COMMON RHEUMATIC DISEASES

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Symptoms and Signs of Joint Diseases

Symptoms

- Pain
- Stiffness
- Deformity
- Loss of function
- Systemic illness

Signs

- Heat
- Redness
- Swelling
- Loss of movement
- Deformity
- Tenderness
- Abnormal movement
- Crepitus
- Functional Abnormality

Juvenile Idiopathic Arthritis

General abbreviations: 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 .

Juvenile Idiopathic Arthritis: Common Exclusions

RHEUMATIC DISEASE	
Post-infectious reactive arthropathy	Psoriatic arthritis
Ankylosing spondylitis	Scleroderma
Reiter's syndrome	Mixed connective tissue disease
Vasculitis syndromes	Chronic active hepatitis
Systemic lupus erythematosus	Inflammatory bowel disease
Rheumatic fever	Sarcoidosis

.

Juvenile Idiopathic Arthritis

NON-RHEUMATIC DISEASE	
Growing pains	Neoplasm's
Benign hypermobility syndrome	Hematologic diseases
Fibrositis	Psychogenic arthralgias
Osteomyelitis	Trauma
Pyogenic arthritis	Slipped capital femoral epiphysis
Osgood-Schlatter disease	Genetic disorders
Chondromalacia patellae	

Pathology

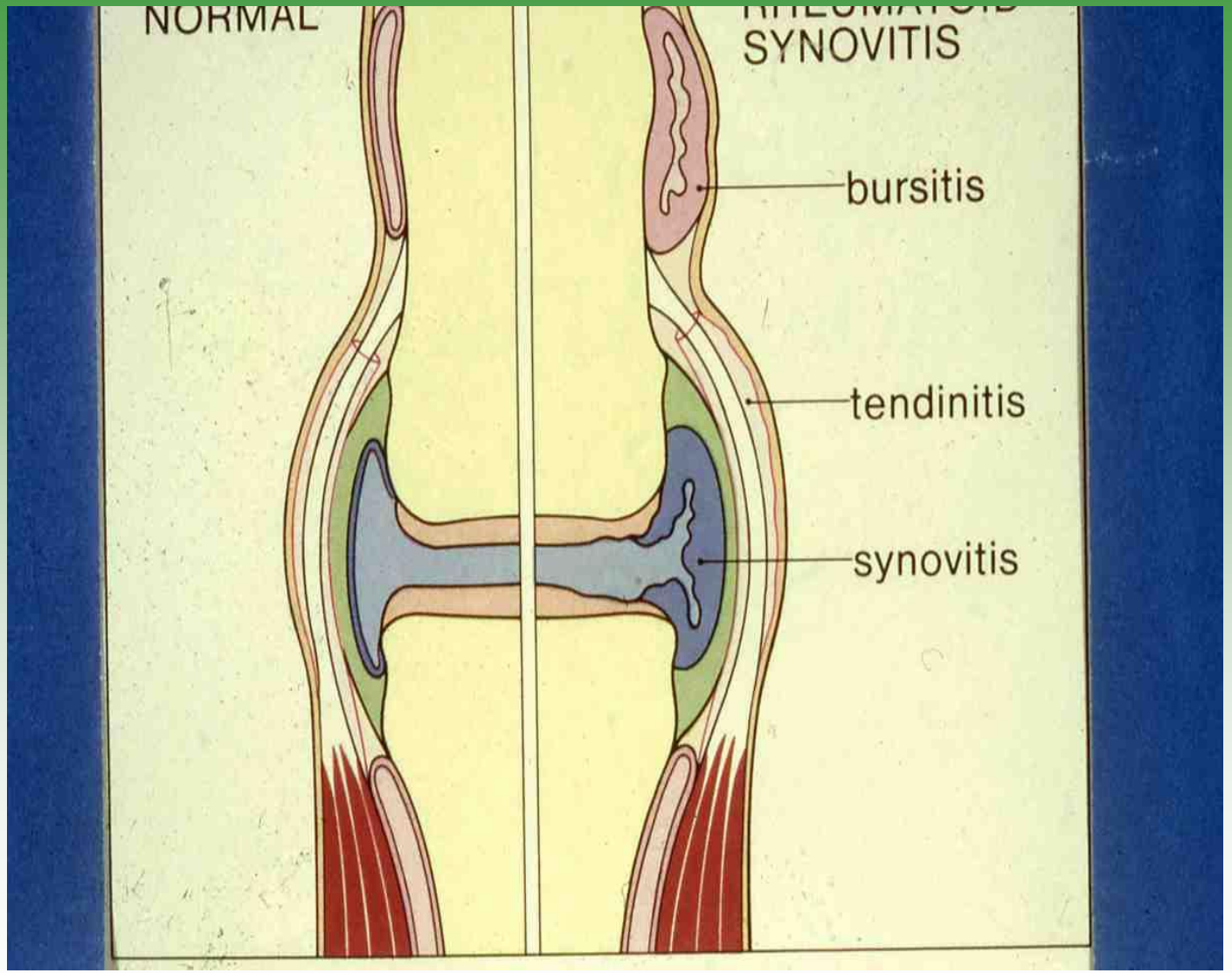
Serositis

1. Synovitis
2. Tendenitis
3. Bursae

Serositis of pleura and pericardium

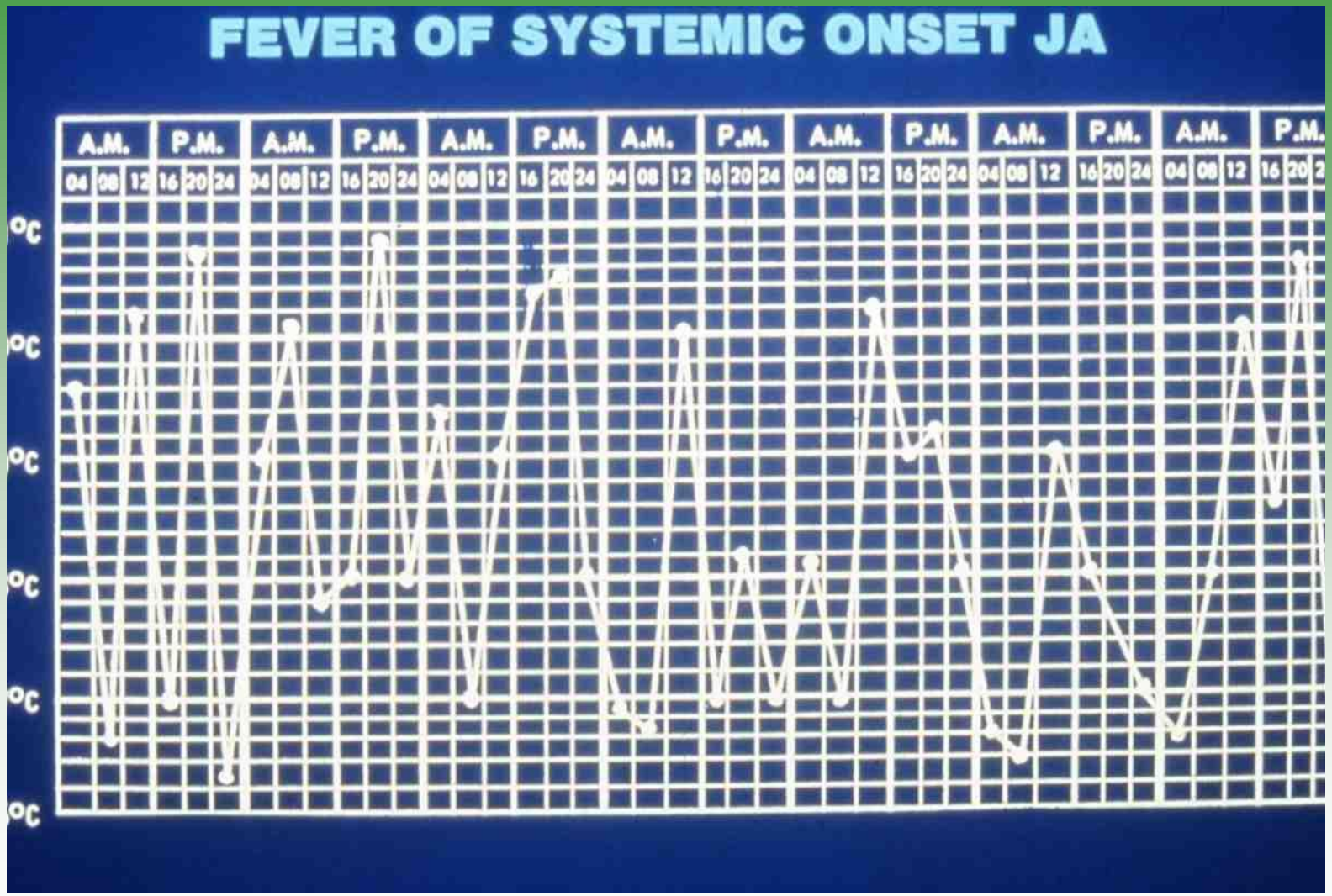
Nodules

Vasculitis


NORMAL
SYNOVITIS
bursitis
tendinitis
synovitis

Juvenile Arthritis with Systemic onset (20% of JA patients)

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%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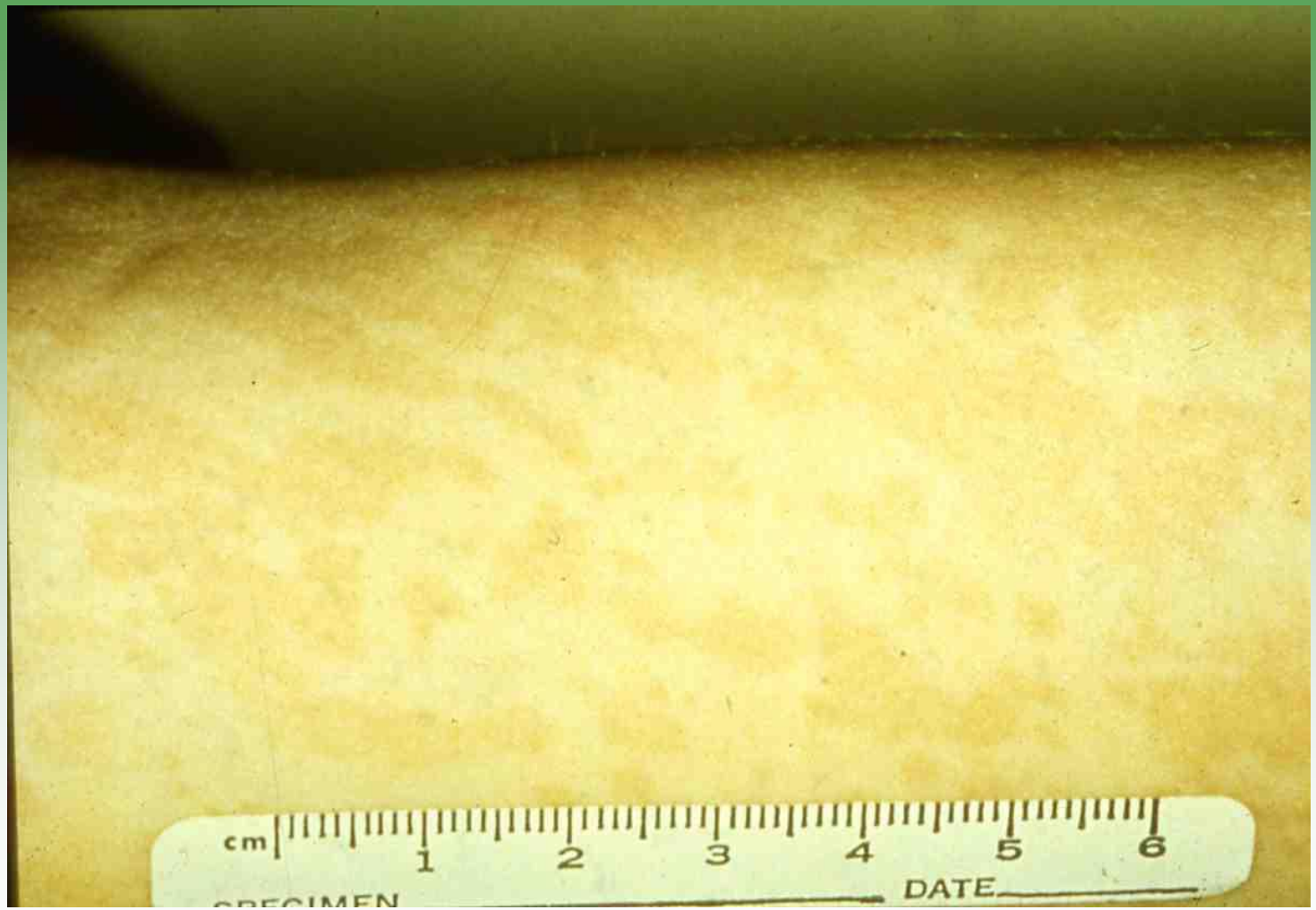
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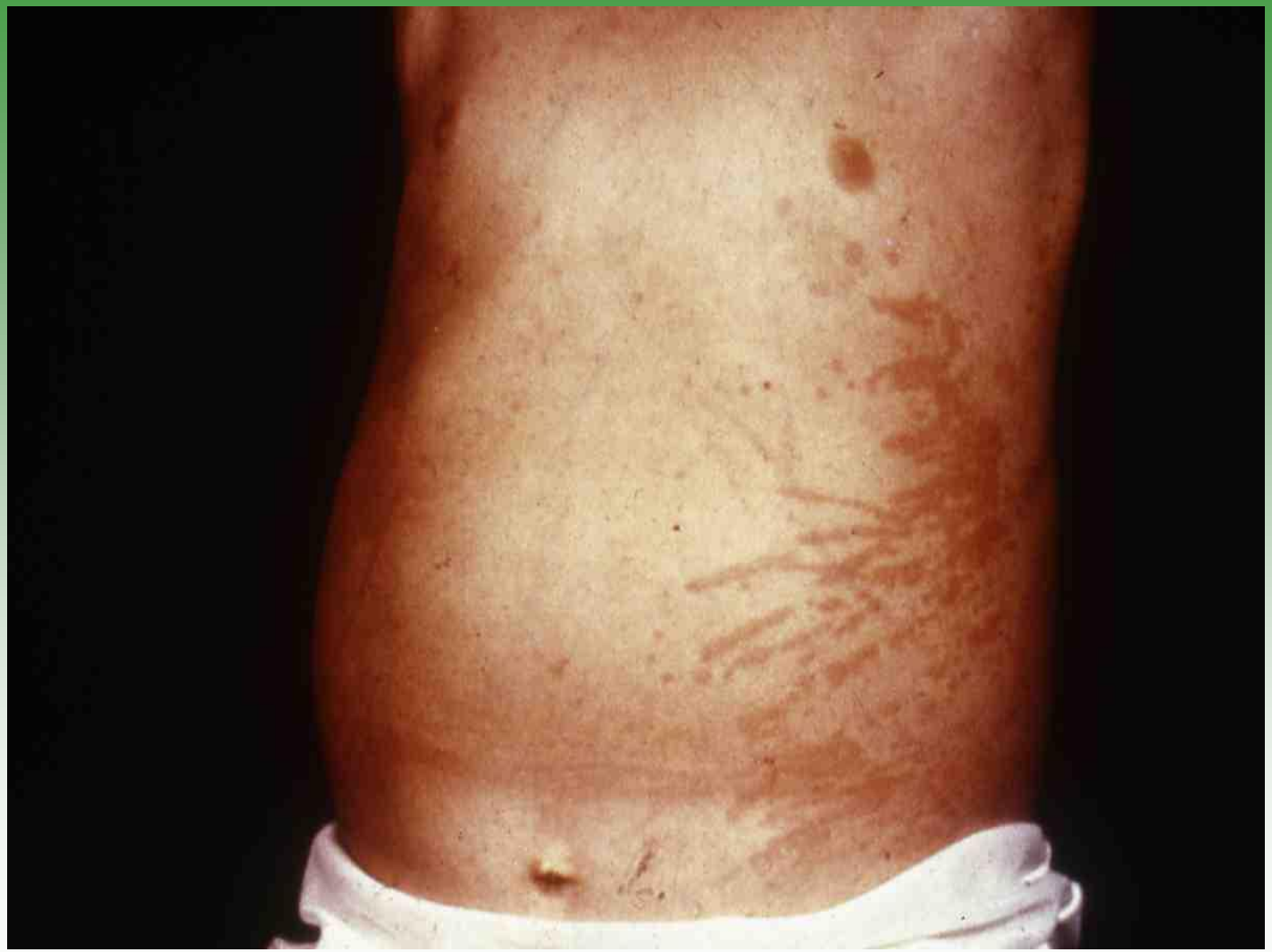
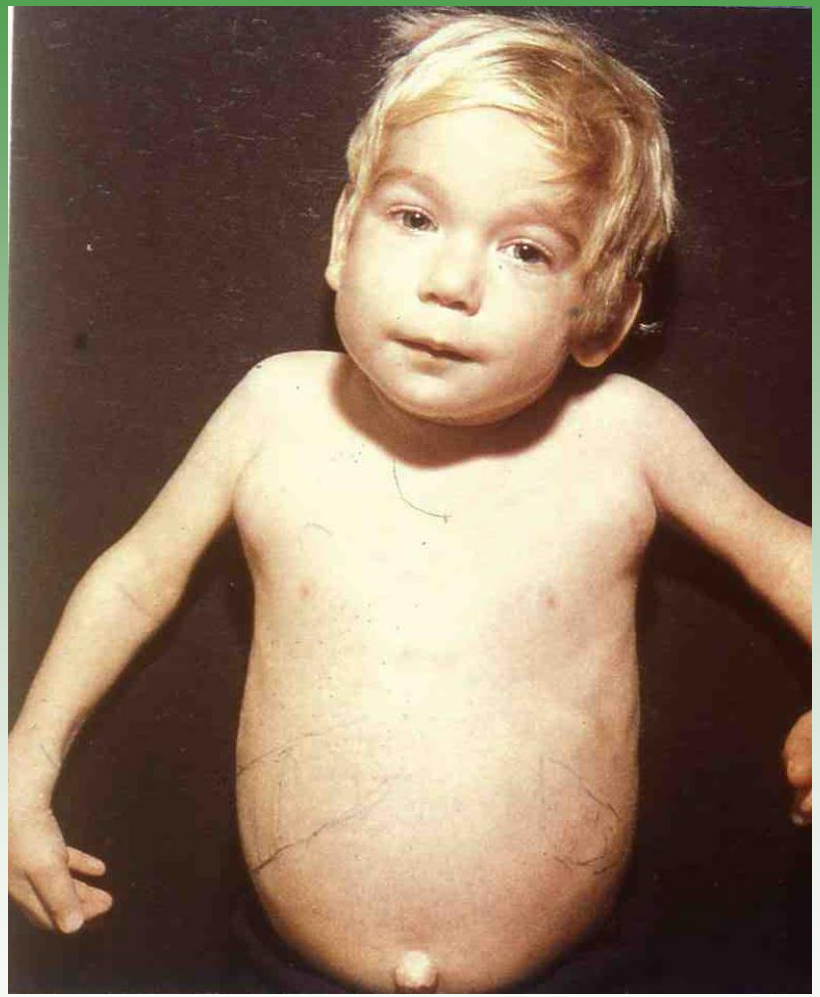
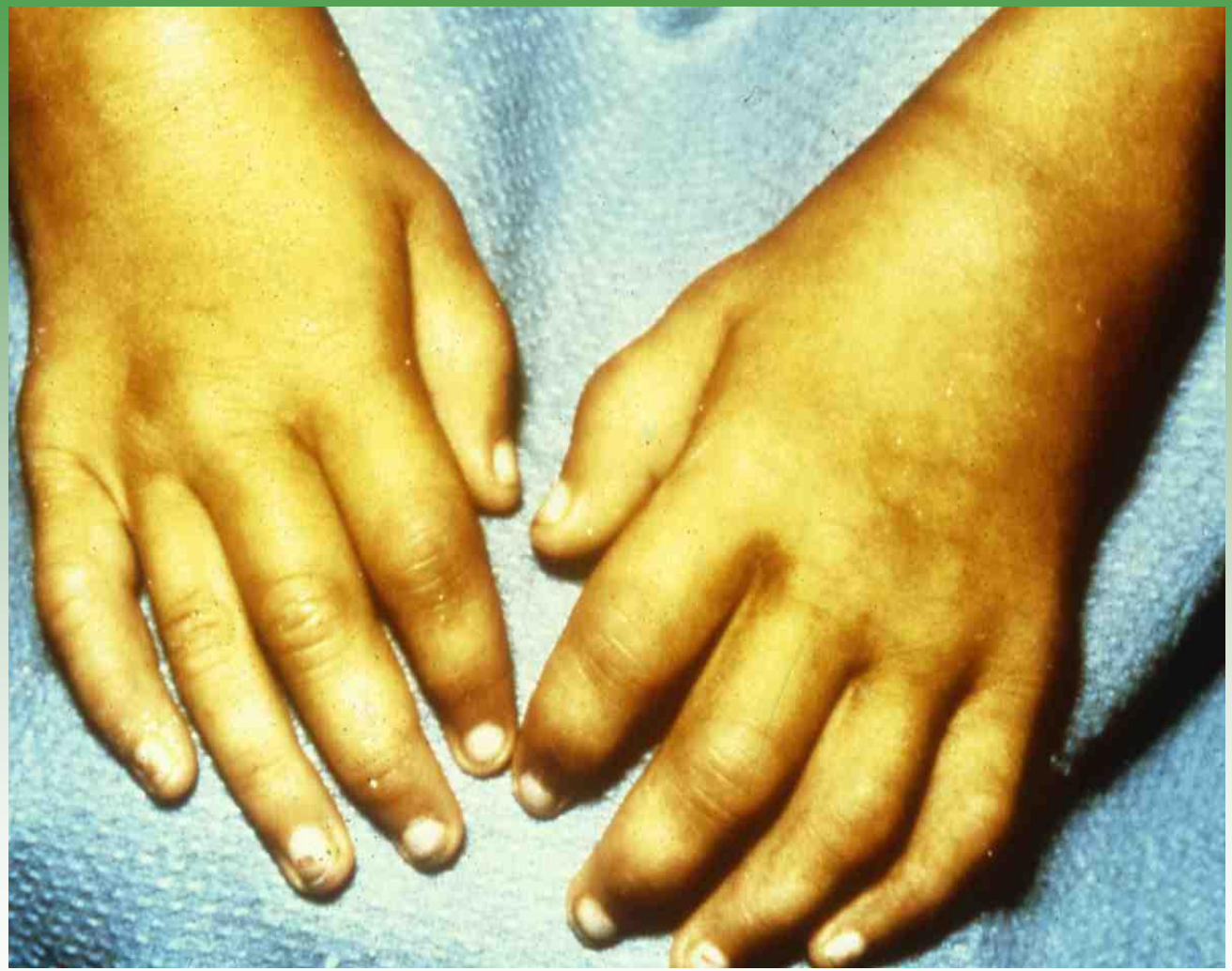
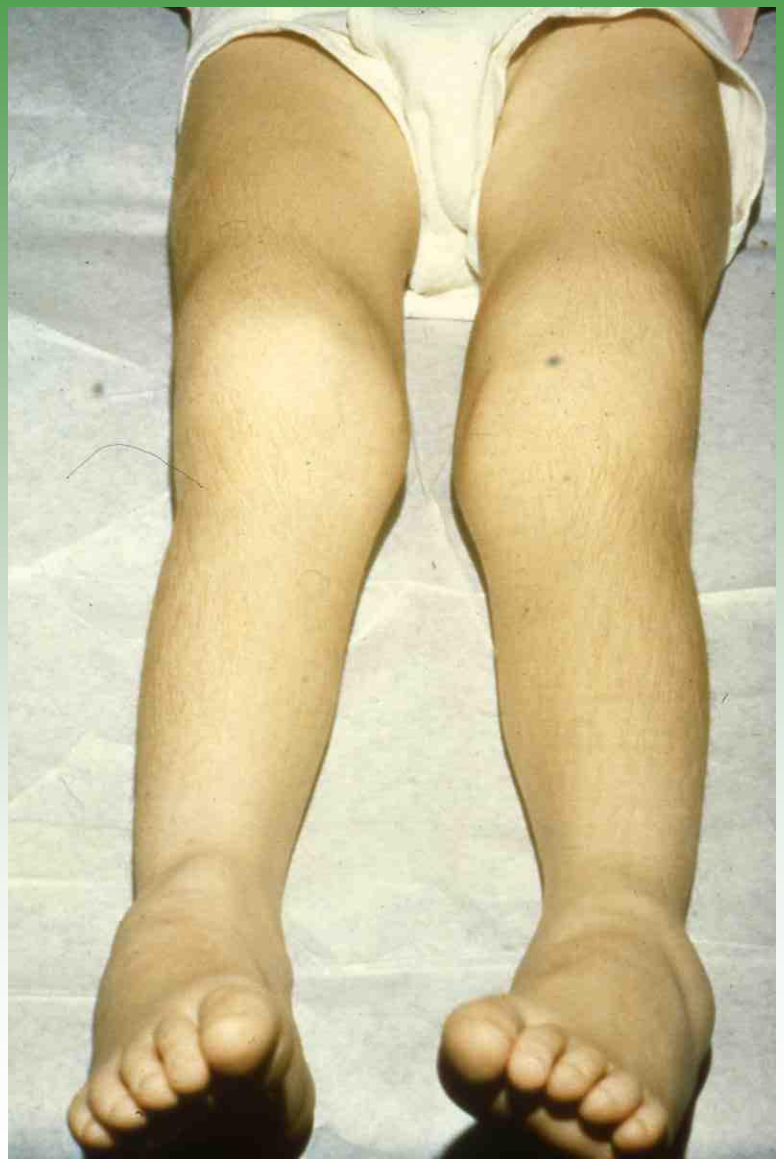


