**Rheumatoid factor-negative form**

- Affects boys and girls equally up to age 9 years; after that girls are more commonly affected.
- Adult-onset Still's disease is rare:
  - Acute phase response,
  - Elevated serum ferritin
  - Tests for RF & ANA are negative.

1. Fever, 2. Soft-tissue swelling (non-erosive) 3. Arthritis / Arthralgia / Myalgia
4. Iritis 5. Polyarthritis
6. Generalized lymphadenopathy
7. Dan Carrot / Polyuria
8. Raised ESR & CRP.

**Systemic JIA (Still's disease)**

<table>
<thead>
<tr>
<th>Oligoarthritis (≤ 4 joints) negative</th>
<th>Polyarthritis (≥ 5 joints) negative</th>
<th>Enthesitis-related</th>
<th>Psoriatic arthritis</th>
</tr>
</thead>
<tbody>
<tr>
<td>1- Arthritis:</td>
<td>1-Arthritis:</td>
<td>Same as adult disease (See below)</td>
<td></td>
</tr>
<tr>
<td>- Affects large joints, knuckles, wrists, ankles &amp; feet initially, eventually larger joints</td>
<td>- Similar distribution to RF negative form</td>
<td>- Joint erosions</td>
<td>- Rare:</td>
</tr>
<tr>
<td>- Eventually it may affect:</td>
<td>- It can be a destructive arthritis</td>
<td>- Enthesitis</td>
<td>- Reactive arthritis</td>
</tr>
<tr>
<td>- Cervical spine</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>- Temporomandibular joints</td>
<td></td>
<td></td>
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<tr>
<td>- Elbows</td>
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<td></td>
</tr>
</tbody>
</table>

**Investigations**

- X-ray: Investigations
- Clinical: Investigations
- 1- Arthritis:
- 2- Polyarthritis
- 3- Arthritis / Arthralgia / Myalgia
- 4- Iritis
- 5- Polyarthritis
- 6- Generalized lymphadenopathy
- 7- Dan Carrot / Polyuria
- 8- Raised ESR & CRP.
- 9- Neutrophilia / Thrombocytosis.

**The principles of management are similar to those in adult inflammatory disease:**
- Systemic JIA: 1- Corticosteroids and 2- methotrexate are required / 1- TNF blockers, 2- is inhibitors. 3- tocolizumab

**Autoimmune-negative**

<table>
<thead>
<tr>
<th>ANA-positive (Anti-Nucleolar antibodies)</th>
<th>RF-negative, ANA-positive</th>
<th>RF-positive, ACAPA-positive</th>
<th>HLA-B27-positive</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rheumatoid arthritis</td>
<td>Rheumatic arthritis</td>
<td>Enteropathic arthritis</td>
<td>Feltty's syndrome</td>
</tr>
</tbody>
</table>

**Feltty's syndrome**

If a patient has the following features:

- A 3-month history of abdominal pain, fever, and weight loss
- Decreased appetite
- Anemia
- Leukopenia
- Thrombocytopenia
- Elevated ESR and CRP
- Abnormal liver function tests
- Positive ANA and RF

- Extra-abdominal manifestations
  - Arthritis
  - Erythema nodosum
  - Gastrointestinal ulcers
  - Pulmonary manifestations
  - Neurological involvement

**Behcet's syndrome**

If a patient has the following features:

- Recurrent oral ulcers
  - Aphthous ulcers, minor 
  - Major aphthous ulcers
  - Recurrent genital ulcers
  - Erythema nodosum
  - Palmar-plantar pustulosis
  - Acroosteolysis
  - Retinal vasculitis
  - Arthritis

**Signs 

- **Rheumatic arthritis**

- Acute anterior uveitis
- B - Oligoarthritis (Persistent).
- 2- Oligoarthritis (Extended).
- - Oligoarthritis specifies to affect many more joints after around 6 weeks, this form can be destructive.

3- Asymmetrical arthritis:
- Arthritis involves small joints of the hands, wrists, ankles & feet initially, eventually larger joints, it may also affect:
- Cervical spine
- Temporomandibular joints
- Elbows
- 2- Uveitis (with or without ANA)

4- Ankylosing spondylitis:
- Ankylosing spondylitis is erosive but the erosions are not symmetrical.
- It targets finger joints and toes.
- Marked periarticular osteolysis and bone shortening ('telescopic' fingers).
- It resembles psoriasis arthropathy.

5- Psoriatic arthritis:
- It targets finger joints and toes.
- Marked periarticular osteolysis and bone shortening ('telescopic' fingers).
- It resembles psoriasis arthropathy.

6- Enteropathic arthritis:
- It targets finger joints and toes.
- Marked periarticular osteolysis and bone shortening ('telescopic' fingers).

7- IgA vasculitis:
- It targets finger joints and toes.
- Marked periarticular osteolysis and bone shortening ('telescopic' fingers).

8- Behcet's syndrome:
- Recurrent oral ulcers
  - Aphthous ulcers, minor 
  - Major aphthous ulcers
  - Recurrent genital ulcers
  - Erythema nodosum
  - Palmar-plantar pustulosis
  - Acroosteolysis
  - Retinal vasculitis
  - Arthritis

- Autoimmune-negative