- Pain or limb dysfunction - **various organ system** involvement

- **Musculoskeletal pain** - common in chidren (4-30% otherwise healthy kids)

- **Arthritis** - small proportion: 108,5/100000 children under 16 (**transitory** conditions in most cases, duration shorter than 6 weeks)

- **Chronic (idiopathic) arthritis** (over 6 weeks duration):

  5,3 - 19,6/100000 dětí

Differencial diagnosis of musculoskeletal pain

Avascular necrosis and degenerative conditions:
Perthes,osteochondritis,chondromalacia patellae,hypermobility

Reactive arthritis: post:-streptococcal,-enteritic, -viral

Trauma, non-accidental injury

Hematological diseases: hemoblastosis,hemofília,lymphoma

Rachitis, other metabolic disorders and endocrinopathies

Infections: septic arthritis, osteomyelitis

Tumors: cartilage, bone, muscle, synovium, blood vessels

Idiopathic pain: localised, generalised

Systemic connective tissue diseases: SLE, vasculitis, dermatomyositis, scleroderma

Rheumatic disease manifestation in orofacial region

- **Juvenile arthritis**
  - TMJ involvement
- **Sytemic connective tissue diseases**
  - Mucosal changes
  - Salivary gland involvement
  - Dentition disorders
  - Neuropathies/palsy
- **Other inflammatory dieseases**
  - Aseptic osteomyelitis (CRMO)

Rheumatic diseases in children- epidemiology

	Incidence	Prevalence
JIA	**5,3 - 19,6**	**minim. 1/1000**
JSLE	0,5 - 0,6	10% new dg.
JDM/JPM	0,2 - 0,5 (0,15)	
SS	0,45 - 1,2 total.	?
	1,0-9,0% do 20 let	
HSP	13,5	?
KD	5,95 - 7,6 do 5 let	?

JUVENILE IDIOPATHIC ARTHRITIS:

Diagnosis per exclusionem

Classification of Juvenile Idiopathic Arthritis

2nd revision-Edmonton 2001

Definition: Arthritis of unknown origin affecting a person younger than 16 years of age of minimum 6 weeks duration in at least one joint

JIA classification

Clinical features during first 6 months

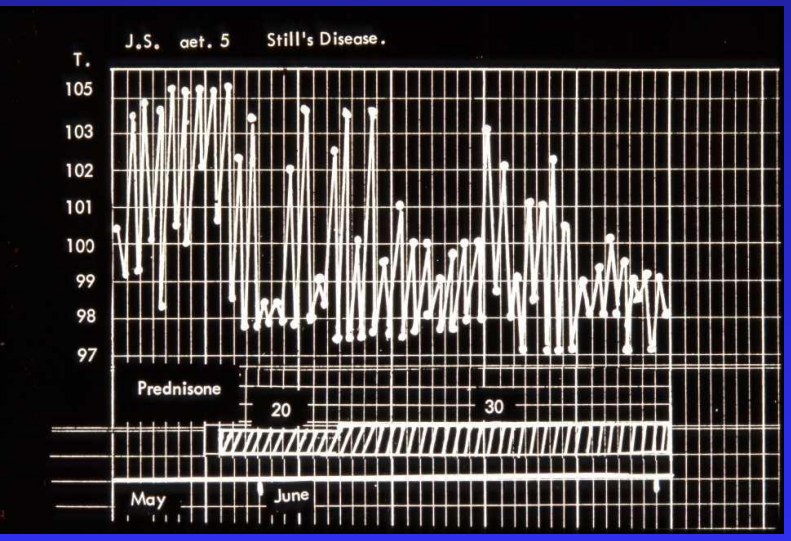
Criteria, descriptors, exclusions

1. Systemic arthritis
2. Polyarthritis IgM RF-
3. Polyarthritis IgM RF+
4. Oligoarthritis: persistent, extended
5. Arthritis with enthesitis
6. Psoriatic arthritis
7. Other arthritis

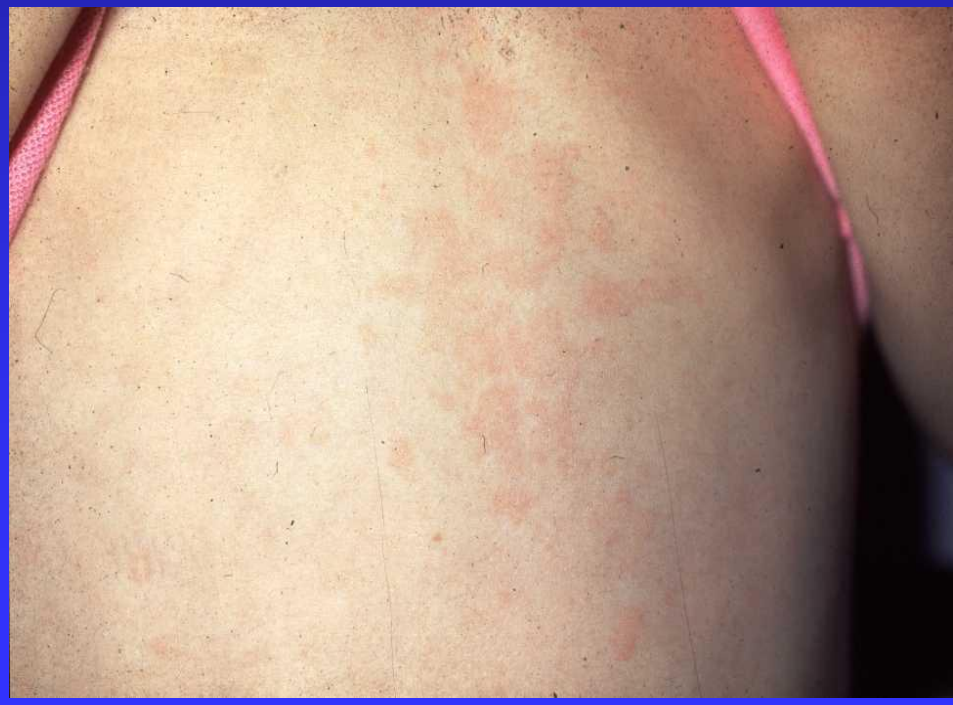
Definition and classification: sJIA

Petty et al, 1998

- **Arthritis** with or preceded by daily **fever** of at least 2 weeks' duration (documented as quotidian for minimum 3 days) accompanied with one or more of:

