Connective tissue disease

Review of system in the differential diagnosis of childhood joint pain and swelling → Its common to have a child with J pain & swelling.

Dermatologic	SLE	Malar rash and hair loss
	Dermatomyositis	Gottron's papules
	Systemic JIA	Evanescent pink macular rash
	HSP	Lower extremity purpuric lesions

Ophthalmologic	Oligoarthritis or psoriatic JIA	Asymptomatic chronic anterior uveitis
	Enthesitis related arthritis	Acute symptomatic uveitis (pain, redness)
	Kawasaki disease	Conjunctival injection without discharge
	Sjogren's syndrome	Dry eyes with keratitis
Oral	SLE	Painless oral ulcers on palate
	Behcet Disease	Large extremely painful oral ulcers

Respiratory	CF or immunodeficiency	Recurrent pneumonia
	Wegener's granulomatosis	Destructive upper tract lesions
	SLE or systemic JIA	Pleuritis
	SLE or scleroderma	Interstitial lung disease
	Churg-Strauss syndrome	Eosinophilic pneumonia
Cardiovascular	ARF or endocarditis	New heart murmur
	SLE, systemic JIA, or ARF	Pericarditis
	SLE or scleroderma	Raynaud phenomenon
	Takayasu arteritis	Absent pulses

١٤٤١/١/١٤

* HSP → not every pt w/ it is admitted
↳ 100% of pts have purpuric skin rash
↳ most common presentation after the rash → arthritis or arthralgia
↳ complications for admission → ① Renal (GN) → Do urinalysis

② abdominal pain (Intussusception)
↳ Emergency
↳ come w/ colicky pain, seizure (rarely)
Bloody urine
⇒ treated w/ NSAIDs, oral prednisolone
(in GI manifestations, CNS, Renal)

Gastrointestinal	IBD, SLE, or vasculitis	Weight loss or poor growth
	IBD	Diarrhea and abdominal pain
	Reactive arthritis	Preceding infectious gastroenteritis
	HSP (vasculitis)	Intermittent colicky abdominal pain
Genitourinary	Gonococcal arthritis	Pustular urethritis or cervicitis
	Reactive arthritis	Non-gonococcal urethritis
	Behcet disease or IBD	Large painful genital ulcerations

+ Joint Pain ⇓

Renal manifestations of HSP appear after 4 months → so you have to follow the pt-up

Hematologic	SLE or hemoglobinopathy (eg, SCD)	Hemolytic anemia
	SLE	Pancytopenia
	Bleeding disorders	Hemarthrosis
Neurologic	SLE	Seizures and psychosis
	SLE or fibromyalgia	Difficulty concentrating
	SLE, vasculitis, or hypercoagulability	Stroke
	Vasculitis	Asymmetric polyneuropathy
	Dermatomyositis and polymyositis	Proximal muscle weakness

- Rheumatic diseases are defined by the constellation of results of the physical examination, autoimmune marker and other serologic tests, tissue pathology, and imaging.

-JIA

- SLE

- JUVENILE DERMATOMYOCITIS

Juvenile rheumatoid arthritis (JRA)

Juvenile idiopathic arthritis (JIA),

Juvenile rheumatoid arthritis (JRA) Juvenile idiopathic arthritis (JIA),

- **is a common, rheumatic disease of children and a major cause of chronic disability.**
- **It is characterized by a synovitis of the peripheral joints manifesting in soft tissue swelling and effusion.**

JRA classification

- Systemic onset JRA (formerly called **Still's disease**)
  - arthritis with fever and rash.
- Pauciarticular (**Oligoarthritis**)

(1-4 J) < 5 joints after 6 months of illness.

- Polyarticular (**Polyarthritis**)

  > 4 joints after 6 months of illness.

١١/٠١/١٤٤١

Psoriatic arthritis	Arthritis and psoriasis, or arthritis and at least 2 of the following: 1. Dactylitis[‡] 2. Nail pitting[§] and onycholysis 3. Psoriasis in a 1st-degree relative
Enthesitis-related arthritis	Arthritis and enthesitis,[‖] or arthritis or enthesitis with at least 2 of the following: 1. Presence of or a history of sacroiliac joint tenderness or inflammatory lumbosacral pain or both[¶] 2. Presence of HLA-B27 antigen 3. Onset of arthritis in a male >6 yr old 4. Acute (symptomatic) anterior uveitis 5. History of ankylosing spondylitis, enthesitis-related arthritis, sacroiliitis with inflammatory bowel disease, Reiter syndrome, or acute anterior uveitis in a 1st-degree relative
Undifferentiated arthritis	Arthritis that fulfills criteria in no category or in ≥2 of the above categories.

inflamation at the site of insertion of a tendon

we can see in this picture :-

① right knee swelling

② flexion contracture ~~position~~ posture to minimize the pain (rest position)

- Eighteen-month-old girl with arthritis in her right knee. Note the flexion contracture of that knee.

