DCVAS SCREENING

Screening Date	D D / M M M / Y Y Y Y
Screening ID								
Gender	☐ Male ☐ Female
Date of birth:	D D / M M M / Y Y Y Y

Inclusion criteria

		Yes	No
1.	Is the patient over the age of 18 years?	☐	☐
2.	Does the patient have a diagnosis of vasculitis? **OR** Is vasculitis a potential diagnosis for their current illness?	☐	☐
3.	Has the patient given informed consent? **OR** The patient does not have capacity to provide informed consent but a 'consultee' ("surrogate") has declared that the patient would want to participate in the study?	☐	☐

If "No" is ticked for any of the inclusion criteria, then patient is NOT eligible for the study.

I confirm that the patient:

Meets **ALL** the inclusion criteria for the study: ☐

Or

Does not meet the inclusion criteria for the study ☐

Signature of investigator :_____ Print name: _____

Clinical scenarios

Tick **all** that applies for this subject for their current illness:

- ☐ 1. Referred to secondary or tertiary care with a suspicion of vasculitis
- ☐ 2. Multi-system disease (presentation of disease with at least 2 organs involved)
- ☐ 3. Fever
- ☐ 4. Chronic signs and/or symptoms of upper airway disease
- ☐ 5. Haemoptysis/pulmonary haemorrhage
- ☐ 6. Acute respiratory distress, exacerbation of asthma, or unexplained pulmonary fibrosis
- ☐ 7. Acute or progressive renal impairment and/or failure
- ☐ 8. Acral ischaemia/necrosis
- ☐ 9. Ischaemic jaw or tongue pain
- ☐ 10. Limb claudication
- ☐ 11. Aortic aneurysm (>5cm, thoracic or abdominal)
- ☐ 12. New-onset hypertension associated with other systemic features
- ☐ 13. Acute or chronic abdominal pain
- ☐ 14. New-onset headache
- ☐ 15. Sudden visual loss
- ☐ 16. Other signs or symptoms of ocular disease (unexplained keratitis, scleritis, uveitis, orbital disease, other)
- ☐ 17. Stroke
- ☐ 18. Chronic headache
- ☐ 19. Peripheral neuropathy (either sensory or motor)
- ☐ 20. Inflammatory polyarthritis
- ☐ 21. Inflammatory shoulder and/or hip girdle symptoms (Polymyalgia-like symptoms)
- ☐ 22. Rash/skin abnormalities (including nodules and ulcers)
- ☐ 23. Peripheral blood eosinophilia
- ☐ 24. Positive serologic test for autoimmune disease (ANCA, ANA, other)

- ☐ 25. Biopsy specimen consistent with vasculitis

- ☐ 26. Other (please specify): ☐

DCVAS STUDY VISIT 1

Visit date:	D D / M M M / Y Y Y Y
Date of onset of symptoms of current illness:	D D / M M M / Y Y Y Y
Date of diagnosis of current illness:	D D / M M M / Y Y Y Y
Date of first assessment of this patient by you :	D D / M M M / Y Y Y Y

Tobacco smoking status

- ☐ Current smoker
- ☐ Previous smoker
- ☐ Never smoked

Pre- existing co-morbidity

Present before the onset of current illness (please tick **all** that apply):

- ☐ Coronary heart disease
- ☐ Heart failure
- ☐ Peripheral vascular disease
- ☐ Hypertension requiring treatment
- ☐ Chronic obstructive pulmonary disease (not including late onset asthma)
- ☐ Asthma
- ☐ Diabetes mellitus
- ☐ Cerebrovascular accident (stroke)
- ☐ Dyslipidaemia

- ☐ Malignancy (please specify): _____
- ☐ Other (please specify): _____

Ethnicity

Tick the appropriate box to indicate ethnic background. If mixed ethnicity more than 1 box may be ticked.

- ☐ African North
- ☐ African Sub Saharan
- ☐ African-American
- ☐ Black Caribbean
- ☐ Chinese Han
- ☐ Chinese Other
- ☐ European North
- ☐ European South (Italian, Spanish, Portuguese, Greek, Balkan)
- ☐ Indian (Indian/Pakistani/Bangladeshi)
- ☐ Japanese
- ☐ Korean
- ☐ Latin American - Native
- ☐ Latin American - Mestizo
- ☐ Middle Eastern
- ☐ Pacific Islander
- ☐ Turkish
- ☐ White Caucasian American
- ☐ Other (please specify):

Clinical Features

1. General

Is this the patients 1st presentation for this illness? Yes ☐ No ☐

Current height		cm
Current weight		Kg
Systolic blood pressure left arm		mmHg
Systolic blood pressure right arm		mmHg

Present at any time since the onset of the current illness = Tick this box if symptoms or sign were present during the current illness. The current illness includes the whole period from when you think the vasculitis or mimic disease started till present day. This includes items that may have occurred and disappeared in the course of this illness.

Please tick if present at any time since the onset of the current illness

SYMPTOMS

- ☐ Light-headedness
- ☐ Syncope / Fainting
- ☐ Fatigue
- ☐ Night sweats
- ☐ Rigors

CLINICAL FEATURES

- ☐ Fever ≥ 38°C (≥ 100.4F)
- ☐ Lymphadenopathy in more than one anatomic chain
- ☐ Scalp tenderness

Weight loss this illness:

0Kg	≤2Kg	2-5Kg	≥5kg

2. Musculoskeletal Not involved ☐

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Please tick if present at any time since the onset of the current illness

SYMPTOMS

- ☐ Arthralgia (Joint pain)
- ☐ Morning stiffness ≥ 1 hour
- ☐ Myalgia (muscle pain) or muscle cramps
- ☐ Other (please specify):

CLINICAL FEATURES

- ☐ Swollen or inflamed joint(s)
- ☐ Muscle tenderness
- ☐ Muscle weakness
- ☐ Other (please specify):

3. Skin Not involved ☐

Present at any time since the onset of the current illness = Tick this box if symptoms or sign were present during the current illness. The current illness includes the whole period from when you think the vasculitis or mimic disease started till present day. This includes items that may have occurred and disappeared in the course of this illness.