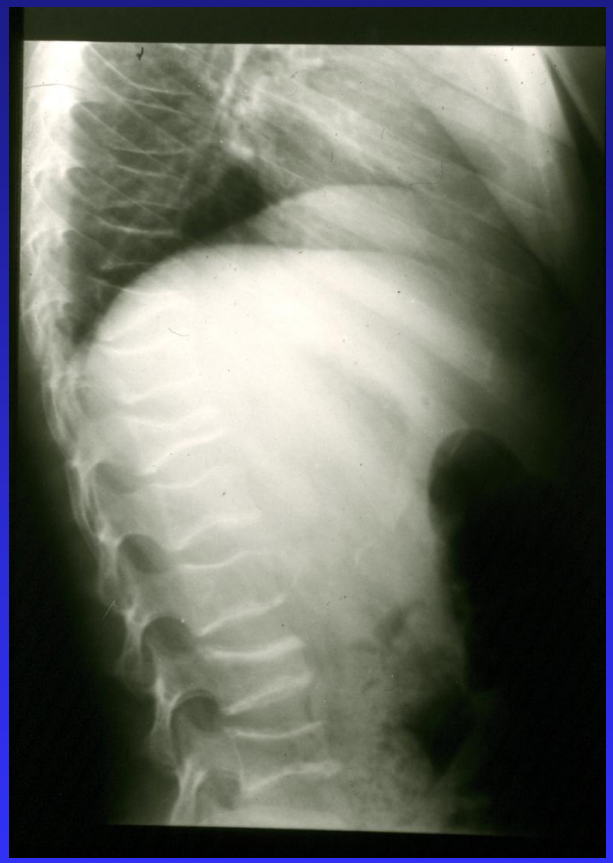


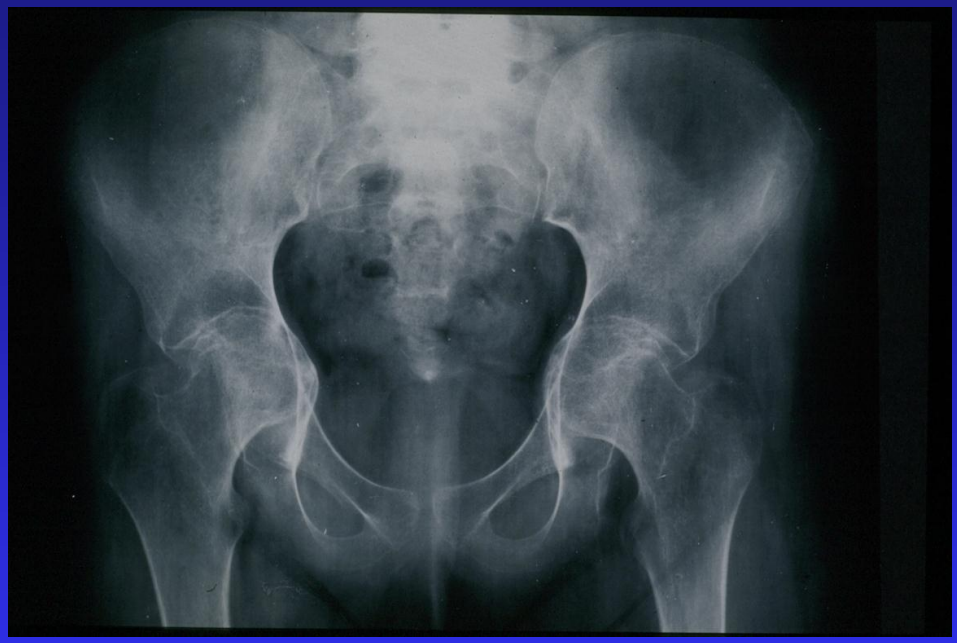
-Evanescent, non-fixed erythematous **rash**



- Generalised **LNpathy**
- **Hepato or splenomegaly**
- **Serositis**

Complications: osteoporosis





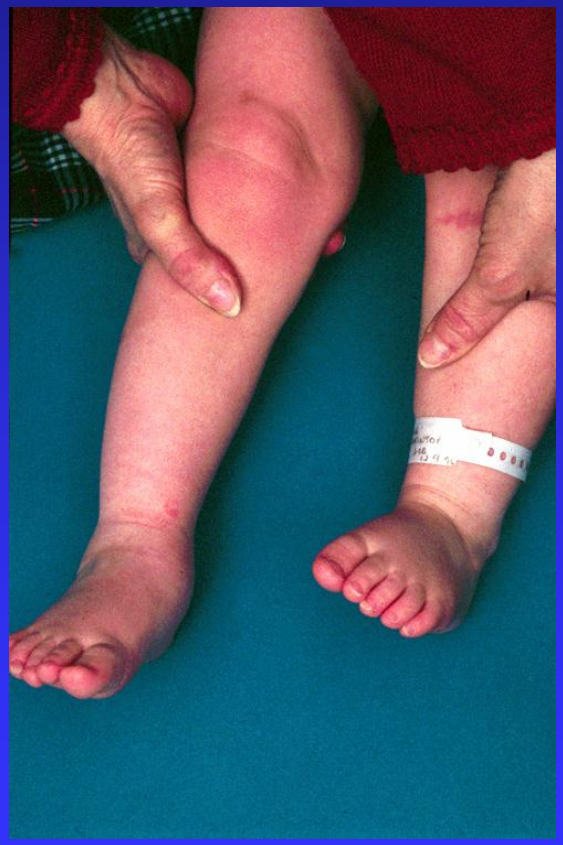
Ca and Vitamin D3 supplementation

- calcitonin, bisphosphonates (oral alendronate, i.v. pamidronate) Noguera et al 2003, Steelman and Zeitler 2003

sJIA disease course and outcome

- 3 **clinical subtypes**
  - Monocyclic (11%)
  - Intermittent (34%)
  - Persistent (55%) Lomater et al 2000
- **Active disease** after >10 years follow-up in 23-58 % of sJIA patients
- 81 adults with JIA, med 21 years duration: 39% active disease, significantly higher **HAQ** and lower **SF-36** than controls Foster et al 2003
- **Mortality** all JRA: <0.29% (0.08% standardised death rate USA), 2/3 sJRA Wallace and Levinson 1991

