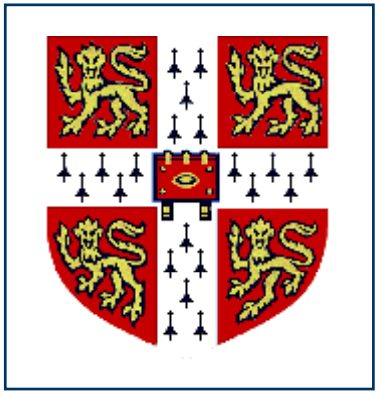
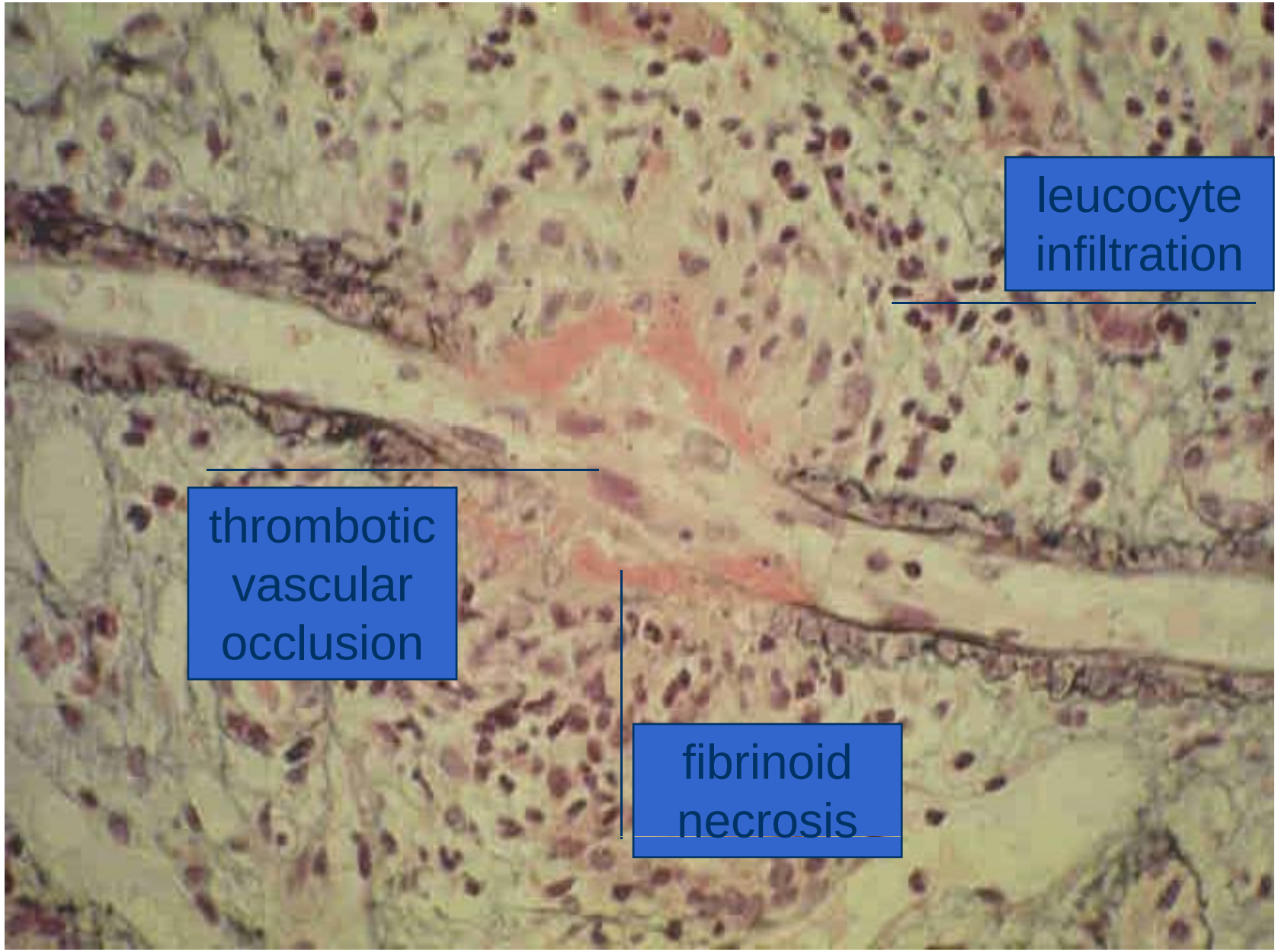
Vasculitis meeting, Tokyo, Japan 2009 January 9, 2009

Classification of Vasculitis

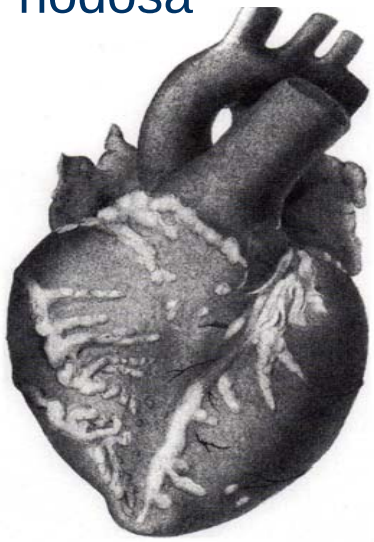
David Jayne
Vasculitis and Lupus Clinic
Addenbrooke's Hospital
Cambridge UK



Vasculitis- a histological triad



Polyarteritis nodosa



Adolf Kussmaul
(1822-1902)

Wegener's granulomatosis

Friedrich Wegener
(1907-1990)

Takayasu aortitis

American College of Rheumatology (ACR) 1990 Classification

- Aim - to promote the uniform description of patients for the purposes of research
- 48 rheumatology centres in USA, Canada and Mexico
- Cases of ‘definite vasculitis’ according to the individual rheumatologist
- 1020 submitted, 807 selected
- 154 discriminators selected
- 182 combinations tested

