



New ACR-EULAR classification criteria for ANCA-associated vasculitis

Lead Investigators

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Peter A. Merkel, University of Pennsylvania  
Richard Watts, University of East Anglia



Disclosures

- Ravi Suppiah – None
- Peter Grayson – None
- Cristina Ponte – None
- Jo Robson – None
- Richard Watts – None
- Peter Merkel – None
- Raashid Luqmani – None

Evidence based

Prior Classification/Nomenclature Schemes in Vasculitis

1. Leavitt RY et al. The American College of Rheumatology 1990 criteria for the classification of Wegener's granulomatosis. *Arthritis Rheum* 1990.
2. Masi AT et al. The American College of Rheumatology 1990 criteria for the classification of Churg-Strauss syndrome (allergic granulomatosis and angiitis). *Arthritis Rheum* 1990.
3. Lightfoot RW et al. The American College of Rheumatology 1990 criteria for the classification of polyarteritis nodosa. *Arthritis Rheum* 1990.
4. Jennette JC et al. Nomenclature of systemic vasculitides. Proposal of an international consensus conference. *Arthritis Rheum* 1994
5. Jennette JC et al. 2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. *Arthritis Rheum* 2013.



Acknowledgements

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DCVAS is *funded* by the American College of Rheumatology, the European League Against Rheumatism and the Vasculitis Foundation



Introduction to the ACR-EULAR classification criteria for ANCA-associated vasculitis symposium

- Why and how?  
Ravi Suppiah, Peter Grayson, Cristina Ponte
- Results of 2017 Provisional classification criteria for Granulomatosis with Polyangiitis (GPA)  
Joanna Robson
- Implications for Clinical Practice / Q&A  
Raashid Luqmani, Peter Merkel, Richard Watts



New ACR-EULAR classification criteria for ANCA-associated vasculitis: *why and how?*

Presenters

Ravi Suppiah, Auckland District Health Board  
Peter Grayson, United States National Institutes of Health  
Cristina Ponte, Lisbon Academic Medical Centre



Overview of this section

1. Why do we need new criteria?

2. How did we develop new criteria?

1. Why?

Example case 1

- 45 year-old female
- Unwell for 2 weeks
- Fevers to 38°C (100°F)
- Weight loss of 4kg (8.8lb)
- Left foot and right wrist drop

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1. Why?

Example case 1

- 45 year-old female
- Unwell for 2 weeks
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- Scleritis



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1. Why?

Example case 1

- 45 year-old female
- Unwell for 2 weeks
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- Weight loss of 4kg (8.8lb)
- Left foot and right wrist drop
- Scleritis
- Progressive breathlessness for 4 months



Blunting of costophrenic angle + opacification

DCVAS

1. Why?

Example case 1

- 45 year-old female
- Unwell for 2 weeks
- Fevers to 38°C (100°F)
- Weight loss of 4kg (8.8lb)
- Left foot and right wrist drop
- Scleritis
- Progressive breathlessness for 4 months



HRCT Chest      Fibrotic NSIP

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1. Why?

Example case 1 - Investigations

- Hb 101 g/L (10.1g/dL)
- Eosinophil count normal
- CRP 180 mg/L
- Creatinine 360 umol/L (3.5 mg/dL)



Red cell casts in urine



Pauci immune crescentic GN on renal biopsy



pANCA positive on IF  
MPO-ANCA positive by ELISA

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1. Why?

Example case 1 - How would you classify this case?

- A. Polyarteritis nodosa (PAN)
- B. Eosinophilic granulomatosis with polyangiitis (EGPA)
- C. Granulomatosis with polyangiitis (GPA)
- D. Microscopic polyangiitis (MPA)
- E. Other type of vasculitis

DCVAS

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1. Why?

Example case 1 - How would you classify this case?

No microscopic polyangiitis (MPA)  
in the 1990 ACR classification criteria

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1. Why?