Oligoarticular JIA



- No affected joints < 5
- Frequency: 60-75% all JIA
- Subgroups: Type I- early (toddlers and preschool)
- Type II- late (prepubertal)

Clinical picture

- Onset 2-3 years, girls predominate
- Knee, ankle
- Limp, stiffness, swelling, flexion deformity
- Lab often normal
- Fever and general symptoms – careful differential dg
  - Malignancies (leukemia, neuroblastoma)
  - Infections, other systemic diseases
- CAVE: ANA
- eyes
- psoriasis
- local growth disturbancies

Prognosis

- Excellent if treated early and appropriately
- Chronic uveitis – limits favourable prognosis

- Therapy:
  - Intraarticular corticosteroid, PT, NSA
  - (MTX)

Polyarthritis RF negative

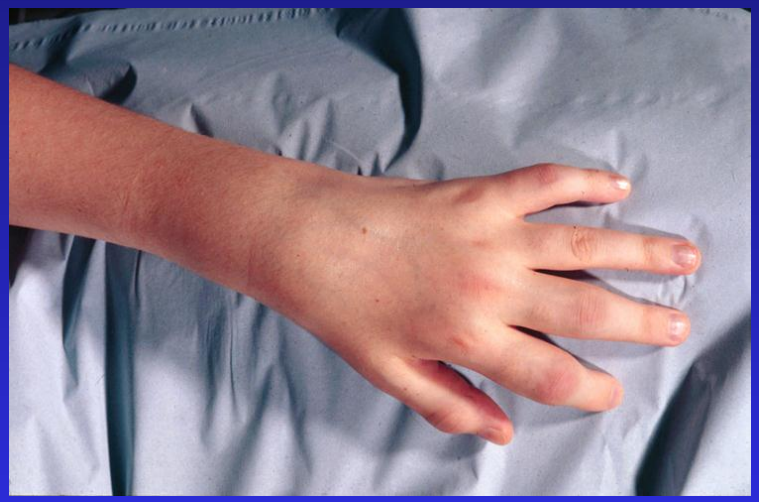
More than 4 affected joints during initial 6 mo