- Validated against ‘other vasculitis’

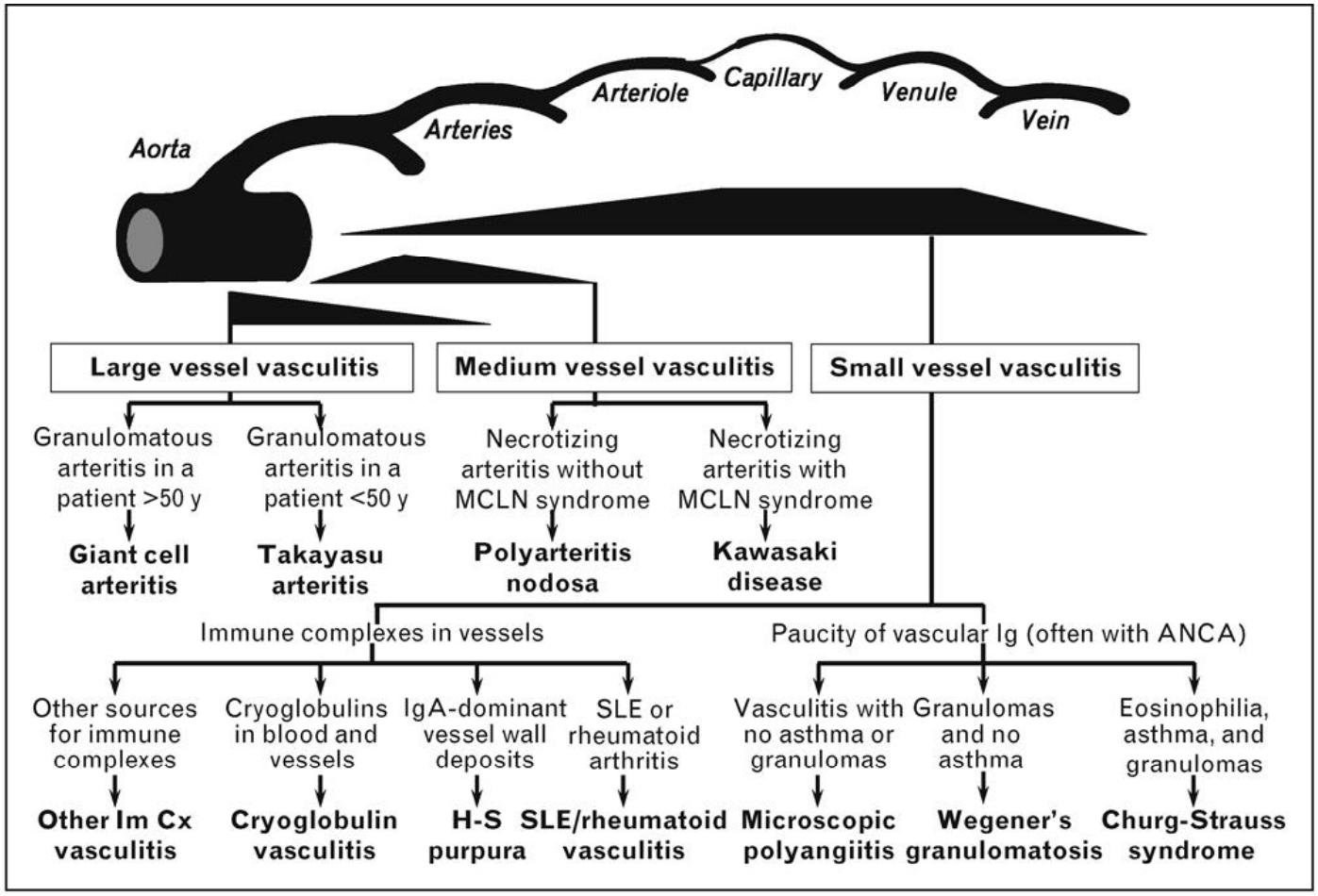
Arthritis & Rheumatism. 1990;33

ACR 1900 Classification

Vasculitis type	Classification method	Sensitivity (%)	Specificity (%)
PAN	Traditional	82.2	86.6
	Tree	87.3	89.3
CSS	Traditional	85.0	99.7
	Tree	98.0	99.2
WG	Traditional	88.2	92.0
	Tree	87.1	93.6
HV	Traditional	71.0	83.9
	Tree	78.5	78.7
HSP	Traditional	87.1	87.7
	Tree	89.4	88.1
GCA	Traditional	93.5	91.2
	Tree	95.3	90.7
TA	Traditional	90.5	97.8
	Tree	92.1	97.0

