VALORACIÓN DE LA ACTIVIDAD EN VASCULITIS ASOCIADAS A ANCA

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Medicina Interna
Hospital Universitari de Bellvitge

Mortalidad

Crónicas
Recidivantes

Fauci AS. Effects of cyclophosphamide upon the immune response in Wegener's granulomatosis. **NEJM 1971;** 285:1494-1496
Fauci AS. Wegener's granulomatosis: prospective clinical and therapeutic experience with 85 patients for 21 years. **Ann Intern Med. 1983** Jan;98(1):76-85
Gayraud M. Long-term followup of polyarteritis nodosa, microscopic polyangiitis, and Churg-Strauss syndrome:. **Arthritis Rheum. 2001** Mar;44(3):666-75

HERRAMIENTAS

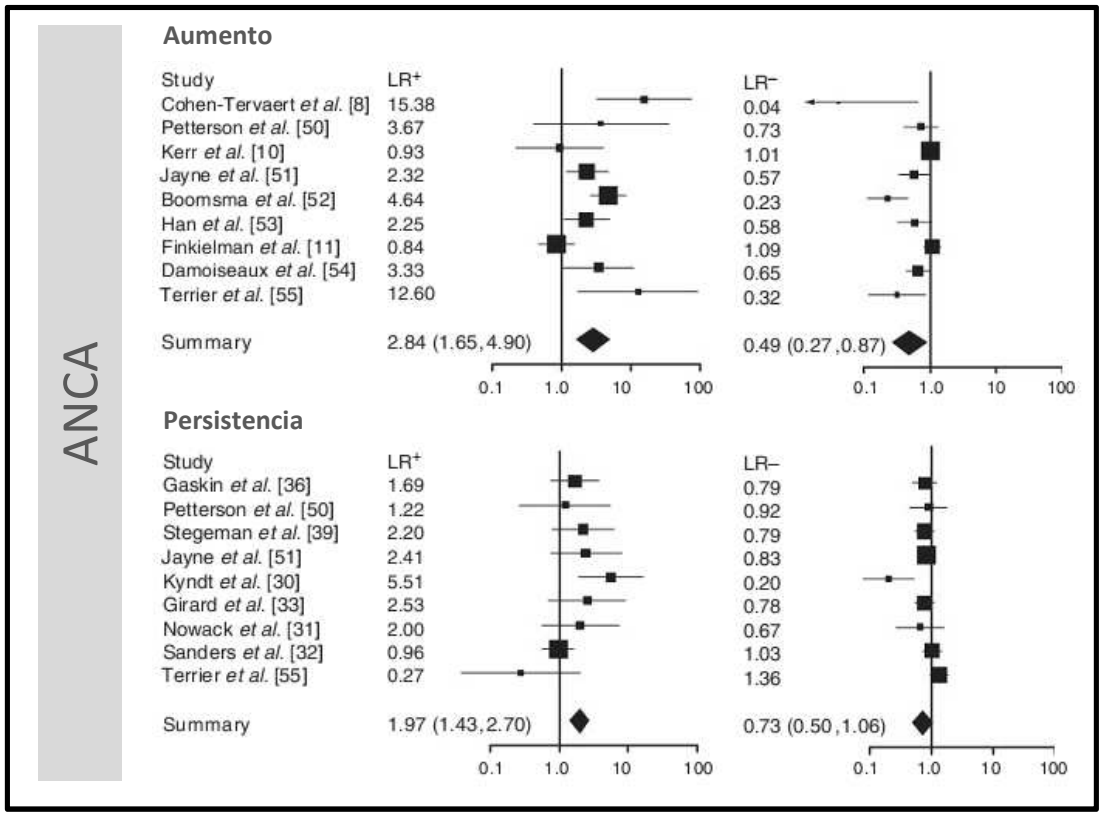
valoración de **aspectos** de las vasculitis relacionados con la **morbilidad**

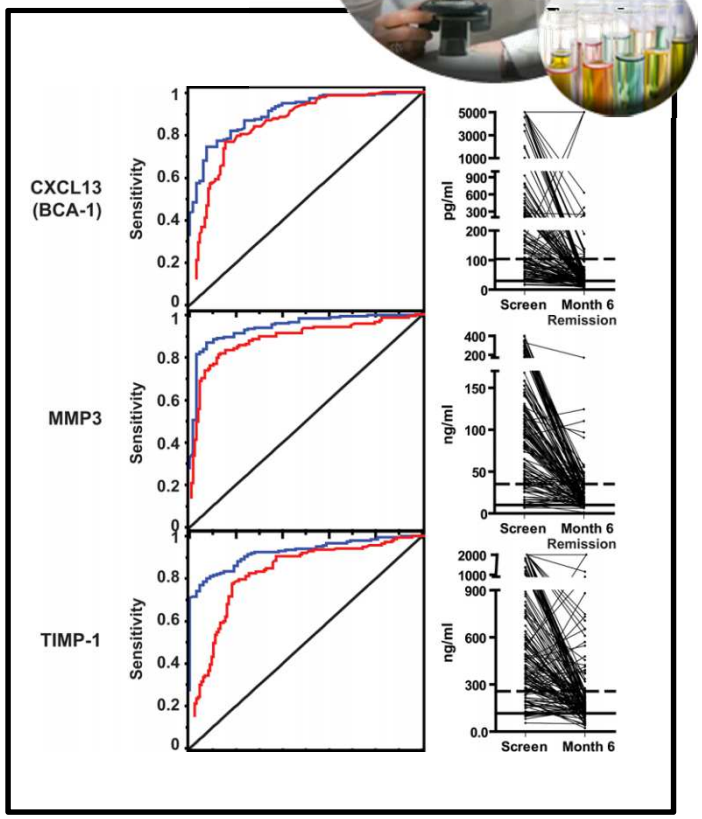
Merkel PA. Current Status of Outcome Measure Development in Vasculitis. J Rheumatol 2014;41;593-598


¿CÓMO
SE MIDE LA
ACTIVIDAD?

¿CÓMO MEDIR LA ACTIVIDAD?

BIOMARCADORES





Van der Woude. Autoantibodies against neutrophils and monocytes. **Lancet. 1985** Feb 23;1(8426):425-9

Nölle B. Anticytoplasmic autoantibodies: their immunodiagnostic value in Wegener granulomatosis. **Ann Intern Med. 1989** Jul 1;111(1):28-40

Tomasson G. Value of ANCA measurements during remission to predict a relapse of ANCA-associated vasculitis-a meta-analysis. **Rheumatology 2012** 51:100-9

Monach PA. Serum proteins reflecting inflammation, injury and repair as biomarkers of disease activity in ANCA-associated vasculitis. **Ann Rheum Dis. 2013**


TEST

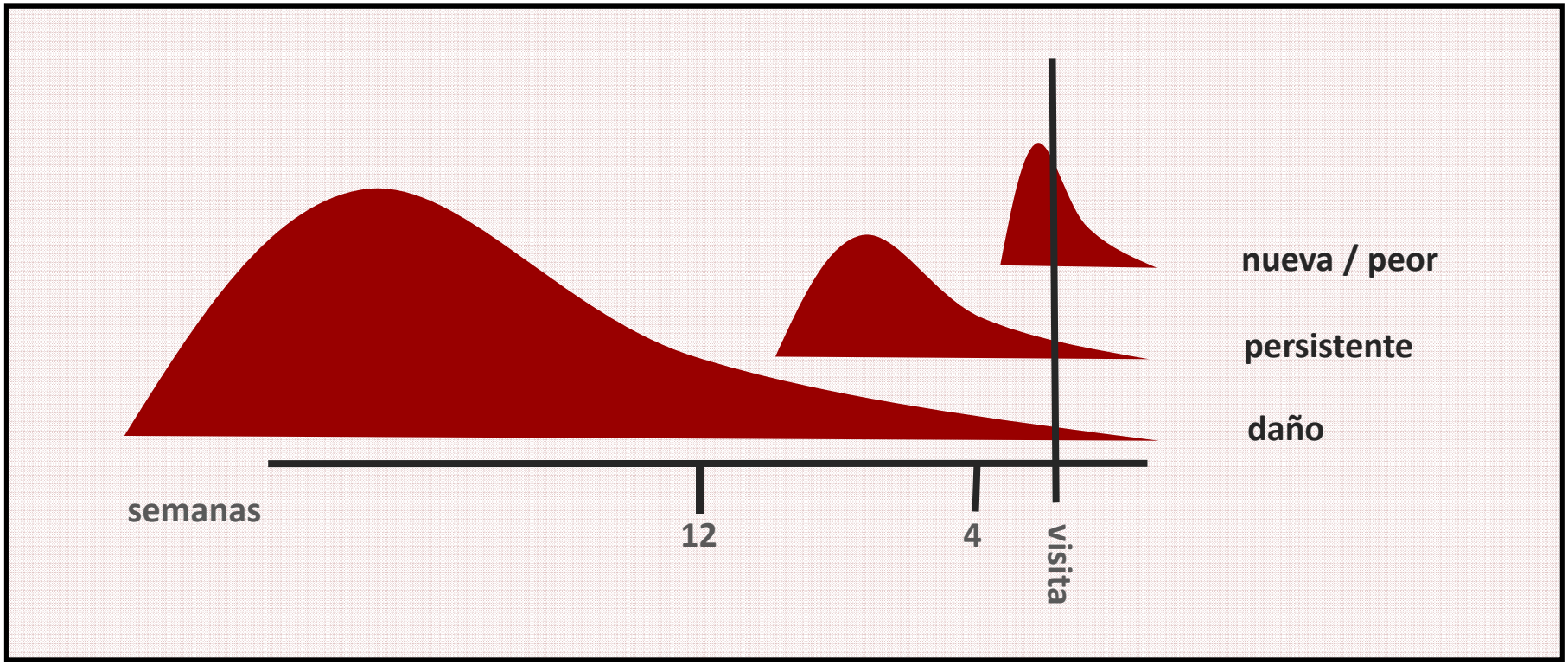
¿Qué significa **ACTIVIDAD**?