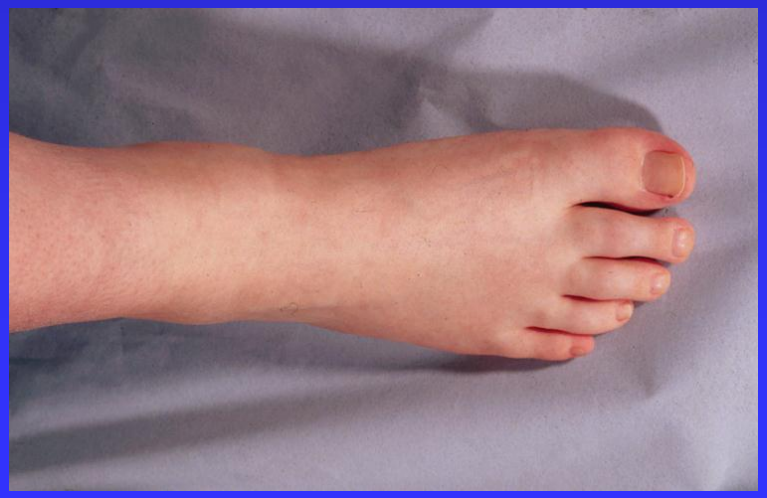
↑ nonspecific inflammation

Often foot, wrist, hip involvement – negative prognostic factor

- Therapy: Methotrexate 15 mg/m2, NSA, locaů CS, unusually systemic CS

Polyarthritis RF positive

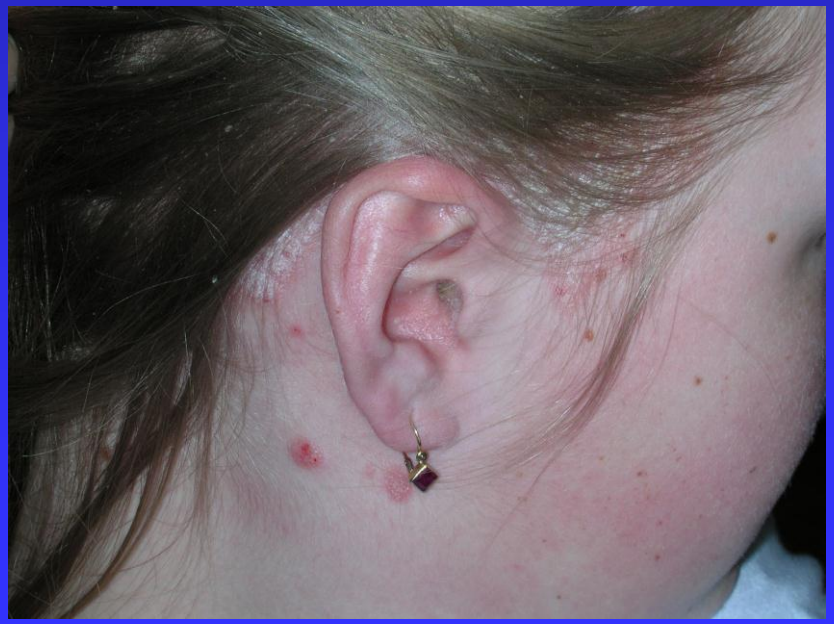


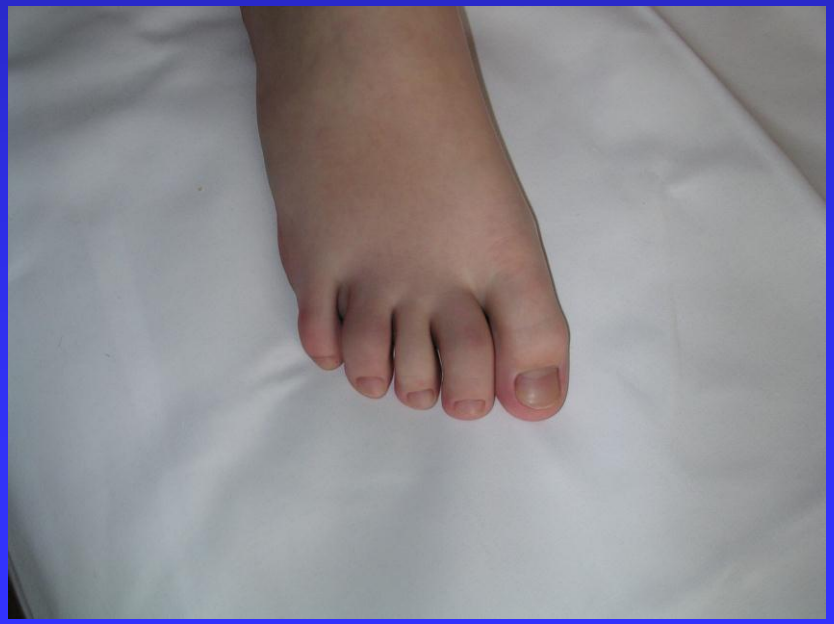


- 1-2% JIA
- Often adolescent girls
- Similar to adult RA
- Rapidly progressive destruction, symmetrical small joint disease

Psoriatic arthritis

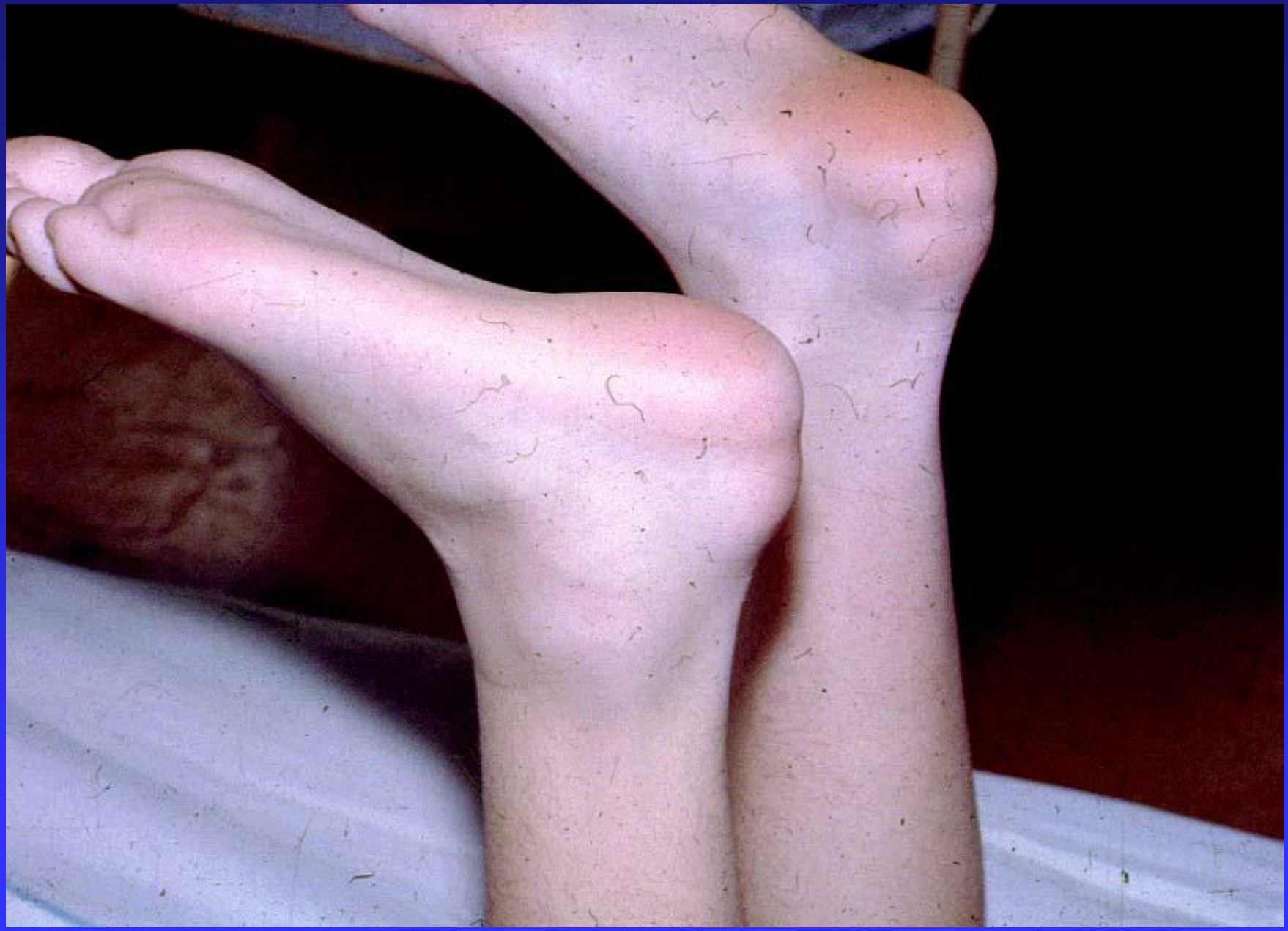
- Artritida + psoriasis or arthritis + nail pitting/psoriasis in 1st degree relative/dactylitis (2 of 3)

