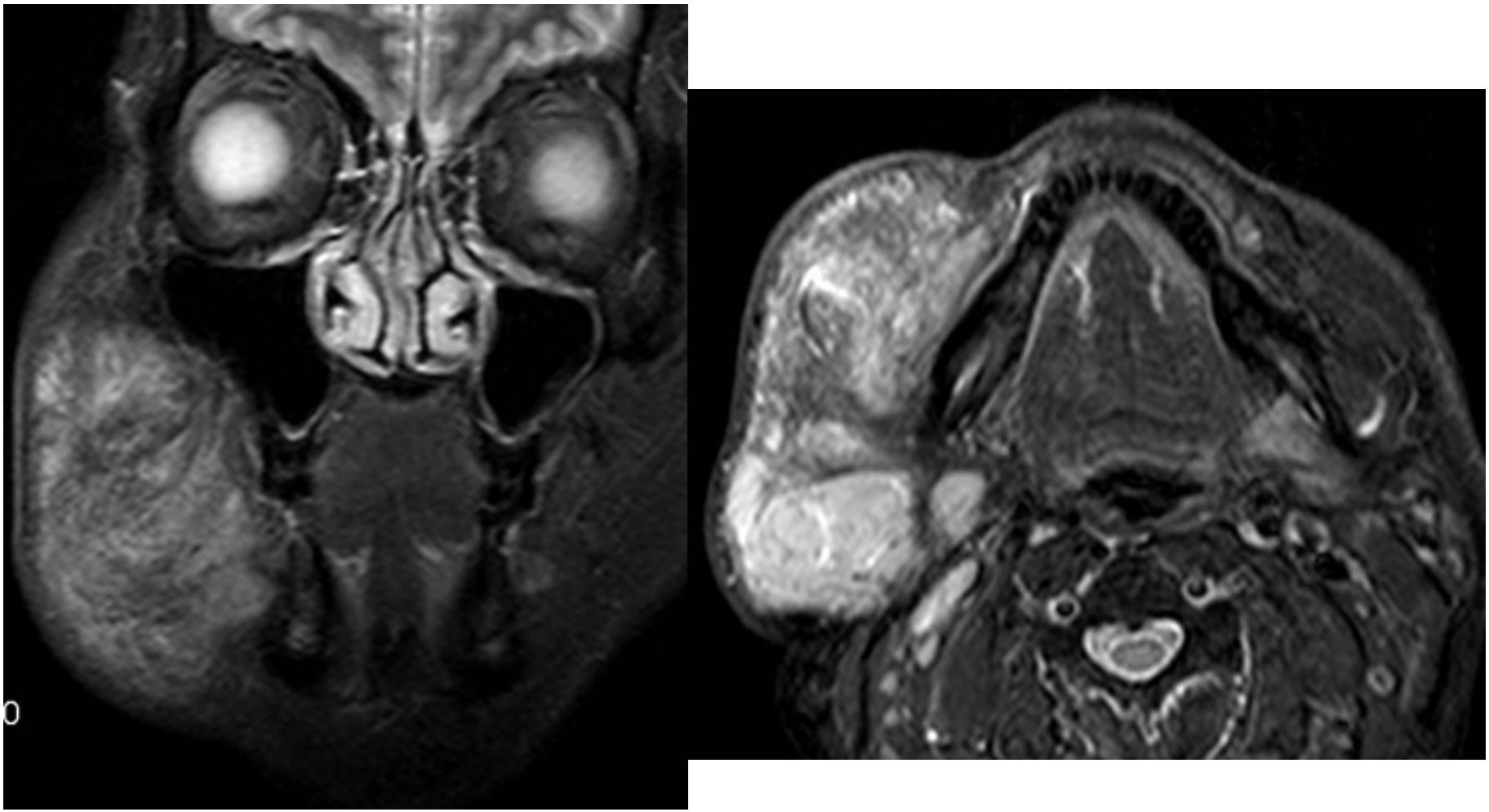
Sclerotherapy Bleomycine



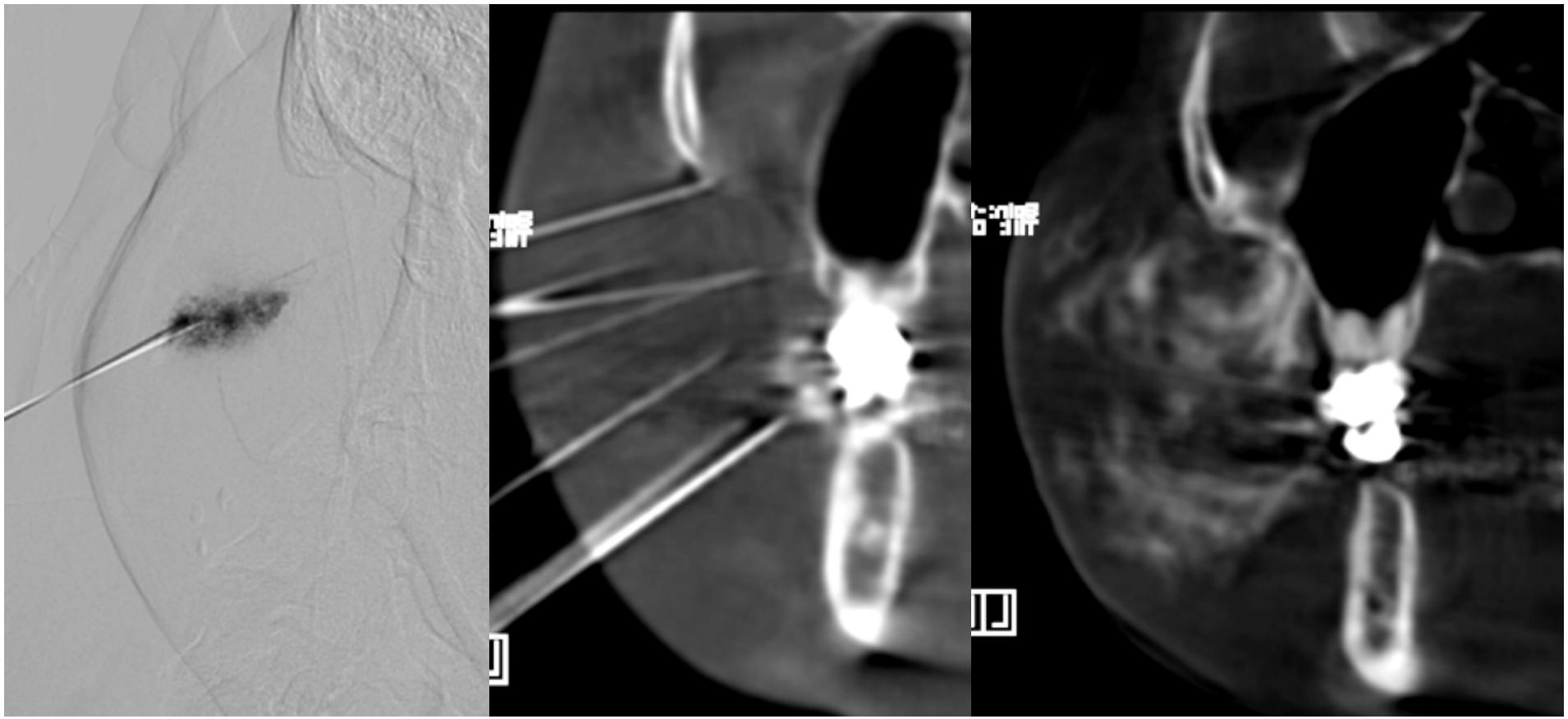




Microcystic lymphangioma



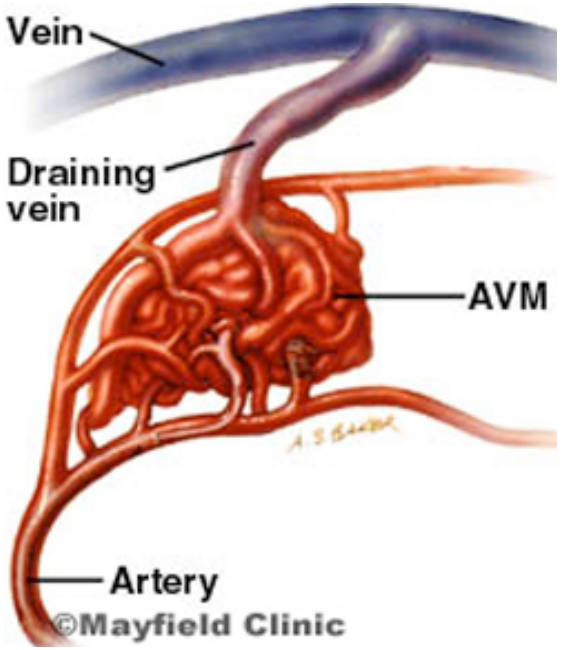
Bleomycine



15 mg bleomycine in 15 ml nacl + 5ml contrast

Arterio-venous malformation

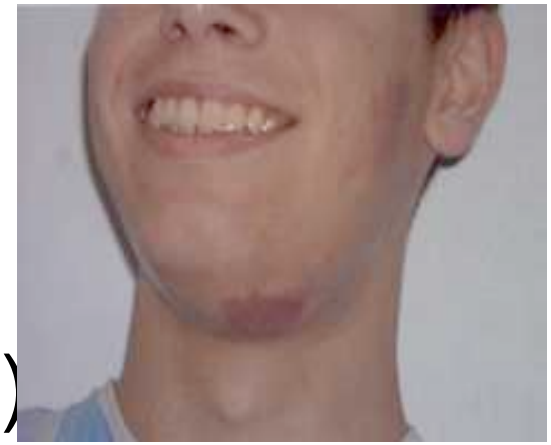
- High-flow malformation
  - AV-shunting
  - Nidus
- Congenital
  - Expansion
    - Teenage
    - Pregnancy



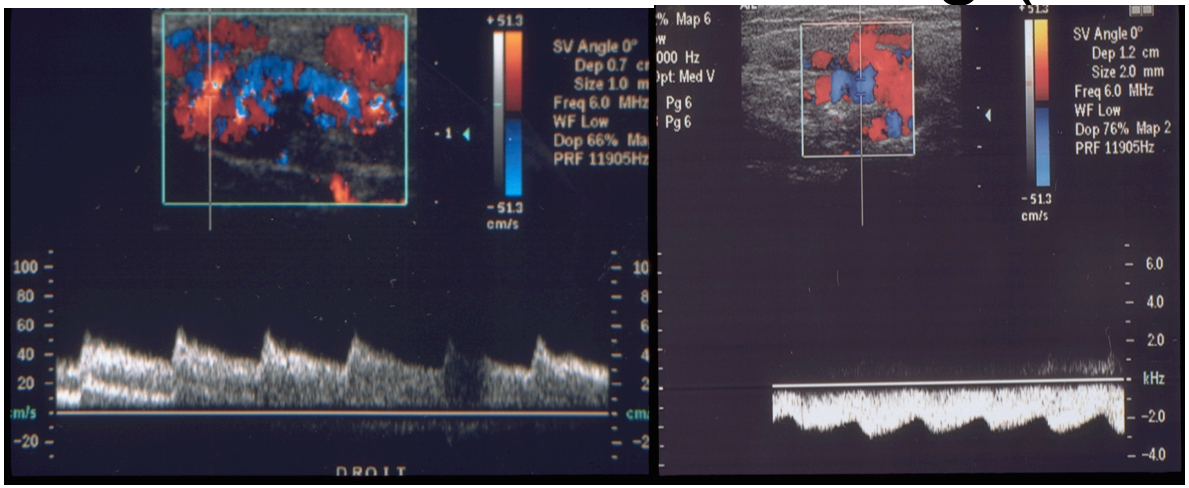
Schobinger classification

(Clinical staging system to grade the evolution of AVMs)

Stage 1: Quiescent



- Pink-bluish stain
- Warm
- Arteriovenous shunting (DUS)