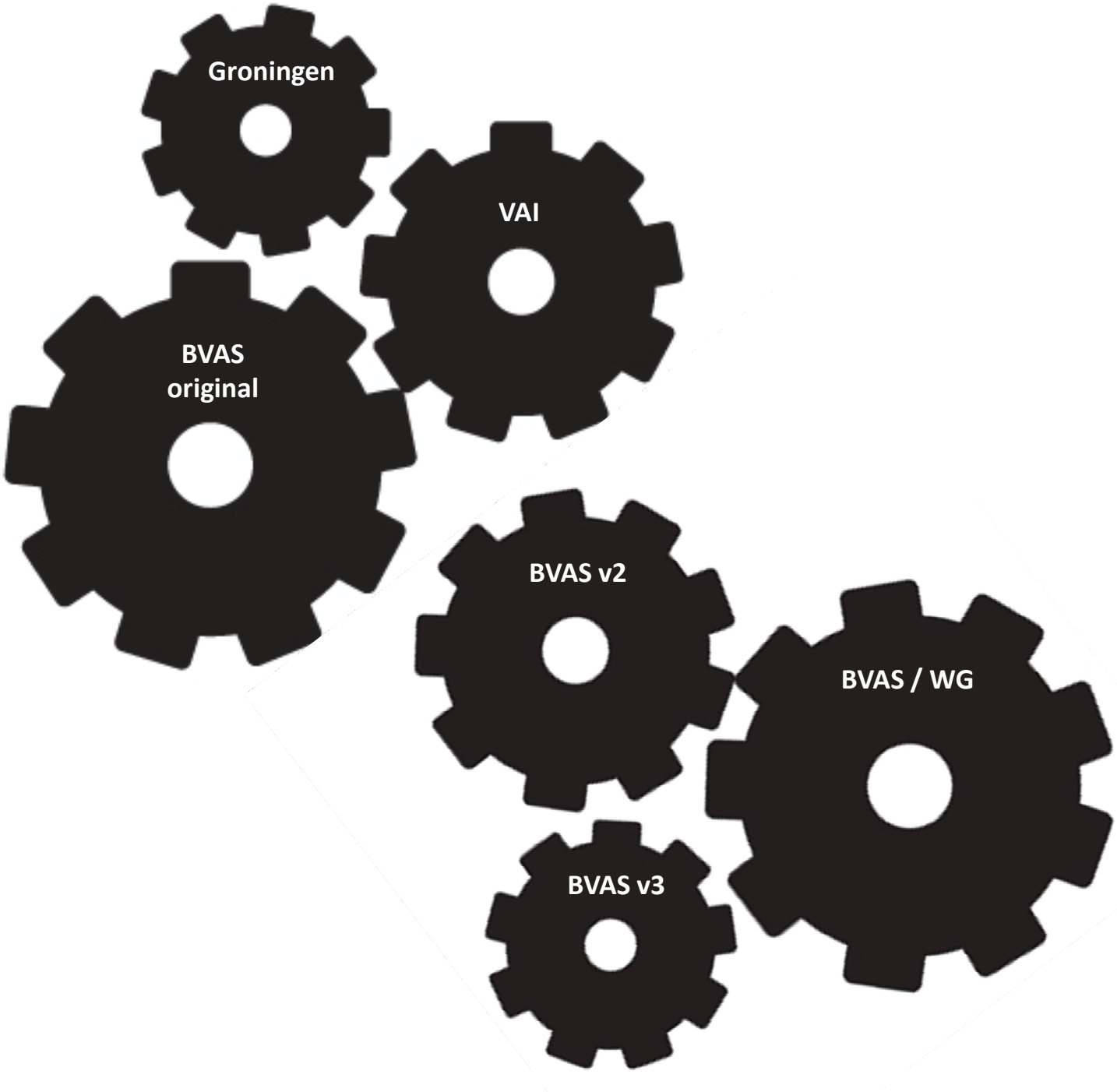
Manifestaciones **secundarias a la vasculitis**
activas (en el diagnóstico se recogen todas)
y de **intensidad suficiente** (intención de tratar y/o mayor monitorización)

NUEVA/PEOR	**PERSISTENTES**
durante las últimas **4 semanas**	que empezaron/empeoraron hace **> 4ª y < 12ª semanas** las últimas 4 semanas están **activas** pero i**gual o mejor**

¿y DAÑO?

Manifestación de **cualquier causa** (vasculitis, tratamiento…)
a partir de debut de la vasculitis (acumulativo)
han estado o están activas durante **> 12 semanas** (aunque sea curable)


nueva / peor
persistente
daño
semanas
12
4
visita


Groningen
VAI
BVAS
original
BVAS v2
BVAS / WG
BVAS v3

GRONINGEN INDEX

Manifestaciones secundarias a **GPA**, **nuevas** y de menos de **12 semanas**

Exacerbación **mayor** → expertos → Prednisona 1mg/kg/d + CYC vo 2mg/kg/d

- decrease of creatinine clearance of more than 30% within a period of 3 months or less in combination with erythrocyturia
- pulmonary infiltrates on chest X-ray in combination with dyspnea and rising CRP
- cerebral vasculitis
- motor and sensory nerve disturbances with rising CRP
- cranial nerve palsy with rising CRP
- orbital pseudotumor
- necrotizing scleritis
- tracheal stenosis with severe dyspnea
- myocardial infarction due to vasculitis
- acute abdomen or massive gastroinestinal hemorrhage due to vasculitis
- progressive disease despite treatment of minor relapses with prednisolone 30 mg and cylophosphamide 75 mg for at least 2 weeks.

Exacerbación **minor** → Score >5 → Prednisona 0.5mg/kg/d + CYC vo 75mg/d

1. ENT-symptoms
   - clinical manifestation, e.g. sinusitis, sanguinous nasal discharge, with
   a. an appearance of nasal ulceration and/or proliferative mass at endoscopy, and
   b. a histologic pattern of granulomatous inflammation at biopsy — 5 points
   - idem, but a histologic pattern of necrotizing inflammation without granulomas — 3 points
   - serous otitis and/or otorrhea — 2 points
   - buccal ulceration — 1 points
   - tracheal stenosis without severe dyspnea — 3 points
   - tracheal stenosis with severe dyspnea — 8 points
2. Pulmonary symptoms
   - pulmonary infiltrates (chest X-ray) — 4 points
   - impending respiratory insufficiency with pulmonary infiltrates (chest X-ray) with or without hemoptysis — 10 points
3. Renal symptoms
   - more than 15 erythrocytes/h.p.f. on more than one occasion in urinary sediment — 2 points
   - cellular casts on more than one occasion in urinary sediment — 3 points
   - decrease of creatinine clearance of less than 30% within a period of 3 months in combination with eryhrocyturia — 4 points
   - decrease of creatinine clearance of more than 30% within a period of 3 months in combination with erythrocyturia — 10 points
4. (Central) nervous system and eyes
   - cerebral vasculitis — 10 points
   - mononeuritis multiplex including paresis and sensory loss — 8 points
   - mononeuritis multiplex without paresis — 3 points
   - cranial nerve palsy — 8 points
   - episcleritis or corneal ulceration — 2 points
   - necrotizing scleritis — 8 points
   - orbital pseudotumor — 8 points
   - recurrent dacryocystitis — 1 points
5. Cardiovascular
   - mycardial infarction due to vasculitis — 10 points
   - peripheral gangrene — 10 points
6. Gastro-intestinal
   - acute abdomen (intestinal perforation) due to vasculitis — 10 points
   - massive hemorrhage due to vasculitis — 10 points
7. Muskuloskeletal
   - arthralgia (one or more joints) — 2 points
   - arthritis (one or more joints) — 3 points
8. Skin
   - ulceration — 2 points
   - purpura — 1 points
   - ulceration and/or purpura with histologically granulomatous inflammation and/or necrotizing vasculitis — 3 points
9. Miscellaneous
   - any manifestation with histologically granulomatous inflammation and/or necrotizing vasculitis at biopsy — 3 points
   - fever of unknown origin of more than one weeks duration
     - ≤ 38.5 C and > 38.0 C rectally — 1 points
     - > 38.5 C rectally — 3 points
   - increase in CRP-levels
     - > 20 mg/l — 1 points
     - > 50 mg/l — 2 points
     - > 100 mg/l — 3 points

Score máximo 137

Kallenberg CG. Criteria for disease activity in Wegener's granulomatosis: a requirement for longitudinal clinical studies. **APMIS Suppl 1990**;19:37–9

