

VCRC-OMERACT ANCA-Vasculitis Outcome Measure Initiative Comparative Outcome Measure Exercise-Training Cases

Introduction to Training Cases

The purpose of this preliminary exercise is to familiarize you with the “rules” and logistics of the five instruments under study. Please do not read the Training Cases until you have reviewed the Manual of Operations for each instrument.

The VCRC-OMERACT Outcomes Measure Initiative practice cases are meant to familiarize you with the use of the instruments and therefore include a variety of items. There are also some deliberately misleading statements in the practice cases, which represent old disease or irrelevant items in order to help you distinguish between current disease activity and what is probably best termed “damage”. You also need to recognize that in addition to including different sets of manifestations, the different instruments also deal with persistent disease differently.

In the following three practice cases, there is a short description of a patient. In all of the case histories, you are provided with “positive” data. If there is no mention of any particular feature, you may assume that it is not present or that it is normal. Please read Training Case I and then score it using each of 5 measures we are testing in the exact order of attached pages. Once you have scored the first case, you can review the “answers” as outlined by members of the group highly familiar with each instrument. Please then repeat this process for Training Cases II and III.

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Training Case I:

A 76-year-old man has noted a three week history of fatigue, 4kg weight loss, and joint pains in his hands and ankles. He has noticed some nasal crusting and bleeding, with severe tenderness over the cheeks. His past medical history includes a twenty-year history of osteoarthritis affecting his knees and DIP joints. Ten years before presentation he suffered a mild left hemiparesis, but subsequently recovered most of his neurological function. Physical examination reveals swollen, boggy ankles and MCP joints. He has increased reflexes in his left arm and leg, and an upgoing plantar response. Urinalysis reveals 15 red cells per high power field and occasional RBC casts. His serum creatinine has risen from 1.69 mg/dl (150 umol/l) one week ago to 2.82 mg/dl (250 umol/l) today. A diagnosis of WG was established on renal and nasal biopsy.

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Training Case II:

A 46-year-old man presents with acute onset of hearing loss, aural discharge, epistaxis, nasal discharge and crusting, and fever. Physical examination reveals a temperature of 38.3° C. He is found to have bilateral conductive hearing loss. Urinalysis and serum creatinine are both normal. A nasal biopsy shows granulomata, and he has a positive test for C-ANCA/anti-PR3. Chest x-ray is normal. He is started on methotrexate and prednisolone.

Three weeks later, he returns to clinic feeling worse. He has been tired, with increasing amounts of epistaxis (daily instead of alternate days), less aural discharge, with some improvement in hearing. The nasal crusting is worse. He now complains of sinus discomfort. Physical examination reveals blood pressure of 175/98 mm/Hg (increased from prior visit). He is afebrile. He has tender maxillary sinuses. Urinalysis shows hematuria (++) , proteinuria (++) , but no RBC casts. Serum creatinine is still normal. He is sent for a renal opinion.

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Training Case III:

A 59-year-old man presented with ischemic fingers, splinter hemorrhages, and gangrene ten months prior to the current visit. At that time, he was found to have a serum creatinine of 5.08 mg/dl (450 umol/L) and his urinalysis had shown too numerous to count RBCs, RBC casts, and marked proteinuria (+++). Several months before that presentation, he had developed an inflammatory arthritis and bilateral hearing difficulties, for which an ENT surgeon had inserted tympanostomy tubes. The diagnosis of Wegener's granulomatosis was confirmed by a renal biopsy, which demonstrated segmental, necrotizing glomerulonephritis of a pauci-immune nature. He was strongly C-ANCA/anti-PR-3 positive. In retrospect, he recalled being told two years ago that he had a "deviated" nasal septum. In fact, at the time of presentation he was found to have a nasal septal perforation and bloody nasal crusts. He responded promptly to cyclophosphamide and prednisolone.