Please tick if present at any time since the onset of the current illness

SYMPTOMS

- ☐ Pruritus
- ☐ Painful skin lesions of any type
- ☐ Other (please specify): _____

CLINICAL FEATURES

- ☐ Petechiae or Purpura (subcutaneous or dermal haemorrhage of any size)
- ☐ Maculopapular or papular rash (exanthema of macules and papules)
- ☐ Livedo reticularis (mottled cyanotic discoloration of the skin, with characteristic network pattern)
- ☐ Livedo racemosa ('broken' livedo - mottled cyanotic discoloration of the skin, with characteristic open network in the shape of lightening)
- ☐ Non tender skin nodules
- ☐ Tender skin nodules
- ☐ Gangrene (necrosis of tissue, usually due to loss of blood supply)
- ☐ Splinter haemorrhage (thin longitudinal lines of blood extravasation in the nail bed)
- ☐ Ulcer (defect in which the epidermis and upper part of dermis are lacking)
- ☐ Urticaria/Wheals/ Hives
- ☐ Other (please specify): _____

4. Eyes Not involved ☐

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Please tick if present at any time since the onset of the current illness

SYMPTOMS

- ☐ Amaurosis fugax (transient/temporary loss of vision)
- ☐ Sudden visual loss - ongoing
- ☐ Blurred vision in either eye
- ☐ Painful eye(s)
- ☐ Other (please specify): _____

CLINICAL FEATURES

- ☐ Conjunctivitis
- ☐ Watery eyes or lacrimal gland enlargement
- ☐ Proptosis / Exophthalmos (bulging out of one or both of the eyes)
- ☐ Ischaemic Optic Neuritis (confirmed by ophthalmologist)

Ocular inflammation (confirmed by an ophthalmologist)

- ☐ Keratitis (inflammation of the cornea)
- ☐ Scleritis or episcleritis (inflammation of the sclera)
- ☐ Uveitis (inflammation of any of the following: iris, anterior chamber, vitreous cavity, retina or choroid)

Retinal pathology (confirmed by an ophthalmologist)

- ☐ Retinal exudates (e.g. cotton wool spots)
- ☐ Retinal aneurysms
- ☐ Retinal haemorrhages (including flame haemorrhages)
- ☐ Retinal vessel thrombosis
- ☐ Other (please specify): _____

5. Ear Nose and Throat Not involved ☐

Present at any time since the onset of the current illness = Tick this box if symptoms or sign were present during the current illness. The current illness includes the whole period from when you think the vasculitis or mimic disease started till present day. This includes items that may have occurred and disappeared in the course of this illness.

Please tick if present at any time since the onset of the current illness

SYMPTOMS

- ☐ Jaw claudication
- ☐ Hearing loss or reduction
- ☐ Bloody nasal discharge
- ☐ Non blood stained nasal discharge
- ☐ Sino nasal congestion or blockage
- ☐ Loss of smell (anosmia)
- ☐ Loss of taste (ageusia)
- ☐ Other (please specify): _____

CLINICAL FEATURES

- ☐ Inflamed ear or nose cartilage
- ☐ Saddle nose deformity (nasal bridge collapse)
- ☐ Conductive hearing loss
- ☐ Sensorineural hearing loss
- ☐ Nasal polyps
- ☐ Nasal ulcers, mucosal abnormalities or crusting
- ☐ Nasal septal defect / perforation
- ☐ Hoarse voice/stridor
- ☐ Salivary gland enlargement
- ☐ Subglottic stenosis with no previous surgery
- ☐ Subglottic stenosis with prior surgery or intubation
- ☐ Other (please specify): _____

6. Chest / Pulmonary Not involved ☐

Present at any time since the onset of the current illness = Tick this box if symptoms or sign were present during the current illness. The current illness includes the whole period from when you think the vasculitis or mimic disease started till present day. This includes items that may have occurred and disappeared in the course of this illness.

Please tick if present at any time since the onset of the current illness

SYMPTOMS

- ☐ Dyspnoea / Shortness of Breath
- ☐ Dry cough
- ☐ Moist cough but unable to expectorate sputum
- ☐ Productive cough with purulent sputum
- ☐ Minor haemoptysis (few streaks of blood only)
- ☐ Major haemoptysis (> 1 teaspoon of blood in a day)
- ☐ Sharp chest pain on inspiration
- ☐ Other (please specify): _____

CLINICAL FEATURES

- ☐ Chest wall tenderness
- ☐ Crackles / râles on auscultation
- ☐ Wheeze or evidence of obstructive airways disease
- ☐ Respiratory compromise requiring oxygen
- ☐ Respiratory failure requiring intubation
- ☐ Other (please specify): _____

Lung Function Not performed ☐

Please tick if present on lung function testing at any time since the onset of the current illness	
☐	Restrictive airways disease
☐	Obstructive airways disease
☐	Reduced DLCO or KCO
☐	Increased KCO
☐	Other (specify):

Bronchoscopy Not performed ☐

Please tick if present on bronchoscopy at any time since the onset of the current illness	
☐	Mass or tumour
☐	Bronchitic changes or mucosal injury
☐	Friable mucosa
☐	Evidence of alveolar haemorrhage
☐	Blood stained broncho alveolar lavage
☐	Purulent or muco-purulent secretions
☐	Other (specify):

7. Cardiovascular Not involved ☐

Present at any time since the onset of the current illness = Tick this box if symptoms or sign were present during the current illness. The current illness includes the whole period from when you think the vasculitis or mimic disease started till present day. This includes items that may have occurred and disappeared in the course of this illness.

