2. DACH ANCA VASKULITIS FORUM 2024

22. & 23. NOVEMBER 2024 | MÜNCHEN

CSL Vifor

Diagnosestellung der AAV (Praktische Erfahrungen)

Prof. Dr. Sabine Adler

Aarau

KSA Kantonsspital Aarau

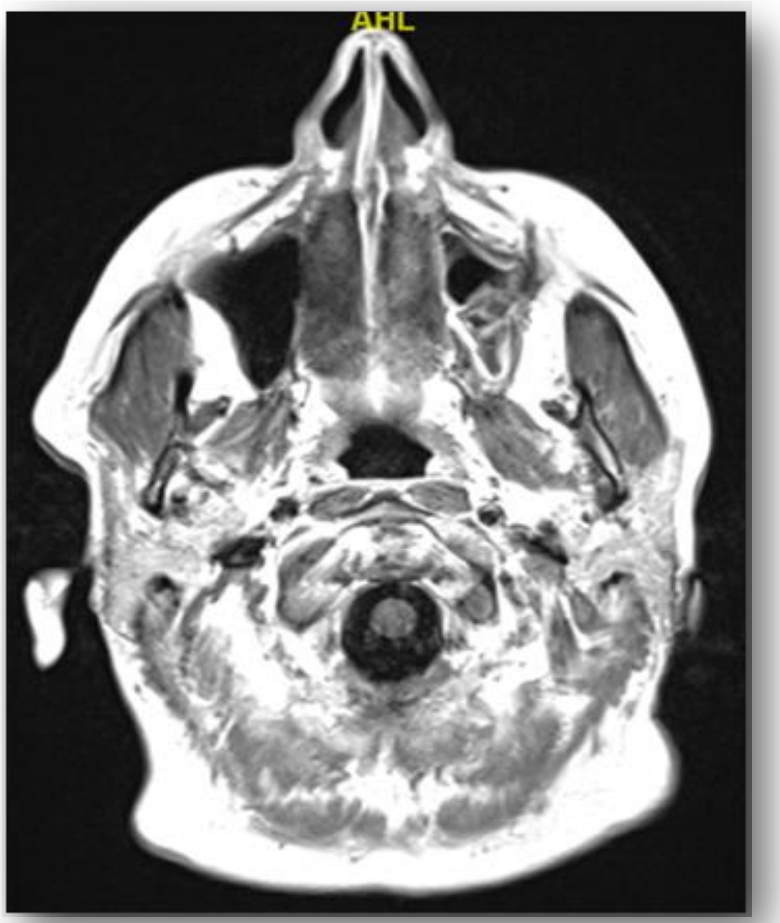
Diagnosestellung der AAV praktische Erfahrungen

2. DACH ANCA-Vaskulitis Forum 2024

Sabine Adler

München
22. November 2024

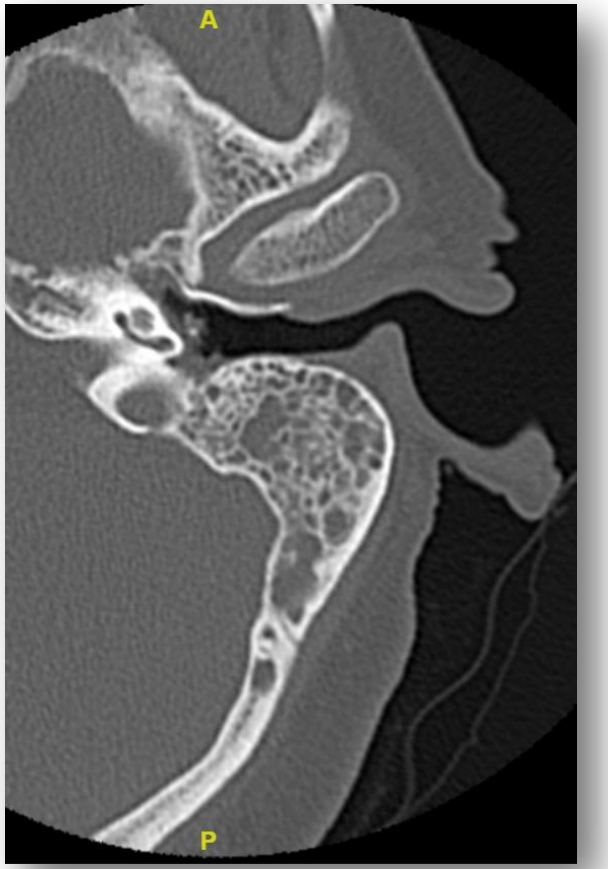
Frau G. R. 68 Jahre



- AZ Verschlechterung
- Hörverlust, Facialisparese, "Nase zu"
- Arthralgien

- vorübergehend "Cortison"
  mit "etwas" Ansprechen

Frau G. R. 68 Jahre



CT Felsenbein

- AZ Verschlechterung
- Hörverlust, Facialisparese
- Arthralgien

- serologische Inflammation
- Anämie
- Gewichtsverlust

- im Verlauf….. Anti-PR3 47 U/ml (Norm < 7)
- renal unauffällig

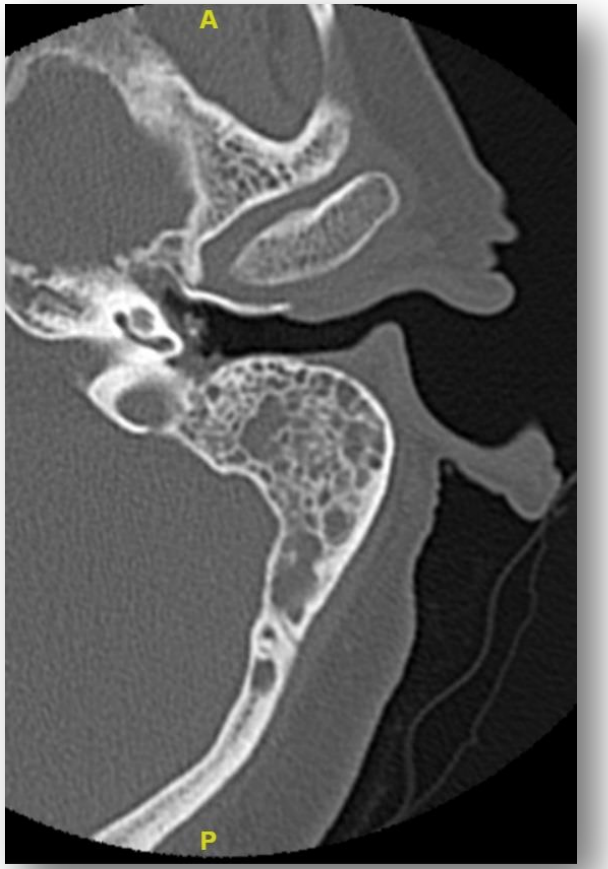
Frau G. R. 68 Jahre

Histologien HNO

NNH links:
Von respiratorischer Schleimhaut überkleidete Gewebsfragmente mit fokaler Nekrose, mässiggradiger chronischer, teils erosiver Entzündung, vereinzelten locker eingestreuten Granulomen und geringgradiger Eosinophilie.
Kein Nachweis von Pilzelementen in der GMS-Färbung.
Kein Dysplasie- oder Malignitätsnachweis.

Die vorliegenden Proben sind in Zusammenschau mit den klinischen Angaben und der erhöhten c-ANCA Titer mit einer Granulomatöse mit Polyangiitis vereinbar. Histologisch sind die Veränderungen einer Vaskulitis nicht (mehr) zu sehen. Auch die Nekrose ist fokal und nicht landkartenartig. Möglicherweise ist dies auf eine vorausgegangene Steroidtherapie zurückzuführen.

Frau G. R. 68 Jahre

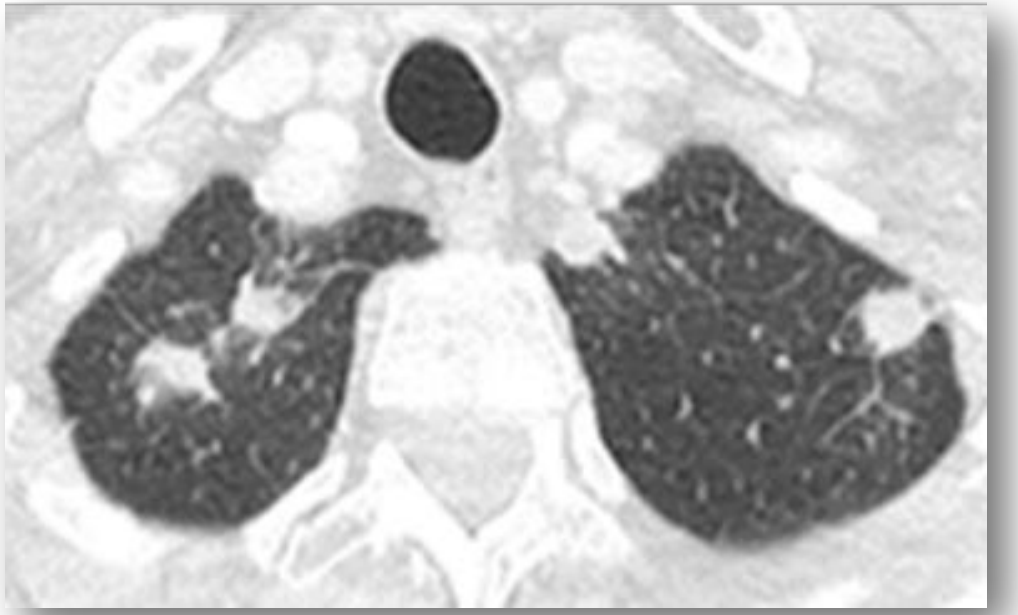


CT Felsenbein

AAV / PR-3 positiv

- HNO Manifestationen
- ausgeprägte B-Symptomatik

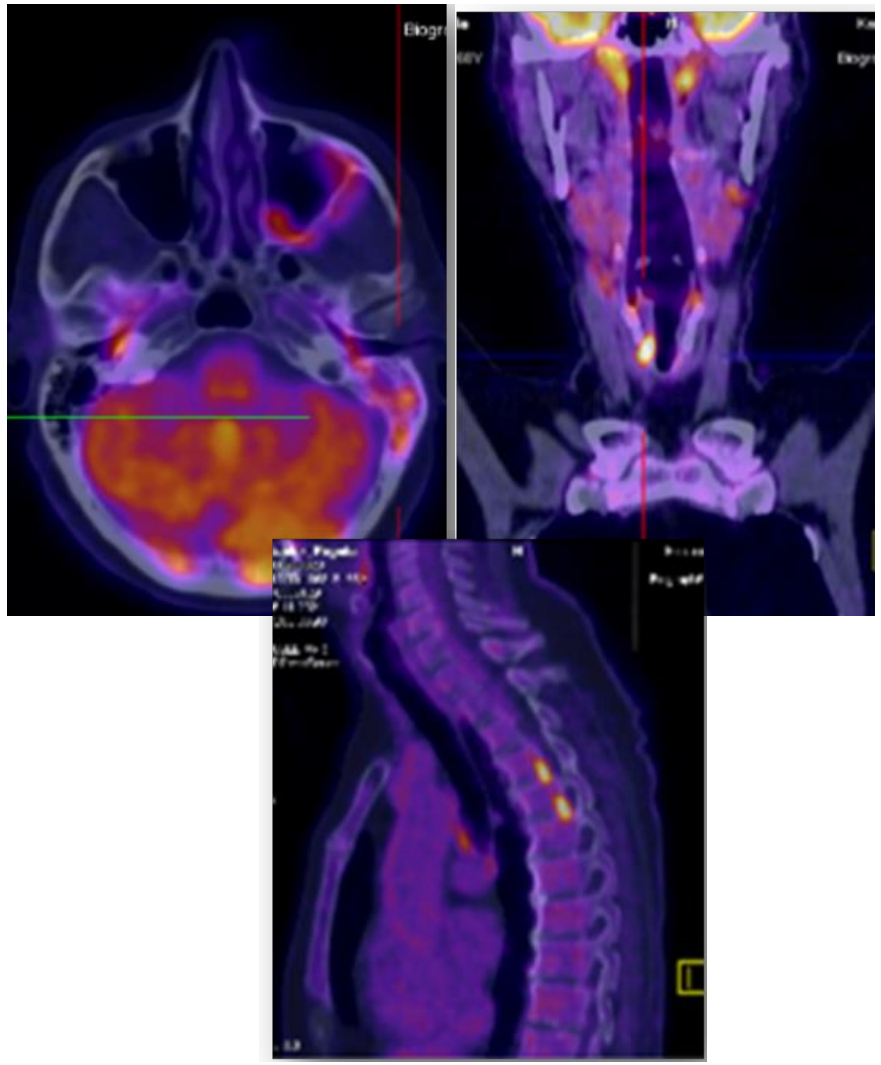
Frau G. R. 68 Jahre



AAV / PR-3 positiv

- HNO Manifestationen
- ausgeprägte B-Symptomatik
- pulmonale Granulomatose
- (DD Tumor??)

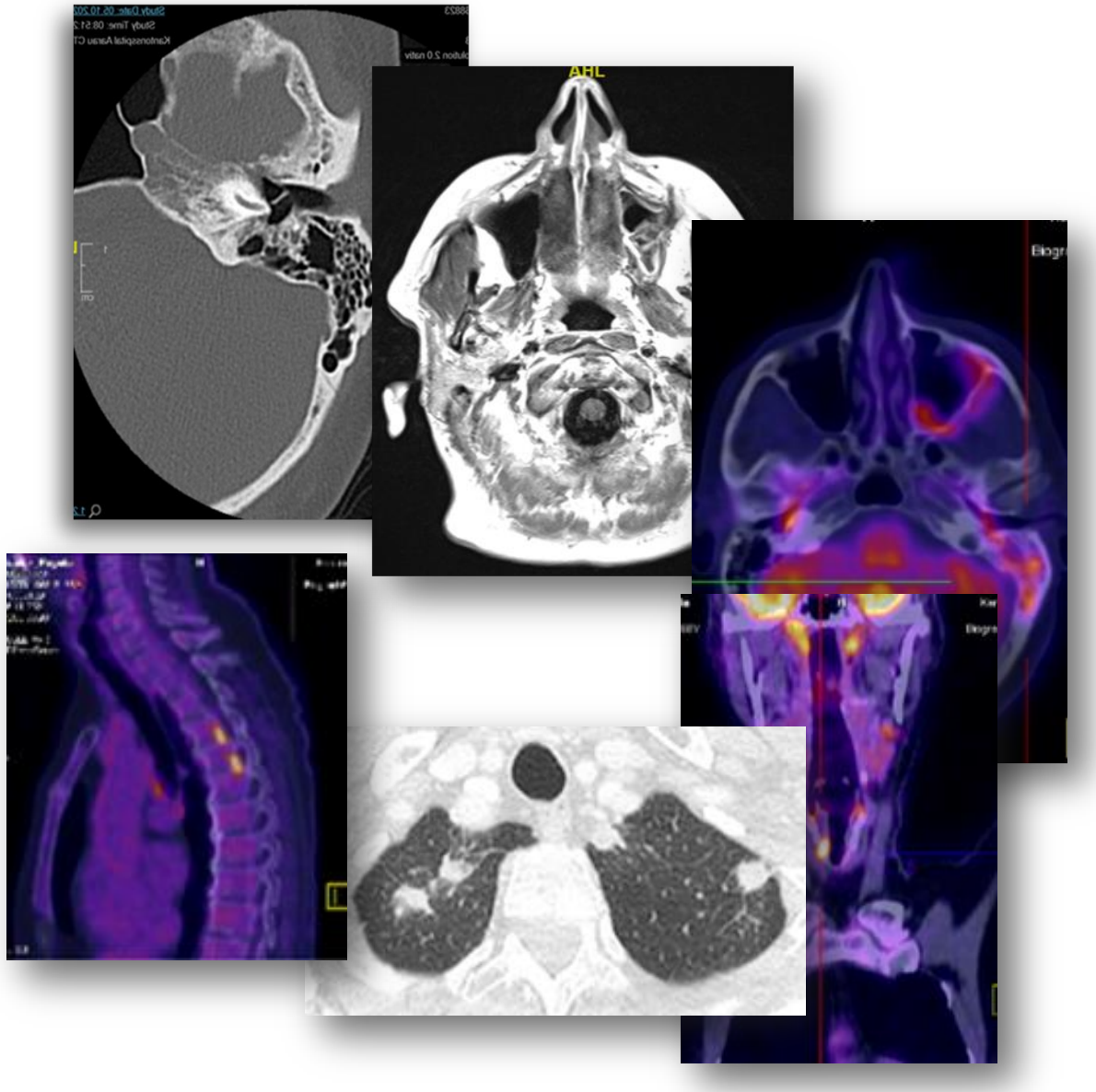
Frau G. R. 68 Jahre



AAV / PR-3 positiv

- HNO Manifestationen
- ausgeprägte B-Symptomatik
- pulmonale Granulomatose
- WK Manifestation?
- Sepsis?
- ANCA paraneoplastisch?

