- **Henoch-Schonlein purpura:**
  - It is an IgA-mediated vasculitis common in males with median age of onset at 5 years. It is usually preceded by an URTI.
  - **Clinical features:**
    ✓ **Skin:** non-thrombocytopenic palpable purpura especially on buttocks and lower extremities. Edema might also be found in feet, scrotum, hand and scalp.
    ✓ **Joints:** arthralgia or arthritis in 80% of patients (commonly knee and ankle joints are involved).
    ✓ **GI:** colicky abdominal pain, GI bleeding and increased risk of intussusceptions.
    ✓ **Renal:** microscopic/macroscopic hematuria (80%); nephrotic syndrome (rare).

- **Investigations:**
  ✓ Notice that diagnosis is mainly based on history and physical examination.
  ✓ Normal platelet count!
  ✓ Increased serum IgA (only in 50% of patients).

- **Management:** steroids. Most patients will recover within 4 weeks.

- **Kawasaki disease:**
  - It is an acute febrile vasculitis occurring more in males of Asian ethnicity at a mean age of 18 months – 24 months.
  - **Diagnostic criteria:**
    ✓ Fever ≥ 5 days with four of the following “CREAM”:
      - **C:** Conjunctivitis (bilateral, non-exudative).
      - **R:** Rash (erythematous, maculopapular).
      - **E:** Extremities (peeling 1 week after onset of fever).
      - **A:** Adenopathy (cervical, unilateral).
      - **M:** Mucosal changes (strawberry-tongue).
- **Complication**: coronary artery aneurysm occurring in 20% of patients. Notice that even if they are large, they will regress with treatment and mortality rate is < 1%. Another complication is hydrops of gallbladder.

- **Clinical course of the disease, investigations and management**:

<table>
<thead>
<tr>
<th></th>
<th>Acute phase (1-2 weeks)</th>
<th>Sub-acute phase (weeks-months)</th>
<th>Convalescent phase (weeks-years)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Investigations</td>
<td>↑ESR, ↑CRP</td>
<td>↑platelet count</td>
<td>Lab values normalize within 6-8 weeks</td>
</tr>
<tr>
<td>Management</td>
<td>IV immunoglobulin + high-dose aspirin (for anti-inflammatory effect)</td>
<td>IV immunoglobulin + low-dose aspirin (for anti-platelet effect)</td>
<td>Continue low-dose aspirin only if aneurysm remains</td>
</tr>
</tbody>
</table>

- **Juvenile Rheumatoid Arthritis (JRA)**:
  - It is a chronic inflammation of joints in children commonly occurring in females between the age of 1-3 years.
  - **Classification**:

<table>
<thead>
<tr>
<th>Pauciarticular (≤ 4 joints are involved); not necessarily symmetrical; 40% of cases</th>
<th>Polyarticular (&gt; 4 joints are involved); symmetrical; 40% of cases</th>
<th>Systemic (still’s disease); 20% of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Sub-classified to</strong></td>
<td><strong>Sub-classified to</strong></td>
<td><strong>High spiking fevers (&gt; 39C) accompanied with salmon-colored rash.</strong></td>
</tr>
<tr>
<td>✓ Early-onset (1-5 years):</td>
<td>✓ RF-negative: presenting early and late in childhood.</td>
<td>✓ Hepatosplenomegaly.</td>
</tr>
<tr>
<td>✓ Females</td>
<td>✓ RF-positive:</td>
<td>✓ Lymphadenopathy.</td>
</tr>
<tr>
<td>✓ Positive ANA (75%)</td>
<td>✓ &gt; 8 years.</td>
<td></td>
</tr>
<tr>
<td>✓ 50% have chronic uveitis</td>
<td>✓ Higher risk of severe arthritis.</td>
<td></td>
</tr>
<tr>
<td>✓ Late-onset (&gt; 8 years):</td>
<td></td>
<td></td>
</tr>
<tr>
<td>✓ Males</td>
<td></td>
<td></td>
</tr>
<tr>
<td>✓ HLA-B27</td>
<td></td>
<td></td>
</tr>
<tr>
<td>✓ Involvement of hip and sacroiliac joints</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Investigations:
- Hypochromic microcytic anemia (anemia of chronic disease).
- ↑ ESR, CRP and platelets.
- RF (-) in majority of patients.

Management: NSAID’s (↓ pain and inflammation).

