

Systemic Vasculitis

Recent Advances – Epidemiology

or “How common is it?”

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Queens College Cambridge 15th September 2007**



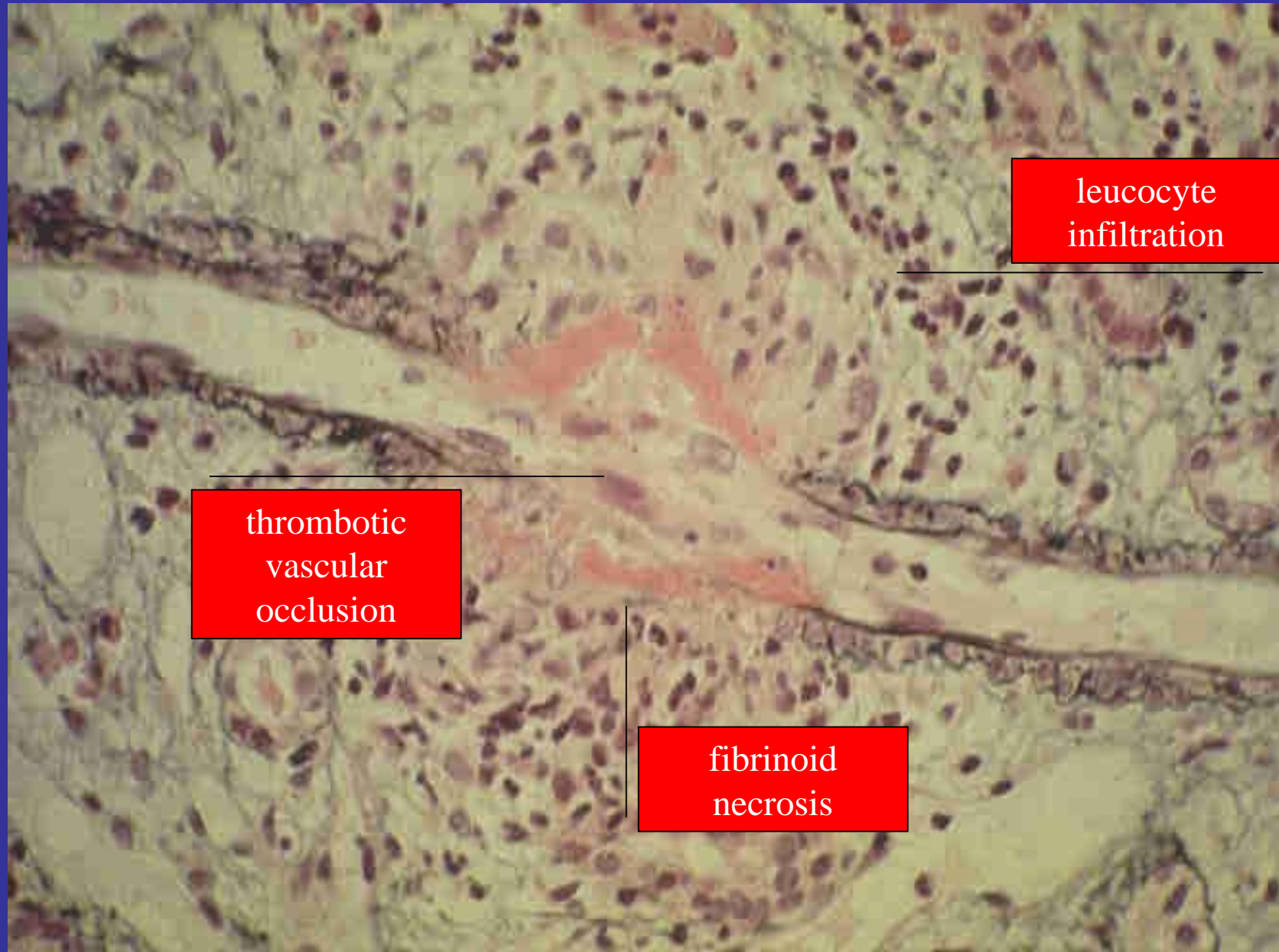


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"Yea, though I walk through the valley of the shadow of death, I will fear no evil" Psalm 23

Vasculitis - a histological triad



leucocyte
infiltration

thrombotic
vascular
occlusion

fibrinoid
necrosis

Classification of Systemic Vasculitis

	Primary	Secondary
LARGE ARTERIES	GCA Takayasu	aortitis - RA, AS infection - syphilis
MEDIUM ARTERIES	cPAN Kawasaki	infection - Hep B malignancy - hairy cell leukaemia
SMALL/MEDIUM VESSELS	Wegener's* MPA* Churg-Strauss*	RA, SLE, SS infection - HIV, Hep C drugs
SMALL VESSELS	HSP ess.cryo.angiitis cut.leuco.vasculitis	RA, SLE, SS infection drugs

