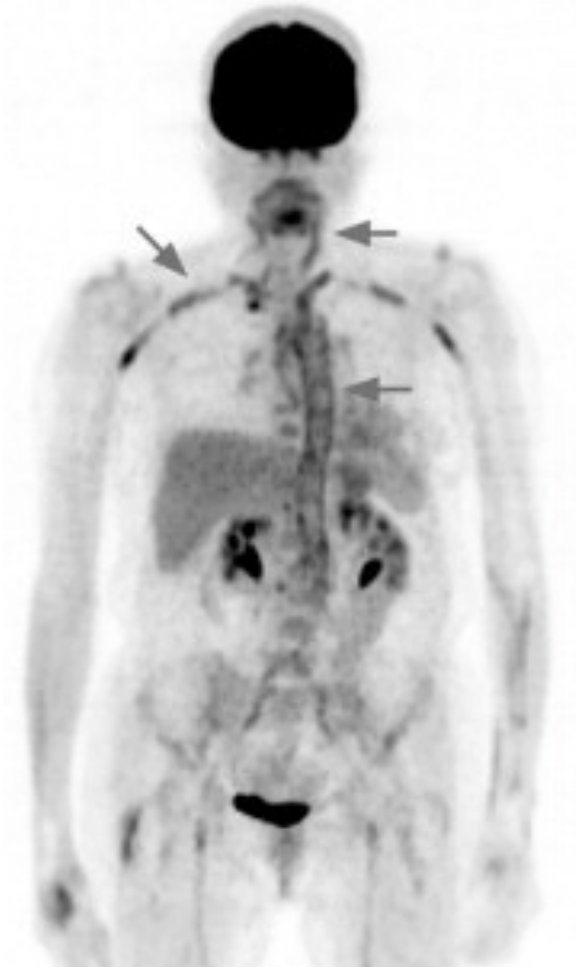
Université Paris Cité

GFEV GROUPE FRANÇAIS D'ÉTUDE DES VASCULARITES

HÔPITAUX UNIVERSITAIRES PARIS CENTRE
Cochin • Port-Royal • Tarnier • Broca
La Collégiale • La Rochefoucauld • Hôtel-Dieu

Artérite à cellules géantes



Benjamin Terrier

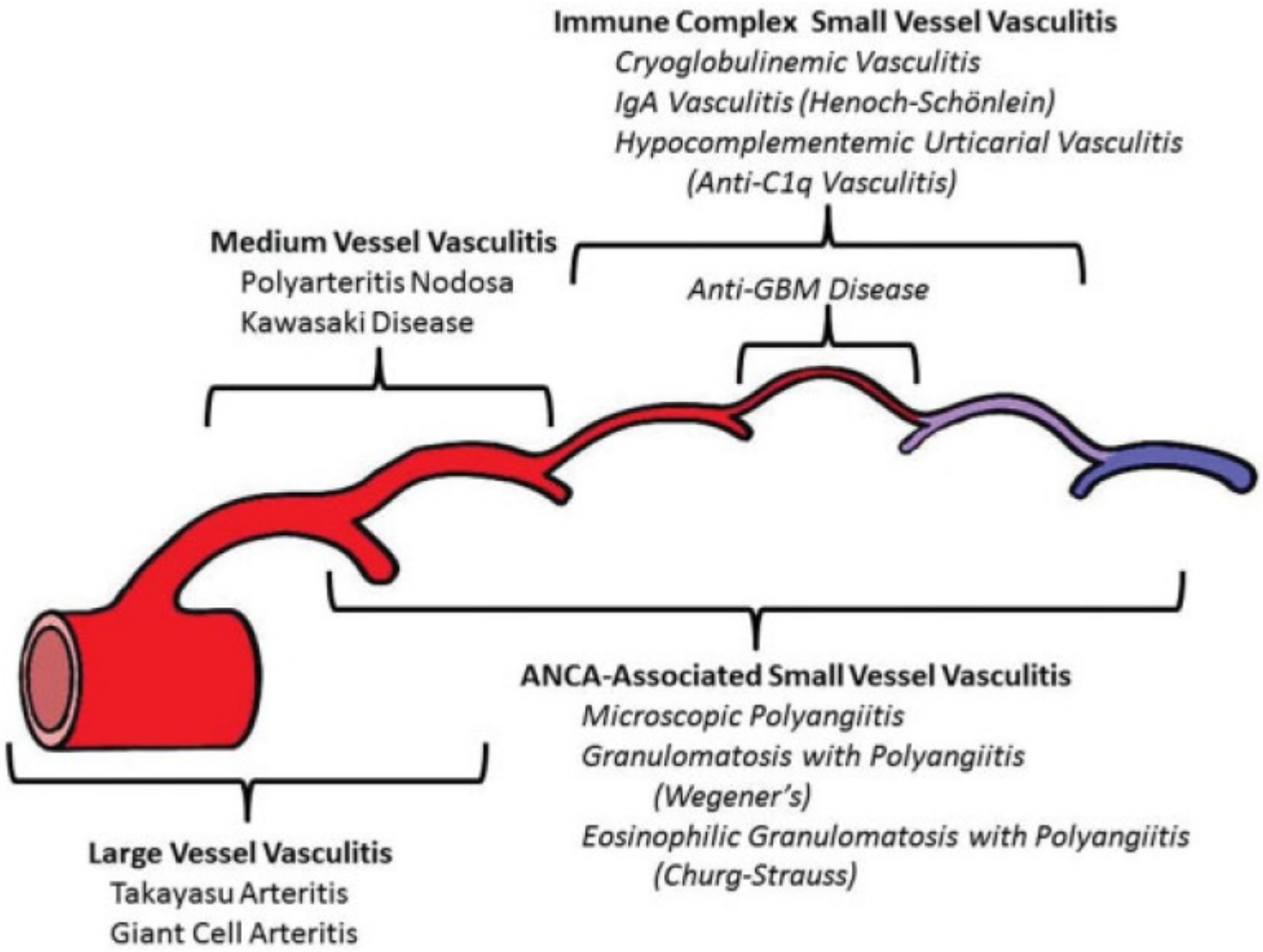
CMR Maladies Autolmmunes et Systémiques Rares d'Ile de France

Hôpital Cochin, Université de Paris

PARCC, INSERM U970

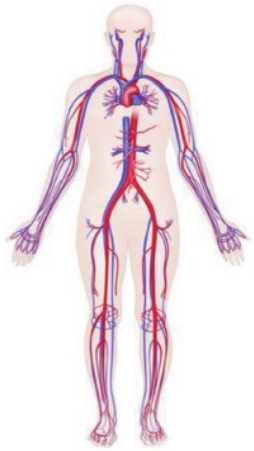
Paris, France

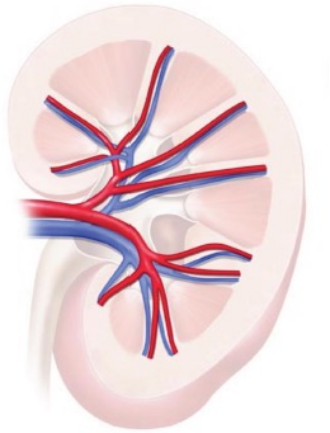
Vascularites des gros vaisseaux



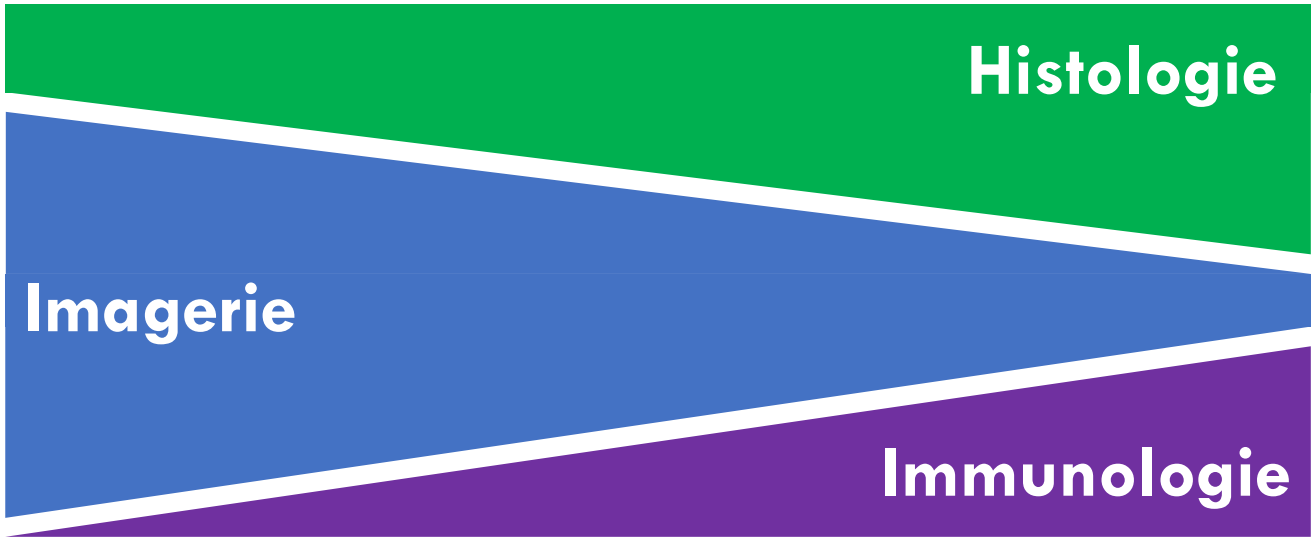
Jennette, Arthritis Rheum, 2013

Taille des vaisseaux atteints

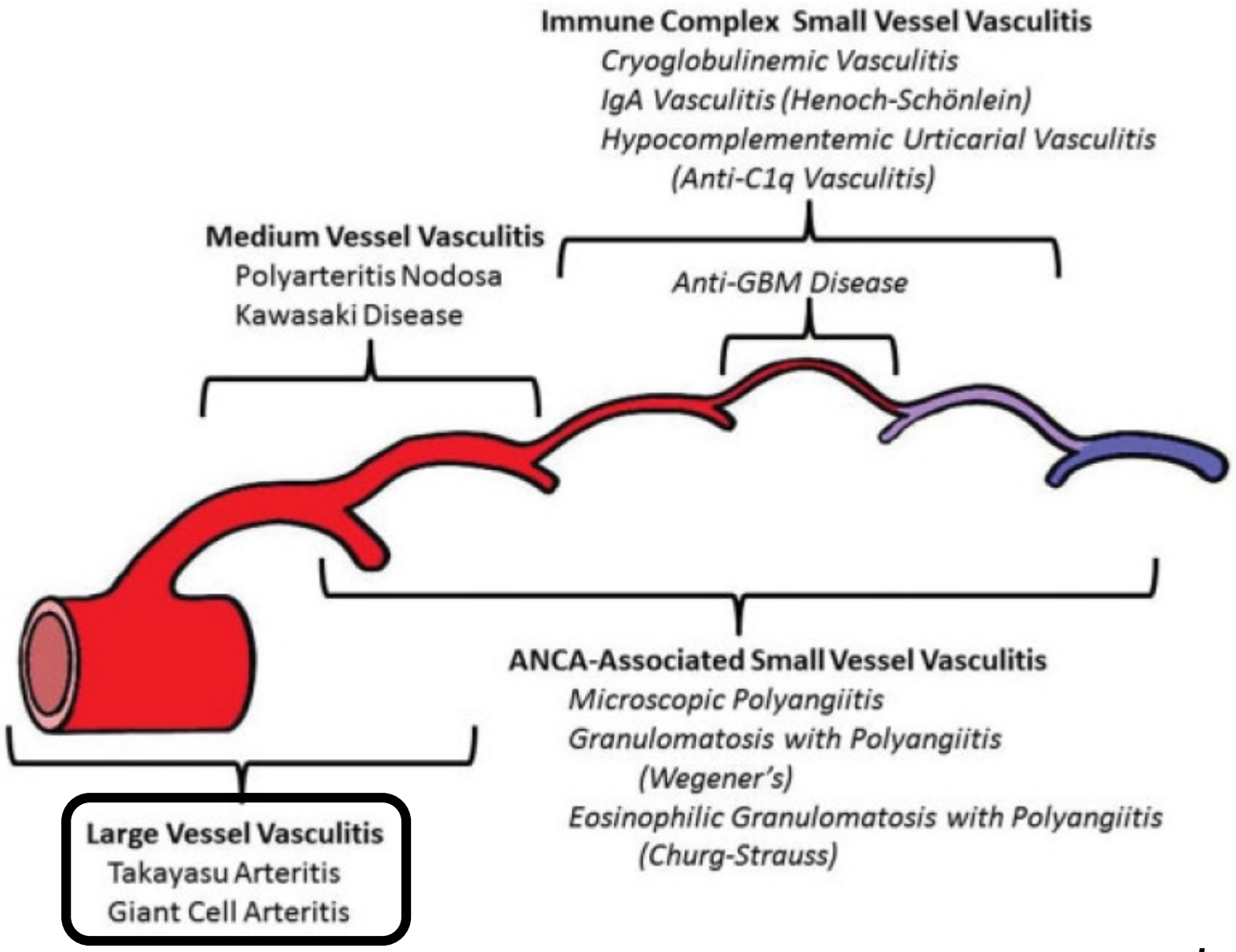




Contribution des approches immunologiques, radiologiques et histologiques pour le diagnostic de vascularite



Vascularites des gros vaisseaux



Jennette, Arthritis Rheum, 2013

Artérite à cellules géantes

Âge > 50 ans : 70-75 ans

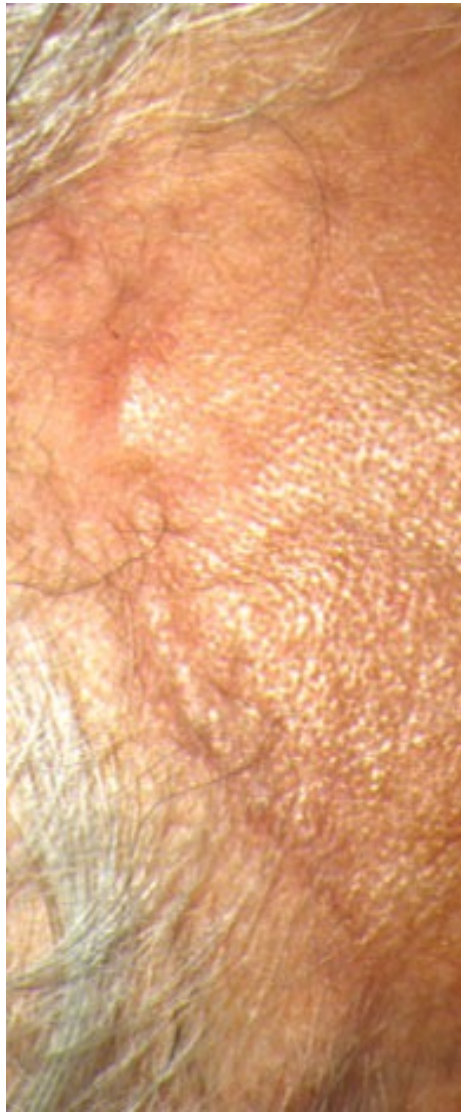
Prédominance féminine : 2 à 3 femmes pour un homme

