2. DACH ANCA VASKULITIS FORUM 2024

22. & 23. NOVEMBER 2024 | MÜNCHEN

CSL Vifor

Wieso sind die Langzeitdaten der frühen EUVAS-Studien wichtig?

Univ.Doz.Dr. Irmgard Neumann

Wien

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ANCA assoziierte Vaskulitis
Wieso sind die Langzeitdaten der frühen EUVAS Studien wichtig?

Irmgard Neumann

Wien und Immunologie Zentrum Zürich

European Vasculitis Society EUVAS

Disclosures

Consulting/ Advisory boards: Vifor, GSK, Otsuka, Glaxo, Novartis

Wieso sind die Langzeitdaten der frühen EUVAS Studien wichtig?

- Langzeit follow-up Studien
- Was haben wir daraus gelernt ?
- Welche Implikationen gibt es ?

Prognose der AAV
History

1950s	Überleben unbehandelt 5 Monate 1-Jahres Mortalität 82%	Walton EW Br Med J 1958
1967	Steroide → 1-Jahres Überleben 34%	Hollander, Ann Intern Med 1967
1970s	Kortikosteroide + CYC → Remission bis zu 90%	Fauci AS, Ann Intern Med 1983
1982-2010	1-J Überleben 80 - 88% 5-J Überleben 70 - 78%	SlotMC, Kidney Int 2003

Prognose der AAV
History

1950s Überleben unbehandelt 5 Monate
1-Jahres Mortalität 82%

Walton EW Br Med J 1958

1967

, Ann Intern Med 1967

1970s

, Ann Intern Med 1983

1982-2

lotMC, Kidney Int 2003

Cyclophosphamide - Treatment associated morbidity

- **Infectious complications 46 %**
- **Haemorrhagic Cystitis 43 %**
- **Infertility (after 1 yr) 57 %**

- **Malignancy compared to NIH overall 2.4-fold**
  bladder-cancer 33-fold
  lymphoma 14-fold

Hoffman et al. Ann Int Med 92

Prognose der AAV
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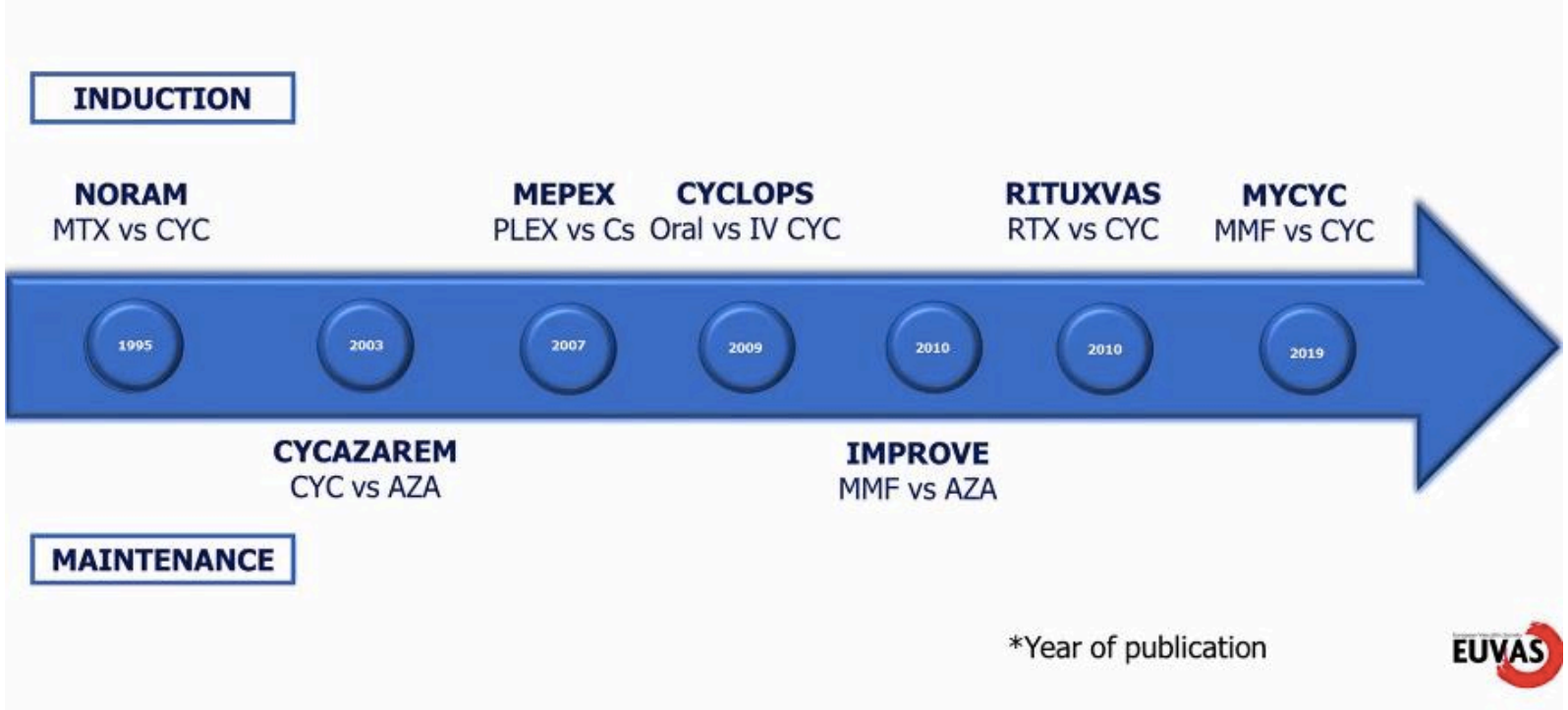
Ersten 4 EUVAS Trials	5-Jahres follow-up Remission nach 6 Monaten 80%-90% 2.6-fach erhöhtes Risiko für Mortalität im Vergleich zur Allgemeinbevölkerung	Flossmann O, Ann Rheum Dis 2011

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

continuation of the previous EUVAS
5-year follow-up



RCTs (1995-2012): **n=848**
MEPEX, NORAM, CYCAZAREM, CYCLOPS, IMPROVE, RITUXVAS and MYCYC,

Sánchez Álamo, EUVAS, NDT 2023

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

continuation of the previous EUVAS
5-year follow-up

Characteristics	NORAM ($N = 95$)	CYCAZAREM ($N = 155$)	CYCLOPS ($N = 148$)	MEPEX ($N = 137$)	RITUXVAS ($N = 44$)	IMPROVE ($N = 156$)	MYCYC ($N = 140$)
Disease stage	Early systemic	Mild–moderate	Mild–moderate	Severe	Mild–moderate	Mild–moderate	Mild–moderate
Age (years), median (IQR)	53 (18–78)	58 (20–77)	57 (18–86)	66 (27–81)	63 (20–84)	55 (19–74)	56 (9–87)
Induction	MTX versus CYC po	CYC po	CYC IV versus po	MEP versus PE	RTX + CYC versus CYC	CYC po	CYC po versus MMF
Maintenance	MTX or CYC po	CYC po versus AZA	AZA	AZA	AZA	AZA versus MMF	AZA
Duration (months)	18	18	18	12	24	48	18
Inclusion period	1995–2000	1995–1997	1998–2001	1995–2001	2006–2007	2002–2005	2008–2011

AZA: azathioprine; CYC: cyclophosphamide; iv: intravenous; MEP: pulse methylprednisolone; MMF: mycophenolate mofetil; MTX: methotrexate; OCS: oral corticosteroids; po: oral.