*ANCA, cyclo responsive,
glomerulonephritis

Scott & Watts 1994 BJR

VASCULITIS 2007

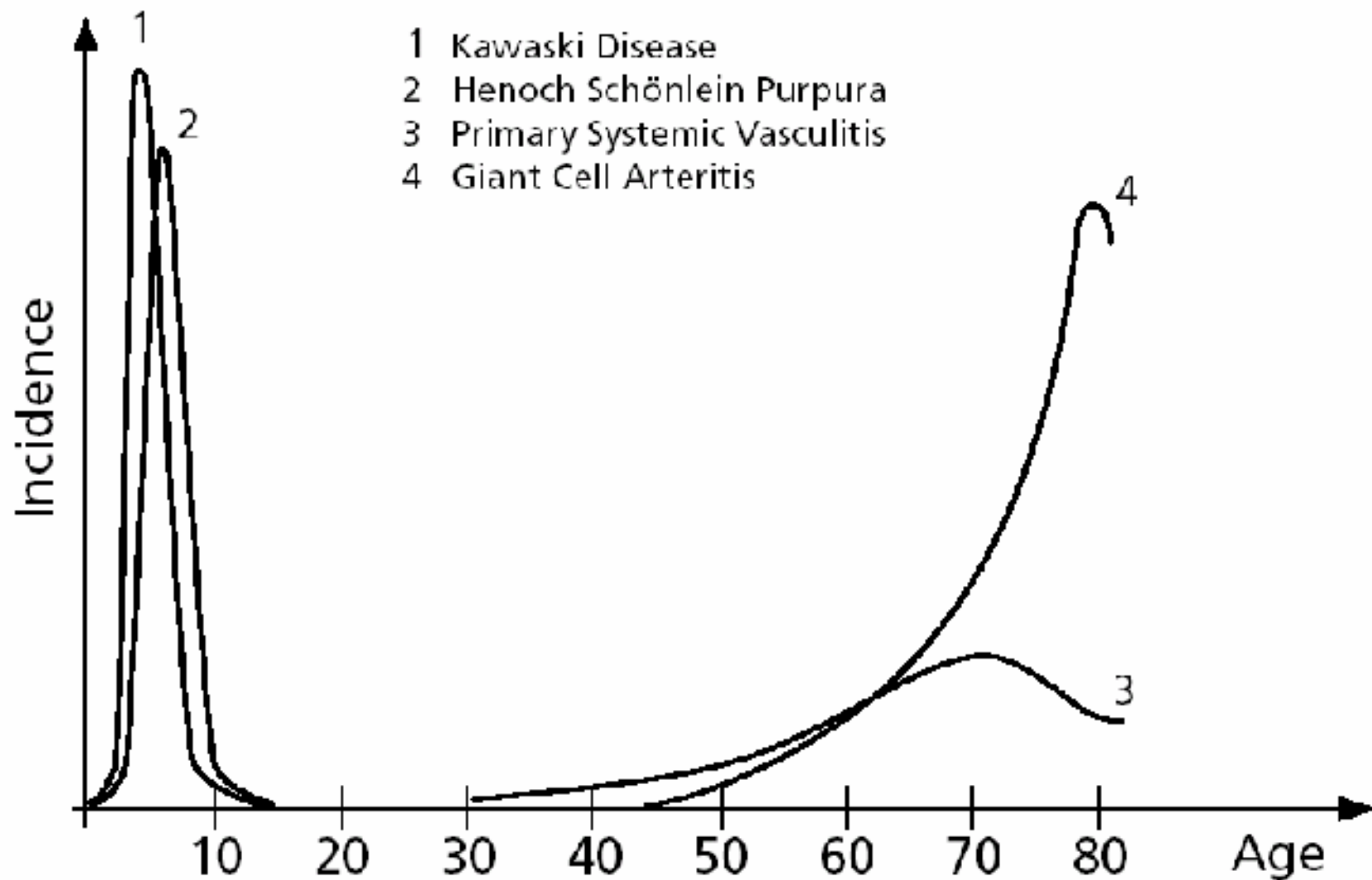
**“Inflammatory cell infiltration and
necrosis of blood vessel walls”**

Age 65-75 years male > female (1.5:1)

