Tips for bruk av BVAS og VDI i oppfølging av pasienter med vaskulitt

Wenche Koldingsnes

NorVas

HELSE NORD

NorVas

Skåring av sykdomsaktivitet og skade

- I oppfølging av pasienter med vaskulitt er vurdering og konklusjon vedr. sykdomsaktivitet helt avgjørende for riktig behandling.

- Skåring av sykdomsaktivitet er vesentlig for å besvare spørsmålene:
  - Er pasienten i remisjon?
  - Har pasienten et residiv?

- Registrering av utviklet organskade er vesentlig for monitorering av gitt behandling og for å vurdere framtidig behandling.

HELSE NORD

NorVas

Skåring av sykdomsaktivitet ved BVAS (Birmingham Vasculitis Activity Score)

- **BVAS** er et velprøvd instrument for evaluering av sykdomsaktivitet ved vaskulitter
- BVAS har vært brukt i en rekke europeiske studier i regi av EUVAS.

- Se: http://www.vasculitis.org

- EUVAS The European Vasculitis Society
  - **EUVAS** is an open collaboration of physicians interested in research and education in vasculitis.
  - **EUVAS** has representatives from many medical specialties based both inside and outside the European Union.
  - **EUVAS** conducts a range of activities including clinical trials and studies into the assessment of vasculitis.
  - **EUVAS** is a partner for interested researchers in the development of collaborative studies and offers organisational support through its office.
  - **EUVAS** has links with other vasculitis research groups:
    -

VASCULITIS CLINICAL RESEARCH CONSORTIUM

Groupe Français d'Etude des Vascularites

HELSE NORD

NorVas

BVAS 2003

Birmingham Vasculitis Activity Score (v 3) (weighting with major items are highlighted)

Patient ID: **Date of birth:** **Total score:**

Assessor: **Date of assessment**

Tick an item **only** if attributable to active vasculitis. If there are no abnormalities in a section, please tick 'None' for that organ-system.

If **all** abnormalities are due to persistent disease (active vasculitis which is not new/worse in the prior 4 weeks), tick the **PERSISTENT** box at the bottom right corner

Is this the patient's first assessment? **Yes ○** **No ○**

		P	N/W
1. General	**Max**	**2**	**3**
Myalgia		1	1
Arthralgia / arthritis		1	1
Fever ≥38° C		2	2
Weight loss ≥2 kg		2	2
2. Cutaneous	**Max**	**3**	**6**
Infarct		1	2
Purpura		1	2
Ulcer		1	4
♦Gangrene		2	6
Other skin vasculitis		1	2
3. Muc membranes/ eyes	**Max**	**3**	**6**
Mouth ulcers		1	2
Genital ulcers		1	1
Adnexal inflammation		2	4
Significant proptosis		2	4
Scleritis / Episcleritis		1	2
Conjunctivitis / Blepharitis / Keratitis		1	1
Blurred vision		2	3
Sudden visual loss		❖	6
Uveitis		2	6
♦Retinal changes (vasculitis / thrombosis / exudate / haemorrhage)		2	6
4. ENT	**Max**	**3**	**6**
Bloody nasal discharge / crusts / ulcers / granulomata		2	4
Paranasal sinus involvement		1	2
Subglottic stenosis		3	6
Conductive hearing loss		1	3
♦Sensorineural hearing loss		2	6
5. Chest	**Max**	**3**	**6**
Wheeze		1	2
Nodules or cavities		❖	3
Pleural effusion / pleurisy		2	4
Infiltrate		2	4
Endobronchial involvement		2	4
♦Massive haemoptysis / alveolar haemorrhage		4	6
♦Respiratory failure		4	6

