## BVAS versjon 3 (2008)

# Birmingham Vasculitis Activity Score (version 3)

**Patient ID:** **Date of birth:**

**Total score:**

**Assessor:** **Date of assessment**

Tick an item **only** if attributable to active vasculitis. If there are no abnormalities in a section, please tick 'None' for that organ-system.

If **all** abnormalities are due to persistent disease (active vasculitis which is not new/worse in the prior 4 weeks), tick the **PERSISTENT** box at the bottom right corner

**Is this the patient's first assessment?** **Yes O** **No O**

| | None | Active disease |
|---|---|---|
| **1. General** | O | |
| Myalgia | | O |
| Arthralgia / arthritis | | O |
| Fever ≥38° C | | O |
| Weight loss ≥2 kg | | O |
| **2. Cutaneous** | O | |
| Infarct | | O |
| Purpura | | O |
| Ulcer | | O |
| Gangrene | | O |
| Other skin vasculitis | | O |
| **3. Mucous membranes / eyes** | O | |
| Mouth ulcers | | O |
| Genital ulcers | | O |
| Adnexal inflammation | | O |
| Significant proptosis | | O |
| Scleritis / Episcleritis | | O |
| Conjunctivitis / Blepharitis / Keratitis | | O |
| Blurred vision | | O |
| Sudden visual loss | | O |
| Uveitis | | O |
| Retinal changes (vasculitis / thrombosis / exudate / haemorrhage) | | O |
| **4. ENT** | O | |
| Bloody nasal discharge / crusts / ulcers / granulomata | | O |
| Paranasal sinus involvement | | O |
| Subglottic stenosis | | O |
| Conductive hearing loss | | O |
| Sensorineural hearing loss | | O |
| **5. Chest** | O | |
| Wheeze | | O |
| Nodules or cavities | | O |
| Pleural effusion / pleurisy | | O |
| Infiltrate | | O |
| Endobronchial involvement | | O |
| Massive haemoptysis / alveolar haemorrhage | | O |
| Respiratory failure | | O |

| | None | Active disease |
|---|---|---|
| **6. Cardiovascular** | O | |
| Loss of pulses | | O |
| Valvular heart disease | | O |
| Pericarditis | | O |
| Ischaemic cardiac pain | | O |
| Cardiomyopathy | | O |
| Congestive cardiac failure | | O |
| **7. Abdominal** | O | |
| Peritonitis | | O |
| Bloody diarrhoea | | O |
| Ischaemic abdominal pain | | O |
| **8. Renal** | O | |
| Hypertension | | O |
| Proteinuria >1+ | | O |
| Haematuria ≥10 RBCs/hpf | | O |
| Serum creatinine 125-249 µmol/L* | | O |
| Serum creatinine 250-499 µmol/L* | | O |
| Serum creatinine ≥500 µmol/L* | | O |
| Rise in serum creatinine >30% or fall in creatinine clearance >25% | | O |
| ***Can only be scored on the first assessment** | | |
| **9. Nervous system** | O | |
| Headache | | O |
| Meningitis | | O |
| Organic confusion | | O |
| Seizures (not hypertensive) | | O |
| Cerebrovascular accident | | O |
| Spinal cord lesion | | O |
| Cranial nerve palsy | | O |
| Sensory peripheral neuropathy | | O |
| Mononeuritis multiplex | | O |
| **10. Other** | O | |
| a. | | O |
| b. | | O |
| c. | | O |
| d. | | O |
| **PERSISTENT DISEASE ONLY:** (Tick here if **all** the abnormalities are due to persistent disease) | | ☐ |


**References:**

**Version 1**: Luqmani, RA, et al. (1994). "Birmingham Vasculitis Activity Score (BVAS) in systemic necrotizing vasculitis." QJM **87**(11):671-8.

**Version 2**: Luqmani, RA, et al. (1997). "Disease assessment and management of the vasculitides." Bailieres Clin Rheumatol **11**(2): 423-46.

**Version 3**: Mukhtyar C, et al (2008). "Modification and validation of the Birmingham Vasculitis Activity Score (version 3) Ann Rheum Dis. 2008 Dec 3. [Epub ahead of print]

## Utfylling/vurdering BVAS

- BVAS fylles ut ved alle besøk på alle pasienter med ANCA-assosiert vaskulitt

- Husk at dette er en vurdering av vaskulittaktivitet - og ikke et symptomskår

- Symptomer som ikke skyldes pågående vaskulittaktivitet, skal ikke registreres (f.eks. artrosesmerter, fibromyalgisymptomer, angina/hypertensjon oppstått før vaskulitten, eller nese-/bihulesymptomer som en mener skyldes infeksjon)