Criteria for the Classification of Juvenile Rheumatoid Arthritis
Age at onset: <16 yr
Arthritis (swelling or effusion, or the presence of 2 or more of the following signs: limitation of range of motion, tenderness or pain on motion, increased heat) in ≥1 joints
Duration of disease: ≥ 6 wk
Onset type defined by type of articular involvement in the 1st 6 mo after onset:
Polyarthritis: ≥5 inflamed joints
Oligoarthritis: ≤4 inflamed joints
Systemic disease: arthritis with a characteristic intermittent fever
Exclusion of other forms of juvenile arthritis

كتك اهم بال criteria ←
كم عدد المفاصل ال
involved →
بستنى ٦ أشهر وبعد
ال Joints
involved

ETIOLOGY.

- **Unknown etiology**
- Autoimmune disease AND Genetic susceptibility factors
- Environmental (infection, trauma,and stress).
- Possible external triggers include viruses (parvovirus B19, rubella, Epstein-Barr virus),
- Host hyperreactivity to specific self antigens (type II collagen), and enhanced T-cell reactivity to bacterial or mycobacterial heat shock proteins.

EPIDEMIOLOGY.

- Describe in all races and geographical areas
- The incidence of JRA is ≈13.9/100,000 children/yr among white children ≤15 yr of age, with a prevalence of ≈113/100,000 children.
- Female predominance 2:1

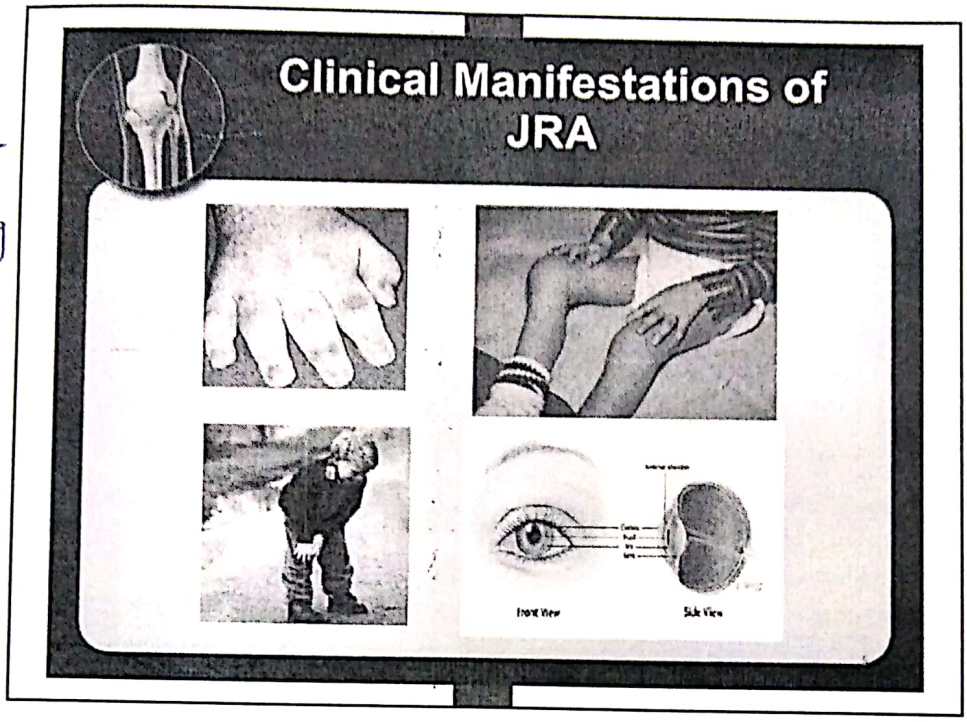
CLINICAL MANIFESTATIONS

can be acute or subacute

Initial symptoms

- subtle or acute
- morning stiffness
- easy fatigability, particularly after school in the early afternoon
- joint pain
- joint swelling
- Joint stiffness
- limp
- Restriction of movement
- Eye symptoms
- Systemic manifestation → like fever, rash, organomegaly, thrombocytopenia