		P	N/W
6. Cardiovascular	**Max**	**3**	**6**
Loss of pulses		1	4
Valvular heart disease		2	4
Pericarditis		1	3
♦Ischaemic cardiac pain		2	4
♦Cardiomyopathy		3	6
♦Congestive cardiac failure		3	6
7. Abdominal	**Max**	**4**	**9**
Peritonitis		3	9
Bloody diarrhoea		3	9
♦Ischaemic abdominal pain		2	6
8. Renal	**Max**	**6**	**12**
Hypertension		1	4
Proteinuria >1+		2	4
♦Haematuria ≥10 RBCs/hpf		3	6
Creatinine 125-249µmol/L(1.41-2.82mg/dl)*		❖	4
Creatinine 250-499 µmol/L(2.83-5.64mg/dl)*		❖	6
♦Creatinine ≥500 µmol/L (≥5.66mg/dl)*		❖	8
♦Rise in serum creatinine >30% or fall in creatinine clearance >25%		❖	6
***Can only be scored on the first assessment**			
9. Nervous system	**Max**	**6**	**9**
Headache		1	1
Meningitis		1	3
Organic confusion		1	3
Seizures (not hypertensive)		3	9
♦Cerebrovascular accident		3	9
♦Spinal cord lesion		3	9
♦Cranial nerve palsy		3	6
Sensory peripheral neuropathy		3	6
♦Mononeuritis multiplex		3	9
10. Other	**Max**	❖	❖
a.		❖	❖
b.		❖	❖
c.		❖	❖
d.		❖	❖

PERSISTENT DISEASE ONLY:
(Tick here if **all** the abnormalities are due to persistent disease) ☐

♦ Indicates major item ❖These items are not scored P=Persistent N/W=New or Worse

Max indicates the maximum score for each section

Maximum persistent score = 33 Maximum new/worse score = 63

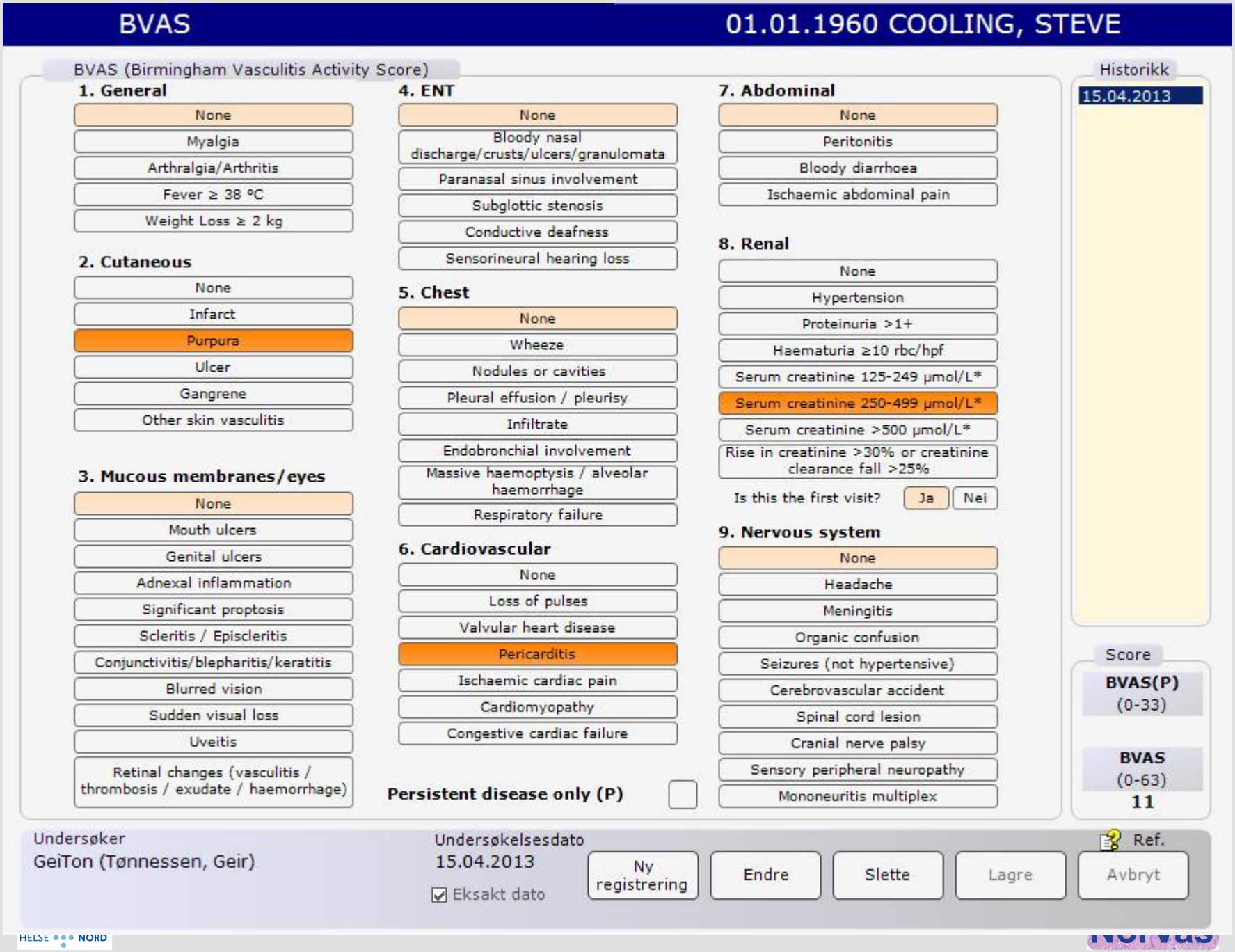
Please note, only score for persistent if all items are persistent; if any items are new/worse score all items as new/worse