Chapel Hill Consensus: 1992

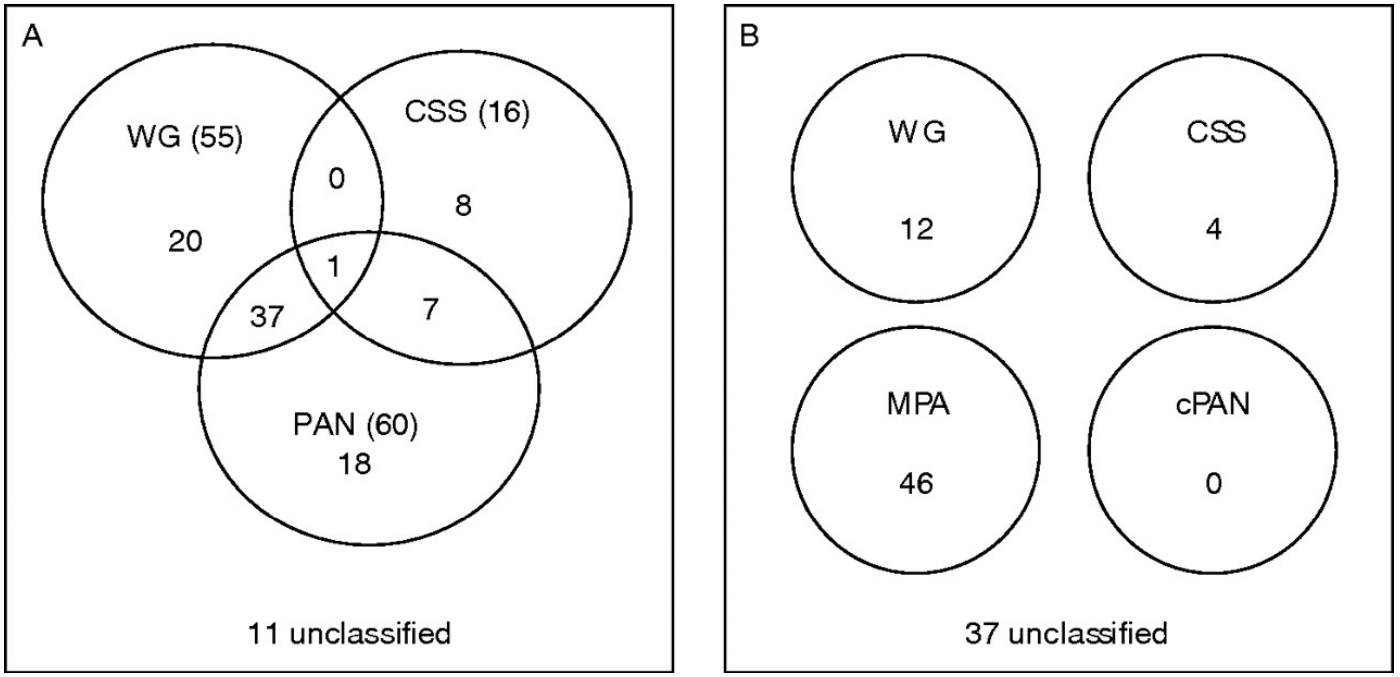
Chapel Hill Consensus Conference (CHCC)



Algorithm showing some of the pathologic and clinical features that allow classification and diagnostic differentiation among the different forms of vasculitis. ANCA, antineutrophil cytoplasm autoantibody; H-S purpura, Henoch-Schönlein purpura; Im Cx, immune complex; MCLN syndrome, mucocutaneous lymph node syndrome; SLE, systemic lupus erythematosus.

Jeannette JC et al. *Arthritis & Rheumatism. 1994;37(2):187-92.*

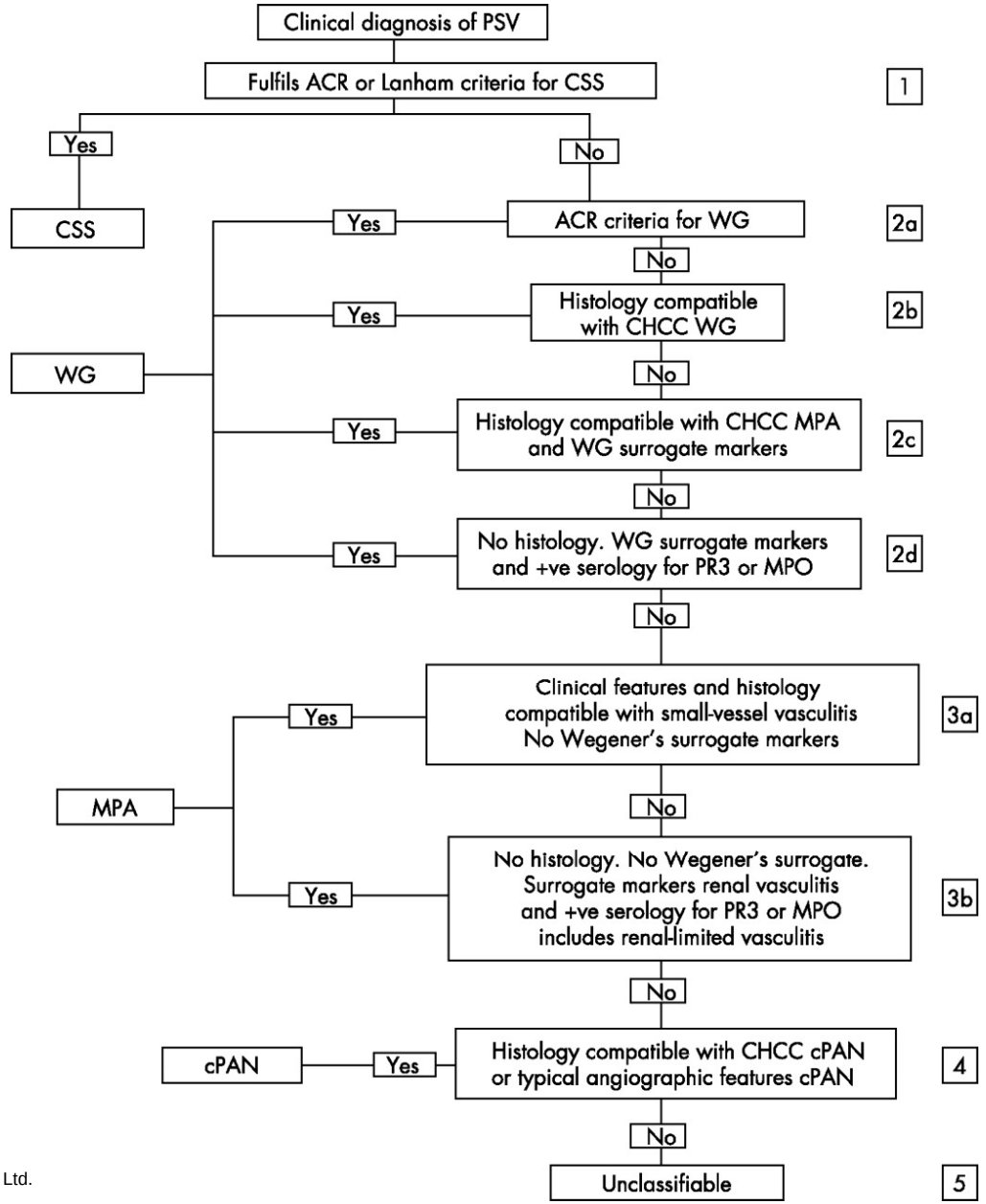
Application of (A) ACR (1990) criteria and (B) CHCC definitions for Wegener's granulomatosis (WG), Churg-Strauss syndrome (CSS), microscopic polyangiitis (MPA) and polyarteritis nodosa (PAN), n = 99



