ABROGATE BVAS/WG Non-Severe Disease Module
Investigator Training Packet

Every investigator involved in ABROGATE who will be completing the BVAS/WG must complete the BVAS/WG non-severe disease module training before he or she is eligible to conduct an assessment of a study subject. At least one investigator at each site must have completed the BVAS/WG non-severe disease module before the site can be opened for recruitment.

Each investigator must do the following:

1. Read the instructions
2. Review the glossary
3. Study the training case and answer explanation
4. Take the 10 question test
5. Self-score the test
6. Return the test case score sheet and training certification form

This packet includes the following documents:

ABROGATE BVAS/WG Training: Introduction and Glossary

ABROGATE BVAS/WG Training: Training Case and Answers

ABROGATE BVAS/WG Training: Test Cases Score Sheet

ABROGATE BVAS/WG Training: Test Cases

ABROGATE BVAS/WG Training: Test Cases Answers

ABROGATE BVAS/WG Training: Investigator Training Certification

Please scan/email or fax the test cases score sheet and investigator training certification to the ABROGATE Data coordinator:

Cristina Burroughs
Fax: +1 813 910-1225
Email: Cristina.Burroughs@epi.usf.edu

Birmingham Vasculitis Activity Score Modified for Wegener's Granulomatosis (BVAS/WG)

An Introduction and Glossary of Terms

Purpose of assessment

BVAS/WG is designed to document clinical features that are directly due to active ANCA-associated vasculitis (AAV): granulomatosis with polyangiitis (Wegener's, GPA) or microscopic polyangiitis (MPA). In addition, the instrument separates the features that represent new or worse disease activity from those that represent persistent activity. In scoring BVAS/WG, it is very important not to confuse *activity* with *damage*. Damage, is defined as the presence of irreversbile scarring/dysfunction, and is a concept distinct from current disease activity. Damage will be scored separately in ABROGATE using another index, the Combined Damage Assessment (CDA), which is not the subject of this exercise.

Recording disease activity

The list of items in BVAS/WG includes clinical symptoms and signs, as well as information obtained from additional tests (e.g., chest x-rays) or subspecialty consultations. When using the BVAS/WG evaluation form, one scores only these items attributable to currently active GPA (after the exclusion of obvious causes such as infection, hypertension, and treatment toxicity). BVAS scores may vary rapidly, and reflect the need for therapy.

Patients' assessment at Month 0

If a patient is being evaluated at the time of diagnosis, all of the abnormalities noted should be recorded as NEW/WORSE (○) regardless of their duration. **However, patients are being recruited to ABROGATE at the time of relapse. Therefore, only disease activity within the previous 28 days should be scored at Month 0.** In order to be eligible for the study subjects must score at least 3 NEW/WORSE minor items or 1 NEW/WORSE major item on BVAS/WG. Only features that represent active disease should be recorded on the BVAS/WG form. As patients are recruited to ABROGATE at the time of relapse, they may have accumulated damage either due to AAV or prior treatment. These elements will be captured on the Combined Damage Assessment (CDA) form. After going through the entire items list, also consider adding any other significant items to the "Other" section, if relevant. A partial list of "Other" items that might be included in these sections is displayed at the end of the glossary. "Other" items are classified as either major or minor by the investigator and do count towards total score and to determine eligibility. If a section has no items present, check the "none" box.

Patients' assessment at subsequent follow up visits

If the patient is being evaluated in follow-up, there may be some abnormalities that are NEW or WORSE (○**)** within the previous 28 days. Other abnormalities may have been present on the previous assessment and are neither new nor worse, but rather still present (PERSISTENT □). By making this distinction, one differentiates new, acute disease activity from persistent disease activity. It is important to remember that *persistent* activity is *activity*, not damage. Thus, persistent purpura should be scored as activity. In contrast, weakness from mononeuritis multiplex of 4 months duration is damage, and should not be scored in BVAS. It can be difficult to be certain whether a symptom or sign is due to persistent activity or to damage; in evaluating such cases one relies on clinical judgment to make this distinction.

Checking the boxes
Check **one** of the boxes for each item (○ or □) only if the abnormality is ascribed to the presence of active vasculitis. If no abnormalities ascribable to vasculitis are present in a given organ system, check the "none" box; this confirms you did not overlook an organ system on the scoring sheet. Sometimes you will have patients in whom abnormalities are present that are not due to AAV (e.g., hematuria due to urinary infection or cyclophosphamide toxicity). In these cases, you should NOT record them in the BVAS/WG list, even though they are present, because they cannot be ascribed to active AAV. In some patients, abnormalities that were due to previous episodes of vasculitis may still be evident, even though the disease is entirely inactive (e.g., stroke). These features should also NOT be recorded on BVAS/WG, since they represent non-healing scars (damage).

○ Check this box only if the abnormality is NEW/WORSE within the **previous 28 days** (unless this is the first presentation of untreated disease).
□ Check this box only if the abnormality is PERSISTENT since the last assessment and not worse within the **previous 28 days.**
□ Check this space if there is not a single major or minor item that is new/worse within a particular organ system.

Necessity for "Judgment Calls"
As in clinical practice, one must sometimes make "judgment calls" in scoring BVAS/WG. For example, persistent sinus symptoms are often notoriously difficult to classify with certainty as either active disease or permanent damage. Similarly, small amounts of hematuria (usually with RBC casts) may persist for months in patients whose disease is otherwise quiescent. In both such cases, the physician is unlikely to intensify treatment in the absence of other indications of active disease. For this reason, these findings (and analogous findings in other organ systems) should not be scored in BVAS/WG. If subsequent events or test results cause you to re-consider your judgment call, you may go back and change your initial decision regarding a particular finding.

Recording Major and Minor Items
Individual items are defined as Major by the presence of an asterisk (*). All other items are defined as Minor. If you list additional items in the "Other" section, you should indicate whether the item is "Major" or "Minor". In general, a Major item is one whose presence would have traditionally prompted the use of cyclophosphamide. Minor items are those more likely to be treated with methotrexate or an increase in prednisone. It is possible to upgrade a minor item to a major item by marking it with an asterisk, if you feel that it is severe enough to merit this.

If you decide that a particular abnormality is due to the presence of active AAV, you must distinguish problems that are new/worse from those problems that are persistent. For each item where there is an abnormality, you need to check either the NEW/WORSE box or the PERSISTENT box, but not both.

Summing Up BVAS/WG
Now add up all of the Major (*) items marked in the New/Worse column, and enter the sum in the appropriate box. Repeat this for the Minor items in the NEW/WORSE column, and then do the same for the Major and Minor items in the Persistent column.

Defining disease status

Severe disease/flare: If any Major item is recorded, the patient has a "Severe Flare".

Limited disease/flare: If only Minor items are recorded, the patient has a "Limited Flare".

Persistent Disease: Persistent disease indicates the presence of 1 or more persistent items attributed to active disease.

Remission: Remission indicates no active disease (i.e., no new/worse and no persistent items present). For the purpose of the ABROGATE trial, the definition of remission is BVAS/WG ≤ 1 (zero or one minor persistent BVAS/WG item).

Physician's Global Assessment

Finally, use the 10 point Likert scale to record your assessment of the overall disease activity in this case. Remember that you should not be influenced by the presence of any accumulated damage, complication of treatment, social/emotional problems, or other issues not related to active vasculitis.

BVAS/WG

GLOSSARY OF TERMS

GENERAL RULE: Disease features are scored only when they are attributable to <u>active</u> GPA/MPA, after exclusion of other obvious causes (e.g., infection, hypertension, toxicity of treatment, etc.). <u>THIS IS THE MOST IMPORTANT ASPECT OF SCORING TO REMEMBER!</u>

If an item is new or represents a deterioration of status occurring in the previous 28 days, it is scored in the NEW/WORSE box.

If the feature was present at the previous evaluation and is not new or worse but still represents ongoing disease activity, record it as PERSISTENT.

Check box (○ or □) only if the abnormality is ascribable to the presence of active WG.

○ Check this circle only if the abnormality is NEW/WORSE within the **previous 28 days.**
□ Check this box only if the abnormality is PERSISTENT since the last assessment and not worse within the **previous 28 days.**

For some features, further information (e.g., a chest radiograph or subspecialty consult) may be required to determine if an abnormality is new or worse.

Glossary definitions used in BVAS/WG

For most patients, you will be able to complete the BVAS evaluation form on the same day you evaluate the patient. However, on other occasions, you may require further information before entering some items. For example, if the patient has new onset of stridor, you would usually ask a colleague in ENT to investigate this further to determine whether or not it is due to active vasculitis. It is suggested that you leave such items blank temporarily, but complete them once the information is available.

1. General	
Arthralgia:	Joint pain without obvious swelling.
Arthritis:	Joint inflammation.
Fever:	Documented temperature elevation. The value refers to oral temperatures (38.0ºC).

2. Cutaneous	
Purpura:	Petechiae (small red spots), palpable purpura, or ecchymoses (large plaques) in skin or oozing (in the absence of trauma) in the mucous membranes.
Ulcer:	Open sore in a skin surface.
***Gangrene:**	Extensive tissue necrosis (e.g., digit). Gangrene refers not to superficial infarction (e.g., a nailbed infarct), but rather to severe ischemia affecting the viability of a substantial portion of tissue, such as an entire fingertip.
** If new/worse, this denotes a major item for assessment of flares.*	

3. Mucous Membranes and Eyes	
Mouth ulcers:	Ulcers localized in the mouth. Exclude other causes, such as drugs, Crohn's disease, pemphigus, etc.
Conjunctivitis:	Inflammation of the conjunctivae (exclude infectious causes).
Episcleritis:	Inflammation of the superficial sclera.
Retro-orbital mass/ Proptosis:	Protrusion of the eye caused by an inflammatory mass behind the globe. This may be associated with diplopia due to infiltration of extra-ocular muscles.
Uveitis:	Inflammation of the uveal tract (iris, ciliary body, choroid) confirmed by ophthalmologist.
***Scleritis**	Inflammation of the deep sclera (specialist opinion usually required).
***Retinal exudates:**	Any area of soft retinal exudates (exclude hard exudates) seen on ophthalmoscopic examination.
***Retinal hemorrhages:**	Any area of retinal hemorrhage seen on ophthalmoscopic examination.
** If new/worse, this denotes a major item for assessment of flares.*	

4. ENT	
Bloody nasal discharge:	Blood stained secretions from the nose, irrespective of severity or frequency, occurring since the last visit.
Nasal crusting:	Discharge of large serous or serosanguinous crusts.
Nasal ulceration:	Nasal mucosal lesions (not due to trauma).
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 the section for "Other" items).
Swollen salivary glands	Tender swelling of one or more major salivary glands not due to an infection, stone, or other non-AAV cause.
Subglottic inflammation:	Inspiratory stridor with significant narrowing of subglottic space confirmed by further examination (usually by an ENT specialist).
Conductive deafness:	Any hearing loss due to middle ear involvement, preferably confirmed by audiometry.
***Sensorineural deafness:**	Deafness caused by damage to the auditory nerve or cochlea.
** If new/worse, this denotes a major item for assessment of flares.*	

5. Cardiovascular	
Pericarditis:	Pericardial pain and/or friction rub on clinical assessment.

6. Abdominal	
***Mesenteric ischemia:**	Defined as severe abdominal pain, bloody diarrhea, gut perforation/ infarction due to AAV.
** If new/worse, this denotes a major item for assessment of flares.*	

7. Chest/Pulmonary	
Pleurisy:	Pleural pain and/or friction rub on clinical assessment or new onset of radiologically confirmed pleural effusion. Other causes (e.g., infection, cancer) should be excluded.
Nodules or cavities:	New lesions, detected by CXR.
***Tracheobronchial involvement:**	Pseudotumour or ulceration of tracheobronchial tree. Requires bronchoscopy to exclude tumor or infection.
***Alveolar hemorrhage:**	Major pulmonary bleeding, with shifting pulmonary infiltrates. Other causes of bleeding should be excluded.
***Respiratory failure:**	Dyspnea requiring artificial ventilation.

** If new/worse, this denotes a major item for assessment of flares.*	
8. Renal	
Hematuria: (no RBC casts)	≥1+ on urinalysis; ≥10 rbc/hpf. Infection should be excluded. The hematuria must be considered due to active renal vasculitis, not just prior damage.
***RBC casts and/or Glomerulonephritis on biopsy**	The appearance of RBC casts in the urinary sediment and/or evidence of active glomerulonephritis on biopsy. RBC casts are essentially the "surrogate" for glomerulonephritis.
***Rise in creatinine > 30% or creatinine clearance fall > 25%:**	Deterioration in renal function that is attributable to active AAV and meets these criteria.
** If new/worse, this denotes a major item for assessment of flares.*	
9. Nervous System	
***Meningitis:**	Severe headache +/- neck stiffness, ascribed to inflammatory meningitis after the exclusion of infection, bleeding, and other causes.
***Stroke:**	Cerebrovascular accident resulting in focal neurological signs such as paresis, weakness, etc.
***Cord lesion:**	Transverse myelitis with extremity weakness or sensory loss.
***Cranial nerve palsy:**	Isolated acute cranial nerve palsy (excluding sensorineural hearing loss, which is listed in ENT).
***Sensory Peripheral neuropathy:**	Neuropathy resulting in glove and/or stocking distribution of sensory loss. Other causes should be excluded (e.g., idiopathic, metabolic, vitamin deficiencies, infectious, toxic, hereditary).
***Motor mononeuritis multiplex:**	Neuritis of named peripheral nerve, only scored if motor involvement. On EMG/NCV evaluation, multiple nerve dysfunction may be documented, but clinical involvement of only one named nerve is required to score this item. Other causes should be excluded (diabetes, sarcoidosis, carcinoma, amyloidosis).
** If new/worse, this denotes a major item for assessment of flares.*	

10. Other:	Significant features attributable to active AAV not listed above. Please provide full details and designate item as Major or Minor items. Potential "Other" items are listed below.
If defined as new/worse, this may denote a major or minor item for assessment of flares.	