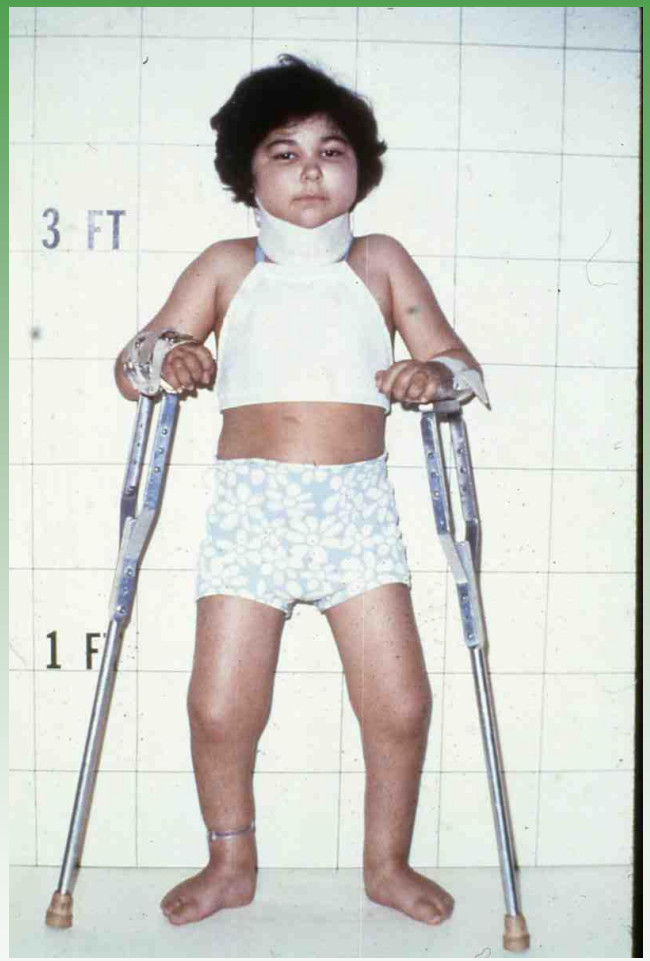


Juvenile arthritis with polyarticular onset (30% of JA patients)

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%



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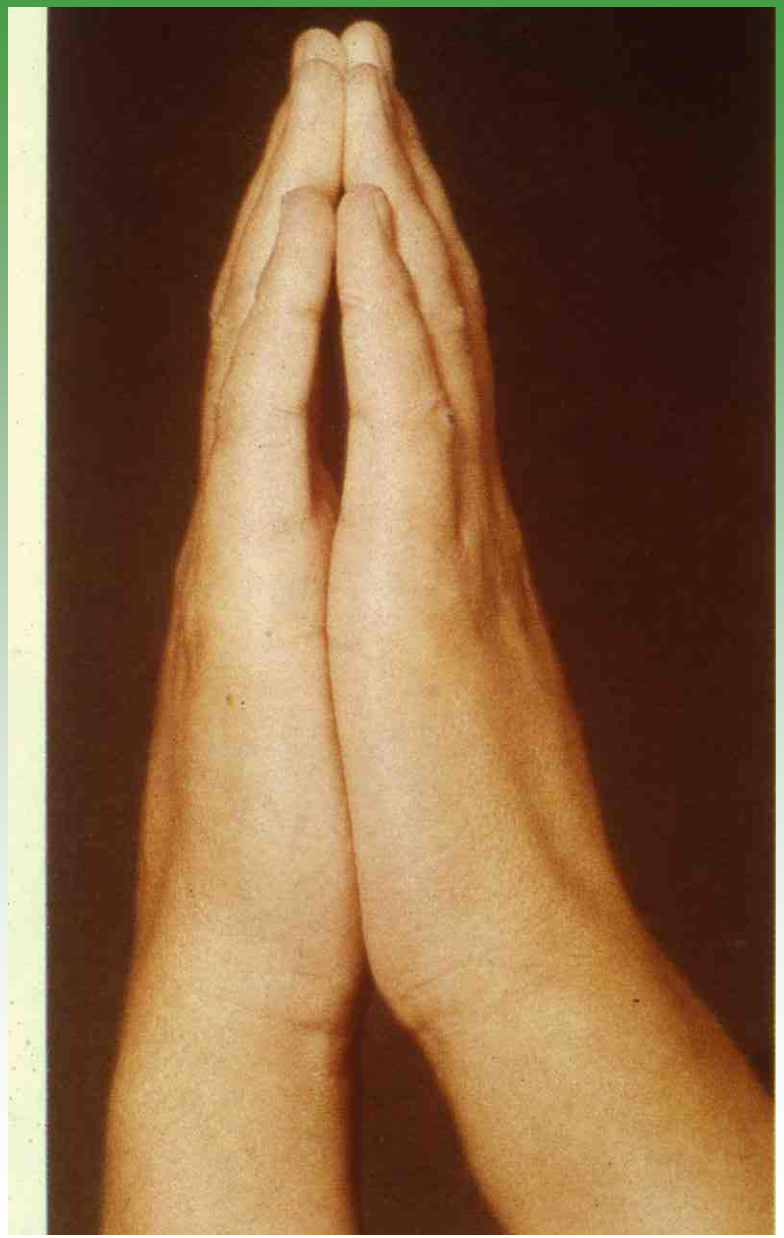