Watts, R. et al. Ann Rheum Dis 2007;66:222-227

EMEA Vasculitis Classification algorithm

Watts, R. et al. Ann Rheum Dis 2007;66:222-227



EULAR/PRES Endorsed Consensus Criteria* for the Classification of Childhood Vasculitides * under review by the ACR

Seza Ozen, Nicolino Ruperto, Michael Dillon, Arvind Bagga, Karyl Barron, Jean Claude Davin, Tomisaku Kawasaki, Carol Lindsley, Ross Petty, Anne-Marie Prieur, Angelo Ravelli and Patricia Woo

Ann Rheum Dis published online 1 Dec 2005;
doi:10.1136/ard.2005.046300

eular
european league against rheumatism

American College of Rheumatology
Education • Treatment • Research

Points to consider in the diagnosis, definition and classification of systemic vasculitis

Richard Watts, Raashid Luqmani

Steering Group Members

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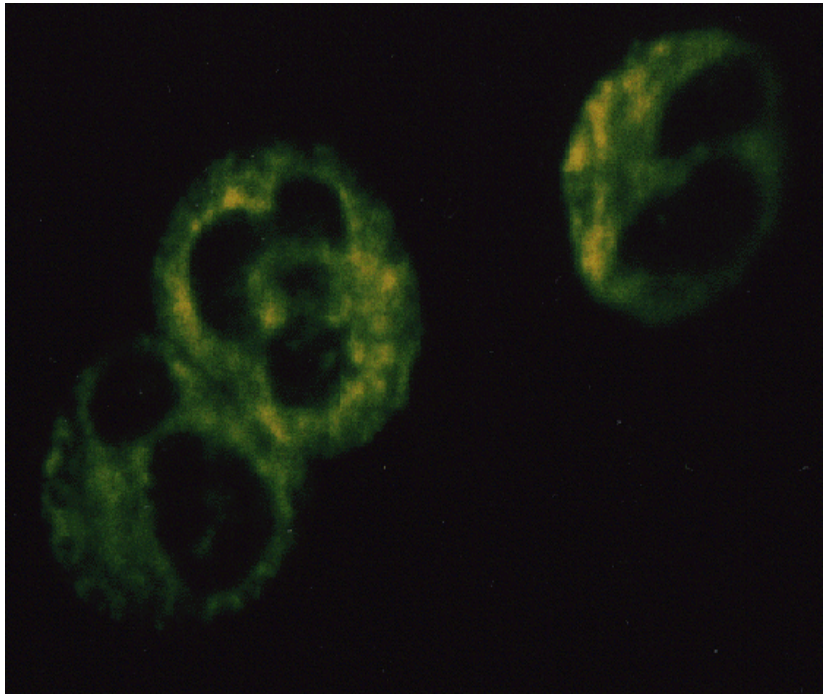
Brogan	Paul
Calabrese	Len
Cid	Maria
Cohen-Tervaert	Jan Wilhelm
Coles	Alastair
de Groot	Kirsten
Gross	Wolfgang
Guillevin	Loic
Hauser	Thomas
Jayne	David
Jennette	Charles
Kallenberg	Cees
Kobayashi	Shigeto
Little	Mark
Luqmani	Raashid
Mahr	Alfred
Merkel	Peter
Puechal	Xavier
Rasmussen	Niels
Salama	Alan
Salvarani	Carlo
Savage	Caroline
Scott	David
Segelmark	Marten
Specks	Ulrich
Sunderkootter	Cord
Suzuki	Kazuo
Tesar	Vladimir
Watts	Richard
Wiik	Alan
Yazici	Hasan

Important concepts

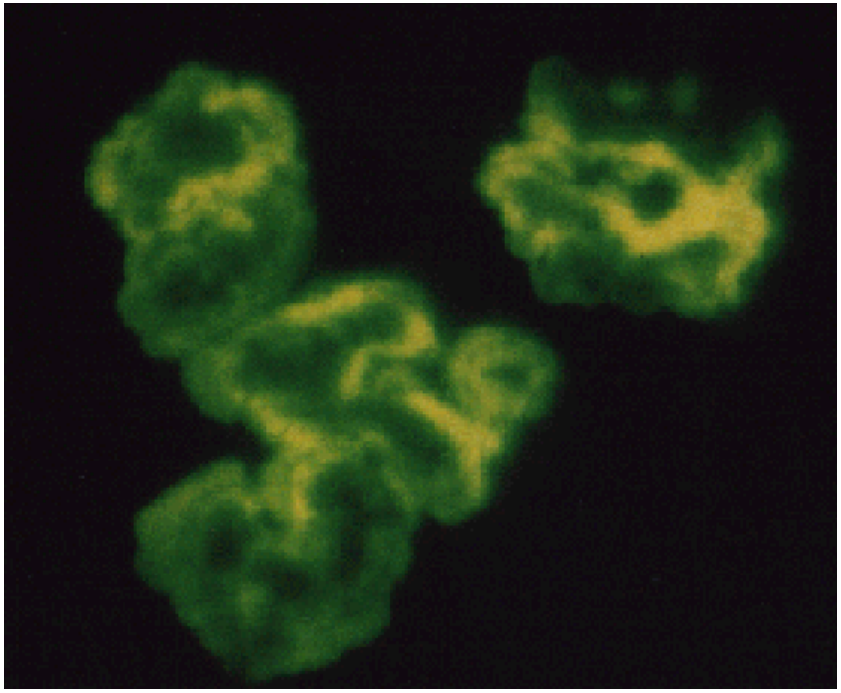
1. Hierarchy of sub-groupings and syndromes
   Refer to pathophysiology where possible
2. Overlaps between syndromes and primary/secondary
   Indicate major links between syndromes
3. Levels of confidence Definite/Probable/Possible
   Allow for migration between confidence levels
4. Consider:
   Pediatric vasculitis
   Mimics and non-inflammatory vasculopathies