Examples of Potential "Other" items:

- Weight loss (>2 kg over 28 day period)
- Genitourinary involvement
- Granulomatous lesion not otherwise mentioned in list (e.g. breast mass)
- Cardiac valvular lesions
- Cutaneous infarctions (splinter hemorrhages, digital infarcts)
- Pulmonary infiltrates (not due to alveolar hemorrhage, cavity)
- Cardiomyopathy
- Pancreatitis
- Mastoiditis
- Many others….

BVAS/WG – Non-severe disease module – Training case

Training Case 1.

A 50 year old man with granulomatosis with polyangiitis (Wegener's)(GPA) was diagnosed 2 years ago with manifestations that included bloody nasal discharge, pulmonary nodules, nodular skin lesions on his elbows, and glomerulonephritis with red blood cell casts. His creatinine values were consistently normal. For this, he was treated with prednisone and rituximab. He achieved remission and his chest CT at the time of remission revealed one residual pulmonary nodular density 1 cm x 1 cm in the right middle lobe.

He now presents with symptoms of new migratory arthralgias, increased nasal crusting without sinus pressure, new nodular lesions on his elbows similar to what he had at the time of his original diagnosis, and cough. On physical examination his nasal mucosa appears inflamed with ulceration, the remainder of the examination is unremarkable. His urinalysis is negative for blood or protein. Chest CT scan shows 2 new pulmonary lesions – a 2 cm x 3 cm right lower lobe cavitary nodule and a 2 cm x 2 cm left upper lobe pulmonary nodule, with the prior 1 cm x 1 cm right middle lobe nodule being unchanged.

Training CASE 1A: What is his current BVAS/WG ?

He is placed on treatment for active GPA with methotrexate and prednisone. At his month 3 visit the arthralgias, nasal crusting, cough, and elbow lesions have resolved. On physical examination his nasal mucosa shows no mucosal inflammation or ulceration. Laboratory testing shows that his urinalysis remains negative for blood and protein. Chest CT scan shows no new pulmonary lesions – the prior 2 cm x 3 cm right lower lobe cavitary nodule is now a 1.2 cm x 1.0 cm nodule, the prior 2 cm x 2 cm left upper lobe pulmonary nodule is now a flattened linear density 1 cm x 3 mm, and the original 1 cm x 1 cm right middle lobe nodule remains unchanged.

Training CASE 1B: What is his current BVAS/WG ?

BVAS/WG – Non-severe disease module – Training case

TRAINING CASE: ANSWERS AND EXPLANATIONS

Training Case 1A

The following manifestations of AAV were all present within 28 days of evaluation:

Arthralgias

Nasal crusting

Nodular elbow skin lesion Because skin nodule is not a set item on the BVAS/WG form it is necessary to write it in the "Other" section. Skin nodule is not considered a major item so it was not starred.

Pulmonary nodules or cavities

Total of 4 new minor items and a limited disease flare

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Training Case 1B

The patient has had resolution of the arthralgias, nasal crusting, and skin nodules. His chest CT shows persistence of the original nodule which had been left as damage from his presentation, with the 2 new nodules seen at the time of relapse reduced to a 1x1 cm small pulmonary nodule and a linear density. These findings can persist as a result of damage and there are no new changes. There is no evidence of active vasculitis within the prior 28 days and his BVAS/WG would be 0 (Remission).

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Participant ID Training Case 1A

Mark only if abnormality is ascribable to the presence of active Wegener's Granulomatosis. Mark "Persistent" or "New/Worse" depending upon if the abnormality is persistent disease activity since the last assessment and not worse within the previous 28 days or if the abnormality is newly present or worse within the previous 28 days correspondingly. If no items are present in any section, tick "none". Major items are in bold and marked with *. All WG-related clinical features need to be documented on this form if they are related to active diseases. Use "OTHER" category as needed.

	Persistent	New/Worse	None
1. GENERAL			☐
a. arthralgia/arthritis	○	⊗	
b. fever (≥ 38 degrees C)	○	○	
2. CUTANEOUS			☒
a. purpura	○	○	
b. skin ulcer	○	○	
c. ***gangrene**	○	○	
3. MUCOUS MEMBRANES/EYES			☒
a. mouth ulcers	○	○	
b. conjunctivitis/episcleritis	○	○	
c. retro-orbital mass/proptosis	○	○	
d. uveitis	○	○	
e. ***scleritis**	○	○	
f. ***retinal exudates/haemorrhage**	○	○	
4. EAR, NOSE & THROAT			☐
a. bloody nasal discharge / nasal crusting / ulcer	○	⊗	
b. sinus involvement	○	○	
c. swollen salivary gland	○	○	
d. subglottic inflammation	○	○	
e. conductive deafness	○	○	
f. ***sensorineural deafness**	○	○	
5. CARDIOVASCULAR			☒
a. pericarditis	○	○	
6. GASTROINTESTINAL			☒
a. ***mesenteric ischemia**	○	○	
7. PULMONARY			☐
a. pleurisy	○	○	
b. nodules or cavities	○	⊗	
c. other infiltrate secondary to WG	○	○	
d. endobronchial involvement	○	○	
e. ***alveolar hemorrhage**	○	○	
f. ***respiratory failure**	○	○	

DETERMINING DISEASE STATUS:
Severe Disease / Flare: ≥ 1 new/worse Major item
Limited Disease / Flare: ≥ new/worse Minor item
Persistent Disease: Continued (but not new/worse) activity
Remission: No active disease, including either new /worse or persistent items

	Persistent	New/Worse	None
8. RENAL			☒
a. hematuria (no RBC casts) (≥ 1 + or ≥ 10 RBC/hpf)	○	○	
b. ***RBC casts**	○	○	
c. ***rise in creatinine > 30% or fall in creatinine clearance > 25%**	○	○	

Note: If both hematuria and RBC casts are present, score only the RBC casts (the major item).

9. NERVOUS SYSTEM			☒
a. ***meningitis**	○	○	
b. ***cord lesion**	○	○	
c. ***stroke**	○	○	
d. ***cranial nerve palsy**	○	○	
e. ***sensory peripheral neuropathy**	○	○	
f. ***motor mononeuritis multiplex**	○	○	

10. OTHER (describe all items and * items deemed major) ☐

Major			
☐	skin nodule	○	⊗
☐	_____	○	○
☐	_____	○	○
☐	_____	○	○

11. TOTAL NUMBER OF ITEMS: ☐

a.	b.	c.	d.
0	04	0	0
Major New / Worse	Minor New / Worse	Major Persistent	Minor Persistent

12. CURRENT DISEASE STATUS (check only one)

Severe Disease/Flare	☐
Limited Disease/Flare	⊗
Persistent Disease	☐
Remission	☐

13. PHYSICIAN'S GLOBAL ASSESSMENT (PGA)
Mark to indicate the amount of WG disease activity (not including longstanding damage) within the previous 28 days:

0	1	2	3	4	5	6	7	8	9	10
○	○	○	⊗	○	○	○	○	○	○	○

Remission **Maximum activity**

Participant ID Training Case 1B

Mark only if abnormality is ascribable to the presence of active Wegener's Granulomatosis. Mark "Persistent" or "New/Worse" depending upon if the abnormality is persistent disease activity since the last assessment and not worse within the previous 28 days or if the abnormality is newly present or worse within the previous 28 days correspondingly. If no items are present in any section, tick "none". Major items are in bold and marked with *. All WG-related clinical features need to be documented on this form if they are related to active diseases. Use "OTHER" category as needed.

	Persistent	New/Worse	None
1. GENERAL			☒
a. arthralgia/arthritis	○	○	
b. fever (≥ 38 degrees C)	○	○	
2. CUTANEOUS			☒
a. purpura	○	○	
b. skin ulcer	○	○	
c. ***gangrene**	○	○	
3. MUCOUS MEMBRANES/EYES			☒
a. mouth ulcers	○	○	
b. conjunctivitis/episcleritis	○	○	
c. retro-orbital mass/proptosis	○	○	
d. uveitis	○	○	
e. ***scleritis**	○	○	
f. ***retinal exudates/haemorrhage**	○	○	
4. EAR, NOSE & THROAT			☒
a. bloody nasal discharge / nasal crusting / ulcer	○	○	
b. sinus involvement	○	○	
c. swollen salivary gland	○	○	
d. subglottic inflammation	○	○	
e. conductive deafness	○	○	
f. ***sensorineural deafness**	○	○	
5. CARDIOVASCULAR			☒
a. pericarditis	○	○	
6. GASTROINTESTINAL			☒
a. ***mesenteric ischemia**	○	○	
7. PULMONARY			☒
a. pleurisy	○	○	
b. nodules or cavities	○	○	
c. other infiltrate secondary to WG	○	○	
d. endobronchial involvement	○	○	
e. ***alveolar hemorrhage**	○	○	
f. ***respiratory failure**	○	○	

DETERMINING DISEASE STATUS:
Severe Disease / Flare: ≥ 1 new/worse Major item
Limited Disease / Flare: ≥ new/worse Minor item
Persistent Disease: Continued (but not new/worse) activity
Remission: No active disease, including either new /worse or persistent items

	Persistent	New/Worse	None
8. RENAL			☒
a. hematuria (no RBC casts) (≥ 1 + or ≥ 10 RBC/hpf)	○	○	
b. ***RBC casts**	○	○	
c. ***rise in creatinine > 30% or fall in creatinine clearance > 25%**	○	○	
Note: If both hematuria and RBC casts are present, score only the RBC casts (the major item).			
9. NERVOUS SYSTEM			☒
a. ***meningitis**	○	○	
b. ***cord lesion**	○	○	
c. ***stroke**	○	○	
d. ***cranial nerve palsy**	○	○	
e. ***sensory peripheral neuropathy**	○	○	
f. ***motor mononeuritis multiplex**	○	○	
10. OTHER (describe all items and * items deemed major)			☒
Major			
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	

11. TOTAL NUMBER OF ITEMS: ☐

a.	b.	c.	d.
0	0	0	0
Major New / Worse	Minor New / Worse	Major Persistent	Minor Persistent

12. CURRENT DISEASE STATUS (check only one)

Severe Disease/Flare	☐
Limited Disease/Flare	☐
Persistent Disease	☐
Remission	☒

13. PHYSICIAN'S GLOBAL ASSESSMENT (PGA)
Mark to indicate the amount of WG disease activity (not including longstanding damage) within the previous 28 days:

0	1	2	3	4	5	6	7	8	9	10
☒	○	○	○	○	○	○	○	○	○	○

Remission **Maximum activity**

VCRC-EUVAS ABROGATE BVAS/WG
Non-severe disease module
Test Cases

Do <u>not</u> score these cases until after you have read the instructions and reviewed the training cases

Please score all 10 questions before you review the answers

Case 1

A 43 year old man with granulomatosis with polyangiitis (Wegener's)(GPA) was diagnosed 2 years ago with manifestations that included bloody nasal discharge, sinusitis, pulmonary nodules, and glomerulonephritis with red blood cell casts. His creatinine values were consistently normal. For this, he was treated with prednisone and rituximab. He was left with residual fatigue and nasal crusting.

He now presents with symptoms of worsened fatigue, new migratory arthralgias, increased nasal crusting, new epistaxis, and sinus pressure. On physical examination his nasal mucosa appears inflamed with ulceration, the remainder of the examination is unremarkable. His ESR is 54 mm/hr (normal: 0-20), CRP 5.1 mg/ L, (normal: 0-4.9), urinalysis is negative for blood or protein. Sinus CT scan reveals increased mucosal thickening in the maxillary and sphenoid sinuses, chest CT scan shows scarring in the right lung base that is unchanged compared to his prior study.

CASE 1A: What is his current BVAS/WG ?

He is placed on treatment for active GPA with methotrexate and prednisone. At his month 3 visit the arthralgias have resolved. His fatigue is better and is back to the pre-relapse baseline but it is not resolved and continues to impact his daily activities. His epistaxis and sinus pressure has resolved but he has residual crusting that he feels is at his baseline. On physical examination his nasal mucosa appears dry with no mucosal inflammation or ulceration. Laboratory testing shows that his ESR is now normal and the urinalysis remains negative for blood and protein. Sinus CT scan shows mucosal thickening in the maxillary and sphenoid sinus with minimal improvement but no bony erosion and no new mucosal thickening.

CASE 1B: What is his current BVAS/WG ?

VCRC-EUVAS ABROGATE BVAS/WG
Training and Practice Cases
BVAS/WG

CASE 1A

Mark only if abnormality is ascribable to the presence of active Wegener's Granulomatosis. Mark "Persistent" or "New/Worse" depending upon if the abnormality is persistent disease activity since the last assessment and not worse within the previous 28 days or if the abnormality is newly present or worse within the previous 28 days correspondingly. If no items are present in any section, tick "none". Major items are in bold and marked with *. All WG-related clinical features need to be documented on this form if they are related to active diseases. Use "OTHER" category as needed.