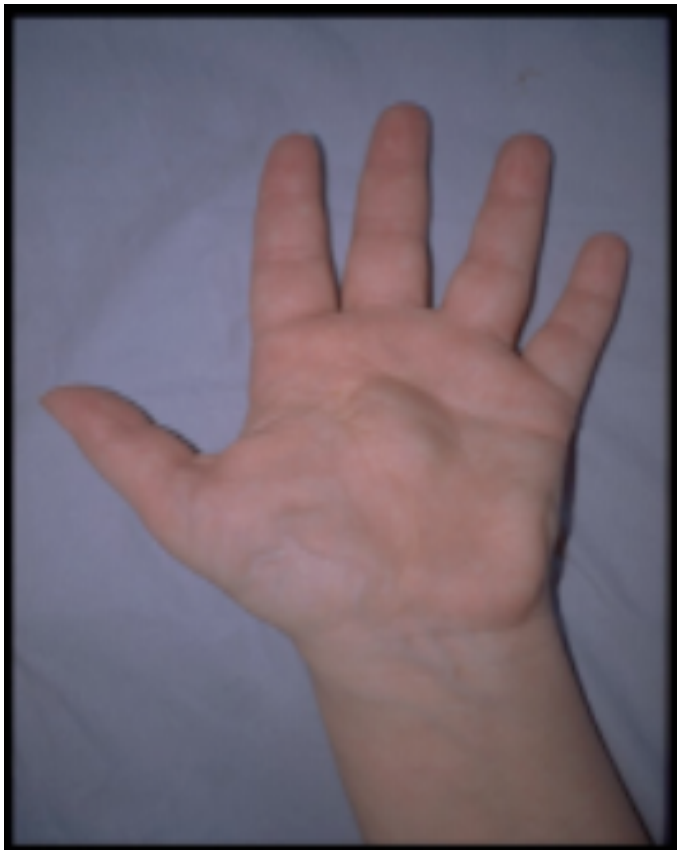
Schobinger classification

Stage 2: Expansion

Stage 1

+

- Darkening blush stain
- Pulsations
- Thrill
- Bruit
- Tortuous/tense veins



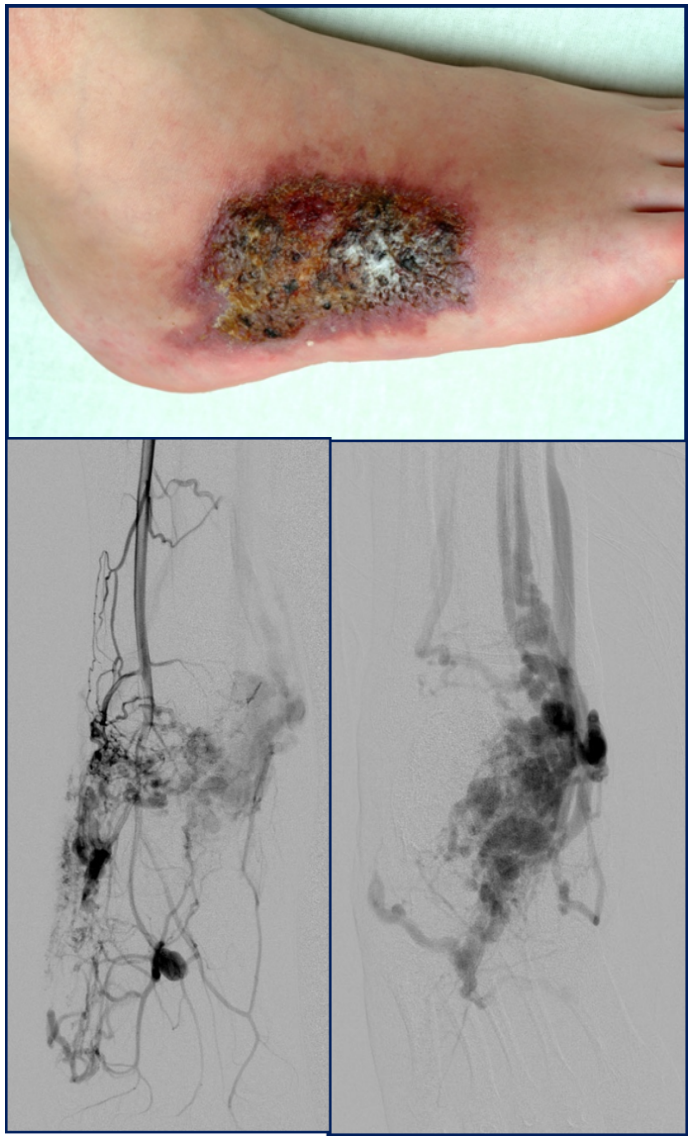
Schobinger classification

Stage 3: Destruction

Stage 2

+

- Steal
- Distal ischemia
- Dystrophic skin changes
- Ulceration
- Bleeding
- Persistent pain
- Tissues necrosis
- Soft tissues and bones changes