Plus fréquente en Europe du Nord

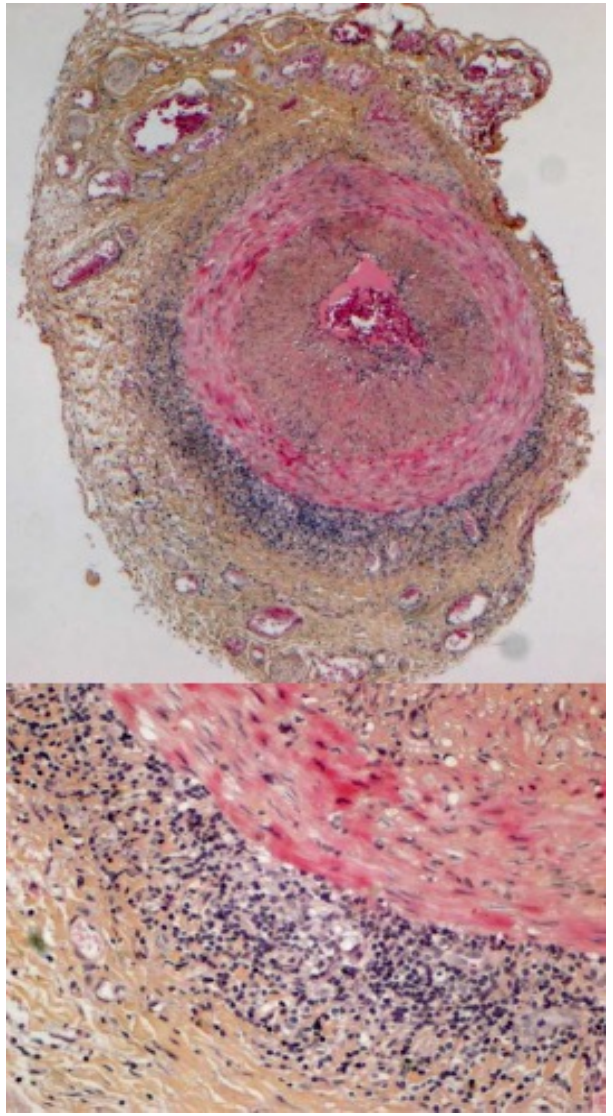
Clinique

- **Signes céphaliques +++**
- **PPR**
- **Signes oculaires**
- **AEG**

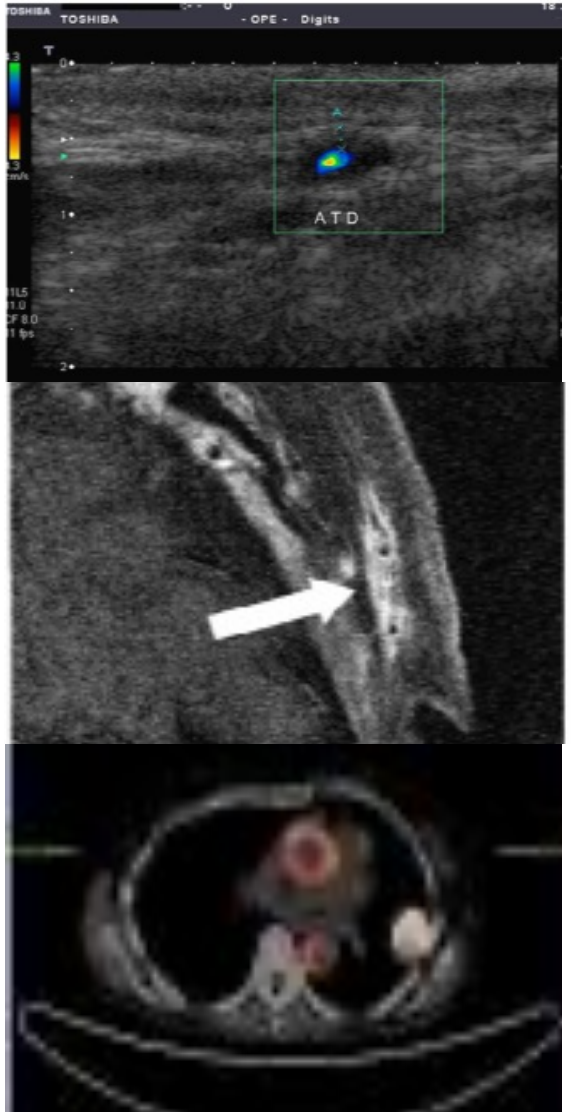
Artère temporale



Histologie



Imagerie



Incidence de l'ACG

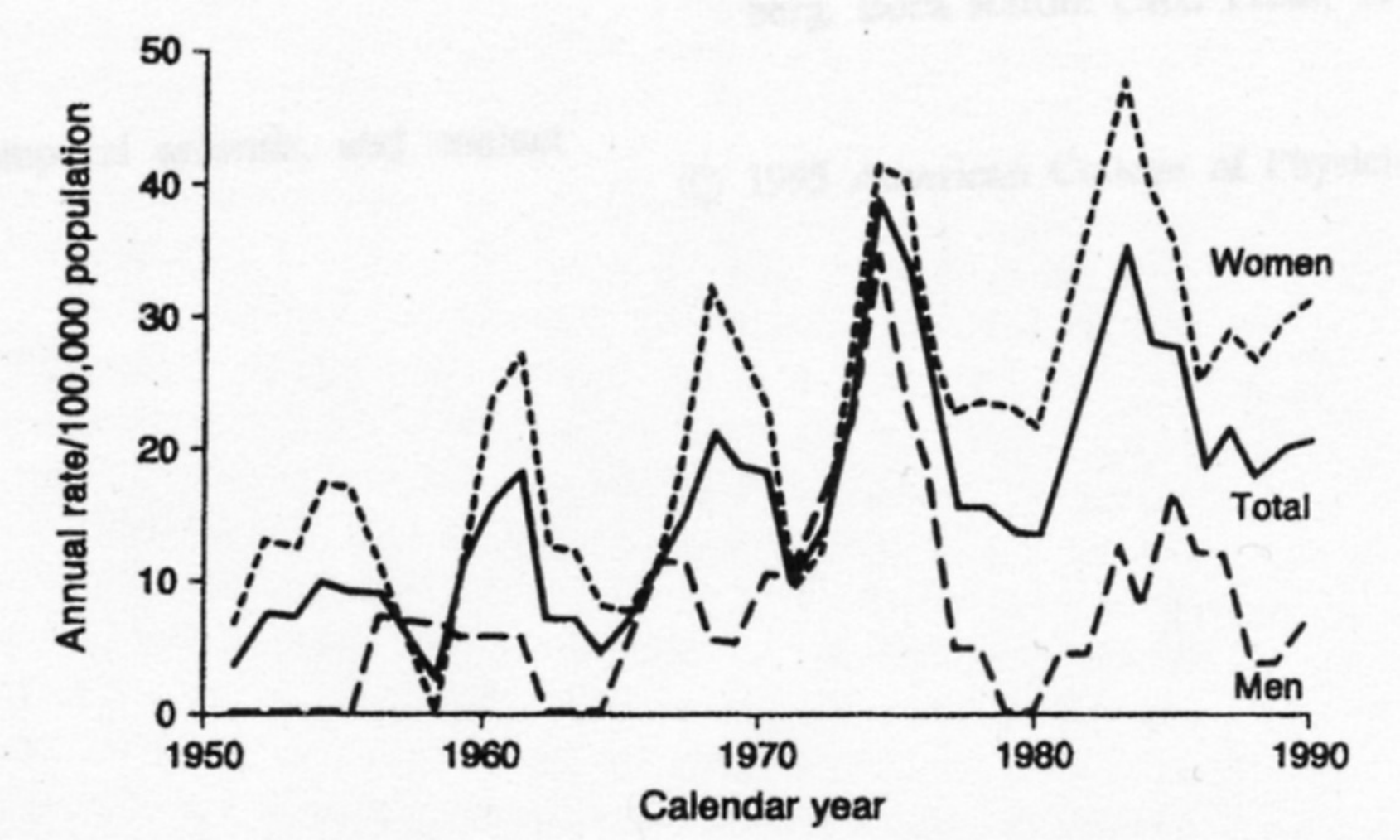
Aust Agder County (Norvège), 1987-94

Age	Incidence (/10^5 hab. ≥ 50 ans)		
	Femmes	Hommes	Tous
50-54	0,0	0,0	0,0
55-59	11,8	5,8	8,8
60-64	26,7	27,1	26,9
65-69	72,2	21,2	47,7
70-74	80,4	36,5	61,1
75-79	71,0	27,9	53,3
80-84	27,0	14,6	27,0
≥ 85	31,1	0,0	21,4
≥ 50	39,9	16,3	29,0

Gran, J Rheum, 1997

Incidence de l'ACG

Olmsted County (Minnesota), 1950-91



Salvarani, Ann Intern Med, 1995

Incidence de l'ACG

Lugo (Espagne), 1981-98

Gonzalez-Gay, Ann Rheum Dis, 2001

Présentation clinique de l'ACG

Performance des symptômes cliniques chez les patients suspects d'ACG

Symptom/References	No. of Patients With Data on Variable†	Positive LR (95% CI)	Negative LR (95% CI)
Anorexia[34,37,39,41,42,46]	674	1.2 (0.96-1.4)	0.87 (0.75-1.0)
Weight loss[31,34,36,37,39,41,42,46,47]	1417	1.3 (1.1-1.5)	0.89 (0.79-1.0)
Arthralgia[33,34,37,39,40,44,46,52]	582	1.1 (0.86-1.4)	1.0 (0.92-1.1)
Diplopia[33,34,42,50,51]	703	3.4 (1.3-8.6)	0.95 (0.91-0.99)
Fatigue[31,33,37,39,41,42,44,46]	1095	1.2 (0.98-1.4)	0.94 (0.86-1.0)
Fever[29,31,34-37,40-42,46,47]	1708	1.2 (0.98-1.4)	0.92 (0.85-0.99)
Temporal headache[36,42]	386	1.5 (0.78-3.0)	0.82 (0.64-1.0)
Any headache[29,31-35,37,39-47,50,51]	2475	1.2 (1.1-1.4)	0.7 (0.57-0.85)
Jaw claudication[29,31-35,37,39,40,42,44-46,50-52]	2314	4.2 (2.8-6.2)	0.72 (0.65-0.81)
Myalgia[31,36,39,40,46]	681	0.93 (0.81-1.1)	1.1 (0.87-1.3)
Polymyalgia rheumatica[29,34,35,37,39,40,42,44,45,47,50]	1383	0.97 (0.76-1.2)	0.99 (0.83-1.2)
Unilateral visual loss[32,50]	341	0.85 (0.58-1.2)	1.2 (1.0-1.3)
Any visual symptom[29,32-37,39-42,44-47,51,52]	2083	1.1 (0.93-1.3)	0.97 (0.9-1.0)
Vertigo[34,36,44]	212	0.71 (0.38-1.3)	1.1 (0.93-1.2)

Smetana, JAMA, 2002

Présentation clinique de l'ACG

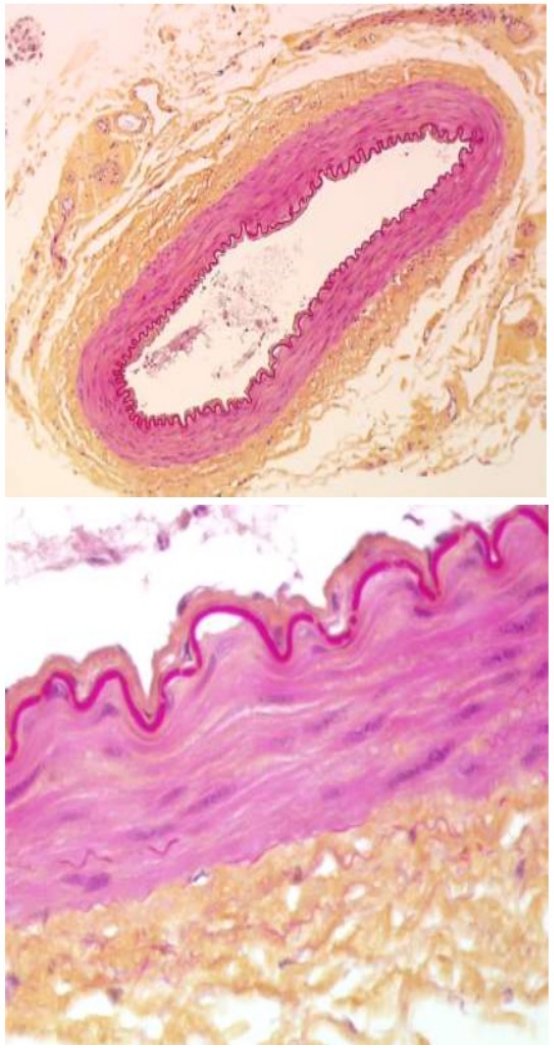
Performance des symptômes cliniques chez les patients suspects d'ACG

Variable/References	No. of Patients With Data on Variable†	Positive LR (95% CI)	Negative LR (95% CI)
	Signs and Demographics		
Optic atrophy or ischemic optic neuropathy[40,50]	142	1.6 (1.0-2.5)	0.8 (0.58-1.1)
Any funduscopic abnormality[29,35,50,52]	745	1.1 (0.8-1.4)	1.0 (0.92-1.1)
Scalp tenderness[31,33-35,52]	923	1.6 (1.2-2.1)	0.93 (0.86-1.0)
Synovitis[29,37,46,52]	734	0.41 (0.23-0.72)	1.1 (1.0-1.2)
Beaded temporal artery[42,52]	323	4.6 (1.1-18.4)	0.93 (0.88-0.99)
Prominent or enlarged temporal artery[36,39,42,44,52]	508	4.3 (2.1-8.9)	0.67 (0.5-0.89)
Tender temporal artery[36,39-42,50-52]	755	2.6 (1.9-3.7)	0.82 (0.74-0.92)
Absent temporal artery pulse[41,52]	68	2.7 (0.55-13.4)	0.71 (0.38-1.3)
Any temporal artery abnormality[29,31-33,37,43,46]‡	1559	2.0 (1.4-3.0)	0.53 (0.38-0.75)
Male sex[29,31-37,40-43,45-47,49-52]	2565	0.83 (0.72-0.96)	
White Race[32,35,37,40,50]	565	1.1 (0.99-1.2)	

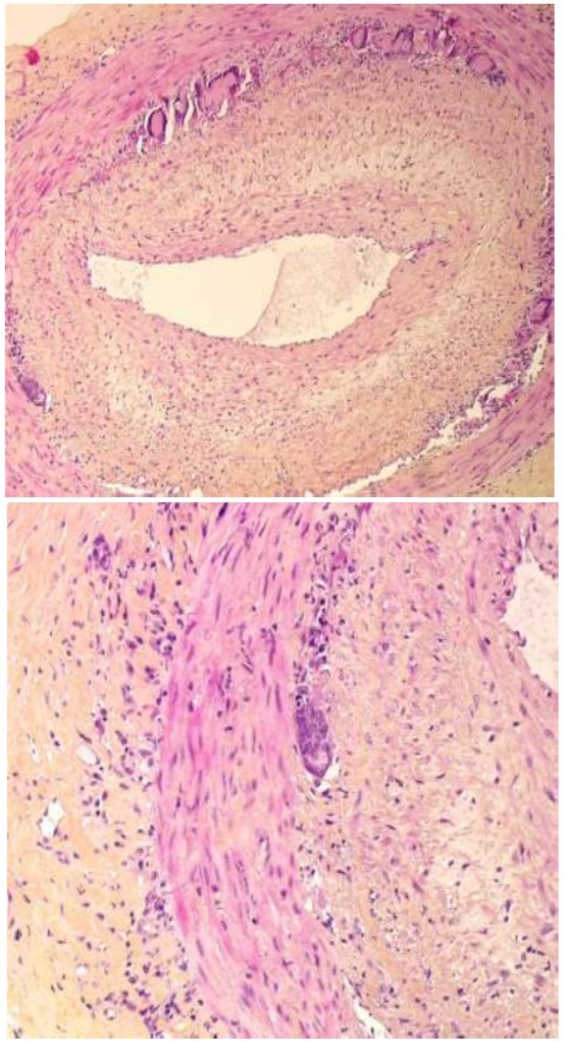
Smetana, JAMA, 2002

Biopsie d'artère temporale

Contrôle



Artérite à cellules géantes



Caractéristiques histologiques

- Epaississement intimal
- Présence de macrophages à proximité des limitantes élastiques
- Infiltrat inflammatoire polymorphe
- Destruction de la LEI
- Néointima, rétrécissement de la lumière artérielle
- Néo-vascularisation de la média