ANCA testing

C-ANCA / PR3-ANCA



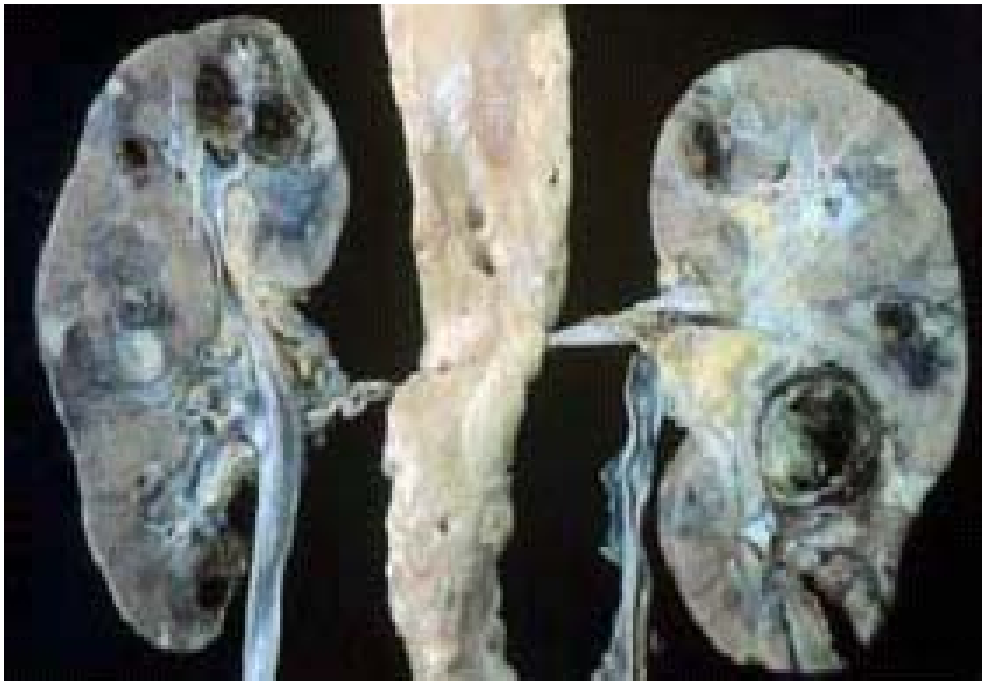
P-ANCA / MPO-ANCA



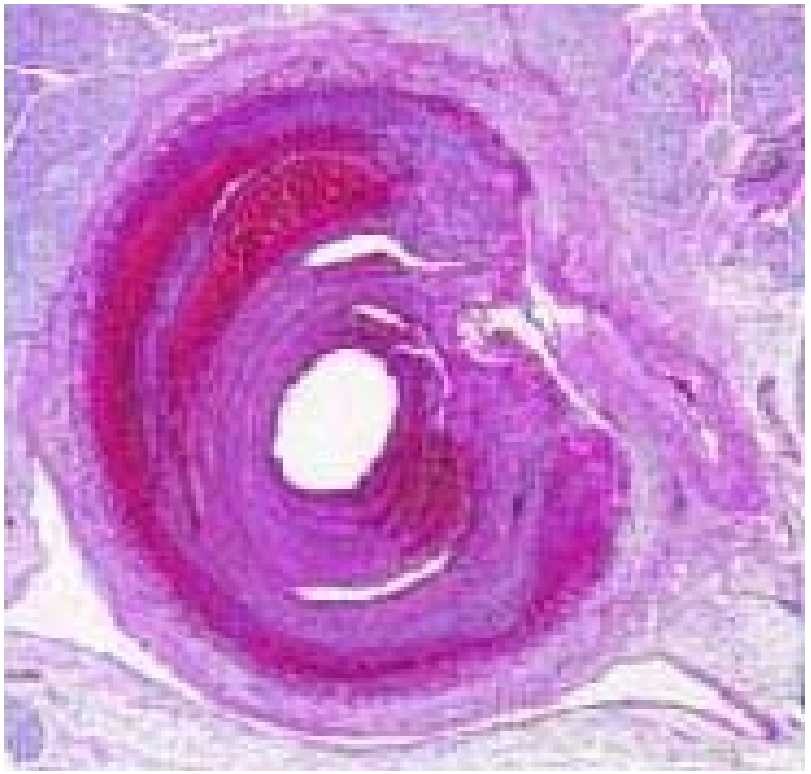
- Sensitivity and specificity depend on clinical setting
- Suspected RPGN: sensitivity > 90%, specificity > 95%
- Negative ANCA does not rule out vasculitis

Polyarteritis nodosa

Necrotizing inflammation of medium-sized or small arteries without glomerulonephritis or vasculitis in arterioles, capillaries or venules.

