
In the name of
God

Systemic Vasculitis

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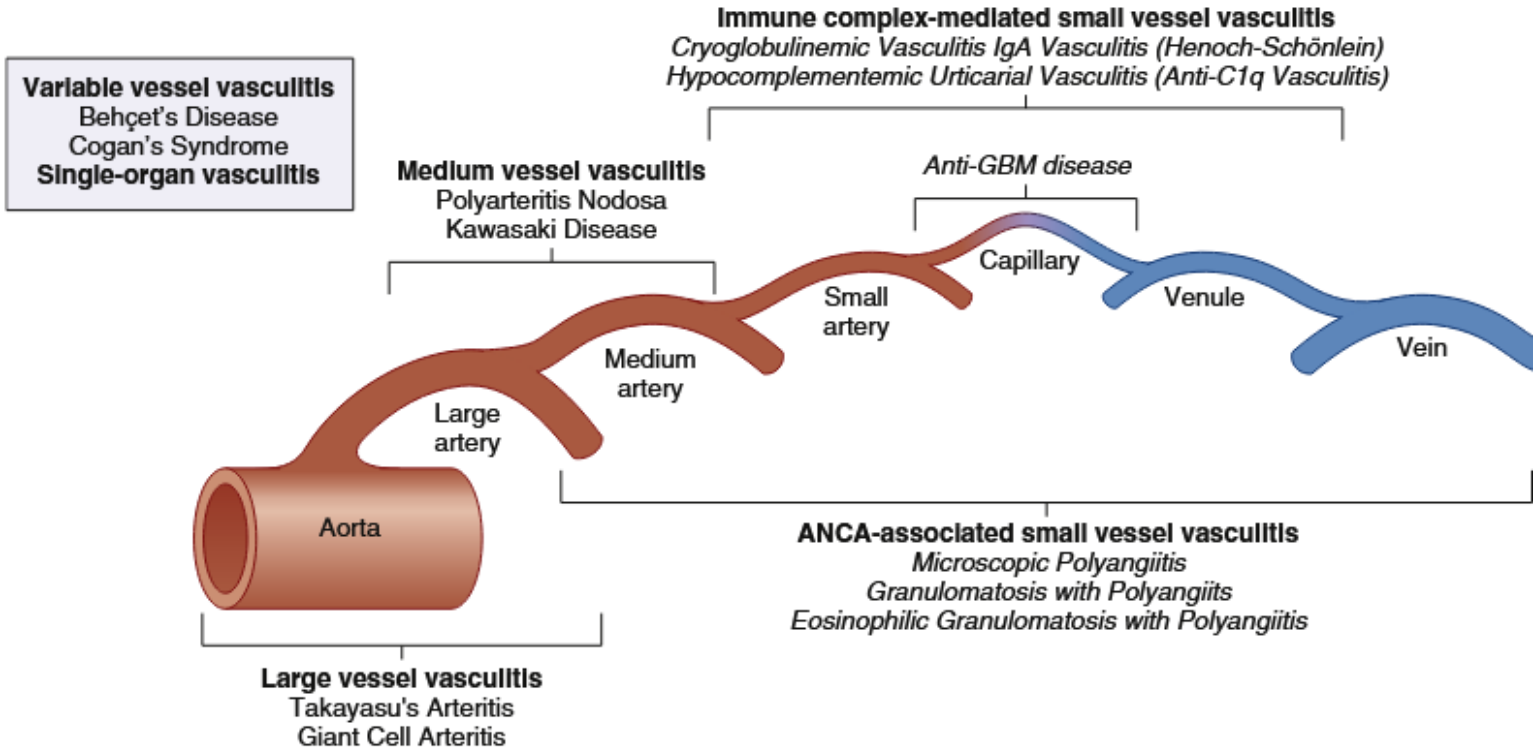
Classification of Vasculitis

- 1994, 2012 International Chapel Hill Consensus Conference (CHCC)


Variable vessel vasculitis
Behçet's Disease
Cogan's Syndrome
Single-organ vasculitis
Immune complex-mediated small vessel vasculitis
Cryoglobulinemic Vasculitis IgA Vasculitis (Henoch-Schönlein)
Hypocomplementemic Urticarial Vasculitis (Anti-C1q Vasculitis)
Medium vessel vasculitis
Polyarteritis Nodosa
Kawasaki Disease
Anti-GBM disease
Capillary
Small artery
Venule
Vein
Medium artery
Large artery
Aorta
ANCA-associated small vessel vasculitis
Microscopic Polyangiitis
Granulomatosis with Polyangiits
Eosinophilic Granulomatosis with Polyangiitis
Large vessel vasculitis
Takayasu's Arteritis
Giant Cell Arteritis

Nomenclature of the Systemic Vasculitides defined during the 2012 International Chapel Hill C

Systemic Vasculitides
Small-vessel vasculitis (SVV)
Anti-neutrophil cytoplasmic antibody (ANCA)–associated vasculitis (AAV)
Microscopic polyangiitis (MPA)
Granulomatosis with polyangiitis (Wegener's) (GPA)
Eosinophilic granulomatosis with polyangiitis (Churg–Strauss) (EGPA)
Immune complex SVV
Anti-glomerular basement membrane (anti-GBM) disease
Cryoglobulinaemic vasculitis (CV)
IgA vasculitis (Henoch–Schonlein) (IgAV)
Hypocomplementaemic urticarial vasculitis (HUV) (anti-C1q vasculitis)
Medium-vessel vasculitis (MVV)
Polyarteritis nodosa (PAN)
Kawasaki disease (KD)
Large-vessel vasculitis
Takayasu arteritis (TA)
Giant cell arteritis (GCA)
Variable vessel vasculitis (VVV)
Behçet's disease (BD)
Cogan's syndrome (CS)

Size of vessels

- Large-vessel → aorta and its main branches and the corresponding veins;
- Medium-vessel → main visceral arteries and veins such as renal, mesenteric, and coronary arteries
- Small-vessel → intraparenchymal arteries, arterioles, capillaries, venules, and veins

Small-Vessel Vasculitis

- ANCA-associated vasculitis

- Immune complex-mediated vasculitis

What is the definition of AAV?

1) Necrotizing
2) Few or no immune complex deposit
3) Affecting small vessels
4) Associated with:

I. MPO ANCA
II. PR3 ANCA
III. ANCA negative

AAV

Feature	GPA	MPA	Eosinophilic GPA
Incidence	0.4–11.9 cases per 1 million person-years	0.5–24.0 cases per 1 million person-years	0.5–2.3 cases per 1 million person-years
Prevalence	2.3–146.0 cases per 1 million persons	9.0–94.0 cases per 1 million persons	2.0–22.3 cases per 1 million persons
Typical age of onset (years)	45–65	55–75	38–54
Male: female ratio	1:1	1:1	1:1
2012 revised CHCC definition[145]	Necrotizing granulomatous inflammation, usually involving the upper and lower respiratory tract; necrotizing vasculitis affecting predominantly small-to-medium vessels (such as capillaries, venules, arterioles, arteries and veins); necrotizing glomerulonephritis is common	Necrotizing vasculitis, with few or no immune deposits, predominantly affecting small vessels (such as capillaries, venules or arterioles); necrotizing arteritis involving small and medium arteries may be present; necrotizing glomerulonephritis is very common; pulmonary capillaritis often occurs; granulomatous inflammation is absent	Eosinophil-rich and necrotizing granulomatous inflammation, often involving the respiratory tract; necrotizing vasculitis predominantly affecting small-to-medium vessels; associated with asthma and eosinophilia; ANCA$^+$ is more frequent when glomerulonephritis is present
Frequency of ANCA	PR3-ANCA$^+$: 65–75% MPO-ANCA$^+$: 20–30% ANCA$^-$: 5%	PR3-ANCA$^+$: 20–30% MPO-ANCA$^+$: 55–65% ANCA$^-$: 5–10%	PR3-ANCA$^+$: <5% MPO-ANCA$^+$: 30–40% ANCA$^-$: 55–65%
Key innate immune cell	Neutrophil	Neutrophil	Eosinophil
Relapse rate	Higher than MPA (or MPO-AAV)	Lower than GPA (or PR3-AAV)	Relapse is frequent