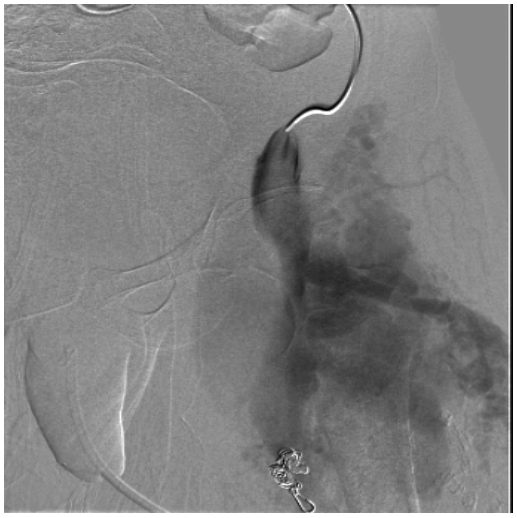


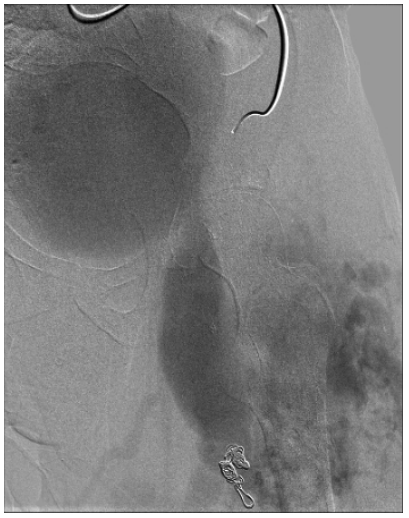
Schobinger classification

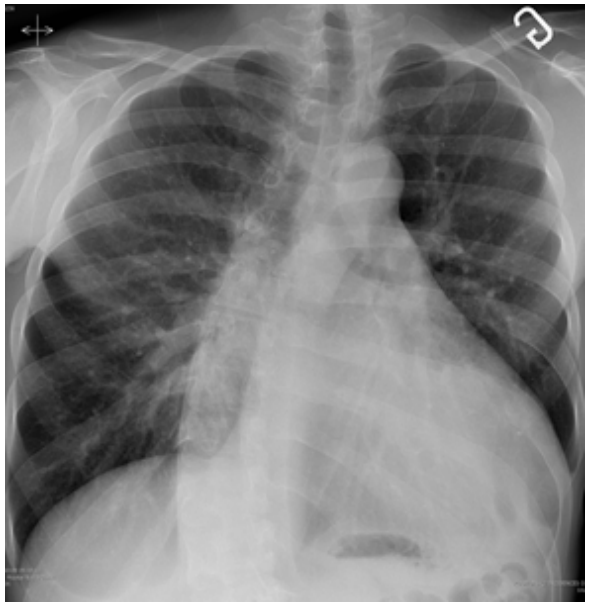
Stage 4: Decompensation

Stage 3 +

- High output cardiac failure







Doppler +++

High flow vascular nidus

–Arterial side

