PATIENT INFORMATION | **RHEUMATOLOGY**

Understanding Lupus (SLE)

An evidence-based guide for patients in Pakistan — what lupus is, how it is diagnosed, and how modern treatment can control the disease and protect your organs for life.

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At a glance

20 – 150	9 : 1	15 – 45	~90%
Cases per 100,000 in Asia	Female : male ratio	Typical age at onset (years)	10-year survival with modern care

Systemic Lupus Erythematosus (SLE) — usually shortened to **lupus** — is a chronic autoimmune disease in which the immune system mistakenly attacks the body's own tissues. It can affect almost any organ: the skin, joints, kidneys, blood, lungs, heart, and brain. The course is typically one of **flares** (periods of active disease) and **remissions** (quiet periods). Lupus is not contagious, not cancer, and not inherited in a simple way. With modern treatment, most patients live full, productive lives.

What is lupus?

Lupus belongs to a family of **connective tissue diseases**. In a healthy person, the immune system distinguishes between the body's own cells (“self”) and harmful invaders such as bacteria and viruses (“non-self”). In lupus this distinction breaks down. The body produces **autoantibodies** — most characteristically antinuclear antibodies (ANA) and anti-double-stranded DNA antibodies (anti-dsDNA) — which form immune complexes that deposit in tissues, activate complement, and drive inflammation in multiple organs.

Different types of lupus

Type	Description
Systemic lupus erythematosus (SLE)	The classical form, in which inflammation can affect any organ. The focus of this leaflet.
Cutaneous lupus erythematosus	Skin-limited disease. Includes **discoid lupus** (scaly disc-shaped plaques) and **subacute cutaneous lupus**. May or may not progress to systemic involvement.
Drug-induced lupus	A lupus-like illness triggered by certain medications (e.g. hydralazine, procainamide, isoniazid, minocycline). Usually resolves within weeks of stopping the drug.
Neonatal lupus	A rare condition in babies of mothers with anti-Ro/SSA antibodies. Mostly transient; the main concern is congenital heart block.

Incidence and epidemiology

Lupus is a global disease but its frequency and severity vary markedly by ethnicity. Reported prevalence ranges from **48–366 per 100,000** in North America, **29–210 per 100,000** in Europe, and **20–103 per 100,000** in Asia. Lupus is up to **three times more common** in people of South Asian, African, Afro-Caribbean, and Hispanic ancestry than in Caucasians, and it also tends to be more severe in these groups — particularly with regard to kidney involvement.

South Asian and Pakistani context

Pakistani patients with lupus show a clinical profile that differs in important ways from Western cohorts, with notably higher rates of renal and neuropsychiatric involvement:

• The landmark Pakistani cohort from the **Aga Khan University Hospital, Karachi** (Rabbani *et al.*, *Lupus* 2004, n = 196) reported a **female-to-male ratio of 7.2:1**, a mean age at presentation of **31 years**, and a mean diagnostic delay of **2.8 years**.
• In that series, **renal involvement** was documented in 33% and **neuropsychiatric features** in 26% — both higher than reported Caucasian rates. **ANA was positive in 86%** and **anti-dsDNA in 74%** of patients.
• A 2023 systematic review and meta-analysis of 13 Pakistani studies (Khan *et al.*, *Front Psychiatry*) estimated the prevalence of **neuropsychiatric lupus at 30.4%**, with cognitive dysfunction the commonest manifestation.
• Infection, renal disease, seizures, and thrombocytopenia were identified as the strongest predictors of poor prognosis in Pakistani patients.

Lupus is significantly under-diagnosed in Pakistan due to non-specific early symptoms, limited rheumatology services outside major cities, and frequent misattribution to other illnesses. A young woman with unexplained fatigue, joint pain, hair loss, and mouth ulcers deserves an urgent rheumatology opinion.

Who is at higher risk?