References: Luqmani, RA, et al. (1994). "Birmingham Vasculitis Activity Score (BVAS) in systemic necrotizing vasculitis." QJM **87**(11):671-8.
Luqmani, RA, et al. (1997). "Disease assessment and management of the vasculitides." Baillieres Clin Rheumatol **11**(2): 423-46.Mukhtyar C. et al (2009). "Modification and validation of the Birmingham Vasculitis Activity Score (version 3) ARD 68:1827

HELSE NORD

NorVas

BVAS i GTI


BVAS
01.01.1960 COOLING, STEVE
BVAS (Birmingham Vasculitis Activity Score)
1. General
None
Myalgia
Arthralgia/Arthritis
Fever ≥ 38 °C
Weight Loss ≥ 2 kg
2. Cutaneous
None
Infarct
Purpura
Ulcer
Gangrene
Other skin vasculitis
3. Mucous membranes/eyes
None
Mouth ulcers
Genital ulcers
Adnexal inflammation
Significant proptosis
Scleritis / Episcleritis
Conjunctivitis/blepharitis/keratitis
Blurred vision
Sudden visual loss
Uveitis
Retinal changes (vasculitis / thrombosis / exudate / haemorrhage)
4. ENT
None
Bloody nasal discharge/crusts/ulcers/granulomata
Paranasal sinus involvement
Subglottic stenosis
Conductive deafness
Sensorineural hearing loss
5. Chest
None
Wheeze
Nodules or cavities
Pleural effusion / pleurisy
Infiltrate
Endobronchial involvement
Massive haemoptysis / alveolar haemorrhage
Respiratory failure
6. Cardiovascular
None
Loss of pulses
Valvular heart disease
Pericarditis
Ischaemic cardiac pain
Cardiomyopathy
Congestive cardiac failure
Persistent disease only (P)
7. Abdominal
None
Peritonitis
Bloody diarrhoea
Ischaemic abdominal pain
8. Renal
None
Hypertension
Proteinuria >1+
Haematuria ≥10 rbc/hpf
Serum creatinine 125-249 µmol/L*
Serum creatinine 250-499 µmol/L*
Serum creatinine >500 µmol/L*
Rise in creatinine >30% or creatinine clearance fall >25%
Is this the first visit?
Ja
Nei
9. Nervous system
None
Headache
Meningitis
Organic confusion
Seizures (not hypertensive)
Cerebrovascular accident
Spinal cord lesion
Cranial nerve palsy
Sensory peripheral neuropathy
Mononeuritis multiplex
Historikk
15.04.2013
Score
BVAS(P)
(0-33)
BVAS
(0-63)
11
Undersøker
GeiTon (Tønnessen, Geir)
Undersøkelsesdato
15.04.2013
Eksakt dato
Ref.
Ny registrering
Endre
Slette
Lagre
Avbryt
HELSE NORD

