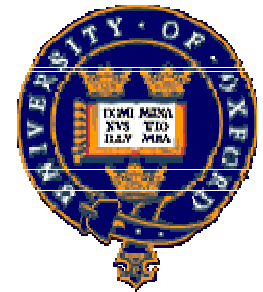


# Vasculitis in children and adolescents



Dr Raashid Luqmani  
Consultant Rheumatologist  
Nuffield Orthopaedic Centre, Oxford  
and University of Oxford



# 8 year old girl

- Red rash on shins
- Persistent
- Ulcerating
- Necrotic
- Resistant to all topical therapy
- 2 years later referred to rheumatology

# 10 year old girl

- Distressed parents +++
- Silent withdrawn child
- Black necrotic rash on both shins
- 3 cm x 5 cm
- Joint pains...

# Further information

- Nose bleeds for 18 months
- Nasal crusts
- Nasal discharge

# Examination

- Urine normal
- Chest x ray normal
- Neurological examination normal
- No weight loss
- No fever

# Examination

- Tender knees – no fluid
- Necrotic ulcerating lesions on shins
- ENT opinion

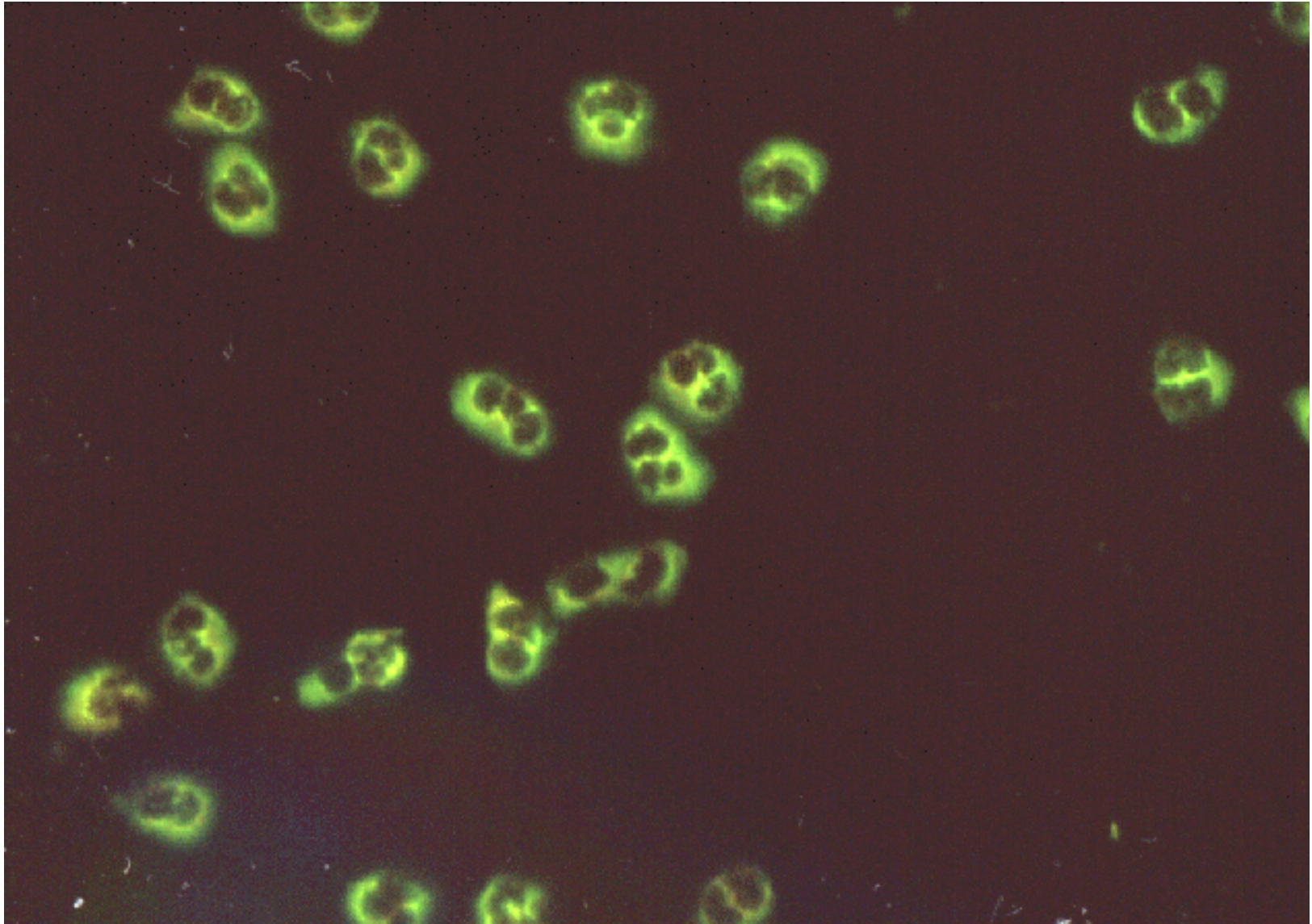
**Diagnosis ?**

# Diagnosis ?

- Non renal Wegener's granulomatosis



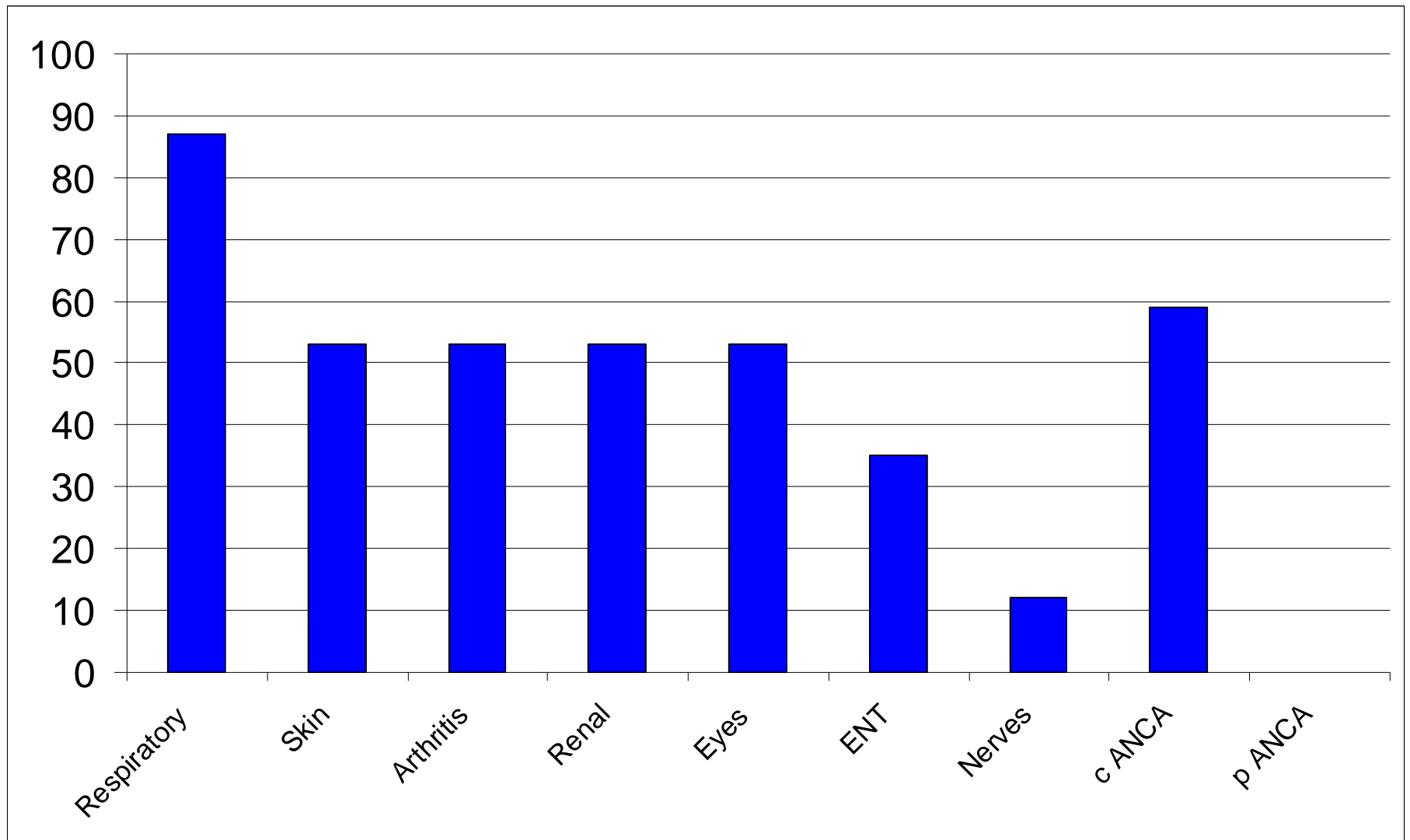
# cANCA



# Wegener's granulomatosis in childhood: presentation with cutaneous manifestations

- Acneiform lesions
- Cutaneous ulcers
  - Chyu et al J Am Acad Dermatol. 1984 10:341-6
  - Brazzelli et al Pediatr Dermatol. 1999 16:277-80