Please tick if present at any time since the onset of the current illness

SYMPTOMS

- ☐ Angina / ischaemic cardiac pain
- ☐ Arm claudication
- ☐ Leg claudication
- ☐ Carotodynia (neck pain in the region of the carotid artery)
- ☐ Raynaud's phenomenon
- ☐ Other (please specify):

CLINICAL FEATURES

- ☐ Any cardiac murmur
- ☐ Congestive cardiac failure / Cardiomyopathy
- ☐ Myocardial infarction confirmed by ECG or blood test results
- ☐ Other (please specify):

Vascular Examination

No abnormality on vascular examination ☐

Artery	Left					Right				
	Pulse		Tenderness	Bruit	Hard 'cord-like'	Pulse		Tenderness	Bruit	Hard 'cord-like'
	Diminished	Absent				Diminished	Absent			
Temporal	☐	☐	☐		☐	☐	☐	☐		☐
Carotid	☐	☐	☐	☐		☐	☐	☐	☐	
Subclavian			☐	☐					☐	
Axillary	☐	☐	☐			☐	☐	☐		
Brachial	☐	☐	☐	☐		☐	☐	☐	☐	
Radial	☐	☐	☐	☐		☐	☐	☐	☐	
Abdominal Aorta			☐	☐				☐	☐	
Renal				☐					☐	
Femoral	☐	☐	☐	☐		☐	☐	☐	☐	
Popliteal	☐	☐	☐	☐		☐	☐	☐	☐	
Posterior tibial	☐	☐	☐			☐	☐	☐		
Dorsalis pedis	☐	☐	☐			☐	☐	☐		

8. Gastrointestinal Not involved ☐

Present at any time since the onset of the current illness = Tick this box if symptoms or sign were present during the current illness. The current illness includes the whole period from when you think the vasculitis or mimic disease started till present day. This includes items that may have occurred and disappeared in the course of this illness.

Please tick if present at any time since the onset of the current illness

SYMPTOMS

- ☐ Abdominal pain (any)
- ☐ Postprandial abdominal pain
- ☐ Claudication of tongue
- ☐ Diarrhoea
- ☐ Dysphagia (difficulty swallowing)
- ☐ Symptoms of GI blood loss (e.g. history of malena)
- ☐ Other (please specify): ☐

CLINICAL FEATURES

- ☐ Mouth ulcers
- ☐ Jaundice
- ☐ Peritonism (rigid abdomen with rebound tenderness)
- ☐ Acute pancreatitis confirmed by elevated amylase
- ☐ Chronic pancreatitis confirmed on blood tests and/or imaging
- ☐ Colitis confirmed by endoscopy
- ☐ Other (please specify): ☐

9. Genitourinary Not involved □

Present at any time since the onset of the current illness = Tick this box if symptoms or sign were present during the current illness. The current illness includes the whole period from when you think the vasculitis or mimic disease started till present day. This includes items that may have occurred and disappeared in the course of this illness.

Please tick if present at any time since the onset of the current illness

SYMPTOMS

- ☐ Macroscopic haematuria (blood visible in urine)
- ☐ Dysuria (pain with passing urine)
- ☐ Difficulty passing urine (e.g. due to urinary obstruction)
- ☐ Passing renal stone / calculi
- ☐ Flank Pain
- ☐ Testicular pain
- ☐ Other (please specify): _____

CLINICAL FEATURES

- ☐ Testicular tenderness
- ☐ Testicular swelling
- ☐ Genital ulcers
- ☐ Other (please specify): _____

10. Neurologic Not involved □

Present at any time since the onset of the current illness = Tick this box if symptoms or sign were present during the current illness. The current illness includes the whole period from when you think the vasculitis or mimic disease started till present day. This includes items that may have occurred and disappeared in the course of this illness.

Please tick if present at any time since the onset of the current illness

SYMPTOMS

- ☐ Seizures
- ☐ Transient ischaemic attack (neurological symptoms consistent with brain ischaemia lasting less than 24 hours)
- ☐ Photophobia
- ☐ **New persistent headache (tick all that apply)**
  - ☐ Frontal
  - ☐ Occipital or cervical
  - ☐ Temporal
- ☐ Other (please specify): _____

CLINICAL FEATURES

- ☐ Cranial nerve palsy
- ☐ Relative afferent pupillary defect
- ☐ Meningism
- ☐ Motor neuropathy (not due to radiculopathy)
- ☐ Sensory neuropathy (not due to radiculopathy)
- ☐ Hemiparesis or hemiplegia
- ☐ Paraparesis or paraplegia
- ☐ Psychosis
- ☐ Other (please specify): _____

Vaccinations, Medications and Other Drugs None apply ☐

Please tick if recently used (current or less than 12 months before onset of illness)

VACCINATION

- ☐ Influenza vaccine
- ☐ H1N1 (swine flu) vaccine
- ☐ Pneumococcal vaccine
- ☐ Hepatitis B vaccine
- ☐ Other Vaccine (specify): _____

SPECIFIC MEDICATIONS (known to be associated with systemic vasculitis)

- ☐ Allopurinol
- ☐ Carbimazole
- ☐ Clozapine
- ☐ Ergot derivative
- ☐ Hydralazine
- ☐ Methysergide
- ☐ Phenytoin
- ☐ Propylthiouracil (PTU)
- ☐ Sulfasalazine
- ☐ Leukotriene antagonist
- ☐ Any other specific medication(s) that you think may be causing patients **systemic** symptoms (not just skin vasculitis):

_____

OTHER DRUGS

- ☐ Cocaine
- ☐ Amphetamine or Methamphetamine
- ☐ Other sympathomimetics (e.g. ephedrine or pseudoephedrine)

Biopsy

No biopsies were done ☐

Please write down the number which corresponds to site/organ biopsied and indicate ALL findings that apply.

