Centre …… Assessor …… Paper case ……

Patient Initials / No …..... Assessment number …… Date ……. *BVAS 2003 Study*

VASCULITIS ACTIVITY SCORE 2003

○ Tick box **only** if abnormality represents active disease (use the Vasculitis Damage Index, VDI to score items of damage). If there are no abnormalities in a system, please tick the "None" box

□ If **all** the abnormalities recorded represent smouldering/low grade/ grumbling disease, and there are no new/worse features, please remember to tick the box at the bottom right corner

	None	Active disease
1. General	○	
Myalgia		○
Arthralgia or arthritis		○
Fever ≥ 38.0 ^{0}C		○
Weight Loss ≥ 2 kg		○
2. Cutaneous	○	
Infarct		○
Purpura		○
Ulcer		○
Gangrene		○
Other skin vasculitis		○
3. Mucous membranes/eyes	○	
Mouth ulcers/granulomata		○
Genital ulcers		○
Adnexal inflammation		○
Significant proptosis		○
Red eye (Epi)scleritis		○
Red eye conjunctivitis/ blepharitis/keratitis		○
Blurred vision		○
Sudden visual loss		○
Uveitis		○
Retinal vasculitis/retinal vessel Thrombosis/retinal exudates/ Retinal haemorrhages		○
4. ENT	○	
Bloody nasal discharge/nasal Crusts/ulcers and/or granulomata		○
Paranasal sinus involvement		○
Subglottic stenosis		○
Conductive hearing loss		○
Sensorineural hearing loss		○
5. Chest	○	
Wheeze		○
Nodules or cavities		○
Pleural effusion/pleurisy		○
Infiltrate		○
Endobronchial involvement		○
Massive haemoptysis/Alveolar haemorrhage		○
Respiratory failure		○

	None	Active disease
6. Cardiovascular	○	
Loss of pulses		○
Valvular heart disease		○
Pericarditis		○
Ischaemic cardiac pain		○
Cardiomyopathy		○
Congestive cardiac failure		○
7. Abdominal	○	
Peritonitis		○
Bloody diarrhoea		○
Ischaemic abdominal pain		○
8. Renal	○	
Hypertension		○
Proteinuria >1+		○
Haematuria ≥10 rbc/hpf		○
Creatinine 125-249 µmol/l		○
Creatinine 250-499 µmol/l		○
Creatinine ≥ 500 µmol/l		○
Rise in creatinine > 30% or Creatinine clearance fall > 25%		○
9. Nervous system	○	
Headache		○
Meningitis		○
Organic confusion		○
Seizures (not hypertensive)		○
Stroke		○
Cord lesion		○
Cranial nerve palsy		○
Sensory peripheral neuropathy		○
Motor mononeuritis multiplex		○
10. OTHER	○	
		○
		○
		○
		○

PERSISTENT DISEASE ONLY:

Tick here if **all** the above abnormalities are due to low grade grumbling disease and not due to new/ worse disease □

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

Glossary and scoring for BVAS 2003. 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 ENT colleague to investigate this further to determine whether or not it is due to active Wegener's granulomatosis.		**BVAS persistent**	**BVAS 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 ≥ 38.0 ^{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	**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
Ulcer	Open sore in a skin surface.	1	4
Gangrene	Extensive tissue necrosis (e.g. digit)	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 and/or "strawberry" gingival hyperplasia, excluding lupus erythematosus, and infection	1	2
Genital ulcers	Ulcers localised 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 preferably required.	2	4
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	2	4
Red eye (Epi)scleritis	Inflammation of the sclerae (specialist opinion usually required). Can be heralded by photophobia.	1	2
Red eye conjunctivitis Blepharitis Keratitis	Inflammation of the conjuctivae (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). Inflammation of eyelids. Exclude other causes (trauma, infection). Usually no specialist opinion is required Inflammation of central or peripheral cornea as evaluated by specialist	1	1
Blurred vision	Altered measurement of best visual acuity from previous or baseline, requiring specialist opinion for further evaluation.	2	3
Sudden visual loss	Sudden loss of vision requiring ophthalmological assessment.		6
Uveitis	Inflammation of the uvea (iris, ciliary body, choroid) confirmed by ophthalmologist.	2	6
Retinal vasculitis	Retinal vessel sheathing on examination by specialist or confirmed by retinal fluoroscein angiography	2	6
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	**Maximum scores**	**3**	**6**
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	3	6
Paranasal sinus involvement	Tenderness or pain over paranasal sinuses usually with pathologic imaging (CT, MR, x-ray, ultrasound)	1	2
Subglottic stenosis	Stridor and 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 confirmed by otoscopy and/or tuning fork examination and/or audiometry	1	3
Sensorineural hearing loss	Hearing loss due to auditory nerve or cochlear damage confirmed by audiometry	2	6
5. Chest	**Maximum scores**	**3**	**6**
Wheeze	Wheeze on clinical examination	1	2
Nodules or cavities	New lesions, detected by CXR		3

		BVAS persistent	BVAS new/ worse
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	2	4
Infiltrate	Detected by CXR or CT scan. Other causes (infection) should be excluded	2	4
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.	2	4
Massive haemoptysis/alveolar haemorrhage	Major pulmonary bleeding, with shifting pulmonary infiltrates; other causes of bleeding should be excluded if possible	4	6
Respiratory failure	Dyspnoea which is sufficiently severe as to require artificial ventilation	4	6
6. Cardiovascular	**Maximum scores**	**3**	**6**
Loss of pulses	Loss of pulses in any vessel detected clinically; this may include loss of pulses leading to threatened loss of limb	1	4
Valvular heart disease	Significant valve abnormalities in the aortic mitral or pulmonary valves detected clinically or echocardiographically.	2	4
Pericarditis	Pericardial pain &/or friction rub on clinical assessment.	1	3
Ischaemic cardiac pain	Typical clinical history of cardiac pain leading to myocardial infarction or angina. Consider the possibility of more common causes (eg atherosclerosis)	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**
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	3	9
Bloody diarrhoea	Of recent onset; inflammatory bowel disease and infectious causes excluded.	3	9
Ischaemic abdominal pain	Severe abdominal pain with typical features of ischaemia confirmed by imaging or at surgery, with typical appearances of aneursyms or abnormal vasculature characteristic of vasculitis.	2	6
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	10 or more RBC per hpf (high power field), excluding urinary infection and urinary lithiasis (stone)	3	6
Creatinine 125-249	Serum creatinine values 125-249 µmol/l at first assessment only.	2	4
Creatinine 250-499	Serum creatinine values 250-499 µmol/l at first assessment only.	3	6
Creatinine ≥ 500	Serum creatinine values 500 µmol/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**
Headache	New, unaccustomed & persistent headache	1	1
Meningitis	Severe headache with neck stiffness ascribed to inflammatory meningitis after excluding infection/bleeding	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)	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
Cranial nerve palsy	Facial nerve palsy, recurrent nerve palsy, oculomotor nerve palsy etc. excluding sensorineural hearing loss and ophthalmic symptoms due to inflammation	3	6
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).	3	6
Motor mononeuritis multiplex	Simultaneous neuritis of single or many peripheral nerves, only scored if motor involvement. Other causes should be excluded (diabetes, sarcoidosis, carcinoma, amyloidosis).	3	9
10. OTHER	Other feature of active vasculitis-please describe		