	Persistent	New/Worse	None
1. GENERAL			☐
a. arthralgia/arthritis	○	○	
b. fever (≥ 38 degrees C)	○	○	
2. CUTANEOUS			☐
a. purpura	○	○	
b. skin ulcer	○	○	
c. ***gangrene**	○	○	
3. MUCOUS MEMBRANES/EYES			☐
a. mouth ulcers	○	○	
b. conjunctivitis/episcleritis	○	○	
c. retro-orbital mass/proptosis	○	○	
d. uveitis	○	○	
e. ***scleritis**	○	○	
f. ***retinal exudates/haemorrhage**	○	○	
4. EAR, NOSE & THROAT			☐
a. bloody nasal discharge / nasal crusting / ulcer	○	○	
b. sinus involvement	○	○	
c. swollen salivary gland	○	○	
d. subglottic inflammation	○	○	
e. conductive deafness	○	○	
f. ***sensorineural deafness**	○	○	
5. CARDIOVASCULAR			☐
a. pericarditis	○	○	
6. GASTROINTESTINAL			☐
a. ***mesenteric ischemia**	○	○	
7. PULMONARY			☐
a. pleurisy	○	○	
b. nodules or cavities	○	○	
c. other infiltrate secondary to WG	○	○	
d. endobronchial involvement	○	○	
e. ***alveolar hemorrhage**	○	○	
f. ***respiratory failure**	○	○	

	Persistent	New/Worse	None
8. RENAL			☐
a. hematuria (no RBC casts) (≥ 1 + or ≥ 10 RBC/hpf)	○	○	
b. ***RBC casts**	○	○	
c. ***rise in creatinine > 30% or fall in creatinine clearance > 25%**	○	○	
Note: If both hematuria and RBC casts are present, score only the RBC casts (the major item).			
9. NERVOUS SYSTEM			☐
a. ***meningitis**	○	○	
b. ***cord lesion**	○	○	
c. ***stroke**	○	○	
d. ***cranial nerve palsy**	○	○	
e. ***sensory peripheral neuropathy**	○	○	
f. ***motor mononeuritis multiplex**	○	○	
10. OTHER (describe all items and * items deemed major)			☐
Major			
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	

11. TOTAL NUMBER OF ITEMS: ☐

a.	b.	c.	d.
___ ___	___ ___	___ ___	___ ___
Major New / Worse	Minor New / Worse	Major Persistent	Minor Persistent

DETERMINING DISEASE STATUS:
Severe Disease / Flare: ≥ 1 new/worse Major item
Limited Disease / Flare: ≥ new/worse Minor item
Persistent Disease: Continued (but not new/worse) activity
Remission: No active disease, including either new /worse or persistent items

12. CURRENT DISEASE STATUS (check only one)

- Severe Disease/Flare ☐
- Limited Disease/Flare ☐
- Persistent Disease ☐
- Remission ☐

	0	1	2	3	4	5	6	7	8	9	10	
Remission	○	○	○	○	○	○	○	○	○	○	○	**Maximum activity**

VCRC-EUVAS ABROGATE BVAS/WG
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CASE 1B

Mark only if abnormality is ascribable to the presence of active Wegener's Granulomatosis. Mark "Persistent" or "New/Worse" depending upon if the abnormality is persistent disease activity since the last assessment and not worse within the previous 28 days or if the abnormality is newly present or worse within the previous 28 days correspondingly. If no items are present in any section, tick "none". Major items are in bold and marked with *. All WG-related clinical features need to be documented on this form if they are related to active diseases. Use "OTHER" category as needed.

	Persistent	New/Worse	None
1. GENERAL			☐
a. arthralgia/arthritis	○	○	
b. fever (≥ 38 degrees C)	○	○	
2. CUTANEOUS			☐
a. purpura	○	○	
b. skin ulcer	○	○	
c. ***gangrene**	○	○	
3. MUCOUS MEMBRANES/EYES			☐
a. mouth ulcers	○	○	
b. conjunctivitis/episcleritis	○	○	
c. retro-orbital mass/proptosis	○	○	
d. uveitis	○	○	
e. ***scleritis**	○	○	
f. ***retinal exudates/haemorrhage**	○	○	
4. EAR, NOSE & THROAT			☐
a. bloody nasal discharge / nasal crusting / ulcer	○	○	
b. sinus involvement	○	○	
c. swollen salivary gland	○	○	
d. subglottic inflammation	○	○	
e. conductive deafness	○	○	
f. ***sensorineural deafness**	○	○	
5. CARDIOVASCULAR			☐
a. pericarditis	○	○	
6. GASTROINTESTINAL			☐
a. ***mesenteric ischemia**	○	○	
7. PULMONARY			☐
a. pleurisy	○	○	
b. nodules or cavities	○	○	
c. other infiltrate secondary to WG	○	○	
d. endobronchial involvement	○	○	
e. ***alveolar hemorrhage**	○	○	
f. ***respiratory failure**	○	○	

DETERMINING DISEASE STATUS:
Severe Disease / Flare: ≥ 1 new/worse Major item
Limited Disease / Flare: ≥ new/worse Minor item
Persistent Disease: Continued (but not new/worse) activity
Remission: No active disease, including either new /worse or persistent items

	Persistent	New/Worse	None
8. RENAL			☐
a. hematuria (no RBC casts) (≥ 1 + or ≥ 10 RBC/hpf)	○	○	
b. ***RBC casts**	○	○	
c. ***rise in creatinine > 30% or fall in creatinine clearance > 25%**	○	○	
Note: If both hematuria and RBC casts are present, score only the RBC casts (the major item).			
9. NERVOUS SYSTEM			☐
a. ***meningitis**	○	○	
b. ***cord lesion**	○	○	
c. ***stroke**	○	○	
d. ***cranial nerve palsy**	○	○	
e. ***sensory peripheral neuropathy**	○	○	
f. ***motor mononeuritis multiplex**	○	○	

10. OTHER (describe all items and * items deemed major) ☐

Major

☐ _____ ○ ○
☐ _____ ○ ○
☐ _____ ○ ○
☐ _____ ○ ○

11. TOTAL NUMBER OF ITEMS: ☐

a.	b.	c.	d.
___ ___	___ ___	___ ___	___ ___
Major New / Worse	Minor New / Worse	Major Persistent	Minor Persistent

12. CURRENT DISEASE STATUS (check only one)

Severe Disease/Flare ☐
Limited Disease/Flare ☐
Persistent Disease ☐
Remission ☐

	0	1	2	3	4	5	6	7	8	9	10	
Remission	○	○	○	○	○	○	○	○	○	○	○	**Maximum activity**

Case 2

A 63 year old woman was diagnosed with granulomatosis with polyangiitis (Wegener's)(GPA) 5 years ago that manifested as blood nasal discharge with crusting, sinusitis, purpura, and glomerulonephritis with a peak creatinine of 4.7 mg/dL (420 μmol/L). For this she was treated with cyclophosphamide and prednisone with a return to a baseline creatinine of 1.4 mg/dL (123 μmol/l). Thereafter she was maintained on azathioprine.

She now presents with weight loss and a new cough. There is no fever, dyspnea, or hemoptysis. On physical examination you confirm a 5 kg weight loss but the remainder of her examination is unremarkable. Her ESR is 67 mm/hr (normal: 0-20), CRP 7.1 mg/ L, (normal: 0-4.9), creatinine is 1.4 mg/dL (124 μmol/L), urinalysis is negative for blood or protein, white blood cell count 12.3 x 10^9/L, hemoglobin 10.2 g/dL, platelet count is 565,000. Her chest CT scan shows 2 new pulmonary nodules: a left upper lobe 1.5 cm solid nodule and a right lower lobe 2.5 cm cavitary nodule. Bronchoscopy shows normal airways, no evidence of bloody return, cytology is negative for malignant cells, and stains and cultures are negative for infection.

CASE 2A: What is her current BVAS/WG ?

The patient is placed on rituximab and prednisone for active GPA. You are seeing her back 3 months later. She is back to her prior baseline weight and the cough has resolved. There are no new symptoms. Physical examination is unremarkable. Laboratory testing show that her ESR and CRP are now normal and the urinalysis remains negative for blood and protein. Repeat chest CT scan shows that the 1.5 cm nodule has resolved and the 2.5 cm cavitary nodule is now a 7 mm solid nodule, with no new nodules, cavities, or infiltrates seen.

CASE 2B: What is her current BVAS/WG ?

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CASE 2A

Mark only if abnormality is ascribable to the presence of active Wegener's Granulomatosis. Mark "Persistent" or "New/Worse" depending upon if the abnormality is persistent disease activity since the last assessment and not worse within the previous 28 days or if the abnormality is newly present or worse within the previous 28 days correspondingly. If no items are present in any section, tick "none". Major items are in bold and marked with *. All WG-related clinical features need to be documented on this form if they are related to active diseases. Use "OTHER" category as needed.

	Persistent	New/Worse	None
1. GENERAL			☐
a. arthralgia/arthritis	○	○	
b. fever (≥ 38 degrees C)	○	○	
2. CUTANEOUS			☐
a. purpura	○	○	
b. skin ulcer	○	○	
c. ***gangrene**	○	○	
3. MUCOUS MEMBRANES/EYES			☐
a. mouth ulcers	○	○	
b. conjunctivitis/episcleritis	○	○	
c. retro-orbital mass/proptosis	○	○	
d. uveitis	○	○	
e. ***scleritis**	○	○	
f. ***retinal exudates/haemorrhage**	○	○	
4. EAR, NOSE & THROAT			☐
a. bloody nasal discharge / nasal crusting / ulcer	○	○	
b. sinus involvement	○	○	
c. swollen salivary gland	○	○	
d. subglottic inflammation	○	○	
e. conductive deafness	○	○	
f. ***sensorineural deafness**	○	○	
5. CARDIOVASCULAR			☐
a. pericarditis	○	○	
6. GASTROINTESTINAL			☐
a. ***mesenteric ischemia**	○	○	
7. PULMONARY			☐
a. pleurisy	○	○	
b. nodules or cavities	○	○	
c. other infiltrate secondary to WG	○	○	
d. endobronchial involvement	○	○	
e. ***alveolar hemorrhage**	○	○	
f. ***respiratory failure**	○	○	

DETERMINING DISEASE STATUS:
Severe Disease / Flare: ≥ 1 new/worse Major item
Limited Disease / Flare: ≥ new/worse Minor item
Persistent Disease: Continued (but not new/worse) activity
Remission: No active disease, including either new /worse or persistent items

	Persistent	New/Worse	None
8. RENAL			☐
a. hematuria (no RBC casts) (≥ 1 + or ≥ 10 RBC/hpf)	○	○	
b. ***RBC casts**	○	○	
c. ***rise in creatinine > 30% or fall in creatinine clearance > 25%**	○	○	
Note: If both hematuria and RBC casts are present, score only the RBC casts (the major item).			
9. NERVOUS SYSTEM			☐
a. ***meningitis**	○	○	
b. ***cord lesion**	○	○	
c. ***stroke**	○	○	
d. ***cranial nerve palsy**	○	○	
e. ***sensory peripheral neuropathy**	○	○	
f. ***motor mononeuritis multiplex**	○	○	
10. OTHER (describe all items and * items deemed major)			☐
Major			
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	

11. TOTAL NUMBER OF ITEMS: ☐

a.	b.	c.	d.
___ ___	___ ___	___ ___	___ ___
Major New / Worse	Minor New / Worse	Major Persistent	Minor Persistent

12. CURRENT DISEASE STATUS (check only one)

- Severe Disease/Flare ☐
- Limited Disease/Flare ☐
- Persistent Disease ☐
- Remission ☐

	0	1	2	3	4	5	6	7	8	9	10	
Remission	○	○	○	○	○	○	○	○	○	○	○	**Maximum activity**

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CASE 2B

Mark only if abnormality is ascribable to the presence of active Wegener's Granulomatosis. Mark "Persistent" or "New/Worse" depending upon if the abnormality is persistent disease activity since the last assessment and not worse within the previous 28 days or if the abnormality is newly present or worse within the previous 28 days correspondingly. If no items are present in any section, tick "none". Major items are in bold and marked with *. All WG-related clinical features need to be documented on this form if they are related to active diseases. Use "OTHER" category as needed.

	Persistent	New/Worse	None
1. GENERAL			☐
a. arthralgia/arthritis	○	○	
b. fever (≥ 38 degrees C)	○	○	
2. CUTANEOUS			☐
a. purpura	○	○	
b. skin ulcer	○	○	
c. ***gangrene**	○	○	
3. MUCOUS MEMBRANES/EYES			☐
a. mouth ulcers	○	○	
b. conjunctivitis/episcleritis	○	○	
c. retro-orbital mass/proptosis	○	○	
d. uveitis	○	○	
e. ***scleritis**	○	○	
f. ***retinal exudates/haemorrhage**	○	○	
4. EAR, NOSE & THROAT			☐
a. bloody nasal discharge / nasal crusting / ulcer	○	○	
b. sinus involvement	○	○	
c. swollen salivary gland	○	○	
d. subglottic inflammation	○	○	
e. conductive deafness	○	○	
f. ***sensorineural deafness**	○	○	
5. CARDIOVASCULAR			☐
a. pericarditis	○	○	
6. GASTROINTESTINAL			☐
a. ***mesenteric ischemia**	○	○	
7. PULMONARY			☐
a. pleurisy	○	○	
b. nodules or cavities	○	○	
c. other infiltrate secondary to WG	○	○	
d. endobronchial involvement	○	○	
e. ***alveolar hemorrhage**	○	○	
f. ***respiratory failure**	○	○	

	Persistent	New/Worse	None
8. RENAL			☐
a. hematuria (no RBC casts) (≥ 1 + or ≥ 10 RBC/hpf)	○	○	
b. ***RBC casts**	○	○	
c. ***rise in creatinine > 30% or fall in creatinine clearance > 25%**	○	○	
Note: If both hematuria and RBC casts are present, score only the RBC casts (the major item).			
9. NERVOUS SYSTEM			☐
a. ***meningitis**	○	○	
b. ***cord lesion**	○	○	
c. ***stroke**	○	○	
d. ***cranial nerve palsy**	○	○	
e. ***sensory peripheral neuropathy**	○	○	
f. ***motor mononeuritis multiplex**	○	○	
10. OTHER (describe all items and * items deemed major)			☐
Major			
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	

11. TOTAL NUMBER OF ITEMS: ☐

a.	b.	c.	d.
___ ___	___ ___	___ ___	___ ___
Major New / Worse	Minor New / Worse	Major Persistent	Minor Persistent

DETERMINING DISEASE STATUS:
Severe Disease / Flare: ≥ 1 new/worse Major item
Limited Disease / Flare: ≥ new/worse Minor item
Persistent Disease: Continued (but not new/worse) activity
Remission: No active disease, including either new /worse or persistent items

12. CURRENT DISEASE STATUS (check only one)

- Severe Disease/Flare ☐
- Limited Disease/Flare ☐
- Persistent Disease ☐
- Remission ☐

	0	1	2	3	4	5	6	7	8	9	10	
Remission	○	○	○	○	○	○	○	○	○	○	○	**Maximum activity**

Case 3

A 20 year old woman has a 1 year history of granulomatosis with polyangiitis (Wegener's)(GPA) with manifestations that have included bloody nasal discharge and crusting, sinus involvement, sensorineural hearing loss, lung nodules, and mononeuritis multiplex with foot drop. During bronchoscopy she was noted to have some subglottic erythema without narrowing. For this, she was treated with rituximab and prednisone followed by methotrexate.

