4.2 Glossary

Glossary and scoring for BVAS (V. 3). GENERAL RULE: disease features are scored only when they are due to active vasculitis, after excluding other causes (e.g. infection, hypertension, etc.). If the feature is due to active disease, it is scored in the boxes. It is essential to apply these principles to each item below. Scores have been weighted according to the severity which each symptom or sign is thought to represent. Tick "Persistent Disease" box if all the abnormalities are due to active (but not new or worse) vasculitis. If any of the abnormalities are due to new/worse disease, DO NOT tick the "Persistent Disease" box. For some features, further information (from specialist opinion or further tests) is required if abnormality is newly present or worse. Remember that in most instances, you will be able to complete the whole record when you see the patient. However, you may need further information before entering some items. Please leave these items blank, until the information is available, and then fill them in. For example, if the patient has new onset of stridor, you would usually ask an otolaryngologist to investigate this further to determine whether or not it is due to active granulomatosis with polyangiitis (Wegener's).

System/item	Description	New/Worse score	Persistent score
1. General		**3**	**2**
Myalgia	Pain in the muscles	1	1
Arthralgia or arthritis	Pain in the joints or joint inflammation	1	1
Fever ≥ 38.0 ^{0}C	Documented oral/axillary temperature elevation. Rectal temps are 0.5 ^{0}C higher	2	2
Weight Loss	At least 2kg loss of body weight (not fluid) having occurred since last assessment or in the 4 weeks not as a consequence of dieting	2	2
2. Cutaneous		**6**	**3**
Infarct	Area of tissue necrosis or splinter haemorrhages	2	1
Purpura	Petechiae (small red spots), palpable purpura, or ecchymoses (large plaques) in skin or oozing (in the absence of trauma) in the mucous membranes.	2	1
Ulcer	Open sore in a skin surface.	4	1
Gangrene	Extensive tissue necrosis (e.g. digit)	6	2
Other skin vasculitis	Livedo reticularis, subcutaneous nodules, erythema nodosum, etc	2	1
3. Mucous membranes/eyes		**6**	**3**
Mouth ulcers/granulomata	Aphthous stomatitis, deep ulcers and/or “strawberry” gingival hyperplasia, excluding lupus erythematosus, and infection	2	1
Genital ulcers	Ulcers localized in the genitalia or perineum, excluding infections.	1	1
Adnexal inflammation	Salivary (diffuse, tender swelling unrelated to meals) or lacrimal gland inflammation. Exclude other causes (infection). Specialist opinion may be required.	4	2
Significant proptosis	Protrusion of the eyeball due to significant amounts of inflammatory in the orbit; if unilateral, there should be a difference of 2 mm between one eye and the other. This may be associated with diplopia due to infiltration of extra-ocular muscles. Developing myopia (measured on best visual acuity, see later) can also be a manifestation of proptosis	4	2

Red eye (Epi)scleritis	Inflammation of the sclerae (specialist opinion usually required). Can be heralded by photophobia.	2	1
Red eye conjunctivitis	Inflammation of the conjunctivae (exclude infectious causes and excluding uveitis as cause of red eye, also exclude conjunctivitis sicca which should not be scored as this is not a feature of active vasculitis); (specialist opinion not usually required).	1	1
Blepharitis	Inflammation of eyelids. Exclude other causes (trauma, infection). Usually no specialist opinion is required		
Keratitis	Inflammation of central or peripheral cornea as evaluated by specialist		
Blurred vision	Altered measurement of best visual acuity from previous or baseline, requiring specialist opinion for further evaluation.	3	2
Sudden visual loss	Sudden loss of vision requiring ophthalmological assessment.	6	*
Uveitis	Inflammation of the uvea (iris, ciliary body, choroid) confirmed by ophthalmologist.	6	2
Retinal vasculitis	Retinal vessel sheathing on examination by specialist or confirmed by retinal fluorescein angiography	6	2
Retinal vessel thrombosis	Arterial or venous retinal blood vessel occlusion		
Retinal exudates	Any area of soft retinal exudates (exclude hard exudates) seen on ophthalmoscopic examination.		
Retinal haemorrhages	Any area of retinal haemorrhage seen on ophthalmoscopic examination.		