Frau G. R. 68 Jahre



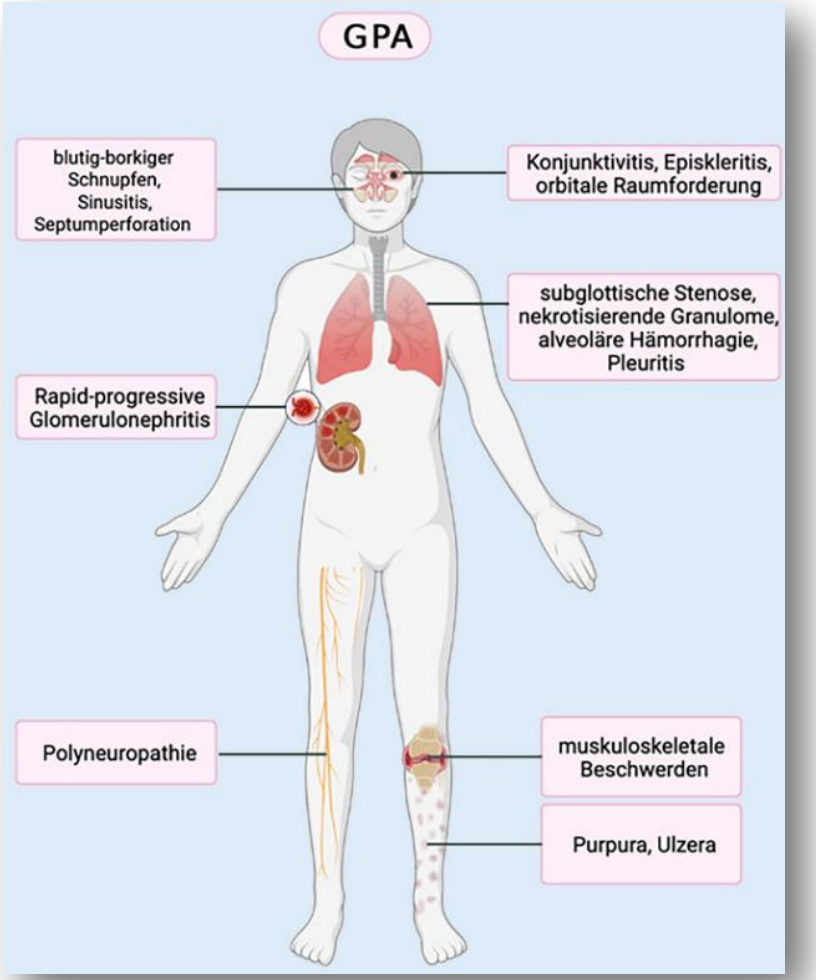
AAV / PR-3 positiv

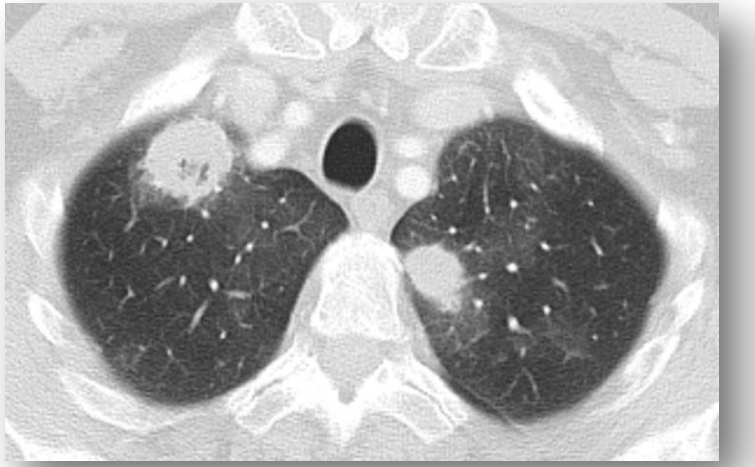
- HNO Manifestationen
- ausgeprägte B-Symptomatik
- pulmonale Granulomatose
- WK Manifestation?

  => "Dura Th5 derb, Pus entleert"

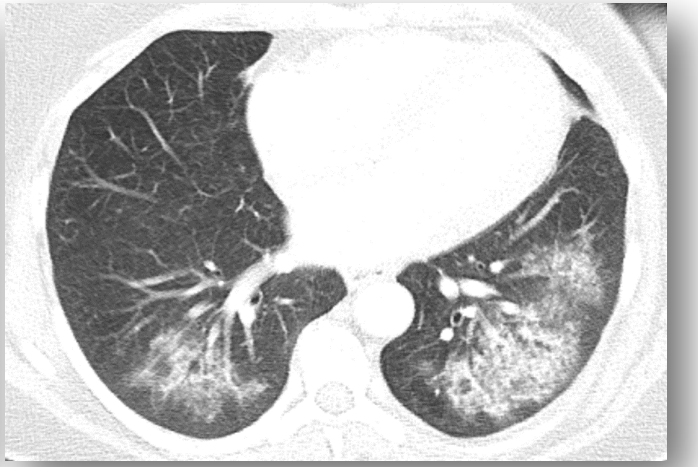
  => MiBi: "steril"

AAV => GPA Manifestationen





Pulmonale Granulome



Pulmonale Hämorrhagie

Krasselt M, Holle J, Innere Med 2022;9:947-960

17 Empfehlungen / Auszug

- Biopsie zur Diagnose/Relapse-sicherung anzustreben
- bei hoher Vortestwahrscheinlichkeit:
  => ANCA für PR3 **UND** MPO (Antigen-spezifische)r assay

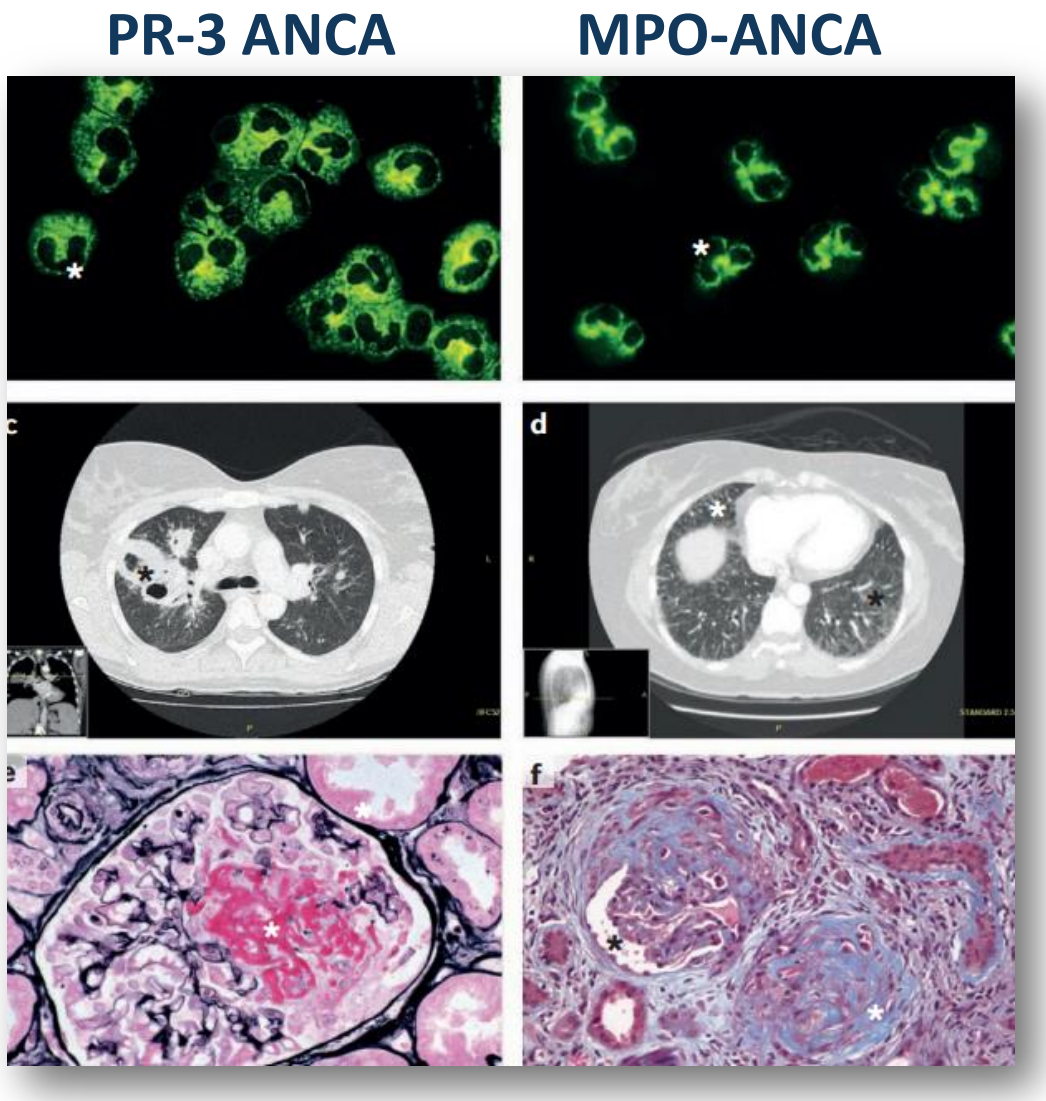
- Remissionsinduktion
- Remissionserhaltung
- Glucocorticoid-Reduktion

modfiziert nach
Hellmich B, et al. Ann Rheum Dis 2023;0:1–18

ANCA-assoziierte Vaskulitiden (AAV)

Erkrankung		ANCA	Zielantigen	
GPA	Granulomatose mit Polyangiitis	c-ANCA	PR-3	spezifisch
MPA	Mikroskopische Polyangiitis	p-ANCA	MPO	spezifisch
eGPA	eosinophile GPA	p-ANCA	MPO (23%)	

