PATIENT INFORMATION | **RHEUMATOLOGY**

Understanding Vasculitis

An evidence-based guide for patients in Pakistan — what vasculitis is, the main forms, how it is diagnosed, and how modern treatment can control inflammation and protect your organs.

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At a glance

> 20	Any vessel	All ages	80 – 90%
Distinct types of vasculitis	Aorta to capillary, any organ	Children to elderly	5-year survival with treatment

Vasculitis is the medical term for inflammation of blood vessels. When the lining of a vessel becomes inflamed, it can swell, narrow, weaken, or block — disrupting the blood supply to the tissues beyond. Because blood vessels reach every organ in the body, vasculitis can produce an astonishing variety of symptoms, from skin rashes and sinusitis to kidney failure, stroke, or sudden loss of vision. The vasculitides are uncommon, but they are **medical emergencies** when major organs are involved. With early diagnosis and modern treatment, most patients now achieve sustained remission.

What is vasculitis?

Vasculitis can be **primary** (the inflammation is the disease itself, with no other cause) or **secondary** (occurring as part of another illness such as rheumatoid arthritis, systemic lupus, hepatitis B or C infection, or as a reaction to a drug). The modern classification — agreed at the **2012 International Chapel Hill Consensus Conference** — groups the vasculitides chiefly by the **size of vessel** predominantly affected:

Group	Main conditions
Large-vessel vasculitis (aorta and its major branches)	**Giant cell arteritis (GCA)** — over 50 years, affects branches of the external carotid **Takayasu arteritis** — typically young women under 40, affects the aorta and its branches
Medium-vessel vasculitis	**Polyarteritis nodosa (PAN)** — often linked to hepatitis B **Kawasaki disease** — children under 5 years, may affect coronary arteries
Small-vessel vasculitis (arterioles, capillaries, venules)	**ANCA-associated vasculitis (AAV):** • **Granulomatosis with polyangiitis (GPA)** — formerly Wegener's • **Microscopic polyangiitis (MPA)** • **Eosinophilic granulomatosis with polyangiitis (EGPA)** — formerly Churg–Strauss **Immune-complex vasculitis:** IgA vasculitis (Henoch–Schönlein purpura), cryoglobulinaemic vasculitis, hypocomplementaemic urticarial vasculitis

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Group	Main conditions
Variable-vessel vasculitis	**Behçet's disease** — affects any vessel size; common along the historic Silk Road, including Pakistan **Cogan's syndrome** — eye and inner-ear involvement
Single-organ vasculitis	Cutaneous leucocytoclastic angiitis, primary central nervous system vasculitis, isolated aortitis
Secondary vasculitis	Vasculitis associated with lupus, rheumatoid arthritis, hepatitis B/C, malignancy, or drugs

Incidence and epidemiology

Taken together, the primary vasculitides affect approximately **40 – 50 people per million per year**. Different types of vasculitis have strikingly different geographic and ethnic patterns: **giant cell arteritis** is most common in Northern Europeans aged over 50; **Takayasu arteritis** and **Kawasaki disease** are commoner in East and South Asian populations; and **Behçet's disease** follows the historic Silk Road from the Mediterranean through Turkey, Iran, and Central Asia into Pakistan, China, and Japan.

South Asian and Pakistani context

Population-level vasculitis data from Pakistan remain limited, but several important observations have been made:

• Pakistan lies along the historic **Silk Road belt** in which **Behçet's disease** is significantly more prevalent than in Western populations. A major Pakistani tertiary-centre series (Hashmi *et al.*, *Pak J Med Sci* 2019) found **oral aphthous ulcers** in 100%, **genital ulcers** in 90%, and **ocular involvement** in 80% of Pakistani Behçet's patients — higher rates than in many international cohorts.
• **Takayasu arteritis** is well-recognised in Pakistani young women, often presenting with hypertension that is difficult to control, unequal pulses, or unexplained stroke; it is frequently misdiagnosed as essential hypertension for years.
• **ANCA-associated vasculitis** is increasingly recognised in Pakistani patients, though under-diagnosed; the combination of sinus disease, lung infiltrates, and kidney dysfunction in any adult should prompt urgent rheumatology and nephrology assessment.
• In regional series, neighbouring Iran reports **Behçet's disease as the single commonest vasculitis** (63.6% of all primary vasculitis cases) — a pattern likely reflected in Pakistan.

