COMMON RHEUMATIC DISEASES CONNECTIVE TISSUE DISEASES

Doctor's Notes

Kaplan's notes

Illustrated notes

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CONNECTIVE TISSUE DISEASES
CTD

. Diseases which affect tissue that supports, binds, and protect organs.

. Two major proteins affected:

- **Collagen**
- **Elastin**

major component of tendons, ligaments and skin

CTD

- >100 Diseases. Most are rare
- Most body systems may be affected.
- Most share common symptoms [joint inflammation, fever, rash, weakness, etc.]
- Etiology unknown [genetic, environment, autoimmunity]
- All male and females affected
- All countries.

Clinical presentation

APPROPRIATE RHEUMATOLOGIC HISTORY

- Pain [joint, muscles, limbs]
- Stiffness [morning]
- Loss of function [limping]
- Systemic illness [fever, rash, weakness, mucous membrane involvement, etc.]

EXAMINATION

- FULL CLINICAL EXAMINATION
- Vital signs
- Growth parameter
- Arthritis [MSK]
- Rash [types]
- Other related systems.

Arthritis: redness, hotness, and tenderness
Arthralgia: pain

Hx and PHx are very important because most are syndromes not specific diseases.
No specific laboratory tests

Juvenile Idiopathic Arthritis
JIA

Abbreviations before : J.C.A. in Europe
J.R.A. in U.S.

Features:

1. Onset under **16** years
2. Persistent arthritis in one or more joints
3. Duration
   - three months or longer (Europe)
   - six weeks or longer (U.S.)
4. Exclude other defined causes of arthritis in childhood.

Simply arthritis for more than 6 weeks with exclusion of other causes

Features in the history are gelling (stiffness after periods of rest, such as long car rides), morning joint stiffness and pain. In the young child, it may present with intermittent limp or deterioration in behavior or mood or avoidance of previously enjoyed activities, rather than complaining of pain.

Juvenile Idiopathic Arthritis: Common Exclusions

RHEUMATIC DISEASE	
Post-infectious reactive arthropathy e.g. after gastroenteritis	Psoriatic arthritis Arthritis can present before skin lesions. FHx is helpful in reaching a diagnosis
Ankylosing spondylitis Mostly older children with back pain	Scleroderma Arthralgia not arthritis
Reiter's syndrome Triad of arthritis, urethritis, and conjunctivitis	Mixed connective tissue disease
Vasculitis syndromes E.g. Henoch schonlein purpura, Kawasaki disease	Hepatitis B and C Patients present with jaundice, vague abdominal pain, and sometimes arthritis
Systemic lupus erythematosus Non destructive arthritis unlike RA which is destructive	Inflammatory bowel disease [ulcerative colitis, crohn's]
Rheumatic fever Migratory non persistent arthritis	Sarcoidosis Arthritis, skin rash, lung nodules

Juvenile idiopathic arthritis
common exclusions
non rheumatic causes of arthritis

Growing pains

3-9 years of age. Usually involves lower limbs
Very active, normal growth. You can do inflammatory marker or cbc and reassure the mother

Benign hypermobility syndrome

Joint hyperextension. Touching the forearm with the thumb, easily reaching the floor when bending. In 5% of the population

Fibromyalgia[fibrositis]

In older females. Previously called fibrositis

Osteomyelitis

Pyogenic arthritis

Bone pain, arthritis, high grade fever, and limping. Aspirate and give ABx

Osgood-Schlatter disease

Avulsion of tibial tubercle under severe stress. Causes pain under the knee. Obvious on Xray.

Patellofemoral pain syndrome [chondromalacia patellae]

Pain on walking up or downstairs, pressure on the patella will cause pain.

Neoplasm

e.g. acute lymphoblastic leukemia

Hematological

e.g. sickle cell anemia

Psychogenic arthritis

Emotional disturbance leading to generalized pain

Trauma

Slipped capital femoral epiphysis

Athletes and obese children. Clear Xray findings

Genetic disorders

Mucopolysaccharidosis

Extra from illustrated

Summary

Diagnostic clues regarding musculoskeletal disorders

'Typical' symptom combinations	Pivotal clinical features	Possible diagnoses
Nocturnal wakening with leg pain	Normal child	'Growing pains'
		Osteoid osteoma
	Anaemia, bruising, irritability, infections	Leukaemia, lymphoma, neuroblastoma (young child)
'Clunk' on hip movement on screening, limp in an older infant	Asymmetrical upper leg skin folds, limited hip abduction	Developmental dysplasia of the hip (DDH)
Febrile, toxic-looking infant, irritability with nappy changing	Restricted joint range (especially hip)	Septic arthritis
		Osteomyelitis
Sudden limp in a otherwise well young child	Unilateral restricted hip movement	Transient synovitis of the hip
		Perthes disease
Fever, erythematous rash, red eyes, irritability in infant or young child	Erythema/oedema of hands and feet, oral mucositis, cervical lymphadenopathy	Kawasaki disease
Irritability, fever, reluctance to move in an infant or young child	Stiff back, 'tripod' sitting	Discitis
		Vertebral osteomyelitis
Joint pain, stiffness and restriction	Persistent joint swelling	Juvenile idiopathic arthritis
Loss of joint function	Loss of joint range	
Hip pain in an obese adolescent boy	Unilateral hip restriction	Slipped capital femoral epiphysis
Lethargy, unwilling to do physical activities, irritability, rash	Eyelid erythema	Juvenile dermatomyositis
	Proximal muscle weakness	
Constitutional symptoms, lethargy, arthralgia in an adolescent female	Multi-system abnormalities, haematuria, facial erythema	Systemic lupus erythematosus

Classification of Juvenile Idiopathic Arthritis[ILAR]

1. Systemic arthritic (Stills disease previously)	10% - 20%
2. Oligoarthritis (<4 joints)	50% - 60%
3. Polyarthritis (RF negative)	20% - 30%
4. Polyarthritis (RF positive)	5% - 10%
5. Psoriatic arthritis	2% - 15%
6. Enthesitis-related arthritis[ERA] [sites where tendons and ligaments insert into bone.]	1% - 7 %
7. Undifferentiated arthritis	-

* ILAR: International League Against Rheumatism

Systemic Arthritis (ILAR)

1. **Arthritis in ≥1 joint [for ≥ 6 weeks].**
   - **with or preceded by**
2. **Fever ≥ 2 weeks, quotidian (≥ 39^0 returns to ≤ 37^0C), documented daily for ≥3days**
   - **with**
3. **At least one of the following:**
   - **Evanescent [not fixed] erythematous rash.**
   - **Generalized lymph node enlargement.**
   - **Hepatomegaly and/or splenomegaly.**
   - **Serositis.**

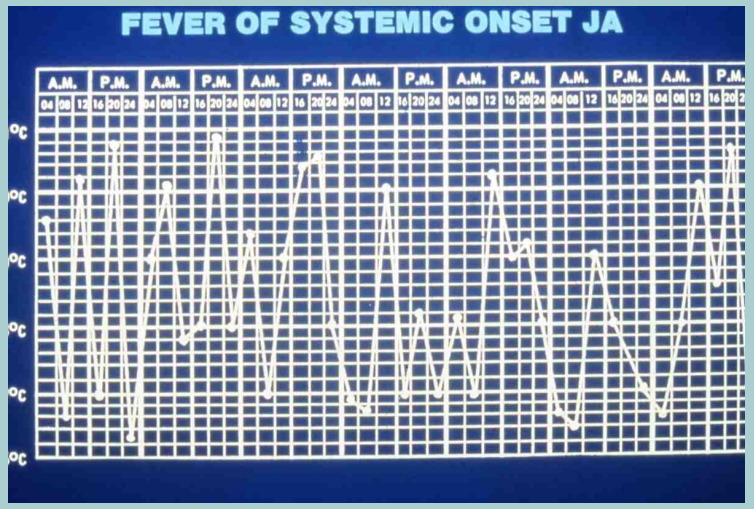
Rash appears when temperature is high and disappears when it goes down

Pleuritis, pericarditis, peritonitis

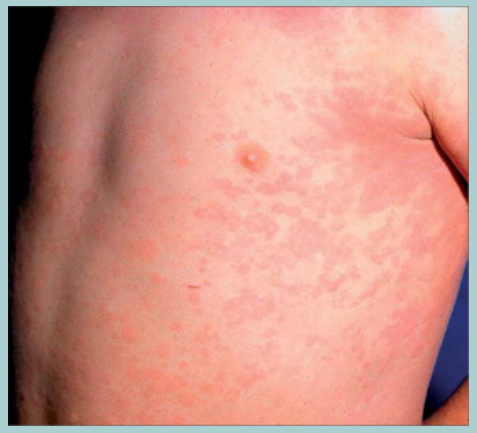
Systemic onset JIA 20%

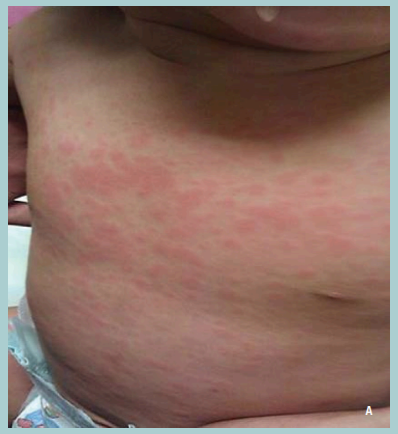
Age at onset	**16** years or younger
Sex ratio	Equal or boys > girls
Articular manifestations	Early – arthritis that may be transient Later – chronic arthritis that is usually polyarticular
Extra-articular manifestations	High intermittent fever; rash, lymphadenopathy, myalgia, serositis, organomegaly
Laboratory tests	Leukocytosis, anemia
Prognosis	Severe arthritis in **25%**

