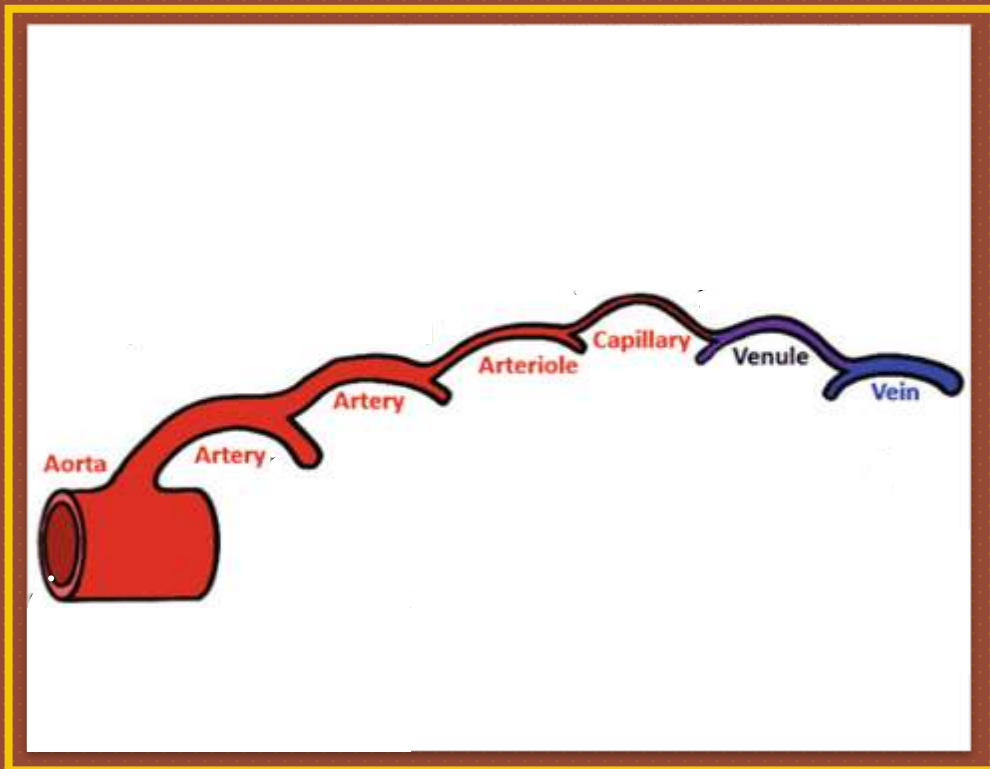


# VASCULITIDES



**CLASSIFICATION,  
SYMPTOMS &  
GENERAL PRINCIPLES OF  
MANAGEMENT**

# VASCULITIDES

Types, Symptoms &  
General Principles of Management

# Introduction

- **Inflammatory** disorders of blood vessels
- Affect any organ
- Presentation depends on the organs involved
- **Primary** condition or **Secondary** to other diseases

# Classification

## Modified Chapel Hill Criteria

### Large Vessels

- Giant cell arteritis
- Takayasu's arteritis

### Medium Vessels

- Polyarteritis nodosa
- Kawasaki disease

### Small Vessels

#### ANCA-associated

- Microscopic polyangiitis
- Granulomatosis with polyangiitis  
(*Wegener's granulomatosis*)
- Eosinophilic granulomatosis with polyangiitis  
(*Churg Strauss syndrome*)

#### Immune complex vasculitis

- Goodpasture's disease
- Cryoglobulinaemic vasculitis
- IgA vasculitis  
(*Henoch-Schonlein purpura*)

### Variable vessel vasculitis

- Behcet's syndrome
- Cogan's syndrome

### Single-organ vasculitis

### Vasculitis associated with systemic diseases

### Vasculitis associated with probable aetiology

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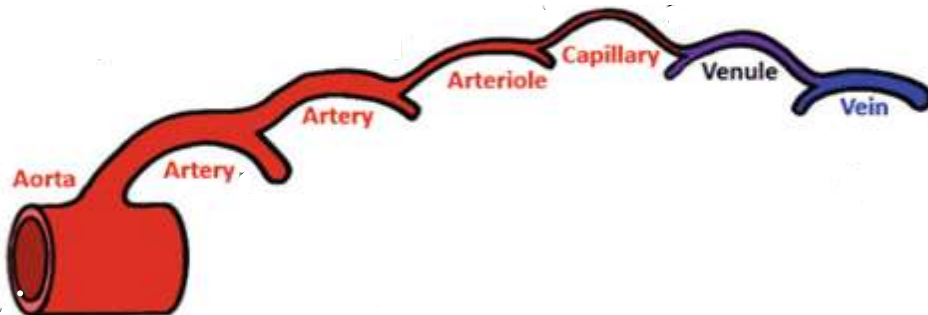
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## Variable vessel vasculitis

- Behcet's syndrome
- Cogan's syndrome

## Single-organ vasculitis

- Central nervous system vasculitis
- Cutaneous small vessel vasculitis
- Isolated aortitis

## Vasculitis associated with Systemic disease

- Systemic Lupus Erythematosus (SLE)
- Rheumatoid Arthritis (RA)
- Relapsing polychondritis
- Others

## Vasculitis associated with probable aetiology

- Hepatitis C virus-associated cryoglobulinemic vasculitis
- Hepatitis B virus-associated polyarteritis nodosa
- Syphilis-associated aortitis
- Hydralazine-associated ANCA-associated vasculitis
- Cancer-associated vasculitis

# Clinical Features

- Different vasculitides affect different organs
- Variable symptoms
- Often, marked fatigue with raised ESR/CRP
- Consider in any unidentified multisystem disorder
- If presentation does not fit into a specific category - Consider malignancy-associated vasculitis

# Clinical Features

- **Systemic:** Fever, Malaise, Weight loss, Arthralgia, Myalgia
- **Skin:** Purpura, Ulcers, Livedo reticularis, Nailbed Infarcts, Digital Gangrene
- **Eyes:** Episcleritis, Scleritis, Visual Loss
- **ENT:** Epistaxis, Nasal Crusting, Stridor, Deafness
- **Pulmonary:** Hemoptysis and Dyspnoea
- **Cardiac:** Angina or MI, Heart failure, Pericarditis

- **GI:** Pain or perforation, malabsorption
- **Renal:** Hypertension, Glomerulonephritis, and Renal failure
- **Neurological:** Stroke, Seizures, Chorea, Psychosis, Confusion, Impaired cognition, Altered mood. Mononeuritis multiplex or Sensorimotor-Polyneuropathy due to arteritis of the vasa nervorum
- **Genitourinary:** Orchitis



# Investigations

- **Angiography +/- Biopsy** - may be diagnostic
- **ESR / CRP** - *Raised*
- **ANCA** - may be *Positive*
- **RFTs** - Creatinine *Raised*, if renal failure
- **Urinalysis** - Proteinuria, Haematuria, Casts

# Management

## Large-vessel

- **Steroids**
- Steroid- sparing agents *later*

## Medium/Small

- **Immunosuppression**
  - Steroids +/- another agent
  - eg Cyclophosphamide *if severe, or* Methotrexate/Azathioprine

- Severe flare is a **medical emergency**
- Seek urgent help, as organ damage may occur rapidly

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