EPIDEMIOLOGY

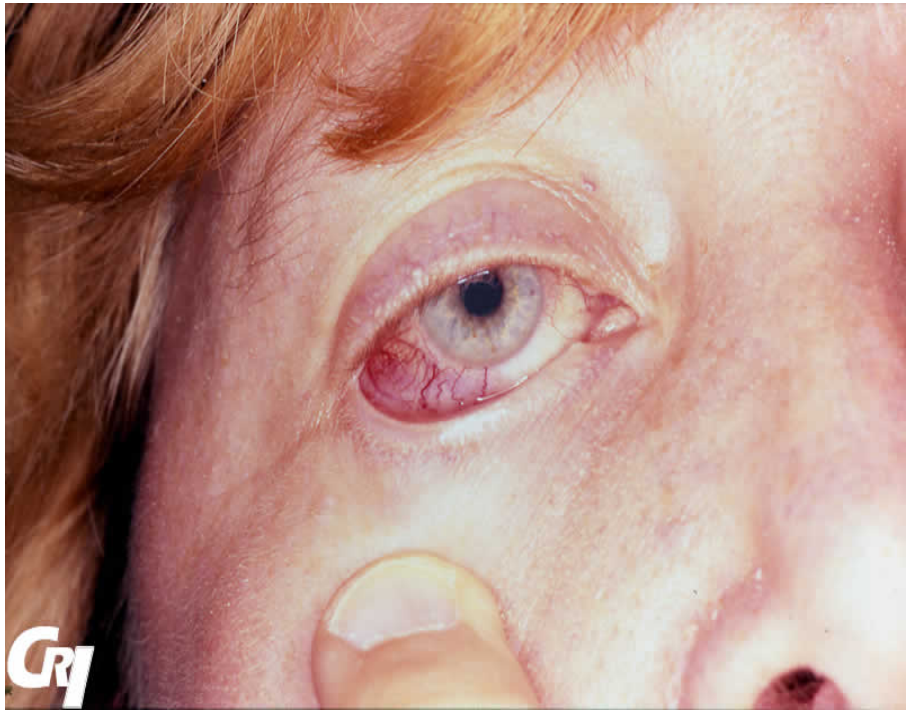
- AAV has an estimated annual incidence of 20 per million in habitants and a prevalence of 100 per million in habitants.
- According to some epidemiological trials, the majority of them from Europe, MPA and GPA have similar incidences of 2.4–10.1 and 2.1–14.4, respectively, and EGPA, less frequent, an estimated incidence of 0.5–3.7

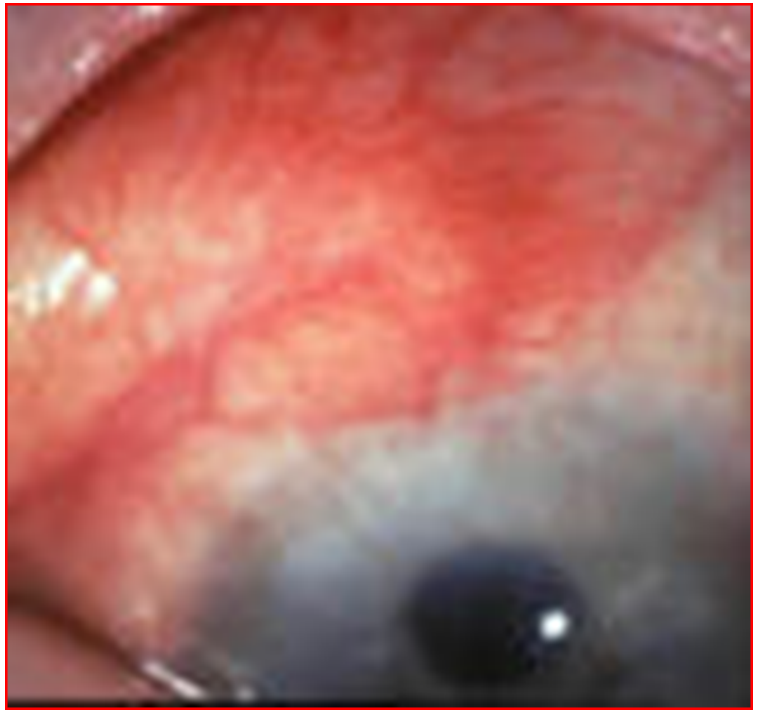
EPIDEMIOLOGY

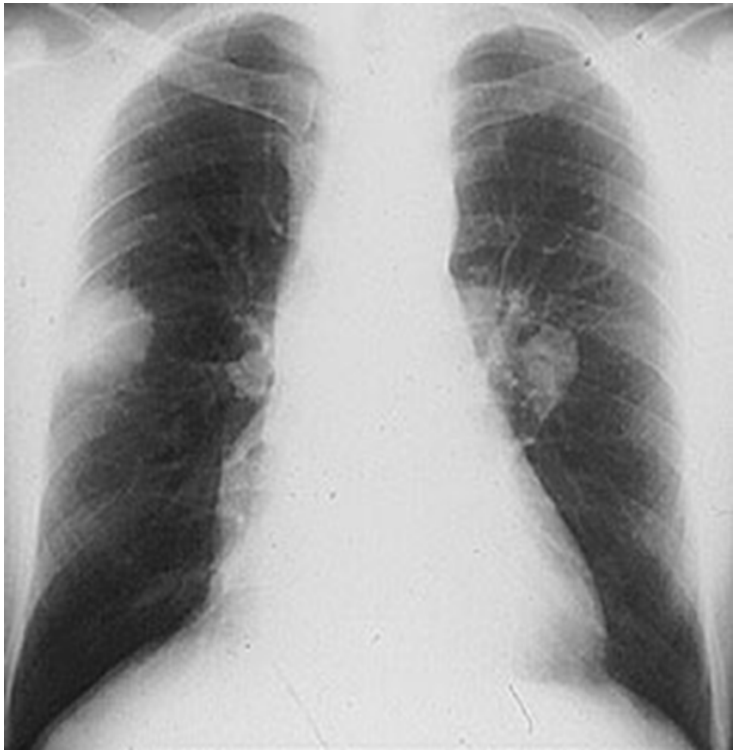
Disease	GEOGRAPHIC AND RACIAL VARIATIONS IN INCIDENCE		Age and Sex Predispositions
	Region or Race With Higher Annual Incidence	Region or Race With Lower Annual Incidence	
Granulomatosis with polyangiitis[54,61–72]	Northern hemisphere and Australia (2-12/million)	Asia and Africa (1-2/million)	Mean age 50-60 years. Female to male ratio 1:1
Microscopic polyangiitis[54,61,64–67,69,71,73]	Asia and Southern Europe (8-18/million)	Northern Europe, North America, Australia (1-7/million)	Mean age 60-70 years. Female to male ratio 1:1.
Eosinophilic granulomatosis with polyangiitis[61,64,66,69,71,73]	Similar across regions (1-2/million)		Mean age 40-50 years. Female to male ratio 1:1.

