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Vascular Anomalies

Anthony Penington MBBS, FRACS



Vascular Birthmarks

- 'Strawberry haemangioma'
- 'Cavernous haemangioma'
- 'Port-wine stain'
- 'Angioma cavernosum'
- 'Angiomatosis'



Hemangiomas and Vascular Malformations in Infants and Children: A Classification Based on Endothelial Characteristics

John B. Mulliken, M.D., and Julie Glowacki, Ph.D.
Boston, Mass.

- Plastic and Reconstructive Surgery, 1982



Vascular Birthmarks

- Haemangiomas
- Malformations



Vascular Birthmarks

- **Haemangiomas**

- Appear at birth or soon after



- **Malformations**

- Usually present at birth but can appear any time



Vascular Birthmarks

- **Haemangiomas**

- Appear at birth or soon after
- Grow rapidly
-

- **Malformations**

- Usually present at birth but can appear any time
- Grow with the child



Vascular Birthmarks

■ Haemangiomas

- Appear at birth or soon after
- Grow rapidly
- Stop growing and involute
-

■ Malformations

- Usually present at birth but can appear any time
- Grow with the child
- Do not involute



Vascular Birthmarks

- **Haemangiomas**
- **Malformations**
- Appear at birth or soon after
- Usually present at birth but can appear any time
- Grow rapidly
- Grow with the child
- Stop growing and involute
- Do not involute
- Pronounced endothelial proliferation
- Stable endothelium
-



Nomenclature

- 'Strawberry haemangioma'
- 'Cavernous haemangioma'
- 'Port-wine stain'
- 'Angioma cavernosum'
- 'Angiomatosis'

- *Naevus maternus*



Cavernous haemangioma



- *it is common for a mother to mark her unborn child with fruit, even “after the infant is entirely formed by the strength of her imagination only”*
- Maubray, 1724



The STRENGTH of
IMAGINATION
IN
Pregnant Women

EXAMIN'D:

And the OPINION that
MARKS and DEFORMITIES

IN CHILDREN arise from thence,
Demonstrated to be a VULGAR ERROR.

By a Member of the *College of Physicians, London.*

*Nihil magis Difficile, quàm Semel Infitam & ab Omnibus Susceptam
Opinionem Evellere, novamque Introducere.*

Jul. Cxl. Arant. de Fœtu Hum.

L O N D O N :

Printed and Sold by J. PEELE, at *Leicester-Head*, in
Pater-Noster-Row. MDCCXXVII.



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Haemangiomas



Haemangioma of Infancy

- Aetiology- unknown
- Risk factors
 - Prematurity (small birth weight)
 - Female
- Growth pattern
 - At or just after birth
 - Variable growth period
- Involution - ?cause
 - Variable extent



Haemangiomas





Regional haemangioma



- PHACE syndrome (PHACES)



Haemangioma treatment 2008

- Conservative
- Steroids
 - Intralesional
 - Systemic
- Interferon
- Vincristine
- Surgery



Haemangioma treatment 2009

- Conservative
- Beta blockers - Propranolol
- Steroids
 - Intralesional
 - Systemic
- Interferon
- Vincristine
- Surgery



Surgery for Haemangioma of Infancy

- Timing
 - End of growth phase
 - Early
 - Late
- Technique
- Indications
 - Complications
 - Obstruction
 - Ulceration
 - Social



Ulceration of Haemangioma



Haemangioma of infancy – specific sites

- Lip
- Eyelid
- Nose
- Trachea
- Liver



Lip







Eyelid



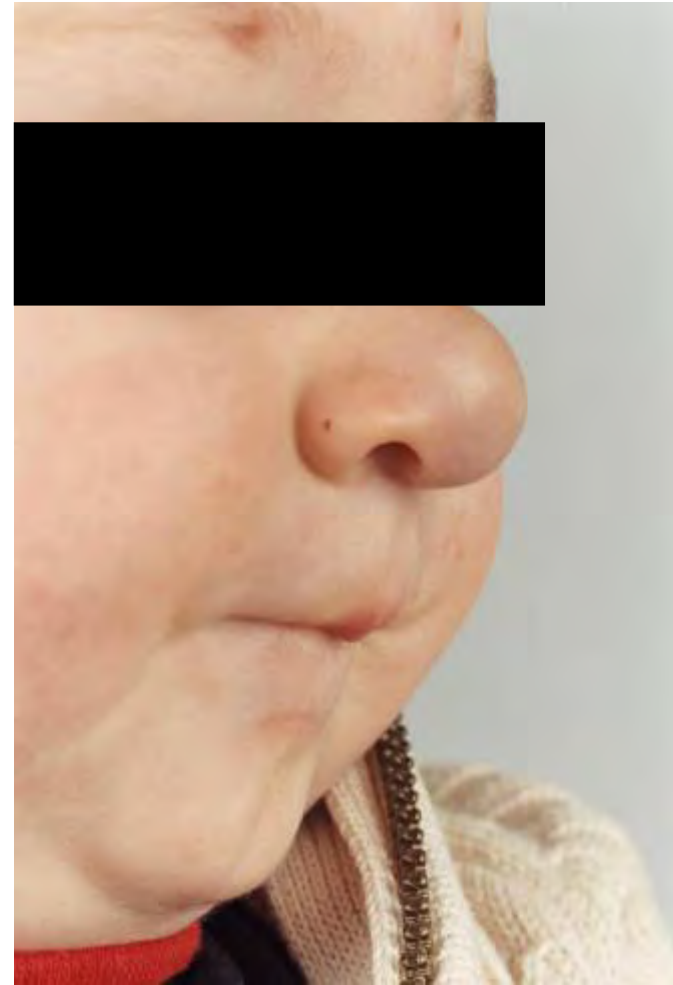
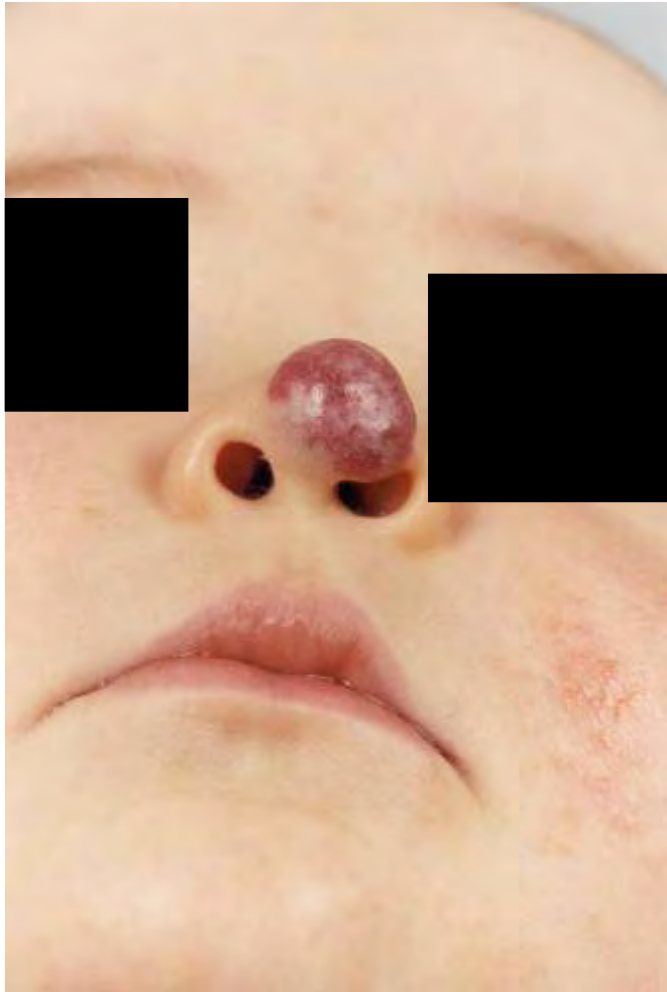
Eyelid