He has now been off cyclophosphamide for five months and is taking methotrexate 20 mg/week. He has also tapered off his prednisolone four months prior to the current visit. His serum creatinine has been stable in the 1.92-2.15 mm/dl range (170-190 umol/l) since discharge from his initial hospitalization. He underwent amputation of a couple of gangrenous fingertips and occasionally has pain at the amputation site, but there is no coolness or evidence of further digital ischaemia. His hematuria has never completely resolved, remaining on the order of 5-10 RBCs per high power field, and he continues to have stable proteinuria (+). He reports occasional sinus stuffiness and a general lack of energy, but has returned to work full-time.

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ANSWERS

Training Case I

BVAS.1 and BVAS.2-CASE I

The relevant abnormalities for Case I are: general: (*malaise, weight loss ≥ 2 kg and arthralgia*), ENT (*bloody nasal discharge and crusting and the cheek tenderness represents sinus involvement*), renal (*haematuria, rise in creatinine $>30\%$ and creatinine 250 - 499*). Since this is the first visit you would score the actual creatinine value. The osteoarthritis and old stroke are irrelevant as are the clinical signs relating to these problems. Therefore the scores should be: BVAS.1 = 21, BVAS.2 = 0

This gives a total calculated BVAS.1 (new/worse) score of 21: general 3, cutaneous 0, mucous membranes 0, ENT 6, chest 0, cardiovascular 0, abdominal 0, renal 12, nervous system 0.

Remember that BVAS 1 score is for new/worse disease and BVAS 2 is for persistent but active disease.

BVAS/WG-CASE I

The appropriate items to score for Case I are: general (arthralgia/arthritis), ENT (nasal discharge, sinus involvement), Renal (RBC casts and rise in creatinine). You should also write in “weight loss” in the “Other” section since the loss is >2 kg. All of the items are considered “New/Worse” since they are present within the past 28 days and no prior BVAS/WG was scored.

Notes:

- Must still fill in “none” for all the other sections
- Both RBC Casts *and* Rise in creatinine are counted but *not* hematuria since by rule, if casts are seen, only cast is scored.
- Most investigators would not score fatigue as an “Other” item since it is vague.

Total number of items (Section 16) includes 2 major items and 4 minor items for a total BVAS/WG score of 10 (3 points for each major and 1 point for each minor).

Current disease status (Section 17) should be scored at Severe Disease/Flare since the patient has one or more major items (in bold, both renal items) and the manifestations are new.

You must fill out the Physician Global Assessment. There is no right or wrong answer for this section.

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BVAS 2003-CASE I

First, consider whether items of active vasculitis have a relevant box to be ticked. The relevant abnormalities for Case I are: general: (*weight loss* $\geq 2\text{kg}$ and *arthralgia*), ENT (*bloody nasal discharge/crusts/ulcers and/or granulomata* and the cheek tenderness represents *sinus involvement*), renal (*haematuria*, and *creatinine 250 - 499*). Since this is the first visit you would score the actual creatinine value, in this example you could instead score the rise in creatinine $> 30\%$ because you have a previous creatinine value, instead of the actual creatinine, but you would not score both. In fact either score 6 and take the renal to the maximum of 12.

Second, consider whether items are new/worse or have all been present before. Here they are new/worse so the persistent box is not ticked.

The osteoarthritis and old stroke are irrelevant as are the clinical signs relating to these problems.

The scores are calculated from the Right hand, new/worse column of the glossary: General $1 + 2 = 3$, ENT $6 + 2$, but max is 6 so = 6, Renal $6 + 6 = 12$; total = $3 + 6 + 12 = 21$.

FFS-CASE I

The FFS is applicable because this patient has newly diagnosed Wegener's granulomatosis. Among the 5 items comprised in this score, the only item present is "creatininemia ≥ 1.58 mg/dl". The item "central nervous system involvement" was not taken into consideration because it appears unlikely that this patient's history of stroke is directly related to Wegener's granulomatosis. Even if we assumed that the neurological disease might correspond to an earlier manifestation of the vasculitis, this item would not be scored because the FFS only considers factors corresponding to active disease. Indeed, the neurological findings at the time of the evaluation would then be considered a sequela.

