Manual of operations for assessment vasculitis using BVAS

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Section 1- Introduction

Welcome to this brief review of assessment of vasculitis using the BVAS (Birmingham Vasculitis Activity Score). This assessment method is a clinical evaluation tool which has been standardised for the management of patients with systemic vasculitis. It is important to standardise how we use this tool so that we agree definitions of disease activity. This brief summary is designed to help you understand the concepts of BVAS as well as providing you with some simple practical exercises on completing BVAS before applying it to patients with systemic vasculitis. A full version of this manual is available on line at the following address:

http://homepages.ed.ac.uk/rluq/vasculitistraining.htm

1.1 Aims and Objectives

Systemic vasculitis is now viewed as chronic disease rather than a fatal condition due to improvement in survival by treating with chemotherapy. Death as the main study come is no longer appropriate, standardized methods of assessment of morbidity are available to accurately measuring outcome of vasculitis and its effects, which consists of:

- current disease activity
- increasing damage, which consists of non-healing scars developing as a consequence to initial disease, eg renal failure, hypertension

There are three aspects of assessment tools used for outcome of clinical trials in vasculitis:

1. BVAS-used to record current disease activity
2. VDI (vasculitis damage index)-used to record damage as a result of vasculitis and its treatment
3. SF-36 patient questionnaire-used to measure physical and mental function

1.2 Methods

This training programme is designed to introduce you to BVAS so that we can standardise the way in which we collect data in clinical trials.

1.3 Assessment of Vasculitis Disease Activity

BVAS (Birmingham Vasculitis Activity Score)

It is designed to document new or worsening clinically active vasculitis that would be likely to require treatment, after exclusion of other causes such as infection, hypertension, etc. It consists of a set of items divided into nine organ based systems. The scoring sheet records the presence or absence of each item. Each item is weighted and a maximum total score applied to each system. The total score on all nine systems gives an indication of the current disease activity.

1.4 Practical Notes on how to Complete BVAS

Modified from Luqmani R. A., Bacon P. A., Moots R. J et al (1994) BVAS in systemic necrotizing vasculitis. Quart J Med 87: 671-678

Three questions should be asked of each item on the scoring sheet on completion.

1. Is the abnormality present?
2. Can it be attributed to active vasculitis?
3. If it can be attributed to active vasculitis, is it newly present or worse or does it simply reflect low grade grumbling disease activity?

- Scoring all items that are present and attributable to vasculitis as New/Worse for patient presented for the first time. In this case the items can have been present for over 3 months or even longer

- Scoring a patient for review with known disease

  if it is newly present or worse the New/Worse circle is ticked
  If it is present and has been present within the last 3 months (even if it is better now), the Persistent box is ticked
  if no item is present, an item has been present for over 3 months, or a patient reports having a symptom but it is not present at the time of scoring on questioning or examination, the None box is ticked

  NB If you tick a New/Worse circle there should be an intention to treat or to act on ticked items.

  eg. If a patient complains of new symptoms of joint pains and on examination there is no obvious joint swelling or tenderness and treatment is not to be changed or commenced, the item should not be ticked. However if on examination there is obvious joint inflammation and it is intended to increase the dose of steroids the New/Worse circle should be ticked

  NB As a general rule the persistent box is only ticked as stated above, if the item has been present and persistent within the last 3 months. There are however occasions when an item has been persistent for longer than 3 months but is obviously of concern and indicates ongoing active disease that needs addressing, the Persistent box should be ticked

  eg. Nose bleeding or bloody nasal discharge which are still occurring on a regular basis. In this instance the Persistent box should be ticked as treatment will need to be monitored. If the Persistent box is not ticked, it would not be apparent that the disease is still active and the steroid dose might be wrongly reduced, which may lead to worsening of the symptoms

1.5 Overview of BVAS

Disease features are scored only when they are due to active vasculitis, after exclusion of other obvious causes (eg infection, hypertension, etc). If the feature has occurred afresh or represents a recent deterioration of status since last visit, it is scored in the New/Worse box. If abnormality indicates the presence of active vasculitis (but not new or worse), the Persistent box is ticked.

For some occasions, further information (from specialist opinion or further tests) is required before entering some items. We would suggest that you leave these items blank, and once the information is available, please remember to fill in the information.

The data from the score sheet will be used to derive indices of disease activity as follows:

BVAS.1 (new/worse) - this represent a score of new/worse disease activity attributable to vasculitis

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BVAS.2 (persistent) - this represents a score of disease activity due to persisting or grumbling disease, which is neither new nor worse, compared to the previous assessment.

