PÉCSI TUDOMÁNYEGYETEM
ÁLTALÁNOS ORVOSTUDOMÁNYI KAR

Kisér vasculitisek

Kumánovics Gábor
PTE, KK, Reumatológiai és Immunológiai Klinika

Klinikai immunológiai és allergológia tanfolvam
2025. október
SERIK, Lukács Épület, Közösségi Központ

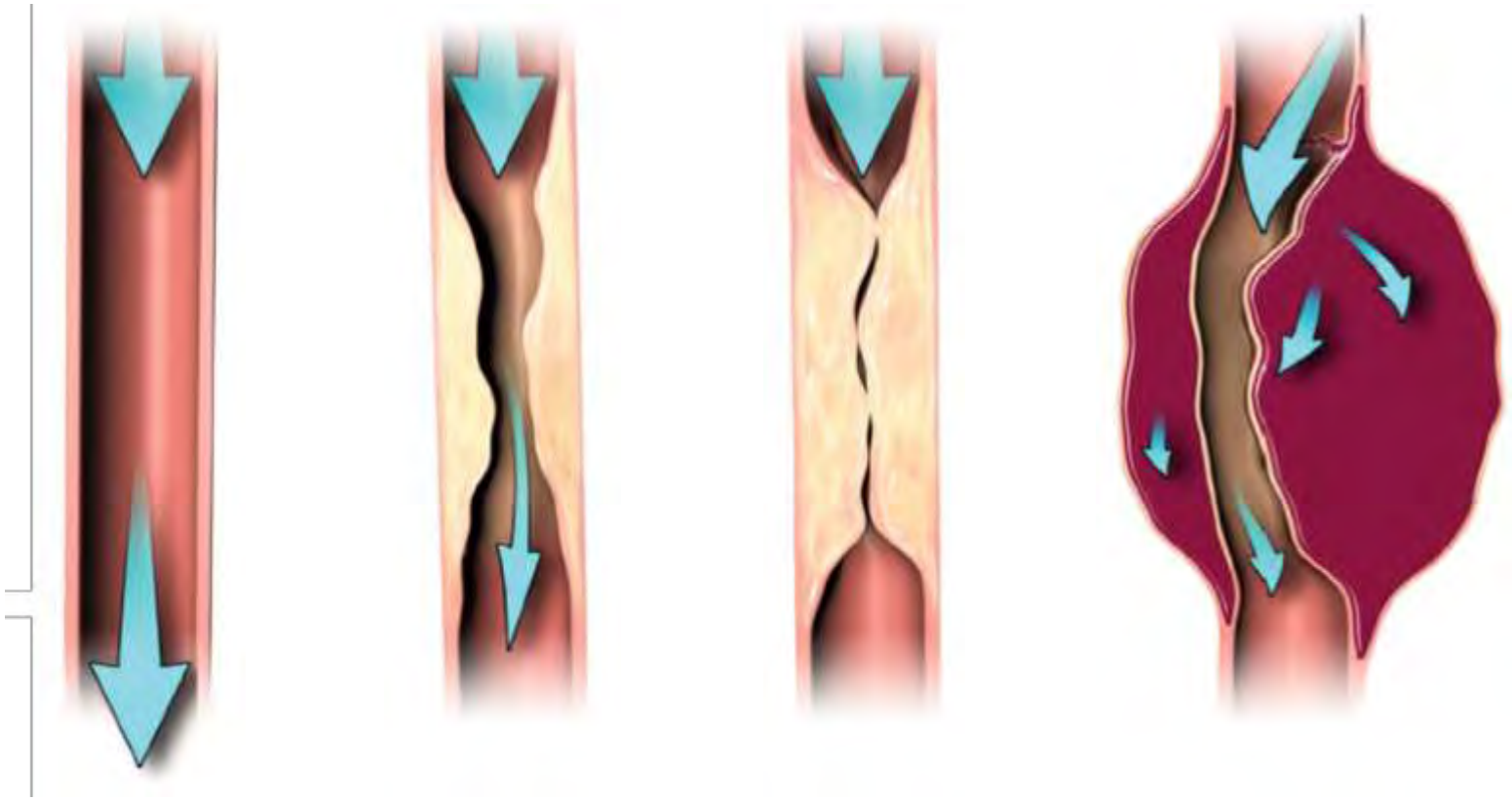
Vasculitis:
az erek falának gyulladásos infiltrációja

Normal artéria
Normál véráramlás

Gyulladt artéria
Csökkent véráramlás

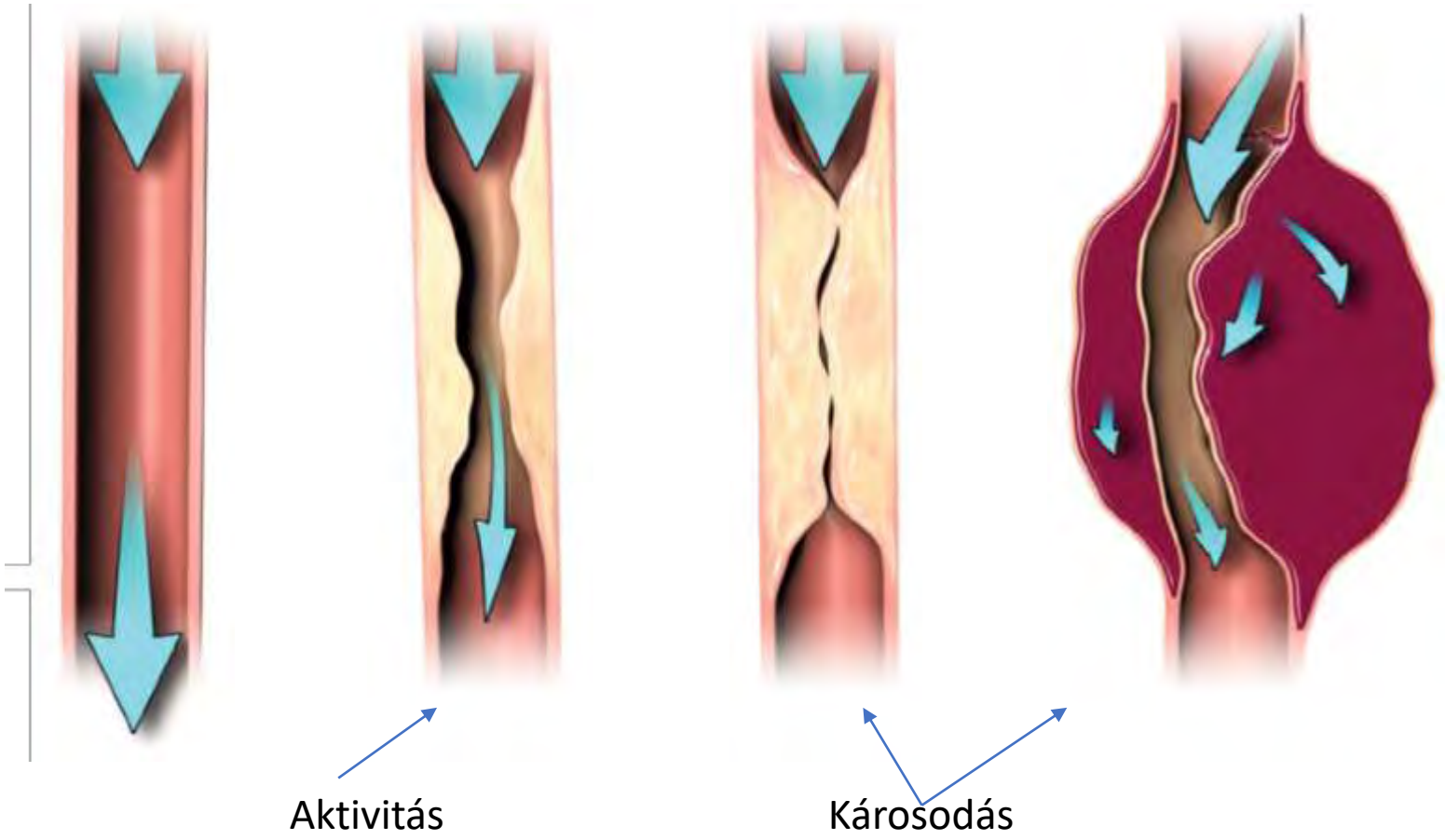
Elzáródott artéria
Nincs véráramlás

Aneurysma



Langford 2010;. Gómez-Puerta & Bosch 2009;. Jennette et al. 1994

Vasculitis:
az erek falának gyulladásos infiltrációja



Langford 2010;. Gómez-Puerta & Bosch 2009;. Jennette et al. 1994

Vasculitis alakulhat ki

- Malignus tumorokban
- Infectiókban
- Autoimmun kórképekben
- Gyógyszer-vegyszer hatás következtében

Large vessel vasculitis:

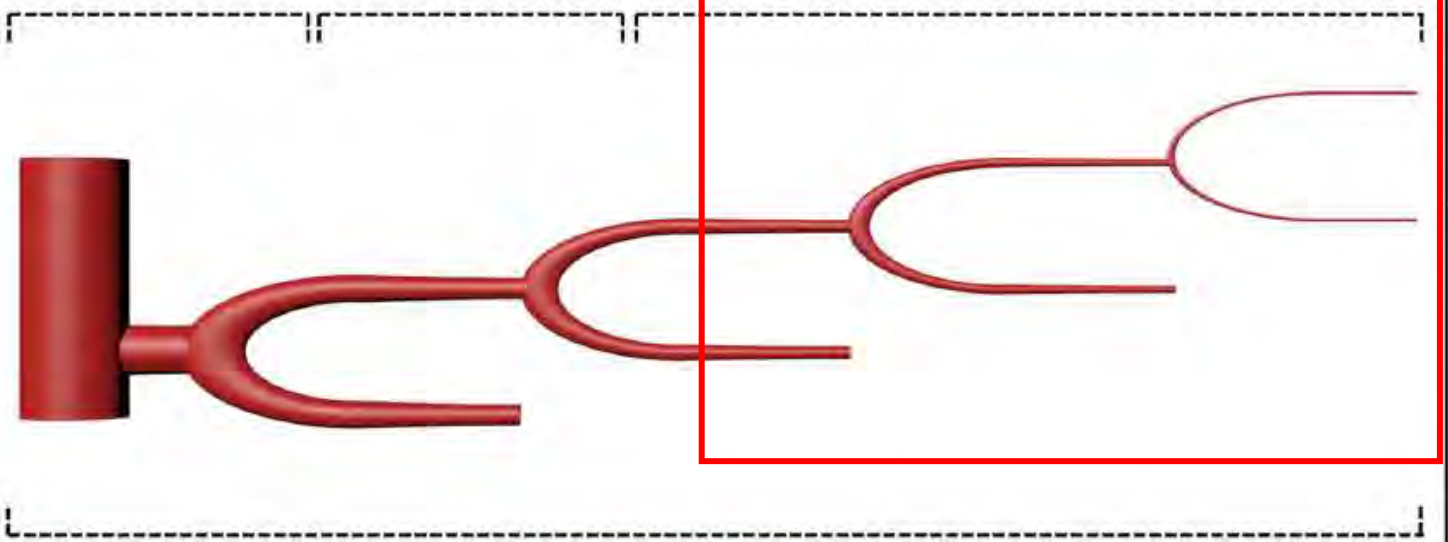
- **Takayasu arteritis**
- **Giant cell arteritis**

Medium vessel vasculitis:

- **Polyarthritis nodosa**
- **Kawasaki disease**

Small vessel vasculitis:

- **ANCA-associated vasculitis:**
  - Microscopic polyangiitis
  - Granulomatosis with polyangiitis
  - Eosinophilic granulomatosis with polyangiitis
- **Immune complex vasculitis**
  - Anti-glomerular basement membrane
  - Cryoglobulinemic vasculitis
  - IgA vasculitis
  - Hypocomplementemia urticarial vasculitis



Variable vessel vasculitis:

- **Behçet disease**
- **Cogan's syndrome**

Other types of vasculitis:

- **Single-organ vasculitis**
  - Cutaneous leukocytoclastic angiitis
  - Cutaneous arteritis
  - Primary central nervous system vasculitis
  - Isolated aortitis
  - Others
- **Vasculitis associated with systemic disease**
  - Lupus vasculitis
  - Rheumatoid vasculitis
  - Sarcoid vasculitis
  - Others
- **Vasculitis associated with probable etiology**
  - Hepatitis C virus–associated cryoglobulinemic vasculitis
  - Hepatitis B virus–associated vasculitis
  - Syphilis-associated aortitis
  - Drug-associated immune complex vasculitis
  - Drug-associated ANCA-associated vasculitis
  - Cancer-associated vasculitis
  - Others

Fig. 1 Diagram of various types of vasculitis based on the size of the affected vessel and etiology

Amouei *et al. Insights into Imaging (2022) 13:143*

A vasculitis syndromák osztályozása

Domináns ér	**Primaer**	**Szekunder**
Nagy arteriák	Óriás sejtes arteritis Takayasu arteritis	RA-hoz, infekcióhoz társuló aortitis (syphilis, TBC)
Közép nagy arteriák	Klasszikus polyarteritis nodosa	Hepatitis B asszociált PAN
	Kawasaki betegség	
Kiserek és közép nagy arteriák	Polyangiitis granulomatosa (korábban: Wegener granulomatosis) * Churg-Strauss syndroma* Microscopicus polyangiitis*	Vasculitis secondary to rheumatoid arthritis, systemás lupus erythematosus, Sjögren syndroma, gyógyszerek, infectiók (pl. HIV)
Kiserek	Henoch-Schönlein purpura	Gyógyszerek
	Cyroglobulinaemia	Hepatitis C asszociált
	Cutan leucocytoclasticus angiitis	Infectio
*ANCA asszociált betegségek. PAN: polyarteritis nodosa; RA, rheumatoid arthritis.		

Vasculitis észlelése során megválaszolandó kérdések

- Mi a diagnózis?

- Melyik erek érintettek?
- A folyamat mely szervekben okoz működészavart?

- Aktív-e a folyamat?
  - Általános tünet: láz, hőemelkedés észlelhető-e?
  - Van-e gyulladásos laboratóriumi eltérés (gyorsult Westergreen, emelkedett CRP)?
  - A complement szintek csökkentek-e?

- Van-e olyan szervi manifesztáció, ami megfelelő kezelés nélkül súlyos tüneteket okozhat?
- A kórlefolyás során várható-e a olyan új szervi manifesztáció jelentkezése, ami rossz prognózist okoz?

- Van-e esetleg vasculitist kiváltani képes valamilyen szekunder betegség (infekció, neoplasma, gyógyszer-vegyszerhatás) a háttérben?

(Gyulladásos betegség) aktivitás ↔ (Irreverzibilis) károsodás

Az ANCA-asszociált vasculitisek új klasszifikációja és terápiás ajánlása

- Szamosi Szilvia, Végh Edit, Bodoki Levente, Horváth Ágnes, Pethő Zsófia, Tari Dóra, Szekanecz Zoltán, Szűcs Gabriella

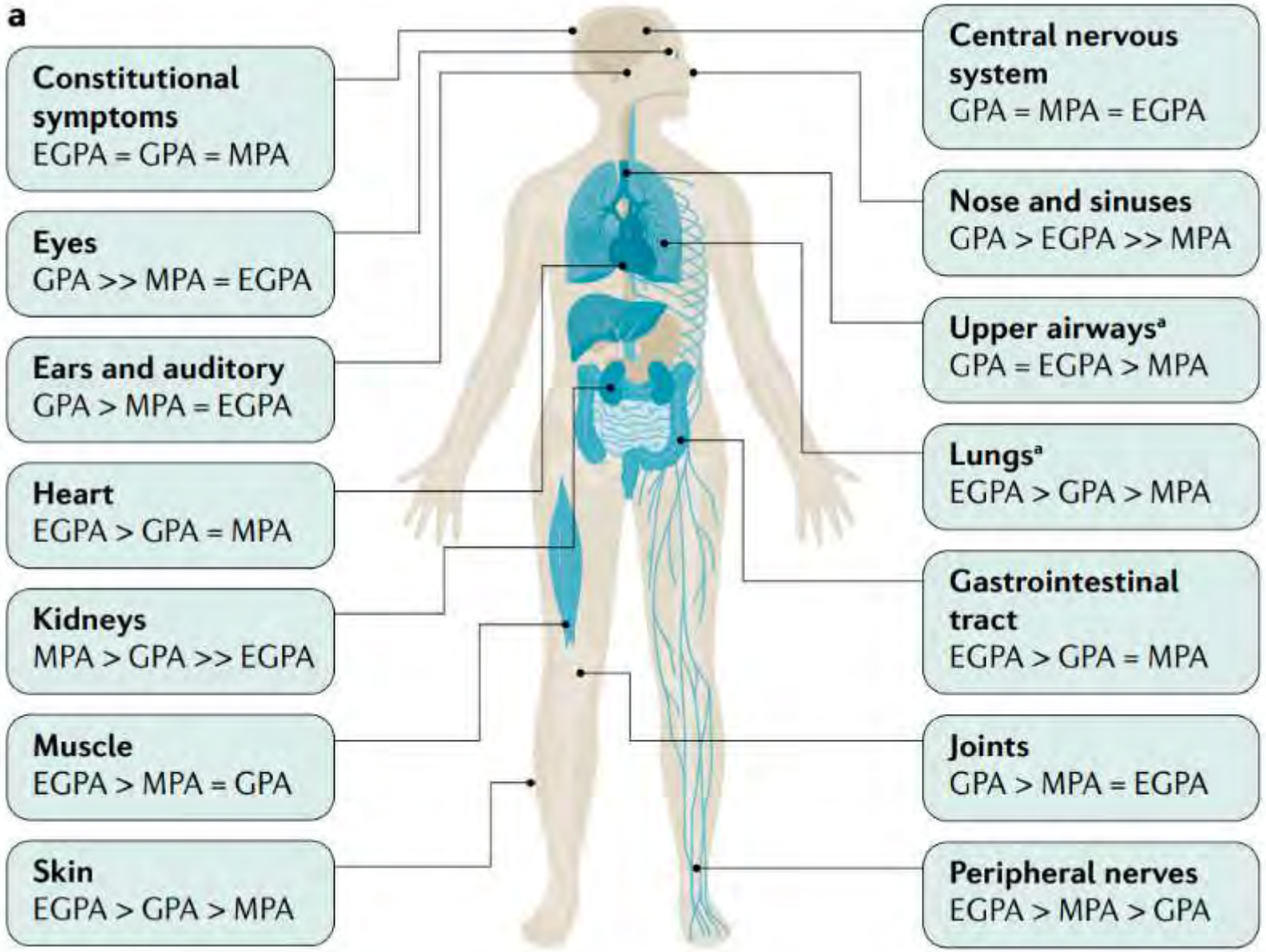
- Immunológiai szemle 2022.04.: 43-56.

Diagnózis:

Klinikai tünetek

- fáradékonyság, láz, fogyás, arthralgia, myalgia

- **Krónikus rhinitis, sinusitis, laryngitis**
- **Pulmonáris hemorrhagia – nehézlégzés, köhögés, haempotysis**
- **Granulomatosus orbitális/retroorbitális eltérések**
- **Purpurák, petechiák**
- **Rapid progresszív glomerulonephritis: proteinuria, haematuria, hypertenzió**
- **Mononeurotis multiplex**



Diagnózis:

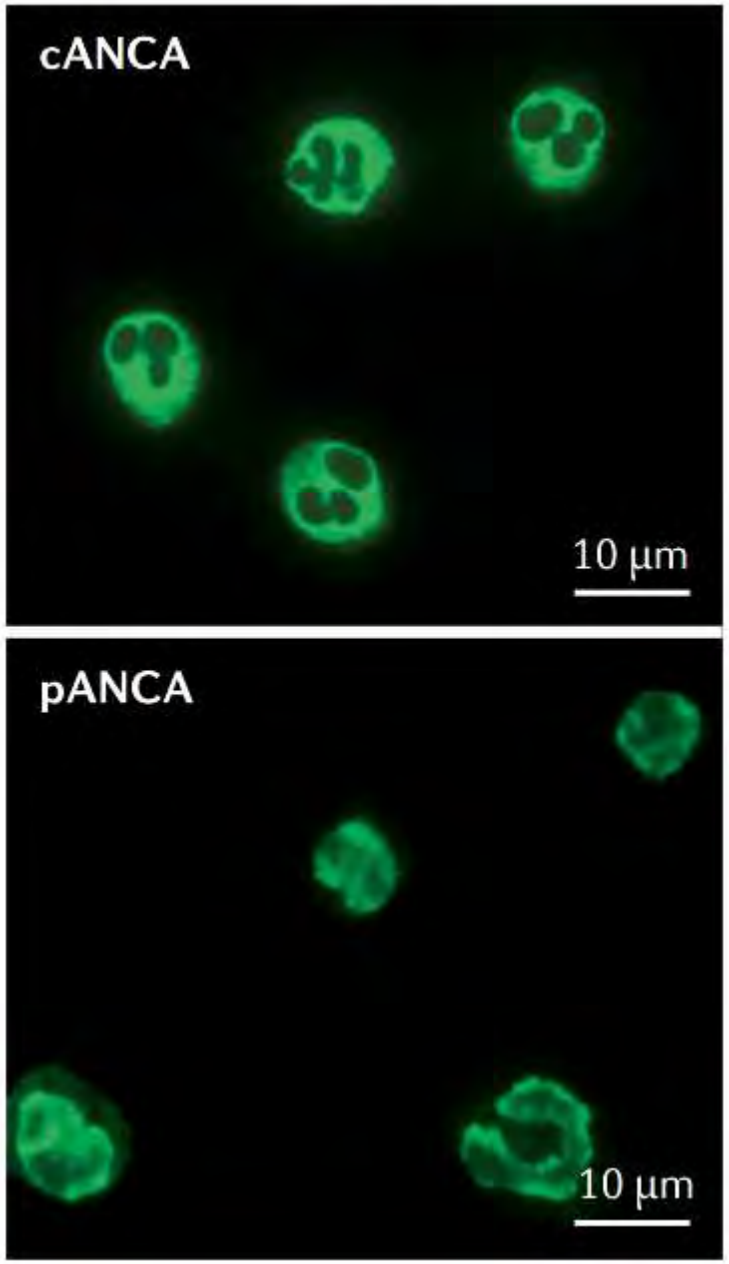
- Biomarkerek - Indirekt immunfluorescencia, ELISA
- Citoplasmatic pattern: c-ANCA (PR3 ANCA)
- Perincuclear pattern: p-ANCA (MPO ANCA)

  Pozitív lehet: infektív endocarditis, colitis ulcerosa, pseudomonas + cystás fibrosis

- Képalkotók: mellkas rtg, CT
- Biopszia: vese, tüdő, bőr, FOG eltérésből

Szövettan: akut - fibrinoid necrosis és gyulladás a kiserekben, olykor thrombosis kíséretében.

