# Rheumatologic Emergencies in the Hospital Setting

Rheumatologic emergencies are life-threatening conditions associated with autoimmune diseases that require prompt recognition and management in the hospital. This pamphlet provides students with a guide to suspect, diagnose, treat, and manage these emergencies, including when to consult rheumatology, with clinical scenarios to apply the knowledge.

## When to Suspect Rheumatologic Emergencies

Rheumatologic emergencies should be suspected in patients with known or suspected autoimmune diseases presenting with acute, severe symptoms involving multiple organ systems. Key conditions include:

- **Systemic Lupus Erythematosus (SLE) Flares:**
  - **Suspect in:** Known SLE patients with new fever, rash (malar or discoid), joint pain, or organ dysfunction (e.g., renal, neurologic, hematologic).
  - **Red Flags:** Acute renal failure (lupus nephritis), seizures/psychosis (CNS lupus), severe cytopenias (hemolytic anemia, thrombocytopenia), or serositis (pleuritis, pericardial).
- **Vasculitis:**
  - **Giant Cell Arteritis (GCA):** Suspect in patients >50 years with new headache (temporal), jaw claudication, vision changes (amaurosis fugax, diplopia), scalp tenderness, or polymyalgia rheumatica (PMR) symptoms (shoulder/hip stiffness).
  - **ANCA-Associated Vasculitis (AAV):** Suspect in patients with rapidly progressive glomerulonephritis (hematuria, proteinuria), pulmonary hemorrhage (hemoptysis, dyspnea), or mononeuritis multiplex (e.g., wrist/foot drop).
- **Catastrophic Antiphospholipid Syndrome (CAPS):** Suspect in: Known antiphospholipid syndrome (APS) or SLE patients with acute multiorgan thrombosis (e.g., DVT, stroke, MI), often with thrombocytopenia and microangiopathic hemolytic anemia (MAHA).
- **Scleroderma Renal Crisis (SRC):** Suspect in: Scleroderma patients (especially diffuse cutaneous systemic sclerosis) with new hypertension (>150/90 mmHg), acute renal failure (Cr rise), and microangiopathic hemolytic anemia (schistocytes, elevated LDH).

- **Septic Arthritis in Rheumatoid Arthritis (RA):** Suspect in: RA patients with acute monoarticular swelling, warmth, and pain (especially knee, hip), often with fever; high risk due to immunosuppression (e.g., on DMARDs, biologics).

## Diagnosis

- **Clinical Assessment:**
- **History:** Prior autoimmune diagnosis (SLE, RA, scleroderma), medication use (steroids, biologics), recent infections (trigger for flares), or symptoms suggestive of organ involvement (e.g., hemoptysis, vision loss, seizures).
- **Physical Exam:**
  - **SLE:** Malar rash, oral ulcers, arthritis, pleural rub, pericardial rub, altered mental status.
  - **GCA:** Temporal artery tenderness, visual acuity loss, PMR symptoms.
  - **AAV:** Purpura (palpable), hemoptysis, neuropathy (mononeuritis multiplex).
  - **CAPS:** Signs of thrombosis (DVT, stroke, livedo reticularis), MAHA (pallor, jaundice).
  - **SRC:** Hypertension, edema, sclerodactyly, Raynaud's phenomenon.
  - **Septic Arthritis:** Hot, swollen joint, limited range of motion, fever.
- Diagnostic Testing (see Testing section for details):
  - **Labs:** CBC (cytopenias, MAHA), CMP (renal/liver function), inflammatory markers (ESR, CRP), autoantibodies (ANA, anti-dsDNA, ANCA, APLAs).
  - **Imaging:** CT chest (pulmonary hemorrhage in AAV), brain MRI (CNS lupus, stroke in CAPS), temporal artery ultrasound (GCA).
- **Procedures:** Joint aspiration (septic arthritis), renal biopsy (lupus nephritis, SRC), temporal artery biopsy (GCA).

## Diagnostic Testing

- **Labs:**
  - **CBC:** Anemia (hemolytic in SLE, CAPS, SRC), thrombocytopenia (SLE, CAPS), leukocytosis (septic arthritis, infection).
  - **CMP:** Elevated Cr (lupus nephritis, SRC, AAV), transaminitis (SLE flare, drug toxicity).
  - **Inflammatory Markers:** ESR/CRP elevated in GCA, AAV, SLE flare; ESR >50 mm/h in GCA (highly suggestive).
  - **Autoantibodies:**
    - **SLE:** ANA (sensitive, >95%), anti-dsDNA (specific, correlates with disease activity), low C3/C4 (complement consumption).