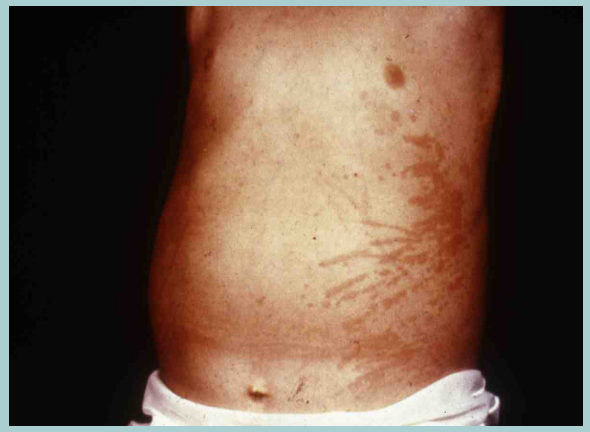
Systemic onset JIA is the most difficult to control in terms of both articular inflammation and systemic manifestations

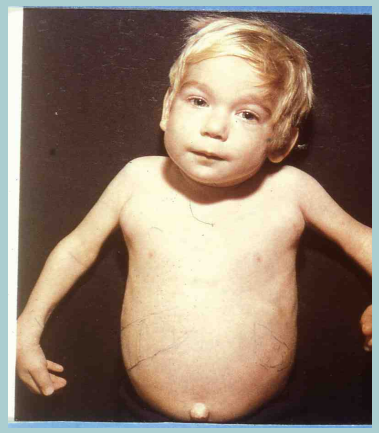


Rash will appear during fever spikes.









Salmon colored maculopapular rash.
Scratching the skin will cause appearance of the rash (koebner's phenomenon)

Oligoarthritis

Pauciarticular

Arthritis in 1-4 joints in the first six months of oncet.

Types

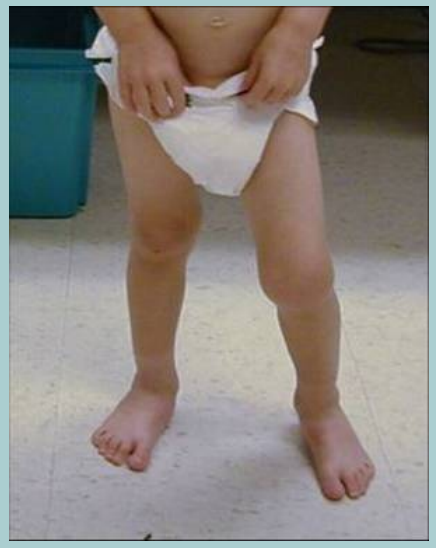
-Persistent disease [1-4 joints throughout the disease]

-Extended disease [≥5 joints after the first six months]. Managed as polyarticular

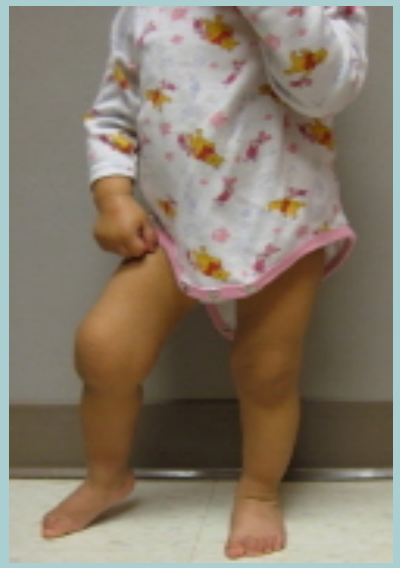
Pauciarticular JIA (50%)

= oligoarticular

SUBGROUP 35%	Iritis is painless at first so any patient with pauciarticular JIA should follow up with ophthalmology every 4-6 months for irits.	SUBGROUP 15%
Early childhood	Age at onset	Late childhood
Girls	Sex predominance	Boys
Knee, ankle, elbow	Typical joints	Lower limb
Chronic iritis	Extra-articular manifestations	Acute iritis, bowel disease, features of Reiter's syndrome
Negative	Rheumatoid factor	Negative
>50%	ANA	0
DR5, 6, 8	HLA	B27
Severe arthritis 10%; severe iridocyclitis possible	prognosis	Chronic spondyloarthropathy possible



Left knee swelling



Swelling and limited extension

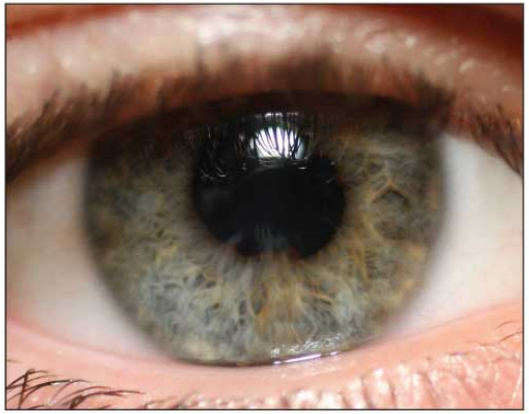
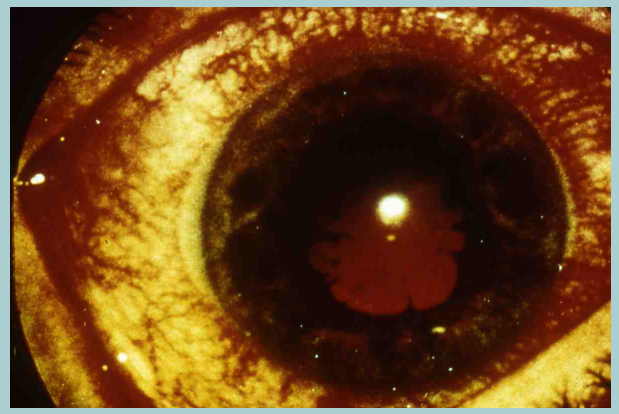


Figure 1 – *Synechiae are seen in a patient with uveitis who has juvenile idiopathic arthritis. Other complications of uveitis include keratotic bands, cataracts, and vision impairment.*



Polyarthritis (RF negative)

Arthritis affecting ≥ 5 joints in the first six months of disease;

- RF negative.

Polyarthritis (RF positive)

Arthritis affecting ≥5 joints in the first six months of disease;

-Positive RF in 2 readings 3 months apart, during the first six months.

Polyarticular JIA 30%

RF-ve (25%)		RF+ve (5%)
16 years or younger	Age at onset	**8** through **16** years
Girls	Sex predominance	Girls
Few	Extra-articular manifestations	Nodules, vasculitis
25% of patients	ANA	**50%** of patients
?	HLA	DW4/DR4
Severe arthritis **10-20%**	Prognosis	Severe arthritis **>50%**

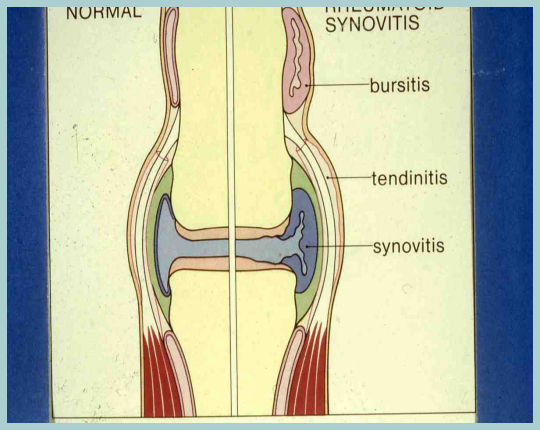
A positive rheumatoid factor is indicative of a poor prognostic outcome

Bad prognosis
Need aggressive management

Pathology

Serositis

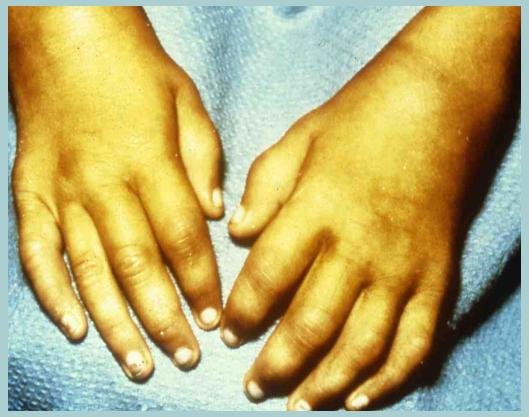
1. Synovitis
2. Tendenitis
3. Bursae



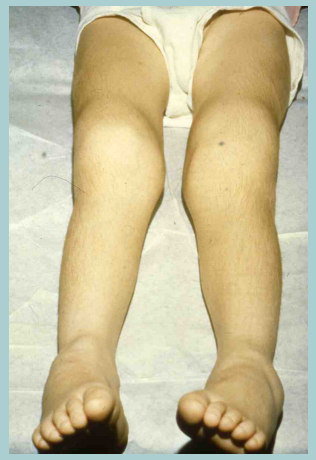
Serositis of pleura and pericardium

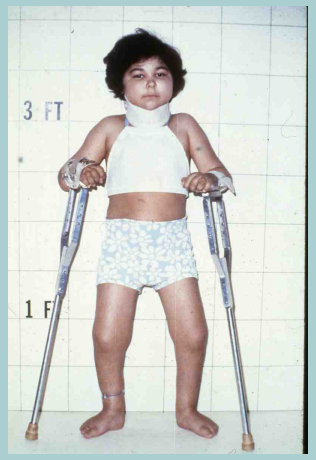
Subcutaneous Nodules

Vasculitis

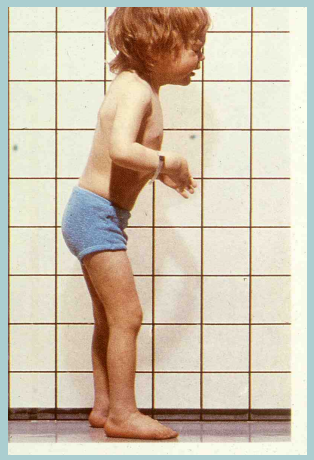


In polyarticular involvement is usually symmetrical.