Example case 1 - 1990 ACR Classification – GPA  
(Wegener's)

- 1. Nasal or oral inflammation
- 2. Abnormal chest radiograph
- 3. Urinary sediment
- 4. Granulomatous inflammation on biopsy

Requires 2 of  
the 4 criteria to  
be classified as  
GPA

Leavitt RY et al. Arthritis Rheum 1990

DCVAS

1. Why?

Example case 1 - 1990 ACR Classification – GPA  
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- 1. Nasal or oral inflammation
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Requires 2 of  
the 4 criteria to  
be classified as  
GPA

Patient meets criteria for GPA

Leavitt RY et al. Arthritis Rheum 1990

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1. Why?

Example case 1 - 1990 ACR Classification - PAN

- 1. Weight loss ≥4 kg
- 2. Livedo reticularis
- 3. Testicular pain or tenderness
- 4. Myalgias, weakness, or leg tenderness
- 5. Mononeuropathy or polyneuropathy
- 6. Diastolic BP >90 mmHg
- 7. Elevated BUN or creatinine
- 8. Hepatitis B virus
- 9. Arteriographic abnormality
- 10. Biopsy of small or medium sized artery containing PMN

Requires 3 of  
the 10 criteria  
to be classified  
as PAN

Lightfoot RW et al. Arthritis Rheum 1990

DCVAS

1. Why?

Example case 1 - 1990 ACR Classification - PAN

- 1. Weight loss  $\geq 4$  kg
- 2. Livedo reticularis
- 3. Testicular pain or tenderness
- 4. Myalgias, weakness, or leg tenderness
- 5. Mononeuropathy or polyneuropathy
- 6. Diastolic BP  $>90$  mmHg
- 7. Elevated BUN or creatinine
- 8. Hepatitis B virus
- 9. Arteriographic abnormality
- 10. Biopsy of small or medium sized artery containing PMN

Requires 3 of the 10 criteria to be classified as PAN

Patient meets criteria for PAN

Lightfoot RW et al. Arthritis Rheum 1990

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1. Why?

Example case 2

- 73 year old female
- Unwell for 2 months
- Fevers to  $38^{\circ}\text{C}$  ( $100^{\circ}\text{F}$ )
- Night sweats
- Weight loss of 3kg (6.6lb)
- Myalgia and arthralgia
- Cough

DCVAS

1. Why?

Example case 2

- 73 year old female
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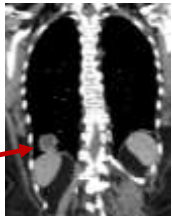
Normal CXR

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1. Why?

Example case 2

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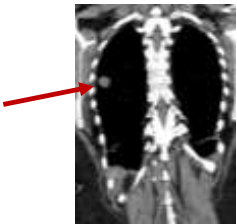
CT Chest—same day as CXR

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1. Why?

Example case 2

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CT Chest—same day as CXR

DCVAS

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CT Chest—same day as CXR

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1. Why?

Example case 2

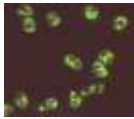
- Sensation of blocked left ear with reduced hearing
- CRP 82 mg/L
- Urine normal
- Blood cultures negative
- Eosinophil count normal

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1. Why?

Example case 2

- Sensation of blocked left ear with reduced hearing
- CRP 82 mg/L
- Urine normal
- Blood cultures negative
- Eosinophil count normal



cANCA positive on IF  
PR3-ANCA strongly positive on ELISA

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1. Why?

Example case 2

- Sensation of blocked left ear with reduced hearing
- CRP 82 mg/L
- Urine normal
- Blood cultures negative
- Eosinophil count normal



Opacified right sphenoid sinus

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1. Why?

Example case 2 - How would you classify this case?

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1. Why?

Example case 2 - 1990 ACR Classification – GPA (Wegener's)

- 1. Nasal or oral inflammation
- 2. Abnormal chest radiograph
- 3. Urinary sediment
- 4. Granulomatous inflammation on biopsy

Requires 2 of the 4 criteria to be classified as GPA

Leavitt RY et al. Arthritis Rheum 1990

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## 1. Why?