Site of biopsy ☐ **Finding(s):** ☐☐☐☐☐☐

Site of biopsy ☐ **Finding(s):** ☐☐☐☐☐☐

Site of biopsy ☐ **Finding(s):** ☐☐☐☐☐☐

Site of biopsy ☐ **Finding(s):** ☐☐☐☐☐☐

Site of biopsy	Findings	
1. None 2. Skin 3. Kidney 4. Temporal artery 5. ENT 6. Nerve 7. Muscle 8. Liver 9. Gut 10. Lung parenchyma 11. Bronchial 12. Heart 13. Brain/meningeal 14. Aorta 15. Bladder 16. Prostate 17. Bone marrow 18. Lymph node 19. Other	**General** 1. Not done 2. Normal 3. Non diagnostic 4. Biopsy consistent with vasculitis but not definite 5. Definite vasculitis **Other findings** 6. Benign tumour 7. Malignant tumour 8. Abscess 9. Organisms seen on microscopy 10. Culture of microorganism from biopsy specimen 11. Unspecified tissue inflammation 12. Tissue fibrosis (e.g. liver or lung fibrosis) 13. Amyloid 14. Age related vascular changes **Vascular specific findings** 15. Arteritis of any artery in a needle, punch or other small biopsy; or of the smallest arteries in a surgical specimen 16. Arteritis in medium sized arteries in a surgical specimen (excluding needle and other small biopsies) 17. Arteritis of a large artery in a surgical specimen (excluding needle and other small biopsies but including arterial segment biopsies, e.g. temporal artery) 18. Aortitis in a surgically removed aortic segment 19. Necrotizing arteritis with fibrinoid necrosis 20. Inflammatory arterial aneurysm 21. Necrotizing or leukocytoclastic arteriolitis 22. Necrotizing or leukocytoclastic venulitis 23. Perivascular inflammation only without vessel wall involvement in a specimen	24. Immune complex deposits in vessels other than glomeruli with prominent IgA 25. Immune complex deposits in vessels other than glomeruli without prominent IgA 26. Absence or paucity of immune complex deposits in vessels other than glomeruli 27. Granulomatous vasculitis 28. Giant cells in vasculitis 29. Prominent neutrophils in vasculitis 30. Prominent eosinophils in vasculitis 31. Predominantly mononuclear leukocytes in vasculitis 32. Vascular scarring consistent with vasculitis but no active inflammation 33. Fragmentation of the internal elastic lamina 34. Intimal thickening 35. Vascular thrombosis **Organ Specific findings** 36. Pulmonary capillaritis or haemorrhage 37. Pauci-immune (≤2+ immunoglobulin) glomerulonephritis 38. Anti-GBM glomerulonephritis 39. Immune complex (>2+ immunoglobulin) glomerulonephritis 40. IgA-dominant Immune complex glomerulonephritis 41. Myelofibrosis or myelodysplasia **Extra vascular findings** 42. Extravascular granulomatous inflammation 43. Extravascular eosinophil-predominant inflammation 44. Extravascular non granulomatous acute or chronic inflammation

Laboratory tests – most representative of this illness & prior to treatment

	Present	Absent	Not done
HAEMATOLOGY			
Significant anaemia (Haemoglobin < 10g/dL or 100 g/L)	☐	☐	☐
Significant thrombocytopenia (platelets <100 x 10^9/L)	☐	☐	☐
Significant thrombocythaemia (platelets >500 x 10^9/L)	☐	☐	☐
Significant leukopenia (Total WBC < 3.0 x 10^9/L)	☐	☐	☐
Significant elevation of WBC (Total WBC >15.0x10^9/L)	☐	☐	☐
Significant neutropenia (PMN <1.5x10^9/L)	☐	☐	☐
Significant neutrophilia (PMN >10x10^9/L)	☐	☐	☐
Maximum eosinophil count (x10^9/L)			☐
Maximum ESR mm/hr			☐
BIOCHEMISTRY			
Maximum CRP Units			☐
Significant elevation of creatinine kinase (>500 IU/L)	☐	☐	☐
AST (SGOT) or ALT (SGPT) >2x upper limit of normal	☐	☐	☐
Alkaline phosphatase >2x upper limit of normal	☐	☐	☐
Hyperglycaemia (glucose > 15 mmol/L or 270 mg/dL)	☐	☐	☐
Maximum creatinine: Units	☐	☐	☐
Albumin below 30g/L	☐	☐	☐
Monoclonal gammopathy	☐	☐	☐

URINE TESTS **Not done** ☐

	Present	Absent	Not done
Protein on urine dipstick*	☐	☐	☐
Blood on urine dipstick*	☐	☐	☐
leucocytes or nitrites on urine dipstick*	☐	☐	☐
Red cell casts in urine	☐	☐	☐
24 hour urine protein >1g/L	☐	☐	☐
*Please tick **present** if there is ≥ a trace amount			

CRYOGLOBULINS AND COMPLEMENT **Not done** ☐

	Present	Absent	Not done
Cryoglobulins (please specify):	☐	☐	☐
Type I (monoclonal immunoglobulin) ☐			
Type II (monoclonal RF (IgM) and polyclonal IgG) ☐			
Type III (polyclonal RF (IgM) and polyclonal IgG) ☐			
Low C3	☐	☐	☐
Low C4	☐	☐	☐

