Service de pneumologie
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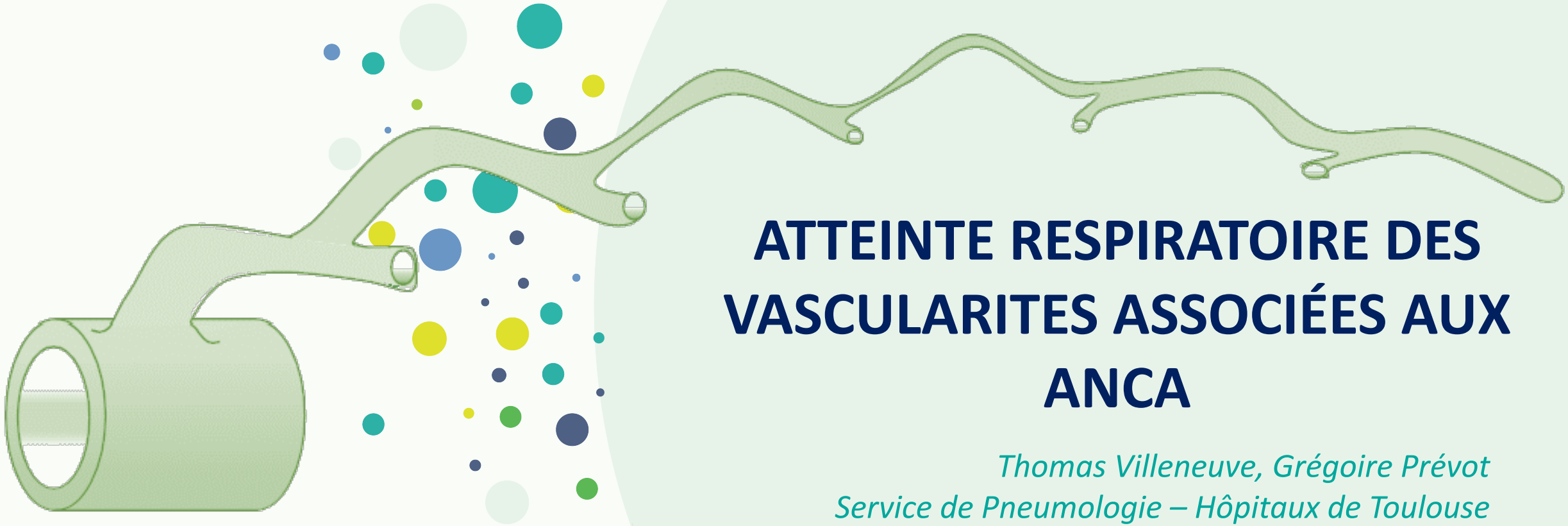

ATTEINTE RESPIRATOIRE DES VASCULARITES ASSOCIÉES AUX ANCA

Thomas Villeneuve, Grégoire Prévot
Service de Pneumologie – Hôpitaux de Toulouse

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Clinique de l'Union

Pôle des voies respiratoires
CHU TOULOUSE
Services pneumologie, chirurgie thoracique, ORL

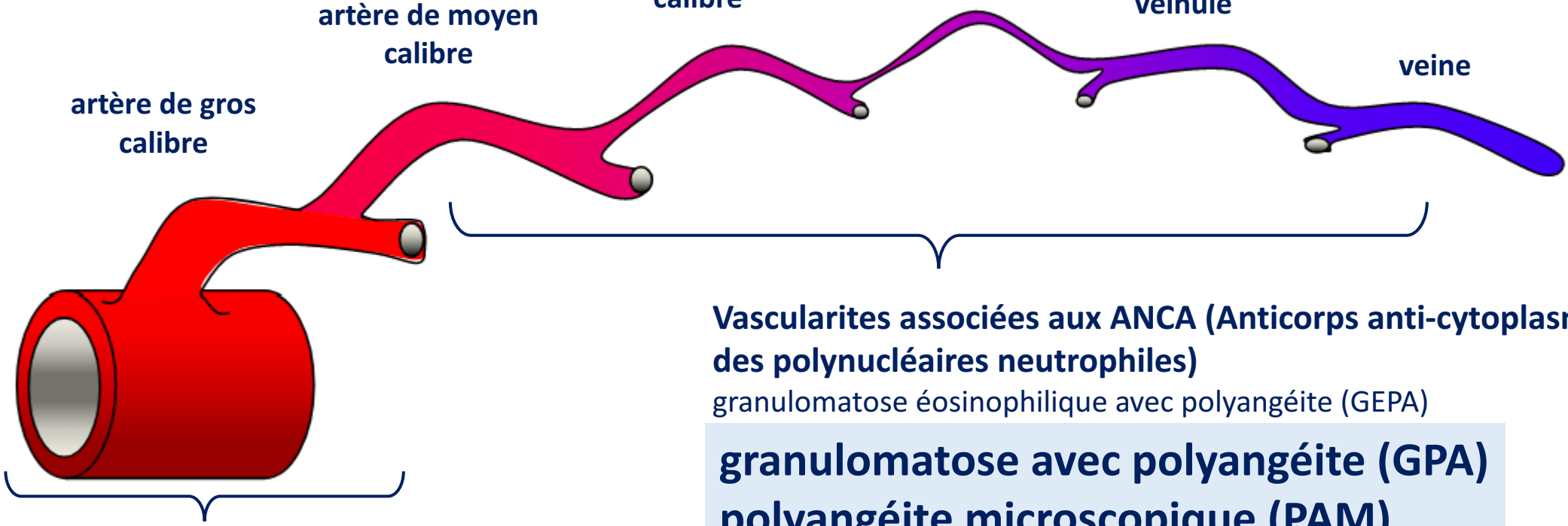
UNIVERSITÉ TOULOUSE III PAUL SABATIER
Université de Toulouse

Classification des vascularites

Vascularite à complexes immuns
Vascularite à IgA, cryoglobulinémique, urticarienne hypocomplémentémique

Périartérite noueuse
Maladie de Kawasaki

Maladie des anticorps anti-membrane basale glomérulaire



Vascularites associées aux ANCA (Anticorps anti-cytoplasme des polynucléaires neutrophiles)
granulomatose éosinophilique avec polyangéite (GEPA)

granulomatose avec polyangéite (GPA)
polyangéite microscopique (PAM)

Artérite de Takayasu
Artérite à cellules géantes

Jennette et al., 2013

Physiopathologie – Vascularite à ANCA

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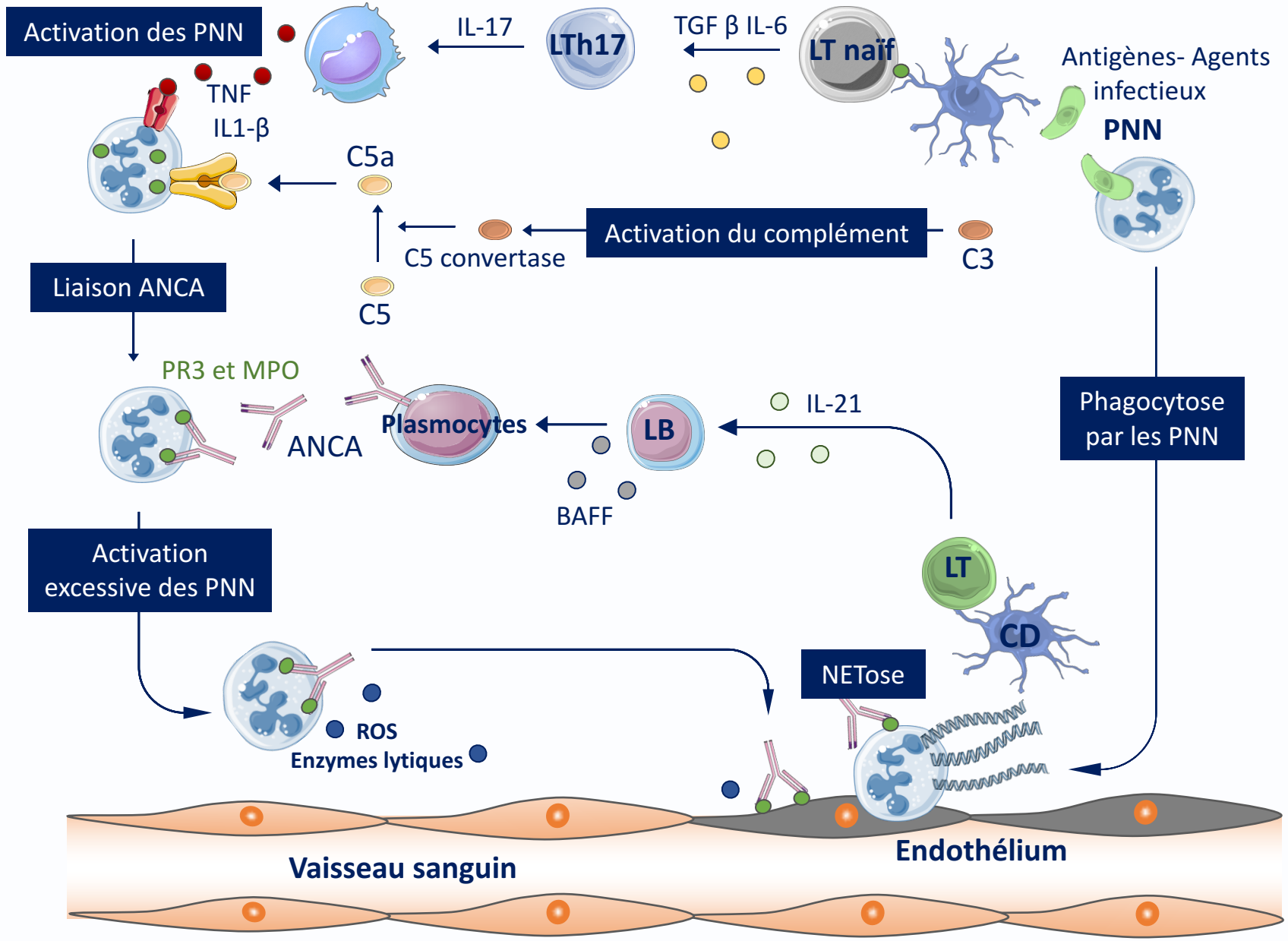
Légende :

PNN : Polynucléaire neutrophile
LTh : Lymphocyte T auxiliaire
LT : Lymphocyte T
LB : Lymphocyte B