The total calculated FFS for this patient is 1.

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DEI-CASE I

The following organ systems are affected by active vasculitis and therefore need to be scored:

- **E** (ENT and upper airway): nasal crusting and bleeding with tenderness over the cheeks, nasal biopsy positive for WG.
- **K** (kidney): red cells in urinalysis with RBC cast, rise in creatinine, renal biopsy positive for WG
- **A** (arthralgias, arthritides): new onset of joint pain, swollen joints
- **B** (constitutional symptoms): fatigue, 4 kg weight loss

All other organ systems (EY, H, L, GI, P, C, S) are not affected by active vasculitis and thus are not scored. Specifically, signs and symptoms attributable to stroke are not counted since these are not related to active vasculitis.

Note, that the notes given on the DEI score sheet are not all inclusive. Thus, items not specifically listed may be scored if there is evidence that they result from active vasculitis. In this patient, this would be the case for rapid weight loss of 4 kg in 3 weeks although the note on the DEI score sheet proposes to record a weight loss of “10% of body weight” (we do not know patient's previous weight here).

This gives a total DEI score of **7**: $2 (E) + 2 (K) + 2 (A) + 1 (B) = 7$

Remember that each organ system is only scored once despite the severity and number of affected items within the respective organ systems.

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Training Case II

BVAS.1 and BVAS.2-CASE II

You were asked you to score this for the current visit but remember that all of the features occurring within the previous month count towards the current visit therefore both sets of assessment are relevant. The relevant features are: general (*fever ≤ 38.5 , malaise*), ENT (*bloody nasal discharge, nasal crusting, sinus involvement, hearing loss, ENT opinion* and the finding of *conductive deafness* and *granulomatous sinusitis*), chest (*none, chest radiology performed* and *no active vasculitis*) renal (*haematuria, proteinuria* and *hypertension*). Although not listed on the BVAS form, *aural discharge* is a relevant finding and should be added to the ‘other’ section although it does not currently score any points. All of these features are new or worse. On this occasion the chest x-ray is normal and there is no active vasculitis. You can either just tick the “none” box or tick “chest radiology performed” and “no active vasculitis” or all three.

The total BVAS.1 (new/worse) score is 20: general 2, cutaneous 0, mucous membranes 0, ENT 6, chest 0, cardiovascular 0, abdominal 0, renal 12, nervous system 0.

The BVAS.2 (grumbling) is zero.

BVAS/WG-CASE II

The appropriate items to score for Case I are: general (fever), ENT (nasal discharge, sinus involvement, conductive hearing loss), Renal (hematuria). You should also write in “aural discharge” in the “Other” section. All of the items are considered “New/Worse” since they are present within the past 28 days and no prior BVAS/WG was scored.

Notes:

- No severe manifestations.
- Must still fill in “none” for all the other sections
- Most investigators would not score “tired” as an “Other” item since it is vague.

Total number of items (Section 16) includes no major items and 5 minor items for a total BVAS/WG score of 5 (3 points for each major and 1 point for each minor).

Current disease status (Section 17) should be scored at Limited Disease/Flare since the manifestations are new but the patient has no major items.

You must fill out the Physician Global Assessment. There is no right or wrong answer for this section.

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BVAS 2003-CASE II

All features are within the previous month so should be scored. They are: general (*fever ≤ 38.5*), ENT (*bloody nasal discharge/nasal crusting/ ulcers and/or granulomata, sinus involvement, Conductive hearing loss*) renal (*haematuria, proteinuria and hypertension*). Although not listed on the BVAS form, *aural discharge* is a relevant finding and should be added to the ‘other’ section although it does not currently score any points. All of these features are new or worse so the persistent box is not ticked.

