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Update on nomenclature and classification of vasculitis

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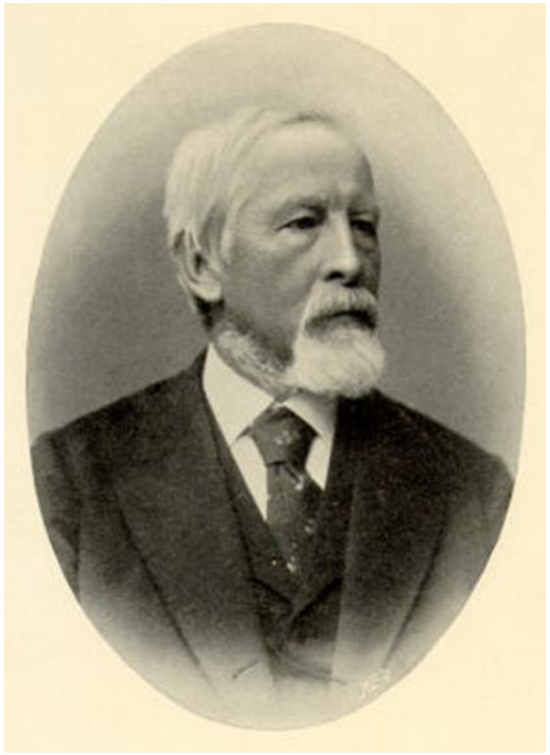
Adolph Kussmaul (1822 –1902)

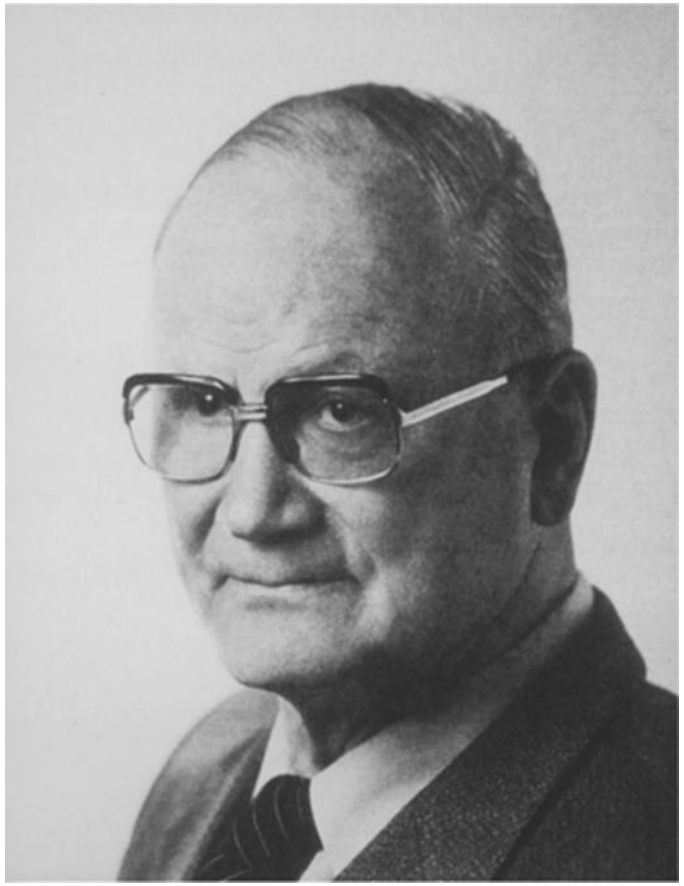


- **First Modern Case: "Periarteritis Nodosa"**
- In 1866 Kussmaul and Maier reported the case of a 27-year-old tailor who died during a month-long hospital stay.
- At autopsy, grossly visible nodules were present along the patient's medium-sized arteries.
- Kussmaul and Maier suggested the name "periarteritis nodosa"

Mikito Takayasu (1860 -1938)

- The first case of Takayasu's arteritis was described in 1908 by Japanese ophthalmologist Mikito Takayasu at the Annual Meeting of the Japan Ophthalmology Society.
- Takayasu described a peculiar "wreathlike" appearance of theblood vessels in the back of the eye (retina).
- Two Japanese physicians at the same meeting (Drs. Onishi and Kagoshima) also reported similar eye findings in patients whose wrist pulses were absent. It is now known that the blood vessel malformations that occur in the retina are an angiogenic response to the arterial narrowings in the neck, and that the absence of pulses noted in some patients occurs because of narrowings of the blood vessels to the arms.