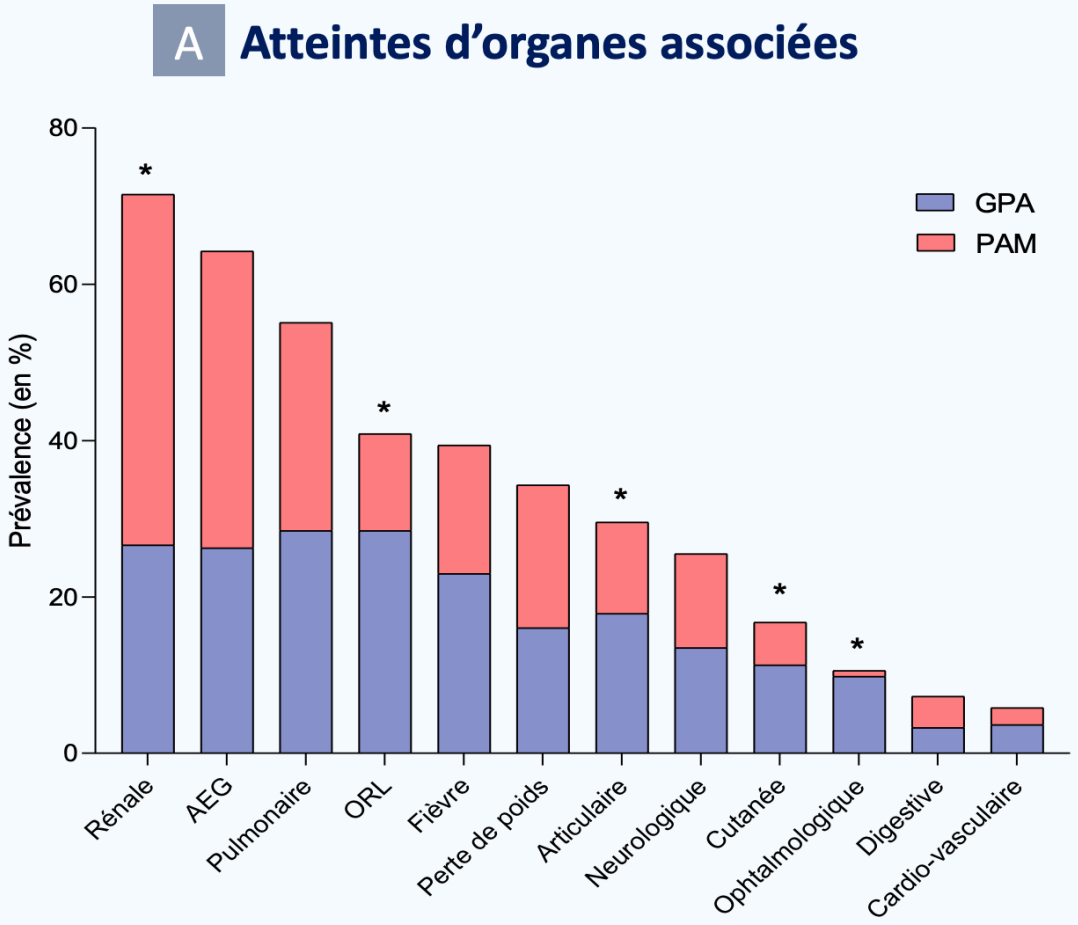
ANCA : Anticorps anti-cytoplasme des polynucléaires neutrophiles
PR3 : Protéinase 3
MPO : Myélopéroxydase

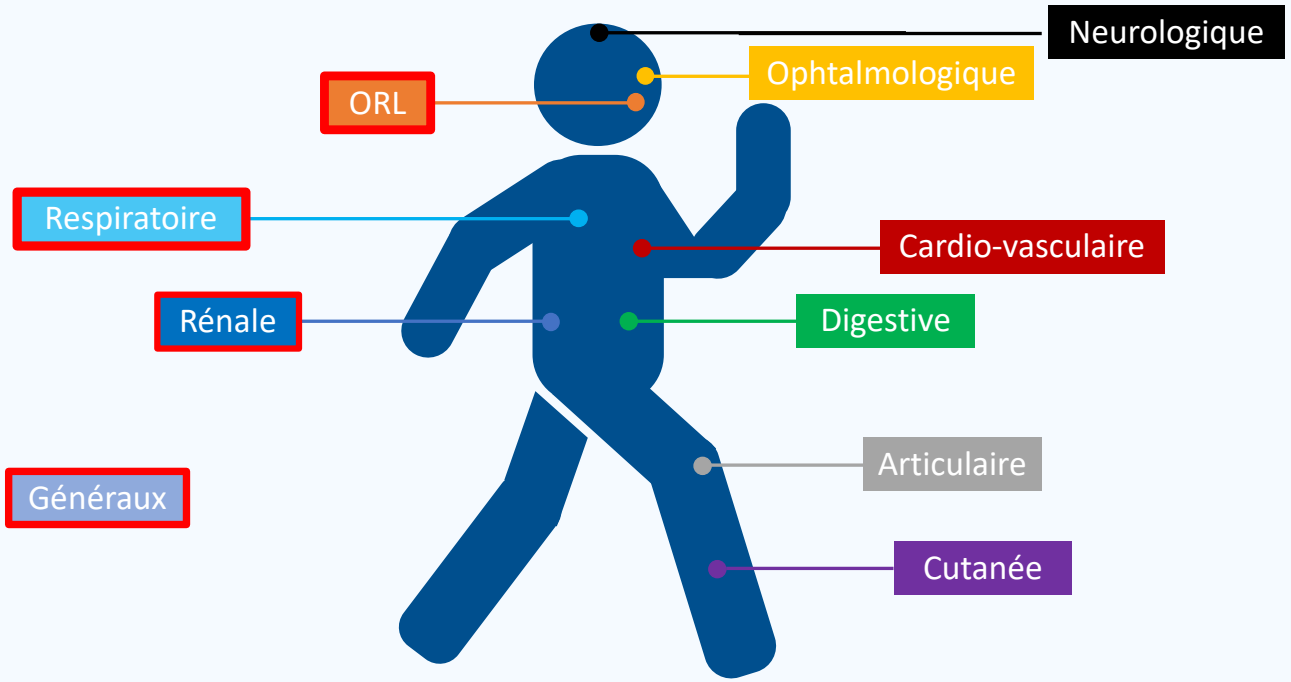
IL- : Interleukine
BAFF :B-cell activating factor
TNF : Tumor Necrosis Factor
ROS : Dérivés réactifs de l'0_2
NET : Neutrophil extracellular traps



D'après Nakazawa et al., 2019

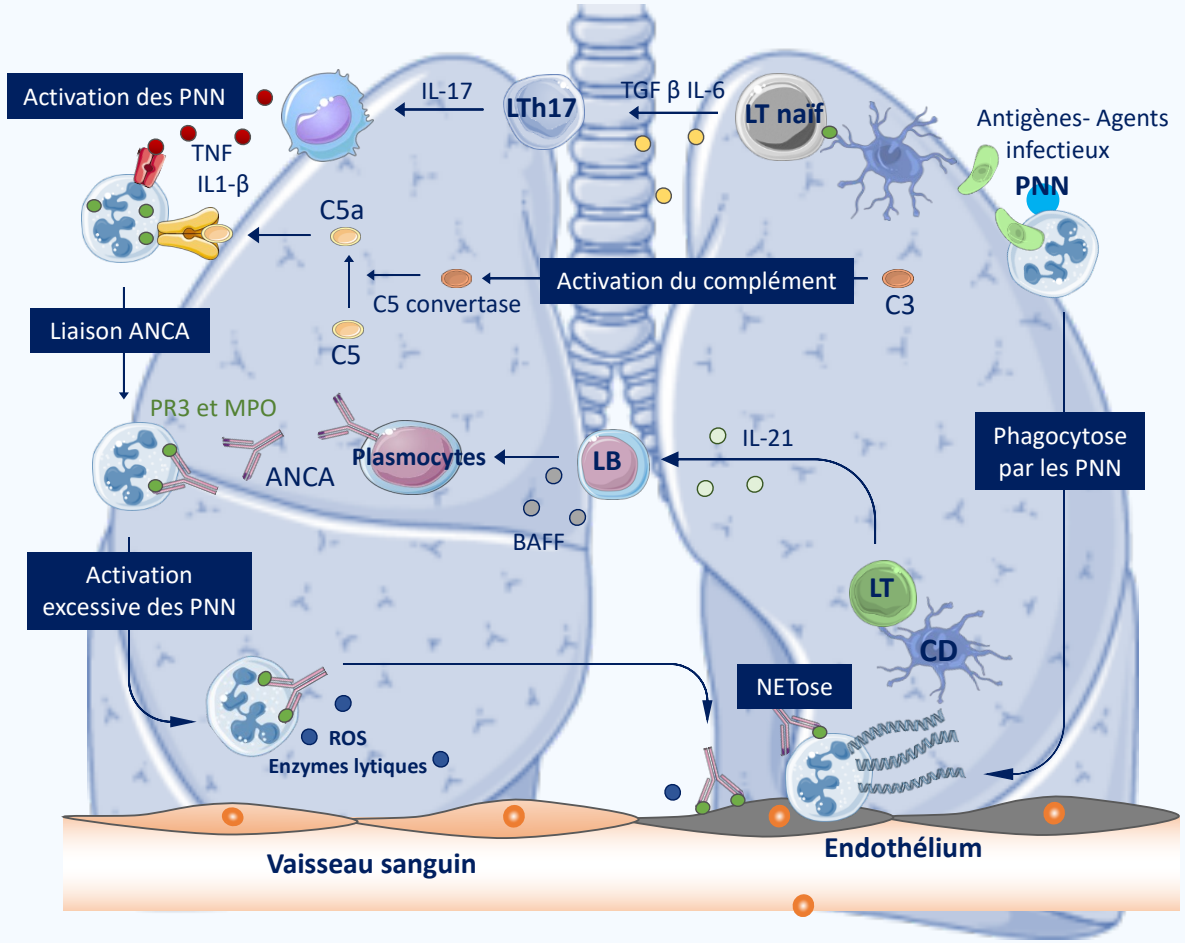
Tropisme d'organe des Vascularites à ANCA





T Villeneuve, thèse 2021

Clinique- Vascularite à ANCA



Vascularite

Hémorragie intra-alvéolaire

Granulomatose

Atteinte nodulaire

Atteinte trachéo-bronchique

Pathologie Infiltrante

Fibrose pulmonaire

PNP à Eosinophiles

Atteintes Respiratoires des vascularites à ANCA

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The Respiratory System in Autoimmune Vascular Diseases

Respiration

Thematic Review Series

Respiration 2018;96:12–28
DOI: 10.1159/000486899

Published online: July 4, 2018

Mouhamad Nasser[a] Vincent Cottin[a, b]