Eyelid



Tip of nose

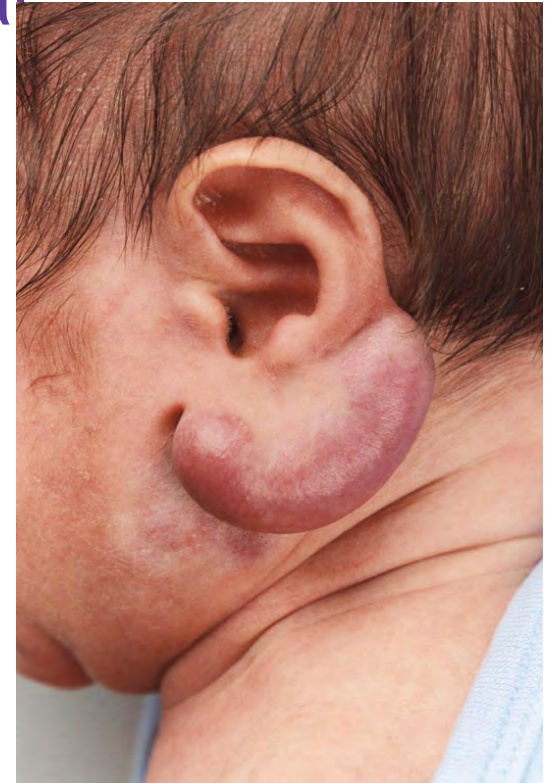




Congenital Haemangiomas

- Rapidly involuting congenital haemangioma (RICH)
- Non-involuting congenital haemangioma (NICH)

- Glut 1 negative





1 month



4 months



8 months





1 month



8 months



3 years



R.I.C.H.



N.I.C.H.

- Haemangioma of Infancy are Glut 1 positive
- Congenital Haemangioma (RICH & NICH) are Glut negative



Other proliferative conditions

- Kaposiform haemangendothelioma (KHE)
- Tufted angioma



Kassebach-Merritt Syndrome

- Platelet trapping
- Associated with KHE and Tufted Angioma
- Episodic fall in platelet count
 - Bleeding
- Steroids



- **Other proliferative conditions**



Other minor conditions

- Pyogenic Granuloma
 - Cheek in young children
 - Recurrent bleeding
 - Excision
- Naevus flammeus
 - 'Stork mark'
- Spider naevus
 - Laser



Vascular Malformations

- Arteriovenous malformations
- Venous malformations
- Glomuvenous malformations (glomangioma)
- Lymphatic malformations
- Capillary malformations



Management

- Make the diagnosis



Management

- Make the diagnosis
- Define the problem



Management

- Make the diagnosis
- Define the problem
 - Pain



Management

- Make the diagnosis
- Define the problem
 - Pain
 - Cosmesis



Management

- Make the diagnosis
- Define the problem
 - Pain
 - Cosmesis
 - Infection



Management

- Make the diagnosis
- Define the problem
 - Pain
 - Cosmesis
 - Infection
 - Loss of function



Management

- Make the diagnosis
- Define the problem
 - Pain
 - Cosmesis
 - Infection
 - Loss of function
 - Other



Management

- Make the diagnosis
- Define the problem
 - Pain
 - Cosmesis
 - Infection
 - Loss of function
 - Other
- Imaging to define lesion

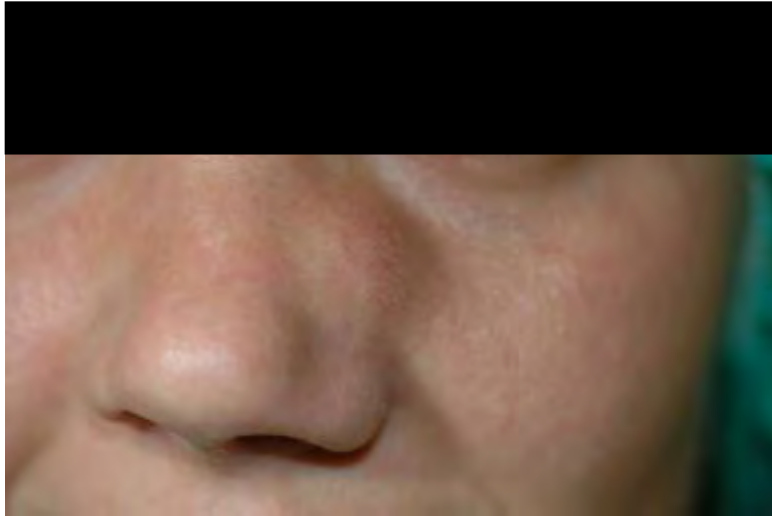


Treatment

- Conservative
 - Surgery
 - Interventional radiology
 - Laser
-
- Treat the problem.