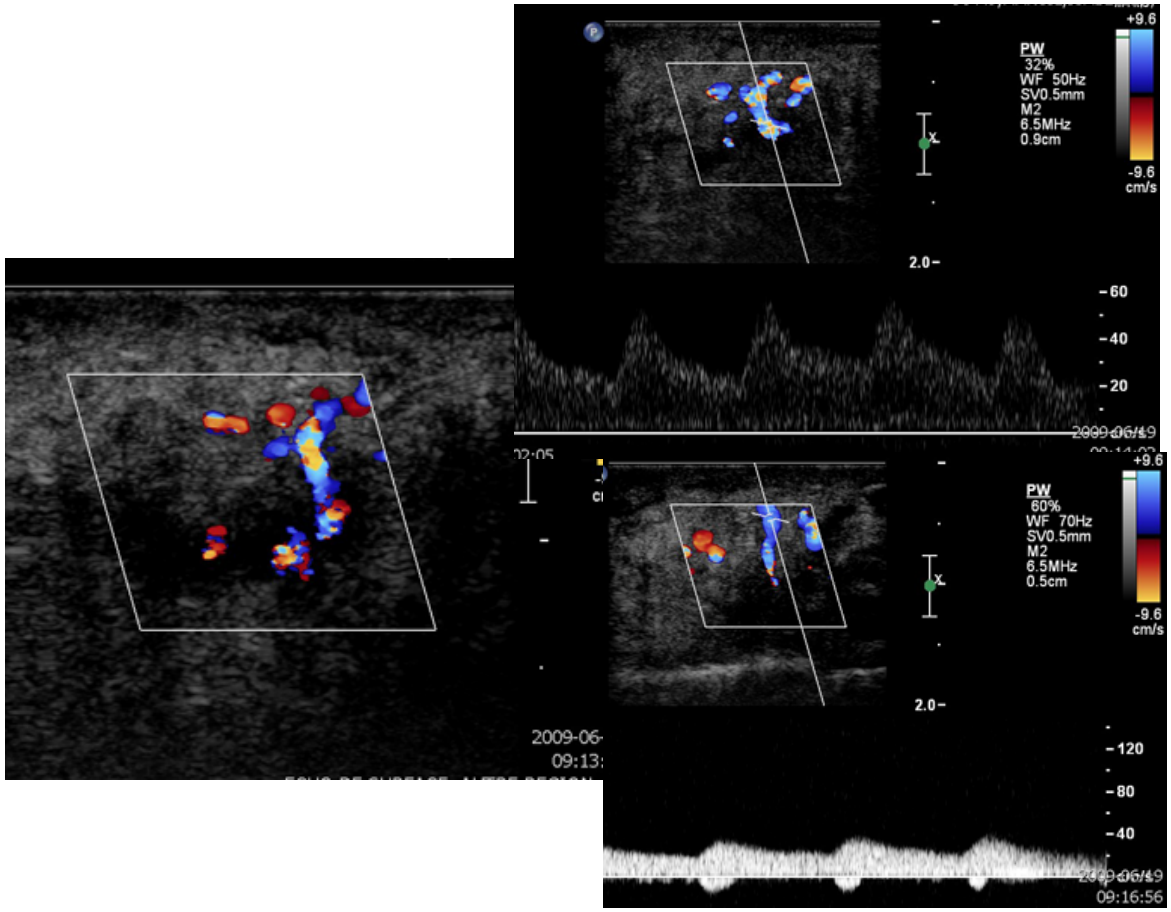
- High velocities and diastolic flow

–Venous side

- Arterialization of venous flow

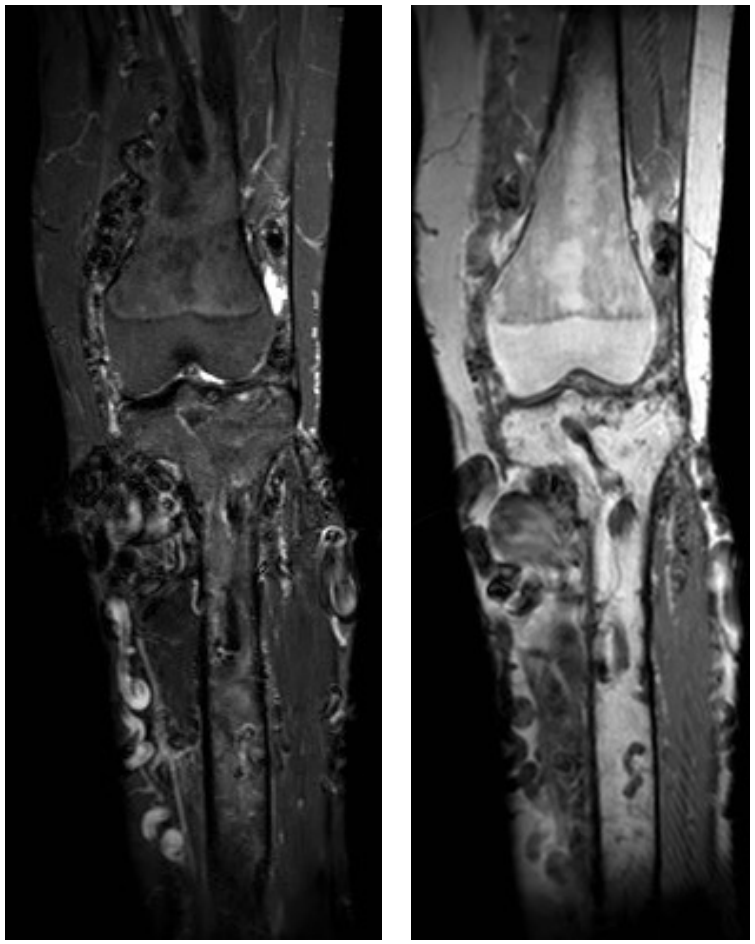
–Evaluate flow imbalance

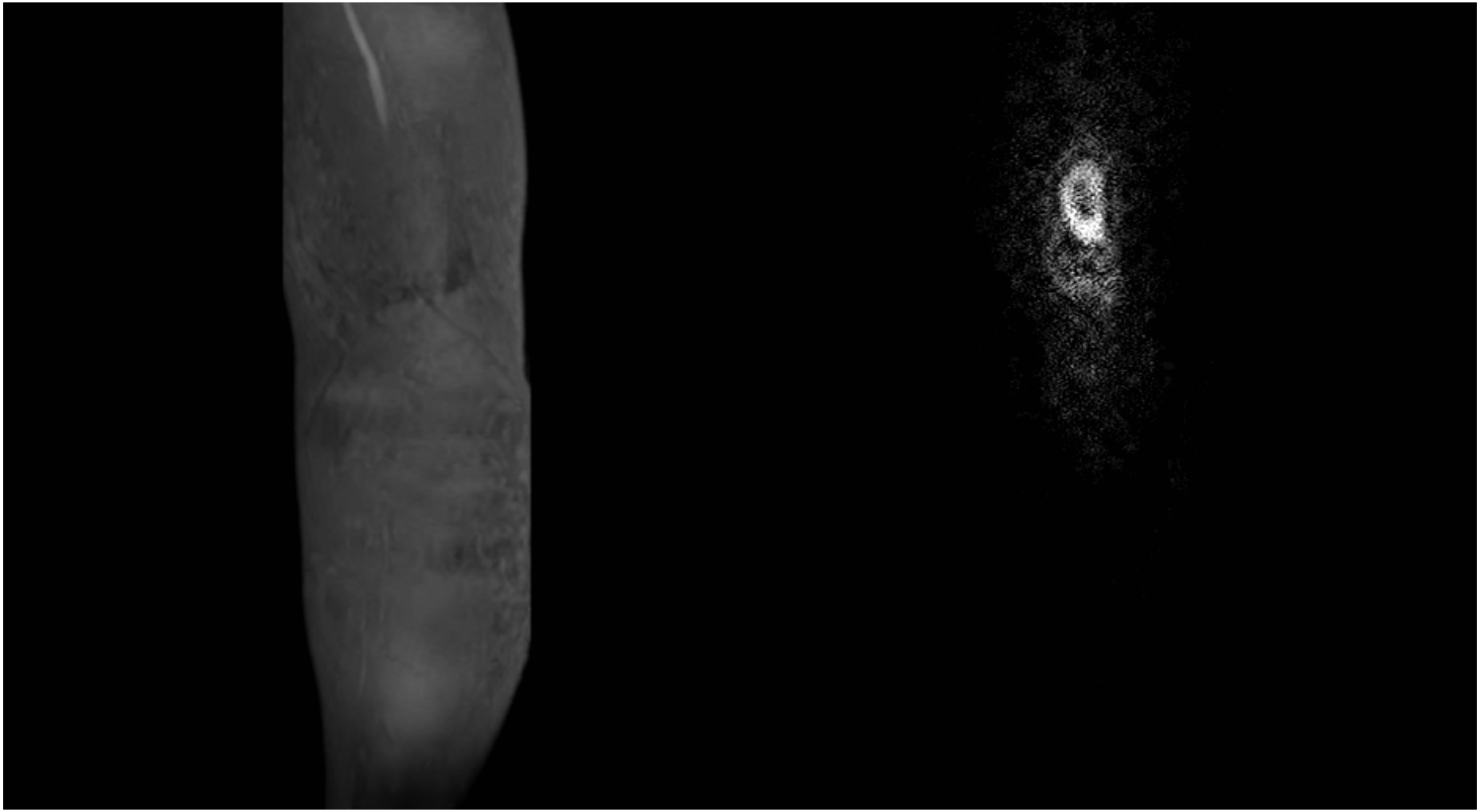
- Arterial feeder
- Venous drainage
- Normal arteries distal to AVM +++