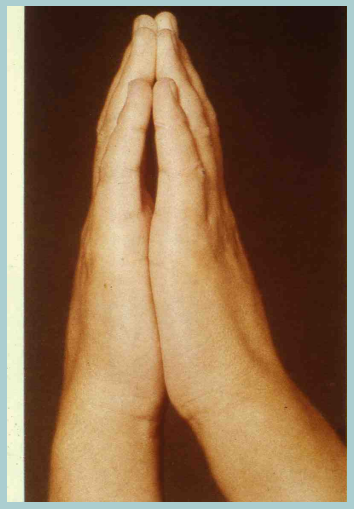




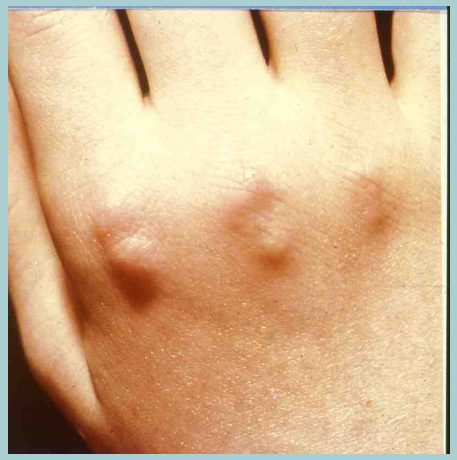
Involvement of the neck. You must be careful with anesthesia



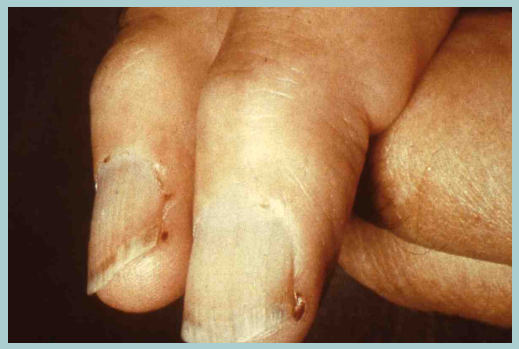
Patients can't stand straight



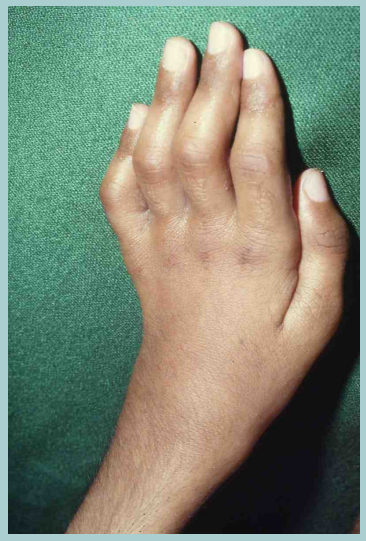
Gap indicates small joint involvement

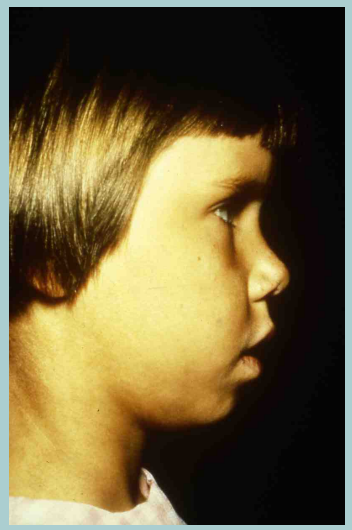


Subcutaneous nodules



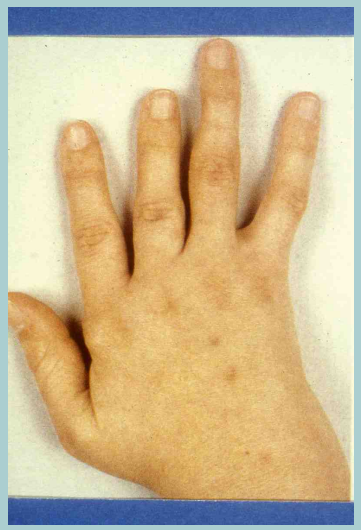
Vasculitis rash



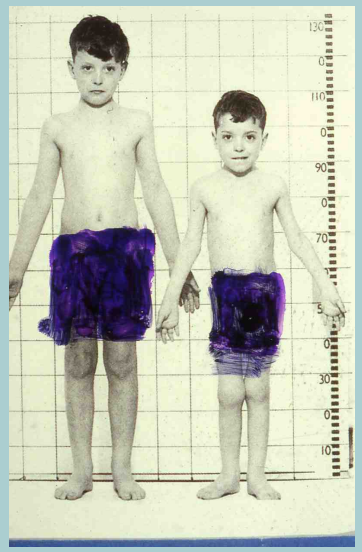


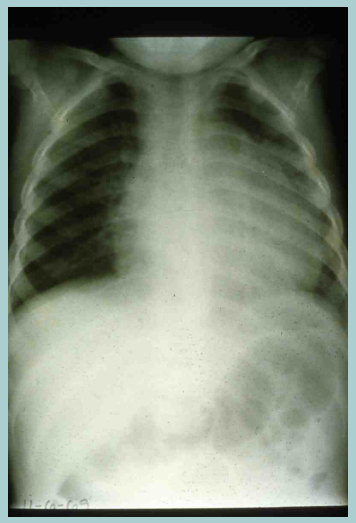
Receding chin can be a complication

They may have c-spine and TMJ involvement



Middle finger was affected early and growth was stunted.





Pericardial effusion

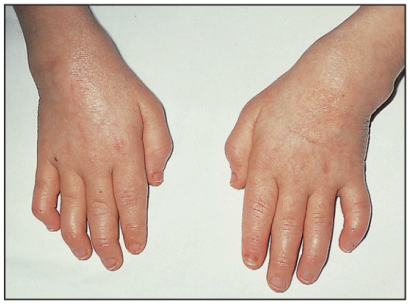


Figure 26.16 Polyarticular juvenile idiopathic arthritis, showing swelling of the wrists, metacarpal and interphalangeal joints and early swan-neck deformities of the fingers.

Extra

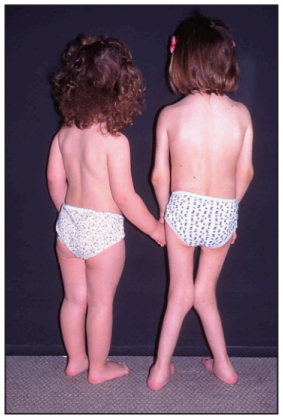
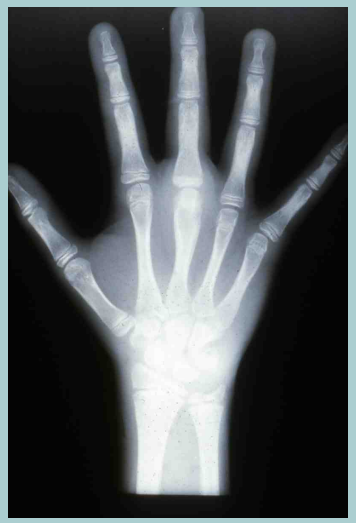


Figure 26.17 Growth failure and marked genu valgum (knock-knees) in an 8-year-old girl with juvenile idiopathic arthritis. For comparison, her sister on the left is 4 years old.

Extra



Joint erosion

Psoriatic Arthritis

Arthritis and psoriasis

OR

Arthritis and at least two of:

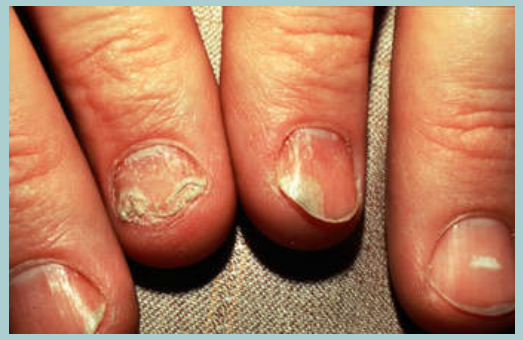
1) Dactylitis,

2) Nail pitting or onycholysis,

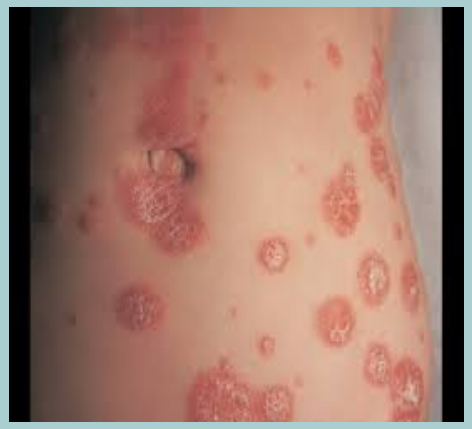
3) Family history of psoriasis.