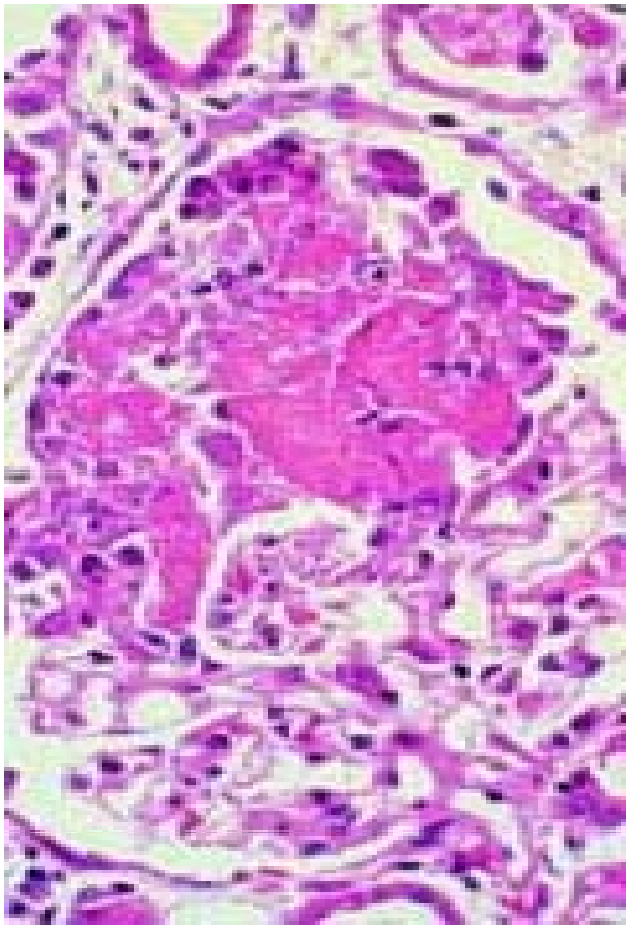


J. Charles Jennette



Microscopic polyangiitis

Necrotizing vasculitis with few or no immune deposits affecting small vessels, i.e. capillaries, venules, or arterioles.
Necrotizing arteritis involving small and medium-sized arteries may be present.
Necrotizing glomerulonephritis is very common.