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

5. Chest		**6**	**3**
Wheeze	Wheeze on clinical examination	2	1
Nodules or cavities	New lesions, detected by CXR or CT.	3	*
Pleural effusion/pleurisy	Pleural pain and/or friction rub on clinical assessment or new onset of radiologically confirmed pleural effusion. Other causes (e.g. infection, malignancy) should be excluded	4	2
Infiltrate	Detected by CXR or CT scan. Other causes (infection) should be excluded	4	2

Endobronchial involvement	Endobronchial pseudotumor or ulcerative lesions. Other causes such as infection or malignancy should be excluded. NB: smooth stenotic lesions to be included in VDI; subglottic lesions to be recorded in the ENT section.	4	2
Massive haemoptysis/ alveolar haemorrhage	Major pulmonary bleeding, with extensive pulmonary infiltrates; other causes of bleeding should be excluded if possible. Patients are usually in extremis. Do not record minor episodes of haemoptysis.	6	4
Respiratory failure	Dyspnoea which is sufficiently severe as to require artificial ventilation	6	4

6. Cardiovascular		**6**	**3**
Loss of pulses	Loss of pulses in any vessel detected clinically; this may include loss of pulses leading to threatened loss of limb	4	1
Valvular heart disease	Significant valve abnormalities in the aortic mitral or pulmonary valves detected clinically or echocardiographically.	4	2
Pericarditis	Pericardial pain &/or friction rub on clinical assessment.	3	1
Ischaemic cardiac pain	Typical clinical history of cardiac pain leading to myocardial infarction or angina. Consider the possibility of more common causes (e.g. atherosclerosis)	4	2
Cardiomyopathy	Significant impairment of cardiac function due to poor ventricular wall motion confirmed on echocardiography	6	3
Congestive cardiac failure	Heart failure by history or clinical examination	6	3

7. Abdominal		**9**	**4**
Peritonism	Acute abdominal pain with peritonism/peritonitis due to perforation/infarction of small bowel, appendix or gallbladder etc., or acute pancreatitis confirmed by radiology/surgery/elevated amylase	9	3
Bloody diarrhoea	Of recent onset; inflammatory bowel disease and infectious causes excluded.	9	3
Ischaemic abdominal pain	Severe abdominal pain with typical features of ischaemia confirmed by imaging or at surgery, with typical appearances of aneurysms or abnormal vasculature characteristic of vasculitis.	6	2

8. Renal		**12**	**6**
Hypertension	Systolic BP>140 or Diastolic BP>95, accelerated or not, with or without retinal changes.	4	1
Proteinuria	>1+ on urinalysis; >0.2g/24 hours Infection should be excluded.	4	2
Haematuria	10 or more RBC per hpf (high power field), excluding urinary infection and urinary lithiasis (stone)or drug side effects (e.g. cyclophosphamide)	6	3
Creatinine 125-249	Serum creatinine values 125-249 µmol/l (1.41-2.82 mg/dL); only used at first assessment.	4	*
Creatinine 250-499	Serum creatinine values 250-499 µmol/l (2.83-5.64 mg/dL); only used at first assessment.	6	*

Creatinine ≥ 500	Serum creatinine values 500 µmol/l(≥5.66mg/dL) or greater; only used at first assessment.	8	*
Rise in creatinine> 30% or creatinine clearance fall > 25%	Significant deterioration in renal function attributable to active vasculitis.	6	*

9. Nervous system		**9**	**6**
Headache	New, unaccustomed & persistent headache	1	1
Meningitis	Severe headache with neck stiffness ascribed to inflammatory meningitis after excluding infection/bleeding	3	1
Organic confusion	Impaired orientation, memory or other intellectual function in the absence of metabolic, psychiatric, pharmacological or toxic causes.	3	1
Seizures (not hypertensive)	Paroxysmal electrical discharges in the brain & producing characteristic physical changes including tonic & clonic movements & certain behavioural changes.	9	3
Stroke	Cerebrovascular accident resulting in focal neurological signs such as paresis, weakness, etc. A stroke due to other causes (e.g. atherosclerosis) should be considered & appropriate neurological advice is recommended	9	3
Cord lesion	Transverse myelitis with lower extremity weakness or sensory loss (usually with a detectable sensory level) with loss of sphincter control (rectal & urinary bladder).	9	3
Cranial nerve palsy	Facial nerve palsy, recurrent nerve palsy, oculomotor nerve palsy etc. excluding sensorineural hearing loss and ophthalmic symptoms due to inflammation	6	3
Sensory peripheral neuropathy	Sensory neuropathy resulting in glove &/or stocking distribution of sensory loss. Other causes should be excluded (e.g. idiopathic, metabolic, vitamin deficiencies, infectious, toxic, hereditary).	6	3
Motor mononeuritis multiplex	Simultaneous neuritis of peripheral nerves, only scored if motor involvement. Other causes should be excluded (diabetes, sarcoidosis, carcinoma, amyloidosis).	9	3

10. Other	Other features of active vasculitis-please describe. Remember that you should review the rest of the BVAS item list before writing the feature in this section because it may already have been described elsewhere on the form. Do not use this section to record laboratory, imaging or pathology findings. Items recorded in this section will not carry any value when calculating the BVAS score	**0**	**0**