Mais 20-40% des BAT sont (faussement ?) négatives

Apport de l'imagerie au cours des vascularites des gros vaisseaux

EULAR recommendations for the use of imaging in large vessel vasculitis in clinical practice

Christian Dejaco,[1,2] Sofia Ramiro,[3] Christina Duftner,[4]
Florent L Besson,[5,6] Thorsten A Bley,[7] Daniel Blockmans,[8] Elisabeth Brouwer,[9]
Marco A Cimmino,[10] Eric Clark,[11] Bhaskar Dasgupta,[12,13] Andreas P Diamantopoulos,[14]
Haner Direskeneli,[15] Annamaria Iagnocco,[16] Thorsten Klink,[7] Lorna Neill,[17]
Cristina Ponte,[18,19] Carlo Salvarani,[20,21] Riemer H J A Slart,[22,23] Madeline Whitlock,[12]
Wolfgang A Schmidt[24]

Dejaco, Ann Rheum Dis, 2018

Apport de l'imagerie au cours des vascularites des gros vaisseaux

Statement	LoE	LoA
1. In patients with suspected GCA, an early imaging test is recommended to complement the clinical criteria for diagnosing GCA, assuming high expertise and prompt availability of the imaging technique. Imaging should not delay initiation of treatment.	1	9.2 (2.1) 90% ≥8
2. In patients in whom there is a high clinical suspicion of GCA and a positive imaging test, the diagnosis of GCA may be made without an additional test (biopsy or further imaging). In patients with a low clinical probability and a negative imaging result, the diagnosis of GCA can be considered unlikely. In all other situations, additional efforts towards a diagnosis are necessary.	2	9.4 (1.0) 90% ≥8
3. Ultrasound of temporal±axillary arteries is recommended as the first imaging modality in patients with suspected predominantly cranial GCA*. A non-compressible 'halo' sign is the ultrasound finding most suggestive of GCA.	1	9.7 (0.6) 100% ≧8
4. High resolution MRI† of cranial arteries‡ to investigate mural inflammation may be used as an alternative for GCA diagnosis if ultrasound is not available or inconclusive.	2	9.2 (1.1) 90% >8
5. CT† and PET† are not recommended for the assessment of inflammation of cranial arteries.	5	9.5 (1.2) 95% >8
6. Ultrasound, PET, MRI and/or CT may be used for detection of mural inflammation and/or luminal changes in extracranial arteries to support the diagnosis of LV-GCA. Ultrasound is of limited value for assessment of aortitis.	3 (PET and CT) and 5 (MRI and ultrasound)	9.8 (0.6) 100% ≧8
7. In patients with suspected TAK, MRI to investigate mural inflammation and/or luminal changes should be used as the first imaging test to make a diagnosis of TAK, assuming high expertise and prompt availability of the technique.	3	9.1 (1.4) 90% >8
8. PET, CT and/or ultrasound may be used as alternative imaging modalities in patients with suspected TAK. Ultrasound is of limited value for assessment of the thoracic aorta.	3 (CT) and 5 (PET and ultrasound)	9.4 (0.8) 100% ≧8
9. Conventional angiography is not recommended for the diagnosis of GCA or TAK as it has been superseded by the previously mentioned imaging modalities.	5	9.8 (0.6) 100% ≥8
10. In patients with LVV (GCA or TAK) in whom a flare is suspected, imaging might be helpful to confirm or exclude it. Imaging is not routinely recommended for patients in clinical and biochemical remission.	5	9.4 (0.8) 100% ≥8
11. In patients with LVV (GCA or TAK), MRA, CTA and/or ultrasound may be used for long-term monitoring of structural damage, particularly to detect stenosis, occlusion, dilatation and/or aneurysms. The frequency of screening as well as the imaging method applied should be decided on an individual basis.	5	9.3 (1.2) 95% ≥8
12. Imaging examination should be done by a trained specialist using appropriate equipment, operational procedures and settings. The reliability of imaging, which has often been a concern, can be improved by specific training. Suggestions for technical and operational parameters are depicted in box 1.	5	9.8 (0.6) 100% ≧8

→ **Imagerie précoce recommandée**

Dejaco, Ann Rheum Dis, 2018

Apport de l'imagerie au cours des vascularites des gros vaisseaux

Statement	LoE	LoA
1. In patients with suspected GCA, an early imaging test is recommended to complement the clinical criteria for diagnosing GCA, assuming high expertise and prompt availability of the imaging technique. Imaging should not delay initiation of treatment.	1	9.2 (2.1) 90% ≥8
2. In patients in whom there is a high clinical suspicion of GCA and a positive imaging test, the diagnosis of GCA may be made without an additional test (biopsy or further imaging). In patients with a low clinical probability and a negative imaging result, the diagnosis of GCA can be considered unlikely. In all other situations, additional efforts towards a diagnosis are necessary.	2	9.4 (1.0) 90% ≥8
3. Ultrasound of temporal±axillary arteries is recommended as the first imaging modality in patients with suspected predominantly cranial GCA*. A non-compressible 'halo' sign is the ultrasound finding most suggestive of GCA.	1	9.7 (0.6) 100% ≧8
4. High resolution MRI† of cranial arteries‡ to investigate mural inflammation may be used as an alternative for GCA diagnosis if ultrasound is not available or inconclusive.	2	9.2 (1.1) 90% >8
5. CT† and PET† are not recommended for the assessment of inflammation of cranial arteries.	5	9.5 (1.2) 95% >8
6. Ultrasound, PET, MRI and/or CT may be used for detection of mural inflammation and/or luminal changes in extracranial arteries to support the diagnosis of LV-GCA. Ultrasound is of limited value for assessment of aortitis.	3 (PET and CT) and 5 (MRI and ultrasound)	9.8 (0.6) 100% ≧8
7. In patients with suspected TAK, MRI to investigate mural inflammation and/or luminal changes should be used as the first imaging test to make a diagnosis of TAK, assuming high expertise and prompt availability of the technique.	3	9.1 (1.4) 90% >8
8. PET, CT and/or ultrasound may be used as alternative imaging modalities in patients with suspected TAK. Ultrasound is of limited value for assessment of the thoracic aorta.	3 (CT) and 5 (PET and ultrasound)	9.4 (0.8) 100% ≧8
9. Conventional angiography is not recommended for the diagnosis of GCA or TAK as it has been superseded by the previously mentioned imaging modalities.	5	9.8 (0.6) 100% ≥8
10. In patients with LVV (GCA or TAK) in whom a flare is suspected, imaging might be helpful to confirm or exclude it. Imaging is not routinely recommended for patients in clinical and biochemical remission.	5	9.4 (0.8) 100% ≥8
11. In patients with LVV (GCA or TAK), MRA, CTA and/or ultrasound may be used for long-term monitoring of structural damage, particularly to detect stenosis, occlusion, dilatation and/or aneurysms. The frequency of screening as well as the imaging method applied should be decided on an individual basis.	5	9.3 (1.2) 95% ≥8
12. Imaging examination should be done by a trained specialist using appropriate equipment, operational procedures and settings. The reliability of imaging, which has often been a concern, can be improved by specific training. Suggestions for technical and operational parameters are depicted in box 1.	5	9.8 (0.6) 100% ≧8

Echo artérielle si signes céphaliques

Dejaco, Ann Rheum Dis, 2018

Apport de l'imagerie au cours des vascularites des gros vaisseaux

Statement	LoE	LoA
1. In patients with suspected GCA, an early imaging test is recommended to complement the clinical criteria for diagnosing GCA, assuming high expertise and prompt availability of the imaging technique. Imaging should not delay initiation of treatment.	1	9.2 (2.1) 90% ≥8
2. In patients in whom there is a high clinical suspicion of GCA and a positive imaging test, the diagnosis of GCA may be made without an additional test (biopsy or further imaging). In patients with a low clinical probability and a negative imaging result, the diagnosis of GCA can be considered unlikely. In all other situations, additional efforts towards a diagnosis are necessary.	2	9.4 (1.0) 90% ≥8
3. Ultrasound of temporal±axillary arteries is recommended as the first imaging modality in patients with suspected predominantly cranial GCA*. A non-compressible 'halo' sign is the ultrasound finding most suggestive of GCA.	1	9.7 (0.6) 100% ≥8
4. High resolution MRI† of cranial arteries‡ to investigate mural inflammation may be used as an alternative for GCA diagnosis if ultrasound is not available or inconclusive.	2	9.2 (1.1) 90% >8
5. CT† and PET† are not recommended for the assessment of inflammation of cranial arteries.	5	9.5 (1.2) 95% >8
6. Ultrasound, PET, MRI and/or CT may be used for detection of mural inflammation and/or luminal changes in extracranial arteries to support the diagnosis of LV-GCA. Ultrasound is of limited value for assessment of aortitis.	3 (PET and CT) and 5 (MRI and ultrasound)	9.8 (0.6) 100% ≥8
7. In patients with suspected TAK, MRI to investigate mural inflammation and/or luminal changes should be used as the first imaging test to make a diagnosis of TAK, assuming high expertise and prompt availability of the technique.	3	9.1 (1.4) 90% >8
8. PET, CT and/or ultrasound may be used as alternative imaging modalities in patients with suspected TAK. Ultrasound is of limited value for assessment of the thoracic aorta.	3 (CT) and 5 (PET and ultrasound)	9.4 (0.8) 100% ≥8
9. Conventional angiography is not recommended for the diagnosis of GCA or TAK as it has been superseded by the previously mentioned imaging modalities.	5	9.8 (0.6) 100% ≥8
10. In patients with LVV (GCA or TAK) in whom a flare is suspected, imaging might be helpful to confirm or exclude it. Imaging is not routinely recommended for patients in clinical and biochemical remission.	5	9.4 (0.8) 100% ≥8
11. In patients with LVV (GCA or TAK), MRA, CTA and/or ultrasound may be used for long-term monitoring of structural damage, particularly to detect stenosis, occlusion, dilatation and/or aneurysms. The frequency of screening as well as the imaging method applied should be decided on an individual basis.	5	9.3 (1.2) 95% ≥8
12. Imaging examination should be done by a trained specialist using appropriate equipment, operational procedures and settings. The reliability of imaging, which has often been a concern, can be improved by specific training. Suggestions for technical and operational parameters are depicted in box 1.	5	9.8 (0.6) 100% ≥8

Angio-IRM des artères superificelles

Dejaco, Ann Rheum Dis, 2018

Apport de l'imagerie au cours des vascularites des gros vaisseaux

Statement	LoE	LoA
1. In patients with suspected GCA, an early imaging test is recommended to complement the clinical criteria for diagnosing GCA, assuming high expertise and prompt availability of the imaging technique. Imaging should not delay initiation of treatment.	1	9.2 (2.1) 90% ≥8
2. In patients in whom there is a high clinical suspicion of GCA and a positive imaging test, the diagnosis of GCA may be made without an additional test (biopsy or further imaging). In patients with a low clinical probability and a negative imaging result, the diagnosis of GCA can be considered unlikely. In all other situations, additional efforts towards a diagnosis are necessary.	2	9.4 (1.0) 90% ≥8
3. Ultrasound of temporal±axillary arteries is recommended as the first imaging modality in patients with suspected predominantly cranial GCA*. A non-compressible 'halo' sign is the ultrasound finding most suggestive of GCA.	1	9.7 (0.6) 100% ≧8
4. High resolution MRI† of cranial arteries‡ to investigate mural inflammation may be used as an alternative for GCA diagnosis if ultrasound is not available or inconclusive.	2	9.2 (1.1) 90% >8
5. CT† and PET† are not recommended for the assessment of inflammation of cranial arteries.	5	9.5 (1.2) 95% >8
6. Ultrasound, PET, MRI and/or CT may be used for detection of mural inflammation and/or luminal changes in extracranial arteries to support the diagnosis of LV-GCA. Ultrasound is of limited value for assessment of aortitis.	3 (PET and CT) and 5 (MRI and ultrasound)	9.8 (0.6) 100% ≧8
7. In patients with suspected TAK, MRI to investigate mural inflammation and/or luminal changes should be used as the first imaging test to make a diagnosis of TAK, assuming high expertise and prompt availability of the technique.	3	9.1 (1.4) 90% >8
8. PET, CT and/or ultrasound may be used as alternative imaging modalities in patients with suspected TAK. Ultrasound is of limited value for assessment of the thoracic aorta.	3 (CT) and 5 (PET and ultrasound)	9.4 (0.8) 100% ≧8
9. Conventional angiography is not recommended for the diagnosis of GCA or TAK as it has been superseded by the previously mentioned imaging modalities.	5	9.8 (0.6) 100% ≥8
10. In patients with LVV (GCA or TAK) in whom a flare is suspected, imaging might be helpful to confirm or exclude it. Imaging is not routinely recommended for patients in clinical and biochemical remission.	5	9.4 (0.8) 100% ≥8
11. In patients with LVV (GCA or TAK), MRA, CTA and/or ultrasound may be used for long-term monitoring of structural damage, particularly to detect stenosis, occlusion, dilatation and/or aneurysms. The frequency of screening as well as the imaging method applied should be decided on an individual basis.	5	9.3 (1.2) 95% ≥8
12. Imaging examination should be done by a trained specialist using appropriate equipment, operational procedures and settings. The reliability of imaging, which has often been a concern, can be improved by specific training. Suggestions for technical and operational parameters are depicted in box 1.	5	9.8 (0.6) 100% ≧8