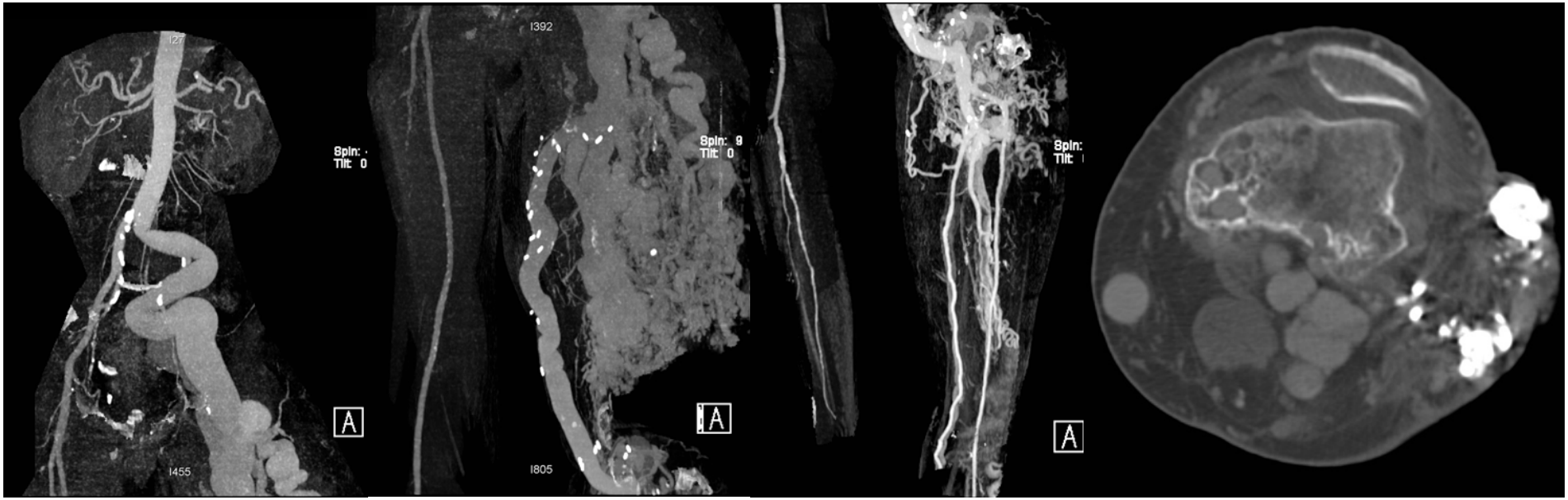
Investigation

- MRI
  - Flow void T1-T2
  - 4D angio
  - High resolution steady state



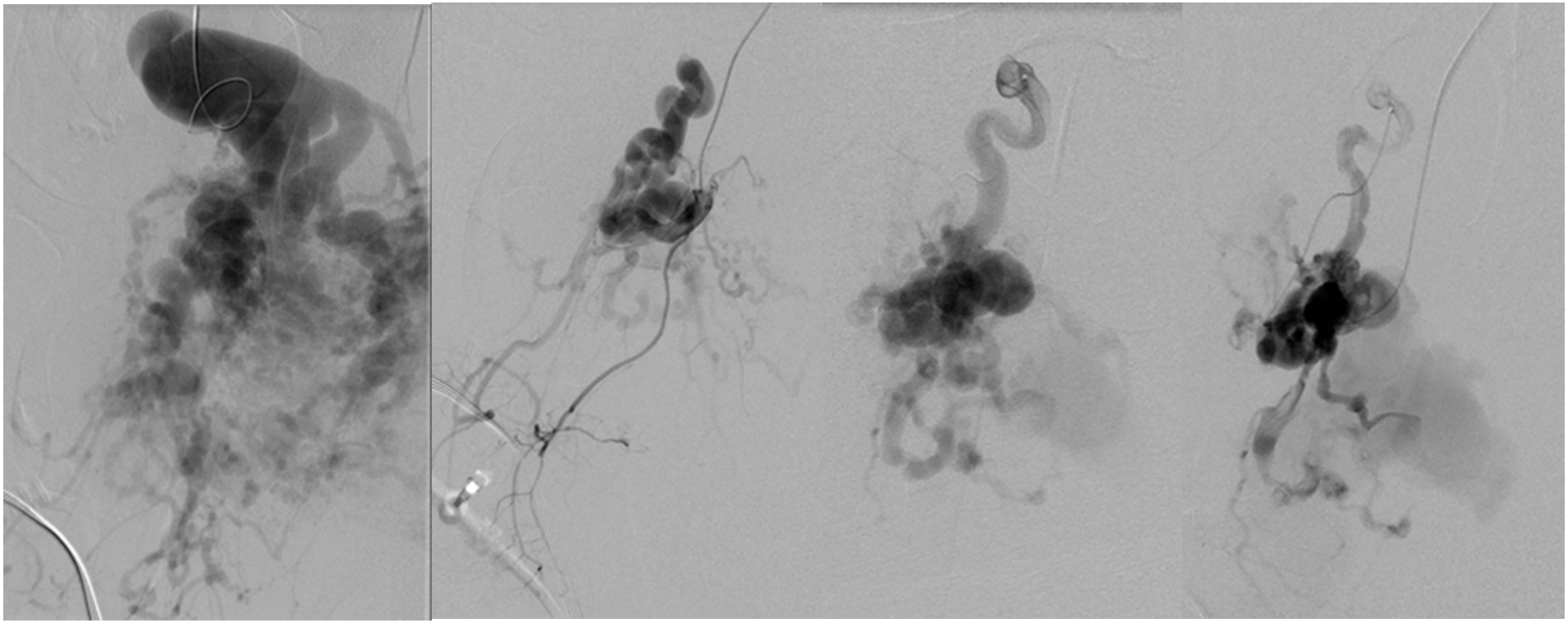


CT-angiography



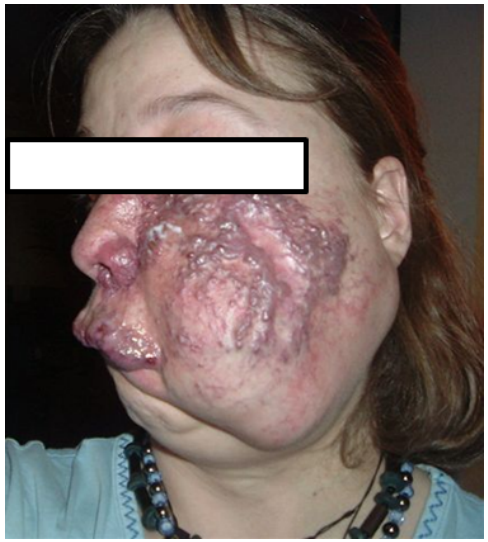
Arterial aneurysm and bone destruction

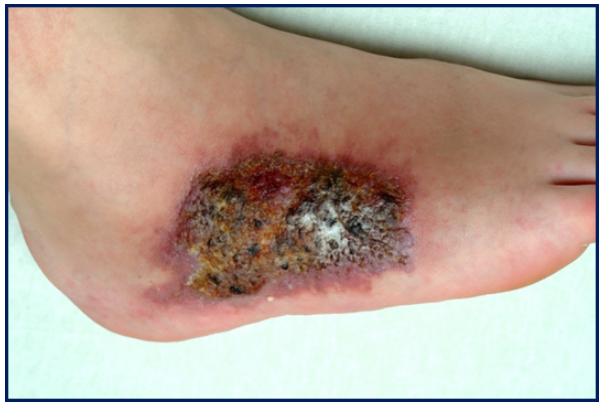
Embolization (ethanol)