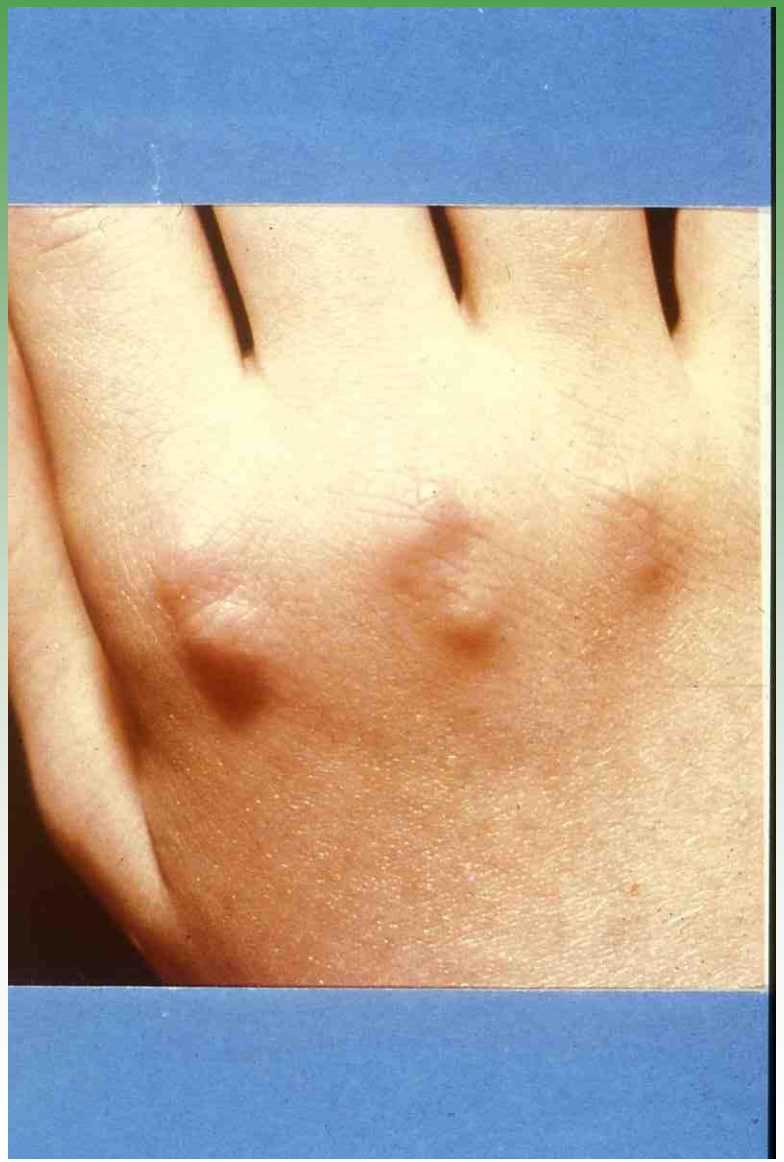

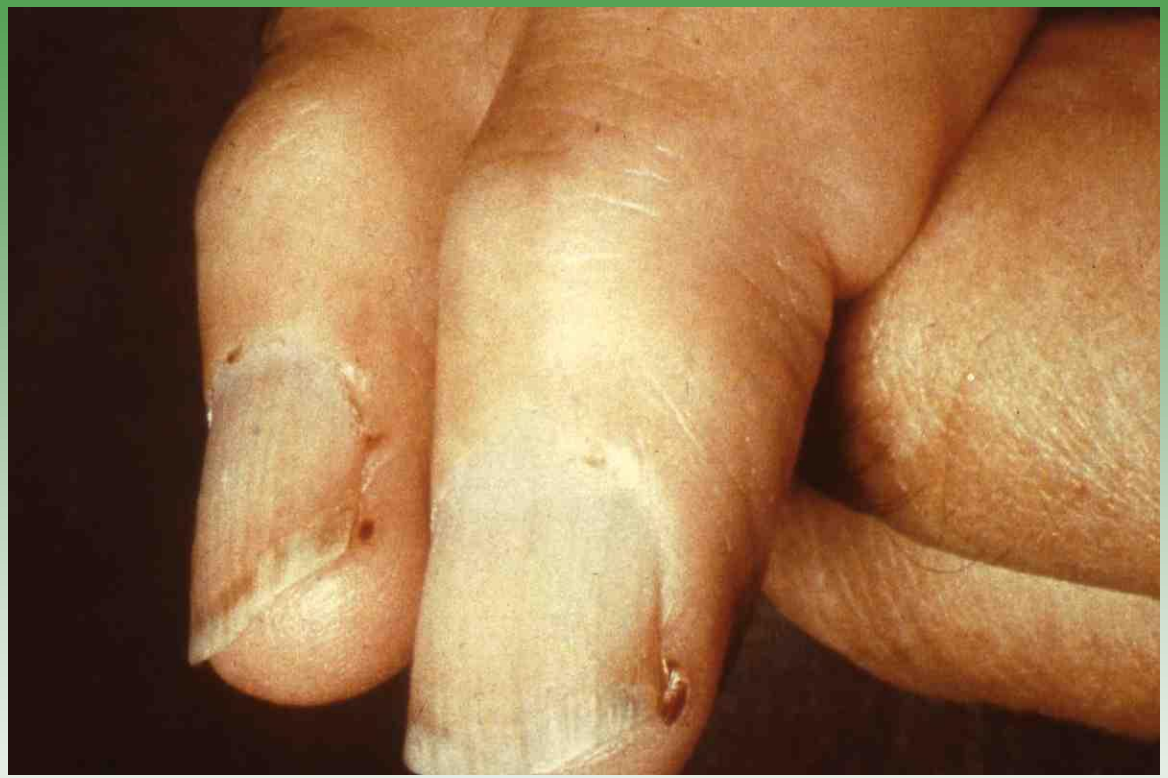

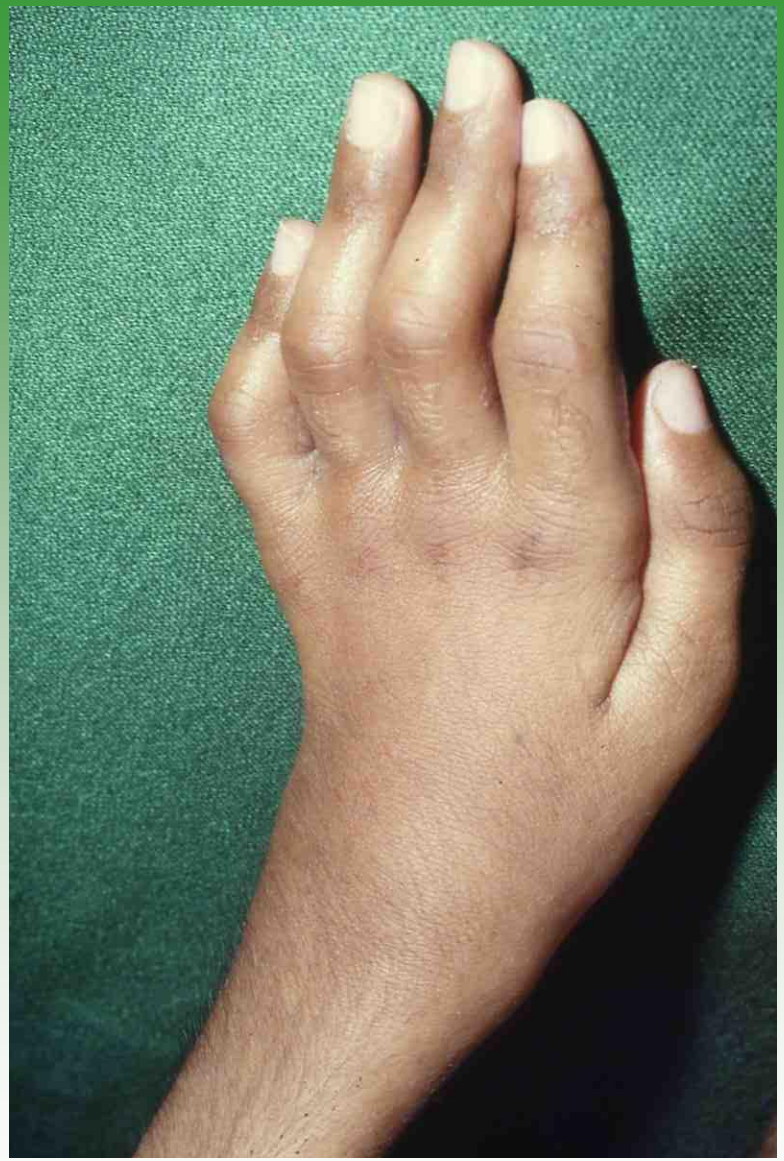

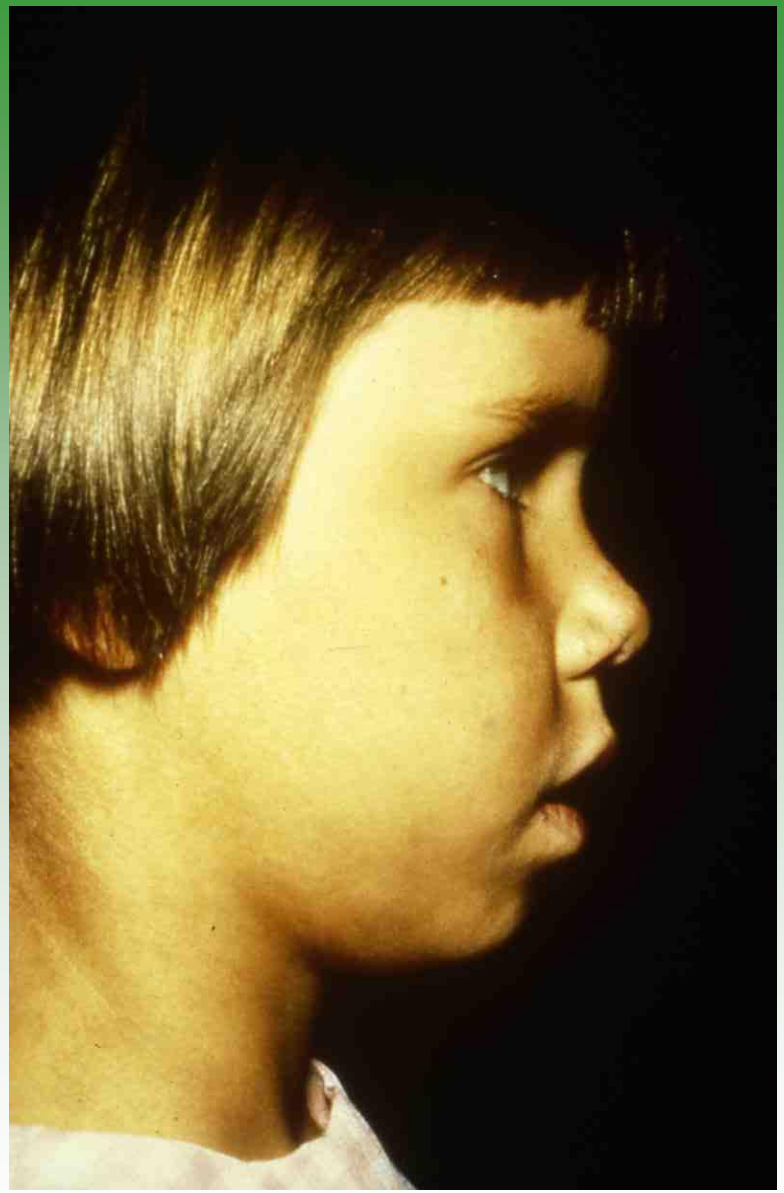

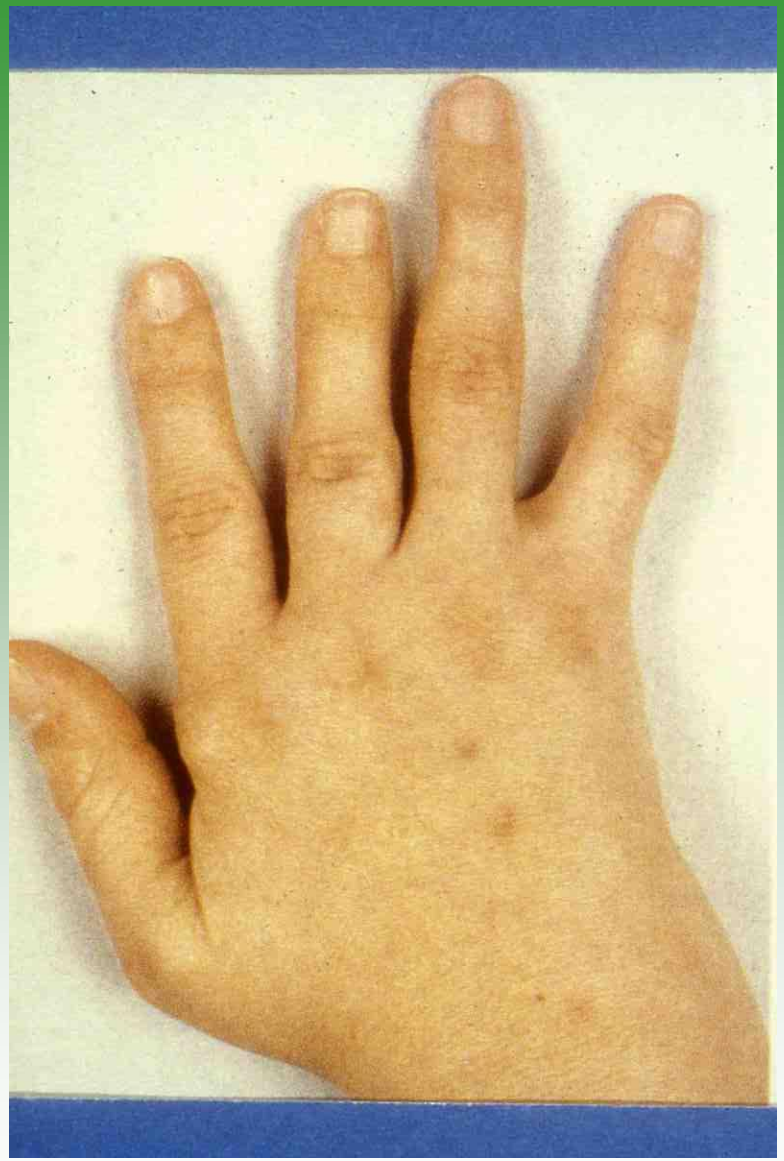

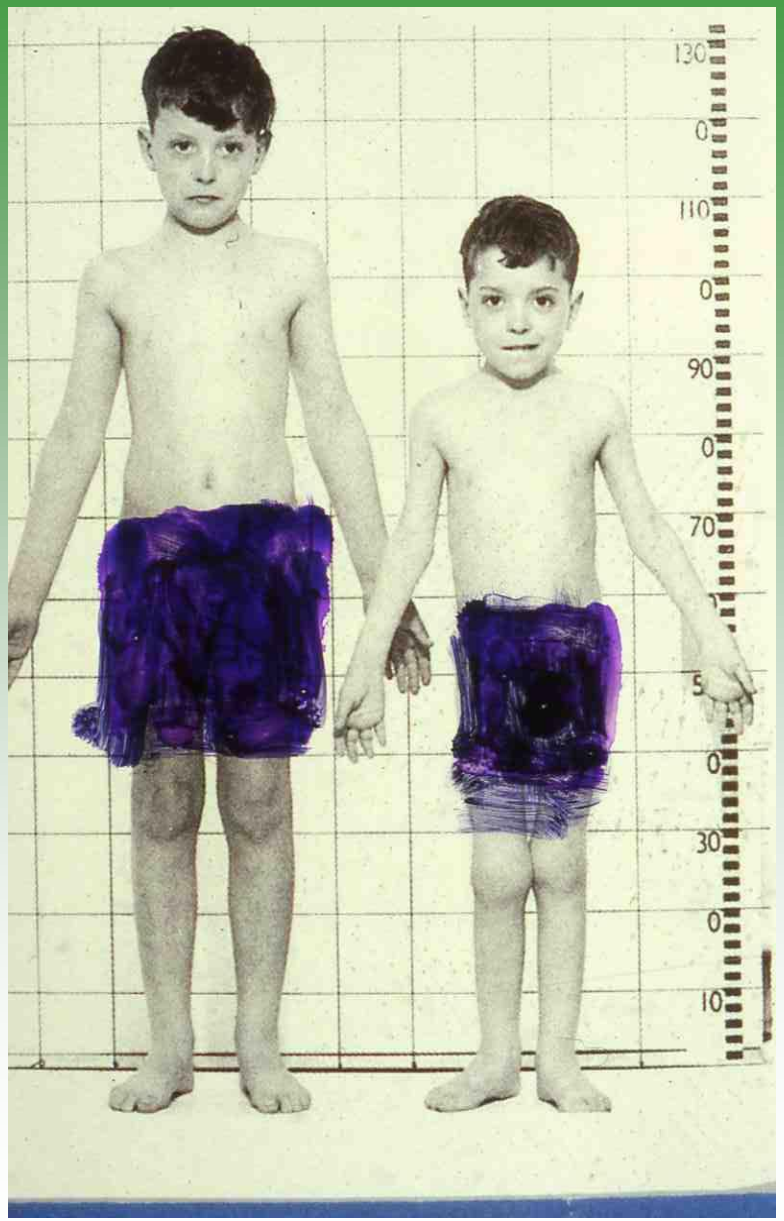
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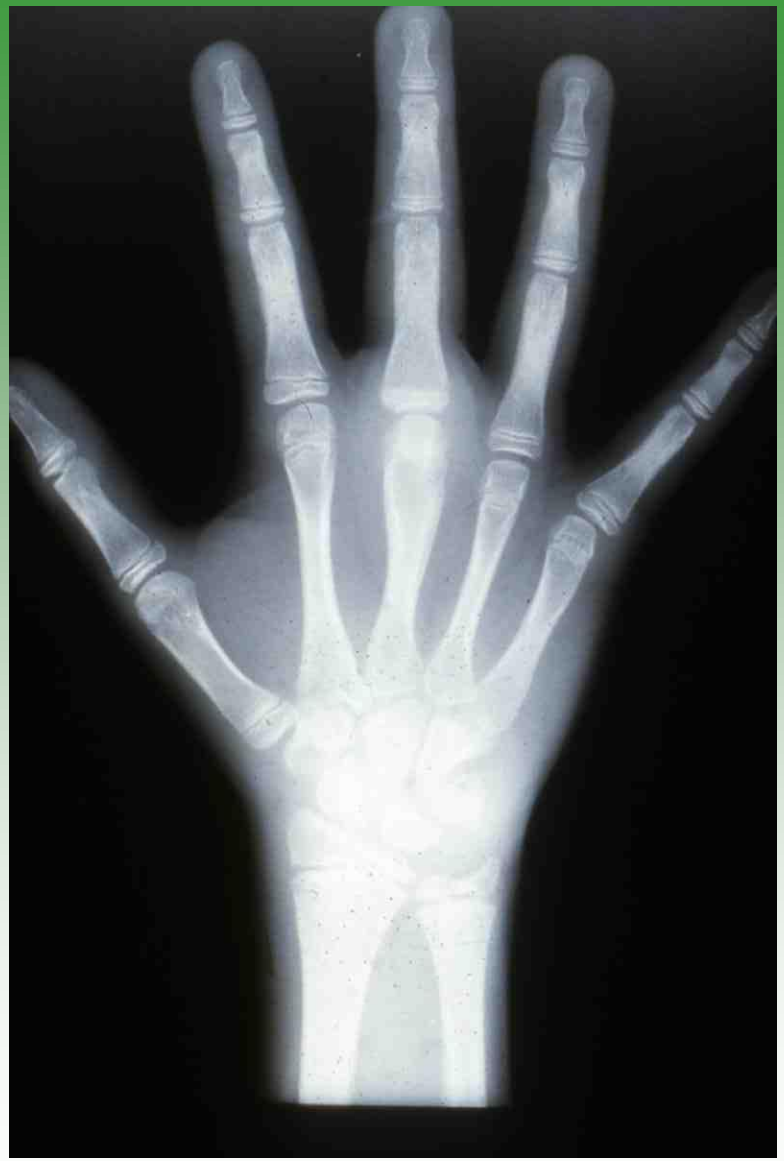
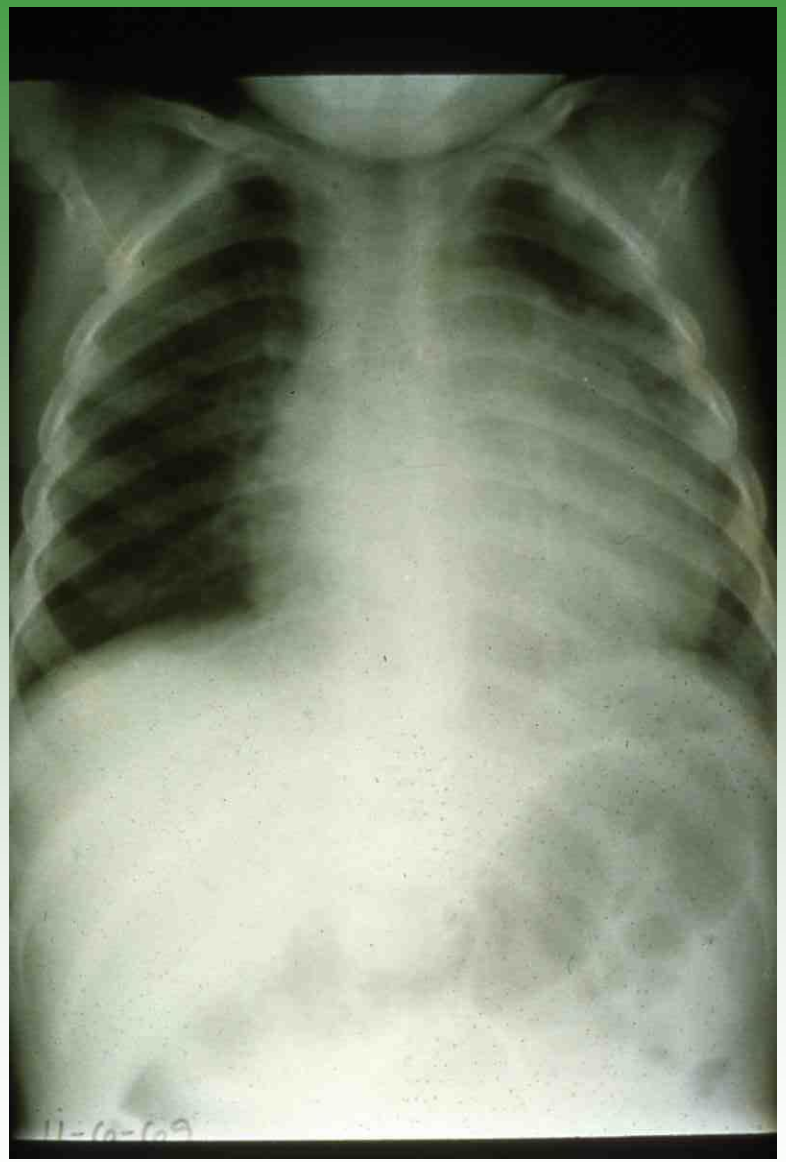
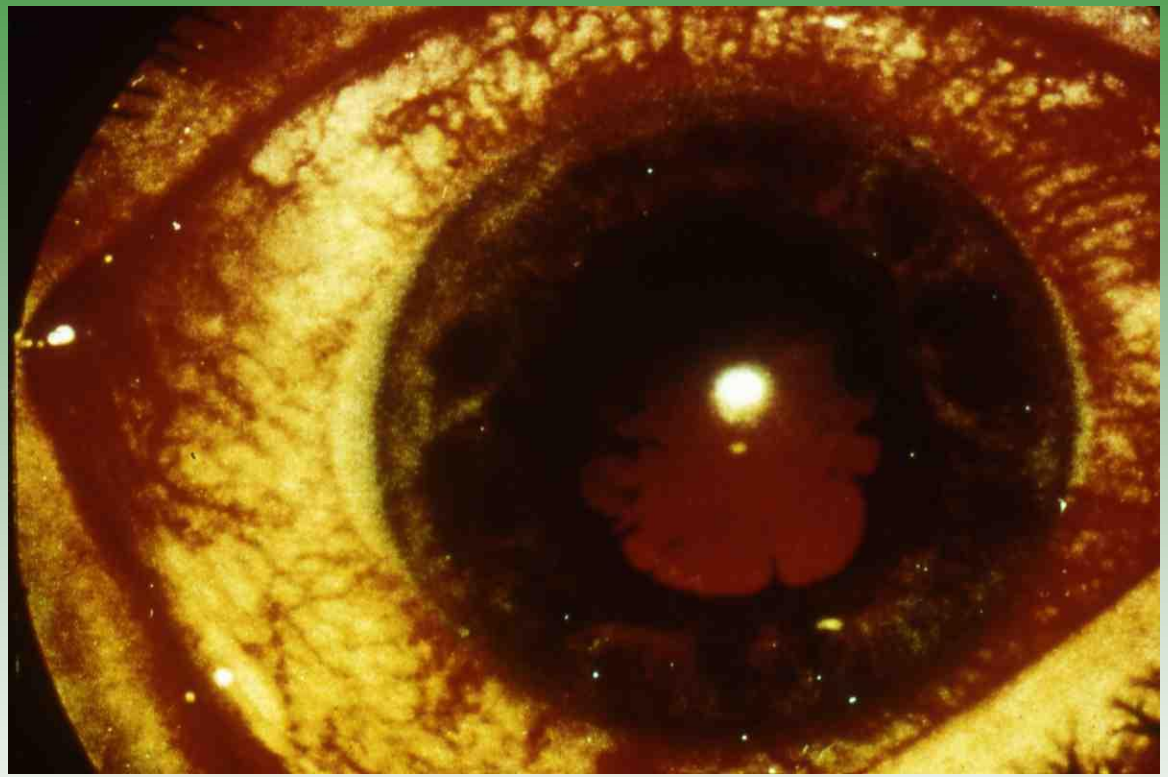