Echo, TEP, IRM et/ou TDM si suspicion d'atteinte de gros vaisseaux

Dejaco, Ann Rheum Dis, 2018

Echographie des artères temporales

Anomalies observées au cours de l'ACG

- Signe du halo +++
- Sténoses et occlusion

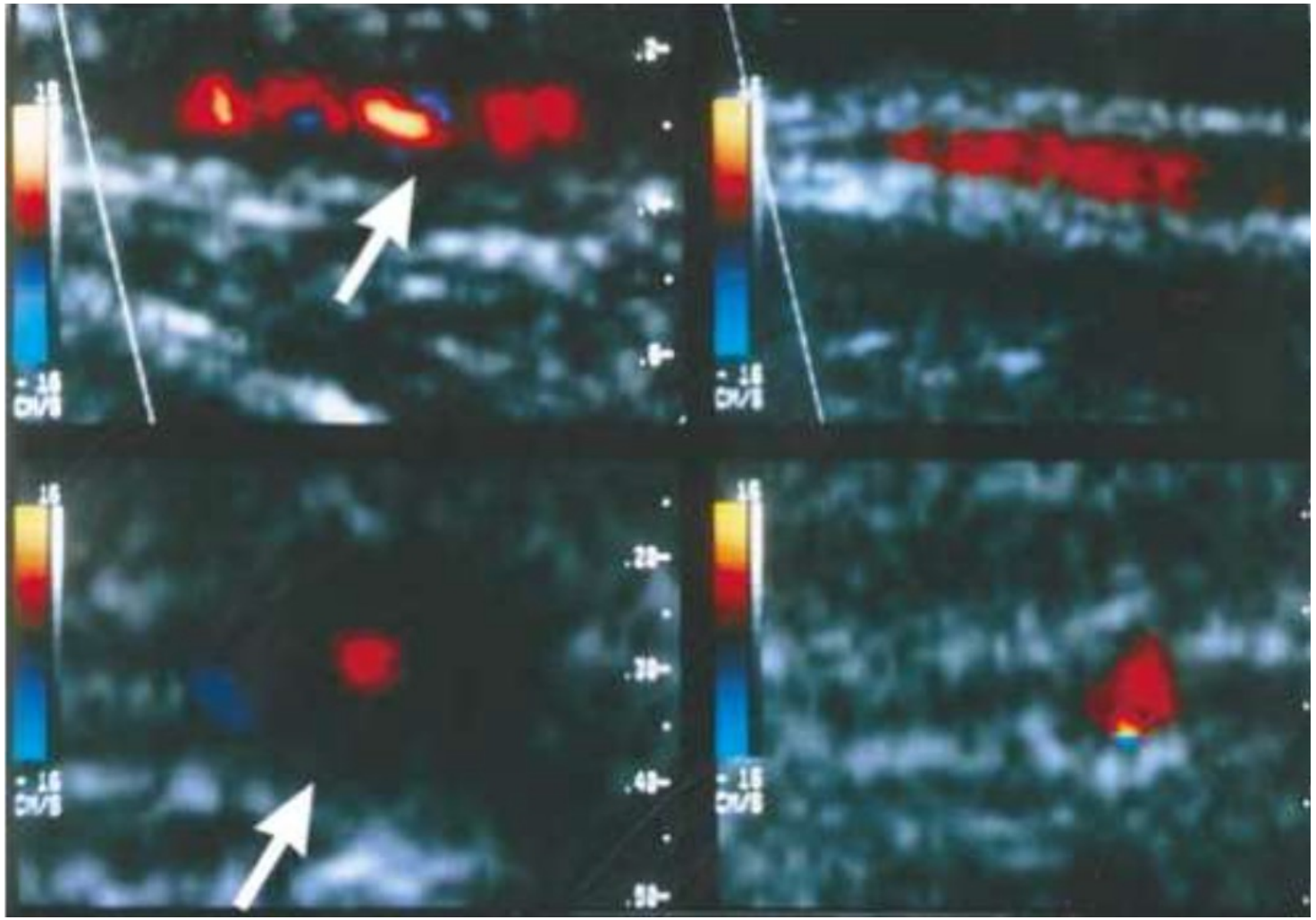
Valeurs diagnostiques

- Halo : Se 10-17%, Sp 100%
- Sténoses et/ou occlusions : Se 50-69°%, Sp 56-60%

Opérateur-dépendant +++

Intérêt ? pour guider BAT

Artérite à cellules géantes *Polyarthrite rhumatoïde*



Maldini, J Rheum, 2010
Schmidt, N Engl J Med, 1997

Echographie des artères temporales

Anomalies observées au cours de l'ACG

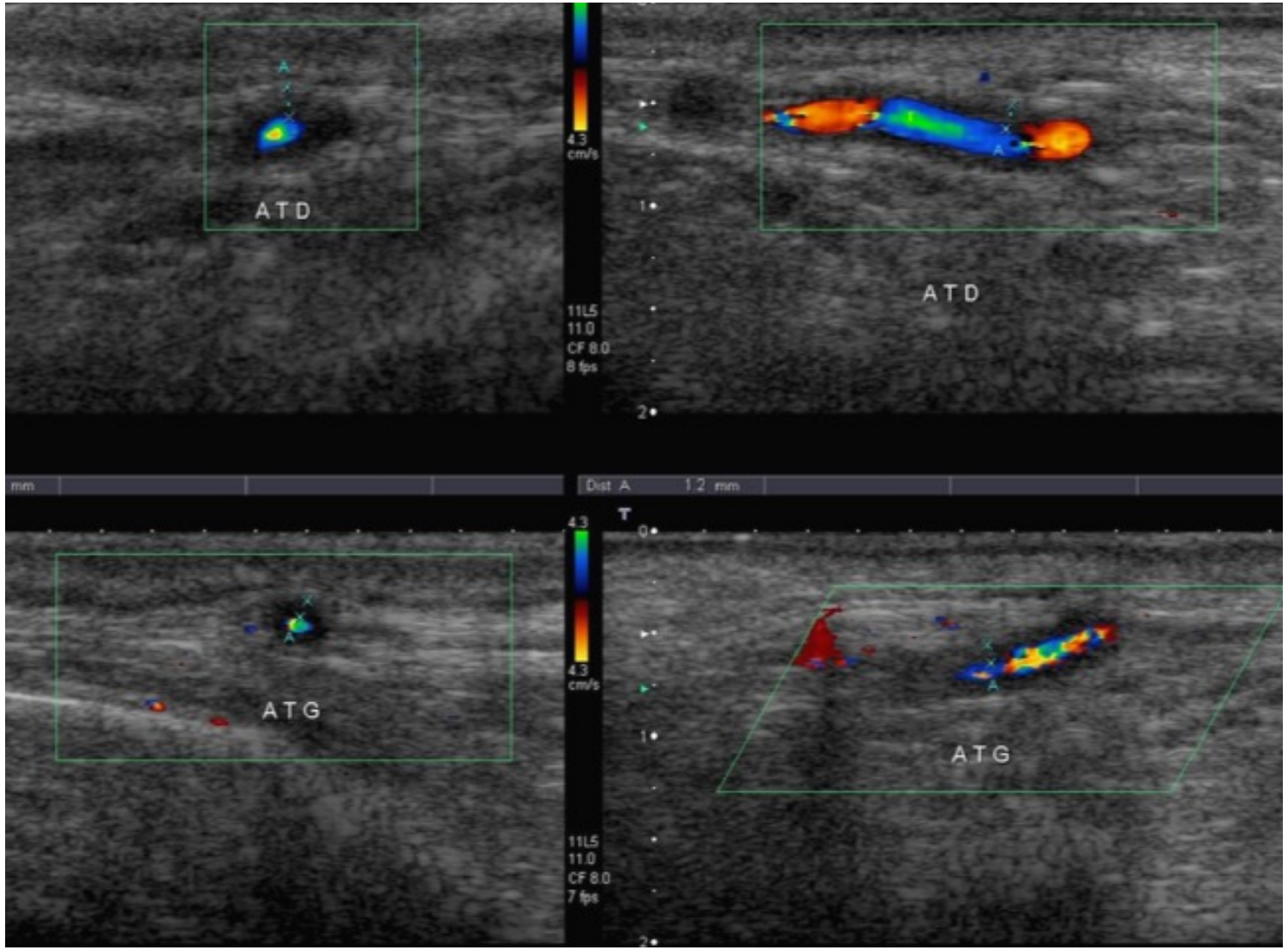
- Signe du halo +++
- Sténoses et occlusion

Valeurs diagnostiques

- Halo : Se 10-17%, Sp 100%
- Sténoses et/ou occlusions : Se 50-69°%, Sp 56-60%

Opérateur-dépendant +++

Intérêt ? pour guider BAT



Maldini, J Rheum, 2010
Schmidt, N Engl J Med, 1997

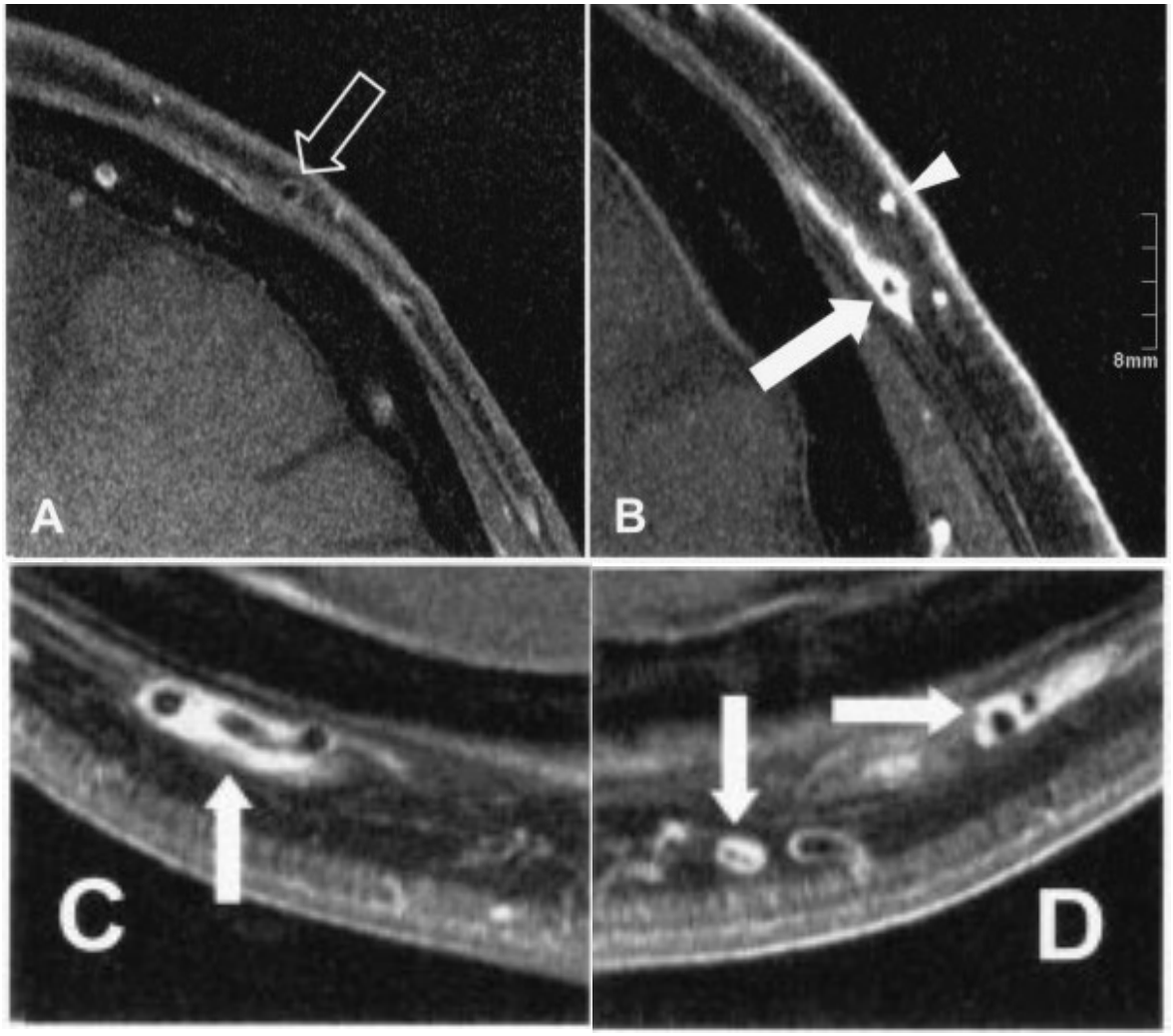
Angio-IRM des artères superificielles

Etude des artères superficielles chez 21 patients

Intérêt de l'IRM de surface à 3T pour le diagnostic d'artérite à cellules géantes

	MRI vs. ACR (n = 21)	MRI vs. TAB (n = 10)
Sensitivity	88.9	100
Specificity	91.7	80
Negative predictive value	91.7	83.3
Positive predictive value	88.9	100