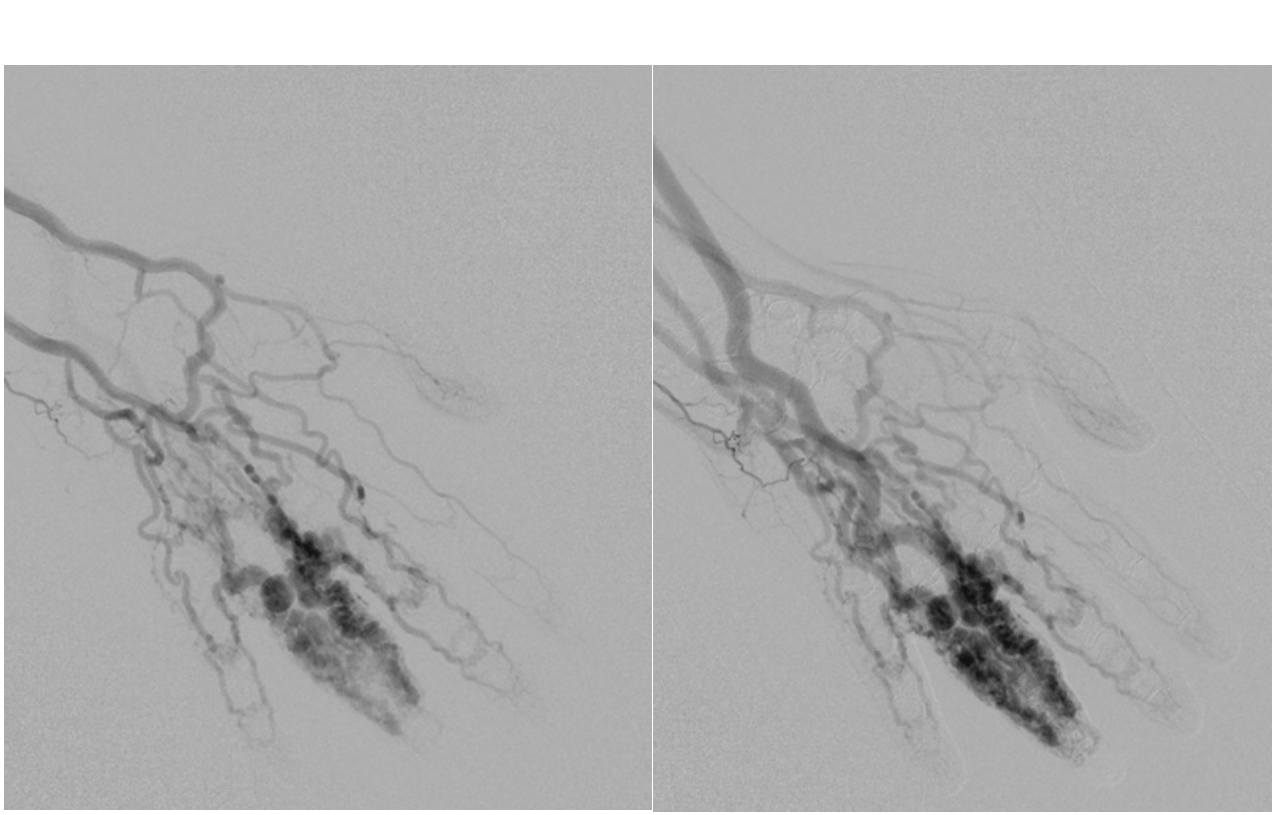
Adapt your strategy to patient symptoms

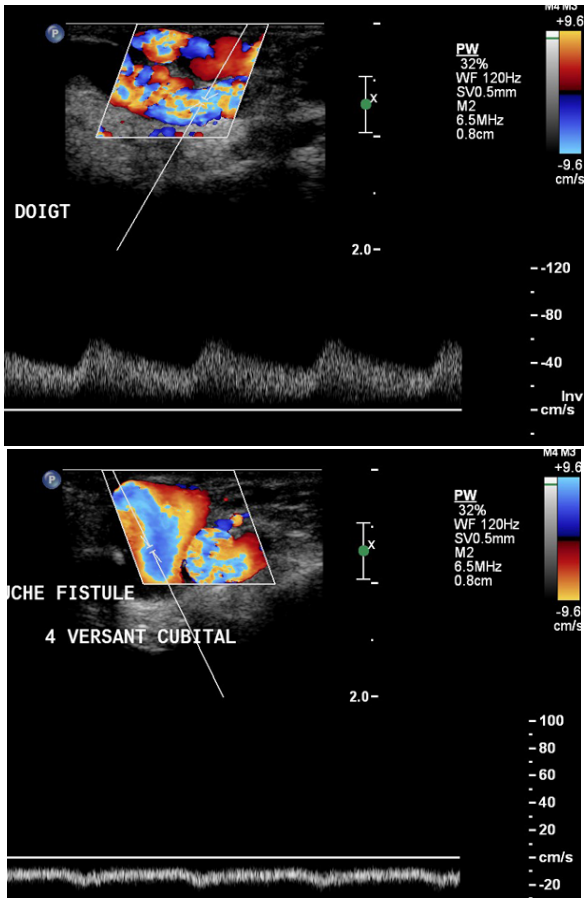
- Venous congestion or haemorrhage
  - Aggressive on the arterial side before occluding the vein
- Tissue necrosis due to capillary shunting
  - Aggressive on the venous side
- Rely on Doppler ultrasound