### Example case 2 - 1990 ACR Classification – GPA (Wegener's)

1. Nasal or oral inflammation
2. Abnormal chest radiograph
3. Urinary sediment
4. Granulomatous inflammation on biopsy

**Requires 2 of the 4 criteria to be classified as GPA**

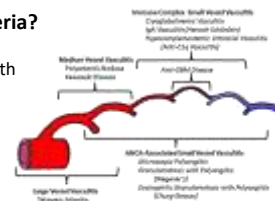
Patient does **NOT** meet criteria for GPA

Leavitt RY et al. *Arthritis Rheum* 1990

## 1. Why?

## Why do we need new criteria?

- Old criteria not consistent with current disease definitions
- No MPA
- ANCA
- Improved imaging



2012 Chapel Hill Consensus Conference

Jennette JC et al. *Arthritis Rheum* 2013

## 1. Why?

### Why do we need new criteria?

- Old criteria not consistent with current disease definitions
- No MPA
- ANCA
- Improved imaging



## TIME FOR AN UPDATE

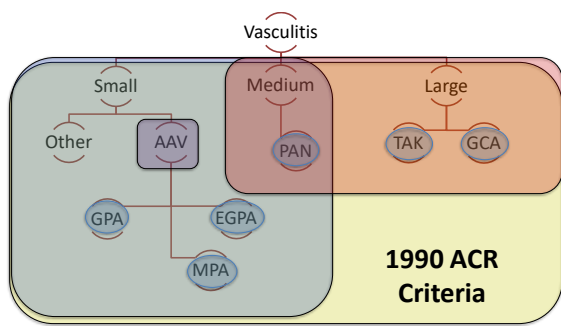


## 2. How?

## Background to the methodology

### Classification vs Diagnostic Criteria

- Classification criteria enable stratification of homogenous groups to standardize recruitment into clinical trials and related studies
- Distinguish amongst different types of vasculitis
  - No inclusion of vasculitis mimics as comparators



**133 sites in 33 countries**

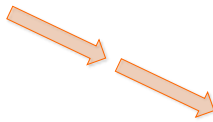
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## 2. How?

### Methodology Advancements

#### Number of Criteria Rule



Weighted Criterion  
with Threshold Score

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## 2. How?

### 1987 ACR Rheumatoid Arthritis Classification Criteria

Table 5. The 1987 revised criteria for the classification of rheumatoid arthritis (traditional format)\*

Criteria	Definition
1. Morning stiffness	Morning stiffness in and around the joints, lasting at least 1 hour before maximal improvement
2. Arthritis of 1 or more joint areas	Arthritis of 1 or more joint areas (swelling or fluid and bony enlargement) observed by a physician. The 14 possible areas are right or left PIP, MCP, wrist, elbow, knee, ankle, and MTP joints
3. Arthritis of hand joints	At least 1 area swollen (as defined above) in a wrist, MCP, or PIP joint
4. Symmetric arthritis	Bilateral involvement of the same joint areas (as defined) in 1 or both sides of the body
5. Rheumatoid nodules	Subcutaneous involvement of PIPs, MCs, or MTPs in asymptomatic (without clinical symptoms) rheumatoid nodules, only bony prominences, or extensor tendons, or in subcutaneous regions, observed by a physician
6. Serum rheumatoid factor	Demonstration of abnormal amounts of serum rheumatoid factor by any method for which the result has been positive in >70% of normal control subjects
7. Radiographic changes	Radiographs (anterior-posterior of rheumatoid arthritis) at posteroanterior hand and wrist radiographs, which must include wrists or metacarpal base dislocations located in or near midline adjacent to the involved joint (metacarpal changes alone do not qualify)

\* For classification purposes, a patient shall be said to have rheumatoid arthritis if 4 or more of these 7 criteria, Criteria 1 through 6 must have been present for at least 6 weeks. Patients with 1 clinical diagnosis are not included. Diagnosis in classic, definite, or probable rheumatoid arthritis is not to be made. See Table 1 for definitions of abbreviations.

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## 2. How?

### 2010 ACR/EULAR Rheumatoid Arthritis Classification Criteria

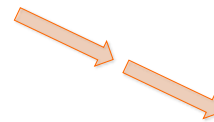
Criteria	Score
1. Morning stiffness (11 items or more)	1
2. Arthritis of 1 or more joint areas	1
3. Arthritis of hand joints	1
4. Symmetric arthritis	1
5. Rheumatoid nodules	1
6. Serum rheumatoid factor	1
7. Radiographic changes	1
8. Erythrocyte sedimentation rate (ESR)	1
9. C-reactive protein (CRP)	1
10. Hemoglobin (Hb)	1
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## 2. How?

