

# MEDIUM VESSEL VASCULITIS

## POLYARTERITIS NODOSA (PAN)

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### Definition

- Systemic necrotizing vasculitis of small to medium muscular arteries.
  - Spares pulmonary circulation.
  - ANCA-negative.
  - Up to 1/3 associated with chronic Hepatitis B, leading to immune complex deposition .
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### Pathogenesis

- Hepatitis B antigens → immune complex formation → deposition in vessel walls → necrotizing inflammation .

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## Morphology

- Segmental transmural necrotizing inflammation of small to medium arteries.
- Mixed infiltrate: neutrophils + mononuclear cells, fibrinoid necrosis, and luminal thrombosis.
- Lesions often at branch points.
- Chronic: fibrous thickening of vessel wall → aneurysms & rupture.
- Organs affected (most → least): kidneys  → heart → liver → GI tract.
- Glomeruli are spared.
- All stages of activity can coexist in different vessels or within the same vessel.

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## Clinical Features

- Usually affects young adults, but can occur in middle/older adults.

- Episodic course with symptom-free intervals.
  - Systemic: fever 🤒, malaise, weight loss.
  - Vascular manifestations:
    - Hypertension from renal artery involvement 💧
    - Abdominal pain, bloody stools from GI lesions
    - Myalgia, arthralgia, peripheral neuropathy
  - Renal involvement is a major cause of death.
  - Untreated → typically fatal; immunosuppression → 5-year survival ~80%.
  - Relapse occurs in up to 25%, more often in non-HBV cases.
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## MEDIUM VESSEL VASCULITIS

### KAWASAKI DISEASE ❤️

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#### Definition

- Acute, febrile, usually self-limited illness of infancy & childhood (<5 years).
  - Vasculitis of large to medium vessels, especially coronary arteries.
  - Can cause coronary aneurysms → rupture or thrombosis → myocardial infarction .
  - More common in children of East Asian descent; genetic susceptibility suspected.
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## Pathogenesis

- Likely delayed-type hypersensitivity reaction against vascular antigens.
  - Infectious triggers (mostly viral; SARS-CoV-2 recently implicated) in genetically susceptible children.
  - Cytokine production + B-cell activation → autoantibodies to endothelial cells and smooth muscle cells → vasculitis .
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## Morphology

- Resembles polyarteritis nodosa: dense transmural inflammation.
  - Fibrinoid necrosis usually less prominent than PAN.
  - Healing → intimal thickening, possible aneurysm formation.
  - SARS-CoV-2 associated cases may involve multiple organs 🌍.
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## Clinical Features

- Mucocutaneous manifestations:
  - Conjunctival & oral erythema/blistering 👁️👄
  - Edema of hands & feet
  - Rash (desquamative)
  - Cervical lymphadenopathy
- Cardiovascular:
  - Coronary artery ectasia → aneurysms (7-8 mm)

- Risk of rupture, thrombosis, myocardial infarction, sudden death 
  - Treatment: IV immunoglobulin + aspirin → drastically reduces symptomatic coronary disease.
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-> The End <-