Características	Groningen Kallenberg 1990	VAI Olsen 1992 Whiting-O'Keefe 1999	BVAS BVAS v2 Luqmani 1994 / 1997	BVAS/WG Stone 2001	BVAS v3 Mukhtyar 2009	BVAS v3 Suppiah 2011
Tipo vasculitis	GPA	Múltiples	Múltiples	GPA	Múltiples (no Horton)	Múltiples
Nº pacientes	¿?	74	213	117	313	238
Face validity	Si	Si	Si	Si	Si	Si
Feasybility	Biopsias	Si	Si	Si	Si	Si
Reliability **- Inter** (reproducible) **- Intra** (repetible)	¿?	ICC=0.45	Si -	ICC = 0.97 ICC = 0.62	ICC = 0.96 ICC = 0.96	ICC=0.99
Construct validity **- PGA** **- VAI** **- Otros indices** **- PCR** **- Decisión tto**	¿? - - - - -	R=0.92 - - - -	T=0.35 T=0.56 T=0.29 (Gron.) ¿? -	R=0.92 - - - -	R=0.91 R=0.88 R=0.94(s1) 0.6 (s2) R=0.43 R=0.66	R=0.85 R=0.82 VDI= -0.1 R=0.18 R=0.54
Sensibilidad al cambio de estadio	-	Si	Si	Si	Si	-

Kallenberg CG. Criteria for disease activity in Wegener's granulomatosis: a requirement for longitudinal clinical studies. **APMIS Suppl 1990**;19:37–9

Whiting-O'Keefe. Validity of a vasculitis activity index for systemic necrotizing vasculitis. Arthritis Rheum **1999**;42:2365-71

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Stone JH. A disease-specific activity index for Wegener's granulomatosis: modification of the Birmingham Vasculitis Activity Score. **Arthritis Rheum 2001**;44:912–20

Mukhtyar C. Modification and validation of the Birmingham vasculitis activity score (version 3). Ann Rheum Dis **2009**;68:1827-32

Suppiah R. A cross-sectional study of the Birmingham Vasculitis Activity Score version 3 in systemic vasculitis. Rheumatology (Oxford). **2011** May;50(5):899-905

VASCULITIS ACTIVITY INDEX

secundarias a **vasculitis**
nuevas / peor

últimas 4 semanas

8 órganos
Ponderación subjetiva

Olsen TL. Validity and precision of a vasculitis activity index (VAI). Arthritis and Rheumatism **1992**;35: S164.

Whiting-O'Keefe. Validity of a vasculitis activity index for systemic necrotizing vasculitis. Arthritis Rheum **1999**;42:2365-71.

Global assessment: 0 1 2 3 4 [ ]

Direct organ system measures: For each organ system, assess the degree of ongoing vasculitis and tissue damage. Preexistent (i.e., older than 4 weeks) damage should not be attributed to active vasculitis and should not be scored. Similarly, clinical findings known or suspected to be due to another disease process, e.g., infection, should not be counted. See attached guidelines for additional details.

	(Absent) 0 1 2 3 4 (Max)		Maximum Value Guidelines
Cutaneous	0 1 2 3 4	[ ]	Ulcerations or ≥10 lesions of palpable purpura.
Peripheral neurologic	0 1 2 3 4	[ ]	Mononeuritis in ≥4 nerves.
Central nervous system	0 1 2 3 4	[ ]	Stroke, cord lesion, ongoing seizures, or meningitis.
Renal	0 1 2 3 4	[ ]	RBC casts, glomerulonephritis on biopsy, or renal microaneurysms.
Pulmonary	0 1 2 3 4	[ ]	Pulmonary infarction, multiple lesions on chest x-ray, hypoxia, or respiratory failure.
Cardiac	0 1 2 3 4	[ ]	Cardiomyopathy, myocardial infarction, or pericarditis requiring prednisone.
Abdominal vasculitis	0 1 2 3 4	[ ]	Mesenteric infarction, microaneurysms, or GI ischemia.
ENT	0 1 2 3 4	[ ]	Sinusitis, mastoiditis, or otitis media requiring hospitalization.
Other	0 1 2 3 4	[ ]	Disease that poses an immediate threat to organ function.

[ ] (Sum of direct measures) / [ ] (Count of direct measures) = [ ] **A**

0.4 × [ ] (Count of direct measures) = [ ] **B**

Indirect measures: (Include only observations made within the last four weeks.)

Fatigue/malaise/arthralgias 0 1 2 3 4 [ ]

Westergren ESR: _____ [ ]

(0 = 0–25, 1 = 26–50, 2 = 51–75, 3 = 76–100, 4 = >100) [ ]

Fevers greater than 38°C: (0 = absent, 4 = present) [ ]

[ ] (Sum of indirect measures) / [ ] (Count of indirect measures) = [ ] **C**

VAI =

Patient Name: _____

Patient ID Number: _____

Date: _____ Observer: _____

Diagnosis: _____

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Reliability **- Inter** (reproducible) **- Intra** (repetible)	¿?	ICC=0.45	Si -	ICC = 0.97 ICC = 0.62	ICC = 0.96 ICC = 0.96	ICC=0.99
Construct validity **- PGA** **- VAI** **- Otros indices** **- PCR** **- Decisión tto**	 - - - - -	 R=0.92 - - - -	 T=0.35 T=0.56 T=0.29 (Gron.) ¿? -	 R=0.92 - - - -	 R=0.91 R=0.88 R=0.94(s1) 0.6 (s2) R=0.43 R=0.66	 R=0.85 R=0.82 VDI= -0.1 R=0.18 R=0.54
Sensibilidad al cambio de estadio	-	Si	Si	Si	Si	-

Kallenberg CG. Criteria for disease activity in Wegener's granulomatosis: a requirement for longitudinal clinical studies. **APMIS Suppl 1990**;19:37–9

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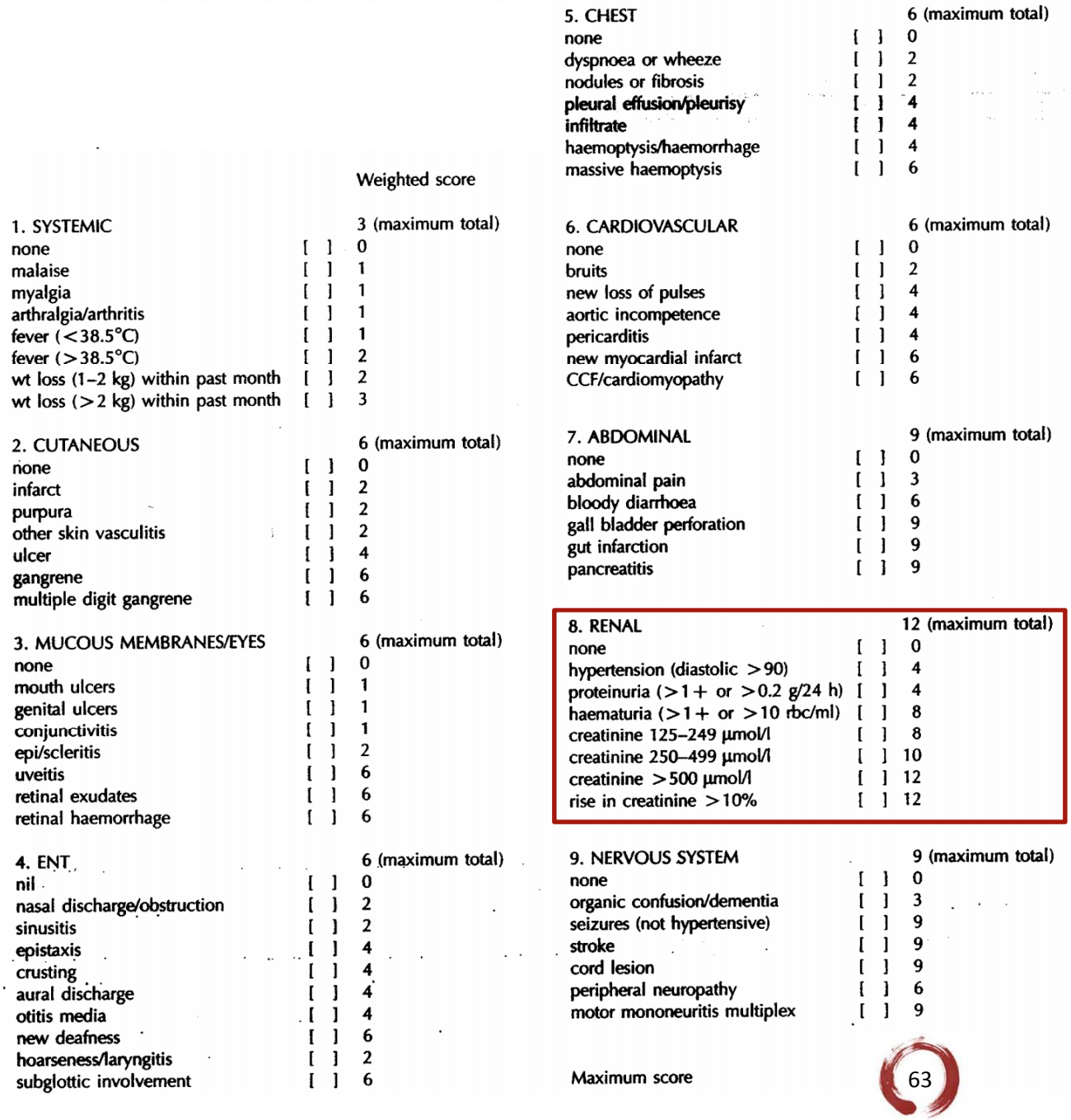
BVAS original

secundarias a vasculitis
nuevas / peor
últimas 4 semanas

59 ítems
(9 órganos)