### Establishing the Gold Standard

#### Submitting Physician Diagnosis



Expert Panel Review

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## 2. How?

RESEARCH IN RHEUMATOLOGY  
Vol. 36, No. 10, October 2016  
DOI: 10.1093/rheumatology/kew016

#### SPECIAL ARTICLE

### 2016 American College of Rheumatology/European League Against Rheumatism Classification Criteria for Primary Sjögren's Syndrome

A Consensus and Data-Driven Methodology Involving Three International Patient Cohorts

Caroline H. Rasmussen,<sup>1</sup> Stephen C. Richman,<sup>1</sup> Raphaela Stern,<sup>2</sup> Lindsay A. Czirak,<sup>3</sup> Marc Lohmeyer,<sup>4</sup> Thomas M. Lipton,<sup>5</sup> Astrid Rasmussen,<sup>6</sup> Hui Szelebi,<sup>7</sup> Chantal Vissel,<sup>8</sup> Bruce J. Goldstein,<sup>9</sup> Xavier Mariette,<sup>10</sup> and the International Sjögren's Syndrome Criteria Working Group

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## 2. How?

### DCVAS Classification criteria methodology: 3 phases

1. Case selection	<ul style="list-style-type: none"> <li>Clinical vignette expert panel review</li> <li>Avoid circularity of applying "gold standard"</li> <li>Quality control</li> </ul>
2. Item selection	<ul style="list-style-type: none"> <li>Data-driven item reduction</li> <li>Clinical consensus item reduction</li> </ul>
3. Final Criteria	<ul style="list-style-type: none"> <li>Combined data-driven and clinical expert review</li> <li>Model performance: face validity, discrimination</li> </ul>

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2. How?

1. Case selection

Production of clinical vignettes

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2. How?

1. Case selection



DCVAS

2. How?

1. Case selection

Production of clinical vignettes

Expert reviewers  
chosen from diverse  
geographical  
locations and  
specialties



Web-based review system

DCVAS

2. How?

1. Case selection

Independent expert review of each clinical vignette

Expert reviewers were asked:



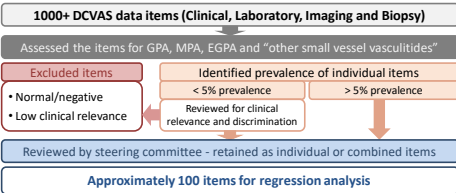
- Level of certainty: **very certain**, **moderately certain**, **uncertain**, or **very uncertain**
- **Match on subtype** ( $\geq$  moderate certainty) - Case passed
- **No match** - Second review by DCVAS committee member

DCVAS

2. How?

2. Item selection

Item selection: data-driven and expert consensus



DCVAS

2. How?

3. Final criteria

Statistical methods

- Development (80%) and validation (20%) datasets
- Too many predictors for standard regression analysis, even after data reduction
- Specific type of analysis - Lasso logistic regression
- Multiple imputation for missing data



Menelaos Pavlou et al. *BMJ Research Methods and Reporting* 2015

DCVAS



Questions?



2017 Provisional Classification Criteria for Granulomatosis with Polyangiitis (GPA)

Presenter

Joanna Robson MBBS PhD MRCP  
Consultant Senior Lecturer in Rheumatology  
University of the West of England, Bristol



Results

DCVAS Classification criteria: 3 phases

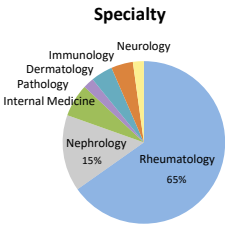
1. Case selection

- Clinical vignette expert panel review
- Avoid circularity of applying "gold standard"
- Quality control

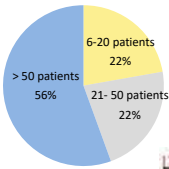


Expert reviewer characteristics

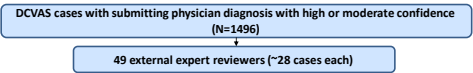
- Forty-nine reviewers from 22 countries