Très fréquent

Fréquent

Peu fréquent

Rare

Très rare

Vasculitis	Lung parenchymal	Bronchial	Pleural	Pulmonary artery	Laryngeal	Nasal/sinus
Granulomatosis with polyangiitis	Cavitated and noncavitated nodule/mass Consolidations Ground glass opacities Alveolar hemorrhage	Stenosis Bronchiectasis	Pleural effusion Pleural nodules Pneumothorax	Inflamed vessel	Strictures and stenosis (usually subglottic) Ulcers Inflamed mucosa	Sinusitis Nasal mucosa ulcers Bone deformity Saddle nose Nasal mass
Eosinophilic granulomatosis with polyangiitis	Consolidations Ground glass opacities Thickening of interlobular septa	Asthma Stenosis Bronchiectasis	Pleural effusion	Not reported	Not reported	Nasal polyposis Eosinophilic rhinitis Chronic/recurrent rhinosinusitis
Microscopic polyangiitis	Alveolar hemorrhage Subpleural reticulations Usual interstitial pneumonia pattern	Not reported	Not reported	Not reported	Not reported	Sinusitis and nasal mucosa ulcers

Atteintes Respiratoires des vascularites à ANCA

Service de pneumologie
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Pôle des voies respiratoires

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Microscopic polyangiitis	Alveolar hemorrhage Subpleural reticulations Usual interstitial pneumonia pattern	Not reported	Not reported	Not reported	Not reported	Sinusitis and nasal mucosa ulcers

TDM thoracique et suspicion de Vascularite à ANCA

Hémorragie intra-alvéolaire

Critères diagnostiques HIA dans l'étude PEVIXAS

DAH due to AAV

Compatible chest X-ray or CT

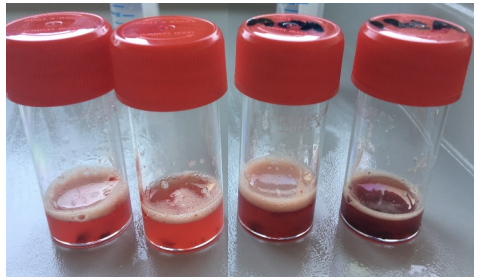
« Diffuse Pulmonary Infiltrate »

+

Absence of alternative explanation for pulmonary infiltrates

+

At least one of the following:
1) Evidence of DAH by bronchoscopy
2) Observed hemoptysis
3) Unexplained anemia (<10 g/dL) or drop in hemoglobin (>1 g/dL)
4) Elevated diffusing capacity of carbon monoxide (DLCO)



RadioGraphics
CHEST IMAGING
Common and Uncommon Manifestations of Wegener Granulomatosis at Chest CT: Radiologic-Pathologic Correlation[1]

Table 4
Differential Diagnosis for Airspace Opacities

Disease	Presentation	Distribution	Adenopathy	Tree-in-Bud Opacities	Cavitation
Wegener granulomatosis	Most often acute	Multifocal, bilateral perihilar and peribronchovascular	Rare	Rare	Rare
Bacterial pneumonia	Acute	Lobar and patchy	Reactive	Yes	Rare
Aspiration	Most often acute	Dependent, associated with bronchiectasis in lower lobes	Reactive	Yes	Some-times
Organizing pneumonia	Chronic	Multifocal, peripheral, migra-tory; atoll sign	Rare	No	No
Neoplasm (adenocarci-noma versus adeno-carcinoma in situ)	Chronic	Uni- or multifocal, ranging from ground-glass opacity to consolidation	Yes	No	Pseudo-cavita-tion

Autres éléments d'orientation:

- Pour HIA :
  - Verre dépoli extensif avec respect du sous-pleural
  - Diminution/majoration spontanée des opacités
- Pour VAA : association lésionnelle

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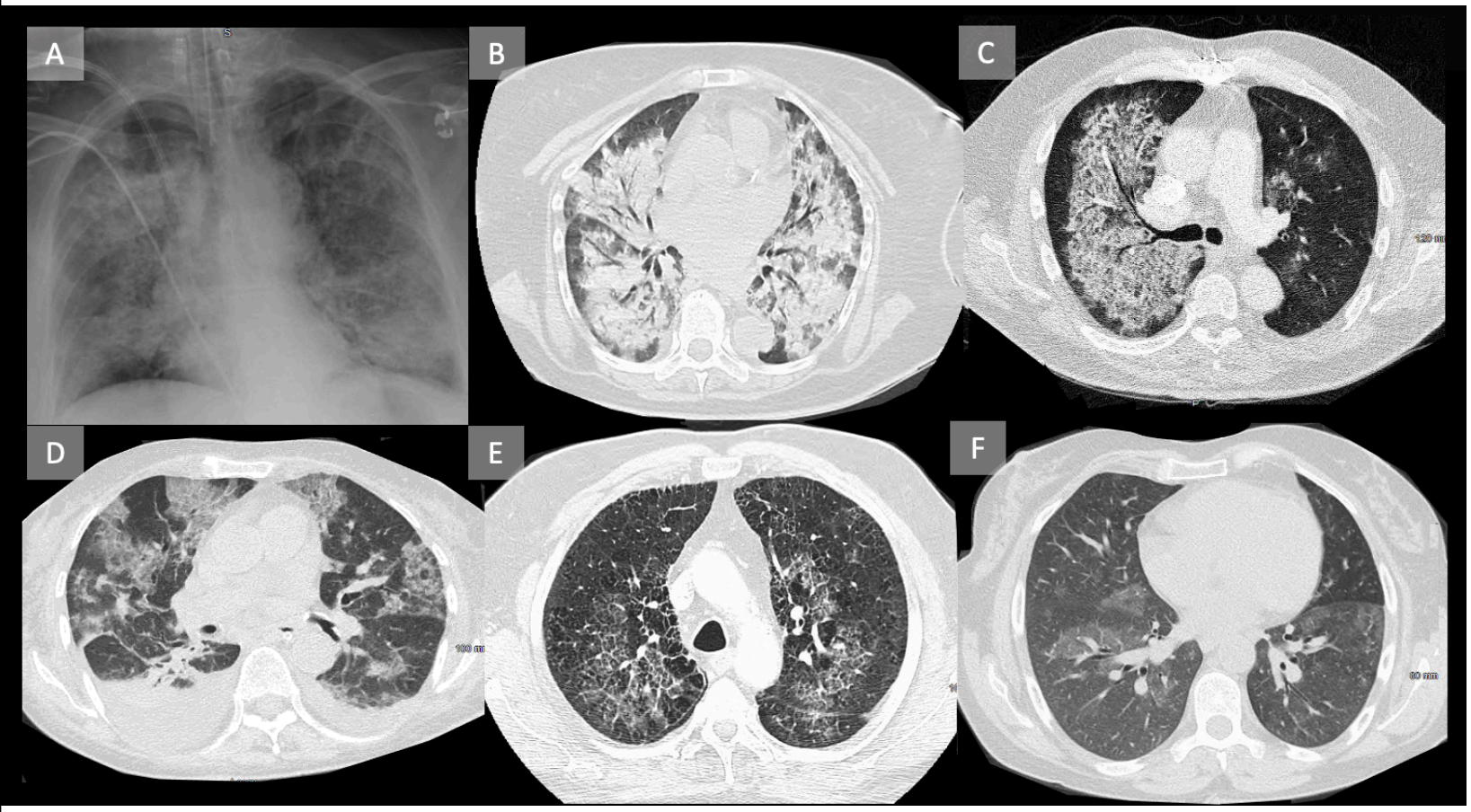
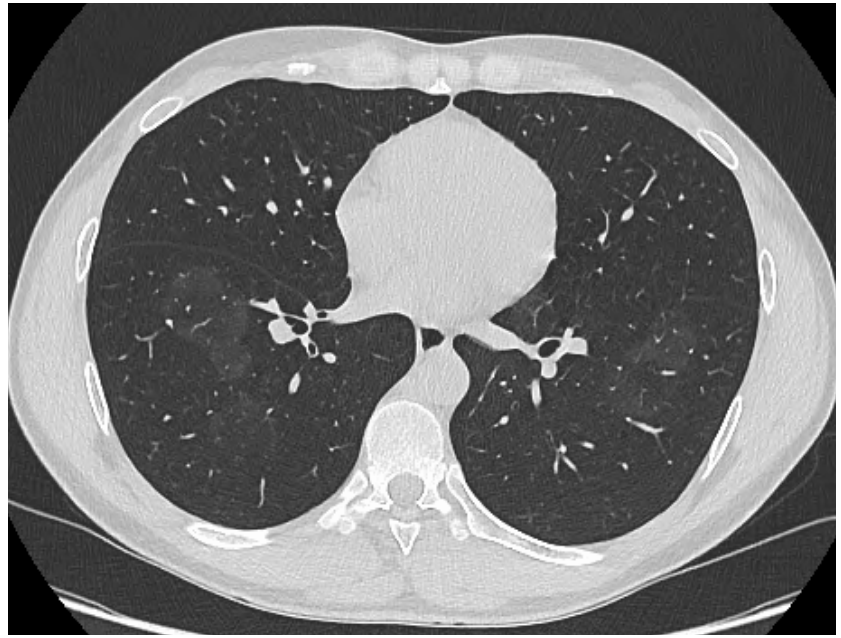
Hémorragie intra-alvéolaire





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Nodules/Masses pulmonaires

RadioGraphics
CHEST IMAGING
Common and Uncommon Manifestations of Wegener Granulomatosis at Chest CT: Radiologic-Pathologic Correlation[1]

Table 3
Differential Diagnosis for Nodules and Masses

Pathologic Condition	Number	Size	Distribution	Cavitation	Ancillary Findings
Wegener granulomatosis	Multiple	Few millimeters up to 10 cm	Usually bilateral and random, but may be peribronchovascular, subpleural, and angiocentric	Seen in up to 50% of lesions >2 cm	CT halo and atoll signs, radiating linear scarring, pleural tags
Metastasis	Multiple	Variable	Bilateral and random	Not common but, if present, suggests squamous, sarcomatous, or transitional cell primary	Lymphadenopathy
Infection	Variable	Usually <10 mm	Peripheral (eg, septic emboli) or miliary (eg, tuberculosis and fungal infection)	Rare	Tree-in-bud opacities, consolidation, reactive lymphadenopathy
Sarcoidosis	Multiple	2–10 mm	Perilymphatic	Rare	Architectural distortion, symmetric adenopathy
Rheumatoid arthritis	Solitary or multiple	5–7 mm	Usually peripheral	Rare	Waxing and waning, subcutaneous nodules