Krónikus: transmurális hegesedés, elasticus lamina interna elvesztésével.

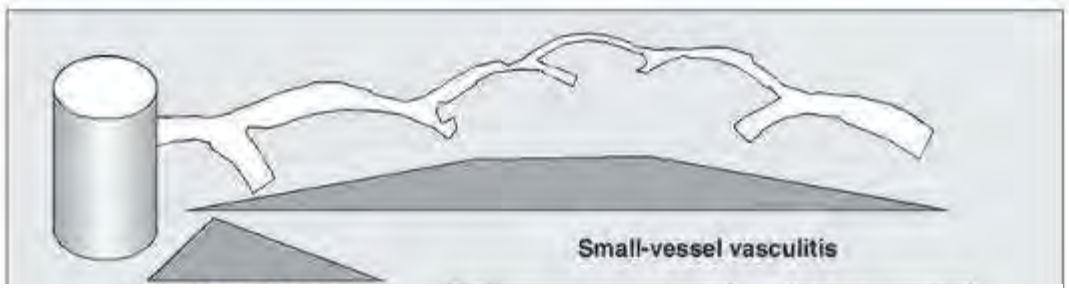


ANCA asszociált vasculitis prevalencia: 48-184 eset / millió

Feature	GPA	MPA	Eosinophilic GPA
Incidence	0.4–11.9 cases per 1 million person-years	0.5–24.0 cases per 1 million person-years	0.5–2.3 cases per 1 million person-years
Prevalence	2.3–146.0 cases per 1 million persons	9.0–94.0 cases per 1 million persons	2.0–22.3 cases per 1 million persons
Typical age of onset (years)	45–65	55–75	38–54
Male: female ratio	1:1	1:1	1:1
2012 revised CHCC definition[145]	Necrotizing granulomatous inflammation, usually involving the upper and lower respiratory tract; necrotizing vasculitis affecting predominantly small-to-medium vessels (such as capillaries, venules, arterioles, arteries and veins); necrotizing glomerulonephritis is common	Necrotizing vasculitis, with few or no immune deposits, predominantly affecting small vessels (such as capillaries, venules or arterioles); necrotizing arteritis involving small and medium arteries may be present; necrotizing glomerulonephritis is very common; pulmonary capillaritis often occurs; granulomatous inflammation is absent	Eosinophil-rich and necrotizing granulomatous inflammation, often involving the respiratory tract; necrotizing vasculitis predominantly affecting small-to-medium vessels; associated with asthma and eosinophilia; ANCA$^+$ is more frequent when glomerulonephritis is present
Frequency of ANCA	PR3-ANCA$^+$: 65–75% MPO-ANCA$^+$: 20–30% ANCA$^-$: 5%	PR3-ANCA$^+$: 20–30% MPO-ANCA$^+$: 55–65% ANCA$^-$: 5–10%	PR3-ANCA$^+$: <5% MPO-ANCA$^+$: 30–40% ANCA$^-$: 55–65%
Key innate immune cell	Neutrophil	Neutrophil	Eosinophil
Relapse rate	Higher than MPA (or MPO-AAV)	Lower than GPA (or PR3-AAV)	Relapse is frequent

AAV, ANCA-associated vasculitis; ANCA, anti-neutrophil cytoplasmic antibody; CHCC, Chapel Hill Consensus Conference; GPA, granulomatosis with polyangiitis; MPA, microscopic polyangiitis; MPO, myeloperoxidase; PR3, leukocyte proteinase 3.

Kisér vasculitisek



Lupushoz társuló vasculitis,
Henoch-Schönlein purpura,
Paraneoplasiás vasculitis
Goodpasture szindróma

Immun komplex depozitum

Nincs immun komplex depozitum

Vasculitis
Nincs asthma
Nincs granuloma

Vasculitis
Nincs asthma
Granuloma

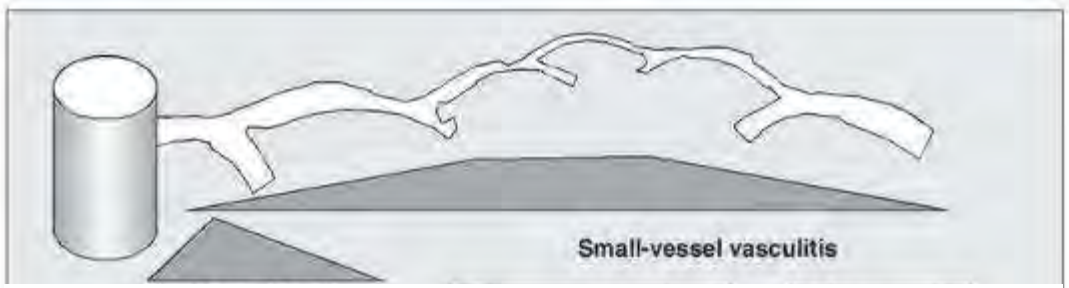
Eosinophilia
Asthma
Granuloma

Microscopicus polyangiitis (MPA)

Polyangiitis granulomatosa (GPA; Wegener)

Churg-Strauss szindróma (eGPA)

Kisér vasculitisek



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Granuloma

Eosinophilia
Asthma
Granuloma

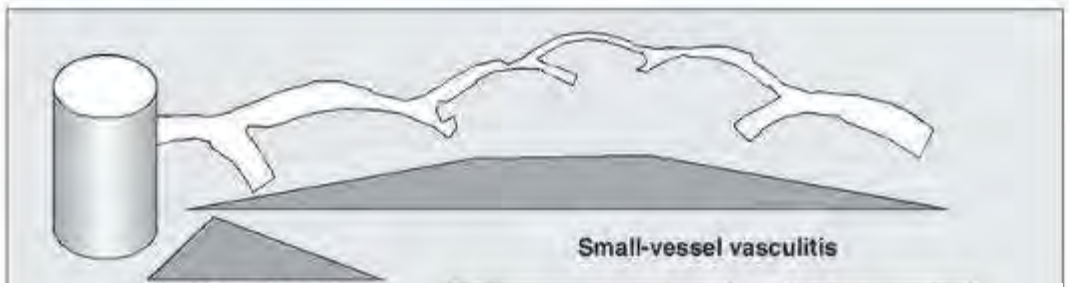
Microscopicus polyangiitis (MPA)

Polyangiitis granulomatosa (GPA; Wegener)

Churg-Strauss szindróma (eGPA)

Nekrotizáló granulomatozus gyulladás a a felső és alsó légutakban. Necrotizáló glomerulonephritis, pulmonaris haemorrhagia illetve ocularis vasculitis szintén gyakori

Kisér vasculitisek



Lupushoz társuló vasculitis,
Henoch-Schönlein purpura,
Paraneoplasiás vasculitis
Goodpasture szindróma

Immun komplex depozitum

Nincs immun komplex depozitum

Vasculitis
Nincs asthma
Nincs granuloma

Vasculitis
Nincs asthma
Granuloma

Eosinophilia
Asthma
Granuloma

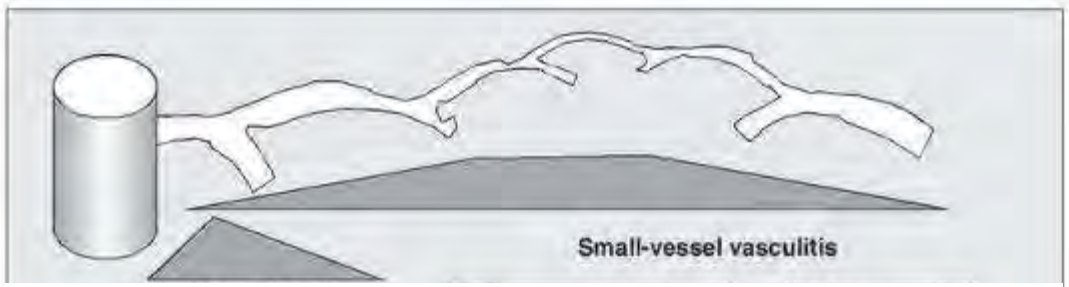
Microscopicus polyangiitis (MPA)

Polyangiitis granulomatosa (GPA; Wegener)

Churg-Strauss szindróma (eGPA)

Nekrotizáló arteritis, de nincs granulomatosus gyulladás. Glomerulonephritis és capillaritis: A tüdőben és egyéb szervekben is lehet.

Kisér vasculitisek



Lupushoz társuló vasculitis,
Henoch-Schönlein purpura,
Paraneoplasiás vasculitis
Goodpasture szindróma

Immun komplex depozitum

Nincs immun komplex depozitum

Vasculitis
Nincs asthma
Nincs granuloma

Vasculitis
Nincs asthma
Granuloma

Eosinophilia
Asthma
Granuloma

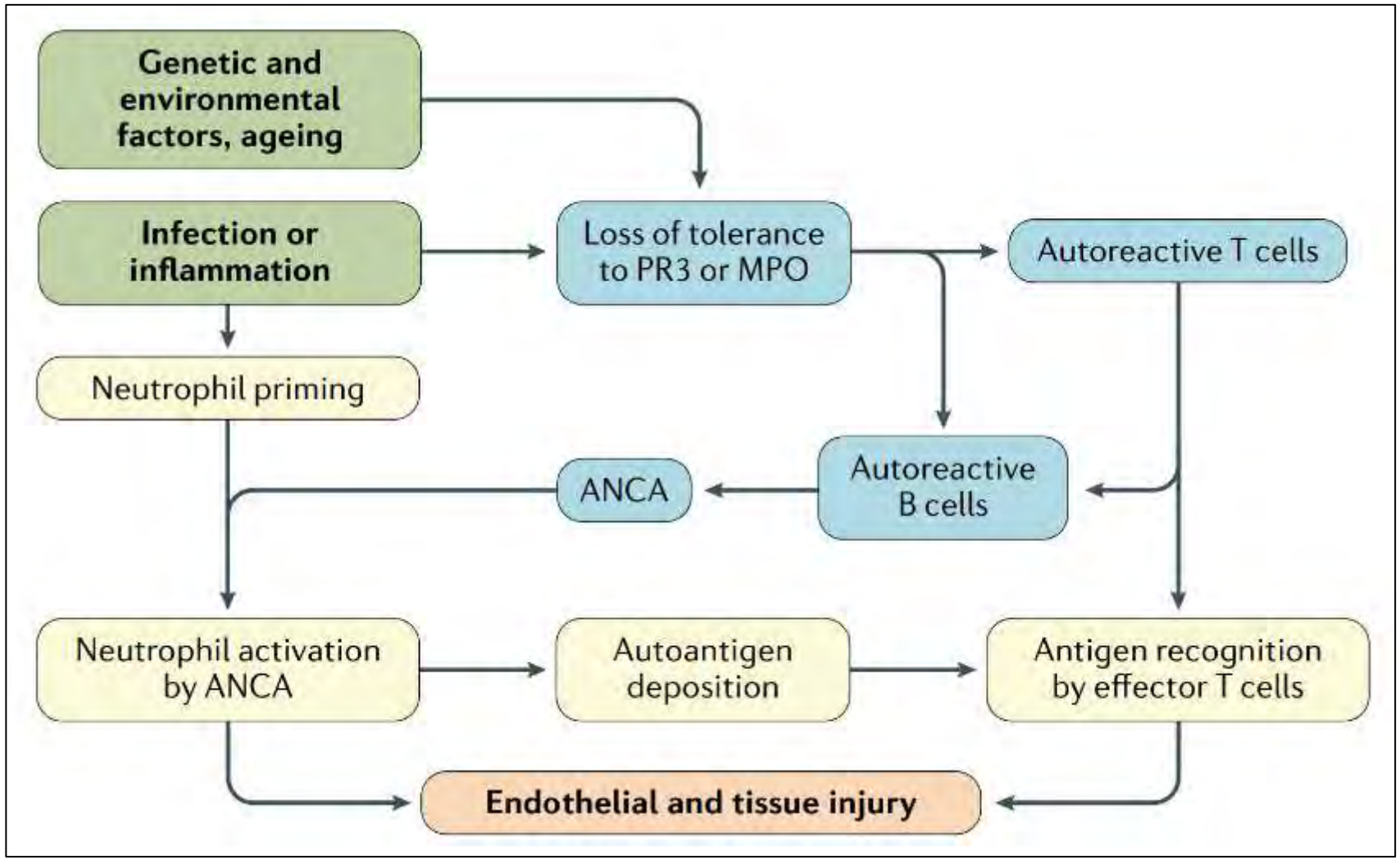
Microscopicus polyangiitis (MPA)

Polyangiitis granulomatosa (GPA; Wegener)

Churg-Strauss szindróma (eGPA)

Nekrotizáló granulomatózus eosinophil sejtekben gazdag gyulladás. A légutak gyakran érintettek beleértve a nasalis polypusokat is. Eosinpophilia.

Pathomechanizmus



Mikrovasculáris endothelialis gyulladás

↓

Extravasculáris gyulladás

↓

Szöveti destrukció

↓

Fibrosis

↓

Funkció kiesés

Rizikófaktorok

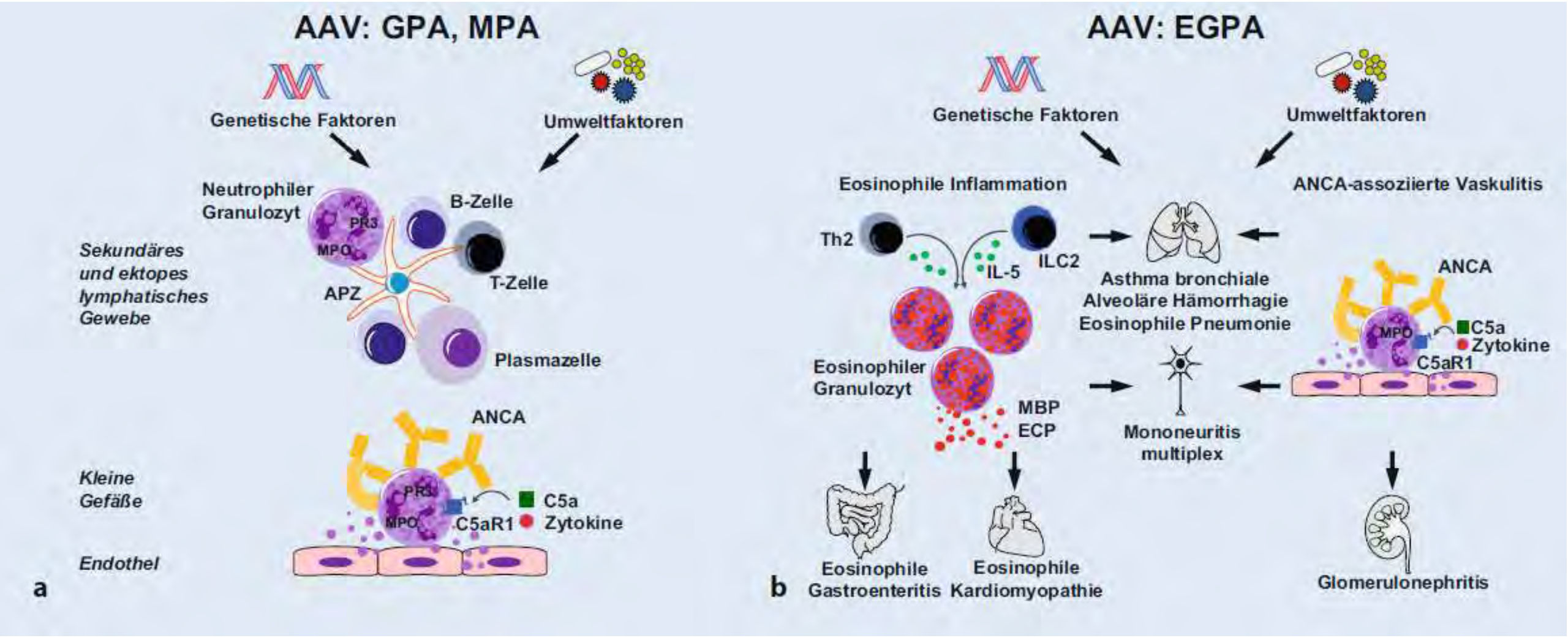
Környezeti

- S. aureus orrüregi hordozása – magasabb GPA relapszus ráta
- Szilícium-dioxid expozícó (pl. cement) – MPO-AAV
- Peszticidek
- UV sugárzás
- Dohányzás
- Kokain

Genetikai

SERPINA1 (alfa1-tripszin)/ PRTN3 (PR3) – mutációjuk magasabb PR3 szinthez vezet, mely PR3 tolerancia elvesztéséhez vezethet

Pathogenesis



Z Rheumatol 2022 81:270–279

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.

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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 $\geq 1 \times 10^9$/liter	-4

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

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CLASSIFICATION CRITERIA FOR **EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS**

CONSIDERATIONS WHEN APPLYING THESE CRITERIA

- **These classification criteria should be applied to classify a patient as having eosinophilic 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

Obstructive airway disease	+3
Nasal polyps	+3
Mononeuritis multiplex	+1

LABORATORY AND BIOPSY CRITERIA

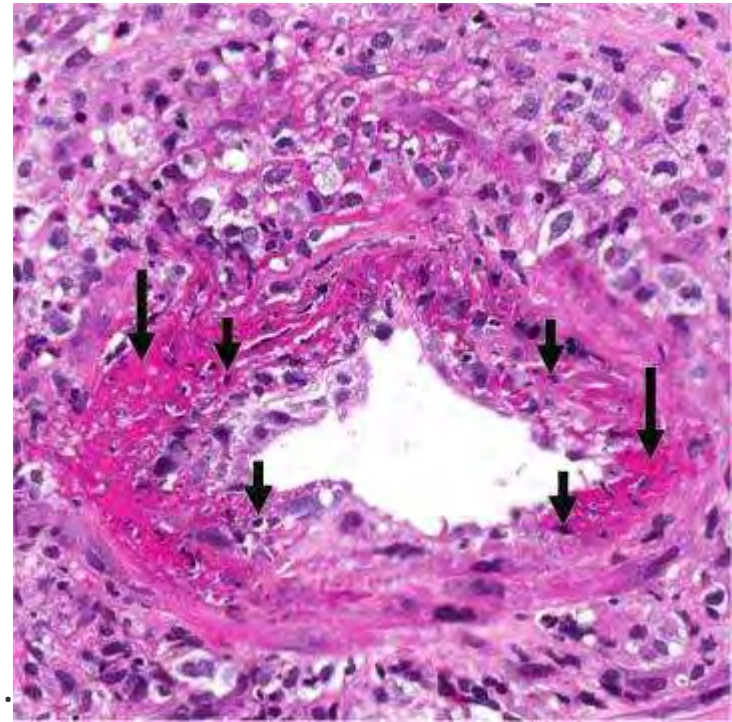
Blood eosinophil count ≥ 1 x10^9/liter	+5
Extravascular eosinophilic-predominant inflammation on biopsy	+2
Positive test for cytoplasmic antineutrophil cytoplasmic antibodies (cANCA) or antiproteinase 3 (anti-PR3) antibodies	-3
Hematuria	-1

Sum the scores for 7 items, if present. A score of ≥ 6 is needed for classification of EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS.

Klasszifikáció

- Kisérvasculitis diagnózisa már megtörtént
- Alernatív diagnózisok kizárásra kerültek

- Klinikai + képalkotó/laboratóriumi/hisztológiai kritériumok
  - Súlyozás jelentőség szerint
    - Pozitív (támogató) krétriumok
    - Negatív (ellene szóló) kritériumok



Arthritis & Rheumatology
Vol. 74, No. 3, March 2022, pp 386-406

Semin Immunopathol **44,** 325–345 (2022).

Klasszifikáció

- GPA
  - Pozitív pANCA vagy anti-MPO antitest -1
  - Vérkép eosinophilszám ≥ $1x10^9/l$ -4
- MPA
  - Orr érintettsége -3
  - Pozitív cANCA vagy anti-PR3 antitest -1
  - Vérkép eosinophilszám ≥ $1x10^9/l$ -4
- EGPA
  - Pozitív cANCA vagy anti-PR3 antitest -3
  - Haematuria -1

Patogenezis

Table 1 Clinical Phenotypes, Genetic Polymorphisms and Biomarkers Associated with PR3 and MPO ANCA Subtypes

	PR3	**MPO**
Proportion of patients with each clinical phenotype thought to display ANCA subtype	GPA 75% MPA 30% EGPA 5%	GPA 20% MPA 60% EGPA 45%
Genetic associations	Polymorphisms in HLA-DP, *PRTN3* and *SERPINA1* genes	Polymorphisms in HLA-DQ
Associated serum biomarkers	IL-6, GM-CSF, IL-15, IL-18, CXCL8/IL-8, CCL17/TARC, IL-18BP, sIL-2Rα, NGFβ	sIL6R, sTNFRII, NGAL, sICAM-1

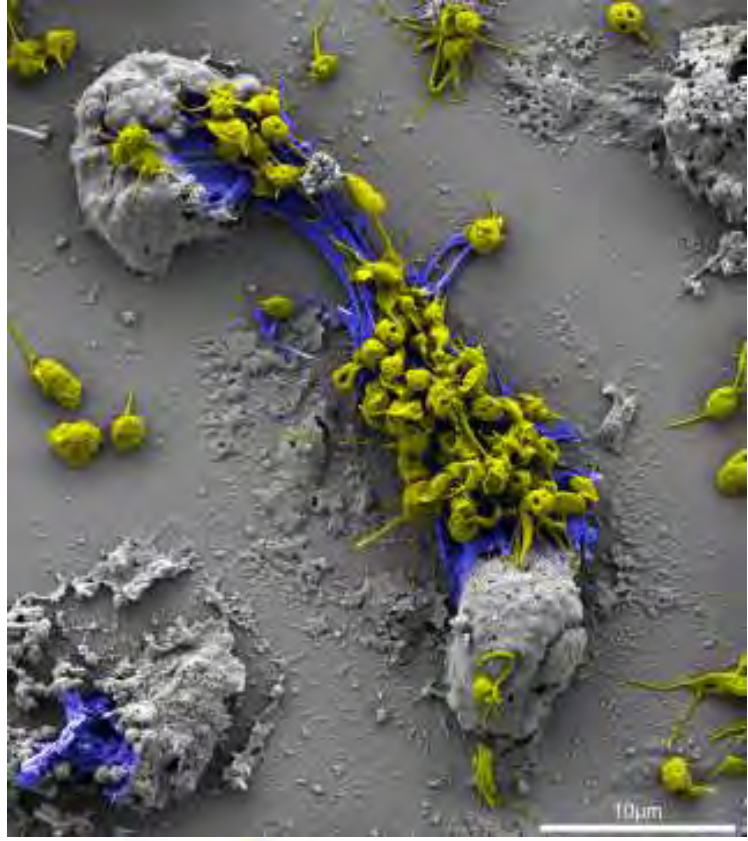
Abbreviations: PR3, anti-proteinase 3 antibody; MPO, anti-myeloperoxidase antibody; GPA, granulomatosis with polyangiitis; MPA, microscopic polyangiitis; EGPA, eosinophilic granulomatosis with polyangiitis; HLA, human leukocyte antigen.