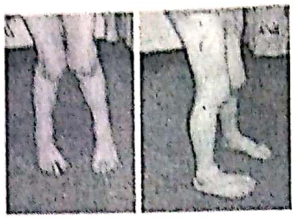
* pediatric presentation mainly ⇒ fever of unknown origin
usually Rheumatic cause.



u can see:-
1- Joint swelling
2- Limitation of Movement, tired fatigue,
3- ant. uveitis.

Oligoarthritis (pauciarticular disease)

- predominantly affects the joints of the lower extremities, such as the knees and ankles .
- only a single joint is involved at onset.
- Isolated involvement of upper extremity large joints is not characteristic of this type of onset.
- Involvement of the hip is almost never a presenting sign of JRA.



Polyarthritis (polyarticular disease)

- is generally characterized by involvement of both large and small joints of both upper and lower extremities .
- As many as 20–40 joints may be affected in the more severely involved child, although inflammation of only ≥5 joints is required as a criterion for classification of this type of onset.
- Rheumatoid nodules on the extensor surfaces of the elbows and over the Achilles tendons, while unusual, are associated with a more severe course
- Micrognathia reflects chronic temporomandibular joint disease.
- Cervical spine involvement of the apophyseal joints occurs frequently with a risk of atlantoaxial subluxation and potential neurologic sequelae.

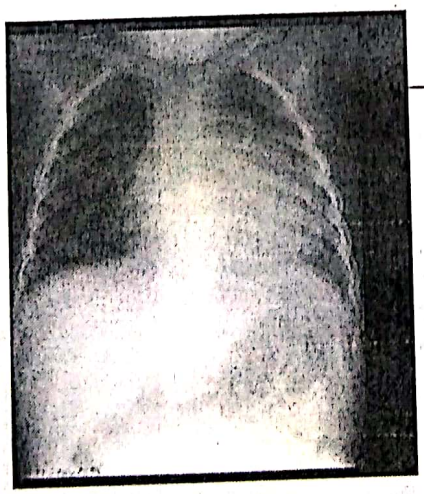
Seen more in adults, but if you see it in younger children → indicates more severe form of the disease.

Fever ≥ on/off, almost daily, Responds to Antibiotics,

Table 1: Systemic JIA ILAR Classification Criteria[1]

daily.

1. Fever ≥2 weeks, quotidien in pattern (≥39°C at least once a day and returns to ≤37°C), documented daily for ≥3 days
2. Arthritis in ≥1 joint (for ≥6 weeks)
3. At least one of the following:
   - >Evanescent erythematous rash;
   - >Generalized lymph node enlargement;
   - >Hepatomegaly and/or splenomegaly; or
   - >Serositis



- Child with pericardial effusion due to systemic onset juvenile idiopathic arthritis (JIA).
- (serositis)

١١/٠١/١٤٤١

Elapsing rash ←
(يظهر بسرعة ويروح
بروح بعد مرور
عدة ساعات)

Systemic juvenile idiopathic arthritis

A) rash

DIAGNOSIS

- no one pathognomonic finding
- The classic intermittent fever , the typical rash and objective arthritis highly suggestive of systemic-onset JRA.
- The diagnosis is based on a history compatible with inflammatory joint disease and a physical examination that confirms the presence of arthritis

↳ most importantly → exclusion of other causes

* If a baby had 2 week onset fever + organomegaly + arthralgia + lymphadenopathy → think about infection, malignancy
DDx — most common — most serious

Laboratory abnormalities

- elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP),
- leukocytosis,
- thrombocytosis,
- anemia of chronic disease. → normocytic, hypochromic or rarely → hypocytic hypochromic
- anti-cyclic citrrullinated peptide (ccp) antibody has very high specificity for rheumatoid arthritis may detected before RF (poor prognosis)
- Carriage of HLA27 antigen is associated with increased risk of developing enthesitis –associated arthritis
- ANA is associated with increased risk of iridocyclitis in pt with oligoarthritis
- RF is positive only in 5% of pt when poly articular disease occur after 8 yrs of age

