VASCULAR MALFORMATIONS

Inborn errors of vasculogenesis of unknown aetiology
All present at birth but only clinically apparent later; never involute, persist for life
May increase in size periodically eg puberty, pregnancy, thrombosis, post-trauma
Classified into low flow (capillary, venous, lymphatic) and high flow (arteriovenous) malformations

LOW FLOW MALFORMATIONS

CAPILLARY MALFORMATIONS

1. Salmon patch (naevus simplex/erythema nuchae)
Red macule/nape of neck, upper eyelids, glabella/non-dermatomal
Present at birth (40%) and fade by 2 years but nuchal lesion persists

2. Port-Wine Stain (naevus flameus)
Red macule/ face + neck, can be trunk or limb/dermatomal
Present at birth (0.3%) and growth rate matches child's/becomes, thick, nodular over time

*Sturg-Weber: port wine stain(V1 distro), ipsilateral leptimeningeal Vm (seizures), choroid Vm (glaucoma)
*only 10% of children with facial P-WS have S-WS
*MRI helpful

VENOUS MALFORMATIONS

Dilated venous spaces of varying size with sluggish blood flow
Most single; Bean syndrome/Blue Rubber Bleb syndrome
Symptoms: **pain** and **swelling** (engorgement/thrombosis)
Signs: blue skin discolouration/pulsatile soft tissue swelling/phleboliths
Other: very large venous malformations → coagulopathy
Imaging: MRI/CT if bone involvement (osseous hypertrophy or atrophy)
Management: treat symptoms and improve cosmesis; depends on size, site, soft or bony nature
-percutaneous sclerotherapy (eg sodium tetradacyl sulphate/absolute alchohol) for large compressible lesions that expand when dependent
-Surgery (Popescu sutural compartmentalisation etc) for small, poorly-compressible lesions with no expansion on dependence

LYMPHATIC MALFORMATIONS

Microcystic (commoner) or macrocystic
From birth, evident years into childhood
MRI; US differentiates between both
Management: percutaneous sclerotherapy (OK-432/bleomycin/doxycycline/alchohol/STS)

HIGH FLOW MALFORMATIONS (AV FISTULAE)

Congenital (present at birth, apparent later) or **Acquired** (penetrating trauma)
Signs: pulsatile swelling/prominent draining veins/skin ulcerates
Imaging: Hand-held Doppler/MRI (extent)/CT (bone involvement)/Angio **not contrast CT**
Management: symptoms/deformity→embolization

Houdart classification (based on arrangement of AV communications)
I ARTERIOVENOUS Venous component supplied by 3- arterial pedicles
II ARTERIOLOVENOUS component supplied by >3 arterial pedicles
III ARTERIOLOVENULOUS: arteriovenous communications indistinguishably small and numerous with first identifiable venous component some distance away

Embolisation - low pressure first venous component is venous sump and must be occluded totally
-can't just embolise feed vessels as will draw blood from collaterals; aim for AV shunts
-also done pre-surgery to reduce vascularity (not extent of excision) with a large particulate that will not pass through AV communciations

Surgery: total exclusion

Vascular malformations

•simple◦
capillary malformation (CM)port-wine stain
telangiectasia
cutis marmorata teleangiectatica congenital
nevus simplex/salmon patch
others

◦venous malformation (VM)common sporadic venous malformation
blue rubber bleb nevus syndrome (BRBNS)
familial cutaneous and mucosal venous malformation
glomuvenous malformation (glomangioma)
cerebral cavernous malformation

◦lymphatic malformation (LM)
common (cystic)
generalized lymphatic anomaly
LM in Gorham-Stout disease
channel-type
primary lymphedema

◦ arteriovenous malformations (AVM) (with a nidus of multiple shunts)

◦ arteriovenous fistulae (AVF)(with one or more shunts)

•those of major named vessels◦various abnormalities affecting origin, course, number, length, diameter, valves, communication and persistence

•those associated with other anomalies◦Klippel-Trenaunay syndrome

◦Parkes-Weber syndrome
◦Servelle-Martorell syndrome
◦Sturge-Weber syndrome
◦Maffucci syndrome
◦CLOVES syndrome
◦Proteus syndrome
◦Bannayan-Riley-Ruvalcaba syndrome