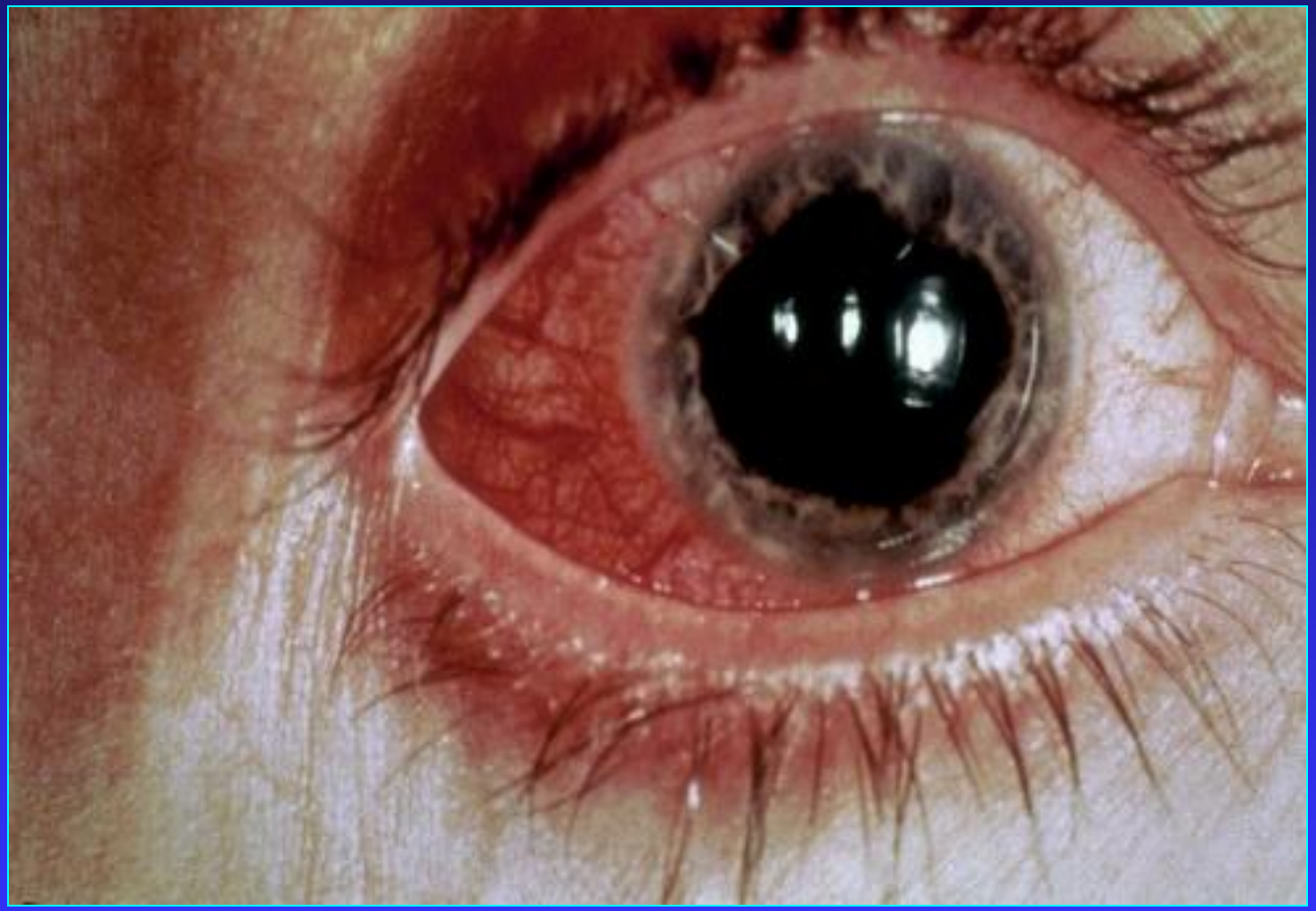




Arthritis with enthesitis

- „Enthesis“ = insertion of tendon or ligament into bone
- Plantar, Achilles tendon, knee, pelvis
- Lower extremities, SI
- Often HLA B-27, enteropathogen-triggered
- Exclude IBD
- Therapy: local CS, Salazopyrin, (MTX, Enbrel)





Acute anterior uveitis (iridocyclitis)

TMJ involvement in JIA

- Frequent in JIA, mainly polyarticular type
- Often asymmetrical, often asymptomatic
- Functional limitation
  - Mandible deviation due to muscle spasm at involved side
  - Mouth opening reduction
- Growth changes
  - „Bird-like“ facial appearance
- Plastic surgery available after growth ends

Juvenile idiopathic inflammatory myopathies

Epidemiology

	Incidence	Prevalence
JIA	5,3 - 19,6	minim. 1/1000
JSLE	0,5 - 0,6	10% nových dg.
JDM/JPM	0,2 - 0,5 (0,15)	
SS	0,45 - 1,2 celk.	?
	1,0-9,0% do 20 let	
HSP	13,5	?
KD	5,95 - 7,6 do 5 let	?