## Clinical features in paediatric Wegener's n=17



Median age 6 years (2 weeks-14 years)

# Wegener's granulomatosis in childhood and adolescence vs adults

Frosch M, Foell D. Eur J Pediatr. 2004 163:425-34

- More
  - subglottic stenosis
  - nasal deformity
- Less
  - Side effects
  - cancer
- Responds to cyclophosphamide and steroids
- Relapses/risk of chronic damage at all ages

# Treatment

- Methotrexate increasing to 20mg per week
- Folic acid
- Prednisolone 40mg per day, reducing progressively

# Methotrexate treatment of Wegener granulomatosis in children

Gottlieb et al J Pediatr. 1996 129:604-7

- 3 cases responding to MTX

## NORAM- non renal Wegener's granulomatosis alternatively treated with methotrexate

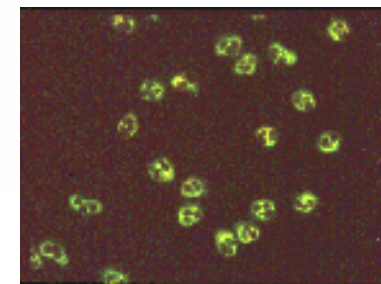
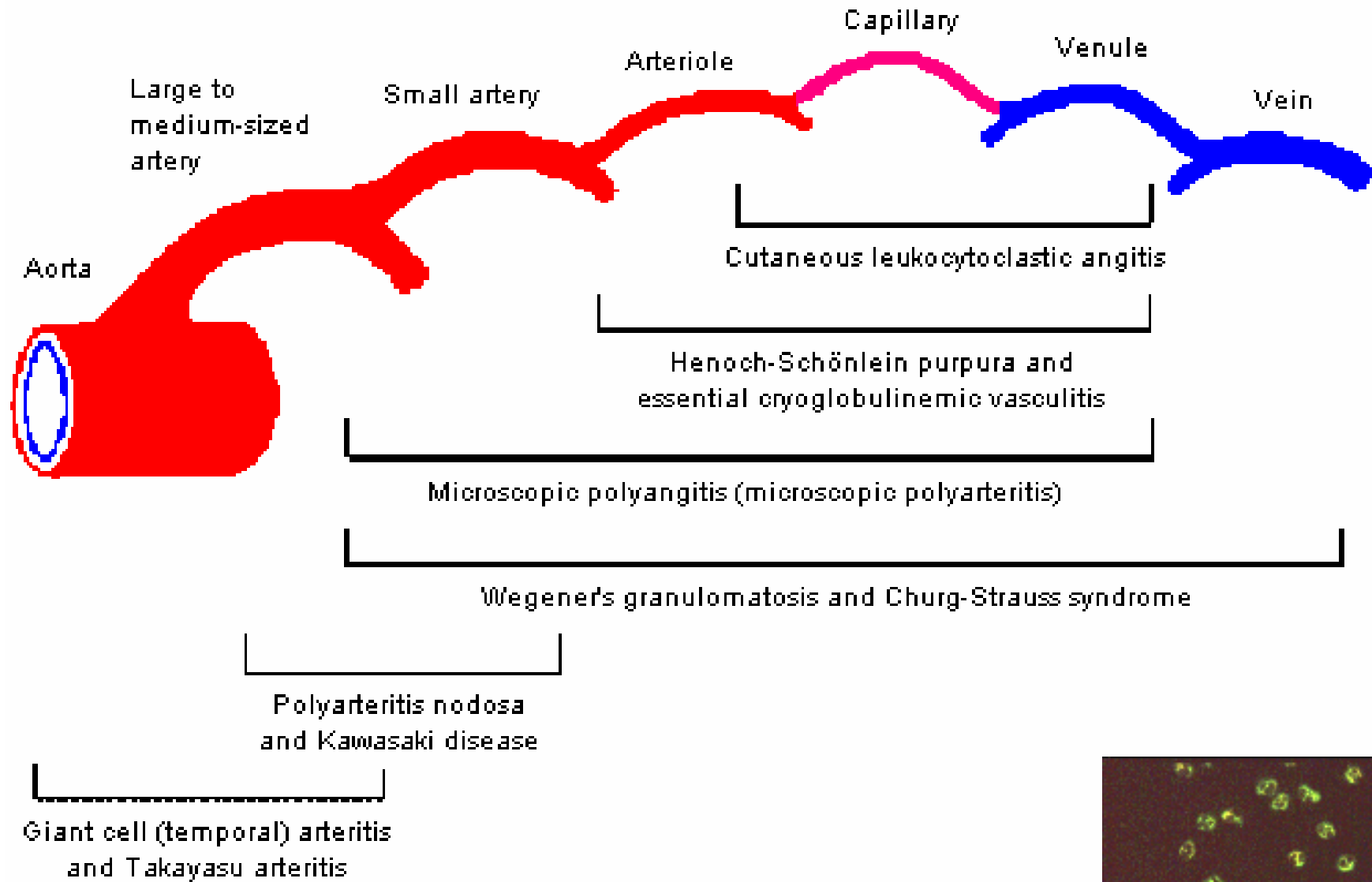
- MTX (49) vs CYC (51) in non renal WG
  - Remission
  - 3 months
  - 6 months
  - Relapse
  - Relapse (months)
  - Prolonged Rx required
  - MTX as safe as CYC
- |                  | MTX  | CYC |
|------------------|------|-----|
| 3 months         | 59%  | 65% |
| 6 months         | 83%  | 84% |
| Relapse          | 69%  | 42% |
| Relapse (months) | 13.5 | 15  |