	Systemic onset JIA	Pauciarticular onset JIA	Polyarticular onset JIA
Percent of JIA patients	10 to 15	50	30 to 40
Sex	F = M	F > M	F > M
Age	any <17 years	peak 2 to 3 years, rare >10	peaks 2 to 5, 10 to 14 years
Joints	any	large joints, but rarely hips	any, rare to start in hip
Fever, rash, lymphadenopathy, hepatosplenomegaly	yes	no	no
Uveitis	rare	20 percent, esp ANA +	less frequent

	Systemic onset JIA	Pauciarticular onset JIA	Polyarticular onset JIA
Laboratory abnormalities			
- Leukocytosis	marked	no	no
- Anemia	marked	no	mild
- Elevated ESR	marked	mild	mild
- ANA	absent	low titer common	low titer common in younger
- Rheumatoid factor	rare	absent	10 to 20 percent in those >10 years
Destructive arthritis	>50 percent	rare	>50 percent
Disease modifying drugs	commonly used	rarely used	commonly used

* Differential diagnosis of joint pain or swelling in children

common presentation

- Trauma
- Irritable hip and transient synovitis
- septic arthritis and osteomyelitis, - fever
- Infection: bacterial ,viral ,Lyme disease
- Hematologic:leukemia ,bleeding diatheses, and hemoglobinopathies , hemophilia.
- Tumor: Musculoskeletal neoplasia (eg, osteosarcoma), lymphoma, and neuroblastoma
- Perthes disease
- Slipped capital femoral epiphysis

- diagnosis of JIA requires the exclusion of all the above diagnoses.

↳ ask if bilateral or unilateral

Management

Its chraic

- No cure but treatable
- Remissions and relapses
- Relieve pain ,reduce inflammation ,preserve joint function ,maintain normal growth and development

↳ They suffer from disability, distruction of the joint structure

USAIDs, ← Management

Methotrixate, Immunosupressant (cayclophosphamide)

- Screen for uveites by periodic slit-lamp ophthalmologic examinations of all patients
- Require multidisciplinary team (MDT) approach
  - dietary evaluation and counseling to ensure appropriate calcium, vitamin D, protein, caloric intake;
  - physical and occupational therapy.
  - A social worker

Medical management

- Nonsteroidal anti-inflammatory drugs (NSAIDs) : Ibuprofen ,Naproxin ,Piroxicam
- Disease modifying anti rheumatic drugs (DMAR)
  - Under supervision of rheumatologist
  - methotrexate the safest, most efficacious, and least toxic given once weekly
  - azathioprine and cyclophosphamide, are reserved for the few children who do not respond to less aggressive therapy.
- Joints steroid injections
- Corticosteroid : oral or IV methylprednisolone
  - Toxicity :-
    - Cushing syndrome, growth retardation, and osteopenia , hyperglycemia, hypertension.
- TNF-α blockers :block the immune protein TNF etanercept infliximab

Systemic lupus erythematosus in children

- Systemic lupus erythematosus (SLE) in children is fundamentally the same disease as in adults with similar etiology, pathogenesis, clinical manifestations, and laboratory findings.
- However, the care of children and adolescents with SLE is very different from that of adults because of the impact of the disease and its therapy on physical and psychological growth and development.

Systemic lupus erythematosis

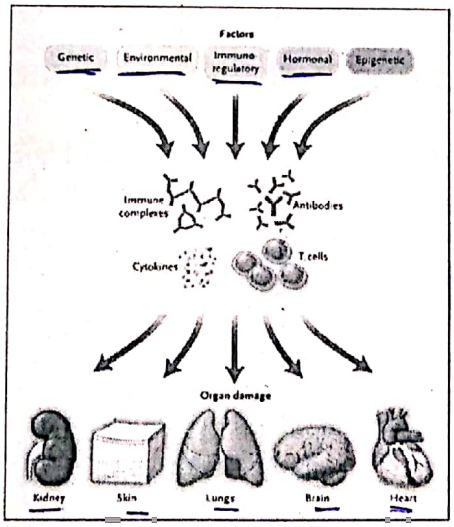
- A chronic inflammatory multi-systemic autoimmune disease characterized by widespread inflammation of blood vessel and connective tissue
- The primary pathology is of persistent polyclonal B-cell stimulation resulting in autoantibody production with wide spread issue antibody production ,with the widespread tissue deposition of immune complex

↳ Its multifactorial

- The pathogenesis of SLE remains largely unknown, but several genetics, hormonal and environmental factors. → multi factorial
- The reported prevalence of SLE in children and adolescents (1-6/100,000) .
- More among African-Americans, Asians, Hispanics, Native Americans, and Pacific Islanders for both adult and pediatric populations.
- SLE predominantly affects females

-Renal → SLE nephritis (proteinurea, hematuria), hypertention, GN.
-Skin → malar rash
-photosensitivety.
-Lungs → serositis, pleuritis
-Brain → psychosis, seizure stroke
-Heart → pericarditis

Overview of the Pathogenesis of Systemic Lupus Erythematosus.