Arteriovenous malformations



Arteriovenous malformations

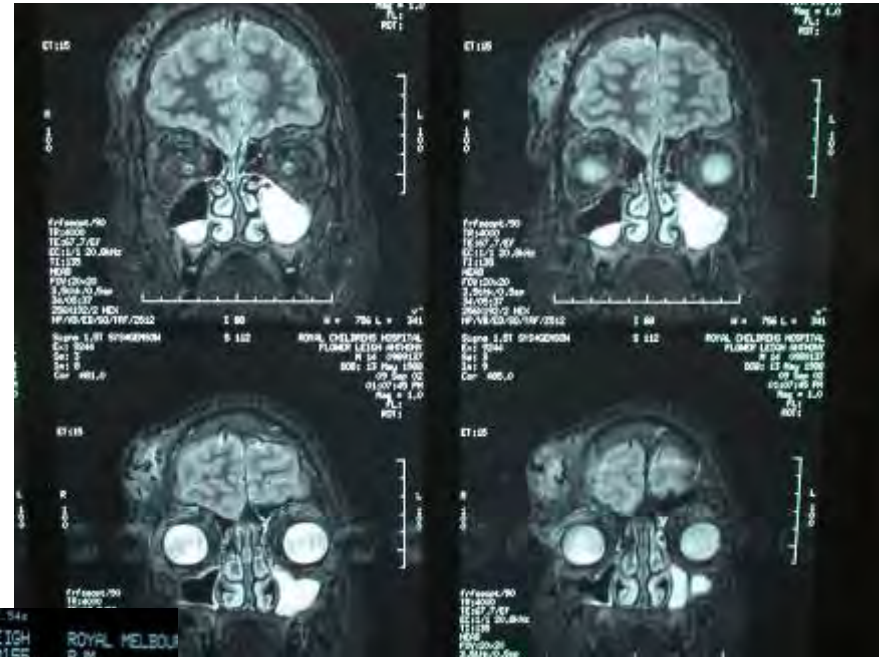


Arteriovenous malformations

Investigation

- MRI (MRA)
- Doppler ultrasound
- Angiography
- (Multislice CT)



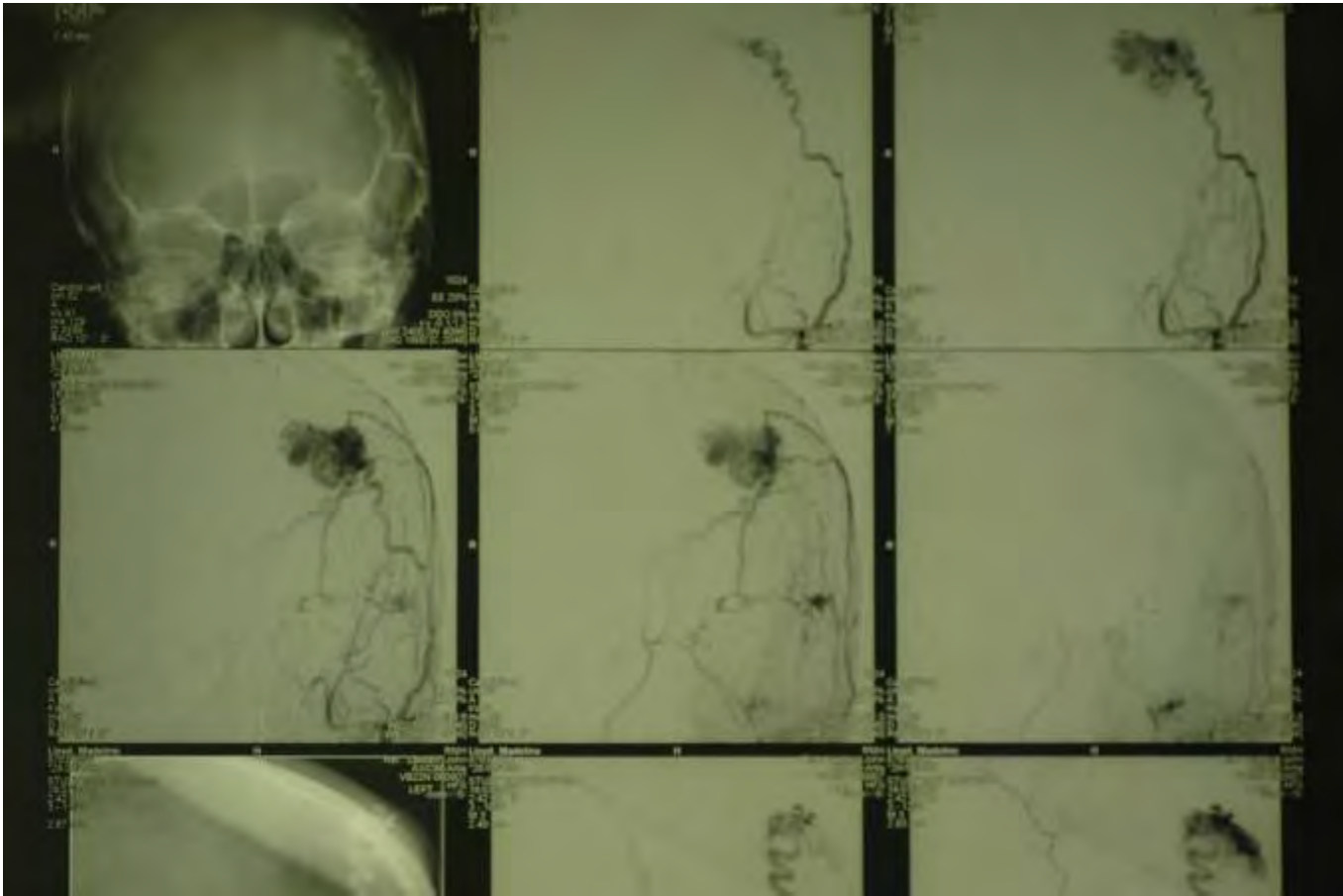


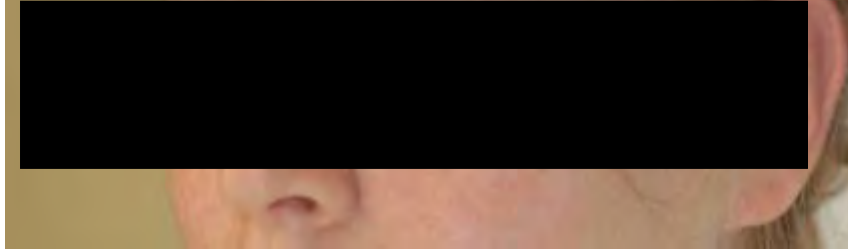
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Arteriovenous malformation

