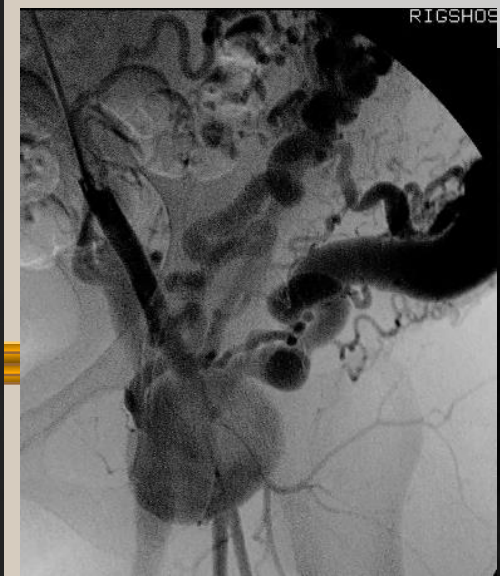
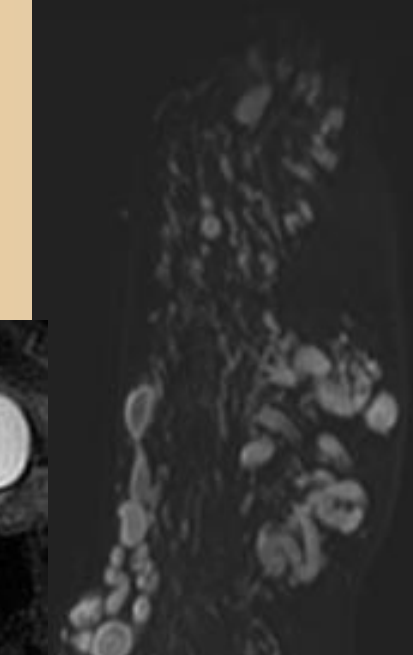
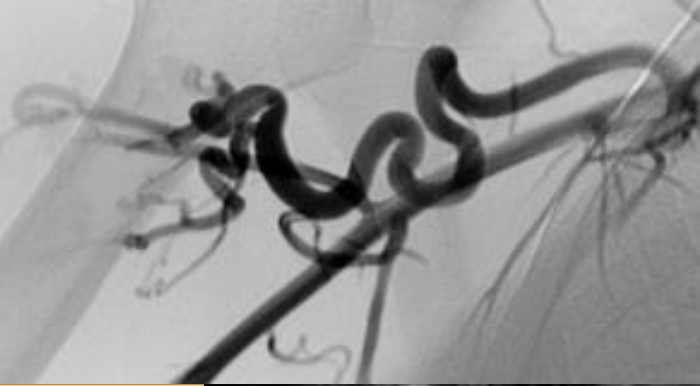


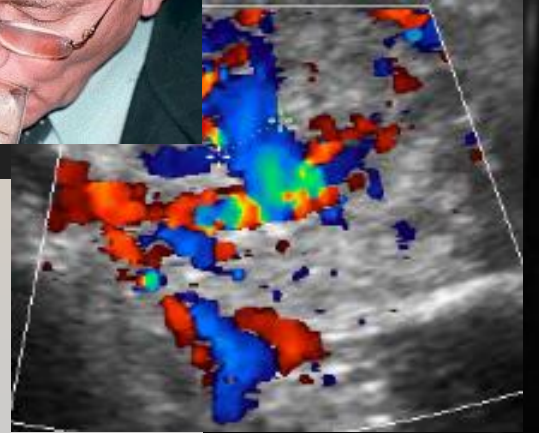
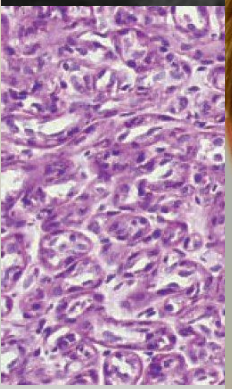
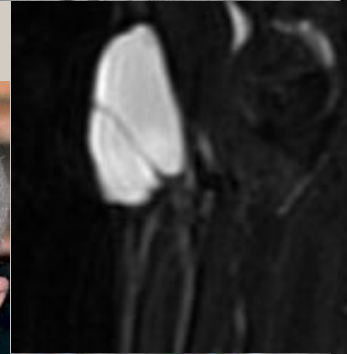
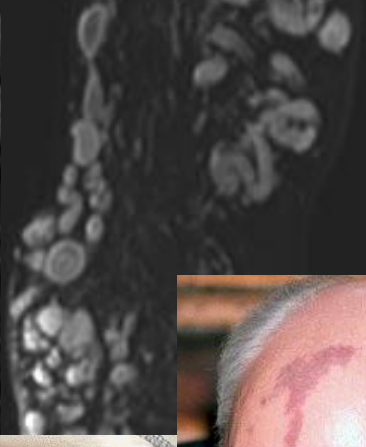
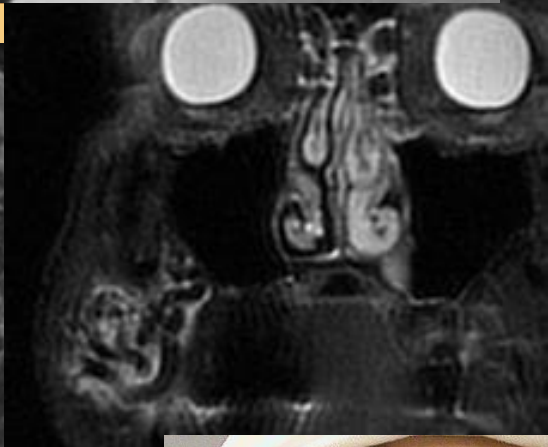
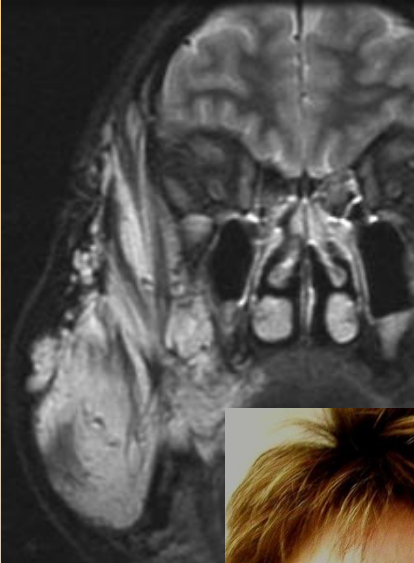
*Kar malformationer*  
*DFIR 2021*

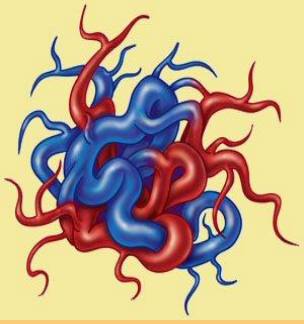
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Susanne Frevert  
Overlæge  
Radiologisk afd. Rigshospitalet



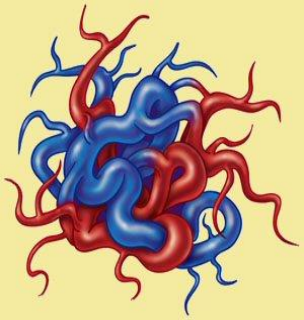
RIGSHOE





# *Klassifikation*

- ★ Mulliken JB, Glowacki J: 1982; *Plast Reconstr Surg*. Hemangiomas and vascular malformations in infants and children. A classification based on endothelial characteristics.



# Vasculær Anomali

Hemangiom

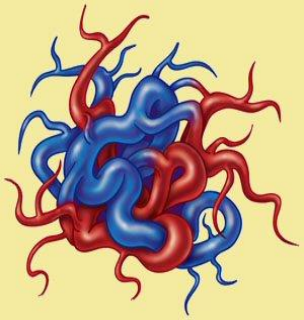
Vasculær malformation

Low flow

High flow

Venøs malformation  
Capillær malformation  
Lymfatisk malformation

Arteriovenøs  
malformation  
Arteriel malformation  
Arteriovenøs fistel



# Vasculær Anomali

Hemangiom

Vasculær malformation

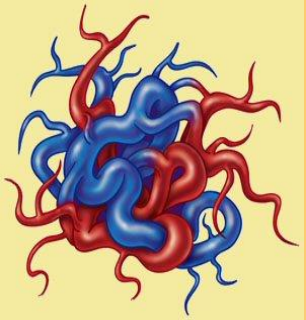
Low flow

High flow

Venøs malformation  
Capillær malformation  
Lymfatisk malformation

Arteriovenøs  
malformation  
Arteriel malformation  
Arteriovenøs fistel





# *Histologi Hemangiom*

## *12% af alle børn*

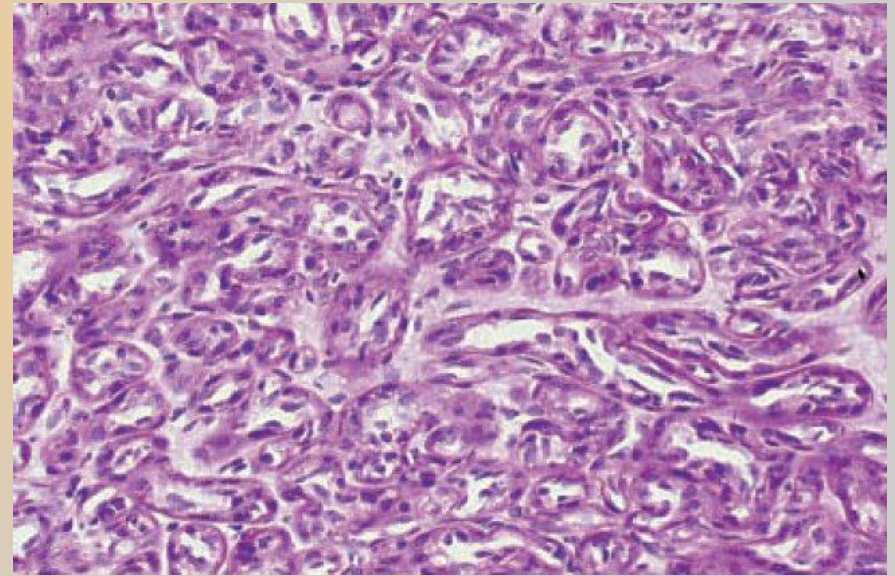
### ★ Proliferation

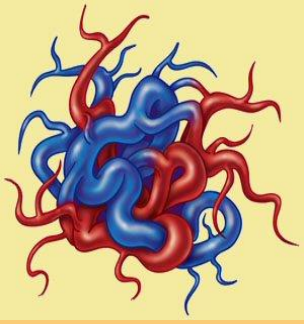
- Plumpe endothelceller med mange mitoser, mast celler og multilaminær basalmembraner

### ★ Involution

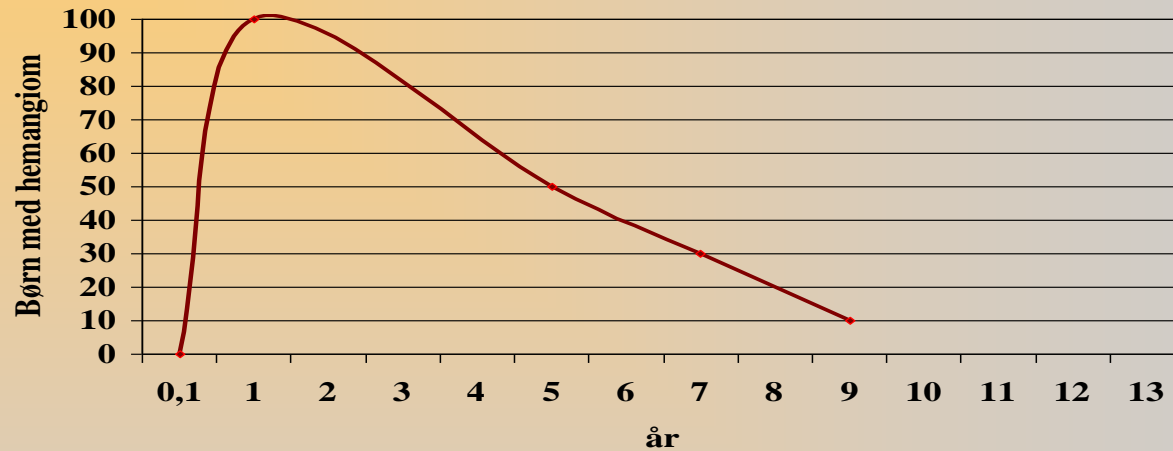
- Flade, inaktive normalt udseende endothel celler i en fibrøs-fedt matrix

### ★ Positive for GLUT-1

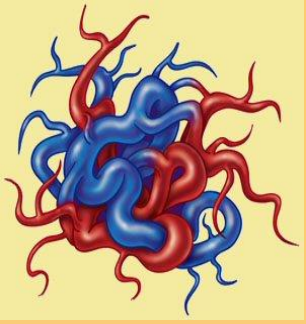




# *Spontan forløb hemangiomer*

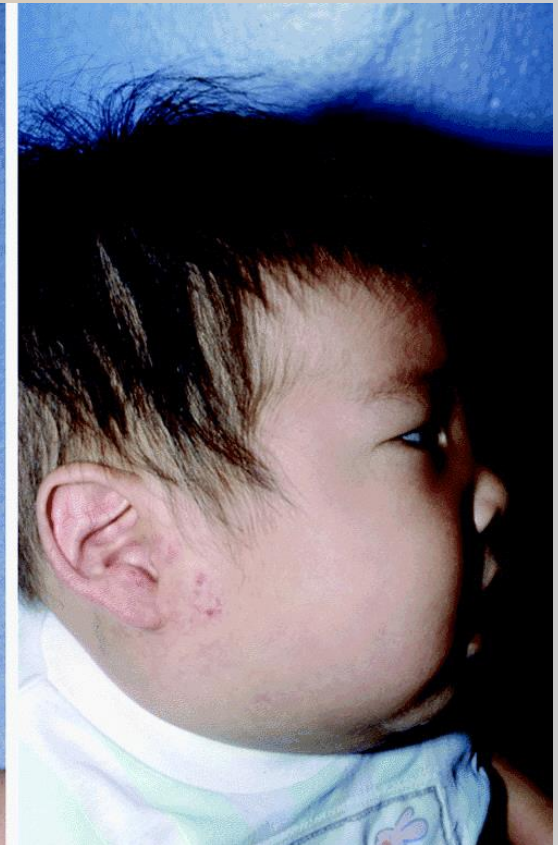


- Kommer hurtigt efter fødslen < 3 mdr.
- Proliferativ fase (vokser mere end barnet)
- Langsom involution

