

Sjögren Disease Clinical Evaluation Pathway

A comprehensive diagnostic, systemic, and surveillance blueprint for primary and secondary epithelitis.

Based on 2016 ACR/EULAR
Guidelines and Australian
Epidemiological Data.

A Chronic Systemic Autoimmune Epithelitis

Sjögren disease (SjD) is driven by lymphocytic infiltration and progressive destruction of exocrine glands. It manifests primarily, or secondarily alongside Rheumatoid Arthritis or SLE.



0.1-0.6%

Estimated Prevalence

Approximately 1 in 1000 adults.



9:1 Ratio

Strong female-to-male predominance



Peak Onset

4th to 6th decades of life.

The Hallmark Sicca Symptoms



Xerostomia (Dry Mouth)

Symptoms:

- Persistent dryness & altered taste
- Difficulty swallowing dry foods
- Oral candidiasis & rampant dental caries

Objective Measures:

- Unstimulated salivary flow rate ≤ 0.1 mL/min
- Abnormal parotid sialography (diffuse sialectasia)
- Salivary gland scintigraphy (delayed uptake/excretion)



Keratoconjunctivitis Sicca (Dry Eyes)

Symptoms:

- Gritty / foreign body sensation
- Burning & photophobia
- Mucoid discharge

Objective Measures:

- Schirmer's test ≤ 5 mm/5 min
- Positive Ocular Staining Score (OSS) ≥ 5
- Tear film break-up time < 10 seconds

Not All Sicca is Sjögren's

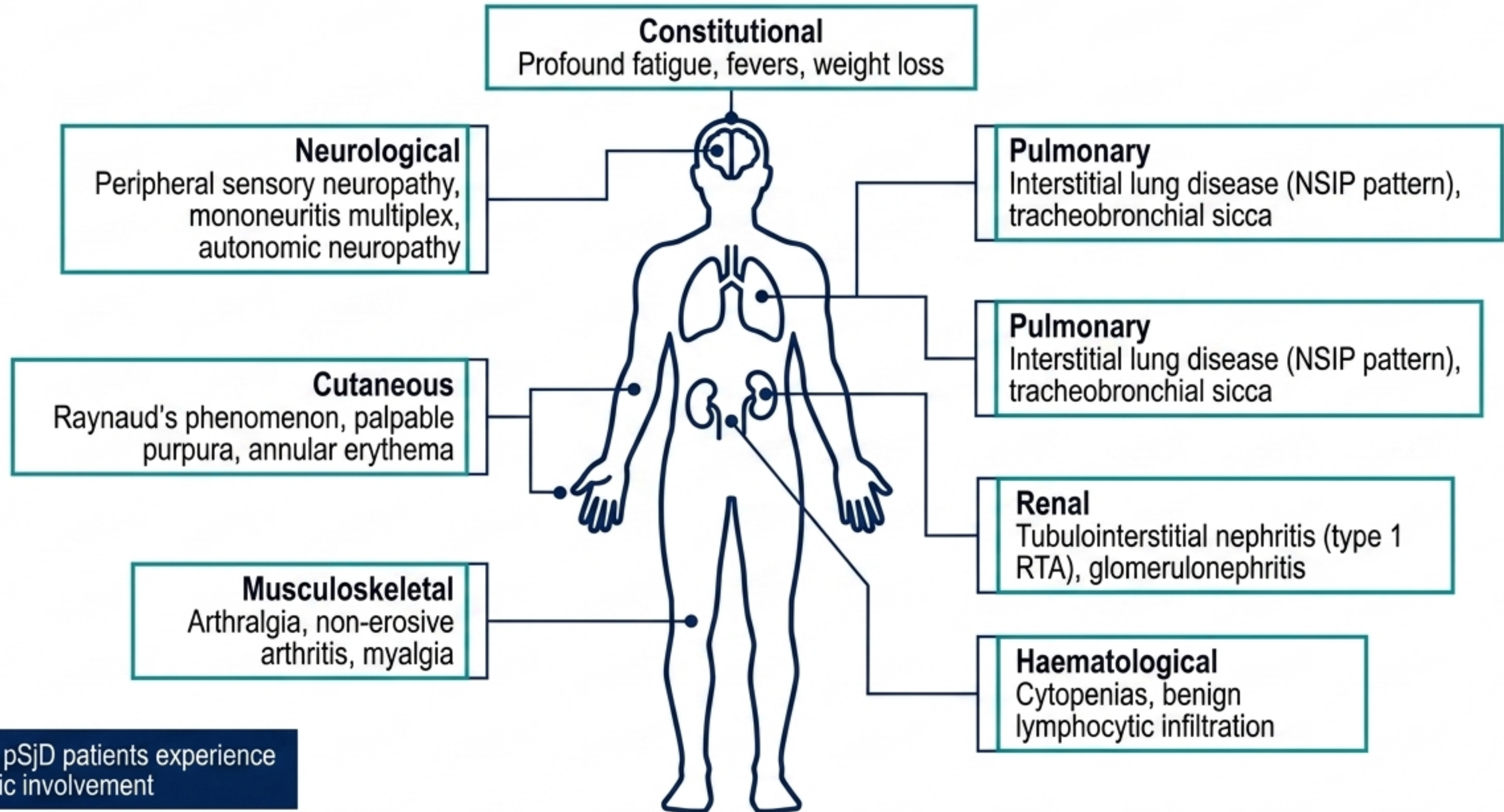
Before proceeding to a systemic diagnostic pathway, explicitly exclude secondary causes of exocrine failure.