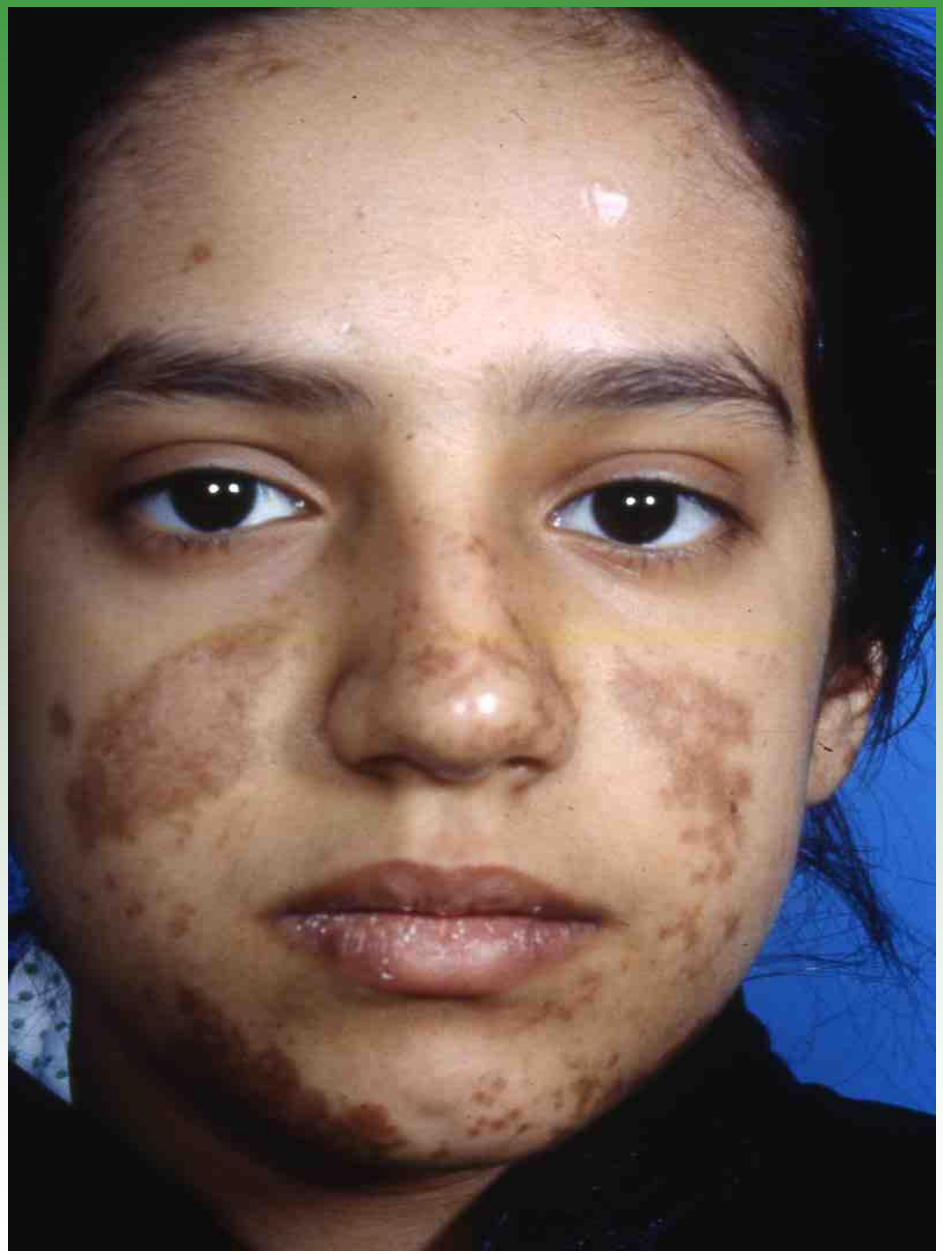
Juvenile arthritis with pauciarticular onset (50% of JA patients)

SUBGROUP ONE (35%)		SUBGROUP TWO (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



Management of Juvenile Arthritis

Accurate assessment of each individual patient	
Treatment for arthritis:	**Treatment for extra-articular manifestations:**
Drugs: **First line –** ▪ nonsteroidal anti-inflammatory drugs (NSAIDs) **Second line** ▪ Intra-articular steroids(pauci-aticular) ▪ antimalerials [hydroxychloroquine] ▪ sulfasalazine **Third line** ▪ steroids ▪ cytotoxic(Methotrexate),Blocking agents (etanercept, infliximab) **Physical and occupation therapy** **Orthopedic therapy**	**Drugs for systemic symptoms:** ▪ NSAIDs ▪ steroids occasionally needed **Drugs for iridocyclitis:** ▪ topical steroids and dilating agents ▪ systemic steroids needed occasionally ▪ Blocking agents
Consideration of whole child and child's family	



GEOGRAPHIC AND RACIAL DISTRIBUTION OF JSLE

RACE

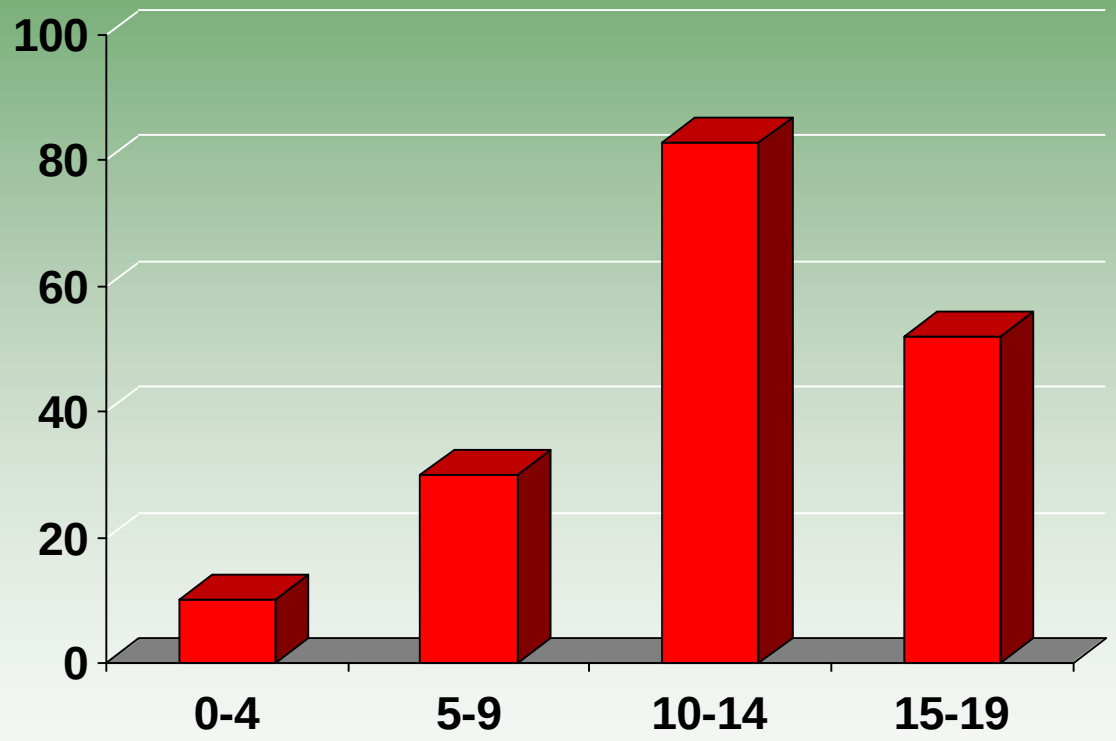
JSLE is common throughout the world

3:1 Incidence rate for black versus white females in USA.

AGE AT ONSET IN JSLE

- Rare before 5 years
- Increasingly more common in adolescence
- JSLE in the first decade: 3.5 – 15% of all cases
- More renal involvement in JSLE
- JSLE in the first decade is a more severe disease .

AGE AT ONSET IN JSLE



Classification criteria of SLE

Malar (butterfly) rash
Discoid-lupus rash
Photosensitivity
Oral or nasal mucocutaneous ulcerations
Nonerosive arthritis
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