        - **AAV:** c-ANCA (anti-PR3, granulomatosis with polyangiitis [GPA]), p-ANCA (anti-MPO, microscopic polyangiitis [MPA]).
        - **APS/CAPS:** Lupus anticoagulant, anti-cardiolipin antibodies, anti-beta-2 glycoprotein I (positive on 2 occasions, 12 weeks apart).
    - **Coagulation Studies:** Prolonged aPTT (lupus anticoagulant in CAPS), elevated D-dimer (thrombosis in CAPS).
    - **Urinalysis:** Hematuria, proteinuria, casts (lupus nephritis, AAV, SRC).
    - **Joint Fluid Analysis:** Septic arthritis → WBC >50,000/µL, >90% neutrophils, positive Gram stain/culture (e.g., S. aureus).
    - **Other:** Elevated LDH, schistocytes on smear (MAHA in CAPS, SRC), CK (myositis in SLE flare).
- **Imaging:**
    - **CT Chest:** Alveolar hemorrhage (AAV), pleural effusion (SLE).
    - **Brain MRI/CT:** Stroke (CAPS), cerebritis (CNS lupus), posterior reversible encephalopathy syndrome (PRES) in SRC.
    - **Temporal Artery Ultrasound:** Halo sign (GCA); alternative to biopsy if available.
- **Echocardiogram:** Pericardial effusion (SLE), vegetations (endocarditis mimic in APS).
- **Procedures:**
    - **Joint Aspiration:** Rule out septic arthritis (cell count, Gram stain, culture).
    - **Renal Biopsy:** Lupus nephritis (class III/IV: proliferative), SRC (onion-skinning of arterioles), AAV (pauci-immune glomerulonephritis).
    - **Temporal Artery Biopsy:** Vasculitis with giant cells (GCA); perform within 7-14 days of starting steroids.
    - **Skin Biopsy:** Vasculitis (palpable purpura in AAV), leukocytoclastic vasculitis (SLE).
- Diagnostic Testing Table

| Test | Condition | Findings | Notes |
|---|---|---|---|
| ANA, anti-dsDNA | SLE | ANA >95% sensitive, anti-dsDNA specific | Low C3/C4 indicates active disease. |
| c-ANCA, p-ANCA | AAV | c-ANCA (GPA), p-ANCA (MPA) | Order with urinalysis for renal involvement. |
| Temporal Artery Biopsy | GCA | Giant cells, vasculitis | Perform within 7-14 days of steroids. |
| Joint Aspiration | Septic Arthritis (RA) | WBC >50,000/µL, positive culture | Rule out gout (crystals) as mimic. |

## Treatment

- **General Principles: Emergent Rheumatology consult!**
  - Stabilize the patient (e.g., airway, fluids, blood pressure control).
  - Initiate high-dose immunosuppression for most emergencies (except septic arthritis).
  - Treat underlying infections or triggers (e.g., infection precipitating SLE flare).
- **SLE Flare (Severe):**
  - **Lupus Nephritis (Class III/IV):** Methylprednisolone 500-1000 mg IV daily x 3 days, then prednisone 1 mg/kg/day PO (taper over 4-6 weeks); add cyclophosphamide 500-1000 mg/m$^2$ IV monthly x 6 or mycophenolate mofetil (MMF) 2-3 g/day PO.
  - **CNS Lupus:** Methylprednisolone 1000 mg IV daily x 3 days, then prednisone 1 mg/kg/day PO; consider cyclophosphamide 750 mg/m$^2$ IV if refractory.
  - **Supportive:** Plasmapheresis (if MAHA or severe flare), IV fluids for renal protection.
- **Giant Cell Arteritis (GCA):**
  - **Vision Threatened:** Methylprednisolone 1000 mg IV daily x 3 days, then prednisone 60 mg PO daily (taper over 6-12 months).
  - **No Vision Threat:** Prednisone 40-60 mg PO daily (taper over 6-12 months).
  - **Supportive:** Aspirin 81 mg PO daily (reduce ischemic risk), PPI (e.g., omeprazole 20 mg PO daily) for GI protection.
- **ANCA-Associated Vasculitis (AAV):**
  - **Pulmonary Hemorrhage or RPGN:** Methylprednisolone 1000 mg IV daily x 3 days, then prednisone 1 mg/kg/day PO; cyclophosphamide 15 mg/kg IV q2-3 weeks x 3-6 doses or rituximab 375 mg/m$^2$ IV weekly x 4.
  - **Plasmapheresis:** Add for severe cases (e.g., pulmonary hemorrhage, dialysis-dependent RPGN), 7-10 sessions over 14 days.
  - **Supportive:** Oxygen, hemodialysis (if renal failure), transfusion (if severe anemia).
- **Catastrophic Antiphospholipid Syndrome (CAPS):**
  - **Anticoagulation:** Heparin 80 units/kg IV bolus, then 18 units/kg/h infusion (target aPTT 60-80 seconds), transition to warfarin (INR 2-3).
  - **Immunosuppression:** Methylprednisolone 1000 mg IV daily x 3 days, then prednisone 1 mg/kg/day PO; IVIG 0.4 g/kg IV daily x 5 days.
  - **Plasmapheresis:** 5-7 sessions over 7-14 days (removes pathogenic antibodies).