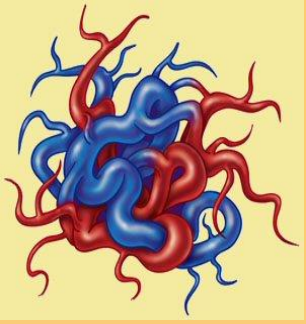


# *Billede af barn med parotis hemangiom*

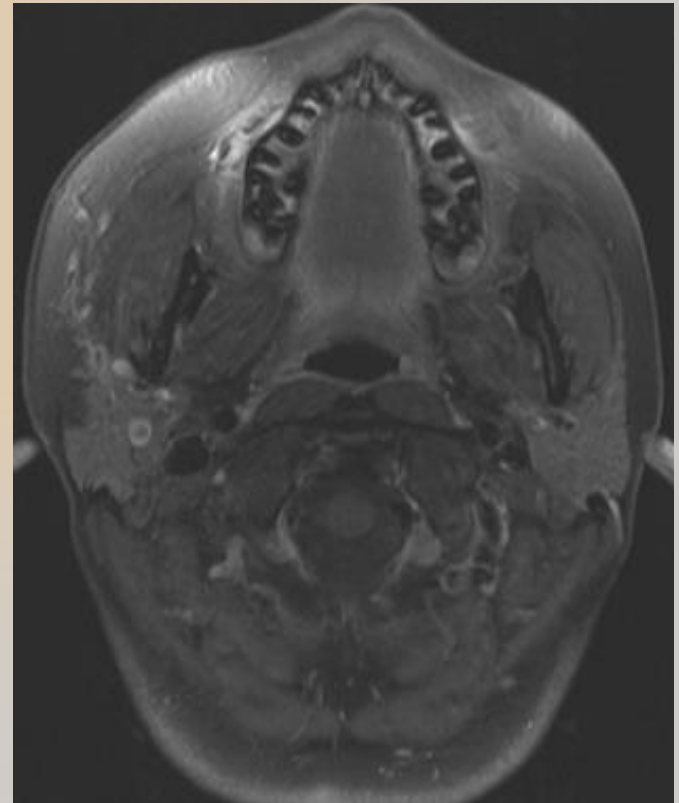
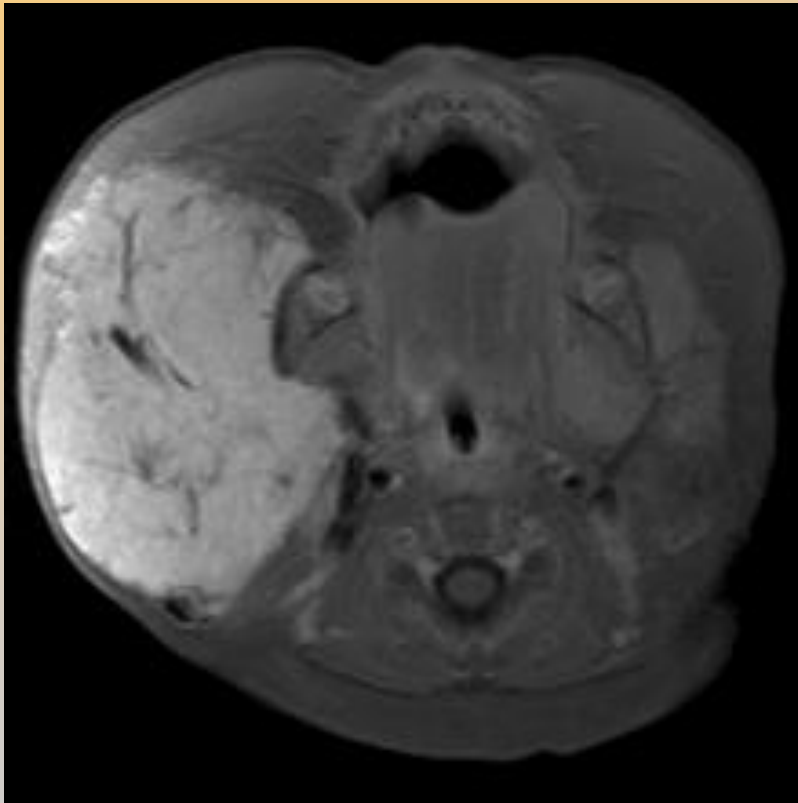
Symptomatiske ptt.  
Behandles med Beta-  
blokker

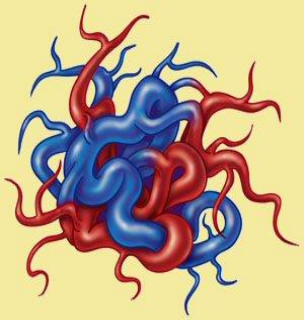






# *MR før og etterinvolution*





Vasculær læsion

Hemangiom

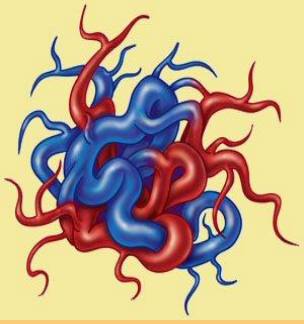
Vasculær malformation

Low flow

High flow

Capillær malformation  
Lymfatisk malformation  
Venøs malformation

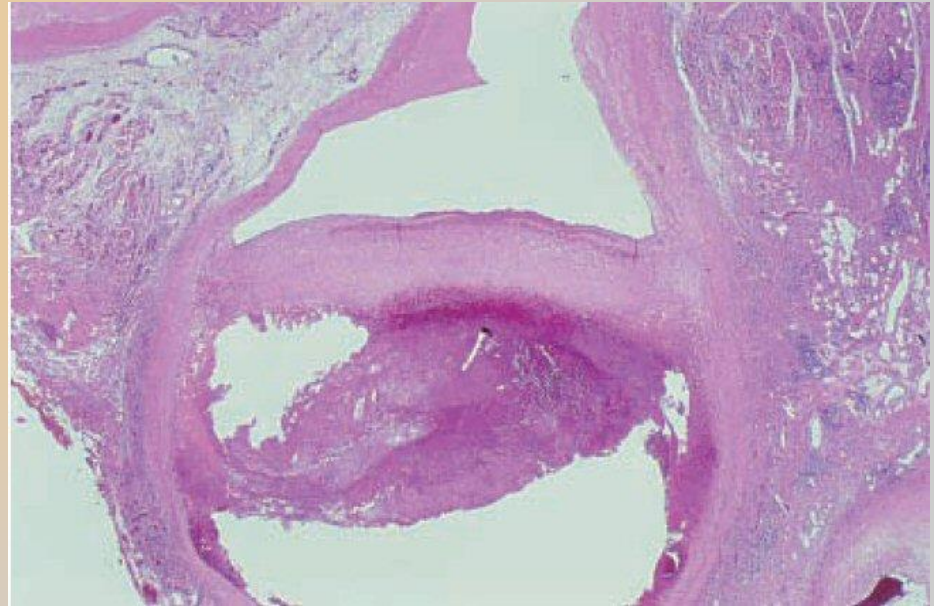
Arteriovenøs  
malformation  
Arteriel malformation  
Arteriovenøs fistel

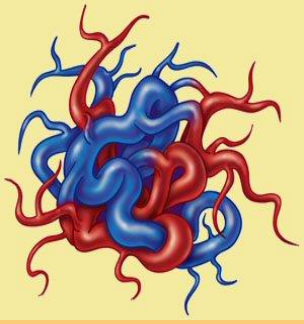


# *Histologi Vasculær malformation*

*Incidens 2-3 %*

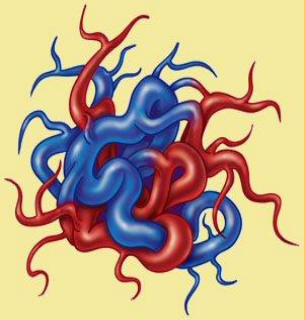
- ★ Flade normale endothelceller og ectatiske kar af en art
- ★ Tilstede ved fødslen
- ★ Vokser sammen med barnet
- ★ Regrediere aldrig





## *Low Flow*

- ★ Kapillær malformation
- ★ Lymfatisk malformation
- ★ Venøs malformation

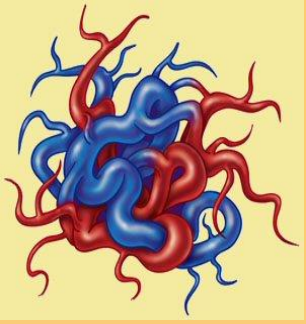


# *Kapillær malformation*

- ★ Kapillær malformation = Port wine stain
- ★ Lyse ved fødslen
- ★ Bliver mørkere med alderen og hypertrofiere
  - Sturge-Weber
- ★ Behandles med laser



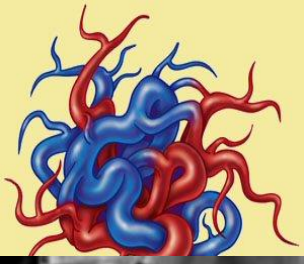




# *Lymfatisk malformation*

- ★ Tidligere cystisk hygrom
- ★ Tilstede ved fødslen
- ★ Sidder oftest på hals eller axil
- ★ Vokser ofte i forbindelse med forkølelse