Semin Immunopathol **44,** 325–345 (2022).

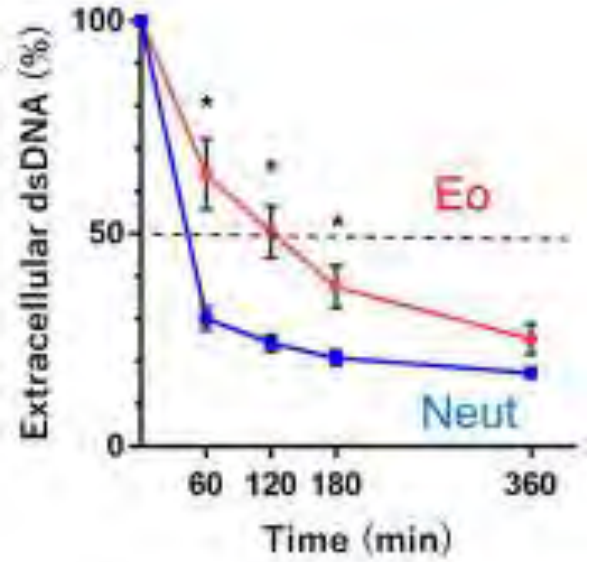
Genome-wide association study

	% of patients with feature		
Clinical feature	MPO+ AAV (non EGPA)	MPO+ EGPA	ANCA– EGPA
Glomerulonephritis	85	29*	9
Neuropathy	20	79*	57
Asthma	n.d.	100	100
Eosinophilia	4.5	100	100
Pulmonary hemorrhage	17	4	4
Ear nose or throat	32	81	88
Pulmonary infiltrates	20	45	61*
Cardiac involvement	3	15	30*
Rituximab response	98	80	38

Nat Commun. 2019; 10: 5120.



Gal-10 - BVAS



Front Immunol. 2021; 12: 801897.

ILD – ANCA asszociált vasculitisekben

- **granulomatosis with polyangiitis (GPA) - 20 - 30%**
  - Cavitated and noncavitated nodules 40-89%
  - Consolidations 30%
  - Ground glass opacities 25-50%
  - Diffuse alveolar haemorrhage 5-10%
- **microscopic polyangiitis (MPA) - 40 - 70%**
  - Diffuse alveolar haemorrhage 10-55%
  - Uni- und bilateral pulmonary infiltrates 26-92%
  - Lung fibrosis / interstitial abnormalities 32-90%
- **eosinophilic granulomatosis with polyangiitis (EGPA) - 30 - 40%**
  - Asthma 95%, kontrollált EGPA mellett is aktív 45%
  - Eosinophilic pneumonia 38-75%
  - Consolidations/Nodules 11-89%
  - Ground glass opacities 39-99%
  - diffuse alveolar haemorrhage 3-8%
- **ILD:**
  - **MPO-ANCA: 70%**
  - **Pr3-ANCA: 30%**
- **Mortalitás ILD-vel: 2-4x**

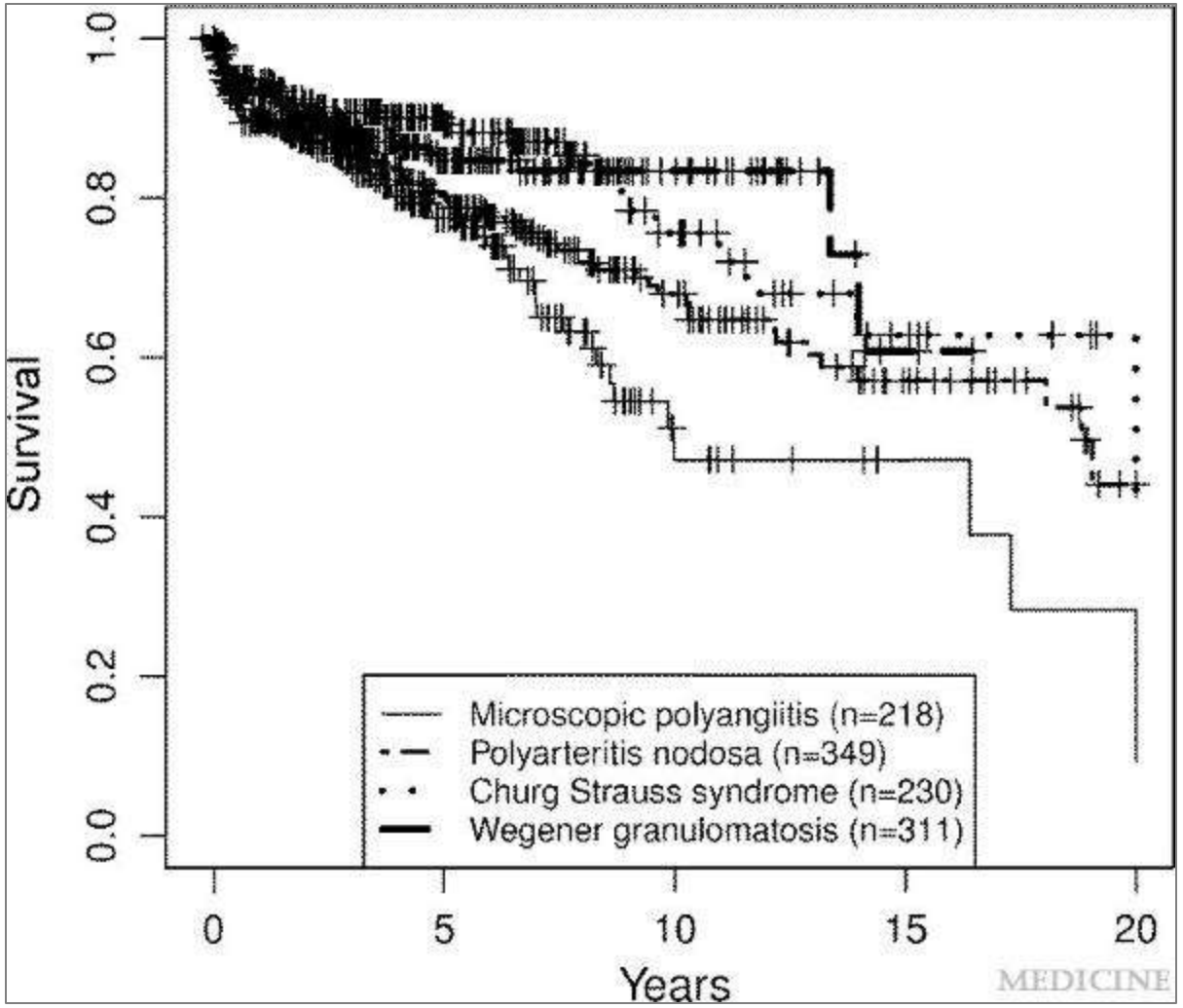
Sebastiani M et al, Clinical and Experimental Rheumatology 2020, 221-231.

ILD – ANCA asszociált vasculitisekben

- **Usual interstitial pneumonia (UIP) - 45.8%,**
- **probable UIP - 4.2%**
- **indeterminate UIP - 8.3%**
- **alternate diagnosis - 41.7%**
  - **chronic hypersensitivity pneumonitis-like pattern - 25%**
  - **nonspecific interstitial pneumonia-like pattern - 12%**
  - **cryptogenic organizing pneumonia-like pattern - 4%**

Kwon M et al, Journal of Clinical Rheumatology: 2021 - 324-330.

Túlélés



Medicine90(1):19-27, January 2011.

Rosszabb prognózis:

-idősebb kor

-ILD

-VTE

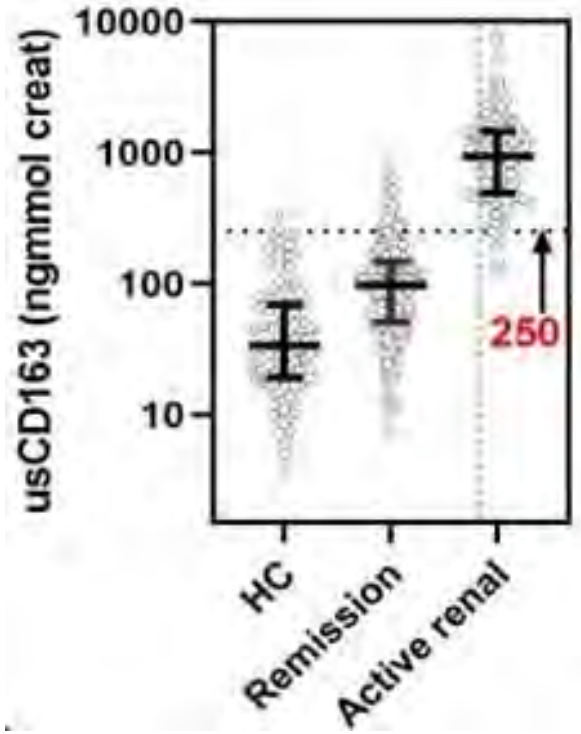
-arteritis a vesebiopsziában

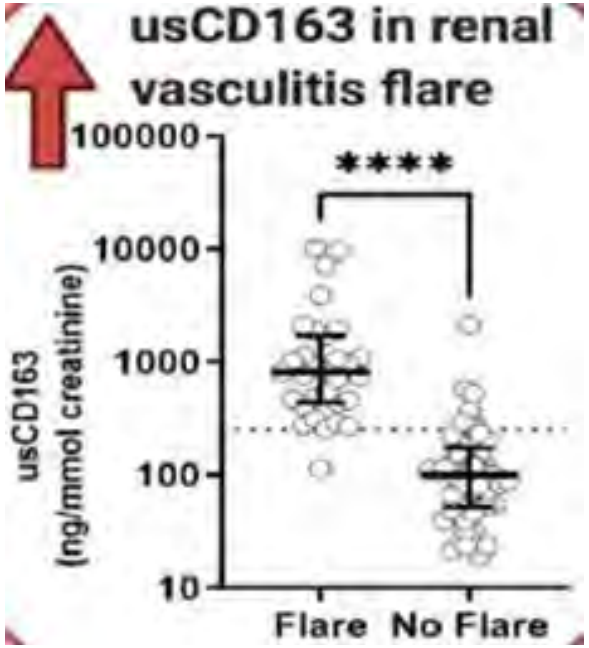
J Am Soc Nephrol . 2021 Sep;32(9):2362-2374.
Rheumatology (Oxford) 2021; 60: 4654-61.

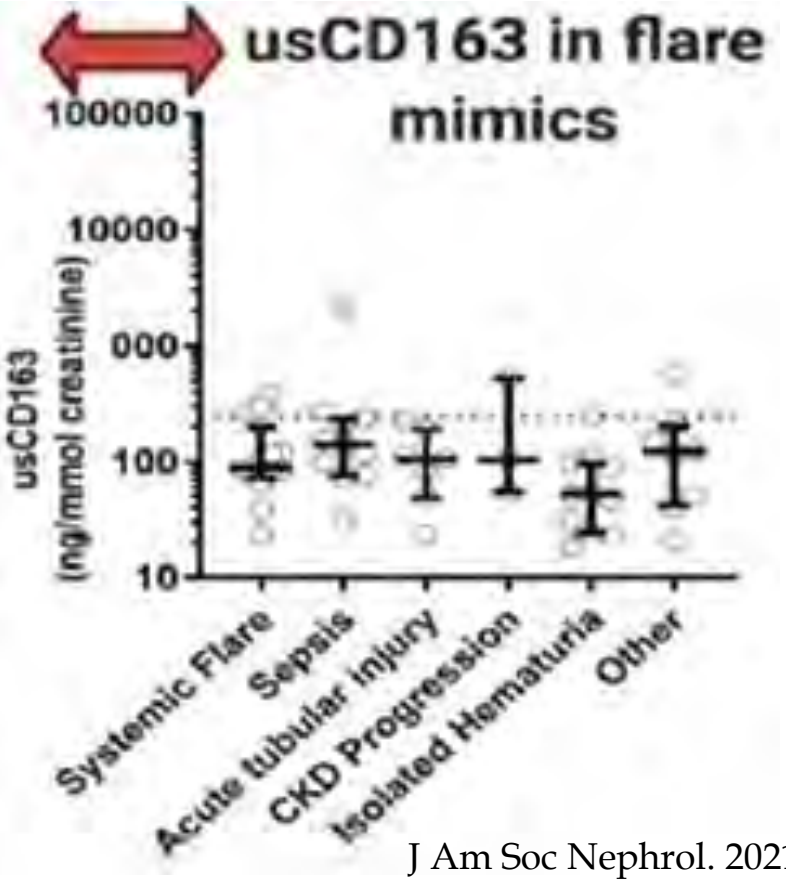
Biomarkerek – aktív nephritis AAV-ben

CD163

scavanger receptor –
monocita/makrofág specifikus
korrelál makrofág aktivációval



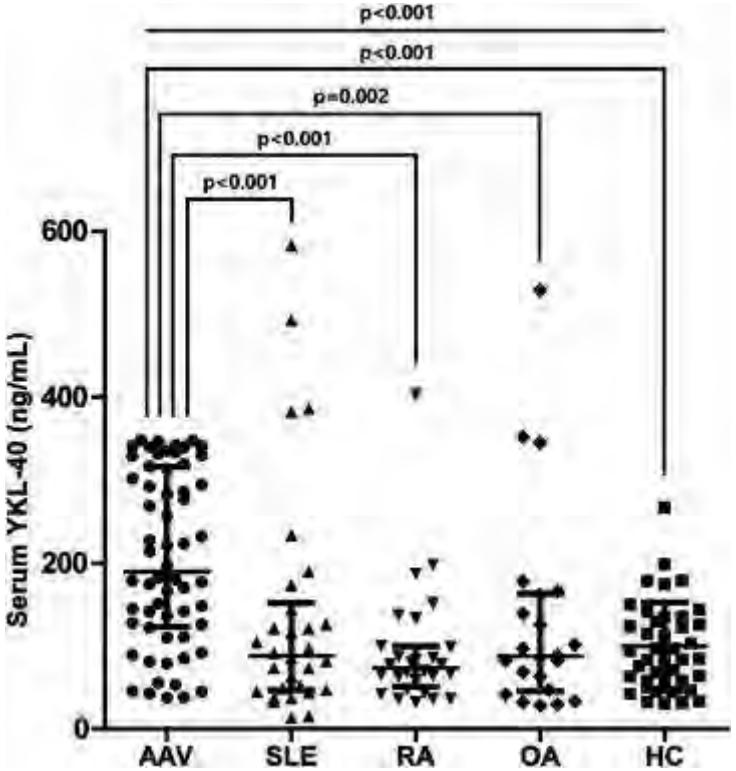


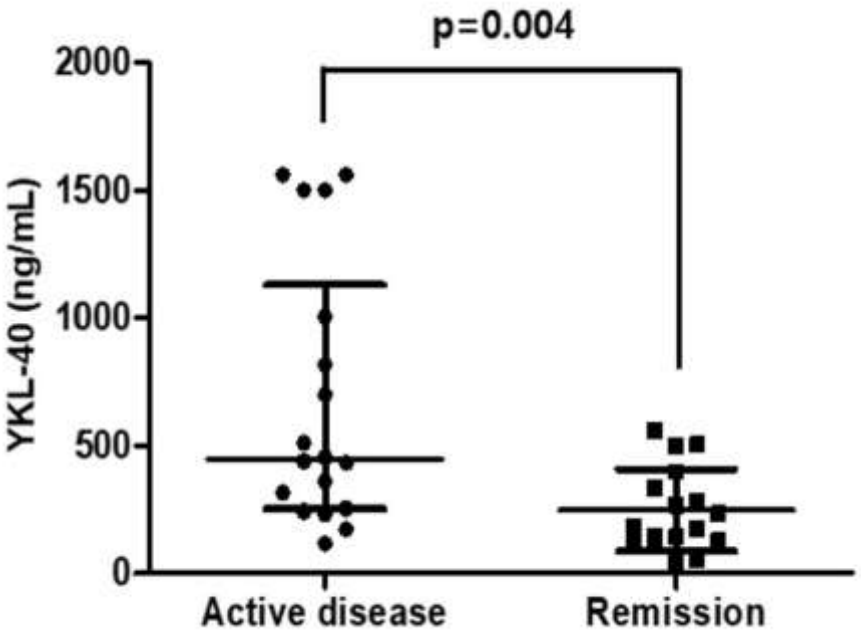


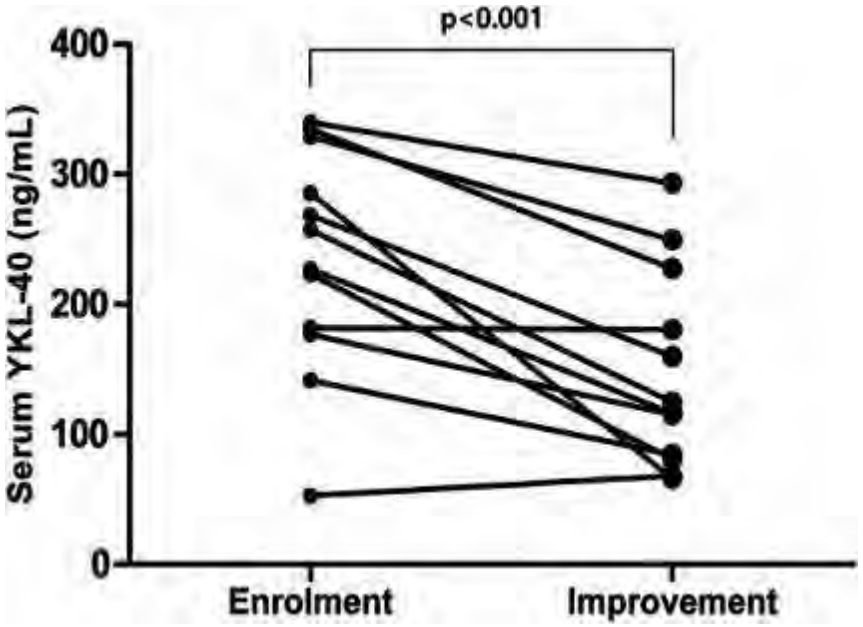
J Am Soc Nephrol. 2021 Nov;32(11):2920-2932.