Primary Sjögren's Disease
Diagnosis proceeds only if secondary causes are absent.

⚠ Exclude Secondary Causes	
Pharmacological	Anticholinergic medications, antidepressants.
Infectious	Active Hepatitis C, HIV/AIDS.
Iatrogenic	Prior head and neck irradiation, Graft-versus-host disease (GVHD).
Infiltrative / Other	Sarcoidosis, Amyloidosis, IgG4-related disease.

Extraglandular Disease Reaches Nearly Every System



50% of pSjD patients experience systemic involvement

The Prognostic Power of Autoantibodies

Anti-Ro / SSA	Anti-La / SSB
Prevalence: 60-70%	Prevalence: 30-40%
Subtypes: Includes Ro52 and Ro60.	Clinical Driver: <ul style="list-style-type: none">• Typically presents in conjunction with anti-Ro• Associated with pronounced glandular dysfunction• Associated with higher lymphoma risk
Clinical Driver: <ul style="list-style-type: none">• Drives earlier disease onset• Severe salivary gland involvement• Extraglandular manifestations (ILD, vasculitis)• Heightened lymphoma risk	

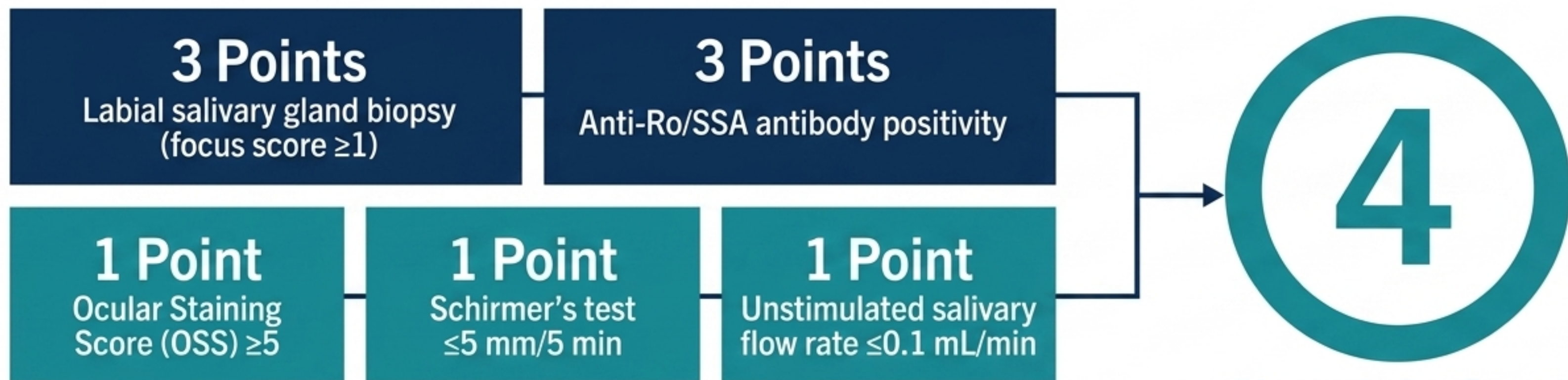


Obstetric Alert: Anti-Ro/SSA antibodies cross the placenta. Critical risk of neonatal lupus erythematosus (NLE) and congenital heart block (CHB).