[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

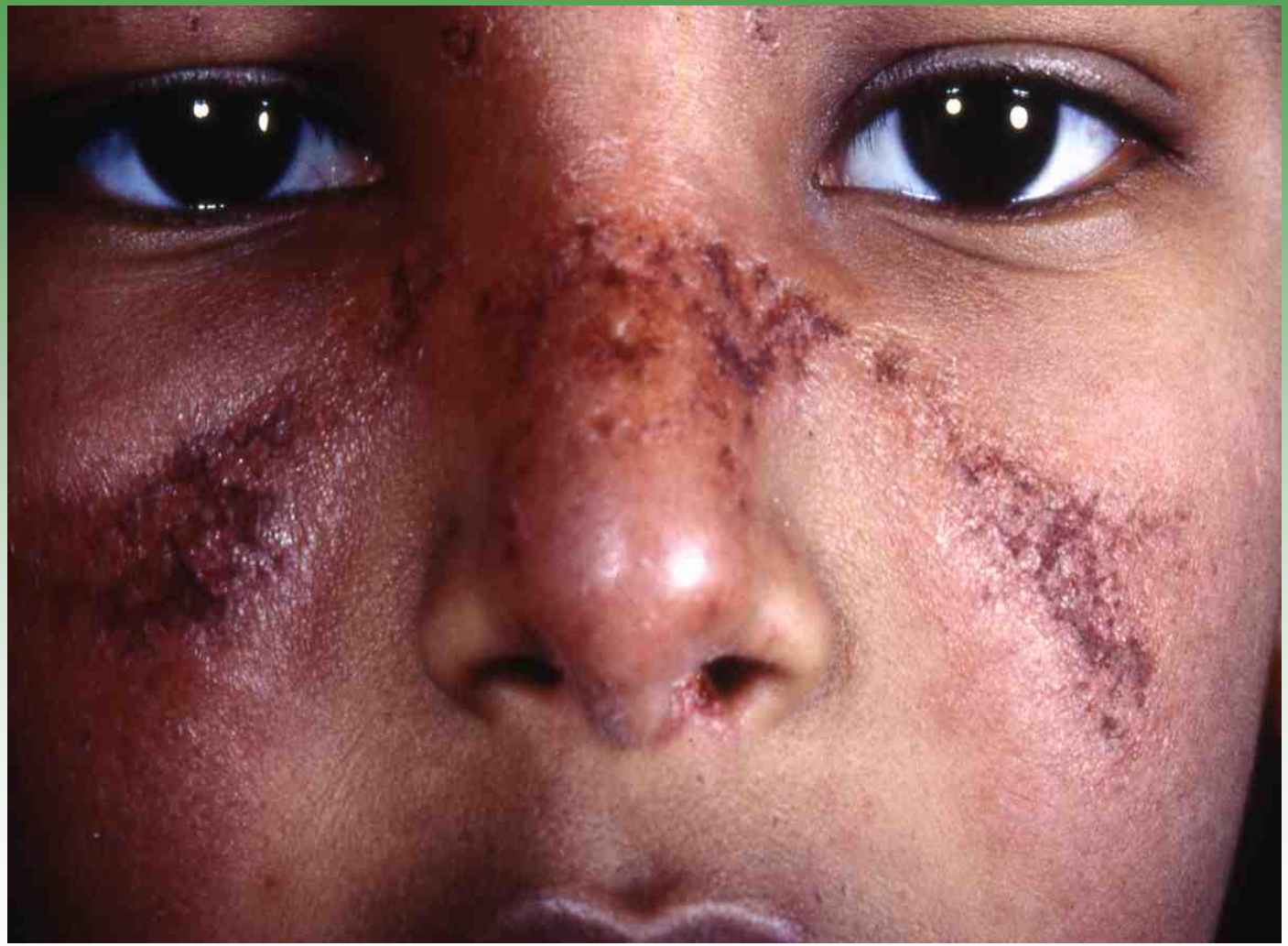
CLINICAL PRESENTATION

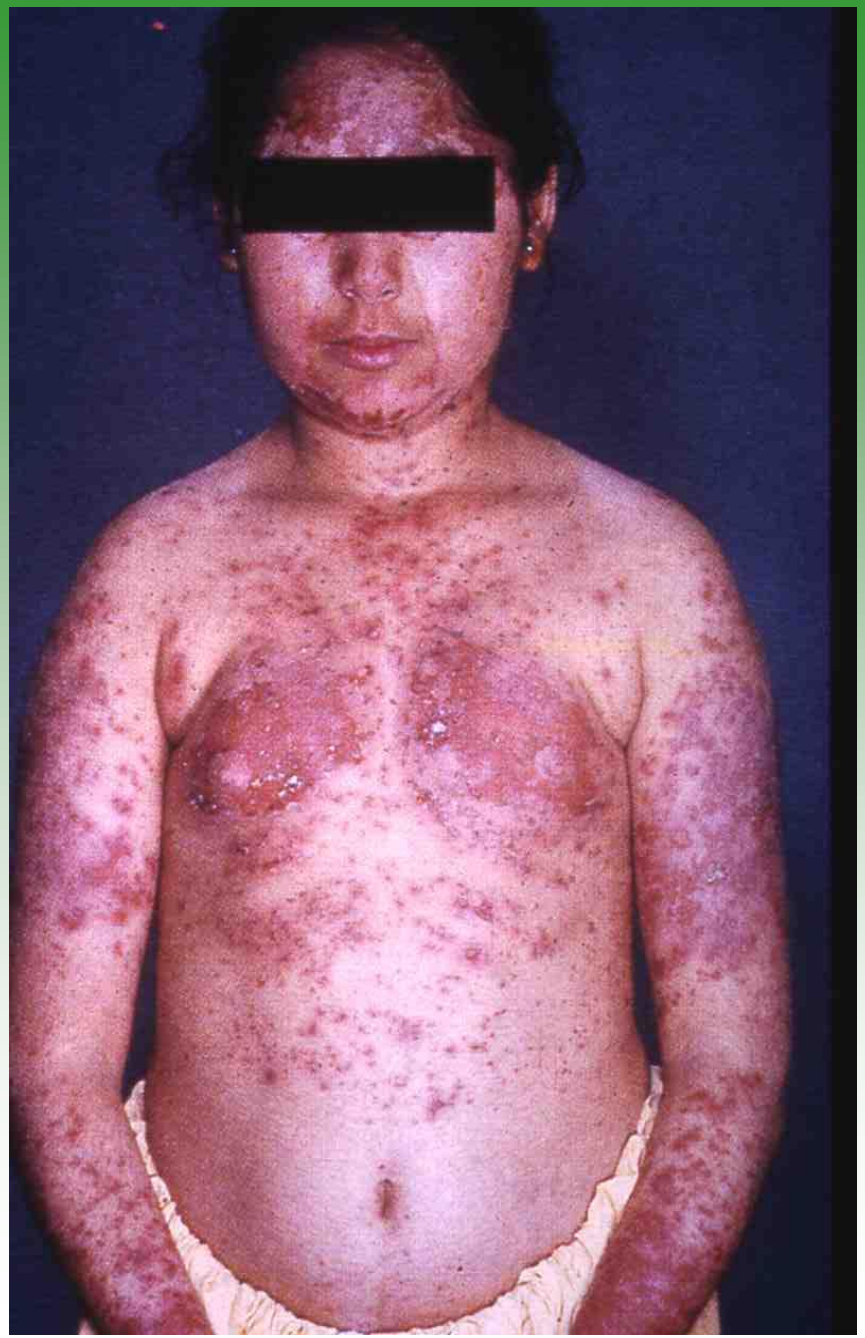
MUCOCUTANEOUS INVOLVEMENT

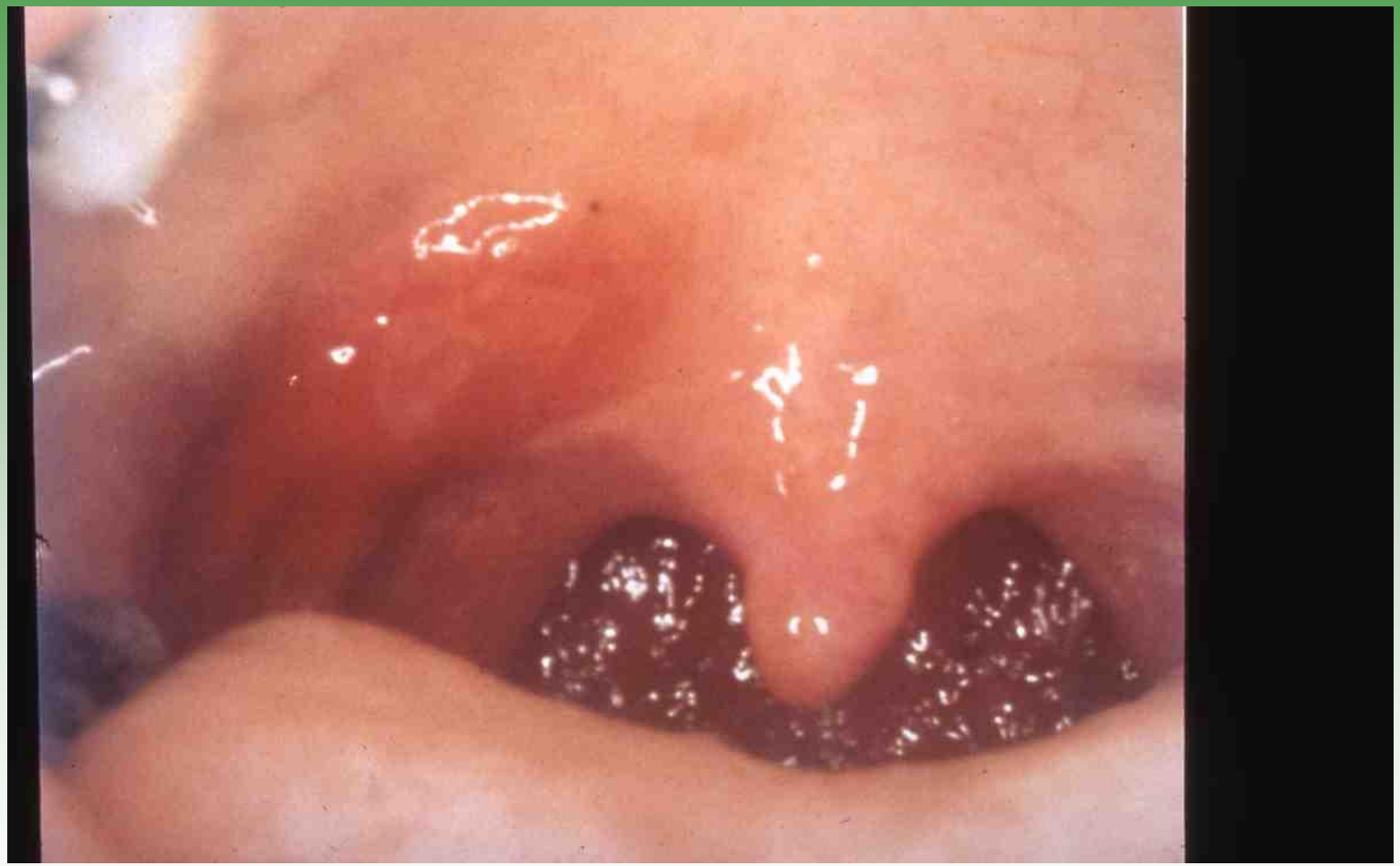
- Malar erythematous rash: Butterfly distribution. 25% of cases at onset and 50% of cases by 3 years .
- Abrupt onset and usually have systemic disease.
- Neonatal Lupus Erythematous: Lesions similar to seborrheic dermatitis, disappear spontaneously in 4-6 months.
- Discoid lupus: Discret, round, erythematous scaly patches with minimal systemic involment

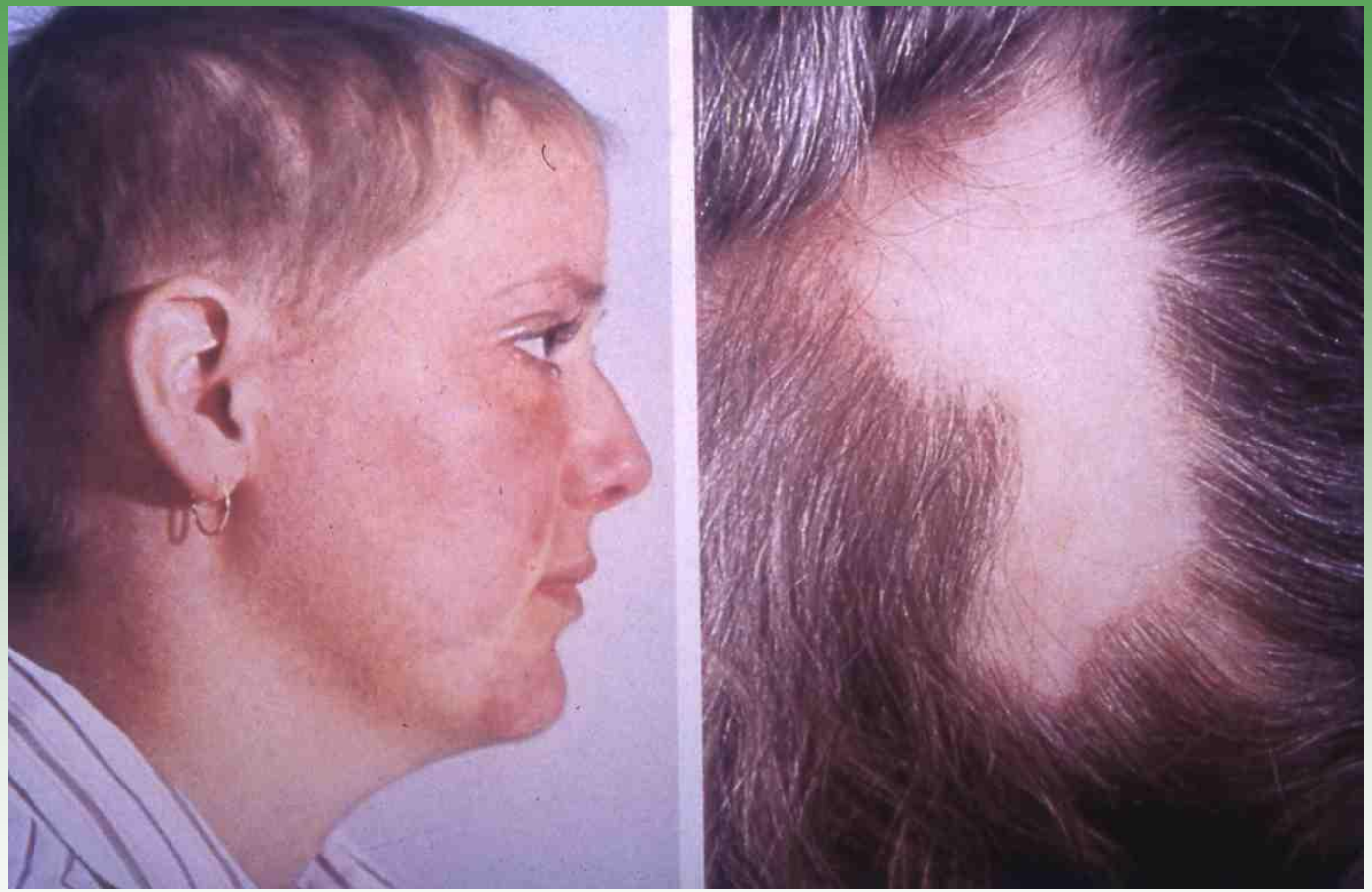
MUCOCUTANEOUS INVOLVEMENT

- Nasal & palatal ulcerations in 50% cases (perforation)
- Alopecia: Generalized thinning with frontal hair.Britle and kinky changes occur frequently in active disease.
- Raynanud’s phenomenon: It may precede the diagnosis by many years.









CARDIOVASCULAR INVOLVEMENT

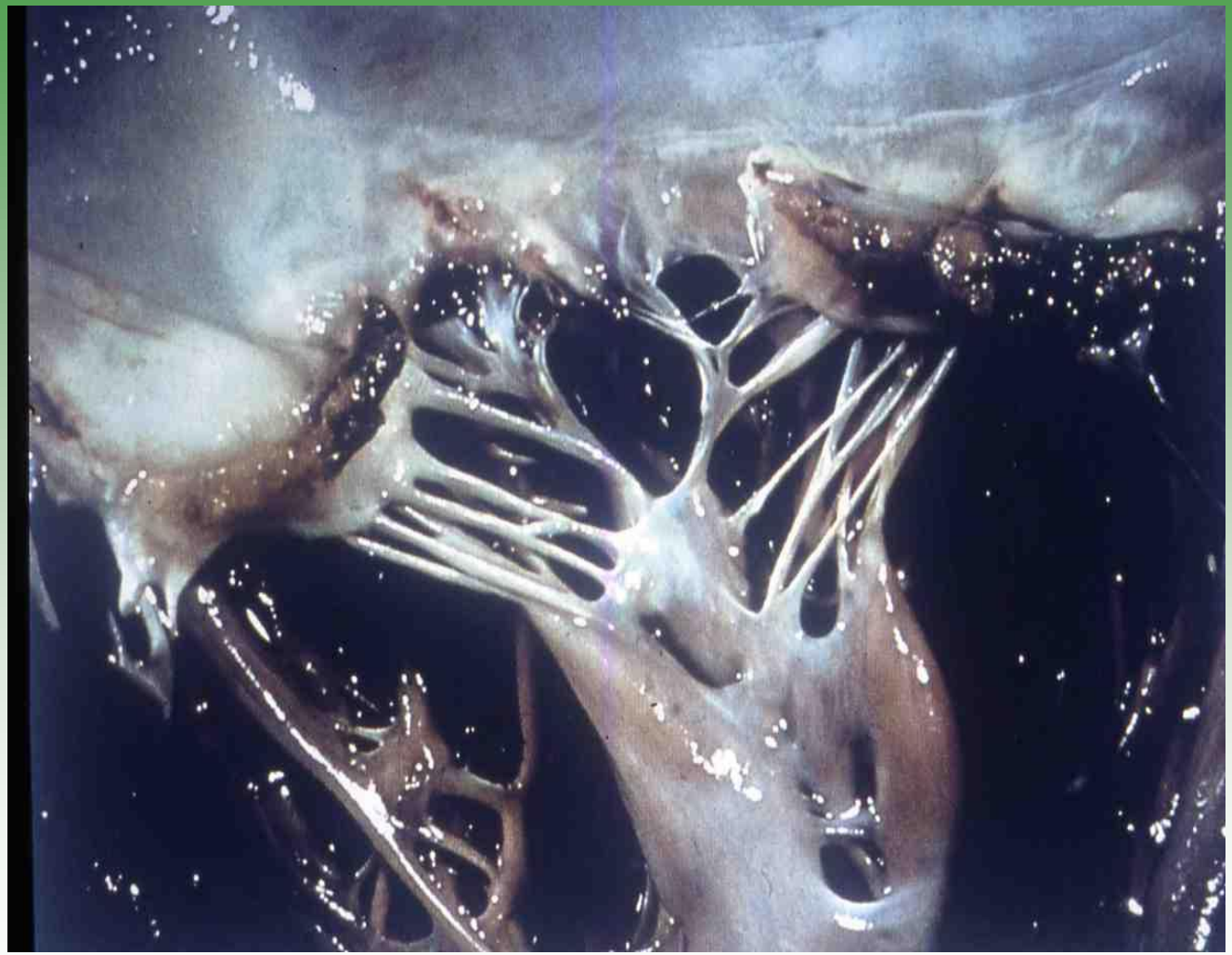
CARDIAC

- Pericarditis
- Myocarditis
- Endocarditis (Libman-Sacks)
- Conduction abnormalities