Biomarkerek

- YKL-40
  - Anti-apoptotikus folyamatok
  - Astrociták migrációs faktora
  - Makrofágok, chondrociták, synoviális sejtek, neutriofilek

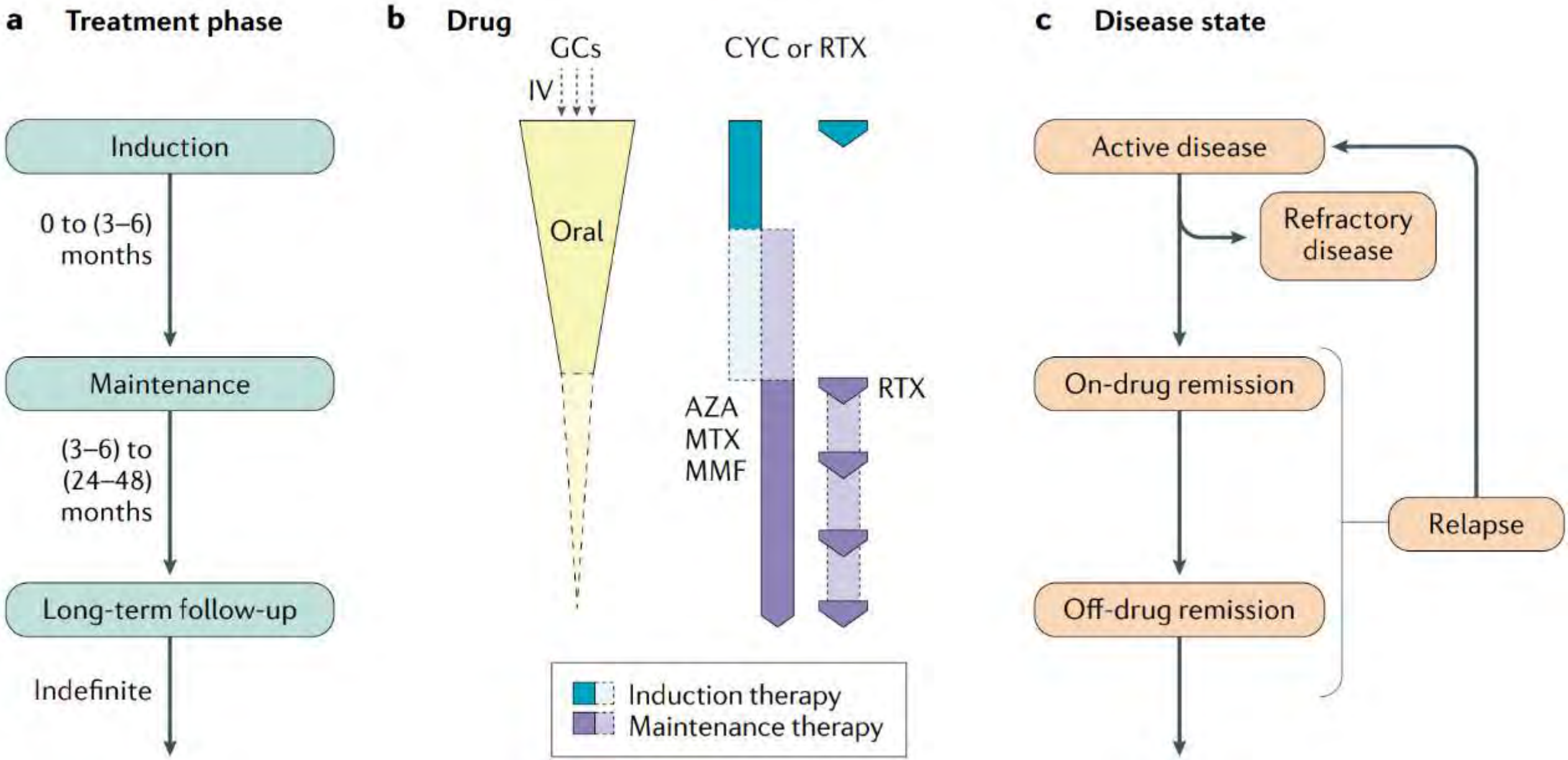




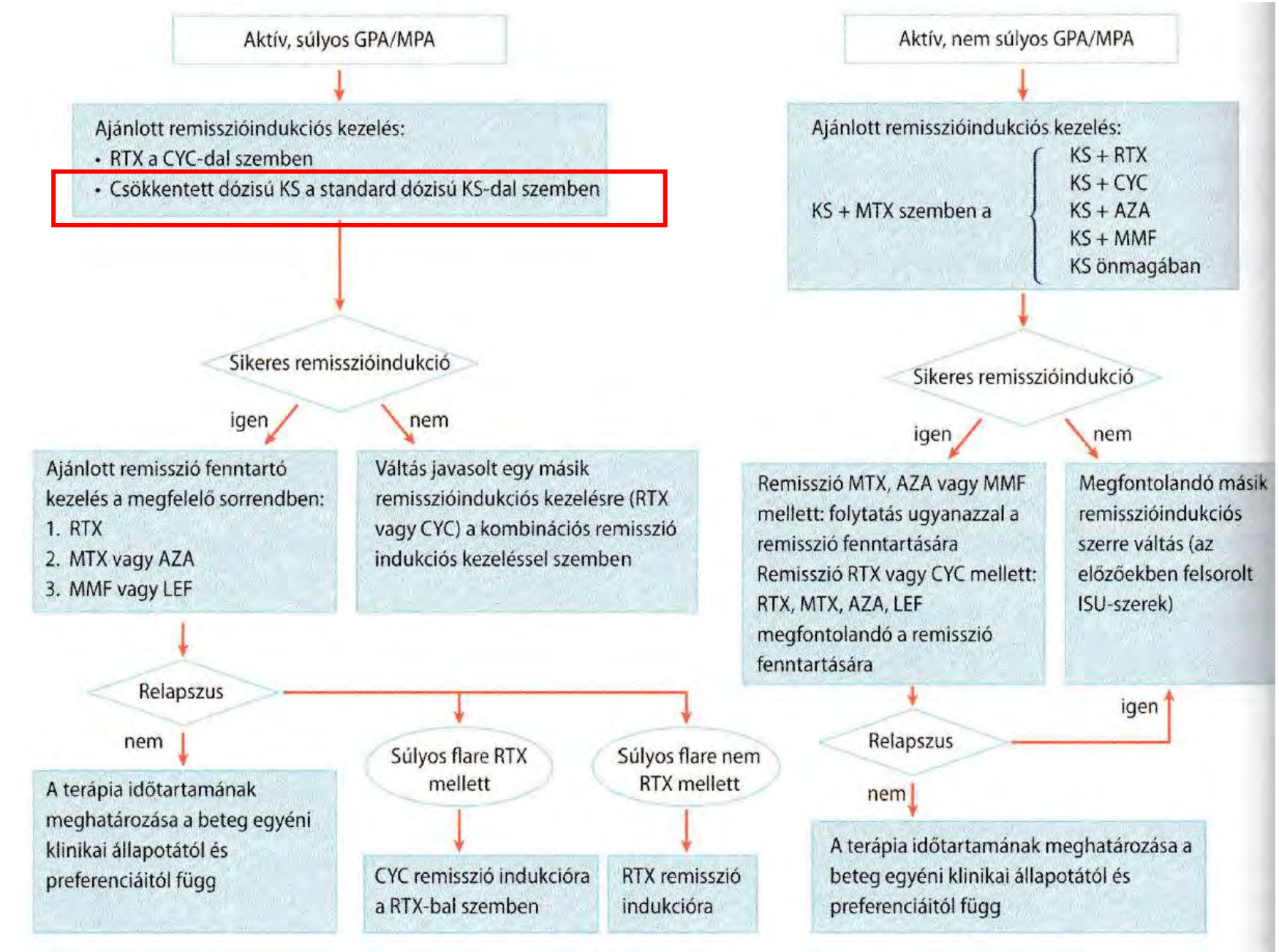


Arthritis Res Ther. 2021; 23: 77.

Terápia

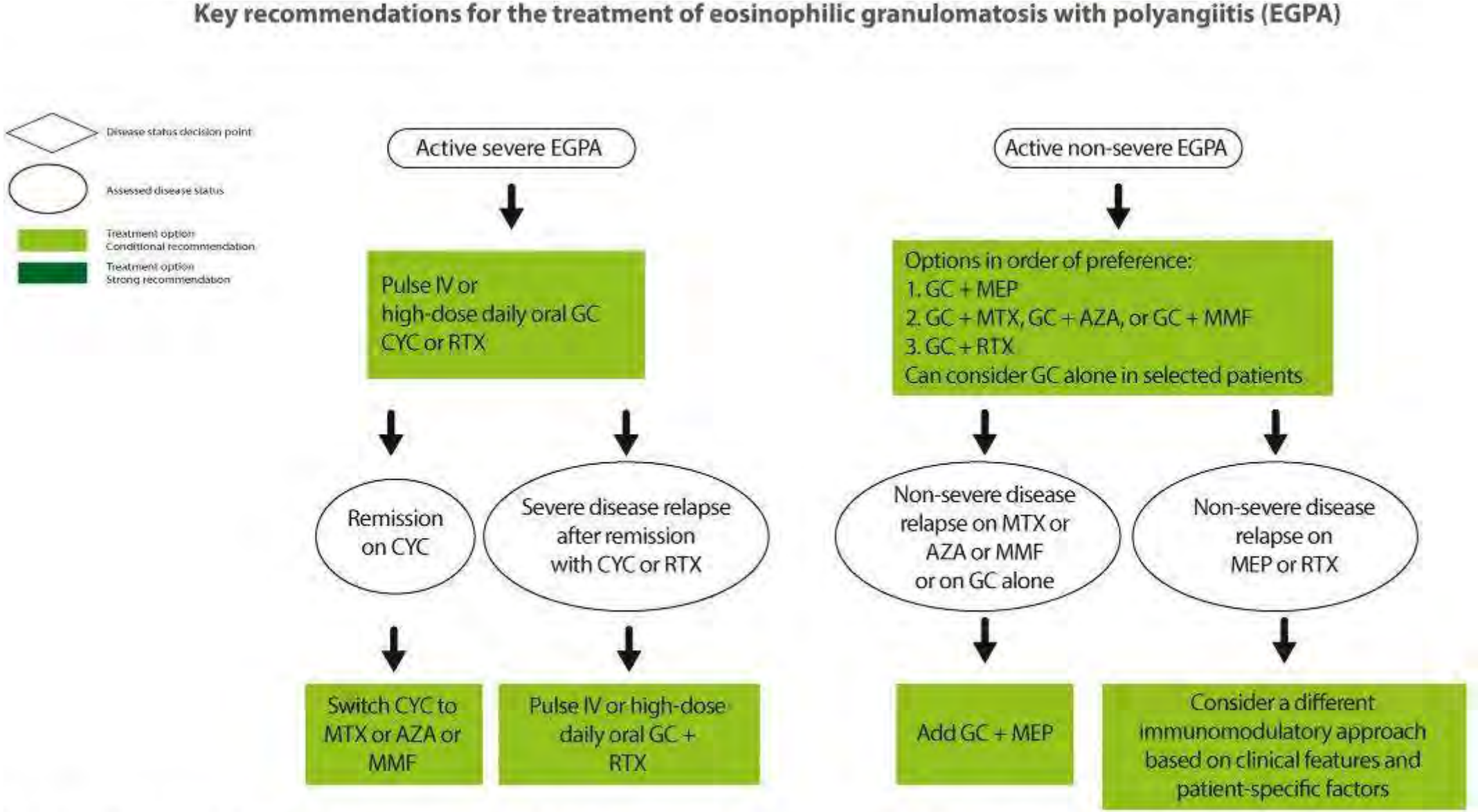

a Treatment phase
Induction
0 to (3–6) months
Maintenance
(3–6) to (24–48) months
Long-term follow-up
Indefinite
b Drug
GCs
IV
Oral
CYC or RTX
AZA
MTX
MMF
RTX
Induction therapy
Maintenance therapy
c Disease state
Active disease
Refractory disease
On-drug remission
Relapse
Off-drug remission

Kezelés



Immunológiai szemle 2022.04.: 43-56.

Kezelés - EGPA



AZA = azathioprine, CYC = cyclophosphamide, GC = glucocorticoids, IV = intravenous, MEP = mepolizumab, MMF = mycophenolate mofetil, MTX = methotrexate, RTX = rituximab

Arthritis Care & ResearchVol. 73, No. 8, August 2021, pp 1088–1105

Kezelés - fogalmak

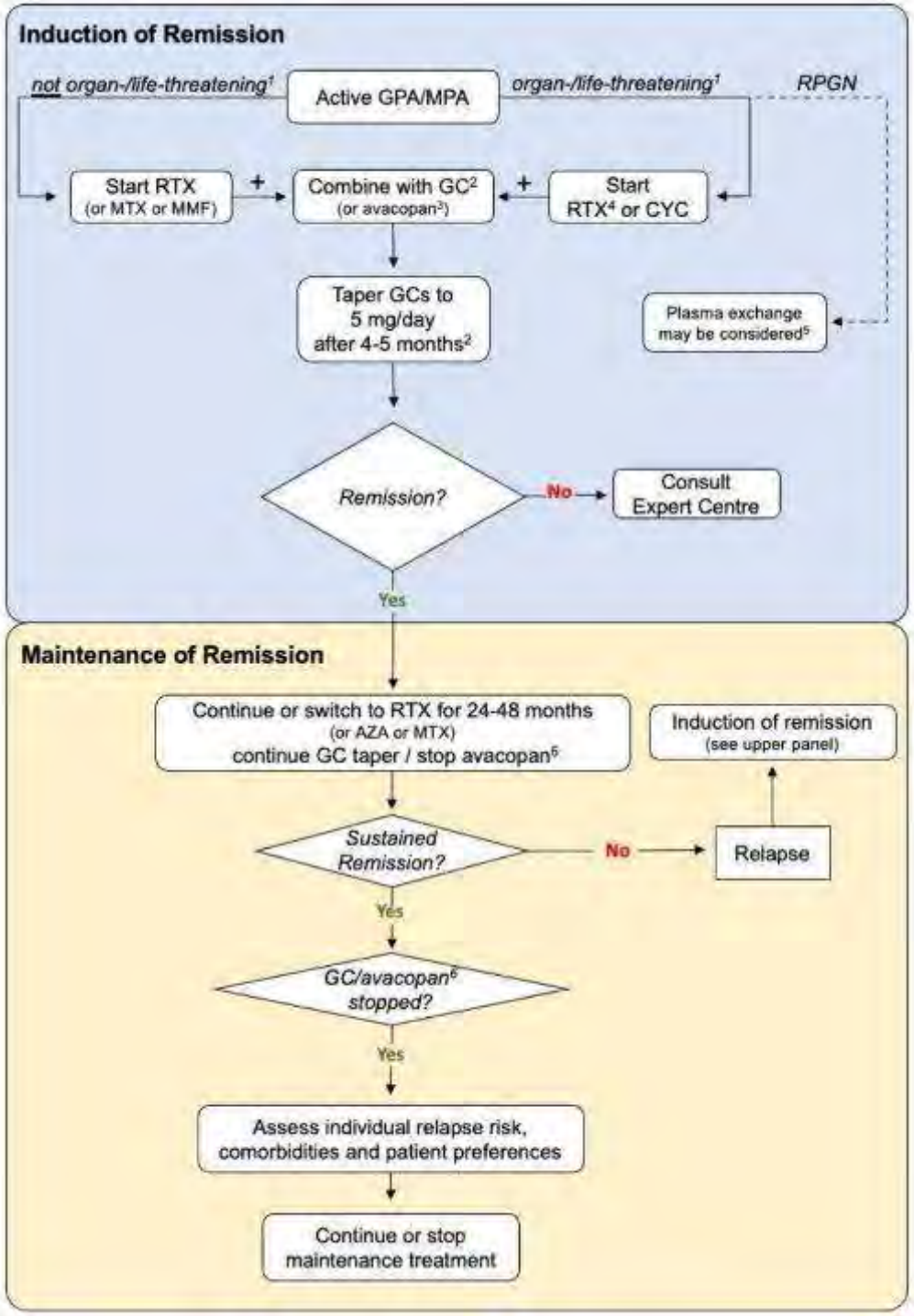
Table 1. Definitions of selected terms used in the recommendations and ungraded position statements for GPA, MPA, and EGPA*

Term	Definition
Disease states	
Active disease	New, persistent, or worsening clinical signs and/or symptoms attributed to GPA, MPA, or EGPA and not related to prior damage
Severe disease	Vasculitis with life- or organ-threatening manifestations (e.g., alveolar hemorrhage, glomerulonephritis, central nervous system vasculitis, mononeuritis multiplex, cardiac involvement, mesenteric ischemia, limb/digit ischemia)
Nonsevere disease	Vasculitis without life- or organ-threatening manifestations (e.g., rhinosinusitis, asthma, mild systemic symptoms, uncomplicated cutaneous disease, mild inflammatory arthritis)
Remission	Absence of clinical signs or symptoms attributed to GPA, MPA, or EGPA, on or off immunosuppressive therapy
Refractory disease	Persistent active disease despite an appropriate course of immunosuppressive therapy
Relapse	Recurrence of active disease following a period of remission
Treatments	
IV pulse GCs	IV methylprednisolone 500–1,000 mg/day (adults) or 30 mg/kg/day (children; maximum 1,000 mg/day) or equivalent for 3–5 days
High-dose oral GCs	Prednisone 1 mg/kg/day (adults; generally up to 80 mg/day) or 1–2 mg/kg/day (children; generally up to 60 mg/day) or equivalent
Remission induction therapy	
Methotrexate	Up to 25 mg/week (SC or oral)
Azathioprine	Up to 2 mg/kg/day
Mycophenolate mofetil	Up to 1,500 mg (oral) twice per day
Cyclophosphamide	Up to 2 mg/kg/day (oral) for 3–6 months; or intermittent 15 mg/kg (IV) every 2 weeks for 3 doses, followed by 15 mg/kg (IV) every 3 weeks for at least 3 doses (adults)
Rituximab	375 mg/m^2 (IV) weekly for 4 doses or 1,000 mg on days 1 and 15 (adults); or 375 mg/m^2 (IV) weekly for 4 doses or 575 mg/m^2 for patients with body surface area ≤1.5m^2 or 750 mg/m^2 for patients with body surface area >1.5m^2 with a typical maximum of 1 gm per infusion (both for 2 doses, days 1 and 15) (children)
Mepolizumab	300 mg (SC) every 4 weeks (adults)
Remission maintenance therapy	
Methotrexate, azathioprine, mycophenolate mofetil	Same dosing regimen as in remission induction therapy
Rituximab	500 mg (IV) every 6 months or 1 gm (IV) every 4 months (adults), 250 mg/m^2 (IV) every 6 months (children), or other doses
Mepolizumab	300 mg (SC) every 4 weeks
Omalizumab	300–600 mg (SC) every 2–4 weeks

* GPA = granulomatosis with polyangiitis; MPA = microscopic polyangiitis; EGPA = eosinophilic granulomatosis with polyangiitis; IV = intravenous; GCs = glucocorticoids; SC = subcutaneous.

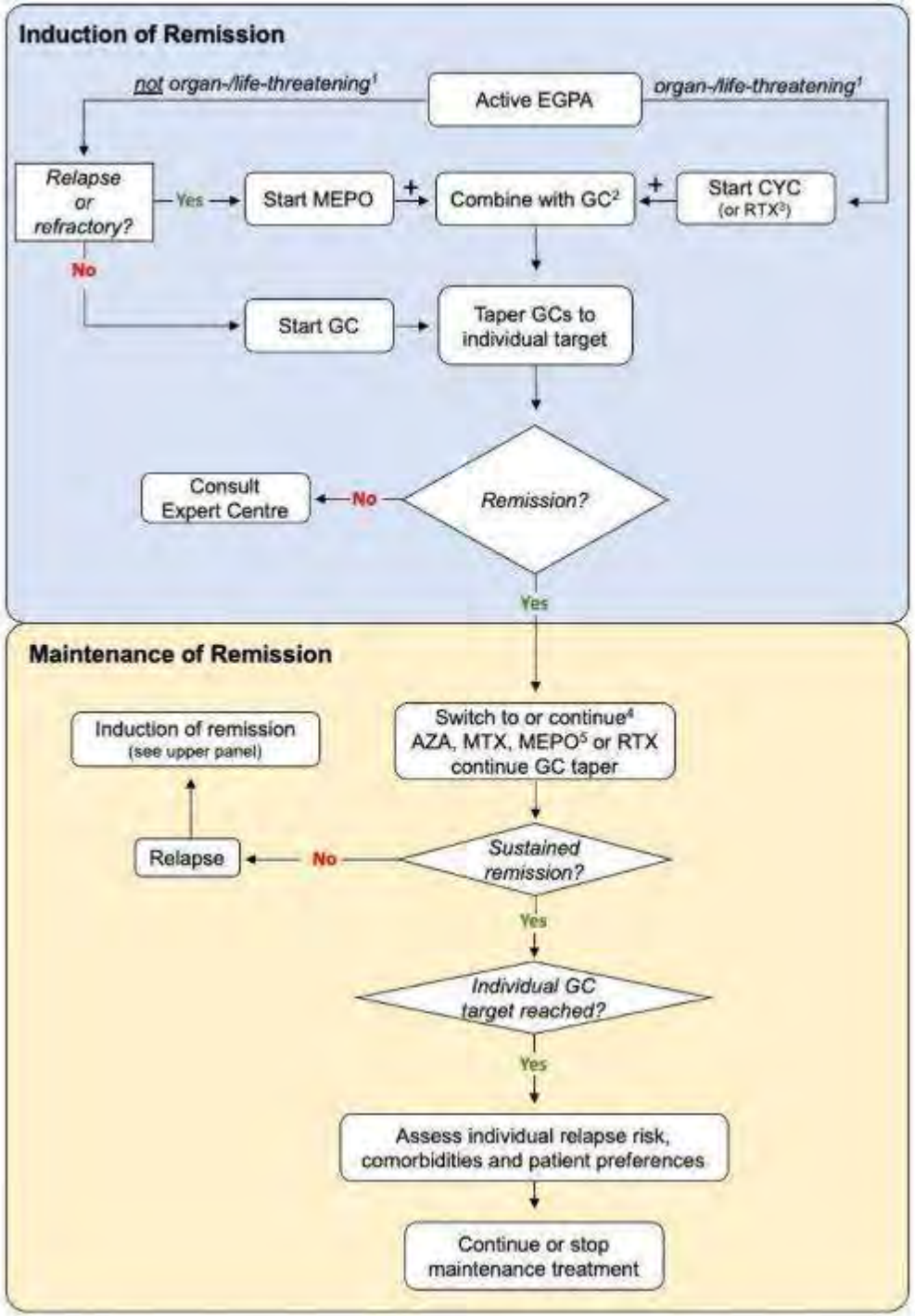
Arthritis Care & ResearchVol. 73, No. 8, August 2021, pp 1088–1105

The 2022 EULAR algorithm for treatment of granulomatosis with polyangiitis (GPA) or microscopic polyangiitis (MPA).



Bernhard Hellmich et al. Ann Rheum Dis 2024;83:30-47

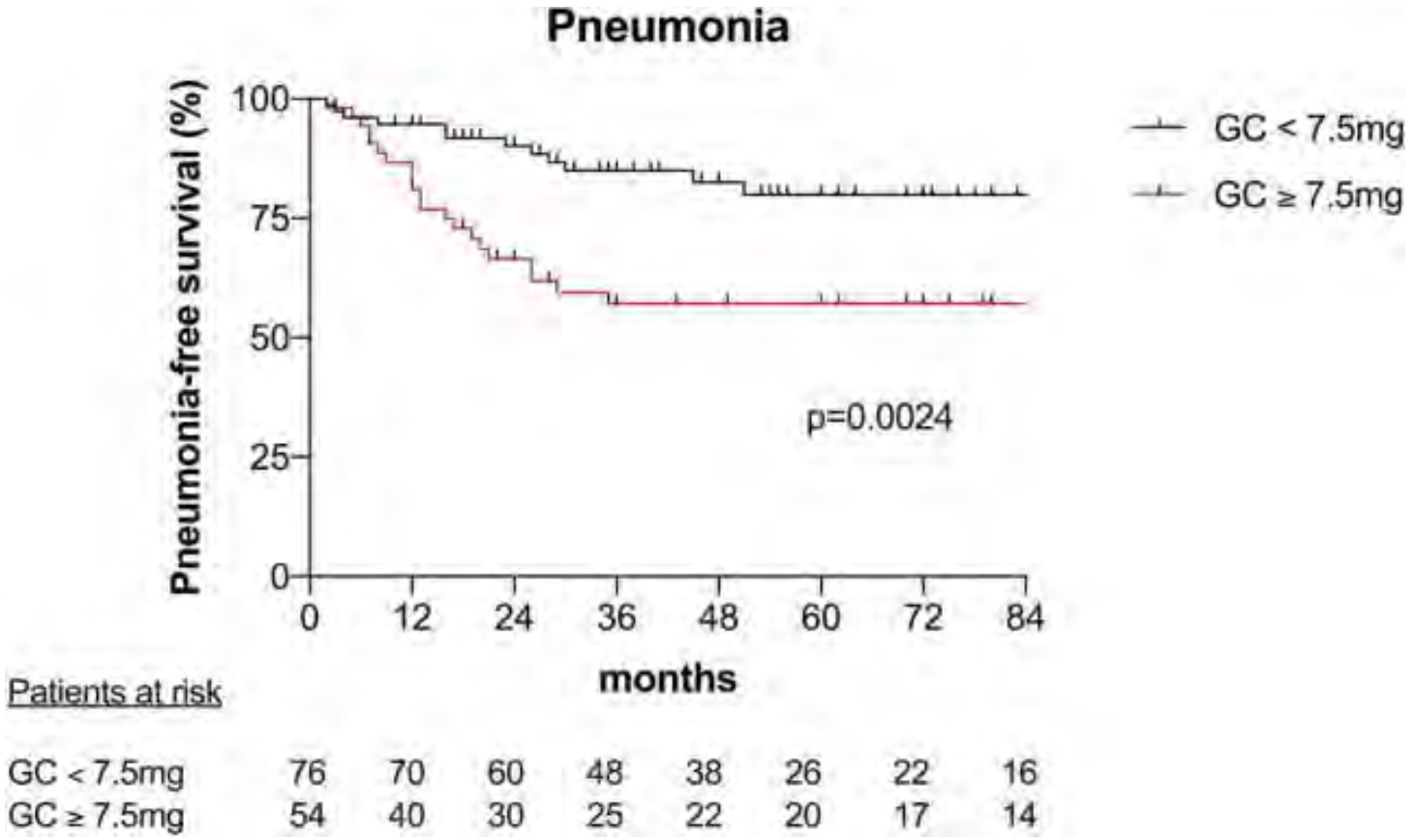
The 2022 EULAR algorithm for treatment of eosinophilic granulomatosis with polyangiitis (EGPA).