ACG - BAT négative ***ACG - BAT positive***



ACG - Artères occipitales

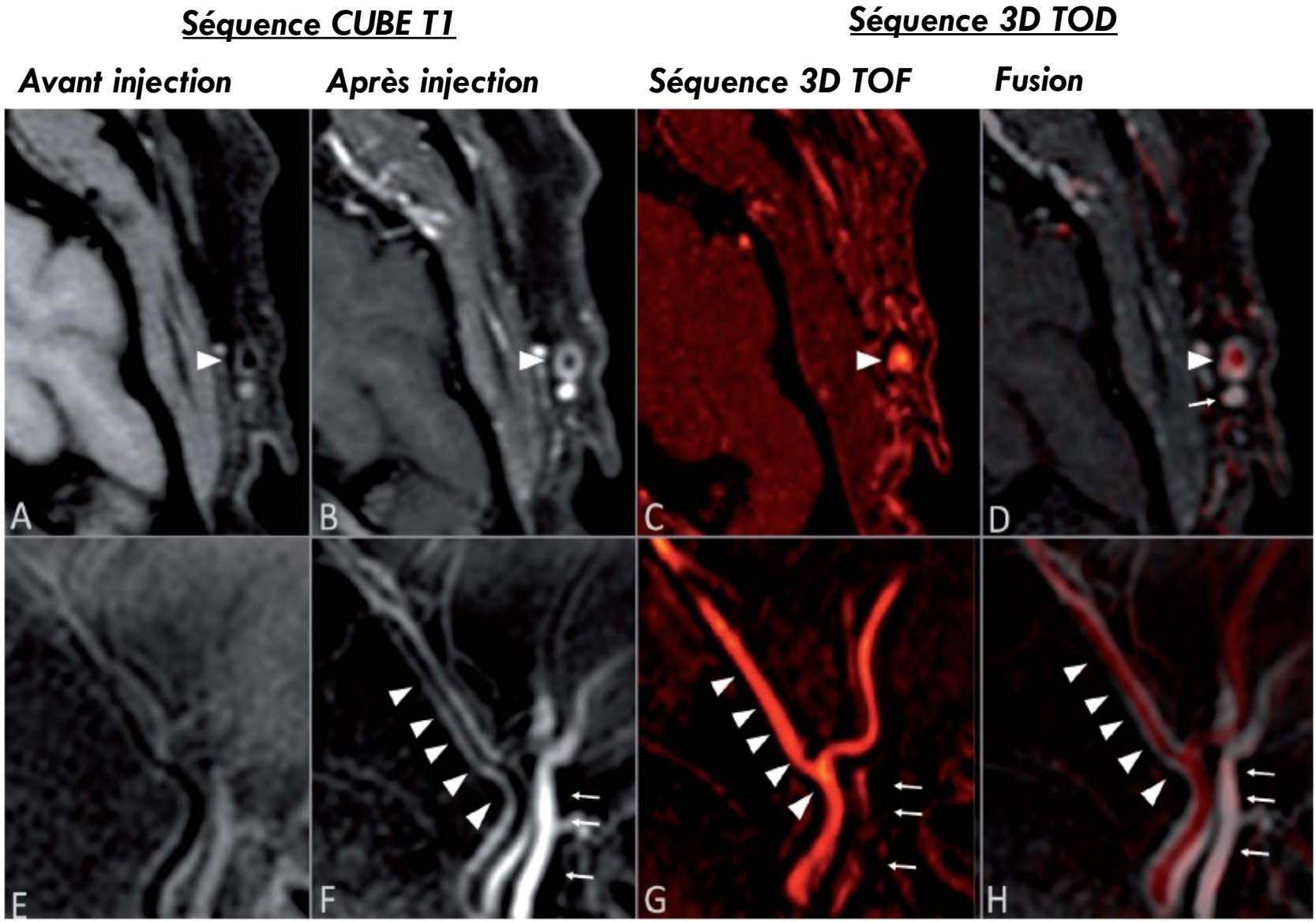
Bley, Arthritis Rheum, 2005

Angio-IRM des artères superificielles

Etude chez 32 patients suspects d'ACG et 10 patients ACG

Intérêt diagnostique de la séquence encéphalique 3D sang noir injectée

Se 80%, Sp 100%, VPP 100%, VPN 92%

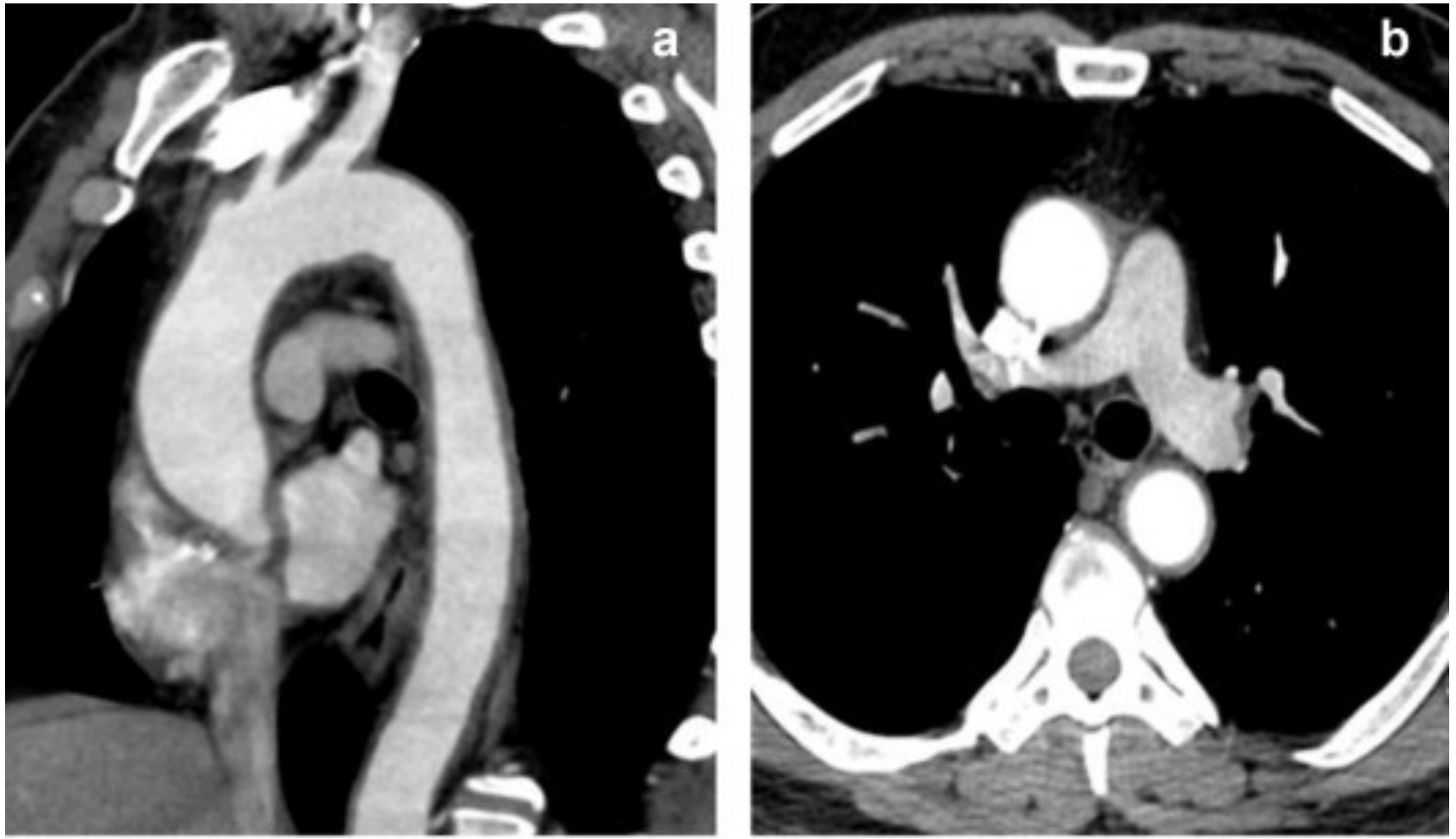


Régent, Clin Exp Rheumatol, 2020

Angio-TDM de l'aorte

Artérite des vaisseaux de gros calibre, avec aortite dans 50%

- Epaississement circonférentiel de l'aorte > 3 mm
- Ectasie/anévrisme aortique
- En l'absence d'athérome et d'infection évolutive



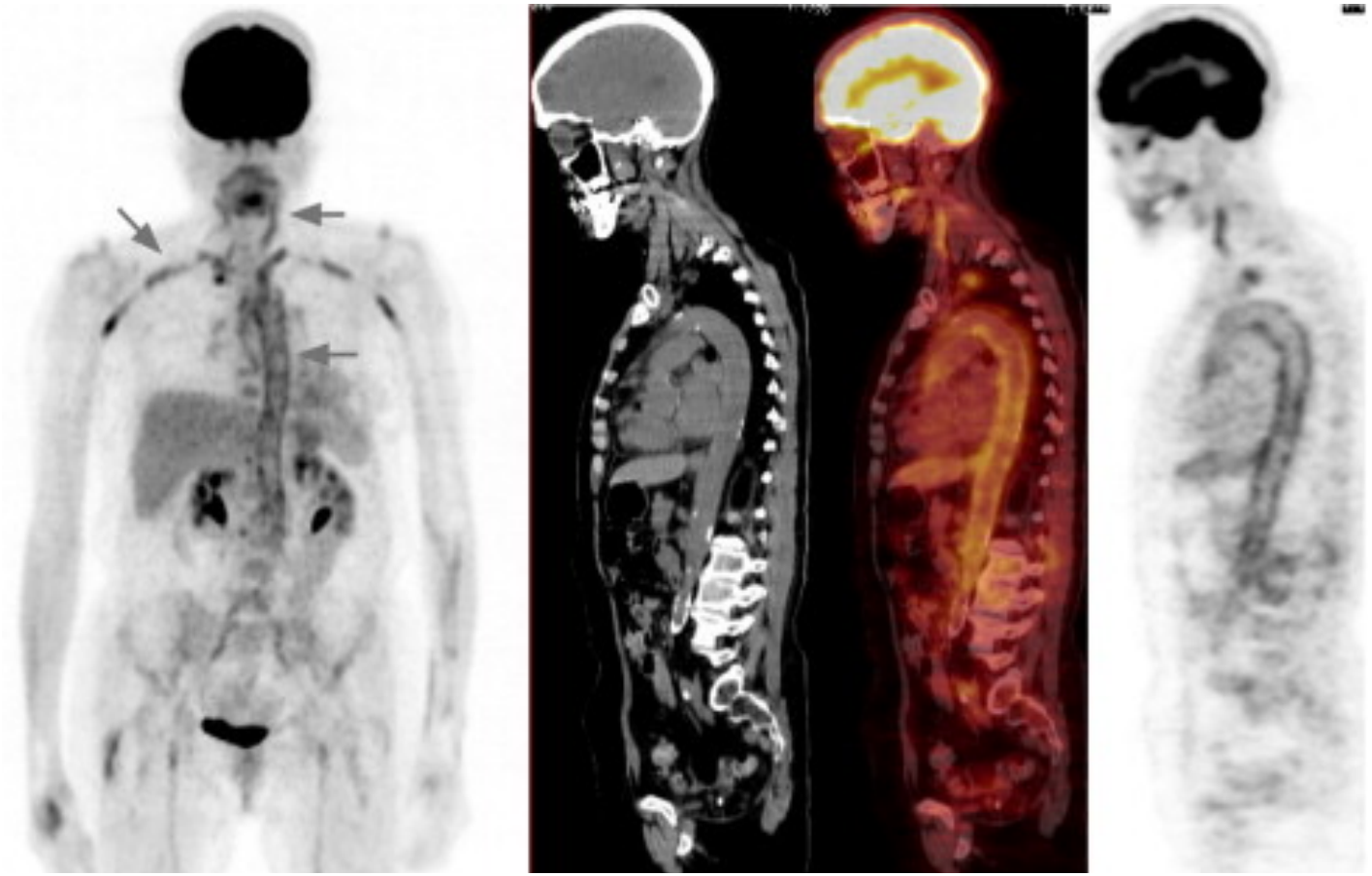
Espitia, Rev Med Interne, 2013
Agard, Arthritis Rheum, 2008

Intérêt diagnostique de la TEP-scanner

Etude prospective de 35 patients avec ACG prouvée histologiquement

Hypermétabolisme vasculaire retrouvé dans 83%

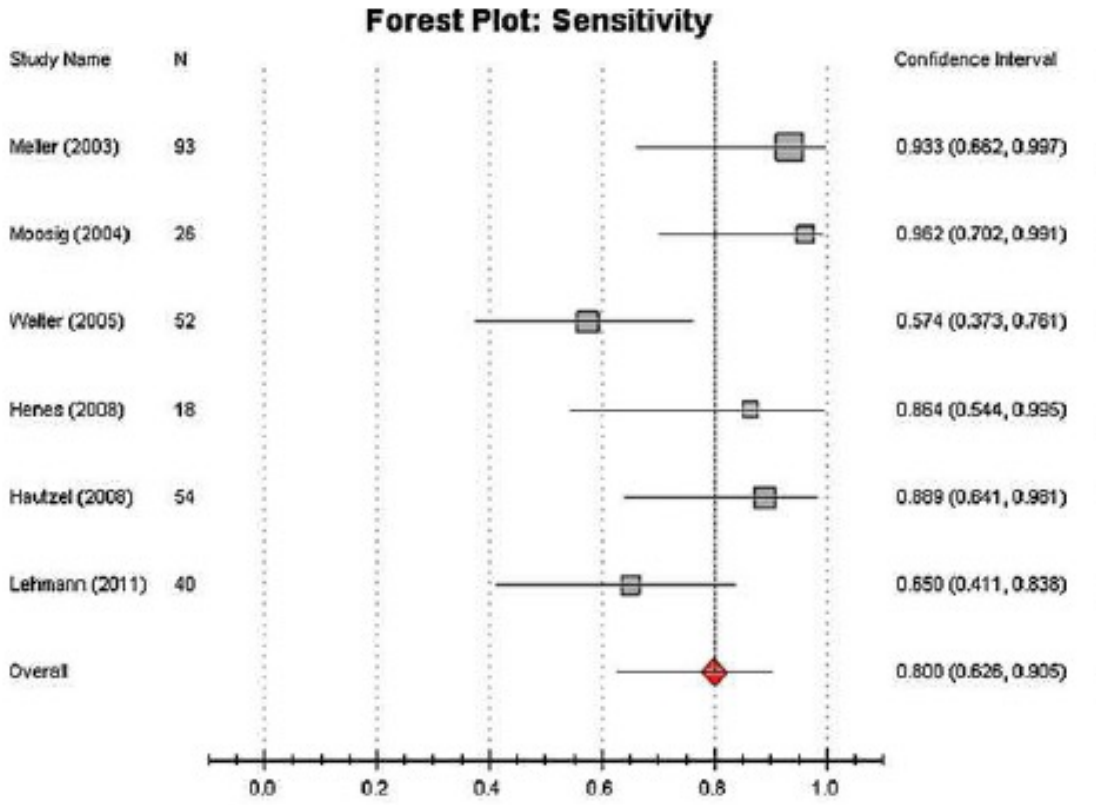
- Artères sous-clavières dans 74%
- Aortique thoracique dans 51%
- Artères fémorales dans 37%



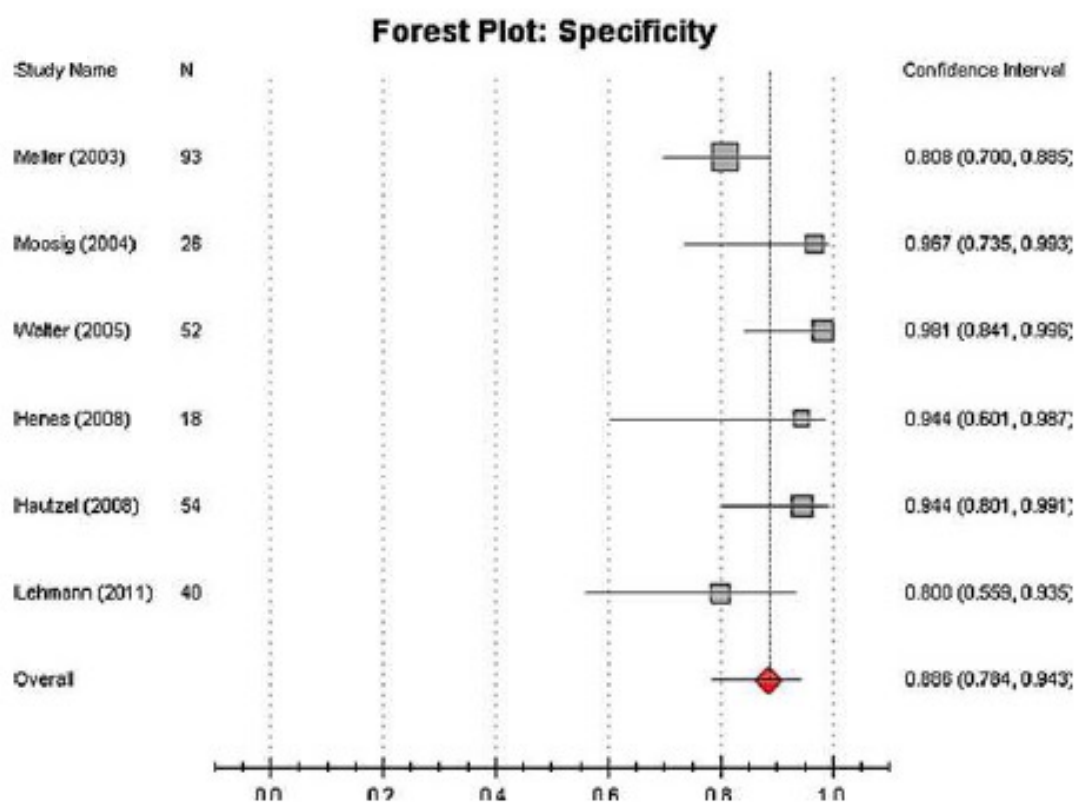
Blockmans, Arthritis Rheum, 2006

Intérêt diagnostique de la TEP-scanner

Sensibilité 80% (63-91)



Spécificité 89% (78-94)



Besson, Eur J Nucl Med Mol Imaging, 2011

Impact pronostique de la TEP-scanner initiale

46 patients évalués par scanner 47 mois après le diagnostic

Hypermétabolisme accru de l'aorte au diagnostic associé significativement à un plus grand diamètre de l'aorte ascendante et de l'aorte descendante

Captation de FDG au niveau de l'aorte thoracique associée à un volume tardif augmenté de l'aorte thoracique

Aortic dimensions (mean ± S.D.)	FDG-uptake negative	FDG-uptake positive	*P*-value
Diameter of the ascending aorta (mm)	37.0 ± 2.8	40.4 ± 6.9	0.025
Diameter of the aortic arch (mm)	30.1 ± 3.6	31.2 ± 3.6	0.281
Diameter of the descending aorta (mm)	30.6 ± 4.0	33.5 ± 5.3	0.044
Volume of the thoracic aorta (cm^3)	253 ± 51	301 ± 81	0.029
Diameter of the suprarenal abdominal aorta (mm)	23.5 ± 3.0	25.5 ± 3.3	0.094
Diameter of the juxtarenal abdominal aorta (mm)	21.7 ± 2.5	22.8 ± 2.1	0.192
Diameter of the infrarenal abdominal aorta (mm)	20.0 ± 2.7	21.1 ± 3.6	0.629
Volume of the abdominal aorta (cm^3)	63 ± 19	64 ± 14	0.925

Blockmans, Rheumatology, 2008

Impact pronostique de la TEP-scanner initiale

Hypermétabolisme vasculaire

- **Aorte thoracique 78%**
- **Artères sous-clavières 72%**
- **Aorte abdominale 55%**
- **Carotides 49%**
- **Axillaires 39%**
- **Fémorales 36%**