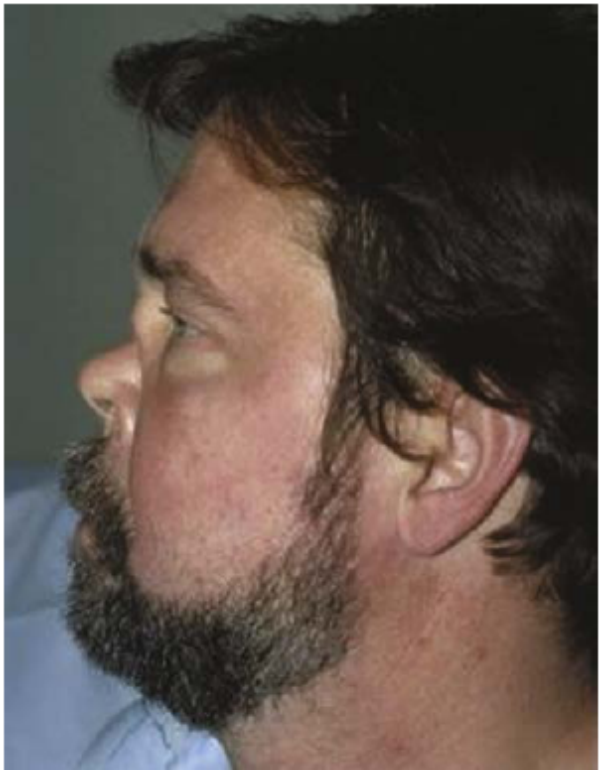
Clinical feature

Organ System	GPA	MPA	EGPA
ENT	83-99	1-20	48-77
Joint/muscle	59-77	14-54+	30-39+
Kidney	66-77	69-100	22-27
Lung	66-85	25-55	51-58
Eye	34-61	1-15	7
Heart	8-25	3-24	16-27
Skin	33-46	11-62	40-57
Peripheral nerve	15-40	13-60	51-76
CNS	8-11	5-12	5-14
GI tract	6-13	3-31	22-31
Constitutional symptoms	58+	67-84	49-68









2022 AMERICAN COLLEGE OF RHEUMATOLOGY / EUROPEAN ALLIANCE OF ASSOCIATIONS FOR RHEUMATOLOGY

CLASSIFICATION CRITERIA FOR **GRANULOMATOSIS WITH POLYANGIITIS**

CONSIDERATIONS WHEN APPLYING THESE CRITERIA

- **These classification criteria should be applied to classify a patient as having granulomatosis with polyangiitis when a diagnosis of small- or medium-vessel vasculitis has been made**
- **Alternate diagnoses mimicking vasculitis should be excluded prior to applying the criteria**

CLINICAL CRITERIA

Nasal involvement: bloody discharge, ulcers, crusting, congestion, blockage, or septal defect / perforation	+3
Cartilaginous involvement (inflammation of ear or nose cartilage, hoarse voice or stridor, endobronchial involvement, or saddle nose deformity)	+2
Conductive or sensorineural hearing loss	+1

LABORATORY, IMAGING, AND BIOPSY CRITERIA

Positive test for cytoplasmic antineutrophil cytoplasmic antibodies (cANCA) or antiproteinase 3 (anti-PR3) antibodies	+5
Pulmonary nodules, mass, or cavitation on chest imaging	+2
Granuloma, extravascular granulomatous inflammation, or giant cells on biopsy	+2
Inflammation, consolidation, or effusion of the nasal/paranasal sinuses, or mastoiditis on imaging	+1
Pauci-immune glomerulonephritis on biopsy	+1
Positive test for perinuclear antineutrophil cytoplasmic antibodies (pANCA) or antimyeloperoxidase (anti-MPO) antibodies	-1
Blood eosinophil count ≥ 1 x10^9/liter	-4

Sum the scores for 10 items, if present. A score of ≥ 5 is needed for classification of GRANULOMATOSIS WITH POLYANGIITIS.

Figure 1 2022 American College of Rheumatology/European Alliance of Associations for Rheumatology classification criteria for granulomatosis with polyangiitis.

2022 AMERICAN COLLEGE OF RHEUMATOLOGY / EUROPEAN ALLIANCE OF ASSOCIATIONS FOR RHEUMATOLOGY

CLASSIFICATION CRITERIA FOR **MICROSCOPIC POLYANGIITIS**

CONSIDERATIONS WHEN APPLYING THESE CRITERIA

- **These classification criteria should be applied to classify a patient as having microscopic polyangiitis when a diagnosis of small- or medium-vessel vasculitis has been made**
- **Alternate diagnoses mimicking vasculitis should be excluded prior to applying the criteria**

CLINICAL CRITERIA

Nasal involvement: bloody discharge, ulcers, crusting, congestion, blockage or septal defect / perforation	-3

LABORATORY, IMAGING, AND BIOPSY CRITERIA

Positive test for perinuclear antineutrophil cytoplasmic antibodies (pANCA) or antimyeloperoxidase (anti-MPO) antibodies ANCA positive	+6
Fibrosis or interstitial lung disease on chest imaging	+3
Pauci-immune glomerulonephritis on biopsy	+3
Positive test for cytoplasmic antineutrophil cytoplasmic antibodies (cANCA) or antiproteinase 3 (anti-PR3) antibodies	-1
Blood eosinophil count ≥ 1 x10^9/liter	-4

Sum the scores for 6 items, if present. A score of ≥ 5 is needed for classification of MICROSCOPIC POLYANGIITIS.

Figure 1 2022 American College of Rheumatology /European Alliance of Associations for Rheumatology classification criteria for microscopic polyangiitis.

2022 AMERICAN COLLEGE OF RHEUMATOLOGY / EUROPEAN ALLIANCE OF ASSOCIATIONS FOR RHEUMATOLOGY

CLASSIFICATION CRITERIA FOR **EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS**

CONSIDERATIONS WHEN APPLYING THESE CRITERIA

- **These classification criteria should be applied to classify a patient as having eosinophilic granulomatosis with polyangiitis when a diagnosis of small- or medium-vessel vasculitis has been made**
- **Alternate diagnoses mimicking vasculitis should be excluded prior to applying the criteria**

CLINICAL CRITERIA

Obstructive airway disease	+3
Nasal polyps	+3
Mononeuritis multiplex	+1

LABORATORY AND BIOPSY CRITERIA

Blood eosinophil count ≥ 1 x10^9/liter	+5
Extravascular eosinophilic-predominant inflammation on biopsy	+2
Positive test for cytoplasmic antineutrophil cytoplasmic antibodies (cANCA) or antiproteinase 3 (anti-PR3) antibodies	-3
Hematuria	-1

Sum the scores for 7 items, if present. A score of ≥ 6 is needed for classification of EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS.

Figure 1 2022 American College of Rheumatology/European Alliance of Associations for Rheumatology Classification Criteria for Eosinophilic Granulomatosis with Polyangiitis.

Induction of Remission for GPA & MPA

Glucocorticoid

- In life- or organ-threatening disease are treated with IV pulse glucocorticoids followed by (prednisone 1 mg/kg/day) that are gradually tapered over many months (e.g., 6 to 18 months)

Induction of remission in GPA & MPA

- **Cyclophosphamide** : alkylating agent
  - until 3-6 months
  - IV: 15 mg/kg
  - Oral: 2 mg/kg/d

- **Rituximab**

Maintenance of remission for GPA & MPA

- **Azathioprine**: 2mg/kg/d
- **MTX**: as efficaceious as AZT
- **MMF**: 2 gr/d (relapses more than AZT)
- **Leflunomide**: 30 mg/d
- **Cotrimoxazole** 800/160: in nasal or upper airway manifestations
- **Rituximab**
- **Glucocorticoids**: 5-10 mg/d
  - Continue for 12-18 months

Treatment of EGPA

- Vasculitis and asthma managed separately
- Initial management of systemic or vasculitic : Corticosteroids
- Do not have (Cr >1.58, Pr>1g, severe GI involvement, cardiomyopathy, or CNS involvement) :Corticosteroids alone
- Immunosuppressive + corticosteroids :severe disease