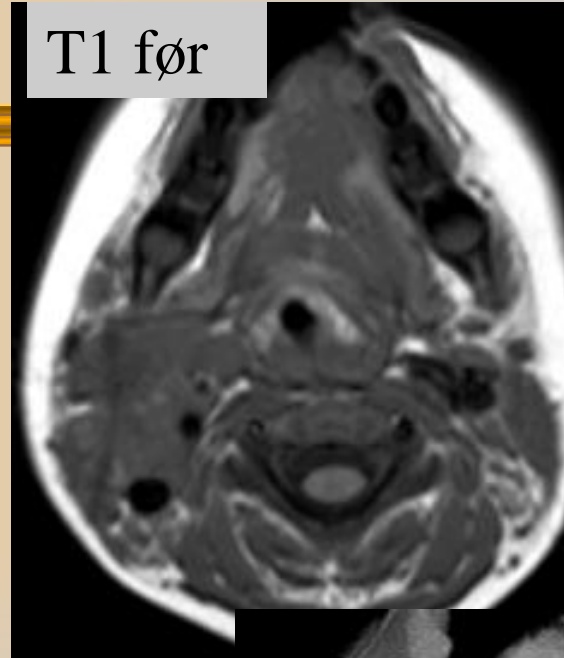


# *Lymfatisk malformation*

STIR



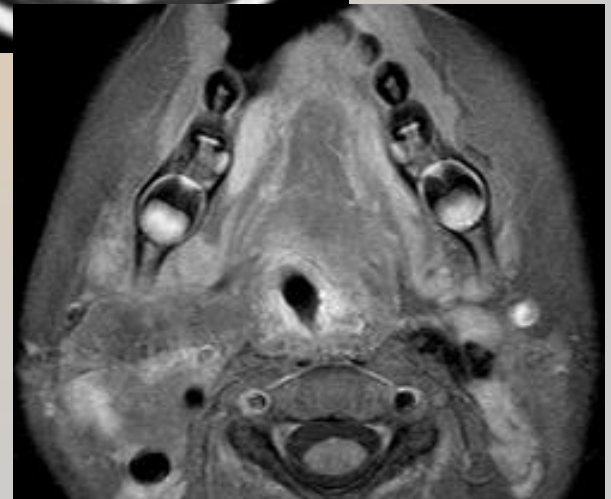
T1 før

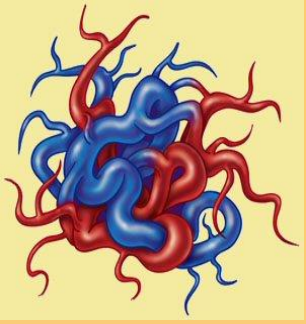


STIR



T1 efter med fatsat





# *Lymfatisk malformation*

## ★ Macrocystisk

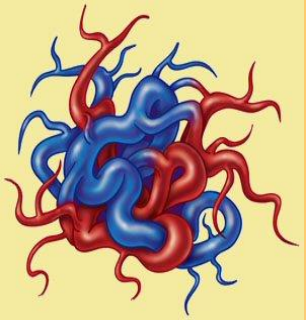
– OK 432

## ★ Microcystisk

– Operation

– Rapamycin





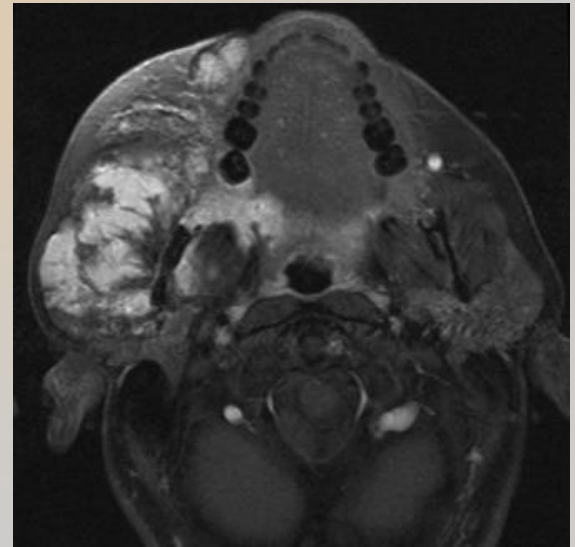
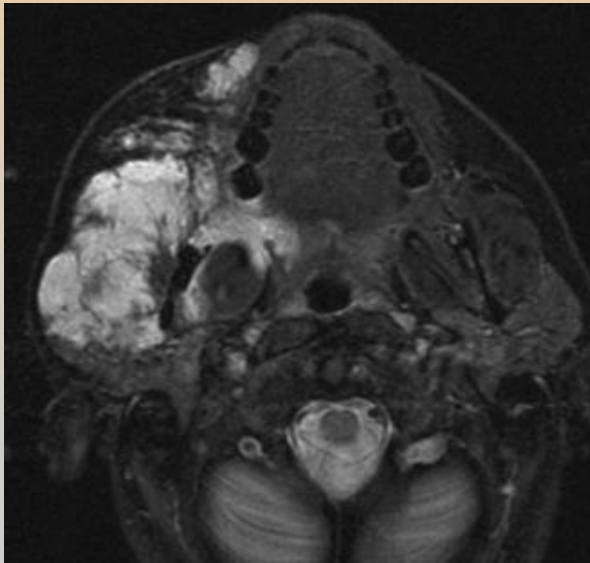
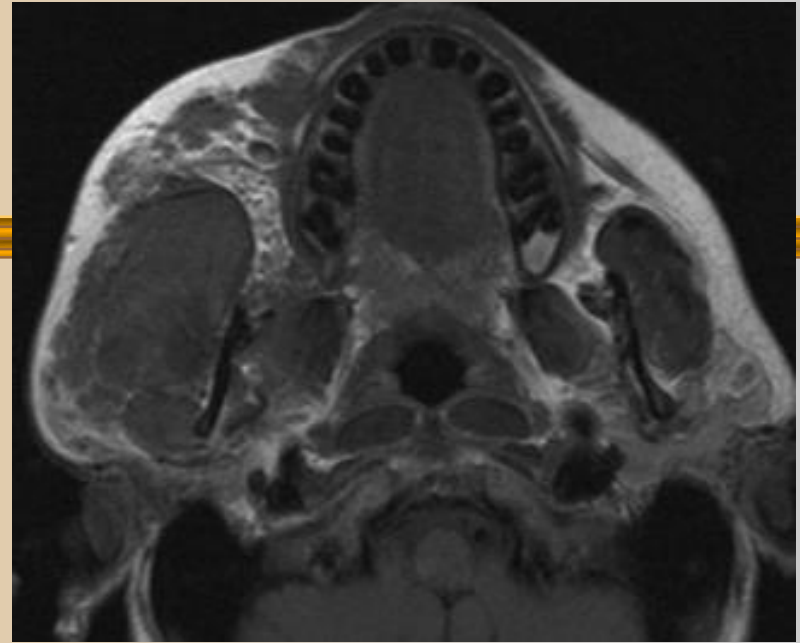
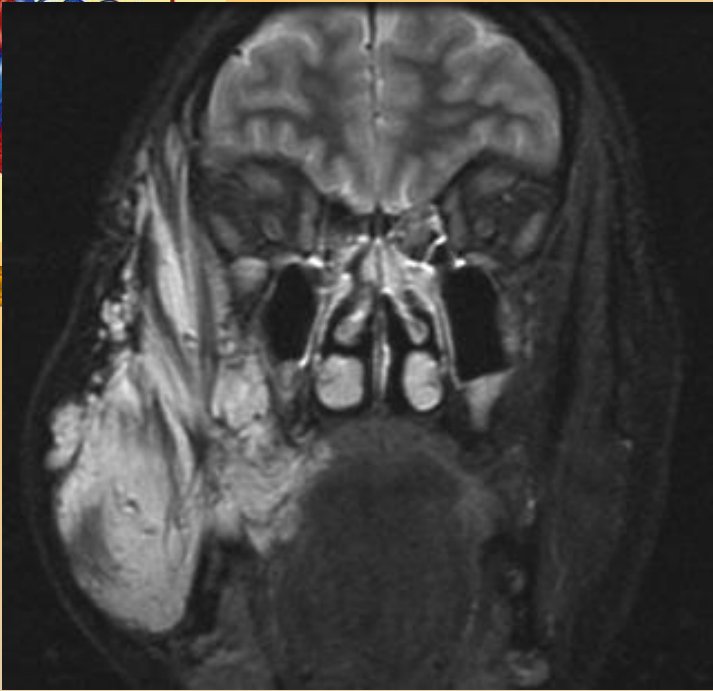
# *Venøse malformationer*

- ★ Phlebolitter på røntgen
- ★ Venøs ectasi
- ★ Afløb i normale/ektatiske vener

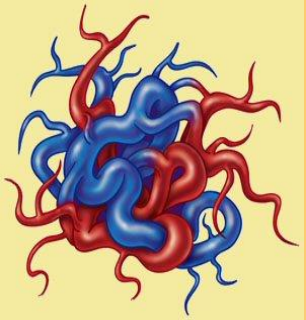




*VM*



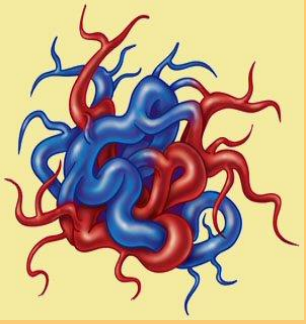




# *Naturhistorie*

- Tilstede ved fødslen
- Vokser langsomt fra fødsel til død
- Særlig symptomatiske ved traumer, hormonale ændringer.
- Lokal thrombose

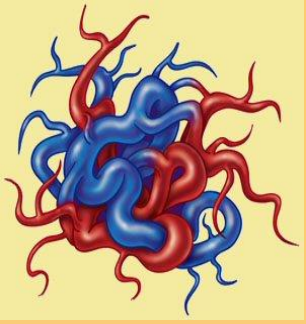




# *Behandling*

- Indikation
- Symptomatisk
  - Smerter
  - Funktions-  
indskrænking
  - Kosmetisk  
gener

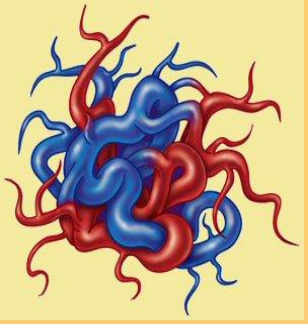




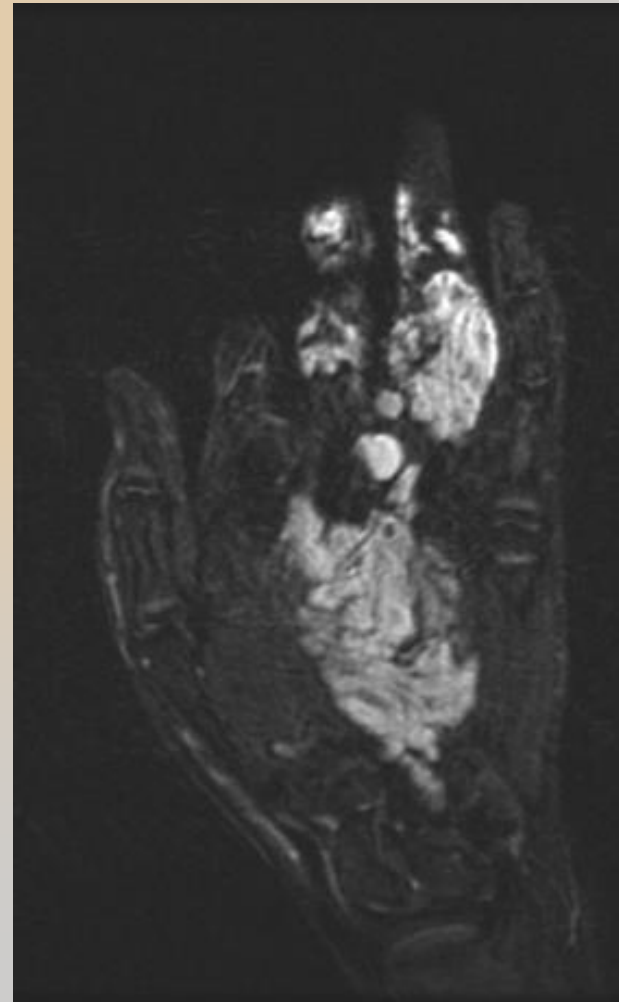
# *Behandling*

- Skleroterapi
  - Fibrovein
  - Polidocanol
  - Alkohol
  - Bleomycin

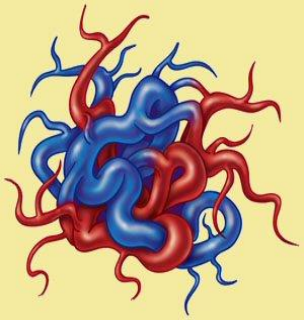




# *Behandling*

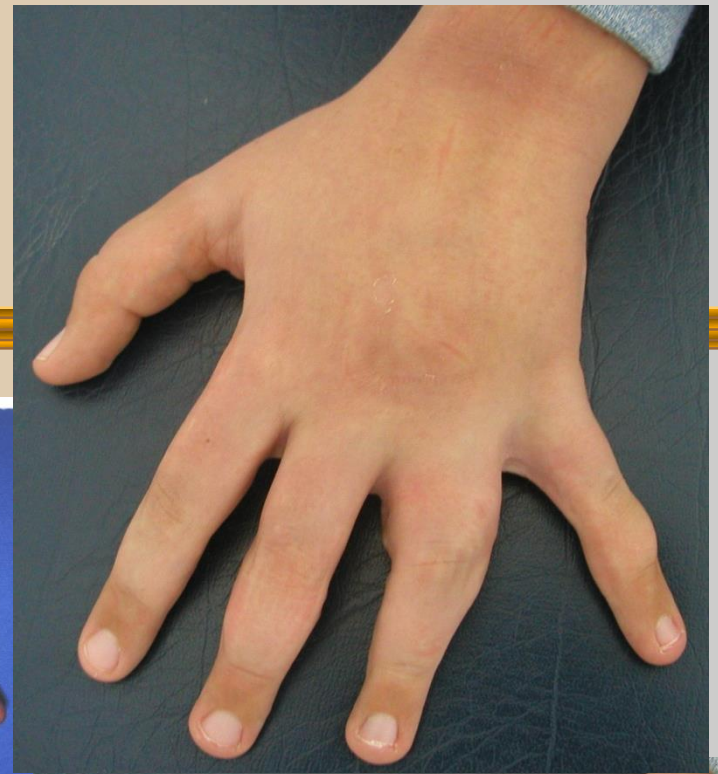




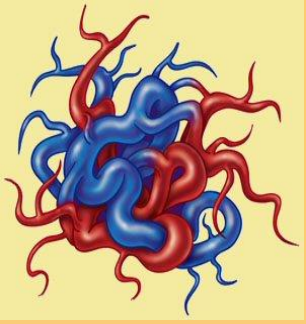


# Resultat

- Skleroterapi ofte mere en en gang.
- Ca. 63 – 75 % oplever symptomfrihed eller betydelig bedring.
- Resten er enten uændret og nogen få forværres.





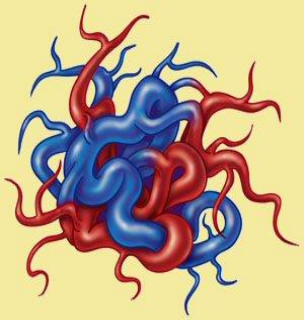


# *Komplikationer*

★ Hyppigst når huden er involveret.

- Sår på huden
- Necrose
- Non target embolisation
- Sædvanligvis opheler det på konservativ behandling med ardannelse

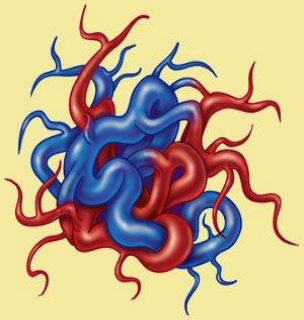




# *AVM: Patofysiologi*

- ★ High-flow Anomali
- ★ Hypertrofierede arterielle feeders
- ★ Central "nidus"  
Bestående af patologiske forbindelser mellem arterier og vener. Med hurtig shunting
- ★ Dilaterede afløbsvener



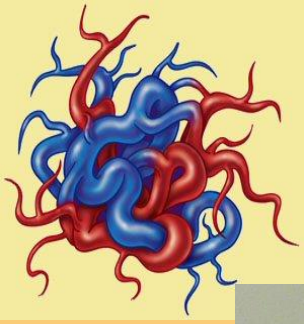


# *Progression fra tidlig barndom til voksen.*



Liu as et al .

Plastic and reconstructive surgery april 2010



# *Schobingers clinical staging*



Stage 1: Quiescence



Stage 2: Expanding, pulsation

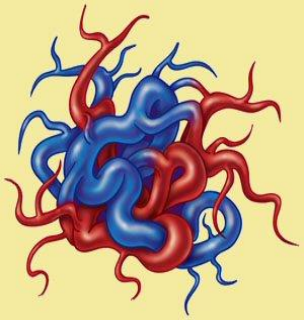


Stage 3: Destruction



Stage 4: Cardiac decompensation





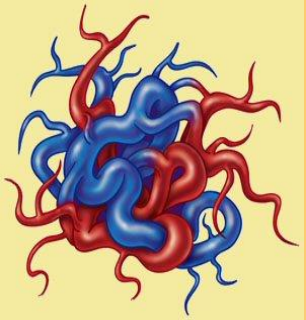
## *Natur historie*

- ★ Alle AVM vil med tiden progredierte til et højere Shobingers stage
- ★ Ca. 43 % før pubertet
- ★ Ca. 83 % før voksenalderen
- ★ Resten progredierte i voksen alder
- ★ Risikoen for hurtig progression er størst i puberteten

Liu as et al .

