Vasculitis Foundation; Cambridge 2007

Churg-Strauss angiitis

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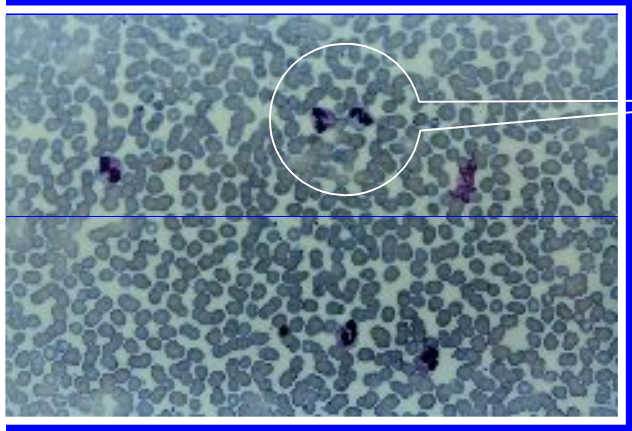
- What is it ?
- What happens if you have it ?
- What is the treatment ?

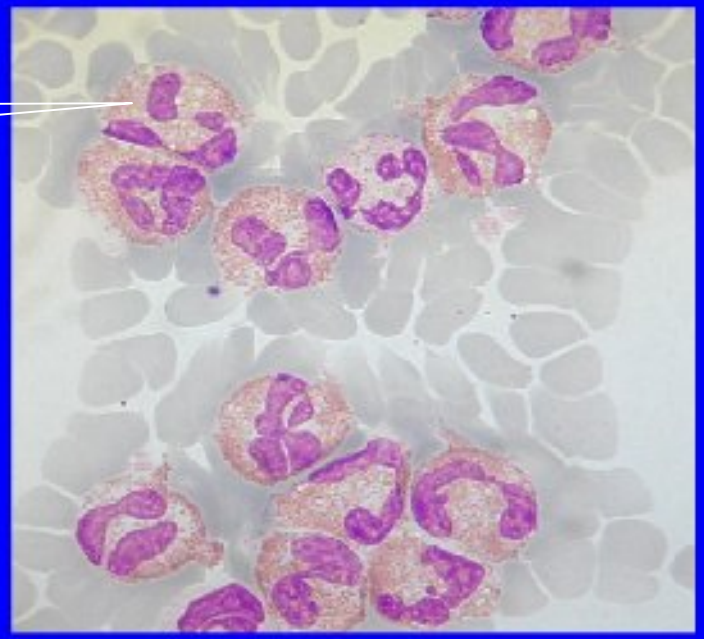
CSS – what is it ?

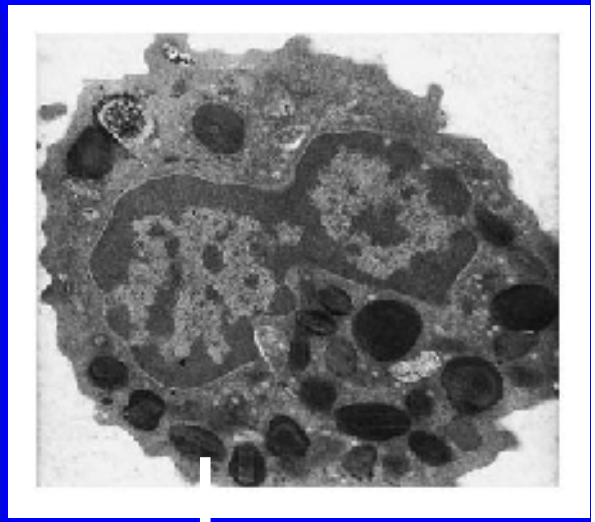
- A form of vasculitis
- Cause unknown, ‘primary vasculitis’
- First described by Jacob Churg and Eli Strauss, pathologists, New York in 1953

- Characterised by eosinophils in affected tissues and increased levels in the blood

What are eosinophils ?