Bernhard Hellmich et al. Ann Rheum Dis 2024;83:30-47

Szteroid csökkentés

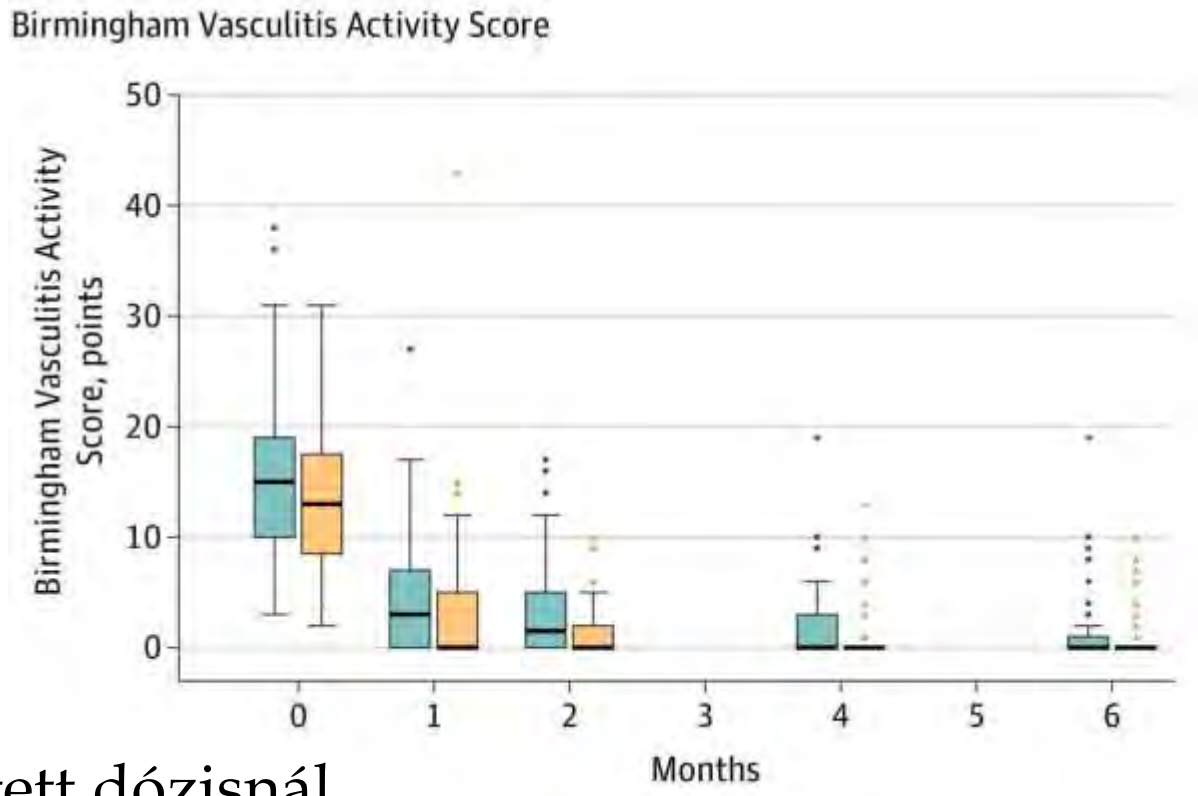
- 130 AAV beteg retrospektíve
  - 70 GPA +60 MPA
  - 76 beteg GC < 7.5 mg, 6 mo
  - 54 beteg GC ≥ 7.5 mg, 6 mo
- Kiindulás azonos
  - Aktivitás
  - báziskezelések



Rheumatol Int. 2021; 41(2): 431–438.

Szteroid csökkentés

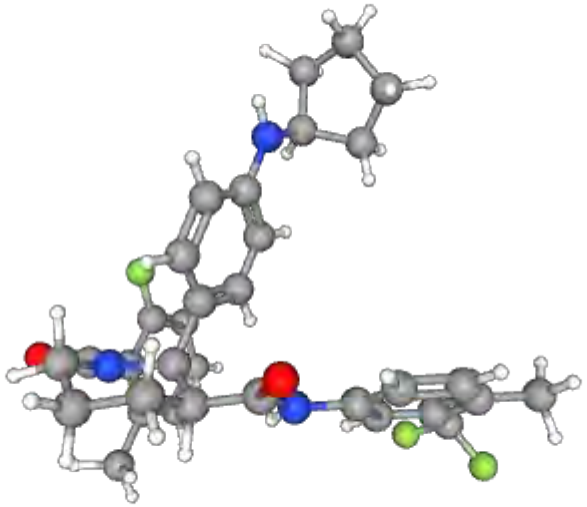
- multicenter, open-label, randomized, noninferiority trial.
- 140 új AAV beteg indukciós kezelése
  - rituximab (375 mg/m2/wk, 4 doses)
    - prednisolone (0.5 mg/kg/d), n= 70
    - prednisolone (1 mg/kg/d), n= 70
- 6 hónap
  - remisszió
    - 49 (71.0%) beteg a csökkentett
    - 45 (69.2%) beteg az alap dózisúban
  - Súlyos AE:
    - 21 AE 13 betegben (18,8%) a csökkentett dózisnál
    - 41 AE 24 betegben 36,9%) az alapdózisnál
    - p=0,02

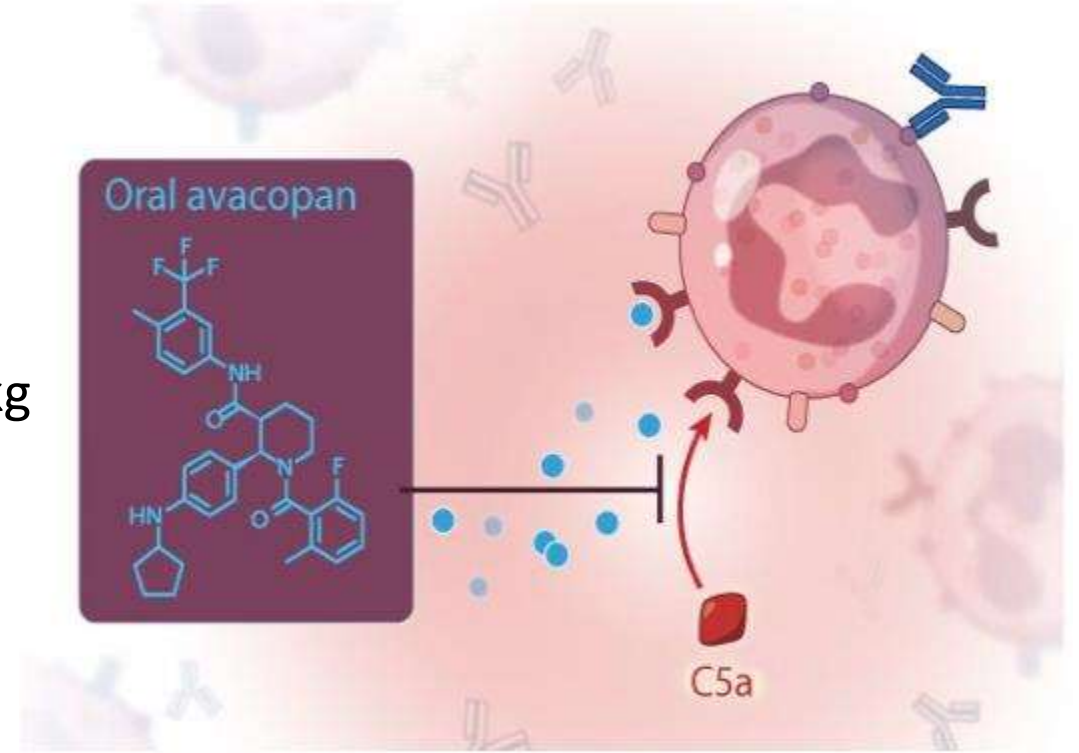


JAMA. 2021. 1;325(21):2178-2187.

Szteroid csökkentés

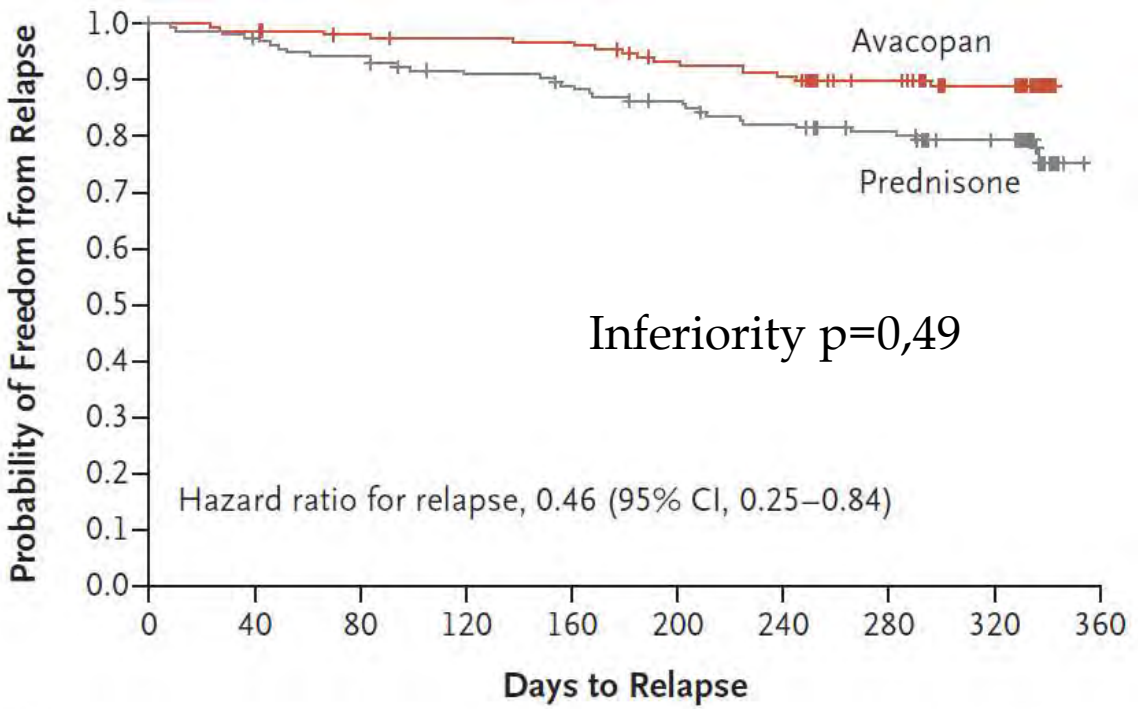
- Avacopan
  - C5a receptor antagonista
  - Kis molekula, p.o. szedhető
  - Neutrofil kemotaxist és aktivációt blokkolja
- Elfogadva
  - FDA 2021.10.
  - EMA 2022.01.
- Prednisolonnal szemben
  - Avacopan 2x30mg/die 52 hétig
  - Prednisolon 60mg, 21. hétre leépítve
- Báziskezelés
  - CYC 15mg/tskg iv. 2-3 hetente 6x, majd AZA 2mg/tskg
  - CYC 2mg/tskg p.o.
  - Rtx 375mg/m2/hét i.v. 4 héten át
- 331 beteg 143 centrumból





N Engl J Med. 2021, 18;384(7):599-609

Avacopan



Inferiority p=0,49

No. at Risk										
Avacopan	158	153	149	146	145	133	129	115	92	0
Prednisone	157	151	146	137	133	126	119	111	90	0

End Point	Avacopan (N=166)	Prednisone (N=164)	Difference (95% CI)
Primary end points			
Remission at wk 26 — no. (%)†	120 (72.3)	115 (70.1)	3.4 (–6.0 to 12.8)‡§
Sustained remission at wk 52 — no. (%)¶	109 (65.7)	90 (54.9)	12.5 (2.6 to 22.3)‡‖

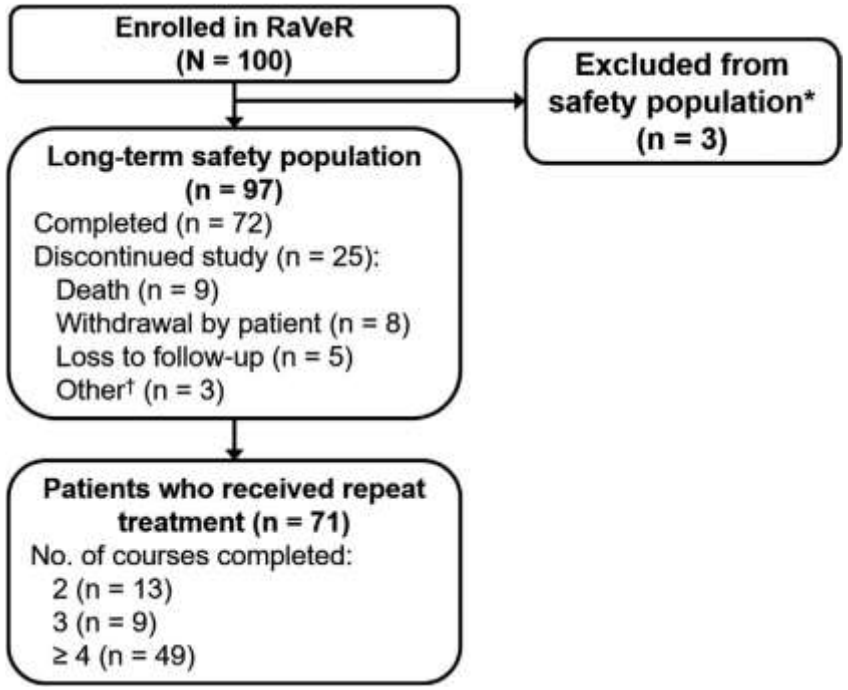
Superiority p=0,007

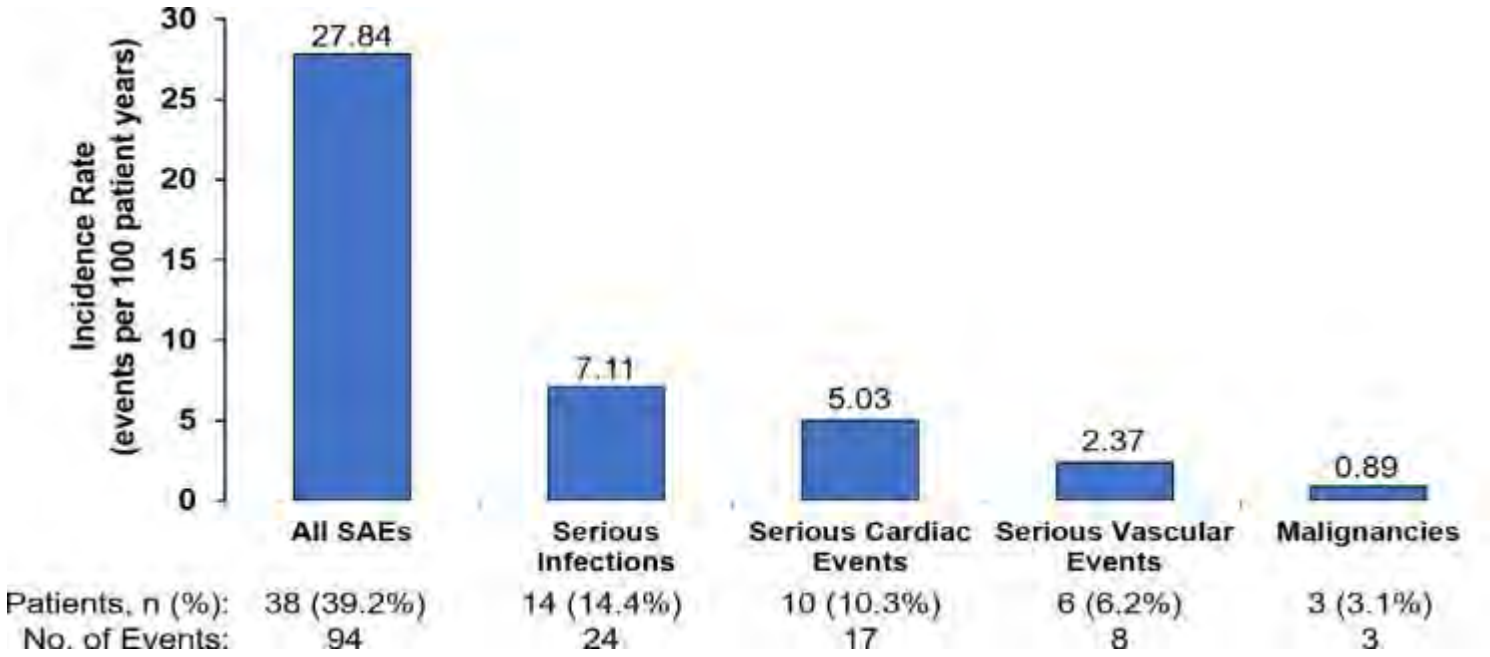
Bármely súlyos, de nem az alapbetegség aktiválódásából származó AE:
33%-kal magasabb volt Prednizolon csoportban

N Engl J Med. 2021, 18;384(7):599-609

Rtx – hosszú távú hatások - RaVeR

phase IV, multicenter, prospective, observational study



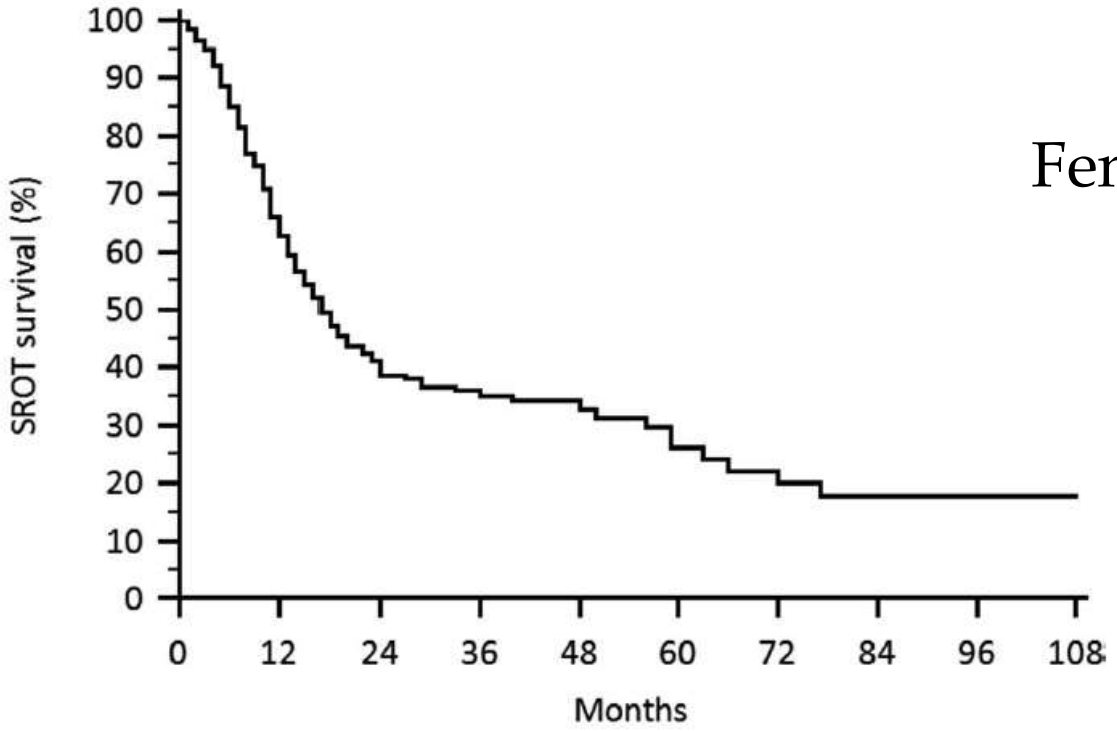


Súlyos fertőzés AAV: IR of 7.11 per 100 Pys
Súlyos fertőzés RA: IR of 7.6 per 100 PYs – Sunstone registry

Arthritis Care & Research. 2021, 9: 1372–1378

Rtx – hosszú távú hatások - FVSG

SROT=≥6 egymáskövető hó GC és IM terápia leállítása után – 259/795 beteg (33%)



Indukció

3 év SROT: CYC= 26% vs Rtx 24%

5 év SROT: több CYC (9 vs 6)