Plastic and reconstructive surgery april 2010





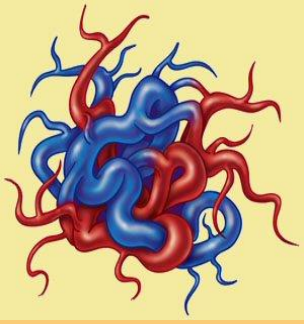
# *Vækst induktion*

## **Traume fx kirurgi**

- ★ 57 årig mand
- ★ Lille protuberance reseceret fra ankel
- ★ Ingen heling
- ★ Nu smerter og blødning

## **2 mdr. Efter kirurgi**

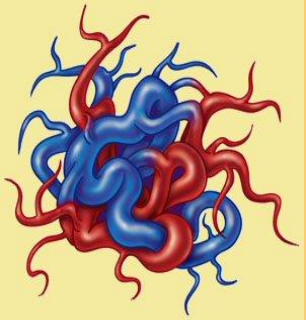




# *Efter flere emboliserings forsøg*

- ★ progression
- ★ Slut resultat:  
Amputation





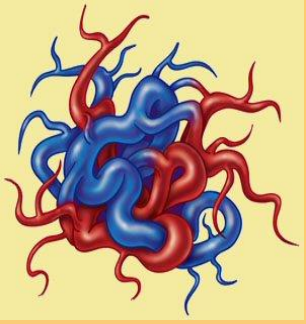
# *Progression under graviditet*

**After 1. pregnancy**



**After 2. pregnancy**





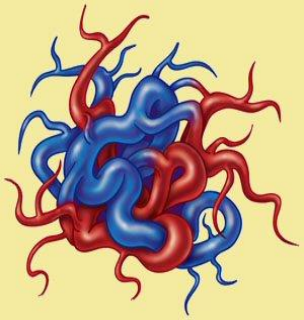
# *Progression under graviditet*

**MR after 1. pregnancy**



**MR after 2. pregnancy**



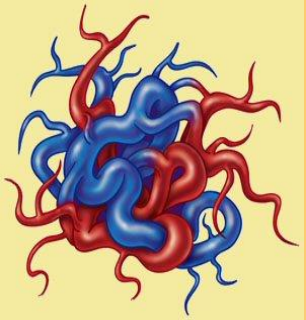


# *Udredning: Diagnose og planlægning af behandling*

## **Diagnosis**

- ★ Klinisk undersøgelse
- ★ Ultralyd
- ★ MR scanning
- ★ Dynamisk MR angiography can være værdifuld
- ★ CT angiografi er især værdifuld i thorax og abdomen



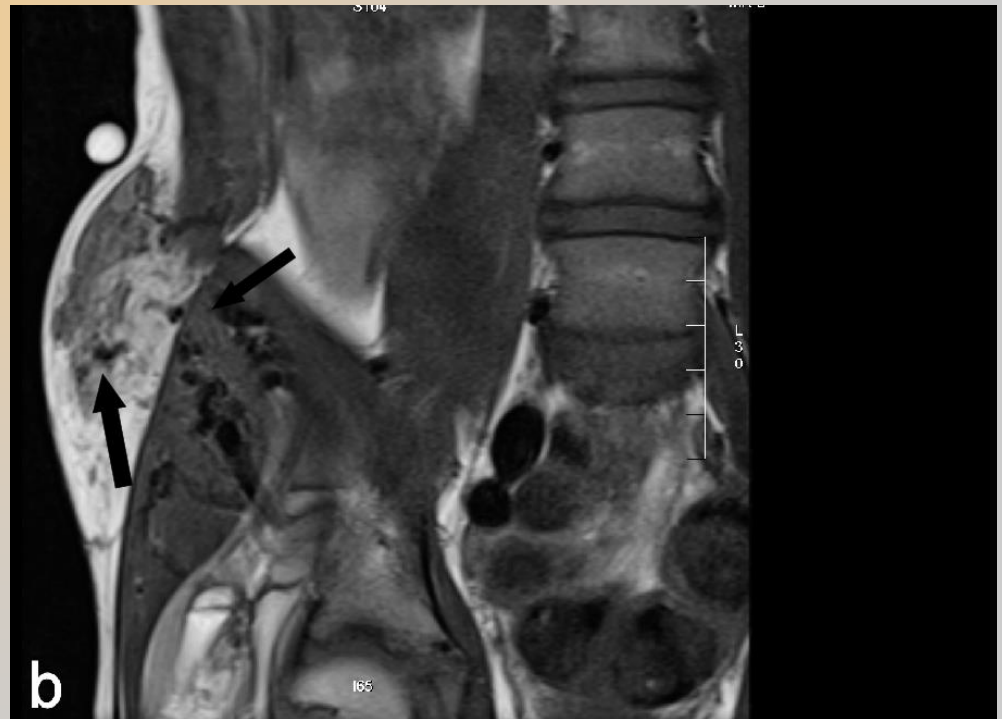


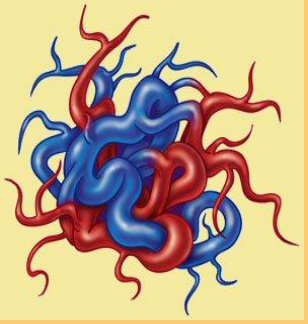
# Imaging

## MR scanning

- ★ T1 vægtet
- ★ Flow void i high flow kar er sorte
- ★ God til vurdering af lokalisation og involvering af vitale strukturer.

## T1 weighted





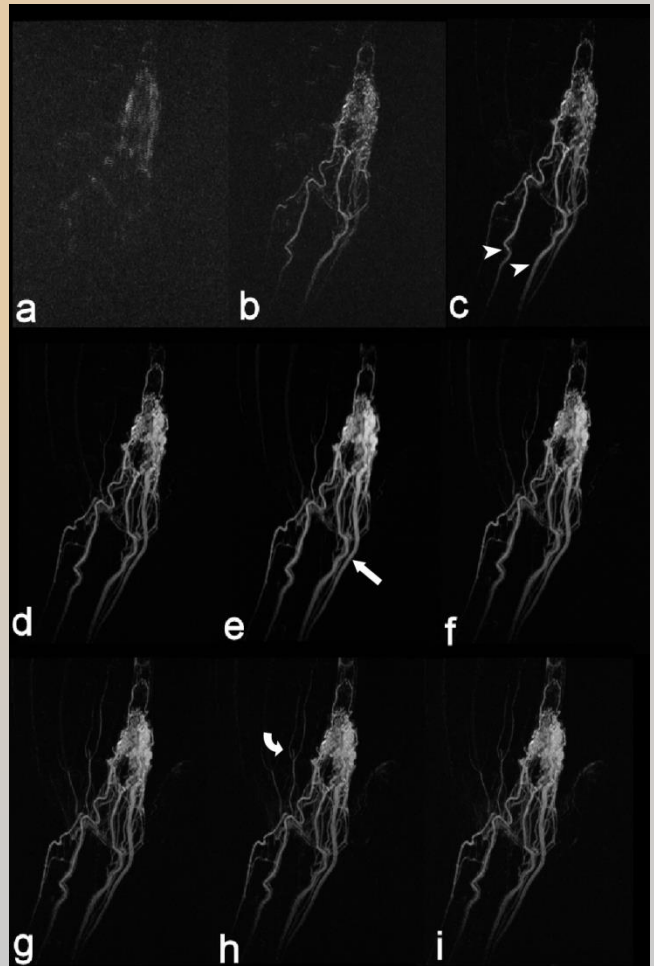
# MR angiografi

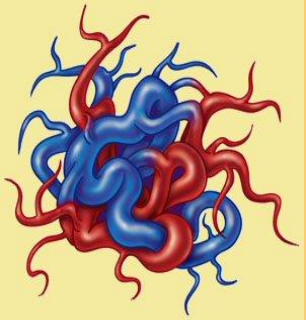
## MR scanning



angiografi  
men  
A

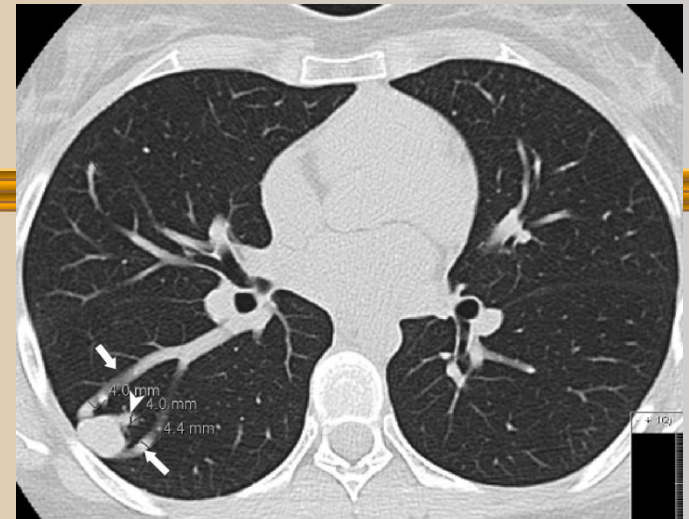
## Dynamic MR angiografi



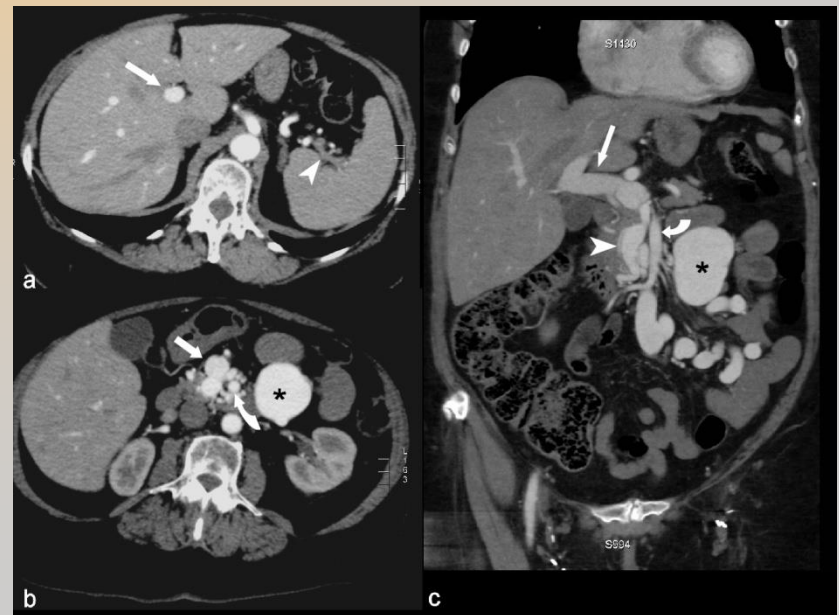


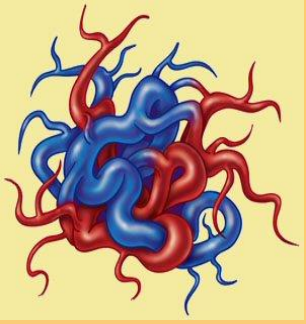
# Billeddiagnostik

## CT scanning



- ★ Hurtig
- ★ Bedst til AVM i lunger og abdomen.