- Surgery
 - Excision of the nidus





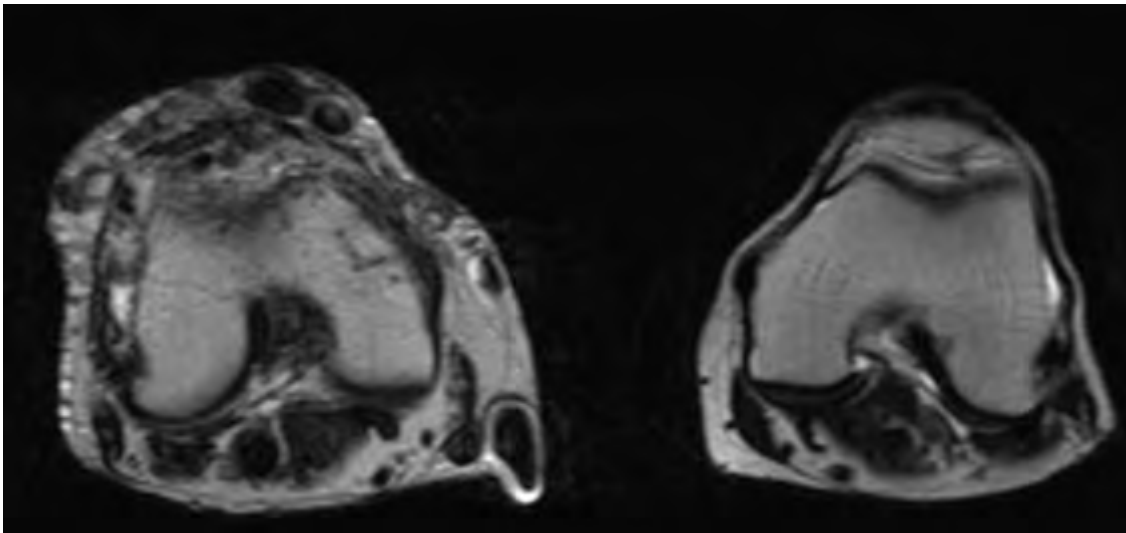
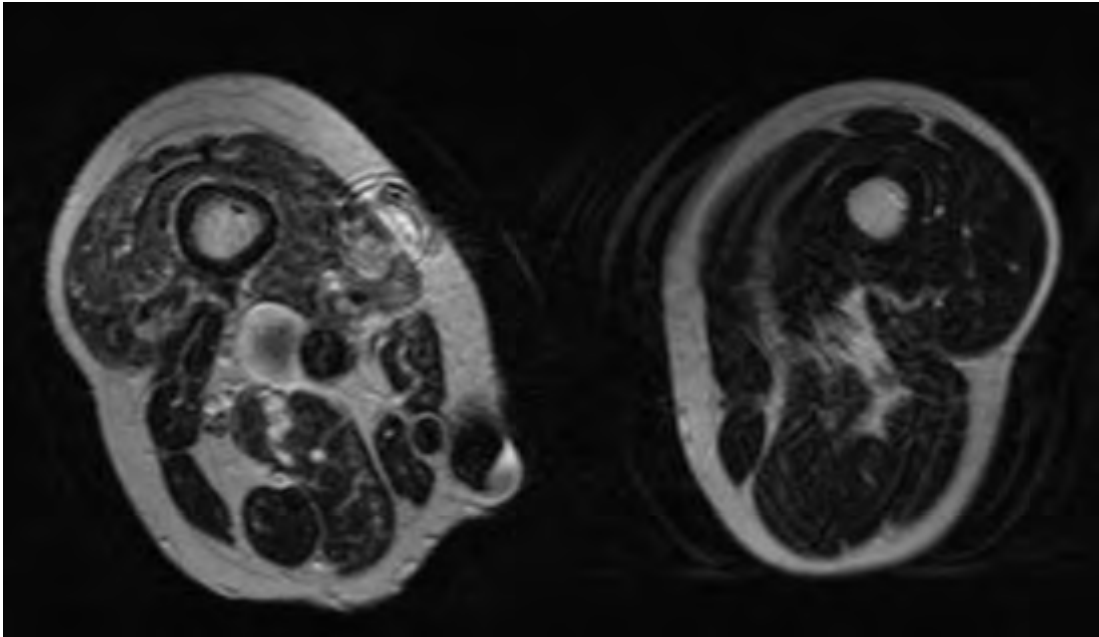


- 59 yo man
- Multiple previous embolizations
- High output cardiac failure
 - Cardiac output 9.5 l/minute
 - Pulmonary artery peak pressure 70-75 mmHg



- MRA







Surgery

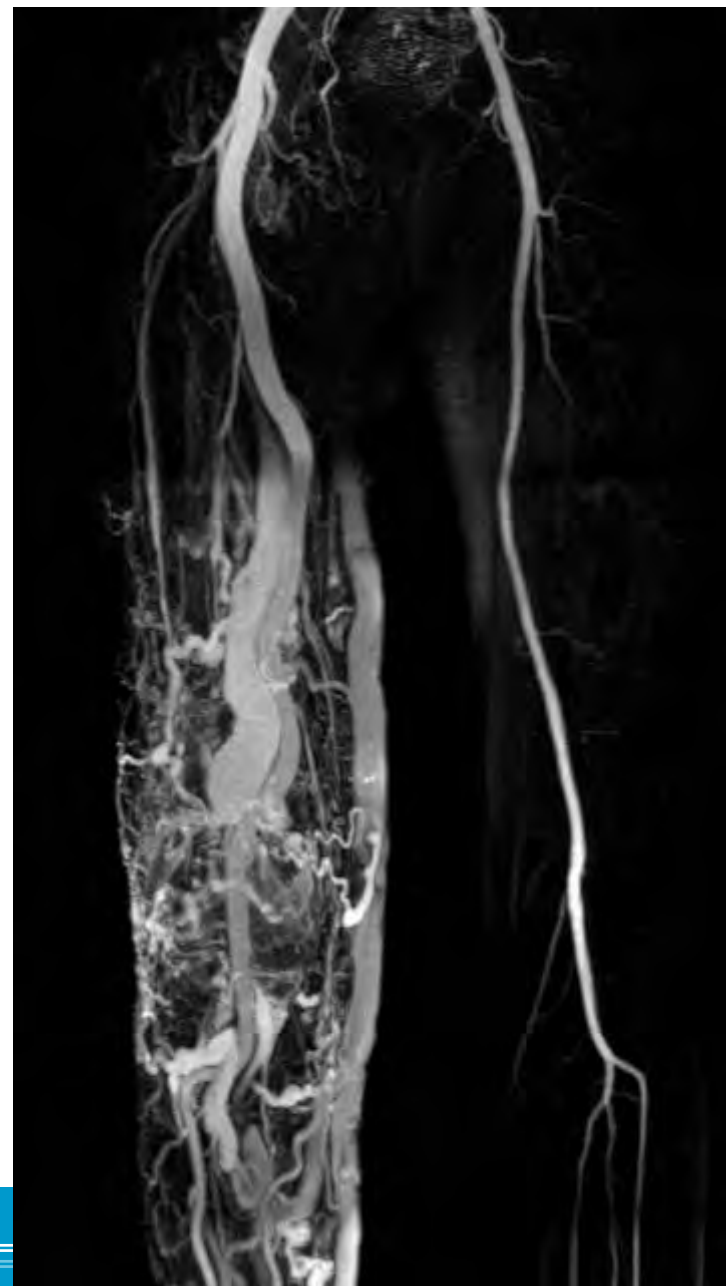
- Resection of vastus intermedius
- Tourniquet
- 10 hour procedure
- Cardiac output measured in theatre fell from 12 l/min to 7 l/min
- Incomplete resection around knee joint
- Ligation of saphenofemoral junction
- No blood intraop but Hb 92 on arrival in ICU