Incidence

- **Primary systemic vasculitis 20/million/year
(Wegener's, MPA, CSS)**
- **Overall ? > 150/million/year**
- **Secondary vasculitis ? 50/million/year**

Relative Incidence of Vasculitis by Age



NORFOLK ARTHRITIS REGISTER (NOAR)



NORFOLK VASCULITIS REGISTER

- Population based
- All GPs in NHA
- Population \approx 500,000
- 99% European Caucasians
- Patients \geq 16 years old
 swelling 2 or more joints
 lasting 4 or more weeks
- Clinical, laboratory, (x-ray) data
 annually for at least 5 years

- Population \approx 500,000
- 9% >75 yrs (> UK average)
- Patients entered prospectively
 from 1988 and confirmed by –
 histopathology database,
 renal biopsies,
 plasma exchange records
 hospital discharges

166 pts PSV (WG, CSS, MPA)
99 from NHA (1990-2000)

Norfolk Vasculitis Register 1988 - 2007

Total 680 patients

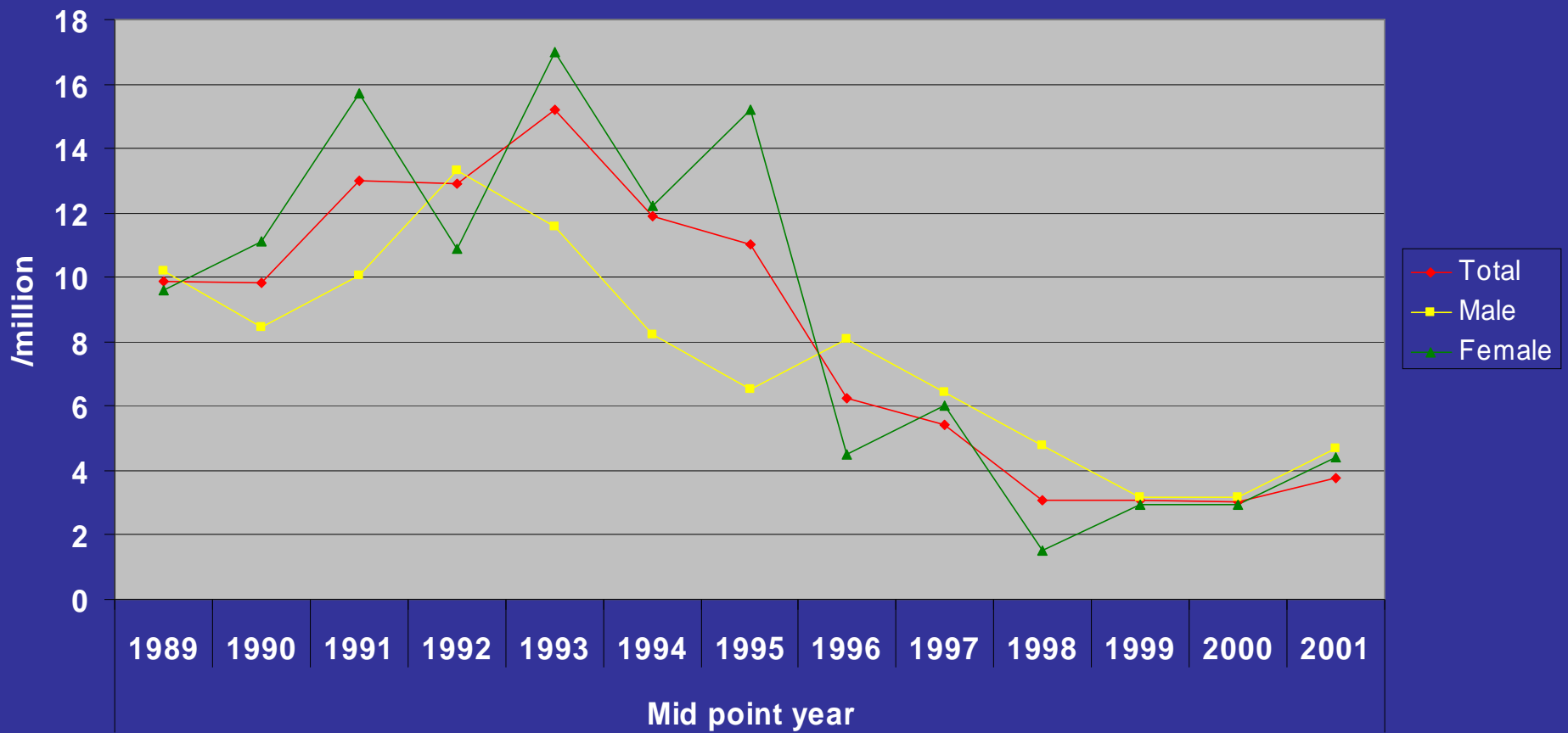
Primary Systemic Vasculitis

WG	132	Aortitis	5
MPA	57	Takayasu	5
CSS	37	Behcets	5

Secondary Vasculitis SRV 80 NFV 48

Small Vessel Vasculitis cutaneous LV 74
 cryo vasculitis 5
 adult HSP 23

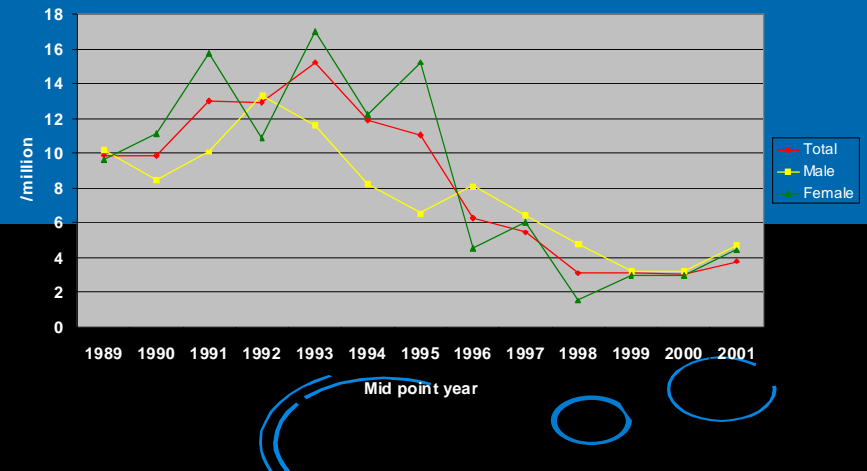
Rolling 3 year average



SYSTEMIC RHEUMATOID VASCULITIS the present



Rolling 3 year average



?12/million/year

• 2 yr mortality 23-46%

• ↑ risk mononeuritis,

gut involvement, arteritis

Scott et al 1981

	n	incidence/ million/yr	m	f
1988-92	24	11.6 (7.4-17)	12	11.2
1993-97	19	8.9 (5.3-13.6)	7.7	9.9
1998-02	8	3.6 (1.6-7.1)	3.7	3.5

Watts, Mooney, Lane & Scott Rheumatology 2004;43:920-3

Epidemiology of PSV

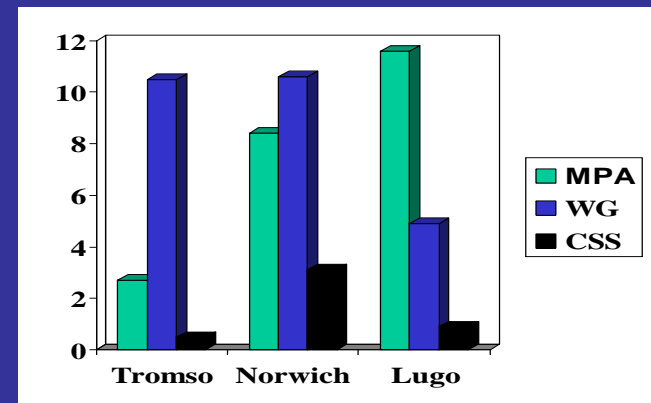
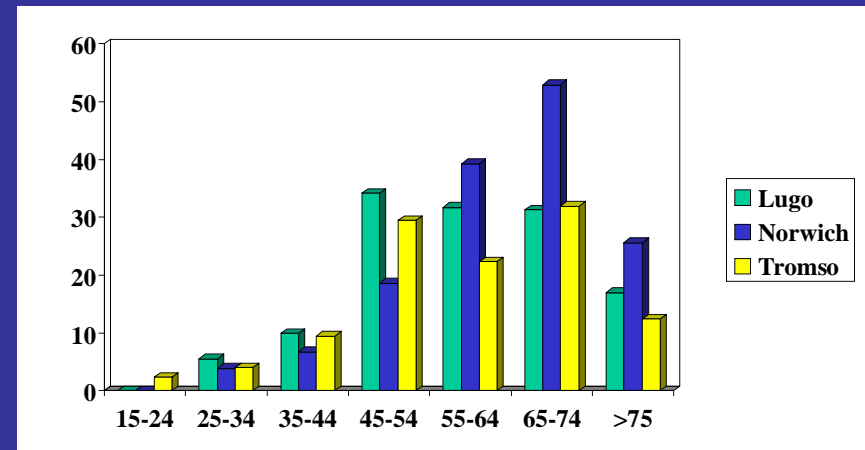
- Age peaks at 60 – 70 yrs