Friedrich Wegener (1907–1990)



- German pathologist who is notable for his description of a rare disease.
- Although this disease was known before Wegener's description, since the 1950s it has been called by the name Wegener's granulomatosis

Summary of historical classification criteria for vasculitis

Author(s)/date	Key points
Zeek, 1954	First classification system, and has served as the basis for all subsequent classification systems
Alarçon-Segovia & Brown, 1964	Similar scheme as Zeek, but GPA and IgAV added
De Shazo, 1975	Similar to Zeek, with the addition of few subgroups
Gilliam & Smiley, 1976	Better appreciation of the degree of overlap in size of vessels involved between vasculitis subgroups
Fauci et al., 1978	Kawasaki's disease added to the vasculitis subtypes
Lie, 1994	Subdivisions into primary or secondary forms, as well as predominant vessel size involvement

Explanation of terminology

Term	Explanation
Diagnosis	The name of a disease
Definition	Disease processes present in any patient that justify assignment of the diagnosis (name)
Classification criteria	Observations that classify a specific patient into a standardized category for study
Diagnostic criteria	Observations that demonstrate or confidently predict the presence of the defining features of the disease in a specific patient

Example

Term	Explanation
Diagnosis	Polyarteritis nodosa
Definition	Necrotizing arteritis of medium or small arteries without glomerulonephritis or vasculitis in arterioles, capillaries, or venules, and not associated with ANCA
Classification criteria	Medium artery necrotizing arteritis seen on biopsy, negative ANCA, no MCLNS, and no evidence of glomerulonephritis
Diagnostic criteria	Medium artery aneurysm seen on imaging or necrotizing arteritis seen on biopsy, negative ANCA, no MCLNS, and no evidence of glomerulonephritis

Considerations in the Classifications of Systemic Vasculitis

- Size of predominant blood vessels affected
- Epidemiologic features
  - Age
  - Gender
  - Ethnic background
- Pattern of organ involvement
- Pathologic features
  - Granulomatous inflammation
  - Immune complex deposition versus pauci-immune histopathology
  - Linear staining along glomerular basement membrane
- Presence of ANCA, anti-GBM antibodies, or rheumatoid factor in serum
- Demonstration of a specific associated infection (hepatitis B or hepatitis C)

Current classification criteria

1990 American College of Rheumatology

1. Hunder GG, Bloch DA, Michel BA, Stevens MB, Arend WP, Calabrese LH, et al. The American College of Rheumatology 1990 criteria for the classification of ***giant cell arteritis***. Arthritis & Rheumatism 1990;33:1122–8.
2. Arend WP, Michel BA, Bloch DA, Hunder GG, Calabrese LH, Edworthy S, et al. The American College of Rheumatology 1990 criteria for the classification of ***Takayasu arteritis***. Arthritis & Rheumatism 1990;33:1129–34.
3. Leavitt RY, Fauci AS, Bloch DA, Michel BA, Hunder GG, Arend WP, et al. The American College of Rheumatology 1990 criteria for the classification of ***Wegener's granulomatosis***. Arthritis & Rheumatism 1990;33:1101–7.
4. Masi AT, Hunder GG, Lie JT, Michel BA, Bloch DA, Arend WP, et al. The American College of Rheumatology 1990 criteria for the classification of ***Churg–Strauss syndrome*** (allergic granulomatosis and angiitis). Arthritis & Rheumatism 1990;33:.١٠٠–١٠٩٤
5. Lightfoot Jr RW, Michel BA, Bloch DA, Hunder GG, Zvaifler NJ, McShane DJ, et al. The American College of Rheumatology 1990 criteria for the classification of ***polyarteritis nodosa***. Arthritis & Rheumatism 1990;33:1088–93.
6. Mills JA, Michel BA, Bloch DA, Calabrese LH, Hunder GG, Arend WP, et al. The American College of Rheumatology 1990 criteria for the classification of ***Henoch–Schönlein purpura***. Arthritis & Rheumatism 1990;33:1114–21.
7. Calabrese LH, Michel BA, Bloch DA, Arend WP, Edworthy SM, Fauci AS, et al. The American College of Rheumatology 1990 criteria for the classification of ***hypersensitivity vasculitis***. Arthritis & Rheumatism 1990;33:1108–13.

American College of Rheumatology Classification Criteria for Giant Cell Arteritis

For purposes of classification, a patient with vasculitis is said to have giant cell (temporal) arteritis if at least three of these five criteria are present. The presence of any three or more criteria yields a sensitivity of 93.5% and a specificity of 91.2%

Criterion[ ]	Definition
Age at disease onset ≥50 yr	Development of symptoms or findings beginning at age 50 or older
New headache	New onset or new type of localized pain in the head
Temporal artery abnormality	Temporal artery tenderness to palpation or decreased pulsation, unrelated to arteriosclerosis of cervical arteries
Elevated erythrocyte sedimentation rate (ESR)	ESR ≥50 mm/hr by the Westergren method
Abnormal artery biopsy	Biopsy specimen with artery showing vasculitis characterized by a predominance of mononuclear cell infiltration or granulomatous inflammation, usually with multinucleated giant cells

Hunder GG, Bloch DA, Michel BA, et al: The American College of Rheumatology 1990 criteria for the classification of giant cell arteritis. Arthritis Rheum 33:1125, 1990

American College of Rheumatology Classification Criteria for Takayasu's Arteritis

The presence of three or more of the six criteria is sensitive (91%) and specific (98%) for the diagnosis of Takayasu's arteritis

CRITERION	DEFINITION
Age at disease onset ≤40 years	Development of symptoms or findings related to Takayasu arteritis at age ≤ 40 years
Claudication of extremities	Development and worsening of fatigue and discomfort in muscles of one or more extremity while in use, especially the upper extremities
Decreased brachial artery pulse	Decreased pulsation of one or both brachial arteries
BP difference >10 mm Hg	Difference of >10 mm Hg in systolic blood pressure between arms
Bruit over subclavian arteries or aorta	Bruit audible on auscultation over one or both subclavian arteries or abdominal aorta
Arteriogram abnormality	Arteriographic narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the proximal upper or lower extremities, not due to arteriosclerosis, fibromuscular dysplasia, or similar causes; changes usually focal or segmental

American College of Rheumatology 1990 criteria for the classification of Takayasu arteritis. Arthritis Rheum 33:1129, 1990.