Scores are calculated using the values given to each item; each section has a maximum score, corresponding to the total value for BVAS (new/worse) and BVAS (persistent).

If the patient scores over the maximum number in a system, they only score the maximum score stipulated. Add the scores for each system together to get the final score.

There are 2 separate final scores

BVAS new/worse	max score 63
BVAS persistent	max score 33

The higher the scores the more active the disease

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VASCULITIS ACTIVITY SCORE

o Tick box **only** if abnormality is **newly present** since last assessment or **worse** in the last **few weeks** (use the Vasculitis Damage Index, VDI to score items of damage)
□ Tick box **only** if abnormality is due to **active** (but not new or worse) vasculitis
◊ Tick box if more information (specialist opinion/tests) is requested
@ oral/axillary temperatures; rectal temperatures are 0.5°C higher

DEMOGRAPHY
Trial Number
Visit Date / /
Investigator

	PERSISTENT	NEW/WORSE
1. GENERAL	□ (none)	
malaise	□	○
myalgia	□	○
arthralgia/arthritis	□	○
headache	□	○
fever (< 38.5°C)@	□	○
fever (≥ 38.5°C)@	□	○
wt loss (≥ 2kg)	□	○
2. CUTANEOUS	□ (none)	
Infarct	□	○
purpura	□	○
other skin vasculitis	□	○
ulcer	□	○
gangrene	□	○
multiple digit gangrene	□	○
3. MUCOUS MEMBRANES/EYES	□ (none)	
mouth ulcers	□	○
genital ulcers	□	○
significant proptosis	□	○
red eye- conjunctivitis	□	○
red eye- epi/scleritis	□	○
blurred vision	□	○
sudden visual loss	□	○
ophthalmic opinion	◊	
no active vasculitis	○	
uveitis	○	
retinal exudates	○	
retinal haemorrhage	○	
4. ENT	□ (none)	
Nasal obstruction	□	○
Bloody nasal discharge	□	○
Nasal crusting	□	○
Sinus involvement	□	○
Hearing loss	□	○
Hoarseness/stridor	□	○
ENT opinion	◊	
no active vasculitis	○	
Granulomatous sinusitis	○	
Conductive hearing loss	○	
Sensorineural hearing loss	○	
Significant Subglottic inflammation	○	

	PERSISTENT	NEW/WORSE
5. CHEST	□ (none)	
persistent cough	□	○
dyspnoea or wheeze	□	○
Haemoptysis/haemorrhage	□	○
chest radiology performed	◊	
no active vasculitis	○	
nodules or cavities	○	
pleural effusion/pleurisy	○	
Infiltrate	○	
massive haemoptysis or alveolar haemorrhage	□	○
respiratory failure	□	○
6. CARDIOVASCULAR	□ (none)	
aortic incompetence	□	○
pericardial pain/rub	□	○
ischaemic cardiac pain	□	○
congestive cardiac failure	□	○
cardiology opinion/tests	◊	
no active vasculitis	○	
pericarditis	○	
myocardial infarct/angina	○	
cardiomyopathy	○	
7. ABDOMINAL	□ (none)	
severe abdominal pain	□	○
bloody diarrhoea	□	○
surgical opinion/tests	◊	
no active vasculitis	○	
gut perforation/infarct	○	
acute pancreatitis	○	
8. RENAL	□ (none)	
hypertension (diastol>95)	□	○
proteinuria >1+/>0.2g/24h)	□	○
haematuria>1+/>10rbc/ml)	□	○
creatinine 125-249 umol/l		○
creatinine 250-499 umol/l		○
creatinine >500 umol/l		○
rise in creatinine >30% or fall in creatinine clearance>25%		○
9. NERVOUS SYSTEM	□ (none)	
organic confusion/dementia	□	○
seizures(not hypertensive)	□	○
stroke	□	○
cord lesion	□	○
sensory peripheral neuropathy	□	○
cranial nerve palsy	□	○
motor mononeuritis multiplex	□	○
10. OTHER		
	□	○

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GLOSSARY for BVAS

GENERAL RULE: disease features are scored only when they are due to active vasculitis, after exclusion of other obvious causes (e.g. infection, hypertension, etc.). If the feature has occurred afresh or represents a recent deterioration of status since last visit, it is scored in the NEW/WORSE 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 box (Persistent) if the abnormality indicates the presence of active (but not new or worse) vasculitis. 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, on occasions, you may require further information before entering some items. We would suggest that you leave these items blank, and once the information is available, please remember to take the time to fill in the information. For example, if the patient has new onset of stridor, you would usually ask an ENT colleague to investigate this further to determine whether or not it is due to active Wegener's granulomatosis.