Todos ponderados según expertos



Weighted score

1. SYSTEMIC		3 (maximum total)
none	[ ]	0
malaise	[ ]	1
myalgia	[ ]	1
arthralgia/arthritis	[ ]	1
fever (<38.5°C)	[ ]	1
fever (>38.5°C)	[ ]	2
wt loss (1–2 kg) within past month	[ ]	2
wt loss (>2 kg) within past month	[ ]	3

2. CUTANEOUS		6 (maximum total)
none	[ ]	0
infarct	[ ]	2
purpura	[ ]	2
other skin vasculitis	[ ]	2
ulcer	[ ]	4
gangrene	[ ]	6
multiple digit gangrene	[ ]	6

3. MUCOUS MEMBRANES/EYES		6 (maximum total)
none	[ ]	0
mouth ulcers	[ ]	1
genital ulcers	[ ]	1
conjunctivitis	[ ]	1
epi/scleritis	[ ]	2
uveitis	[ ]	6
retinal exudates	[ ]	6
retinal haemorrhage	[ ]	6

4. ENT		6 (maximum total)
nil	[ ]	0
nasal discharge/obstruction	[ ]	2
sinusitis	[ ]	2
epistaxis	[ ]	4
crusting	[ ]	4
aural discharge	[ ]	4
otitis media	[ ]	4
new deafness	[ ]	6
hoarseness/laryngitis	[ ]	2
subglottic involvement	[ ]	6

5. CHEST		6 (maximum total)
none	[ ]	0
dyspnoea or wheeze	[ ]	2
nodules or fibrosis	[ ]	2
pleural effusion/pleurisy	[ ]	4
infiltrate	[ ]	4
haemoptysis/haemorrhage	[ ]	4
massive haemoptysis	[ ]	6

6. CARDIOVASCULAR		6 (maximum total)
none	[ ]	0
bruits	[ ]	2
new loss of pulses	[ ]	4
aortic incompetence	[ ]	4
pericarditis	[ ]	4
new myocardial infarct	[ ]	6
CCF/cardiomyopathy	[ ]	6

7. ABDOMINAL		9 (maximum total)
none	[ ]	0
abdominal pain	[ ]	3
bloody diarrhoea	[ ]	6
gall bladder perforation	[ ]	9
gut infarction	[ ]	9
pancreatitis	[ ]	9

8. RENAL		12 (maximum total)
none	[ ]	0
hypertension (diastolic >90)	[ ]	4
proteinuria (>1+ or >0.2 g/24 h)	[ ]	4
haematuria (>1+ or >10 rbc/ml)	[ ]	8
creatinine 125–249 µmol/l	[ ]	8
creatinine 250–499 µmol/l	[ ]	10
creatinine >500 µmol/l	[ ]	12
rise in creatinine >10%	[ ]	12

9. NERVOUS SYSTEM		9 (maximum total)
none	[ ]	0
organic confusion/dementia	[ ]	3
seizures (not hypertensive)	[ ]	9
stroke	[ ]	9
cord lesion	[ ]	9
peripheral neuropathy	[ ]	6
motor mononeuritis multiplex	[ ]	9

Maximum score

Luqmani RA.. Birmingham Vasculitis Activity Score (BVAS) in systemic necrotizing vasculitis. QJM **1994**;87:671–8

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Sensibilidad al cambio de estadio	-	Si	Si	Si	Si	-

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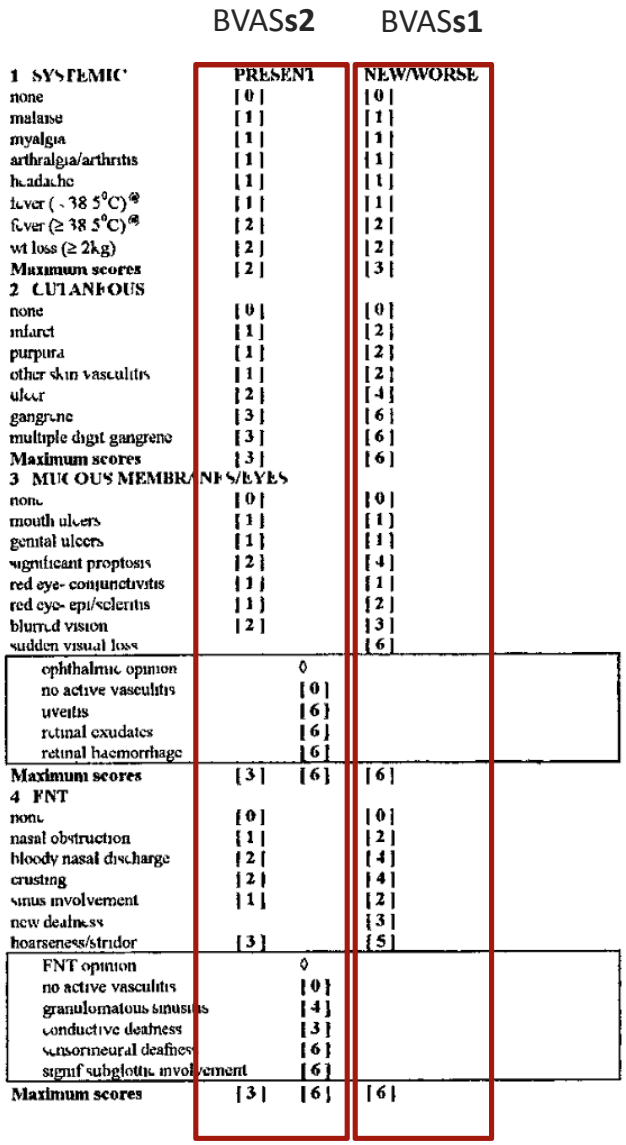
BVAS versión 2



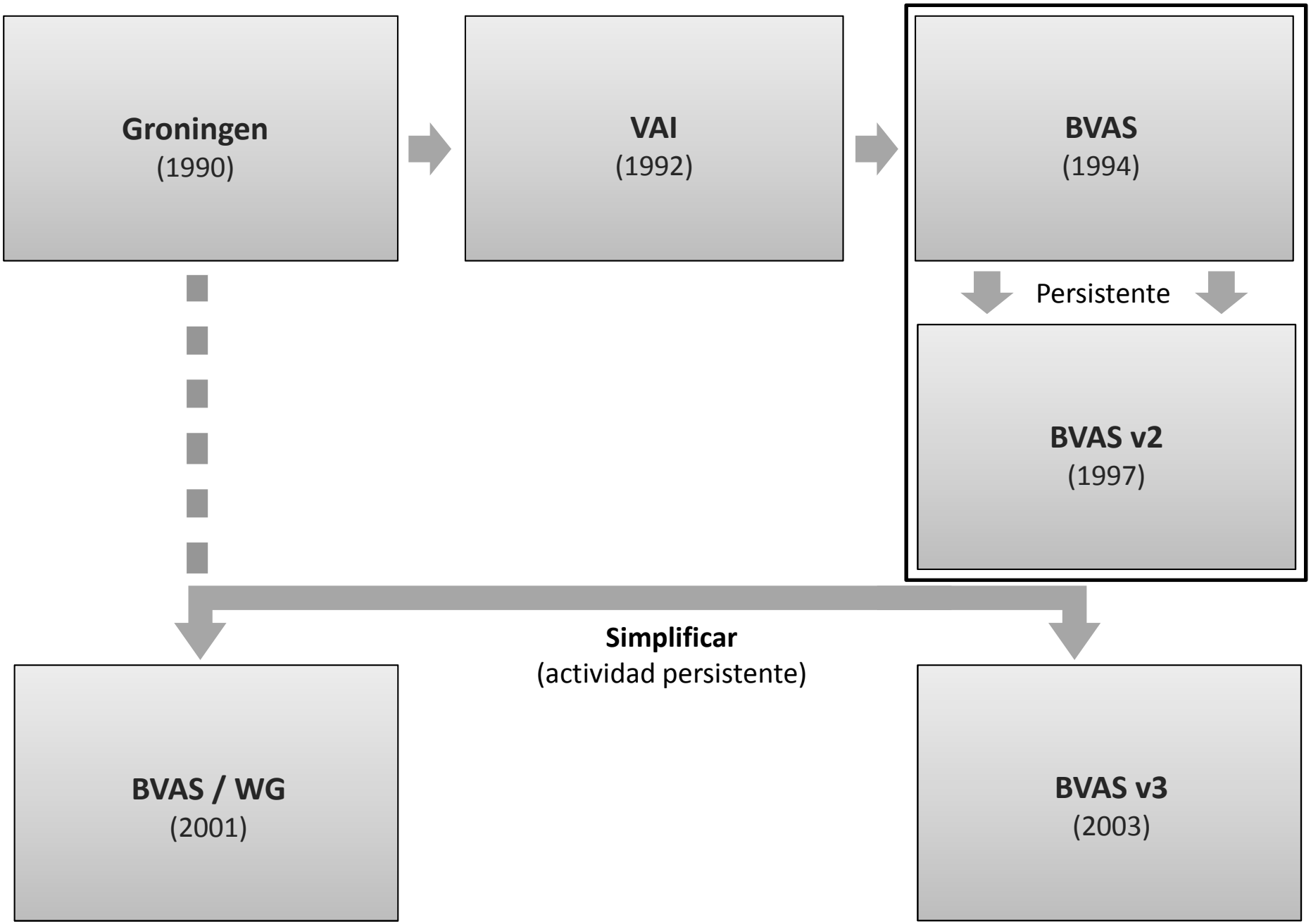
BVASs2		BVASs1
1 SYSTEMIC	**PRESENT**	**NEW/WORSE**
none	[0]	[0]
malaise	[1]	[1]
myalgia	[1]	[1]
arthralgia/arthritis	[1]	[1]
headache	[1]	[1]
fever (< 38.5°C)*	[1]	[1]
fever (≥ 38.5°C)*	[2]	[2]
wt loss (≥ 2kg)	[2]	[2]
Maximum scores	[2]	[3]
2 CUTANEOUS		
none	[0]	[0]
infarct	[1]	[2]
purpura	[1]	[2]
other skin vasculitis	[1]	[2]
ulcer	[2]	[4]
gangrene	[3]	[6]
multiple digit gangrene	[3]	[6]
Maximum scores	[3]	[6]
3 MUCOUS MEMBRANES/EYES		
none	[0]	[0]
mouth ulcers	[1]	[1]
genital ulcers	[1]	[1]
significant proptosis	[2]	[4]
red eye- conjunctivitis	[1]	[1]
red eye- epi/scleritis	[1]	[2]
blurred vision	[2]	[3]
sudden visual loss		[6]
ophthalmic opinion	◊	
no active vasculitis	[0]	
uveitis	[6]	
retinal exudates	[6]	
retinal haemorrhage	[6]	
Maximum scores	[3] [6]	[6]
4 ENT		
none	[0]	[0]
nasal obstruction	[1]	[2]
bloody nasal discharge	[2]	[4]
crusting	[2]	[4]
sinus involvement	[1]	[2]
new deafness		[3]
hoarseness/stridor	[3]	[5]
ENT opinion	◊	
no active vasculitis	[0]	
granulomatous sinusitis	[4]	
conductive deafness	[3]	
sensorineural deafness	[6]	
signif subglottic involvement	[6]	
Maximum scores	[3] [6]	[6]