American College of Rheumatology Classification Criteria for wegener's granulomatosis

For purposes of classifi cation, a patient shall be said to have Wegener's granulomatosis if at least two of these four criteria are present. The presence of any two or more criteria yields a sensitivity of 88.2% and a specifi city of 92.0%.

CRITERION	DEFINITION
Nasal or oral inflammation	Development of painful or painless oral ulcers or purulent or bloody nasal discharge
Abnormal chest radiograph	Chest radiograph showing the presence of nodules, fixed infiltrates, or cavities
Urinary sediment	Microhematuria (> 5 red blood cells per high power field) or red cell casts in urine sediment
Granulomatous inflammation on biopsy	Histologic changes showing granulomatous inflammation within the wall of an artery or in the perivascular or extravascular area (artery or arteriole

Leavitt RY, Fauci AS, Bloch DA, et al. The American College of Rheumatology 1990 criteria for the classifi cation of Wegener's granulomatosis. Arthritis Rheum 1990;33:1101–1107

American College of Rheumatology Classification Criteria for churg–strauss syndrome

For classifi cation purposes, a patient shall be said to have Churg–Strauss syndrome if at least four of these six criteria are positive. The presence of any four or more of the six criteria yields a sensitivity of 85% and a specifi city of 99.7%.

CRITERION	DEFINITION
Asthma	History of wheezing or diffuse high-pitched rales on expiration
Eosinophilia	Eosinophilia >10% on white blood cell differential count
Mononeuropathy or polyneuropathy	Development of mononeuropathy, multiple mononeuropathies, or polyneuropathy (i.e., glove/stocking distribution) attributable to a systemic vasculitis
Pulmonary infiltrates, nonfixed	Migratory or transitory pulmonary infiltrates on radiographs (not including fixed infiltrates), attributable to a systemic vasculitis
Paranasal sinus abnormality	History of acute or chronic paranasal sinus pain or tenderness or radiographic opacification of the paranasal sinuses
Extravascular eosinophils	Biopsy including artery, arteriole, or venule, showing accumulations of eosinophils in extravascular areas

Masi AT, Hunder GG, Lie JT, et al. The American College of Rheumatology 1990 criteria for the classifi cation of Churg–Strauss syndrome (allergic granulomatosis and angiitis). Arthritis Rheum 1990;33:1094–1100,

American College of Rheumatology Classification Criteria for polyarteritis nodosa

For classifi cation purposes, a patient shall be said to have polyarteritis nodosa if at least 3 of these 10 criteria are present. The presence of any three or more criteria yields a sensitivity of 82.2% and a specifi city of 86.6%.

CRITERION	DEFINITION
Weight loss ≥4 kg	Loss of 4 kg or more of body weight since illness began, not due to dieting or other factors
Livedo reticularis	Mottled reticular pattern over the skin of portions of the extremities or torso
Testicular pain or tenderness	Pain or tenderness of the testicles, not due to infection, trauma, or other causes
Myalgias, weakness, or legtenderness	Diffuse myalgias (excluding shoulder and hip girdle) or weakness of muscles or tenderness of leg muscles
Mononeuropathy orpolyneuropathy	Development of mononeuropathy, multiple mononeuropathies, or polyneuropathy
Diastolic BP >90 mm Hg	Development of hypertension with the diastolic BP higher than 90 mm Hg
Elevated BUN or creatinine	Elevation of BUN >40 mg/dL or creatinine >1.5 mg/dL, not due to dehydration or obstruction
Hepatitis B virus	Presence of hepatitis B surface antigen or antibody in serum
Arteriographic abnormality	Arteriogram showing aneurysms or occlusions of the visceral arteries, not due to arteriosclerosis fibromuscular dysplasia, or other non-inflammatory causes
Biopsy of small or medium-sized	Histologic changes showing the presence of granulocytes or artery containing polymorphonuclear leukocytes and mononuclear leukocytes in the artery wall

Lightfoot RW Jr, Michel BA, Bloch DA, et al. The American College of Rheumatology 1990 criteria for the classifi cation of polyarteritis nodosa. Arthritis Rheum 1990;33:1088–1093

American College of Rheumatology Classification Criteria for henoch–schonlein purpura

For purposes of classifi cation, a patient shall be said to have Henoch–Schonlein purpura if at least two of these four criteria are present. The presence of any two or more criteria yields a sensitivity of 87.1% and a specifi city of 87.7%.