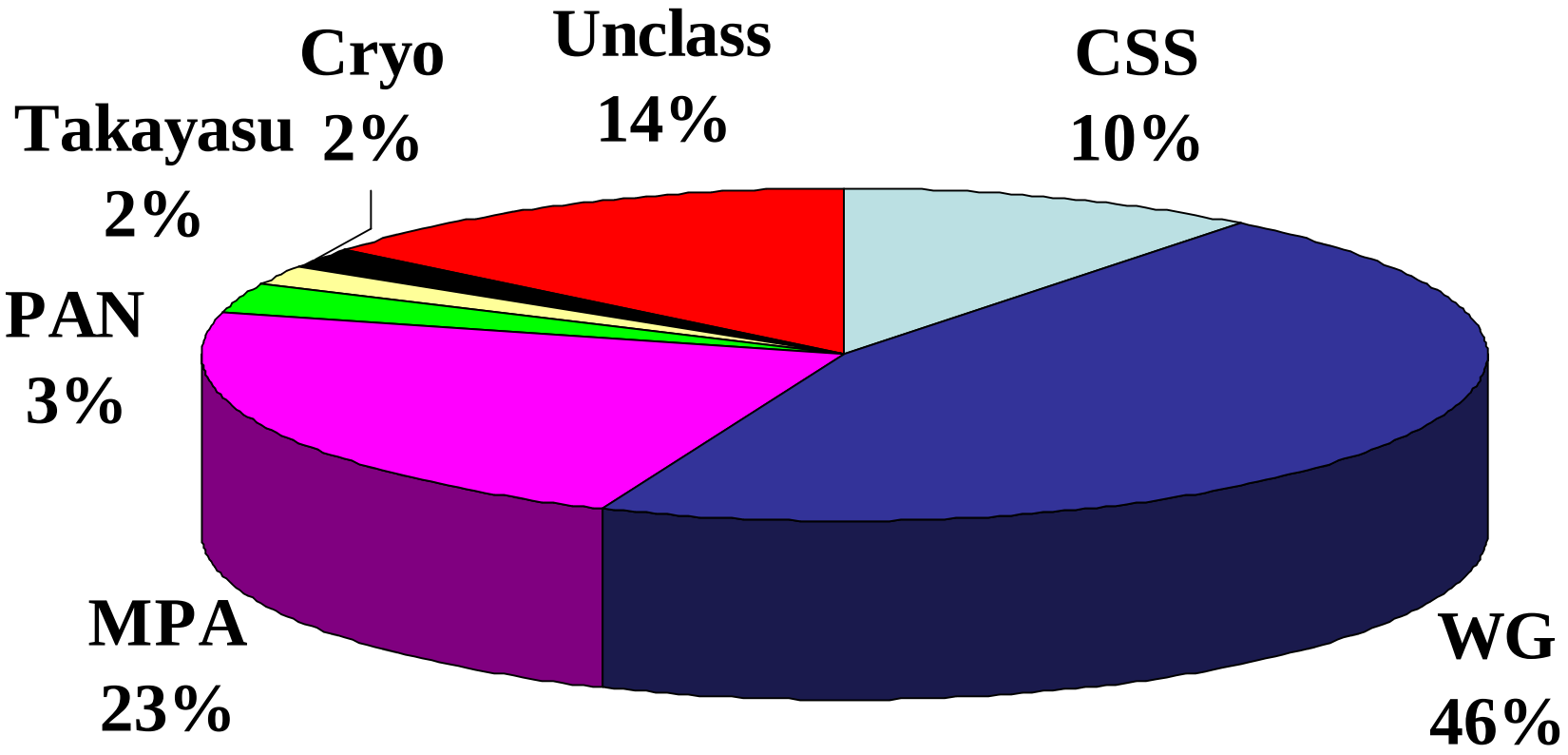
Level reported in ‘Full Blood Count’

Destructive enzymes

Other eosinophil diseases

- Allergy
- Parasitic infections
- Eosinophilic pneumonia
- Blood disorders
  - Hypereosinophil syndrome (HES)
  - Leukaemia

Vasculitis clinic: n=500



CSS – what happens if you have it ?