The 2016 ACR/EULAR Classification Criteria

Entry Requirement: Positive ANA $\geq 1:320$, RF positive, or any systemic feature suggestive of SjD.

Path to 4



Diagnosis Confirmed
(Score ≥ 4)



Biopsy [3]

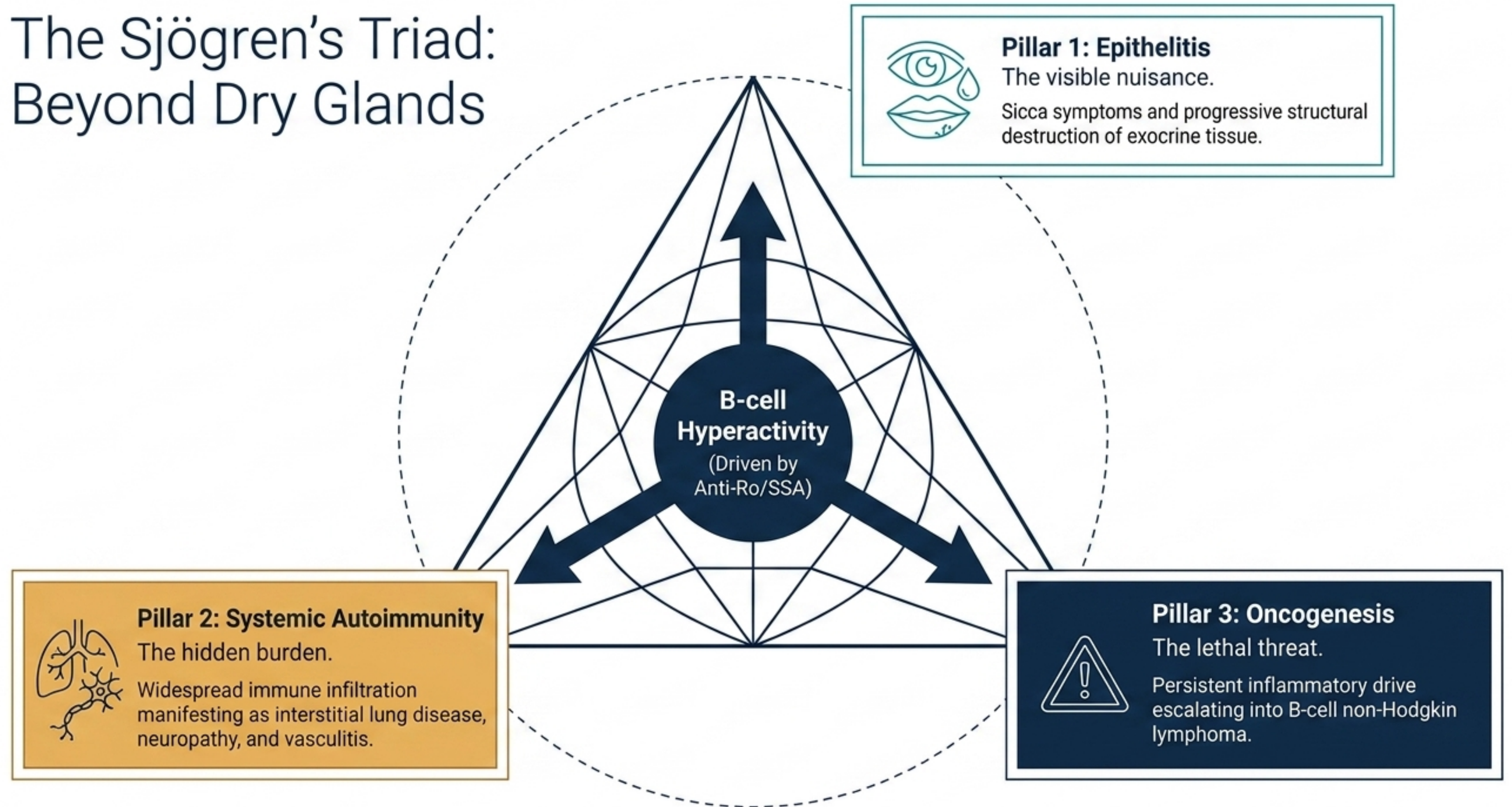


OSS [1]

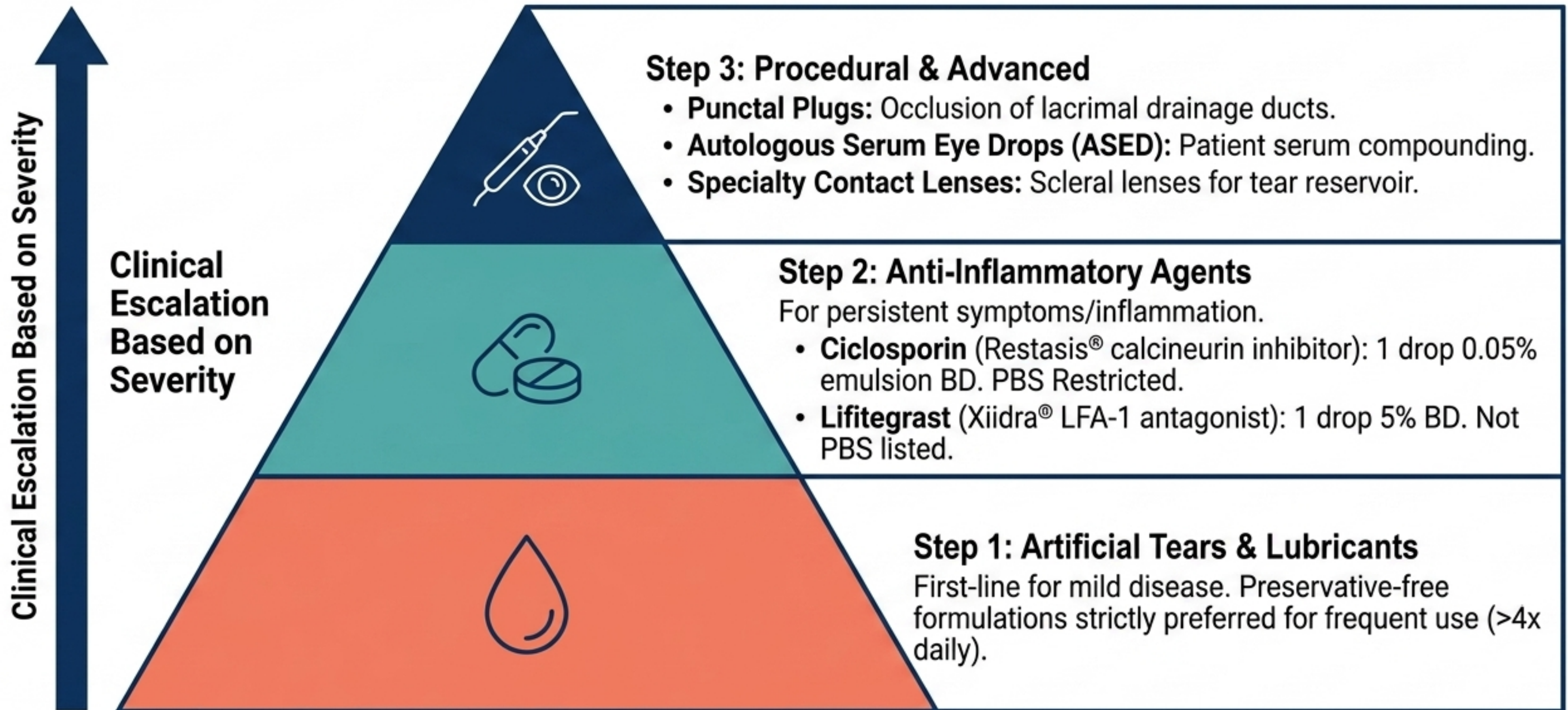


(Criteria Met)

The Sjögren's Triad: Beyond Dry Glands



Stepwise Management of Dry Eye Disease



Systemic Treatment Protocols for Extraglandular Disease

Baseline Systemic Therapy

Hydroxychloroquine acts as the first-line anchor for systemic control and baseline immunomodulation.