BVAS 2003

- Skår kun symptomer/funn som skyldes aktiv vaskulitt (max 63)
  - Ekskluder andre årsaker
    - Infeksjoner, hypertensjon, allergi, etc
    - Varig skade av tidligere aktiv vaskulitt
- "Persistent disease" (max 33)
  - kun når all patologi er kronisk og skyldes aktiv vaskulitt
  - Dvs. intet er nytt eller forverret
- Det kan være nødvendig å avvente før endelig skåring (gjør notater i tekstfeltet i BVAS skjema)
  - spesialundersøkelser
  - utvikling av symptomer

Skill mellom aktiv vaskulitt og skade
BVAS vs VDI (Vasculitis Damage Index)

- BVAS
  - All skåring gjelder aktiv vaskulitt
- VDI
  - "Non-healing scars", oppstått etter debut av sd
  - Skal ha stått i min 3 mndr
    - Eksl: hjerteinfarkt, operasjoner og lignende (skåres etter 3 mndr)
    - ikke bare vaskulitt relatert skade
      - Behandlingsrelatert skade
      - Andre interkurrente skader etter diagnosen vaskulitt
    - Skår forandringer som har vart i 3 mndr selv om de så går tilbake (hudulcera)
- Eksempel:
  - Skoper og tetthet i nese
    - Kan skyldes skadde slimhinner, uten aktiv vaskulitt
  - Nevropati,
    - gir ofte langvarige symptomer, som skyldes den initiale skade

VDI

VASCULITIS DAMAGE INDEX (VDI)

This is for recording organ damage that has accurred in patients *since the onset of vasculitis*
Patients often have co-morbidity before they develop vasculitis, **which must not be scored**
Record features of active disease using the Birmingham Vasculitis Activity Score (BVAS)
A new patient should ***ususlly have a VDI score of zero,*** unless:
(a) they have had vasculitis for more than three months of onset of disease. **and**
(b) the damage has developed or become worse sinse the onset of vasculitis

1. Musculoskeletal	**No**	**Yes**
None	☐	
Significant muscle atrophy or weakness		○
Deforming/erosive arthritis		○
Osteoporosis/vertebral collapse		○
Avascular necrosis		○
Osteomyelitis		○
2. Skin/Mucous membranes		
None	☐	
Alopecia		○
Cutaneous ulcers		○
Mouth ulcers		○
3. Ocular		
None	☐	
Cataract		○
Retinal change		○
Optic atrophy		○
Visual impairment/diplopia		○
Blindness in one eye		○
Blindness in second eye		○
Orbital wall destruction		○
4. ENT		
None	☐	
Hearing loss		○
Nasal blockage/chronic discharge/crusting		○
Nasal bridge collapse/septal perforation		○
Chronic sinusitis/radiological damage		○
Subglottic stenosis (no surgery)		○
Subglottic stenosis (with surgery)		○
5. Pulmonary		
None	☐	
Pulmonary hypertension		○
Pulmonary fibrosis		○
Pulmonary infarction		○
Pleural fibrosis		○
Chronic asthma		○
Chronic breathlessness		○
Impaired lung function		○
6. Cardiovascular		
None	☐	
Angina/angioplasty		○
Myocardial infarction		○
Subsequent myocardial infarction		○
Cardiomyopathy		○
Valvular disease		○
Pericarditis≥3 months or pericardiectomy		○
Diastolic BP≥95 or requiring antihypertensive		○

Name
Trial Number
Date
Centre

7. Peripheral vascular disease	**No**	**Yes**
None	☐	
Absent pulses in one limb		○
2nd episode of absent pulses in one limb		○
Major vessel stenosis		○
Claudication>3 months		○
Minor tissue loss		○
Major tissue loss		○
Subsequent major tissue loss		○
Complicated venous thrombosis		○
8. Gastrointestinal		
None	☐	
Gut infarction/resection		○
Mesenteric insufficiency/pancreatitis		○
Chronic peritonitis		○
Oesophageal stricture/surgery		○
9. Renal		
None	☐	
Estimated/measured GFR≤50%		○
Proteinuria≥0.5 g/24 h		○
End stage renal disease		○
10. Neuropsychiatric		
None	☐	
Cognitive impairment		○
Major psychosis		○
Seizures		○
Cerebrovascular accident		○
2nd cerebrovascular accident		○
Cranial nerve lesion		○
Peripheral neuropathy		○
Transverse myelitis		○
11. Other		
None	☐	
Gonadal failure		○
Marrow failure		○
Diabetes		○
Chemical cystitis		○
Malignancy		○
Other		○