Dactylitis

Nail pitting



Onycholysis



Psoriatic lesions

Enthesitis Related Arthritis

Arthritis and enthesitis,

OR

arthritis or enthesitis with ≥ 2 of :

[1] Sacroiliac joint tendernes and/or inflammatory lumbosacral pain.

[2] HLA-B27

[3] Family history of HLA-B27 associated disease

[4] Acute anterior uveitis

[5] Oncet of oligoarthritis in a boy aged ≥8 years.

Undifferentiated Arthritis

Arthritis that does not fulfills any criteria.

OR

Arthritis that fulfills two OR more criteria.

Management of Juvenile idiopathic Arthritis

TEAMWORK

PEDIATRIC RHUMATOLOGY

PHYSIOTHERAPY

OCCUPATIONAL THERAPY

PEDIATRIC ORTHOPEDIC

SOCIAL WORKER

CLINICAL PHARMACOLOGY

Treatment for JIA

SUBTYPE	first-line therapy	Severe or refractory
Oligoarticular		
- Persistent	**IAS injection ± NSAID; may repeat IAS injections up to 4 per year**	**MTX; Anti-TNF-α agent may be needed**
- Extended	**If fewer than 8 joints, manage as persistent; if more, manage as polyarticular**	
Polyarticular	**MTX ± IAS ± NSAID** Start with NSAIDs	**Add Anti-TNF-α agent; may need different DMARD (leflunomide, SSZ) ± IAS injection ± prednisone, rituximab**
Enthesitis-related	**IAS injection and NSAID**	**MTX or SSZ; Anti-TNF-α agent may be needed**
Systemic		
Active systemic	**Anti-IL-1 agent; prednisone (IV pulse/daily oral) may be needed**	**Cyclosporine, thalidomide** Usually Anakinra (anti IL1) is enough
Active arthritis	**Manage as polyarticular or oligoarticular, depending on number of joints involved**	**Manage as polyarticular**

Extra from illustrated

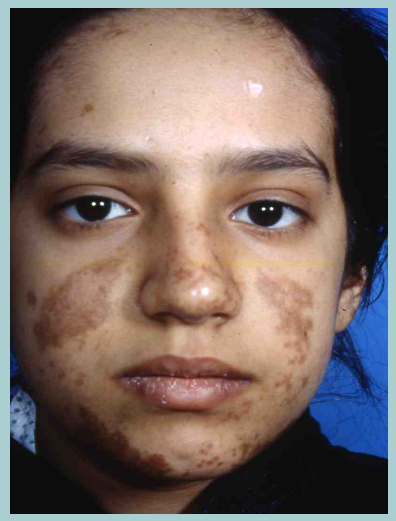
Table 26.4 Classification and clinical features of JIA (juvenile idiopathic arthritis)

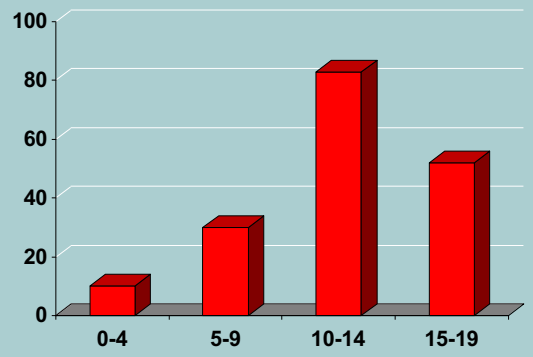
JIA subtype (approximate %)	Onset age	Sex ratio (F:M)	Articular pattern	Extra-articular features	Laboratory abnormalities
Oligoarthritis (persistent) (49%)	1–6 years	5:1	1–4 (max) joints involved; knee, ankle or wrist most common	Chronic anterior uveitis in 20%, leg length discrepancy Prognosis excellent	ANA+/–
Oligoarthritis (extended) (8%)	1–6 years	5:1	>4 joints involved after first 6 months. Asymmetrical distribution of large and small joints	Chronic anterior uveitis 20%, asymmetrical growth Prognosis moderate	ANA+/–
Polyarthritis (RF negative) (16%)	1–6 years	5:1	Symmetrical large and small joint arthritis, often with marked finger involvement Cervical spine and temporomandibular joint may be involved	Low-grade fever, chronic anterior uveitis 5%, late reduction of growth rate Prognosis moderate	
Polyarthritis (RF) positive) (3%)	10–16 years	5:1	Symmetrical large and small joint arthritis, often with marked finger involvement	Rheumatoid nodules 10% Similar to adult rheumatoid arthritis Prognosis poor	RF+ (long term)
Systemic arthritis (9%)	1–10 years	1:1	Oligoarthritis or polyarthritis. May have aches and pains in joints and muscles (arthralgia/myalgia) but initially no arthritis	Acute illness, malaise, high daily fever initially, with salmon-pink, macular rash, lymphadenopathy, hepatosplenomegaly, serositis Prognosis variable to poor	Anaemia, raised neutrophils and platelets, high acute-phase reactants (see Case History 26.1)
Psoriatic arthritis (7%)	1–16 years	1:1	Usually asymmetrical distribution of large and small joints, dactylitis	Psoriasis, nail pitting or dystrophy, chronic anterior uveitis 20% Prognosis moderate	
Enthesitis-related arthritis (7%)	6–16 years	1:4	Lower limb, large joint arthritis initially, mild lumbar spine or sacroiliac involvement later on	Enthesitis – localised inflammation at insertion of tendons or ligaments into bone, often in feet, Achilles insertion Occasional acute uveitis Prognosis moderate	HLAB27+
Undifferentiated arthritis (1%)	1–16 years	2:1 (variable)	Overlapping articular and extra-articular patterns between ≥2 subtypes or insufficient criteria for sub-classification	Prognosis variable	

Juvenile systemic lupus erythematous JSLE

Same presentation and criteria as adults

- **Autoimmune disease**
- **Autoantibodies, immune complex formation, immune dysregulation leading to tissue damage.**
- **Etiology unknown [environmental and hormonal trigger to a genetically susceptible person].**
- **Natural history unpredictable.**
- **All races affected**
- **Females > males**
- **Incidence in children <15 years 0.5-0.6 per100,000**





AGE AT ONSET IN JSLE

JSLE

- Rare before 5 years.
- Neonatal lupus

Complete bundle branch block can be detected in utero. Skin rash similar to seborrheic dermatitis, raised ANA. Resolves after 6 months

- More common in adolescence.
- JSLE in the first decade: 3.5 – 15% of all SLE cases.
- More **renal involvement** in JSLE

SLE in adults is less severe

- More sever in the first decade

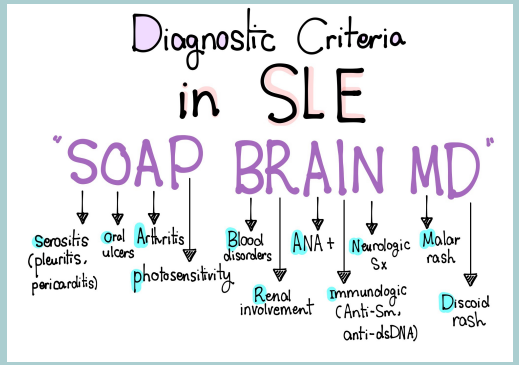
Classification criteria of SLE

Malar (butterfly) rash
Discoid-lupus rash
Photosensitivity
Oral or nasal mucocutaneous ulcerations
Nonerosive arthritis الدكتور عادها ٣ مرات
Nephritis[b]
- **Proteinuria > 0.5 g/day**
- **Cellular casts**

Encephalopathy[b]
- **Seizures**
- **Psychosis**

Pleuritis or pericarditis
Cytopenia
Positive immunoserology
- **Antibodies to nDNA**
- **Antibodies to Sm nuclear antigen**
- **Positive LE-cell preparation**
- **Biologic false-positive test for syphilis**

Positive antinuclear antibody test



Extra

[a] **Four of 11 criteria provide a sensitivity of 96% and a specificity of 96%.**

SEROLOGICAL TESTS

Test	% positive of SLE
ANA by indirect immunofluorescence	95 – 100
Antibody DNA	60
Antibodies to soluble ribonucleoproteins	80
Anti nRNP	30
Anti SM	20
Anti Ro (SSA)	30
Anti La (SSB)	10

DsDNA very specific especially in lupus nephritis

Increased with Raynaud's and pHTN

Especially in neonatal lupus and drug induced lupus

CLINICAL PRESENTATION
Mucocutaneous

Malar rash, Butterfly distribution. 25% of cases.