At a return clinic visit, she comments on increased dyspnea. She enjoys running as exercise and finds she can go for a shorter distance than previously but can do daily activities, including stairs, without difficulty. She has no cough, hemoptysis, nasal symptoms, or sinus pressure and she states that she is otherwise at her usual baseline health status. On physical examination she has mild stridor but is in no respiratory distress and the lungs are clear to auscultation. The remainder of her examination is unremarkable. She has a normal complete blood count, creatinine, ESR, CRP, and urinalysis. Chest CT scan shows clear lung fields.

She is seen by an otolaryngologist who finds evidence of a 20% subglottic stenosis. The mucosa has a pale pink appearance and there is no inflammation or ulceration.

CASE 3A: What is her current BVAS/WG ?

Two months later she returns to the otolaryngologist as she feels the dyspnea is now impacting her ability to do daily activities. She has no other new symptoms and her laboratories and chest x-ray are unremarkable. The otolaryngologist performs direct laryngoscopy with therapeutic dilation and injection. At surgery the lesion shows 60% narrowing of 1 cm in length in the subglottis just below the vocal cords. The subglottic mucosa now has an inflamed appearance. The visible bronchi below are normal without stenosis or mucosal inflammation.

CASE 3B: What is her current BVAS/WG ?

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CASE 3A

Mark only if abnormality is ascribable to the presence of active Wegener's Granulomatosis. Mark "Persistent" or "New/Worse" depending upon if the abnormality is persistent disease activity since the last assessment and not worse within the previous 28 days or if the abnormality is newly present or worse within the previous 28 days correspondingly. If no items are present in any section, tick "none". Major items are in bold and marked with *. All WG-related clinical features need to be documented on this form if they are related to active diseases. Use "OTHER" category as needed.

	Persistent	New/Worse	None
1. GENERAL			☐
a. arthralgia/arthritis	○	○	
b. fever (≥ 38 degrees C)	○	○	
2. CUTANEOUS			☐
a. purpura	○	○	
b. skin ulcer	○	○	
c. ***gangrene**	○	○	
3. MUCOUS MEMBRANES/EYES			☐
a. mouth ulcers	○	○	
b. conjunctivitis/episcleritis	○	○	
c. retro-orbital mass/proptosis	○	○	
d. uveitis	○	○	
e. ***scleritis**	○	○	
f. ***retinal exudates/haemorrhage**	○	○	
4. EAR, NOSE & THROAT			☐
a. bloody nasal discharge / nasal crusting / ulcer	○	○	
b. sinus involvement	○	○	
c. swollen salivary gland	○	○	
d. subglottic inflammation	○	○	
e. conductive deafness	○	○	
f. ***sensorineural deafness**	○	○	
5. CARDIOVASCULAR			☐
a. pericarditis	○	○	
6. GASTROINTESTINAL			☐
a. ***mesenteric ischemia**	○	○	
7. PULMONARY			☐
a. pleurisy	○	○	
b. nodules or cavities	○	○	
c. other infiltrate secondary to WG	○	○	
d. endobronchial involvement	○	○	
e. ***alveolar hemorrhage**	○	○	
f. ***respiratory failure**	○	○	

	Persistent	New/Worse	None
8. RENAL			☐
a. hematuria (no RBC casts) (≥ 1 + or ≥ 10 RBC/hpf)	○	○	
b. ***RBC casts**	○	○	
c. ***rise in creatinine > 30% or fall in creatinine clearance > 25%**	○	○	
Note: If both hematuria and RBC casts are present, score only the RBC casts (the major item).			
9. NERVOUS SYSTEM			☐
a. ***meningitis**	○	○	
b. ***cord lesion**	○	○	
c. ***stroke**	○	○	
d. ***cranial nerve palsy**	○	○	
e. ***sensory peripheral neuropathy**	○	○	
f. ***motor mononeuritis multiplex**	○	○	
10. OTHER (describe all items and * items deemed major)			☐
Major			
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	

11. TOTAL NUMBER OF ITEMS: ☐

a.	b.	c.	d.
___ ___	___ ___	___ ___	___ ___
Major New / Worse	Minor New / Worse	Major Persistent	Minor Persistent

DETERMINING DISEASE STATUS:
Severe Disease / Flare: ≥ 1 new/worse Major item
Limited Disease / Flare: ≥ new/worse Minor item
Persistent Disease: Continued (but not new/worse) activity
Remission: No active disease, including either new /worse or persistent items

12. CURRENT DISEASE STATUS (check only one)

- Severe Disease/Flare ☐
- Limited Disease/Flare ☐
- Persistent Disease ☐
- Remission ☐

	0	1	2	3	4	5	6	7	8	9	10	
Remission	○	○	○	○	○	○	○	○	○	○	○	**Maximum activity**

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CASE 3B

Mark only if abnormality is ascribable to the presence of active Wegener's Granulomatosis. Mark "Persistent" or "New/Worse" depending upon if the abnormality is persistent disease activity since the last assessment and not worse within the previous 28 days or if the abnormality is newly present or worse within the previous 28 days correspondingly. If no items are present in any section, tick "none". Major items are in bold and marked with *. All WG-related clinical features need to be documented on this form if they are related to active diseases. Use "OTHER" category as needed.

	Persistent	New/Worse	None
1. GENERAL			☐
a. arthralgia/arthritis	○	○	
b. fever (≥ 38 degrees C)	○	○	
2. CUTANEOUS			☐
a. purpura	○	○	
b. skin ulcer	○	○	
c. ***gangrene**	○	○	
3. MUCOUS MEMBRANES/EYES			☐
a. mouth ulcers	○	○	
b. conjunctivitis/episcleritis	○	○	
c. retro-orbital mass/proptosis	○	○	
d. uveitis	○	○	
e. ***scleritis**	○	○	
f. ***retinal exudates/haemorrhage**	○	○	
4. EAR, NOSE & THROAT			☐
a. bloody nasal discharge / nasal crusting / ulcer	○	○	
b. sinus involvement	○	○	
c. swollen salivary gland	○	○	
d. subglottic inflammation	○	○	
e. conductive deafness	○	○	
f. ***sensorineural deafness**	○	○	
5. CARDIOVASCULAR			☐
a. pericarditis	○	○	
6. GASTROINTESTINAL			☐
a. ***mesenteric ischemia**	○	○	
7. PULMONARY			☐
a. pleurisy	○	○	
b. nodules or cavities	○	○	
c. other infiltrate secondary to WG	○	○	
d. endobronchial involvement	○	○	
e. ***alveolar hemorrhage**	○	○	
f. ***respiratory failure**	○	○	

	Persistent	New/Worse	None
8. RENAL			☐
a. hematuria (no RBC casts) (≥ 1 + or ≥ 10 RBC/hpf)	○	○	
b. ***RBC casts**	○	○	
c. ***rise in creatinine > 30% or fall in creatinine clearance > 25%**	○	○	
Note: If both hematuria and RBC casts are present, score only the RBC casts (the major item).			
9. NERVOUS SYSTEM			☐
a. ***meningitis**	○	○	
b. ***cord lesion**	○	○	
c. ***stroke**	○	○	
d. ***cranial nerve palsy**	○	○	
e. ***sensory peripheral neuropathy**	○	○	
f. ***motor mononeuritis multiplex**	○	○	
10. OTHER (describe all items and * items deemed major)			☐
Major			
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	

11. TOTAL NUMBER OF ITEMS: ☐

a.	b.	c.	d.
___ ___	___ ___	___ ___	___ ___
Major New / Worse	Minor New / Worse	Major Persistent	Minor Persistent

DETERMINING DISEASE STATUS:
Severe Disease / Flare: ≥ 1 new/worse Major item
Limited Disease / Flare: ≥ new/worse Minor item
Persistent Disease: Continued (but not new/worse) activity
Remission: No active disease, including either new /worse or persistent items

12. CURRENT DISEASE STATUS (check only one)

- Severe Disease/Flare ☐
- Limited Disease/Flare ☐
- Persistent Disease ☐
- Remission ☐

	0	1	2	3	4	5	6	7	8	9	10	
Remission	○	○	○	○	○	○	○	○	○	○	○	**Maximum activity**

Case 4

A 55 year old man has a 10 year history of granulomatosis with polyangiitis (Wegener's)(GPA) with manifestations of bloody nasal discharge and crusting, sinusitis, conductive hearing loss, migratory arthritis, nodular skin lesions on the elbows, and cavitary pulmonary nodules. He was treated with prednisone and methotrexate. He has had 3 prior relapses and has had very significant nasal and sinus disease with collapse of the nasal bridge (saddlenose deformity) and bony erosion of the sinuses on CT. As a result of this he has mild chronic nasal crusting that he clears with twice a day irrigations. His last relapse was 2 years ago treated with rituximab and prednisone.

Today he presents with new diplopia and increased bloody nasal discharge. On physical examination he has mild proptosis and assessment of extraocular movement is abnormal with his right eye revealing a lack of lateral gaze. The optic nerve appears sharp and healthy. The nasal mucosa has an ulcerated appearance but the remainder of his examination is otherwise normal. Laboratory tests reveal mild anemia, creatinine is normal, ESR 22 mm/hr (0-20), CRP is normal, urinalysis is negative for protein and blood. Sinus/orbit CT scan reveals chronic unchanged thickening of the bilateral maxillary sinuses and unchanged erosion along the right medial orbital wall but there is new soft tissue extending into the medial orbit abutting the medial rectus muscle which also extends posteriorly behind the eye. Chest radiograph is normal.

CASE 4A: What is his current BVAS/WG ?

He is begun on treatment for active GPA with prednisone and rituximab and three months later his epistaxis has resolved and his nasal crusting has returned to its prior baseline. Diplopia has resolved. Physical examination shows resolution of the prior nasal membrane inflammation with mild crusting. Extraocular movement is much improved but there remains mild asymmetry in the right eye on the end of lateral gaze but there is no proptosis. Sinus/orbit CT scan is unchanged from the prior study.

CASE 4B: What is his current BVAS/WG ?

VCRC-EUVAS ABROGATE BVAS/WG
Training and Practice Cases
BVAS/WG

CASE 4A

Mark only if abnormality is ascribable to the presence of active Wegener's Granulomatosis. Mark "Persistent" or "New/Worse" depending upon if the abnormality is persistent disease activity since the last assessment and not worse within the previous 28 days or if the abnormality is newly present or worse within the previous 28 days correspondingly. If no items are present in any section, tick "none". Major items are in bold and marked with *. All WG-related clinical features need to be documented on this form if they are related to active diseases. Use "OTHER" category as needed.

	Persistent	New/Worse	None
1. GENERAL			☐
a. arthralgia/arthritis	○	○	
b. fever (≥ 38 degrees C)	○	○	
2. CUTANEOUS			☐
a. purpura	○	○	
b. skin ulcer	○	○	
c. ***gangrene**	○	○	
3. MUCOUS MEMBRANES/EYES			☐
a. mouth ulcers	○	○	
b. conjunctivitis/episcleritis	○	○	
c. retro-orbital mass/proptosis	○	○	
d. uveitis	○	○	
e. ***scleritis**	○	○	
f. ***retinal exudates/haemorrhage**	○	○	
4. EAR, NOSE & THROAT			☐
a. bloody nasal discharge / nasal crusting / ulcer	○	○	
b. sinus involvement	○	○	
c. swollen salivary gland	○	○	
d. subglottic inflammation	○	○	
e. conductive deafness	○	○	
f. ***sensorineural deafness**	○	○	
5. CARDIOVASCULAR			☐
a. pericarditis	○	○	
6. GASTROINTESTINAL			☐
a. ***mesenteric ischemia**	○	○	
7. PULMONARY			☐
a. pleurisy	○	○	
b. nodules or cavities	○	○	
c. other infiltrate secondary to WG	○	○	
d. endobronchial involvement	○	○	
e. ***alveolar hemorrhage**	○	○	
f. ***respiratory failure**	○	○	

DETERMINING DISEASE STATUS:
Severe Disease / Flare: ≥ 1 new/worse Major item
Limited Disease / Flare: ≥ new/worse Minor item
Persistent Disease: Continued (but not new/worse) activity
Remission: No active disease, including either new /worse or persistent items

	Persistent	New/Worse	None
8. RENAL			☐
a. hematuria (no RBC casts) (≥ 1 + or ≥ 10 RBC/hpf)	○	○	
b. ***RBC casts**	○	○	
c. ***rise in creatinine > 30% or fall in creatinine clearance > 25%**	○	○	
Note: If both hematuria and RBC casts are present, score only the RBC casts (the major item).			
9. NERVOUS SYSTEM			☐
a. ***meningitis**	○	○	
b. ***cord lesion**	○	○	
c. ***stroke**	○	○	
d. ***cranial nerve palsy**	○	○	
e. ***sensory peripheral neuropathy**	○	○	
f. ***motor mononeuritis multiplex**	○	○	
10. OTHER (describe all items and * items deemed major)			☐
Major			
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	

11. TOTAL NUMBER OF ITEMS: ☐

a.	b.	c.	d.
___ ___	___ ___	___ ___	___ ___
Major New / Worse	Minor New / Worse	Major Persistent	Minor Persistent

12. CURRENT DISEASE STATUS (check only one)

- Severe Disease/Flare ☐
- Limited Disease/Flare ☐
- Persistent Disease ☐
- Remission ☐

	0	1	2	3	4	5	6	7	8	9	10	
Remission	○	○	○	○	○	○	○	○	○	○	○	**Maximum activity**

VCRC-EUVAS ABROGATE BVAS/WG
Training and Practice Cases
BVAS/WG

CASE 4B

Mark only if abnormality is ascribable to the presence of active Wegener's Granulomatosis. Mark "Persistent" or "New/Worse" depending upon if the abnormality is persistent disease activity since the last assessment and not worse within the previous 28 days or if the abnormality is newly present or worse within the previous 28 days correspondingly. If no items are present in any section, tick "none". Major items are in bold and marked with *. All WG-related clinical features need to be documented on this form if they are related to active diseases. Use "OTHER" category as needed.