Treatment of EGPA

Rituximab

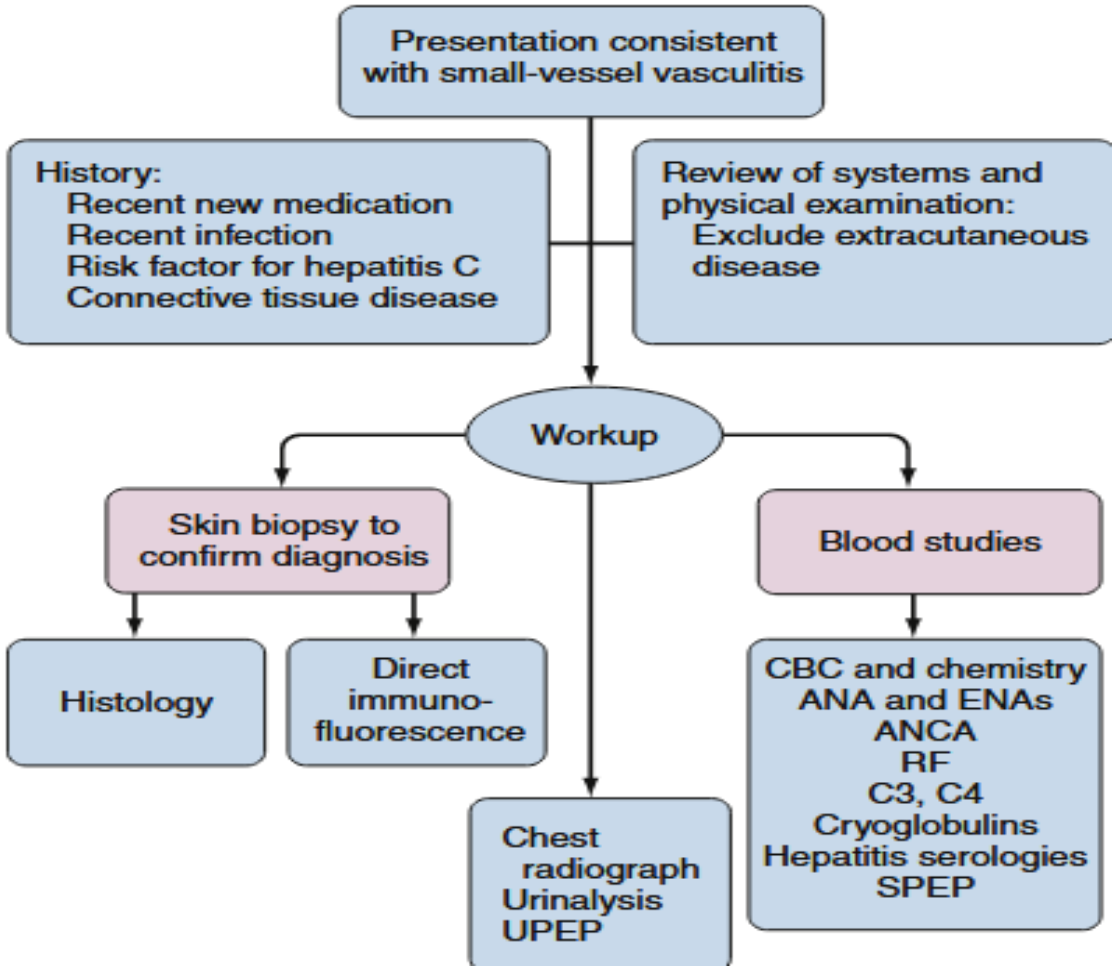
- In refractory

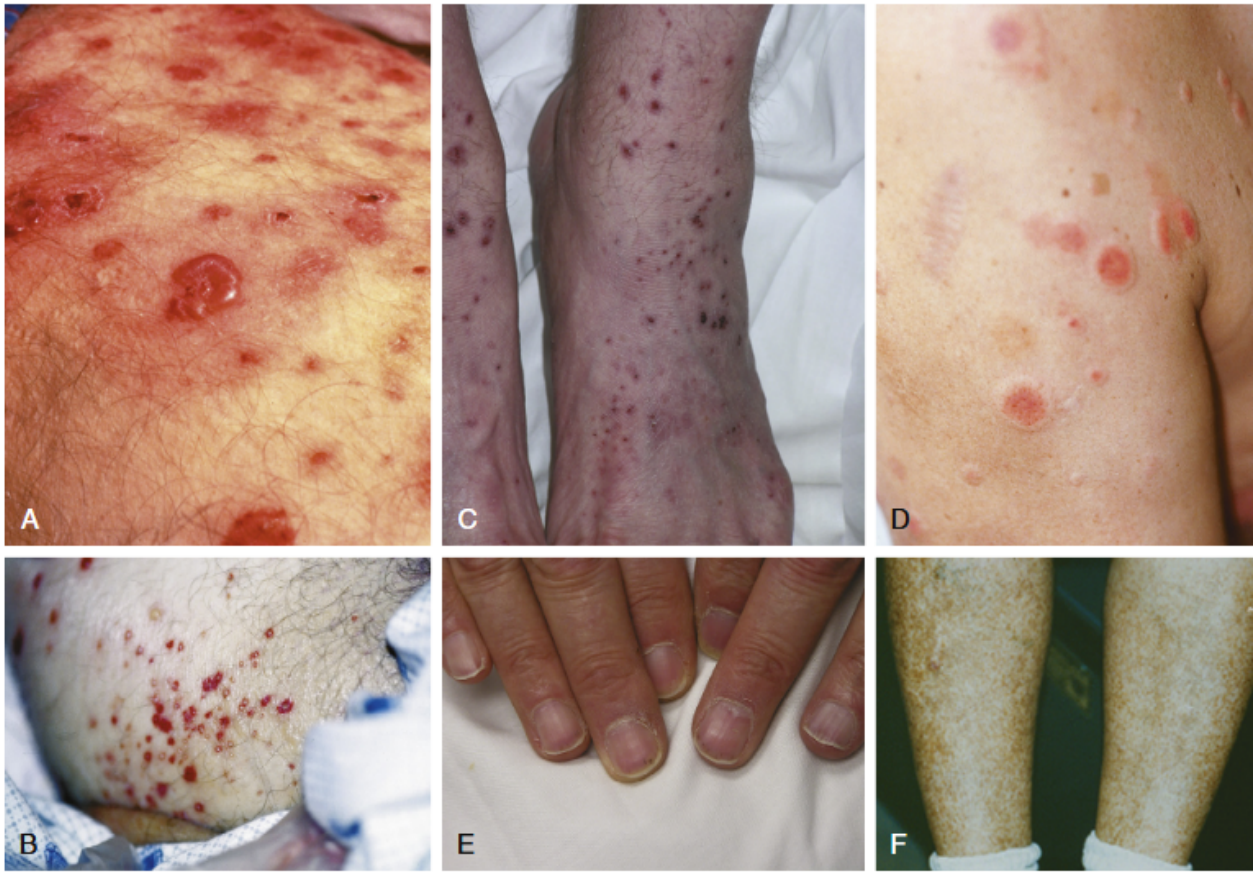
Mepolizumab

- Anti-IL5
- Maintenance non-organ-threatening flares,

Immune Complex–Mediated Small Vessel Vasculitis

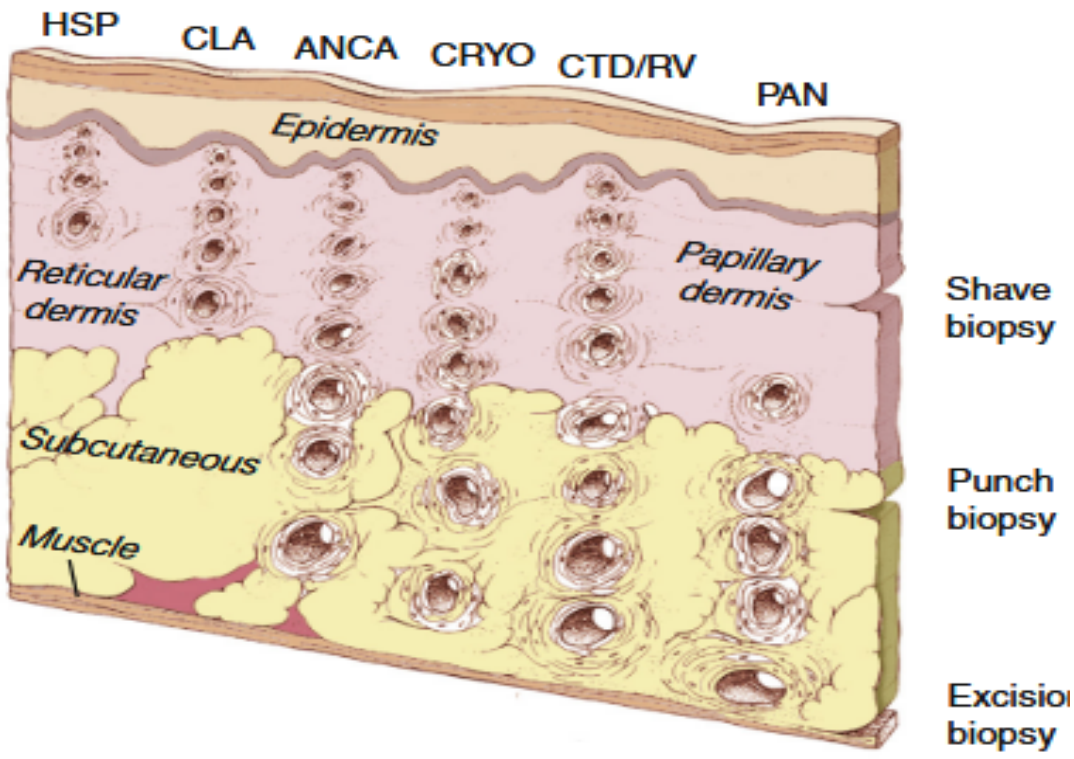
- Hypersensitivity Vasculitis
- Cryoglobulinaemic vasculitis
- IgA vasculitis
- Hypocomplementaemic urticarial vasculitis
- IgG4-Related Vasculitis
- Anti GBM