Fenntartó

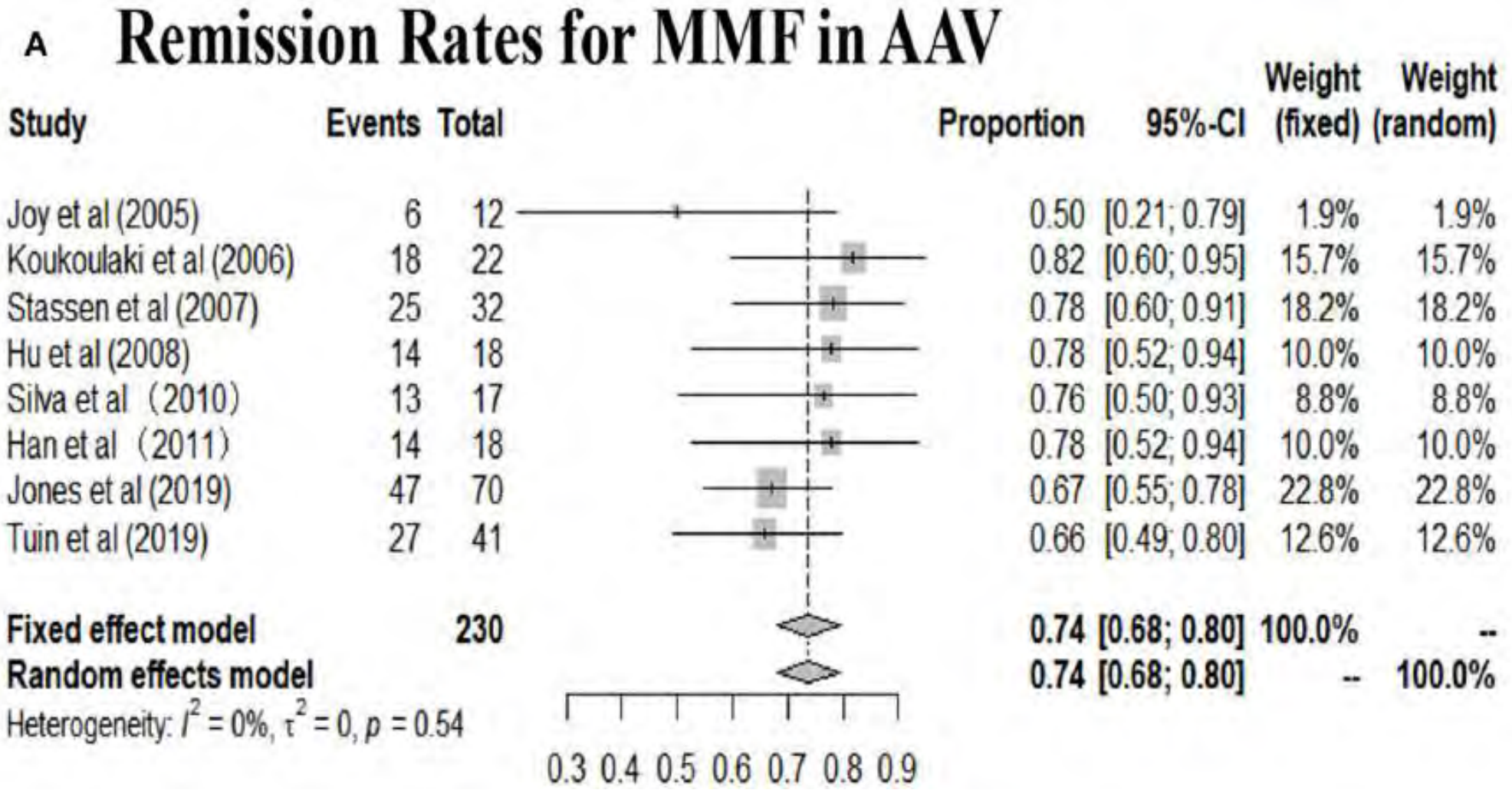
3 év SROT: Rtx 30% vs egyéb 20%

vs AZA 20%

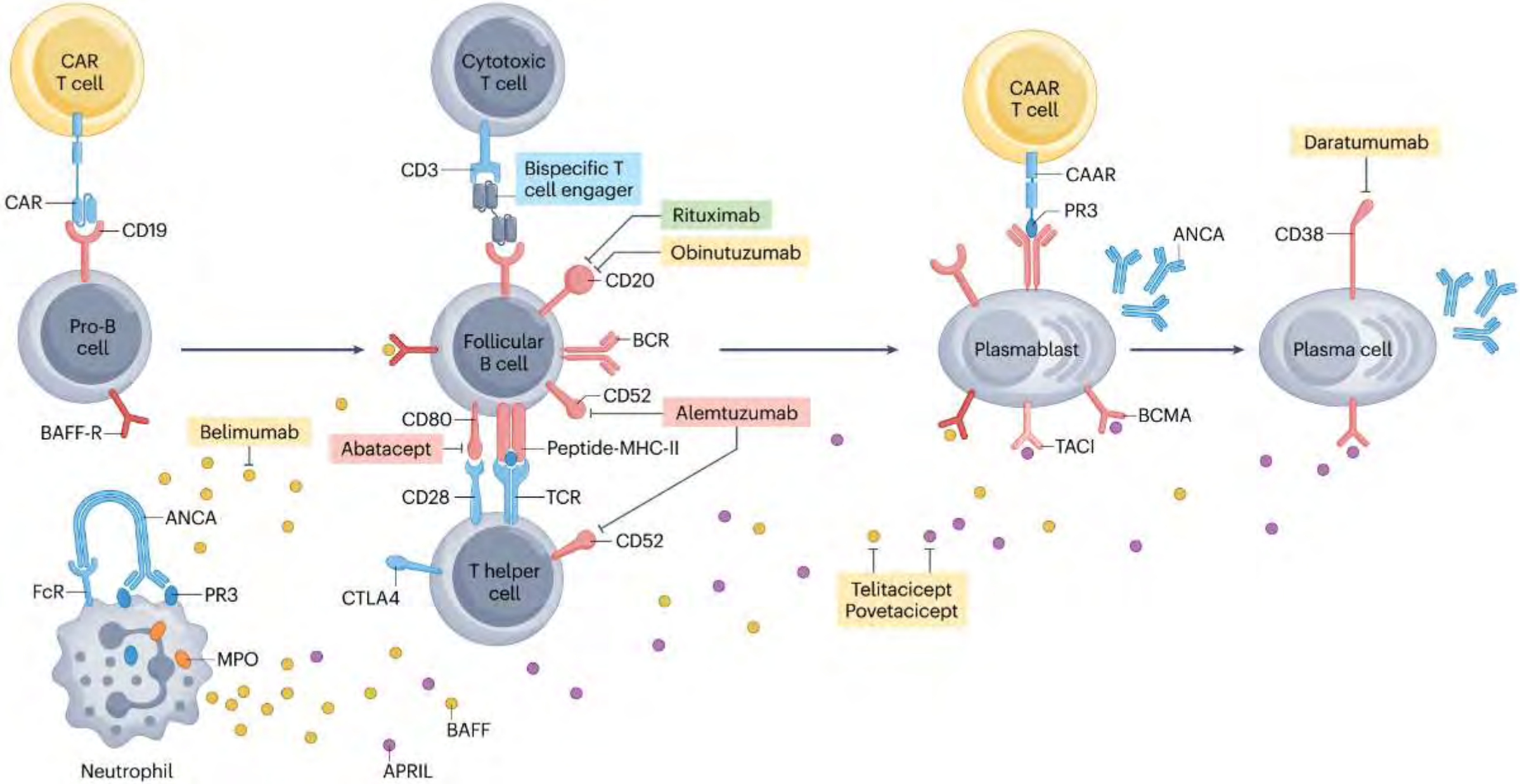
vs Mtx 17%

Arthritis & Rheumatology 2021, 4: 641–650

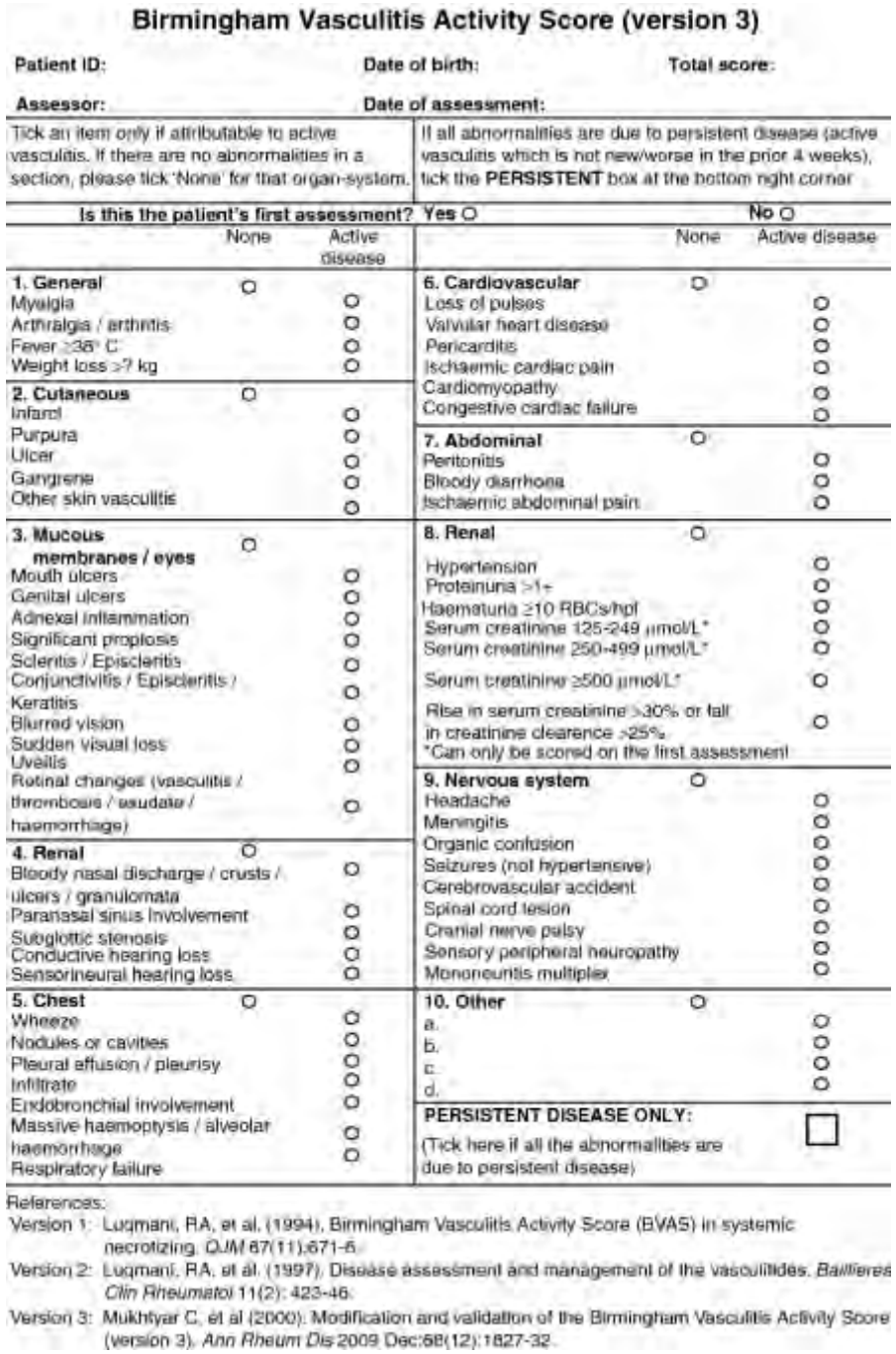
Remisszió indukció MMF-vel?



Front Med (Lausanne). 2021; 8: 609924.



Nature Reviews Rheumatology | Volume 21 | July 2025 | 396–413



Birmingham Vasculitis Activity Score (version 3)

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

Assessor: **Date of assessment:**

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

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

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

	None	Active disease
1. General	○	
Myalgia		○
Arthralgia / arthritis		○
Fever ≥38° C		○
Weight loss >7 kg		○
2. Cutaneous	○	
Infarct		○
Purpura		○
Ulcer		○
Gangrene		○
Other skin vasculitis		○
3. Mucous membranes / eyes	○	
Mouth ulcers		○
Genital ulcers		○
Adnexal inflammation		○
Significant proptosis		○
Scleritis / Episcleritis		○
Conjunctivitis / Episcleritis / Keratitis		○
Blurred vision		○
Sudden visual loss		○
Uveitis		○
Retinal changes (vasculitis / thrombosis / exudate / haemorrhage)		○
4. Renal	○	
Bloody nasal discharge / crusts / ulcers / granulomata		○
Paranasal sinus involvement		○
Subglottic stenosis		○
Conductive hearing loss		○
Sensorineural hearing loss		○
5. Chest	○	
Wheeze		○
Nodules or cavities		○
Pleural effusion / pleurisy		○
Infiltrate		○
Endobronchial involvement		○
Massive haemoptysis / alveolar haemorrhage		○
Respiratory failure		○

	None	Active disease
6. Cardiovascular	○	
Loss of pulses		○
Valvular heart disease		○
Pericarditis		○
Ischaemic cardiac pain		○
Cardiomyopathy		○
Congestive cardiac failure		○
7. Abdominal	○	
Peritonitis		○
Bloody diarrhoea		○
Ischaemic abdominal pain		○
8. Renal	○	
Hypertension		○
Proteinuria >1+		○
Haematuria ≥10 RBCs/hpf		○
Serum creatinine 125-249 µmol/L*		○
Serum creatinine 250-499 µmol/L*		○
Serum creatinine ≥500 µmol/L*		○
Rise in serum creatinine >30% or fall in creatinine clearance >25%		○
*Can only be scored on the first assessment		
9. Nervous system	○	
Headache		○
Meningitis		○
Organic confusion		○
Seizures (not hypertensive)		○
Cerebrovascular accident		○
Spinal cord lesion		○
Cranial nerve palsy		○
Sensory peripheral neuropathy		○
Mononeuritis multiplex		○
10. Other	○	
a.		○
b.		○
c.		○
d.		○

PERSISTENT DISEASE ONLY: ☐

(Tick here if all the abnormalities are due to persistent disease)

References:

Version 1: Luqmani, RA, et al. (1994). Birmingham Vasculitis Activity Score (BVAS) in systemic necrotizing. *QJM* 87(11):671-6.

Version 2: Luqmani, RA, et al. (1997). Disease assessment and management of the vasculitides. *Baillieres Clin Rheumatol* 11(2): 423-46.

Version 3: Mukhtyar C, et al (2000). Modification and validation of the Birmingham Vasculitis Activity Score (version 3). *Ann Rheum Dis* 2009 Dec;68(12):1827-32.

Betegségaktivitás követése

Birmingham Vasculitis Activity Score (BVAS)

Five factor score (FFS) 1996.

1. Cardiac involvement
2. Gastrointestinal disease
3. Plasma creatinine levels >1.6 mg/dl (141 mmol/l)
4. Proteinuria >1 g/day
5. Central nervous system involvement

Revised 2011 five-factor score [17]

- Age> 65 years
- Cardiac insufficiency
- Renal insufficiency (creatinine >1.7 mg/dl)
- Gastrointestinal involvement
- Absence of ear, nose and throat manifestation

VASCULITIS DAMAGE INDEX (VDI)

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

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

Name
Trial Number
Date
Centre

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

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

VDI. Modified from: Exley AR, Bacon PA, Luqmani RA et al (1997). Development and initial validation of the VDI. *Ann Rheum Dis* 56: 371–380

Quality Of Life

- Sort Form Health Survey (SF36)
- EuroQol-5 (EQ5)
- Health Assesmetn Questionaire (HAQ)

Long-term outcomes and prognostic factors for survival of patients with ANCA-associated vasculitis (AAV)

Background

Despite advances in diagnosis and treatment, patients with AAV have a poor prognosis, and the predicative factors are not well categorized. Evaluation of long-term outcomes in major European RCTs and identifying prognostic factors.

Methods

Multicenter
74 centers, 17 countries in Europe

848 patients
Enrolled 1995–2012 in 7 EUVAS (European Vasculitis Society) randomized clinical trials

- Newly diagnosed with AAV
- Compared to matched background population

GPA 56% **MPA 44%**

Median long-term follow-up
8 years (IQR: 2.9–13.6)

→ Survival
Causes of death
Prognostic factors

Results

Median survival from diagnosis: 17.8 years
*95% CI 15.7–20 years

Excess mortality compared to general population

14% at 1 year
20% at 10 years
29% at 15 years

Main causes of death


26% Infection


14% Cardiovascular disease


13% Malignancy

Negative prognostic factors:
Advanced age 
Male sex ♂
Low eGFR 
Low platelet count 

Conclusion

Patients with AAV have an increased risk of mortality compared to the general population. Treatment complications and organ damage are the main causes of limited survival. Infections are the leading cause of death.

ERA NDT NEPHROLOGY DIALYSIS TRANSPLANTATION

Sánchez Álamo B., Moi L. et al., on the behalf of the EUVAS. NDT (2022)
@NDTSocial

PROTEINURIA AND HEMATURIA AFTER REMISSION INDUCTION ARE ASSOCIATED WITH OUTCOME IN ANCA-ASSOCIATED VASCULITIS

kidney INTERNATIONAL

ISN INTERNATIONAL SOCIETY OF NEPHROLOGY

Patients and exposures

Data from 5 European RCTs (RITUXVAS, IMPROVE, MYCYC, MAINRITSAN 1&2)

- 571 patients with AAV
- 60% PR3, 35% MPO
- 77% with kidney involvement

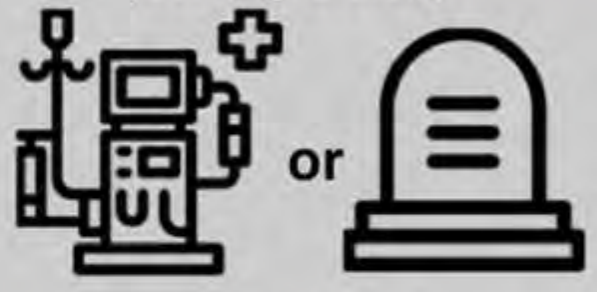
Induction therapy → 4-6 months → Maintenance therapy

Median follow-up 28months

PERSISTENT PROTEINURIA :
PCR (protein/creat ratio) ≥ 0.05 g/mmol

PERSISTENT HEMATURIA :
> 10 red blood cells/mm³
If missing data, Hu ≥ 1x on urine dipstick

Kidney failure or death (Cox regression)



HR 3.06
(CI 95% 1.09-8.59)

PERSISTENT PROTEINURIA
Is associated with kidney outcome

Renal relapse (competing risk regression)

sHR 2.16
(CI 95% 1.13-4.11)

PERSISTENT HEMATURIA
Is associated with kidney relapse

sHR 2.22
(CI 95% 1.16-4.24)

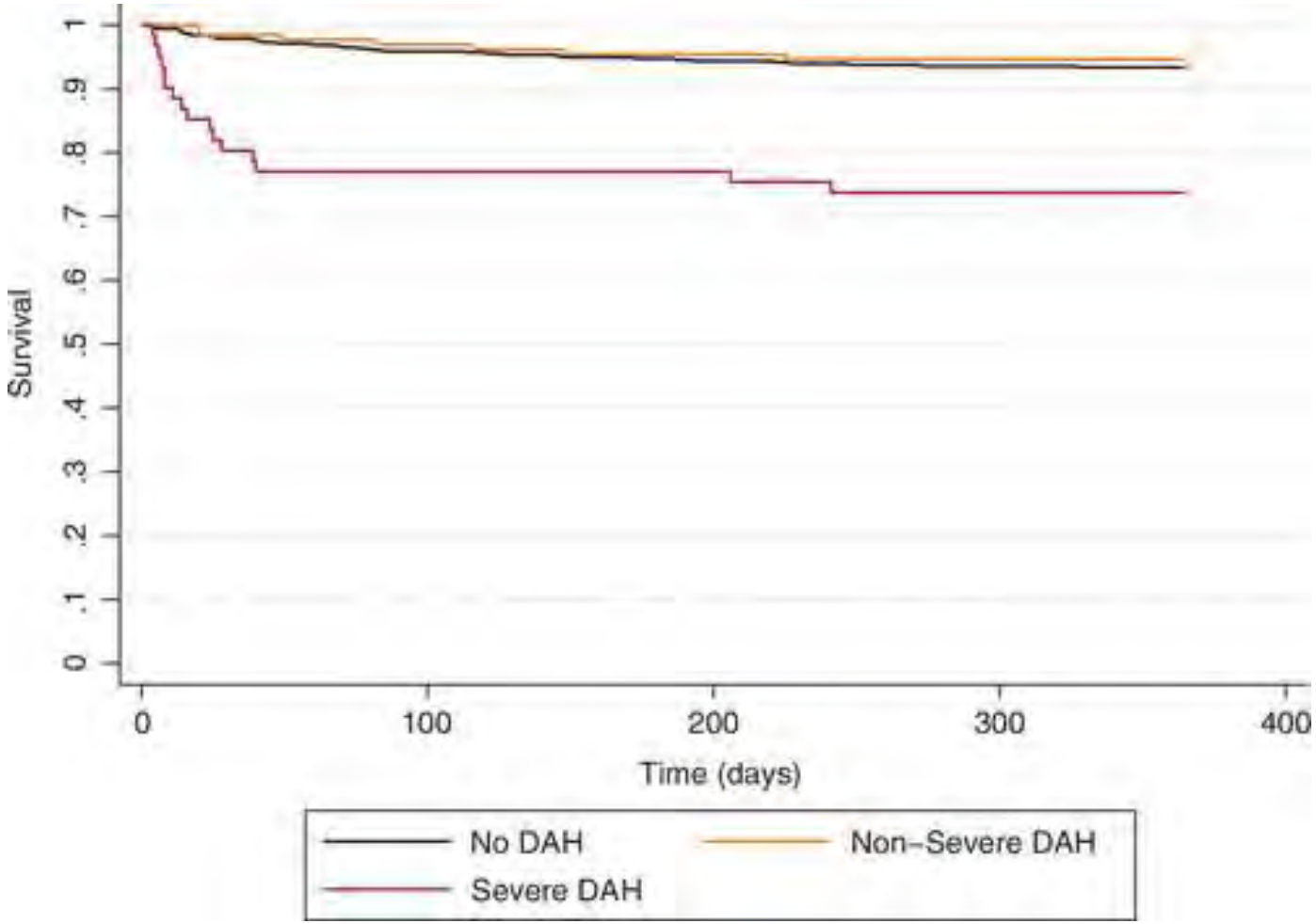
PERSISTENT PROTEINURIA
Is associated with kidney relapse