CORONARY ARTERY DISEASE

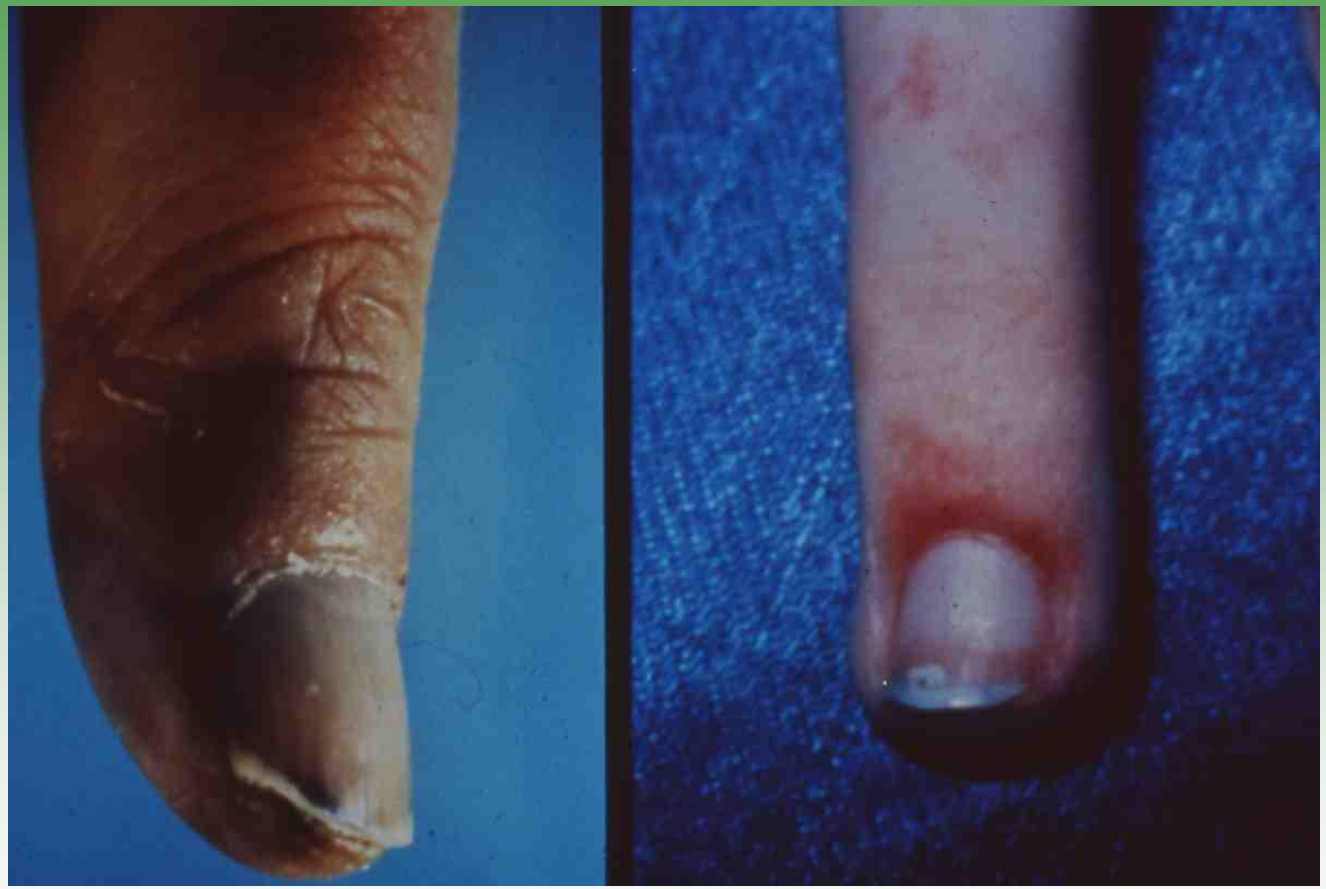
OTHER VASCULAR MANIFESTATIONS

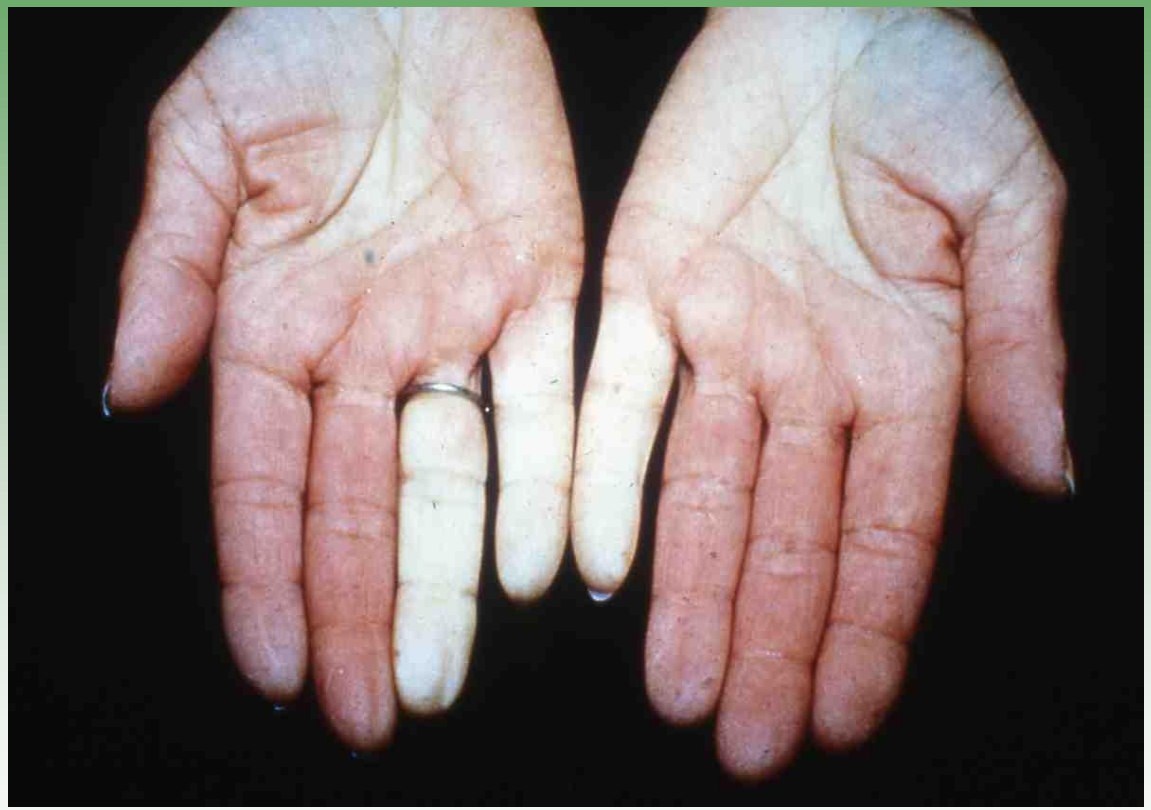
- Raynaud's phenomenon
- Hypertension
- Arteritis
- Venous disease

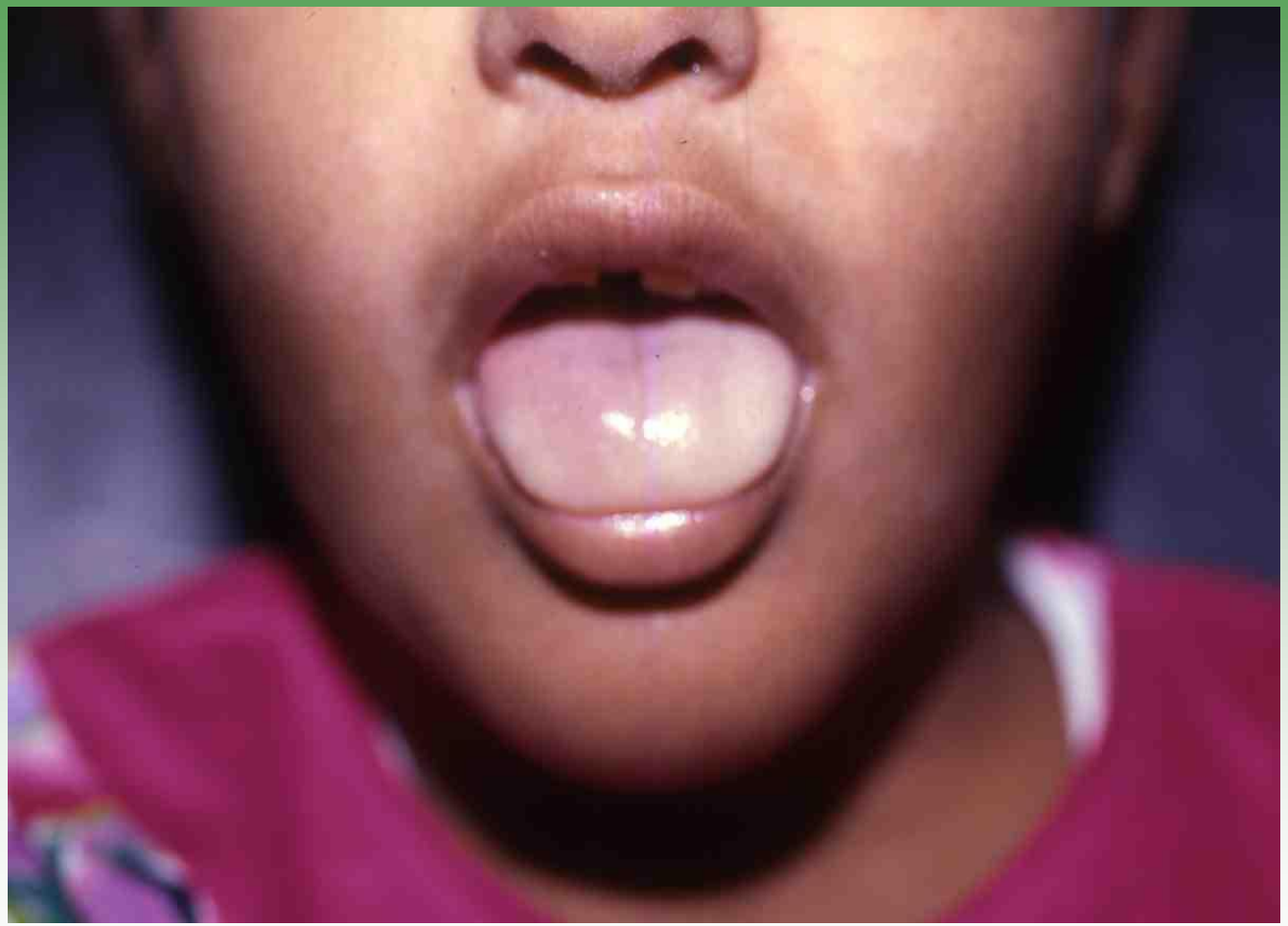


VASCULITIS IN SLE

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







FREQUENCY OF HEMATOLOGIC ABNORMALITIES IN CHILDREN WITH SLE AT ONSET

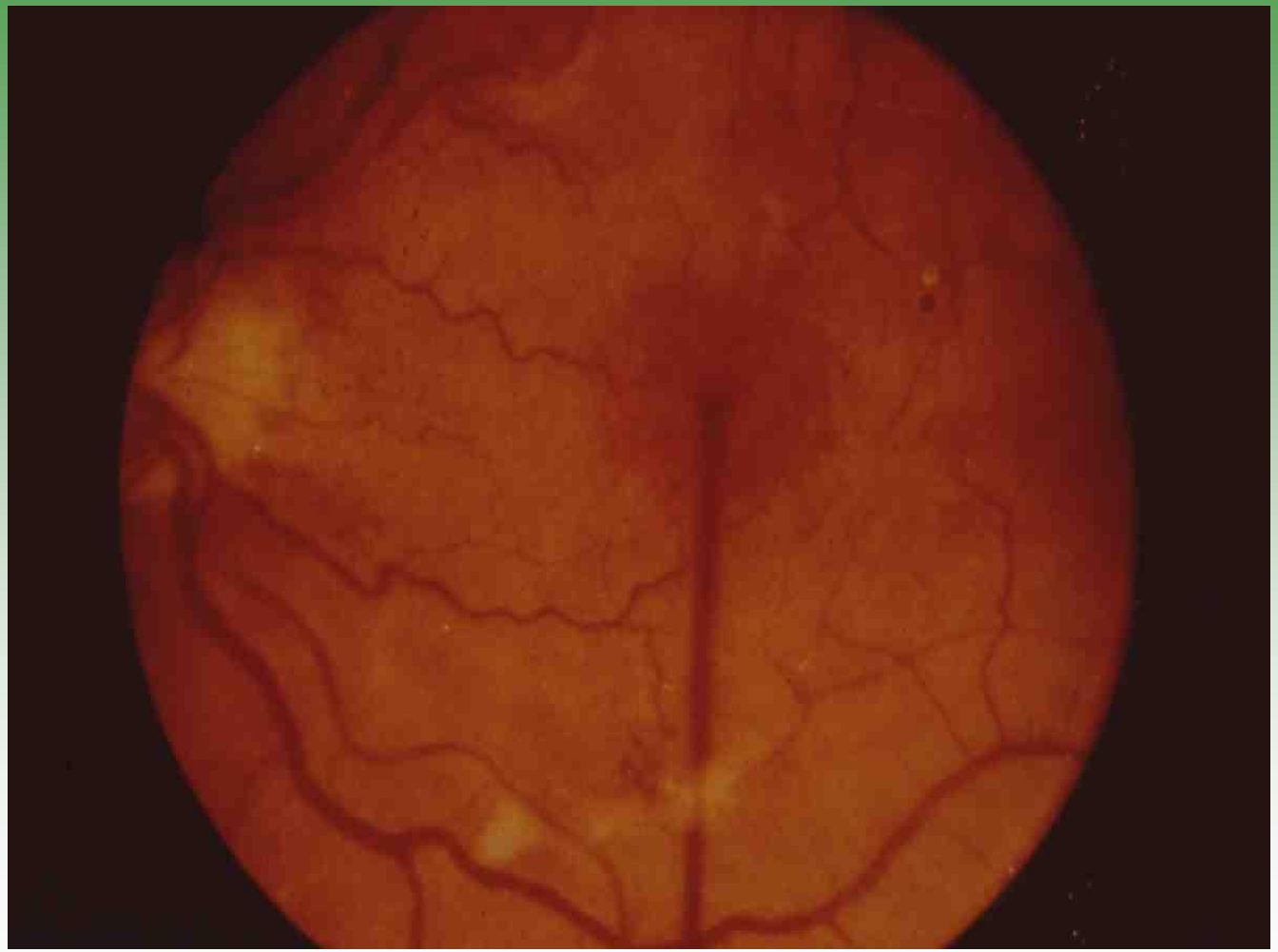
Abnormality	Patients (%)
Anemia (hematocrit < 30%)	50
Acute hemolytic anemia	5
Leukopenia <2,000 WBC/mm^3	10
Leukopenia <4,500 WBC/mm^3	40
Thrombocytopenia <150,000 pts/mm^3	30
Thrombocytopenia <100,000 pts/mm^3	5

G.I. MANIFESTATIONS

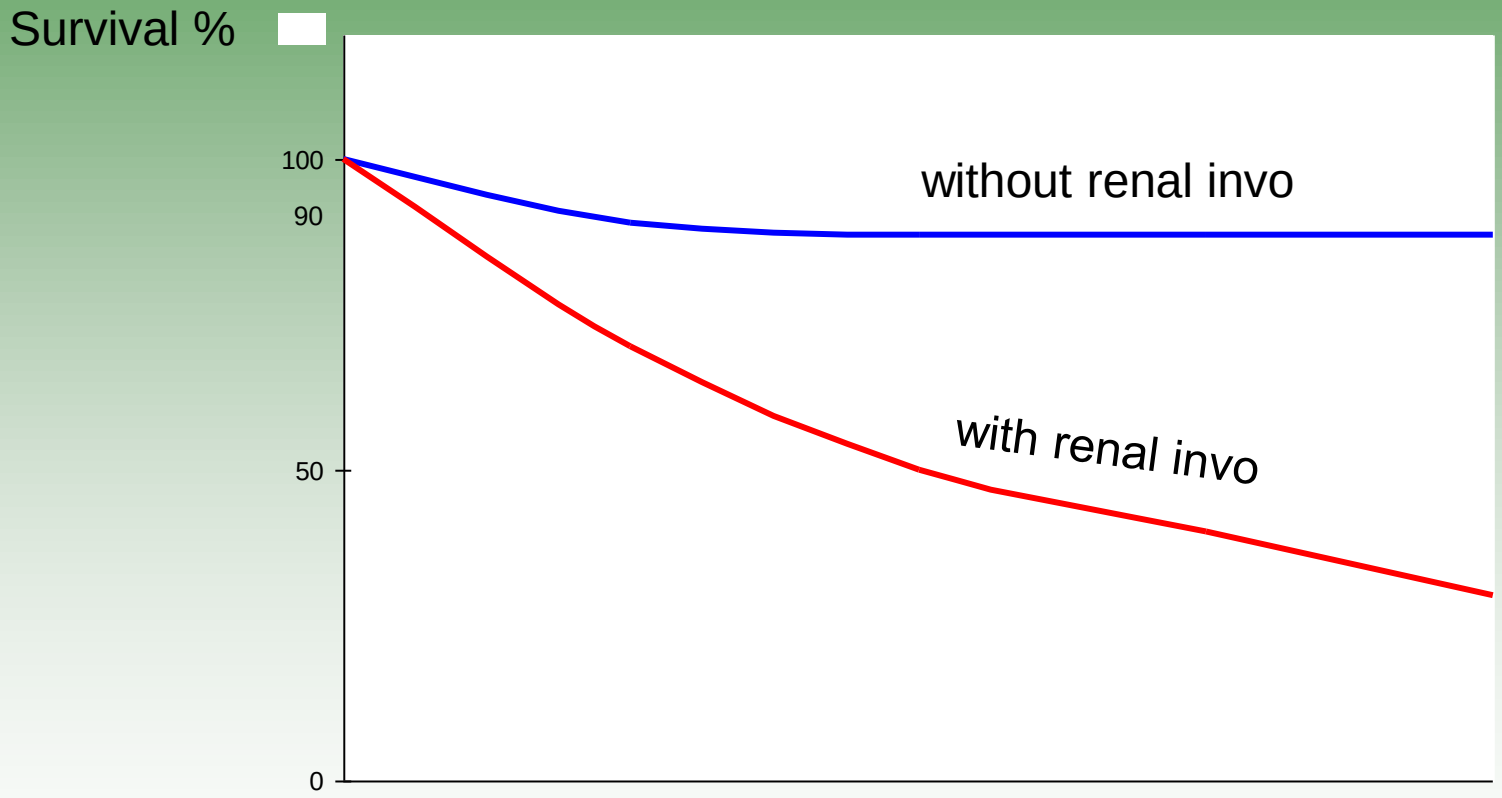
- 31% of cases have abdominal pain.
- Abnormal esophageal motility.
- Ascitis and pertonitis: 8-11%, peritoneal fluid shows high DNA, low component.
- Acute pancreatitis: de novo or steroids related.
- Mesentric artery thrombosis
- Malabsorption
- GI vasculitis: Edema, ulceration, gangrene , perforation

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. headach , aseptic meningitis, mysthenia gravis



Prognosis in SLE



Management of SLE

Question: What system is/are affected?

(history, clinical examination and investigations)

Non-immunosupressants	**Immunosuppresants**
- antimalarial (Hydroxychloroquine - NSAIDs (Ibuprofen, Naproxen)	- Corticosteroids (Prednisone) - DMARDs (MTX, Imuran, cellcept) - Biologics (Rituximab, Tocilizumab, Belimumab) - Cytotoxins (cyclophosphamide)

Management of SLE (cont.)