ANCA



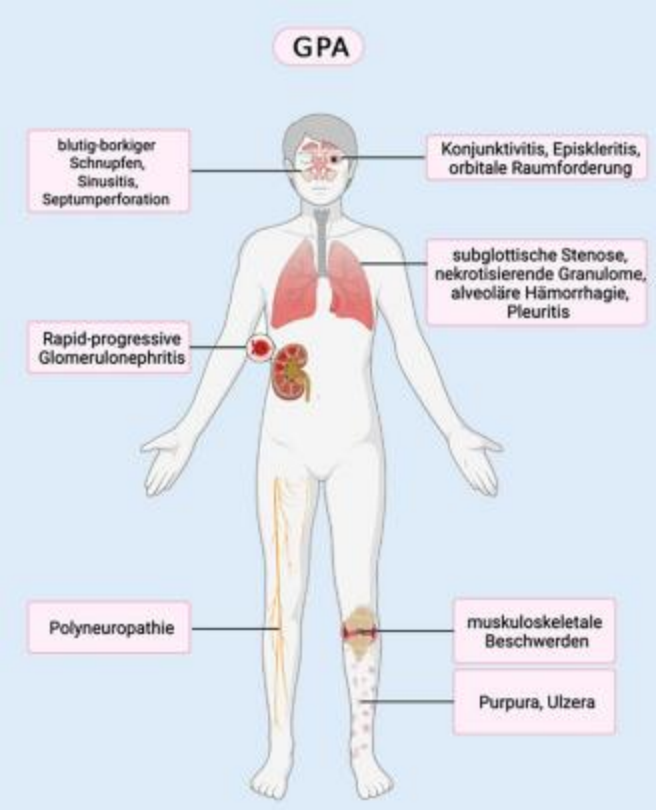
Granulome

Inflammation
Nekrose

Infiltrate
Fibrosierung

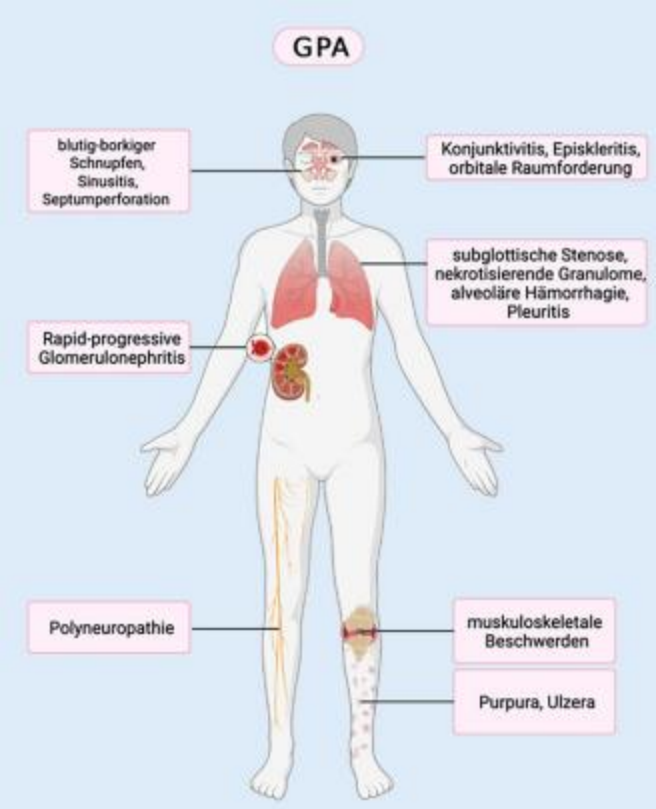
Fibrosierung

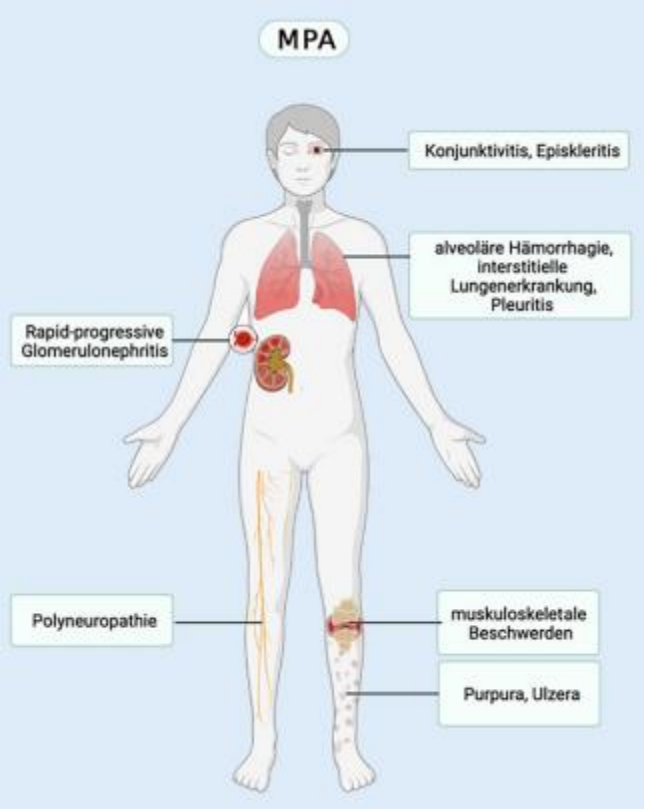
AAV



Krasselt M, Holle J, Innere Med 2022;9:947-960

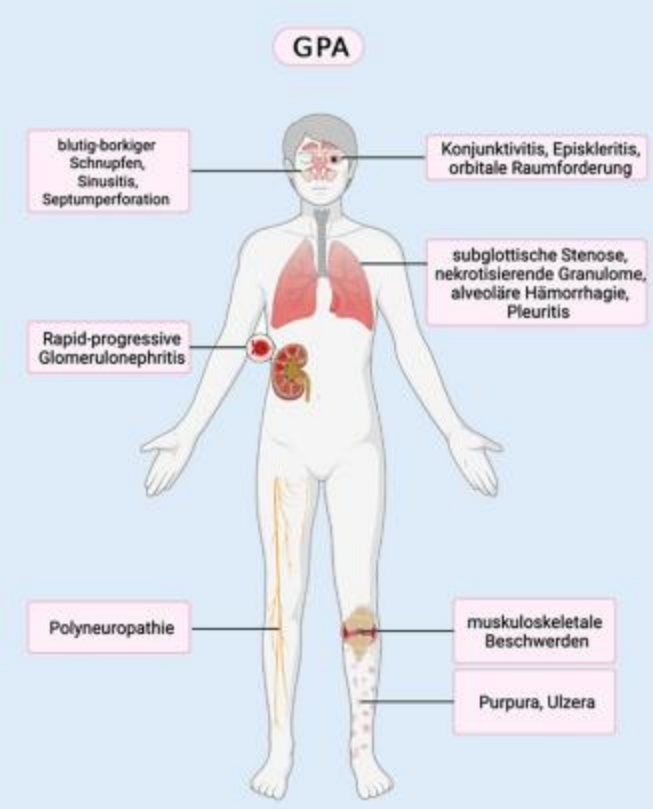
AAV

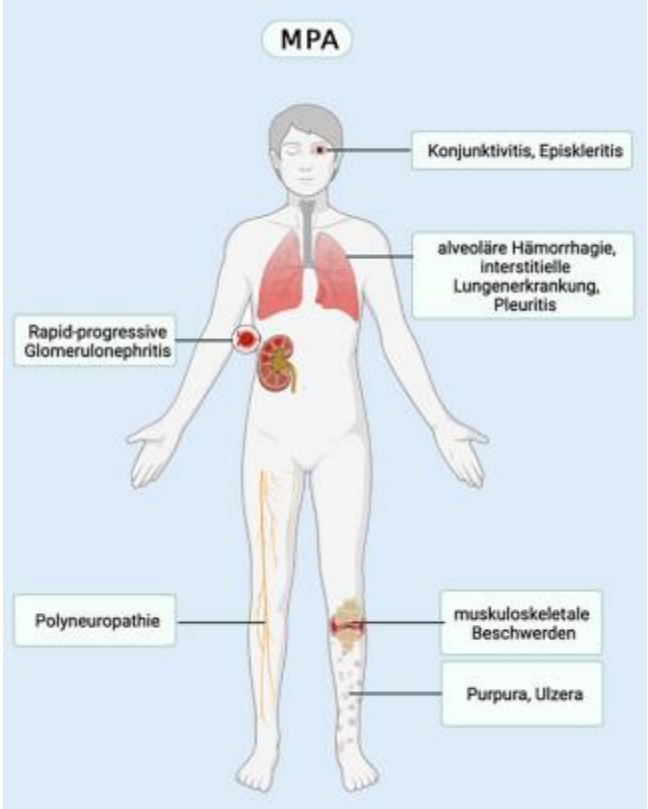


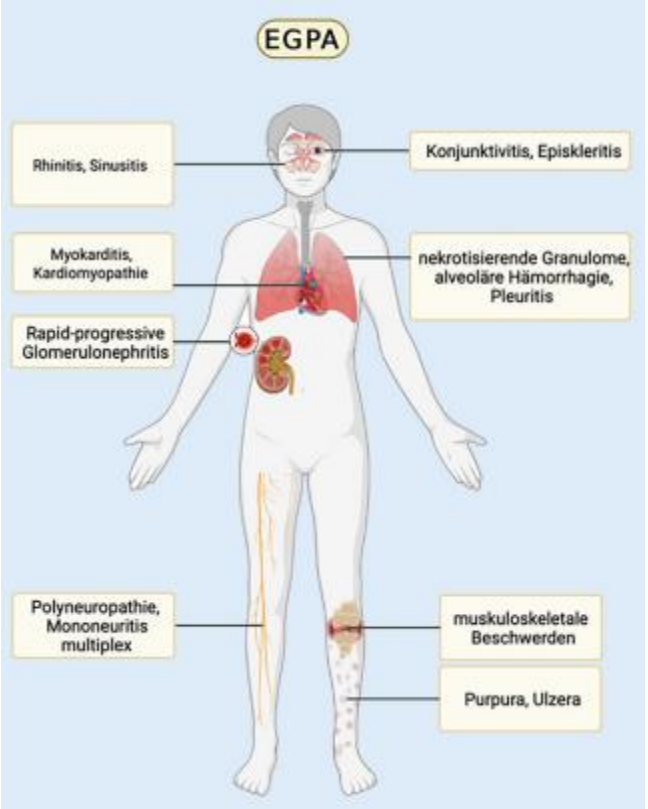


Krasselt M, Holle J, Innere Med 2022;9:947-960

AAV

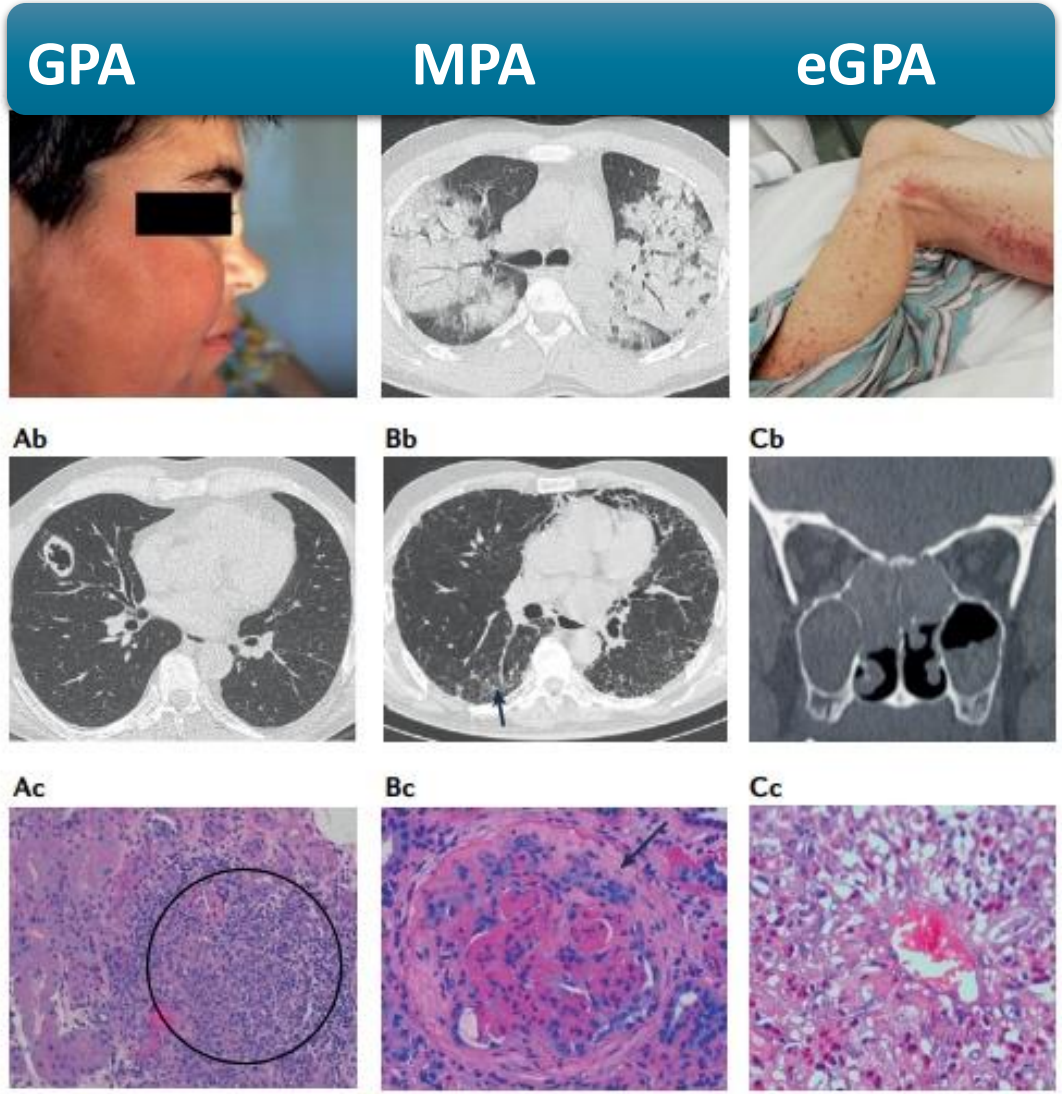






Krasselt M, Holle J, Innere Med 2022;9:947-960

GPA / MPA / eGPA Haupt-Manifestationen



Trivioli G, et al
Nat Rev Rheum 2022; 559-574

AAV Histologie

	Access route or method	Diagnostic yield	Most frequent lesion*	Sampling error issues and aspects to consider
Kidney[14]	Percutaneous	≥99%	Crescentic glomerulonephritis	Low number of glomeruli; atypical lesions (tubulointerstitial nephritis)
Lung[15]	Open	90%	Vasculitic features*	Non-specific inflammation; invasive procedure with a high complication rate
Lung[15]	Transbronchial or CT-guided	≥50%†	Features of mixed inflammatory infiltrate	Biopsy of necrotic areas; complications associated with the procedure (possible pneumothorax)
Ear, nose, and throat[13,16]	Nasal	>30%	Non-specific inflammation and granulomatous or vasculitic features	Inadequate sampling (improve accuracy by taking biopsies >5 mm at the edge of the inflamed area)
Ear, nose, and throat[13,16]	Tracheal-subglottic stenosis	90%	Features of mixed inflammatory infiltrate	Vasculitis features are rare and only present in 10–15% of patients
Eye[17]	Orbit fine needle aspiration or open	>60%	Features of mixed inflammatory infiltrate	Rare disease feature as the eye is generally a non-inflamed area
Skin[18]	Punch biopsy	70–90%	Features of mixed inflammatory infiltrate	Non-specific findings—eg, perivasculitis and acute and chronic inflammation without characteristic features of ANCA-associated vasculitis
Muscle[19]	Open	55–60%	Features of mixed inflammatory infiltrate	More likely positive in women and MPO-ANCA vasculitis

Biopsies taken from other sites are uncommon, but help to differentiate an ANCA-associated vasculitis diagnosis from other pathologies (ie, liver, prostate, or parotid gland diseases). ANCA=antineutrophil cytoplasmic antibody. MPO=myeloperoxidase. PR3=proteinase 3. *Only present in active vasculitis. †The diagnostic yield might have improved over the past decades and depends on the lesion (ie, higher yield when bronchial stenosis and active inflammation is visible). For the biopsy of other lesions, such as pulmonary granulomas, CT-guided biopsy might be preferred.

Table 1: **Diagnostic yield of biopsies taken from patients with PR3-ANCA-associated vasculitis and MPO-ANCA-associated vasculitis**

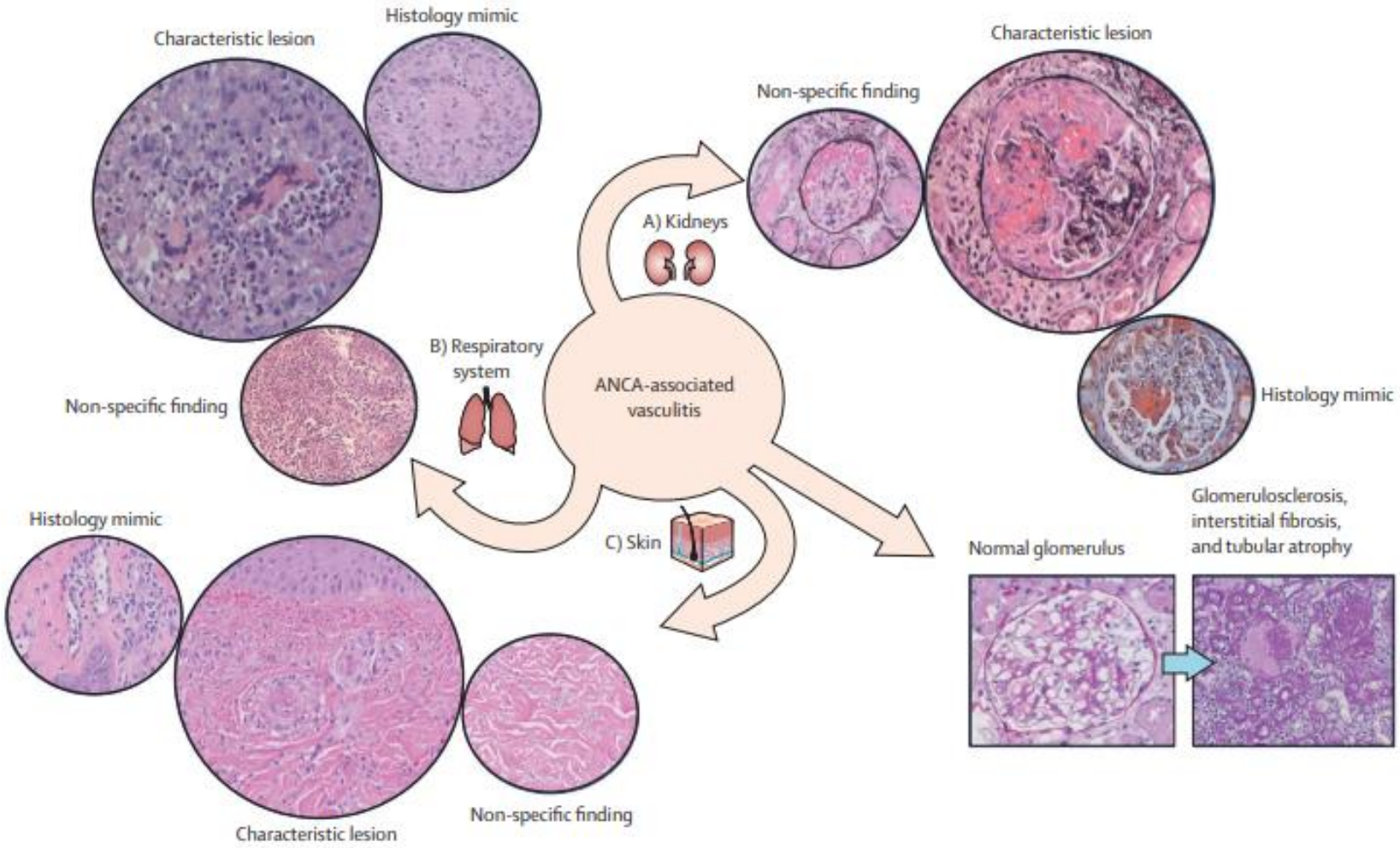
teilweise hohe diagnostische "Ausbeute"

wenig diagnostische Aussagekraft

Nierenhistologie meist sehr hilfreich

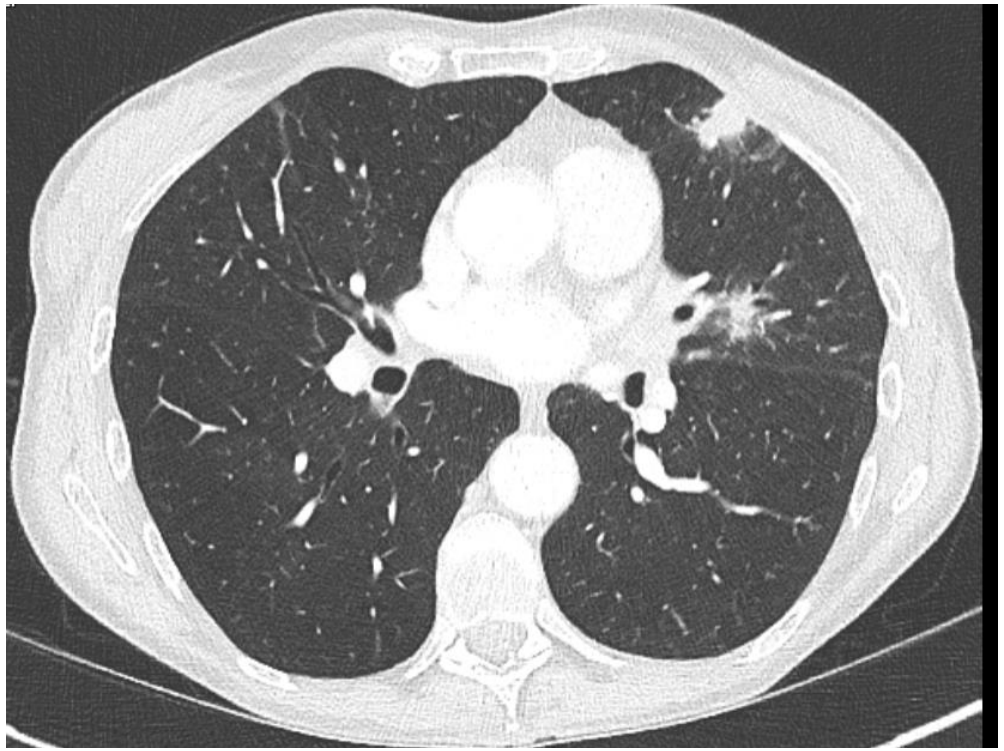
Kronbichler A, et al.
Lancet 2024;403(10427):683-698