Similarities

- Prodrome
- General symptoms
- Overlaps with -
  WG, MPA, PAN

- Prognosis

Differences

- ↑ nerve 70%
- ↑ Gut 50%
- ↑ Heart 40%
- ↓ kidney 30%

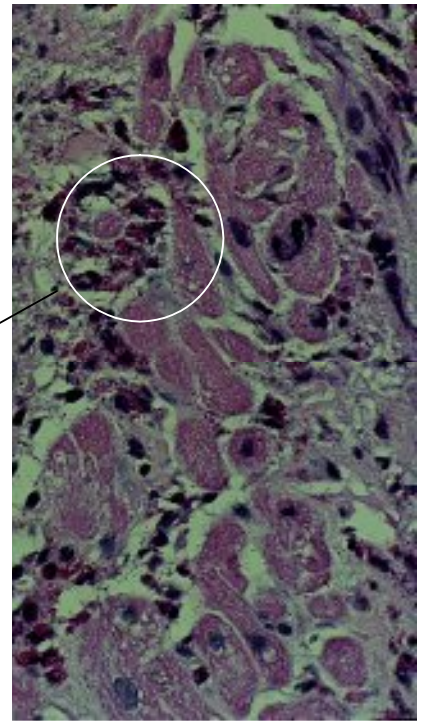
- ANCA 1/3
- Steroid responsive

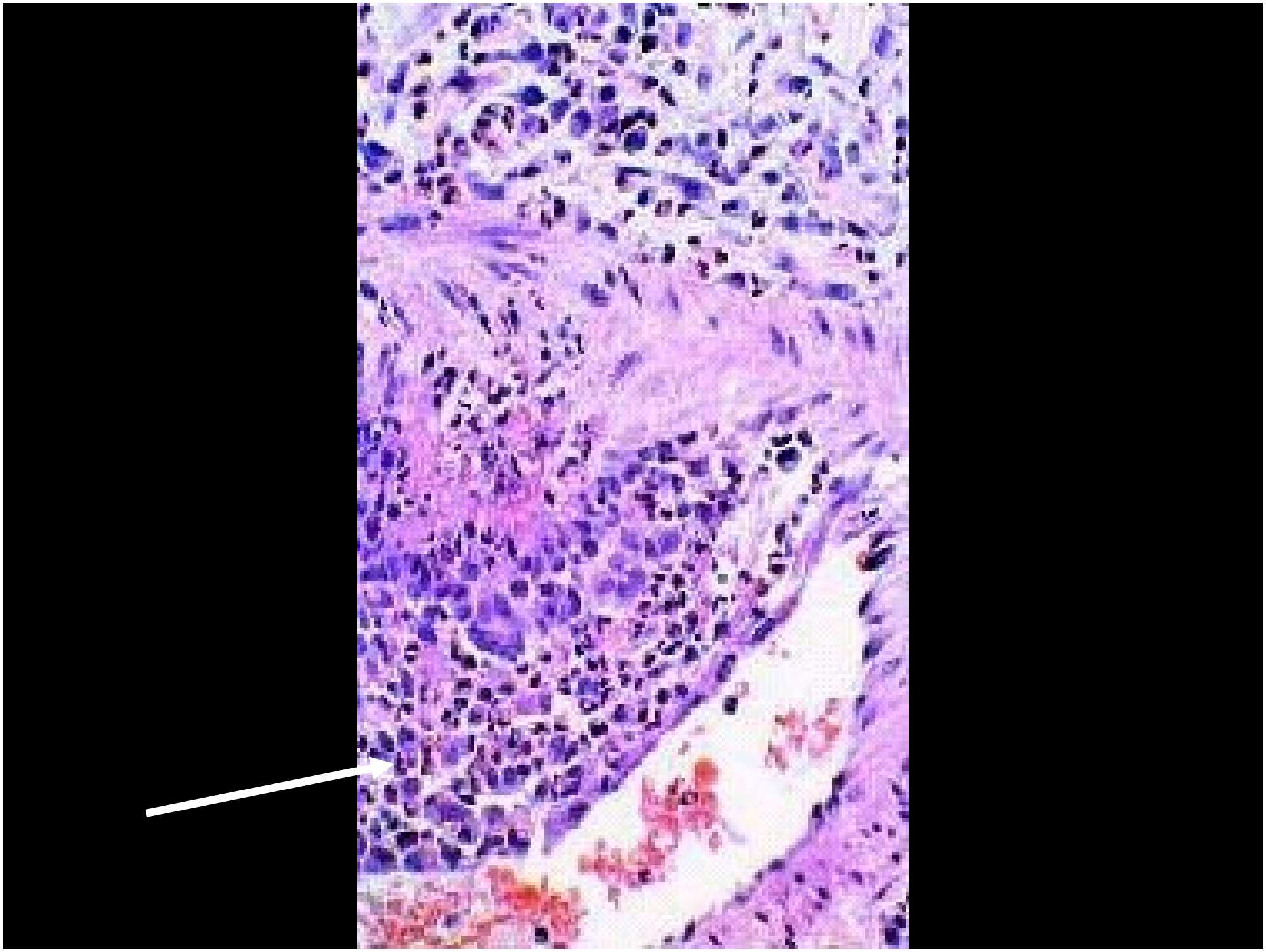
Criteria for diagnosis

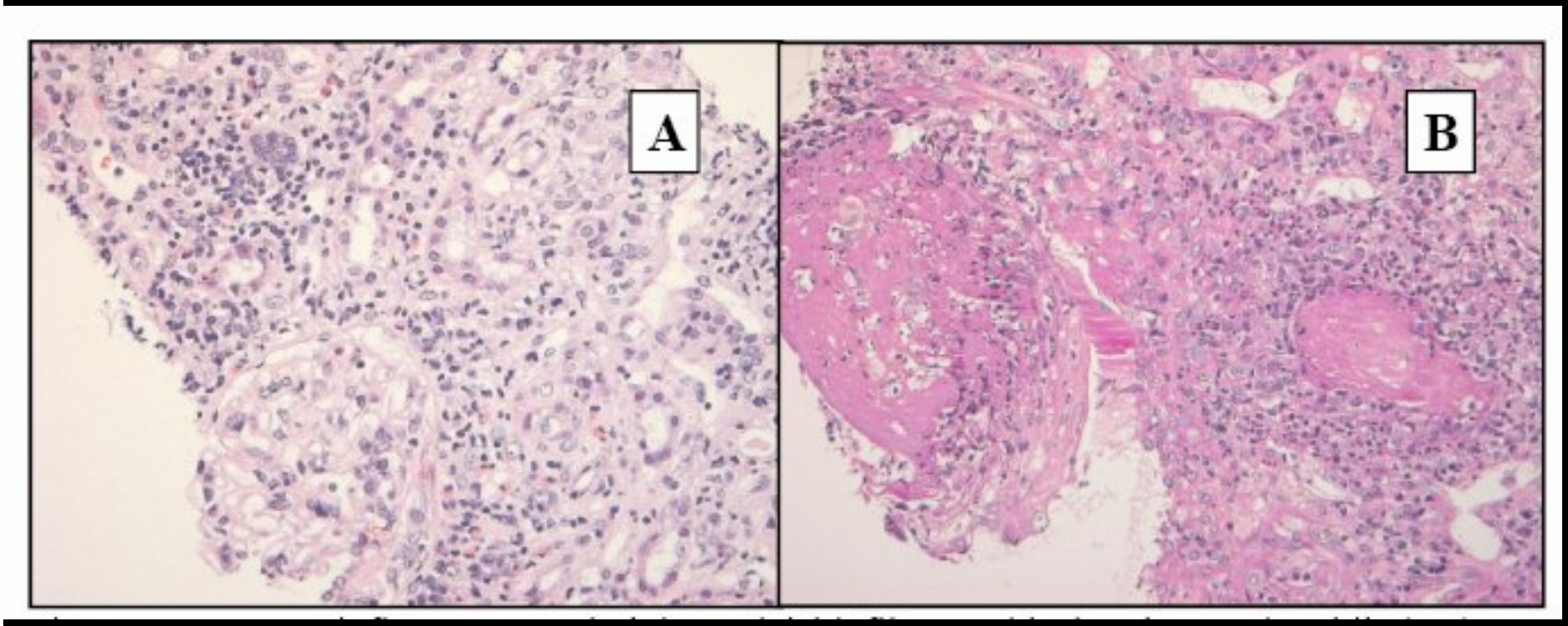
American College of Rheumatology 1990

** Need 4 out of 6 criteria:*

- Asthma