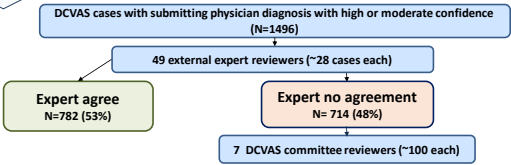
No. of AAV patients seen per year

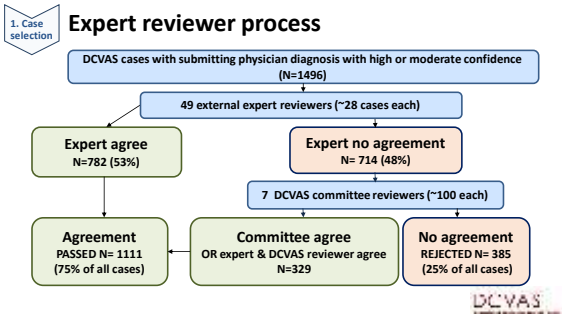


Expert reviewer process



Expert reviewer process





1. Case selection

### Expert review of cases

Type of vasculitis	Submitting physician cases	Total cases passed on expert review
Granulomatosis with polyangiitis	578	492
Microscopic polyangiitis	275	205
Eosinophilic granulomatosis with polyangiitis	214	156
AAV-cannot subtype	31	20
Anti-GBM	9	10
Cryoglobulinaemic vasculitis	32	24
IgA vasculitis	170	108
Other small vessel - cannot subtype	46	1
Polyarteritis nodosa	22	7
Giant Cell Arteritis	43	38
Takayasu arteritis	39	30
Vasculitis mimics	37	20

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1. Case selection

### Expert review of cases

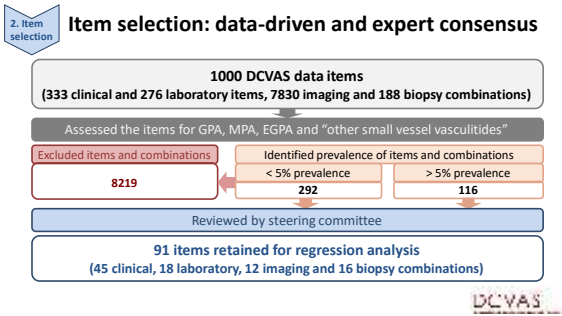
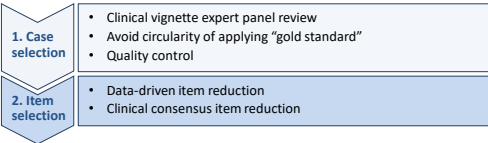
Comparators for GPA

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DCVAS

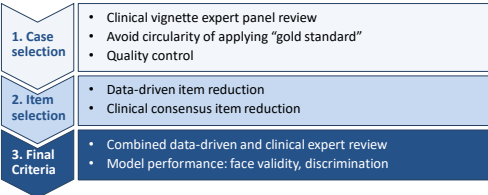
## Results

### DCVAS Classification criteria: 3 phases



## Results

### DCVAS Classification criteria: 3 phases



3. Final criteria

Patient characteristics for GPA analysis

	GPA n = 492	Comparators n = 531	p-value
Age (SD)	53.4 (16.4)	57.8 (16.9)	<0.001
Gender (F)	(47.2% of 492)	(49.5% of 531)	0.32
Creatinine umol/L	159.3	203.0	
mg/dL	1.8	2.3	<0.001
c-ANCA positive (%)	72.4%	5.5%	<0.01
p-ANCA positive (%)	10.4%	47.3%	<0.01
PR3- antibody positive (%)	80.9%	3.4%	<0.01
MPO- antibody positive (%)	8.30%	53.9%	<0.01

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3. Final criteria

Data split of development and validation sets (80% : 20%)

	Development set	Validation set
GPA	395 (48.2%)	97 (47.5%)
Comparators	424 (51.8%)	107 (52.5%)
Total	819	204

DCVAS

3. Final criteria

Results (all 22 independent variables)