AAV Histologie



Kronbichler A, et al.
Lancet 2024;403(10427):683-698

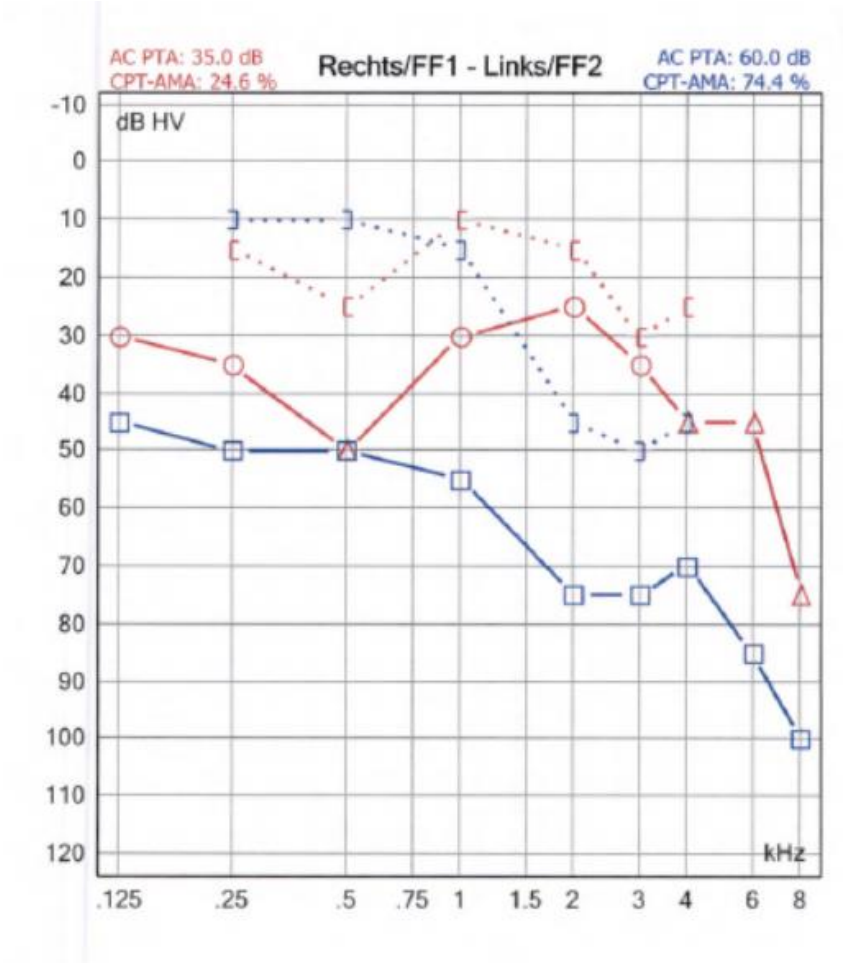
Frau S. C. 60 Jahre



- August 2023
- Husten, Auswurf
- Diagnose COP

- Antibiose
- kurzzeitige Glucocorticoidgabe

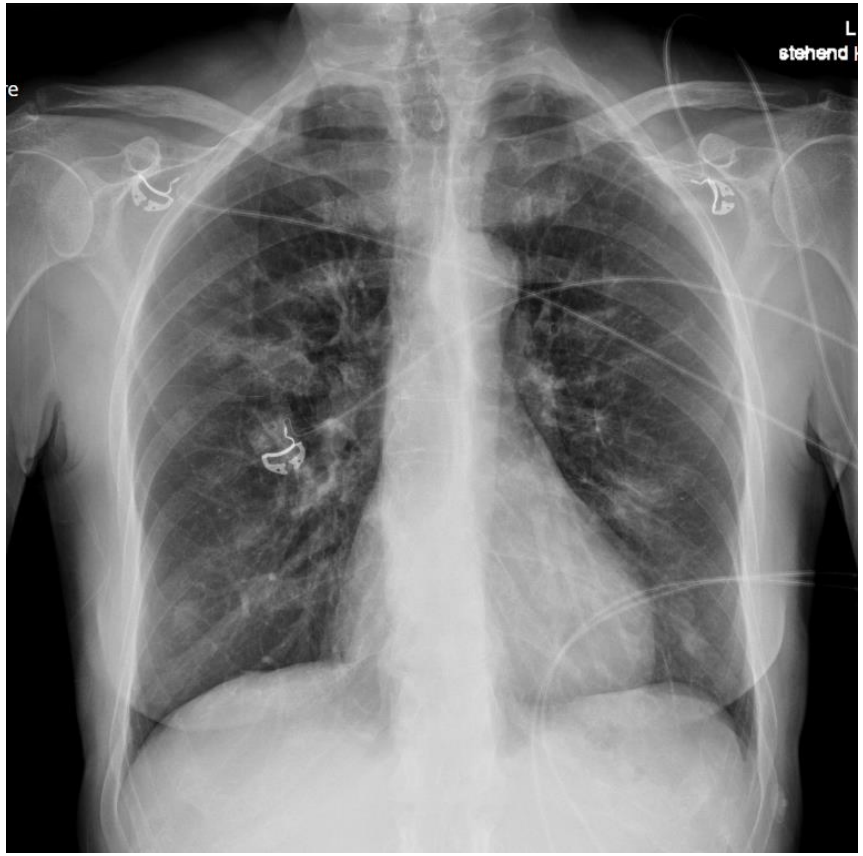
Frau S. C. 60 Jahre



- September 2024
- Ohrenschmerzen
- Hörminderung li > re
- Sekretion aus beiden Ohren

- Antibiose
- bei Diagnose Otitis media

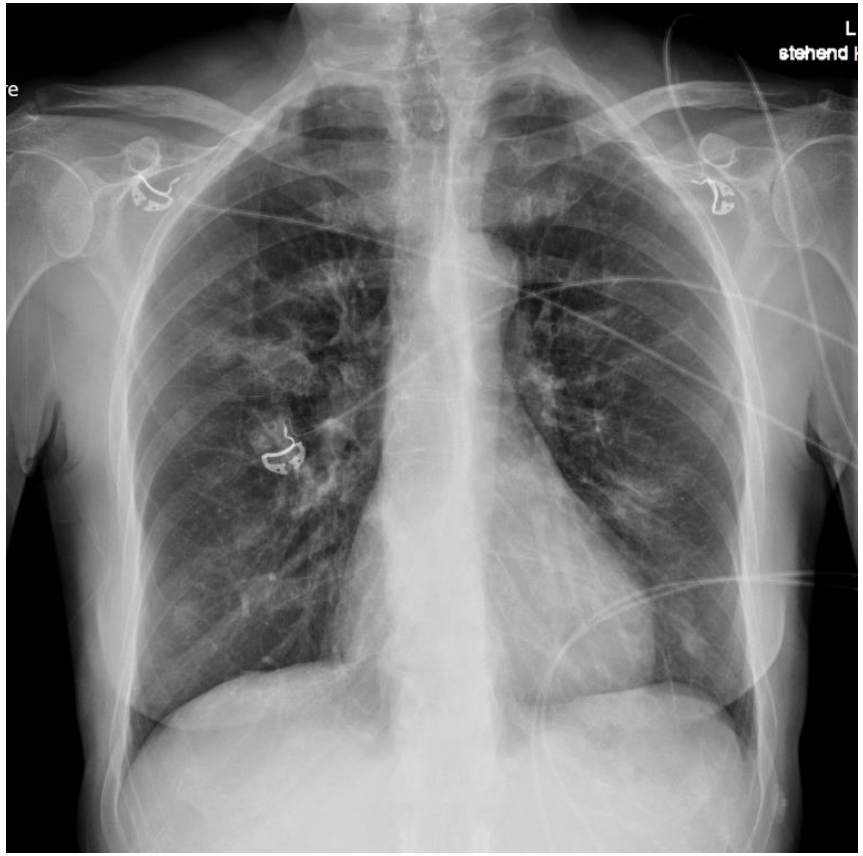
Frau S. C. 60 Jahre



- Oktober/November 2024
- Ohrenschmerzen therapierefraktär
- Hörminderung persistierend
- Sekretion persistierend

- Gelenkschmerzen stark ausgeprägt
- Schwellung Knie / OSG
- "Cortison" gering besser ! Osteoporose
- fraglich Infekt/Antibiotikaassoziiert

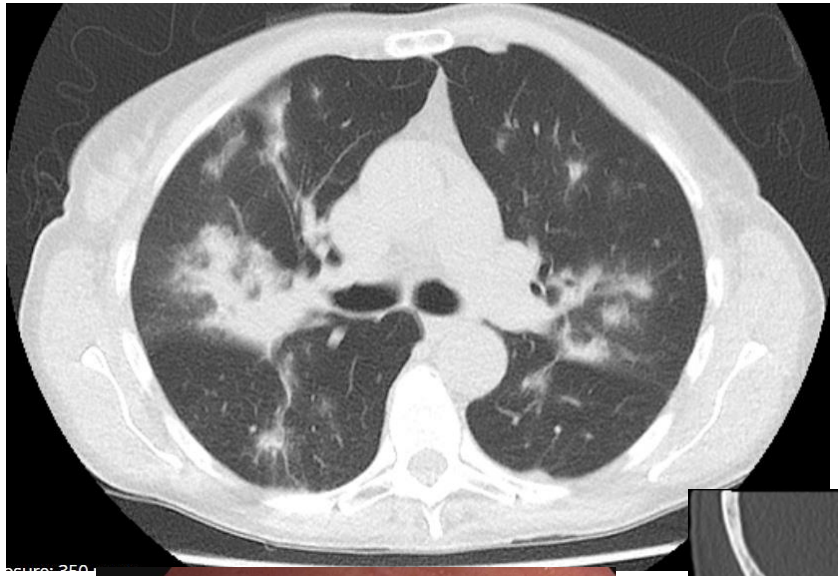
Frau S. C. 60 Jahre

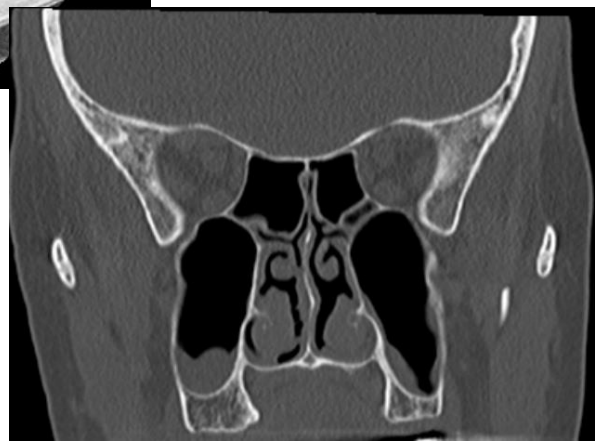


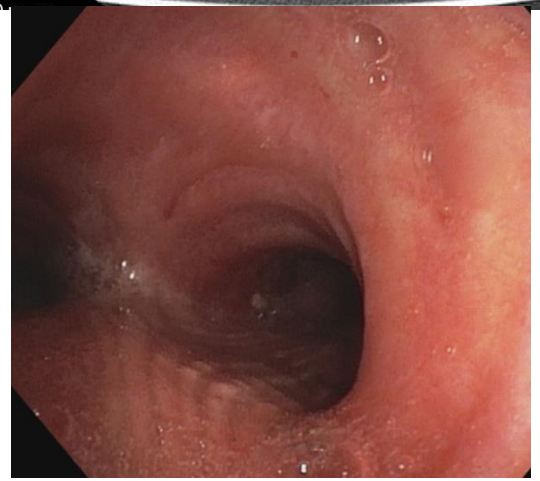
- November 2024
- Husten, Auswurf, teilweise blutig
- CrP Anstieg

- SARS-CoV2 positiv

Frau S. C. 60 Jahre



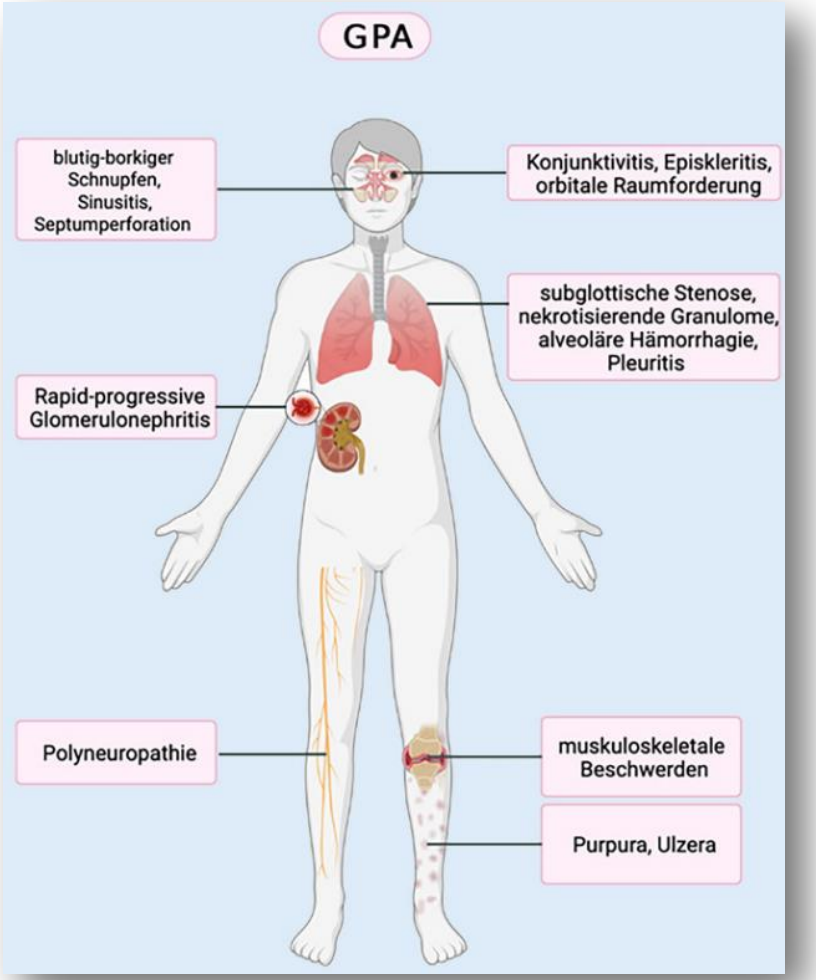




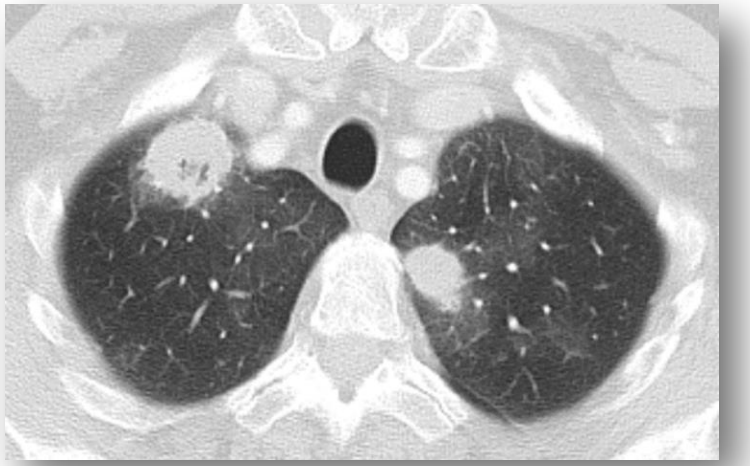
- November 2024
- Husten, Auswurf, teilweise blutig
- CrP Anstieg

- SARS-CoV2 positiv
- PR-3 ANCA positiv (203 U/ml, Norm <7)
- diskrete Erythrozyturie…

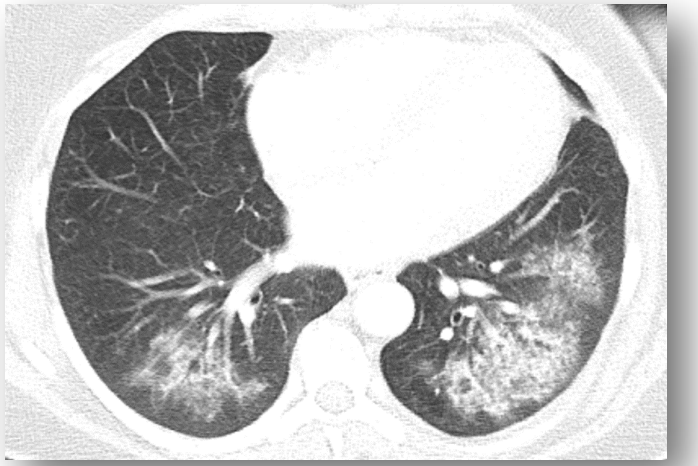
AAV => GPA



- klinisches "recognition pattern"
- ANCA
- Histologie
- Komplementdiagnostik?