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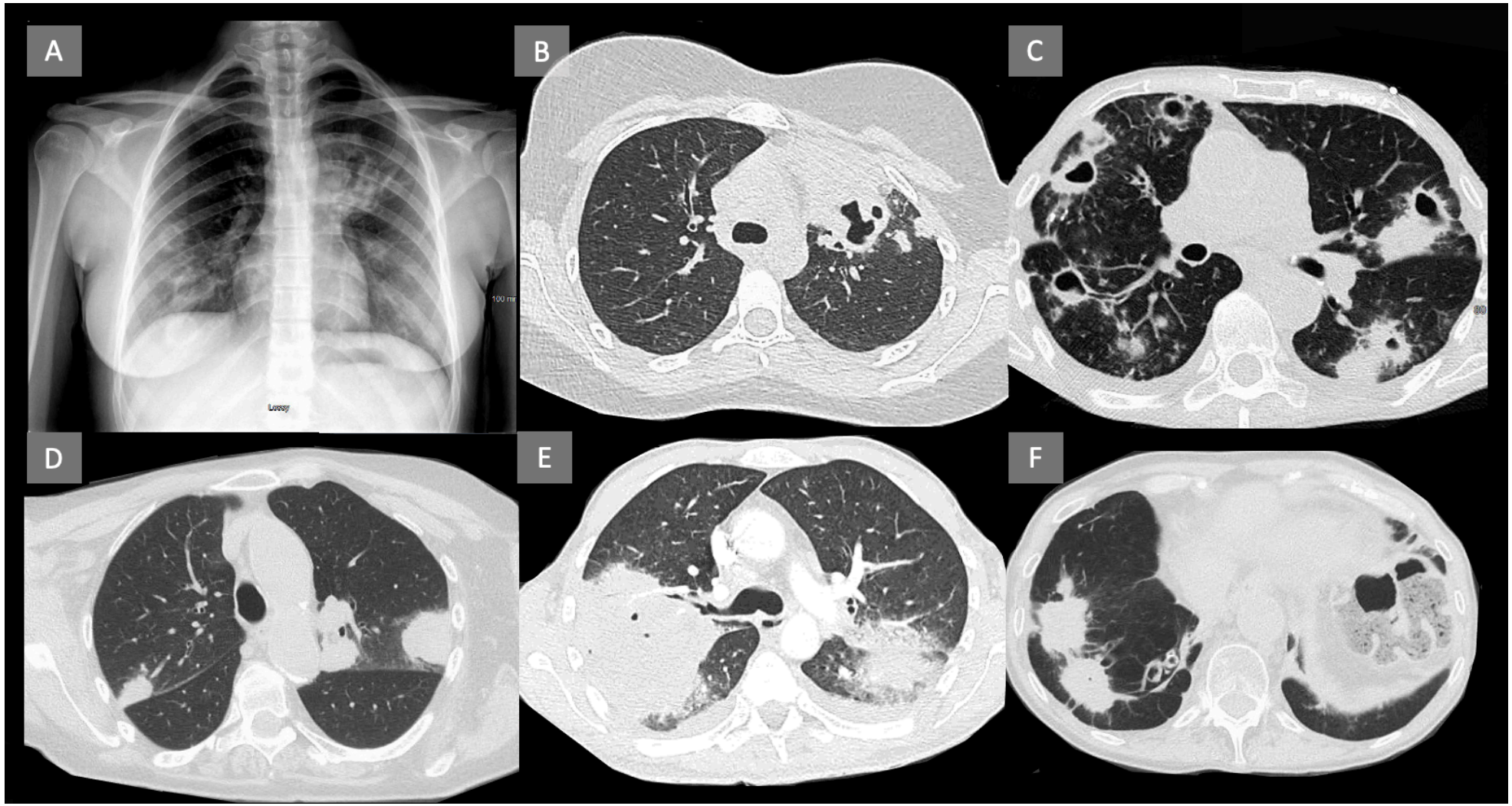
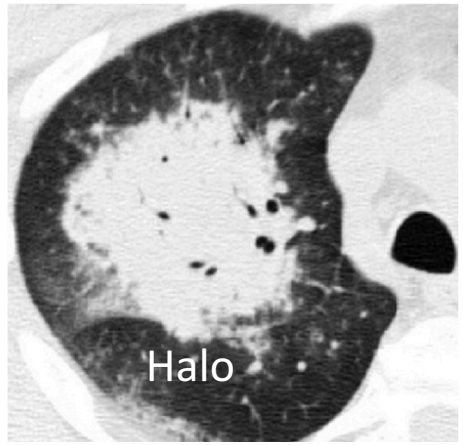
Nodules/Masses pulmonaires





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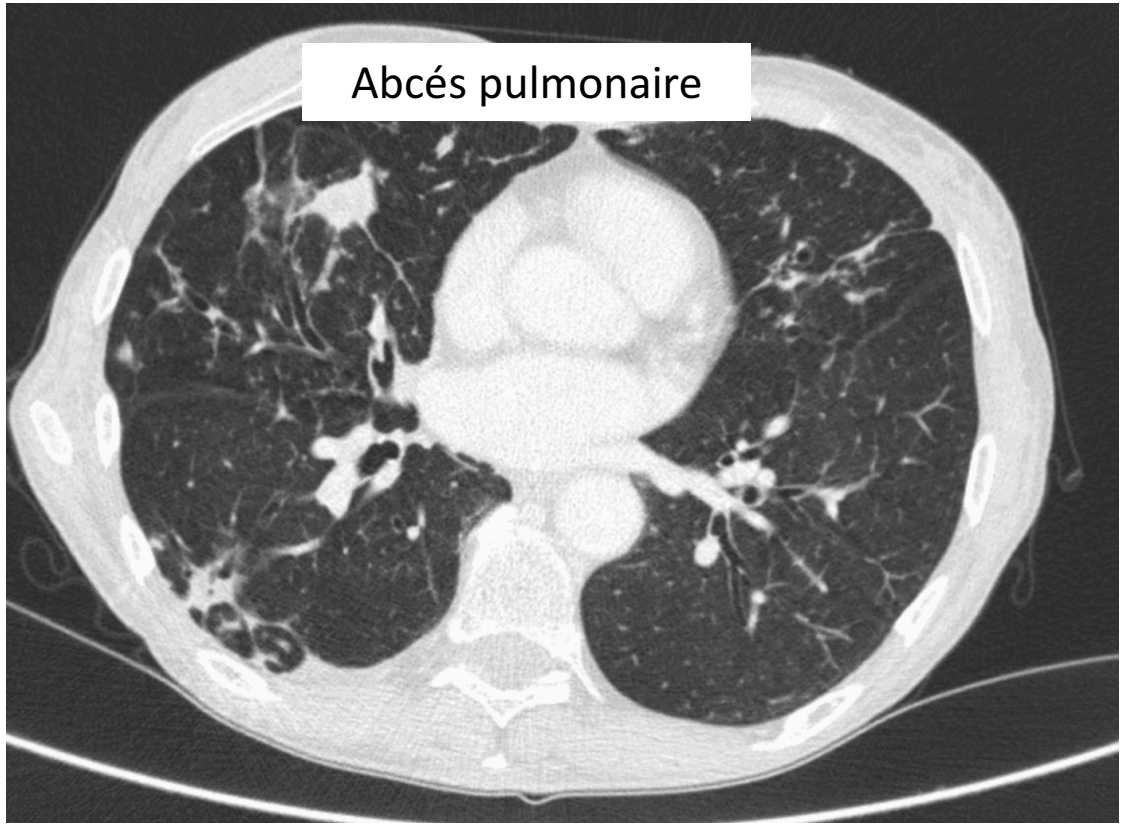
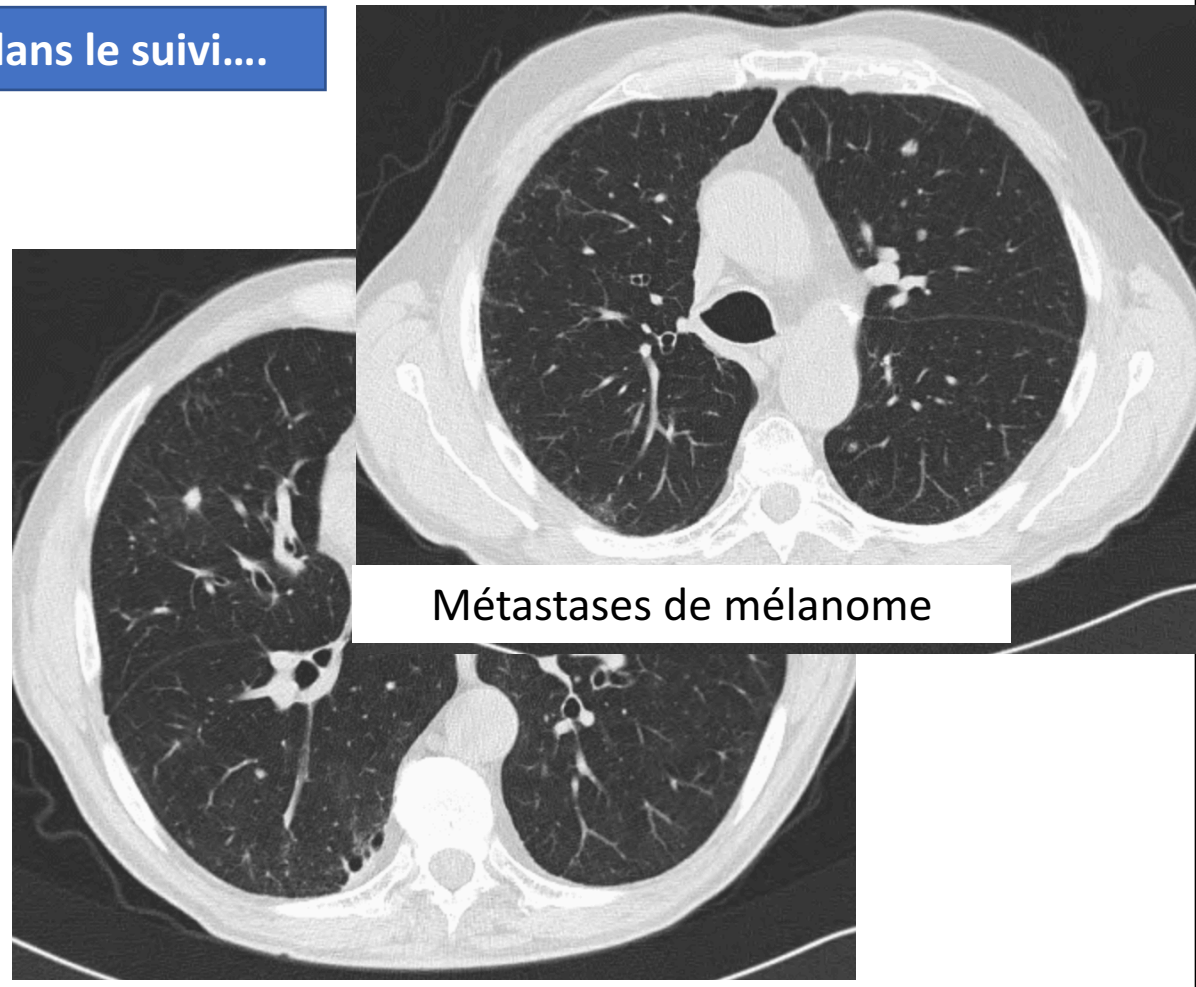
Pôle des voies respiratoires

Nodules/Masses pulmonaires

Les diagnostics différentiels valent au diagnostic de la maladie et dans le suivi….