Rank	Description	OR (95% CI)
1	cANCA or PR3	59.3 (22.2, 158.4)
2	Bloody nasal discharge	18.1 (6.00, 54.6)
3	Nasal ulcers, crusting or sino nasal congestion or blockage	5.79 (2.28, 14.7)
4	Eosinophil count $\geq 1 \times 10^9/L$	0.05 (0.02, 0.17)
5	Nasal polyps	0.03 (0.00, 0.23)
6	Nodules/ mass or cavitation on chest imaging	7.44 (2.76, 20.0)
7	Hearing loss or reduction	5.90 (1.96, 17.8)
8	IgA deposition on biopsy	0.02 (0.00, 0.25)
9	Granuloma on biopsy	5.48 (1.30, 23.1)
10	pANCA or MPO	0.25 (0.10, 0.64)
11	Red or painful eyes	3.87 (1.15, 13.0)

Rank	Description	OR (95% CI)
12	Cartilagenous involvement	3.96 (1.03, 15.3)
13	Abdominal pain	0.40 (0.13, 1.25)
14	Obstructive airways	0.18 (0.06, 0.59)
15	Mononeuritis multiplex OR Motor neuropathy (not radiculopathy)	0.30 (0.12, 0.78)
16	Anaemia (Hb <10g/dL)	0.36 (0.16, 0.79)
17	Non-productive cough	2.89 (1.12, 7.50)
18	Inflammation, consolidation or effusion of the nasal/ paranasal sinuses on imaging	1.39 (0.47, 4.09)
19	Non blood stained nasal discharge	2.01 (0.61, 6.67)
20	Fever, night sweats or rigors	2.54 (1.16, 5.55)
21	Fibrosis or ILD on imaging of the chest	0.11 (0.02, 0.68)
22	Fatigue	0.54 (0.25, 1.17)

DCVAS

3. Final criteria

Results (all 22 independent variables)

Data driven only top 5

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DCVAS

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DCVAS

3. Final criteria

Results (all 22 independent variables)

Clinical and data driven

Rank	Description	OR (95% CI)
2	Bloody nasal discharge	134.8 (10.05, 291.1)
3	Nasal ulcers, crusting or sino nasal congestion or blockage	5.79 (2.28, 14.1)
4	Eosinophil count $\geq 1 \times 10^9/L$	0.08 (0.03, 0.17)
5	Nasal polyps	0.03 (0.01, 0.23)
6	Nodules/ mass or cavitation on chest imaging	7.44 (2.76, 20.0)
7	Hearing loss or reduction	5.90 (1.96, 17.8)
8	IgA deposition on biopsy	0.02 (0.00, 0.25)
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Clinical and data driven

Rank	Description	OR (95% CI)
12	Cartilagenous involvement	3.96 (1.03, 15.2)
13	Abdominal pain	0.40 (0.13, 1.25)
14	Obstructive airways	0.18 (0.06, 0.59)
15	Mononeuritis multiplex OR Motor neuropathy (not radiculopathy)	0.30 (0.12, 0.78)
16	Anaemia (Hb $<10g/dL$ )	0.36 (0.16, 0.79)
17	Non-productive cough	2.89 (1.12, 7.50)
18	Inflammation, consolidation or effusion of the nasal/ paranasal sinuses on imaging	1.39 (0.47, 4.09)
19	Non blood stained nasal discharge	2.01 (0.61, 6.67)
20	Fever, night sweats or rigors	2.54 (1.16, 5.55)
21	Fibrosis or ILO on imaging of the chest	0.11 (0.02, 0.68)
22	Fatigue	0.54 (0.25, 1.17)

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3. Final criteria

Data-driven and clinically selected 9 item model

Description	Odds Ratio (95%CI)	P-value
cANCA or PR3	134.8 (62.4, 291.1)	<0.001
Bloody nasal discharge/ ulcers/ crusting or sino-nasal congestion	16.0 (7.90, 32.2)	<0.001
Granuloma on biopsy	12.9 (3.76, 44.4)	<0.001
Nodules/ mass or cavitation on chest imaging	5.77 (2.63, 12.7)	<0.001
Cartilagenous involvement*	5.50 (1.87, 16.2)	<0.01
Red or painful eyes	4.42 (1.67, 11.7)	<0.01
Hearing loss or reduction	3.74 (1.60, 8.73)	<0.01
Maximum eosinophil count ( $\times 10^9/L$ ) ( $\geq 1$ vs. $<1$ )	0.08 (0.03, 0.19)	<0.001
Nasal polyps	0.02 (0.01, 0.10)	<0.001