CRITERION	DEFINITION
Palpable purpura	Slightly raised “palpable” hemorrhagic skin lesions, not related to thrombocytopenia
Age at disease onset	Patient 20 years or younger at onset of first symptoms
Bowel angina	Diffuse abdominal pain, worse after meals, or the diagnosis of bowel ischemia, usually including bloody diarrhea
Wall granulocytes on biopsy	Histologic changes showing granulocytes in the walls of arterioles or venules

Mills JA, Michel BA, Bloch DA, et al. The American College of Rheumatology 1990 criteria for the classifi cation of Henoch–Schonlein purpura. Arthritis Rheum 1990;33:1114–1121,,

American College of Rheumatology Classification Criteria for hypersensitivity vasculitis

For purposes of classifi cation, a patient shall be said to have hypersensitivity vasculitis if at least three of these fi ve criteria are present. The presence of any three or more criteria yields a sensitivity of 71.0% and a specifi city of 83.9%.

CRITERION	DEFINITION
Age at disease onset >16 years	Development of symptoms after age 16
Medication at disease onset	Medication was taken at the onset of symptoms that may have been a precipitating factor
Palpable purpura	Slightly elevated purpuric rash over one or more areas of the skin; does not blanch with pressure and is not related to thrombocytopenia
Maculopapular rash	Flat and raised lesions of various sizes over one or more areas of the skin
Biopsy including arteriole and venule	Histologic changes showing granulocytes in a perivascular or extravascular location

Calabrese LH, Michel BA, Bloch DA, et al. The American College of Rheumatology 1990 criteria for the classifi cation of hypersensitivity vasculitis. Arthritis Rheum 1990;33:1108–1113,

Summary of ACR criteria and their limitations

Type of Vasculitis	Sensitivity	Specificity	Limitations
GCA	93.5%	91.2%	Temporal artery biopsy is an important diagnostic tool but is not an obligatory criterion
TAK	90.5%	97.7%	Newer imaging modalities, such as CT PET maybe useful, but are not included
GPA	88.2%	92%	No clear discrimination between GPA and MPA, or other mimics of GPA. Does not incorporate ANCA test.
EGPA	85%	99.7%	No inclusion of common features such as cardiac manifestations and rash. Does not incorporate ANCA test.
PAN	82.2%	86.6%	No absolute requirement for arteriography, or biopsy findings. No clear discrimination between PAN and MPA.
IgA V	87.1%	87.7%	Do not distinguish between IgAV from allergic reactions, or infectious related purpura. Common features; arthritis and nephritis are excluded. Age set as important criteria, but almost 30% of patients were above the age of 20.
HSV	71%	83.9%	Difficult to distinguish from IgAV
MPA	-	-	Not recognized by ACR

Paediatric vasculitis

- In 2006, EULAR and the Paediatric Rheumatology European Society (PReS) produced consensus criteria for the classification of childhood vasculitis.
- The EULAR/PReS guidelines use vessel size to categorise vasculitis based on the CHCC definitions.
- The criteria have been validated using a retrospective and prospective web-based database of children with primary vasculitis first diagnosed before the age of 18.
- A total of 1398 children were enrolled. An expert consensus panel, who were blinded to the primary physician diagnosis, classified a representative sample of 280 of the cases. Sensitivity ranges from 89.6%to 100% and specificity ranges from 87% to 99.9% for the classification of vasculitis

EULAR/PreS classification of childhood vasculitis

Vessel size	Vasculitis subtypes
Large	TAK
Medium	Childhood PAN Cutaneous Polyarteritis Kawasaki disease
Small	**Granulomatous** GPA EGPA **Non-granulomatous** MPA IgA vasculitis Isolated cutaneous leucocytoclastic vasculitis Hypocomplementaemic urticarial vasculitis
Other	Behcet's disease Secondary vasculitis Vasculitis associated with connective tissue diseases Isolated vasculitis of the central nervous system Cogan syndrome Unclassified

Ann Rheum Dis, Ozen S, Ruperto N, Dillon et al., 65, 936–941, 2006

Current definitions of the vasculitides

Chapel Hill Consensus Conference CHCC Nomenclature (1994)

- In1994, a group of experts convened at the CHCC with the goal of determining names and definitions for the common systemic vasculitides, with efforts made to use already widely accepted terms.

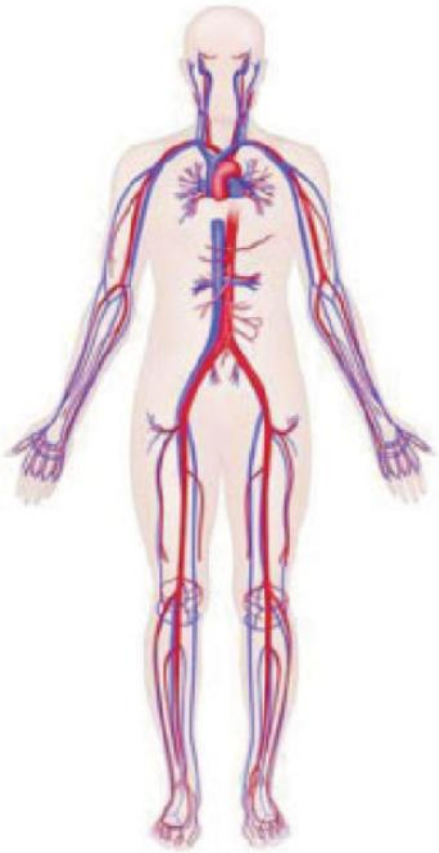
Chapel Hill Consensus Conference CHCC Nomenclature (1994)

- This was as a reflection of the ACR criteria, where the categorization of patients had been determined by physician judgment; however, the clinicians giving their judgment had not been provided with a strict uniform definition for each vasculitis subtype.
- The panel of multidisciplinary experts emphasized that their objectives were not to determine the classification of vasculitis, nor to provide diagnostic criteria.