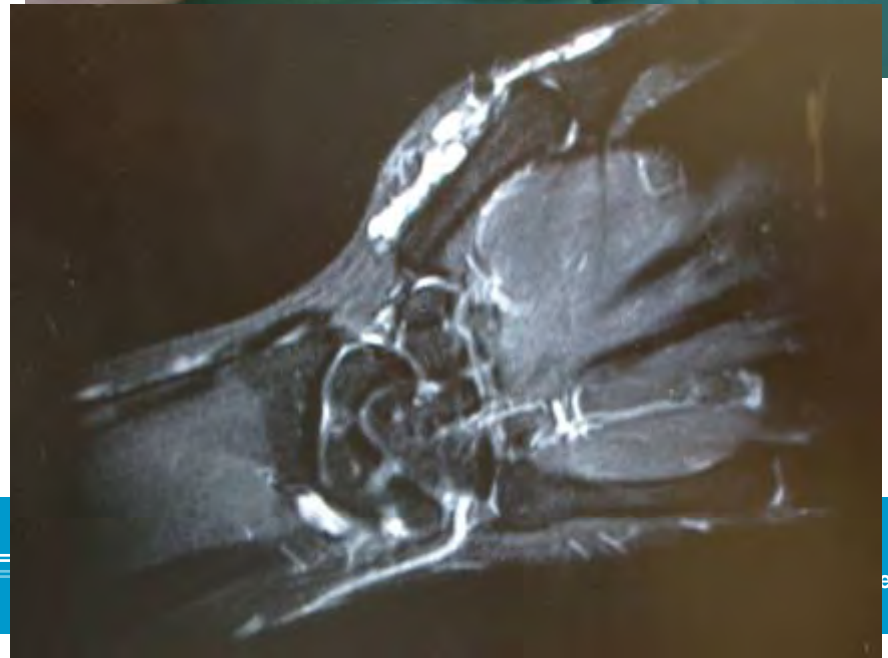
Post op

- Cardiac output 7.6 l/min
- Pulmonary artery pressure 45/22 mmHg
- 4 unit blood transfusion
- 1 day in ICU, 15 days inpatient
- Follow up 6 months, shortness of breath improving
- MRI thigh





Venous Malformations



Venous Malformations

- Aetiology
 - >95% sporadic
 - <5% familial
 - Activating mutation in the Tie 2 gene