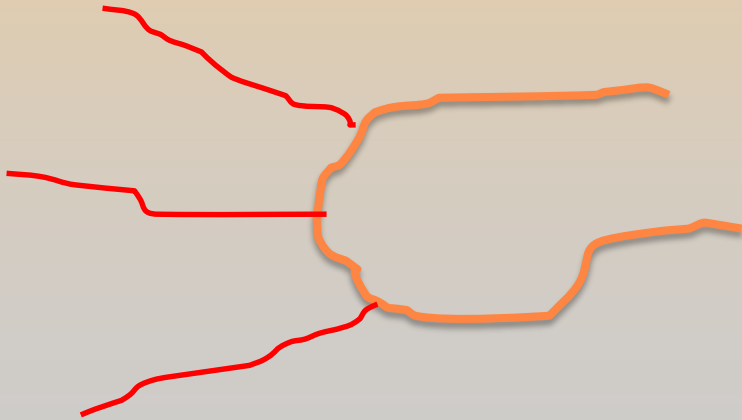




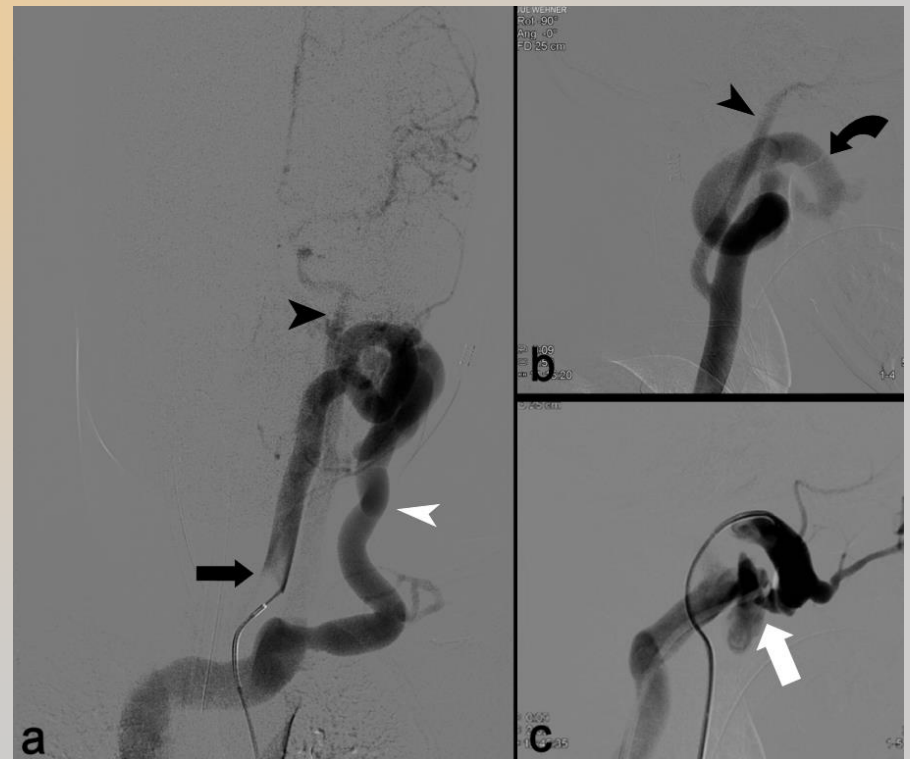
# *Imaging angio-architecture*

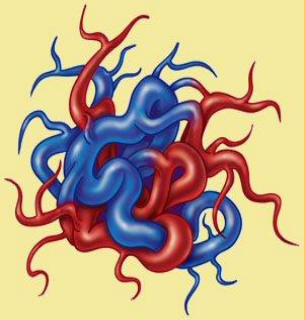
## Digital subtraction angiography

- 1 dominating vein
- Up to 3 feeding arteries



## Arterio- venous malformation (fistula)

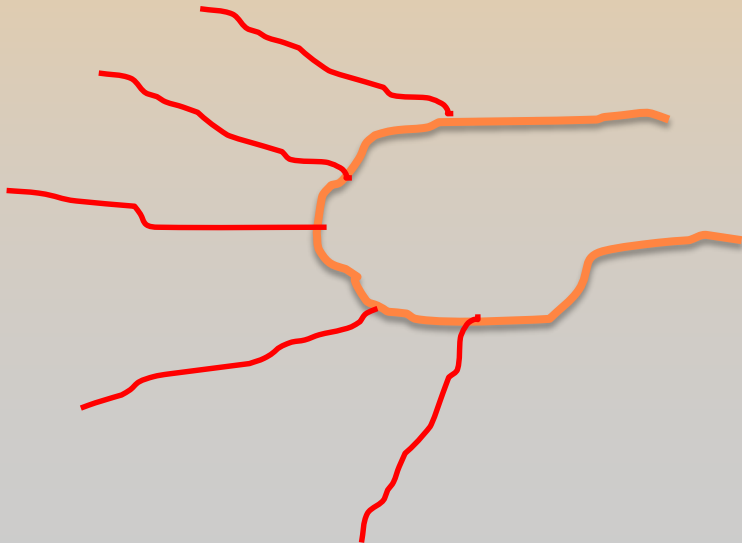




# Imaging angio-architecture

## Digital subtraction angiography

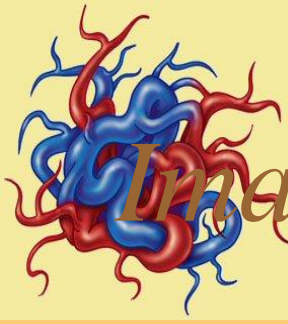
- 1 dominating vein
- More than 3 feeding arteries



## Arterio- venous malformation







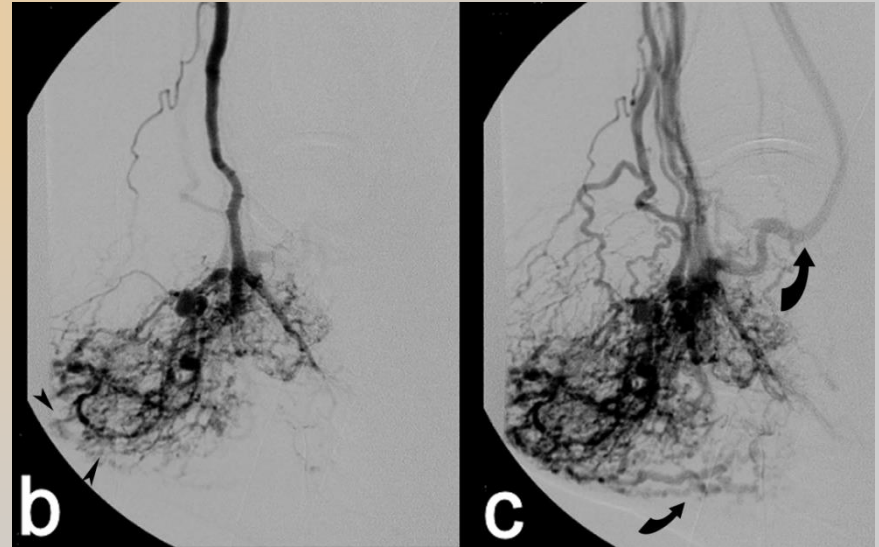
# *Imaging angio-architecture*

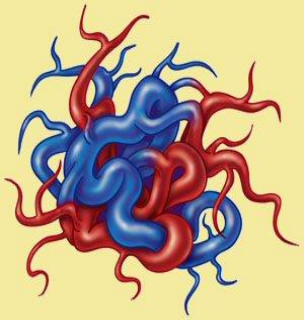
## Digital subtraction angiography

- **Multiple draining veins**
- **Multiple feeding arteries**



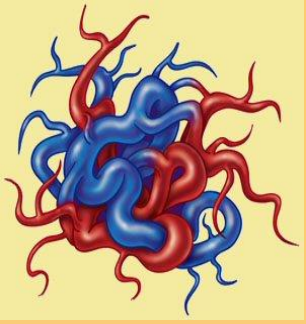
## Arterio- venolous malformation





# *Behandling*

- ★ Ekspekterende
- ★ Aktiv behandlig
  - Embolisering
  - Resection ofte efter preoperativ embolisering
  - Medicinsk behandling
    - Targetet medical treatment.

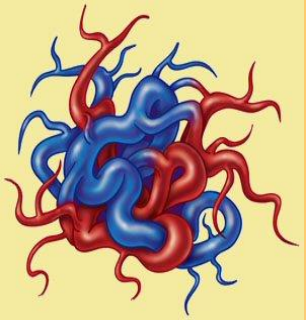


# *Embolic agents*

## **Flydende agents**

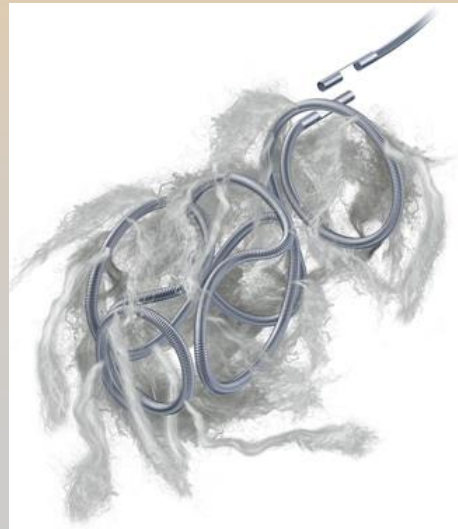
- ★ Ethanol
- ★ Histoacryl
- ★ Onyx





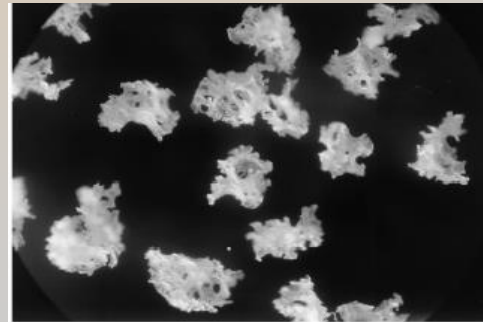
# *Embolic agents*

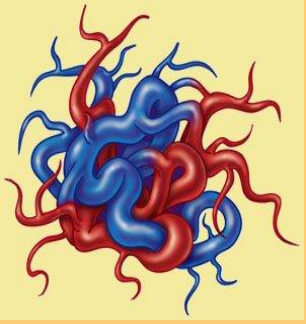
## Examples



## Mechanical occlusion

- ★ Coils, plugs and particles
- ★ Many designs and sizes
  - Preoperative embolisation
  - “Simple angio-architecture”