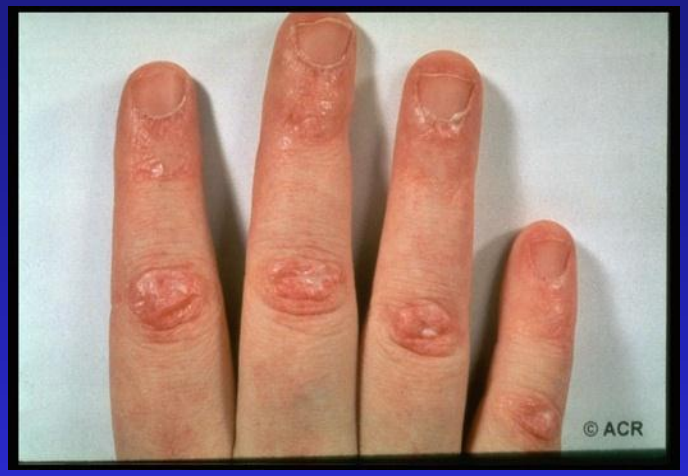
Classification
(Rider a Miller, 1997)

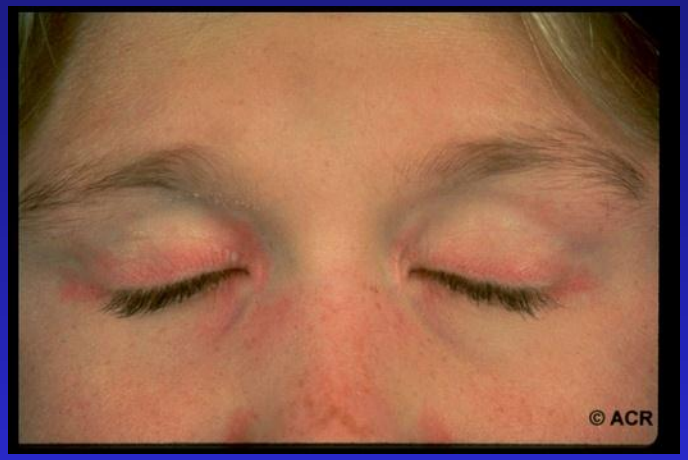
IIM = Idiopathic Inflammatory Myopathies:
Chronic striated muscle inflammation of unknown origin

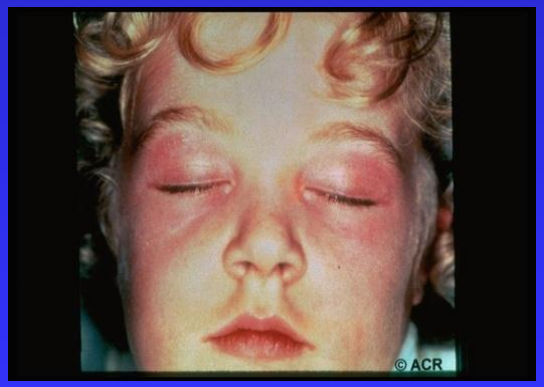
JDM: 85% JPM: 8%

DG criteria JDM/JPM

(Bohan a Peter, 1975)