- I.V. immunoglobulin (IV Ig)
- Plasmapheresis
- **Other treatment:**
  - sunscreen
  - physical and occupational therapy
  - treatment of complications

DERMATOMYOSITIS AND POLYMYOSITIS

- Symmetrical progressive proximal weakness
- Muscle biopsy showing inflammatory changes
- Raised muscle enzymes (**CPK,AST,Aldolase**)
- Electromyography abnormalities
  (e.g. polyphasic potentials)
- Characteristic dermatological changes

Juvenile Dermatomyositis

Expanded criteria for diagnosis

- Typical findings on muscle MRI and ultrasonography
- Nail fold capillaroscopy abnormalities
- Calcinosis
- Dysphonia

Juvenile Dermatomyositis
Clinical Course

- Monocyclic (remission within 2-3 years)
- Polycyclic
- Chronic
- Ulcerative

Investigations

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

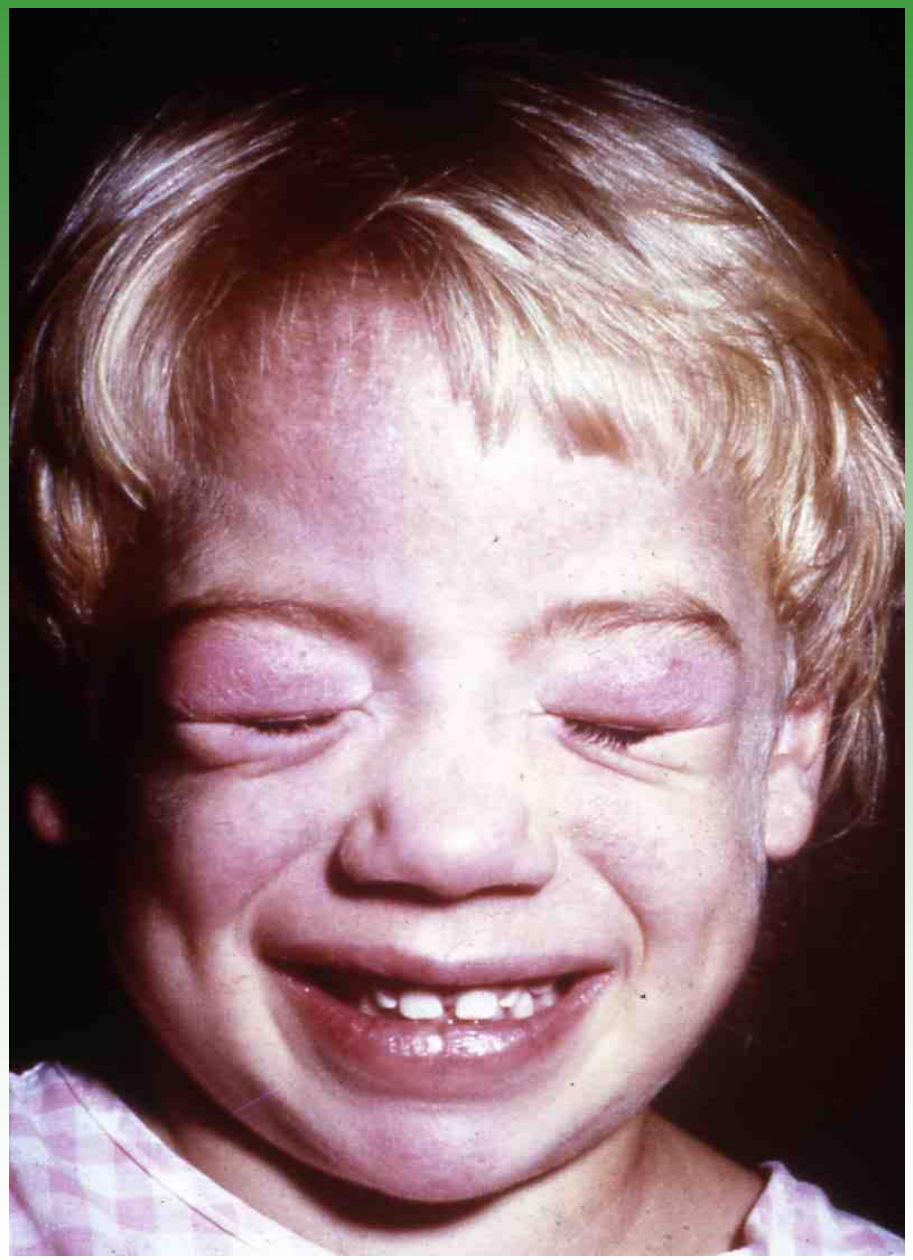
JDM - Treatment

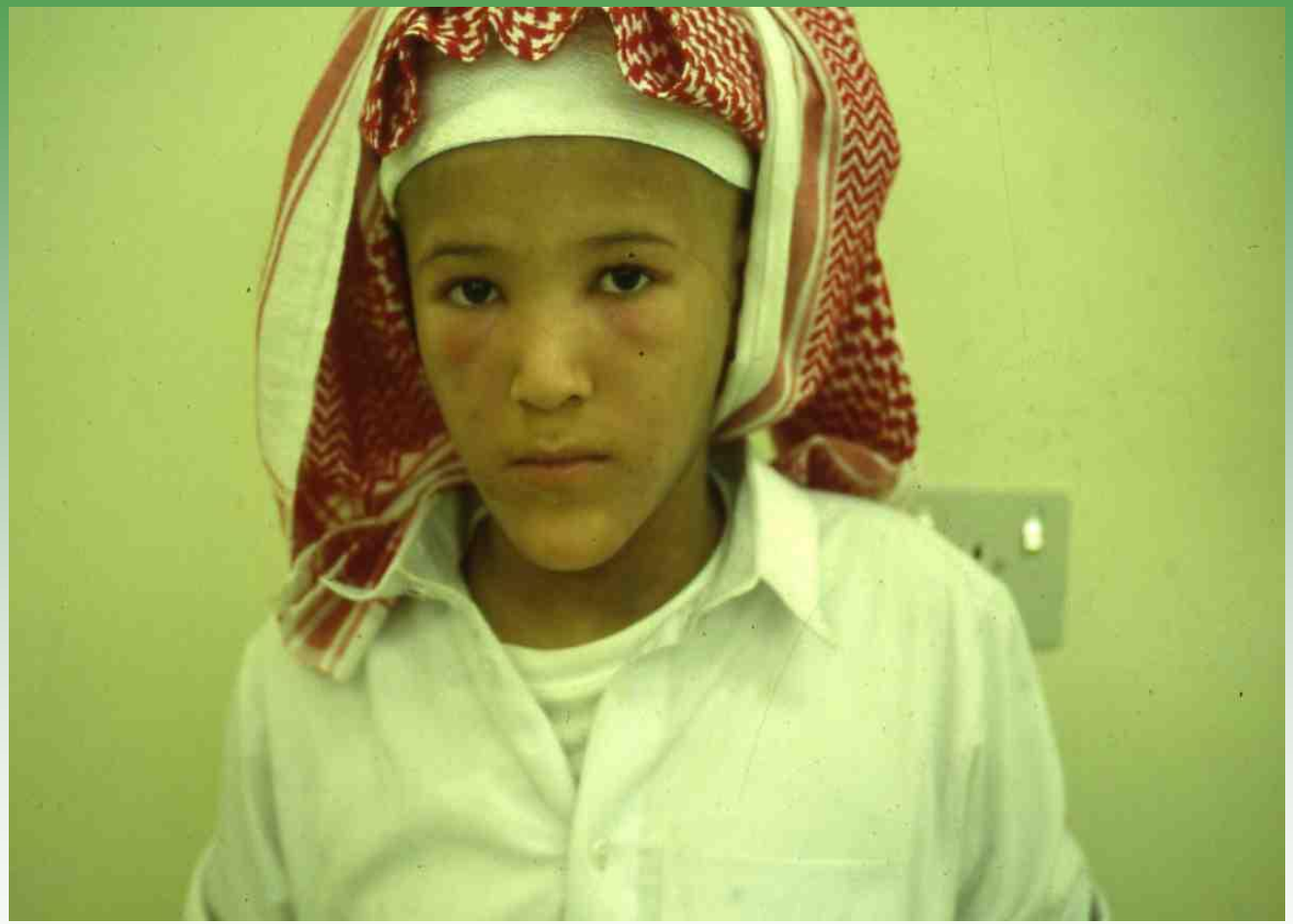
- Aims - minimize inflammation
  - improve function
  - prevent disability
- Early teamwork (Rheumatologist, Dermatologist, Physical Therapist and Primary Care Doctor)
- Drugs:
  - Corticosteroids (Prednisolone, IV, MP)
  - Immunoglobulin (IV Ig)
  - Methotrexate
  - Others (Hydroxychloroquine, anti-TNF therapy)
  - severe cases (Cyclophosphamide, Rituximab)

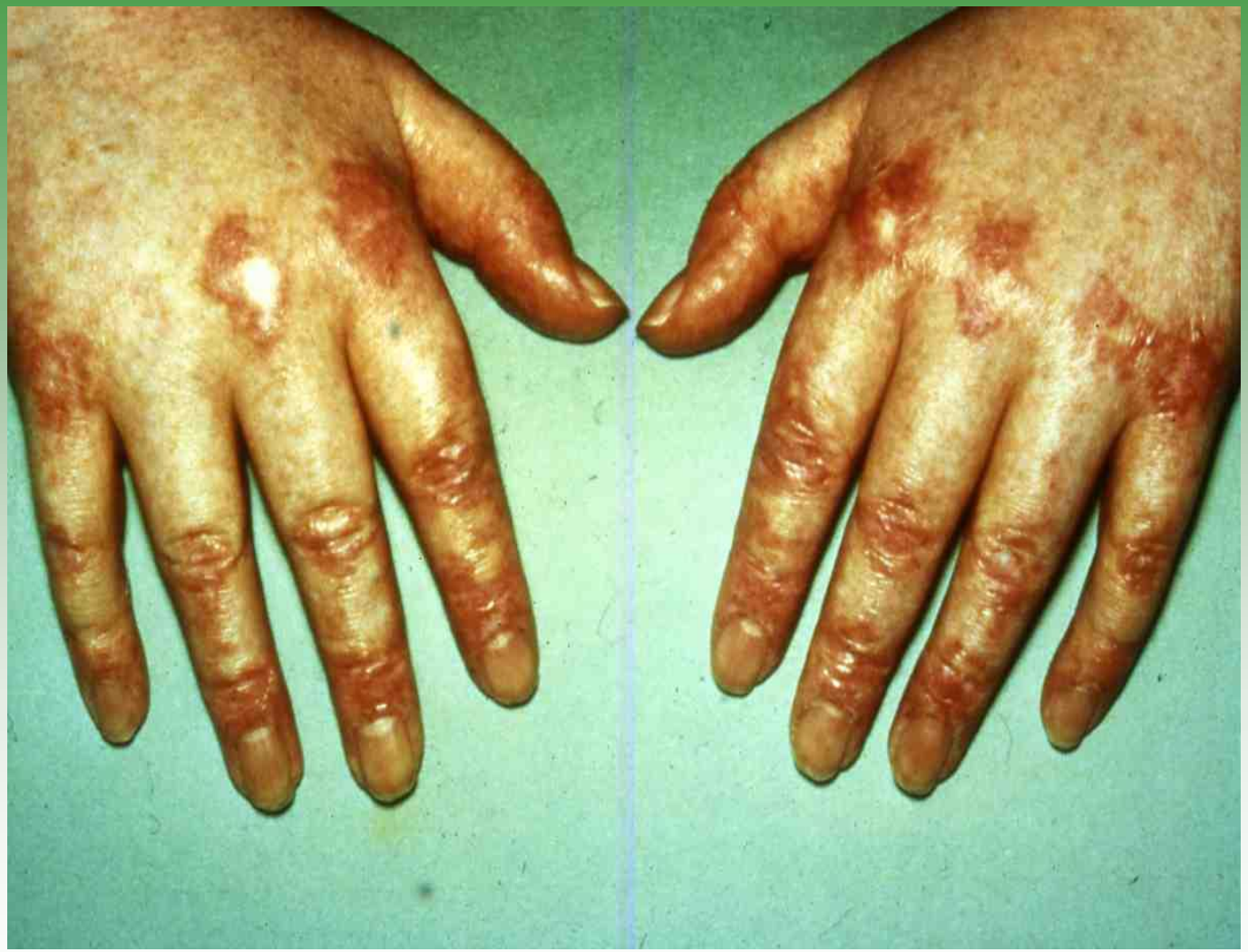
JDM – Treatment (cont.)

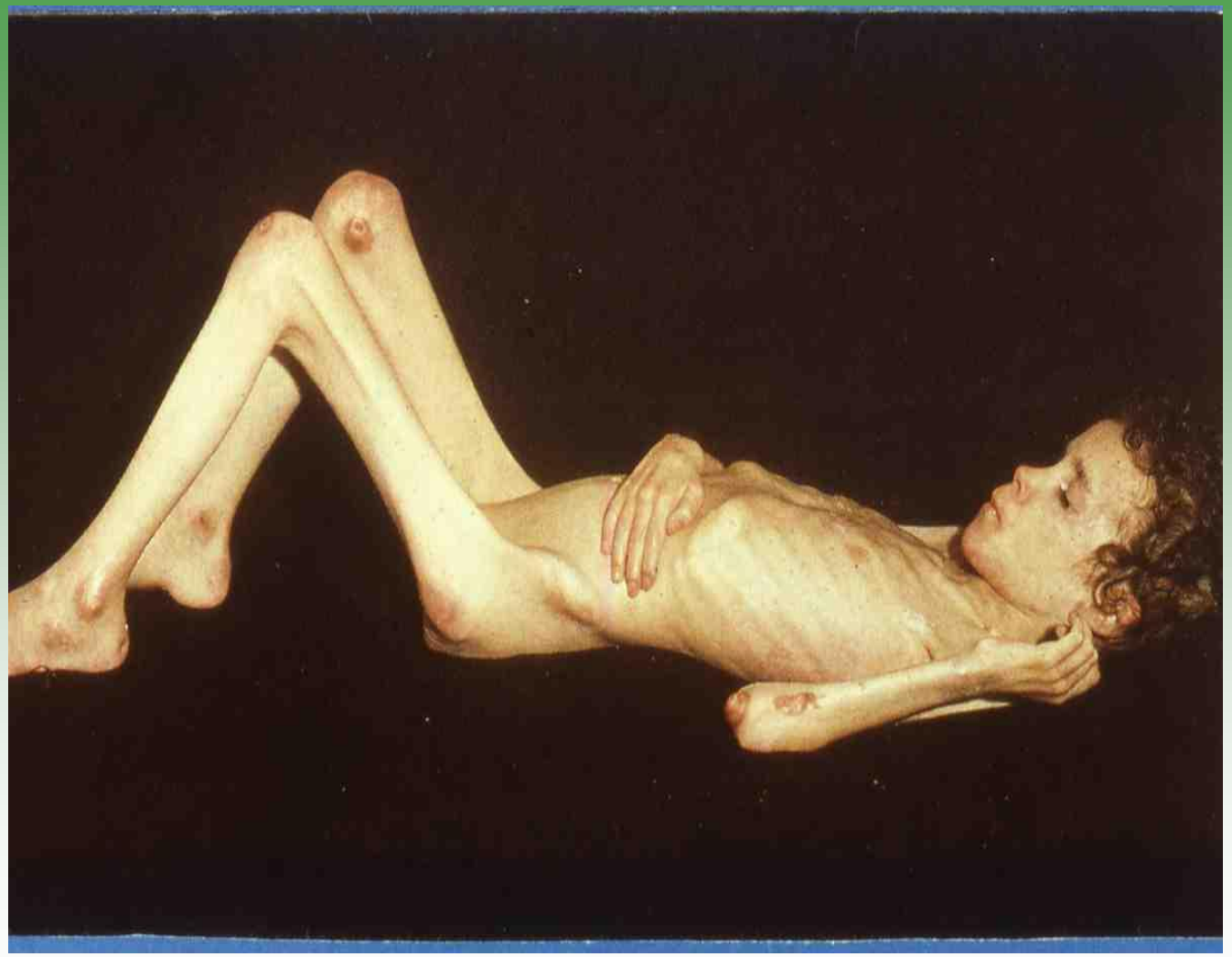
Other aspects of care:

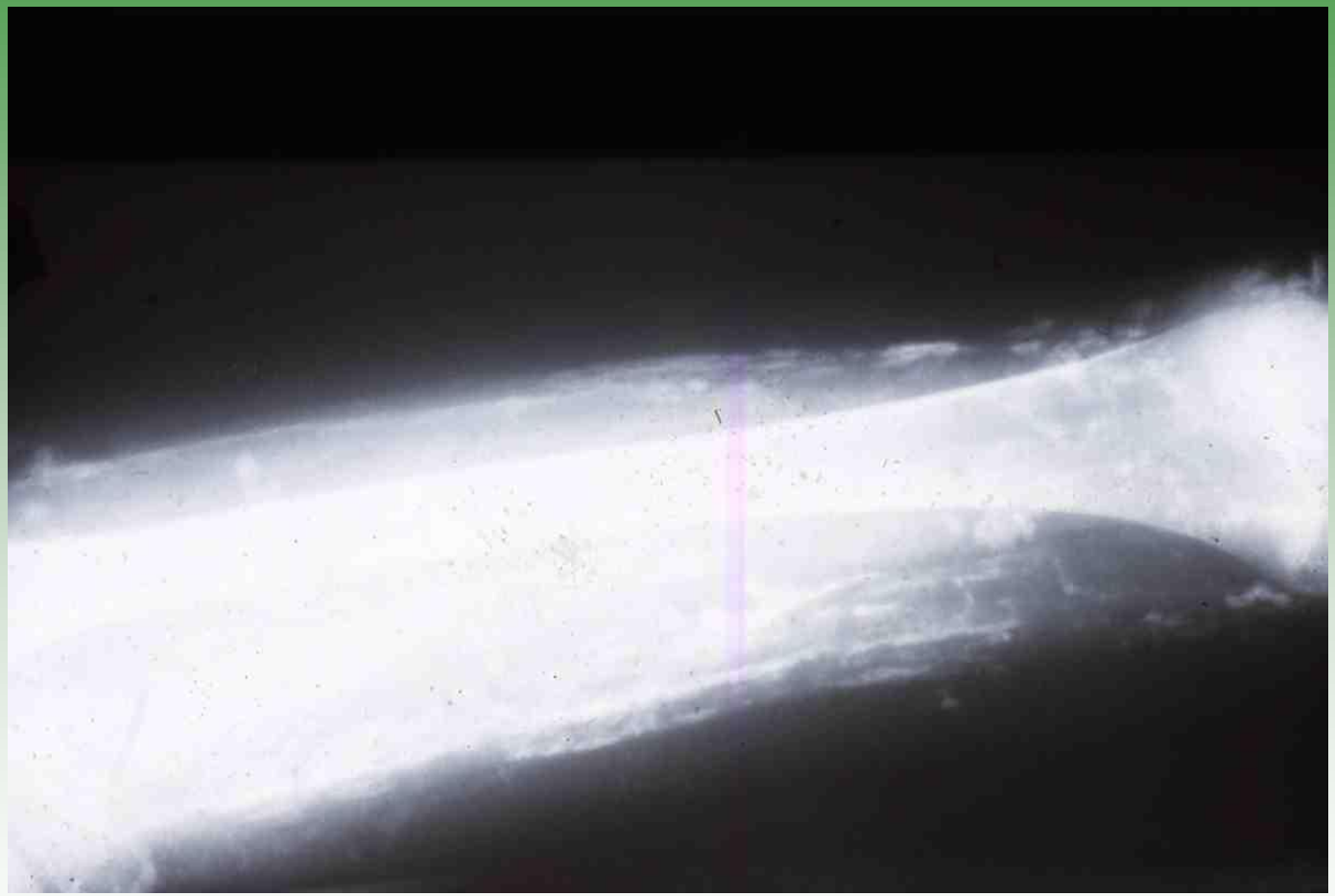
- skin protection
- physical therapy
- speech therapy
- dietetic assessment
- management of calcinosis