Prevalence 2x in European
(104.7/million) compared to
non European origin (52.5)
within France

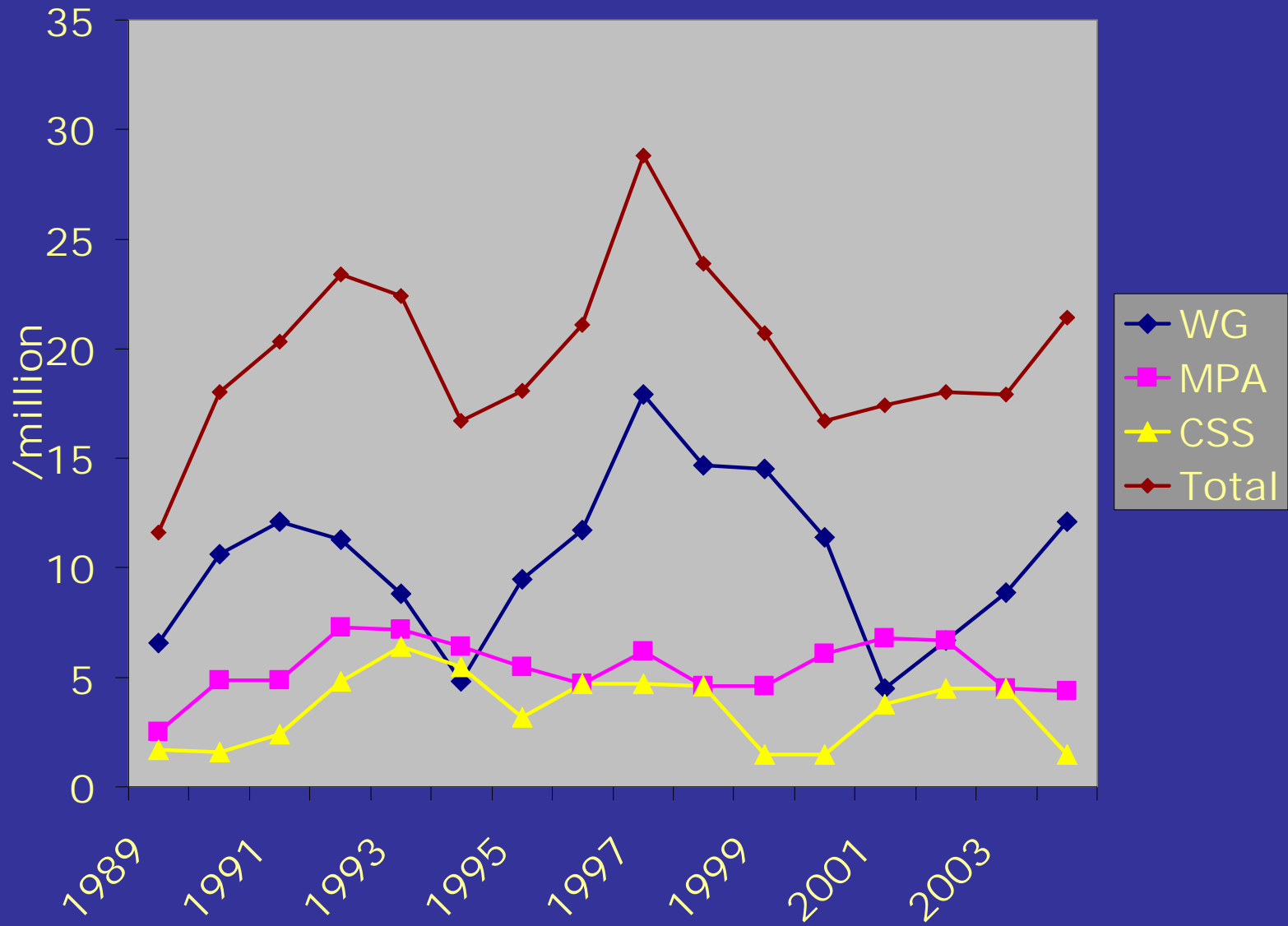
(Mahr et al 2004 ACR 2004;51:92-9)

- In Japan 90% RPGN MPO ANCA +,
PR3 ANCA (presumably WG) rare
- In India WG (54%) > MPA(18%)
PR3(48%) and MPO(43%) almost equal

Incidence (/million)



Rolling 3 year average 1988-2005



Wegener's Granulomatosis

Microscopic Polyangiitis