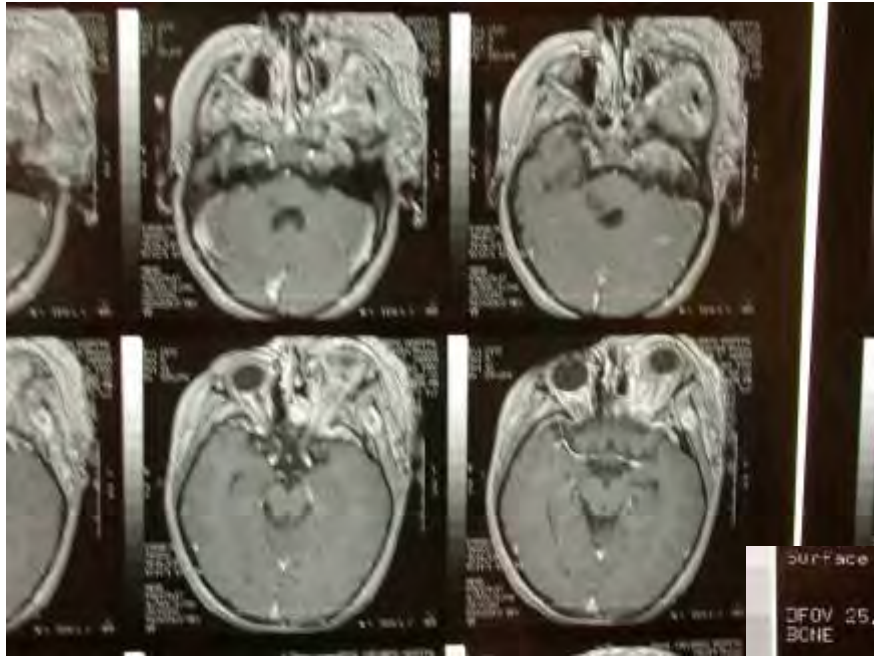
Venous malformation Investigation

- MRI
- Ultrasound

- Direct puncture venography







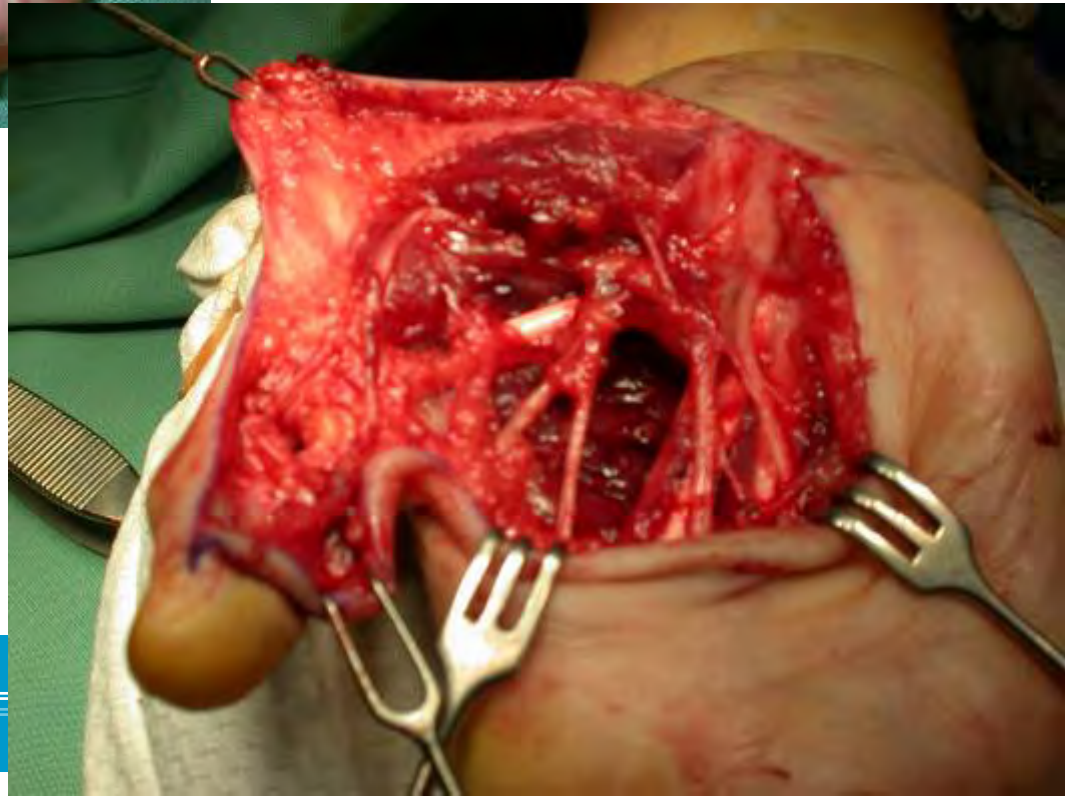
Venous Malformation - Treatment

- Injection Sclerotherapy
 - Ethanol
 - Sodium tetradecol (Sotracol)
 - Polidocanol (Aethoxysclerol)

- Surgery







Lymphatic malformation



Lymphatic Malformations

- Incidence 1 to 2 per 1,000 live births
- Location – all areas of the body
- Growth – grow with the child but some fluctuation in size
- Aetiology – unknown
 - Not associated with syndromes
 - ? Endothelial cell abnormality