	Persistent	New/Worse	None
1. GENERAL			☐
a. arthralgia/arthritis	○	○	
b. fever (≥ 38 degrees C)	○	○	
2. CUTANEOUS			☐
a. purpura	○	○	
b. skin ulcer	○	○	
c. ***gangrene**	○	○	
3. MUCOUS MEMBRANES/EYES			☐
a. mouth ulcers	○	○	
b. conjunctivitis/episcleritis	○	○	
c. retro-orbital mass/proptosis	○	○	
d. uveitis	○	○	
e. ***scleritis**	○	○	
f. ***retinal exudates/haemorrhage**	○	○	
4. EAR, NOSE & THROAT			☐
a. bloody nasal discharge / nasal crusting / ulcer	○	○	
b. sinus involvement	○	○	
c. swollen salivary gland	○	○	
d. subglottic inflammation	○	○	
e. conductive deafness	○	○	
f. ***sensorineural deafness**	○	○	
5. CARDIOVASCULAR			☐
a. pericarditis	○	○	
6. GASTROINTESTINAL			☐
a. ***mesenteric ischemia**	○	○	
7. PULMONARY			☐
a. pleurisy	○	○	
b. nodules or cavities	○	○	
c. other infiltrate secondary to WG	○	○	
d. endobronchial involvement	○	○	
e. ***alveolar hemorrhage**	○	○	
f. ***respiratory failure**	○	○	

DETERMINING DISEASE STATUS:
Severe Disease / Flare: ≥ 1 new/worse Major item
Limited Disease / Flare: ≥ new/worse Minor item
Persistent Disease: Continued (but not new/worse) activity
Remission: No active disease, including either new /worse or persistent items

	Persistent	New/Worse	None
8. RENAL			☐
a. hematuria (no RBC casts) (≥ 1 + or ≥ 10 RBC/hpf)	○	○	
b. ***RBC casts**	○	○	
c. ***rise in creatinine > 30% or fall in creatinine clearance > 25%**	○	○	
Note: If both hematuria and RBC casts are present, score only the RBC casts (the major item).			
9. NERVOUS SYSTEM			☐
a. ***meningitis**	○	○	
b. ***cord lesion**	○	○	
c. ***stroke**	○	○	
d. ***cranial nerve palsy**	○	○	
e. ***sensory peripheral neuropathy**	○	○	
f. ***motor mononeuritis multiplex**	○	○	
10. OTHER (describe all items and * items deemed major)			☐
Major			
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	

11. TOTAL NUMBER OF ITEMS: ☐

a.	b.	c.	d.
___ ___	___ ___	___ ___	___ ___
Major New / Worse	Minor New / Worse	Major Persistent	Minor Persistent

12. CURRENT DISEASE STATUS (check only one)

Severe Disease/Flare	☐
Limited Disease/Flare	☐
Persistent Disease	☐
Remission	☐

	0	1	2	3	4	5	6	7	8	9	10	
Remission	○	○	○	○	○	○	○	○	○	○	○	**Maximum activity**

Case 5

A 35 year old woman has a 4 year history of granulomatosis with polyangiitis (Wegener's)(GPA) with manifestations that included blood nasal discharge, sinusitis, purpura, sensorineural hearing loss, and scleritis. For this she was treated with cyclophosphamide and prednisone followed by methotrexate, which she is continuing to receive.

One month ago she had a sinus infection that completely resolved after treatment with antibiotics. Two days ago she went to her otolaryngologist as she had worsened hearing loss without ear pain or pressure that was present ever since she had the sinus infection. On physical examination there was evidence of a clear serous otitis behind the right eardrum. There was no redness or purulence and her nasal mucosa were not inflamed. Audiogram revealed worsened conductive hearing loss and stable sensorineural hearing loss. The otolaryngologist started her on decongestants and oxymetazoline nasal spray.

At her follow-up with you today, she feels the hearing symptoms are slightly improved. On physical examination there is only a small residual amount of serous fluid. There remains no redness or purulence and her nasal mucosa are not inflamed. There are no features of active disease involving other organ sites by history, examination, labs or imaging.

CASE 5: What is her current BVAS/WG ?

VCRC-EUVAS ABROGATE BVAS/WG
Training and Practice Cases
BVAS/WG

CASE 5

Mark only if abnormality is ascribable to the presence of active Wegener's Granulomatosis. Mark "Persistent" or "New/Worse" depending upon if the abnormality is persistent disease activity since the last assessment and not worse within the previous 28 days or if the abnormality is newly present or worse within the previous 28 days correspondingly. If no items are present in any section, tick "none". Major items are in bold and marked with *. All WG-related clinical features need to be documented on this form if they are related to active diseases. Use "OTHER" category as needed.

	Persistent	New/Worse	None
1. GENERAL			☐
a. arthralgia/arthritis	○	○	
b. fever (≥ 38 degrees C)	○	○	
2. CUTANEOUS			☐
a. purpura	○	○	
b. skin ulcer	○	○	
c. ***gangrene**	○	○	
3. MUCOUS MEMBRANES/EYES			☐
a. mouth ulcers	○	○	
b. conjunctivitis/episcleritis	○	○	
c. retro-orbital mass/proptosis	○	○	
d. uveitis	○	○	
e. ***scleritis**	○	○	
f. ***retinal exudates/haemorrhage**	○	○	
4. EAR, NOSE & THROAT			☐
a. bloody nasal discharge / nasal crusting / ulcer	○	○	
b. sinus involvement	○	○	
c. swollen salivary gland	○	○	
d. subglottic inflammation	○	○	
e. conductive deafness	○	○	
f. ***sensorineural deafness**	○	○	
5. CARDIOVASCULAR			☐
a. pericarditis	○	○	
6. GASTROINTESTINAL			☐
a. ***mesenteric ischemia**	○	○	
7. PULMONARY			☐
a. pleurisy	○	○	
b. nodules or cavities	○	○	
c. other infiltrate secondary to WG	○	○	
d. endobronchial involvement	○	○	
e. ***alveolar hemorrhage**	○	○	
f. ***respiratory failure**	○	○	

	Persistent	New/Worse	None
8. RENAL			☐
a. hematuria (no RBC casts) (≥ 1 + or ≥ 10 RBC/hpf)	○	○	
b. ***RBC casts**	○	○	
c. ***rise in creatinine > 30% or fall in creatinine clearance > 25%**	○	○	
Note: If both hematuria and RBC casts are present, score only the RBC casts (the major item).			
9. NERVOUS SYSTEM			☐
a. ***meningitis**	○	○	
b. ***cord lesion**	○	○	
c. ***stroke**	○	○	
d. ***cranial nerve palsy**	○	○	
e. ***sensory peripheral neuropathy**	○	○	
f. ***motor mononeuritis multiplex**	○	○	
10. OTHER (describe all items and * items deemed major)			☐
Major			
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	

11. TOTAL NUMBER OF ITEMS: ☐

a.	b.	c.	d.
___ ___	___ ___	___ ___	___ ___
Major New / Worse	Minor New / Worse	Major Persistent	Minor Persistent

DETERMINING DISEASE STATUS:
Severe Disease / Flare: ≥ 1 new/worse Major item
Limited Disease / Flare: ≥ new/worse Minor item
Persistent Disease: Continued (but not new/worse) activity
Remission: No active disease, including either new /worse or persistent items

12. CURRENT DISEASE STATUS (check only one)

- Severe Disease/Flare ☐
- Limited Disease/Flare ☐
- Persistent Disease ☐
- Remission ☐

	0	1	2	3	4	5	6	7	8	9	10	
Remission	○	○	○	○	○	○	○	○	○	○	○	**Maximum activity**

Case 6

A 50 year old man has an 8 year history of granulomatosis with polyangiitis (Wegener's)(GPA) with prior manifestations that included bloody nasal discharge and crusting with development of saddlenose deformity, episcleritis, and pulmonary nodules treated with prednisone and methotrexate. He has had frequent relapses of nasal and sinus disease from his GPA. He comes in today out of concern for excess tearing. He otherwise feels at his baseline and his chronic nasal symptoms are unchanged. He has no other vision abnormalities and he denies pain, redness, diplopia, field cuts, or change in visual acuity. On physical examination he has visible tearing and frequently wipes away tears with a facial tissue. Ocular examination reveals no proptosis or periorbital redness or swelling. The conjunctiva is clear, extraocular movements are normal, and there is no lacrimal duct fullness or purulence. Laboratory tests are unremarkable. Sinus/orbit CT scan shows stable mucosal thickening in the maxillary and ethmoid sinuses without orbital disease and chest CT scan is unchanged. Examination by otolaryngology reveals no evidence of nasal mucosa inflammation but there is abundant mucosal scarring with nasolacrimal duct obstruction.

CASE 6: What is his current BVAS/WG ?

VCRC-EUVAS ABROGATE BVAS/WG
Training and Practice Cases
BVAS/WG

CASE 6

Mark only if abnormality is ascribable to the presence of active Wegener's Granulomatosis. Mark "Persistent" or "New/Worse" depending upon if the abnormality is persistent disease activity since the last assessment and not worse within the previous 28 days or if the abnormality is newly present or worse within the previous 28 days correspondingly. If no items are present in any section, tick "none". Major items are in bold and marked with *. All WG-related clinical features need to be documented on this form if they are related to active diseases. Use "OTHER" category as needed.

	Persistent	New/Worse	None
1. GENERAL			☐
a. arthralgia/arthritis	○	○	
b. fever (≥ 38 degrees C)	○	○	
2. CUTANEOUS			☐
a. purpura	○	○	
b. skin ulcer	○	○	
c. ***gangrene**	○	○	
3. MUCOUS MEMBRANES/EYES			☐
a. mouth ulcers	○	○	
b. conjunctivitis/episcleritis	○	○	
c. retro-orbital mass/proptosis	○	○	
d. uveitis	○	○	
e. ***scleritis**	○	○	
f. ***retinal exudates/haemorrhage**	○	○	
4. EAR, NOSE & THROAT			☐
a. bloody nasal discharge / nasal crusting / ulcer	○	○	
b. sinus involvement	○	○	
c. swollen salivary gland	○	○	
d. subglottic inflammation	○	○	
e. conductive deafness	○	○	
f. ***sensorineural deafness**	○	○	
5. CARDIOVASCULAR			☐
a. pericarditis	○	○	
6. GASTROINTESTINAL			☐
a. ***mesenteric ischemia**	○	○	
7. PULMONARY			☐
a. pleurisy	○	○	
b. nodules or cavities	○	○	
c. other infiltrate secondary to WG	○	○	
d. endobronchial involvement	○	○	
e. ***alveolar hemorrhage**	○	○	
f. ***respiratory failure**	○	○	

DETERMINING DISEASE STATUS:
Severe Disease / Flare: ≥ 1 new/worse Major item
Limited Disease / Flare: ≥ new/worse Minor item
Persistent Disease: Continued (but not new/worse) activity
Remission: No active disease, including either new /worse or persistent items

	Persistent	New/Worse	None
8. RENAL			☐
a. hematuria (no RBC casts) (≥ 1 + or ≥ 10 RBC/hpf)	○	○	
b. ***RBC casts**	○	○	
c. ***rise in creatinine > 30% or fall in creatinine clearance > 25%**	○	○	
Note: If both hematuria and RBC casts are present, score only the RBC casts (the major item).			
9. NERVOUS SYSTEM			☐
a. ***meningitis**	○	○	
b. ***cord lesion**	○	○	
c. ***stroke**	○	○	
d. ***cranial nerve palsy**	○	○	
e. ***sensory peripheral neuropathy**	○	○	
f. ***motor mononeuritis multiplex**	○	○	
10. OTHER (describe all items and * items deemed major)			☐
Major			
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	

11. TOTAL NUMBER OF ITEMS: ☐

a.	b.	c.	d.
___ ___	___ ___	___ ___	___ ___
Major New / Worse	Minor New / Worse	Major Persistent	Minor Persistent

12. CURRENT DISEASE STATUS (check only one)

- Severe Disease/Flare ☐
- Limited Disease/Flare ☐
- Persistent Disease ☐
- Remission ☐

	0	1	2	3	4	5	6	7	8	9	10	
Remission	○	○	○	○	○	○	○	○	○	○	○	**Maximum activity**

ANSWERS AND EXPLANATIONS FOR THE BVAS/WG NON-SEVERE TEST CASE ANSWERS

Please do not review these answers until AFTER you have completed the training cases and ALL 10 test cases

ANSWERS AND EXPLANATIONS

Case 1A

The following manifestations of AAV were all present within 28 days of evaluation:

Arthralgias

Bloody nasal discharge

Sinusitis

Fatigue. Because the fatigue is not a set item on the BVAS/WG form it is necessary to write it in the "Other" section. Fatigue is not considered a major item so it was not starred.