Visit Date / /
Investigator

5. CHEST	**PRESENT**	**NEW/WORSE**
none	[0]	[0]
persistent cough	[1]	[2]
dyspnoea or wheeze	[1]	[2]
haemoptysis/haemorrhage	[1]	[3]
chest radiology performed	◊	
no active vasculitis	[0]	
nodules or cavities	[3]	
pleural effusion/pleurisy	[4]	
infiltrate	[4]	
massive haemoptysis		[6]
respiratory failure	[3]	[6]
Maximum scores	[3] [6]	[6]
6 CARDIOVASCULAR		
none	[0]	[0]
bruits	[1]	[2]
new loss of pulses		[4]
new loss of pulses with threatened loss of limb	[4]	
aortic incompetence	[2]	[4]
pericardial pain/rub	[2]	[3]
ischaemic cardiac pain	[2]	[4]
congestive cardiac failure	[2]	[4]
cardiology opinion/tests	◊	
no active vasculitis	[0]	
pericarditis	[4]	
myocardial infarct/angina	[6]	
cardiomyopathy	[6]	
Maximum scores	[3] [6]	[6]
7 ABDOMINAL		
none	[2]	[3]
severe abdominal pain	[2]	[3]
bloody diarrhoea	[2]	[3]
surgical opinion/tests	◊	
no active vasculitis	[0]	
gut perforation/infarct	[9]	
acute pancreatitis	[9]	
Maximum scores	[5] [9]	[9]
8 RENAL		
none	[0]	[0]
hypertension (diastol>95)	[1]	[4]
proteinur (>1+/>0.2g/24h)	[2]	[4]
haematur (>1+/>10rbc/ml)	[3]	[6]
rise in creatinine >30% or fall in creatinine clearance>25%		[6]
Maximum scores	[6]	[12]
9 NERVOUS SYSTEM		
none	[0]	[0]
organic confusion/dement	[1]	[3]
seizures(not hypertensive)	[3]	[9]
stroke	[3]	[9]
cord lesion	[3]	[9]
sensory periphl neuropath	[3]	[6]
cranial nerve palsy	[3]	[6]
motor mononeurit multipl	[3]	[9]
Maximum scores	[6]	[12]
10. OTHER		
Describe -		
MAXIMUM TOTAL SCORE	32	63

Inicio (66 ítems)
Seguimiento (62 ítems)

Luqmani RA,. Disease assessment and management of the vasculitides. **Baillieres Clin Rheumatol 1997;**11:423–46


Groningen
(1990)
VAI
(1992)
BVAS
(1994)
Persistente
BVAS v2
(1997)
Simplificar
(actividad persistente)
BVAS / WG
(2001)
BVAS v3
(2003)

BVAS v3 (2003)

secundario a vasculitis (56 ítems)

Simplifica recogida información:

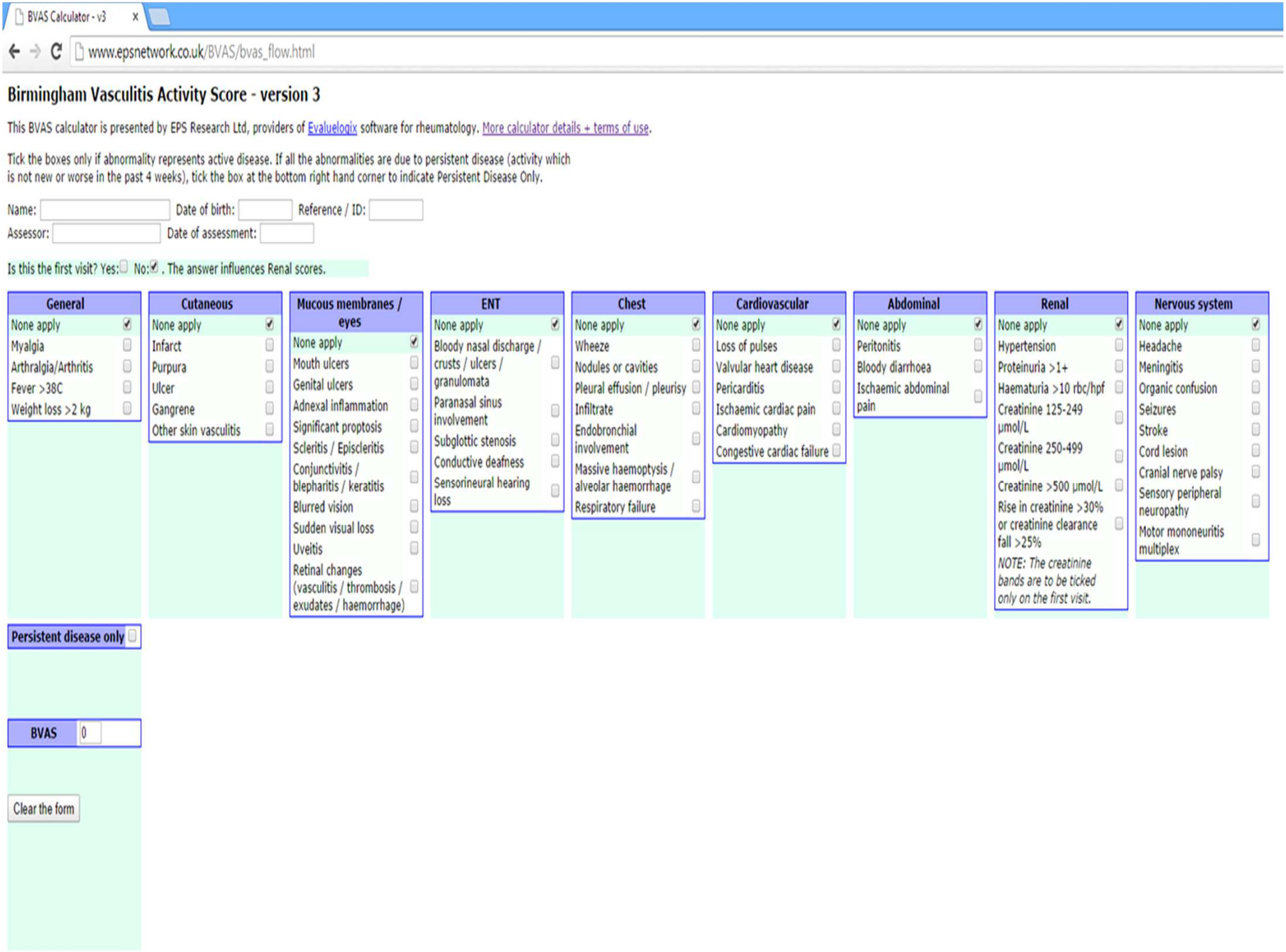
✓Inicio / Seguimiento
✓**Activas** = nuevas o persistentes
✓Casilla (**solo todo persistente**)