- **Female sex** — women account for around 90% of cases; the female-to-male ratio peaks between menarche and menopause, when oestrogen levels are highest.
- **Age 15 to 45 years** — the typical window of onset, although lupus can occur in children and the elderly.
- **South Asian, African, Afro-Caribbean, or Hispanic ancestry** — disease is more common and tends to be more severe.
- **Family history** — a first-degree relative with lupus or another autoimmune disease increases the risk modestly; identical twin concordance is approximately 25%.
- **Environmental triggers** — ultraviolet (sunlight) exposure, Epstein-Barr virus infection, smoking, silica dust, and certain medications.
- **Hormonal factors** — flares are more frequent during pregnancy and the post-partum period, and around the menopause.

How does lupus develop? — the science explained simply

Lupus arises when a genetically predisposed individual encounters an environmental trigger that switches on a self-perpetuating immune response against the body's own cells. The process unfolds in several linked steps:

Step	What happens in your body
1. Genetic susceptibility	Over 100 genetic variants — including in **HLA, complement (C1q, C2, C4)**, and **interferon-pathway genes** — predispose to lupus. Genetics alone are not enough.
2. Environmental trigger	**Ultraviolet light**, viral infection (especially Epstein-Barr virus), smoking, or certain drugs cause cellular injury and release of nuclear material into the circulation.
3. Loss of self-tolerance	B-cells, T-cells, and plasmacytoid dendritic cells fail to ignore self-antigens. **Type I interferon** (IFN-α) drives the disease forward — the target of newer biologics.
4. Autoantibody production	B-cells produce autoantibodies (ANA, anti-dsDNA, anti-Sm, anti-Ro, anti-La, antiphospholipid antibodies) which form **immune complexes**.
5. Tissue damage	Immune complexes deposit in skin, joints, kidneys, and blood vessels, activating complement and recruiting inflammatory cells — producing the multi-organ features of lupus.

*Modern treatments interrupt this pathway at several points: **hydroxychloroquine** dampens immune activation, **belimumab** targets B-cells, and **anifrolumab** blocks type I interferon.*

Symptoms — what to look out for

Lupus is famously called “the great mimic” because its symptoms can resemble many other illnesses. No two patients are exactly alike, and the same patient may experience different symptoms during different flares. The most important features are described below by organ system:

Organ system	Typical features
Constitutional	**Fatigue** (the single most common symptom), fever, weight loss, and lymph node swelling. Fatigue is often disproportionate to other findings and disabling.
Skin and mucosa	**Butterfly (malar) rash** across the cheeks and nose, **discoid plaques**, **photosensitivity** (rash or flare triggered by sunlight), painless **oral or nasal ulcers**, and patchy **hair loss (alopecia)**.
Joints and muscles	**Arthritis** in the small joints of the hands, wrists, and knees — typically painful and stiff but, unlike rheumatoid arthritis, **non-erosive**. Generalised muscle aches (myalgia) are common.
Kidneys (lupus nephritis)	Often silent in early stages. Look for **frothy urine** (proteinuria), **ankle swelling**, high blood pressure, or blood in the urine. Affects up to 50% of patients and is the single most important determinant of long-term prognosis.
Blood	**Anaemia**, low white-cell count (leucopenia), low platelets (thrombocytopenia), and lymph node enlargement. Bleeding or unusual bruising warrants prompt review.
Lungs and heart	**Pleurisy** (sharp chest pain on breathing in), **pericarditis**, and rarely lupus pneumonitis or pulmonary embolism. Accelerated atherosclerosis increases long-term cardiovascular risk.
Nervous system	Headaches, mood disturbance, cognitive difficulties (**“lupus fog”**), seizures, stroke, or peripheral neuropathy. Neuropsychiatric lupus affects approximately 30% of Pakistani patients.
Vascular	**Raynaud's phenomenon** — fingers turning white, then blue, then red in the cold. Antiphospholipid antibodies (in ~30%) increase the risk of blood clots and miscarriage.