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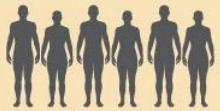
Long-term outcomes and prognostic factors for survival of patients with ANCA-associated vasculitis

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%

848 neu diagnostizierte AAV
gut charakterisiert
Alter 58 ± 14 Jahre

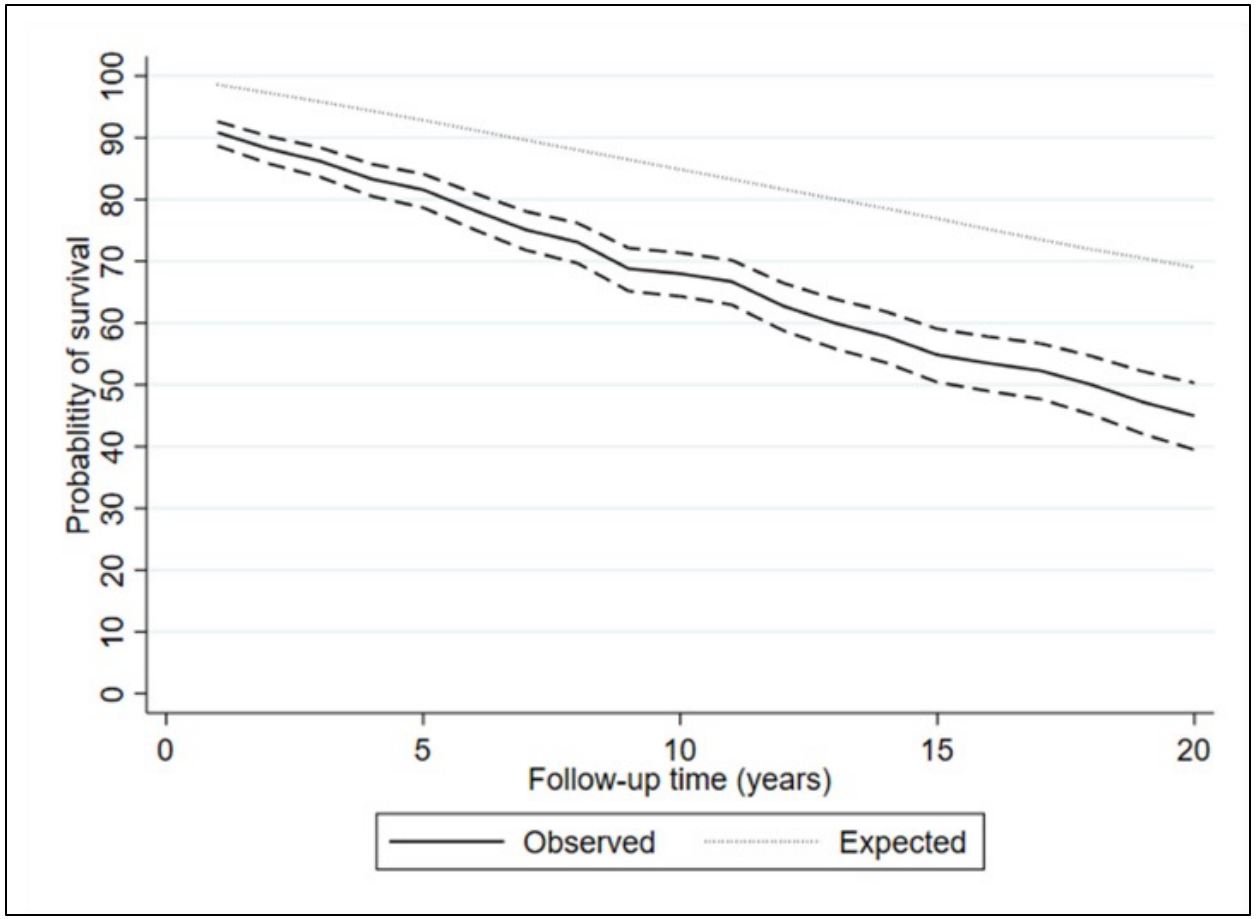
Median long-term follow-up → 8 years (IQR: 2.9–13.6)

Survival
Causes of death
Prognostic factors

Sánchez Álamo, EUVAS, NDT 2023

Patienten Überleben

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



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Patienten Überleben

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

Survival	
at 1 yr	88.2%
5 yrs	78.2%
10 yrs	66.7%
15 yrs	53.5%

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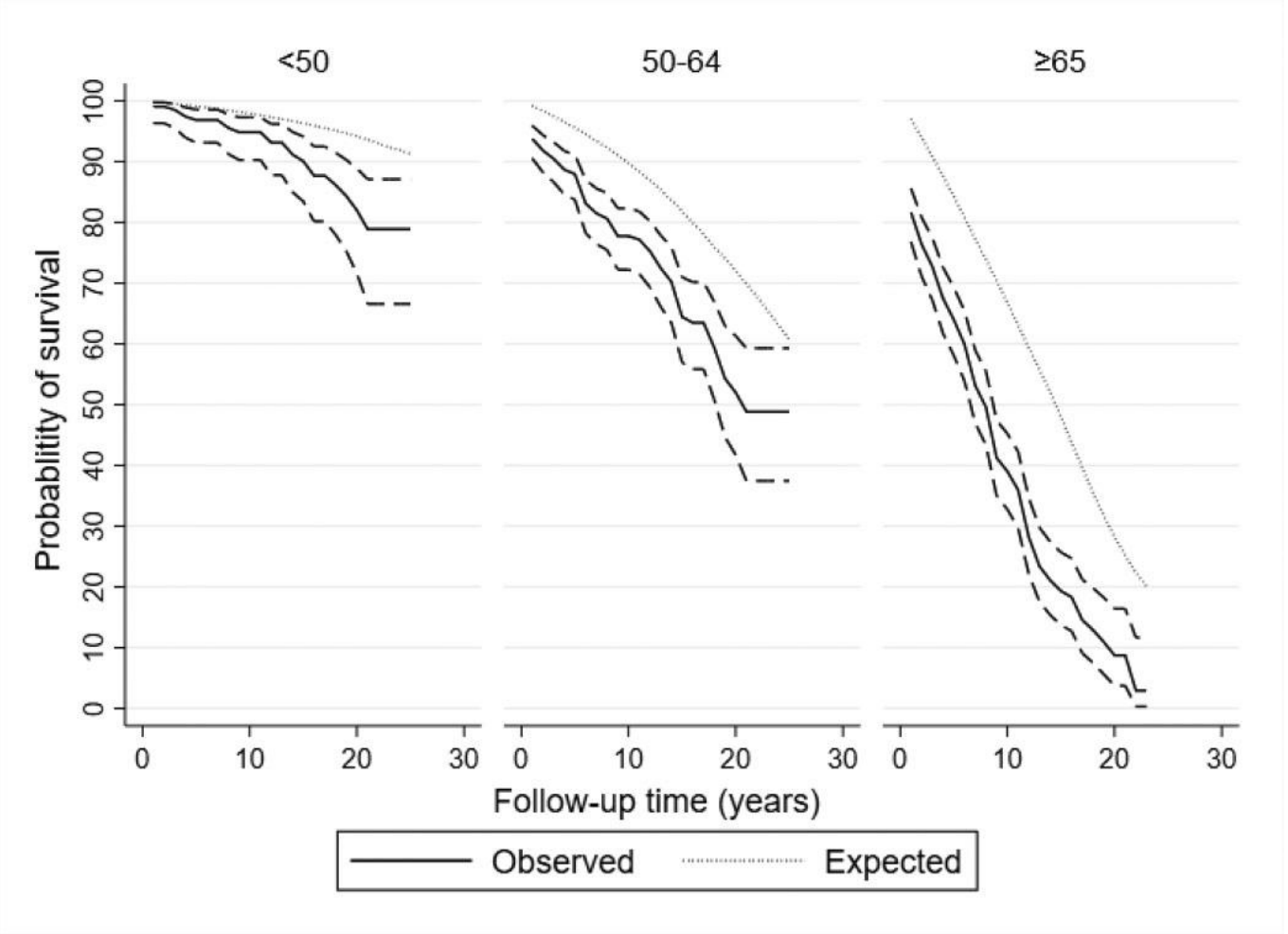
Patienten Überleben nach Alter