Presentation consistent with small-vessel vasculitis
History:
Recent new medication
Recent infection
Risk factor for hepatitis C
Connective tissue disease
Review of systems and physical examination:
Exclude extracutaneous disease
Workup
Skin biopsy to confirm diagnosis
Histology
Direct immuno-fluorescence
Chest radiograph
Urinalysis
UPEP
Blood studies
CBC and chemistry
ANA and ENAs
ANCA
RF
C3, C4
Cryoglobulins
Hepatitis serologies
SPEP



• **Fig. 96.3** Cutaneous findings of immune complex–mediated small vessel vasculitis. (A) Vesicles. (B) Pustules. (C) Superficial ulcerations. (D) Urticaria. (E) Splinter hemorrhages. (F) Hyperpigmentation.

Skin biopsy


HSP
CLA
ANCA
CRYO
CTD/RV
PAN
Epidermis
Papillary dermis
Reticular dermis
Subcutaneous
Muscle
Shave biopsy
Punch biopsy
Excisio
biopsy

Hypersensitivity Vasculitis

- Cutaneous leukocytoclastic angiitis
- Drug-induced immune complex vasculitis—under a new category of Vasculitis Associated With Probable Etiology.
- IC deposition in capillaries, postcapillary venules, and arterioles
- Medications, infections, and antibiotics (particularly penicillins and cephalosporins)

Treatment

- Depends on the inciting cause.
- Treatment with glucocorticoids is reserved for patients with extensive disease and can usually be discontinued within several weeks. Patients who experience repeated disease flares may need low-dose glucocorticoids to prevent recurrences.

IgA Vasculitis (Henoch-Schönlein Purpura)

- IgA deposition within blood vessel walls.

- Many IgA vasculitis cases occur after upper respiratory tract infections. Multiple bacterial, viral, and other infectious agents have been suggested as the cause of IgA vasculitis, but the true cause remains unknown.

- Abdominal pain , arthralgia, palpable purpura ,hematuria

- The annual incidence of IgA vasculitis is estimated at 3–26.7/100000 for children and 0.8–1.8/100000 for adults.
- A seasonal pattern with predominant IgA vasculitis onset during the fall and winter is consistently observed for children but not for adults.

- **Treatment :**
- In mild cases of IgA vasculitis, no specific therapy is necessary.
- glucocorticoids or immunosup-pressive agents???

Cryoglobulinemic Vasculitis

TABLE 96.6 Types of Cryoglobulins

Cryoglobulin	RF Positivity	Monoclonality	Associated Diseases
Type I	No	Yes (IgG or IgM)	Hematopoietic malignancy (multiple myeloma, Waldenström's macroglobulinemia)
Type II	Yes	Yes (polyclonal IgG, monoclonal IgM)	Hepatitis C Other infection Sjögren's syndrome SLE
Type III	Yes	No (polyclonal IgG and IgM)	Hepatitis C Other infection Sjögren's syndrome SLE

- Type II and III cryoglobulinemias often present with a triad of signs and symptoms: purpura, arthralgia, and myalgia

- **kidneys** (Membranoproliferative glomerulonephritis)

- **peripheral nerves**

Cryoglobulinemia Treatment

- Anti-viral therapies and B cell depletion approaches with rituximab

- Alveolar hemorrhage or with symptoms of hyperviscosity: plasma exchange

Rheumatoid Vasculitis

- RV is a potentially devastating complication that may involve both medium and small blood vessels and requires the most aggressive therapeutic interventions.(RTX/CYC)

- In the nodular, rheumatoid factor–positive, joint-destructive disease

Hypocomplementemic Urticarial Vasculitis

- HUV is more likely to be a chronic disorder that has certain overlapping features with SLE: low serum complements, autoantibodies, and an interface dermatitis characterized by immunoreactant deposition (complement and immunoglobulins) at the dermal-epidermal junction

- Some cases of HUV respond to therapies commonly used for the treatment of SLE, including low-dose prednisone, hydroxychloroquine, dapsone, or other immunomodulatory agents.

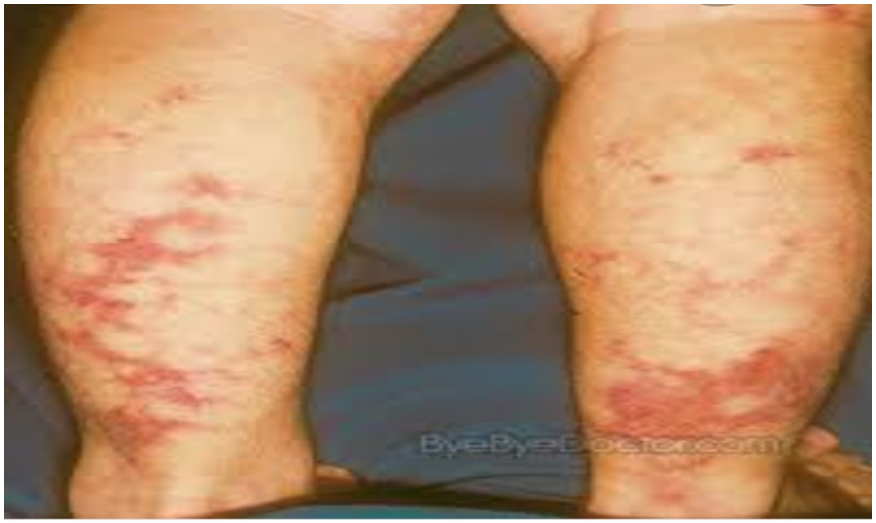
- SLE, COPD, angioedema, and valvular abnormalities are all known to occur in association with this disorder

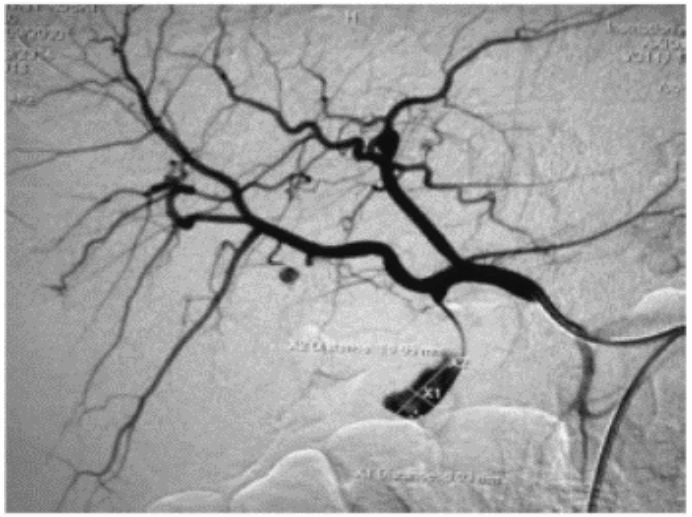
PAN

The annual incidence of PAN in European countries is approximately 2-10 cases per million population.