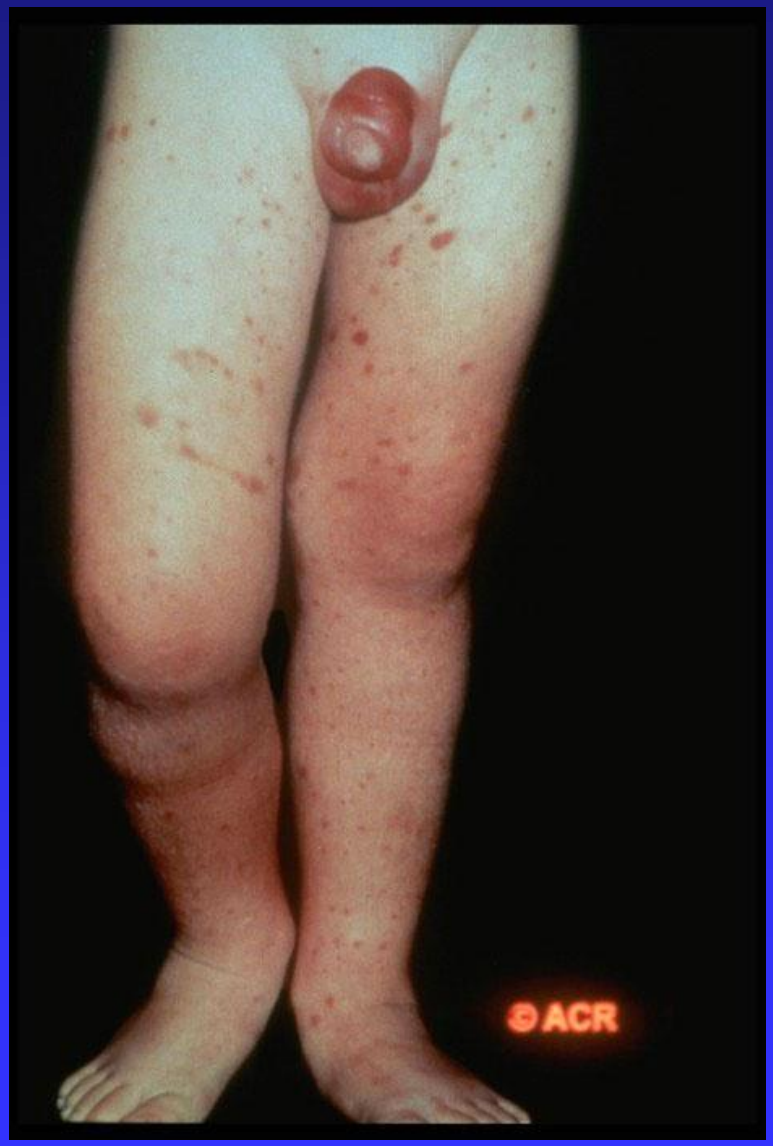


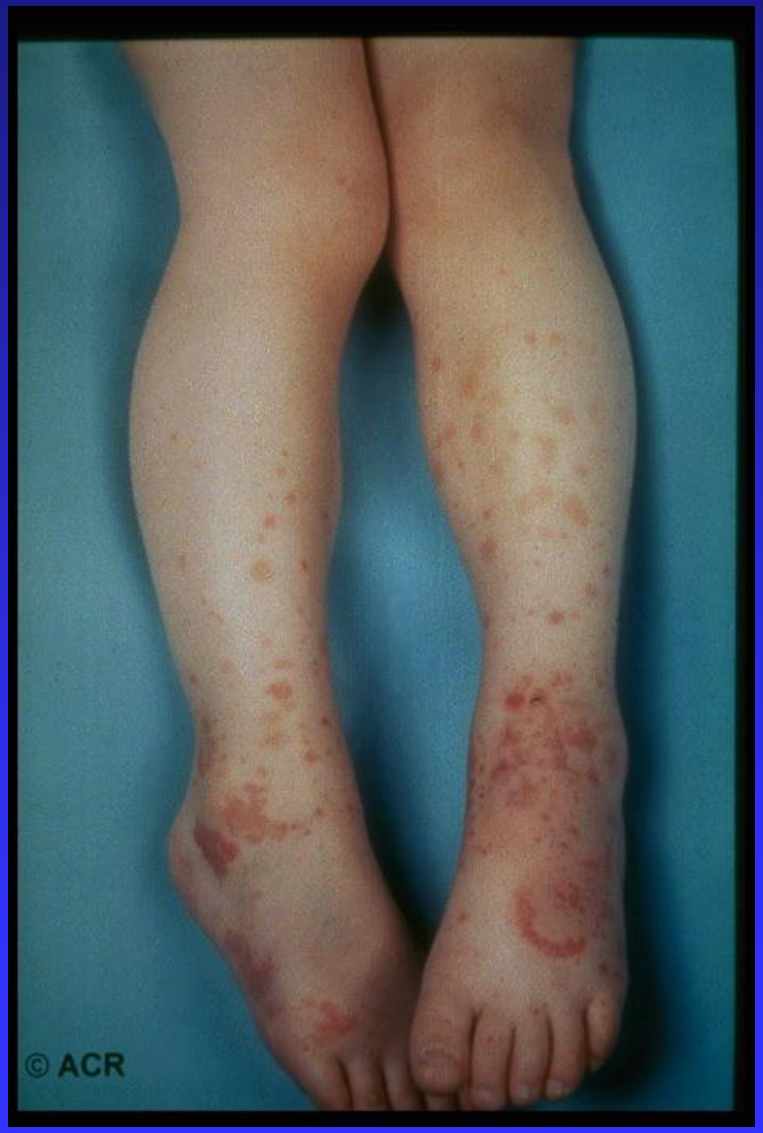


	JDM	JPM
Charakteristické kožní projevy	+	-
Symetrická proximální svalová slabost *	+	+
Svalové enzymy	+	+
EMG změny	+	+
Histopatologie **	+	+