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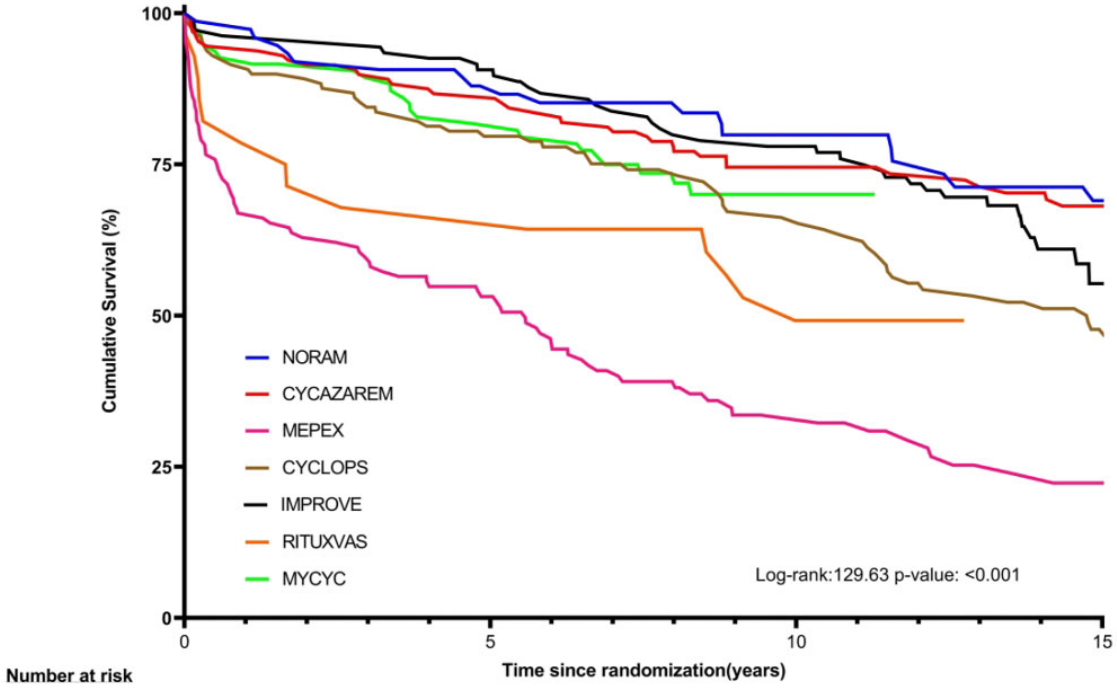
Patienten Überleben nach RCTs



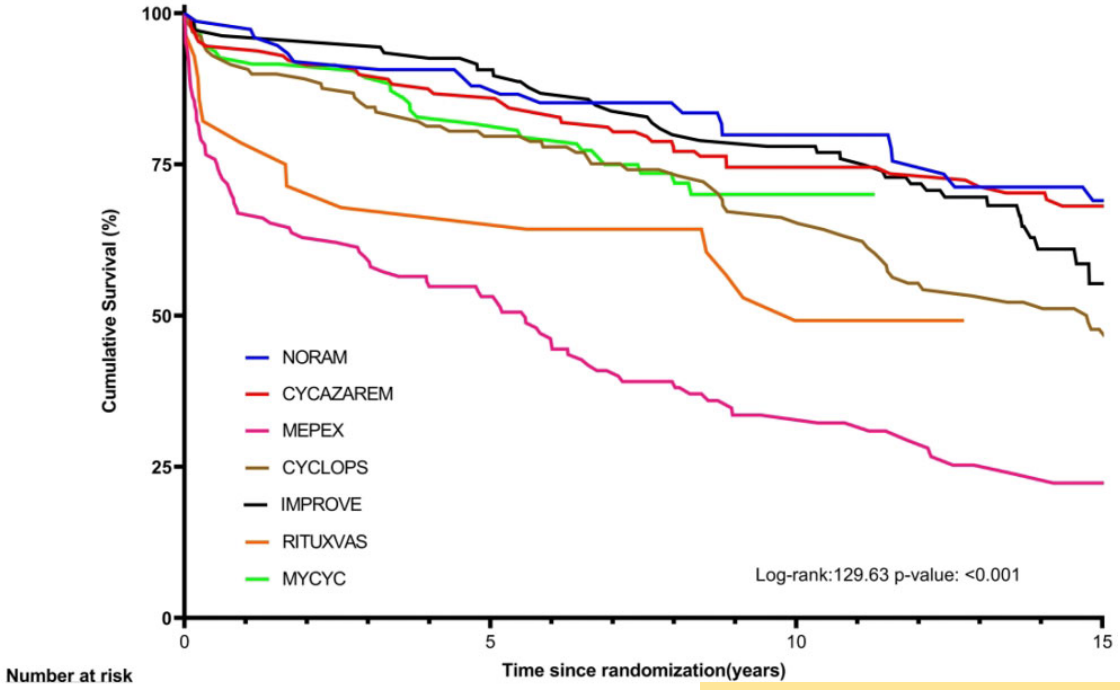
CYCLOPS
RITUXIVAS

MEPEX

	Survival at 10 years(%)	95 % CI
MEPEX	33.7	24.9 - 42.7
RITUXVAS	53.7	33.7 - 70
CYCLOPS	66.1	56.7 - 73.9
MYCYC	73.1	62.6- 81.1
CYCAZAREM	75.9	67.6 - 82.4
IMPROVE	81.3	73 - 87.2
NORAM	81.4	70.4 - 88.6

Sánchez Álamo, EUVAS, NDT 2023

Patienten Überleben nach RCTs



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NORAM	81.4	70.4 - 88.6

638 patients received CYC
63% p.o. 37% i.v.
→ better survival compared i.v. with oral
(log rank 11.0, P = 0.001)

Sánchez Álamo, EUVAS, NDT 2023

Woran sind die Patienten verstorben ??

305 deaths

Main cause of death

- Infection 26%
- CVD 14%
- Malignancy 13%

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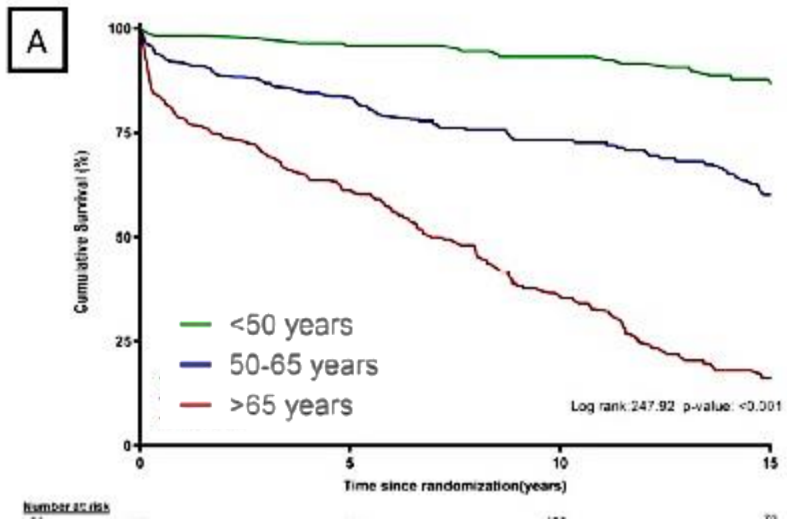
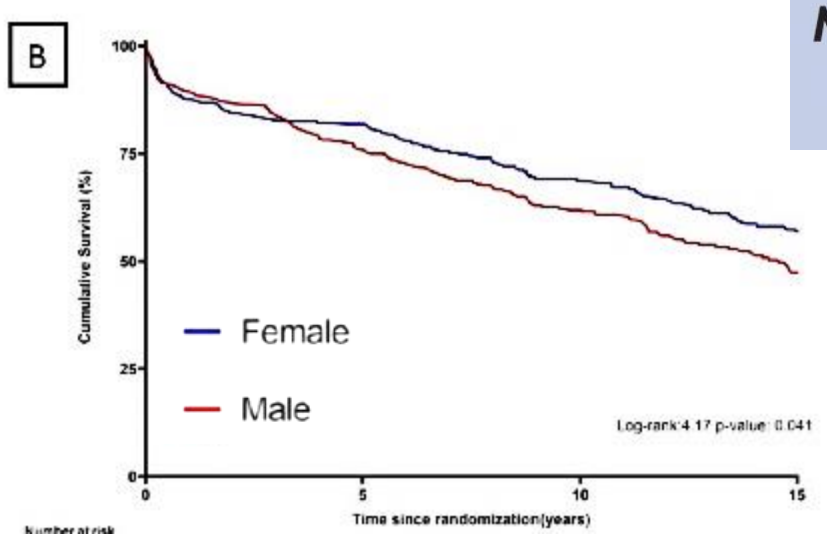
Negative prognostic factors for patient survival