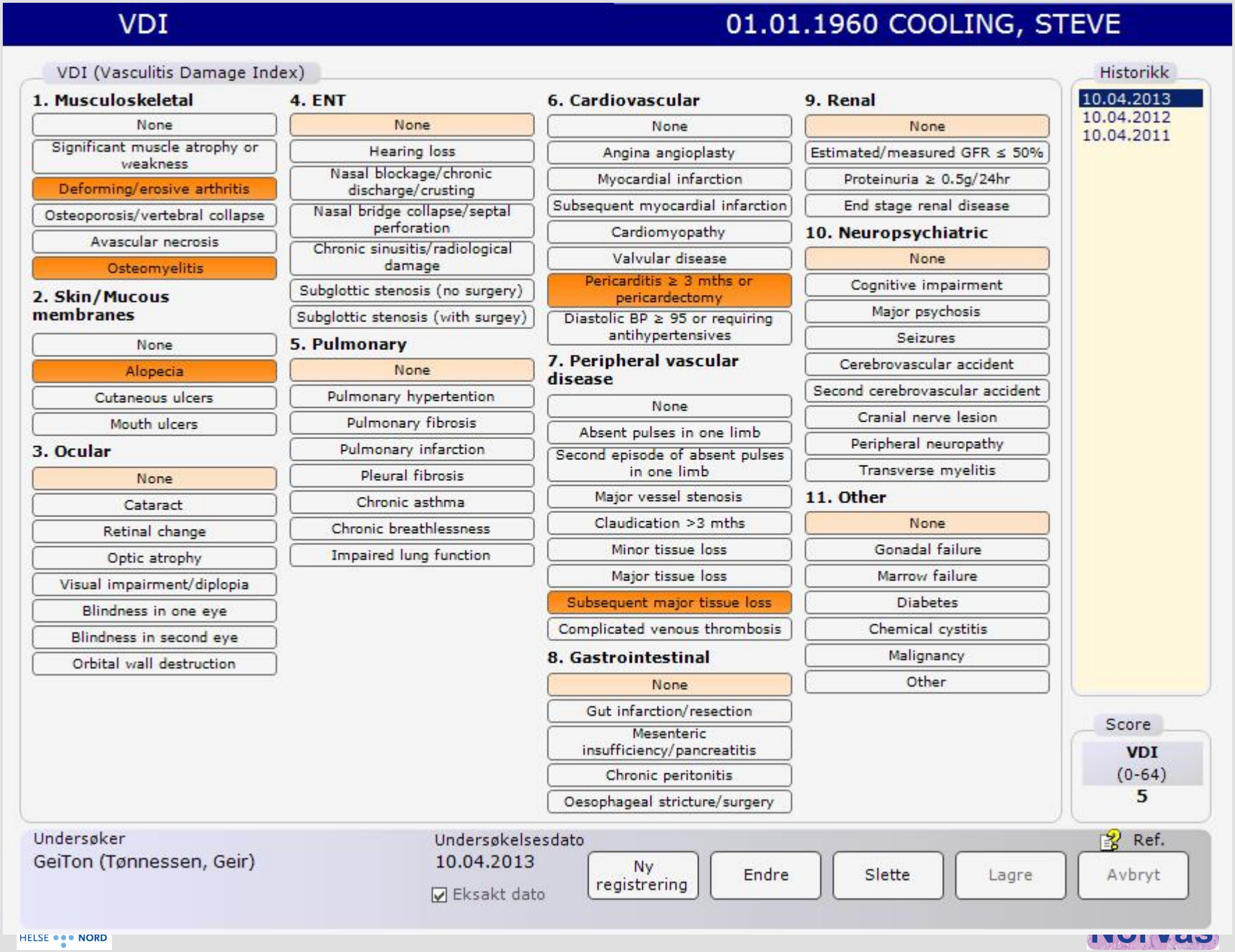
Total VDI score. Record the number of positive items (1 point for each). The VDI score can either increase or remain the same over time. Remember to carry forward any previous items of damage. ☐