The BVAS 2003 score is General 2 = 2, ENT 6 + 2 + 3, but max is 6 so = 6, Renal 4 + 4 + 6, but max is 12 so = 12; total = 2 + 6 + 12 = 20.

FFS-CASE II

For this patient’s evaluation, the FFS is relevant with all the noted signs being attributable to newly diagnosed Wegener’s granulomatosis.

The only appropriate FFS item is “proteinuria ≥ 1 g/day”. Indeed, this item should be accorded 1 point because proteinuria of 2+ or more in a dipstick urine analysis corresponds to a proteinuria ≥ 1 g/day.

The total calculated FFS for this patient is 1.

DEI-CASE II

The following organ systems are affected by active vasculitis and therefore need to be scored:

- **E** (ENT and upper airway): new onset of hearing loss, epistaxis, nasal discharge and crusting, hearing loss, sinus discomfort, positive biopsy showing granulomata.
- **K** (kidney): hematuria, proteinuria, hypertension
- **B** (constitutional symptoms): fever

All other organ systems (EY, H, L, GI, P, C, S, A) are not affected by active vasculitis and thus are not scored. Specifically, signs and symptoms attributable to stroke are not counted since these are not related to active vasculitis.

This gives a total DEI score of **5**: 2 (E) + 2 (K) + 1 (B) = **5**

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Training Case III

BVAS.1 and BVAS.2-CASE III

This patient has had three separate assessments and we are really asking you to concentrate on the most recent assessment when he has been off treatment for several months. All of the presenting features would have been scored at the time but are not currently relevant since they do not represent current disease activity. Therefore the ischaemic fingers, splinter haemorrhages, gangrene, haematuria, red cell casts, proteinuria, inflammatory arthritis, bilateral conductive hearing loss, septal perforation and bloody nasal crusts do not count for the current assessment. This man has a lot of damage but no current disease activity. The creatinine level is stable and does not count. The gangrenous fingers have been treated with amputation; the haematuria is chronic and has not increased. The proteinuria is stable and the nasal stuffiness is damage and the general lack of energy is also not relevant.

His BVAS.1 (new/worse) score is therefore 0 and no items should be scored on this current visit. The BVAS.2 (grumbling/persistent) score is also zero.

BVAS/WG-CASE III

There are no items that are appropriate to be scored since there is no evidence of ongoing disease activity. Therefore you should fill in “none” for all the sections.

Notes:

- The hematuria is months old and the renal function is stable. We can assume that without a more active urinary sediment, this represents either renal scarring or cyclophosphamide toxicity.

Total number of items (Section 16) includes no major items and no minor items for a total BVAS/WG score of 0 (3 points for each major and 1 point for each minor).

Current disease status (Section 17) should be scored at Remission.

You must fill out the Physician Global Assessment. For this case, you should have put a mark at zero.

BVAS 2003-CASE III

There are no convincing features of active vasculitis at the current assessment. All sections should be checked as ‘none’ and do not tick the persistent box.

The BVAS 2003 score would be 0.

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FFS-CASE III

The FFS is not applicable for this patient's current assessment. Indeed, although he complains of some persistent general and ENT symptoms, his vasculitis is not newly diagnosed and he is not experiencing a clear-cut disease flare.

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DEI-CASE III

None of the features present at the current visit are likely to result from active vasculitis. Since no organ system is affected by active vasculitis the DEI score for this visit is 0.

Hematuria and proteinuria are chronic and creatinine is stable. Since we are not provided with information suggesting active glomerulonephritis (e.g. RBC casts, recent biopsy results) hematuria, proteinuria and elevated creatinine are due to damage and thus are not scored in the DEI. Since there is no evidence for further digital ischemia (possibly related to active vasculitis), pain at the amputation site is not recorded in the DEI at the present visit. All other items present several month ago (S, K, A, E) do not count for the current assessment of DEI since there are no signs of active vasculitis in these organ systems at the time of the present assessment.