- >10% blood eosinophilia
- Pulmonary infiltrates on CXR
- Nose & sinus abnormalities
- Eosinophils in tissue biopsies
- Nerve involvement






A
B

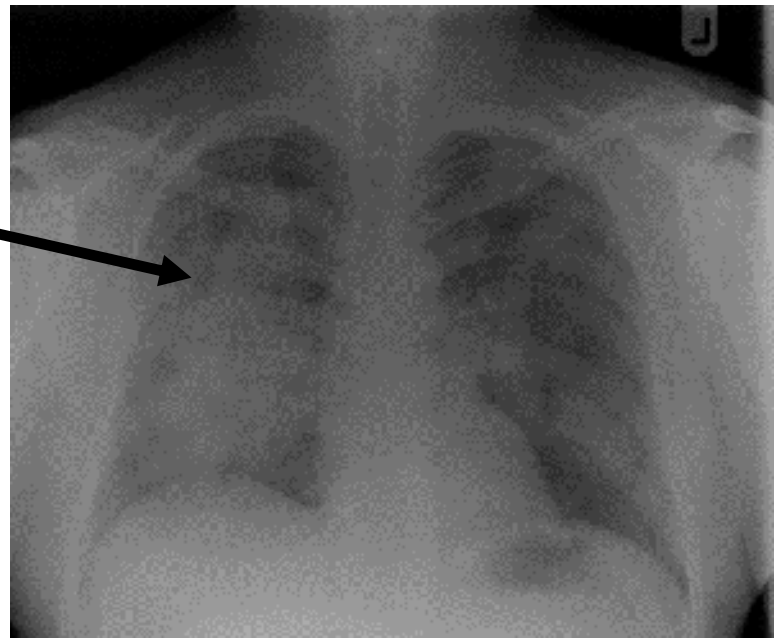
The montelukast story

- Montelukast is a newer treatment for asthma
- CSA patients more likely to have taken montelukast than healthy controls
- But more likely to have taken other asthma treatments