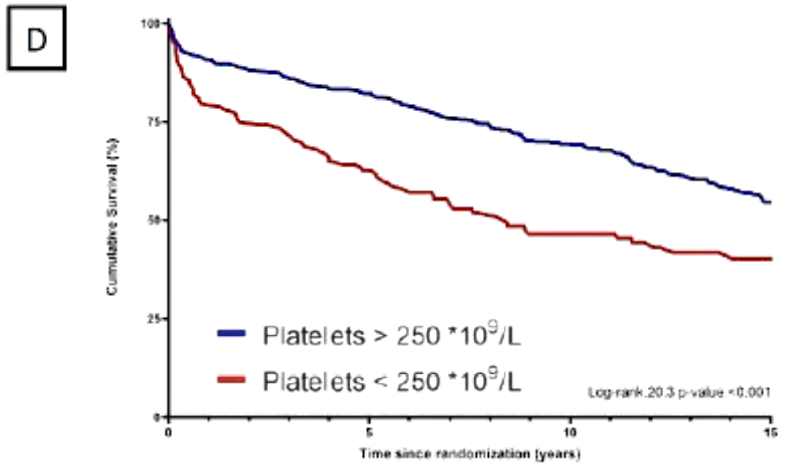
Kaplan Meier curves

Advanced age

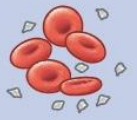


Male sex ♂





Low platelet count



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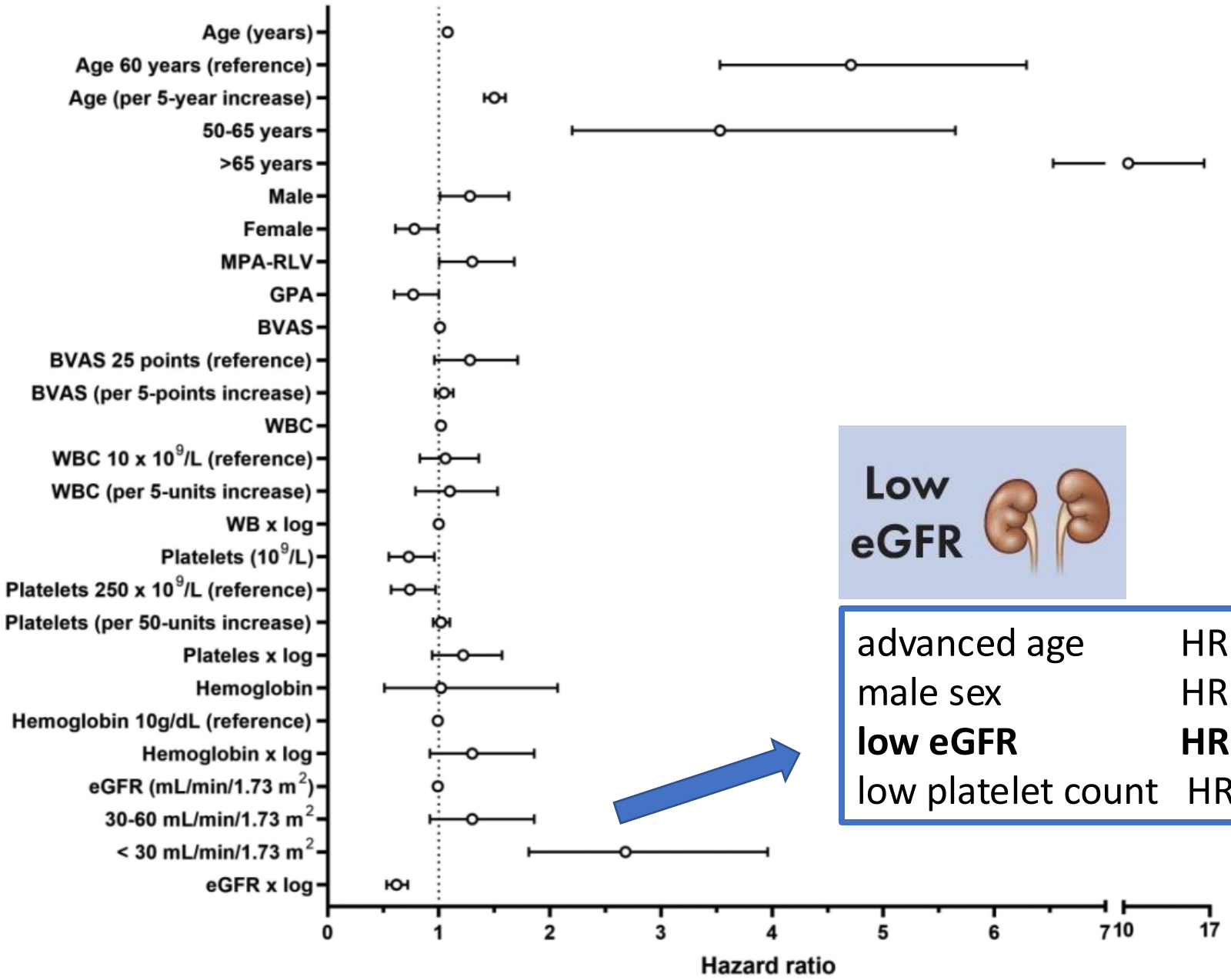
Negative prognostic factors for patient survival

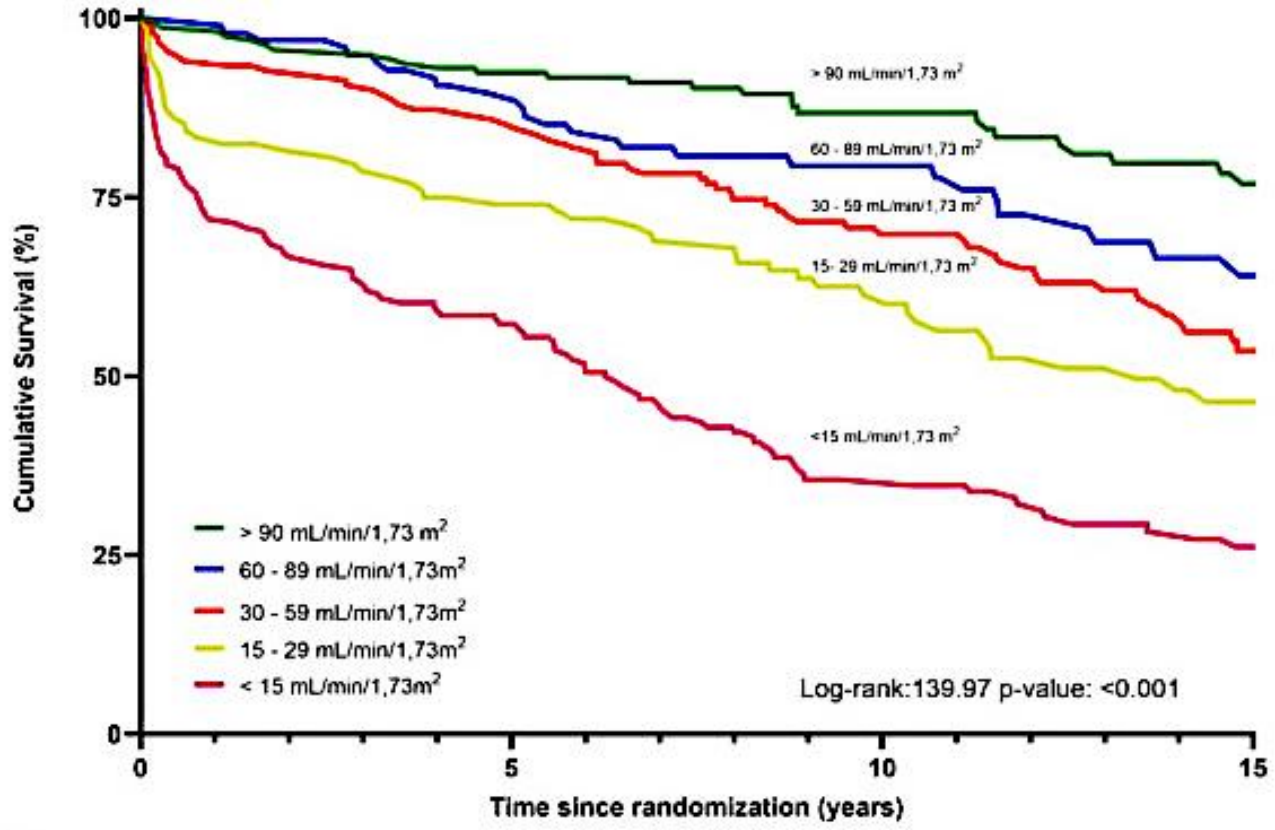


20.6% reached ESKD

advanced age	HR 9.9	(95% CI 6.2–15.8), P < .001
male sex	HR 1.3	(95% CI 1.1–1.7), P = .02
low eGFR	**HR 2.63**	(95% CI 1.77–3.91), P = .001
low platelet count	HR 1.7	(95% CI 1.2–2.4), P = .004