\*Inflamed ear or nose cartilage or hoarse voice/stridor, endobronchial involvement or saddle nose deformity

DCVAS

3. Final criteria

Points based risk score

Risk factor at baseline	Coefficient	Risk score
cANCA or PR3	4.90	+ 5
Bloody nasal discharge/ ulcers or crusting or sino-nasal congestion	2.77	+ 3
Granuloma on biopsy	2.56	+ 3
Nodules/ mass or cavitation on chest imaging	1.75	+ 2
Cartilagenous involvement	1.71	+ 2
Red or painful eyes	1.49	+ 1
Hearing loss or reduction	1.32	+ 1
Maximum eosinophil count $\geq 1$ ( $\times 10^9/L$ )	- 2.56	- 3
Nasal polyps	- 3.70	- 4

DCVAS

3. Final criteria

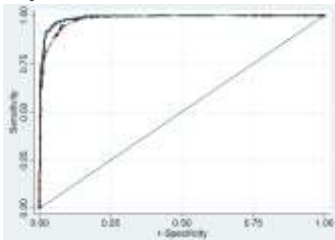
Model Performance

Model Performance	Development	Validation
AUC (95% CI)	0.98 (0.97, 0.99)	0.98 (0.96, 0.99)
Cut-off $\geq 5$ points		
Sensitivity	89.9%	90.7%
Specificity	94.1%	93.5%
Cut-off $\geq 6$ points		
Sensitivity	78.2%	75.3%
Specificity	98.4%	99.1%

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3. Final criteria

Discrimination curves



22 variables model (AUC 0.99)

9 variables model (AUC 0.98)

Risk Score (AUC 0.98)

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ACR/ EULAR Provisional 2017 Classification Criteria for Granulomatosis with Polyangiitis (GPA)

This criteria set is for use when a diagnosis of small or medium vessel vasculitis has been made, in order to classify the patient as having GPA

Clinical	Bloody nasal discharge, ulcers, crusting or sino-nasal congestion	3
	Nasal polyps	- 4
	Hearing loss or reduction	1
	Cartilagenous involvement	2
	Red or painful eyes	1
Tests	cANCA or PR3- antibody positive	5
	Eosinophil count $\geq 1$ ( $\times 10^9/L$ )	- 3
	Nodules, mass or cavitation on chest imaging	2
	Granuloma on biopsy	3

Add scores for 9 items; a score of  $\geq 5$  is needed for classification of GPA

DCVAS

Acknowledgements

Expert panel reviewers

Alojzija Hocevar	Eamonn Molloy	Marco A Alba	Pavel Novikov
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Bo Baslund	Joerg Henes	Neil Basu	Shouichi Fujimoto
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Carlo Salvarani	Kazuo Matsui	Nina Brown	Thomas Hauser
Carol Langford	Lillian Barra	Ömer Karadağ	Thomas Neumann
Cees Kallenberg	Liza Rajasekhar	Ora Gewurz-Singer	Vicky Quincey
Charles Jennette	Loic Guillevin	Paul A Monach	Vladimir Tesar
Cord Sunderkötter	Luis Felipe Flores-Suárez	Paul Gatenby	Wolfgang Schmidt
David Jayne			



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	<b>Site communications &amp; payments</b> Marian Montgomery
	<b>Database design</b> Joe Barrett, Joe Rosa, Nathanael Gray



Questions?