AUTOANTIBODIES **Not done** ☐

	Present	Absent	Not done
cANCA on immunofluorescence	☐	☐	☐
pANCA on immunofluorescence	☐	☐	☐
PR3 ANCA (ELISA)	☐	☐	☐
MPO ANCA (ELISA)	☐	☐	☐
Other ANCA (specify):	☐	☐	☐
Anti GBM antibodies	☐	☐	☐
RF (Rheumatoid factor)	☐	☐	☐
ACPA (Anti CCP antibody)	☐	☐	☐
ANA (Anti nuclear antibody)	☐	☐	☐
Anti dsDNA	☐	☐	☐
ENA (extractable nuclear antigen)	☐	☐	☐
Ro (SS-A)	☐	☐	☐
La (SS-B)	☐	☐	☐
RNP	☐	☐	☐
Sm	☐	☐	☐
Scl-70	☐	☐	☐
Jo 1	☐	☐	☐
Lupus anticoagulant	☐	☐	☐
Anti-cardiolipin IgG	☐	☐	☐
Anti-cardiolipin IgM	☐	☐	☐
Anti Beta- 2- glycoprotein 1 (B2GP1)	☐	☐	☐

INFECTION SCREEN **Not done** ☐

		Positive	Negative	Not done
Blood culture (organism):		☐	☐	☐
Urine culture (organism):		☐	☐	☐
Sputum culture (organism):		☐	☐	☐
CSF culture (organism):		☐	☐	☐
HIV test		☐	☐	☐
Hepatitis A (evidence of acute infection only)		☐	☐	☐
Hepatitis B (evidence of acute or chronic infection)		☐	☐	☐
Hepatitis C (evidence of acute or chronic infection)		☐	☐	☐
CMV (evidence of acute or chronic infection)		☐	☐	☐
EBV (evidence of acute or chronic infection)		☐	☐	☐
Lyme disease (*Borrelia burgdorferi)*		☐	☐	☐
Other (specify):		☐	☐	☐

OTHER LABORATORY INVESTIGATIONS **Not done** ☐

		Present	Not done
CSF: elevated protein		☐	☐
CSF: oligoclonal bands		☐	☐
CSF: pleocytosis (increased white cell count in CSF)		☐	☐
CSF: xanthochromia (degraded blood in CSF indicating previous bleed)		☐	☐
		☐	☐
Other (specify):		☐	
Other (specify):		☐	
Other (specify):		☐	

Vascular imaging (most representative for this illness) Not done ☐

Please write down the number which corresponds to modality of imaging used, vessel imaged and findings based on imaging. More than one abnormal finding is possible, please indicate ALL that apply. Circle the side imaging was performed. L =Left, R=Right, NA=Not applicable.

Imaging Modality: ☐ **Vessel:** ☐ **Side: L/R/NA** **Finding(s):** ☐☐☐☐

Imaging Modality: ☐ **Vessel:** ☐ **Side: L/R/ NA** **Finding(s):** ☐☐☐☐

Imaging Modality: ☐ **Vessel:** ☐ **Side: L/R/NA** **Finding(s):** ☐☐☐☐

Imaging Modality: ☐ **Vessel:** ☐ **Side: L/R/ NA** **Finding(s):** ☐☐☐☐

Imaging Modality	Vessel imaged		Findings
1. None 2. CT 3. CT angiogram 4. MRI 5. MR angiogram 6. FDG-PET 7. Digital Subtraction Angiogram (DSA) 8. Ultrasound 9. Flourescein angiogram 10. Echocardiogram	1. None 2. Temporal artery 3. Intracranial artery 4. Retinal artery 5. Sagittal sinus or veins 6. Ascending thoracic aorta 7. Descending thoracic aorta 8. Abdominal aorta 9. Subclavian artery 10. Axillary artery 11. Carotid artery 12. Radial artery 13. Ulnar artery 14. Iliac artery 15. Femoral artery 16. Popliteal artery 17. Posterior tibial artery 18. Dorsalis pedis artery 19. Distal hand artery 20. Distal foot artery	21. Pulmonary artery 22. Coronary arteries 23. Coronary sinus 24. Renal artery 25. Splenic artery 26. Superior mesenteric artery 27. Inferior mesenteric artery 28. Popliteal vein 29. Femoral vein 30. Iliac vein 31. Inferior vena cava 32. Superior vena cava 33. Axillary vein 34. Other	1. Not done 2. Vessel narrowing 3. Vessel occlusion 4. Wall thickening 5. Delayed contrast enhancement 6. Enhanced FDG uptake 7. Aneurysm 8. Dissection 9. Beading or micro aneurysms 10. Calcification of vessel 11. Thrombus present 12. Halo sign positive 13. Normal 14. Other

Other imaging (most representative for this illness) Not done ☐

Please write down the number which corresponds to modality of imaging used, site imaged and imaging finding(s). More than one finding is possible, please indicate ALL that apply. (List is on opposite page) Use as many or as few boxes as required.

Imaging Modality: ☐ **Site imaged:** ☐ **Finding(s):** ☐ ☐ ☐ ☐

Imaging Modality: ☐ **Site imaged:** ☐ **Finding(s):** ☐ ☐ ☐ ☐

Imaging Modality: ☐ **Site imaged:** ☐ **Finding(s):** ☐ ☐ ☐ ☐

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Other imaging (most representative for this illness)