Sánchez Álamo, EUVAS, NDT 2023

Negative prognostic factors for patient survival



The highest number of deaths was found in the group of patients with lowest GFR

Sánchez Álamo, EUVAS, NDT 2023

Long-term outcome of kidney function in patients with ANCA-associated vasculitis

848 AAV Patienten

Renale Beteiligung 644 (76%)

follow-up 7.96 Jahre

Sánchez Álamo, EUVAS, NDT 2024

Long-term outcome of kidney function in patients with ANCA-associated vasculitis

848 AAV Patienten

Renale Beteiligung 644 (76%)

follow-up 7.96 Jahre

GFR at baseline

199 eGFR < 15 mL/min/ 1.73 m2

144 eGFR 15–30 mL/min/1.73 m2

183 eGFR 30–60 mL/min/1.73 m2

107 dialysis at presentation

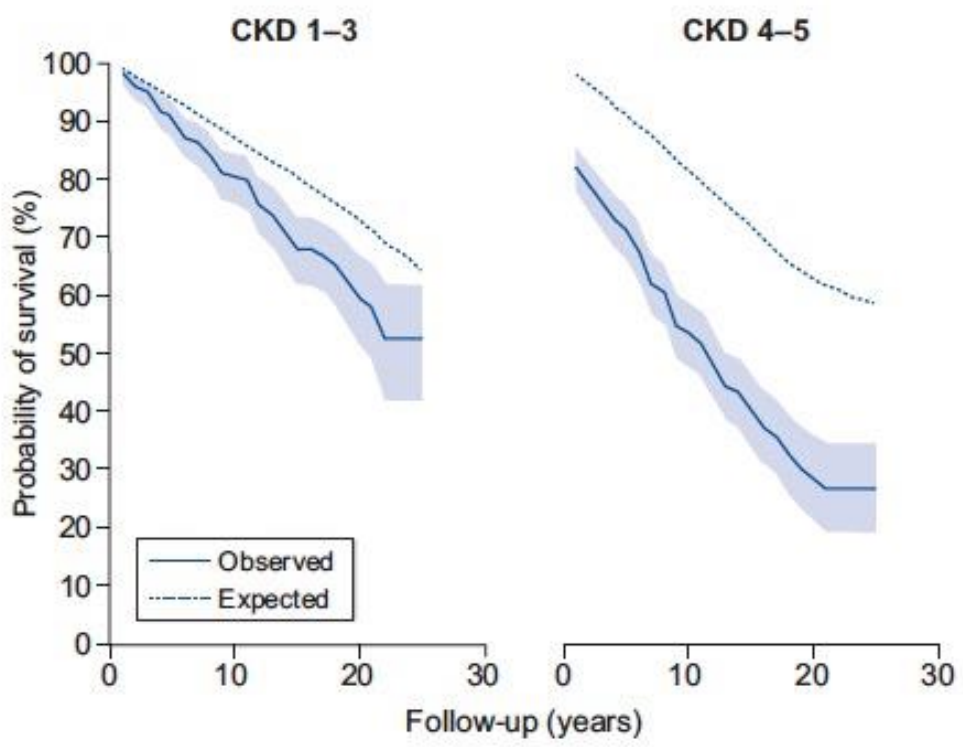
Gibt es Risikofaktoren für ESKD ?

Haben histologische Veränderungen eine prognostische Bedeutung ?

Sánchez Álamo, EUVAS, NDT 2024

Long-term outcome of kidney function in patients with ANCA-associated vasculitis

Patienten Überleben nach CKD-Stadium



Baseline GFR is important !!

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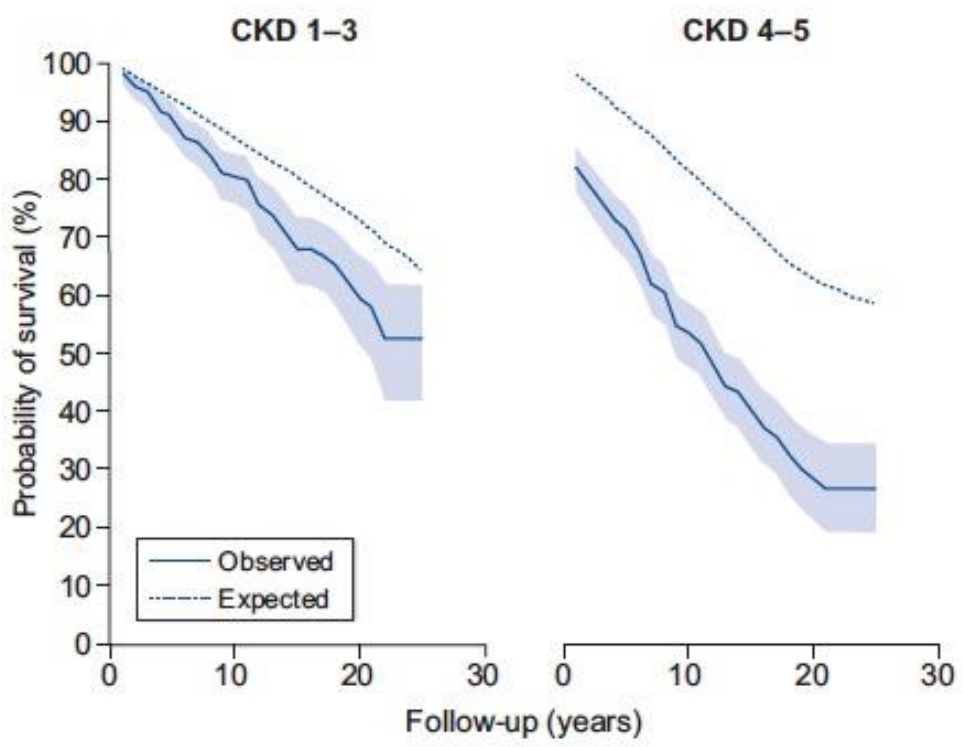
Long-term outcome of kidney function in patients with ANCA-associated vasculitis

Patienten Überleben nach CKD-Stadium



- ESKD nach 5 Jahren 17%
  nach 10 Jahren 22%
  nach 15 Jahren 27%

- 34% Recovery von initialer Dialyse

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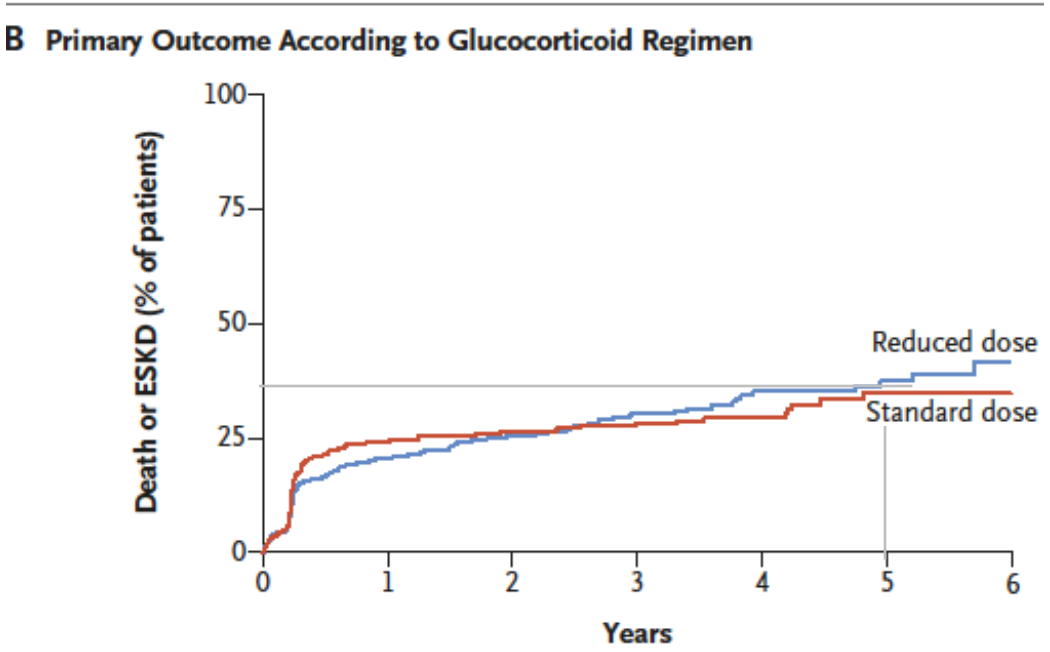
ANCA Vaskulitis- Renale Prognose