Total of 4 new minor items and a limited disease flare

.....

Case 1B

The patient has had resolution of the arthralgias, fatigue, and sinus pressure. His nasal symptoms are back to his baseline and his nasal membranes do not appear inflamed. Although his sinus CT continues to show thickening, this can persist as a result of damage and there are no new changes. There is no evidence of active vasculitis within the prior 28 days and his BVAS/WG would be 0 (Remission).

.....

CASE 1A

Mark only if abnormality is ascribable to the presence of active Wegener's Granulomatosis. Mark "Persistent" or "New/Worse" depending upon if the abnormality is persistent disease activity since the last assessment and not worse within the previous 28 days or if the abnormality is newly present or worse within the previous 28 days correspondingly. If no items are present in any section, tick "none". Major items are in bold and marked with *. All WG-related clinical features need to be documented on this form if they are related to active diseases. Use "OTHER" category as needed.

	Persistent	New/Worse	None
1. GENERAL			☐
a. arthralgia/arthritis	○	●	
b. fever (≥ 38 degrees C)	○	○	
2. CUTANEOUS			☒
a. purpura	○	○	
b. skin ulcer	○	○	
c. ***gangrene**	○	○	
3. MUCOUS MEMBRANES/EYES			☒
a. mouth ulcers	○	○	
b. conjunctivitis/episcleritis	○	○	
c. retro-orbital mass/proptosis	○	○	
d. uveitis	○	○	
e. ***scleritis**	○	○	
f. ***retinal exudates/haemorrhage**	○	○	
4. EAR, NOSE & THROAT			☐
a. bloody nasal discharge / nasal crusting / ulcer	○	●	
b. sinus involvement	○	●	
c. swollen salivary gland	○	○	
d. subglottic inflammation	○	○	
e. conductive deafness	○	○	
f. ***sensorineural deafness**	○	○	
5. CARDIOVASCULAR			☒
a. pericarditis	○	○	
6. GASTROINTESTINAL			☒
a. ***mesenteric ischemia**	○	○	
7. PULMONARY			☒
a. pleurisy	○	○	
b. nodules or cavities	○	○	
c. other infiltrate secondary to WG	○	○	
d. endobronchial involvement	○	○	
e. ***alveolar hemorrhage**	○	○	
f. ***respiratory failure**	○	○	

DETERMINING DISEASE STATUS:
Severe Disease / Flare: ≥ 1 new/worse Major item
Limited Disease / Flare: ≥ new/worse Minor item
Persistent Disease: Continued (but not new/worse) activity
Remission: No active disease, including either new /worse or persistent items

	Persistent	New/Worse	None
8. RENAL			☒
a. hematuria (no RBC casts) (≥ 1 + or ≥ 10 RBC/hpf)	○	○	
b. ***RBC casts**	○	○	
c. ***rise in creatinine > 30% or fall in creatinine clearance > 25%**	○	○	
Note: If both hematuria and RBC casts are present, score only the RBC casts (the major item).			
9. NERVOUS SYSTEM			☒
a. ***meningitis**	○	○	
b. ***cord lesion**	○	○	
c. ***stroke**	○	○	
d. ***cranial nerve palsy**	○	○	
e. ***sensory peripheral neuropathy**	○	○	
f. ***motor mononeuritis multiplex**	○	○	
10. OTHER (describe all items and * items deemed major)			☐
Major			
☐ FATIGUE_____	○	●	
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	

11. TOTAL NUMBER OF ITEMS: ☐

a.	b.	c.	d.
0 0	0 4	0 0	0 0
Major New / Worse	Minor New / Worse	Major Persistent	Minor Persistent

12. CURRENT DISEASE STATUS (check only one)

Severe Disease/Flare	☐
Limited Disease/Flare	☒
Persistent Disease	☐
Remission	☐

13. PHYSICIAN'S GLOBAL ASSESSMENT (PGA)
Mark to indicate the amount of WG disease activity (not including longstanding damage) within the previous 28 days:

0	1	2	3	4	5	6	7	8	9	10
○	○	●	○	○	○	○	○	○	○	○

Remission **Maximum activity**

Participant ID **CASE 1B**

Mark only if abnormality is ascribable to the presence of active Wegener's Granulomatosis. Mark "Persistent" or "New/Worse" depending upon if the abnormality is persistent disease activity since the last assessment and not worse within the previous 28 days or if the abnormality is newly present or worse within the previous 28 days correspondingly. If no items are present in any section, tick "none". Major items are in bold and marked with *. All WG-related clinical features need to be documented on this form if they are related to active diseases. Use "OTHER" category as needed.

	Persistent	New/Worse	None
1. GENERAL			☒
a. arthralgia/arthritis	○	○	
b. fever (≥ 38 degrees C)	○	○	
2. CUTANEOUS			☒
a. purpura	○	○	
b. skin ulcer	○	○	
c. ***gangrene**	○	○	
3. MUCOUS MEMBRANES/EYES			☒
a. mouth ulcers	○	○	
b. conjunctivitis/episcleritis	○	○	
c. retro-orbital mass/proptosis	○	○	
d. uveitis	○	○	
e. ***scleritis**	○	○	
f. ***retinal exudates/haemorrhage**	○	○	
4. EAR, NOSE & THROAT			☒
a. bloody nasal discharge / nasal crusting / ulcer	○	○	
b. sinus involvement	○	○	
c. swollen salivary gland	○	○	
d. subglottic inflammation	○	○	
e. conductive deafness	○	○	
f. ***sensorineural deafness**	○	○	
5. CARDIOVASCULAR			☒
a. pericarditis	○	○	
6. GASTROINTESTINAL			☒
a. ***mesenteric ischemia**	○	○	
7. PULMONARY			☒
a. pleurisy	○	○	
b. nodules or cavities	○	○	
c. other infiltrate secondary to WG	○	○	
d. endobronchial involvement	○	○	
e. ***alveolar hemorrhage**	○	○	
f. ***respiratory failure**	○	○	

DETERMINING DISEASE STATUS:
Severe Disease / Flare: ≥ 1 new/worse Major item
Limited Disease / Flare: ≥ new/worse Minor item
Persistent Disease: Continued (but not new/worse) activity
Remission: No active disease, including either new /worse or persistent items

	Persistent	New/Worse	None
8. RENAL			☒
a. hematuria (no RBC casts) (≥ 1 + or ≥ 10 RBC/hpf)	○	○	
b. ***RBC casts**	○	○	
c. ***rise in creatinine > 30% or fall in creatinine clearance > 25%**	○	○	
Note: If both hematuria and RBC casts are present, score only the RBC casts (the major item).			
9. NERVOUS SYSTEM			☒
a. ***meningitis**	○	○	
b. ***cord lesion**	○	○	
c. ***stroke**	○	○	
d. ***cranial nerve palsy**	○	○	
e. ***sensory peripheral neuropathy**	○	○	
f. ***motor mononeuritis multiplex**	○	○	
10. OTHER (describe all items and * items deemed major)			☒
Major			
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	

11. TOTAL NUMBER OF ITEMS: ☒

a.	b.	c.	d.
0 0	0 0	0 0	0 0
Major New / Worse	Minor New / Worse	Major Persistent	Minor Persistent

12. CURRENT DISEASE STATUS (check only one)

Severe Disease/Flare	☐
Limited Disease/Flare	☐
Persistent Disease	☐
Remission	☒

13. PHYSICIAN'S GLOBAL ASSESSMENT (PGA)
Mark to indicate the amount of WG disease activity (not including longstanding damage) within the previous 28 days:

0	1	2	3	4	5	6	7	8	9	10
●	○	○	○	○	○	○	○	○	○	○

Remission **Maximum activity**

Case 2A

The following manifestations of AAV were present within 28 days of evaluation:

Pulmonary nodule

Weight loss Because the weight loss is not a set item on the BVAS/WG form it is necessary to write it in the "Other" section. Weight loss is not considered a major item so it was not starred.

Total of 2 minor new items and a limited disease flare

.....

Case 2B

The patient has regained the lost weight. Although her chest CT shows that the prior 2.5 cm cavitary nodule is now a 7 mm solid nodule, persistent radiographic nodules can occur as a result of damage and there are no new infiltrates, nodules, or cavities. There is no evidence of active vasculitis within the prior 28 days and his BVAS/WG would be 0 (Remission).

.....

Participant ID **CASE 2A**

Mark only if abnormality is ascribable to the presence of active Wegener's Granulomatosis. Mark "Persistent" or "New/Worse" depending upon if the abnormality is persistent disease activity since the last assessment and not worse within the previous 28 days or if the abnormality is newly present or worse within the previous 28 days correspondingly. If no items are present in any section, tick "none". Major items are in bold and marked with *. All WG-related clinical features need to be documented on this form if they are related to active diseases. Use "OTHER" category as needed.

	Persistent	New/Worse	None
1. GENERAL			☒
a. arthralgia/arthritis	○	○	
b. fever (≥ 38 degrees C)	○	○	
2. CUTANEOUS			☒
a. purpura	○	○	
b. skin ulcer	○	○	
c. ***gangrene**	○	○	
3. MUCOUS MEMBRANES/EYES			☒
a. mouth ulcers	○	○	
b. conjunctivitis/episcleritis	○	○	
c. retro-orbital mass/proptosis	○	○	
d. uveitis	○	○	
e. ***scleritis**	○	○	
f. ***retinal exudates/haemorrhage**	○	○	
4. EAR, NOSE & THROAT			☒
a. bloody nasal discharge / nasal crusting / ulcer	○	○	
b. sinus involvement	○	○	
c. swollen salivary gland	○	○	
d. subglottic inflammation	○	○	
e. conductive deafness	○	○	
f. ***sensorineural deafness**	○	○	
5. CARDIOVASCULAR			☒
a. pericarditis	○	○	
6. GASTROINTESTINAL			☒
a. ***mesenteric ischemia**	○	○	
7. PULMONARY			☐
a. pleurisy	○	○	
b. nodules or cavities	○	●	
c. other infiltrate secondary to WG	○	○	
d. endobronchial involvement	○	○	
e. ***alveolar hemorrhage**	○	○	
f. ***respiratory failure**	○	○	

DETERMINING DISEASE STATUS:
Severe Disease / Flare: ≥ 1 new/worse Major item
Limited Disease / Flare: ≥ new/worse Minor item
Persistent Disease: Continued (but not new/worse) activity
Remission: No active disease, including either new /worse or persistent items

	Persistent	New/Worse	None
8. RENAL			☒
a. hematuria (no RBC casts) (≥ 1 + or ≥ 10 RBC/hpf)	○	○	
b. ***RBC casts**	○	○	
c. ***rise in creatinine > 30% or fall in creatinine clearance > 25%**	○	○	
Note: If both hematuria and RBC casts are present, score only the RBC casts (the major item).			
9. NERVOUS SYSTEM			☒
a. ***meningitis**	○	○	
b. ***cord lesion**	○	○	
c. ***stroke**	○	○	
d. ***cranial nerve palsy**	○	○	
e. ***sensory peripheral neuropathy**	○	○	
f. ***motor mononeuritis multiplex**	○	○	
10. OTHER (describe all items and * items deemed major)			☐
Major			
☐ WEIGHT LOSS_	○	●	
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	

11. TOTAL NUMBER OF ITEMS: ☐

a.	b.	c.	d.
0 0	0 2	0 0	0 0
Major New / Worse	Minor New / Worse	Major Persistent	Minor Persistent

12. CURRENT DISEASE STATUS (check only one)

Severe Disease/Flare	☐
Limited Disease/Flare	☒
Persistent Disease	☐
Remission	☐

13. PHYSICIAN'S GLOBAL ASSESSMENT (PGA)
Mark to indicate the amount of WG disease activity (not including longstanding damage) within the previous 28 days:

0	1	2	3	4	5	6	7	8	9	10
○	○	○	●	○	○	○	○	○	○	○

Remission **Maximum activity**

Participant ID **CASE 2B**

Mark only if abnormality is ascribable to the presence of active Wegener's Granulomatosis. Mark "Persistent" or "New/Worse" depending upon if the abnormality is persistent disease activity since the last assessment and not worse within the previous 28 days or if the abnormality is newly present or worse within the previous 28 days correspondingly. If no items are present in any section, tick "none". Major items are in bold and marked with *. All WG-related clinical features need to be documented on this form if they are related to active diseases. Use "OTHER" category as needed.