Prevalence has been estimated between 2 and 31 cases/million

sixth decade of life





Poly arteritis nodosa (PAN), is a multisystem disease characterized by a necrotizing vasculitis of predominantly medium-sized arteries, with few or no deposits.

PAN is a rare and life-threatening disease with heterogeneous clinical presentations.

TABLE 95.3 **Organ Involvement in Polyarteritis Nodosa**

System	Comment	Frequency	Reference(s)
Constitutional	Fever and weight loss (current and previous)	>90%	40
Musculoskeletal	Arthritis, arthralgia, myalgia, or weakness; when muscle is involved, it provides a useful site for biopsy	24%-80%	19,28
Skin	Purpura, nodules, livedo reticularis, ulcers, bullous or vesicular eruptions, and segmental skin edema	44%-50%	19,40,125,126
Cardiovascular	Cardiac ischemia, cardiomyopathy, hypertension	35%	28,35,40
Ear, nose, and throat	No involvement; nasal crusting, sinusitis, and hearing loss suggest an alternative diagnosis such as granulomatosis with polyangiitis	None	
Respiratory	Lung involvement not seen in PAN; abnormal respiratory findings suggest an alternative diagnosis	None	
Abdominal	Pain is an early feature of mesenteric artery involvement; progressive involvement may cause bowel, liver, or splenic infarction, bowel perforation, or bleeding from a ruptured arterial aneurysm; less common presentations include appendicitis, pancreatitis, or cholecystitis as a result of ischemia or infarction; the presence of abdominal tenderness or peritonitis and blood loss on rectal examination should be assessed	33%-36%	19,40
Renal	Vasculitis involving the renal arteries is present in many cases but does not commonly give rise to clinical features; it can present with renal impairment, renal infarcts, or rupture of renal arterial aneurysms; glomerular ischemia may result in mild proteinuria or hematuria, but red cell casts are absent because glomerular inflammation is not a feature; if evidence of glomerular inflammation exists, then an alternative diagnosis such as microscopic polyangiitis or granulomatosis with polyangiitis must be considered; hypertension is a manifestation of renal ischemia causing activation of the renin-angiotensin system	11%-66%	19,40
Nervous system	Mononeuritis multiplex, with sensory symptoms preceding motor deficits; CNS involvement is a less frequent finding and can present with encephalopathy, seizures, and stroke	55%-79%	19,40
Ocular	Visual impairment, retinal hemorrhage, and optic ischemia	Rare	28,35
Other	Breast or uterine involvement is rare; testicular pain from ischemic orchitis is a characteristic feature, albeit an uncommon presentation	Rare	127,128

- **Conventional angiography** is the gold-standard with a sensitivity and specificity of ≈90%.

- **CT-angiography** also has a high yield for demonstrating the aneurysms of PAN.

- The most common CT-angiography finding was **aneurysm**, followed by stenosis/occlusion.

- The most commonly involved arteries were the renal arteries

- The most common visceral abnormality was splenic and renal infarct

- **FDG-PET** may show the increased uptake in the affected arteries

PAN Treatment

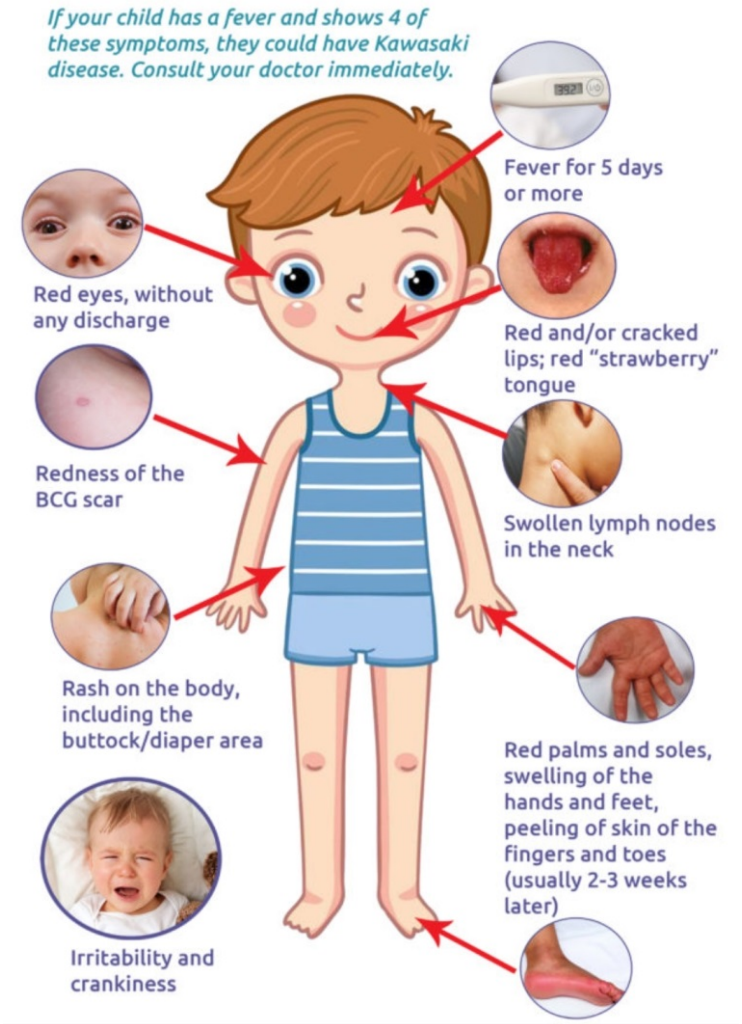
- A combination of **cyclophosphamide** and **glucocorticoids** for remission induction was recommended.

- **For mild cases** 1 mg/kg/day of prednisolone alone and tapering the dose after the control of manifestations is advised.
- **Plasmapheresis** may be considered in catastrophic cases unresponsive to aggressive immunosuppressive therapies and may have a role in the management of HBV-related PAN.

KAWASAKI

In the United States, the estimated incidence of KD among children ,5 years of age is in the range of 19-25 cases per 100,000

Incidence varies by race, being markedly more frequent in East Asia, particularly in Japan.



Large vessel vasculitis

- Giant Cell Arteritis
- POLYMYALGIA RHEUMATICA
- TAKAYASU'S ARTERITIS

GCA

- The most common form of systemic vasculitis in adults.
- The disease affects primarily the thoracic aorta and its major branches, especially the extracranial segments of the carotid artery in patients older than 50 years.
- The most feared complication of GCA is irreversible loss of vision

EPIDEMIOLOGY

- Less than **0.1 to 77** per **100,000**
- Age 79 Y **(50 TO 90 Y)**
- F/M 2/1

Clinical Features

Classic Manifestations

The most common manifestations of GCA are constitutional symptoms, headache, visual symptoms, jaw claudication, and PMR

Headache

is the most common symptom in GCA,The most consistent characteristic is that the patient experiences the headache as something new and unusual. Abnormalities of the temporal artery—including enlargement, nodular swelling, tendernes,or loss of pulse

Visual symptoms

Visual symptoms are common in GCA, especially loss of vision and diplopia.