The major diagnostic challenges for Pakistani patients are delay (symptoms attributed to infection, hypertension, or other diseases for months or years) and limited access to specialist rheumatology and immunology services outside major cities. Vasculitis should be considered in any patient with unexplained systemic illness, recurrent ulcers, sinus disease, or organ dysfunction.

How does vasculitis develop?

The mechanisms differ between the various types of vasculitis, but the common end-point is the same: inflammation damages the wall of the blood vessel, leading to **narrowing**, **occlusion**, **aneurysm formation**, or **rupture**. Several distinct immune mechanisms have been identified:

Mechanism	Main examples and explanation
Granulomatous inflammation	T-cells and macrophages form granulomas inside the vessel wall, as in **giant cell arteritis**, **Takayasu arteritis**, and **GPA**. The vessel wall thickens and the lumen narrows.
ANCA-driven neutrophil activation	Antineutrophil cytoplasmic antibodies (ANCA) — **c-ANCA / anti-PR3** or **p-ANCA / anti-MPO** — activate neutrophils, which then damage small vessels. Characteristic of **GPA**, **MPA**, and **EGPA**.
Immune complex deposition	Antigen-antibody complexes lodge in small-vessel walls, activating complement. Examples: **IgA vasculitis**, **cryoglobulinaemic vasculitis** (often hepatitis C-related).
Variable / heterogeneous	**Behçet's disease** is unique — it affects vessels of any size and any type (arteries and veins), driven by a complex interplay of innate immunity, neutrophils, and HLA-B*51 genetic predisposition.

*Understanding the mechanism guides treatment: granulomatous and ANCA-associated forms respond to immunosuppression (steroids, rituximab, cyclophosphamide); immune-complex disease may require treating the underlying trigger (e.g. hepatitis); and Behçet's disease often responds to **colchicine** and **biologics** such as anti-TNF agents.*

Symptoms — what to look out for

Because vasculitis can affect any organ, symptoms are extraordinarily varied. There are, however, certain **constitutional** features common to almost all forms, and characteristic **organ-specific** patterns that should raise suspicion:

Constitutional symptoms (common to most vasculitides)

- Persistent fever of unknown cause, particularly with raised inflammatory markers.
- Severe fatigue, weight loss, night sweats, and loss of appetite.
- Generalised muscle and joint aches, often without obvious swelling.

Organ-specific clues by type of vasculitis

Vasculitis	Key clinical clues
Giant cell arteritis	New severe headache (often temporal), **jaw claudication** (pain on chewing), scalp tenderness, **sudden visual loss** in one eye, polymyalgia rheumatica.
Takayasu arteritis	Young woman with **unequal pulses**, asymmetric blood pressure between arms, **arterial bruits**, claudication of arms or legs, dizziness, refractory hypertension.
Polyarteritis nodosa	Mononeuritis multiplex (sudden weakness of a single nerve, e.g. foot drop), abdominal pain, testicular pain, livedo reticularis, hypertension, raised creatinine.
GPA	**Sinusitis, nasal crusting, bloody nasal discharge**, saddle-nose deformity, deafness, cough, haemoptysis, breathlessness, and **glomerulonephritis**.
MPA	Rapidly progressive glomerulonephritis (raised creatinine, blood and protein in the urine), alveolar haemorrhage, peripheral neuropathy. Generally **no granulomas**.
EGPA	**Adult-onset asthma**, nasal polyps, very high **blood eosinophils**, peripheral neuropathy, skin nodules, cardiac involvement.
IgA vasculitis	Palpable purpuric rash on legs and buttocks, joint pain, abdominal pain, and microscopic blood in the urine — commonest in children but can occur in adults.
Behçet's disease	Recurrent **painful oral ulcers**, **genital ulcers**, uveitis, acneiform skin lesions, deep-vein thrombosis, pathergy (skin pustules at needle-prick sites).