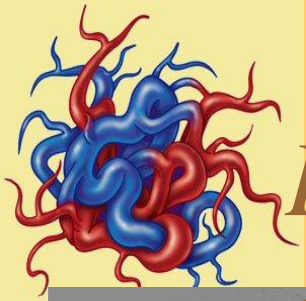




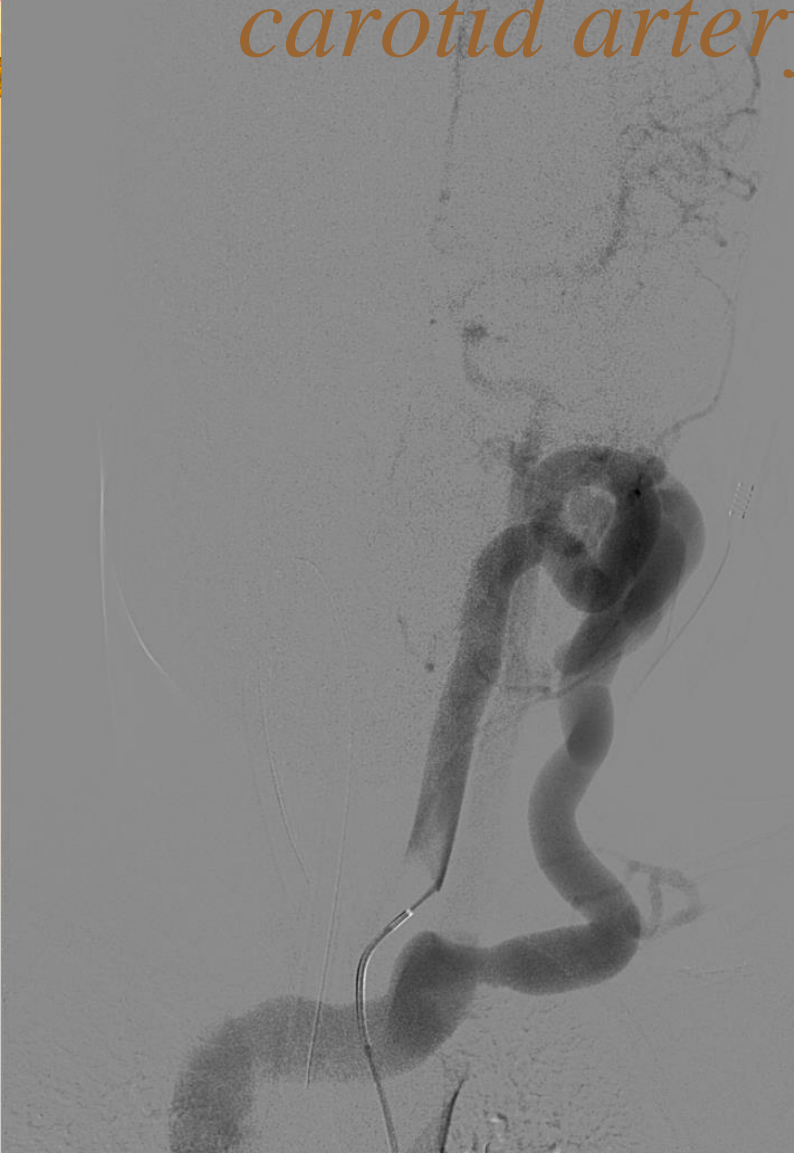
# *9 years old boy*







# *Fistula between the external carotid artery and the jugular vein*

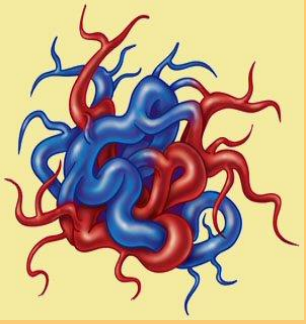


D 25 cm

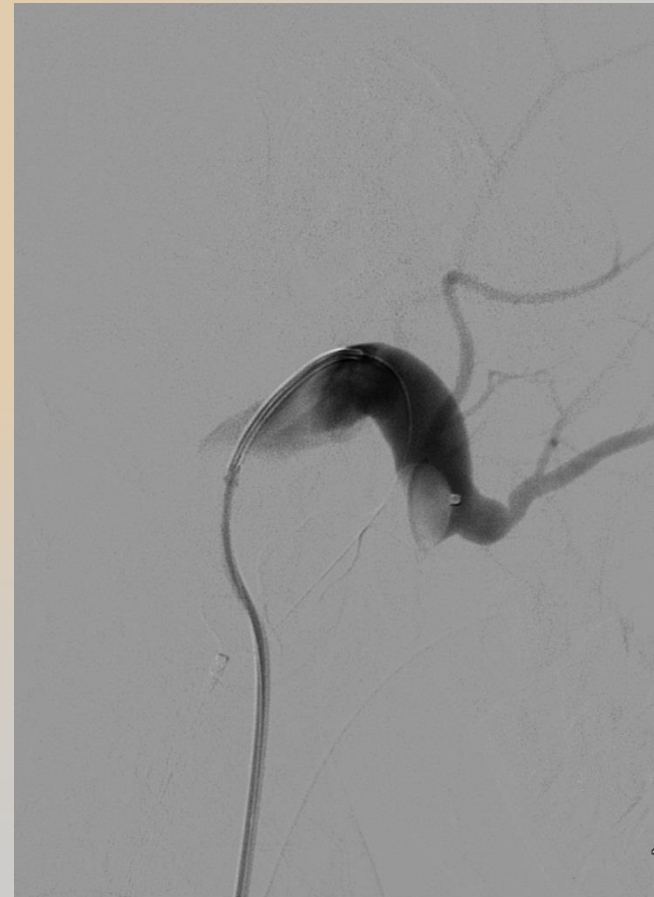
P 0:09  
D 2:09  
M 12:45:35

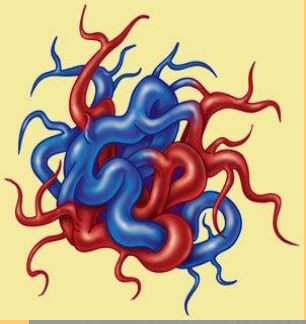


2  
1-5



*The plug was placed from the vein*

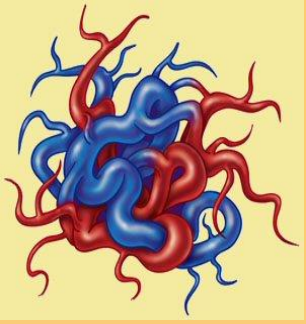




*End result*

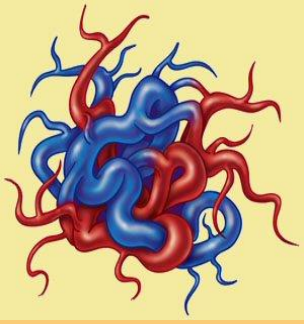




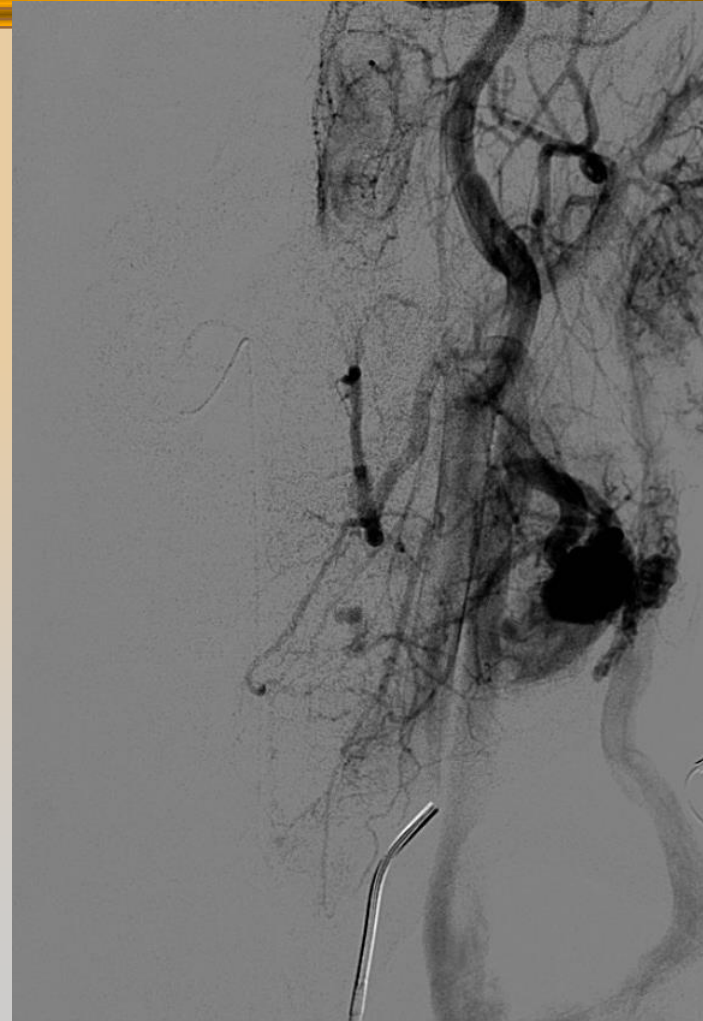


*12 years old girl*

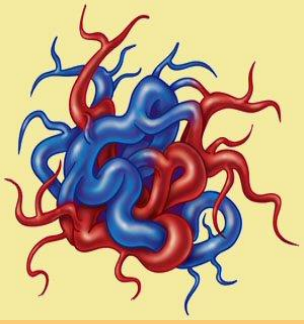




# *AVM in external carotid artery territory*

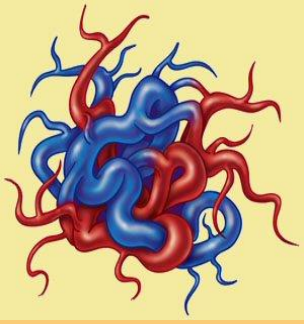




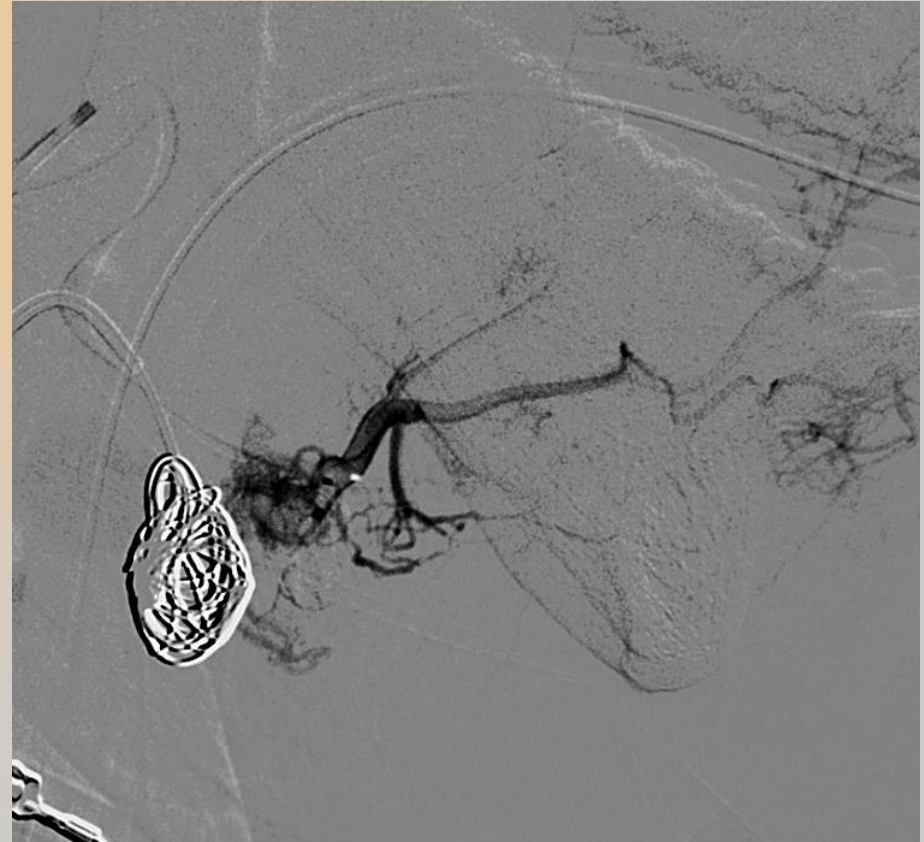
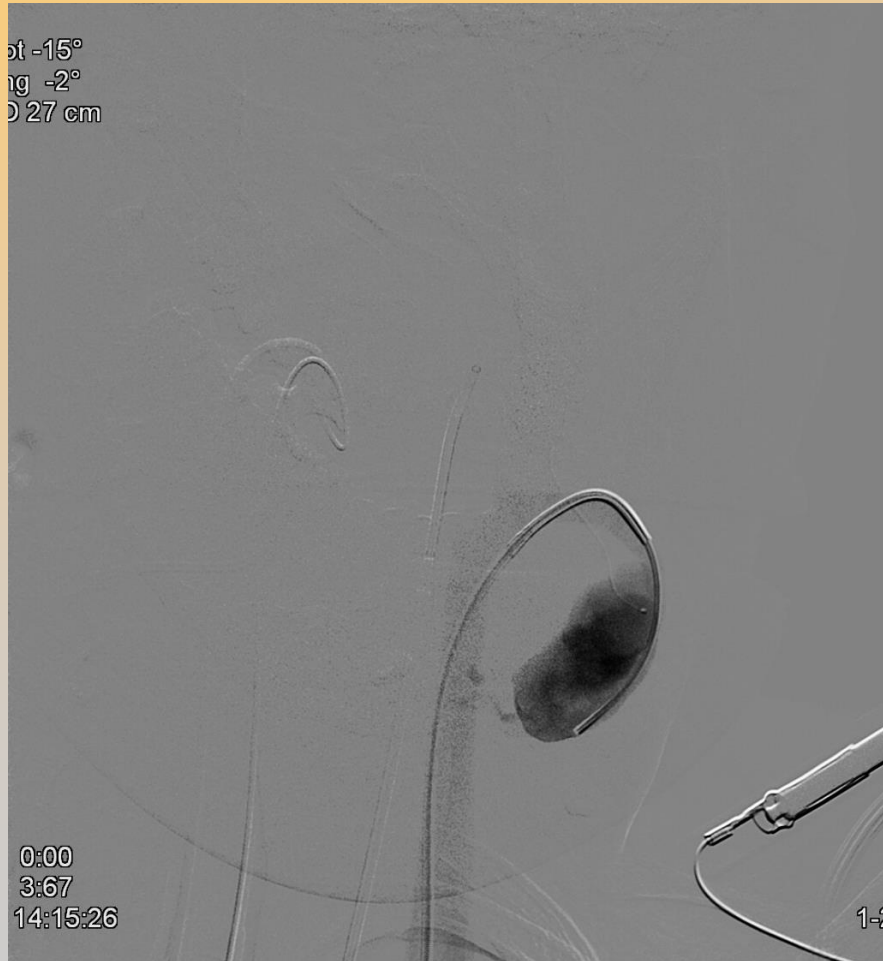


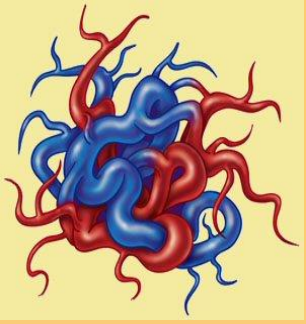
# *Arteriolo-venous malformation*



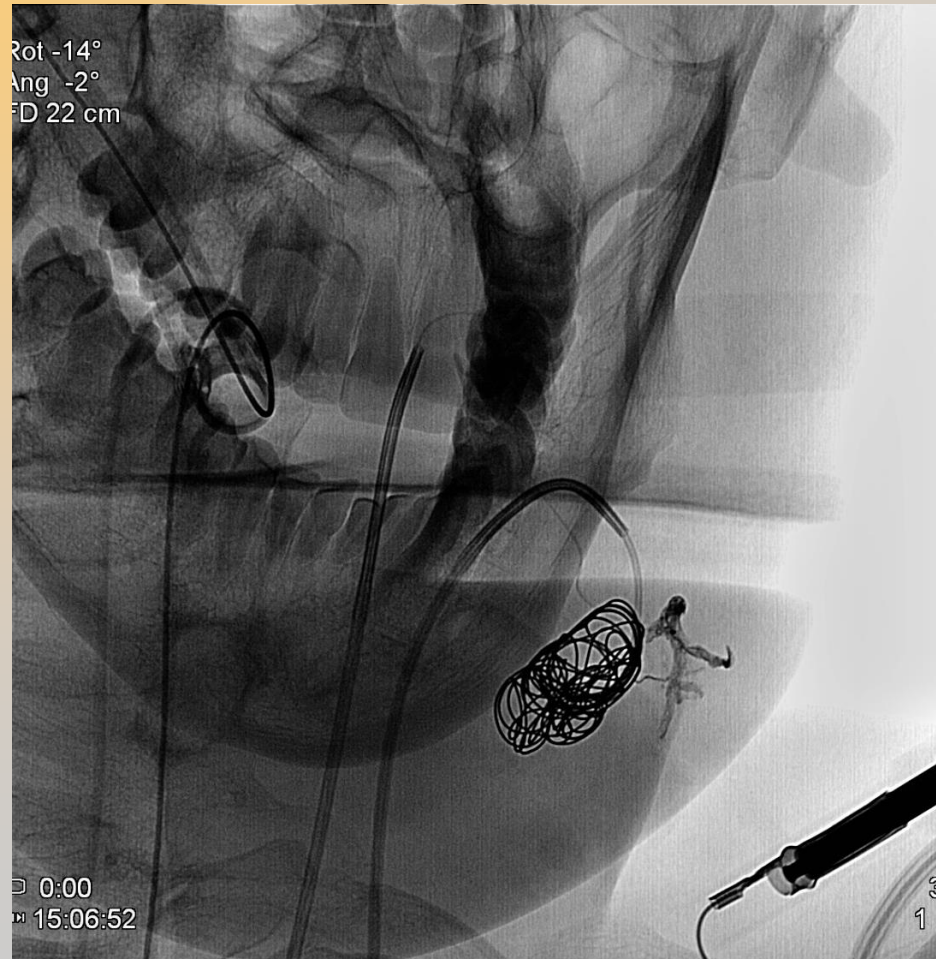


# *Coil embolisation of the dominant vein*

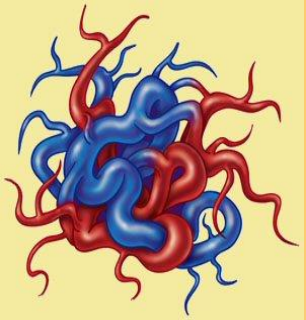




# *Onyx from the artery*

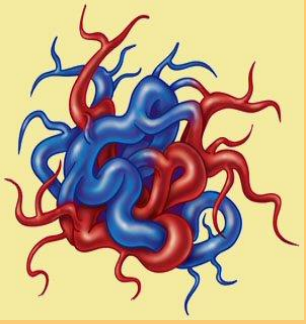






# Completion arteriography

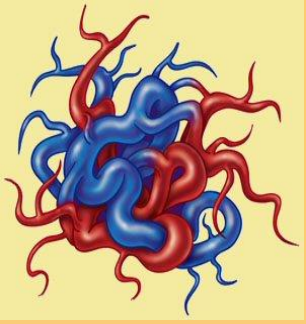




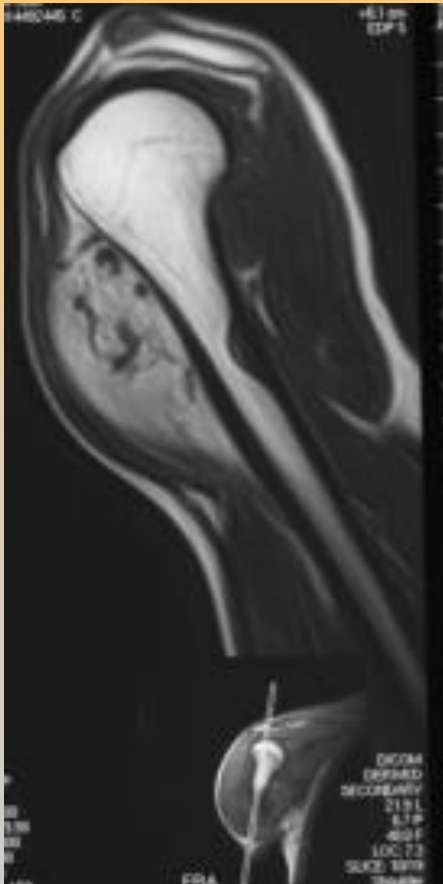
# *Arteriography 3 month post embolisation*

