

# The Birmingham Vasculitis Activity Score (BVAS)



## INTRODUCTION TO THE BVAS

> The BVAS is a **validated** tool and the **most widely accepted** measure of disease activity in major studies of vasculitis, including AAV<sup>1</sup>

**First published in 1994<sup>2</sup>**

**Developed by consensus expert opinion, based on clinical practice<sup>2</sup>**

**Clinical features revised over the years, with version 3 being the most recent version<sup>3</sup>**



## THE BVAS CALCULATION

- > The BVAS version 3 comprises **56 individual items** in **9 organ systems** (1 general and 8 tissue-specific)<sup>3</sup>
  - Each item and organ system are given a **predetermined numerical score** (item) or ceiling score (organ system) that is **weighted** according to its perceived clinical relevance<sup>3</sup>
- > The BVAS captures a **broad spectrum of clinical manifestations** for new, worsening, or persistent disease<sup>3</sup>
  - Disease manifestations are scored only if active in the previous 4 weeks and directly attributable to vasculitis<sup>3</sup>
  - Disease is marked as “persistent” only if all disease manifestations are attributable to active disease that is not new/worse in the previous 4 weeks<sup>3</sup>



## INTERPRETATION OF THE BVAS

> The BVAS quantifies a patient’s disease activity: the higher the score, the more active the vasculitic disease at the time of evaluation<sup>3,4</sup>

**Maximum possible score of 63<sup>4,5</sup>**

**Score of 0 represents remission<sup>5</sup>**

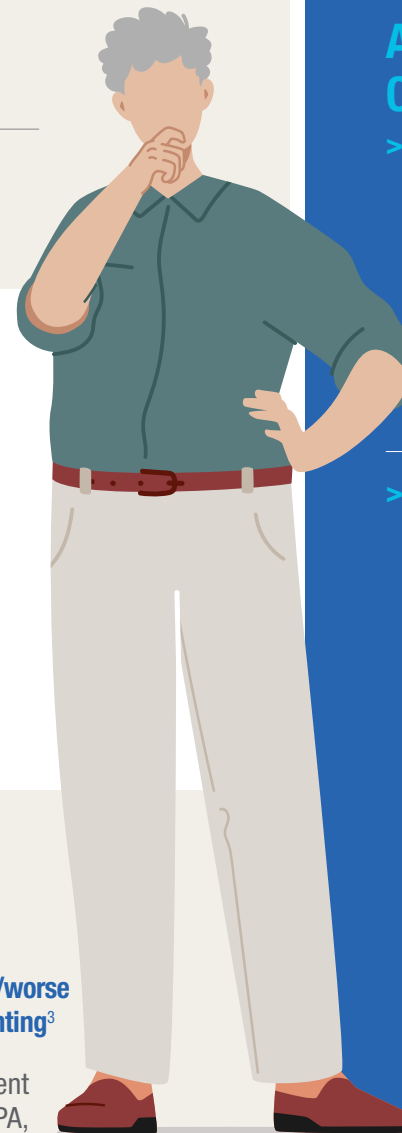
**Score of  $\geq 1$  represents active disease<sup>5</sup>**

**Score based on new/worse or persistent weighting<sup>3</sup>**

> The BVAS is not reflected in the definition of disease states in the 2021 ACR/VF AAV treatment guidelines, which define remission as no clinical signs or symptoms attributed to GPA or MPA, on or off immunosuppressive therapy<sup>6</sup>

## APPLICATIONS OF THE BVAS

- > The BVAS is a **clinical tool** to standardize disease assessment in clinical studies<sup>3,7</sup>
  - In vasculitis studies, the BVAS can be used to **differentiate between remission and active disease** and assess **response to therapy**<sup>3,7</sup>
- > In clinical practice, the BVAS may be used as a **checklist of parameters to examine** following diagnosis and may not be used to guide treatment course decisions<sup>3,6-8</sup>
  - In daily practice, the diagnosis of AAV is based upon the combination of clinical findings, laboratory tests or histology, and/or imaging studies, and the disease severity classification drives treatment decisions<sup>8</sup>
  - Specific training on the use of the BVAS is needed for doctors to appropriately use the tool in clinical practice<sup>3</sup>



AAV = anti-neutrophil cytoplasmic antibody-associated vasculitis; ACR = American College of Rheumatology; BVAS = Birmingham Vasculitis Activity Score; GPA = granulomatosis with polyangiitis; MPA = microscopic polyangiitis, VF = Vasculitis Foundation.