Tsokos GC. N Engl J Med 2011;365:2110-2121.

Table 158-1 | Potential Clinical Manifestations of Systemic Lupus Erythematosus

TARGET ORGAN	POTENTIAL CLINICAL MANIFESTATIONS
Constitutional	Fatigue, anorexia, weight loss, fever, lymphadenopathy
Musculoskeletal	Arthritis, myositis, tendonitis, arthralgias, myalgias, avascular necrosis, osteoporosis
Skin	Malar rash, discoid (annular) rash, photosensitive rash, cutaneous vasculitis (petechiae, palpable purpura, digit ulcers, gangrene, urticaria), livedo reticularis, periungual capillary abnormalities, Raynaud phenomenon, alopecia, oral and nasal ulcers, panniculitis, chilblains, alopecia
Renal	Hypertension, proteinuria, hematuria, edema, nephrotic syndrome, renal failure
Cardiovascular	Pericarditis, myocarditis, conduction system abnormalities, Libman-Sacks endocarditis
Neurologic	Seizures, psychosis, cerebritis, stroke, transverse myelitis, depression, cognitive impairment, headaches, migraines, pseudotumor, peripheral neuropathy (mononeuritis multiplex), chorea, optic neuritis, cranial nerve palsies, acute confusional states, dural sinus thrombosis
Pulmonary	Pleuritis, interstitial lung disease, pulmonary hemorrhage, pulmonary hypertension, pulmonary embolism
Hematologic	Immune-mediated cytopenias (hemolytic anemia, thrombocytopenia or leukopenia), anemia of chronic inflammation, [illegible]
Gastroenterology	Hepatosplenomegaly, pancreatitis, vasculitis affecting bowel, protein-losing enteropathy, peritonitis
Ocular	Retinal vasculitis, scleritis, episcleritis, papilledema, dry eyes, optic neuritis

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11 criteria, should have at least 4/11 for DX

American College of Rheumatology Criteria for the Diagnosis of Systemic Lupus Erythematosus (SLE).

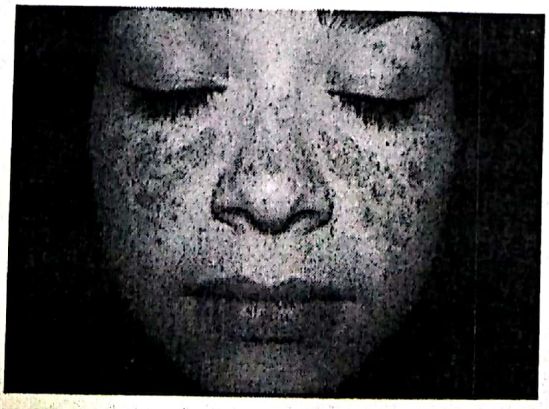
Table 1. American College of Rheumatology Criteria for the Diagnosis of Systemic Lupus Erythematosus (SLE).*

Criterion	Definition
Malar rash	A rash on the cheeks and nose, often in the shape of a butterfly
Discoid rash	A rash that appears as red, raised, disk-shaped patches
Photosensitivity	A reaction to sunlight that causes a rash to appear or get worse
Oral ulcers	Sores in the mouth
Arthritis	Joint pain and swelling of two or more joints
Serositis	Inflammation of the lining around the lungs (pleuritis) or inflammation of the lining around the heart that causes chest pain, which is worse with deep breathing (pericarditis)
Kidney disorder	Persistent protein or cellular casts in the urine
Neurologic disorder	Seizures or psychosis
Blood disorder	Anemia (low red-cell count), leukopenia (low white-cell count), lymphopenia (low level of specific white cells), or thrombocytopenia (low platelet count)
Immunologic disorder	Positive test for anti–double-stranded DNA, anti-Sm, or antiphospholipid antibodies
Abnormal antinuclear antibodies	Positive antinuclear-antibody test

* Four of the 11 criteria are needed for the formal diagnosis of SLE.