New ACR/EULAR Classification Criteria for ANCA-Associated Vasculitis: *Implications for Clinical Practice*

**Presenters**

Raashid Luqmani, University of Oxford  
Peter A. Merkel, University of Pennsylvania  
Richard Watts, University of East Anglia



ACR/ EULAR Provisional 2017 Classification Criteria for Granulomatosis with Polyangiitis (GPA)

This criteria set is for use when a diagnosis of small or medium vessel vasculitis has been made, in order to classify the patient as having GPA

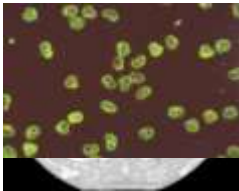
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	Nasal polyps	— 4
	Hearing loss or reduction	1
	Cartilagenous involvement	2
	Red or painful eyes	1
Tests	cANCA or PR3- antibody positive	5
	Eosinophil count ≥ 1 (x10 <sup>9</sup> /L)	— 3
	Nodules, mass or cavitation on chest imaging	2
	Granuloma on biopsy	3

Add scores for 9 items; a score of ≥ 5 is needed for classification of GPA



Key points

- Provide modern criteria for 21<sup>st</sup> century
- Clinically based
  - Reflects current investigative practice
  - Includes, BUT not critically dependent on
    - ANCA
    - Biopsy
  - Provide a risk score
  - Good specificity and sensitivity



Clinical Implications

- Clinical trials based on modern classification criteria for ANCA-associated vasculitis
- Improvement in evidence base for decisions about therapy

DCVAS

Example Case 1

Bloody nasal discharge, ulcers or crusting or sino-nasal congestion	3
Nasal polyps	4
Hearing loss or reduction	1
Cartilagenous involvement	2
Red or painful eyes	1
cANCA or PR3- antibody positive	5
Eosinophil count $\geq 1$ ( $\times 10^9/L$ )	3
Nodules, mass or cavitation on chest imaging	2
Granuloma on biopsy	3

Risk score = 1; classified as *not* GPA  
(score of  $\geq 5$  is needed for classification of GPA)

DCVAS

Example Case 2

Bloody nasal discharge, ulcers or crusting or sino-nasal congestion	3
Nasal polyps	4
Hearing loss or reduction	1
Cartilagenous involvement	2
Red or painful eyes	1
cANCA or PR3- antibody positive	5
Eosinophil count $\geq 1$ ( $\times 10^9/L$ )	3
Nodules, mass or cavitation on chest imaging	2
Granuloma on biopsy	3

Risk score = 8; classified as GPA  
(score of  $\geq 5$  is needed for classification of GPA)

DCVAS

Example Case 1

- 45 year-old woman
- Constitutional symptoms
- Left foot and right wrist drop
- Interstitial lung disease



Scleritis



Red cell casts in urine



Pauci immune crescentic GN on renal biopsy



pANCA positive on IF  
MPO-ANCA positive by ELISA

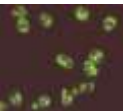
DCVAS

Example Case 2

- 73 year-old woman
- Constitutional symptoms
- Cough
- Reduced hearing
- CRP 82 mg/L
- Eosinophil count normal



Normal CXR, but  
CT chest showing  
cavitating nodules



cANCA positive on IF  
PR3-ANCA strongly  
positive on ELISA

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ACR-EULAR 2017 GPA criteria

Impact on the two cases

Case 1

- ACR 1990: Classified as PAN *and* GPA (WG)
- ACR/EULAR 2017: Risk score = 1; Not classified as GPA

Case 2

- ACR 1990: Not classified as GPA (WG)
- ACR/EULAR 2017: Risk score = 8; Classified as GPA

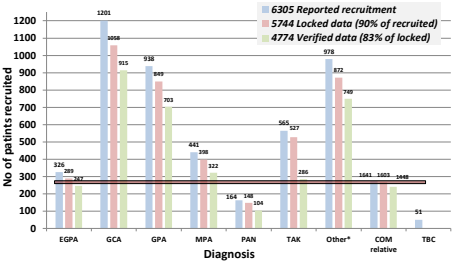
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Future Analyses

- Develop criteria for AAV, PAN, TAK, GCA, and IgA
- Performance in different datasets
  - e.g.
    - physician-submitted cases
    - external datasets
- Comparison with previous ACR criteria
- Overall clinical features of each group and outcomes
- Diagnostic criteria



DCVAS: Recruitment by Diagnosis



Questions?