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Atteinte trachéo-bronchique

RadioGraphics
CHEST IMAGING 51
Common and Uncommon Manifestations of Wegener Granulomatosis at Chest CT: Radiologic-Pathologic Correlation[1]

Table 5
Differential Diagnosis for Airway Thickening

Tracheal Disease	CT Appearance	Differentiating Features
Wegener granulomatosis	Circumferential tracheal wall thickening, most often involving the subglottic region	Subglottic tracheal narrowing; history of sinus or renal disease, pulmonary cavitary nodules, or pulmonary hemorrhage; posterior wall involvement; no wall calcification
Relapsing polychondritis	Thickening of the cartilaginous trachea and tracheal wall	Sparing of the posterior tracheal wall, cartilaginous abnormalities of the ears or nose, tracheal narrowing, wall calcification (occasionally)
Tracheobronchopathia osteochondroplastica	Calcified/ossified nodules in the cartilaginous trachea	Nodular calcified/ossified tracheal wall (almost always) with wall thickening and sparing of the posterior tracheal membrane and superior trachea
Amyloidosis	Calcified/ossified, nodular concentric tracheal wall thickening	Nodular concentric calcified/ossified wall (often present) with wall thickening and without posterior sparing, possibly involving the larynx and upper trachea

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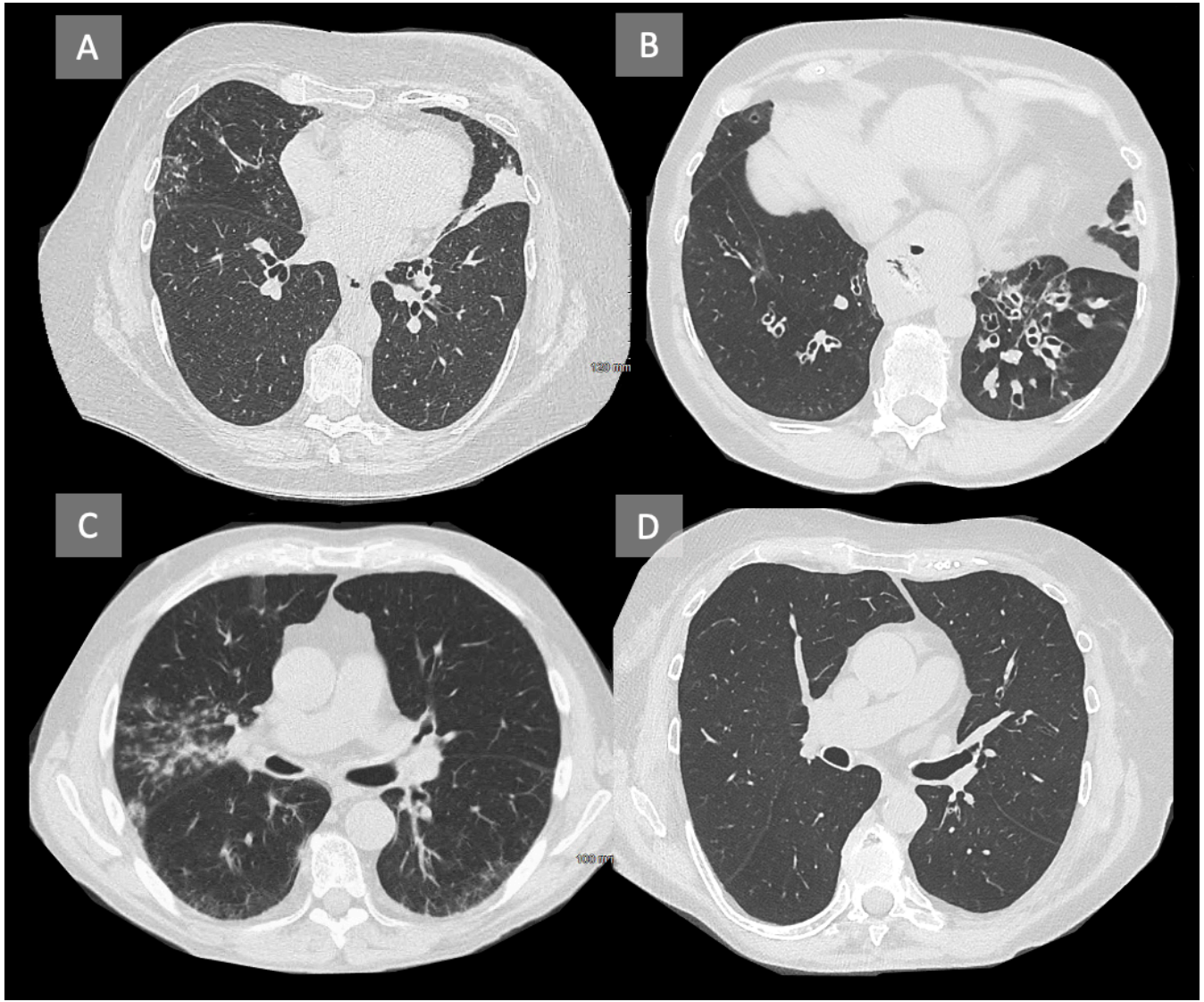
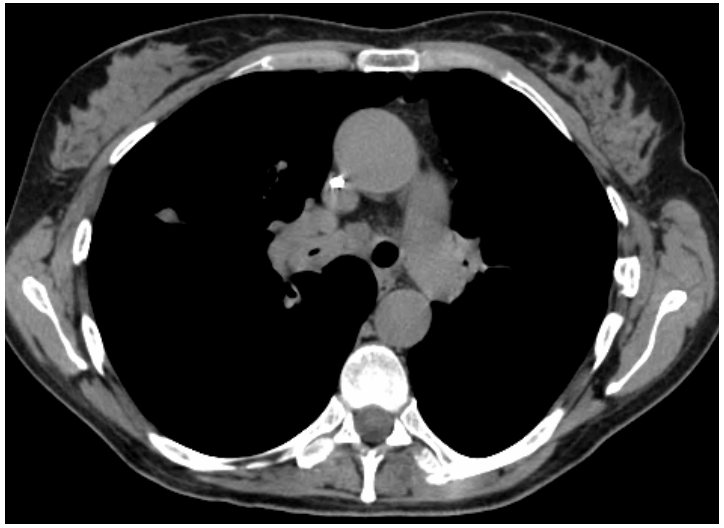
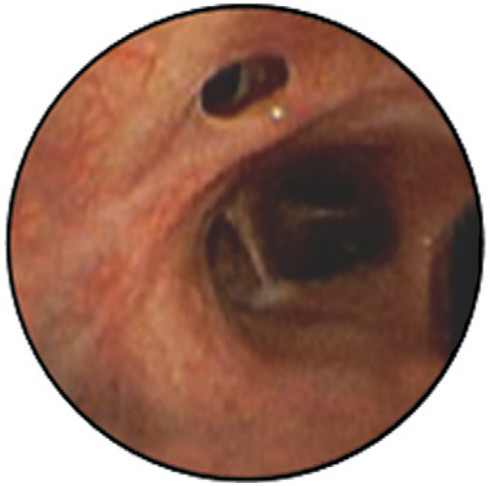
Atteinte trachéo-bronchique







Polychronopoulos Rheum Dis Clin N Am 200

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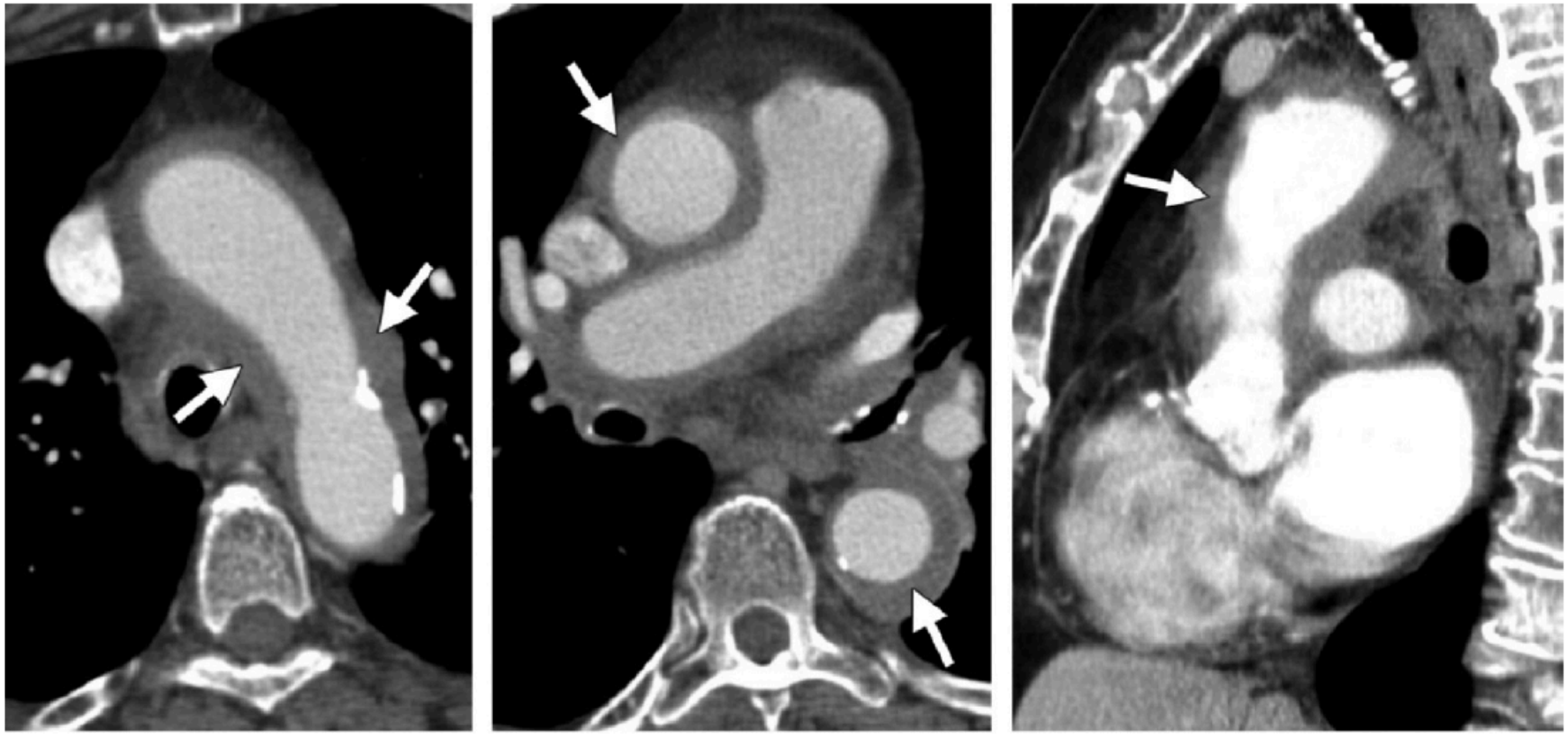
Atteinte des gros vaisseaux



a. b. c.