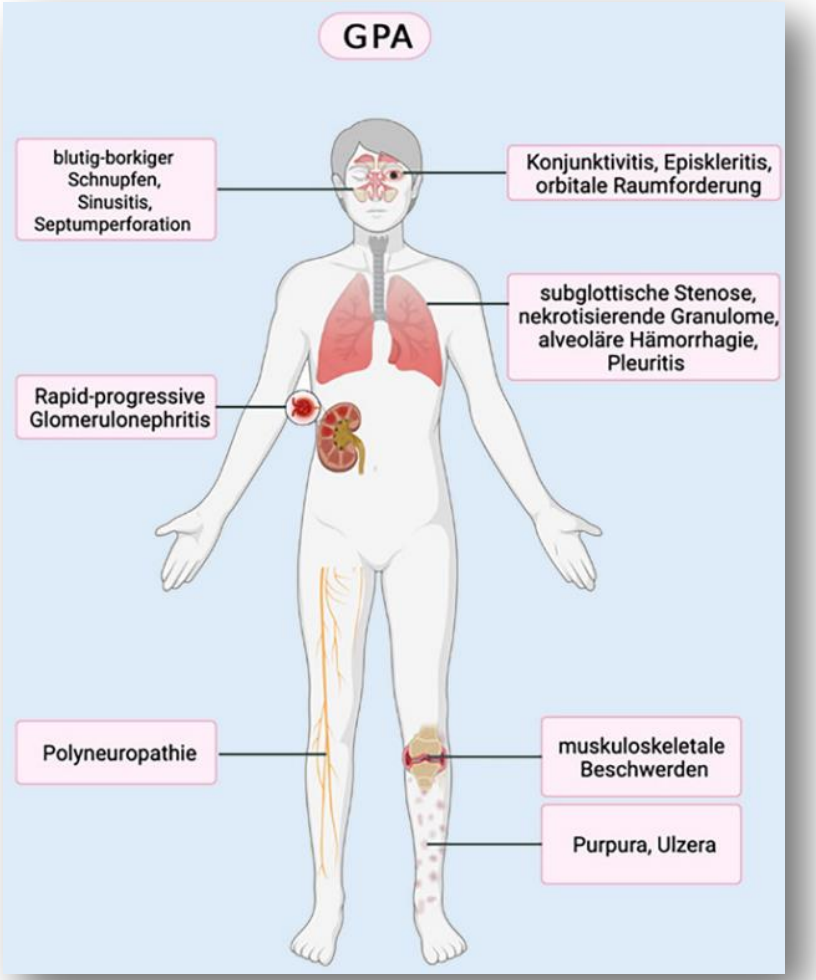
Pulmonale Granulome



Pulmonale Hämorrhagie

Krasselt M, Holle J, Innere Med 2022;9:947-960

AAV => GPA



Erkrankung	ACR/EULAR
Granulomatose mit Polyangiitis	*Klinisch:*
	Nasenbeteiligung mit blutigem Ausfluss, Ulzerationen, Verkrustungen, Verstopfung oder Septumdefekt/-perforation +3
	Knorpelbeteiligung mit Entzündung von Nasen-/Ohrknorpel, Heiserkeit, Stridor, endobronchialer Beteiligung oder Sattelnase +2
	Schallleitungs- oder sensorineuraler Hörverlust +1
	Laborchemisch/bildgebend/histologisch:
	Positive c-ANCA oder PR3-Antikörper +5
	Pulmonale Knötchen, Infiltrate oder Kavernen +2
	Granulome, extravaskuläre granulomatöse Entzündung, Riesenzellen +2
	Entzündung, Konsolidierung oder Erguss der (para-)nasalen Sinus oder des Mastoids +1
	Pauci-immune GN +1
	Positive p-ANCA oder MPO-Antikörper −1
	Periphere Eosinophilie ≥ $1 \cdot 10^9$/l −4
	≥ 5 Punkte für Klassifikation erforderlich

Krasselt M, Holle J, Innere Med 2022;9:947-960

AAV => Otitis media

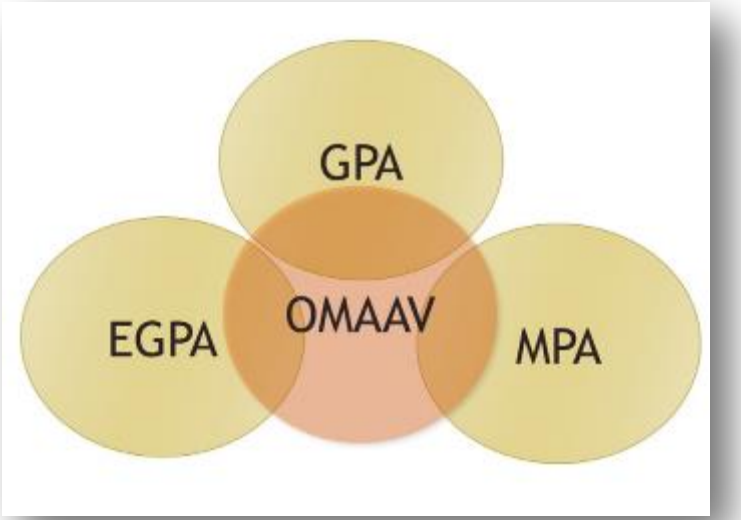
Contents lists available at ScienceDirect

Auris Nasus Larynx

journal homepage: www.elsevier.com/locate/anl

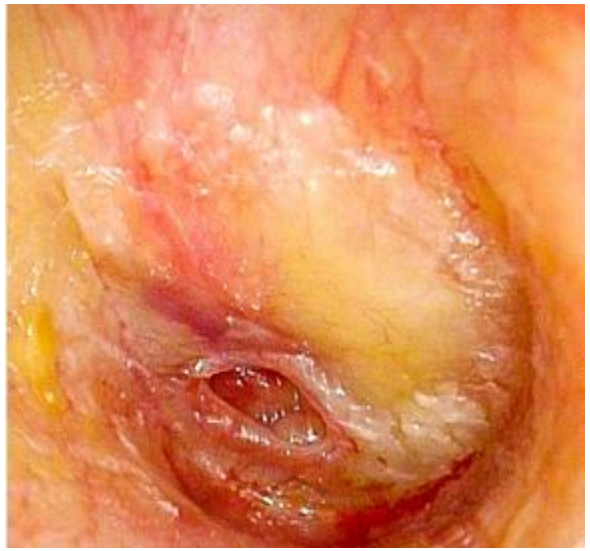
Clinical characteristics, the diagnostic criteria and management recommendation of otitis media with antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (OMAAV) proposed by Japan Otological Society

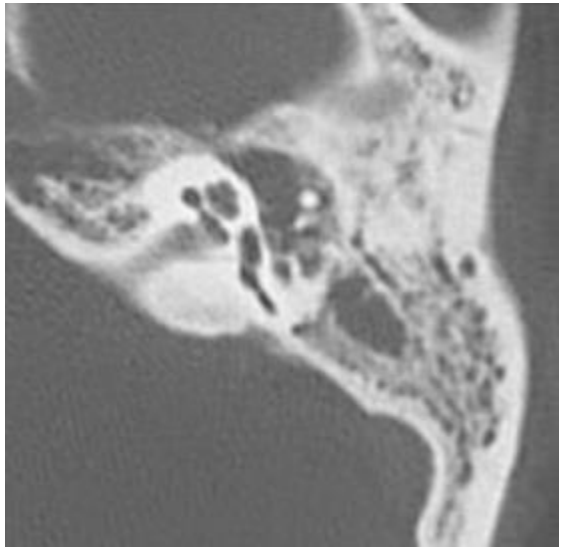
Yasuaki Harabuchi[a,*], Kan Kishibe[a], Kaori Tateyama[b], Yuka Morita[c],

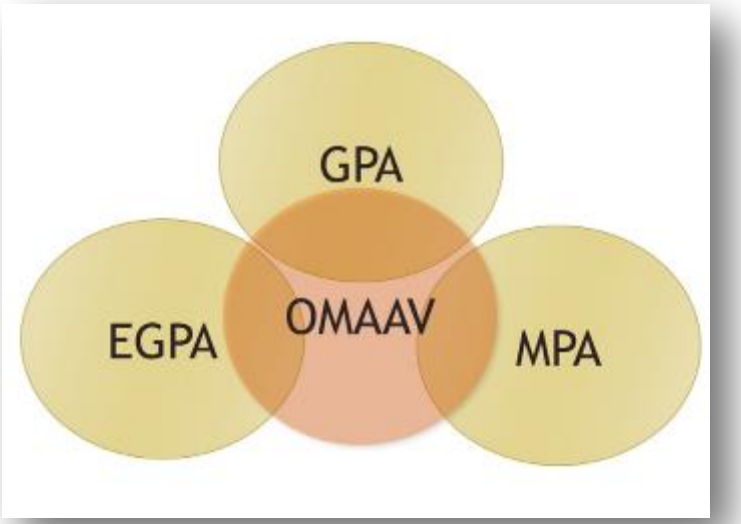


Yasuaki Harabuchi, et al, Auris Nasus Larynx 2021;2-14

AAV => Otitis media

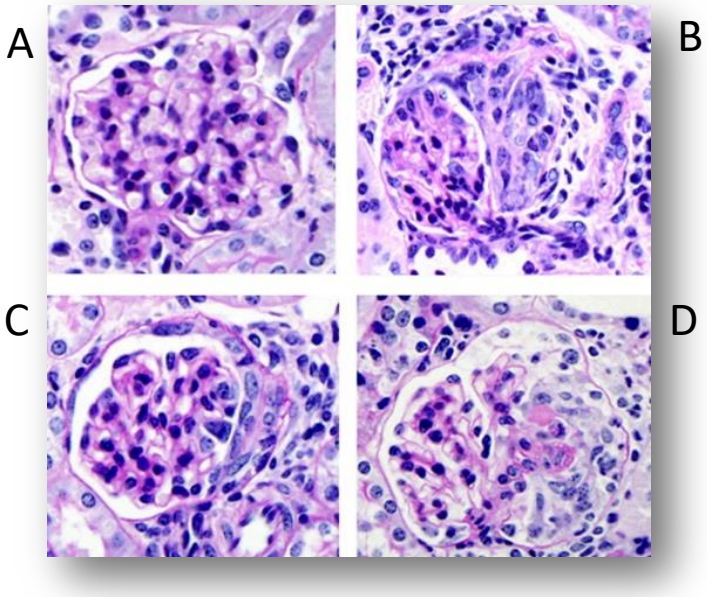






Yasuaki Harabuchi, et al, Auris Nasus Larynx 2021;2-14

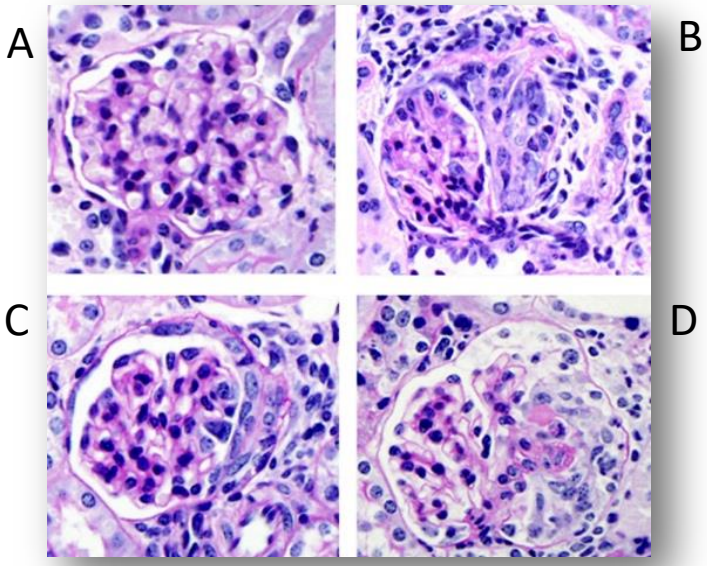
AAV Anti MPO und C5a Blockade -Mausmodell-

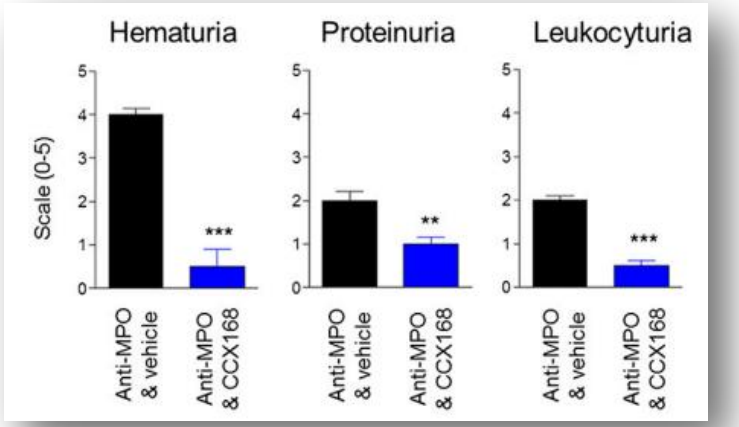


- Anti MPO induzierte AAV-GN in Mäusen

Xiao H et al 2014;25:225-231

AAV Anti MPO und C5a Blockade -Mausmodell-





- Anti MPO induzierte AAV-GN in Mäusen

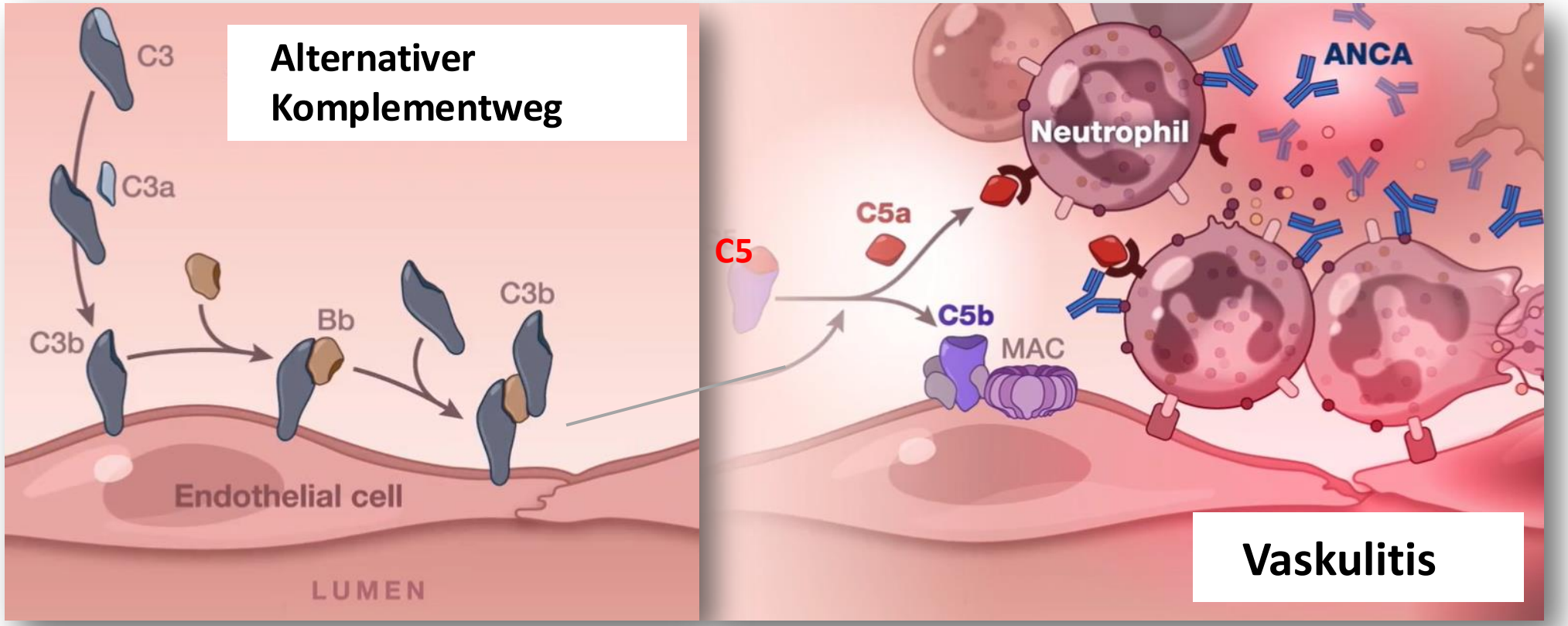
Halbmondbildung

- C5aR -/- Maus (A)
- C5L2 -/- Maus (B)
- C6 -/- Maus (C)
- hC5aR Maus (D)