Imaging Modality	Site imaged	Findings	
1. None 2. Plain radiograph 3. CT 4. MRI 5. MRCP 6. ERCP 7. FDG-PET 8. PET CT 9. Isotope Bone scan 10. Ventilation/Perfusion scan 11. Echocardiogram 12. Ultrasound 13. Vascular Doppler 14. Labelled white cell scan	1. None 2. Whole body 3. Brain 4. Head/Neck 5. Whole spine 6. Cervical spine 7. Thoracic spine 8. Lumbar/sacral spine 9. Chest 10. Heart 11. Abdomen a. Liver b. Gallbladder c. Bowel d. Spleen e. Kidney f. Retro peritoneum 12. Pelvis 13. Arms / Hands 14. Legs /Feet	**General** 1. Not done 2. Normal 3. Mass /tumour 4. Metastasis 5. Abscess 6. Haematoma 7. Bony abnormality 8. Enlarged lymph nodes (localised) 9. Enlarged lymph nodes (widespread) 10. Soft tissue abnormality **Head and neck** 11. Para nasal sinus inflammation 12. Other Para nasal sinus abnormality 13. Pachymeningitis 14. Brain haemorrhage 15. Brain infarct 16. Brain patchy white matter ischaemia 17. Spinal cord abnormality 18. Orbital wall destruction **Abdomen** 19. Bowel infarction 20. Thickened bowel wall (target sign) 21. Renal infarction 22. Shrunken kidney 23. Swollen/large kidney 24. Hydronephrosis 25. Splenomegaly 26. Enlarged liver 27. Shrunken liver 28. Liver infarction 29. Splenic infarction 30. Gallstones 31. Bile duct obstruction 32. Pancreatitis	**Respiratory** 33. Pulmonary infiltrate 34. Airspace / Pulmonary consolidation 35. Pleural effusion 36. Pleural calcification 37. Pulmonary nodules 38. Pulmonary cavities 39. Pericardial effusion 40. Alveolar haemorrhage 41. Lung infarction 42. Pulmonary hypertension **Heart** 43. Myocardial infarction 44. Heart valve stenosis 45. Heart valve incompetence 46. Heart valve vegetations or mass 47. Cardiomegaly 48. Ventilation/Perfusion mismatch 49. Other soft tissue abnormality 50. Other **Musculoskeletal** 51. Joint space narrowing 52. Joint erosion 53. Skeletal muscle infarction 54. Muscle oedema 55. Other Muscle abnormality

BVAS v3 (Please tick all that apply for this illness, irrespective of aetiology)

General? ☐ Yes ☐ No

If yes:

- ☐ Myalgia
- ☐ Arthralgia / arthritis
- ☐ Fever ≥ 38℃
- ☐ Weight loss ≥ 2kg

Cutaneous? ☐ Yes ☐ No

If yes:

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

Mucous membranes / eyes? ☐ Yes ☐ No

If yes:

- ☐ Mouth ulcers
- ☐ Genital ulcers
- ☐ Adnexal inflammation
- ☐ Significant proptosis
- ☐ Scleritis / Episcleritis
- ☐ Conjunctivitis / Blepharitis / Keratitis
- ☐ Blurred vision
- ☐ Sudden visual loss
- ☐ Uveitis
- ☐ Retinal changes (vasculitis / thrombosis / exudate / haemorrhage)

ENT? ☐ Yes ☐ No

If yes:

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

Chest? ☐ Yes ☐ No

If yes:

- ☐ Wheeze
- ☐ Nodules or cavities
- ☐ Pleural effusion / pleurisy
- ☐ Infiltrate
- ☐ Endobronchial involvement
- ☐ Massive haemoptysis / alveolar haemorrhage
- ☐ Respiratory failure

Cardiovascular? ☐ Yes ☐ No

If yes:

- ☐ Loss of pulses
- ☐ Vascular heart disease
- ☐ Pericarditis
- ☐ Ischaemic cardiac pain
- ☐ Cardiomyopathy
- ☐ Congestive cardiac failure

Abdominal? ☐ Yes ☐ No

If yes:

- ☐ Peritonitis
- ☐ Bloody diarrhoea
- ☐ Ischaemic abdominal pain

Renal? ☐ Yes ☐ No

If yes:

- ☐ Hypertension
- ☐ Proteinuria > 1+
- ☐ Haematuria ≥ 10 RBCs/hpf
- ☐ Serum creatinine 125-249 µmol/L*
- ☐ Serum creatinine 250-499 µmol/L*
- ☐ Serum creatinine ≥500 µmol/L*
- ☐ Rise in serum creatinine >30% or fall in creatinine clearance >25%

* Can only be scored on the first assessment

Nervous system? ☐ Yes ☐ No

If yes:

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

Other? ☐ Yes ☐ No

If yes, specify:

PERSISTENT DISEASE ONLY*

☐ Tick if ALL abnormalities are due to persistent disease.

VDI (Please tick all that apply for this illness, irrespective of aetiology)

Musculoskeletal? ☐ Yes ☐ No

If yes:

- ☐ Significant muscle atrophy or weakness
- ☐ Deforming/erosive arthritis
- ☐ Osteoporosis/vertebral collapse
- ☐ Avascular necrosis
- ☐ Osteomyelitis

Skin/Mucous membranes? ☐ Yes ☐ No

If yes:

- ☐ Alopecia
- ☐ Cutaneous ulcers
- ☐ Mouth ulcers

Ocular? ☐ Yes ☐ No

If yes:

- ☐ Cataract
- ☐ Retinal change
- ☐ Optic atrophy
- ☐ Visual impairment/diplopia
- ☐ Blindness in one eye
- ☐ Blindness in second eye
- ☐ Orbital wall destruction

ENT? ☐ Yes ☐ No

If yes:

- ☐ Hearing loss
- ☐ Nasal blockage/chronic discharge/crusting
- ☐ Nasal bridge collapse/septal perforation
- ☐ Chronic sinusitis/radiological damage
- ☐ Subglottic stenosis (no surgery)
- ☐ Subglottic stenosis (with surgery)

Pulmonary? ☐ Yes ☐ No

If yes:

- ☐ Pulmonary hypertension
- ☐ Pulmonary fibrosis
- ☐ Pulmonary infarction
- ☐ Pleural fibrosis
- ☐ Chronic asthma
- ☐ Chronic breathlessness
- ☐ Impaired lung function