Henoch-Schonlin Purpura (HSP)

- **Small-vessel vasculitis**
- **Benign self-limiting disorder**
- **Deposition of IgA, C3, immune complex in small vessels HSP and IgA nephropathy (both have ↑ IgA and identical findings on renal biopsy)**
- **Diagnosis is clinical**
- **Laboratory investigations to exclude other causes**

HENOCH-SCHONLEIN PURPURA

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

HSP - Treatment

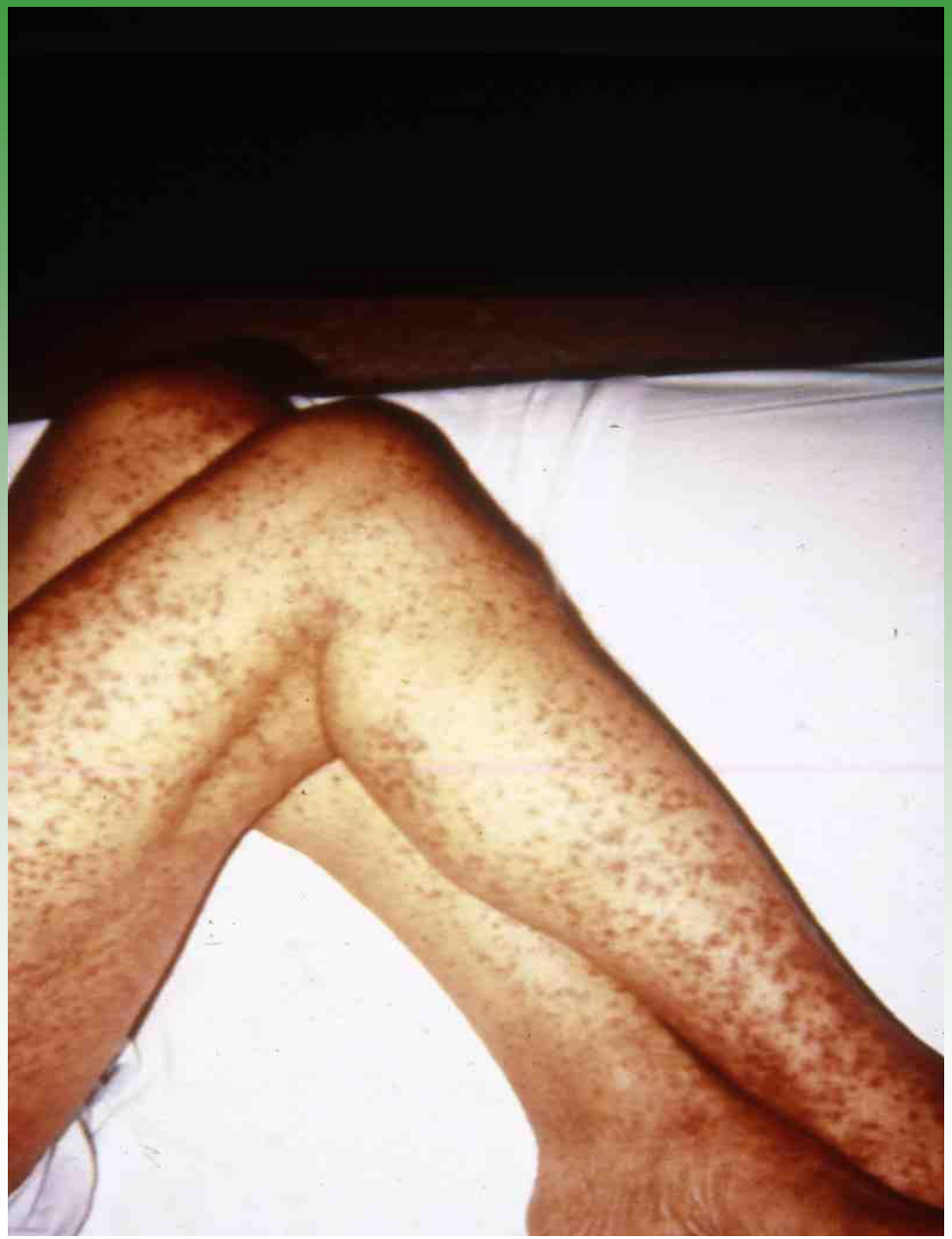
- **Resolve spontaneously**
- **Treatment according to system involved**

MSK - Ibuprofen

GIT - Corticosteroids (Prednisolone, IV MP)

Renal - Cyclophosphamide (Cytoxan)

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



Kawasaki's Disease (KD)

- **Small vessel vasculitis (coronary artery)**
- **Unknown etiology**
- **Children under 5 years**
- **Diagnoses is clinical**
- **Laboratory studies to exclude other causes and look for complications**

  (CBC – CRP – ECHO)

KAWASAKI'S DISEASE

Fever	95%
Conjuctival congestion	90%
Exanthema	90%
Oral mucosa involvement	90%
Desquamation	90%
Cervical lymphadenopathy	75%

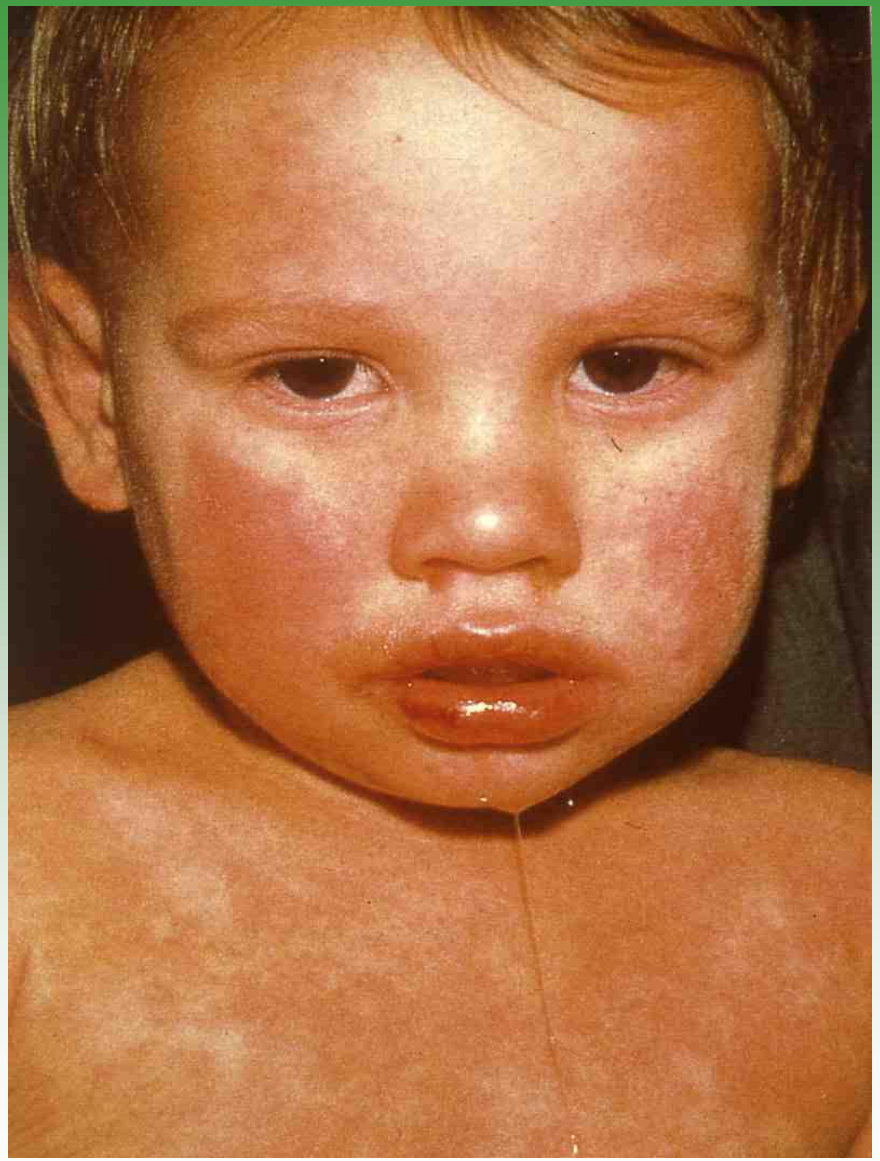
Diagnostic Criteria

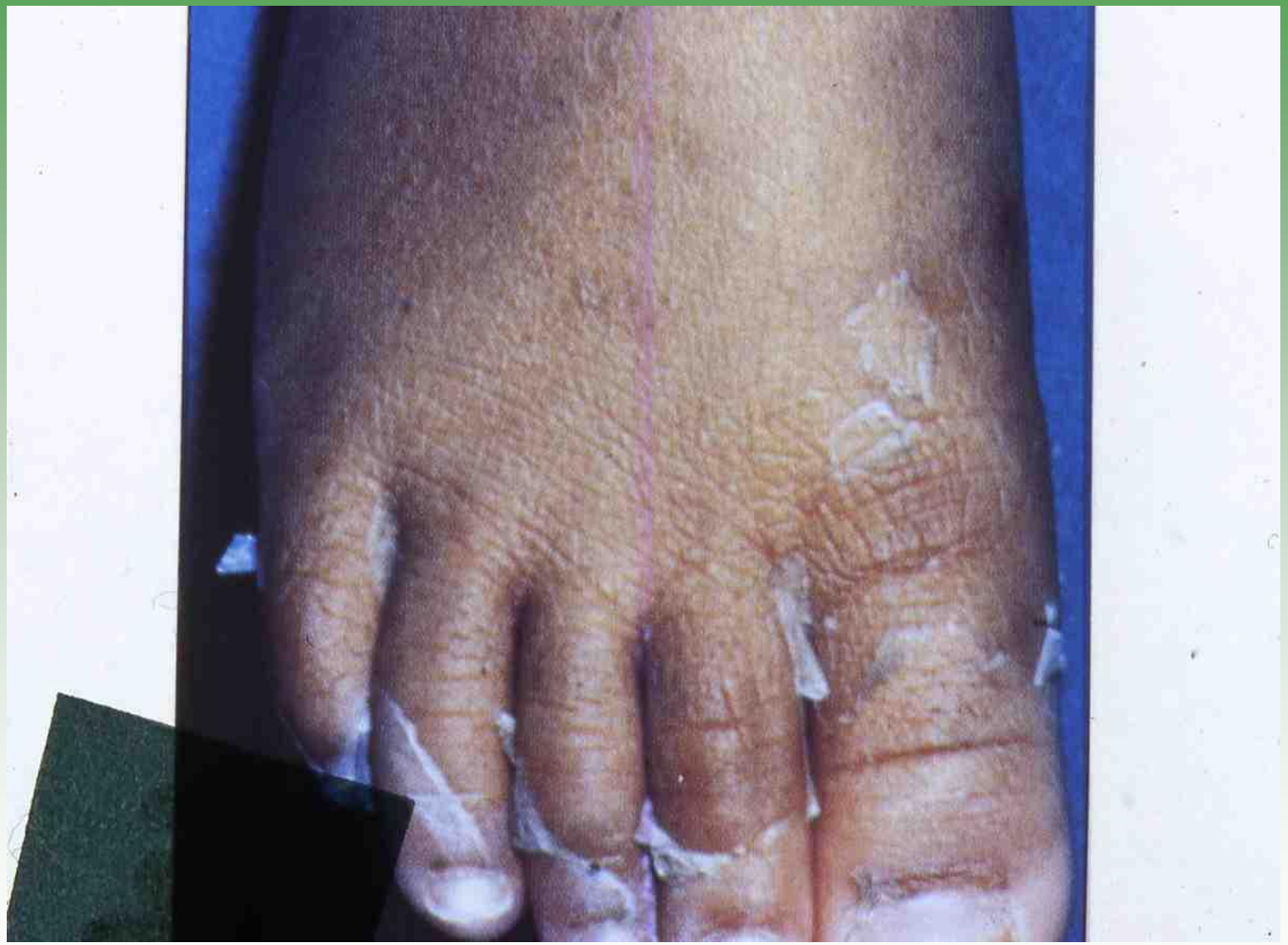
For a definitive diagnosis the patients must have **5** of the following **6** criteria:

1. **Splking fever for at least 5 days**
2. **Bilateral conjunctival injection**
3. **One orpharyngeal sign**
   - Diffuse oropharyngeal Erythema
   - Strawberry tongue
   - Redness, dryness, and fissures of lips
4. **Polymorphous erythematous rash**
5. **cervical lymphadenopathy**
6. **One or more of the following signs**
   - Indurative edema of hands and feet
   - Erythema of palms and sole
   - Desquamation of fingers and toes
   - About 2 weeks after onset
   - Transverse grooves in nails
   - 2 or 3 months after onset

Treatment of Kawasaki Disease

- High dose aspirin
- Low dose aspirin
- High dose IV immunoglobulin
- ± 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

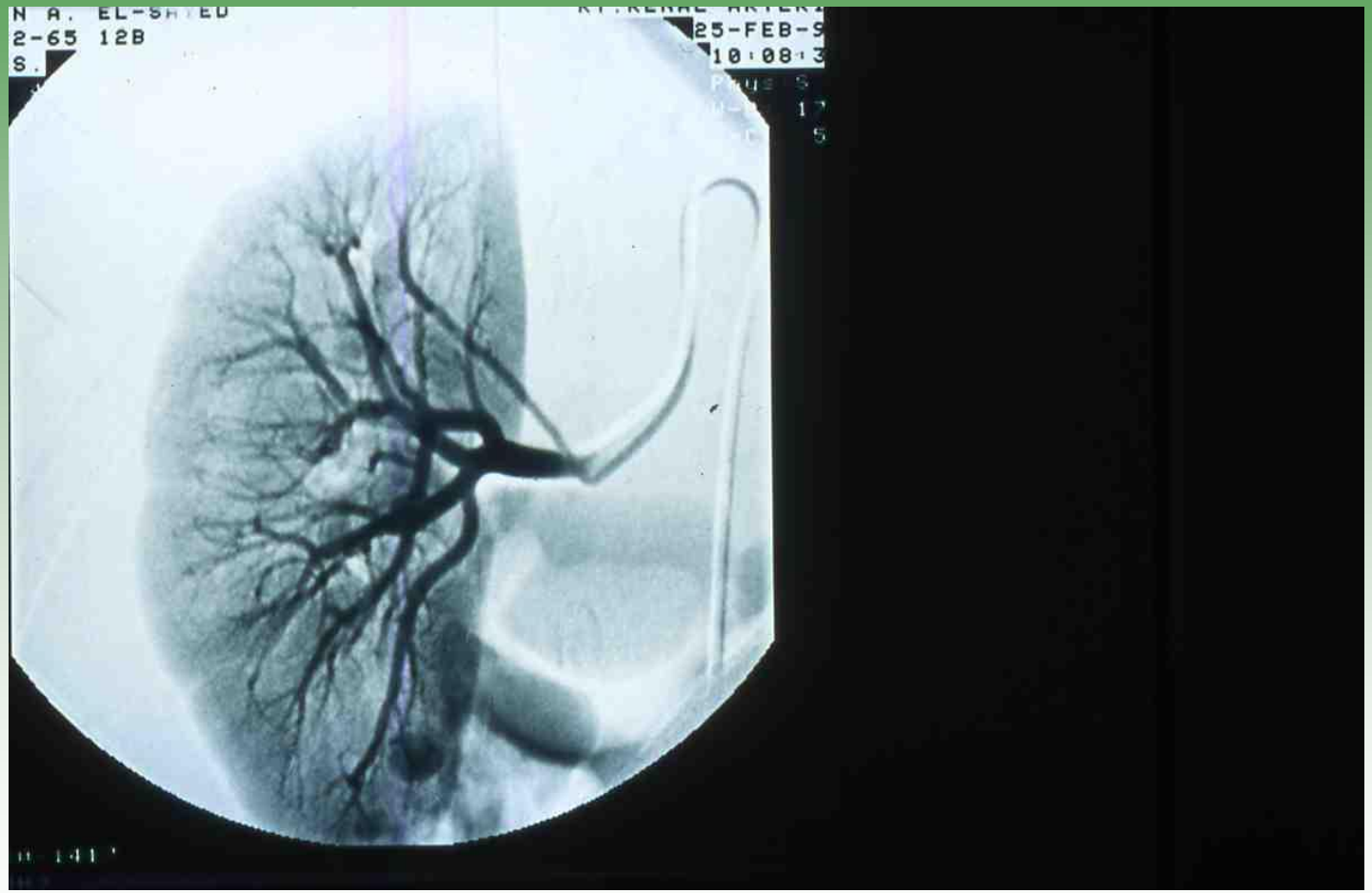
Familial clustering

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
- Reiters disease
- (Behçets Syndrome)
- Reactive arthritis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Thank You