Red-flag features needing urgent assessment

- Severe headache with vomiting, confusion, or new seizure — possible neuropsychiatric lupus or cerebral venous thrombosis.
- Sudden breathlessness or chest pain — exclude pulmonary embolism, pleurisy, or pericardial effusion.
- Frothy urine, swelling around the eyes, or rapidly rising blood pressure — possible lupus nephritis flare.
- High fever in a patient on immunosuppressive treatment — exclude infection.
- Severe abdominal pain — lupus mesenteric vasculitis or pancreatitis is rare but serious.

How is lupus diagnosed?

There is no single test for lupus. Diagnosis rests on the **combination of clinical features and characteristic autoantibodies**. The most widely used framework is the **2019 EULAR/ACR classification criteria**, which require a positive ANA (≥1:80) as the entry criterion, plus a weighted score of additional clinical and immunological features.

Tests your rheumatologist will arrange

Test	Why it matters
Antinuclear antibody (ANA)	Positive in >95% of SLE patients — the entry criterion. A negative ANA makes SLE very unlikely.
Anti-dsDNA and anti-Sm antibodies	Highly specific for lupus. Anti-dsDNA levels often track with disease activity, especially in lupus nephritis.
Anti-Ro/SSA, anti-La/SSB, anti-RNP	Help define lupus subsets. Anti-Ro is important in pregnancy planning (risk of neonatal lupus and congenital heart block).
Complement (C3, C4)	Low complement levels suggest active disease, particularly active lupus nephritis.
Antiphospholipid antibodies	Lupus anticoagulant, anti-cardiolipin, and anti-β2-glycoprotein-I. Positive in ~30%; predict thrombosis and pregnancy loss.
Full blood count, ESR, CRP	Look for cytopenias. ESR is usually raised; CRP is often *normal* in active lupus (a sharp CRP rise suggests infection instead).
Urine analysis & urine protein:creatinine ratio	Essential at every visit. Proteinuria or red-cell casts mandate urgent renal investigation.
Renal biopsy	The gold standard when nephritis is suspected. Defines the histological class (I–VI) and guides treatment.

Treatment — a modern, target-driven approach

Treatment follows the **2023 EULAR recommendations** for SLE management. The treat-to-target strategy aims for **remission** or, at minimum, **low disease activity**, with the lowest possible glucocorticoid dose. Modern care has transformed lupus from a frequently fatal disease — **10-year survival has risen from approximately 50% in the 1950s to over 90% today**.

Core principles

- **Hydroxychloroquine for everyone.** Unless contraindicated, every lupus patient should take hydroxychloroquine — it reduces flares, organ damage, thrombosis, and mortality.
- **Minimise glucocorticoids.** The EULAR target for long-term maintenance is ≤5 mg/day prednisolone equivalent. Steroids are bridging therapy, not maintenance.

- **Use immunosuppressants and biologics earlier.** Adding belimumab or anifrolumab should be considered for any patient unable to taper steroids below 5 mg/day.
- **Protect against ultraviolet light.** Strict sun protection prevents both skin and systemic flares.

The stepped-care ladder

Tier	Class & examples	Role
1	**Hydroxychloroquine** 5 mg/kg/day (max 400 mg/day)	Backbone therapy for all patients. Annual ophthalmology screening from year 5 to detect rare retinal toxicity.
2	**Glucocorticoids** Oral prednisolone, IV methylprednisolone pulses for severe flares	Rapid control of active disease; tapered as quickly as possible to ≤5 mg/day.
3	**Conventional immunosuppressants** Methotrexate, azathioprine, mycophenolate mofetil, cyclophosphamide, ciclosporin, tacrolimus	Steroid-sparing; used for moderate-severe disease and organ involvement. Mycophenolate and cyclophosphamide are mainstays in lupus nephritis.
4	**Biologic therapies** • **Belimumab** (anti-BLyS) — for active non-renal SLE and lupus nephritis • **Anifrolumab** (anti-type-I interferon receptor) — approved 2021 for active non-renal SLE • **Rituximab** (anti-CD20) — refractory disease, off-label	Considered early, alongside immunosuppressants — no longer reserved for treatment failure.
5	**Voclosporin** (calcineurin inhibitor) — newest agent for active lupus nephritis, used with mycophenolate and low-dose steroids	Approved 2021. Combination therapy improves renal response rates.