Photosensetivity. Recurrence of symptoms after sun exposure e.g. fever and arthritis

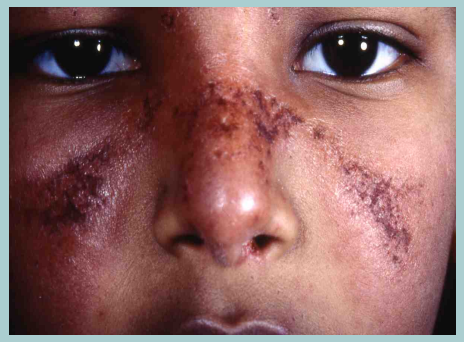
Discoid rash Similar to psoriasis

Nasal and oral ulceration [**painless** may perforate]

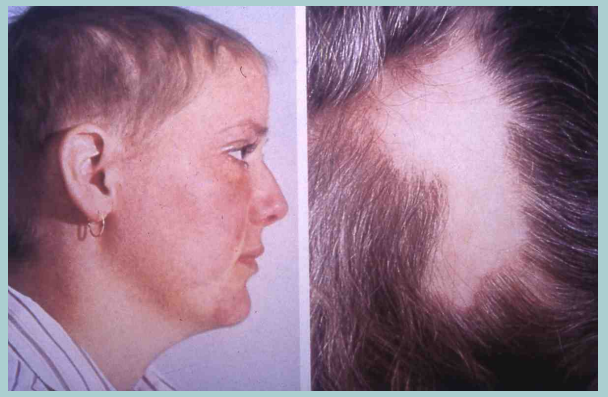
Small vessel vasculitis [digital ulcer, livedo-reticularis, raynauds phenomenon]

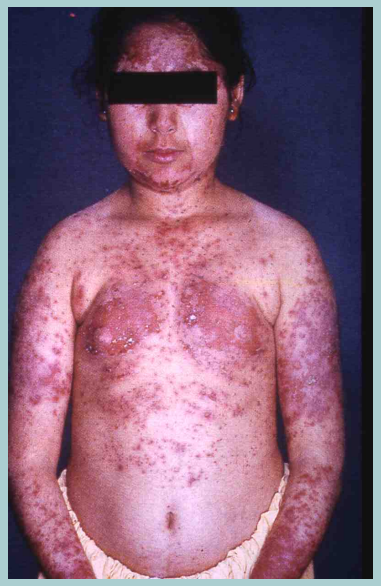
Alpoecia

Neonatal Lupus Erythematous: Lesions similar to seborrheic dermatitis, disappear spontaneously in 4-6 months.

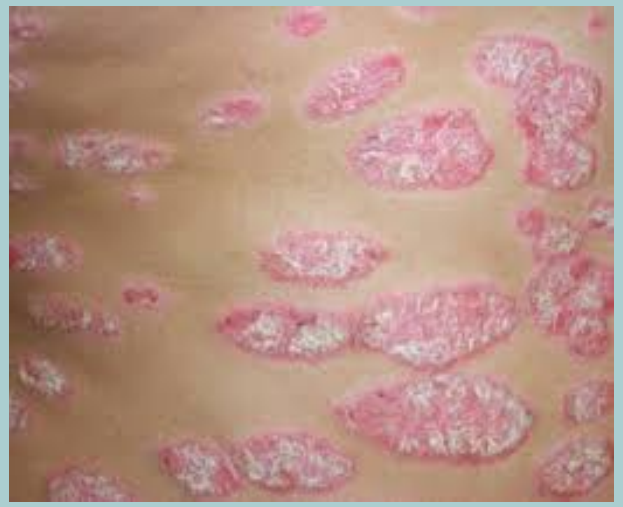


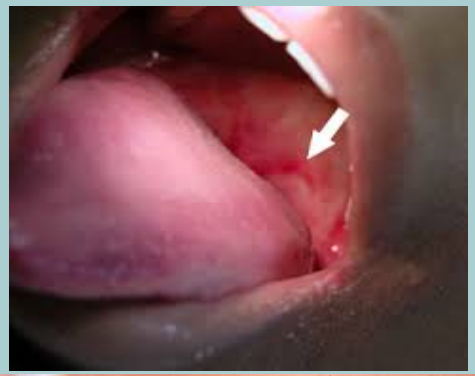
Butterfly rash

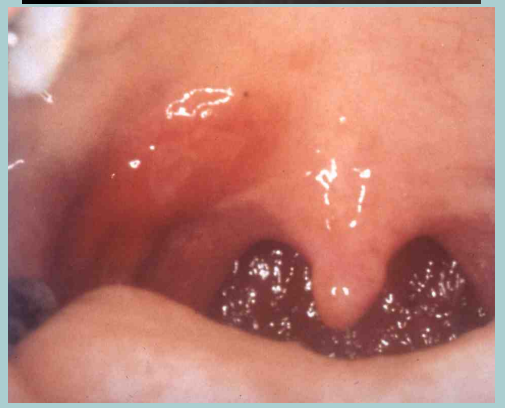




Discoid rash







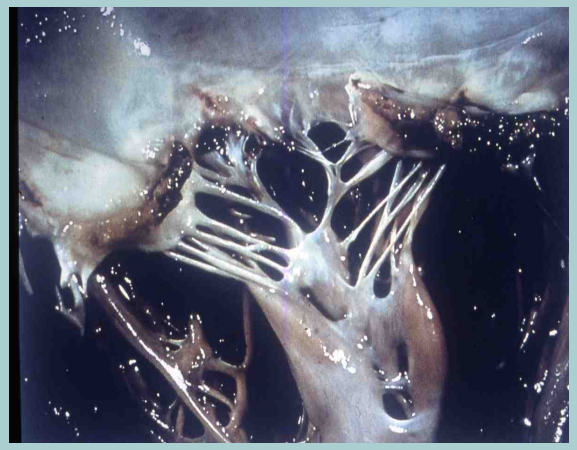
Oral ulcers indicates active disease

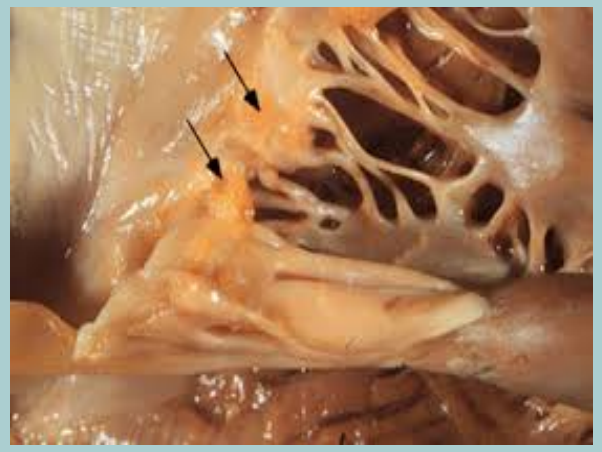
CARDIOVASCULAR

Pancarditis

- Myocarditis
- Pericarditis
- Endocarditis (Libman-Sacks)
- Conduction defect [CBBB] in neonate.
- Neonatal lupus [Rash similar to seborrheic dermatitis

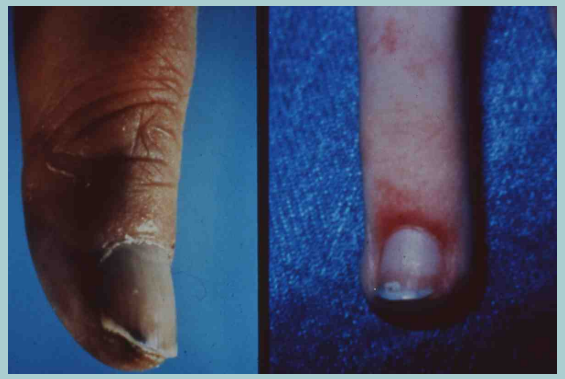
A pregnant woman with SLE will transfer IgG autoantibodies (usually anti-Ro) across the placenta at 12 to 16 weeks. This can cause a variety of manifestations, the most common being congenital heart block. All are temporary, except for the heart block, which may require permanent pacing.



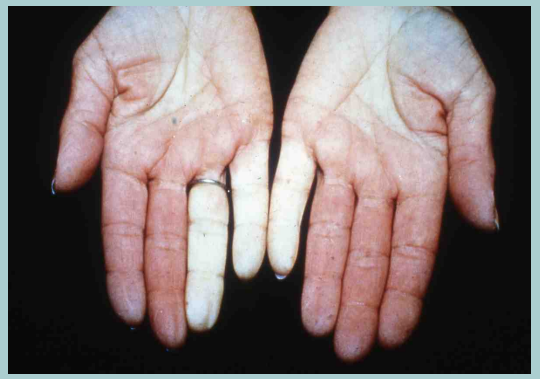


VASCULITIS IN SLE

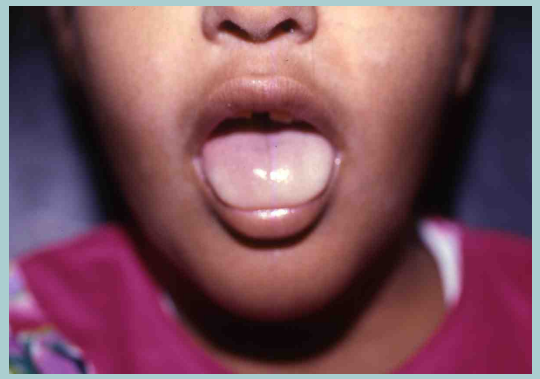
- SIZE
  - Small Vessel Vasculitis
- CLINICAL PRESENTATION:
  - Lupus Crisis (wide spread vasculitis + polyserositis)
  - Raynaud's phenomenon
  - Digital involvement
  - Recurrent thrombophlebitis In older females
  - Livedo reticularis



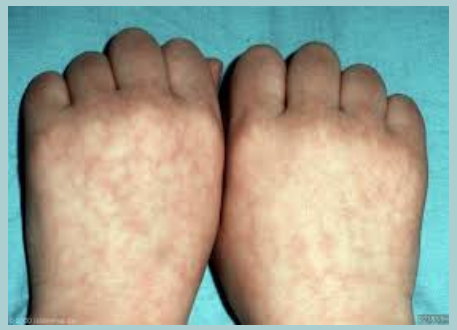
Vasculitic rash

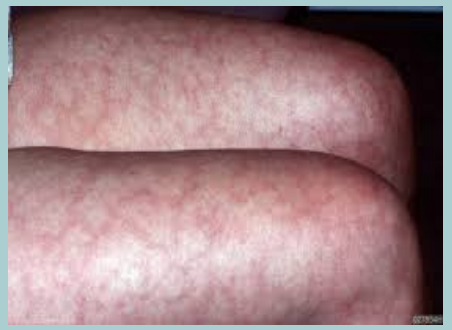


Raynaud's phenomenon

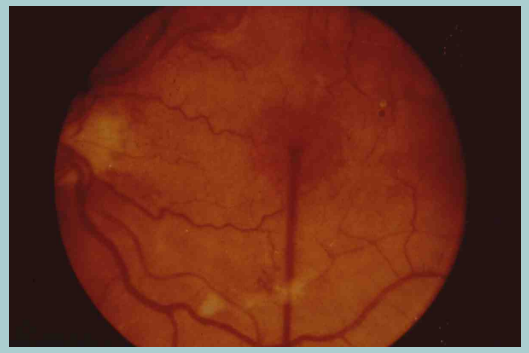


Tongue raynaud's (very rare)