Hauser, *13th International ANCA workshop* 2007

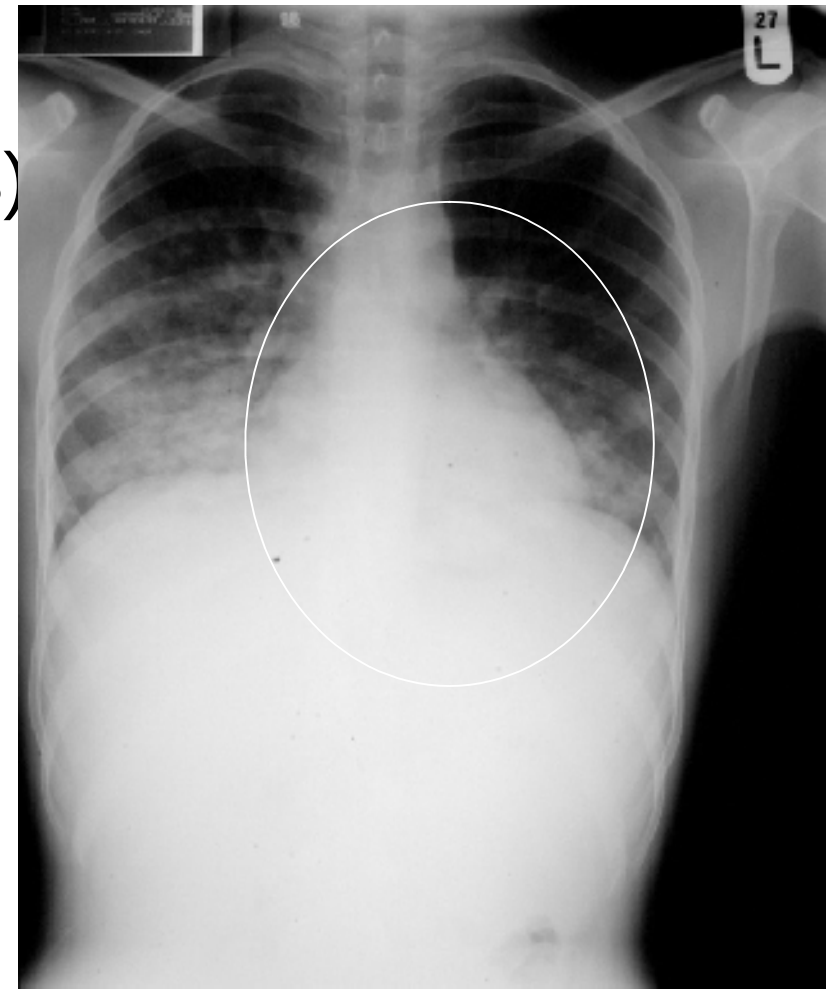
CSS – the lung

- Maturity onset asthma (wheeze)
- Lung infiltrates

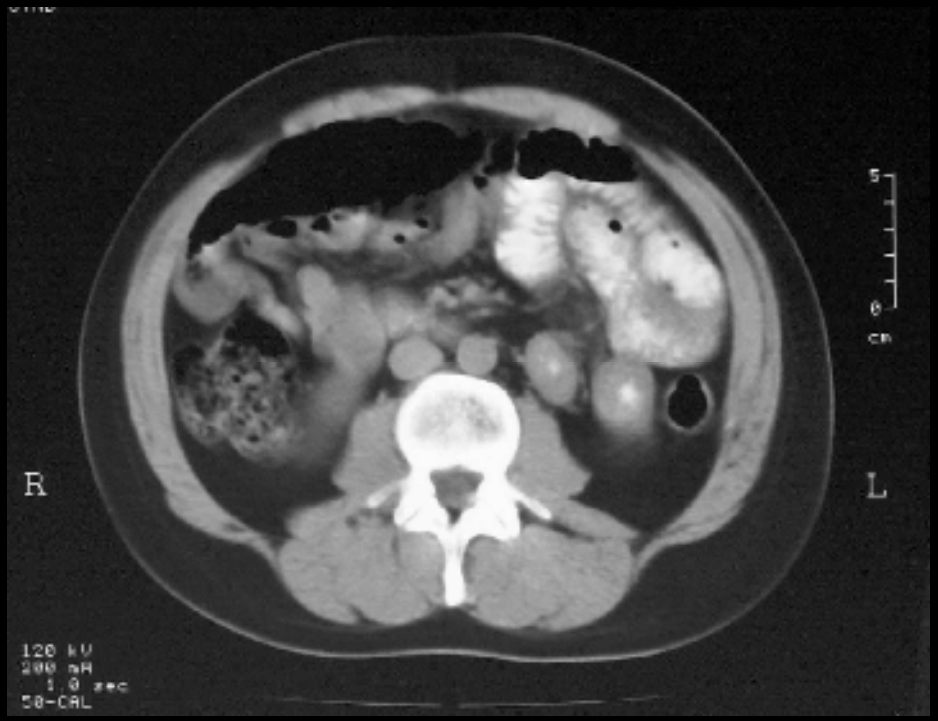


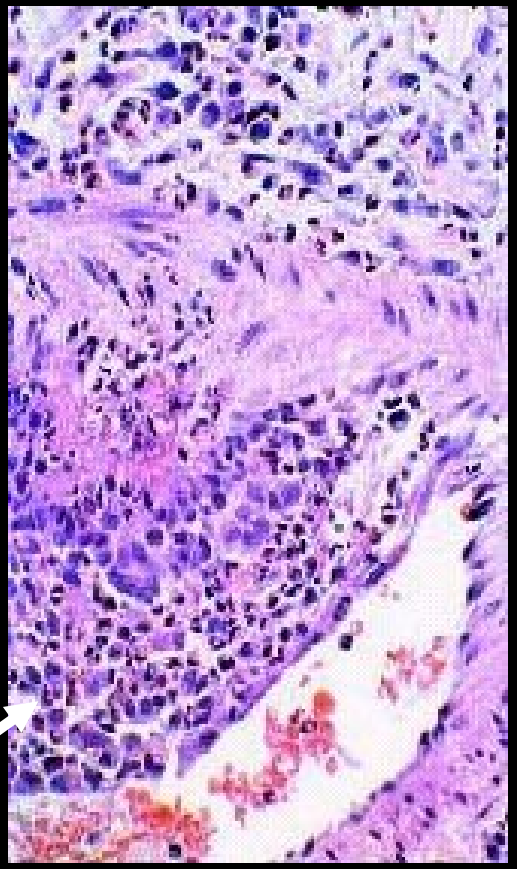
CSS and the heart

- 40 %
- Heart muscle (myocarditis) pump failure
- Heart sack (pericarditis)



CSS – the gut





What is the treatment ?

Prognostic factors (n=82)

	R.R. mortality
CNS	1.8
Cardiac	2.2
Renal	2.6
GI (surgery)	2.8

Guillevin 1995 Medicine;75:17

CHUSPAN trial – good prognosis