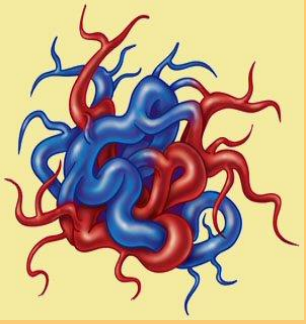




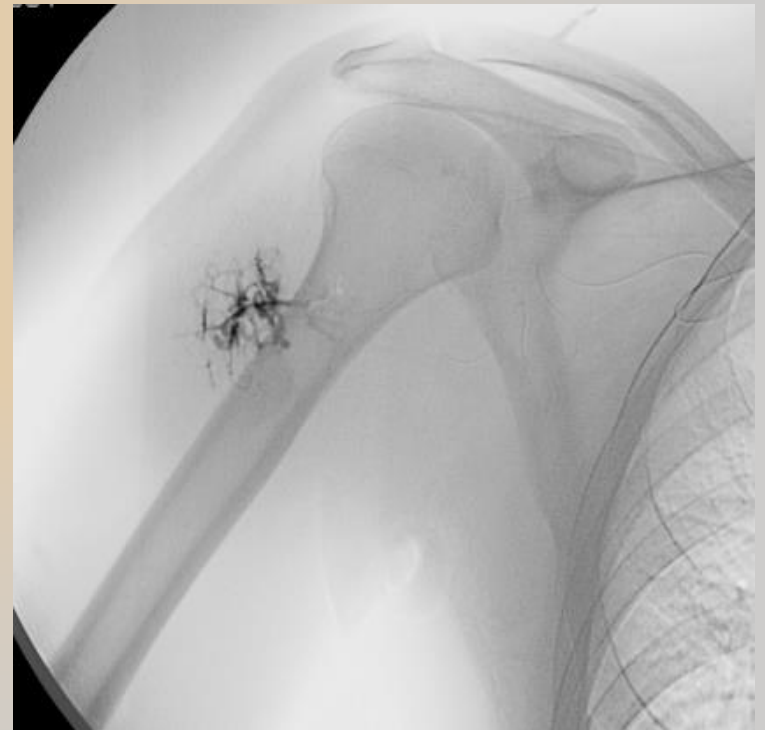


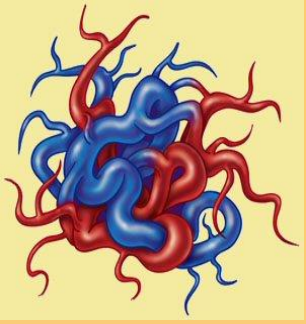
# *Shoulder, resectable AVM*





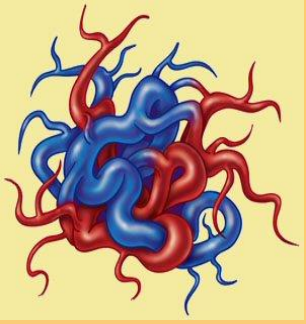
# *Pre operative embolisation using Onyx*





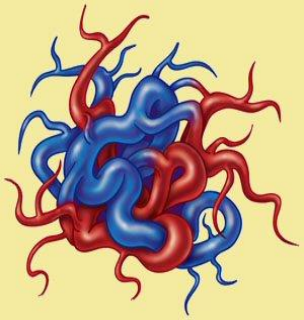
*Super selectively with micro catheter*





# *Embolisation using Onyx*



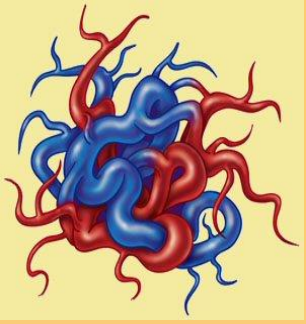


# *Slut arteriografi*

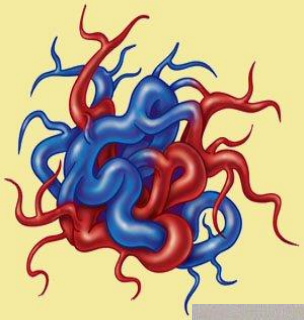
- ★ Nye kollateraler dukker op.
- ★ Læsionen blev resekeret med godt resultat.





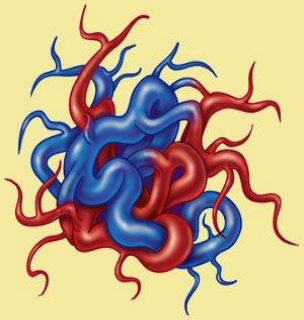


- ★ 25 årig kvinde
- ★ Stor AVM i venstre lyske og flanke
- ★ Smerter de sidste 5 år tiltaget efter fødsel.

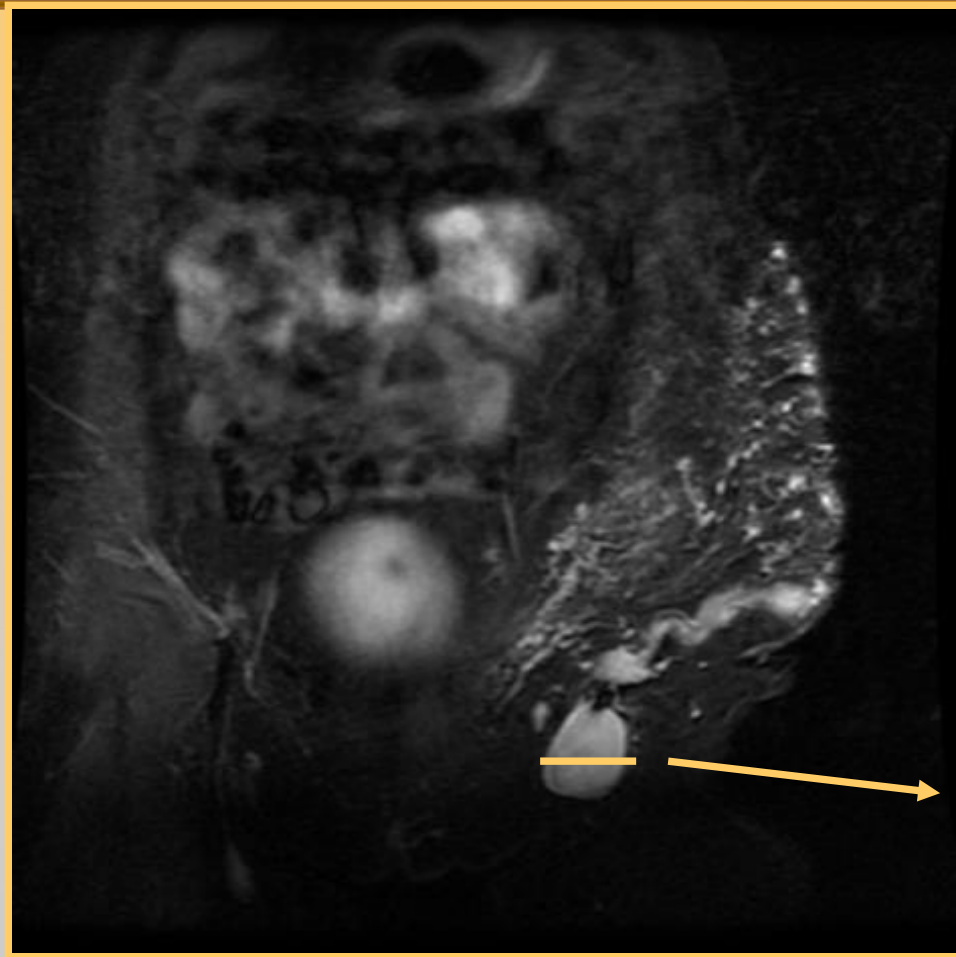


# *Klinisk foto*

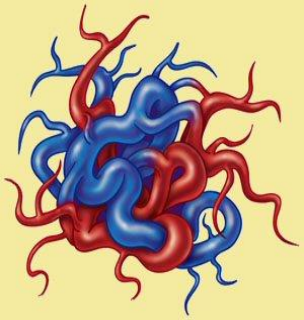




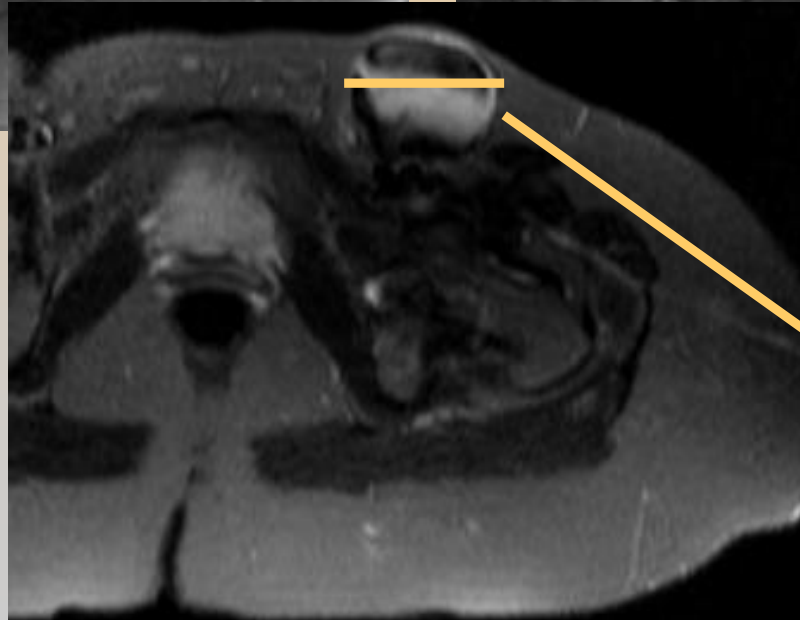
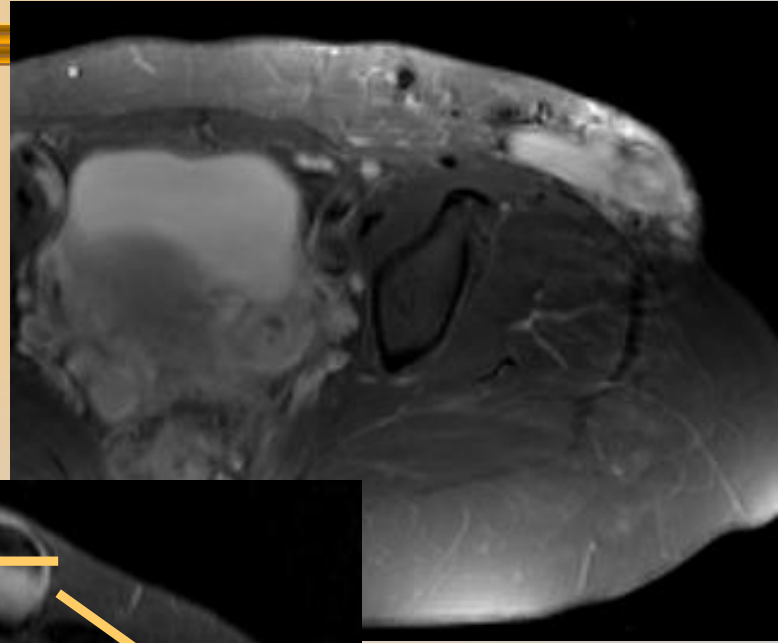
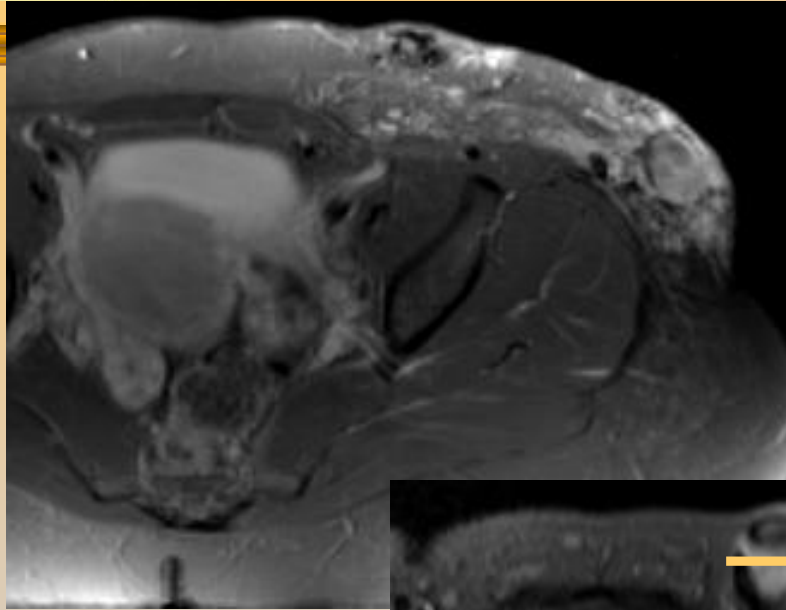
*MR 2001*