	Persistent	New/Worse	None
1. GENERAL			☒
a. arthralgia/arthritis	○	○	
b. fever (≥ 38 degrees C)	○	○	
2. CUTANEOUS			☒
a. purpura	○	○	
b. skin ulcer	○	○	
c. ***gangrene**	○	○	
3. MUCOUS MEMBRANES/EYES			☒
a. mouth ulcers	○	○	
b. conjunctivitis/episcleritis	○	○	
c. retro-orbital mass/proptosis	○	○	
d. uveitis	○	○	
e. ***scleritis**	○	○	
f. ***retinal exudates/haemorrhage**	○	○	
4. EAR, NOSE & THROAT			☒
a. bloody nasal discharge / nasal crusting / ulcer	○	○	
b. sinus involvement	○	○	
c. swollen salivary gland	○	○	
d. subglottic inflammation	○	○	
e. conductive deafness	○	○	
f. ***sensorineural deafness**	○	○	
5. CARDIOVASCULAR			☒
a. pericarditis	○	○	
6. GASTROINTESTINAL			☒
a. ***mesenteric ischemia**	○	○	
7. PULMONARY			☒
a. pleurisy	○	○	
b. nodules or cavities	○	○	
c. other infiltrate secondary to WG	○	○	
d. endobronchial involvement	○	○	
e. ***alveolar hemorrhage**	○	○	
f. ***respiratory failure**	○	○	

DETERMINING DISEASE STATUS:
Severe Disease / Flare: ≥ 1 new/worse Major item
Limited Disease / Flare: ≥ new/worse Minor item
Persistent Disease: Continued (but not new/worse) activity
Remission: No active disease, including either new /worse or persistent items

	Persistent	New/Worse	None
8. RENAL			☒
a. hematuria (no RBC casts) (≥ 1 + or ≥ 10 RBC/hpf)	○	○	
b. ***RBC casts**	○	○	
c. ***rise in creatinine > 30% or fall in creatinine clearance > 25%**	○	○	
Note: If both hematuria and RBC casts are present, score only the RBC casts (the major item).			
9. NERVOUS SYSTEM			☒
a. ***meningitis**	○	○	
b. ***cord lesion**	○	○	
c. ***stroke**	○	○	
d. ***cranial nerve palsy**	○	○	
e. ***sensory peripheral neuropathy**	○	○	
f. ***motor mononeuritis multiplex**	○	○	
10. OTHER (describe all items and * items deemed major)			☒
Major			
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	

11. TOTAL NUMBER OF ITEMS: ☒

a.	b.	c.	d.
0 0	0 0	0 0	0 0
Major New / Worse	Minor New / Worse	Major Persistent	Minor Persistent

12. CURRENT DISEASE STATUS (check only one)

Severe Disease/Flare	☐
Limited Disease/Flare	☐
Persistent Disease	☐
Remission	☒

13. PHYSICIAN'S GLOBAL ASSESSMENT (PGA)
Mark to indicate the amount of WG disease activity (not including longstanding damage) within the previous 28 days:

0	1	2	3	4	5	6	7	8	9	10
●	○	○	○	○	○	○	○	○	○	○

Remission **Maximum activity**

Case 3A

This patient now has evidence of a 20% subglottic narrowing with non-inflamed pale appearing mucosa. At the time of her presentation she was noted on bronchoscopy to have some subglottic erythema without narrowing. The presence of new narrowing in the setting of prior inflammation could represent scarring from her past disease, which would be supported by the current absence of mucosal inflammation. There is no evidence of active vasculitis within the prior 28 days and her BVAS/WG would be 0 (Remission).

.....

Case 3B

Two months later, this patient now has evidence of a 60% subglottic narrowing with inflamed appearing mucosa.

The following manifestations of AAV were present within 28 days of evaluation:

Subglottic inflammation

Total of 1 minor new item and a limited disease flare

.....

Participant ID **CASE 3A**

Mark only if abnormality is ascribable to the presence of active Wegener's Granulomatosis. Mark "Persistent" or "New/Worse" depending upon if the abnormality is persistent disease activity since the last assessment and not worse within the previous 28 days or if the abnormality is newly present or worse within the previous 28 days correspondingly. If no items are present in any section, tick "none". Major items are in bold and marked with *. All WG-related clinical features need to be documented on this form if they are related to active diseases. Use "OTHER" category as needed.

	Persistent	New/Worse	None
1. GENERAL			☒
a. arthralgia/arthritis	○	○	
b. fever (≥ 38 degrees C)	○	○	
2. CUTANEOUS			☒
a. purpura	○	○	
b. skin ulcer	○	○	
c. ***gangrene**	○	○	
3. MUCOUS MEMBRANES/EYES			☒
a. mouth ulcers	○	○	
b. conjunctivitis/episcleritis	○	○	
c. retro-orbital mass/proptosis	○	○	
d. uveitis	○	○	
e. ***scleritis**	○	○	
f. ***retinal exudates/haemorrhage**	○	○	
4. EAR, NOSE & THROAT			☒
a. bloody nasal discharge / nasal crusting / ulcer	○	○	
b. sinus involvement	○	○	
c. swollen salivary gland	○	○	
d. subglottic inflammation	○	○	
e. conductive deafness	○	○	
f. ***sensorineural deafness**	○	○	
5. CARDIOVASCULAR			☒
a. pericarditis	○	○	
6. GASTROINTESTINAL			☒
a. ***mesenteric ischemia**	○	○	
7. PULMONARY			☒
a. pleurisy	○	○	
b. nodules or cavities	○	○	
c. other infiltrate secondary to WG	○	○	
d. endobronchial involvement	○	○	
e. ***alveolar hemorrhage**	○	○	
f. ***respiratory failure**	○	○	

DETERMINING DISEASE STATUS:
Severe Disease / Flare: ≥ 1 new/worse Major item
Limited Disease / Flare: ≥ new/worse Minor item
Persistent Disease: Continued (but not new/worse) activity
Remission: No active disease, including either new /worse or persistent items

	Persistent	New/Worse	None
8. RENAL			☒
a. hematuria (no RBC casts) (≥ 1 + or ≥ 10 RBC/hpf)	○	○	
b. ***RBC casts**	○	○	
c. ***rise in creatinine > 30% or fall in creatinine clearance > 25%**	○	○	
Note: If both hematuria and RBC casts are present, score only the RBC casts (the major item).			
9. NERVOUS SYSTEM			☒
a. ***meningitis**	○	○	
b. ***cord lesion**	○	○	
c. ***stroke**	○	○	
d. ***cranial nerve palsy**	○	○	
e. ***sensory peripheral neuropathy**	○	○	
f. ***motor mononeuritis multiplex**	○	○	
10. OTHER (describe all items and * items deemed major)			☒
Major			
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	

11. TOTAL NUMBER OF ITEMS: ☒

a.	b.	c.	d.
0 0	0 0	0 0	0 0
Major New / Worse	Minor New / Worse	Major Persistent	Minor Persistent

12. CURRENT DISEASE STATUS (check only one)

Severe Disease/Flare	☐
Limited Disease/Flare	☐
Persistent Disease	☐
Remission	☒

13. PHYSICIAN'S GLOBAL ASSESSMENT (PGA)
Mark to indicate the amount of WG disease activity (not including longstanding damage) within the previous 28 days:

0	1	2	3	4	5	6	7	8	9	10
●	○	○	○	○	○	○	○	○	○	○

Remission **Maximum activity**

Participant ID **CASE 3B**

Mark only if abnormality is ascribable to the presence of active Wegener's Granulomatosis. Mark "Persistent" or "New/Worse" depending upon if the abnormality is persistent disease activity since the last assessment and not worse within the previous 28 days or if the abnormality is newly present or worse within the previous 28 days correspondingly. If no items are present in any section, tick "none". Major items are in bold and marked with *. All WG-related clinical features need to be documented on this form if they are related to active diseases. Use "OTHER" category as needed.

	Persistent	New/Worse	None
1. GENERAL			☒
a. arthralgia/arthritis	○	○	
b. fever (≥ 38 degrees C)	○	○	
2. CUTANEOUS			☒
a. purpura	○	○	
b. skin ulcer	○	○	
c. ***gangrene**	○	○	
3. MUCOUS MEMBRANES/EYES			☒
a. mouth ulcers	○	○	
b. conjunctivitis/episcleritis	○	○	
c. retro-orbital mass/proptosis	○	○	
d. uveitis	○	○	
e. ***scleritis**	○	○	
f. ***retinal exudates/haemorrhage**	○	○	
4. EAR, NOSE & THROAT			☐
a. bloody nasal discharge / nasal crusting / ulcer	○	○	
b. sinus involvement	○	○	
c. swollen salivary gland	○	○	
d. subglottic inflammation	○	●	
e. conductive deafness	○	○	
f. ***sensorineural deafness**	○	○	
5. CARDIOVASCULAR			☒
a. pericarditis	○	○	
6. GASTROINTESTINAL			☒
a. ***mesenteric ischemia**	○	○	
7. PULMONARY			☒
a. pleurisy	○	○	
b. nodules or cavities	○	○	
c. other infiltrate secondary to WG	○	○	
d. endobronchial involvement	○	○	
e. ***alveolar hemorrhage**	○	○	
f. ***respiratory failure**	○	○	

DETERMINING DISEASE STATUS:
Severe Disease / Flare: ≥ 1 new/worse Major item
Limited Disease / Flare: ≥ new/worse Minor item
Persistent Disease: Continued (but not new/worse) activity
Remission: No active disease, including either new /worse or persistent items

	Persistent	New/Worse	None
8. RENAL			☒
a. hematuria (no RBC casts) (≥ 1 + or ≥ 10 RBC/hpf)	○	○	
b. ***RBC casts**	○	○	
c. ***rise in creatinine > 30% or fall in creatinine clearance > 25%**	○	○	
Note: If both hematuria and RBC casts are present, score only the RBC casts (the major item).			
9. NERVOUS SYSTEM			☒
a. ***meningitis**	○	○	
b. ***cord lesion**	○	○	
c. ***stroke**	○	○	
d. ***cranial nerve palsy**	○	○	
e. ***sensory peripheral neuropathy**	○	○	
f. ***motor mononeuritis multiplex**	○	○	
10. OTHER (describe all items and * items deemed major)			☒
Major			
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	

11. TOTAL NUMBER OF ITEMS: ☐

a.	b.	c.	d.
0 0	0 1	0 0	0 0
Major New / Worse	Minor New / Worse	Major Persistent	Minor Persistent

12. CURRENT DISEASE STATUS (check only one)

Severe Disease/Flare	☐
Limited Disease/Flare	☒
Persistent Disease	☐
Remission	☐

13. PHYSICIAN'S GLOBAL ASSESSMENT (PGA)
Mark to indicate the amount of WG disease activity (not including longstanding damage) within the previous 28 days:

0	1	2	3	4	5	6	7	8	9	10
○	●	○	○	○	○	○	○	○	○	○

Remission **Maximum activity**

Case 4A

The following manifestations of AAV were present within 28 days of evaluation:

Retro-orbital mass/proptosis

Bloody nasal discharge

Total of 2 minor new items and a limited disease flare

.....

Case 4B

The patient has had resolution of the bloody nasal drainage. There is persistent crusting but he had that prior to the relapse and has represented past damage. His protosis has resolved and other ocular symptoms have improved. Although his orbit CT continues to show presence of the mass, fibrosis with persistence of the retro-orbital mass commonly occurs as a result of damage and there are no new changes. There is no evidence of active vasculitis within the prior 28 days and his BVAS/WG would be 0 (Remission).

.....

Participant ID **CASE 4A**

Mark only if abnormality is ascribable to the presence of active Wegener's Granulomatosis. Mark "Persistent" or "New/Worse" depending upon if the abnormality is persistent disease activity since the last assessment and not worse within the previous 28 days or if the abnormality is newly present or worse within the previous 28 days correspondingly. If no items are present in any section, tick "none". Major items are in bold and marked with *. All WG-related clinical features need to be documented on this form if they are related to active diseases. Use "OTHER" category as needed.

	Persistent	New/Worse	None
1. GENERAL			☒
a. arthralgia/arthritis	○	○	
b. fever (≥ 38 degrees C)	○	○	
2. CUTANEOUS			☒
a. purpura	○	○	
b. skin ulcer	○	○	
c. ***gangrene**	○	○	
3. MUCOUS MEMBRANES/EYES			☐
a. mouth ulcers	○	○	
b. conjunctivitis/episcleritis	○	○	
c. retro-orbital mass/proptosis	○	●	
d. uveitis	○	○	
e. ***scleritis**	○	○	
f. ***retinal exudates/haemorrhage**	○	○	
4. EAR, NOSE & THROAT			☐
a. bloody nasal discharge / nasal crusting / ulcer	○	●	
b. sinus involvement	○	○	
c. swollen salivary gland	○	○	
d. subglottic inflammation	○	○	
e. conductive deafness	○	○	
f. ***sensorineural deafness**	○	○	
5. CARDIOVASCULAR			☒
a. pericarditis	○	○	
6. GASTROINTESTINAL			☒
a. ***mesenteric ischemia**	○	○	
7. PULMONARY			☒
a. pleurisy	○	○	
b. nodules or cavities	○	○	
c. other infiltrate secondary to WG	○	○	
d. endobronchial involvement	○	○	
e. ***alveolar hemorrhage**	○	○	
f. ***respiratory failure**	○	○	

DETERMINING DISEASE STATUS:
Severe Disease / Flare: ≥ 1 new/worse Major item
Limited Disease / Flare: ≥ new/worse Minor item
Persistent Disease: Continued (but not new/worse) activity
Remission: No active disease, including either new /worse or persistent items

	Persistent	New/Worse	None
8. RENAL			☒
a. hematuria (no RBC casts) (≥ 1 + or ≥ 10 RBC/hpf)	○	○	
b. ***RBC casts**	○	○	
c. ***rise in creatinine > 30% or fall in creatinine clearance > 25%**	○	○	
Note: If both hematuria and RBC casts are present, score only the RBC casts (the major item).			
9. NERVOUS SYSTEM			☒
a. ***meningitis**	○	○	
b. ***cord lesion**	○	○	
c. ***stroke**	○	○	
d. ***cranial nerve palsy**	○	○	
e. ***sensory peripheral neuropathy**	○	○	
f. ***motor mononeuritis multiplex**	○	○	
10. OTHER (describe all items and * items deemed major)			☒
Major			
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	

11. TOTAL NUMBER OF ITEMS: ☐

a.	b.	c.	d.
0 0	0 2	0 0	0 0
Major New / Worse	Minor New / Worse	Major Persistent	Minor Persistent

12. CURRENT DISEASE STATUS (check only one)

Severe Disease/Flare	☐
Limited Disease/Flare	☒
Persistent Disease	☐
Remission	☐

13. PHYSICIAN'S GLOBAL ASSESSMENT (PGA)
Mark to indicate the amount of WG disease activity (not including longstanding damage) within the previous 28 days:

0	1	2	3	4	5	6	7	8	9	10
○	○	○	●	○	○	○	○	○	○	○

Remission **Maximum activity**

VCRC-EUVAS ABROGATE BVAS/WG
Training and Practice Cases
BVAS/WG

CASE 4B

Mark only if abnormality is ascribable to the presence of active Wegener's Granulomatosis. Mark "Persistent" or "New/Worse" depending upon if the abnormality is persistent disease activity since the last assessment and not worse within the previous 28 days or if the abnormality is newly present or worse within the previous 28 days correspondingly. If no items are present in any section, tick "none". Major items are in bold and marked with *. All WG-related clinical features need to be documented on this form if they are related to active diseases. Use "OTHER" category as needed.