Livido reticularis



Cotton wool spots indicate retinal vasculitis

HAEMATOLOGICAL ABNORMALITIES

Think of lupus as a differential for pancytopenia

Abnormality	Patients (%)
Anemia (hematocrit < 30%)	**50**
Acute hemolytic anemia	5
Leukopenia <2,000 WBC/mm^3	10
Leukopenia <4,500 WBC/mm^3	**40**
Thrombocytopenia <150,000 pts/mm^3	**30**
Thrombocytopenia <100,000 pts/mm^3	5

G.I. MANIFESTATIONS

- 31% of cases have abdominal pain.
- Abnormal esophageal motility.
- Ascites and peritonitis.
- Acute pancreatitis.
- Mesenteric artery thrombosis
- Malabsorption
- GI vasculitis: edema, ulceration, gangrene, perforation

NEUROPSYCHIATRIC MANIFESTATIONS

- Non-Focal Cerebral Dysfunction (35-60%)
  organic brain syndrome.
  Psychosis.
  Neurosis.
- Movement Disorders (10-35%)
- Seizures (15-35%)
- Focal Deficits (10-35%)
- Peripheral Neuropathies (10-25%)
- Others: e.g. headache, aseptic meningitis, myasthenia gravis

Management of SLE

In lupus nephritis it is important to refer to nephrology for biopsy

Depend on system affected.

(history, clinical examination, and investigations)

Non-immunosupressants

- antimalarial (Hydroxychloroquine)

  Steroid sparing especially helpful with discoid rash

- NSAIDs (Ibuprofen, Naproxen)

  For musculoskeletal involvement

Immunosuppresants

- Corticosteroids (Prednisone/ivMP)

  Most cases need steroids

- DMARDs (MTX, Imuran, cellcept)

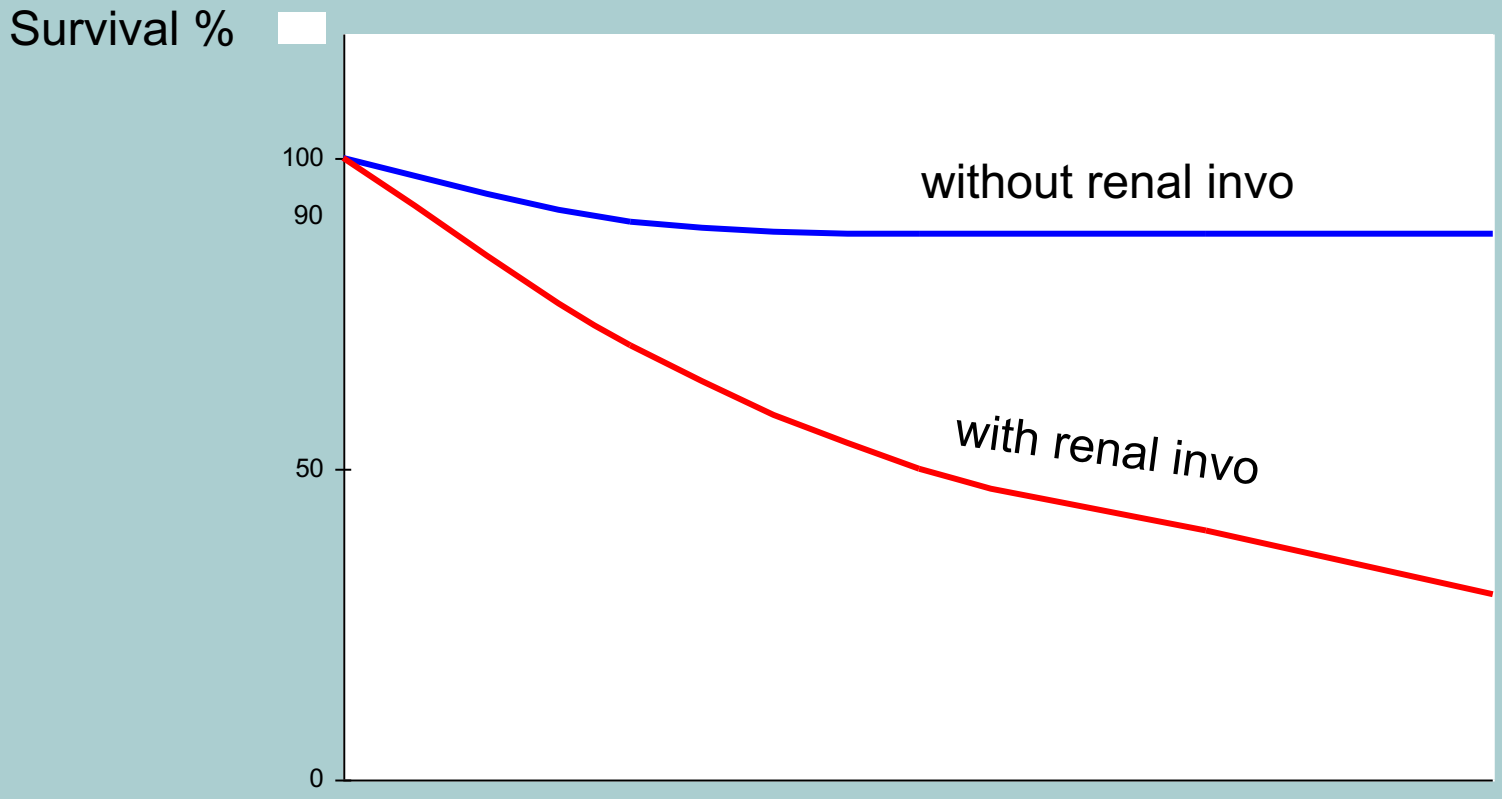
- Biologics (Rituximab, Tocilizumab, Belimumab)

- Cytotoxins (cyclophosphamide)

Management of SLE (cont.)

- I.V. immunoglobulin (IVIG). If steroids failed
- Plasmapheresis.
- **Other treatment:**
  - sunscreen.
  - physical and occupational therapy
  - treatment of complications.

Prognosis in SLE



JUVENILE DERMATOMYOSITIS JDM

Idiopathic inflammatory myopathy

Has characteristic cutaneous lesions

Affect skin and muscles

May affect [joints, oesphagus, lungs]

Calcinosis is common

Association with malignancy.

DERMATOMYOSITIS / POLYMYOSITIS

If no skin manifestations think of polymyositis

- Symmetrical progressive proximal muscle weakness.
- Characteristic rashes[Gottron papules, heliotrope rash]
- Biopsy showing inflammatory changes
- Raised muscle enzymes (**CPK**, **AST**, **Aldolase**)
- Electromyography abnormalities
  (e.g. polyphasic potentials)

Juvenile Dermatomyositis

Expanded criteria for diagnosis

- Nail fold capillaroscopy abnormalities
- Calcinosis
- Dysphonia Involvement of vocal cords
- Typical findings on MRI of muscle and ultrasonography

JDM Clinical Course

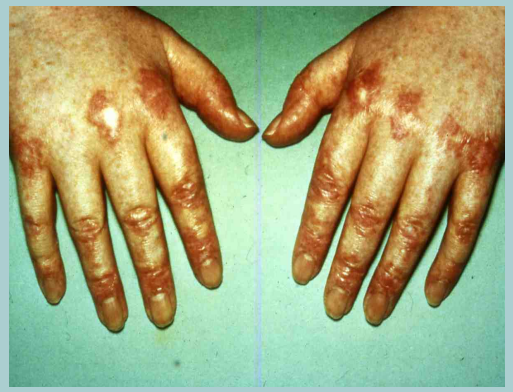
- **Monocyclic (remission within 2-3 years)**
- **Polycyclic**
- **Chronic**
- **Ulcerative[GI-system]**

One attack lasting a year or two then disease resolution

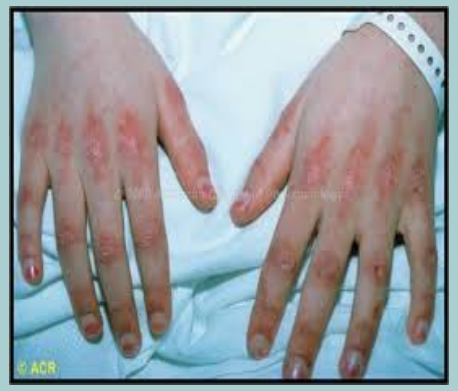
JDM Investigations

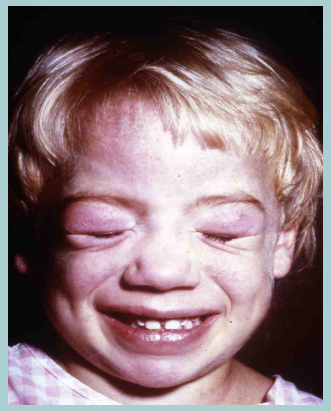
- **EMG**
- **Muscle biopsy**
- **Muscle enzymes (CPK, Aldolase)**
- **Nail fold capillaroscopy**
- **MRI**

