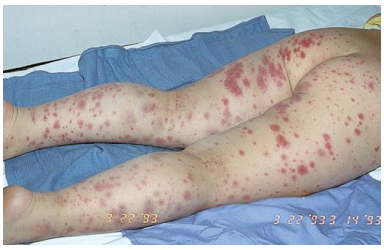
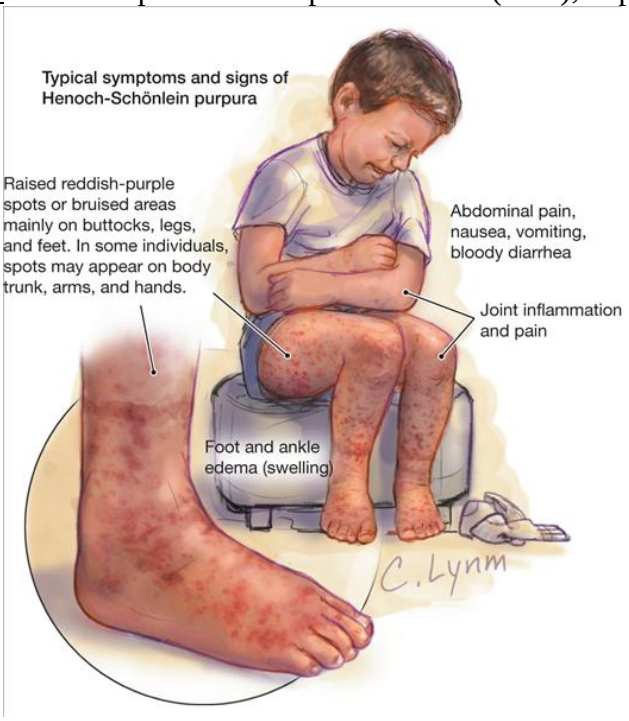
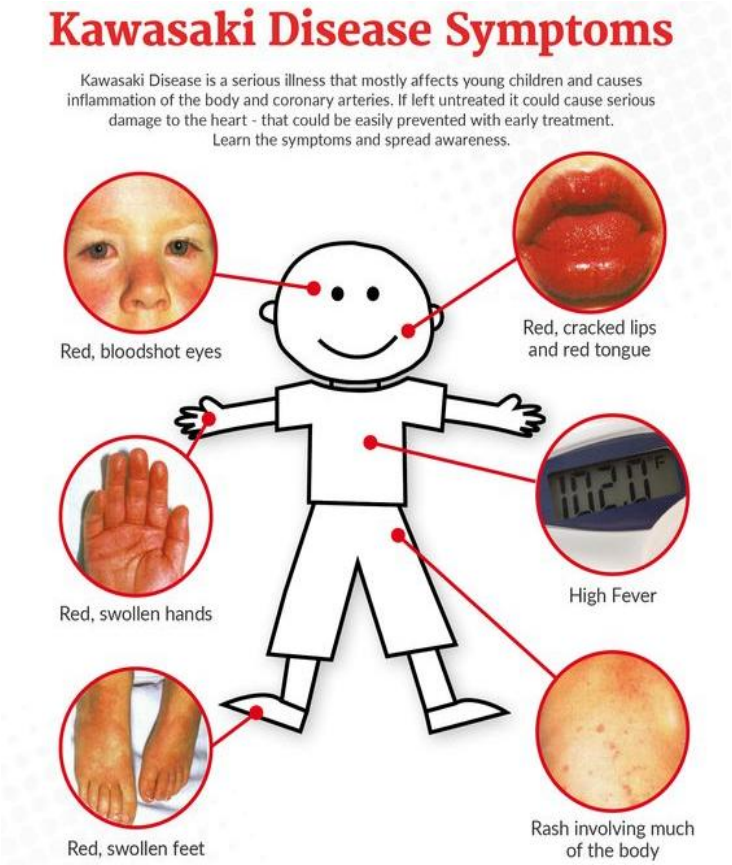
- **Henoch-Schonlein purpura:**
  - **It is an IgA-mediated vasculitis common in males with median age of onset at 5 years. It is usually preceded by an URTI.**
  - **Clinical features:**
    - ✓ Skin: non-thrombocytopenic palpable purpura especially on buttocks and lower extremities. Edema might also 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 (eryhtematous, 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:**