Targeted Immunosuppression

Deployed for severe organ manifestations (e.g., ILD, vasculitis).

Includes: Methotrexate, Azathioprine, or Mycophenolate.

The Multidisciplinary Requirement

Systemic management demands a coordinated network:



Rheumatology
(Systemic oversight)



Ophthalmology
(Corneal preservation)

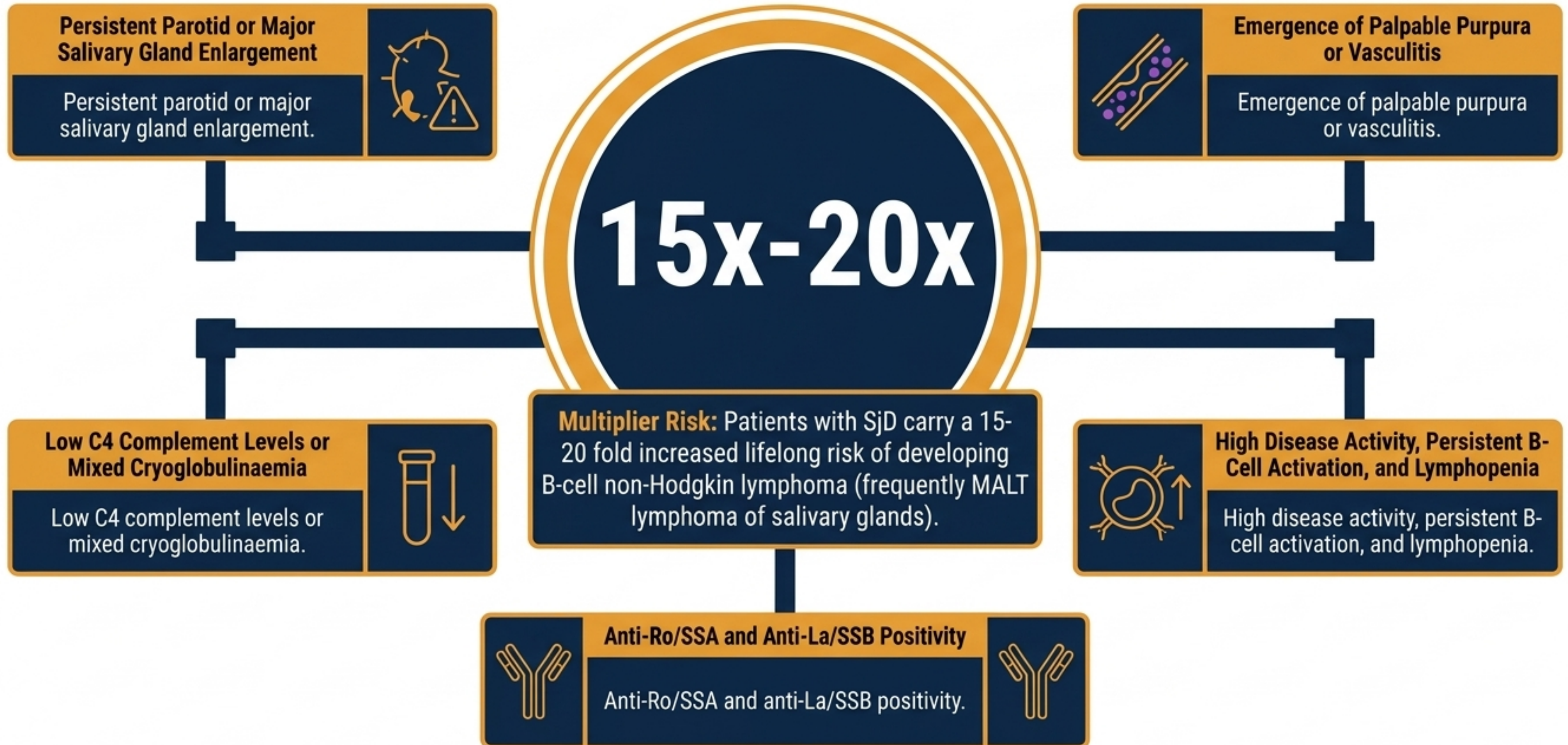


Dentistry
(Caries/oral care)