Red-flag features needing same-day assessment

- **Sudden loss of vision** in one or both eyes, double vision, or new severe headache in anyone over 50 — possible giant cell arteritis. **This is a medical emergency**; high-dose steroids must be started without delay to prevent blindness.
- **Coughing up blood**, breathlessness with new lung infiltrates, or a rapid fall in haemoglobin — possible alveolar haemorrhage.
- **Rapidly rising creatinine, blood or protein in the urine**, or oliguria — possible rapidly progressive glomerulonephritis.
- A new painful, purpuric rash with abdominal pain or joint swelling.
- Sudden weakness or numbness in a single limb — possible mononeuritis multiplex.
- Severe abdominal pain, bloody diarrhoea, or new chest pain.

How is vasculitis diagnosed?

There is no single test for vasculitis. Diagnosis is built from a careful history and examination, blood and urine tests, imaging, and — wherever possible — a **biopsy** of an affected vessel or organ to confirm the diagnosis histologically.

Tests your rheumatologist may arrange

- **Full blood count, ESR, CRP, urea, creatinine, electrolytes, liver function** — to assess inflammation, organ function, and screen for anaemia or eosinophilia.
- **Urinalysis** at every visit — blood, protein, and red-cell casts in the urine are early warning signs of kidney involvement.
- **ANCA testing** (c-ANCA/PR3 and p-ANCA/MPO) — central to diagnosing GPA, MPA, and EGPA.
- **Autoimmune screen** — ANA, anti-dsDNA, complement, rheumatoid factor, anti-CCP to identify secondary vasculitis.
- **Infection screen** — hepatitis B and C, HIV; cryoglobulins where indicated.
- **Chest X-ray and HRCT chest** — to detect nodules, cavities, or alveolar haemorrhage.
- **Imaging of large vessels** — temporal artery **ultrasound** or **biopsy** for GCA; **MRA, CTA, or PET-CT** for Takayasu arteritis and aortitis.
- **Biopsy** — of skin, kidney, temporal artery, or lung as appropriate. Histology remains the gold standard wherever feasible.
- **HLA-B*51 testing** — strongly associated with Behçet's disease; **pathergy testing** may also be performed.

Treatment — a modern, target-driven approach

Treatment has improved dramatically over the past two decades. Most modern regimens follow the **2022 EULAR recommendations** for ANCA-associated vasculitis, the **2021 ACR/Vasculitis Foundation guidelines** for GCA, Takayasu arteritis, PAN, and Kawasaki disease, and the **2018 EULAR recommendations** for Behçet's disease. Treatment is divided into two phases: **remission induction** (urgent suppression of active inflammation) and **remission maintenance** (long-term prevention of relapse with the lowest possible steroid dose).

Stepped care across phases

Phase	Approach	Examples
Induction	Rapid, high-intensity immunosuppression to stop ongoing damage.	**High-dose oral or intravenous glucocorticoids** (typically 0.5–1 mg/kg/day prednisolone, or methylprednisolone pulses) combined with: • **Rituximab** (anti-CD20) — first-line for GPA and MPA • **Cyclophosphamide** — for severe, organ-threatening disease or where rituximab is unsuitable • **Plasma exchange** — in life-threatening alveolar haemorrhage or rapidly progressive renal failure • **Avacopan** (C5a receptor inhibitor) — newer agent that allows substantial steroid reduction in GPA/MPA
Maintenance	Lower-intensity immunosuppression to prevent relapse and protect organs.	**Rituximab** (preferred for AAV maintenance), **azathioprine**, **methotrexate**, or **mycophenolate mofetil**. Prednisolone tapered to ≤ 5 mg/day by 4–5 months.
Specific agents for specific diseases	Targeted biologics for selected conditions.	• **Tocilizumab** (IL-6 inhibitor) — giant cell arteritis • **Mepolizumab** (anti-IL-5) — relapsing/refractory EGPA • **Colchicine, apremilast, anti-TNF agents** — Behçet's disease • **Antiviral therapy** — hepatitis-B-associated PAN, hepatitis-C cryoglobulinaemia • **IV immunoglobulin (IVIG) and aspirin** — Kawasaki disease
Supportive care	Reduces long-term morbidity from disease and treatment.	Pneumocystis prophylaxis (co-trimoxazole) while on heavy immunosuppression, vaccination before treatment, bone protection, cardiovascular risk reduction, and contraception while on teratogenic drugs.