Pas d'utilité du TEP-scanner pour le diagnostic de rechute dans 92%

Characteristic	PET+ (n=69)	PET− (n=61)	P
Demographic characteristics			
Women	48 (70)	37 (61)	0.29
Age	69 [50–86]	72 [53–86]	0.13
Cardiovascular risk factors			
Hypertension	27 (39)	33 (54)	0.09
Dyslipidemia	20 (29)	14 (23)	0.43
Diabetes mellitus	7 (10)	10 (15)	0.29
Smoking	11 (16)	15 (25)	0.22
Clinical manifestations			
Fever	20 (29)	25 (41)	0.15
Cephalic manifestations	52 (75)	56 (92)	0.01
Headaches	47 (68)	53 (87)	0.01
Ophthalmologic signs	11 (16)	18 (30)	0.06
Extracephalic manifestations	41 (60)	23 (38)	0.01
Polymyalgia rheumatica	32 (46)	17 (28)	0.03
Vascular bruits	8 (12)	7 (11)	0.99
Limb claudication	4 (6)	1 (2)	0.37
Laboratory tests			
Erythrocyte sedimentation rate, mm	75 [15–130]	80 [14–135]	0.85
C-reactive protein, mg/L	99 [3–390]	123 [11–400]	0.24
Histological results			
Positive TAB	40/67 (60)	35/61 (57)	0.79
Other positive vascular histology	2 (3)	—	—

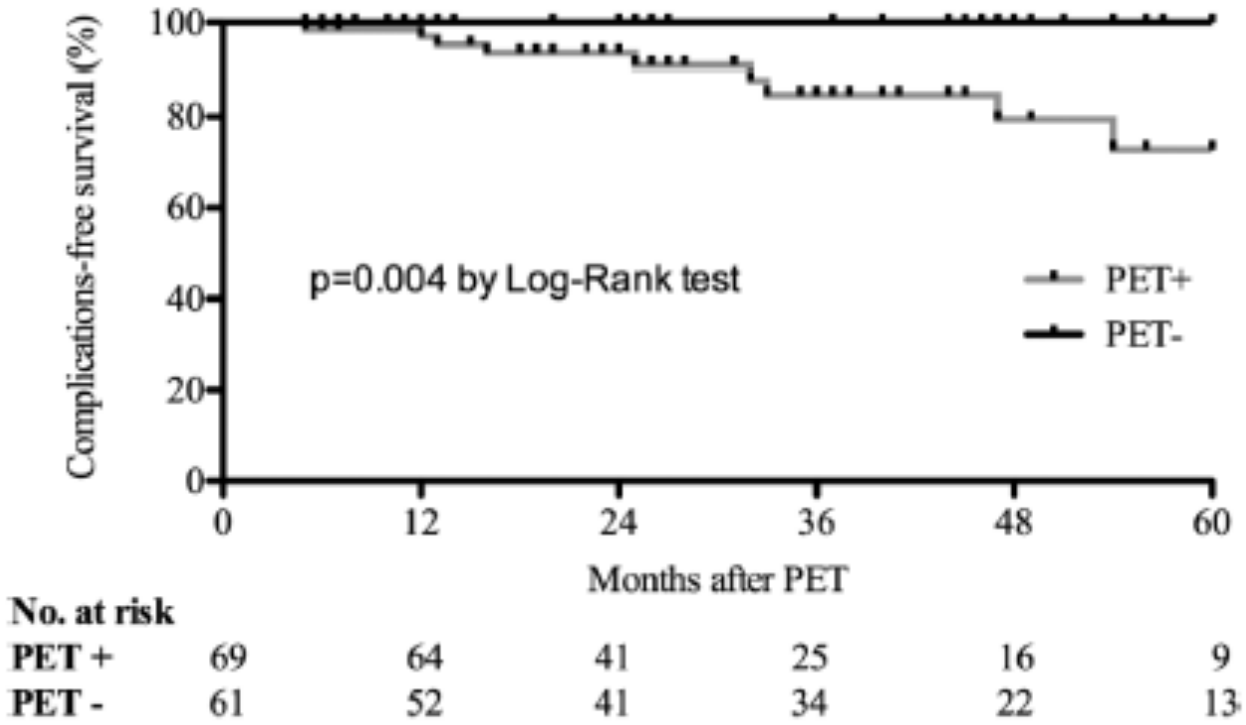
Hypermétabolisme aortique au TEP-scanner associé à un sur-risque de complications aortiques (dissection, …)

De Boysson, Medicine, 2016

Impact pronostique de la TEP-scanner initiale

Hypermétabolisme vasculaire

- **Aorte thoracique 78%**
- **Artères sous-clavières 72%**
- **Aorte abdominale 55%**
- **Carotides 49%**
- **Axillaires 39%**
- **Fémorales 36%**



Pas d'utilité du TEP-scanner pour le diagnostic de rechute dans 92%

9 complications aortiques, toutes avec TEP-scanner positif au niveau aortique

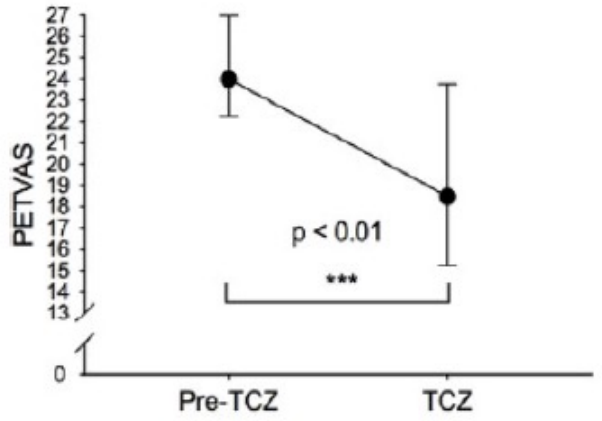
Hypermétabolisme aortique au TEP-scanner associé à un sur-risque de complications aortiques (dissection, …)

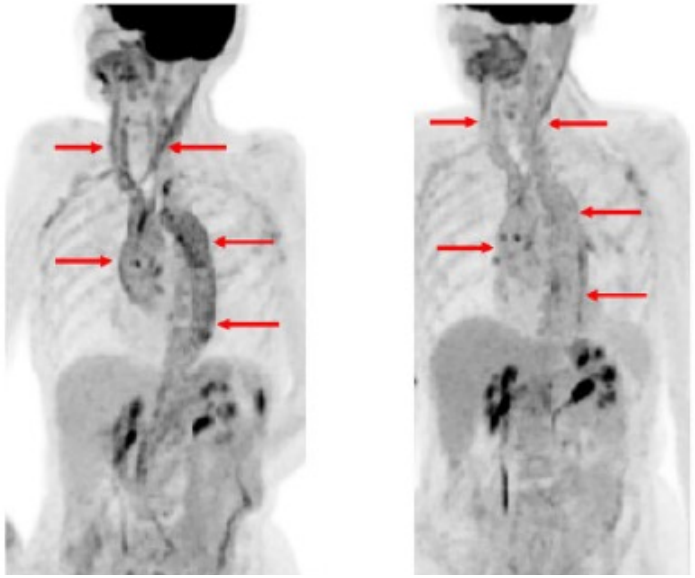
De Boysson, Medicine, 2016

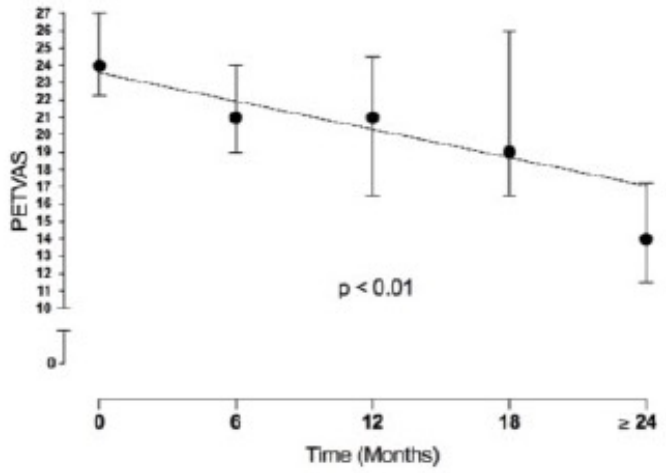
Evolution de la TEP-scanner sous traitement

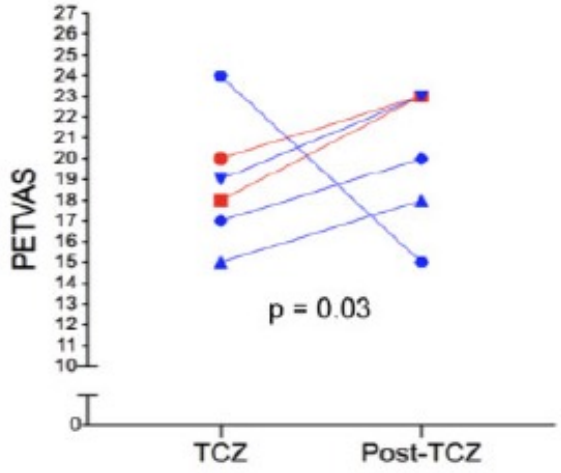
Activité TEP significativement réduite en réponse au TCZ

Amélioration similaire de l'inflammation vasculaire au cours de la première et de la deuxième année de traitement par TCZ





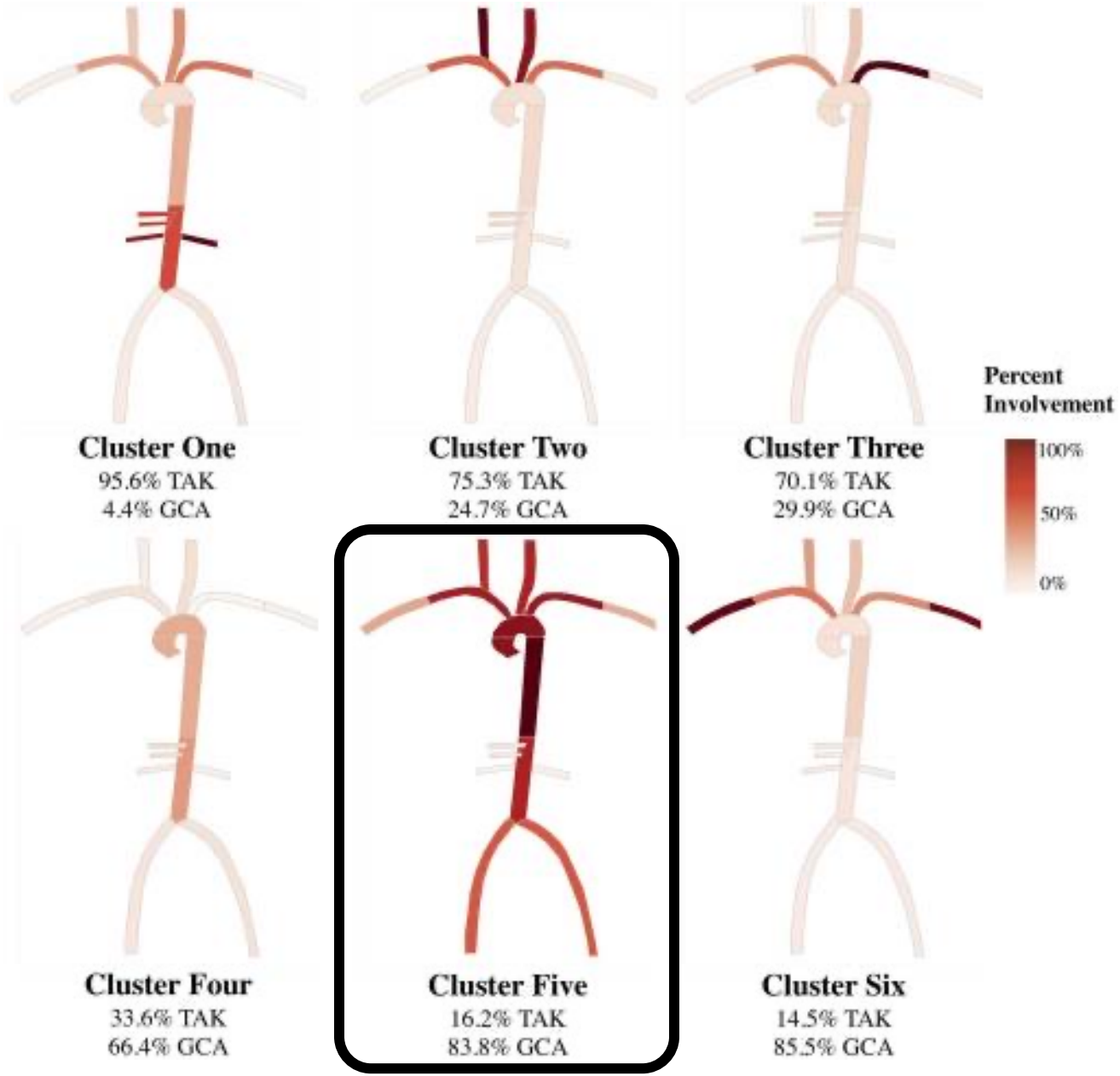


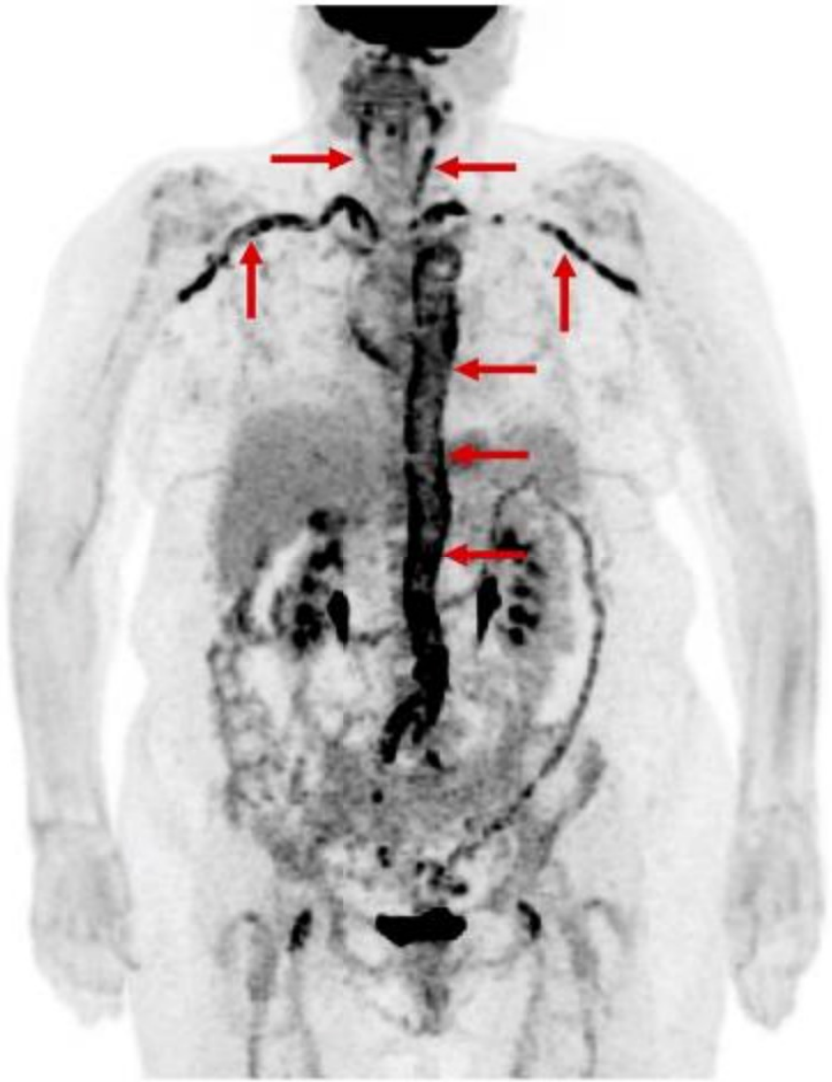


TEP-FDG répétés après arrêt du tocilizumab montrant une aggravation de l'activité TEP chez la plupart des patients