Figure 27. Axial CT scans at the level of the aortic arch **(a)** and right pulmonary artery **(b)** and sagittal reformatted CT image **(c)** obtained in a 45-year-old patient with known Wegener granulomatosis show extensive periaortitis (arrows).

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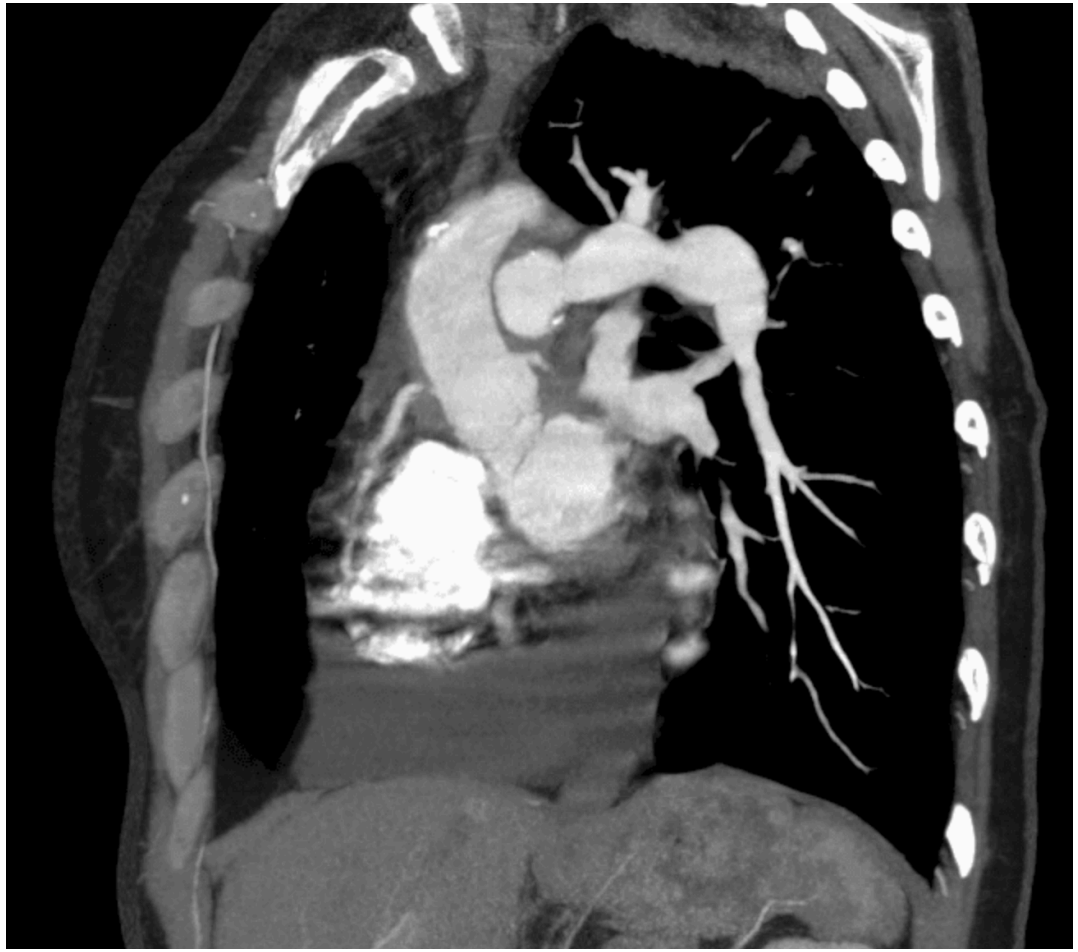
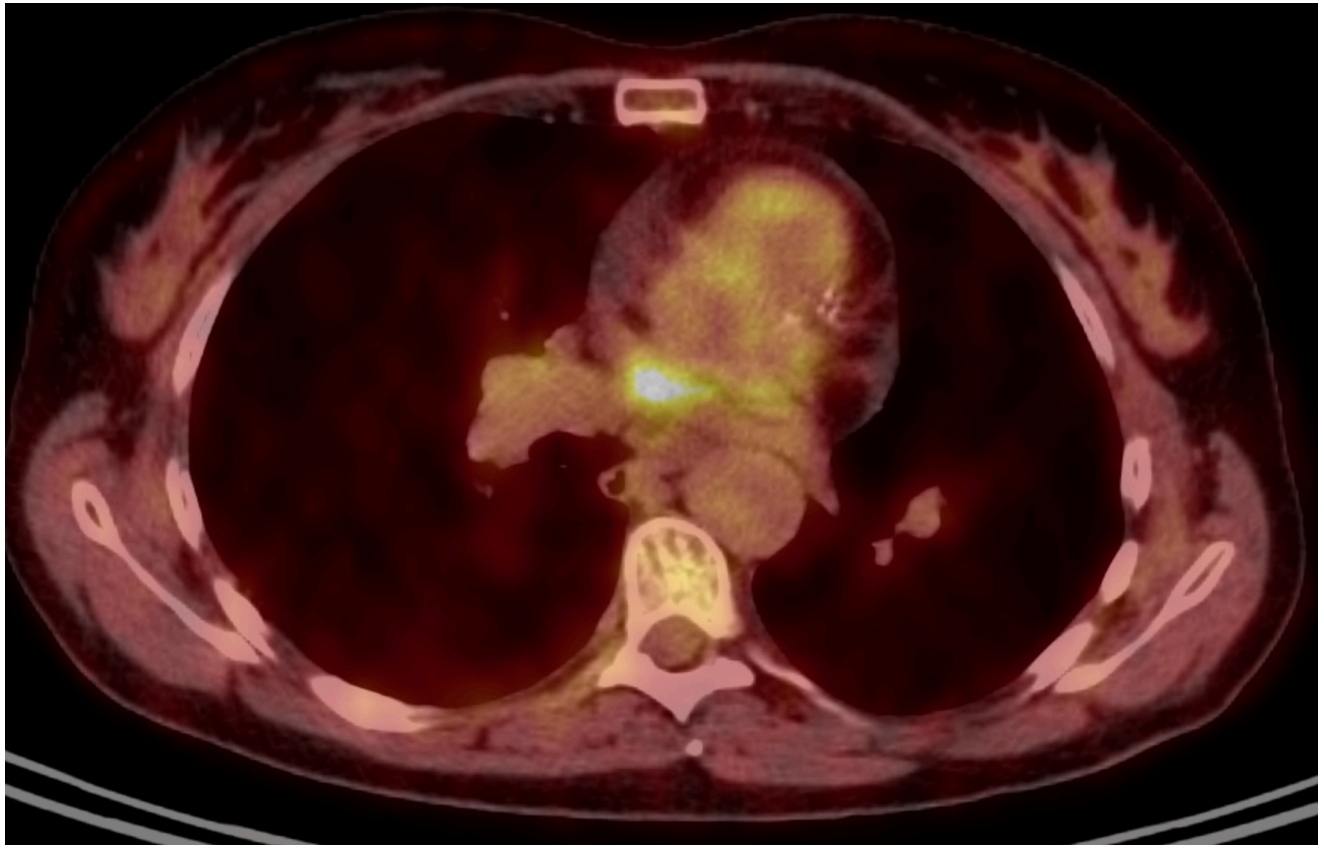
Atteinte des gros vaisseaux





TDM thoracique et suspicion de Vascularite à ANCA

Fibrose pulmonaire

Epidémiologie

- La fibrose pulmonaire concerne plus souvent la MPA que la GPA et les VAA anti-MPO que les anti-PR3
- MPA avec fibrose sont plus âgés : 66 ans
- Ratio H/F débattu
- La fibrose apparaît généralement avant la MPA, souvent simultanément, plus rarement aprés

Autoimmun Rev. 2017 July ; 16(7): 722–729

Imagerie

- PIC > PINS
- Autre : indéterminé ++ et DIP
- Pas d'élément évident en dehors de signes associés qui distinguerait PI-ANCA+ de PI-ANCA-

Anatomopathologie

- PIC > 50% mais avec souvent des zones de PINS et un infiltrat inflammatoire avec atteinte des petites voies aériennes et des follicules lymphoïdes plus fréquent que dans la PIC de FPI
- Lésions de vascularite associée : rarement

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Fibrose pulmonaire

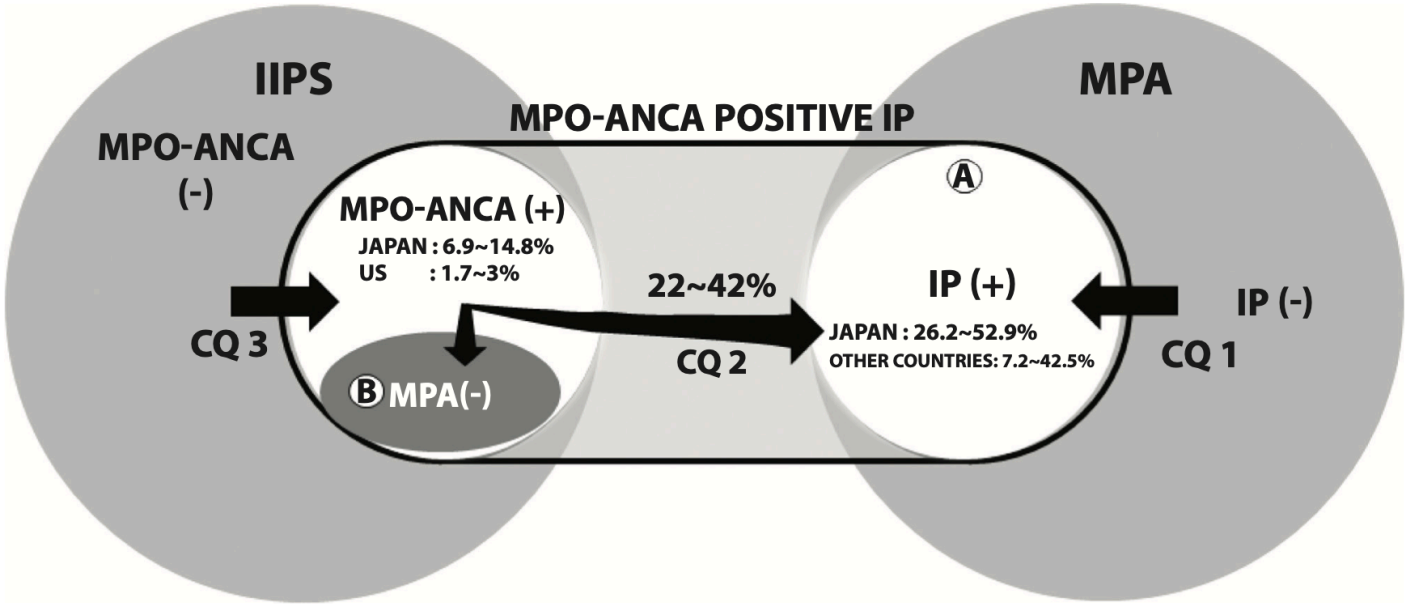

Figure 2. Disease concept and related CQs for
CQ 1: Can MPA or MPO-ANCA cause IP?
CQ 2: Can MPO-ANCA positive IP cause MPA?
CQ 3: Can IP trigger MPO-ANCA production?