DERIVATION of BVAS.1 (new/worse) BVAS.2 (persistent) scores. The data from the score sheet will be used to derive indices of disease activity as follows:

BVAS.1 - This represents a score of new/worse disease activity attributable to vasculitis

BVAS.2 - This represents a score of disease activity due to persisting or grumbling disease, which is neither new nor worse, compared to the previous assessment..

Scores are calculated using the values given to each item as shown; each section has a maximum score, corresponding to the total value for BVAS (new/worse) and BVAS (persistent).

TERM	DEFINITION	BVAS persis-tent	BVAS new/ worse
1. General	**Maximum scores**	**2**	**3**
Malaise	A general feeling of tiredness, illness & discomfort.	1	1
Myalgia	Pain in the muscles	1	1
Arthralgia or arthritis	Pain in the joints or joint inflammation;	1	1
Headache	New, unaccustomed & persistent	1	1
Fever <38.5	Documented oral/axillary temperature elevation. Rectal temperatures are 0.5 C higher	1	1
Fever >=38.5	Documented oral/axillary temperature elevation. Rectal temperatures are 0.5 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	**Maximum scores**	**3**	**6**
Infarct	Area of tissue necrosis or splinter haemorrhages	1	2
Purpura	Petechiae (small red spots), palpable purpura, or ecchymoses (large plaques) in skin or oozing (in the absence of trauma) in the mucous membranes.	1	2
Other skin vasculitis	e.g., livedo reticularis, nodules etc.	2	2
Ulcer	Open sore in a skin surface.	1	4
Gangrene	Extensive tissue necrosis (e.g. digit)	1	6
Multiple digit gangrene	Extensive tissue necrosis occurring in more than one digit or limb.	2	6
3. Mucous membranes/eyes	**Maximum score**	**3**	**6**
Mouth ulcers	Ulcers localised in the mouth. Exclude other causes, such as drugs, Crohn's disease, pemphigus etc.	1	1
Genital ulcers	Ulcers localised in the genitalia or perineum.	1	1
Significant proptosis	Protrusion of the eyeball due to significant amounts of inflammatory in the orbit. This may be associated with diplopia due to infiltration of extra-ocular muscles.	2	4
Red eye conjunctivitis	Inflammation of the conjuctivae (exclude infectious causes); (specialist opinion not usually required).	1	1
Red eye (Epi)scleritis	Inflammation of the sclerae (specialist opinion not usually required).	1	2
Blurred vision	Significant impairment of vision.	2	3
Sudden visual loss	Sudden loss of vision requiring ophthalmological assessment.	-	6
Ophthalmic opinion	To diagnose & score retinal exudates, haemorrhages, uveitis & reason for sudden visual loss. This data must be entered on score sheets subsequently.		
Uveitis*	Inflammation of the uvea (iris, ciliary body, choroid) confirmed by ophthalmologist.	-	6
Retinal exudates*	Any area of soft retinal exudates (exclude hard exudates) seen on ophthalmoscopic examination.	-	6
Retinal haemorrhages*	Any area of retinal haemorrhage seen on ophthalmoscopic examination.	-	6
4. ENT	**Maximum scores**	**3**	**6**
Nasal obstruction	A history of nasal blockage	1	2
Bloody nasal discharge	Blood stained secretions from the nose, irrespective of severity, or frequency & severity of previously occurring bleeding since last visit.	2	4
Nasal crusting	Discharge of large serous or serosanguinous crusts from either nostril.	2	4
Sinus involvement	Tenderness or pain over paranasal sinuses or X-ray evidence of sinusitis. If nasal bridge collapse is observed, this may be recorded separately (in 10. Other)	1	2
Hearing loss	Significant new hearing loss requiring specialist opinion.	-	3
Hoarseness/stridor	Increasing hoarseness & inspiratory stridor.	2	5
ENT opinion	To ascribe otitis media, deafness, or diagnose subglottic involvement due to vasculitis. This data can be entered on score sheets subsequently.		
Granulomatous sinusitis*	Characteristic appearance on nasal examination	-	4
Conductive hearing loss*	Any hearing loss due to middle ear involvement preferably confirmed by audiometry.	-	3
Sensorineural hearing loss*	Deafness attributable to auditory nerve or cochlear damage.	-	6
Significant subglottic inflammation*	Inspiratory stridor with significant narrowing of subglottic space confirmed by further examination (usually by an ENT specialist) or by radiological assessment	-	6