- Steroids alone work in 2/3
- The others require an immunesuppressive

CHUSPAN – poor prognosis (48 patients)

- All have steroids
- 6 months vs. 12 months cyclophosphamide
  - High remission rates with both
  - Fewer relapses with 12 months

Cohen, *Arthritis Rheum* 2007

What do we do ? – ‘Standard’

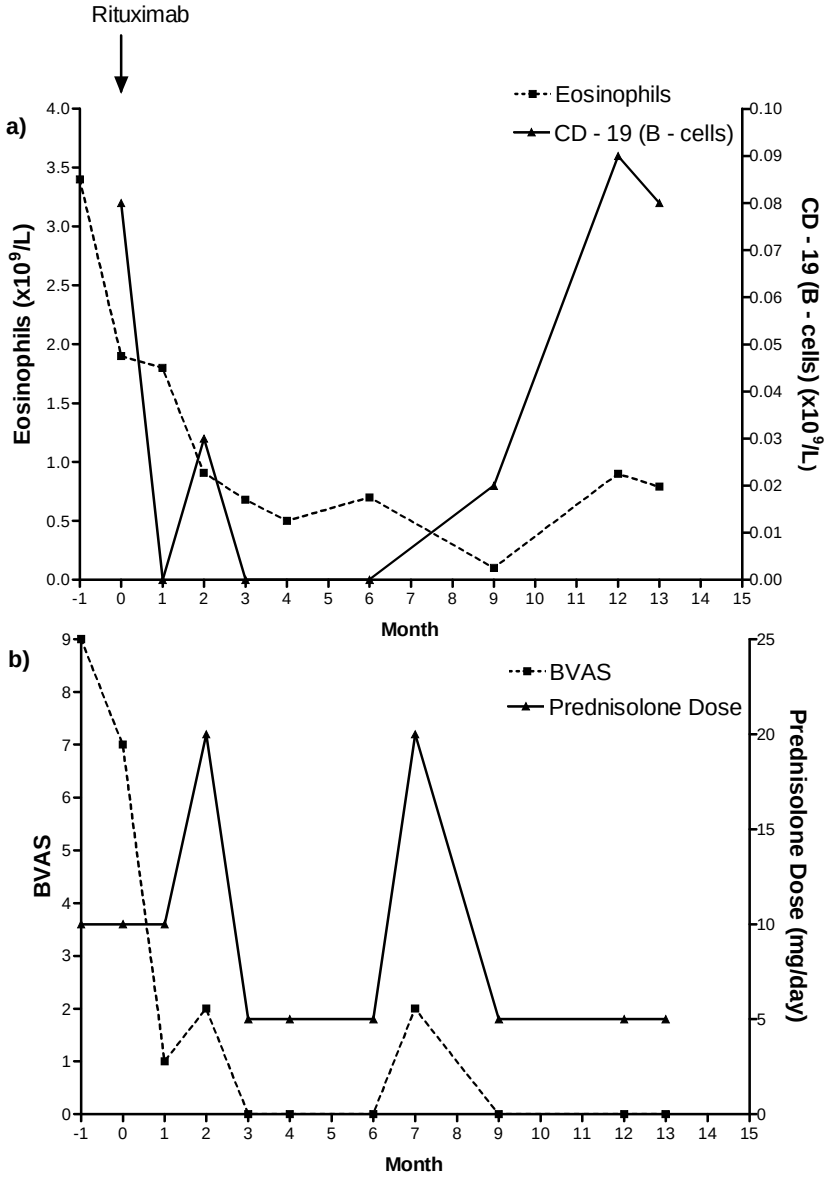
- Steroids
  - Intravenous (IV) and oral
- Immune suppressive
  - Usually start with IV cyclophosphamide, 3-6 months
  - Then ‘safer’ immunesuppressive for 2 years +
    - Azathioprine, methotrexate, mycophenolate