- + Placebo / + CCX 168 (C5a Inhibitor)
- => "C5a Rezeptor Blockade schützt vor MPO-ANCA Glomerulonephritis"

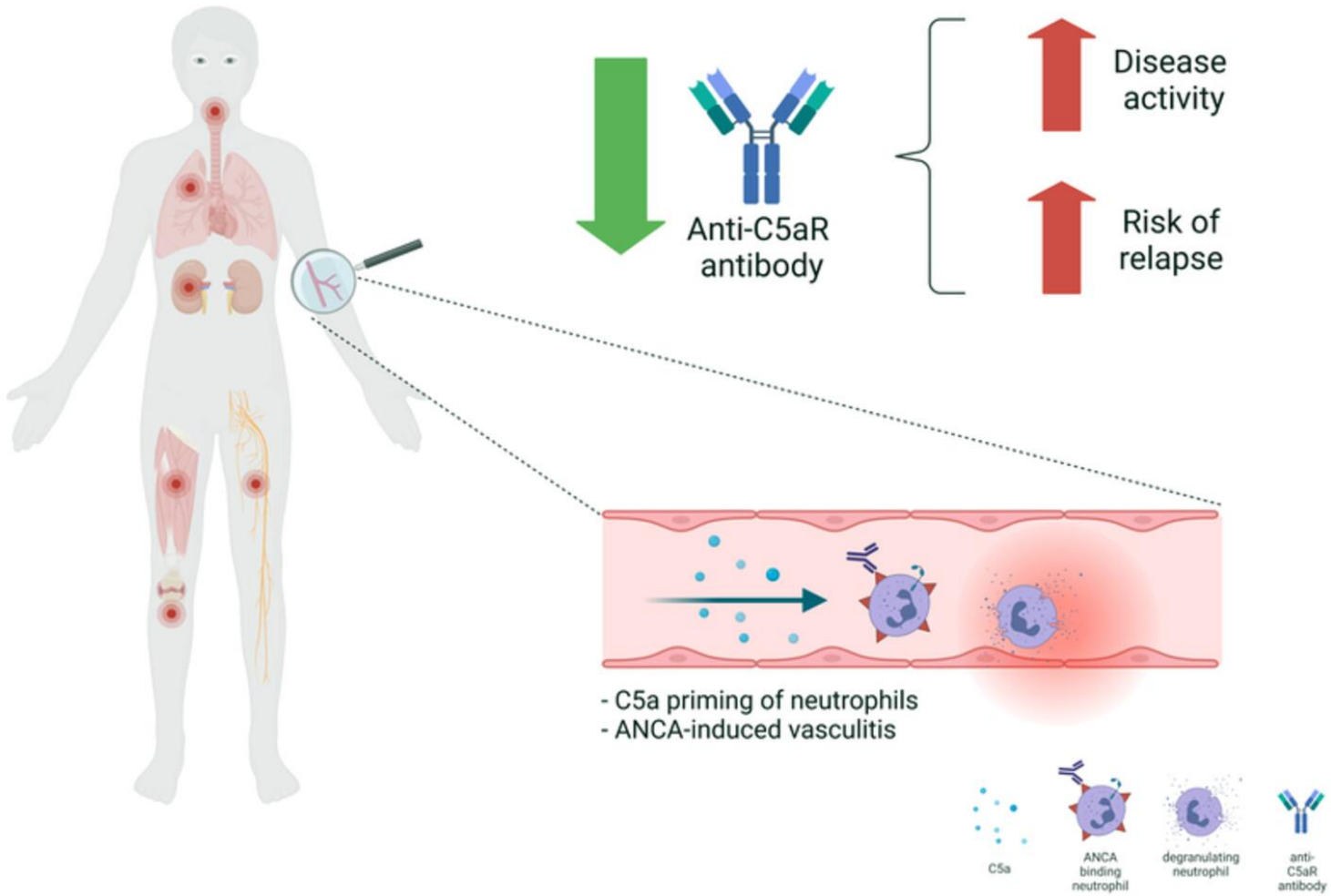
Xiao H et al 2014;25:225-231

AAV Rolle des Komplementsystems



Quick Take adaptiert aus
Jayne D et al 2021:NEJM 384:599-609

Low Concentrations of C5a Receptor Antibodies are Linked to Disease Activity and Relapse in ANCA-Associated Vasculitis



- Decreased concentrations of anti-C5aR antibodies are associated with high disease activity
- Anti-C5aR antibody concentrations <0.45 U/ml are associated with relapse

Klapa S, Müller A, Koch A, et al. Low concentrations of C5a complement receptor antibodies are linked to disease activity and relapse in ANCA-associated vasculitis. *Arthritis Rheumatol* 2023.

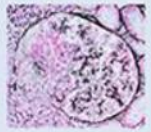
Arthritis & Rheumatology ACR American College of Rheumatology

Performance of urinary soluble CD163 to detect active ANCA glomerulonephritis

CJASN
Clinical Journal of the American Society of Nephrology

Limburg Renal Registry

ANCA GN (assessed by kidney biopsy)



Histological evaluation

Urinary soluble CD163

n = 95 patients

125 Kidney tissue sections

First kidney biopsy
n = 67

Repeat kidney biopsy
n = 58

Increased levels of urinary soluble CD163

87% Active ANCA GN (96/110)

7% Without active ANCA GN (1/15)

6% Healthy controls (1/17)

Urinary soluble CD163 correlates with

$r_s = 0.48$ *Spearman's rank (ρ) correlation analysis*
Fibrinoid Necrosis
(p<0.001)

$r_s = 0.70$ *Spearman's rank (ρ) correlation analysis*
Cellular Crescents
(p<0.001)

Recognition of relapsing ANCA GN

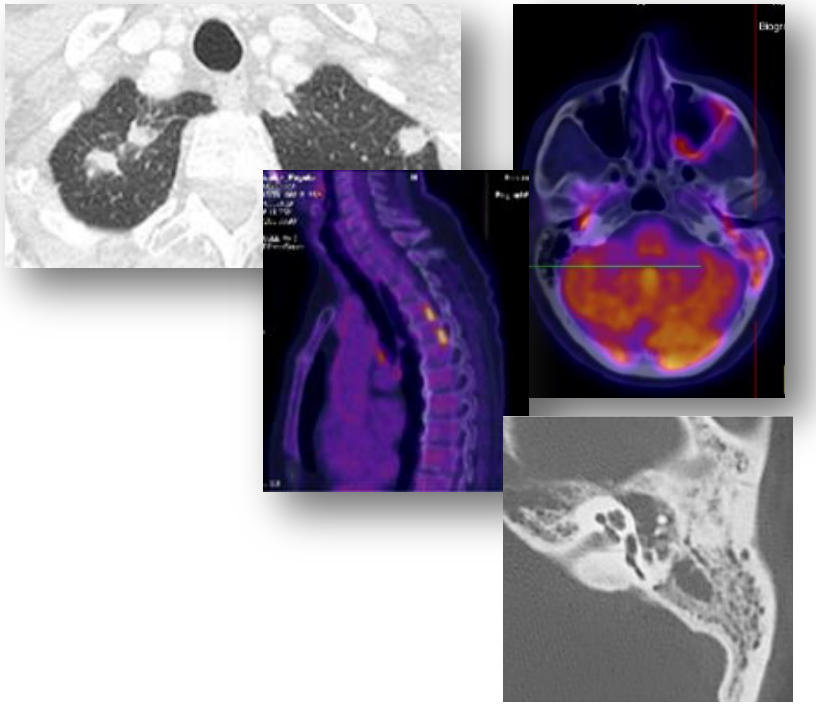
0.94 Sensitivity

0.91 Specificity

Conclusion Urinary soluble CD163 is associated with active ANCA GN and correlates with histological features as seen in ANCA GN.

Joop P. Aendekerk, Sjoerd A.M.E.G. Timmermans, Matthias H. Busch, et al. ***Urinary Soluble CD163 and Disease Activity in Biopsy Proven ANCA-Associated Glomerulonephritis***. CJASN doi: 10.2215/CJN.07210520.
Visual Abstract by Edgar Lerma, MD, FASN

…take home…



- pattern der AAV — kennen und erkennen
- bei klinischem Verdacht — ANCA incl Zielantigen
  antigenspezifischer assay
- "fast track clinic" — interdisziplinär/ Histologie / Bildgebung
  KEINE Therapieverzögerung durch Histologie
- Expertenzentrum — Zuweisung zu Dg /Therapieeinleitung
- zukünftige Diagnostik — Urinaktivitätsmarker ?
  komplementspezifische assays

«Mimicry» von Liis Roden

Liste der Referenzen

Alberici, Federico et al. “Treatment goals in ANCA-associated vasculitis: defining success in a new era.” Frontiers in immunology vol. 15 1409129. 13 Jun. 2024, doi:10.3389/fimmu.2024.1409129

Assmann, Gerd et al. “Effects of ezetimibe, simvastatin, atorvastatin, and ezetimibe-statin therapies on non-cholesterol sterols in patients with primary hypercholesterolemia.” Current medical research and opinion vol. 24,1 (2008): 249-59. doi:10.1185/030079908x253663

Basu N, et al. Fatigue: a principal contributor to impaired quality of life in ANCA-associated vasculitis. Rheumatology (Oxford). 2010 Jul;49(7):1383-90. doi: 10.1093/rheumatology/keq098.

Basu, Neil et al. Markers for work disability in anti-neutrophil cytoplasmic antibody-associated vasculitis, Rheumatology, Volume 53, Issue 5, May 2014, Pages 953–956, https://doi.org/10.1093/rheumatology/ket483

Bate, Sebastian et al. “The Improved Kidney Risk Score in ANCA-Associated Vasculitis for Clinical Practice and Trials.” Journal of the American Society of Nephrology : JASN vol. 35,3 (2024): 335-346. doi:10.1681/ASN.0000000000000274

Benichou, Nicolas et al. “Proteinuria and hematuria after remission induction are associated with outcome in ANCA-associated vasculitis.” Kidney international vol. 103,6 (2023): 1144-1155. doi:10.1016/j.kint.2023.02.029

Berden, Annelies E et al. “Histopathologic classification of ANCA-associated glomerulonephritis.” Journal of the American Society of Nephrology : JASN vol. 21,10 (2010): 1628-36. doi:10.1681/ASN.2010050477

Brar, Sandeep et al. “Association of Angiotensin-Converting Enzyme Inhibitor or Angiotensin Receptor Blocker Use With Outcomes After Acute Kidney Injury.” JAMA internal medicine vol. 178,12 (2018): 1681-1690. doi:10.1001/jamainternmed.2018.4749

rix SR. The Challenge of Assessing Remission and Relapse in ANCA Kidney Disease. J Am Soc Nephrol. 2024 Apr 1;35(4):395-397. doi: 10.1681/ASN.0000000000000331.

Caravaca-Fontán, Fernando et al. Sodium-glucose cotransporter 2 inhibition in primary and secondary glomerulonephritis, Nephrology Dialysis Transplantation, Volume 39, Issue 2, February 2024, Pages 328–340, https://doi.org/10.1093/ndt/gfad175

Cartin-Ceba, Rodrigo et al. “Diffuse Alveolar Hemorrhage Secondary to Antineutrophil Cytoplasmic Antibody-Associated Vasculitis: Predictors of Respiratory Failure and Clinical Outcomes.” Arthritis & rheumatology (Hoboken, N.J.) vol. 68,6 (2016): 1467-76. doi:10.1002/art.39562

Chalkia, Aglaia et al. “Avacopan for ANCA-associated vasculitis with hypoxic pulmonary haemorrhage.” Nephrology, dialysis, transplantation : official publication of the European Dialysis and Transplant Association - European Renal Association vol. 39,9 (2024): 1473-1482. doi:10.1093/ndt/gfae020

Chapman, Lara et al. Management of foot health in people with inflammatory arthritis: British Society for Rheumatology guideline scope, Rheumatology, Volume 61, Issue 10, October 2022, Pages 3907–3911, https://doi.org/10.1093/rheumatology/keac340

Clifford, Alison H, and Jan Willem Cohen Tervaert. “Cardiovascular events and the role of accelerated atherosclerosis in systemic vasculitis.” Atherosclerosis vol. 325 (2021): 8-15. doi:10.1016/j.atherosclerosis.2021.03.032

Liste der Referenzen

Cortazar, Frank B et al. “Renal Recovery for Patients with ANCA-Associated Vasculitis and Low eGFR in the ADVOCATE Trial of Avacopan.” Kidney international reports vol. 8,4 860-870. 3 Feb. 2023, doi:10.1016/j.ekir.2023.01.039

Davidson, Michael H et al. “Ezetimibe coadministered with simvastatin in patients with primary hypercholesterolemia.” Journal of the American College of Cardiology vol. 40,12 (2002): 2125-34. doi:10.1016/s0735-1097(02)02610-4

Dirikgil E, van Leeuwen JR, Bredewold OW, Ray A, Jonker JT, Soonawala D, Remmelts HHF, van Dam B, Bos WJ, van Kooten C, Rotmans J, Rabelink T, Teng YKO. ExploriNg DUrable Remission with Rituximab in ANCA-associatEd vasculitis (ENDURRANCE trial): protocol for a randomised controlled trial. BMJ Open. 2022 Sep 21;12(9):e061339. doi: 10.1136/bmjopen-2022-061339.

Engesser, Jonas et al. “Immune profiling-based targeting of pathogenic T cells with ustekinumab in ANCA-associated glomerulonephritis.” Nature communications vol. 15,1 8220. 19 Sep. 2024, doi:10.1038/s41467-024-52525-w

Falde, Sam D et al. “Treatment of Antineutrophil Cytoplasmic Antibody-Associated Vasculitis With Diffuse Alveolar Hemorrhage With Avacopan.” ACR open rheumatology vol. 6,10 (2024): 707-716. doi:10.1002/acr2.11726

Falde, Sam D et al. “Treatment of Antineutrophil Cytoplasmic Antibody-Associated Vasculitis With Diffuse Alveolar Hemorrhage With Avacopan.” ACR open rheumatology vol. 6,10 (2024): 707-716. doi:10.1002/acr2.11726

Fauci, A S et al. “Wegener's granulomatosis: prospective clinical and therapeutic experience with 85 patients for 21 years.” Annals of internal medicine vol. 98,1 (1983): 76-85. doi:10.7326/0003-4819-98-1-76

Ference, Brian A et al. “Low-density lipoproteins cause atherosclerotic cardiovascular disease. 1. Evidence from genetic, epidemiologic, and clinical studies. A consensus statement from the European Atherosclerosis Society Consensus Panel.” European heart journal vol. 38,32 (2017): 2459-2472. doi:10.1093/eurheartj/ehx144

Floege, Jürgen. “A new alternative: inhibiting complement activation in patients with IgA nephropathy.” Kidney international vol. 105,1 (2024): 28-30. doi:10.1016/j.kint.2023.10.012

Flossmann, Oliver et al. “Long-term patient survival in ANCA-associated vasculitis.” Annals of the rheumatic diseases vol. 70,3 (2011): 488-94. doi:10.1136/ard.2010.137778

Floyd, Lauren et al. “A systematic review of patient-reported outcome measures in patients with anti-neutrophil cytoplasmic antibody associated vasculitis.” Rheumatology (Oxford, England) vol. 63,10 (2024): 2624-2637. doi:10.1093/rheumatology/keae069

Furuta, Shunsuke et al. “Reduced-dose versus high-dose glucocorticoids added to rituximab on remission induction in ANCA-associated vasculitis: predefined 2-year follow-up study.” Annals of the rheumatic diseases vol. 83,1 96-102. 2 Jan. 2024, doi:10.1136/ard-2023-224343

Fussner, Lynn A et al. “Alveolar Hemorrhage in Antineutrophil Cytoplasmic Antibody-Associated Vasculitis: Results of an International Randomized Controlled Trial (PEXIVAS).” American journal of respiratory and critical care medicine vol. 209,9 (2024): 1141-1151. doi:10.1164/rccm.202308-1426OC

Gisslander K, et al. Data-driven subclassification of ANCA-associated vasculitis: model-based clustering of a federated international cohort. Lancet Rheumatol. 2024 Nov;6(11):e762-e770. doi: 10.1016/S2665-9913(24)00187-5.