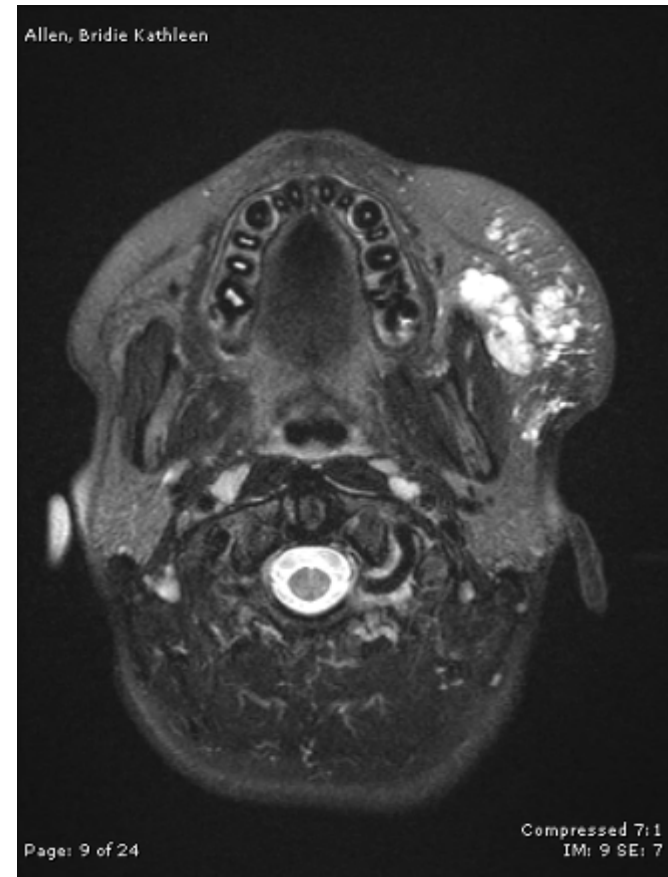
Lymphatic Malformations

- Macrocystic
 - V
- Microcystic



Investigations

- Ultrasound
- MRI



Define the problem

- Appearance
- Loss of function
- Infection
- Pain



Infection



Acute infection →
resolution of large
cystic lesions



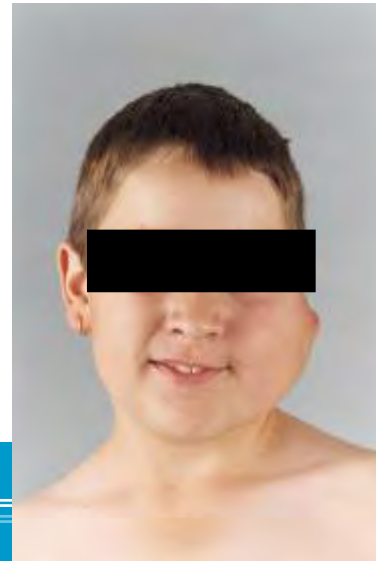
Infection



Acute infection → resolution of large cystic lesions



Chronic infection →



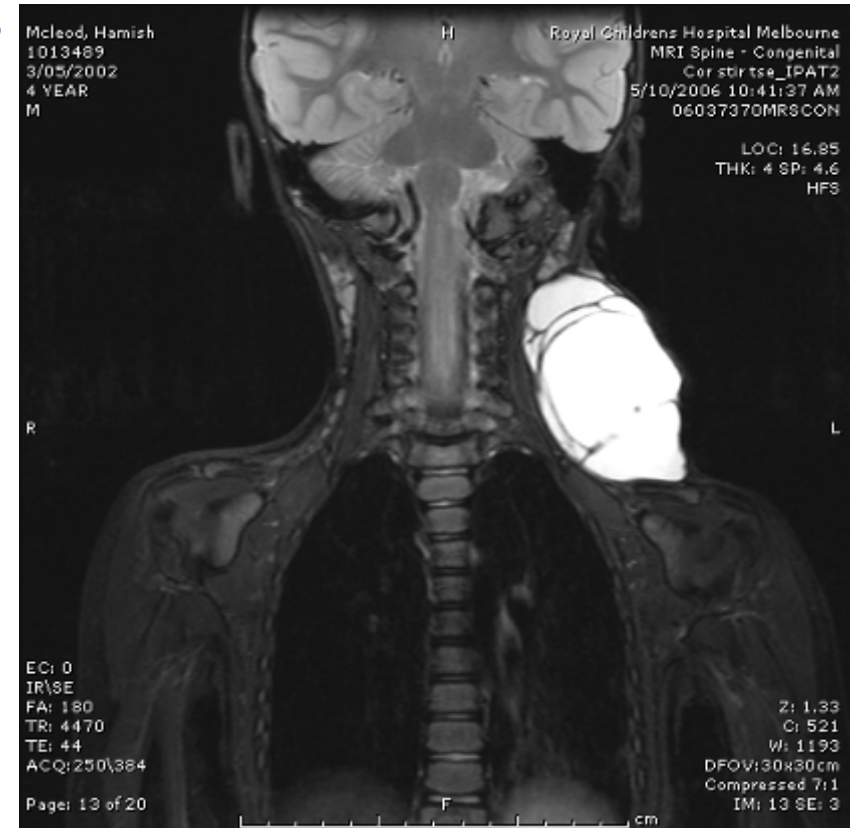
Treatment Options

- Conservative
- Sclerotherapy
- Surgery



Sclerotherapy

- Macrocytic lesions
 - Must be visible on ultrasound

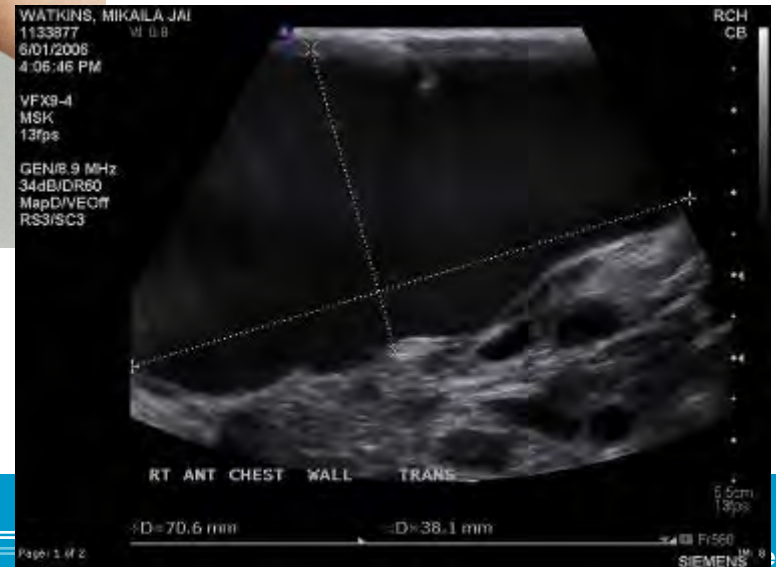


Sclerotherapy

- Multiple agents
 - OK432
 - Doxycycline
 - Bleomycin
 - Ethanol
- Good correction of contour
 - Significant post-op swelling



Sclerotherapy



Sclerotherapy





Surgery

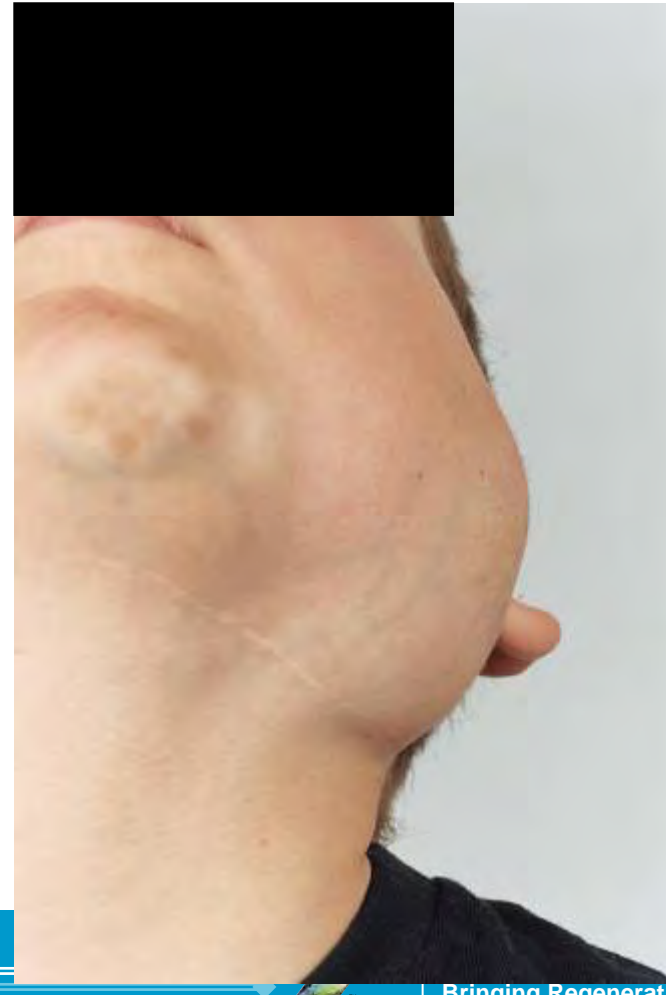
- Technically challenging
 - Nerve dissection
 - Preserve contour
 - Bleeding
 - Airway
- Clear one region as completely as possible