SARCOIDOSIS VASCULITIS AND DIFFUSE LUNG DISEASES 2021; 38 (4);

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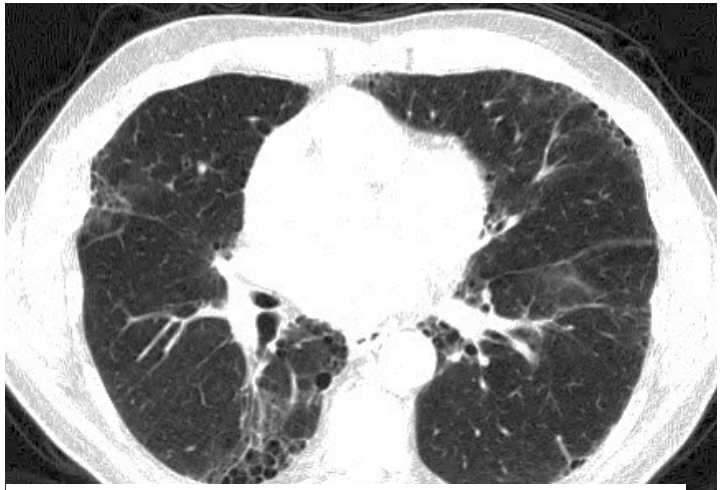
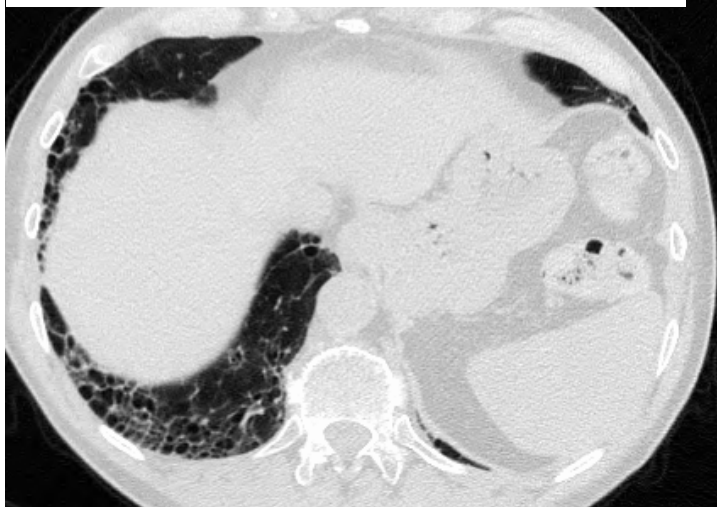
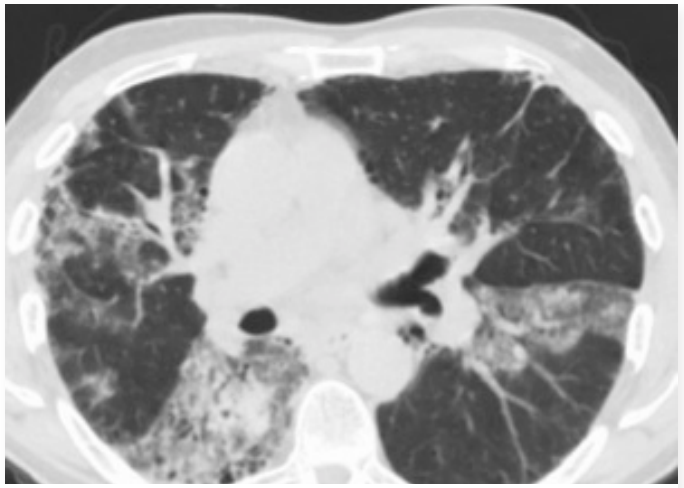
Fibrose pulmonaire

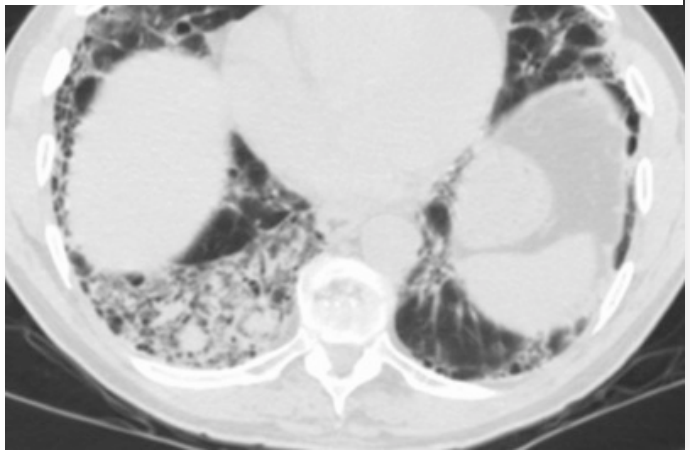


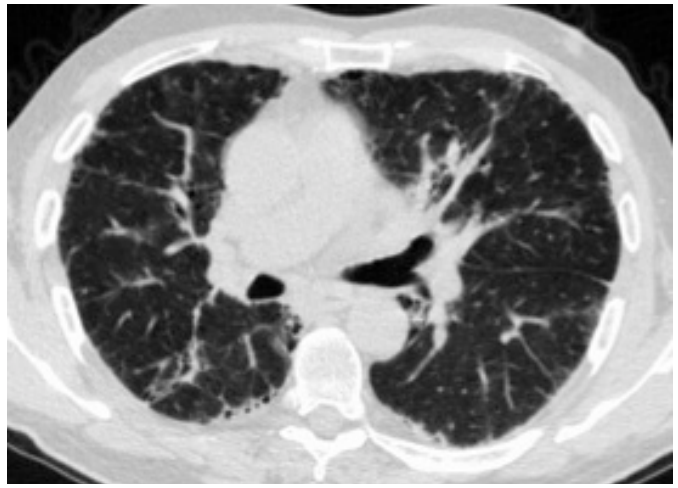
Au diagnostic : FPI Anti MPO+



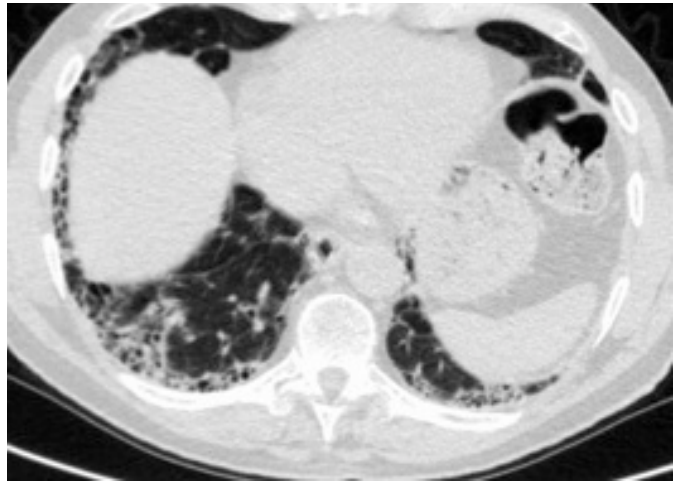


+ 3 ans : VAA Anti MPO+ avec HIA et Fibrose pulmonaire





Après CYP



TDM thoracique et suspicion de Vascularite à ANCA

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Fibrose pulmonaire

Chez les patients VAA avec PID:

- seules les PIC sont associées à un plus mauvais pronostic
- Les traitements immunosuppresseurs n'influencent pas le pronostic