Liste der Referenzen

Gopaluni, Seerapani et al. “Effect of Disease Activity at Three and Six Months After Diagnosis on Long-Term Outcomes in Antineutrophil Cytoplasmic Antibody-Associated Vasculitis.” Arthritis & rheumatology (Hoboken, N.J.) vol. 71,5 (2019): 784-791. doi:10.1002/art.40776

Grahammer, Florian et al. “The podocyte slit diaphragm--from a thin grey line to a complex signalling hub.” Nature reviews. Nephrology vol. 9,10 (2013): 587-98. doi:10.1038/nrneph.2013.169

Hakroush, Samy et al. “Bowman's capsule rupture links glomerular damage to tubulointerstitial inflammation in ANCA-associated glomerulonephritis.” Clinical and experimental rheumatology vol. 39 Suppl 129,2 (2021): 27-31. doi:10.55563/clinexprheumatol/7eol6d

Harper, Lorraine et al. “Treatment of fatigue with physical activity and behavioural change support in vasculitis: study protocol for an open-label randomised controlled feasibility study.” BMJ open vol. 8,10 e023769. 30 Oct. 2018, doi:10.1136/bmjopen-2018-023769

Harper, Lorraine et al. Management of fatigue with physical activity and behavioural change support in vasculitis: a feasibility study, Rheumatology, Volume 60, Issue 9, September 2021, Pages 4130–4140, https://doi.org/10.1093/rheumatology/keaa890

Hayek, Salim S et al. “Soluble Urokinase Receptor and Acute Kidney Injury.” The New England journal of medicine vol. 382,5 (2020): 416-426. doi:10.1056/NEJMoa1911481

Heijl, Caroline et al. “Long-term patient survival in a Swedish population-based cohort of patients with ANCA-associated vasculitis.” RMD open vol. 3,1 e000435. 13 Jul. 2017, doi:10.1136/rmdopen-2017-000435

Hellmich, Bernhard et al. “EULAR recommendations for the management of ANCA-associated vasculitis: 2022 update.” Annals of the rheumatic diseases vol. 83,1 30-47. 2 Jan. 2024, doi:10.1136/ard-2022-223764

Heron, Vanessa et al. “The impact of antineutrophil cytoplasmic antibody-associated vasculitis on employment and work disability in an Australian population.” International journal of rheumatic diseases vol. 24,7 (2021): 904-911. doi:10.1111/1756-185X.14131

Hiepe, Falk, and Andreas Radbruch. “Plasma cells as an innovative target in autoimmune disease with renal manifestations.” Nature reviews. Nephrology vol. 12,4 (2016): 232-40. doi:10.1038/nrneph.2016.20

Hoffman, G S et al. “Wegener granulomatosis: an analysis of 158 patients.” Annals of internal medicine vol. 116,6 (1992): 488-98. doi:10.7326/0003-4819-116-6-488

Hollander, D, and R T Manning. “The use of alkylating agents in the treatment of Wegener's granulomatosis.” Annals of internal medicine vol. 67,2 (1967): 393-8. doi:10.7326/0003-4819-67-2-393

Holle et al. S3-Leitlinie der Deutschen Gesellschaft für Rheumatologie und Klinische Immunologie e. V. (DGRh). Diagnostik und Therapie der ANCA-assoziierten Vaskulitiden, Version: 1.1; Stand: 12.08.2024

Hruskova, Zdenka et al. “Characteristics and Outcomes of Granulomatosis With Polyangiitis (Wegener) and Microscopic Polyangiitis Requiring Renal Replacement Therapy: Results From the European Renal Association-European Dialysis and Transplant Association Registry.” American journal of kidney diseases : the official journal of the National Kidney Foundation vol. 66,4 (2015): 613-20. doi:10.1053/j.ajkd.2015.03.025

Iudici, Michele et al. “Granulomatosis with polyangiitis: Study of 795 patients from the French Vasculitis Study Group registry.” Seminars in arthritis and rheumatism vol. 51,2 (2021): 339-346. doi:10.1016/j.semarthrit.2021.02.002

Becherucci, Francesca et al. “A Clinical Workflow for Cost-Saving High-Rate Diagnosis of Genetic Kidney Diseases.” Journal of the American Society of Nephrology : JASN vol. 34,4 (2023): 706-720. doi:10.1681/ASN.0000000000000076

Liste der Referenzen

Jayne, David R W et al. “Avacopan for the Treatment of ANCA-Associated Vasculitis.” The New England journal of medicine vol. 384,7 (2021): 599-609. doi:10.1056/NEJMoa2023386

Jayne, David R W et al. “Randomized Trial of C5a Receptor Inhibitor Avacopan in ANCA-Associated Vasculitis.” Journal of the American Society of Nephrology : JASN vol. 28,9 (2017): 2756-2767. doi:10.1681/ASN.2016111179

Jayne, David R W et al. “Avacopan for the Treatment of ANCA-Associated Vasculitis.” The New England journal of medicine vol. 384,7 (2021): 599-609. doi:10.1056/NEJMoa2023386

Jayne, David R W et al. “Randomized trial of plasma exchange or high-dosage methylprednisolone as adjunctive therapy for severe renal vasculitis.” Journal of the American Society of Nephrology : JASN vol. 18,7 (2007): 2180-8. doi:10.1681/ASN.2007010090

Jayne, David R W et al. “Avacopan for the Treatment of ANCA-Associated Vasculitis.” The New England journal of medicine vol. 384,7 (2021): 599-609. doi:10.1056/NEJMoa2023386

June, Carl H et al. “CAR T cell immunotherapy for human cancer.” Science (New York, N.Y.) vol. 359,6382 (2018): 1361-1365. doi:10.1126/science.aar6711

Junek, Mats L et al. “Risk of Relapse of Antineutrophil Cytoplasmic Antibody-Associated Vasculitis in a Randomized Controlled Trial of Plasma Exchange and Glucocorticoids.” Arthritis & rheumatology (Hoboken, N.J.) vol. 76,9 (2024): 1431-1438. doi:10.1002/art.42843

Kaufeld, Jessica et al. “Atypical Hemolytic and Uremic Syndrome Triggered by Infection With SARS-CoV2.” Kidney international reports vol. 6,10 (2021): 2709-2712. doi:10.1016/j.ekir.2021.07.004

Klapa, Sebastian et al. “Low Concentrations of C5a Complement Receptor Antibodies Are Linked to Disease Activity and Relapse in Antineutrophil Cytoplasmic Autoantibody-Associated Vasculitis.” Arthritis & rheumatology (Hoboken, N.J.) vol. 75,5 (2023): 760-767. doi:10.1002/art.42410

Kochi, Masako et al. “Chronic kidney disease, inflammation, and cardiovascular disease risk in rheumatoid arthritis.” Journal of cardiology vol. 71,3 (2018): 277-283. doi:10.1016/j.jjcc.2017.08.008

Krasselt, Marco et al. “48-jährige Patientin mit krustenartigen Veränderungen an den Füßen” [48-year-old woman with crusty alterations on the feet]. Deutsche medizinische Wochenschrift (1946) vol. 148,18 (2023): 1155-1156. doi:10.1055/a-2061-5060

Krasselt, Marco L, and Julia U Holle. “ANCA-assoziierte Vaskulitis” [ANCA-associated vasculitis]. Innere Medizin (Heidelberg, Germany) vol. 63,9 (2022): 947-960. doi:10.1007/s00108-022-01386-w

Kronbichler, Andreas et al. “Diagnosis and management of ANCA-associated vasculitis.” Lancet (London, England) vol. 403,10427 (2024): 683-698. doi:10.1016/S0140-6736(23)01736-1

Kronbichler, Andreas et al. “Plasma exchange in ANCA-associated vasculitis: the pro position.” Nephrology, dialysis, transplantation : official publication of the European Dialysis and Transplant Association - European Renal Association vol. 36,2 (2021): 227-231. doi:10.1093/ndt/gfaa311

Kronbichler, Andreas et al. “Diagnosis and management of ANCA-associated vasculitis.” Lancet (London, England) vol. 403,10427 (2024): 683-698. doi:10.1016/S0140-6736(23)01736-1

Liste der Referenzen

L'Imperio, Vincenzo et al. “Bowman's capsule rupture on renal biopsy improves the outcome prediction of ANCA-associated glomerulonephritis classifications.” Annals of the rheumatic diseases vol. 81,6 (2022): e95. doi:10.1136/annrheumdis-2020-217979

Lejeune, Margaux et al. “Bispecific, T-Cell-Recruiting Antibodies in B-Cell Malignancies.” Frontiers in immunology vol. 11 762. 7 May. 2020, doi:10.3389/fimmu.2020.00762

Little, Mark A et al. “Early mortality in systemic vasculitis: relative contribution of adverse events and active vasculitis.” Annals of the rheumatic diseases vol. 69,6 (2010): 1036-43. doi:10.1136/ard.2009.109389

Mach, François et al. “2019 ESC/EAS Guidelines for the management of dyslipidaemias: lipid modification to reduce cardiovascular risk.” European heart journal vol. 41,1 (2020): 111-188. doi:10.1093/eurheartj/ehz455

Manolio, Teri A et al. “Finding the missing heritability of complex diseases.” Nature vol. 461,7265 (2009): 747-53. doi:10.1038/nature08494

Maunz, Annika et al. “Association of the AAV-PRO questionnaire with established outcome measures in AAV.” Rheumatology (Oxford, England) vol. 63,1 (2024): 174-180. doi:10.1093/rheumatology/kead199

McAdoo, Stephen P et al. “Long-term follow-up of a combined rituximab and cyclophosphamide regimen in renal anti-neutrophil cytoplasm antibody-associated vasculitis.” Nephrology, dialysis, transplantation : official publication of the European Dialysis and Transplant Association - European Renal Association vol. 34,1 (2019): 63-73. doi:10.1093/ndt/gfx378

McGregor, JulieAnne G et al. “Adverse events and infectious burden, microbes and temporal outline from immunosuppressive therapy in antineutrophil cytoplasmic antibody-associated vasculitis with native renal function.” Nephrology, dialysis, transplantation : official publication of the European Dialysis and Transplant Association - European Renal Association vol. 30 Suppl 1,Suppl 1 (2015): i171-81. doi:10.1093/ndt/gfv045

Monti, Sara et al. “Factors influencing patient-reported outcomes in ANCA-associated vasculitis: correlates of the Patient Global Assessment.” Seminars in arthritis and rheumatism vol. 56 (2022): 152048. doi:10.1016/j.semarthrit.2022.152048

Moran, Sarah M et al. “The Clinical Application of Urine Soluble CD163 in ANCA-Associated Vasculitis.” Journal of the American Society of Nephrology : JASN vol. 32,11 (2021): 2920-2932. doi:10.1681/ASN.2021030382

Casal Moura, Marta et al. “Management of antineutrophil cytoplasmic antibody-associated vasculitis with glomerulonephritis as proposed by the ACR 2021, EULAR 2022 and KDIGO 2021 guidelines/recommendations.” Nephrology, dialysis, transplantation : official publication of the European Dialysis and Transplant Association - European Renal Association vol. 38,11 (2023): 2637-2651. doi:10.1093/ndt/gfad090

Casal Moura, Marta et al. “Maintenance of Remission and Risk of Relapse in Myeloperoxidase-Positive ANCA-Associated Vasculitis with Kidney Involvement.” Clinical journal of the American Society of Nephrology : CJASN vol. 18,1 (2023): 47-59. doi:10.2215/CJN.06460622

Casal Moura, Marta et al. “Maintenance of Remission and Risk of Relapse in Myeloperoxidase-Positive ANCA-Associated Vasculitis with Kidney Involvement.” Clinical journal of the American Society of Nephrology : CJASN vol. 18,1 (2023): 47-59. doi:10.2215/CJN.06460622

Nicholls, Stephen J et al. “Effect of Evolocumab on Progression of Coronary Disease in Statin-Treated Patients: The GLAGOV Randomized Clinical Trial.” JAMA vol. 316,22 (2016): 2373-2384. doi:10.1001/jama.2016.16951

O'Malley, Lucy et al. “The Longitudinal Course of Fatigue in Antineutrophil Cytoplasmic Antibody-associated Vasculitis.” The Journal of rheumatology vol. 47,4 (2020): 572-579. doi:10.3899/jrheum.190113

Liste der Referenzen

Hildreth, Andrew D et al. “Single-cell sequencing of human white adipose tissue identifies new cell states in health and obesity.” Nature immunology vol. 22,5 (2021): 639-653. doi:10.1038/s41590-021-00922-4

O'Sullivan, Kim M, and Stephen R Holdsworth. “Neutrophil Extracellular Traps: A Potential Therapeutic Target in MPO-ANCA Associated Vasculitis?.” Frontiers in immunology vol. 12 635188. 15 Mar. 2021, doi:10.3389/fimmu.2021.635188

Odler, Balazs et al. “Risk factors for serious infections in ANCA-associated vasculitis.” Annals of the rheumatic diseases vol. 82,5 (2023): 681-687. doi:10.1136/ard-2022-223401

Odler, Balazs et al. “Challenges of defining renal response in ANCA-associated vasculitis: call to action?.” Clinical kidney journal vol. 16,6 965-975. 11 Jan. 2023, doi:10.1093/ckj/sfad009

Oristrell, J et al. “Relapse rate and renal prognosis in ANCA-associated vasculitis according to long-term ANCA patterns.” Clinical and experimental immunology vol. 203,2 (2021): 209-218. doi:10.1111/cei.13530

Ostendorf, Lennard et al. “Daratumumab for the treatment of refractory ANCA-associated vasculitis.” RMD open vol. 9,1 (2023): e002742. doi:10.1136/rmdopen-2022-002742

Perkovic, Vlado et al. “Alternative Complement Pathway Inhibition with Iptacopan in IgA Nephropathy.” The New England journal of medicine, 10.1056/NEJMoa2410316. 25 Oct. 2024, doi:10.1056/NEJMoa2410316

Perna, Alessandro et al. “Kidney transplantation in patients with ANCA-associated vasculitis is associated with a high incidence of post-transplant cancer.” Journal of nephrology vol. 37,6 (2024): 1611-1619. doi:10.1007/s40620-024-01951-6

Pittam, Bradley et al. “The prevalence and impact of depression in primary systemic vasculitis: a systematic review and meta-analysis.” Rheumatology international vol. 40,8 (2020): 1215-1221. doi:10.1007/s00296-020-04611-7

Prskalo, Luka et al. “Urinary CD4 + T Cells Predict Renal Relapse in ANCA-Associated Vasculitis.” Journal of the American Society of Nephrology : JASN vol. 35,4 (2024): 483-494. doi:10.1681/ASN.0000000000000311

Quartuccio, Luca et al. “Unmet needs in ANCA-associated vasculitis: Physicians' and patients' perspectives.” Frontiers in immunology vol. 14 1112899. 23 Feb. 2023, doi:10.3389/fimmu.2023.1112899

Quartuccio, Luca et al. “Alveolar haemorrhage in ANCA-associated vasculitis: Long-term outcome and mortality predictors.” Journal of autoimmunity vol. 108 (2020): 102397. doi:10.1016/j.jaut.2019.102397

Quinn, Kaitlin A et al. “An international Delphi exercise to identify items of importance for measuring response to treatment in ANCA-associated vasculitis.” Seminars in arthritis and rheumatism vol. 55 (2022): 152021. doi:10.1016/j.semarthrit.2022.152021

Ridker, Paul M et al. “Inhibition of Interleukin-1β by Canakinumab and Cardiovascular Outcomes in Patients With Chronic Kidney Disease.” Journal of the American College of Cardiology vol. 71,21 (2018): 2405-2414. doi:10.1016/j.jacc.2018.03.490

Ridker, Paul M et al. “Inhibition of Interleukin-1β by Canakinumab and Cardiovascular Outcomes in Patients With Chronic Kidney Disease.” Journal of the American College of Cardiology vol. 71,21 (2018): 2405-2414. doi:10.1016/j.jacc.2018.03.490