- Symptomer og funn som er resultat av skade oppstått som følge av tidligere aktiv vaskulitt (f.eks. skorper i nesen, redusert hørsel, nevropatisk smerte, proteinuri) skal en heller ikke krysse av for i BVAS såfremt de ikke har blitt verre siste 4 uker
    - Skadeutvikling som følge av tidligere vaskulittaktivitet registreres i skadeindeksen VDI

- Dersom ingen av de registrerte symptomer/funn er nye eller forverrede siste 4 uker (vanligvis skal de ha vedvart i 3 md.), skal man markere for «Persistent disease only», og poengsummen blir da nærmest halvert i forhold til skår ved aktiv/ny sykdom
    - Eksempel: pasienten kommer til kontroll og er i bedring etter debut eller residiv, men det er fortsatt noe vaskulittaktivitet til stede. «Persistent disease» forutsetter at man mener det foreligger vaskulittaktivitet, og at symptomer/funn ikke bare skyldes skade av tidligere aktiv vaskulitt
    - Det er avkrysningen «Persistent disease only» som avgjør poengsum og må avkrysses hvis man mener dette er persistent disease. Det er ikke nok å angi «mener dette er persisterende sykdom» i legens vurdering (den er ikke koblet til utregning av VDI-skår)

- Legen må konkludere klinisk og krysse av på sykdomsvurdering:
  Debut (gjelder kun ved sykdomsdebut), lett residiv, alvorlig residiv, persisterende sykdom (vedvarende vaskulittaktivitet etter debut eller residiv) eller remisjon. Det bør naturligvis være samsvar mellom avkryssing i BVAS og legens vurdering

## GLOSSARY AND SCORING FOR BVAS version 3

**Rules for scoring BVAS**

1. Disease manifestations are scored **only when they are attributable to active vasculitis**. The manifestation should not be scored if there is reasonable evidence of another aetiology for the symptoms e.g. infection, drug reaction, other co-morbidity.
2. Tick "Persistent Disease" box if **all** the abnormalities are due to active (but not new or worse) vasculitis.
3. Specialist opinion, or the results of laboratory or imaging investigations will be required for some items. Excepting those circumstances, the whole form should be completed at the time of the consultation.
4. The bands of serum creatinine should be scored **only** on the first visit.
5. Items marked with an asterisk (*) are not compatible with 'persistent' disease. These manifestations always suggest new or worse disease when due to active vasculitis.

| Manifestation | Definition | Persistent | New / Worse |
|---|---|---|---|
| **1. General** | **Maximum scores** | **2** | **3** |
| Myalgia | Pain in the muscles | 1 | 1 |
| Arthralgia or arthritis | Pain in the joints or joint inflammation | 1 | 1 |
| Fever ≥38° C | Documented oral / axillary temperature. If rectal temperature is measured, raise threshold to 38.5° C | 2 | 2 |
| Weight Loss ≥2 kg | Loss of dry body weight without dieting | 2 | 2 |

| **2. Cutaneous** | **Maximum scores** | **3** | **6** |
|---|---|---|---|
| Infarct | Area of tissue necrosis or splinter haemorrhages | 1 | 2 |
| Purpura | Subcutaneous or submucosal haemorrhage in the absence of trauma | 1 | 2 |
| Ulcer | A disruption in the continuity of the skin | 1 | 4 |
| Gangrene | Extensive tissue necrosis | 2 | 6 |
| Other skin vasculitis | Livedo reticularis, subcutaneous nodules, erythema nodosum, etc | 1 | 2 |

| **3. Mucous Membranes / eyes** | **Maximum scores** | **3** | **6** |
|---|---|---|---|
| Mouth ulcers / granulomata | Aphthous stomatitis, deep ulcers, strawberry gingival hyperplasia | 1 | 2 |
| Genital ulcers | Ulcers on the genitalia or perineum | 1 | 1 |
| Adnexal inflammation | Salivary or lacrimal gland inflammation. | 2 | 4 |
| Significant proptosis | >2 mm protrusion of the eyeball | 2 | 4 |
| Scleritis / Episcleritis | Inflammation of the sclera | 1 | 2 |
| Conjunctivitis / Blepharitis / Keratitis | Inflammation of the conjunctiva, eyelids or cornea - but not due to sicca syndrome | 1 | 1 |
| Blurred vision | Deterioration of visual acuity from previous or baseline | 2 | 3 |
| Sudden visual loss* | Acute loss of vision | * | 6 |
| Uveitis | Inflammation of the uvea (iris, ciliary body, choroid) | 2 | 6 |
| Retinal changes (vasculitis, thrombosis / exudate / haemorrhage) | Sheathing of retinal vessels or evidence of retinal vasculitis on fluorescein angiography; thrombotic retinal arterial or venous occlusion; soft retinal exudate (exclude hard exudates) / retinal haemorrhage | 2 | 6 |