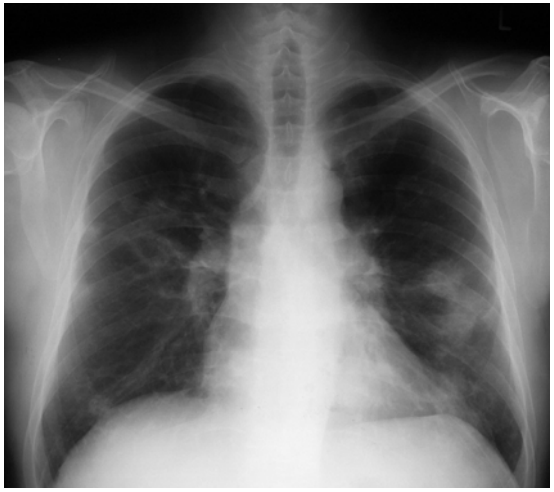


J. Charles Jennette

ANCA associated vasculitis

Wegener's granulomatosis

- Nose, lung, kidney 70%
- PR3 >> MPO-ANCA



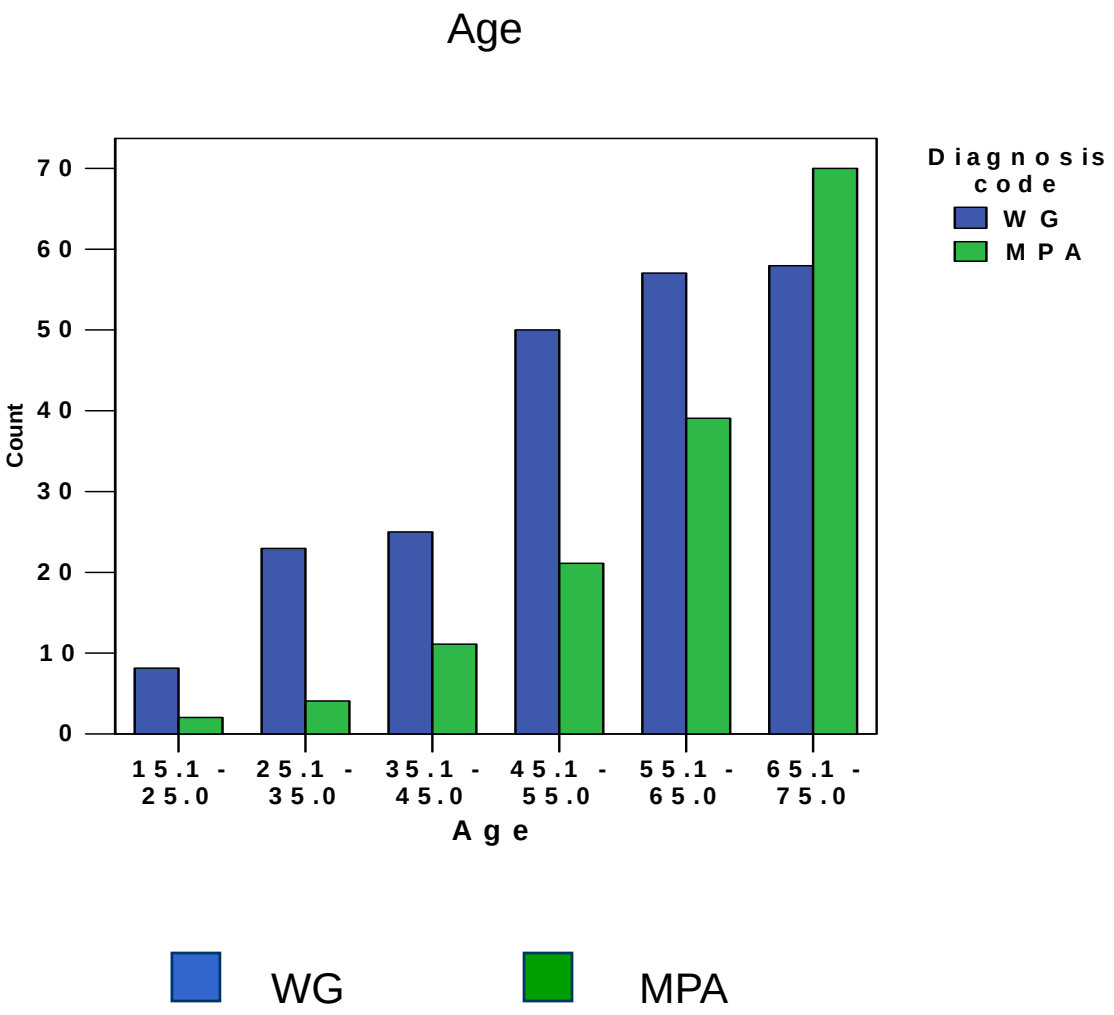
Microscopic polyangiitis

- Kidney 90%
- MPO>PR3-ANCA
- Renal-limited variant

Churg-Strauss angiitis

- Kidney 15%
- MPO>>PR3-ANCA

Epidemiology



Incidence:
AAV 15-20/million

Prevalence:
100-200/million

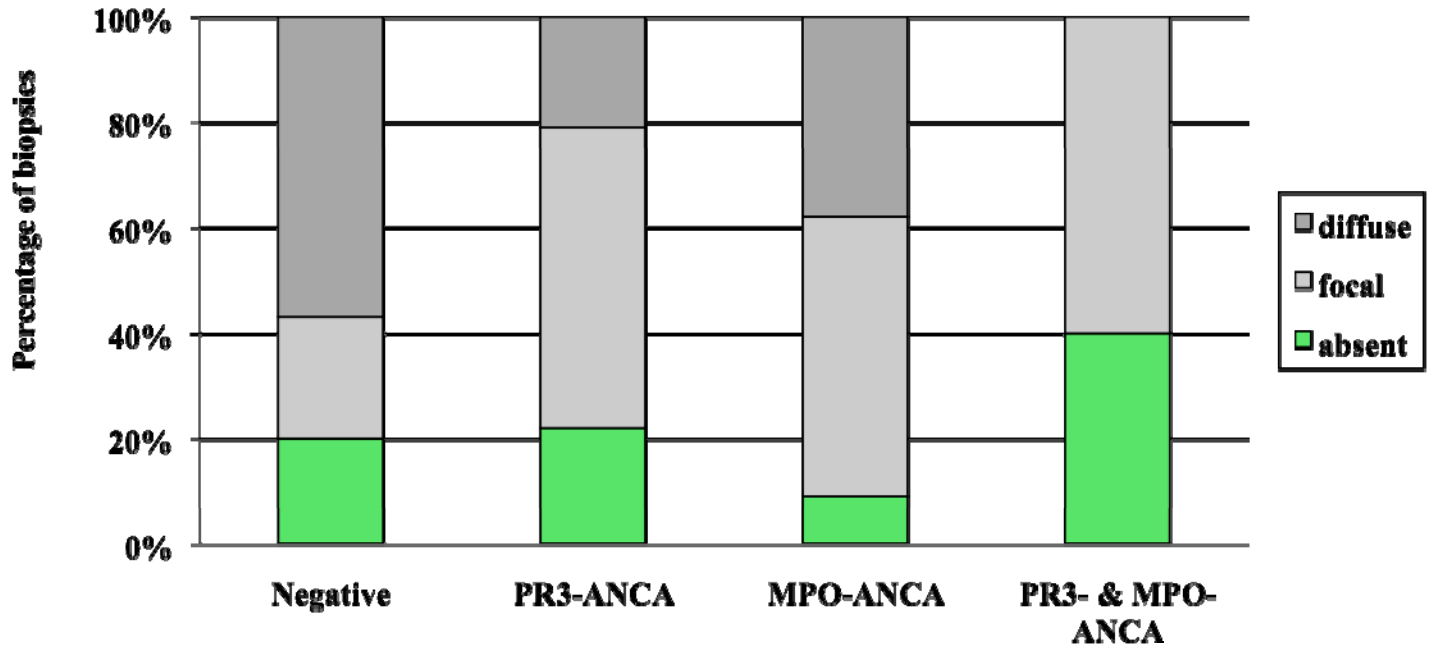
Ethnicity:
Caucasian>Asian>African

Sex:
M=F

Renal-limited
<5%

EUVAS
Lane 2004, Arthritis Rheum

Interstitial fibrosis in the diagnostic biopsy



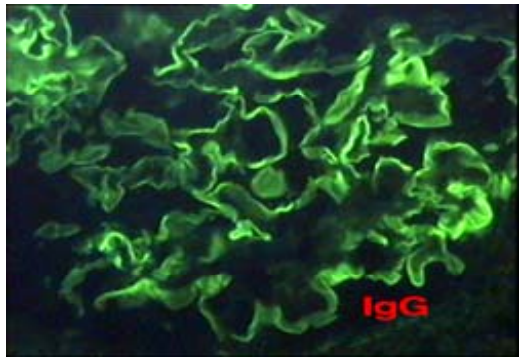
Neg. vs. PR3-ANCA: p = 0.029
Neg. vs. MPO-ANCA: p = 0.403
Neg. vs. PR3- & MPO-ANCA: p = 0.053

PR3-ANCA vs. MPO-ANCA: p = 0.008
PR3- vs. PR3- & MPO-ANCA: p = 0.220
MPO- vs. PR3- & MPO-ANCA: p = 0.026