- **Supportive:** Treat thrombosis (e.g., tPA for massive PE, 100 mg IV over 2h), manage MAHA (transfusion if Hgb <7 g/dL).

- **Scleroderma Renal Crisis (SRC):**
  - **BP Control:** ACE inhibitor (e.g., captopril 25 mg PO q6-8h, titrate to BP <130/80 mmHg; max 150 mg/day).
  - **Immunosuppression:** Avoid high-dose steroids (can worsen SRC); if needed, prednisone <15 mg/day PO.
  - **Supportive:** Hemodialysis (if renal failure), monitor for MAHA (schistocytes, LDH).
- **Septic Arthritis in RA:**
  - **Antibiotics:** Vancomycin 15-20 mg/kg IV q12h (S. aureus coverage) + ceftriaxone 2 g IV daily (Gram-negative coverage) until culture results; adjust based on sensitivities (e.g., methicillin-sensitive S. aureus → nafcillin 2 g IV q4h).
  - **Surgical Drainage:** Urgent orthopedic consult for joint aspiration or arthroscopy (especially hip, knee).
  - **Supportive:** Hold immunosuppressive RA meds (e.g., methotrexate, biologics) until infection resolves; low-dose prednisone (5-10 mg PO daily) if needed for RA flare.

### Treatment Guidelines Table

| Condition | Treatment Agent/Dose | Notes |
|---|---|---|
| SLE Flare (Severe) | Methylprednisolone 1000 mg IV x 3 days - Cyclophosphamide 500-1000 mg/m$^2$ IV | Add MMF for lupus nephritis maintenance. |
| GCA (Vision Threat) | High-Dose Steroids Methylprednisolone 1000 mg IV x 3 days | Transition to prednisone 60 mg PO daily. |
| AAV (Pulmonary Hemorrhage) | Immunosuppression + Plasmapheresis Rituximab 375 mg/m$^2$ IV weekly x 4 | Oxygen, hemodialysis if needed. |
| Septic Arthritis (RA) | Antibiotics + Drainage Vancomycin 15-20 mg/kg IV q12h | Urgent orthopedic consult for drainage. |

## When to Consult Rheumatology

- **Indications:**
  - **Diagnostic Uncertainty:** Suspected autoimmune disease with unclear diagnosis (e.g., overlapping features of SLE and AAV).
  - **Severe Organ Involvement:** Lupus nephritis (Cr rise, proteinuria), CNS lupus (seizures, psychosis), pulmonary hemorrhage (AAV), vision loss (GCA).
  - **Refractory Disease:** Flares not responding to initial therapy (e.g., persistent hemoptysis in AAV despite steroids).

    - **Complex Therapy Needs:** Need for biologics (e.g., rituximab in AAV), plasmapheresis (CAPS, AAV), or long-term immunosuppression plans.
    - **CAPS or SRC:** Urgent consult for multiorgan involvement and specialized management (e.g., anticoagulation in CAPS, ACE inhibitors in SRC).
- **Timing:**
    - **Urgent:** Life-threatening conditions (e.g., pulmonary hemorrhage, vision loss, CAPS, SRC, severe lupus flare).
    - **Routine:** Stable patients needing long-term management plans or confirmation of diagnosis (e.g., GCA with no vision loss).

## Complications

- **SLE Flare:**
    - Renal failure (lupus nephritis → ESRD in 10-20% if untreated), stroke (CNS lupus), infection (immunosuppression-related, e.g., PNA), thrombosis (APS overlap).
- **GCA:**
    - Permanent vision loss (anterior ischemic optic neuropathy, 15-20% if untreated), stroke (vertebrobasilar), aortic aneurysm (long-term risk).
- **ANCA-Associated Vasculitis (AAV):**
    - Respiratory failure (pulmonary hemorrhage), ESRD (RPGN, 20-40% progress to dialysis), neuropathy (permanent mononeuritis multiplex).
- **Catastrophic Antiphospholipid Syndrome (CAPS):**
    - Multiorgan failure (thrombosis in kidneys, lungs, brain), DIC (20% of cases), death (mortality 30-50% if untreated).
- **Scleroderma Renal Crisis (SRC):**
    - ESRD (50% progress to dialysis if untreated), hypertensive emergency (encephalopathy, seizures), heart failure (HTN-related).
- **Septic Arthritis in RA:**
    - Joint destruction (cartilage loss), sepsis (mortality 10-15%), osteomyelitis (if untreated).