| **4. ENT** | **Maximum scores** | **3** | **6** |
|---|---|---|---|
| Bloody nasal discharge / crusts / ulcers / granulomata | Bloody, mucopurulent, nasal secretion, light or dark brown crusts frequently obstructing the nose, nasal ulcers or granulomatous lesions observed on rhinoscopy | 2 | 4 |
| Paranasal sinus involvement | Tenderness or pain over paranasal sinuses (usually confirmed by imaging) | 1 | 2 |
| Subglottic stenosis | Stridor or hoarseness due to inflammation and narrowing of the subglottic area observed by laryngoscopy | 3 | 6 |
| Conductive hearing loss | Hearing loss due to middle ear involvement (usually confirmed by audiometry) | 1 | 3 |
| Sensorineural hearing loss | Hearing loss due to auditory nerve or cochlear damage (usually confirmed by audiometry) | 2 | 6 |

| 5. Chest | Maximum scores | 3 | 6 |
|---|---|---|---|
| Wheeze | Wheeze on clinical examination | 1 | 2 |
| Nodules or cavities* | New lesions detected on imaging | * | 3 |
| Pleural effusion / pleurisy | Pleural pain and/or friction rub on clinical assessment; radiologically confirmed pleural effusion. | 2 | 4 |
| Infiltrate | Detected on chest X-ray or CT scan | 2 | 4 |
| Endobronchial involvement | Endobronchial pseudotumor or ulcerative lesions. NB: smooth stenotic lesions to be included in VDI; subglottic lesions to be recorded in the ENT section. | 2 | 4 |
| Massive haemoptysis / alveolar haemorrhage | Major pulmonary bleeding, with shifting pulmonary infiltrates | 4 | 6 |
| Respiratory failure | The need for artificial ventilation | 4 | 6 |

| 6. Cardiovascular | Maximum scores | 3 | 6 |
|---|---|---|---|
| Loss of pulses | Clinical absence of peripheral arterial pulsation in any limb | 1 | 4 |
| Valvular heart disease | Clinical or echo detection of aortic / mitral / pulmonary valve involvement | 2 | 4 |
| Pericarditis | Pericardial pain / friction rub on clinical assessment | 1 | 3 |
| Ischaemic cardiac pain | Typical clinical history of cardiac pain leading to myocardial infarction or angina. | 2 | 4 |
| Cardiomyopathy | Significant impairment of cardiac function due to poor ventricular wall motion confirmed on echocardiography | 3 | 6 |
| Congestive cardiac failure | Heart failure by history or clinical examination | 3 | 6 |

| 7. Abdominal | Maximum scores | 4 | 9 |
|---|---|---|---|
| Peritonitis | Typical abdominal pain suggestive of peritoneal involvement | 3 | 9 |
| Bloody diarrhoea | Of recent onset | 3 | 9 |
| Ischaemic abdominal pain | Typical abdominal pain suggestive of bowel ischaemia, confirmed by imaging or surgery | 2 | 6 |

| 8. Renal | Maximum scores | 6 | 12 |
|---|---|---|---|
| Hypertension | Diastolic >95 mm Hg | 1 | 4 |
| Proteinuria | >1+ on urinalysis or >0.2g/24 hours | 2 | 4 |
| Haematuria | 'Moderate' on urinalysis or ≥10 RBC per high power field, usually accompanied by red cell casts | 3 | 6 |
| Serum creatinine 125-249 µmol/L | At first assessment only | 2 | 4 |
| Serum creatinine 250-499 µmol/L | | 3 | 6 |
| Serum creatinine ≥500 µmol/L | | 4 | 8 |
| >30% rise in creatinine or >25% fall in creatinine clearance * | Progressive worsening of renal function. Can be used at each assessment if the renal function has deteriorated from prior value | * | 6 |

| 9. Nervous system | Maximum scores | 6 | 9 |
|---|---|---|---|
| Headache | Unaccustomed & persistent headache | 1 | 1 |
| Meningitis | Clinical evidence of meningism | 1 | 3 |
| Organic confusion | Impaired orientation, memory or other intellectual function in the absence of metabolic, psychiatric, pharmacological or toxic causes. | 1 | 3 |
| Seizures (not hypertensive) | Clinical or EEG evidence of aberrant electrical activity in the brain | 3 | 9 |
| Stroke | Focal neurological signs lasting >24 hours due to a CNS vascular event | 3 | 9 |
| Spinal cord lesion | Clinical or imaging evidence of spinal cord involvement | 3 | 9 |
| Cranial nerve palsy | Clinical evidence of cranial nerve palsy – score VIII nerve palsy as sensorineural hearing loss, do not score ocular palsies if they secondary to pressure effects | 3 | 6 |
| Sensory peripheral neuropathy | Objective sensory deficit in a non-dermatomal distribution | 3 | 6 |
| Mononeuritis multiplex | Single or multiple specific motor nerve palsies | 3 | 9 |