What you can do yourself

- **Strict sun protection.** Broad-spectrum SPF 50+ sunscreen daily, wide-brimmed hats, long sleeves, and avoiding midday sun. This is non-negotiable.
- **Stop smoking.** Smoking worsens lupus, reduces the effectiveness of hydroxychloroquine, and dramatically raises cardiovascular risk.
- **Take your medication every day**, even when you feel well. Stopping hydroxychloroquine is the single most preventable cause of flares.
- **Stay up to date with vaccinations** — annual influenza, pneumococcal, COVID-19, and (where indicated) shingles vaccines. Avoid live vaccines if on biologic or strong immunosuppressive therapy.
- **Eat a Mediterranean-style diet**, maintain a healthy weight, and exercise regularly to protect against cardiovascular disease and osteoporosis.
- **Plan pregnancies carefully.** Lupus should be in remission for at least six months before conception, and medication needs adjustment — hydroxychloroquine is continued, but mycophenolate, methotrexate, and cyclophosphamide must be stopped before pregnancy.
- **Mind your mental health.** Depression and anxiety are common and treatable.

What is the outlook?

The outlook for lupus has improved dramatically. Early diagnosis, hydroxychloroquine for all, modern immunosuppressants, biologic agents, and aggressive cardiovascular risk management have transformed lupus into a manageable chronic disease for most patients. **Ten-year survival now exceeds 90%** in

well-resourced settings. Long-term outcomes depend heavily on three things: **early diagnosis**, **adherence to treatment**, and **regular monitoring** — particularly of the kidneys, which can flare silently. Lupus nephritis, infection, and cardiovascular disease remain the principal causes of death; all three are largely preventable with current standards of care.

When to contact your rheumatologist promptly

- A new rash, mouth ulcers, or hair loss — possible flare.
- Swelling around the eyes or ankles, frothy urine, or rising blood pressure.
- Severe headache, confusion, weakness on one side, or a new seizure.
- Chest pain, breathlessness, or coughing up blood.
- Fever above 38°C, especially if on biologic or immunosuppressive treatment — urgent assessment to exclude infection.
- Unexpected pregnancy — early review allows safe medication adjustment.

Frequently asked questions

Question	Answer
Is lupus contagious?	No. Lupus is an autoimmune disease, not an infection. You cannot catch it from another person and you cannot pass it on by close contact.
Can lupus be cured?	Lupus cannot yet be cured, but it can be very effectively **controlled**. With modern treatment, most patients achieve long periods of remission and lead full lives.
Will I pass lupus on to my children?	Lupus is not inherited in a simple way. The risk to a child of a parent with lupus is approximately 5%, compared to 0.1% in the general population — modestly raised, but the vast majority of children remain unaffected.
Can I have children?	Yes. Fertility is generally normal in lupus, but pregnancy must be planned carefully — ideally during a period of at least 6 months of disease remission, with medication reviewed in advance. Hydroxychloroquine is safe and should be continued.
Can I fast during Ramadan?	Most stable lupus patients can fast safely, but you must stay well hydrated outside fasting hours and discuss medication timing with your rheumatologist. Patients with active disease, kidney involvement, or on diuretics may be exempt.
Is hydroxychloroquine safe long-term?	Yes, for the vast majority of patients. The main concern is a very rare retinal toxicity after several years of use, which is why annual eye checks are recommended from year 5 of treatment onwards.

Key references and further reading

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Speak to a specialist

Lupus is treatable, and early specialist care protects your organs for life — 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.

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