Is this the patient's first assessment? Yes ☐ No ☐

	None	Active disease
1. General	☐	
Myalgia		☐
Arthralgia / arthritis		☐
Fever ≥38° C		☐
Weight loss ≥2 kg		☐
2. Cutaneous	☐	
Infarct		☐
Purpura		☐
Ulcer		☐
♦Gangrene		☐
Other skin vasculitis		☐
3. Mucous membranes / eyes	☐	
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	☐	
Bloody nasal discharge / crusts / ulcers / granulomata		☐
Paranasal sinus involvement		☐
Subglottic stenosis		☐
Conductive hearing loss		☐
♦Sensorineural hearing loss		☐
5. Chest	☐	
Wheeze		☐
Nodules or cavities		☐
Pleural effusion / pleurisy		☐
Infiltrate		☐
Endobronchial involvement		☐
♦Massive haemoptysis / alveolar haemorrhage		☐
♦Respiratory failure		☐

	None	Active disease
6. Cardiovascular	☐	
Loss of pulses		☐
Valvular heart disease		☐
Pericarditis		☐
♦Ischaemic cardiac pain		☐
♦Cardiomyopathy		☐
♦Congestive cardiac failure		☐
7. Abdominal	☐	
Peritonitis		☐
Bloody diarrhoea		☐
♦Ischaemic abdominal pain		☐
8. Renal	☐	
Hypertension		☐
Proteinuria >1+		☐
♦Haematuria ≥10 RBCs/hpf		☐
Creatinine 125-249µ/L(1.41-2.82mg/dl)*		☐
Creatinine 250-499 µ/L(2.83-5.64mg/dl)*		☐
♦Creatinine ≥500 µ/L (≥5.66mg/dl)*		☐
♦Rise in serum creatinine >30% or fall in creatinine clearance >25%		☐
***Can only be scored on the first assessment**		
9. Nervous system	☐	
Headache		☐
Meningitis		☐
Organic confusion		☐
Seizures (not hypertensive)		☐
♦Cerebrovascular accident		☐
♦Spinal cord lesion		☐
♦Cranial nerve palsy		☐
Sensory peripheral neuropathy		☐
♦Mononeuritis multiplex		☐
10. Other	☐	
a.		☐
b.		☐
c.		☐
d.		☐
PERSISTENT DISEASE ONLY: (Tick here if **all** the abnormalities are due to persistent disease)		☐

Mukhtyar C. Modification and validation of the Birmingham vasculitis activity score (version 3). Ann Rheum Dis **2009**;68:1827-32
Suppiah R. A cross-sectional study of the Birmingham Vasculitis Activity Score version 3 in systemic vasculitis. Rheumatology. **2011** May;50(5):899-905


BVAS Calculator - v3
www.epsnetwork.co.uk/BVAS/bvas_flow.html
Birmingham Vasculitis Activity Score - version 3
This BVAS calculator is presented by EPS Research Ltd, providers of Evaluelogix software for rheumatology. More calculator details + terms of use.
Tick the boxes only if abnormality represents active disease. If all the abnormalities are due to persistent disease (activity which is not new or worse in the past 4 weeks), tick the box at the bottom right hand corner to indicate Persistent Disease Only.
Name:
Date of birth:
Reference / ID:
Assessor:
Date of assessment:
Is this the first visit? Yes: No: . The answer influences Renal scores.
General
None apply
Myalgia
Arthralgia/Arthritis
Fever >38C
Weight loss >2 kg
Cutaneous
None apply
Infarct
Purpura
Ulcer
Gangrene
Other skin vasculitis
Mucous membranes / eyes
None apply
Mouth ulcers
Genital ulcers
Adnexal inflammation
Significant proptosis
Scleritis / Episcleritis
Conjunctivitis / blepharitis / keratitis
Blurred vision
Sudden visual loss
Uveitis
Retinal changes (vasculitis / thrombosis / exudates / haemorrhage)
ENT
None apply
Bloody nasal discharge / crusts / ulcers / granulomata
Paranasal sinus involvement
Subglottic stenosis
Conductive deafness
Sensorineural hearing loss
Chest
None apply
Wheeze
Nodules or cavities
Pleural effusion / pleurisy
Infiltrate
Endobronchial involvement
Massive haemoptysis / alveolar haemorrhage
Respiratory failure
Cardiovascular
None apply
Loss of pulses
Valvular heart disease
Pericarditis
Ischaemic cardiac pain
Cardiomyopathy
Congestive cardiac failure
Abdominal
None apply
Peritonitis
Bloody diarrhoea
Ischaemic abdominal pain
Renal
None apply
Hypertension
Proteinuria >1+
Haematuria >10 rbc/hpf
Creatinine 125-249 µmol/L
Creatinine 250-499 µmol/L
Creatinine >500 µmol/L
Rise in creatinine >30% or creatinine clearance fall >25%
NOTE: The creatinine bands are to be ticked only on the first visit.
Nervous system
None apply
Headache
Meningitis
Organic confusion
Seizures
Stroke
Cord lesion
Cranial nerve palsy
Sensory peripheral neuropathy
Motor mononeuritis multiplex
Persistent disease only
BVAS 0
Clear the form

Disease Status	BVAS items	Old BVAS	BVAS (V. 3)
Remission	*New/worse items:* None *Persistent items:* None	BVAS.1=0 BVAS.2=0	BVAS new/worse = 0 BVAS persistent = 0
Active Disease, No persistent items	*New/worse items:* Malaise Myalgia Arthralgia/arthritis Headache/meningitis Fever ≥ 38.5^{0}C Weight loss ≥ 2kg Nasal obstruction Bloody nasal discharge Nasal crusting Persistent cough Infiltrate Proteinuria Haematuria Creatinine 125-249µmol/L Rise in creatinine >30% *Persistent items:* None	BVAS.1=27 BVAS.2=0	BVAS new/worse = 29 BVAS persistent = 0
Persistent Disease, No new/worse items	*New/worse items:* None *Persistent items:* Arthralgia/arthritis Other skin vasculitis Pericardial pain/rub	BVAS.1=0 BVAS.2=5	BVAS new/worse = 0 BVAS persistent = 3
Persistent Disease, 1 new item	*New/worse items:* Stroke *Persistent items:* Fever ≥ 38.5^{0}C Weight loss ≥ 2kg	BVAS.1=9 BVAS.2=5	BVAS new/worse = 18 BVAS persistent = 0

	Nuevas / peores	Persistentes
8. Renal	**12**	**6**
Hypertension	4	1
Proteinuria	4	2
Haematuria	6	3
Creatinine 125-249	4	*
Creatinine ≥ 500	8	*
Rise in creatinine> 30% or creatinine clearance fall > 25%	6	*

ponderadas expertos

Características	Groningen Kallenberg 1990	VAI Olsen 1992 Whiting-O'Keefe 1999	BVAS **BVAS v2** Luqmani 1994 / 1997	BVAS v3 Mukhtyar 2009	BVAS v3 Suppiah 2011	BVAS/WG Stone 2001
Tipo vasculitis	GPA	Múltiples	Múltiples	Múltiples (no Horton)	Múltiples	GPA
Nº pacientes	¿?	74	213	313	238	117
Face validity	Si	Si	Si	Si	Si	Si
Feasybility	Biopsias	Si	Si	Si	Si	Si
Reliability **- Inter** (reproducible) **- Intra** (repetible)	¿? - -	 ICC=0.45 -	 Si -	 ICC = 0.96 ICC = 0.96	 ICC=0.99	 ICC = 0.97 ICC = 0.62
Construct validity **- PGA** **- VAI** **- Otros indices** **- PCR** **- Decisión tto**	 - - - - -	 R=0.92 - - - -	 T=0.35 T=0.56 T=0.29 (Gron.) ¿? -	 R=0.91 R=0.88 R=0.94(s1) 0.6 (s2) R=0.43 R=0.66	 R=0.85 R=0.82 VDI= -0.1 R=0.18 R=0.54	 R=0.92 - - - -
Sensibilidad al cambio de estadio	-	Si	Si	Si	-	Si

Kallenberg CG. Criteria for disease activity in Wegener's granulomatosis: a requirement for longitudinal clinical studies. **APMIS Suppl 1990**;19:37–9

Whiting-O'Keefe. Validity of a vasculitis activity index for systemic necrotizing vasculitis. Arthritis Rheum **1999**;42:2365-71

Luqmani RA.. Birmingham Vasculitis Activity Score (BVAS) in systemic necrotizing vasculitis. QJM **1994**;87:671–8

Luqmani RA,. Disease assessment and management of the vasculitides. Baillieres Clin Rheumatol **1997;**11:423–46

Stone JH. A disease-specific activity index for Wegener's granulomatosis: modification of the Birmingham Vasculitis Activity Score. **Arthritis Rheum 2001**;44:912–20