Robson, Joanna et al. “Damage in the anca-associated vasculitides: long-term data from the European vasculitis study group (EUVAS) therapeutic trials.” Annals of the rheumatic diseases vol. 74,1 (2015): 177-84. doi:10.1136/annrheumdis-2013-203927

Liste der Referenzen

Sagmeister, Michael S et al. “Kidney transplantation in ANCA-associated vasculitis.” Journal of nephrology vol. 32,6 (2019): 919-926. doi:10.1007/s40620-019-00642-x

Salmela, Anna et al. “Prognostic Factors for Survival and Relapse in ANCA-Associated Vasculitis with Renal Involvement: A Clinical Long-Term Follow-Up Study.” International journal of nephrology vol. 2018 6369814. 16 Oct. 2018, doi:10.1155/2018/6369814

Sachez-Alamo, Beatriz et al. “Long-term outcome of kidney function in patients with ANCA-associated vasculitis.” Nephrology, dialysis, transplantation : official publication of the European Dialysis and Transplant Association - European Renal Association vol. 39,9 (2024): 1483-1493. doi:10.1093/ndt/gfae018

Sachez-Alamo, Beatriz et al. “Long-term outcome of kidney function in patients with ANCA-associated vasculitis.” Nephrology, dialysis, transplantation : official publication of the European Dialysis and Transplant Association - European Renal Association vol. 39,9 (2024): 1483-1493. doi:10.1093/ndt/gfae018

Sánchez Álamo, Beatriz et al. “Long-term outcomes and prognostic factors for survival of patients with ANCA-associated vasculitis.” Nephrology, dialysis, transplantation : official publication of the European Dialysis and Transplant Association - European Renal Association vol. 38,7 (2023): 1655-1665. doi:10.1093/ndt/gfac320
Sayer, Matthew et al. “Cardiovascular Disease in Anti-neutrophil Cytoplasm Antibody-Associated Vasculitis.” Current rheumatology reports vol. 26,1 (2024): 12-23. doi:10.1007/s11926-023-01123-8

Schäfer, Ann-Kathrin et al. “Case Report: High-dose immunoglobulins prior to plasma exchange in severe pulmonary renal syndrome.” Frontiers in immunology vol. 14 1210321. 9 Jun. 2023, doi:10.3389/fimmu.2023.1210321

Scherbacher, Paul J et al. “Prospective study of complications and sequelae of glucocorticoid therapy in ANCA-associated vasculitis.” RMD open vol. 10,1 e003956. 29 Feb. 2024, doi:10.1136/rmdopen-2023-003956

Schunk, Stefan J et al. “Measurement of urinary Dickkopf-3 uncovered silent progressive kidney injury in patients with chronic obstructive pulmonary disease.” Kidney international vol. 100,5 (2021): 1081-1091. doi:10.1016/j.kint.2021.06.029

Schunk, Stefan J et al. “Association between urinary dickkopf-3, acute kidney injury, and subsequent loss of kidney function in patients undergoing cardiac surgery: an observational cohort study.” Lancet (London, England) vol. 394,10197 (2019): 488-496. doi:10.1016/S0140-6736(19)30769-X

Speer, Thimoteus et al. “Urinary DKK3 as a biomarker for short-term kidney function decline in children with chronic kidney disease: an observational cohort study.” The Lancet. Child & adolescent health vol. 7,6 (2023): 405-414. doi:10.1016/S2352-4642(23)00049-4

Schunk, Stefan J et al. “WNT-β-catenin signalling - a versatile player in kidney injury and repair.” Nature reviews. Nephrology vol. 17,3 (2021): 172-184. doi:10.1038/s41581-020-00343-w

Schupp, Jonas Christian et al. “Usefulness of Cyclophosphamide Pulse Therapy in Interstitial Lung Diseases.” Respiration; international review of thoracic diseases vol. 91,4 (2016): 296-301. doi:10.1159/000445031

Shochet, Lani et al. “Animal Models of ANCA Associated Vasculitis.” Frontiers in immunology vol. 11 525. 9 Apr. 2020, doi:10.3389/fimmu.2020.00525

Silva, Rita M et al. “Renal Transplantation in Antineutrophil Cytoplasmic Antibody-Associated Vasculitis: A Single-Center 10-Year Experience.” Transplantation proceedings vol. 55,6 (2023): 1396-1399. doi:10.1016/j.transproceed.2023.04.018

Liste der Referenzen

Simms-Williams, Nikita et al. “Effect of combination treatment with glucagon-like peptide-1 receptor agonists and sodium-glucose cotransporter-2 inhibitors on incidence of cardiovascular and serious renal events: population based cohort study.” BMJ (Clinical research ed.) vol. 385 e078242. 25 Apr. 2024, doi:10.1136/bmj-2023-078242

Slot, Marjan C et al. “Renal survival and prognostic factors in patients with PR3-ANCA associated vasculitis with renal involvement.” Kidney international vol. 63,2 (2003): 670-7. doi:10.1046/j.1523-1755.2003.00769.x

Smith, Rona M et al. “Rituximab versus azathioprine for maintenance of remission for patients with ANCA-associated vasculitis and relapsing disease: an international randomised controlled trial.” Annals of the rheumatic diseases vol. 82,7 (2023): 937-944. doi:10.1136/ard-2022-223559

Sonnemann, Janis et al. “Urinary T Cells Identify Renal Antineutrophil Cytoplasmic Antibody-Associated Vasculitis and Predict Prognosis: A Proof of Concept Study.” Kidney international reports vol. 8,4 871-883. 18 Jan. 2023, doi:10.1016/j.ekir.2023.01.013

Specks, Ulrich et al. “Efficacy of remission-induction regimens for ANCA-associated vasculitis.” The New England journal of medicine vol. 369,5 (2013): 417-27. doi:10.1056/NEJMoa1213277

Stone, John H et al. “Rituximab versus cyclophosphamide for ANCA-associated vasculitis.” The New England journal of medicine vol. 363,3 (2010): 221-32. doi:10.1056/NEJMoa0909905

Specks, Ulrich et al. “Efficacy of remission-induction regimens for ANCA-associated vasculitis.” The New England journal of medicine vol. 369,5 (2013): 417-27. doi:10.1056/NEJMoa1213277

Strand, Vibeke et al. “The impact of treatment with avacopan on health-related quality of life in antineutrophil cytoplasmic antibody-associated vasculitis: a post-hoc analysis of data from the ADVOCATE trial.” The Lancet. Rheumatology vol. 5,8 (2023): e451-e460. doi:10.1016/S2665-9913(23)00092-9

Tampe, Désirée et al. “Different Patterns of Kidney Fibrosis Are Indicative of Injury to Distinct Renal Compartments.” Cells vol. 10,8 2014. 6 Aug. 2021, doi:10.3390/cells10082014

SPRINT Research Group et al. “A Randomized Trial of Intensive versus Standard Blood-Pressure Control.” The New England journal of medicine vol. 373,22 (2015): 2103-16. doi:10.1056/NEJMoa1511939

Trivioli, Giorgio et al. “Slowly progressive anti-neutrophil cytoplasmic antibody-associated renal vasculitis: clinico-pathological characterization and outcome.” Clinical kidney journal vol. 14,1 332-340. 6 Sep. 2020, doi:10.1093/ckj/sfaa139

Trivioli, Giorgio et al. “Genetics of ANCA-associated vasculitis: role in pathogenesis, classification and management.” Nature reviews. Rheumatology vol. 18,10 (2022): 559-574. doi:10.1038/s41584-022-00819-y

van Eeden, Charmaine et al. “Fatigue in ANCA-associated vasculitis (AAV) and systemic sclerosis (SSc): similarities with Myalgic encephalomyelitis/Chronic Fatigue Syndrome (ME/CFS). A critical review of the literature.” Expert review of clinical immunology vol. 18,10 (2022): 1049-1070. doi:10.1080/1744666X.2022.2116002

van Eeden, Charmaine et al. “Myalgic encephalomyelitis/chronic fatigue syndrome (ME/CFS) and fibromyalgia: PR3-versus MPO-ANCA-associated vasculitis, an exploratory cross-sectional study.” Lancet regional health. Americas vol. 20 100460. 27 Feb. 2023, doi:10.1016/j.lana.2023.100460

Visseren, Frank L J et al. “2021 ESC Guidelines on cardiovascular disease prevention in clinical practice.” European heart journal vol. 42,34 (2021): 3227-3337. doi:10.1093/eurheartj/ehab484

Liste der Referenzen

Wallace, Zachary S et al. “Disease Activity, Antineutrophil Cytoplasmic Antibody Type, and Lipid Levels in Antineutrophil Cytoplasmic Antibody-Associated Vasculitis.” Arthritis & rheumatology (Hoboken, N.J.) vol. 71,11 (2019): 1879-1887. doi:10.1002/art.41006

Walsh, C L et al. “Imaging intact human organs with local resolution of cellular structures using hierarchical phase-contrast tomography.” Nature methods vol. 18,12 (2021): 1532-1541. doi:10.1038/s41592-021-01317-x

Walsh, Michael et al. “Plasma Exchange and Glucocorticoids in Severe ANCA-Associated Vasculitis.” The New England journal of medicine vol. 382,7 (2020): 622-631. doi:10.1056/NEJMoa1803537

Walsh, Michael et al. “The effects of plasma exchange in patients with ANCA-associated vasculitis: an updated systematic review and meta-analysis.” BMJ (Clinical research ed.) vol. 376 e064604. 25 Feb. 2022, doi:10.1136/bmj-2021-064604

WALTON, E W. “Giant-cell granuloma of the respiratory tract (Wegener's granulomatosis).” British medical journal vol. 2,5091 (1958): 265-70. doi:10.1136/bmj.2.5091.265

Weiner, Maria et al. “Outcome and treatment of elderly patients with ANCA-associated vasculitis.” Clinical journal of the American Society of Nephrology : CJASN vol. 10,7 (2015): 1128-35. doi:10.2215/CJN.00480115

Weppner, Gesche et al. “In situ detection of PR3-ANCA+ B cells and alterations in the variable region of immunoglobulin genes support a role of inflamed tissue in the emergence of auto-reactivity in granulomatosis with polyangiitis.” Journal of autoimmunity vol. 93 (2018): 89-103. doi:10.1016/j.jaut.2018.07.004

Windpessl, Martin et al. “Preventing infections in immunocompromised patients with kidney diseases: vaccines and antimicrobial prophylaxis.” Nephrology, dialysis, transplantation : official publication of the European Dialysis and Transplant Association - European Renal Association vol. 38,Supplement_2 (2023): ii40-ii49. doi:10.1093/ndt/gfad080

Windpessl, Martin et al. “Preventing infections in immunocompromised patients with kidney diseases: vaccines and antimicrobial prophylaxis.” Nephrology, dialysis, transplantation : official publication of the European Dialysis and Transplant Association - European Renal Association vol. 38,Supplement_2 (2023): ii40-ii49. doi:10.1093/ndt/gfad080

Xiao, Hong et al. “C5a receptor (CD88) blockade protects against MPO-ANCA GN.” Journal of the American Society of Nephrology : JASN vol. 25,2 (2014): 225-31. doi:10.1681/ASN.2013020143

Zimmermann, Jonas et al. “Avacopan in Anti-Neutrophil Cytoplasmic Autoantibodies-Associated Vasculitis in a Real-World Setting.” Kidney international reports vol. 9,9 2803-2808. 6 Jul. 2024, doi:10.1016/j.ekir.2024.07.007

Zimmermann J, Sonnemann J, Jabs WJ, et al. Avacopan in Anti-Neutrophil Cytoplasmic Autoantibodies-Associated Vasculitis in a Real-World Setting. Kidney Int Rep. 2024;9(9):2803-2808. Published 2024 Jul 6. doi:10.1016/j.ekir.2024.07.007

Zonozi, Reza et al. “Real-World Experience With Avacopan in Antineutrophil Cytoplasmic Autoantibody-Associated Vasculitis.” Kidney international reports vol. 9,6 1783-1791. 26 Mar. 2024, doi:10.1016/j.ekir.2024.03.022

Gekürzte Verschreibungsinformationen

Schweiz:

Tavneos®. Z: Avacopan. **I:** Tavneos, als ergänzende Therapie zu einer immunsuppressiven Standardbehandlung auf Basis von Rituximab oder Cyclophosphamid mit Glukokortikoiden, ist für die Behandlung erwachsener Patienten mit schwerer aktiver ANCA Vaskulitis (GPA/MPA) indiziert. **D:** Orale Einnahme morgens und abends 2x täglich 30 mg (3 Kapseln zu je 10 mg) mit Nahrung. **KI:** Überempfindlichkeit gegen den Wirkstoff oder einen der Hilfsstoffe. **VM:** Hepatotoxizität; Angioödem; Überwachung des Blutbildes (weisse Blutkörperchen); Schwere Infektionen; Reaktivierung des Hepatitis-B-Virus; Herzbeschwerden; Bösartige Tumore; Macroglycerinhydroxystearat. **S/S:** Eine Anwendung während der Schwangerschaft und bei Frauen im gebärfähigen Alter, die keine Verhütungsmethode anwenden, ist nicht empfohlen. Es ist nicht bekannt, ob Avacopan in die Muttermilch ausgeschieden wird. Der Nutzen des Stillens für das Kind sollte gegen den Nutzen der Behandlung für die Patientin abgewogen werden. **UW:** Sehr häufig: Infektion der oberen Atemwege, Nasopharyngitis; Kopfschmerzen; Erbrechen, Durchfall, Übelkeit; erhöhter Lebertest; verminderte Anzahl weisser Blutkörperchen. Häufig: Lungenentzündung, Infektion der unteren Atemwege, Influenza, Bronchitis, Zellulitis, Infektion der Harnwege, Herpes zoster, Sinusitis, orale Candidose, Herpes im Mundbereich, Otitis media, Rhinitis, Gastroenteritis; Neutropenie; Oberbauchschmerzen; Anstieg der Kreatinphosphokinase im Blut. Gelegentlich: Angioödeme. **IA:** Avacopan ist ein Substrat von CYP3A4. Die gleichzeitige Verabreichung von Induktoren oder Inhibitoren dieses Enzyms kann die Pharmakokinetik von Avacopan beeinflussen. Siehe Fachinformation. **P:** Tavneos 10 mg: 30 und 180 Hartkapseln. **Liste B**. Detaillierte Informationen**:** www.swissmedicinfo.ch. Stand der Information**:** Januar 2024. **Zulassungsinhaberin:** Vifor Fresenius Medical Care Renal Pharma Ltd., St. Gallen. **Vertrieb:** Vifor Pharma Switzerland AG, CH-1752 Villars-sur-Glâne |

▼Dieses Arzneimittel unterliegt einer zusätzlichen Überwachung. Für weitere Informationen, siehe Fachinformation TAVNEOS® auf www.swissmedicinfo.ch.

Gekürzte Verschreibungsinformationen

Österreich:

Tavneos® Fachkurzinformation
Tavneos®10mg Hartkapsel

Zusammensetzung: Jede Hartkapsel enthält 10 mg Avacopan. Sonstige Bestandteile mit bekannter Wirkung:245 mg Macrogolglycerolhydroxystearat(Ph.Eur). **Anwendungsgebiete:** Tavneos® ist in Kombination mit einem Rituximab- oder Cyclophopamid-Dosierungsschema indiziert zur Behandlung erwachsener Patienten mit schwerer aktiver Granulomatose mit Polyangiitis (GPA) oder mikroskopischer Polyangiitis (MPA). **Gegenanzeigen:** Überempfindlichkeit gegen den Wirkstoff oder einen der sonstigen Bestandteile. **Pharmakotherapeutische Gruppe:** Komplement-Inhibitoren **ATC- Code:** L04AJ05 **Inhaber der Zulassung:** Vifor France, 100-101 Terrasse Boieldieu Tour Franklin La Defense 8 92042 Paris La Defense Cedex, Frankreich. Rezept- und apothekenpflichtig. Weitere Angaben zu Warnhinweisen und Vorsichtsmaßnahmen für die Anwendung, Wechselwirkungen mit anderen Arzneimitteln oder sonstigen Wechselwirkungen, Schwangerschaft und Stillzeit und Nebenwirkungen sowie Gewöhnungseffekten sind der veröffentlichten Fachinformation zu entnehmen. Stand der Information: Mai 2023

▼Dieses Arzneimittel unterliegt einer zusätzlichen Überwachung. Dies ermöglicht eine schnelle Identifizierung neuer Sicherheitsdaten. Angehörige der Gesundheitsberufe werden gebeten, alle Verdachtsfälle von unerwünschten Wirkungen zu melden.