ANA

Malar rash



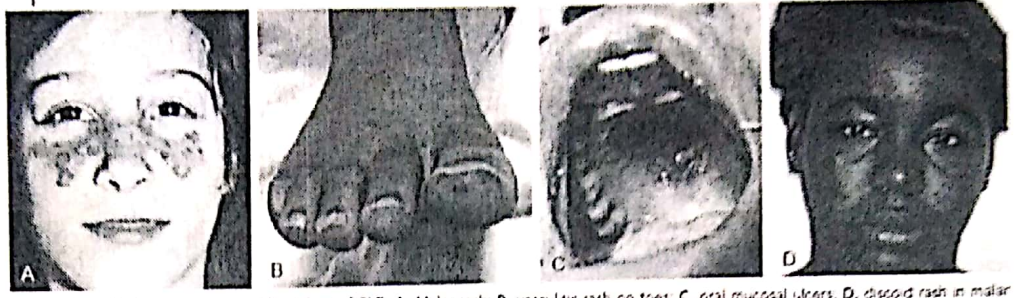


Figure 158-1 Mucocutaneous manifestations of SLE. A, Malar rash; B, vasculitic rash on toes; C, oral mucosal ulcers; D, discoid rash in malar distribution.

لها بالصورة الخاصة بكل واحد.

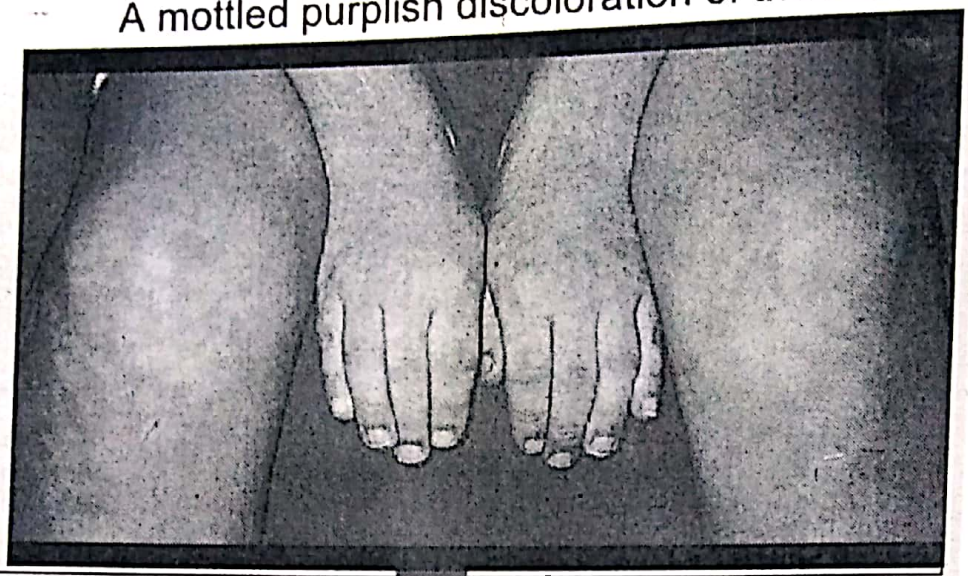
Signs and symptoms

- The disease can affect a wide range of organ systems
- Generalized symptoms such as fever , weight loss , and malaise are common
- Other common signs and symptoms include fever,anorexia ,Raynauds phenomenon , vasculitis , chorea, neuropathy, depression, and cognitive changes

* Its also one of the causes of fever of unknown origin

Livedo reticularis

A mottled purplish discoloration of the skin



Laboratory findings

- Complete blood counts
  - Leukopenia ,anemia ,thrombocytopenia , or pancytopenia
- 15% coombs test- positive
- ESR frequently elevated
- CRP with active SLE is normal but elevated may be due to infectious causes especially bacterial infection
- Urine analysis for proteinuria and hematuria

highly specific ←

Laboratory findings

- Complements :- **C3& C4** are frequently reduced due to increased consumption
- **ANA** is positive in 95-100% of patients usually at titer 1:320 or above
- **Anti double -strand DNA** sensitive and specific , raised particularly in lupus nephritis
- **Anti-smith antibody** are specific for lupus and are related to CNS involvement, positive in only 30% ,lack sensitivity as diagnostic test
- **Antiphospholipid antibodies** found in approximately 50-60% of pediatric SLE patients , increase risk of thrombosis
- **Anticardiolipin antibodies** are detected in up to 50% of children with lupus ,associated with episode of arterial and venous thrombosis