## Key Pearls

- Suspect rheumatologic emergencies in autoimmune patients with acute multiorgan symptoms (e.g., hemoptysis, vision loss, renal failure).
- **SLE Flares:** High-dose steroids (methylprednisolone IV) for severe cases; add cyclophosphamide or MMF for lupus nephritis.
- **GCA:** Start steroids immediately if vision loss suspected; don’t delay for biopsy (treat first, biopsy within 7-14 days).

- **AAV:** Pulmonary hemorrhage is a medical emergency; use steroids, rituximab/cyclophosphamide, and plasmapheresis.
- **CAPS:** Triple therapy (anticoagulation, steroids, IVIG/plasmapheresis) for multiorgan thrombosis.
- **SRC:** ACE inhibitors are first-line; avoid high-dose steroids (worsen crisis).
- **Septic Arthritis:** Urgent joint aspiration and antibiotics; hold RA immunosuppressants during infection.
- **Rheumatology Consult:** Urgent for life-threatening cases (e.g., CAPS, SRC, pulmonary hemorrhage); routine for diagnostic uncertainty.

## References


- **UpToDate:** "Rheumatologic Emergencies: Diagnosis and Management" (2025).
- **Arthritis & Rheumatology:** "Management of Giant Cell Arteritis and Polymyalgia Rheumatica" (2023).
- **NEJM:** "Catastrophic Antiphospholipid Syndrome: Diagnosis and Treatment" (2024).
- **Annals of the Rheumatic Diseases:** "Scleroderma Renal Crisis: Updates in Management" (2024).


## Clinical Scenarios

### Case 1: A 30-Year-Old Female with Fever and Rash

- **Presentation:** A 30-year-old female with known SLE presents with fever, malar rash, joint pain, and confusion for 2 days. Exam: BP 110/70 mmHg, HR 100 bpm, altered mental status, arthritis (wrists, knees).
- **Labs**: Hgb 9 g/dL, platelets 80,000/µL, Cr 2.5 mg/dL (baseline 0.9), ANA positive, anti-dsDNA 1:320, C3/C4 low, urinalysis: hematuria, proteinuria.
- **Diagnosis:** Severe SLE Flare (CNS Lupus + Lupus Nephritis) → Altered mental status (CNS lupus), renal dysfunction (nephritis), cytopenias.
- **Management:** Start methylprednisolone 1000 mg IV daily x 3 days, then prednisone 1 mg/kg/day PO. Add cyclophosphamide 750 mg/m$^2$ IV for nephritis. Order brain MRI (rule out cerebritis, stroke). Urgent rheumatology consult for severe flare. Monitor renal function, mental status; consider plasmapheresis if refractory.

## Case 2: A 65-Year-Old Male with Headache and Vision Loss

- **Presentation:** A 65-year-old male presents with a new temporal headache, jaw claudication, and sudden vision loss in the right eye for 1 day. Exam: Scalp tenderness, right visual acuity 20/200, ESR 70 mm/h.
- **Imaging:** Temporal artery ultrasound: halo sign.
- **Diagnosis:** Giant Cell Arteritis (GCA) with Vision Loss → Classic symptoms, vision loss, elevated ESR.
- **Management:** Start methylprednisolone 1000 mg IV daily x 3 days, then prednisone 60 mg PO daily. Aspirin 81 mg PO daily, omeprazole 20 mg PO daily (GI protection). Urgent rheumatology consult for vision-threatening GCA. Schedule temporal artery biopsy within 7-14 days. Refer to ophthalmology for vision assessment.

## Case 3: A 50-Year-Old Male with Hemoptysis and Renal Failure

- **Presentation:** A 50-year-old male presents with hemoptysis, dyspnea, and decreased urine output for 3 days. Exam: BP 150/90 mmHg, crackles in lungs, palpable purpura on legs, foot drop (neuropathy).
- **Labs:** Cr 3.5 mg/dL, urinalysis: hematuria, proteinuria, c-ANCA positive (anti-PR3), Hgb 8 g/dL.
- **Imaging:** CT chest: alveolar hemorrhage.
- **Diagnosis:** ANCA-Associated Vasculitis (GPA) with Pulmonary Hemorrhage and RPGN → Hemoptysis (pulmonary hemorrhage), renal failure (RPGN), c-ANCA positive.
- **Management:** Start methylprednisolone 1000 mg IV daily x 3 days, then prednisone 1 mg/kg/day PO. Rituximab 375 mg/m$^2$ IV weekly x 4. Initiate plasmapheresis (7 sessions over 14 days). Oxygen support, consider hemodialysis. Urgent rheumatology consult for life-threatening AAV. Monitor respiratory status, renal function, and response to therapy.

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