Causes of vision loss

Causes of vision loss ;AION,retinal ischemia, visual cortex ischemia

Clinical feature of PMR

- Systemic manifestations such as malaise, low-gradefever, and weight loss.
- Arthralgia and myalgias, In most patients, the shoulder girdle is the ˜ become symptomatic; in the remainder, the hip or neck is involved

- Morning stiffness resembling that of RA and “gelling” after inactivi prominent

- Transient synovitis of the knees, wrists and SCJ. The shoulders and hips are covered by

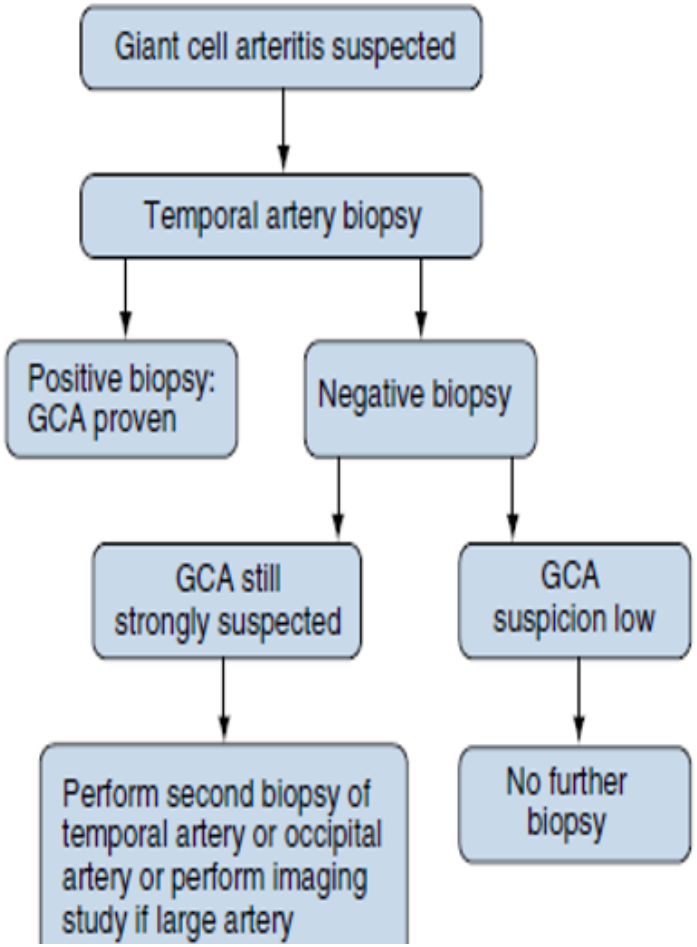
Imaging

- Patients who have signs of subclavian and axillary disease manifested by arm claudication, unequal arm blood pressures, and supraclavicular or axillary bruits may be diagnosed by angiogram, **MRA**, or **CTA.**

- **Color duplex ultrasonography** showed abnormalities of the temporal artery (sensitivity of 93%).

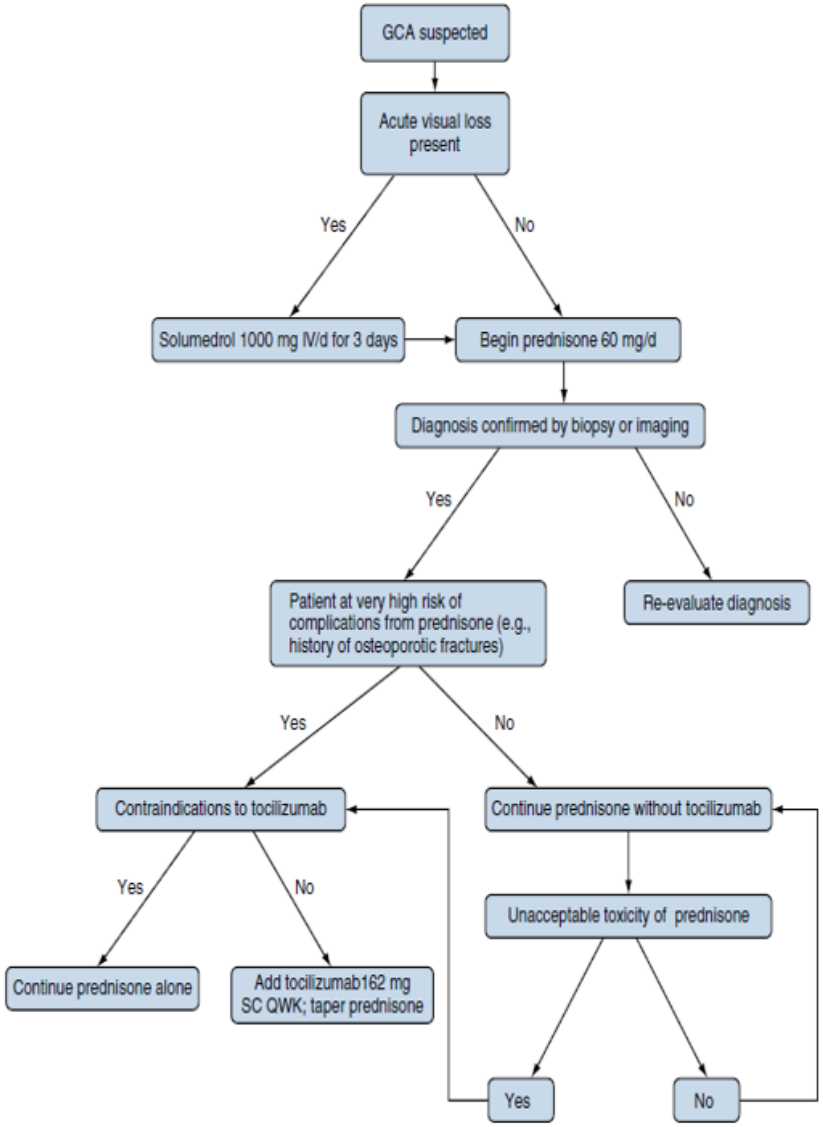
- **PET** has shown promise in detecting occult involvement of the aorta and great vessels.

Ultrasound



Algorithm for diagnosing giant cell arteritis (GCA)

Algorithm for treating giant cell arteritis



Takayasu's Arteritis

- Known as pulseless disease or

- The median patient age at onset is 25 years; however, approximately 25% of cases begin before age 20, and 10% to 20% of patients are seen after age 40 years.

- F/M=8/1

Clinical Features

- The most common presenting vascular symptoms were claudication (35%), reduced or absent pulse , carotid bruit , hypertension , carotidynia , lightheadedness & syncope and asymmetric arm blood pressures
- Headache, which is commonin TAK, does not correlate with carotid or vertebral disease, Which develops in nearly 40% of patients.
- Constitutional, musculoskeletal, and other symptoms of systemic inflammation are also common presenting complaints.

Imaging

1) MRA
2) US
3) CTA

MRA: preferred imaging method for assessing the extent of vascular involvement and damage, both initially and during follow-up

Treatment

- **Corticosteroids are the cornerstone of treatment of active TAK**
- **Criteria for active disease(85% of paitent) include new onset or worsening of two or more of the following:**

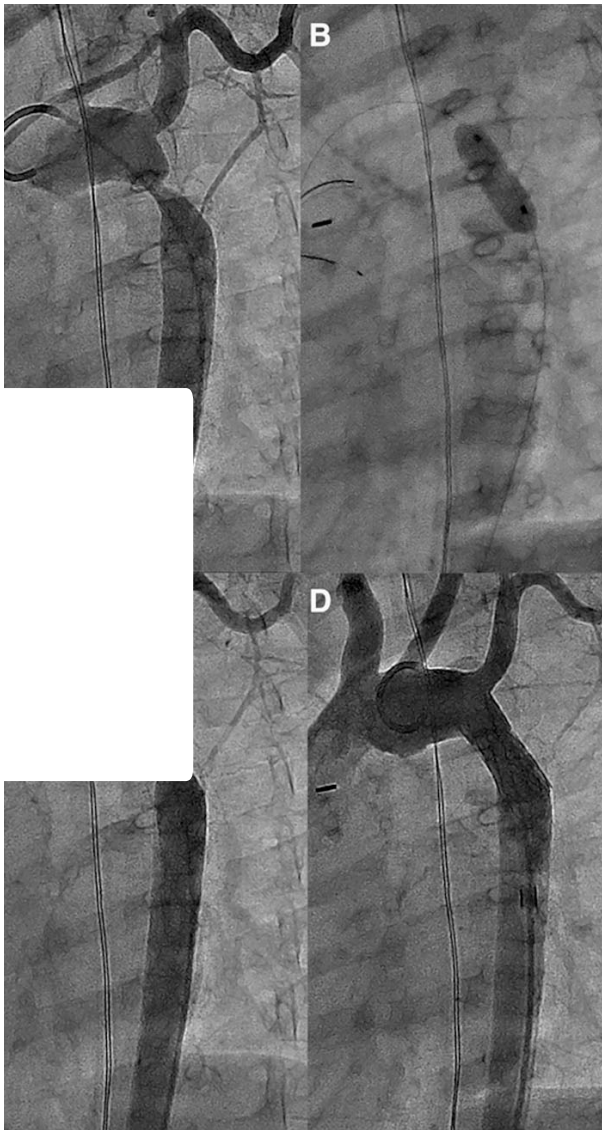
- **Fever or other systemic features**
- **Elevated ESR**
- **Symptoms or signs of vascular ischemia or inflammation**
- **Typical angiographic lesions**