In multivariate analysis, after adjustment for
age, ANCA type, maintenance therapy, serum creatinine, PCR and hematuria after induction therapy

Benichou et al., 2022

CONCLUSION: In this large cohort of AAV patients, persistent proteinuria after induction therapy was associated with death/kidney failure and kidney relapse, whereas persistent hematuria was an independent predictor of kidney relapse

Alveolar Hemorrhage in AAV

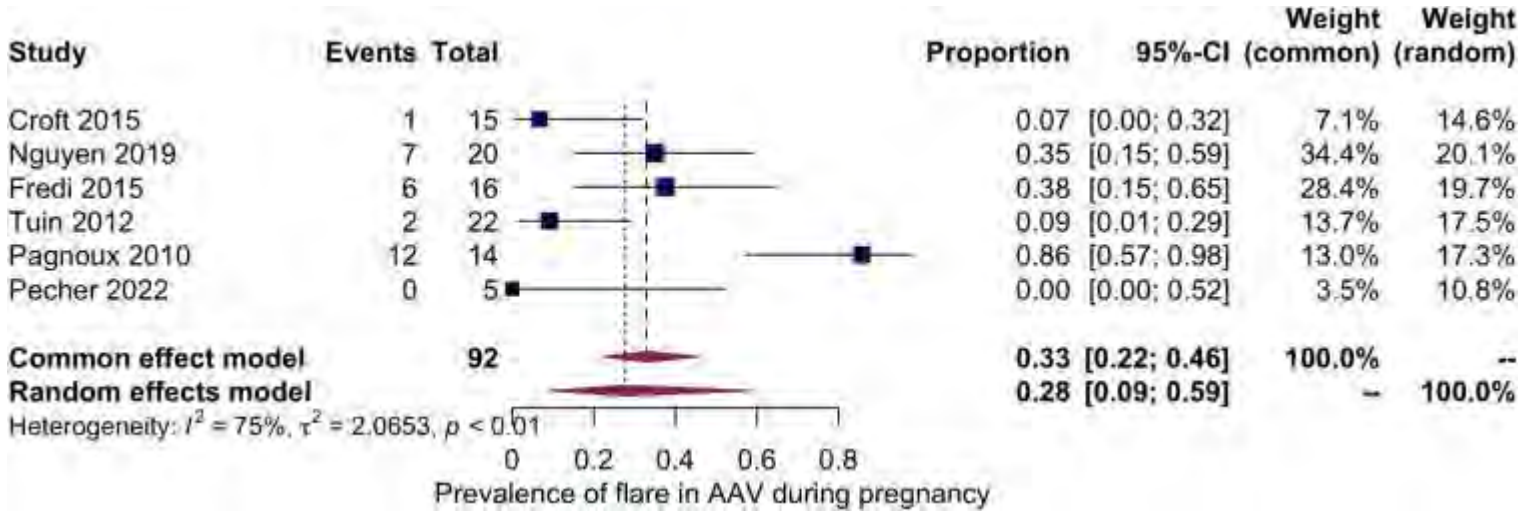


- DAS vs no-DAS
  - Fiatalabbak
  - Gyakrabban PR3-ANCA+
  - Gyakoribb relapsus
- PLEX vs. no-PLEX
  - Halál – 8% vs 15% - HR 0,52
- Csökkentett szteroid vs standard
  - 13% vs 10% - HR 1.33

Am J Respir Crit Care Med. 2024 May 1;209(9):1141-1151

Terhesség AAV-ben

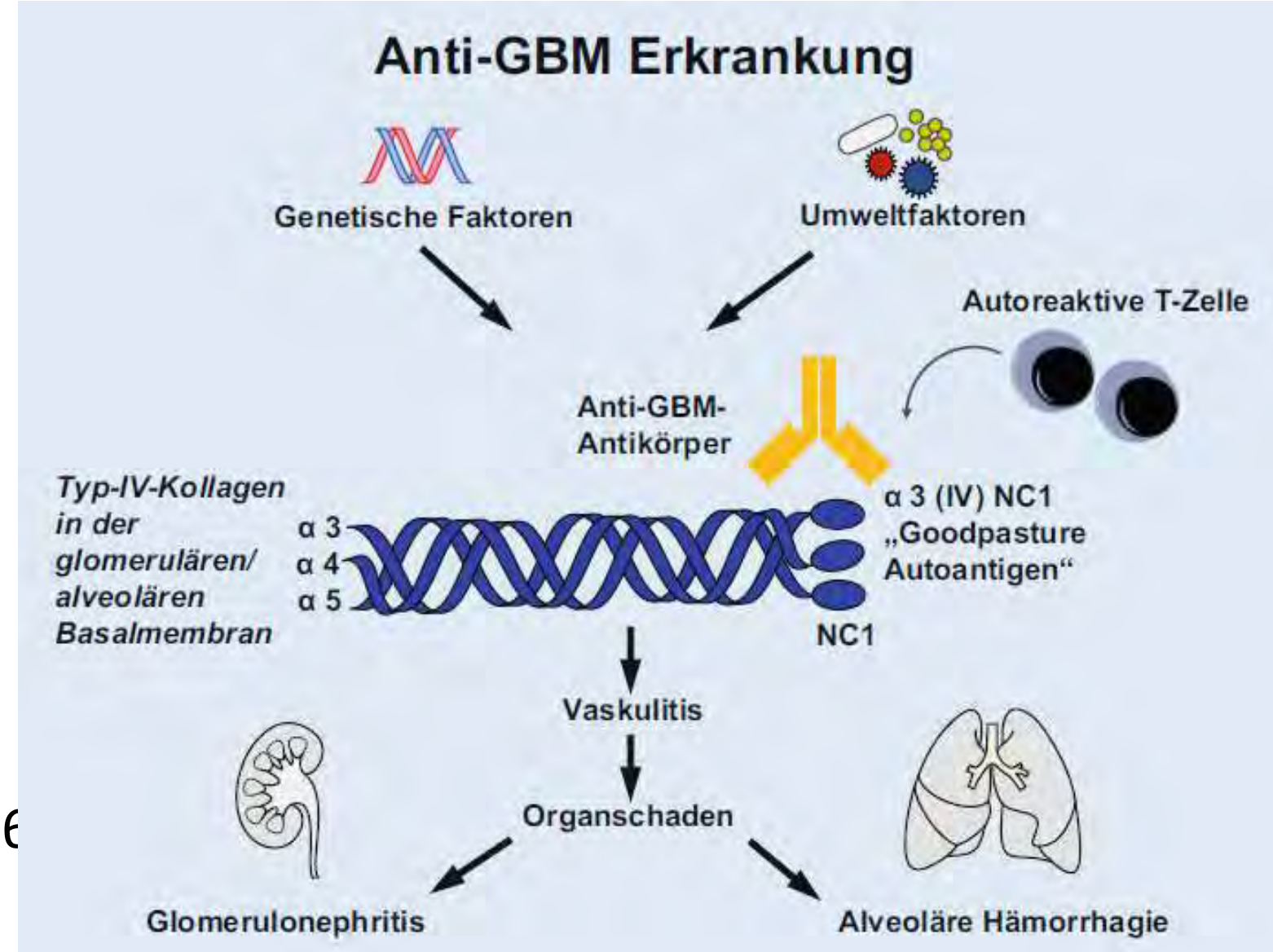
- The prevalence of pre-term delivery - 18% (CI: 0.10-0.30, P=non-significant),
- IUGR neonates -20% (CI: 0.11-0.33, P=non-significant)
- Disease flare 28% (CI: 0.09-0.59, P<0.01)



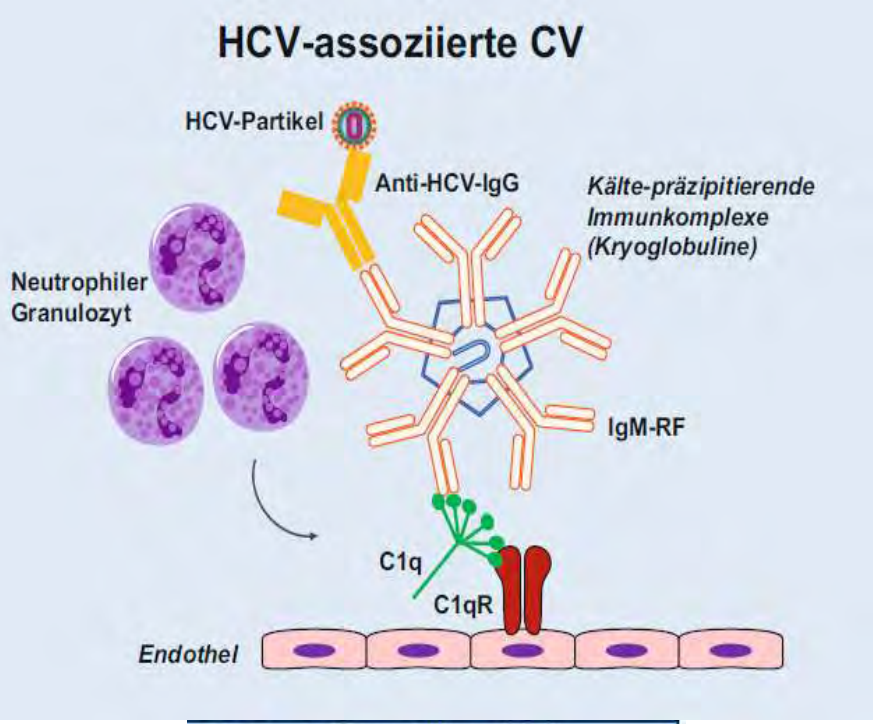
Joint Bone Spine. 2023 Dec;90(6):105609 - metanalízis

Anti-glomeruláris bazálmembrán vasculitis (Goodpasture)

- Incidencia: 1-2/1 millió /év
- GN-ek 1-5%-a
- Féldholdképződéssel járó nekrotizáló GN-ek 20-30%-a
- Pulmo-renalis sy 10-20%-a
- 3. évtized
- Férfi túlsúly
- 50%-ban ANCA is+ (MPO)
- 30-40% intenzív ellátásra
- 70-80% dialízisre
- Csoportosítás
  - Betegség- vese
  - Szindróma- vese+tüdő (40-6
  - Izolált tüdő – 10%



Kryoglobulinaemiás vasculitisek



Tab. 1 Klassifikation der Kryoglobulinämien nach Brouet [4]

	Klonalität	Assoziierte Erkrankungen
Typ I	Monoklonale Immunglobuline Typischerweise IgG oder IgM, seltener IgA oder freie Ig-Leichtketten	Monoklonale Gammopathien, multiples Myelom, Lymphome
Typ II	Gemischte monoklonale und polyklonale Immunglobuline IgM (oder IgG oder IgA), oft mit Rheumafaktoraktivität	Hepatitis C (und andere chronische Virusinfekte) SLE, Sjögren-Syndrom Lymphoproliferative Erkrankungen Essenziell (idiopathisch)
Typ III	Gemischte polyklonale Immunglobuline IgG und IgM	

Ig Immunglobulin, *SLE* systemischer Lupus erythematodes

HCV – 40-60%
Egyéb krónikus infekció – 15-25%
Egyéb szisztémás betegség – 10-20%

Tab. 2 Spektrum CV(kryoglobulinämische Vaskulitis)-assoziierter Erkrankungen und Ursachen [3, 5, 22]

Infektionen	Hepatitis-C-Virus (HCV)
	Hepatitis-B-Virus (HBV)
	Humanes Immundefizienzvirus (HIV)
	Subakute bakterielle Endokarditis
Kollagenosen	Primäres Sjögren-Syndrom
	Systemischer Lupus erythematodes (SLE)
	Mischkollagenose (MCTD)
Arthritiden	Rheumatoide Arthritis (RA)
Hämatologische Erkrankungen	Lymphome
	Plasmozytom
	Leukämien

Z Rheumatol 2022 · 81:300–304

Kryoglobulinaemiás vasculitisek

Tab. 2 Allgemeine und klinische Symptome der kryoglobulinämischen Vaskulitis. (Nach [6])

	%
Allgemeinsymptome	
Leistungsminderung	50–100
Arthralgien	>90
Sicca-Symptomatik	~50
Raynaud-Symptomatik	~50
Klinische Symptome	
Purpura	54–98
Polyneuropathie	60–80
Leberbeteiligung	~70
Hautulzera	~50
Nierenbeteiligung	~30
Arthritis	6–10
Gastrointestinale Beteiligung	2–6
Pulmonale Beteiligung	1–6
Hyperviskositätssyndrom	0,50

- Lefolyás
  - 50% benignus
  - 30% súlyos
  - 20% malignoma háttér dominál
- Kezelés
  - 2 év antivirális terápia
    - 80%-ban HCV- lesz
      - 70%-ukban kryoglobulin eltűnik
      - 20%-ban magas marad
        - 90%-uk klinikai remisszióban lesz
  - Szteroid
  - DMARD
  - Plazmaferezis
  - Rtx

Kryoglobulinaemiás vasculitisek

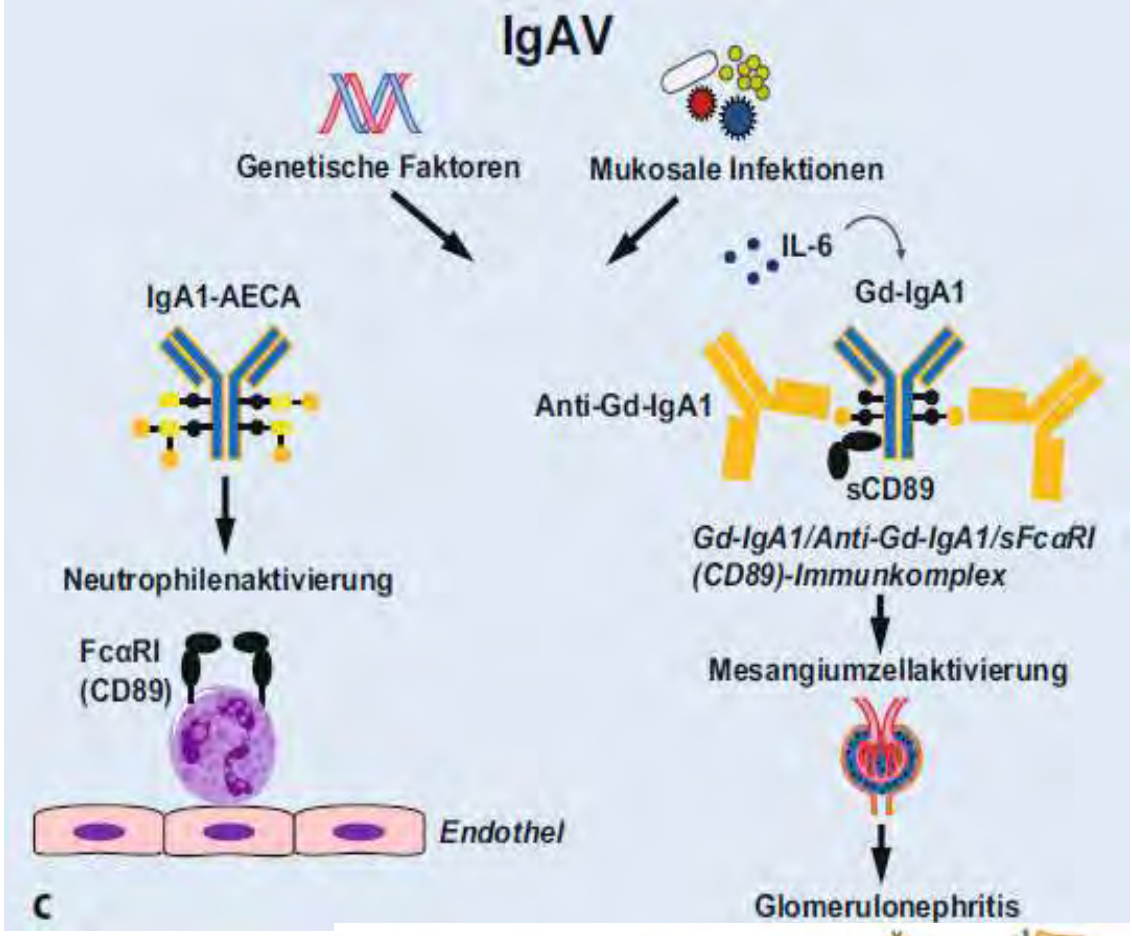
Table 2 The final set of recommendations for the management of mixed cryoglobulinemia with rituximab from the Italian Study Group of Cryoglobulinemia (GISC)

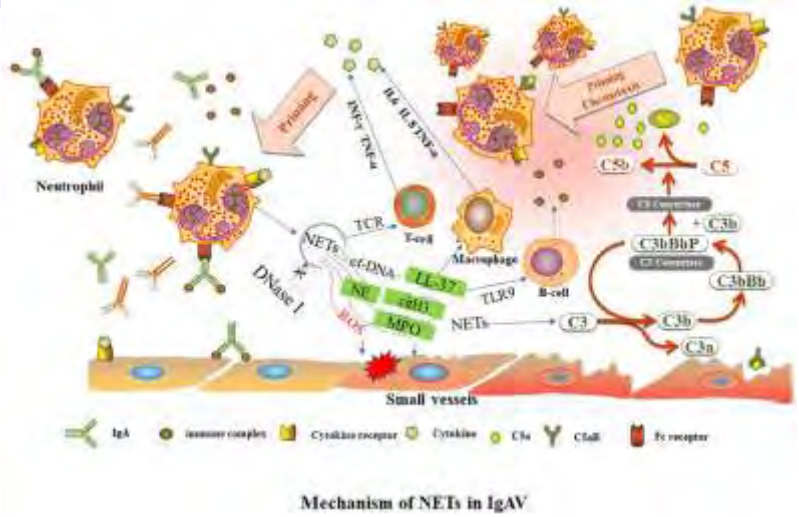
Statements	Mean	Standard deviation
1. Overall, rituximab is effective (and safe) on the severe, not immediately life-threatening, clinical manifestations of cryoglobulinemic vasculitis (LoE 1A)	92,33	7,42
2. In particular, rituximab is effective (and safe) on the glomerulonephritis of cryoglobulinemic vasculitis (LoE 2B)	91,92	8,62
3. In particular, rituximab is effective (and safe) on the peripheral neuropathy of cryoglobulinemic vasculitis (LoE 2C)	77,71	14,51
4. In particular, rituximab is effective (and safe) on the skin ulcers of cryoglobulinemic vasculitis (LoE 1A)	85,21	13,08
5. Rituximab is equally effective on other, not severe manifestations (purpura, arthralgia, fatigue) of cryoglobulinemic vasculitis (LoE 2B)	80,00	16,39
6. Rituximab may be equally effective in infectious and non-infectious cryoglobulinemic vasculitis (LoE 5C)	76,92	16,69
7. Rituximab should be used cautiously in patients carrying latent HBV infection, provided that and an adequate prophylactic therapy for HBV infection, or monitoring HBV DNA or HBsAg title should be done (LoE 5C)	92,42	26,54
8. Re-treatment at clinical relapse with rituximab, after the first cycle, is effective (and safe) in patients with severe, not immediately life-threatening, clinical manifestations of cryoglobulinemic vasculitis (LoE 2B)	90,17	10,71
9. Rituximab shows a "steroid-sparing" effect in patients with severe, not immediately life-threatening, clinical manifestations of cryoglobulinemic vasculitis (LoE 2B)	92,17	8,68
10. Rituximab does not usually carry an increased risk of serious adverse events compared to other immunosuppressants or high-dose glucocorticoids. Attention should be paid for repeated courses and multiple comorbidities (LoE 1,A)	90,31	21,17
11. Rituximab given alone is not associated with an increased risk of hepatitis C reactivation, even if a transient elevation of the viral load can be seen (LoE 1B)	89,50	19,68
12. The risk of severe infusion reactions during rituximab administration is very low (LoE 1A)	87,58	10,94
13. Rituximab is effective and safe in combination with antivirals in some cases of cryoglobulinemic vasculitis (LoE 5C)	91,38	11,55
14. Rituximab is effective in patients with HCV-related cryoglobulinemic vasculitis showing persistent and severe clinical course, despite virological clearance by antivirals (LoE 5C)	89,92	9,05
15. Rituximab given at low doses (250 mg/mq weekly for 2 weeks) is equally effective as given at high doses (375 mg/mq/weekly for 4 weeks or 1 g 2 weeks apart) in somecases of cryoglobulinemic vasculitis (LoE 5C)	72,00	27,16
16. Maintenance treatment with rituximab is required in severe or life-threatening cryoglobulinemic vasculitis (LoE 5C)	74,58	29,47

Clinical Rheumatology September 2022

IgA vasculitis (Henoch-Schönlein)

- Incidencia:
  - Gyerekkor (5-8 évesek): 30-260/1millió/év
  - Felnőttkor (40-60 évesek): 1-18/1millió/év
  - Férfi>nő
- Kiváltó:
  - Fertőzés felsőlégúti 30-40%
  - Fertőzés GI (HP) 20-30%
  - Gyógyszerek 20%
  - Malignómák 10%
- Szövettan:
  - Leukocitoklasztikus vaszkulitisz
  - IgA (+C3) depozíció
    - Bőrben
    - Mesangiumban
- Szervek – bőr 80-100% izület 90% vese 70%, GI 50%
- Dialízis
  - Gyerekkorban 8%
  - Felnőtteknél 18%