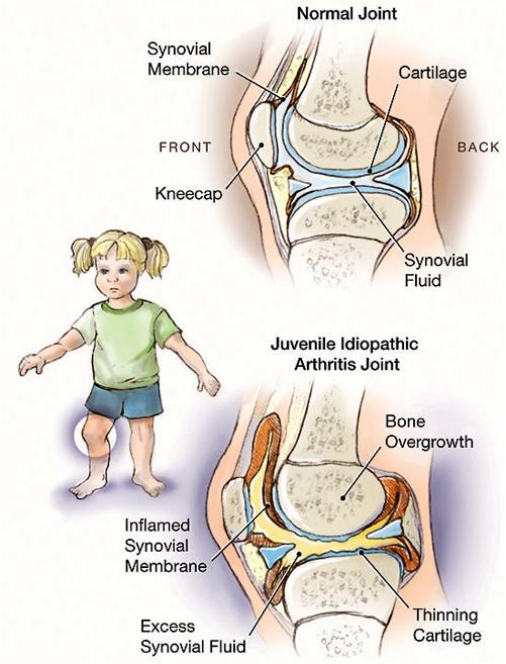
	Acute phase (1-2 weeks)	**Sub-acute phase (weeks-months)**	**Convalescent phase (weeks-years)**
Investigations	↑ESR, ↑CRP	↑platelet count	Lab values normalize within 6-8 weeks
Management	IV immunoglobulin + high-dose aspirin (for anti-inflammatory effect)	IV immunoglobulin + low-dose aspirin (for anti-platelet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

- **Juvenile Rheumatoid Arthritis (JRA):**

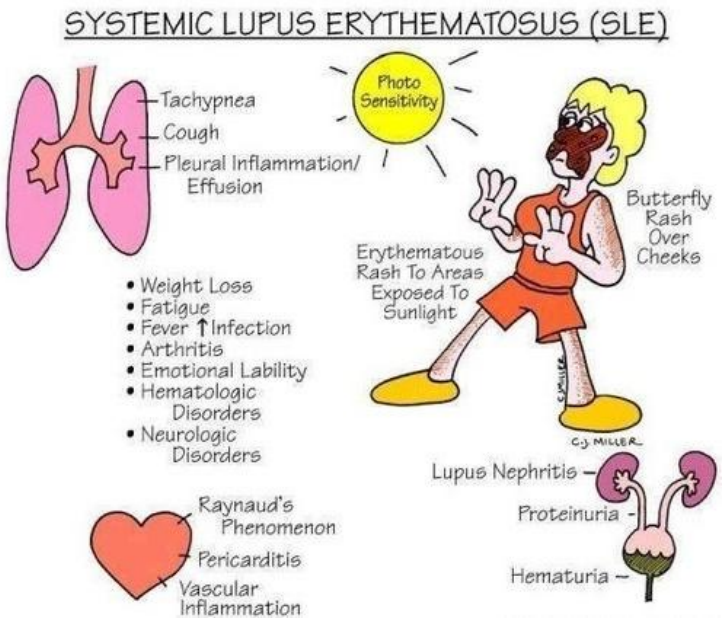
- **It is a chronic inflammation of joints in children commonly occurring in females between the age of 1-3 years.**
- **Classification:**

Pauciarticular (≤ 4 joints are involved); not necessarily symmetrical; 40% of cases	**Polyarticular (> 4 joints are involved); symmetrical; 40% of cases**	**Systemic (still's disease); 20% of cases**
Sub-classified to: • Early-onset (1-5 years): ✓ Females ✓ Positive ANA (75%) ✓ 50% have chronic uveitis • Late-onset (> 8 years): ✓ Males ✓ HLA-B27 ✓ Involvement of hip and sacroiliac joints	**Sub-classified to:** • RF-negative: presenting early and late in childhood. • RF-positive: ✓ > 8 years. ✓ Higher risk of severe arthritis.	• High spiking fevers (> 39C) accompanied with salmon-colored rash. • Hepatosplenomegaly. • Lymphadenopathy.