eGFR <50 mL/min → >30% ESKD

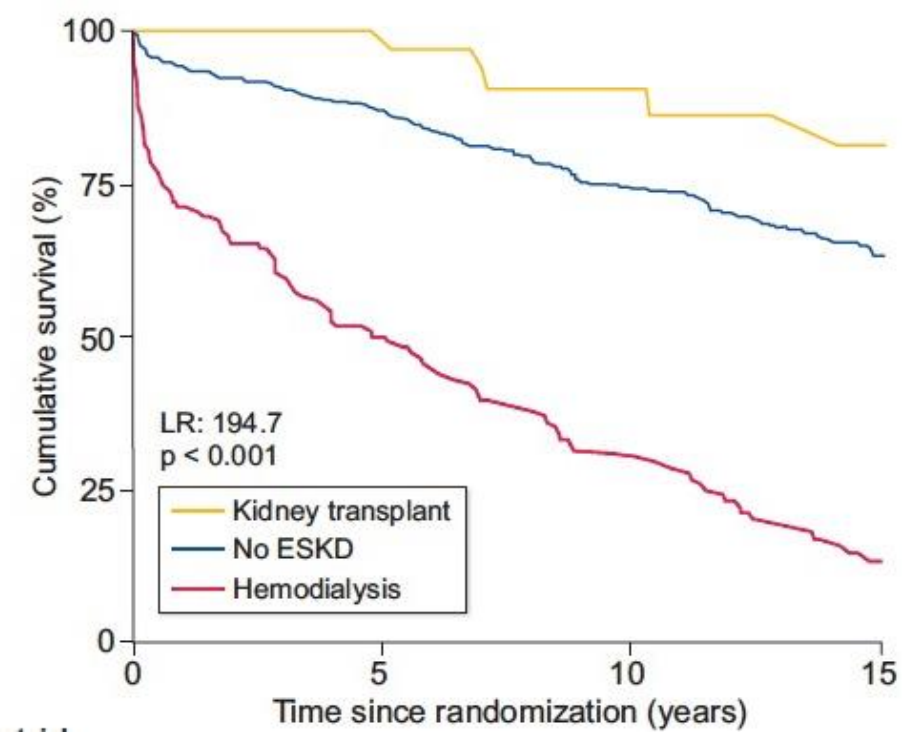


Walsh M., NEJM 2020

Patienten Überleben - ESKD

35	Transplantiert
140	Dialyse

Patienten Überleben



- **Erhöhte Mortalität mit ESKD** (HR) 2.8; *P* < .001
  Prognose besser mit GPA als mit MPA (HR 5.4; *P* = .02)

- **Ursachen für die Mortalität**
  Infektionen (29.7%)
  CVD (16.9%)
  Maligne Erkrankungen (5.9%).

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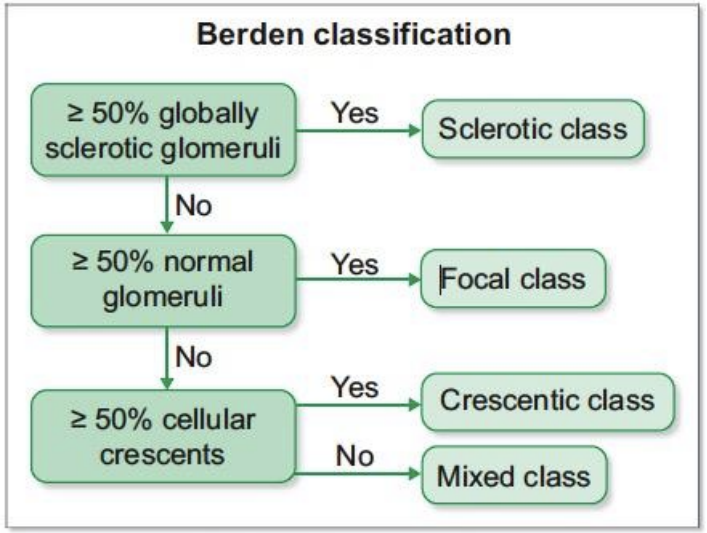
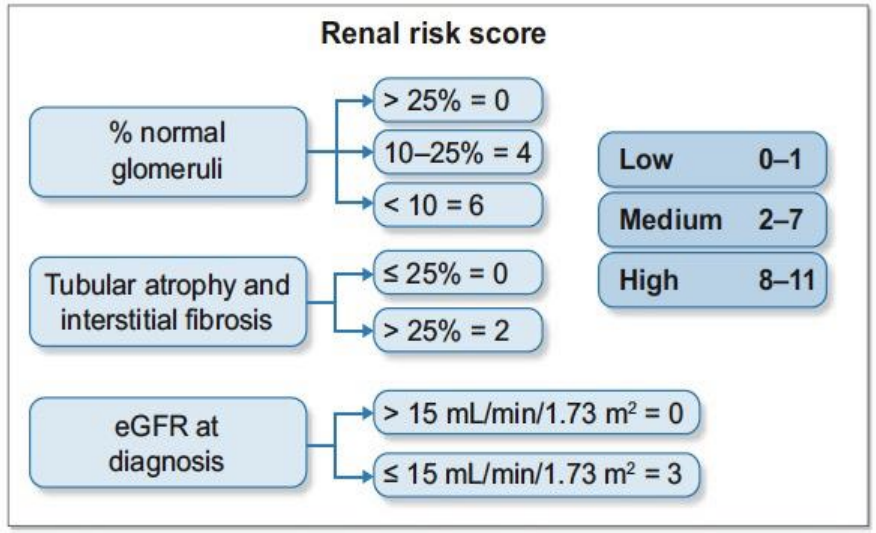
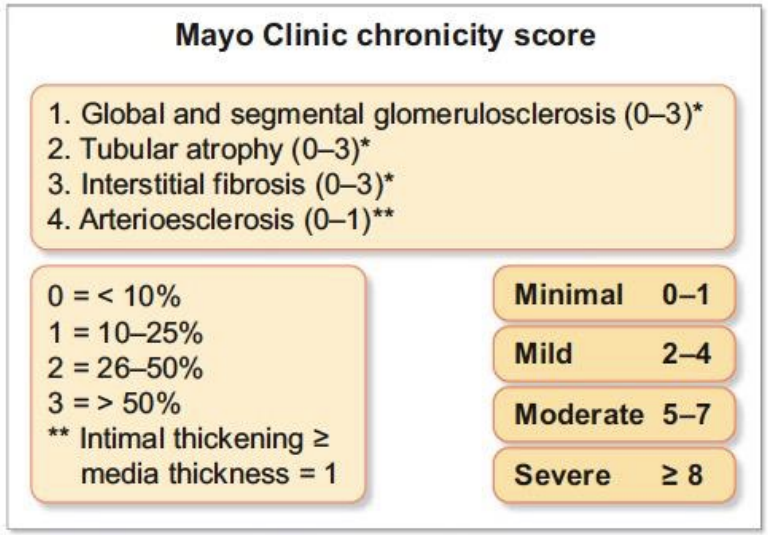
Was ist der Stellenwert der Nierenhistologie bezüglich Outcome ?

Different prediction models for kidney survival







Fokus
Glomeruläre Veränderungen

Berden A , JASN 2010

Fokus
Glomerulär und tubulo-interstitiell
+ eGFR !!