Quinn, Rheumatology, 2021

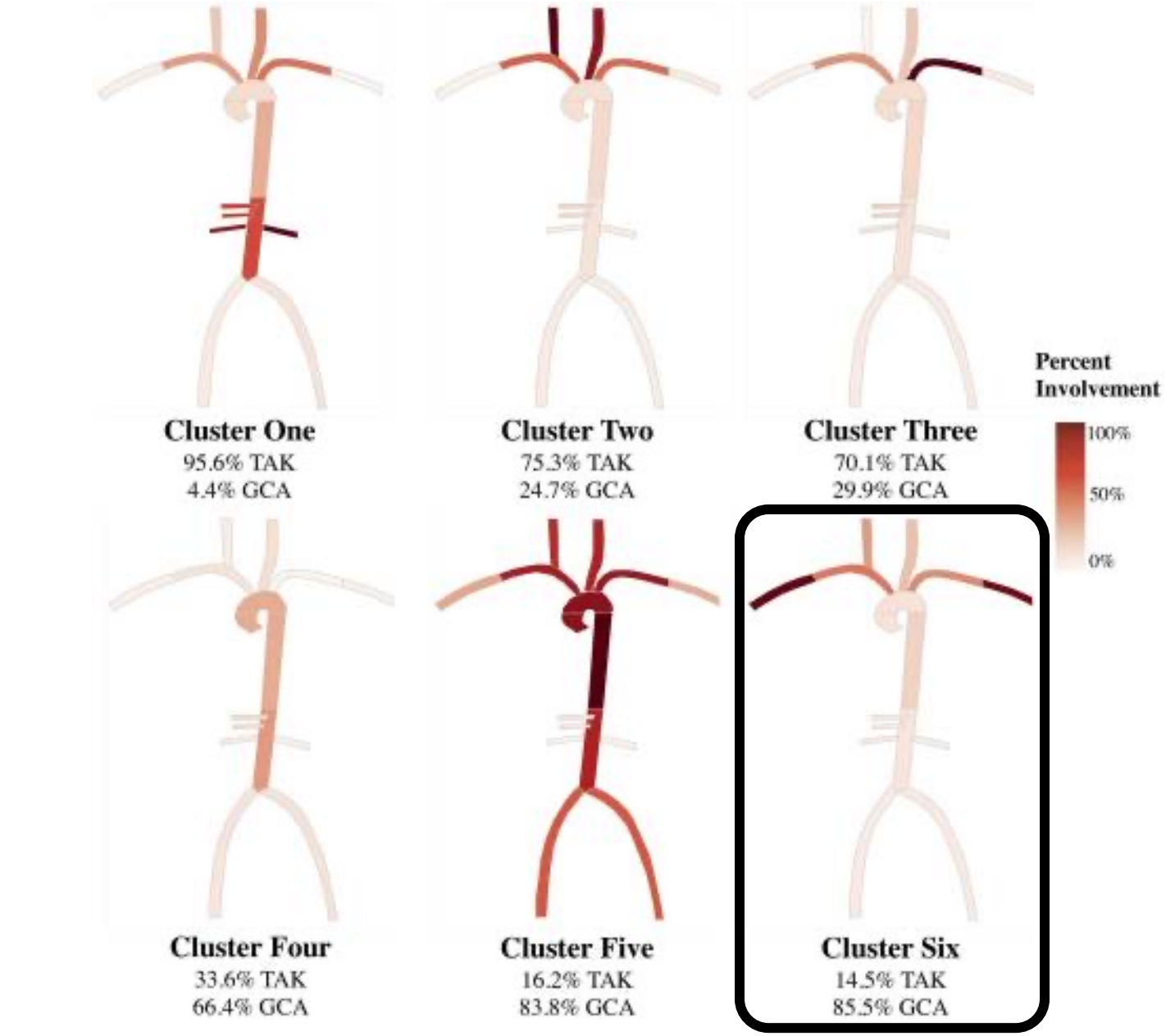
Impact du type d'atteinte vasculaire en imagerie

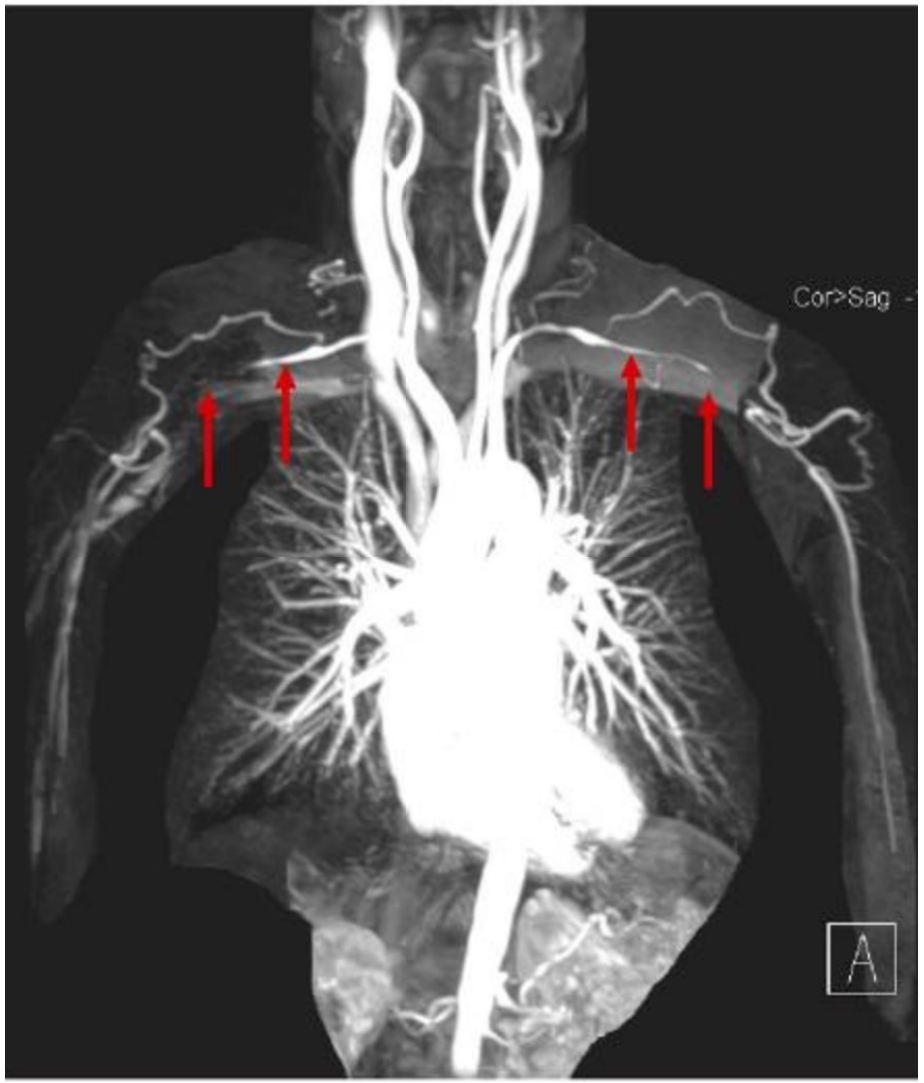




Gribbons, Arthritis Care Res, 2020

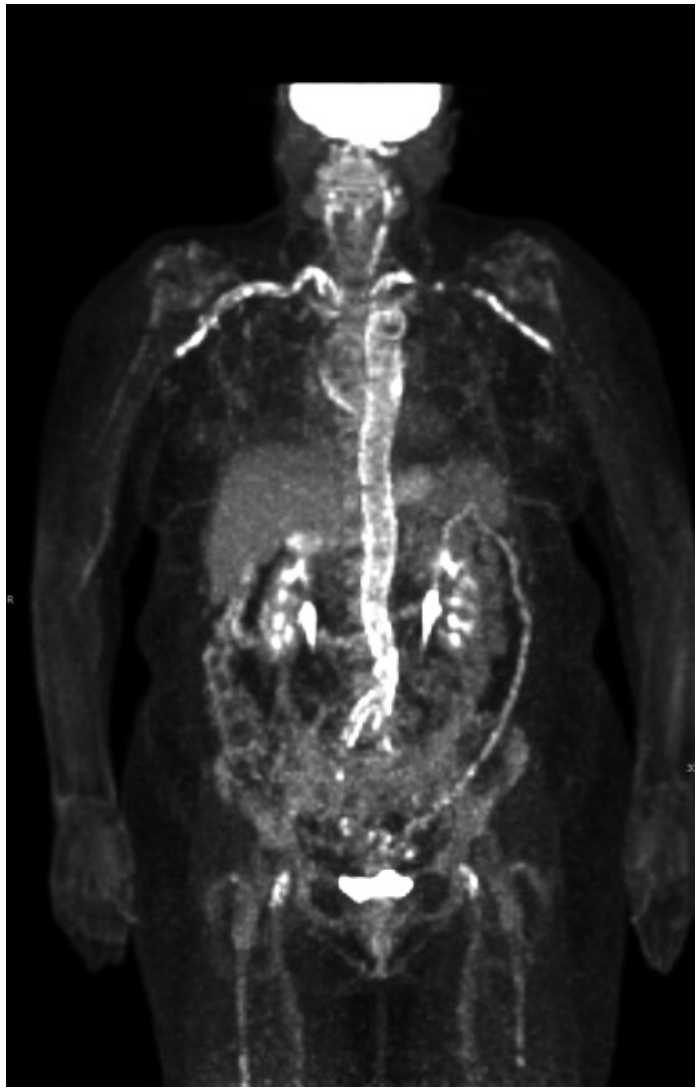
Impact du type d'atteinte vasculaire en imagerie





Gribbons, Arthritis Care Res, 2020

Impact du type d'atteinte vasculaire en imagerie



Activité métabolique au TEP-scanner

Activité métabolique **diffuse** le long de l'aorte et de ses branches au cours de l'**artérite à cellules géantes**

Activité métabolique plus **focale** au cours de l'**artérite de Takayasu**

Grayson, ACR Congress, 2021

Critères de classification de l'ACG

2022 AMERICAN COLLEGE OF RHEUMATOLOGY / EUROPEAN ALLIANCE OF ASSOCIATIONS FOR RHEUMATOLOGY

CLASSIFICATION CRITERIA FOR **GIANT CELL ARTERITIS**

CONSIDERATIONS WHEN APPLYING THESE CRITERIA

- **These classification criteria should be applied to classify the patient as having giant cell arteritis when a diagnosis of large-vessel vasculitis has been made**
- **Alternate diagnoses mimicking vasculitis should be excluded prior to applying the criteria**

CRITERIA ABSOLUTE REQUIREMENTS

Age ≥ 50 years at time of diagnosis	

ADDITIONAL CLINICAL CRITERIA

Morning stiffness in shoulders/neck	+2
Sudden visual loss	+3
Jaw or tongue claudication	+2
New temporal headache	+2
Scalp tenderness	+2
Abnormal examination of the temporal artery[1]	+2

LABORATORY, IMAGING, AND BIOPSY CRITERIA

Maximum ESR ≥ 50 mm/hour or maximum CRP ≥ 10 mg/liter[2]	+2
Positive temporal artery biopsy or halo sign on temporal artery ultrasound[3]	+5
Bilateral axillary involvement[4]	+2
FDG-PET activity throughout aorta[5]	+2

Sum the scores for 10 items, if present. A score of ≥ 6 points is needed for the classification of GIANT CELL ARTERITIS.

