**Adult onset Still's disease**
- systemic inflammatory disease, unknown etiology
- dx of exclusion

**Fever:**
- quotidian or double quotidian
- highest temps usually in late afternoon or early evening

**Sore throat:**
- onset of disease usually heralded by sore throat and constitutional manifestations
- sore throat is cardinal sign! May be a/w odynophagia – throat cxs neg, viral serologies nondiagnostic, abx ineffective

**Arthralgia/arthritis**
- 64-100% of patients – flares during fever spikes
- knees, wrists, ankles, and elbows MC

**LAD:**
- 44-90%
- can raise suspicion of lymphoma initially

**Labs**
- ESR elevation
- WBC>15K
- LFTs elevated (transaminitis) in 75%
- Anemia of chronic disease
- RF and ANA neg
- **High ferritin** seems characteristic
  o >2000 in 38%
  o higher ferritin = greater disease activity
  o Ddx of high ferritin: adult Still’s, HLH (hemophagocytic lymphohistiocytosis (excessive immune activation)), disseminated histo, lymphoma

**Diagnosis (of exclusion):**
- Yamaguchi’s criteria most widely used and most sensitive (93.5%)
- 2002 – new criteria by Fautrel et al – 2 new markers: ferritin and its glycosylated fraction (sens 80.6%, spec 98.5%)

<table>
<thead>
<tr>
<th>Table 2: Diagnostic criteria for adult onset Still's disease</th>
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<tbody>
<tr>
<td><strong>Yamaguchi</strong></td>
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<tr>
<td>Major</td>
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<tr>
<td>Arthralgia &gt;2 weeks</td>
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<tr>
<td>Fever &gt;39 intermittent ≥ 1 week</td>
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<tr>
<td>Typical rash</td>
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<tr>
<td>WBC &gt;10000 (&gt;80% granulocyte)</td>
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<tr>
<td>Minor</td>
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<tr>
<td>Sore throat</td>
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<tr>
<td>Lymphadenopathy and/or splenomegaly</td>
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<tr>
<td>RF and ANA: Neg</td>
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<td><strong>Abnormal LFT</strong></td>
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<td><strong>ANA:</strong> antinuclear antibody; RF: rheumatoid factor; LFT: liver function test, Neg: negative, RES: reticuloendothelial system.</td>
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**Treatment:**
- ASA or NSAIDs as initial treatment – but response rate 20-25%
  o Monitor LFTs
- Steroids – 95% efficacy (use prednisolone)
- DMARDs for maintenance, ?TNF-blocking agents