- **Relapses :**
- Increasing the prednisone dose or adding an immunosuppressive agent
- **MTX**
- TNF inhibitors (etanercept and infliximab)
- Tocilizumab
- azathioprine, mycophenolate mofetil, cyclosporine, tacrolimus, leflunomide, and cyclophosphamide
- Tofacitinib: cure & complete remission at 6 and 12 months
- prevent osteoporosis should be initiated early
- Modifiable risk factors for atherosclerosis

Medical therapy rarely reduces or reverses stenotic lesions

bypass surgery yields better results than angioplasty.

autologous vessels give better results than synthetic graft



Behcet's disease

- Multisystem
- Chronic inflammatory
- Attack & remission
- Most prevalent in silk road (middle east, mediterranian,…)
- Turkey: 80-370 in 100000
- Iran: 80 in 100000

- Characterized clinically by oral and genital aphtus lesion

Vascular disorder

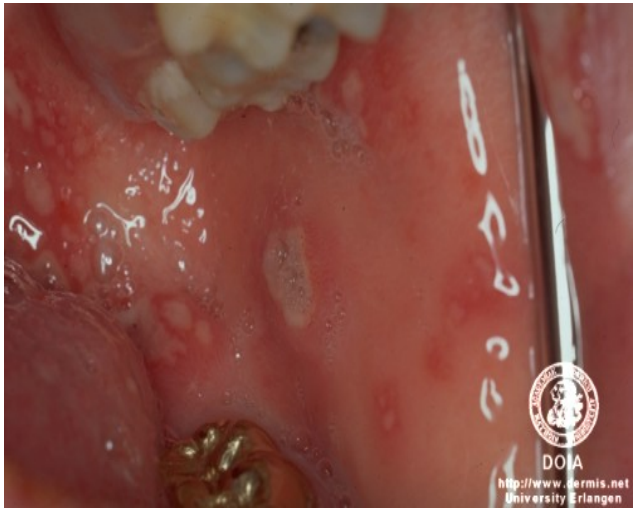
- **Any size**

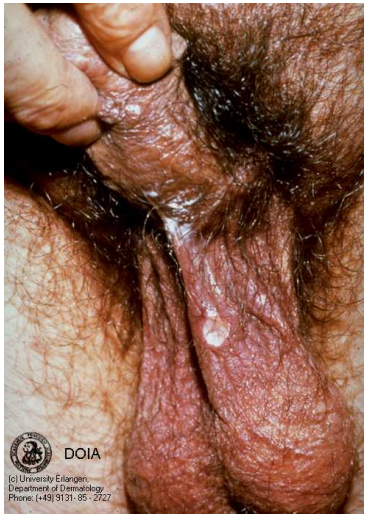
- **Venous: <u>more common</u>**

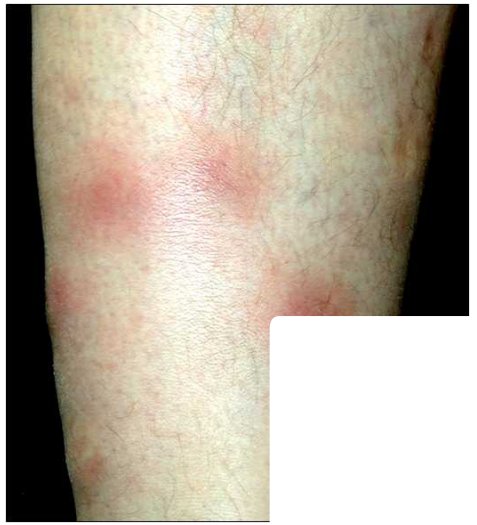
  ***superficial phlebitis, DVT, large vein t**

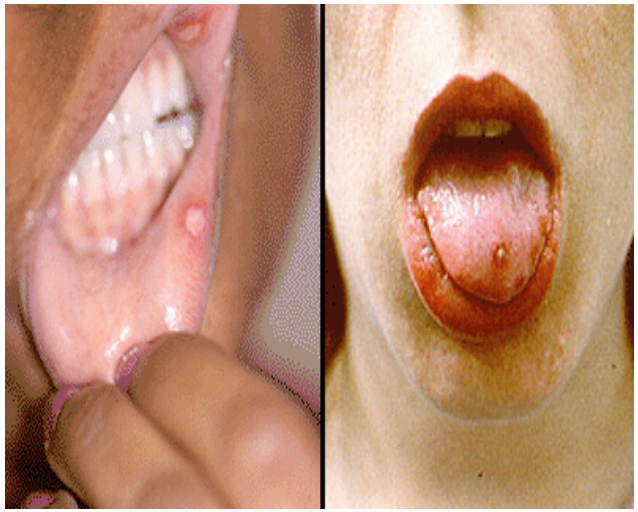
- **Arterial:**

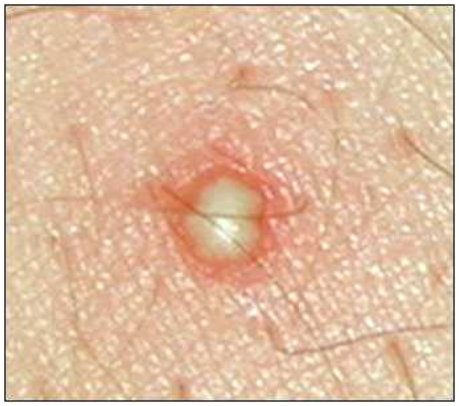
  ***aneurism, thrombosis**

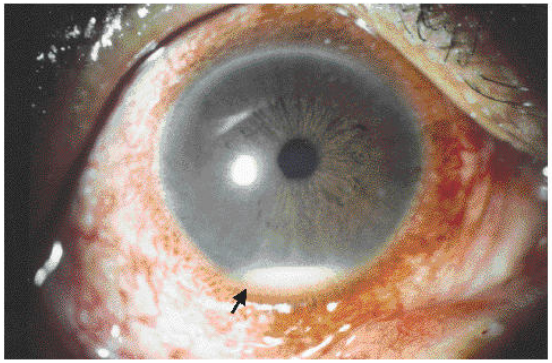

DOIA
http://www.dermis.net
University Erlangen


DOIA
(c) University Erlangen
Department of Dermatology
Phone: (+49) 9131- 85 - 2727









ISG diagnostic criteria

Diagnostic Criteria for Behcet's Disease†

Criterion	Required features
Recurrent oral ulceration	Aphthous (idiopathic) ulceration, observed by physician or patient, with at least three episodes in any 12 month period
Plus any two of the following	
Recurrent genital ulceration	Aphthous ulceration or scarring, observed by physician or patient
Eye lesions	Anterior or posterior uveitis cells in vitreous in slit lamp examination; or retinal vasculitis documented by ophthalmologist
Skin lesions	Erythema nodosum-like lesions observed by physician or patient; papulopustular skin lesions or pseudofolliculitis with characteristic acneiform nodules observed by physician
Pathergy test	Interpreted at 24 to 48 hours by physician

†Adapted from International Study Group for Behcet's Disease. Criteria for diagnosis of Behcet's disease. Lancet 1990; 335:1078.

Treatment

- **Based on organ involvement**

Topical and systemic

Glucocorticoid and immunosuppressive


Thank
you!