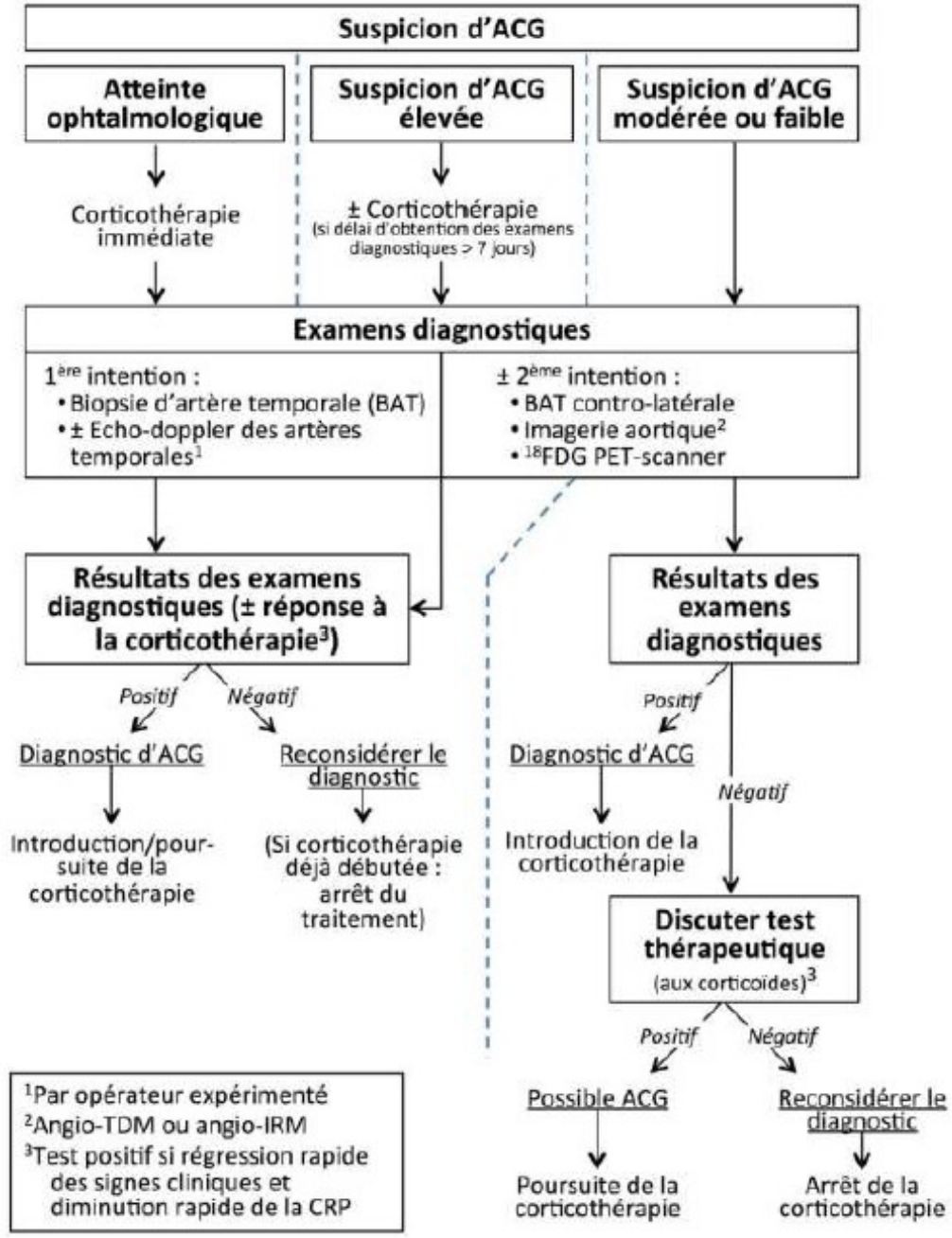
Sensibilité 87%

Spécificité 95%

Grayson, ACR Congress, 2021

Prise en charge thérapeutique

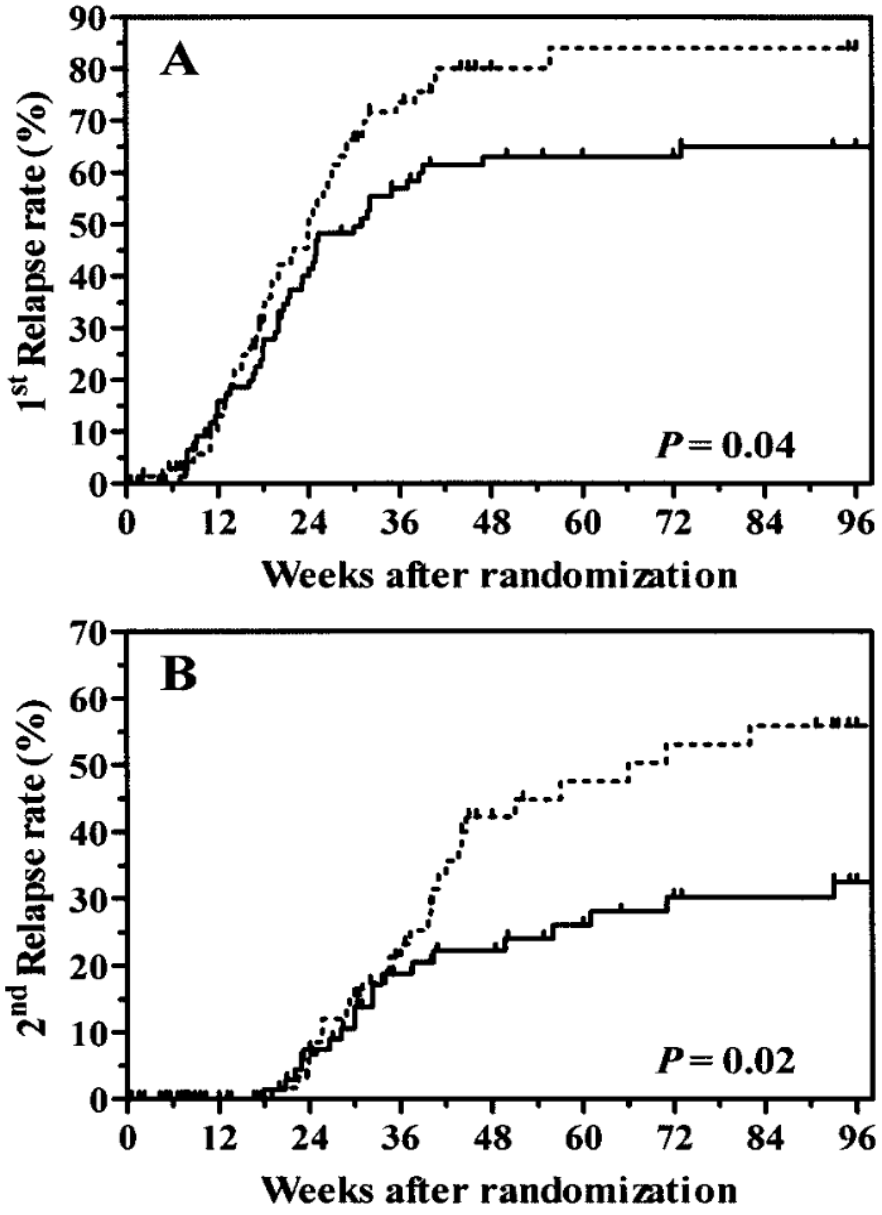
- **Corticothérapie**
  - Prednisone 0,7 mg/kg/j, et 1 mg/kg/j dans les formes compliquées (atteinte oculaire, dilatation, anévrysme ou dissection aortique, ischémie d'un membre)
  - 15 à 20 mg à 3 mois
  - 7,5 à 10 mg à 6 mois
  - 5 mg à 12 mois
  - Puis baisse de 1 mg par mois
- **Bolus de méthylprednisolone ?**
  - Réservés aux formes sévères, mais sans preuve de haut niveau
- **Traitements adjuvants en cas de recherche d'épargne cortisonique (≥ 7,5 mg/j)**



PNDS Artérite à cellules géantes

Place du méthotrexate

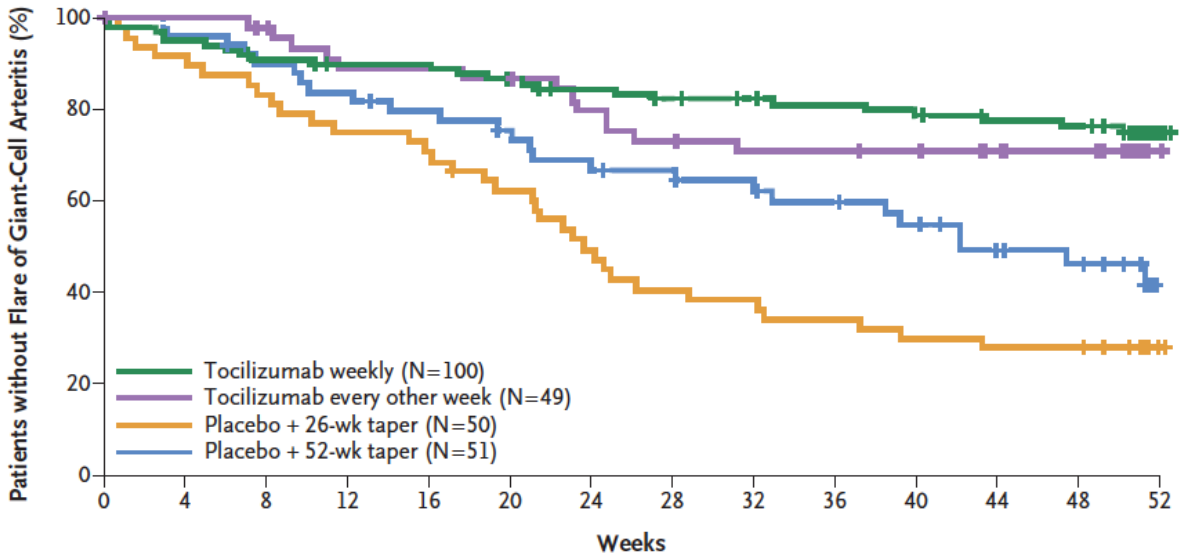
- **Méta-analyse de 3 essais prospectifs randomisés contre placebo**
- Méthotrexate à la dose (faible) de 7,5-15 mg/semaine
- Diminution du risque de 1ère et 2ème rechute
- Diminution de la dose cumulée de corticoïdes
- **En pratique, souvent utilisé en 2ème ligne en cas de cortico-dépendance ou de rechute**



Mahr, Arthritis Rheum, 2007

Place du tocilizumab

- **Effet bénéfique du tocilizumab pendant 12 mois démontré par 2 RCT contre placebo**
- Essai GiACTA : TCZ 162 mg/sem par voie SC
- Essai Villiger et al. : TCZ 8 mg/kg/4 sem. par voie IV



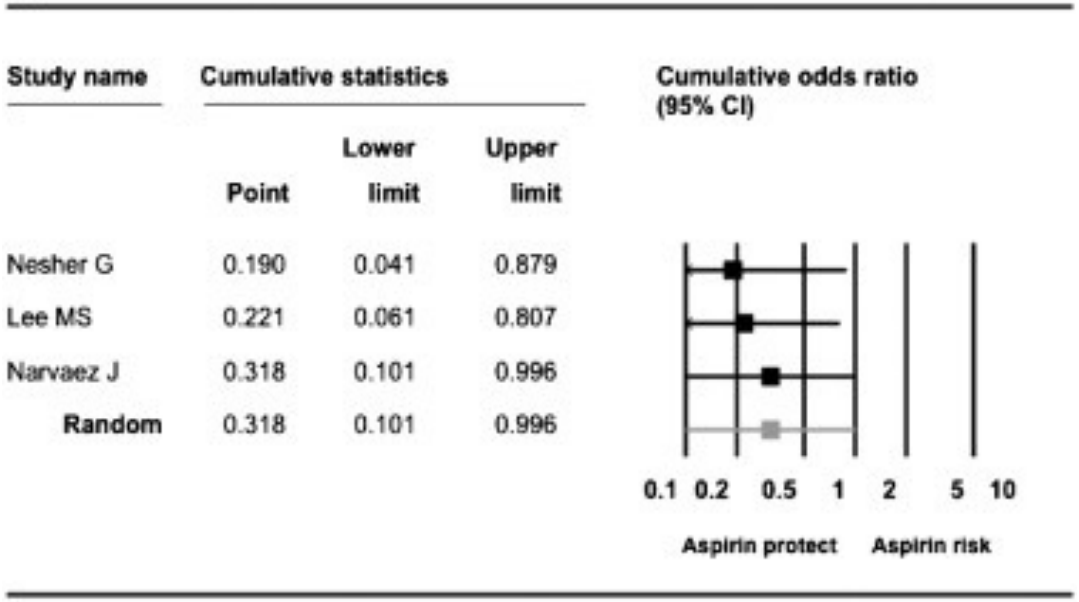
- Profil de sécurité du TCZ satisfaisant (mais attention à la diverticulite et l'absence d'élévation de la CRP +++)
- Arrêt du TCZ après 12 mois s'accompagnant d'un risque de rechute de la maladie chez environ 50% des patients
- **AMM du tocilizumab pour le traitement de l'ACG par voie SC à la posologie 162 mg par semaine**
- Effet d'épargne cortisonique du tocilizumab paraissant supérieur à celui du méthotrexate (essai METOGIA)

Stone, NEJM, 2017
Villiger, Lancet, 2016

Place de l'anti-agrégation plaquettaire

Méta-analyse de 6 études rétrospectives

- Pas de diminution du risque de complications ischémiques <u>avant</u> le diagnostic d'ACG
- Diminution du risque de complications ischémiques <u>après</u> le diagnostic d'ACG
- Pas de majoration du risque hémorragique



PNDS Artérite à cellules géantes

- **Aspirine (75 à 300 mg/jour) en cas d'atteinte ophtalmologique**
- **Sinon : pas de prescription systématique d'aspirine, mais décision au cas par cas**
- Pas de place pour les anti-coagulants

PNDS Artérite à cellules géantes
Martinez-Taboada, Autoimmun Rev, 2014

Quelles perspectives thérapeutiques ?

Sécukinumab (anti-IL-17)

- Essai de phase 2 (***TITAN***) : résultats très positifs (comparables à anti-IL-6R)
- Profil de sécurité satisfaisant
- Essai de phase 3 en cours (***GCAptain***)

Mavrilumab (anti-GM-CSF)

- Essai de phase 2 : résultats positifs
- Profil de sécurité satisfaisant
- Diminution du risque de complications ischémiques <u>après</u> le diagnostic d'ACG

Inhibiteurs de JAK (baricitinib, upadacitinib)

- Essai de phase 2 monocentrique : efficacité chez les rechuteurs
- Profil de sécurité satisfaisant
- Essai de phase 3 en cours avec l'upadacitinib (***Select-GCA***)

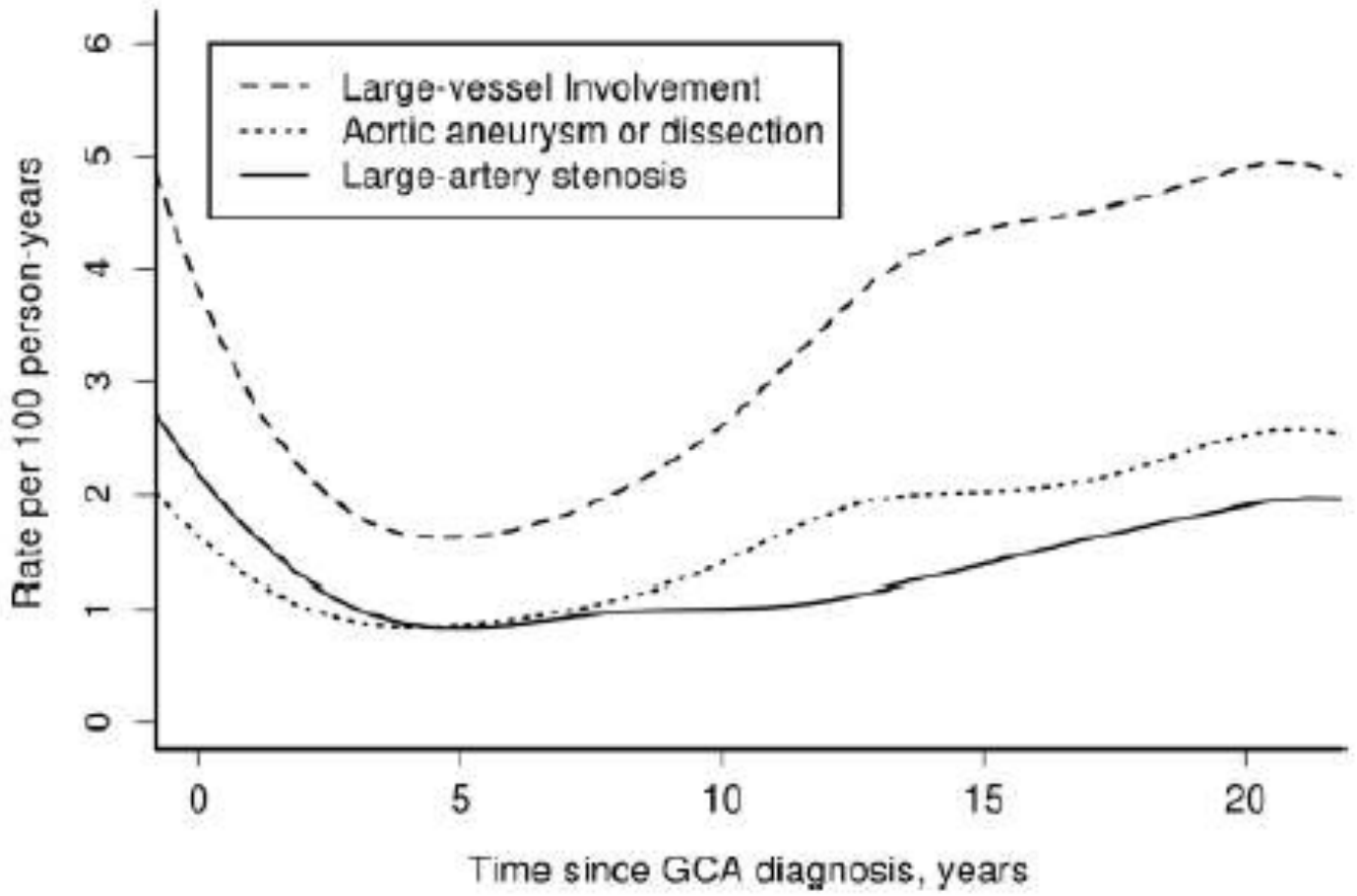
Anti-IL12/IL-23, anti-IL23

- Etude pilote ouverte encourageante
- Essai de phase 2 avec l'ustekinumab (***ULTRA***)
- Essai de phase 2 en cours avec le guselkumab (***THEIA***)

Risque évolutif à long-terme

- Dépistage de l'anévrysme aortique non consensuel, ni sur le délai ni sur les modalités de dépistage

- Proposition d'une imagerie aortique au diagnostique et dans le suivi à 2 et 5 ans puis tous les 5 ans



Kermani, Ann Rheum Dis, 2012

Messages clés

- **Diagnostic clinique le plus souvent simple, mais caractère peu spécifique des formes avec AEG au premier plan**

- **Suspicion diagnostique nécessitant une confirmation diagnostique**
  - Par une biopsie d'artère temporale
  - Par un écho-doppler artériel
  - Par une TEP-TDM (attention : fixation aspécifique de l'athérome)
  - Par un angio-TDM (attention : épaississement d'origine athéromateuse)
  - Par une angio-IRM des artères superificielles

- **Corticothérapie seule restant le traitement de 1ère intention, mais stratégies d'épargne cortisonique +++ (anti-IL-6R, MTX, …)**

Remerciements

CMR
CENTRE MALADIES RARES
VASCULARITES | SCLÉRODERMIES
GOUGEROT-SJÖGREN | LUPUS

CENTRE DE REFERENCE GROUPE I
MALADIES AUTO-IMMUNES ET MALADIES SYSTEMIQUES RARES
Vascularites, Sclerodermies Systemiques,
Histiocytose langerhansienne non pulmonaire de l'adulte

www.maladiesautoimmunes-cochin.org

GFEV | GROUPE FRANÇAIS D'ÉTUDE DES VASCULARITES

www.vascularites.org

Conseil scientifique du GFEV

Collaborateurs et investigateurs du GFEV

Loïc Guillevin, Xavier Puéchal

Patients