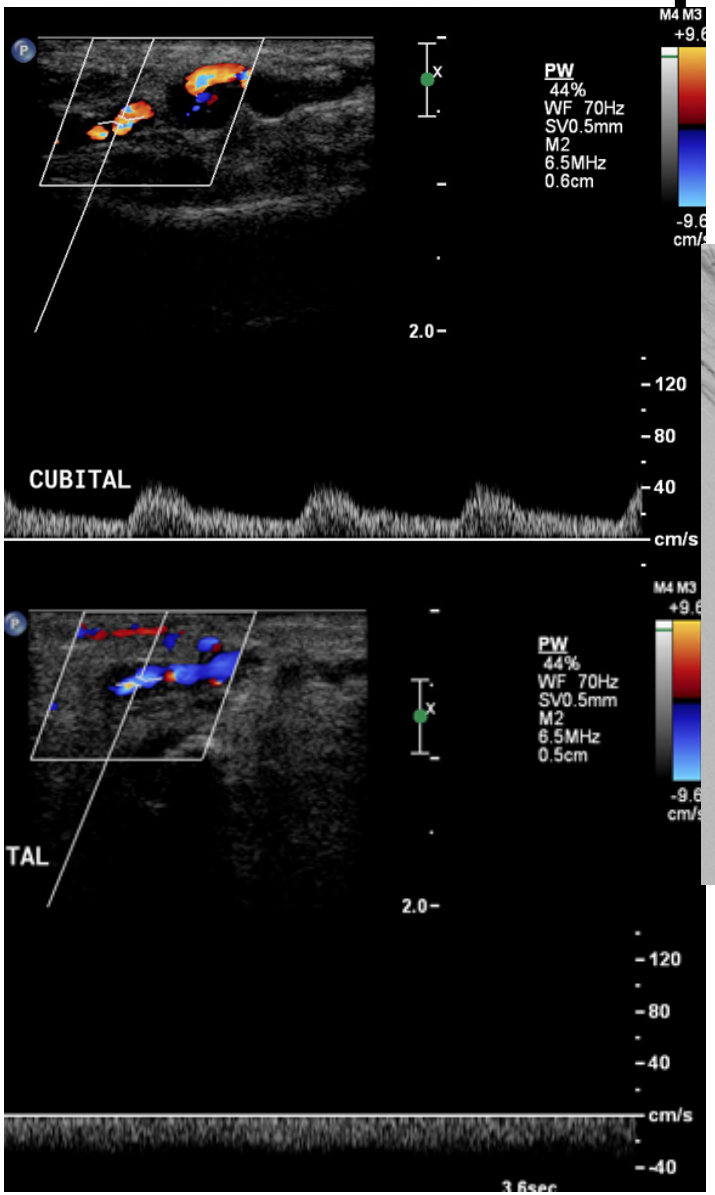


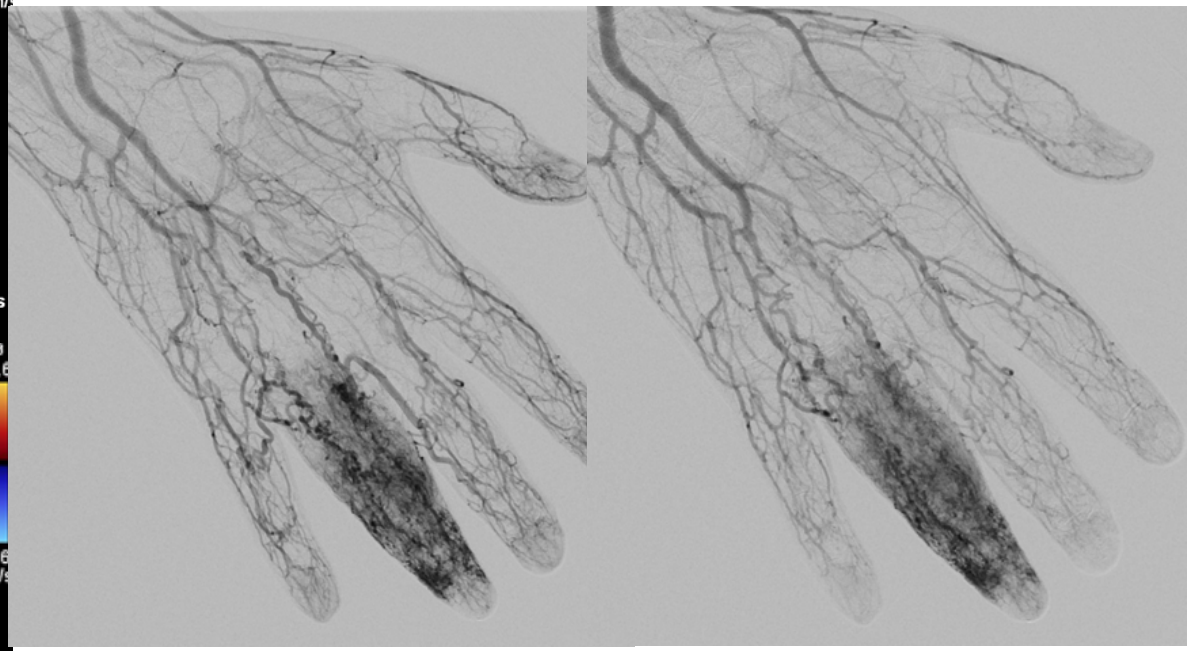
Correlation between Doppler and DSA





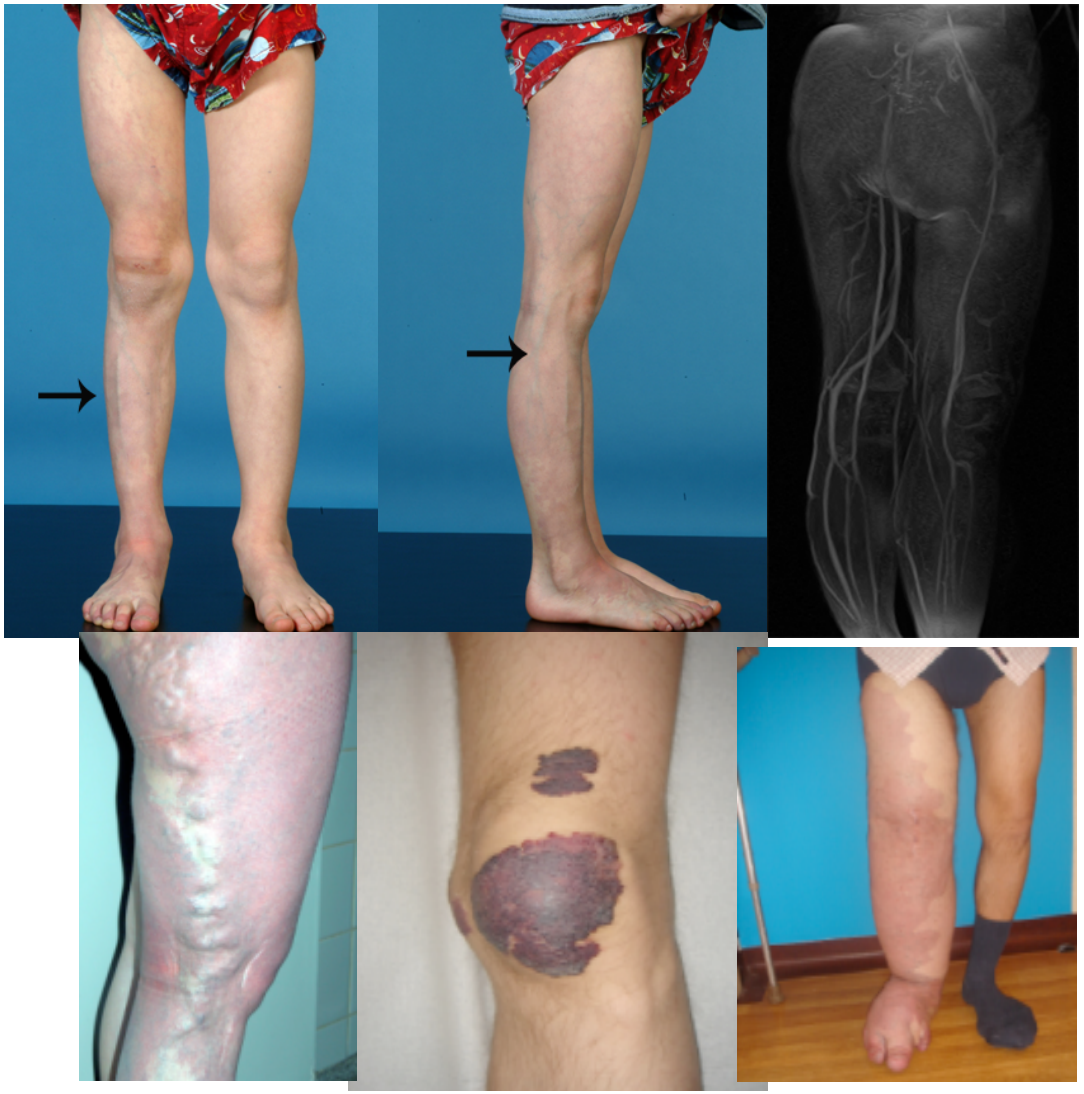
Follow-Up


PW
44%
WF 70Hz
SV0.5mm
M2
6.5MHz
0.6cm
2.0
120
80
40
cm/s
CUBITAL
PW
44%
WF 70Hz
SV0.5mm
M2
6.5MHz
0.5cm
2.0
120
80
40
cm/s
-40
3.6sec