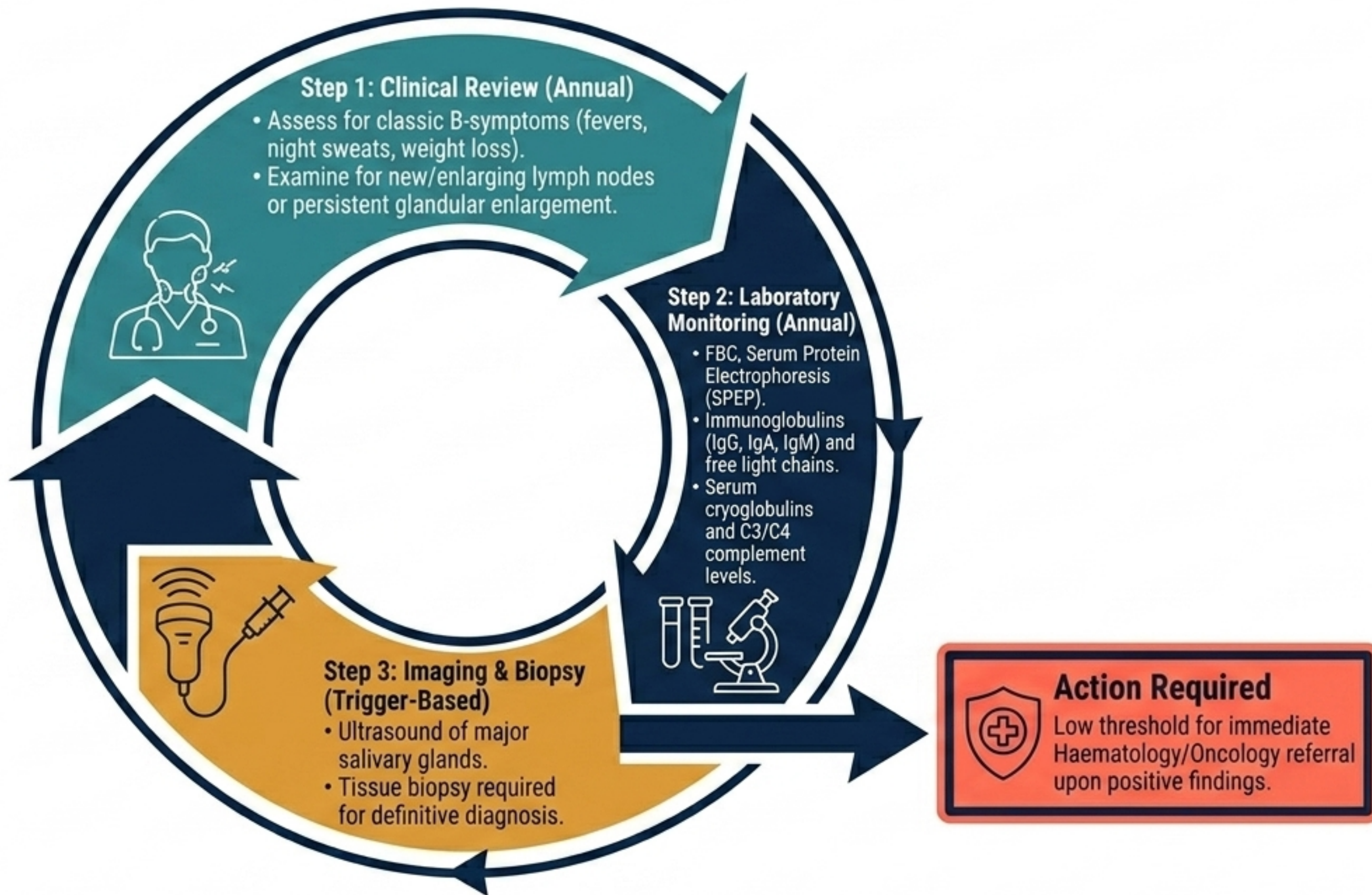


Physiotherapy
(MSK/fatigue management)