Rarely needed

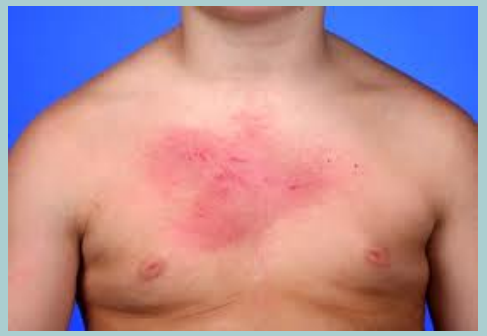


Gottron's papules

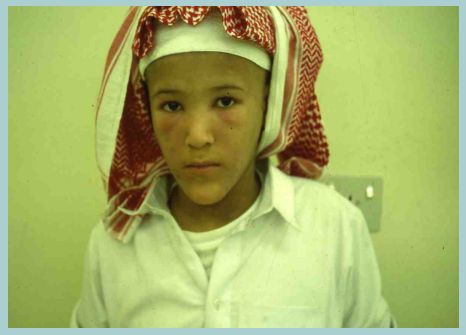




Heliotrope rash

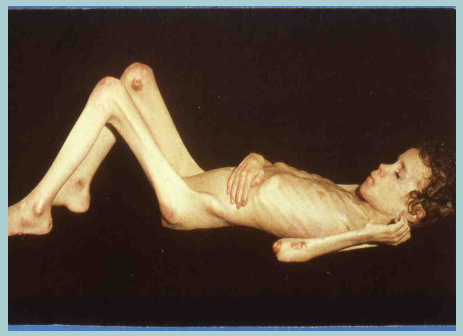


V sign

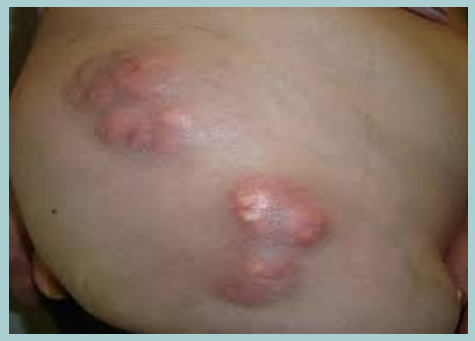


هذا ماي بيشنت لما نقرل بترفلاي مو دايما لوبس فهذا عنده شويه كذا لوك لايك بس مو نازل على اللوبس؟؟

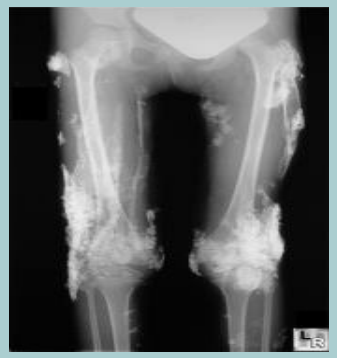
Capillaroscopy



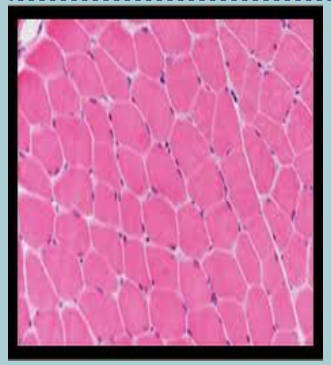
Severe muscle destruction (rare)



Calcinosis



Calcinosis + severe destruction



Muscle destruction

JDM - Treatment

- **Aims** - minimize inflammation
  - improve function
  - prevent disability
- Early teamwork (rheumatologist, dermatologist, physical therapist and primary care doctor)
- Drugs:
  - Corticosteroids (Prednisolone, IV MP)
  - Immunoglobulin (IV Ig)
  - Methotrexate
  - Others (Hydroxychloroquine, anti-TNF therapy)
  - Severe cases (Cyclophosphamide, Rituximab)

Steroids:
In lupus: 1-2 mg/kg
In JDM: 2-3 mg/kg or pulse steroids

If signs of vasculitis add methotrexate

JDM – Treatment (cont.)

Other aspects of care:

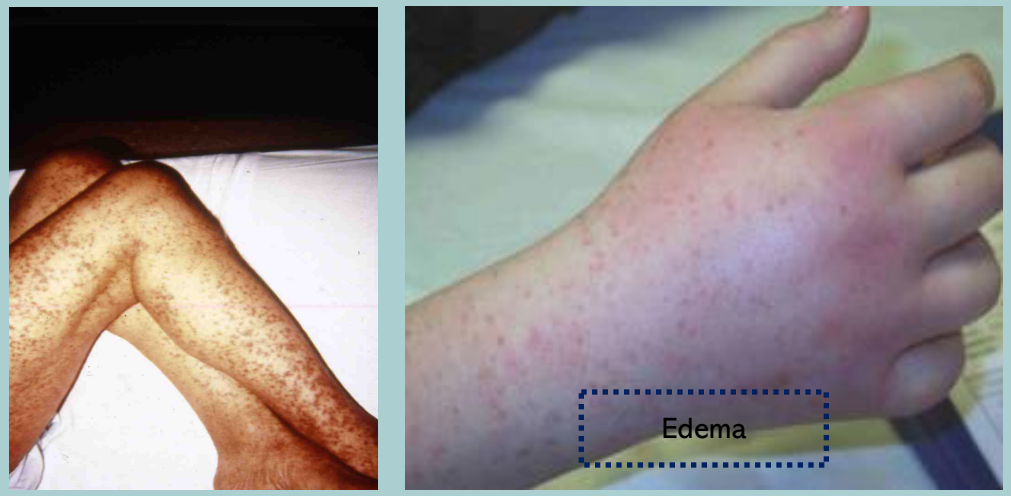
- Skin protection
- Physical therapy
- Speech therapy
- Dietetic assessment
- Management of calcinosis

If calcinosis is near a joint or an organ remove surgically

different drugs used with poor response such as pamidronate, probenecid, warfarin, colchicine, aluminum hydroxide, infliximab.

Henoch-Schonlin Purpura

- **Small-vessel vasculitis**
- **Benign self-limiting, unknown etiology, multifactorial causes [genetic, environment, infection] [group A stept, mycoplasma, EBV, Hepatitis c, adenovirus, parvovirus, measles]**
- **Deposition of [IgA, C3] immune complex in small vessels.**
- **HSP and IgA nephropathy (both have ↑ IgA and identical renal biopsy)**
- **Diagnosis is clinical**
- **Laboratory investigations to exclude other causes**



Characteristic rash in HSP on the buttocks, upper, and lower limbs

HENOCH-SCHONLEIN PURPURA HSP

Purpura	100%
Arthritis	71%
Gastrointestinal involvement	68%
Renal involvement	45%
Fever	75%
Hypertension	13%
Subcutaneous oedema	20-50%
Scrotal oedema	2 – 35%

Severe abdominal pain either due to vasculitis or intussusception. Do barium studies to exclude intussusception and treat with steroids.

HSP - Treatment

- Resolve spontaneously
- Treatment according to system involved

MSK – Ibuprofen[NSAID]

GIT - Corticosteroids (Prednisolone, IV MP)

Renal - Cyclophosphamide (Cytoxan)

- Azathioprine (Imuran)
- Mycophenolate mofetil (Cellcept)
- High dose IV immunoglobulin
- Plasmapheresis

Kawasaki's Disease (KD)=MCLS

Mucocutaneous lymph node syndrome

- **Small and medium vessel vasculitis (coronary artery)**
- **Unknown etiology [infection, genetic, autoim]**
- **Children under 5 years**
- **Diagnoses is clinical**
- **Laboratory studies to exclude other causes and look for complications (CBC – CRP – ECHO)**

Inflammation and aneurysm of coronary arteries

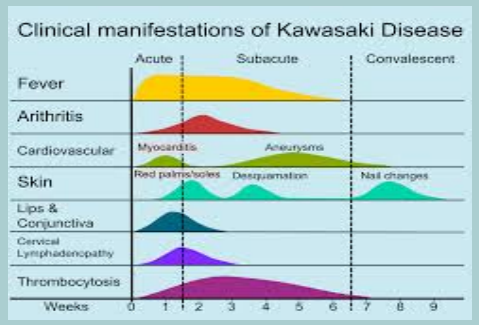
Any child with suspected Kawasaki should have an echocardiogram

Poor outcome predictors with respect to coronary artery disease: very young age, male, neutrophilia, decreased platelets, increased liver enzymes, decreased albumin, hyponatremia, increased CRP, prolonged fever.