Chapel Hill Consensus Conference CHCC Nomenclature (1994)

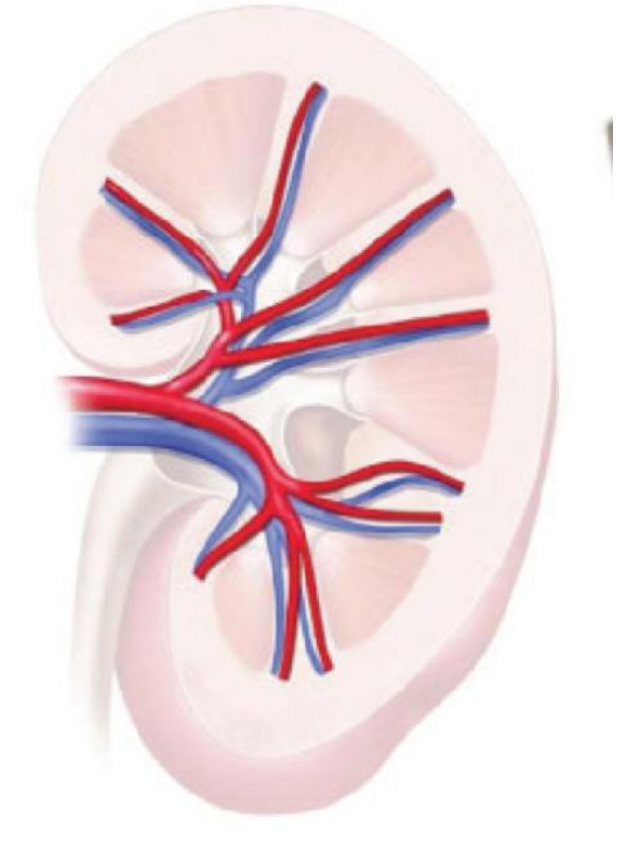
- **Ten** vasculitis syndromes were defined using clinical and histological criteria and these were grouped according to vessel size.

Large vessels



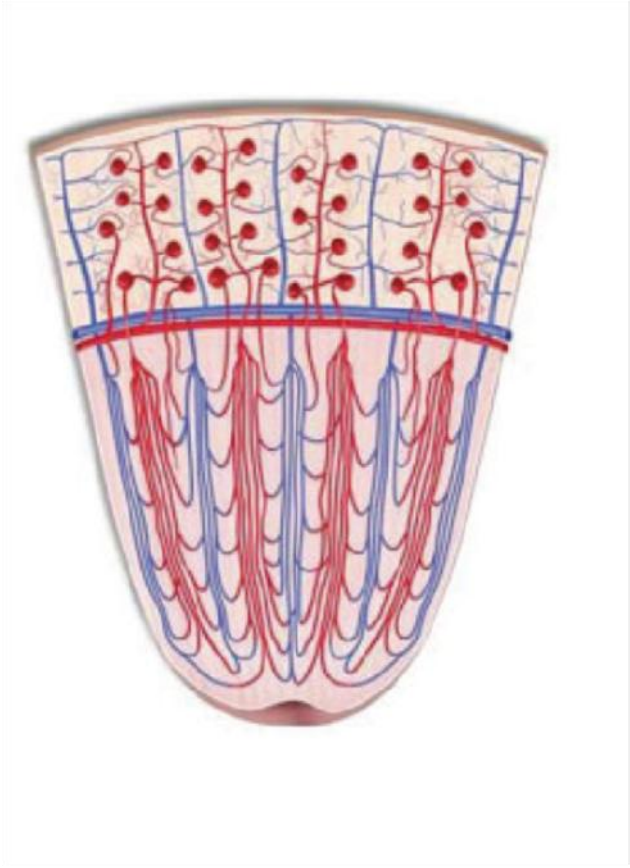
- Large vessels are the aorta and its major branches and the analogous veins.

Medium vessels



- Medium vessels are the main visceral arteries and veins and their initial branches.
- (smaller than the major aortic branches yet still large enough to contain four elements: (1) an intima, (2) a continuous internal elastic lamina, (3) a muscular media, and (4) an adventitia.)

Small vessels



- Small vessels are intraparenchymal arteries, arterioles, capillaries, venules, and veins.
- (below macroscopic disease, includes capillaries, postcapillary venules, and arterioles. Such vessels all are typically less than 500 ε m in outer diameter)

Chapel Hill Consensus Conference CHCC Nomenclature (1994)

- The CHCC, unlike the ACR criteria, recognised MPA as a distinct entity.
- The CHCC definitions, in particular, addressed the differences between classical PAN and MPA.
- The CHCC incorporated ANCA testing into their definitions in contrast to the ACR criteria.
- A **limitation of the CHCC criteria** is that the definitions are based on histological criteria.