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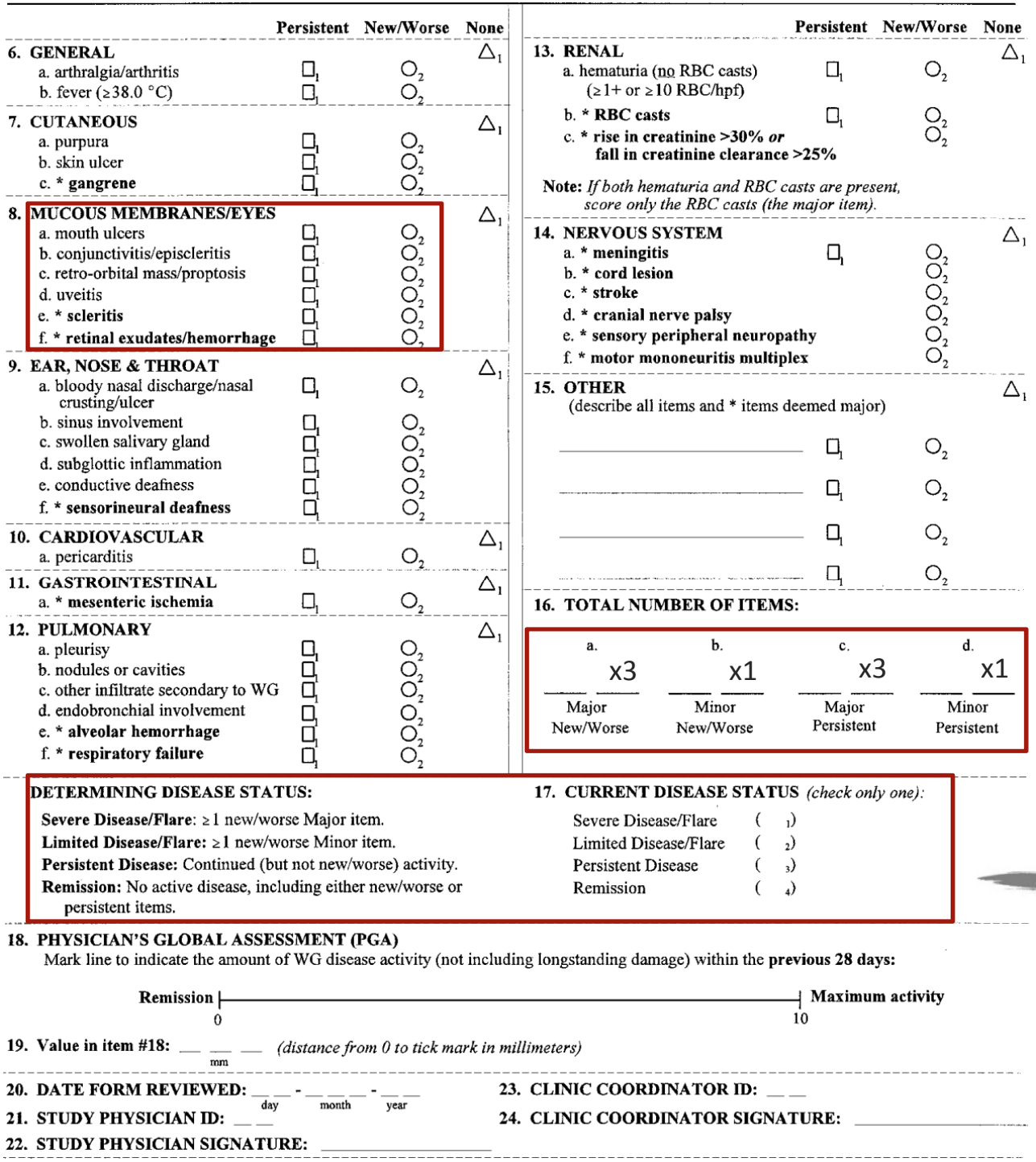

	Persistent	New/Worse	None
6. GENERAL			△1
a. arthralgia/arthritis	□1	○2	
b. fever (≥38.0 °C)	□1	○2	
7. CUTANEOUS			△1
a. purpura	□1	○2	
b. skin ulcer	□1	○2	
c. * **gangrene**	□1	○2	
8. MUCOUS MEMBRANES/EYES			△1
a. mouth ulcers	□1	○2	
b. conjunctivitis/episcleritis	□1	○2	
c. retro-orbital mass/proptosis	□1	○2	
d. uveitis	□1	○2	
e. * **scleritis**	□1	○2	
f. * **retinal exudates/hemorrhage**	□1	○2	
9. EAR, NOSE & THROAT			△1
a. bloody nasal discharge/nasal crusting/ulcer	□1	○2	
b. sinus involvement	□1	○2	
c. swollen salivary gland	□1	○2	
d. subglottic inflammation	□1	○2	
e. conductive deafness	□1	○2	
f. * **sensorineural deafness**	□1	○2	
10. CARDIOVASCULAR			△1
a. pericarditis	□1	○2	
11. GASTROINTESTINAL			△1
a. * **mesenteric ischemia**	□1	○2	
12. PULMONARY			△1
a. pleurisy	□1	○2	
b. nodules or cavities	□1	○2	
c. other infiltrate secondary to WG	□1	○2	
d. endobronchial involvement	□1	○2	
e. * **alveolar hemorrhage**	□1	○2	
f. * **respiratory failure**	□1	○2	

	Persistent	New/Worse	None
13. RENAL			△1
a. hematuria (no RBC casts) (≥1+ or ≥10 RBC/hpf)	□1	○2	
b. * **RBC casts**	□1	○2	
c. * **rise in creatinine >30% *or* fall in creatinine clearance >25%**		○2	

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

14. NERVOUS SYSTEM			△1
a. * **meningitis**	□1	○2	
b. * **cord lesion**		○2	
c. * **stroke**		○2	
d. * **cranial nerve palsy**		○2	
e. * **sensory peripheral neuropathy**		○2	
f. * **motor mononeuritis multiplex**		○2	
15. OTHER (describe all items and * items deemed major)			△1
_____	□1	○2	
_____	□1	○2	
_____	□1	○2	
_____	□1	○2	

16. TOTAL NUMBER OF ITEMS:

a.	b.	c.	d.
____ x3	____ x1	____ x3	____ x1
Major New/Worse	Minor New/Worse	Major Persistent	Minor Persistent

DETERMINING DISEASE STATUS:

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

17. CURRENT DISEASE STATUS *(check only one)*:

Severe Disease/Flare (1)
Limited Disease/Flare (2)
Persistent Disease (3)
Remission (4)

18. PHYSICIAN'S GLOBAL ASSESSMENT (PGA)

Mark line to indicate the amount of WG disease activity (not including longstanding damage) within the **previous 28 days:**

Remission (0) — (10) **Maximum activity**

19. Value in item #18: ___ ___ ___ mm *(distance from 0 to tick mark in millimeters)*

20. DATE FORM REVIEWED: ___ - ___ ___ - ___ (day - month - year)

21. STUDY PHYSICIAN ID: ___ ___

22. STUDY PHYSICIAN SIGNATURE: _____

23. CLINIC COORDINATOR ID: ___ ___

24. CLINIC COORDINATOR SIGNATURE: _____

BVAS / WG(GPA)

secundario a **GPA** (34 ítems)
nuevas o persistentes
mayores* (x3) y **menores** (x1)

Máximo 68
- 64 predeterminados
- 1 *otro* mayor (x3)
- 1 *otro* menor (x1)

Decisiones terapéuticas

Stone JH. A disease-specific activity index for Wegener's granulomatosis: modification of the Birmingham Vasculitis Activity Score. **Arthritis Rheum 2001**;44:912–20.

Características	**Groningen** Kallenberg 1990	**VAI** Olsen 1992 Whiting-O'Keefe 1999	**BVAS** **BVAS v2** Luqmani 1994 / 1997	**BVAS v3** Mukhtyar 2009	Suppiah 2011	**BVAS/WG** Stone 2001
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Feasybility	Biopsias	Si	Si	Si	Si	Si
Reliability **- Inter** (reproducible) **- Intra** (repetible)	¿? - -	 ICC=0.45 -	 Si -	 ICC = 0.96 ICC = 0.96	 ICC=0.99	 ICC = 0.97 ICC = 0.62
Construct validity **- PGA** **- VAI** **- Otros indices** **- PCR** **- Decisión tto**	 - - - - -	 R=0.92 - - - -	 T=0.35 T=0.56 T=0.29 (Gron.) ¿? -	 R=0.91 R=0.88 R=0.94(s1) 0.6 (s2) R=0.43 R=0.66	 R=0.85 R=0.82 VDI= -0.1 R=0.18 R=0.54	 R=0.92 - - - -
Sensibilidad al cambio de estadio	-	Si	Si	Si	-	Si