The Oncogenic Threat: Non-Hodgkin Lymphoma



The Annual Lymphoma Surveillance Engine



Special Populations: Pregnancy & Paediatrics



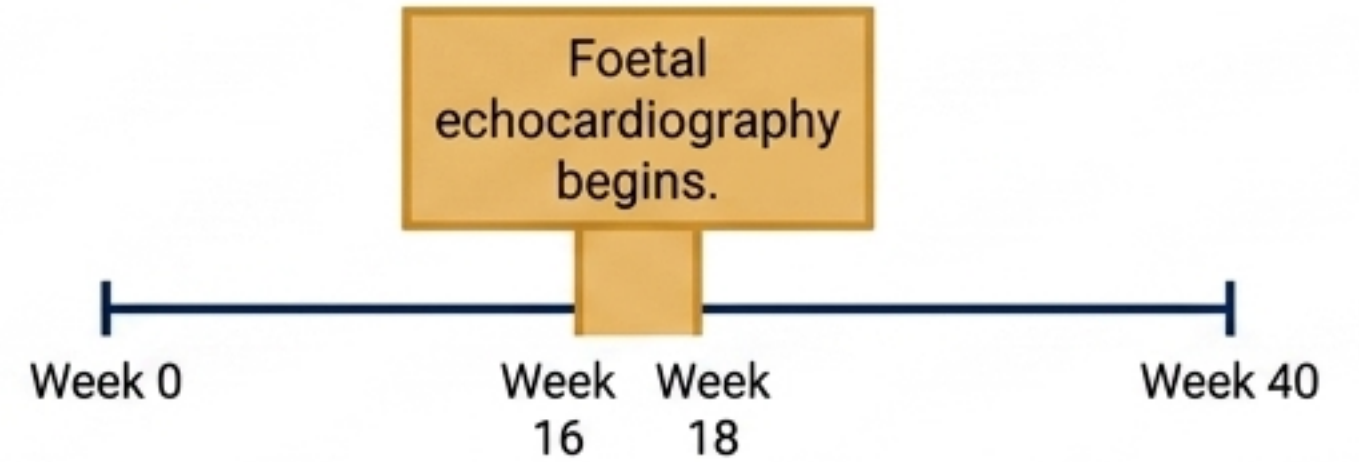
Pregnancy & Obstetrics

Key Threat

Anti-Ro/SSA mediated neonatal lupus and congenital heart block (CHB).

Pharmacology

Hydroxychloroquine should be continued. Strictly avoid methotrexate and mycophenolate.



Protocol: Mandatory pre-conception counselling.