# Treatment

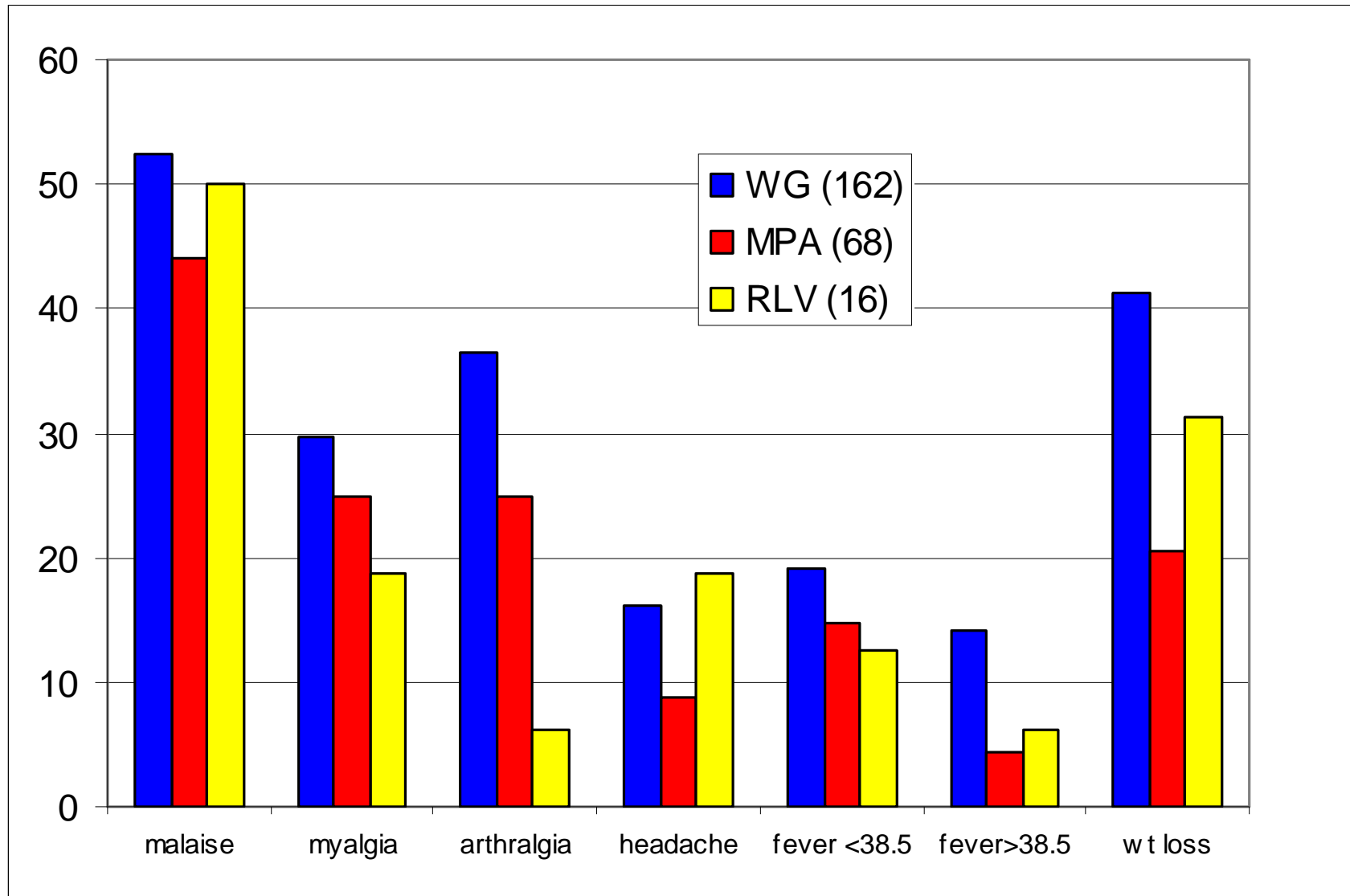
- Ulcers healed, nasal symptoms resolved
- Prednisolone eventually stopped
- Methotrexate tailed off after 3 years
- No relapse