What do we do ? – ‘Alternative’

- Intravenous immunoglobulin
- Rituximab
- CAMPATH

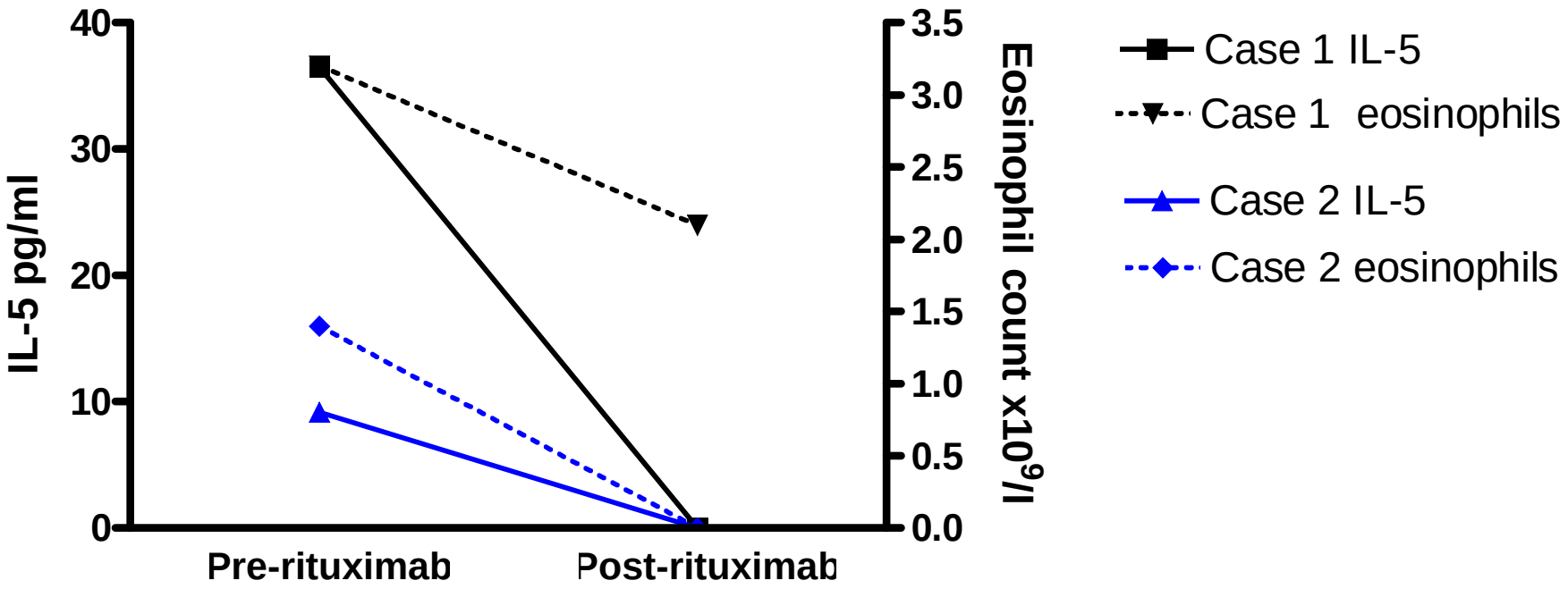
- Ciclosporin, interferon α

Rituximab in Churg Strauss Syndrome



Koukoulaki, *Ann Rheum Dis* 2005

CSS and IL-5



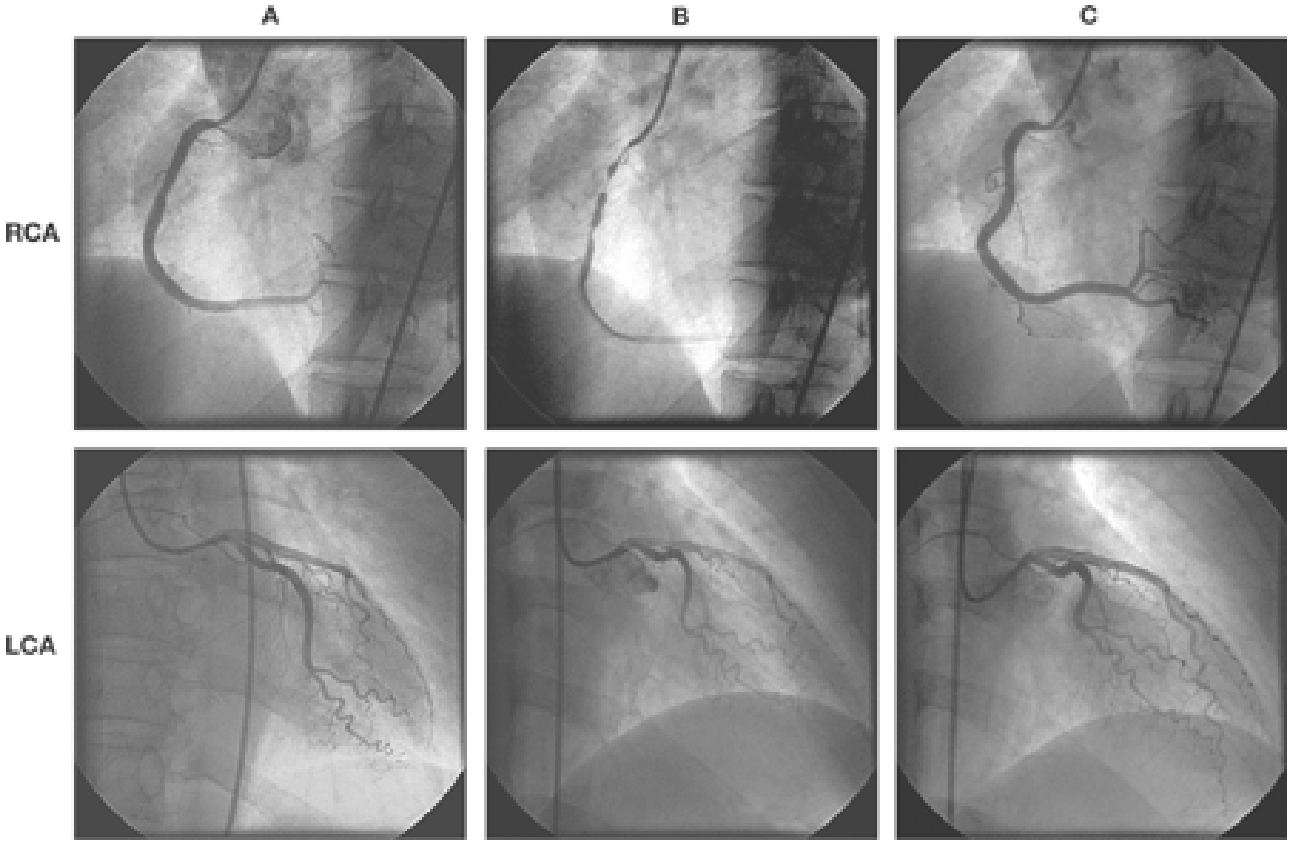
Salama et al

CSS therapy – the future

- Rituximab
- Anti-IL5 therapy – Mepolimumab

- More and better studies !!!

Figure 2 Coronary arteriogram showing epicardial vasomotor response before after infusion with intracoronary acetylcholine, and after administration of intracoronary bolus nitroglycerin



Petrakopoulou P *et al.* (2005) Vasospastic angina pectoris associated with Churg–Strauss syndrome
Nat Clin Pract Cardiovasc Med **2:** 484–489 doi:10.1038/ncpcardio0299

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