Updated CHCC 2012 nomenclature

- Notable difference within this update:
  - The definition of **PAN**, which now includes a **negative ANCA** in its definition, helping to clearly distinguish it from MPA.
  - The greatest change in nomenclature has been within the predominant small-vessel vasculitides group, with subdivision into those with a paucity of vessel-wall immunoglobulin (**ANCA-associated vasculitides**), and those with prominent vessel-wall immunoglobulin (**immune complex small-vessel vasculitides**).
  - Additionally anti-glomerular basement membrane disease and hypocomplementaemic urticarial vasculitis have now been incorporated.

Updated CHCC 2012 nomenclature

- The definition of **MPA** has been refined with the addition of the statement "granulomatous inflammation is absent," presumably to help differentiate the respiratory tract involvement in MPA compared with GPA.
- **Limited GPA** has been recognized in the updated CHCC 2012, and it is suggested that where there is good evidence of clinical and pathological features of GPA confined to the respiratory tract, especially if associated with a positive ANCA result, they should be defined within this category.
- The 2012 CHCC has incorporated other vasculitides now well recognised and includes definitions for Behçet's disease and Cogan's syndrome.
- Furthermore, there has been effort made towards incorporating and recognising other single-organ disease and secondary vasculitides, which are incorporated under the definitions of '**vasculitis associated with systemic disease**' and 'vasculitis associated with probable aetiology'.

Names for vasculitides adopted by the 2012 International Chapel Hill Consensus Conference on the Nomenclature of Vasculitides

- **Large vessel vasculitis (LVV)**
  - Takayasu arteritis (TAK)
  - Giant cell arteritis (GCA)
- **Medium vessel vasculitis (MVV)**
  - Polyarteritis nodosa (PAN)
  - Kawasaki disease (KD)
- **Small vessel vasculitis (SVV)**
  - Antineutrophil cytoplasmic antibody (ANCA)–associated vasculitis (AAV)
    - Microscopic polyangiitis (MPA)
    - Granulomatosis with polyangiitis (Wegener's) (GPA)
    - Eosinophilic granulomatosis with polyangiitis (Churg-Strauss) (EGPA)
  - Immune complex SVV
    - Anti–glomerular basement membrane (anti-GBM) disease
    - Cryoglobulinemic vasculitis (CV)
    - IgA vasculitis (Henoch-Scho¨nlein) (IgAV)
    - Hypocomplementemic urticarial vasculitis (HUV) (anti-C1q vasculitis)

Names for vasculitides adopted by the 2012 International Chapel Hill Consensus Conference on the Nomenclature of Vasculitides

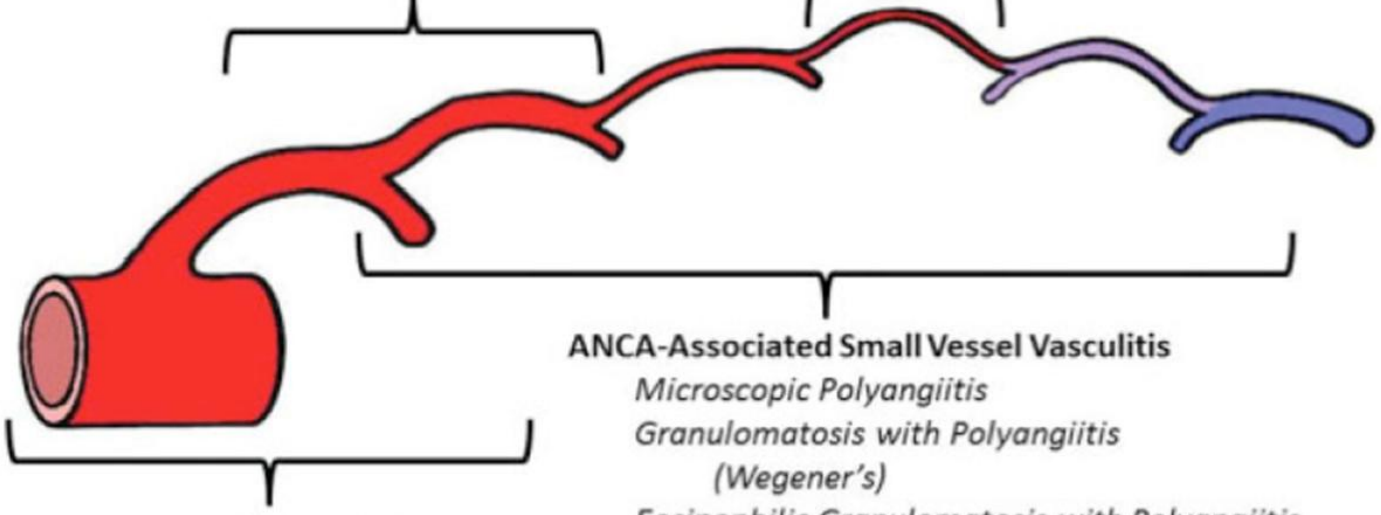
- **Variable vessel vasculitis (VVV)**
  - Behcet's disease (BD)
  - Cogan's syndrome (CS)
- **Single-organ vasculitis (SOV)**
  - Cutaneous leukocytoclastic angiitis
  - Cutaneous arteritis
  - Primary central nervous system vasculitis
  - Isolated aortitis
  - Others
- **Vasculitis associated with systemic disease**
  - Lupus vasculitis
  - Rheumatoid vasculitis
  - Sarcoid vasculitis
  - Others