Brix SR, KI 2018

Fokus
Chronische Veränderungen

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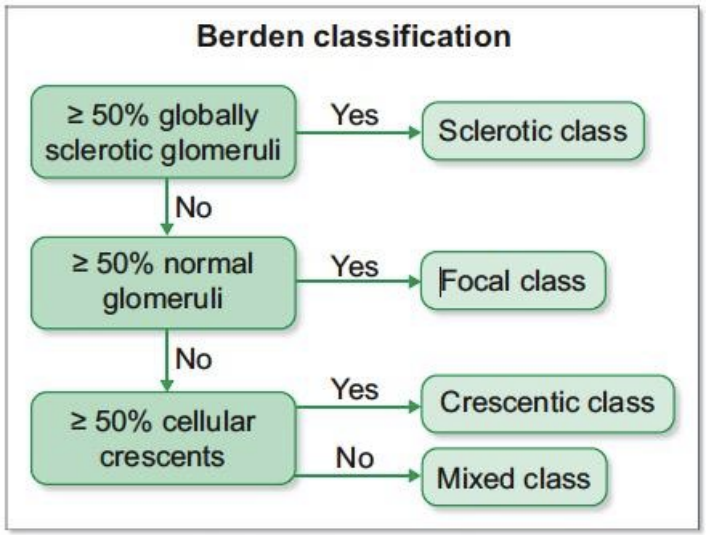
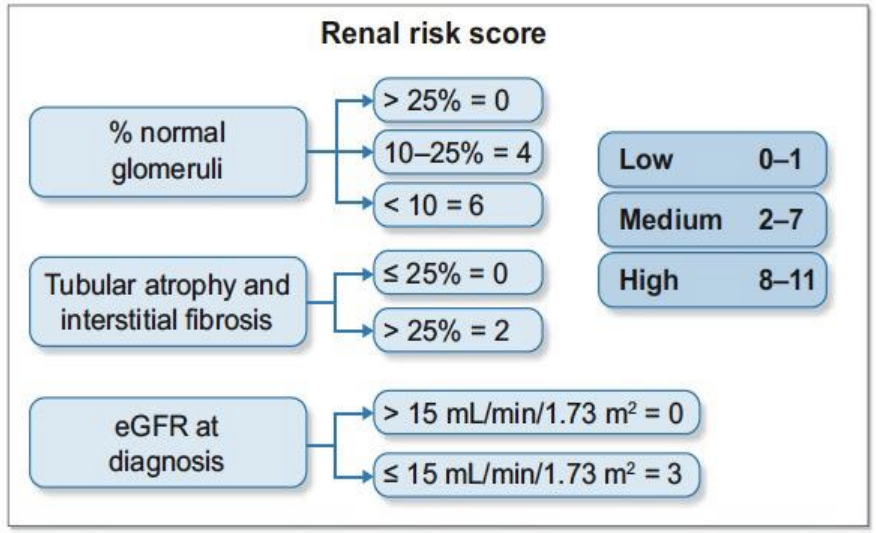
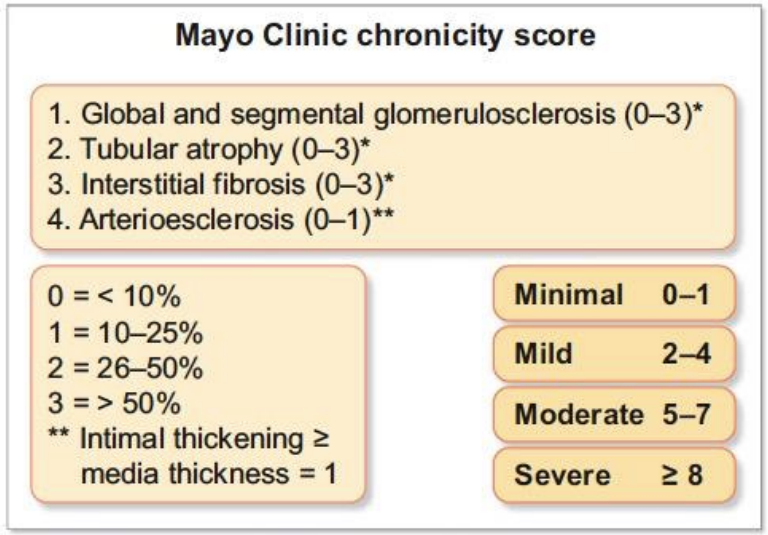
Casal Moura M, NDT 22

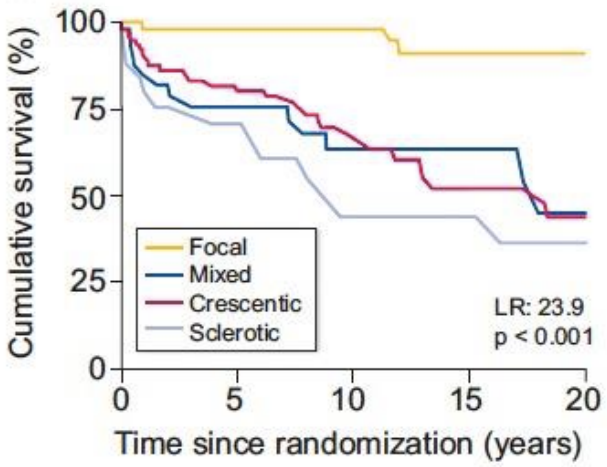
Was ist der Stellenwert der Nierenhistologie bezüglich Outcome ?

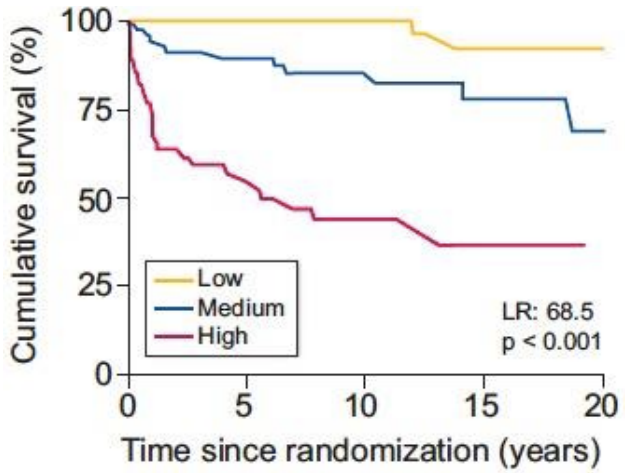




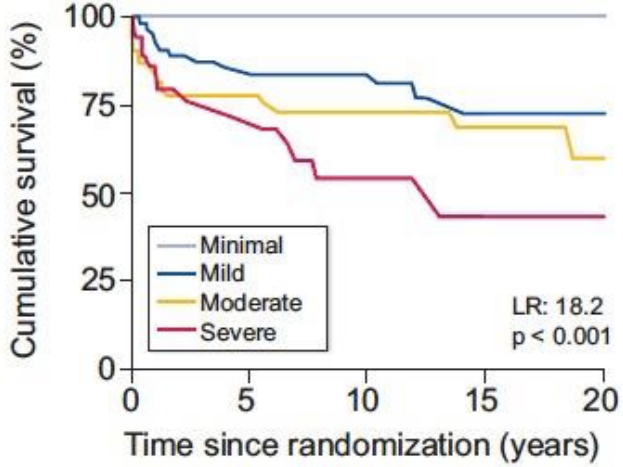


(A) Kidney endpoint according to the histological classifications





(C)



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Prognostic factors for kidney endpoints

Variables	Univariate			Multivariate		
	HR	95% CI	P-value	HR	95% CI	P-value
Kidney endpoint—all the patients						
MPA	2.02	1.44–2.86	<.001	1.14	0.80–1.64	.5
Male	1.34	0.94–1.91	.1	1.28	0.89–1.84	.2
eGFR (mL/min/1.73 m^2)	0.97	0.96–0.97	<.001	**0.97**	0.93–0.99	**<.001**
Age >65 years old	2.49	1.76–3.53	<.001	**1.68**	1.18–2.41	**.005**
Hemoglobin (g/dL)	0.75	0.68–0.83	<.001	**0.88**	0.78–0.98	**.02**