# Follow up

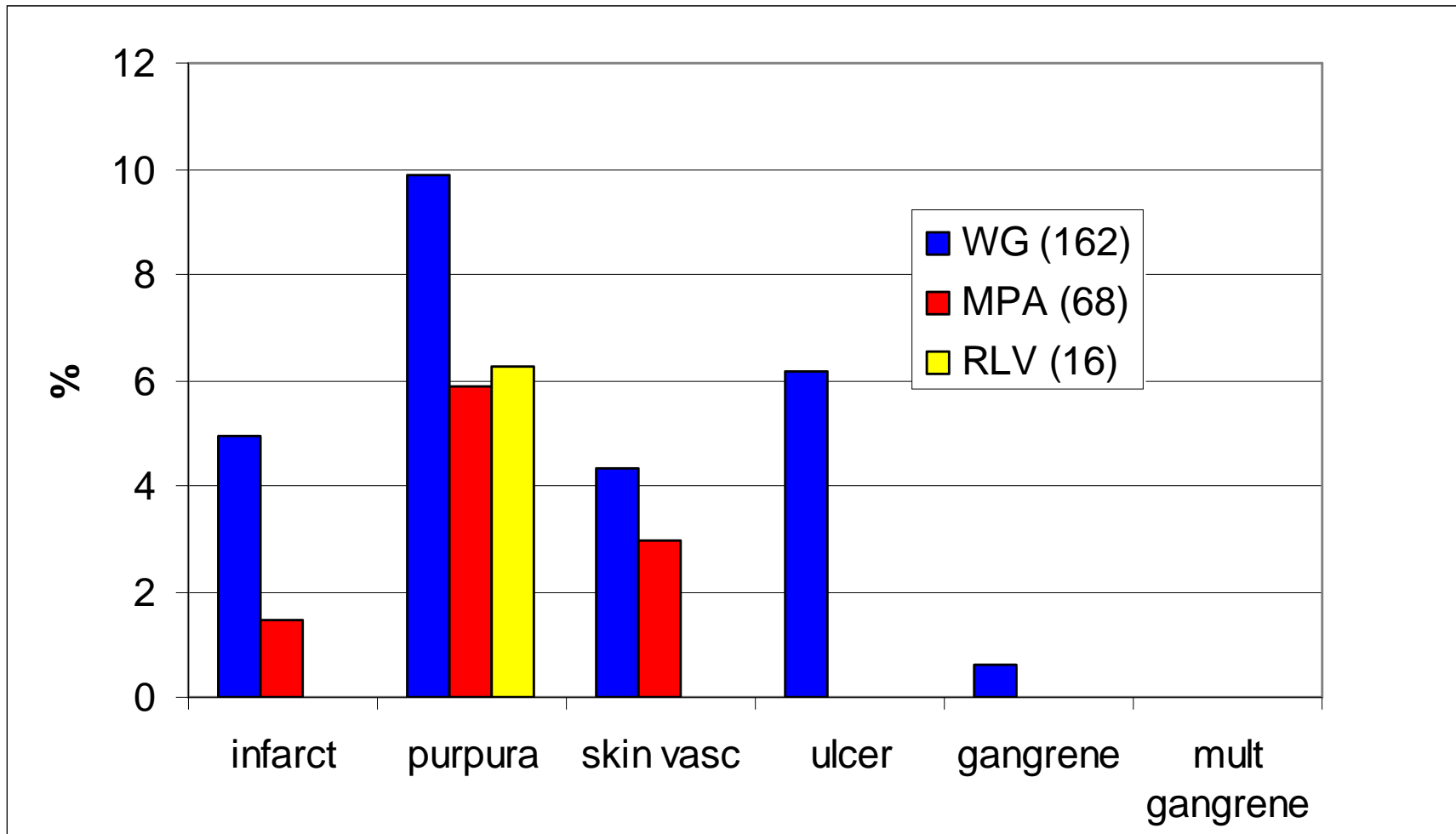
- Continued to be well



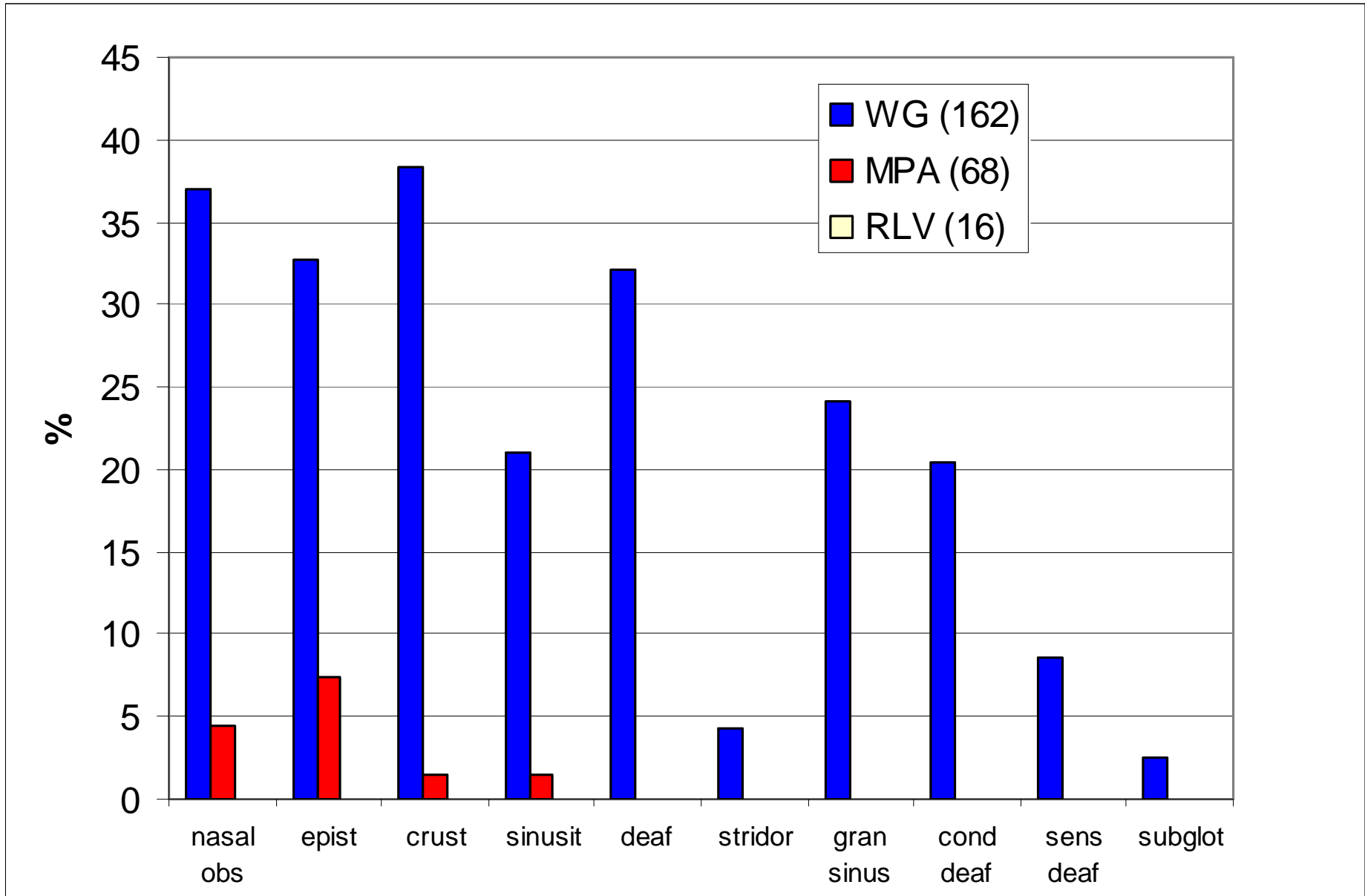
# Systemic features of vasculitis



# Cutaneous features of vasculitis



# Ear nose and throat features of vasculitis

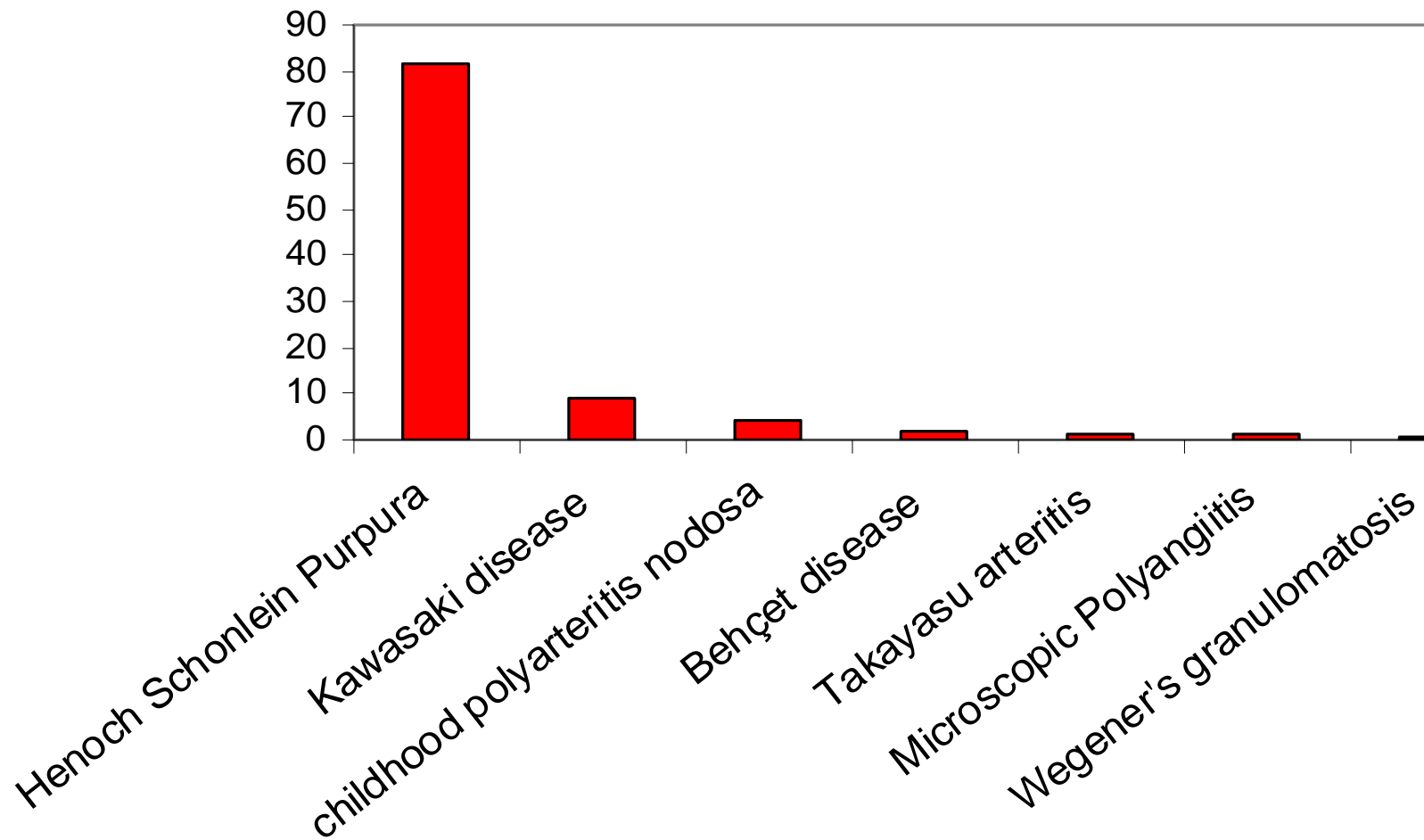