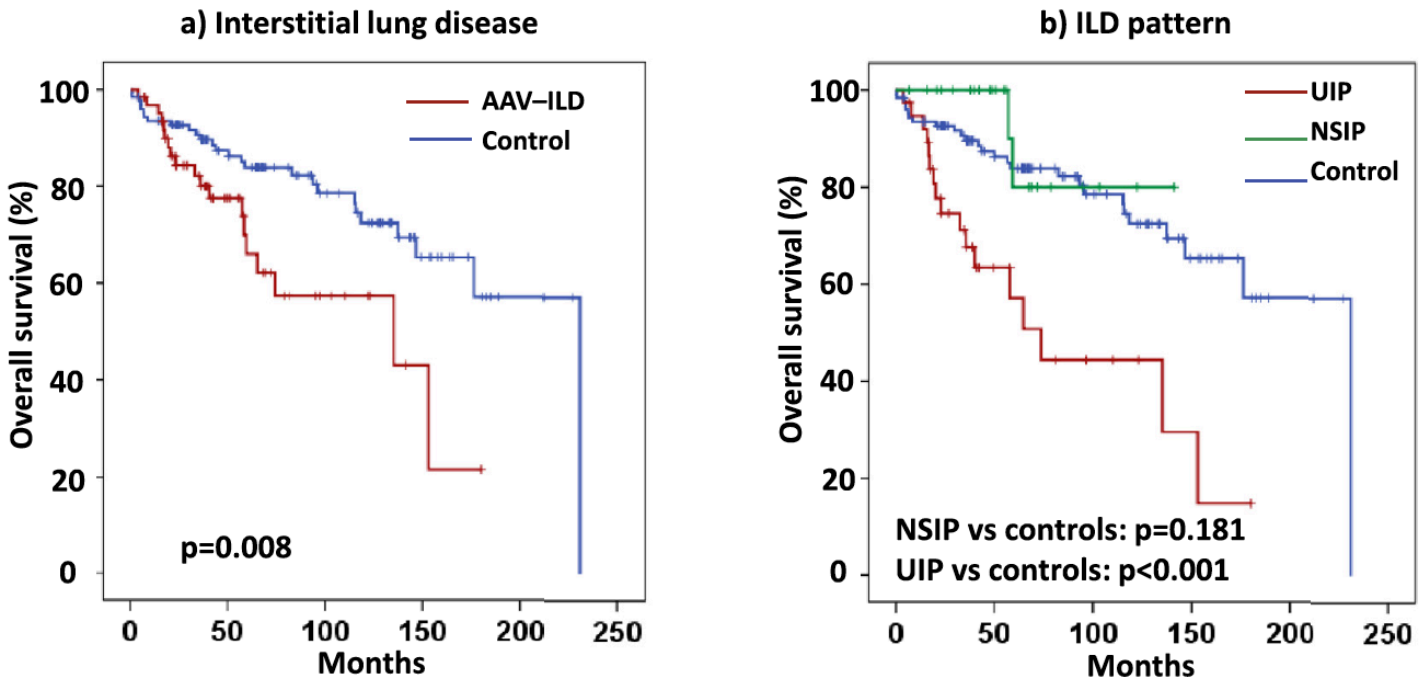
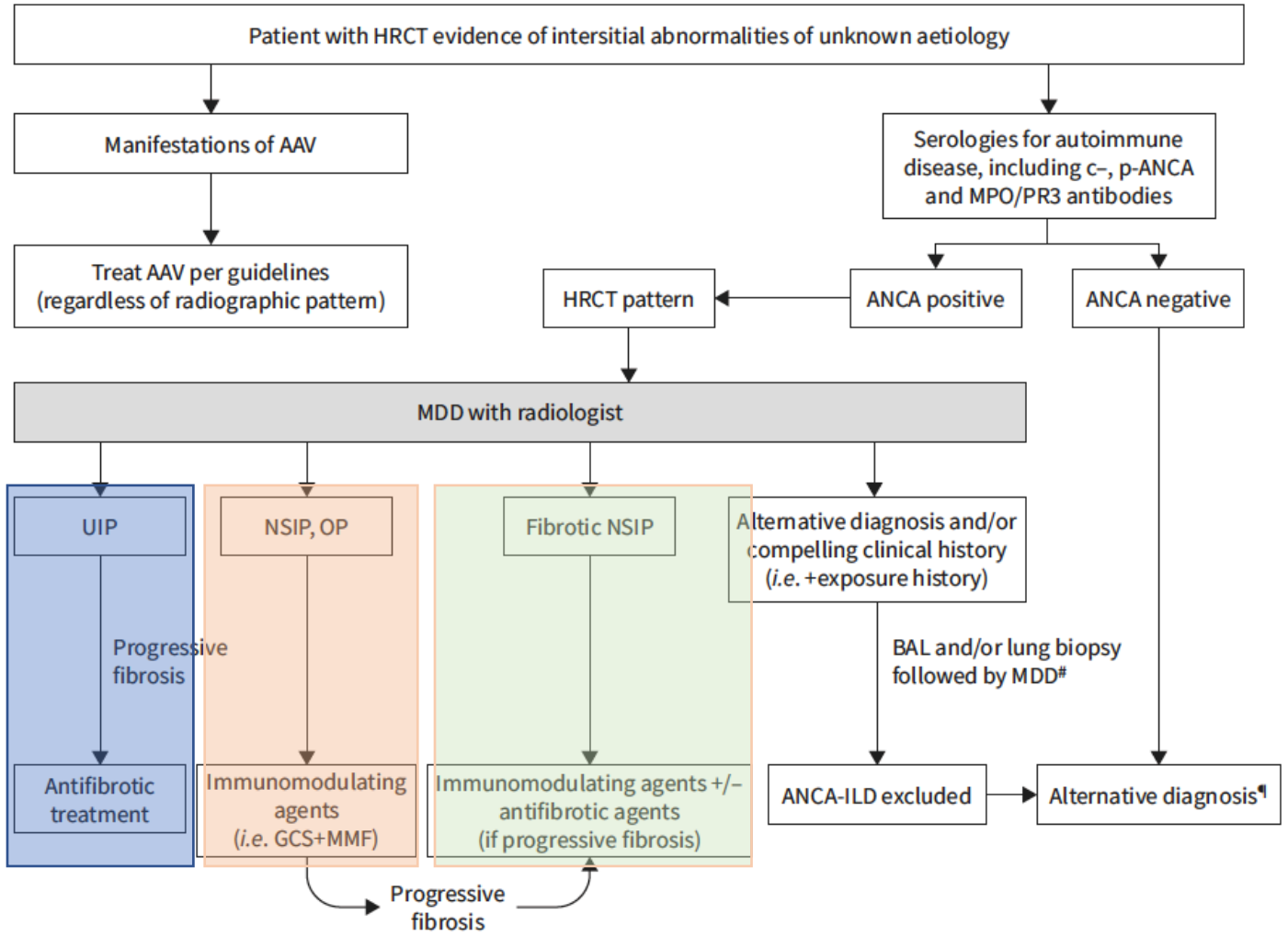


Fig. 3. Overall survival according to the according to the analysis of AAV–ILD cases vs AAV controls. P value derived from log-rank tests. AAV–ILD: ANCA-associated vasculitis–progressive fibrosing interstitial lung disease; NSIP: non-specific interstitial pneumonia; UIP: Usual interstitial pneumonia.

Maillet et al J Autoimmu. 2020

TDM thoracique et suspicion de Vascularite à ANCA

Fibrose pulmonaire



Kadura ERR 2021

TDM thoracique et classification de la VAA

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

CLASSIFICATION CRITERIA FOR GRANULOMATOSIS WITH POLYANGIITIS

Arthritis & Rheumatology
Vol. 74, No. 3, March 2022, pp 393–399
DOI 10.1002/art.41986

AMERICAN COLLEGE of RHEUMATOLOGY
Empowering Rheumatology Professionals

2022 American College of Rheumatology/European Alliance of Associations for Rheumatology Classification Criteria for Granulomatosis With Polyangiitis

CONSIDERATIONS WHEN APPLYING THESE CRITERIA

- These classification criteria should be applied to classify a patient as having granulomatosis with polyangiitis when a diagnosis of small- or medium-vessel vasculitis has been made
- Alternate diagnoses mimicking vasculitis should be excluded prior to applying the criteria

CLINICAL CRITERIA

Nasal involvement: bloody discharge, ulcers, crusting, congestion, blockage, or septal defect / perforation	+3
Cartilaginous involvement (inflammation of ear or nose cartilage, hoarse voice or stridor, endobronchial involvement, or saddle nose deformity)	+2
Conductive or sensorineural hearing loss	+1

LABORATORY, IMAGING, AND BIOPSY CRITERIA

Positive test for cytoplasmic antineutrophil cytoplasmic antibodies (cANCA) or antiproteinase 3 (anti-PR3) antibodies	+5
Pulmonary nodules, mass, or cavitation on chest imaging	+2
Granuloma, extravascular granulomatous inflammation, or giant cells on biopsy	+2
Inflammation, consolidation, or effusion of the nasal/paranasal sinuses, or mastoiditis on imaging	+1
Pauci-immune glomerulonephritis on biopsy	+1
Positive test for perinuclear antineutrophil cytoplasmic antibodies (pANCA) or antimyeloperoxidase (anti-MPO) antibodies	-1
Blood eosinophil count ≥ 1 x10^9/liter	-4

Sum the scores for 10 items, if present. A score of ≥ 5 is needed for classification of GRANULOMATOSIS WITH POLYANGIITIS.

TDM thoracique et classification de la VAA

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

CLASSIFICATION CRITERIA FOR MICROSCOPIC POLYANGIITIS

American College of Rheumatology
Empowering Rheumatology Professionals

Arthritis & Rheumatology
Vol. 74, No. 3, March 2022, pp 400–406
DOI 10.1002/art.41983

2022 American College of Rheumatology/European Alliance of Associations for Rheumatology Classification Criteria for Microscopic Polyangiitis

CONSIDERATIONS WHEN APPLYING THESE CRITERIA

- **These classification criteria should be applied to classify a patient as having microscopic polyangiitis when a diagnosis of small- or medium-vessel vasculitis has been made**
- **Alternate diagnoses mimicking vasculitis should be excluded prior to applying the criteria**

CLINICAL CRITERIA

Nasal involvement: bloody discharge, ulcers, crusting, congestion, blockage or septal defect / perforation	-3

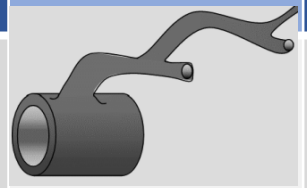
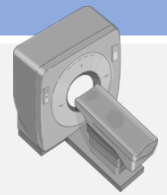
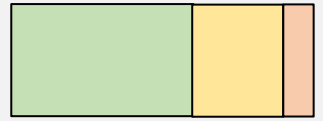
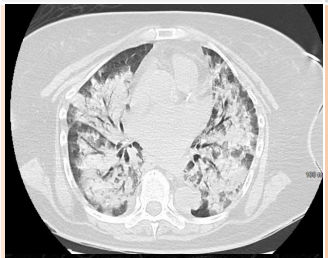
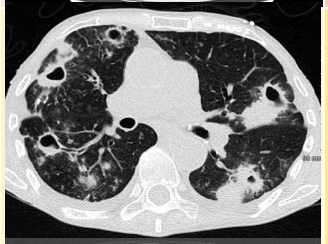
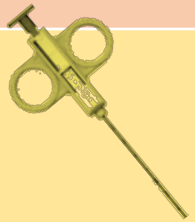
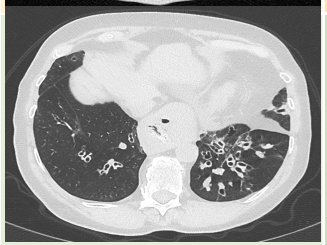
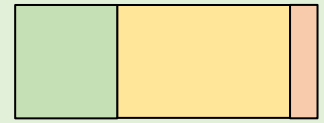
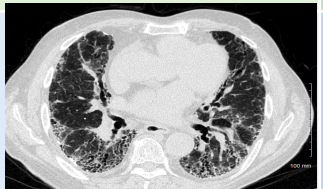
LABORATORY, IMAGING, AND BIOPSY CRITERIA

Positive test for perinuclear antineutrophil cytoplasmic antibodies (pANCA) or antimyeloperoxidase (anti-MPO) antibodies ANCA positive	+6
Fibrosis or interstitial lung disease on chest imaging	+3
Pauci-immune glomerulonephritis on biopsy	+3
Positive test for cytoplasmic antineutrophil cytoplasmic antibodies (cANCA) or antiproteinase 3 (anti-PR3) antibodies	-1
Blood eosinophil count ≥ 1 x10^9/liter	-4

Sum the scores for 6 items, if present. A score of ≥ 5 is needed for classification of MICROSCOPIC POLYANGIITIS.

Conclusion – thèse Thomas Villeneuve

Service de pneumologie
CHU TOULOUSE
Pôle des voies respiratoires

Atteinte	% *littérature*	Terrain	Type de VAA	Imagerie	Diagnostics différentiels	Evolution sous ttt
Respiratoire	55% *25-74%*				23% Examens	
Hémorragie intra-alvéolaire	52% *28-41%*	65 ans H/F 1,2	MPA > GPA		16% 	
Nodulaire	32% *25-59%*	57 ans H/F 0,9	GPA >> MPA		22% 	
Trachéo-bronchique	16% *12%*	63 ans H/F 0,9	GPA >> MPA		56% 	
Interstitielle	15% *2%*	74 ans H/F 1,8	MPA >> GPA		15%