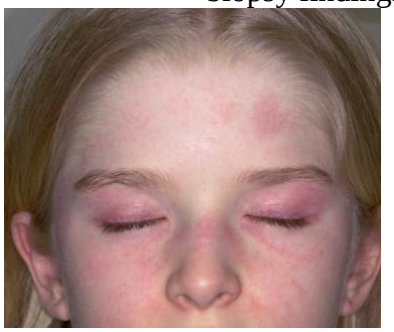
- **Investigations:**
  - ✓ Hypochromic microcytic anemia (anemia of chronic disease).
  - ✓ ↑ ESR, CRP and platelets.
  - ✓ RF (-) in majority of patients.
- **Management**: NSAID's (↓ pain and inflammation).

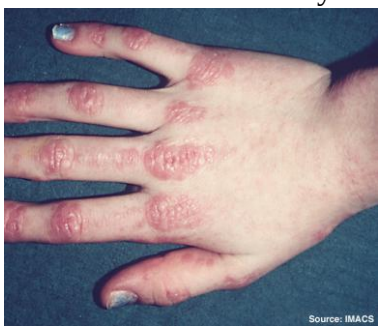


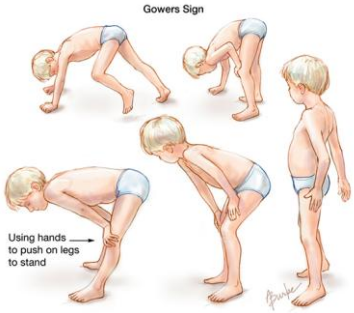
- **Systemic Lupus Erythematosus (SLE):**
  - **It is a multisystem autoimmune disease in which there is immune complex-mediated vasculitis (type-III hypersensitivity). Common in females after 10 years of age.**
  - **Diagnostic criteria**: 4 of these 11 criteria will provide a sensitivity and specificity 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.



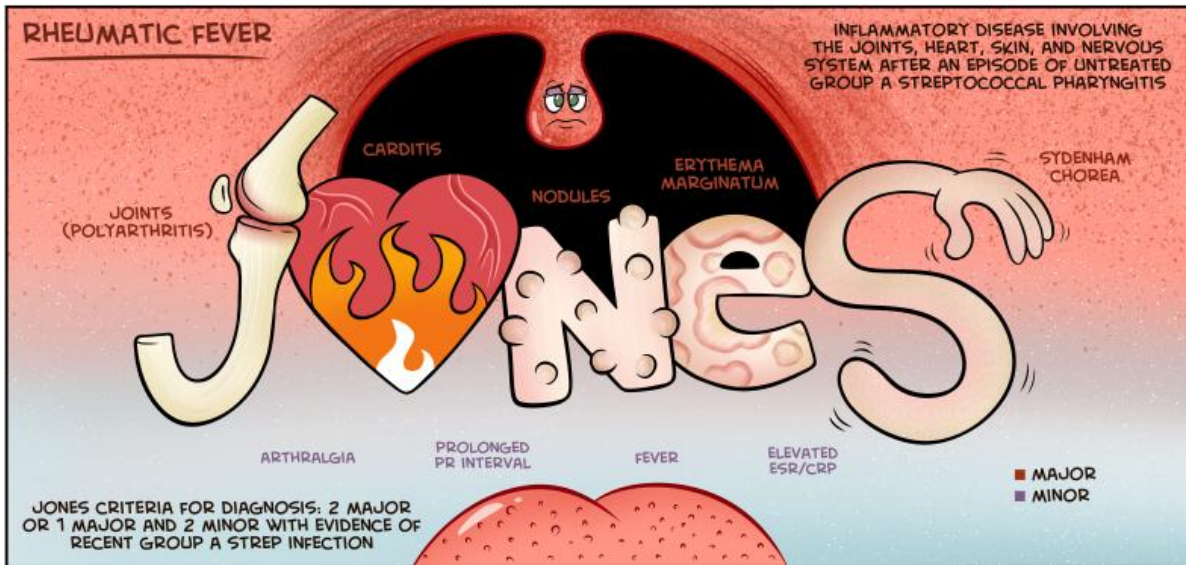




  - **Management:**
    - ✓ Corticosteroids.
    - ✓ Vitamin D and calcium: to prevent osteopenia (resulting from steroid therapy) and reducing frequency of fractures.
- **Rheumatic fever:**
  - **It is an autoimmune complication of pharyngitis caused by Group A β-Hemolytic Streptococcus GABHS (Streptococcus pyogens). 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	Arthralgia
Erythema marginatum	↑ESR and CRP
Subcutaneous nodules (rare!)	Prolonged PR interval on ECG



- **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:**

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

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