	Persistent	New/Worse	None
1. GENERAL			☐
a. arthralgia/arthritis	○	○	
b. fever (≥ 38 degrees C)	○	○	
2. CUTANEOUS			☐
a. purpura	○	○	
b. skin ulcer	○	○	
c. ***gangrene**	○	○	
3. MUCOUS MEMBRANES/EYES			☐
a. mouth ulcers	○	○	
b. conjunctivitis/episcleritis	○	○	
c. retro-orbital mass/proptosis	○	○	
d. uveitis	○	○	
e. ***scleritis**	○	○	
f. ***retinal exudates/haemorrhage**	○	○	
4. EAR, NOSE & THROAT			☐
a. bloody nasal discharge / nasal crusting / ulcer	○	○	
b. sinus involvement	○	○	
c. swollen salivary gland	○	○	
d. subglottic inflammation	○	○	
e. conductive deafness	○	○	
f. ***sensorineural deafness**	○	○	
5. CARDIOVASCULAR			☐
a. pericarditis	○	○	
6. GASTROINTESTINAL			☐
a. ***mesenteric ischemia**	○	○	
7. PULMONARY			☐
a. pleurisy	○	○	
b. nodules or cavities	○	○	
c. other infiltrate secondary to WG	○	○	
d. endobronchial involvement	○	○	
e. ***alveolar hemorrhage**	○	○	
f. ***respiratory failure**	○	○	

DETERMINING DISEASE STATUS:
Severe Disease / Flare: ≥ 1 new/worse Major item
Limited Disease / Flare: ≥ new/worse Minor item
Persistent Disease: Continued (but not new/worse) activity
Remission: No active disease, including either new /worse or persistent items

	Persistent	New/Worse	None
8. RENAL			☐
a. hematuria (no RBC casts) (≥ 1 + or ≥ 10 RBC/hpf)	○	○	
b. ***RBC casts**	○	○	
c. ***rise in creatinine > 30% or fall in creatinine clearance > 25%**	○	○	
Note: If both hematuria and RBC casts are present, score only the RBC casts (the major item).			
9. NERVOUS SYSTEM			☐
a. ***meningitis**	○	○	
b. ***cord lesion**	○	○	
c. ***stroke**	○	○	
d. ***cranial nerve palsy**	○	○	
e. ***sensory peripheral neuropathy**	○	○	
f. ***motor mononeuritis multiplex**	○	○	
10. OTHER (describe all items and * items deemed major)			☐
Major			
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	

11. TOTAL NUMBER OF ITEMS: ☐

a.	b.	c.	d.
___ ___	___ ___	___ ___	___ ___
Major New / Worse	Minor New / Worse	Major Persistent	Minor Persistent

12. CURRENT DISEASE STATUS (check only one)

Severe Disease/Flare	☐
Limited Disease/Flare	☐
Persistent Disease	☐
Remission	☐

	0	1	2	3	4	5	6	7	8	9	10	
Remission	○	○	○	○	○	○	○	○	○	○	○	**Maximum activity**

Case 5

This patient presents with decreased hearing. She has had past nasal and sinus disease and recently had a sinus infection. On exam there is evidence of a clear serous otitis without redness or purulence and thus this is not suggestive of infection. There was no evidence by audiogram of worsened sensorineural hearing loss and this was consistent with a conductive hearing loss. She has had improvement already just 2 days into treatment with decongestants and oxymetazoline. This suggests that she had a serous otitis media secondary to the recent sinus infection, which can commonly occur in patients with sinus disease. There is no evidence of active vasculitis within the prior 28 days and her BVAS/WG would be 0 (Remission).

.....

Case 6

This patient presents with excess tearing and has evidence of nasolacrimal duct obstruction. He has longstanding nasal and sinus disease with evidence of prior damage manifest as saddlenose deformity. There are no other features of active disease and on his exam there is evidence of mucosal scarring obstructing the nasolacrimal duct. Although the clinical symptom that the patient is experiencing with tearing is new, this does not mean that this is due to active disease and can be the result of scarring as was the case in this patient. There is no evidence of active vasculitis within the prior 28 days and his BVAS/WG would be 0 (Remission).

Participant ID **CASE 5**

Mark only if abnormality is ascribable to the presence of active Wegener's Granulomatosis. Mark "Persistent" or "New/Worse" depending upon if the abnormality is persistent disease activity since the last assessment and not worse within the previous 28 days or if the abnormality is newly present or worse within the previous 28 days correspondingly. If no items are present in any section, tick "none". Major items are in bold and marked with *. All WG-related clinical features need to be documented on this form if they are related to active diseases. Use "OTHER" category as needed.

	Persistent	New/Worse	None
1. GENERAL			☒
a. arthralgia/arthritis	○	○	
b. fever (≥ 38 degrees C)	○	○	
2. CUTANEOUS			☒
a. purpura	○	○	
b. skin ulcer	○	○	
c. ***gangrene**	○	○	
3. MUCOUS MEMBRANES/EYES			☒
a. mouth ulcers	○	○	
b. conjunctivitis/episcleritis	○	○	
c. retro-orbital mass/proptosis	○	○	
d. uveitis	○	○	
e. ***scleritis**	○	○	
f. ***retinal exudates/haemorrhage**	○	○	
4. EAR, NOSE & THROAT			☒
a. bloody nasal discharge / nasal crusting / ulcer	○	○	
b. sinus involvement	○	○	
c. swollen salivary gland	○	○	
d. subglottic inflammation	○	○	
e. conductive deafness	○	○	
f. ***sensorineural deafness**	○	○	
5. CARDIOVASCULAR			☒
a. pericarditis	○	○	
6. GASTROINTESTINAL			☒
a. ***mesenteric ischemia**	○	○	
7. PULMONARY			☒
a. pleurisy	○	○	
b. nodules or cavities	○	○	
c. other infiltrate secondary to WG	○	○	
d. endobronchial involvement	○	○	
e. ***alveolar hemorrhage**	○	○	
f. ***respiratory failure**	○	○	

DETERMINING DISEASE STATUS:
Severe Disease / Flare: ≥ 1 new/worse Major item
Limited Disease / Flare: ≥ new/worse Minor item
Persistent Disease: Continued (but not new/worse) activity
Remission: No active disease, including either new /worse or persistent items

	Persistent	New/Worse	None
8. RENAL			☒
a. hematuria (no RBC casts) (≥ 1 + or ≥ 10 RBC/hpf)	○	○	
b. ***RBC casts**	○	○	
c. ***rise in creatinine > 30% or fall in creatinine clearance > 25%**	○	○	
Note: If both hematuria and RBC casts are present, score only the RBC casts (the major item).			
9. NERVOUS SYSTEM			☒
a. ***meningitis**	○	○	
b. ***cord lesion**	○	○	
c. ***stroke**	○	○	
d. ***cranial nerve palsy**	○	○	
e. ***sensory peripheral neuropathy**	○	○	
f. ***motor mononeuritis multiplex**	○	○	
10. OTHER (describe all items and * items deemed major)			☒
Major			
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	

11. TOTAL NUMBER OF ITEMS: ☒

a.	b.	c.	d.
0 0	0 0	0 0	0 0
Major New / Worse	Minor New / Worse	Major Persistent	Minor Persistent

12. CURRENT DISEASE STATUS (check only one)

Severe Disease/Flare	☐
Limited Disease/Flare	☐
Persistent Disease	☐
Remission	☒

13. PHYSICIAN'S GLOBAL ASSESSMENT (PGA)
Mark to indicate the amount of WG disease activity (not including longstanding damage) within the previous 28 days:

0	1	2	3	4	5	6	7	8	9	10
●	○	○	○	○	○	○	○	○	○	○

Remission **Maximum activity**

Participant ID **CASE 6**

Mark only if abnormality is ascribable to the presence of active Wegener's Granulomatosis. Mark "Persistent" or "New/Worse" depending upon if the abnormality is persistent disease activity since the last assessment and not worse within the previous 28 days or if the abnormality is newly present or worse within the previous 28 days correspondingly. If no items are present in any section, tick "none". Major items are in bold and marked with *. All WG-related clinical features need to be documented on this form if they are related to active diseases. Use "OTHER" category as needed.

	Persistent	New/Worse	None
1. GENERAL			☒
a. arthralgia/arthritis	○	○	
b. fever (≥ 38 degrees C)	○	○	
2. CUTANEOUS			☒
a. purpura	○	○	
b. skin ulcer	○	○	
c. ***gangrene**	○	○	
3. MUCOUS MEMBRANES/EYES			☒
a. mouth ulcers	○	○	
b. conjunctivitis/episcleritis	○	○	
c. retro-orbital mass/proptosis	○	○	
d. uveitis	○	○	
e. ***scleritis**	○	○	
f. ***retinal exudates/haemorrhage**	○	○	
4. EAR, NOSE & THROAT			☒
a. bloody nasal discharge / nasal crusting / ulcer	○	○	
b. sinus involvement	○	○	
c. swollen salivary gland	○	○	
d. subglottic inflammation	○	○	
e. conductive deafness	○	○	
f. ***sensorineural deafness**	○	○	
5. CARDIOVASCULAR			☒
a. pericarditis	○	○	
6. GASTROINTESTINAL			☒
a. ***mesenteric ischemia**	○	○	
7. PULMONARY			☒
a. pleurisy	○	○	
b. nodules or cavities	○	○	
c. other infiltrate secondary to WG	○	○	
d. endobronchial involvement	○	○	
e. ***alveolar hemorrhage**	○	○	
f. ***respiratory failure**	○	○	

DETERMINING DISEASE STATUS:
Severe Disease / Flare: ≥ 1 new/worse Major item
Limited Disease / Flare: ≥ new/worse Minor item
Persistent Disease: Continued (but not new/worse) activity
Remission: No active disease, including either new /worse or persistent items

	Persistent	New/Worse	None
8. RENAL			☒
a. hematuria (no RBC casts) (≥ 1 + or ≥ 10 RBC/hpf)	○	○	
b. ***RBC casts**	○	○	
c. ***rise in creatinine > 30% or fall in creatinine clearance > 25%**	○	○	
Note: If both hematuria and RBC casts are present, score only the RBC casts (the major item).			
9. NERVOUS SYSTEM			☒
a. ***meningitis**	○	○	
b. ***cord lesion**	○	○	
c. ***stroke**	○	○	
d. ***cranial nerve palsy**	○	○	
e. ***sensory peripheral neuropathy**	○	○	
f. ***motor mononeuritis multiplex**	○	○	
10. OTHER (describe all items and * items deemed major)			☒
Major			
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	
☐ _____	○	○	

11. TOTAL NUMBER OF ITEMS: ☒

a.	b.	c.	d.
0 0	0 0	0 0	0 0
Major New / Worse	Minor New / Worse	Major Persistent	Minor Persistent

12. CURRENT DISEASE STATUS (check only one)

Severe Disease/Flare	☐
Limited Disease/Flare	☐
Persistent Disease	☐
Remission	☒

13. PHYSICIAN'S GLOBAL ASSESSMENT (PGA)
Mark to indicate the amount of WG disease activity (not including longstanding damage) within the previous 28 days:

0	1	2	3	4	5	6	7	8	9	10
●	○	○	○	○	○	○	○	○	○	○

Remission **Maximum activity**

Test Cases Score Sheet

Name: _____

Institution: _____

Date: __ __ / __ __ __ / __ __ __ __

BVAS/WG includes 10 sections documenting clinical manifestations of disease:

1. General
2. Cutaneous
3. Mucous Membrane/Eyes
4. Ear, Nose & Throat
5. Cardiovascular
6. Gastrointestinal
7. Pulmonary
8. Renal
9. Nervous System
10. Other

When self-scoring the BVAS/WG test cases, you score 1 point for each section correctly completed.
Correctly completed means that all the correct items and none of the incorrect items were selected.
If there are no active items in a section then "none" is the correct answer.
The maximum you can score on each case is 10.

Test Case 1A: Score _____/10

Test Case 1B: Score _____/10

Test Case 2A: Score _____/10

Test Case 2B: Score _____/10

Test Case 3A: Score _____/10

Test Case 3B: Score _____/10

Test Case 4A: Score _____/10

Test Case 4B: Score _____/10

Test Case 5: Score _____/10

Test Case 6: Score _____/10

Total score (sum of the score on each case): _____/100

Please scan/email or fax this form to the ABROGATE Data coordinator:

Cristina Burroughs
Fax: +1 813 910-1225
Email: Cristina.Burroughs@epi.usf.edu

ABROGATE BVAS/WG Investigator Training Certification

This form must be completed by *every* investigator who will be completing the BVAS/WG before he or she is eligible to conduct an assessment of a study subject. At least one investigator at each site must be certified for BVAS/WG before the site can be opened for recruitment.

☐ I have fully read and understand the "BVAS/WG-Introduction, Instructions, and Glossary"

☐ I have reviewed the "BVAS/WG-Training Cases"

☐ I have completed, on my own, the 10 BVAS/WG test cases

- ☐ I received a passing score
- ☐ I did not achieve a passing score

_____
Signature of investigator

_____
Printed name of investigator

_____
Study site

_____
Date

Please scan/email or fax this form AND the test cases score sheet to the ABROGATE Data coordinator:

Cristina Burroughs
Fax: +1 813 910-1225
Email: Cristina.Burroughs@epi.usf.edu