5. Chest	**Maximum scores**	**3**	**6**
Persistent cough	Cough for more than 2 weeks (other causes for the cough having been excluded e.g. infection)	1	2
Dyspnoea or wheeze	Shortness of breath or audible wheeze on exercise, by history &/or clinical examination.	1	2
Haemoptysis/ haemorrhage	Production of blood stained sputum. Other causes (e.g. infection, cancer) should be excluded.	1	3
Chest radiology performed	A chest radiograph should be performed if there are significant signs or symptoms to suggest chest disease or in the presence of a generalised flare - to determine the following three:		
Nodules or cavities*	New lesions, detected by CXR.	-	3
Pleural effusion/pleurisy*	Pleural pain &/or friction rub on clinical assessment or new onset of radiologically confirmed pleural effusion. Other causes (e.g. infection, cancer) should be excluded.	-	4
Infiltrate	By CXR, CT scan.	-	4
Massive haemoptysis/ Alveolar haemorrhage	Major pulmonary bleeding, with shifting pulmonary infiltrates & usually associated with signs of shock; other causes of bleeding should be excluded.	-	6
Respiratory failure	Dyspnoea which is sufficiently severe as to require artificial ventilation; arterial blood gases should be performed to confirm the presence of hypoxaemia & or hypercapnia.	3	6
6. Cardiovascular	**Maximum scores**	**3**	**6**
Aortic incompetence	Significant aortic valve regurgitation, detected clinically or echocardiographically.	2	4
Pericardial pain/rub	Pericardial pain &/or friction rub on clinical assessment.	2	3
Ischaemic cardiac pain	Typical clinical history of cardiac pain. Consider the possibility of more common causes (e.g. atherosclerosis)	2	4
Congestive cardiac failure	By history or clinical examination	2	4
Cardiology opinion or tests	Specialist opinion/tests are usually required to determine the following features		
Pericarditis*	Pericardial pain &/or friction rub on clinical assessment.	-	4
Myocardial infarction/angina*	Typical history of cardiac pain.	-	6
Cardiomyopathy*	Heart failure by history or clinical examination		6
7. Abdominal	**Maximum scores**	**4**	**9**
Severe abdominal pain	Of recent onset & attributed to vasculitis.	2	3
Bloody diarrhoea	Of recent onset, not due to known inflammatory bowel disease, etc.	2	3
Surgical opinion/ tests	Specialist opinion/tests required to determine the cause of abdominal pain or diarrhoea if they are of recent onset or worse since last visit.		
Gut perforation/ infarction*	Typical pain & peritonism includes gall bladder or appendix. Confirmed by X-ray or at surgery.	-	9
Acute pancreatitis*	Typical history & clinical examination findings of acute abdominal pain & tenderness with guarding. Confirmed by elevated serum amylase & a surgical opinion	-	9
8. Renal	**Maximum scores**	**6**	**12**
Hypertension	Diastolic BP>95, accelerated or not, with or without retinal changes.	1	4
Proteinuria	>1+ on urinalysis; >0.2g/24 hours. Infection should be excluded.	2	4
Haematuria	>1+ on urinalysis; >10 rbc/ml, or red cell casts seen on urine microscopy. Infection should be excluded.	3	6
Creatinine 125-249	Serum creatinine values 125-249 umol/l at first assessment only.	2	4
Creatinine 250-499	Serum creatinine values 250-499 umol/l at first assessment only.	3	6
Creatinine >=500	Serum creatinine values 500 umol/l or greater at first assessment only.	4	8
Rise in creatinine > 30% or creatinine clearance fall > 25%	Significant deterioration in renal function attributable to active vasculitis.	-	6
9. Nervous system	**Maximum scores**	**6**	**9**
Organic confusion/ Dementia	Impaired orientation, memory or other intellectual function in the absence of metabolic, psychiatric, pharmacological or toxic causes.	1	3
Seizures (not hypertensive)	Paroxysmal electrical discharges in the brain & producing characteristic physical changes including tonic & clonic movements & certain behavioural changes.	3	9
Stroke	Cerebrovascular accident resulting in focal neurological signs such as paresis, weakness, etc. A stroke due to other causes (eg atherosclerosis) should be considered & appropriate neurological advice is recommended	3	9
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).	3	9
Sensory Peripheral neuropathy	Sensory neuropathy resulting in glove &/or stoking distribution of sensory loss. Other causes should be excluded (e.g. idiopathic, metabolic, vitamin deficiencies, infectious, toxic, hereditary).	3	6
Cranial nerve palsy	Isolated acute cranial nerve palsy, excluding sensorineural hearing loss, or optic nerve lesion secondary to retro-orbital mass.	3	6
Motor mononeuritis multiplex	Simultaneous neuritis of many peripheral nerves, only scored if motor involvement. Other causes should be excluded (diabetes, sarcoidosis, carcinoma, amyloidosis).	3	9
10. Other	**Significant features attributable to active vasculitis not listed above.**		
	Total maximum score	**33**	**63**