* za vyloučení jiných onemocnění ** odlišná u JDM a JPM

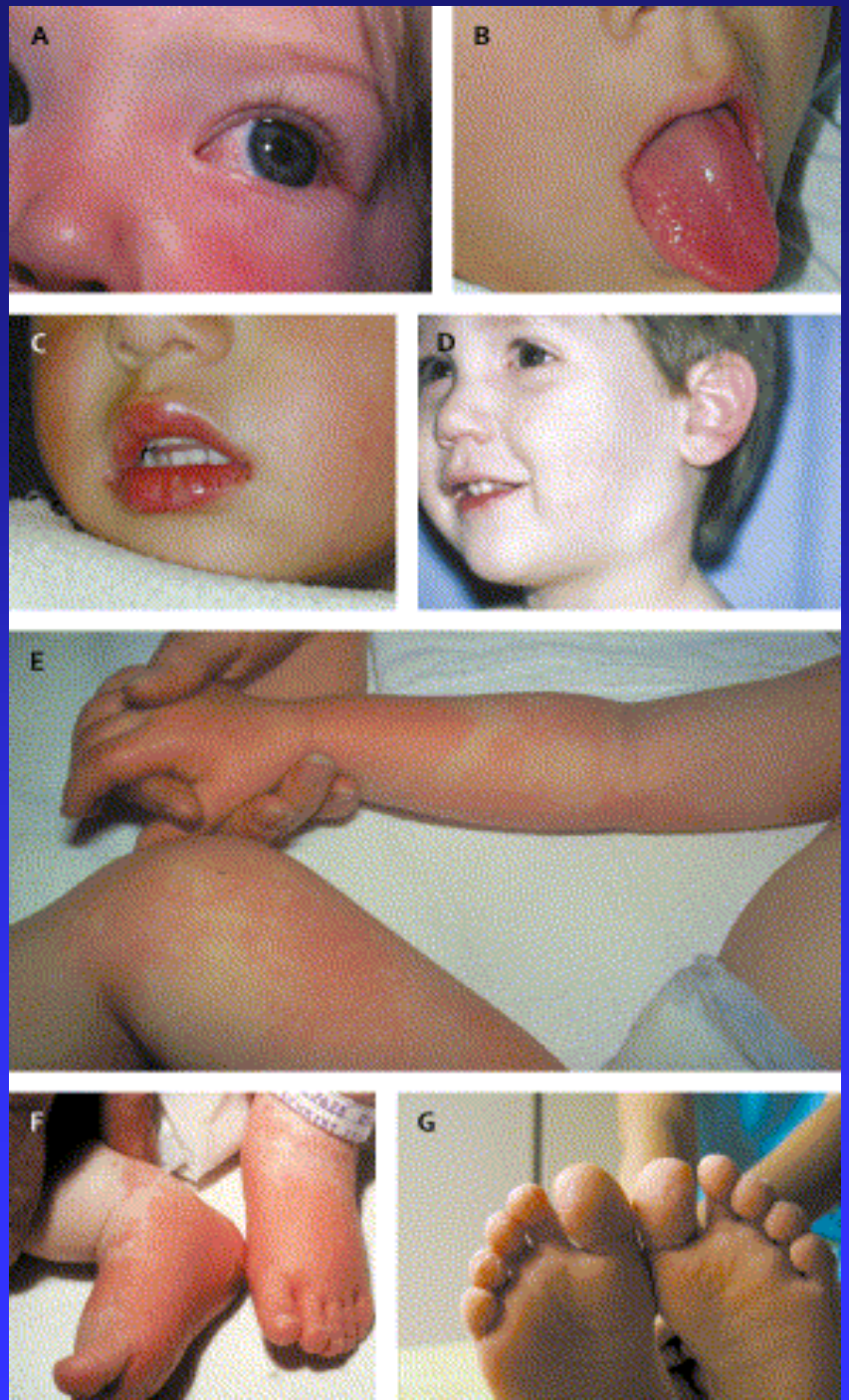
Henoch-Schönlein purpura

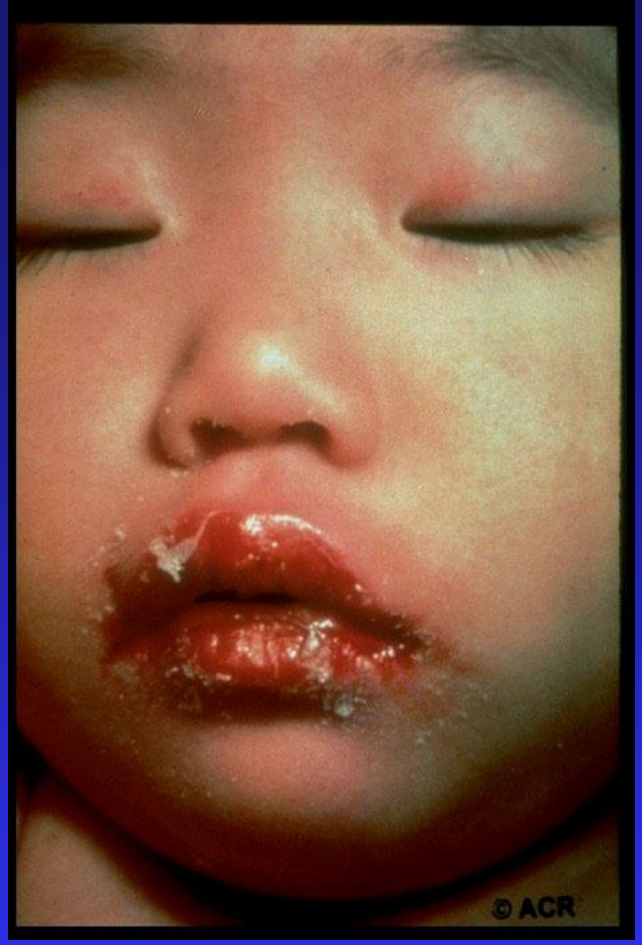




Kawasaki disease

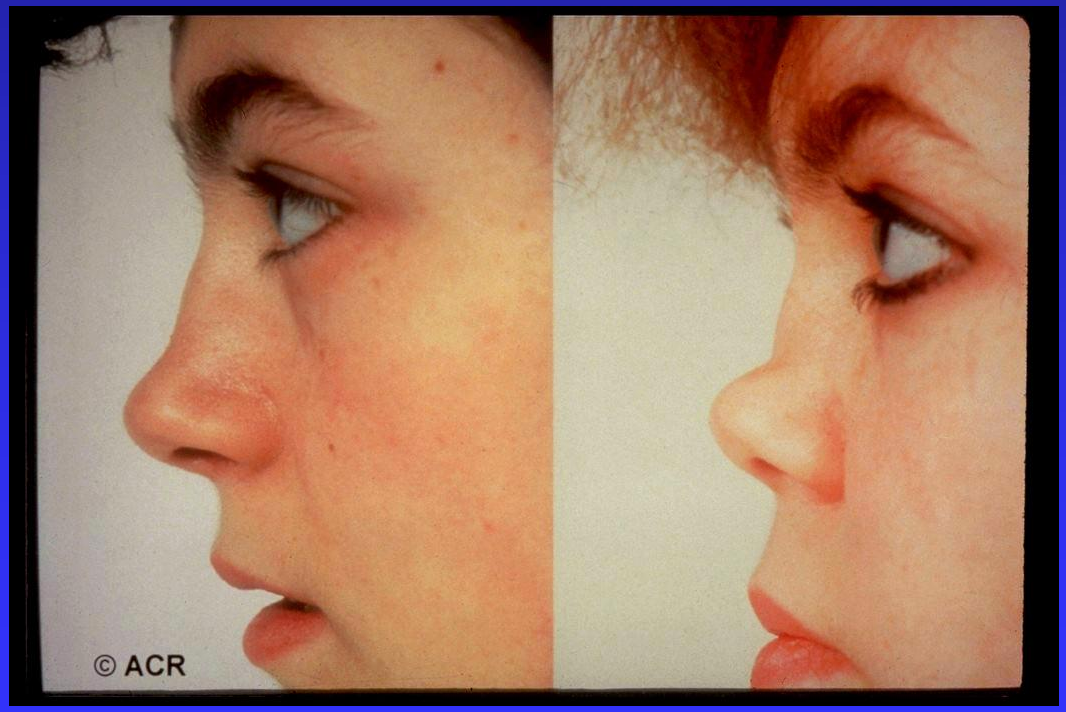
- Fever (100%)
  - >5 days
- A: Conjunctivitis (85%)
  - Bilat., bulbar, non-exsudative
- B,C: Mucosal changes (90%)
  - Red lips, strawberry tongue, oropharyngeal erythema
- D: Lymphadenopathy(70%)
  - Cervical, acute, non-suppurative, >1,5 cm
- E: Rash (80%)
  - Polymorphous
- F,G: Extremity changes.(70%)
  - Erythema palms and soles, edema, peeling





Kawasaki disease

Wegener´s granulomatosis



SLE - DG CRITERIA (ACR)

- Malar rash
- Discoid lupus
- Mucocutaneous ulcerations
- Non-erosive arthritis
- Nephritis
- Encephalopathy
- Pleuritis or pericarditis
- Cytopenia
- Positive immune serology
- Positive ANA