VDI Modified from Exley AR, Bacon PA, Luqmani et al (1997) Development and initial validation of the VDI, Arthritis Rheum 40 : 371–380

HELSE NORD

NorVas

VDI i GTI


VDI
01.01.1960 COOLING, STEVE
VDI (Vasculitis Damage Index)
1. Musculoskeletal
None
Significant muscle atrophy or weakness
Deforming/erosive arthritis
Osteoporosis/vertebral collapse
Avascular necrosis
Osteomyelitis
2. Skin/Mucous membranes
None
Alopecia
Cutaneous ulcers
Mouth ulcers
3. Ocular
None
Cataract
Retinal change
Optic atrophy
Visual impairment/diplopia
Blindness in one eye
Blindness in second eye
Orbital wall destruction
4. ENT
None
Hearing loss
Nasal blockage/chronic discharge/crusting
Nasal bridge collapse/septal perforation
Chronic sinusitis/radiological damage
Subglottic stenosis (no surgery)
Subglottic stenosis (with surgey)
5. Pulmonary
None
Pulmonary hypertention
Pulmonary fibrosis
Pulmonary infarction
Pleural fibrosis
Chronic asthma
Chronic breathlessness
Impaired lung function
6. Cardiovascular
None
Angina angioplasty
Myocardial infarction
Subsequent myocardial infarction
Cardiomyopathy
Valvular disease
Pericarditis ≥ 3 mths or pericardectomy
Diastolic BP ≥ 95 or requiring antihypertensives
7. Peripheral vascular disease
None
Absent pulses in one limb
Second episode of absent pulses in one limb
Major vessel stenosis
Claudication >3 mths
Minor tissue loss
Major tissue loss
Subsequent major tissue loss
Complicated venous thrombosis
8. Gastrointestinal
None
Gut infarction/resection
Mesenteric insufficiency/pancreatitis
Chronic peritonitis
Oesophageal stricture/surgery
9. Renal
None
Estimated/measured GFR ≤ 50%
Proteinuria ≥ 0.5g/24hr
End stage renal disease
10. Neuropsychiatric
None
Cognitive impairment
Major psychosis
Seizures
Cerebrovascular accident
Second cerebrovascular accident
Cranial nerve lesion
Peripheral neuropathy
Transverse myelitis
11. Other
None
Gonadal failure
Marrow failure
Diabetes
Chemical cystitis
Malignancy
Other
Historikk
10.04.2013
10.04.2012
10.04.2011
Score
VDI
(0-64)
5
Undersøker
GeiTon (Tønnessen, Geir)
Undersøkelsesdato
10.04.2013
Eksakt dato
Ny registrering
Endre
Slette
Lagre
Avbryt
Ref.
HELSE NORD

BVAS

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

- If all the abnormalities recorded represent smouldering/low grade/grumbling disease, and there are no new/worse features, please remember to tick the box for "persistent disease"

HELSE NORD

NorVas

BVAS "new or worse" versus "persistent"

- If you score the items as New/Worse there should be **an intention to treat or to act on at least one of the ticked items.**

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

BVAS v/debut vs v/kontroll

- **Scoring a new patient presenting for the first time**
  - Score all items that are present and attributable to vasculitis as **New/ Worse.**
  - In this case the items can have been present for over 3 months or even longer.
- **Scoring a patient for review with known disease**
- New/Worse :
  - If any item is newly present or worse, all the items will be scored as **New/Worse** and the "persistent disease only box" **is not** ticked
- Persistent disease only :
  - If **all** the ticked items have been present **within** the last 3 months and are not new or worse, the "Persistent disease only box" is ticked and the calculated score for BVAS will contain only values representing persistent disease.
- The **none** box :
  - If none of the items are present,
  - an item has been present for **over** 3 months,
  - a patient reports having had a symptom but it is not present at the time of scoring on questioning or examination, ➔ the **none** box is ticked

Remission / Relapse

- Remission
  - BVAS = 0
- Relapse
  - after remission for ≥ 3 months
- Major relapse
  - Recurrence or first appearance of ≥ 1 BVAS item indicating treathened vital organ function due to active vasculitis (se BVAS-liste)
- Minor relapse
  - Recurrence or first appearance of ≥ 1 (3) other BVAS items related to non-vital organs

BVAS med markering av "major items" som representerer "Major Relapse"

Birmingham Vasculitis Activity Score (v 3) (weighting with major items are highlighted)

Patient ID: **Date of birth:** **Total score:**

Assessor: **Date of assessment**

Tick an item **only** if attributable to active vasculitis. If there are no abnormalities in a section, please tick 'None' for that organ-system.