Hauer H, Kidney International 2001

Renal histology; classification by immune deposits

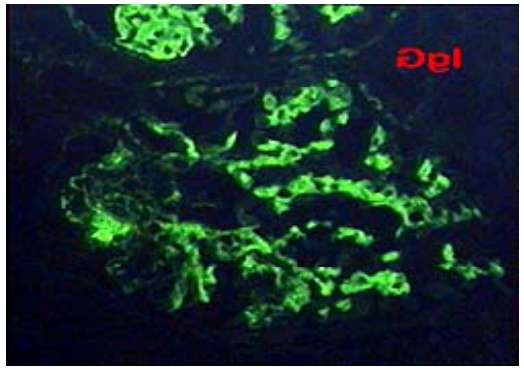
Type I

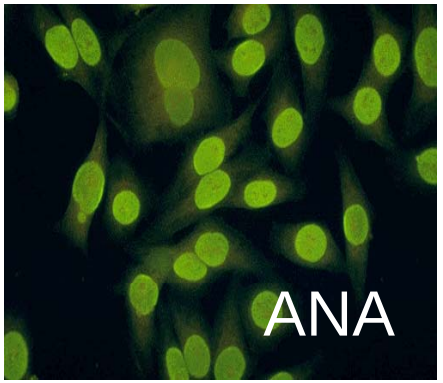


Anti-GBM antibodies

Anti-GBM disease

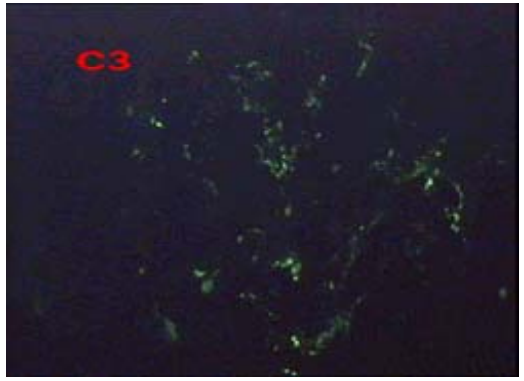
Type II

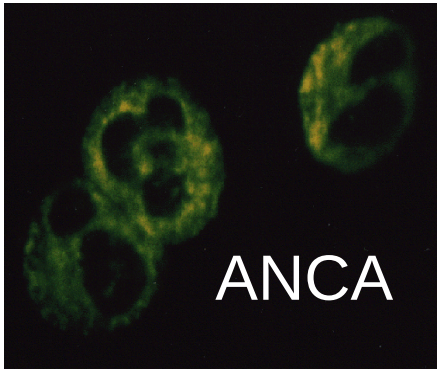




SLE
Cryoglobulin
HSP

Type III



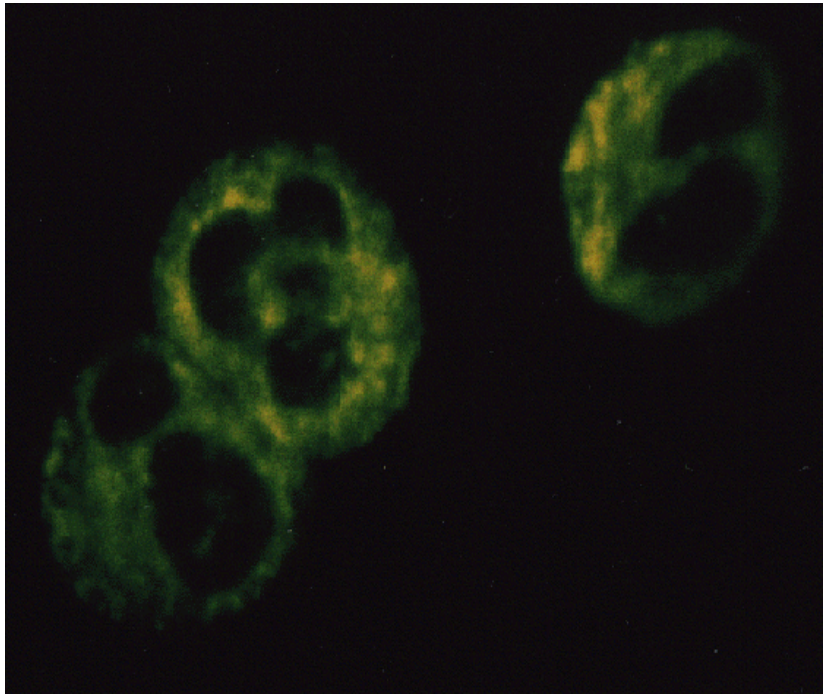


Vasculitis
‘pauci-immune’

Causer, Am J Kid Dis 1988

ANCA testing

C-ANCA / PR3-ANCA



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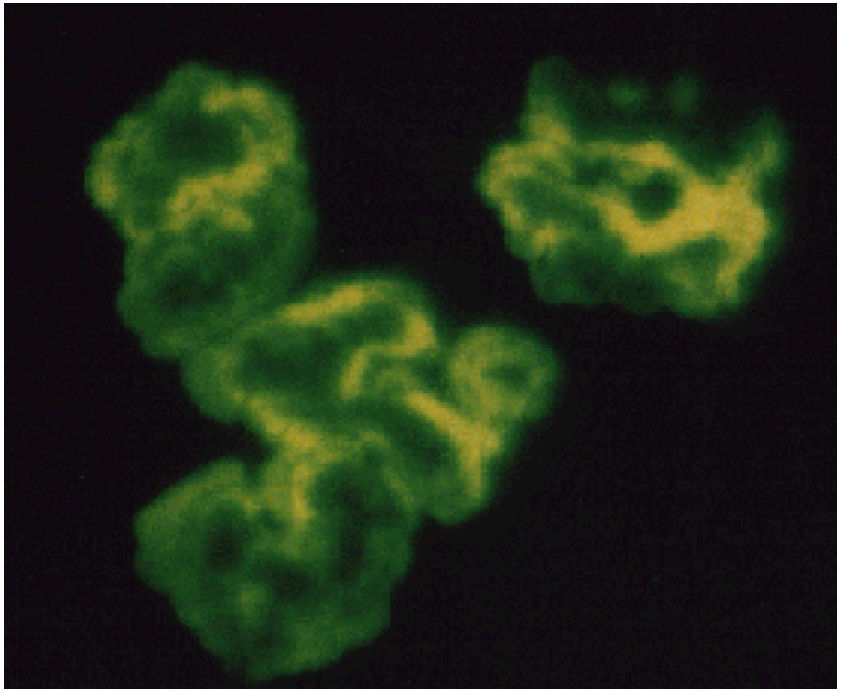

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Thank you