Emergency Department Guideline
Fever in the Immunosuppressed Rheumatology Patient

Inclusion criteria:
- Patient with measured or reported fever (≥38.0°C/100.4°F) and immunosuppression, including:
  - Patients with rheumatologic conditions (e.g. JIA, SLE, etc.) currently being treated with immunosuppressive medications, regardless of ANC.
    - Medications which are considered immunosuppressive include:
      - Any biologic (e.g. etanercept, adalimumab, infliximab, anakinra, canakinumab, abatacept, rituximab, tocilizumab)
      - Mycophenolate
      - Azathioprine
      - Cyclophosphamide
      - Tacrolimus or sirolimus
      - IVIG
      - Corticosteroids greater than 2 mg/kg or 20 mg per day.
    - Medications which are NOT considered immunosuppressive include:
      - Methotrexate
      - Hydroxychloroquine
      - Sulfasalazine
      - Leflunomide
      - Colchicine.
  - In the absence of immunosuppressive medications, some rheumatologic diseases (such as SLE) in and of themselves could result in a suppressed immune system if disease is not well-controlled. If this is a concern, discuss with the Pediatric Rheumatologist.

Triage considerations:
- NO rectal temperature measurements for these patients
- ESI triage level 2
  - Triage level 1 and immediate placement in resuscitation room if altered mental status, significant VS abnormalities, other clinical concern
- Full set of vitals including blood pressure
- Place LMX on port site if patient/family desires
  - Do NOT wait for LMX in ill-appearing/septic patients
- Acetaminophen if current fever and none given in 4 hours
• NSAIDs also ok if patient not already on a scheduled NSAID
• Room immediately if at all possible; avoid waiting room exposure
• NOT all immunosuppressed rheumatology patients will need antibiotics.
  • If patient is ill-appearing or neutropenic (ANC<500), goal should be IV antibiotics within 60 minutes of arrival.
  • If well-appearing and not neutropenic, antibiotics can be discussed with on-call Pediatric Rheumatologist.

Laboratory studies:
• In all patients:
  o Obtain IV access or access indwelling line in all patients
  o CBC with diff, CRP, ESR
  o Blood culture from each port of indwelling line
    ▪ Peripheral cultures NOT routinely indicated in patients with lines
  o Single peripheral blood culture if no line
• In patients who are significantly ill-appearing:
  o CMP
  o Coagulation studies: INR, PTT, fibrinogen, D-dimer
  o Type and screen
  o iStat CG8
  o Lactate
  o Procalcitonin
  o Ferritin
• In patients with abdominal pain or vomiting:
  o CMP
  o Lipase
• In patients with urinary symptoms or no evident source of infection:
  o UA/UC from clean catch or bag; NO catheterization
• In patients with systemic onset juvenile idiopathic arthritis or any suspicion of macrophage activation syndrome:
  o CMP
  o Ferritin
  o D-dimer
  o Fibrinogen
• In patients with juvenile dermatomyositis
  o AST
  o CK
  o LDH
  o Aldolase

Imaging:
• CXR if respiratory symptoms

Medications/interventions:
• Severely ill-appearing or toxic patients:
  o Supportive care for sepsis as needed
    ▪ IV NS Bolus 20 ml/kg rapid push x 3 if needed to support circulation
- Additional circulatory support per sepsis guideline
  - IV antibiotics as soon as possible:
    - Vancomycin 15 mg/kg IV AND
    - Cefepime 50 mg/kg IV OR
    - Meropenem 20 mg/kg/ IV for penicillin or cephalosporin allergy
- Well-appearing patients who ARE neutropenic:
  - Cefepime 50 mg/kg IV
- Well-appearing patients who are NOT neutropenic:
  - No clear indication for antibiotics. Discuss need with on call Pediatric Rheumatologist.
- Acetaminophen (Tylenol) prn fever
  - 15 mg/kg PO/IV (no PR medications in these patients)
- Ibuprofen (Motrin) prn fever if not already on scheduled NSAID
  - 10 mg/kg PO

Consultations:
- Pediatric Rheumatology fellow or attending on call

Reassessments:
- Continuous cardiorespiratory monitoring with pulse ox for ill-appearing patients
- VS with BP q1h for other patients

Differential diagnosis:
- Fever and neutropenia
- Sepsis/septic shock
- Pneumonia
- UTI
- Line infection
- Viral illness
- Septic joint or osteomyelitis
- Macrophage activation syndrome

Discharge or Admission criteria:
- PICU
  - Intubated patient
  - Vital sign instability
  - Sepsis/septic shock requiring significant resuscitation
- Inpatient floor
  - Ill-appearing or need for further workup/monitoring
  - Concern for respiratory compromise
  - Poor PO intake
  - Rheumatologist recommendation
- Discharge home
  - ANC > 500 (Discuss with Pediatric Rheumatologist whether admission is indicated in patients with ANC < 500)
  - Well-appearing
  - Tolerating PO intake
- Reliable family with transportation and access to close (usually 1 day) follow-up
- Agreement of rheumatologist

**Quality measures:**
- First dose of IV antibiotics within 60 minutes for any ill-appearing or neutropenic patients.
- Documented discussion with Pediatric Rheumatologist within 90 minutes for any well-appearing patient who is NOT neutropenic.
- Blood cultures (as outlined above) prior to antibiotics.