Autoantibodies Found in Systemic Lupus Erythematosus

Antibody	Manifestation
Coombs antibodies	→ Hemolytic anemia
Antiphospholipid antibodies	
Antiphospholipid antibody syndrome	
Lupus anticoagulant	→ Coagulopathy
Antithyroid antibodies	→ Hypothyroidism
Antiribosomal P antibody	→ Lupus cerebritis

Treatment

- 'Mild SLE'
  - nonsteroidal antiinflammatory drugs
  - hydroxychloroquine → anti-malarial → for skin manifestations
- 'Moderate SLE'
  - high-dose glucocorticoids
  - mycophenolate mofetil
- 'Severe SLE'
  - cyclophosphamide – Immunosuppressant
  - prednisone

Complications

Table 158-6	Morbidity in Childhood Lupus
Renal	Hypertension, dialysis, transplantation
Central nervous system	Organic brain syndrome, seizures, psychosis, neurocognitive dysfunction
Cardiovascular	Atherosclerosis, myocardial infarction, cardiomyopathy, valvular disease
Immune	Recurrent infection, functional asplenia, malignancy
Musculoskeletal	Osteopenia, compression fractures, avascular necrosis
Ocular	Cataracts, glaucoma, retinal detachment, blindness
Endocrine	Diabetes, obesity, growth failure, infertility, fetal wastage

Neonatal lupus

↳ happens in children whose moms have lupus

- Clinical manifestations of neonatal lupus include a characteristic annular or macular rash typically affecting the face (especially the periorbita larea), trunk, and scalp
- The rash typically appears within the 1st 6 wk of life after exposure to ultraviolet light and lasts 3-4 mo.
- Infants may also have cytopenias and hepatitis, congenital heart blocks the most feared complication.
- Conduction system abnormalities range from prolongation of the PR interval to complete heart block, with development of progressive cardiomyopathy
- Neonatal lupus results from the passive transfer of maternal immunoglobulin G autoantibodies to the fetus. The vast majority of neonatal lupus cases are associated with maternal anti-Ro (also known as SSA) and anti-La antibodies (also known as SSB).

↳ If a mother w/ lupus gave birth to a baby you have to do [ECG]

↳ for heart complications

also → CBC, Renal function test, liver enzymes

perminant ← heart prob ال ماعدا كلهم بتحسنوا ←

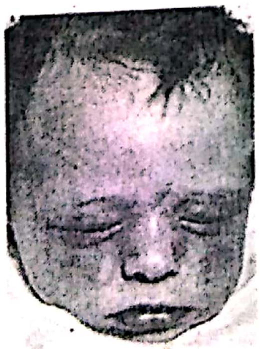


Figure 158-3 Neonatal lupus syndrome. Typical rash, often photosensitive with a malar distribution, appearing as annular plaques with erythema and scaling. Reproduced with written parental permission from Pan C, [illegible]: Neonatal lupus syndrome, Pediatr Child [illegible]

Course and prognosis

- Cutaneous and hematological manifestations transient
- Congenital heart block permanent
- Hepatic fibrosis occasional
- Some risk of SLE in teenage or adult year

Management:

- Symptomatic for transient manifestations
- Heart block may need pace maker

juvenile dermatomyositis and polymyositis

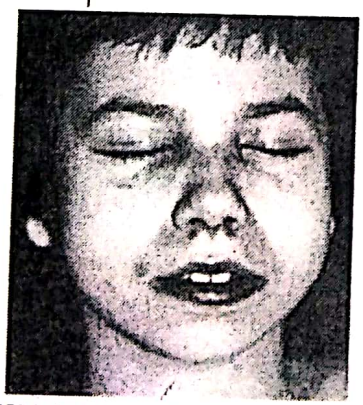


Figure 159-1 The facial rash of juvenile dermatomyositis. There is erythema over the bridge of the nose and malar areas with violaceous (heliotropic) discolorations of the upper eyelids.