Systemic Lupus Erythematosus (SLE):
- It is a multisystem autoimmune disease in which there is immune complex-mediated vasculitis (type-III hypersensitivity). Common in females after 10 years of age.
- Diagnostic criteria: 4 of these 11 criteria will provide a sensitivity and specificity of 96% “SOAP BRAIN MD”:
  - S: Serositis (pleuritis and inflammation of pericardium).
  - O: Oral ulcers.
  - A: Arthritis (non-erosive, migratory and transient).
  - P: Photosensitivity.
  - B: Blood cytopenias (hemolytic anemia, thrombocytopenia and leucopenia).
  - R: Renal disease.
  - A: ANA-positive.
  - I: Immunoserology abnormalities (anti-dsDNA antibodies).
  - N: Neurologic symptoms (encephalitis, seizures and psychosis).
  - M: Malar rash (butterfly rash).
  - D: Discoid lupus.
- Investigations:
  - ↑ ESR and CRP.
  - Hemolytic anemia, leucopenia and thrombocytopenia.
  - ANA and RF: ↑ in SLE but not specific.
  - Anti-dsDNA and anti-Sm are very specific.
  - Antiphospholipid antibodies: associated with increased risk of thrombotic events → this is treated with low-molecular weight heparin or warfarin.
  - ↓ C3 and C4: indicating immune complex-mediated complement activation.
- Management: glucocorticoids are the mainstay of therapy for children with SLE.
- Prognosis: mortality commonly due to infection (caused by immunosuppression), renal failure or CNS complications.
- **Dermatomyositis:**
  - It is an inflammatory condition which results in progressive proximal muscle weakness that is preceded by characteristic skin findings. It is common in females between the age of 5-14 years.
  - **Clinical features:**
    ✓ Skin findings:
      - Periorbital violaceous heliotrope rash.
      - Gottron’s papules: skin over metacarpal and proximal interphalangeal joints will become erythematous and thickened.
    ✓ Muscle weakness: with positive Gower’s sign (which means that the patient is unable to stand from a sitting position unless climbing his thighs). Muscle weakness can be confirmed with abnormal EMG findings, abnormal muscle biopsy findings or abnormal muscle enzymes.

- **Management:**
  ✓ Corticosteroids.
  ✓ Vitamin D and calcium: to prevent osteopenia (resulting from steroid therapy) and reducing frequency of fractures.

- **Rheumatic fever:**
  - It is an autoimmune complication of pharyngitis caused by Group A β-Hemolytic Streptococcus GABHS (Streptococcus pyogenes). Commonly affecting children between the age of 5-15 years.
  - **Diagnosis:** through Jones criteria (2 major or 1 major + 2 minor):
    | Major criteria | Minor criteria |
    |----------------|---------------|
    | Migratory asymmetric polyarthritis (70% of patients) | Fever |
    | Carditis (50% of patients): commonly endocarditis which results in insufficiency of left-sided valves (aortic or mitral) | Leukocytosis |
Sydenham’s Chorea
Erythema marginatum
Subcutaneous nodules (rare!)

<table>
<thead>
<tr>
<th>Sydenham’s Chorea</th>
<th>Arthralgia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Erythema marginatum</td>
<td>↑ESR and CRP</td>
</tr>
<tr>
<td>Subcutaneous nodules (rare!)</td>
<td>Prolonged PR interval on ECG</td>
</tr>
</tbody>
</table>

- **Investigations:** ↑ASO titer (indicating recent GABHS infection).
- **Management:**
  - **Eradication of GABHS infection:** single dose IM injection of benzathine penicillin.
  - **Control of inflammation:** NSAIDs.
  - **Corticosteroids used when there is severe cardiac involvement.**

**- Lyme disease:**
- It is caused by an infection with the spirochete Borrelia burgdorferi that is transmitted by a tick bite from Ixodes species which has to be attached to skin for more than 36-48 hours for transmission to occur.
- **Clinical features:**

<table>
<thead>
<tr>
<th>Early disease</th>
<th>Late disease</th>
</tr>
</thead>
</table>
| **Early localized disease (1-4 months):**
  - Erythema migrans which is annular and target-like.
  - Constitutional symptoms: fever, fatigue, myalgias and arthralgias.
| The hallmark is arthritis |
| **Early disseminated disease (5-12 months):**
  - Multiple secondary erythema migrans (smaller than initial lesion).
  - Neurologic manifestations: facial nerve palsy (3%) and aseptic meningitis (1%) |

- **Investigations:** serology measuring antibodies to B. burgdorferi by ELISA and conformation by Western blot.
- **Management:** amoxicillin or doxycycline (for children ≥ 9 years).