Paediatric Considerations

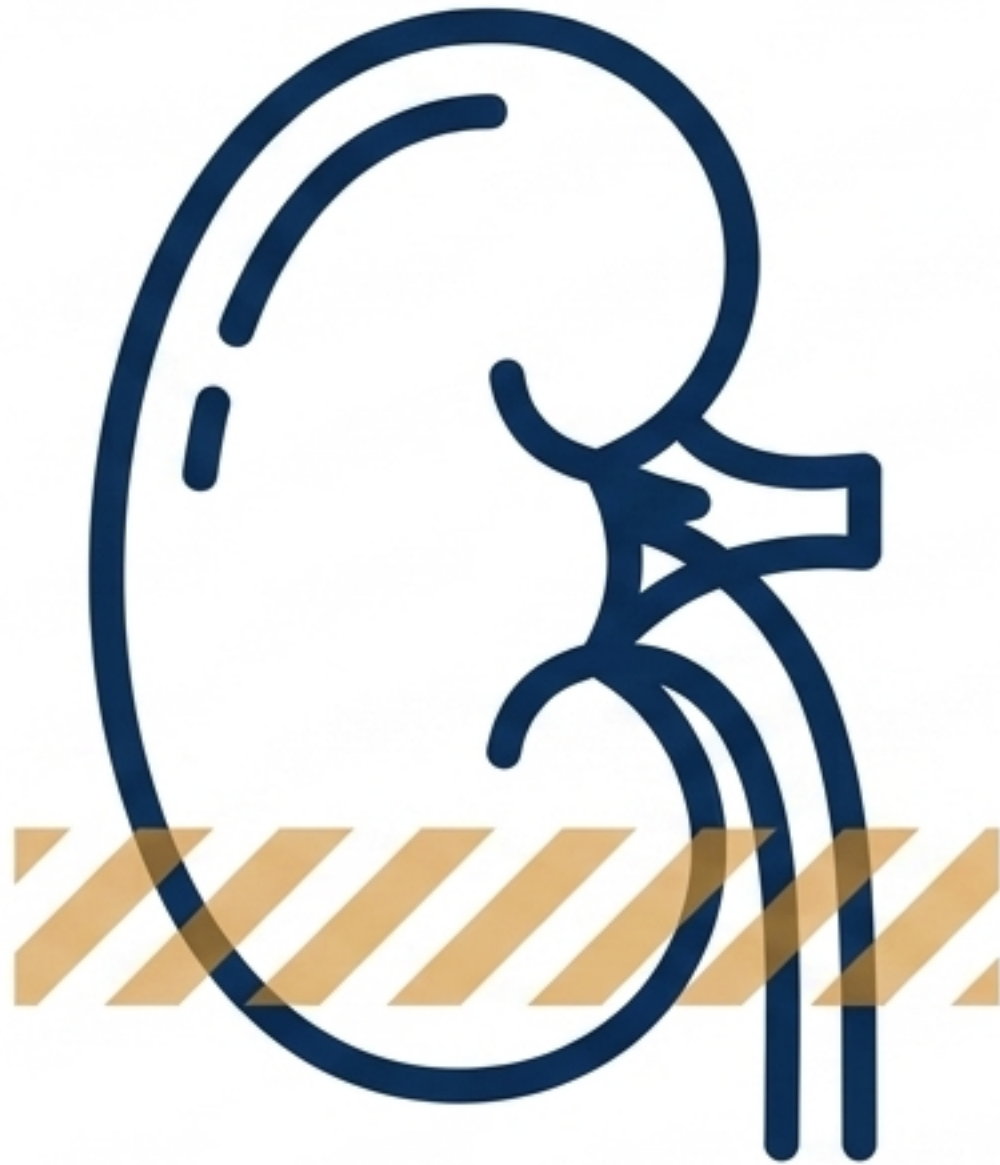
Presentation:

Often presents primarily with parotitis and sicca. Demonstrates a strong association with anti-Ro/SSA and familial autoimmunity.

Management:

Dose adjustments for weight are strictly required; biologic data remains limited in this demographic.

Special Populations: Renal Impairment



Pathological Link

Type 1 Renal Tubular Acidosis (RTA) is a common, often silent complication of SjD.

Monitoring Protocol

Require strict, routine monitoring of electrolytes and systemic acid-base status.

Pharmacological Adjustments



- NSAIDs should be actively avoided if possible to protect remaining renal function.
- Mandatory dose adjustments required for nephrotoxic or renally cleared drugs, specifically methotrexate and ciclosporin.

Aboriginal and Torres Strait Islander Health Considerations



Remote & Rural Access

Address specialist shortages by utilizing telehealth and store-and-forward for ocular imaging. Collaborate tightly with local Aboriginal Medical Services (AMS) and visiting specialists.



Oral Health Interventions

High baseline rates of dental disease exacerbate xerostomia. Promote fluoride varnish, sugar-free gum, and frequent AMS dental checks.



Medication Continuity

Ensure continuous access to PBS-listed mainstays (hydroxychloroquine, artificial tears). Consider long-term dispensing protocols to overcome travel and cost barriers.



Cultural Safety & Health Literacy

Employ jargon-free visual aids. Integrate Aboriginal Health Workers into education to build long-term, high-trust therapeutic relationships.