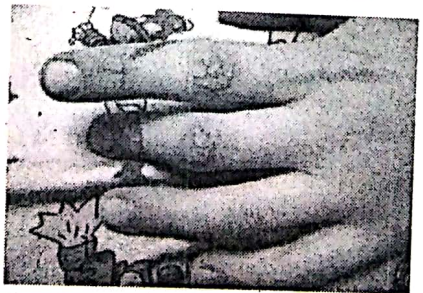


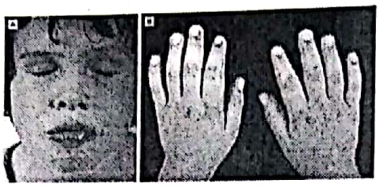
Figure 159-2 The rash of juvenile dermatomyositis. The skin over the metacarpal and proximal interphalangeal joints may be hypertrophic and pale red (Gottron papules).

- Non –suppurative myositis with characteristics skin rash and vasculitis
- Girls more than boys
- Peak incidence 4-10 yrs of age
- juvenile polymyositis involves direct T-cell invasion of muscle fibers similar to that seen in adult polymyositis ,accounts 3-6% of cases.

CLASSIFICATION AND DIAGNOSTIC CRITERIA

- Symmetrical weakness of the proximal muscles
- Characteristic cutaneous changes :-
  - heliotrope dermatitis (reddish-purple rash on the upper eyelids with periorbital edema) and
  - Gottron's papules (erythematous, papulosquamous eruption over the dorsal surfaces of the knuckles)

Clinical images of typical juvenile dermatomyositis



- A) Heliotrope discolouration of the eyelids, and malar or facial erythema and
- (B) scaly, red rash on the knuckles with Gottron's papules.

Table 159-2 Clinical Features of Juvenile Dermatomyositis During the Course of the Disease

FEATURE	%
Muscle weakness → almost all pts	90-100
Dysphagia or dysphonia	13-40
Muscle atrophy	10
Muscle pain and tenderness	30-83
Skin lesions	85-100
Heliotrope rash of eyelids	66-83
Gottron papules	57-91
Erythematous rash of malar/facial area	42-100
Periungual capillary changes	80
Photosensitive rash	5-42
Ulcerations	22-30
Calcinosis	12-30
Lipodystrophy	11-14
Raynaud phenomenon	2-15
Arthritis and arthralgia	22-58
Joint contractures	26-27
Fever	16-46
Gastrointestinal signs and symptoms	8-22
Restrictive pulmonary disease	4-32
Interstitial lung disease	1-7
Cardiac involvement	0-3

Bulbar involvment ←

least manifestation ←

Table 159-1 Diagnostic Criteria for Juvenile Dermatomyositis

Classic rash	Heliotrope rash of the eyelids
	Gottron papules
Plus 3 of the following:	
Weakness	Symmetric
	Proximal
Muscle enzyme elevation (≥1)	Creatine kinase
	Aspartate aminotransferase
	Lactate dehydrogenase
	Aldolase
Electromyographic changes	Short, small polyphasic motor unit potentials
	Fibrillations
	Positive sharp waves
	Insertional irritability
	Bizarre, high-frequency repetitive discharges
Muscle biopsy	Necrosis
	Inflammation

Investigation

- Elevation of the serum level of one of the muscle enzymes
  - Creatine kinase (CK)
  - Lactate *dehydrogenase (LDH)*
  - AST → has muscle & liver origin
    ↳ AST > ALT
- ANA positive in some
- **Electromyography:** useful to distinguish myopathic from neuropathic causes of muscle weakness

Magnetic resonance imaging

- MRI scan of quadriceps muscle can be used in equivocal cases to confirm the presence of inflammatory myositis

Muscle biopsy

- Is indicated in cases of myositis without the pathognomonic rash.
- Muscle biopsy displaying fiber necrosis and inflammation, small vessel occlusive vasculitis

Treatment

- Suppression of inflammatory response and prevention of the loss of muscle function and joint range of motion
- Assessment of the ventilatory effort and swallowing
- Corticosteroids prednisone or pulse methylprednisolone
- Methotrexate.
- In severe cases cyclosporine or cyclophosphamide
- For skin manifestation :Hydroxychloroquine and intravenous immunoglobulin

Prognosis

- Variable
- Usually good with adequate treatment
- Recurrent rate 10-20%
- Small percent develop extensive muscle wasting ,severe contracture and wide spread calcinosis
- Dermatomyositis in children is not associated with increase risk of cancer as in adult

Thank you