# Turkish survey of paediatric vasculitis

Ozen et al 2007

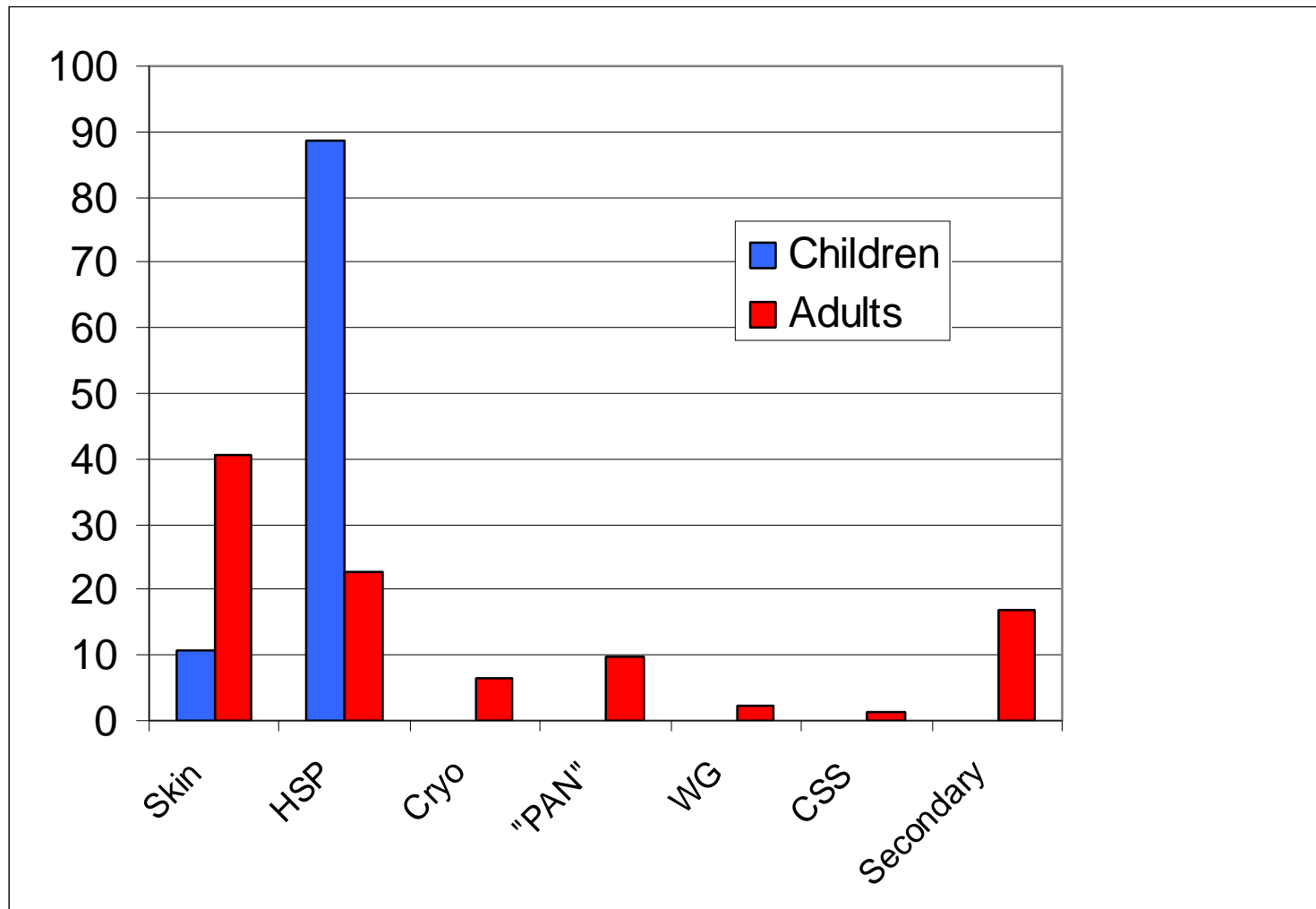
- Males and females equally affected
- mean age 11.05 +/-4.89 years
- Increased acute phase reactants
- Kidney involvement
  - 28.6% of HSP
  - 53% of the Classical - PAN.
- Abdominal aorta involved in all TA patients

# % of different childhood vasculitides in Turkey



# Cutaneous vasculitis in children and adults n= 303

Blanco et al Medicine (Baltimore) 1998 77:403-18.



# Conclusions

- Childhood forms of vasculitis are similar in spectrum to adult disease
- Emphasis on KD, HSP, less WG and MPA
- Long term treatment is similar
- Outcomes are better (but only small studies)
- Effects of disease and treatment on growth and development need to be addressed