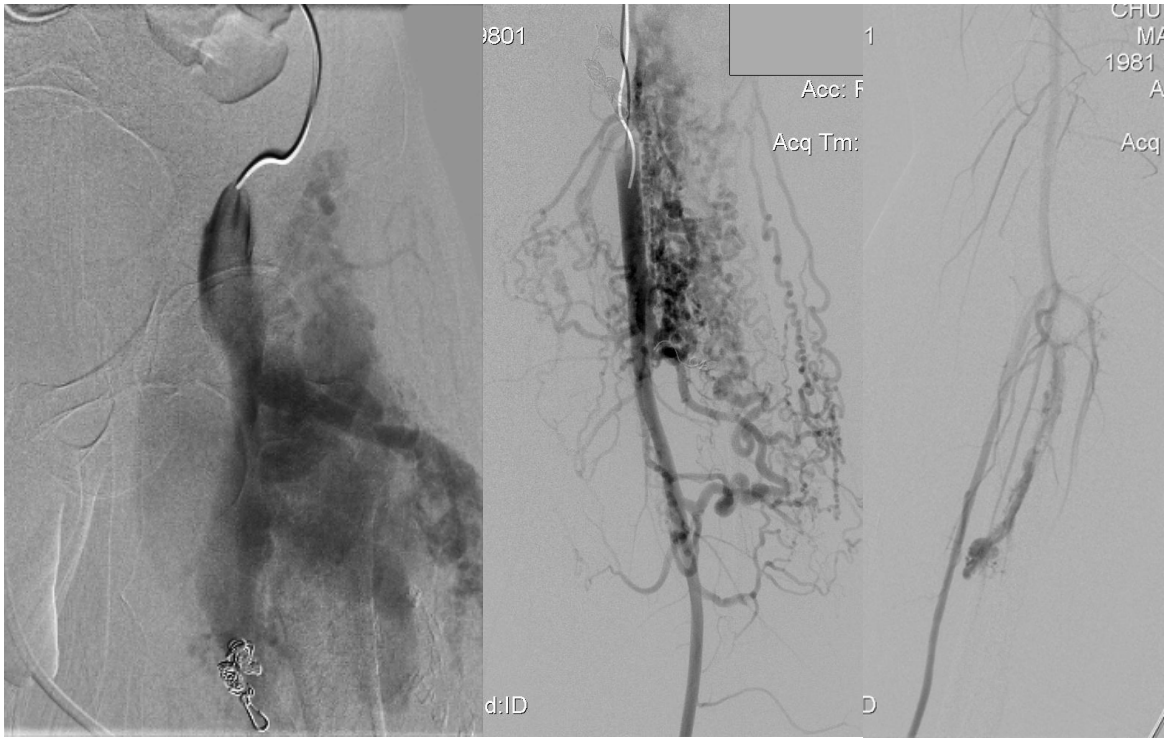
Klippel trenaunay

- Limb hypertrophy
- Cutaneous angioma
- Venous and or lymphatic
- R/O hypoplasia deep venous system
- Sclerosis of varicose vein

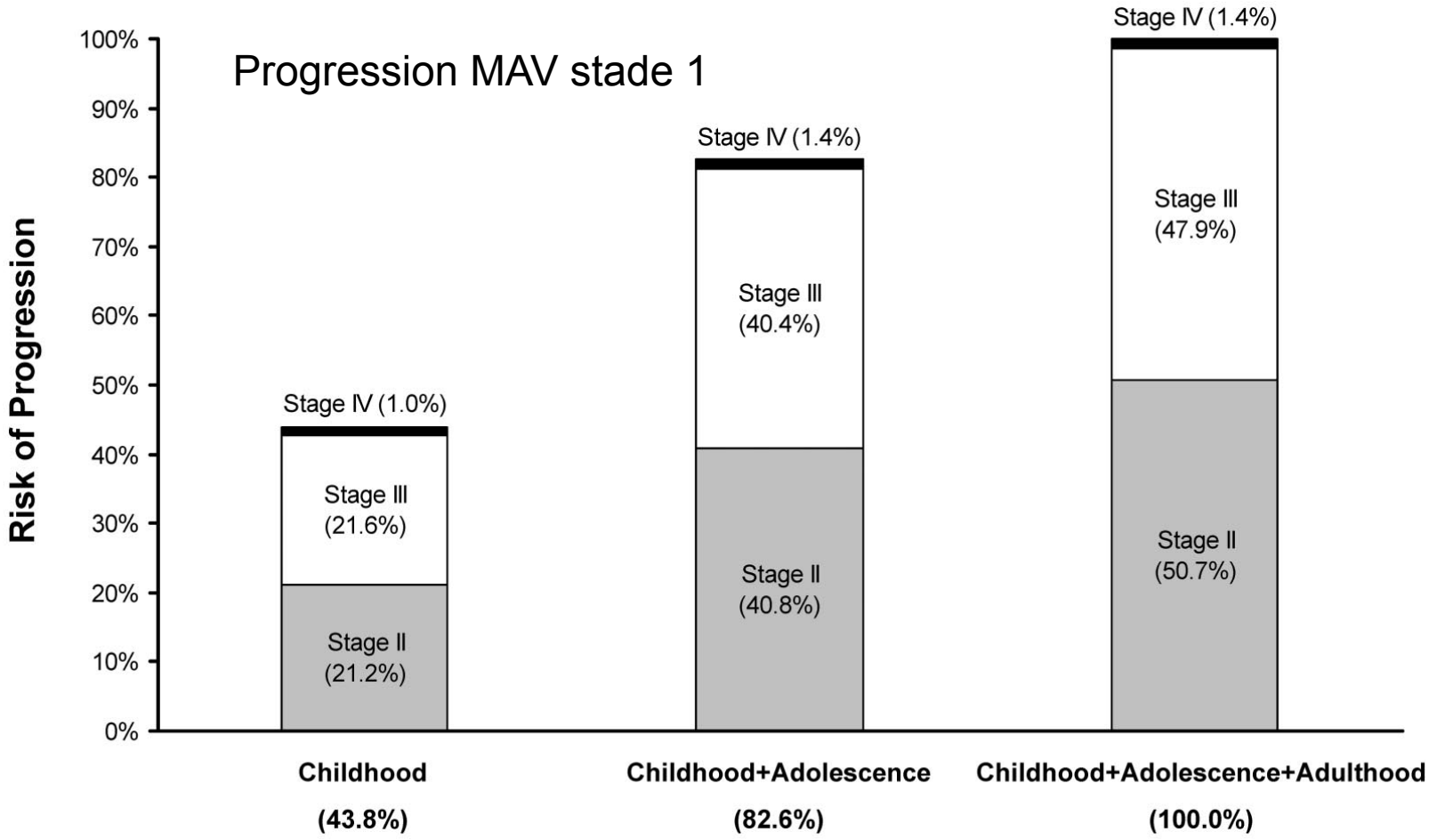


Parkes Weber

- AVM
- Cutaneous angioma
- Limb hypertrophy



AVM evolution: Boston Study: n=272



Liu AS et al Extracranial arteriovenous malformations: natural progression and recurrence after treatment .Plastic and Reconstructive surgery 2010:1185

Recurrence embo versus surgery (Boston study n=272)

- Mean FU: 8.9y ±5.2y
  - Recurrence all patients 93%

- Predictor of recurence:
  - Embo vs surgery (combined or not with embo)
  - Shobinger stage at treatment

- Resection
  - recurrence rate = 81%
  - Time to recurrence: 42.7% > 1y
- Embo alone
  - Recurrence rate= 98%
  - Time to recurrence: 14.4 % > 1y
- Selection bias +++

Liu et al Extracranial arteriovenous malformations: natural progression and recurrence after treatment . Plastic and Reconstructive surgery 2010:1185

Conclusion

- Make the diagnosis
- Clinical examination, Doppler, MRI
- Treat symptoms
- Multidisciplinary approach
- IR pivotal role
  - Must be involved in clinical evaluation and imaging work-up and follow-up