What you can do yourself

- **Take every dose of medication on time.** Stopping or missing immunosuppressants is the single commonest cause of relapse.
- **Stay up to date with vaccinations** — pneumococcal, annual influenza, COVID-19, and (where indicated) shingles. Avoid live vaccines while on rituximab, high-dose steroids, or strong immunosuppressants.
- **Stop smoking** — smoking worsens vasculitis and dramatically raises cardiovascular and thrombotic risk.
- **Watch for infection** — fever, productive cough, urinary symptoms, or shingles rash while on immunosuppression deserves prompt medical review.
- **Protect your bones** — adequate calcium and vitamin D, weight-bearing exercise, and (where appropriate) bisphosphonates to prevent steroid-induced osteoporosis.
- **Monitor blood pressure and lipids** — accelerated cardiovascular disease is a long-term complication of both vasculitis and its treatment.
- **Plan pregnancies carefully.** Several vasculitis medications (cyclophosphamide, methotrexate, mycophenolate) must be stopped before conception. Discuss pregnancy plans with your rheumatologist well in advance.

What is the outlook?

Once almost uniformly fatal, the major systemic vasculitides are now treatable chronic diseases. With modern induction therapy, **5-year survival in ANCA-associated vasculitis exceeds 80–90%**, and most

patients achieve sustained remission. Outcomes depend heavily on three things: **early diagnosis**, **prompt initiation of immunosuppression**, and **regular long-term monitoring** for relapse, infection, and treatment side effects. Late or missed diagnosis remains the principal cause of avoidable disability — particularly blindness in giant cell arteritis, end-stage kidney disease in ANCA vasculitis, and stroke or aneurysm rupture in Takayasu arteritis.

When to contact your rheumatologist promptly

- New visual disturbance, headache, jaw or scalp pain — possible flare of GCA.
- Frothy urine, ankle swelling, or rising blood pressure — possible renal flare.
- New cough, breathlessness, or coughing up blood.
- Fever above 38°C, particularly while on rituximab, cyclophosphamide, or high-dose steroids — urgent assessment to exclude infection.
- New rash, mouth or genital ulcers, painful red eye, or numbness/weakness in a limb.
- Unexpected pregnancy — many vasculitis drugs require urgent adjustment.

Frequently asked questions

Question	Answer
Is vasculitis hereditary?	Most forms of vasculitis are **not** directly inherited. Some — notably Behçet's disease — have a strong genetic predisposition (HLA-B*51), but most patients have no affected relatives.
Can vasculitis be cured?	Most vasculitides cannot be “cured” in the strictest sense, but modern treatment can induce **complete remission** lasting many years. Some patients can eventually stop all immunosuppression, while others need lifelong therapy.
Will I need treatment forever?	Many patients can taper off active treatment after several years in remission, particularly with ANCA-associated vasculitis and Behçet's disease. Others — especially with Takayasu arteritis or relapsing disease — need long-term low-dose maintenance.
Is rituximab safe?	Yes, in the vast majority of patients. The main risk is infection, particularly while B-cells are depleted. Pre-treatment screening (hepatitis B, immunoglobulins) and vaccination are essential.
Can I fast during Ramadan?	Most stable patients in remission can fast safely, but those on diuretics, with active kidney involvement, or on multiple medications need individualised advice. Discuss timing of doses with your rheumatologist before Ramadan begins.
Can I have children?	Yes. Fertility is usually preserved with modern treatment, although cyclophosphamide can reduce ovarian and testicular function. Pregnancy should be planned during a period of stable remission, with medication reviewed in advance.

Key references and further reading

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Speak to a specialist

Vasculitis is treatable, and the earlier it is diagnosed the better the outcome — book a consultation today.

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Important disclaimer. This leaflet provides general information for patients and does not replace individual medical advice from a qualified rheumatologist. Treatment decisions must take into account your full medical history, examination findings, and investigations. Please discuss any concerns directly with Dr Saira Khan or your treating physician.

Reviewed by Dr Saira Elaine Anwer Khan, May 2026.
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