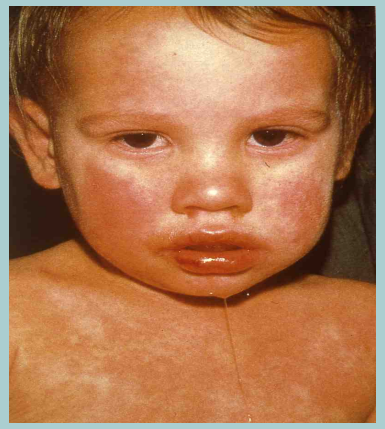
KAWASAKI'S DISEASE

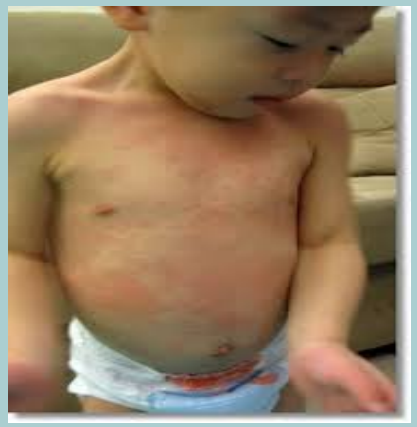
Fever	Continuous high-grade fever for at least 5 days	95%
Conjunctival congestion		90%
Exanthema		90%
Oral mucosa involvement		90%
Desquamation		90%
Cervical lymphadenopathy		75%

+ Erythema of the hands and soles

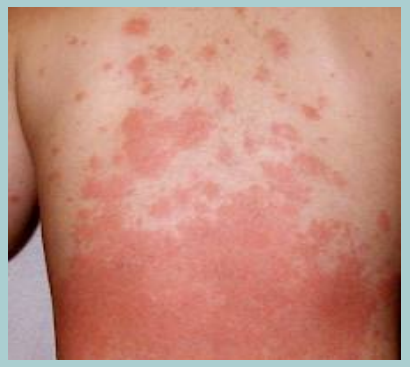


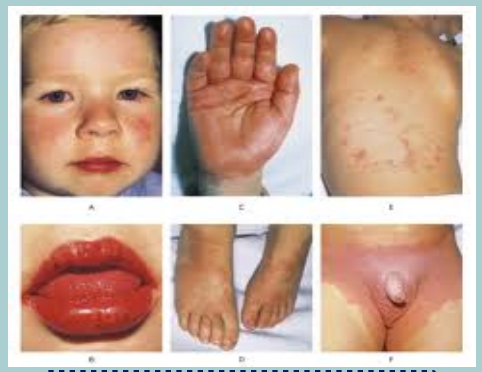
The doctor read this picture



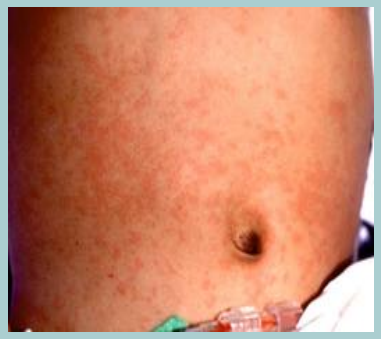


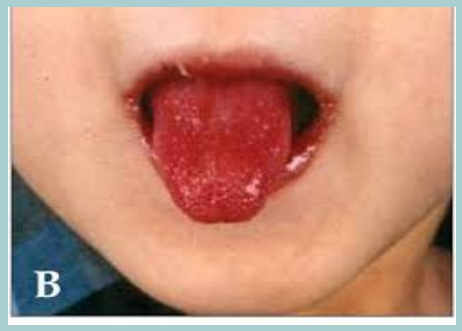
Generalized erythematous rash





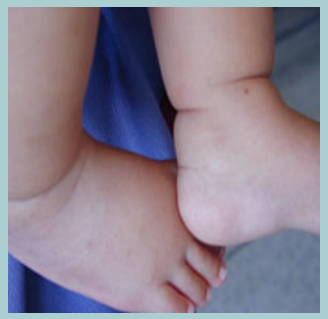
Fissuring of the lips
Swelling and erythema of lower and upper limbs





Strawberry tongue

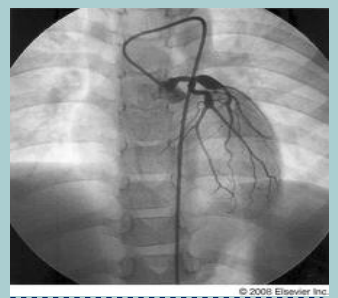
Swelling of the lower limbs, erythema, and desquamation



Arthritis

Conjunctival injection

Desquamation



Coronary involvement

MCLS Diagnostic Criteria

1. **Spiking fever 5 days or more + 4/5 criteria**

2. **Bilateral conjunctival injection**
3. **One orpharyngeal sign**
    1. Diffuse oropharyngeal erythema
    2. Strawberry tongue
    3. Redness, dryness, and fissures of lip
4. **Polymorphous erythematous rash**
5. **Cervical lymphadenopathy**
6. **One or more of the following signs**
    1. Indurative edema of hands and feet
    2. Erythema of palms and sole
    3. Desquamation of fingers and toes [2 weeks after onset]
    4. Transverse grooves in nails [2 or 3 months after onset]

Treatment of Kawasaki Disease

- **High dose aspirin**
- **Low dose aspirin**

If high grade fever, inflammation, arthritis give high dose 60-70 mg/kg
After fever subsides give low dose aspirin (platelets decrease later in the disease course)

- **High dose IV immunoglobulin**

Start with a dose of IVIG if failed a second dose
if failed steroids. If failed Others e.g. infliximab.

- **± IV methylprednisolone**
- **Non-steroidal anti-inflammatory drugs**
- **Plasmapheresis (non-responding to IV Ig)**
- **Tumor necrosis factor (TNF) blocking drugs**
  - **- Infliximab (Remicade)**
  - **- Etanercept (Enbrel)**

SPONDYLOARTHROPATHIES

Absence of rheumatoid factor(seronegative)

Involvement of sacroiliac and joints

Peripheral arthritis (predominantly lower limb)

Enthesopathy

Familial clustering

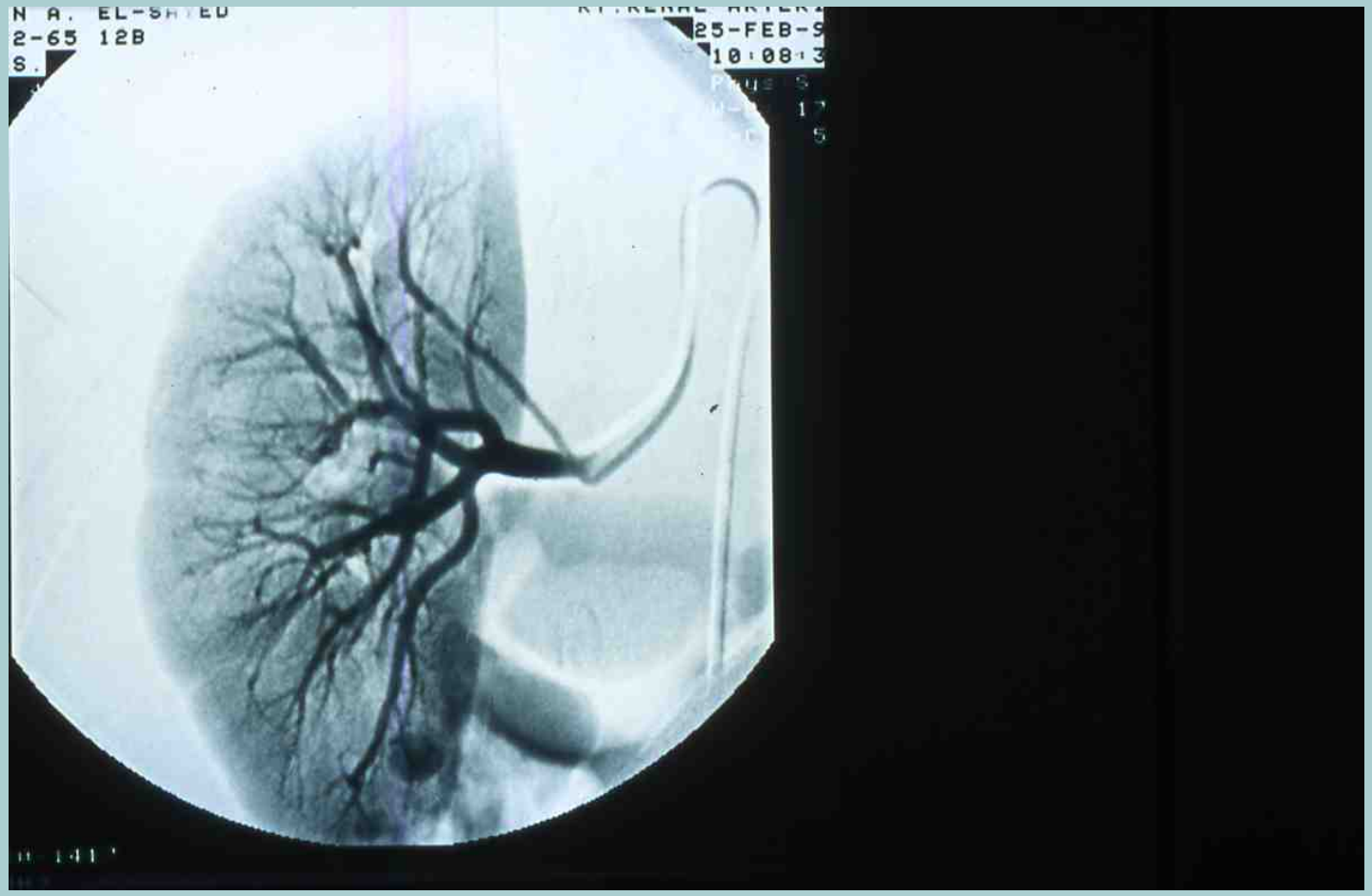
Increased incidence of HLA-B27

Common spectrum of extra- articular features (predominantly muco-cutaneous)

SPONDYLOARTHROPATHIES

- Ankylosing spondylitis
- Psoriasis
- Whipple's disease
- Ulcerative colitis
- Crohn's disease
- Reiter's disease
- Behçet's Syndrome
- Reactive arthritis

Thank YOU FOR LISTENING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