2,5 cm

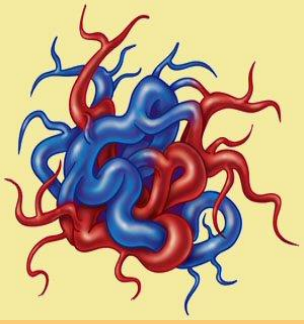


# *MR ax T1 post CM 2004*



5 cm

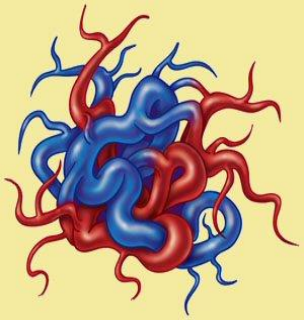




# *Arteriografi*

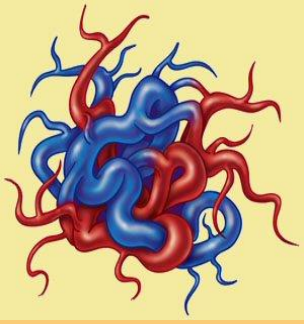






# *A. Iliaca Interna*



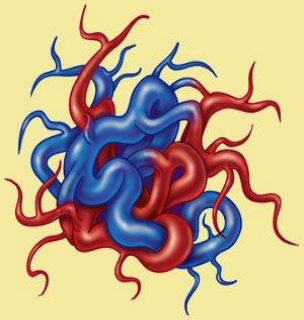


# *“Natur historie” Efter behandling*

- ★ Resection hvis muligt giver den bedste chance for long term control
- ★ Overall risiko for recidiv er høj. Op til 93%

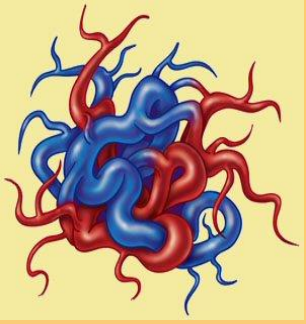
Liu as et al .

Plastic and reconstructive surgery april 2010



# *Syndromer associeret med vasculære læsioner*

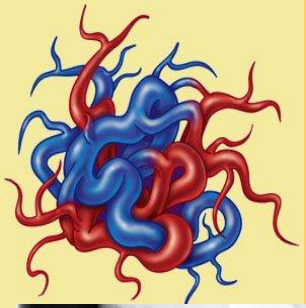
- ★ Osler-Weber-Rendu syndrom
- ★ Blue rubber bleb syndrom
- ★ Parkes-Weber syndrom
- ★ Bannayan's syndrom
- ★ Sturge-Weber-Krabbe syndrom
- ★ Klippel-Trenaunay syndrom
- ★ Servelle-Martorell syndrom
- ★ Maffucci's syndrom



# *Klippel-Trenaunay*

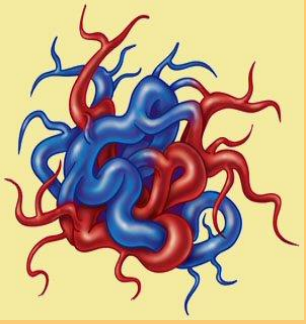




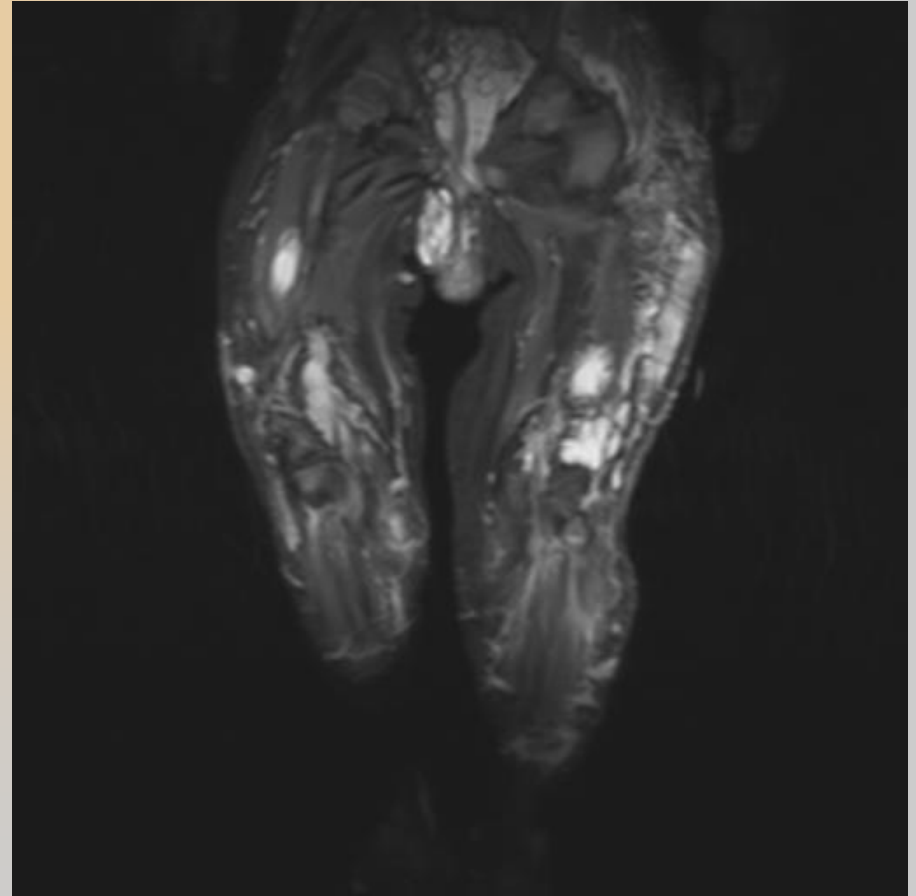
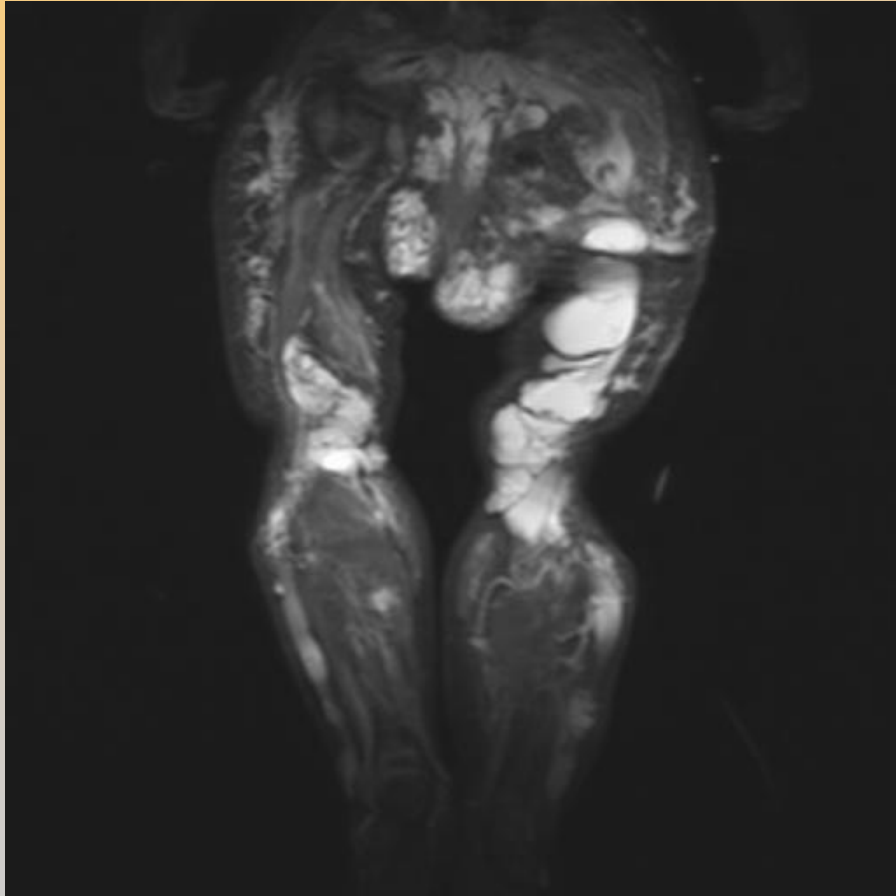


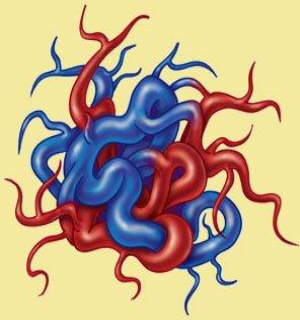
# *Klippel-Trenaunay*





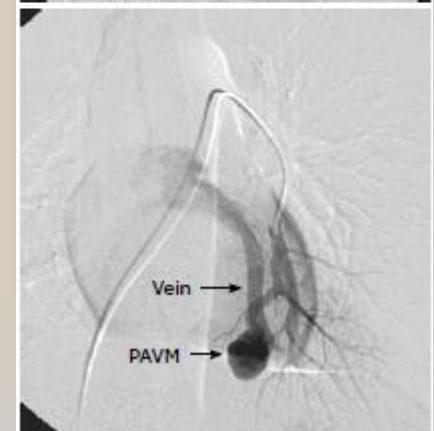
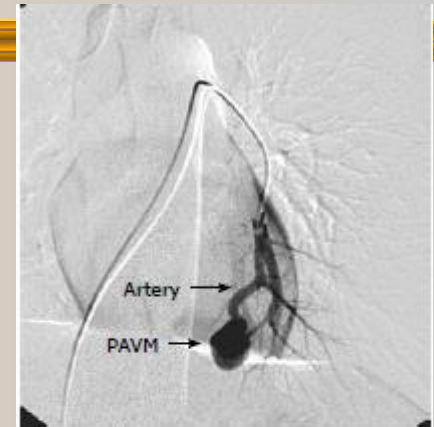
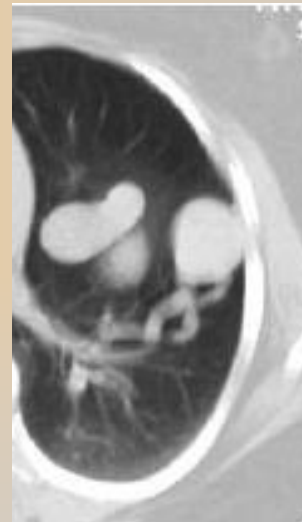
# *Klippel-Trenaunay*

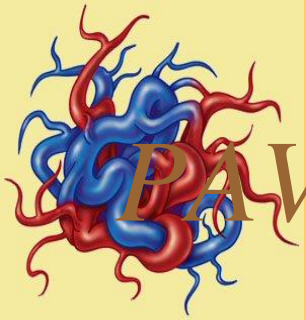




# *Pulmonary Arterio-venous malformation*

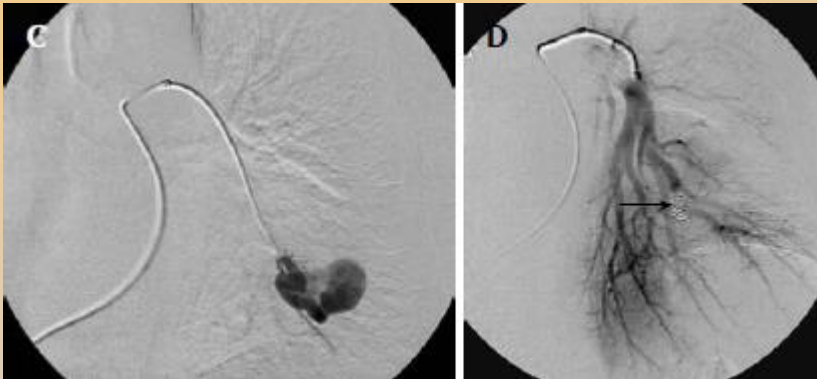
- ★ 30 % of Osler patients
- ★ Symptoms
  - Hypoxia
  - Paradoxical emboli to brain
  - Brain abscess



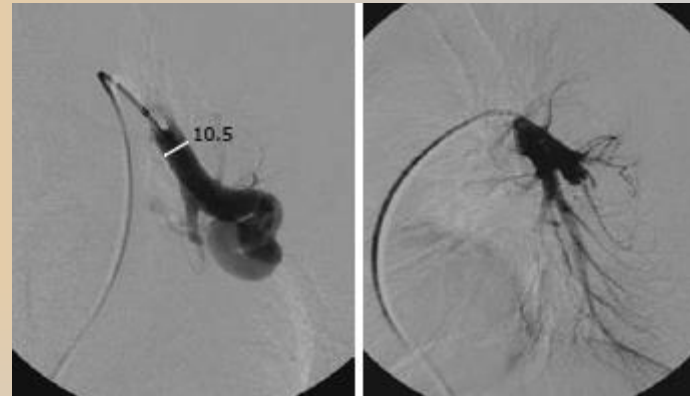


# *PAVM embolisation*

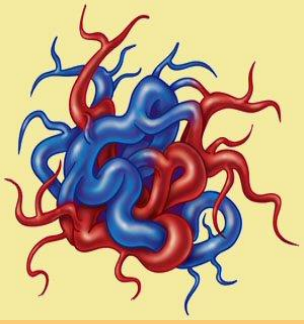
## **Embolised using coils**



## **Embolised using plug**







# *NSVA meeting*

- ★ Nordic Society for Vascular Anomalies
- ★ København 8-9 November 2021
- ★ Rigshospitalet.
  
- ★ Registrering
- ★ Email: [SFRE0008@regionh.dk](mailto:SFRE0008@regionh.dk)