Names for vasculitides adopted by the 2012 International Chapel Hill Consensus Conference on the Nomenclature of Vasculitides

- **Vasculitis associated with probable etiology**
  - Hepatitis C virus–associated cryoglobulinemic vasculitis
  - Hepatitis B virus–associated vasculitis
  - Syphilis-associated aortitis
  - Drug-associated immune complex vasculitis
  - Drug-associated ANCA-associated vasculitis
  - Cancer-associated vasculitis
  - Other

Classification of Vasculitis

Chapel Hill Consensus Criteria
Nomenclature update 2012



Immune Complex Small Vessel Vasculitis
Cryoglobulinemic Vasculitis
IgA Vasculitis (Henoch-Schönlein)
Hypocomplementemic Urticarial Vasculitis (Anti-C1q Vasculitis)

Medium Vessel Vasculitis
Polyarteritis Nodosa
Kawasaki Disease

Anti-GBM Disease

ANCA-Associated Small Vessel Vasculitis
Microscopic Polyangiitis
Granulomatosis with Polyangiitis (Wegener's)
Eosinophilic Granulomatosis with Polyangiitis (Churg-Strauss)

Large Vessel Vasculitis
Takayasu Arteritis
Giant Cell Arteritis

Revised CHCC 2012 nomenclature

- Name: MPA
- Definition:
  - Necrotizing vasculitis, with few or no immune deposits, predominantly affecting small vessels i.e. capillaries, venules, or arterioles).
  - Necrotizing arteritis involving small and medium arteries may be present.
  - Necrotizing glomerulonephritis is very common. Pulmonary capillaritis often occurs.
  - Granulomatous inflammation is absent

Revised CHCC 2012 nomenclature

- Name: GPA (*ANCA-associated vasculitis (AAV)*)
- Definition:
  - Necrotizing granulomatous inflammation usually involving the upper and lower respiratory tract, and necrotizing vasculitis affecting predominantly small to medium vessels (e.g., capillaries, venules, arterioles, arteries and veins).
  - Necrotizing glomerulonephritis is common.

Revised CHCC 2012 nomenclature

- Name: Limited GPA (*ANCA associated vasculitis (AAV)*)
- Definition:
- Limited expressions of GPA occur, especially disease confined to the upper or lower respiratory tract , or the eye.
- These patients may have no identifiable evidence of systemic vasculitis.

Revised CHCC 2012 nomenclature

- Name: EGPA (*ANCA-associated vasculitis (AAV)*)
- Definition:
  - Eosinophil-rich and necrotizing granulomatous inflammation often involving the respiratory tract, and necrotizing vasculitis predominantly affecting small to medium vessels, and associated with asthma and eosinophilia.
  - ANCA is more frequent when glomerulonephritis is present.