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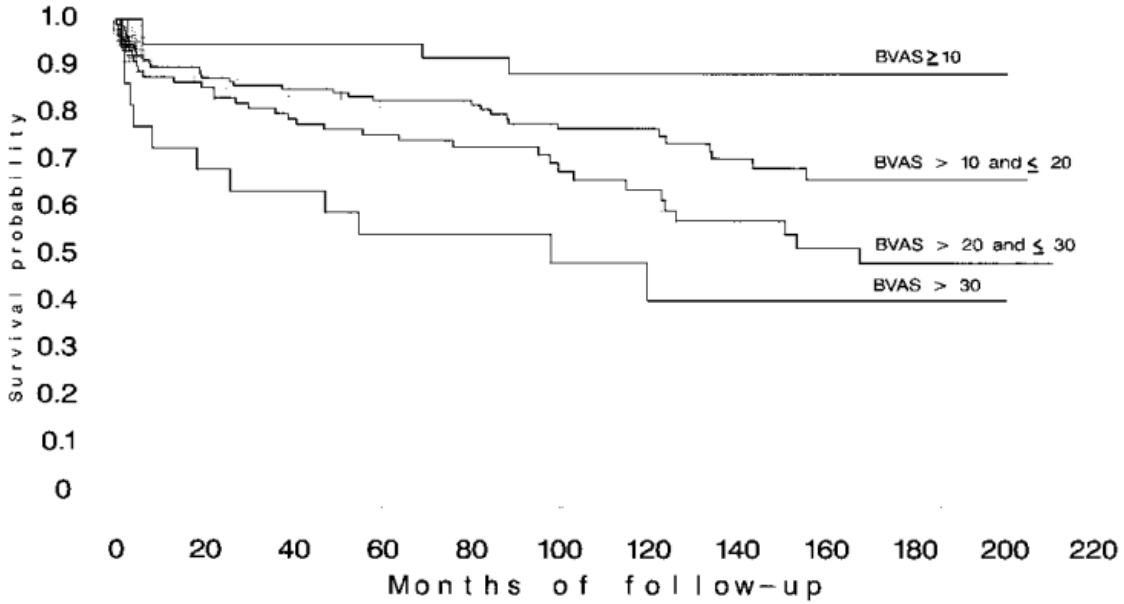
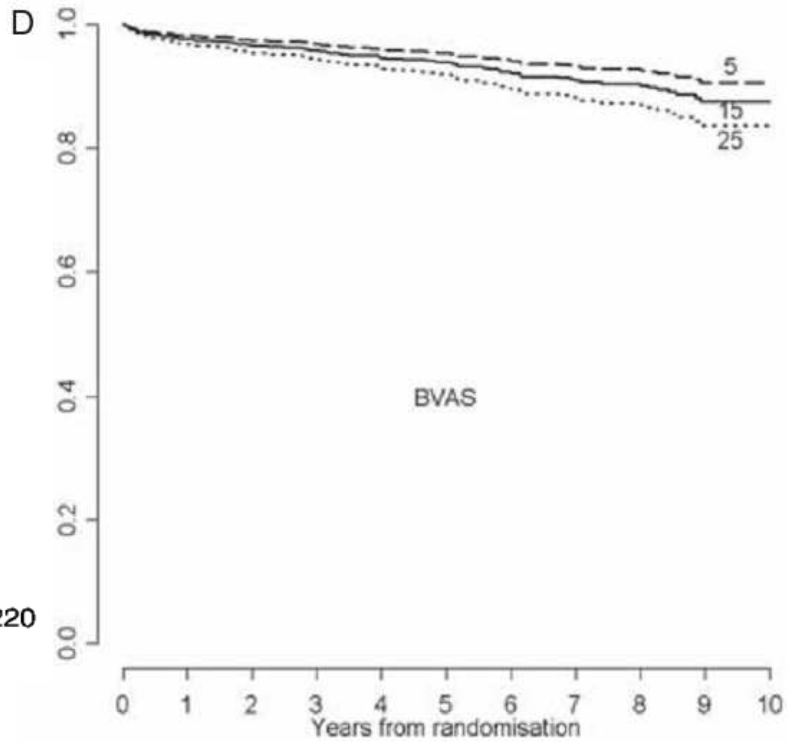
UTILIDAD del BVAS

Estandarizar en los ensayos / práctica clínica

Información **pronóstica** (al igual que el FFS)





Merkel PA. Current status of outcome measures in vasculitis: focus on Wegener's granulomatosis and microscopic polyangiitis. **JRheumatol. 2005** 32:2488-95

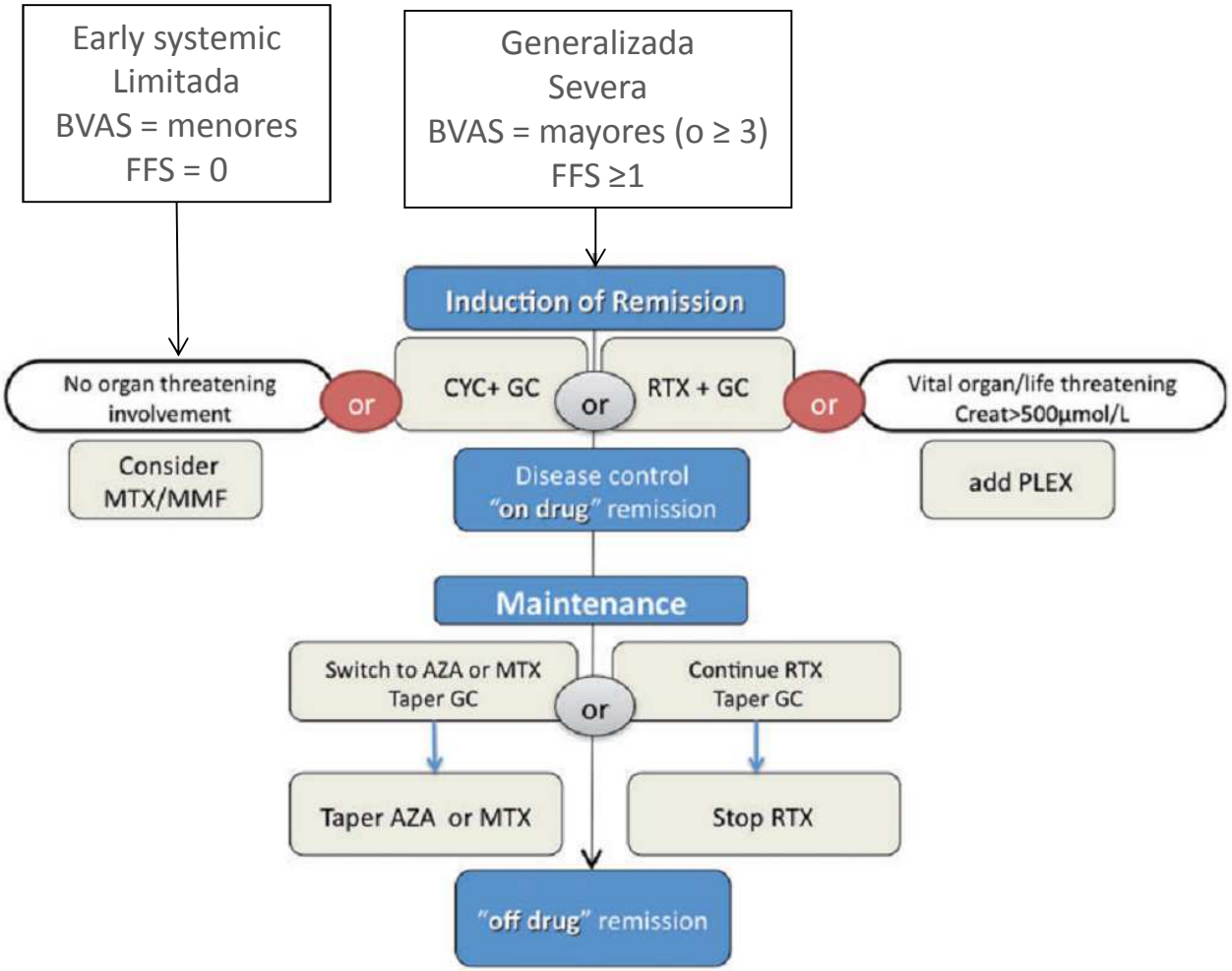
Hellmich B, EULAR recommendations for conducting clinical studies and/or clinical trials in systemic vasculitis. **Ann Rheum Dis. 2007** May;66(5):605-17

Gayraud M. Long-term followup of polyarteritis nodosa, microscopic polyangiitis, and Churg-Strauss syndrome:. **Arthritis Rheum. 2001** Mar;44(3):666-75

Flossmann O. Long-term patient survival in ANCA-associated vasculitis. **Ann Rheum Dis. 2011** Mar;70(3):488-94

UTILIDAD del BVAS

Decisión terapéutica inicial / recidiva



Ntatsaki E. BSR and BHPR guideline for the management of adults with ANCA-associated vasculitis. **Rheumatology (Oxford). 2014** Apr 11.

UTILIDAD del BVAS

Definir **estadios evolutivos** de la vasculitis

Remisión:	ausencia completa de actividad = BVAS 0
Respuesta:	descenso del BVAS >50% y ausencia de nuevas manifestaciones
Refractario:	BVAS igual o mayor tras 4 semanas de tratamiento estándar, o falta respuesta, descenso del BVAS <50% tras 6 semanas de tratamiento, o cronicidad, BVAS con ≥ 1 ítem mayor o ≥ 3 menores tras ≥ 12 sem de tto.
Baja actividad:	Persistencia de síntomas menores que responden a dosis bajas de CS.
Recidiva:	Reaparición o nuevo síntoma atribuible a la vasculitis (mayor o menor)

Determinar la **eficacia del tratamiento** durante evolución

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Gracias