Cardiovascular? ☐ Yes ☐ No

If yes:

- ☐ Angina/angioplasty
- ☐ Myocardial infarction
- ☐ Subsequent myocardial infarction
- ☐ Cardiomyopathy
- ☐ Valvular disease
- ☐ Pericarditis ≥ 3 mths or pericardectomy
- ☐ Diastolic BP ≥ 95 or requiring antihypertensives

Peripheral vascular disease? ☐ Yes ☐ No

If yes:

- ☐ Absent pulses in one limb
- ☐ 2nd episode of absent pulses in one limb
- ☐ Major vessel stenosis
- ☐ Claudication >3 mths
- ☐ Minor tissue loss
- ☐ Major tissue loss
- ☐ Subsequent major tissue loss
- ☐ Complicated venous thrombosis

Gastrointestinal? ☐ Yes ☐ No

If yes:

- ☐ Gut infarction/resection
- ☐ Mesenteric insufficiency / pancreatitis
- ☐ Chronic peritonitis
- ☐ Oesophageal stricture/surgery

Renal? ☐ Yes ☐ No

If yes:

- ☐ Estimated/measured GFR < 50%
- ☐ Proteinuria > 0.5g/24hr
- ☐ End stage renal disease

Neuropsychiatric? ☐ Yes ☐ No

If yes:

- ☐ Cognitive impairment
- ☐ Major psychosis
- ☐ Seizures
- ☐ Cerebrovascular accident
- ☐ 2nd cerebrovascular accident
- ☐ Cranial nerve lesion
- ☐ Peripheral neuropathy
- ☐ Transverse myelitis

Other? ☐ Yes ☐ No

If yes:

- ☐ Gonadal failure ☐ Chemical cystitis
- ☐ Marrow failure ☐ Malignancy
- ☐ Diabetes ☐ Other

Total VDI Score*

☐ Record the number of positive items (1 point for each).

Working Diagnosis – Baseline

Do you think that the patient's current illness is due to:

Primary systemic vasculitis	☐	Please answer section A and B
Secondary vasculitis	☐	Please answer section C (next page)
Other illness	☐	Please answer section C (next page)

A. In your opinion, what is this patient's diagnosis?

☐ Takayasu arteritis	☐ Systemic granulomatous vasculitis (Wegener's granulomatosis)	☐ Behçet's disease
☐ Giant cell arteritis	☐ Microscopic polyangiitis	☐ Other primary vasculitis with no specific vessel size
☐ Isolated Aortitis	☐ Churg- Strauss syndrome	☐ Henoch-Schönlein purpura
☐ Other large vessel vasculitis	☐ Polyarteritis nodosa	☐ Cryoglobulinaemic vasculitis
☐ Single organ vasculitis: ☐☐☐☐☐☐☐☐☐☐	☐ Other small vessel vasculitis : ☐☐☐☐☐☐☐☐☐☐	

B. How confident are you about the diagnosis? (tick one box only)

☐ Very certain (75% or more)

☐ Moderately certain (50%-74%)

☐ Uncertain (25%-49%)

☐ Very uncertain (less than 25%)

C. If not primary systemic vasculitis, what is underlying diagnosis for this illness?

Dermatologic
- ☐ Livedoid vasculopathy
- ☐ Ischaemic skin lesion not due to vasculitis*
- ☐ Non-ischaemic skin lesion
- ☐ Other dermatologic*

Endocrine/Metabolic
- ☐ Calciphylaxis
- ☐ Scurvy
- ☐ Thyroid Disease
- ☐ Other endocrine/metabolic*

Gastrointestinal
- ☐ Mesenteric ischaemia
- ☐ Other gastrointestinal*

Genitourinary
- ☐ Nephrolithiasis
- ☐ Nephritic / nephrotic syndrome (non-vasculitic)*
- ☐ Other genitourinary*

Haematology
- ☐ Coagulopathy / Thrombosis
- ☐ Hypereosinophilic Syndrome
- ☐ Multiple myeloma /paraproteinemia
- ☐ Systemic amyloidosis
- ☐ Thrombotic thrombocytopenic purpura
- ☐ Other haematologic*

Infectious Disease
- ☐ Bacterial endocarditis / bacteraemia
- ☐ Bacterial or viral pneumonia
- ☐ Eosinophilic pneumonia
- ☐ Upper respiratory tract infection
- ☐ Urinary tract Infection
- ☐ Tuberculosis
- ☐ Other infection*

Malignancy
- ☐ Solid malignancy*
- ☐ Hematologic malignancy*

Neurologic
- ☐ Guillain-Barre syndrome
- ☐ Hearing loss not due to vasculitis*
- ☐ Migraine or other headache syndrome
- ☐ Multiple sclerosis
- ☐ Myasthenia gravis
- ☐ Neurodegenerative disorder
- ☐ Neuropathy not due to vasculitis*
- ☐ Stroke not due to vasculitis*
- ☐ Other neurologic*

Ophthalmologic
- ☐ Vision loss not due to vasculitis*
- ☐ Other ophthalmologic*

☐ **Other ***

Respiratory
- ☐ Asthma
- ☐ Airway stenosis not due to vasculitis*
- ☐ Cavitary lung lesion not due to vasculitis*
- ☐ Chronic obstructive pulmonary disease
- ☐ Other respiratory*

Rheumatologic
- ☐ Antiphospholipid antibody syndrome
- ☐ Polymyositis/Dermatomyostis
- ☐ Sarcoidosis
- ☐ Systemic lupus erythematosis
- ☐ Sjögren's syndrome
- ☐ Other Connective Tissue *
- ☐ Rheumatoid Arthritis
- ☐ Other inflammatory arthritis*
- ☐ Periodic fever syndrome
- ☐ Other rheumatologic*

Vascular
- ☐ Atherosclerosis
- ☐ Fibromuscular Dysplasia
- ☐ Thromboangiigitis obliterans (Buerger's disease)
- ☐ Other vascular*

☐ **Toxic**
- ☐ Cocaine-induced disease
- ☐ Other drug-induced disease*

*Please provide as precise a diagnosis as you can:

DCVAS STUDY 6 MONTH UPDATE

Today's date: D D / M M M / Y Y Y Y

Is the patient still alive?