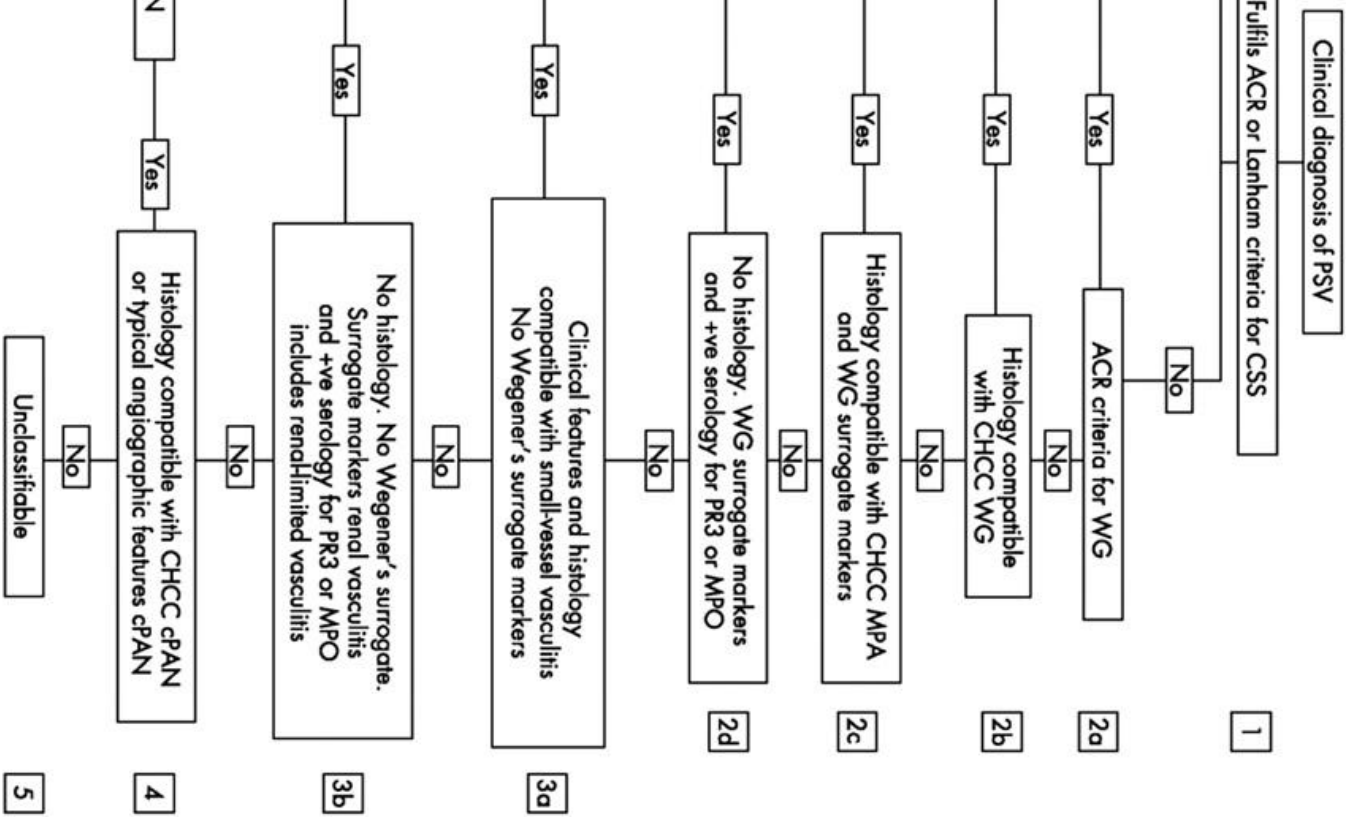
Revised CHCC 2012 nomenclature

- Name: PAN
- Definition:
- Necrotizing arteritis of medium or small arteries without glomerulonephritis or vasculitis in arterioles, capillaries, or venules; and not associated with ANCA

Application of current classification criteria and definitions

- Watts et al. developed an algorithm for the purpose of categorising patients with GPA, MPA, EGPA and PAN for epidemiological studies in 2006.
- The algorithm combined the ACR and 1994 CHCC tools, included ANCA testing and other surrogate markers such as radiographic evidence of fixed pulmonary infiltrates, nodules or cavitations present for more than 1 month as symptoms suggestive of granulomatous disease.
- It also incorporates Lanham's criteria for EGPA.

Watts et al. classification algorithm. Reproduced from [Annals Rheumatic Diseases, Watts R, Lane S, Hanslik T et al., 66, 222–227, 2007] with permission from BMJ Publishing Group Ltd.



Problems

- At present:
  - There remains major controversy, and incompatibility between the ANCA-associated vasculitides (AAV): Wegener's granulomatosis (WG), microscopic polyangiitis (MPA), and Churg Straus Syndrome (CSS), as well as polyarteritis nodosa (PAN) in the current classification criteria and disease definitions.
  - The classification criteria for the large vessel vasculitides (giant cell arteritis (GCA) and Takayasu's arteritis (TAK)are dated and considered not fit for purpose by experts in the field.
  - There are currently no diagnostic criteria for primary systemic vasculitis.

Solution

- A new validated set of classification criteria for the primary systemic vasculitides.
- A validated set of diagnostic criteria for the primary systemic vasculitides.

Diagnostic and Classification Criteria in Vasculitis Study (DCVAS)

From 2010 - 2013

DCVAS

ACR/EULAR endorsed study to develop classification and diagnostic criteria for primary systemic vasculitis

Chief Investigators

Prof Raashid Luqmani, University of Oxford

Prof Peter Merkel, University of Pennsylvania

Prof Richard Watts, University of East Anglia

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- Vasculitis UK
- ClinicalTrials.gov
- UKCRN Portfolio

Publications

ACR/EULAR endorsed study to develop classification and diagnostic criteria for primary systemic vasculitis

Site enrolment has been extended until December 2014
Last scheduled patient follow-up December 2015

DCVAS has recruited 2551 patients
from 101 sites worldwide

We aim to increase recruitment to the comparator group.
Please see the **Recruitment Guide** for further information.

New sites joining DCVAS in last quarter

- York Hospital, UK
- Dartmouth-Hitchcock Medical Centre, USA
- Gartnavel Hospital, Glasgow, UK
- Nizam's Institute of Medical Sciences, Hyderabad, India
- Karolinska Institute, Stockholm, Sweden

Thank you to all sites for continuing to support DCVAS.

Study Objective

Develop and validate diagnostic and classification criteria for systemic vasculitis which can be used in daily clinical practice and for use in clinical trials .

Recruitment Target :

␣ ¬ ⁄patients from > ʻ ¬ sites

Study population

¸ ⁄ ⁄patients over / ⁼ years with a new or an established diagnosis of vasculitis

/ ¬ ⁄patients over / ⁼ years with a similar presentation to vasculitis, but an alternative diagnosis

Study Start Date	January 2011
Estimated Completion Date	December 2015
Estimated date last patient recruited	June 2015