Lymphatic Malformations

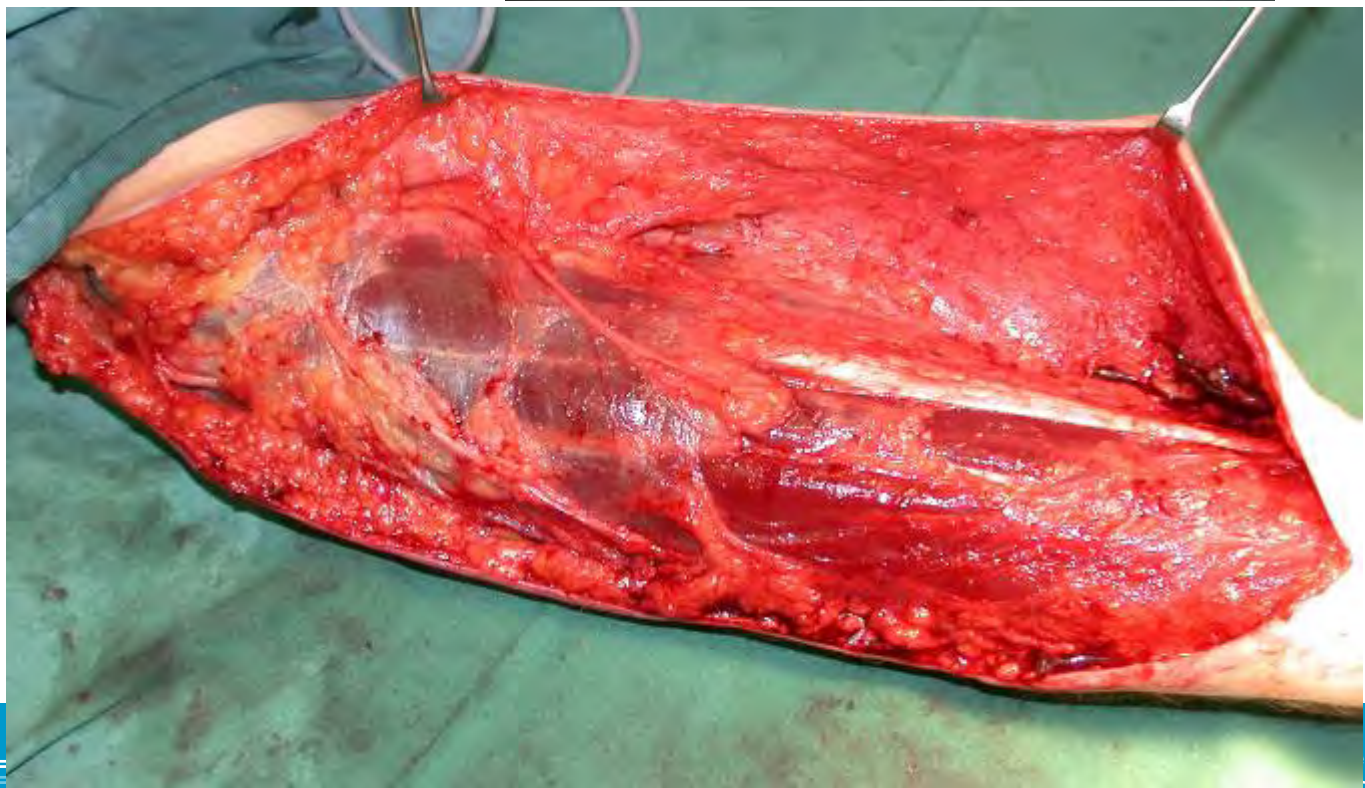


Redo Face dissection

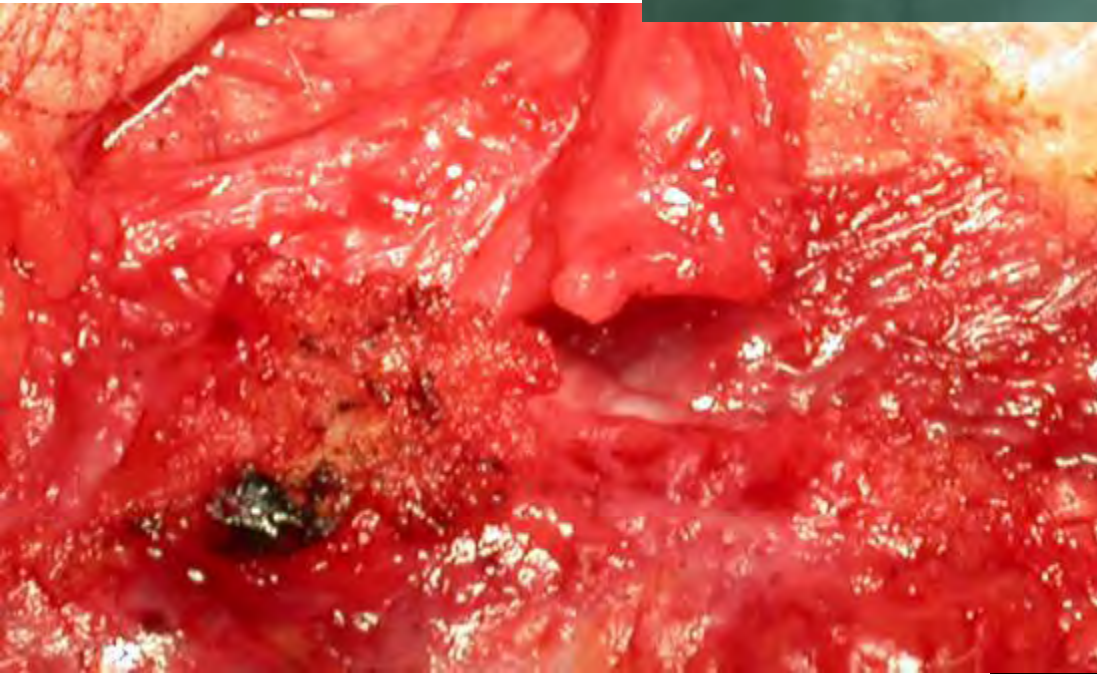








Lympha



Glomuvenous malformation (Glomangioma)

- High incidence of familial inheritance
 - Autosomal dominant
 - 'Glomulin' gene



Capillary Malformations



Capillary Malformation



Syndromes

- Klippel-Trenaunay syndrome
- Parkes Weber syndrome
- Maffucci
- Blue Rubber Bleb Naevus
- Sturge Weber
- Bannayan Riley Rubalcava
 - Cowden Syndrome (PTEN)
- Proteus



- MÉMOIRES ORIGINAUX

- DU NŒVUS VARIQUEUX ~~OSTÉO~~-HYPERTROPHIQUE

- PAR MM.

- M. KLIPPEL ET P. TRENAUNAY

- Medecin des Hôpitaux

- Externe des Hôpitaux

Archive Générales de Médecine, 1900



Klippel-Trenaunay Syndrome

- Vascular birthmark
- Overgrowth
- Deep vein absence



Parkes Weber Syndrome

- Limb Hypertrophy
- Small red patches on skin
- Multiple small AV fistulae

- RASA 1 gene



Syndromes

- Maffucci's syndrome



Syndromes

- Blue Rubber Bleb Naevus syndrome



Syndromes

- Sturge Webber
 - Facial CM
 - Epilepsy



Syndromes

- Klippel-Trenaunay syndrome
- Parkes Weber syndrome
- Maffuci
- Blue Rubber Bleb Naevus
- Sturge Weber
- Bannayan Riley Rubalcava
 - Cowden Syndrome (PTEN)
- Proteus



Interventional Radiology

- AVM
- Venous Malformation
- Lymphatic Malformation



Arteriovenous Malformation

- Interventional radiology
 - Embolization – high recurrence rate
- Sclerotherapy
 - Alcohol
 - STD
 - Aims to obliterate the nidus
 - Skin necrosis, nerve damage



Venous Malformation

- Surgery
 - Complete excision impossible
- Interventional radiology
 - Percutaneous puncture
 - Alcohol, STD, Polidocanol



Lymphatic malformation

- Macrocystic v microcystic
- Macrocystic
 - Injection sclerotherapy
 - OK432, Tetracycline, Bleomycin, Alcohol
- Microcystic
 - Surgery



Conclusion

- Classification
 - Mulliken/ISSVA
 - Haemangioma v Vascular Malformation
- (Treatment)