If **all** abnormalities are due to persistent disease (active vasculitis which is not new/worse in the prior 4 weeks), tick the **PERSISTENT** box at the bottom right corner

Is this the patient's first assessment? **Yes ○** **No ○**

	P	N/W
1. General **Max**	**2**	**3**
Myalgia	1	1
Arthralgia / arthritis	1	1
Fever ≥38° C	2	2
Weight loss ≥2 kg	2	2
2. Cutaneous **Max**	**3**	**6**
Infarct	1	2
Purpura	1	2
Ulcer	1	4
♦**Gangrene**	2	6
Other skin vasculitis	1	2
3. Muc membranes/ eyes **Max**	**3**	**6**
Mouth ulcers	1	2
Genital ulcers	1	1
Adnexal inflammation	2	4
Significant proptosis	2	4
Scleritis / Episcleritis	1	2
Conjunctivitis / Blepharitis / Keratitis	1	1
Blurred vision	2	3
Sudden visual loss	❖	6
Uveitis	2	6
♦**Retinal changes (vasculitis / thrombosis / exudate / haemorrhage)**	2	6
4. ENT **Max**	**3**	**6**
Bloody nasal discharge / crusts / ulcers / granulomata	2	4
Paranasal sinus involvement	1	2
Subglottic stenosis	3	6
Conductive hearing loss	1	3
♦**Sensorineural hearing loss**	2	6
5. Chest **Max**	**3**	**6**
Wheeze	1	2
Nodules or cavities	❖	3
Pleural effusion / pleurisy	2	4
Infiltrate	2	4
Endobronchial involvement	2	4
♦**Massive haemoptysis / alveolar haemorrhage**	4	6
♦**Respiratory failure**	4	6

	P	N/W
6. Cardiovascular **Max**	**3**	**6**
Loss of pulses	1	4
Valvular heart disease	2	4
Pericarditis	1	3
♦**Ischaemic cardiac pain**	2	4
♦**Cardiomyopathy**	3	6
♦**Congestive cardiac failure**	3	6
7. Abdominal **Max**	**4**	**9**
Peritonitis	3	9
Bloody diarrhoea	3	9
♦**Ischaemic abdominal pain**	2	6
8. Renal **Max**	**6**	**12**
Hypertension	1	4
Proteinuria >1+	2	4
♦**Haematuria ≥10 RBCs/hpf**	3	6
Creatinine 125-249µmol/L(1.41-2.82mg/dl)*	❖	4
Creatinine 250-499 µmol/L(2.83-5.64mg/dl)*	❖	6
♦**Creatinine ≥500 µmol/L (≥5.66mg/dl)***	❖	8
♦**Rise in serum creatinine >30% or fall in creatinine clearance >25%**	❖	6
***Can only be scored on the first assessment**		
9. Nervous system **Max**	**6**	**9**
Headache	1	1
Meningitis	1	3
Organic confusion	1	3
Seizures (not hypertensive)	3	9
♦**Cerebrovascular accident**	3	9
♦**Spinal cord lesion**	3	9
♦**Cranial nerve palsy**	3	6
Sensory peripheral neuropathy	3	6
♦**Mononeuritis multiplex**	3	9
10. Other **Max**	❖	❖
a.	❖	❖
b.	❖	❖
c.	❖	❖
d.	❖	❖
PERSISTENT DISEASE ONLY: (Tick here if **all** the abnormalities are due to persistent disease)	☐	

♦ **Indicates major item** ❖**These items are not scored** **P=Persistent N/W=New or Worse**

HELSE NORD