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VASCULITIS ACTIVITY SCORE

o Tick box **only** if abnormality is newly present since last assessment or **worse** in the last **few weeks** (use the Vasculitis Damage Index, VDI to score items of damage)

Tick box **only** if abnormality is due to **active** (but not new or worse) vasculitis

[] Tick box if more information (specialist opinion/tests) is requested

@ oral/axillary temperatures; rectal temperatures are 0.5 C higher

1. Subject ID:

2. Clinic ID:

3. Date form completed: day / month / year

4. Investigator:

	PERSISTENT	NEW/WORSE
1. GENERAL	☐ (none)	
malaise	☐	○
myalgia	☐	○
arthralgia/arthritis	☐	○
headache	☐	○
fever (<38.5C) @	☐	○
fever (>=38.5C) @	☐	○
wt loss (>=2kg)	☐	○
2. CUTANEOUS	☐ (none)	
Infarct	☐	○
purpura	☐	○
other skin vasculitis	☐	○
ulcer	☐	○
gangrene	☐	○
multiple digit gangrene	☐	○
3. MUCOUS MEMBRANES/EYES	☐ (none)	
mouth ulcers	☐	○
genital ulcers	☐	○
significant proptosis	☐	○
red eye conjunctivitis	☐	○
red eye- epi/scleritis	☐	○
blurred vision	☐	○
sudden visual loss	☐	○
ophthalmic opinion	[]	
no active vasculitis	○	
uveitis	○	
retinal exudates	○	
retinal haemorrhage	○	
4. ENT	☐ (none)	
Nasal obstruction	☐	○
Bloddy nasal discharge	☐	○
Nasal crusting	☐	○
Sinus involvement	☐	○
Hearing loss	☐	○
Hoarseness/stridor	☐	○
ENT opinion	[]	
no active vasculitis	○	
Granulomatous sinusitis	○	
Conductive hearing loss	○	
Sensorineural hearing loss	○	
Significant Subglottic inflammation	○	

	PERSISTENT	NEW/WORSE
5. CHEST	☐ (none)	
persistent cough	☐	○
dyspnoea or wheeze	☐	○
Haemoptysis/haemorrhage	☐	○
chest radiology performed	[]	
no active vasculits	○	
nodules or cavities	○	
pleural effusion/pleurisy	○	
Infiltrate	○	
massive haemoptysis or alveolar haemorrhage	☐	○
respiratory failure	☐	○
6. CARDIOVASCULAR	☐ (none)	
aortic incompetence	☐	○
pericardial pain/rub	☐	○
ischaemic cardiac pain	☐	○
congestive cardiac failure	☐	○
cardiology opinion/tests	[]	
no active vasculitis	○	
pericarditis	○	
myocardial infarct/angina	○	
cardiomyopathy	○	
7. ABDOMINAL	☐ (none)	
severe abdominal pain	☐	○
bloddy diarrhoea	☐	○
surgical opinion/tests	[]	
no active vasculitis	○	
gut perforation/infarct	○	
acute pancreatitis	○	
8. RENAL	☐ (none)	
hypertension (diastol>95)	☐	○
proteinuria > 1+/>0.2g/24h)	☐	○
haematurial>1+/>10rbc/ml)	☐	○
creatinine 125-249 µmol/l		○
creatinine 250-499 µmol/l		○
creatinine > 500 µmol/l		○
rise in creatinine > 30% or fall in creatinine clearance > 25%		○
9. NERVOUS SYSTEM	☐ (none)	
organic confusion/dementia	☐	○
seizures (not hypertensive)	☐	○
stroke	☐	○
cord lesion	☐	○
sensory peripheral neuropathy	☐	○
cranial nerve palsy	☐	○
motor mononeuritis multiplex	☐	○
10. OTHER	☐	○