Yes	☐	Please answer section 2 and 3
No	☐	Please answer section 1, 2 and 3
Don't know	☐	What was the date of last follow up? D D / M M M / Y Y Y Y Please answer section 2 and 3

Section 1: Date and cause of death

Date of death: D D / M M M / Y Y Y Y

Cause of Death:

- ☐ Pulmonary Haemorrhage
- ☐ Renal Failure
- ☐ Myocardial infarction
- ☐ Stroke
- ☐ Malignancy (please specify):
- ☐ Infection (please specify):
- ☐ Unknown
- ☐ Other (please specify):

Section 2: Diagnosis at 6 months

Do you think that the patient's current illness is due to:

Primary systemic vasculitis	☐ Please answer section A and B
Secondary vasculitis	☐ Please answer section C (next page)
Other illness	☐ Please answer section C (next page)

A. In your opinion, what is this patient's diagnosis?

☐ Takayasu arteritis	☐ Systemic granulomatous vasculitis (Wegener's granulomatosis)	☐ Behçet's disease
☐ Giant cell arteritis	☐ Microscopic polyangiitis	☐ Other primary vasculitis with no specific vessel size
☐ Isolated Aortitis	☐ Churg- Strauss syndrome	☐ Henoch-Schönlein purpura
☐ Other large vessel vasculitis	☐ Polyarteritis nodosa	☐ Cryoglobulinaemic vasculitis
☐ Single organ vasculitis: ☐☐☐☐☐☐☐☐☐☐	☐ Other small vessel vasculitis : ☐☐☐☐☐☐☐☐☐☐	

B. How confident are you about the diagnosis? (tick one box only)

☐ Very certain (75% or more)
☐ Moderately certain (50%-74%)
☐ Uncertain (25%-49%)
☐ Very uncertain (less than 25%)

If the diagnosis has changed from baseline, please answer section 3

C. If not primary systemic vasculitis, what is underlying diagnosis for this illness?

Dermatologic

- ☐ Livedoid vasculopathy
- ☐ Ischaemic skin lesion not due to vasculitis*
- ☐ Non-ischaemic skin lesion
- ☐ Other dermatologic*

Endocrine/Metabolic

- ☐ Calciphylaxis
- ☐ Scurvy
- ☐ Thyroid Disease
- ☐ Other endocrine/metabolic*

Gastrointestinal

- ☐ Mesenteric ischaemia
- ☐ Other gastrointestinal*

Genitourinary

- ☐ Nephrolithiasis
- ☐ Nephritic / nephrotic syndrome (non-vasculitic)*
- ☐ Other genitourinary*

Haematology

- ☐ Coagulopathy / Thrombosis
- ☐ Hypereosinophilic Syndrome
- ☐ Multiple myeloma /paraproteinemia
- ☐ Systemic amyloidosis
- ☐ Thrombotic thrombocytopenic purpura
- ☐ Other haematologic*

Infectious Disease

- ☐ Bacterial endocarditis / bacteraemia
- ☐ Bacterial or viral pneumonia
- ☐ Eosinophilic pneumonia
- ☐ Upper respiratory tract infection
- ☐ Urinary tract Infection
- ☐ Tuberculosis
- ☐ Other infection*

Malignancy

- ☐ Solid malignancy*
- ☐ Hematologic malignancy*

Neurologic

- ☐ Guillain-Barre syndrome
- ☐ Hearing loss not due to vasculitis*
- ☐ Migraine or other headache syndrome
- ☐ Multiple sclerosis
- ☐ Myasthenia gravis
- ☐ Neurodegenerative disorder
- ☐ Neuropathy not due to vasculitis*
- ☐ Stroke not due to vasculitis*
- ☐ Other neurologic*

Ophthalmologic

- ☐ Vision loss not due to vasculitis*
- ☐ Other ophthalmologic*

☐ **Other ***

Respiratory

- ☐ Asthma
- ☐ Airway stenosis not due to vasculitis*
- ☐ Cavitary lung lesion not due to vasculitis*
- ☐ Chronic obstructive pulmonary disease
- ☐ Other respiratory*

Rheumatologic

- ☐ Antiphospholipid antibody syndrome
- ☐ Polymyositis/Dermatomyostis
- ☐ Sarcoidosis
- ☐ Systemic lupus erythematosis
- ☐ Sjögren's syndrome
- ☐ Other Connective Tissue *
- ☐ Rheumatoid Arthritis
- ☐ Other inflammatory arthritis*
- ☐ Periodic fever syndrome
- ☐ Other rheumatologic*

Vascular

- ☐ Atherosclerosis
- ☐ Fibromuscular Dysplasia
- ☐ Thromboangiigitis obliterans (Buerger's disease)
- ☐ Other vascular*

Toxic

- ☐ Cocaine-induced disease
- ☐ Other drug-induced disease*

*Please provide as precise a diagnosis as you can:

Section 3: Reasons for change in diagnosis

(Only answer this section if diagnosis has changed from baseline)

What factors influenced the change in diagnosis? (please tick all that apply)

- ☐ Evolution of patients symptoms or signs
- ☐ Radiology result(s)
- ☐ Biopsy result(s)
- ☐ Laboratory test result(s)
- ☐ Post mortem examination
- ☐ Other (please specify): _____