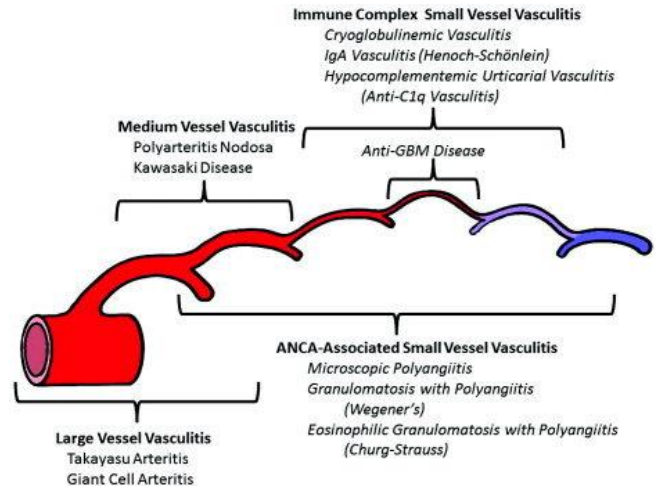


## What is large vessel vasculitis?

'Large vessel vasculitis' (LVV) encompasses a spectrum of diseases causing chronic granulomatous inflammation of large vessels. The two main well-defined conditions are Takayasu Arteritis and Giant Cell Arteritis. It is worth noting that any other large vessel can be affected and this is even more rare. Presentation will depend on which artery is affected.



## **Takayasu Arteritis** AKA Pulseless Disease

### Definition

Chronic, progressive, inflammatory, occlusive disease of the aorta and its branches, resulting in stenosis, occlusion, dilation or aneurysm formation.

### Epidemiology

More common in Asia (Japan) & in women. Usually under the age of 40.

### Presentation

**Systemic stage** (prior to occlusion):

Fever, fatigue, weight loss, malaise, arthralgia

**Occlusive stage** (ischaemic phenomena)

Limb claudication, hypertension, back pain, headaches, dyspnoea, syncope, abdo pain

**Key findings:** systolic BP difference >10 between arms, non-palpable peripheral pulses, tender carotid artery, ↑ ESR/CRP (unlikely to be any LVV without this)

### Diagnosis

Angiography, USS, histology of post-op specimen

### Management

Steroids, DMARDs e.g. methotrexate/azathioprine, biologics e.g. tocilizumab.

Angioplasty or grafts might be required

May be a single episode or relapsing/remitting disease

## **Giant Cell Arteritis** AKA Temporal Arteritis

### Definition

Systemic, immune-mediated inflammatory disease of the carotid artery & its extra-cranial branches, usually the temporal artery. (Note that temporal arteritis just means inflammation of the temporal artery and could be caused by conditions other than GCA).

### Epidemiology

Usually in Northern European countries & in women Always aged >50, usually age 60-80

### Presentation

**Systemic:** Myalgia, malaise, fever, night sweats

**Inflammatory/ischaemic:** Temporal headache, jaw claudication, scalp tenderness, visual disturbance

N.B. 50% of patients with GCA have symmetrical shoulder pain/stiffness with a picture similar to PMR\*

**Key findings:** ↑ ESR/CRP, hardened and enlarged temporal artery and absent temporal pulse.

### Diagnosis

Temporal artery biopsy, ultrasound temporal artery Results sometimes obscured by effects of steroids as delaying treatment results in permanent blindness.

### Management

High dose steroids, wean down gradually.

Methotrexate/tocilizumab if refractory

## \***Polymyalgia Rheumatica (PMR)**

**Epidemiology:** As with GCA.

**Presentation:** Acute severe bilateral pain and morning stiffness of the shoulder, neck and pelvic girdle.

**Diagnosis:** Clinical picture with ↑ ESR/CRP (exclude cancer/infection/GCA which needs urgent treatment) Inflammation of subclavian/axillary arteries may be seen on USS – indication of pathogenesis.

**Management:** Usually 1 year of low dose steroids (compared to high dose with GCA).

## **Resources**

- Oxford Textbook of Rheumatology
- Patient UK Professional <https://patient.info>
- 2018 Update of the EULAR recommendations for the management of large vessel vasculitis

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