1. Merkel PA, et al. *J Rheumatol*. 2011;38:1480–1486. 2. Luqmani RA, et al. *QJM*. 1994;87:671–678. 3. Mukhtyar C, et al. *Ann Rheum Dis*. 2009;68:1827–1832 and Supplementary online appendix. 4. Stone JH, et al. *N Engl J Med*. 2010;363:221–232. 5. Suppiah R, et al. *Rheumatology (Oxford)*. 2011;50:899–905. 6. Chung SA, et al. *Arthritis Rheumatol*. 2021;73:1366–1383. 7. Kitching AR, et al. *Nat Rev Dis Primers*. 2020;6:71. 8. Miller A, et al. *Clin Exp Immunol*. 2010;160:143–160.

**AMGEN**<sup>®</sup>

# The Birmingham Vasculitis Activity Score (Version 3) Checklist

## General

- Myalgia
- Arthralgia/arthritis
- Fever  $\geq 38^{\circ}\text{C}$
- Weight loss  $\geq 2$  kg

## Ears, Nose, and Throat

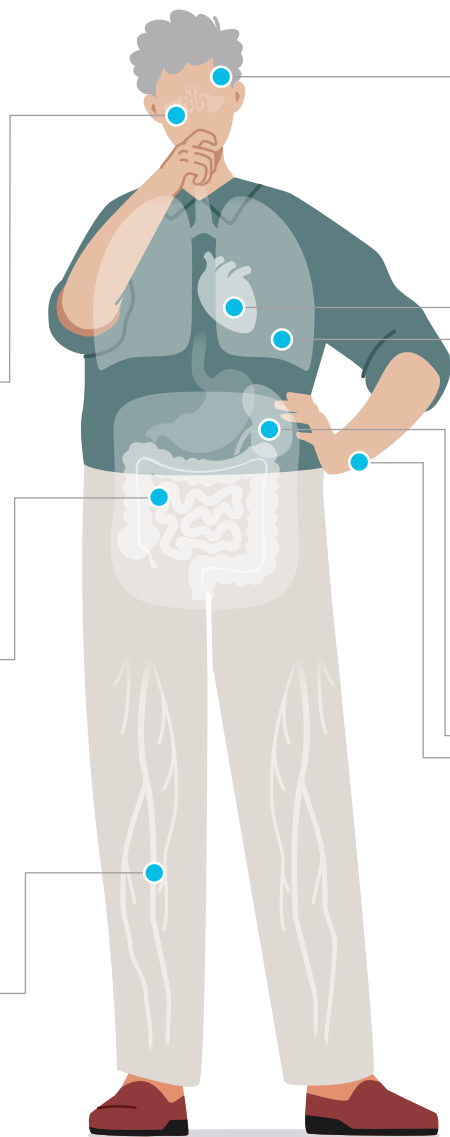
- Bloody nasal discharge/crusts/ulcers/granulomata
- Paranasal sinus involvement
- Subglottic stenosis
- Conductive hearing loss
- Sensorineural hearing loss

## Abdominal

- Peritonitis
- Bloody diarrhea
- Ischemic abdominal pain

## Nervous System

- Headache
- Meningitis
- Organic confusion
- Seizures (not hypertensive)
- Cerebrovascular accident
- Spinal cord lesion
- Cranial nerve palsy
- Sensory peripheral neuropathy
- Mononeuritis multiplex



## Mucous Membranes/Eyes

- Mouth ulcers
- Genital ulcers
- Adnexal inflammation
- Significant proptosis
- Scleritis/episcleritis
- Conjunctivitis/blepharitis/keratitis
- Blurred vision
- Sudden visual loss
- Uveitis
- Retinal changes\*

## Renal

- Hypertension
- Proteinuria  $> 1+$
- Hematuria  $\geq 10$  RBC/hpf
- Serum creatinine  $125-249 \mu\text{mol/L}^{\dagger}$
- Serum creatinine  $250-499 \mu\text{mol/L}^{\dagger}$
- Serum creatinine  $\geq 500 \mu\text{mol/L}^{\dagger}$
- Rise in serum creatinine  $> 30\%$  or fall in creatinine clearance  $> 25$

## Cutaneous

- Infarct
- Purpura
- Ulcer
- Gangrene
- Other skin vasculitis

## Cardiovascular

- Loss of pulses
- Valvular heart disease
- Pericarditis
- Ischemic cardiac pain
- Cardiomyopathy
- Congestive cardiac failure

## Chest

- Wheeze
- Nodules or cavities
- Pleural effusion/pleurisy
- Infiltrate
- Endobronchial involvement
- Massive hemoptysis/alveolar hemorrhage
- Respiratory failure

## Persistent Disease Only

- All the abnormalities are due to persistent disease

The Birmingham Vasculitis Activity Score also includes an "other" category to capture additional symptoms attributed to active vasculitis.

\*Vasculitis/thrombosis/exudate/hemorrhage; †can only be scored on the first assessment.

RBC = red blood cells; hpf = high power field.

Mukhtyar C, et al. *Ann Rheum Dis*. 2009;68:1827-1832. Supplementary online appendix.

© 2023 Amgen Inc. All rights reserved. USA-569-80336. 8/23.