Mechanism of NETs in IgAV

Z Rheumatol 2022 · 81:305–312

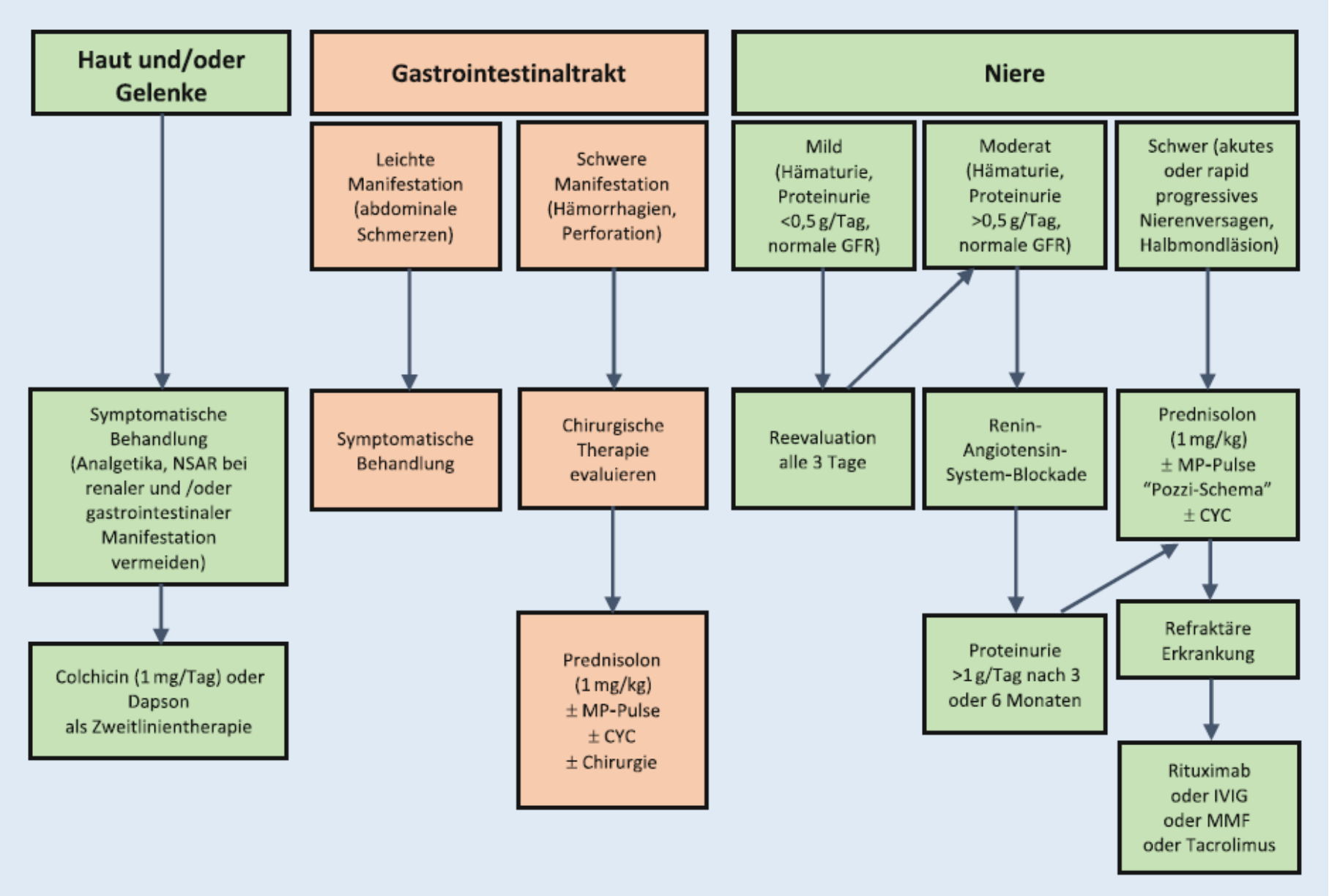
IgA vasculitis (Henoch-Schönlein)

Tab. 1 ACR(American College of Rheumatology)- und EULAR(European League against Rheumatism)/PRINTO(Paediatric Rheumatology International Trials Organization)/PRES(Paediatric Rheumatology European Society)-Klassifikationskriterien und CHCC(Chapel Hill Consensus Conference)-Nomenklatur der IgA(Immunglobulin A)-Vaskulitis

ACR-Klassifikationskriterien (1990)	EULAR/PRINTO/PRES-Klassifikationskriterien 2008	Chapel Hill Consensus Conference (CHCC) 2012
Alter ≤ 20 Jahre bei Krankheitsbeginn	Palpable Purpura, dominierend an den unteren Extremitäten	Vaskulitis mit IgA1-dominanten Ablagerungen
Palpable Purpura	Diffuse, kolikartige abdominelle Schmerzen mit akutem Beginn (Invagination oder Blutung möglich)	Betroffen sind die kleinen Gefäße (Kapillaren, Venolen oder Arteriolen) der Haut und des Gastrointestinaltraktes
Akute abdominelle Schmerzen	Leukozytoklastische Vaskulitis mit dominierend IgA-Ablagerungen oder proliferative Glomerulonephritis mit IgA-Ablagerungen	Arthritiden sind häufig
Histologischer Nachweis von Granulozyten in der Wand der kleinen Arteriolen oder Venolen	Arthritis mit akutem Beginn (Schmerzen, Schwellung und Funktionseinschränkung), Arthralgien mit akutem Beginn (Schmerzen ohne Schwellung oder Funktionseinschränkung)	Glomerulonephritis (nicht von der IgA-Nephropathie zu unterscheiden) kann auftreten
–	Proteinurie > 0,3 g/24 h oder > 30 mmol/mg Albumin/Kreatinin-Ratio im Spontanurin, Hämaturie oder Erythrozytenzylinder: > 5 Erythrozyten/HPF oder Erythrozytenzylinder oder Proteinurie ≥ 2+ im Spontanurin	–
≥ 2 von 4 Kriterien	Kriterium 1 (obligatorisch) und ≥ 1 weiteres Kriterium	–
Sensitivität 87,1 %, Spezifität 87,7 %	Sensitivität 100 %, Spezifität 87 %	–

HPF „High Power Field", Hauptgesichtsfeld

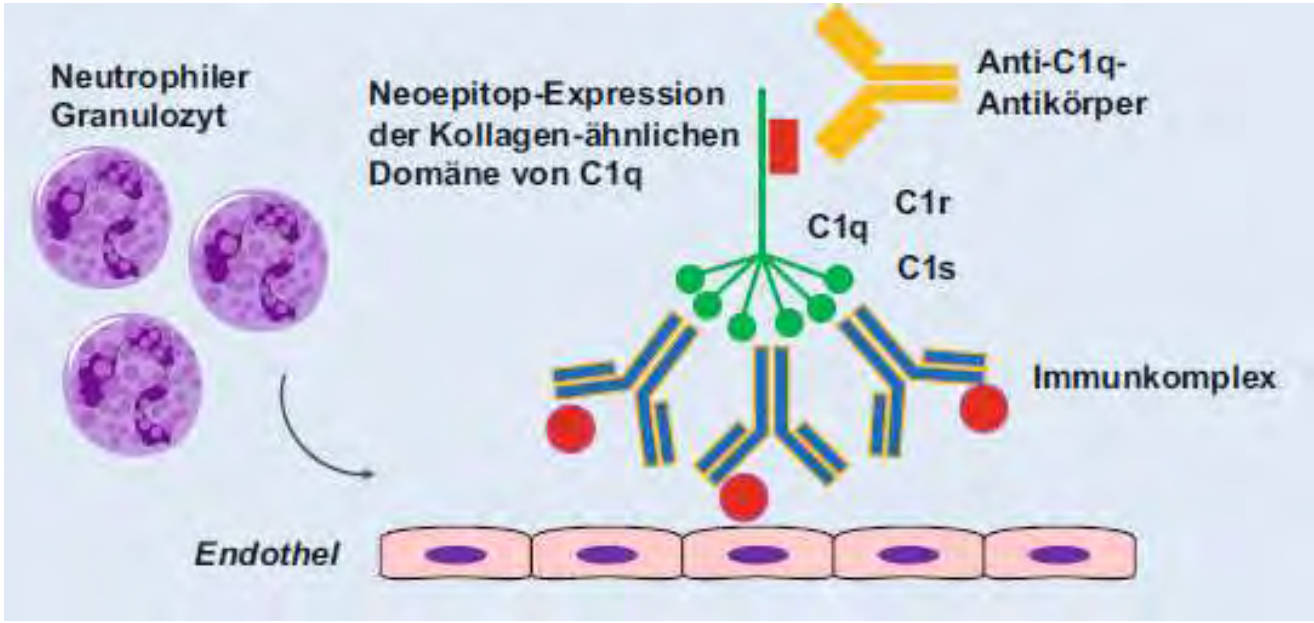
IgA vasculitis (Henoch-Schönlein)



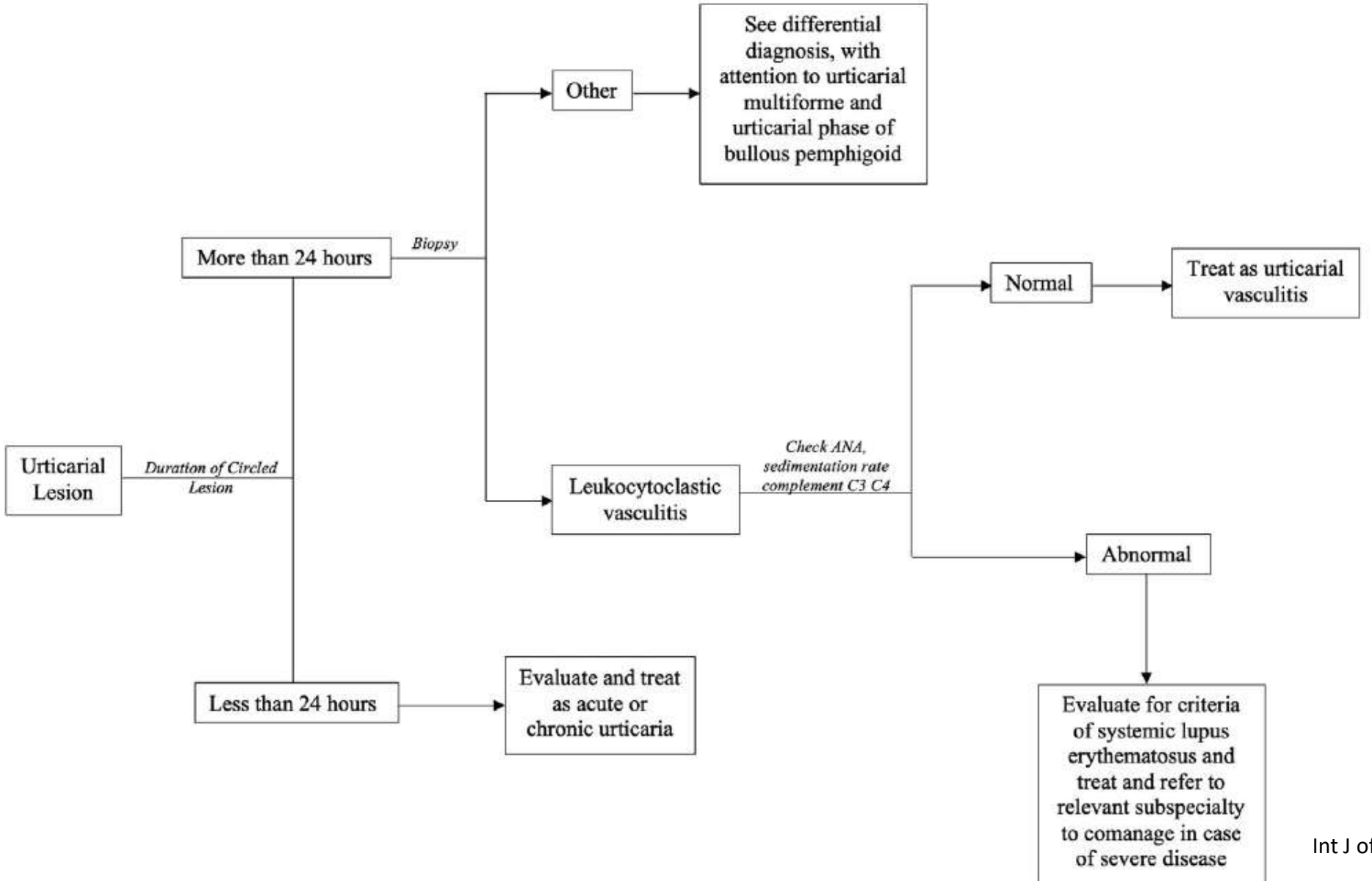
Z Rheumatol 2022 · 81:305–312

Hypokomplementemiás ulticaria vasculitis

- Incidencia: 1-2/1millió/év
- Átlagos életkor: 50. év
- Nő:férfi= 8:1
- Lefolyás
  - 4-8 hét alatt szűnik egy epizód
  - 30-40%-ban egy év alatt megszűnik
  - 2-10%-ban válik krónikussá
- Anti-C1q antitest pozitivitás 50-100%
- >24h át megmaradó ulticariák
- Leukocitoklasztikuss vaszkulitisz
- GN
  - Membranózus
  - Mesangioproliferatív



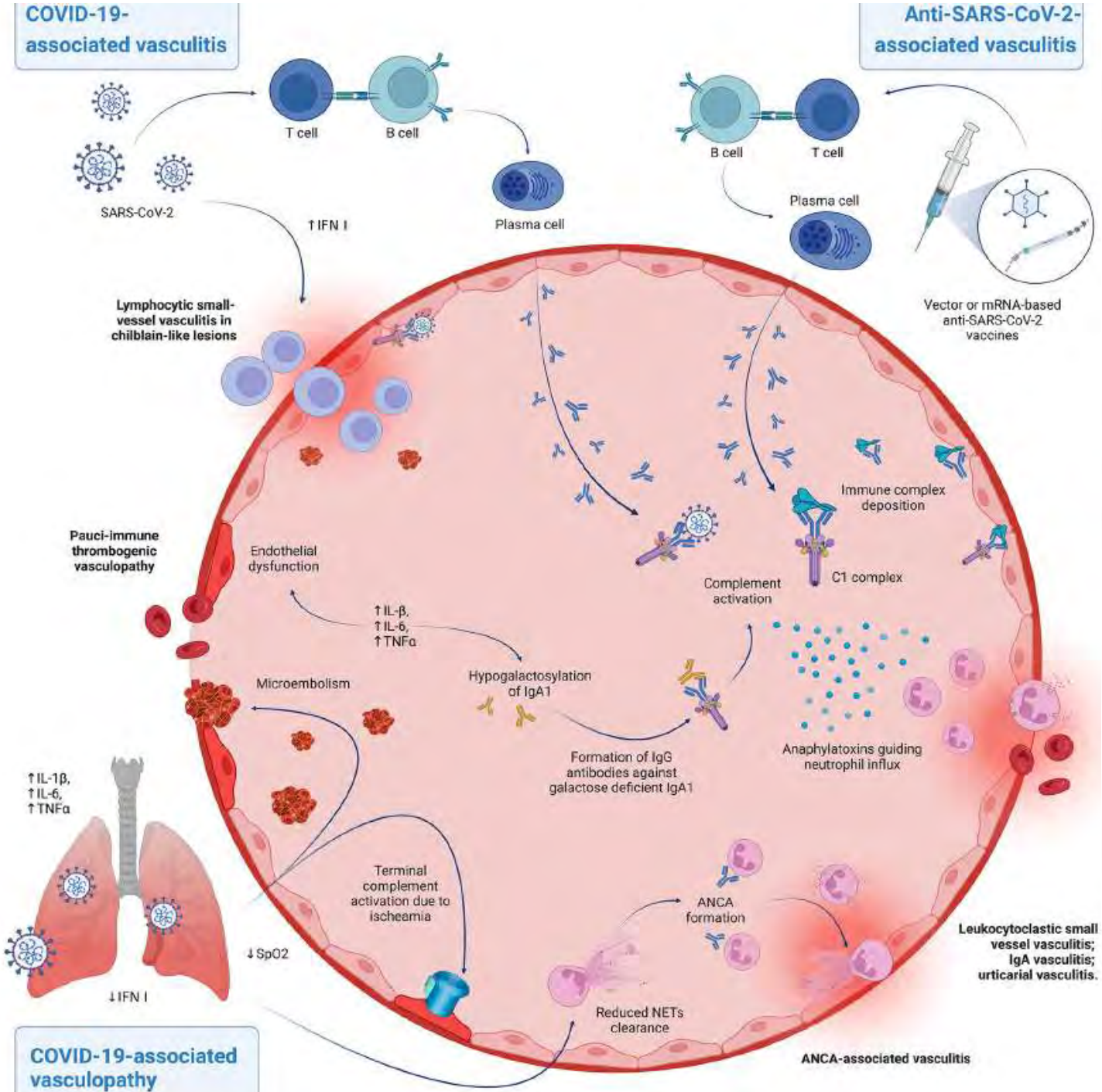
Hypokomplementemiás ulticaria vasculitis



Int J of Women's Dermatology 7 (2021) 290–297

Table 1 Most common causes of drug-induced vasculitis

Drug-induced vasculitis as per vessel size	Associated drugs
Large vessel vasculitis (LVV)/aortitis	- Granulocyte colony-stimulating factor (G-CSF) - Immune check point inhibitors
Medium vessel vasculitis (MVV)	- Minocycline - Empagliflozin - Gemcitabine -Metolazone
Small vessel vasculitis • ANCA-associated vasculitis (AAV)	- Hydralazine - Cocaine - Propylthiouracil -Methimazole - TNF-inhibitors - Sulfasalazine - Allopurinol - D-penicillamine - Isoniazid - Phenytoin - immune check point inhibitors
• IgA vasculitis (IgAV)	- Antibiotics - NSAIDs - TNF-inhibitors - Statin - Warfarin
Single organ vasculitis • Leukocytoclastic vasculitis	- NSAIDs - Antibiotics - TNF-inhibitors - Allopurinol - Valproic acid - Sulfonamides - Warfarine - Amiodarone - Anti tuberculosis drugs - Immune checkpoint inhibitors
• Cerebral vasculitis	- Amphetamines - Cocaine - Immune checkpoint inhibitors
• Peripheral vasculitic neuropathy	- Minocycline - Leflunomide - TNF-inhibitors - Amphetamines - Immune checkpoint inhibitors - Phenytoin



	Clinical features
Cutaneous small-vessel vasculitis*	Palpable purpura, petechiae and/or hemorrhagic macules or (rarely) blisters. Occasionally, ulcerations can be observed. Lower extremities are commonly affected. Extracutaneous involvement is uncommon and usually mild.
Skin-limited IgA vasculitis	Erythematous macules or papules evolving into palpable purpura predominantly on the lower limbs, thighs, and buttocks. Hemorrhagic bullae and targetoid lesions can also be observed.
Urticarial vasculitis**	Erythematous, oedematous wheal-like lesions persisting more than 24 h, associated with non-blanchable purpura and resolving with hyperpigmented *sequelae*, most commonly on the trunk and proximal extremities. Burning, rather than itching is typically reported.
Lymphocytic vasculitis	Maculo-papular erythemato-violaceous lesions with purpuric aspects located on lower and upper limbs. Chilblain-like appearance (i.e., "COVID toes").
Pauci-immune thromboembolic vasculopathy***	Necrotic lesions, retiform purpura, finger or toe cyanosis, gangrene, blisters or livedoid rash.

Frontiers in Medicine 2022 10.3389

Table 1 Classification and causes of leukocytoclastic vasculitis

CHCC 2012 category	Causes and/or associated diseases	CHCC 2012 definition	Histopathology
ANCA-associated vasculitis	Granulomatosis with polyangiitis (GPA) Microscopic polyangiitis (MPA) Eosinophilic granulomatosis with polyangiitis (EGPA)	Necrotizing vasculitis with few or no immune deposits, predominantly affecting small vessels (i.e., capillaries, venules, arterioles, and small arteries); associated with ANCAs	Vasculitis of small-to-medium vessels in the skin, often with leukocytoclasia with or without granulomatous inflammation
Immune complex vasculitis		Vasculitis with moderate-to-marked vessel wall deposits of immunoglobulin and/or complement components, predominantly affecting small vessels (i.e., capillaries, venules, arterioles, and small arteries)	LCV of small vessels (mostly postcapillary venules, occasionally small veins or arterioles)
	Cryoglobulinemic Vasculitis (CV)	Vasculitis with cryoglobulin immune deposits affecting small vessels (predominantly capillaries, venules, or arterioles); associated with serum cryoglobulins	LCV of small vessels (postcapillary venules, small veins, or arterioles); associated with serum cryoglobulins (usually type II and type III)
	IgA-Vasculitis (Henoch–Schonlein purpura, HSP)	Vasculitis with IgA 1-dominant immune deposits, affecting small vessels (predominantly capillaries, venules, or arterioles)	Leukocytoclastic IgA 1-dominant vasculitis of mostly postcapillary venules and also veins or arterioles in the skin, with vascular IgA deposits
	Hypocomplementemic Urticarial Vasculitis (anti-C1q vasculitis, HUV)	Vasculitis accompanied by urticaria and hypocomplementemia, affecting small vessels (i.e., capillaries, venules, or arterioles) and associated with anti-C1q antibodies; common forms include glomerulonephritis, arthritis, obstructive pulmonary disease, and ocular inflammation	Cutaneous LCV of mostly postcapillary venules with vascular deposits of immunoglobulins, and manifesting with lasting urticarial lesions; anti C1q antibodies may be present
	IgM/IgG immune complex vasculitis*	Vasculitis with IgM and/or IgG-dominant immune deposits, affecting small vessels (predominantly capillaries, venules, or arterioles)	Leukocytoclastic IgM and/or IgG-dominant vasculitis of mostly postcapillary venules and also veins or arterioles in the skin, with vascular deposits
Vasculitis associated with systemic diseases	Rheumatoid arthritis Systemic lupus erythematosus Sjögren syndrome Sarcoidosis	Vasculitis that is associated with and maybe secondary to (caused by) a systemic disease (e.g., rheumatoid vasculitis, SLE, sarcoid vasculitis, etc.); the name (diagnosis) should have a prefix term specifying the systemic disease (e.g., rheumatoid vasculitis, lupus vasculitis, etc.)	Cutaneous LCV as a component of systemic vasculitis; the type of cutaneous vasculitis (small vessel or medium vessel vasculitis) varies depending on the underlying systemic disease
Vasculitis associated with probable etiology	Drugs Infection Sepsis Neoplasms	Vasculitis that is associated with a probable specific etiology, e.g., drug, infection, sepsis, neoplasm, etc	Cutaneous LCV as a component of systemic vasculitis that is associated with a probable specific etiology, e.g., drug, sepsis, etc

Leukocytoclastic vasculitis
palpable purpura

inflammatory infiltrate is composed
of neutrophils with fibrinoid necrosis and disintegration of nuclei into fragments ("leukocytoclasia").

Internal and Emergency Medicine (2021) 16:831–841

Összefoglalás

- Olvassunk – mindig van új adat
- Fontos új adatok vannak
  - Patogenezis
  - Klasszifikáció
  - Rizikóbecslés
  - Kezelés
- Szteroid spórolás előtérbekerülése
- RCT vs RWE

Köszönöm a figyelmet!