Sánchez Álamo, EUVAS, NDT 2024

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Variables	Univariate			Multivariate		
	HR	95% CI	P-value	HR	95% CI	P-value
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Male	1.34	0.94–1.91	.1	1.28	0.89–1.84	.2
eGFR (mL/min/1.73 m^2)	0.97	0.96–0.97	<.001	**0.97**	0.93–0.99	**<.001**
Age >65 years old	2.49	1.76–3.53	<.001	**1.68**	1.18–2.41	**.005**
Hemoglobin (g/dL)	0.75	0.68–0.83	<.001	**0.88**	0.78–0.98	**.02**
Kidney endpoint—patients with kidney biopsy						
Normal glomeruli (%)	0.94	0.92–0.96	<.001	**0.96**	0.93–0.98	**.002**
Mild glomeruloesclerosis	1.55	0.68–3.53	.3	2.20	0.94–5.14	.07
Moderate glomeruloesclerosis	3.52	1.54–8.05	.003	1.93	0.84–4.42	.1
Severe glomeruloesclerosis	3.87	1.73–8.64	.001	**2.87**	1.26–6.57	**.01**
Age >65 years old	2.37	1.67–3.37	<.001	1.41	0.75–2.65	.3
eGFR (mL/min/1.73 m^2)	0.97	0.96–0.97	<.001	**0.96**	0.93–0.99	**.005**

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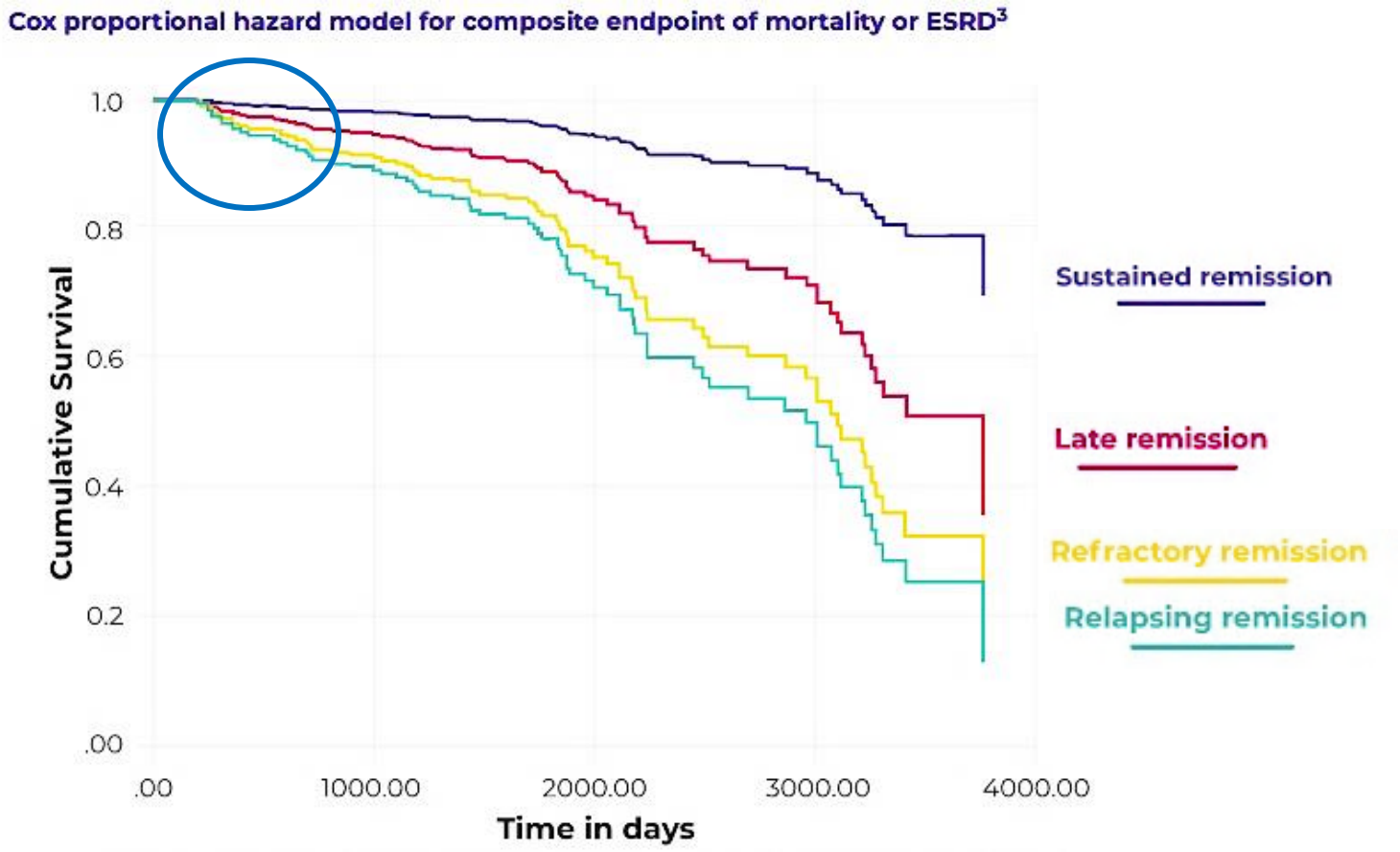
Time and Treatment

4 EUVAS trials



- **Frühe anhaltende Remission**
- **Umso besser die Nierenfunktion umso besser der outcome!**
- **Relapse → ESKD, Mortalität**

Gopaluni S, Arthritis Rheum 2019

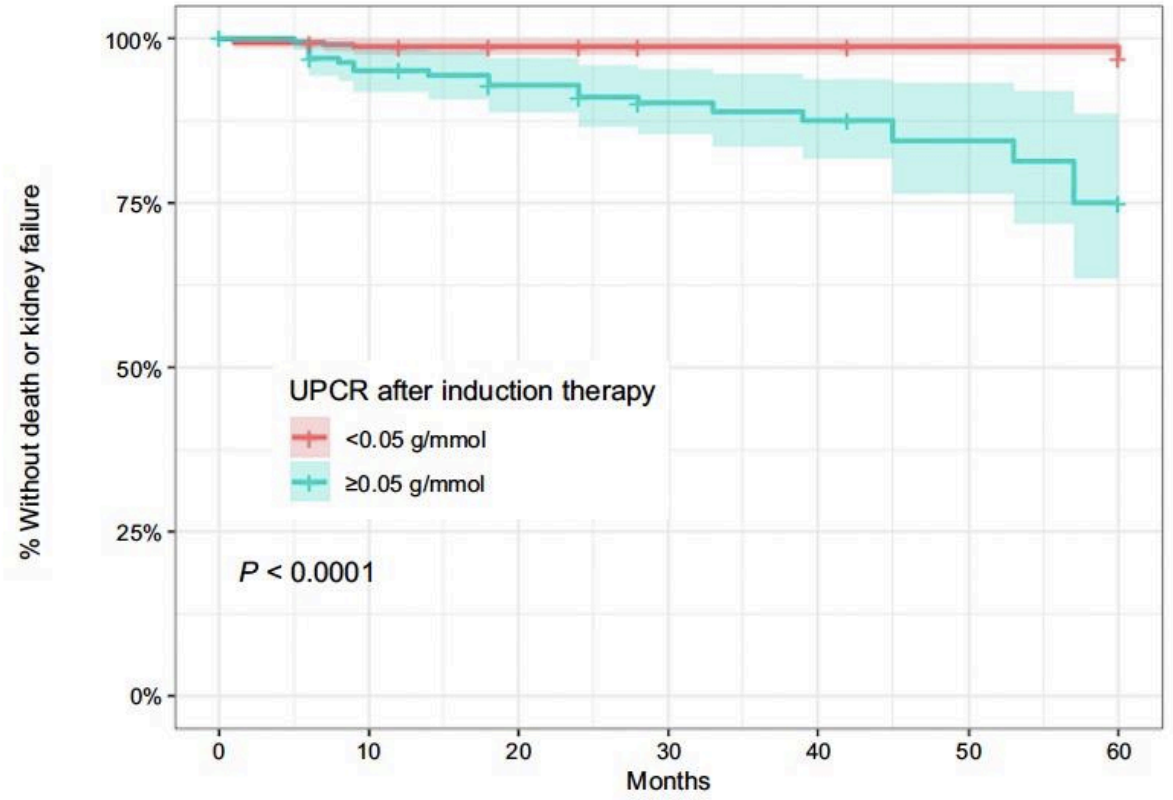
Renal Outcome - Proteinurie

5 European RCT on AAV
(MAINRITSAN, MAINRITSAN2, RITUXVAS, MYCYC, IMPROVE), n=571

Nach der Induktions-Therape
- Persistierende Hämaturie 30 %
- UPCR >0.05 g/mmol 34 %

Proteinurie

Composite outcome of **death or kidney failure**



Benichou N, KI 2023

5 European RCT on AAV
(MAINRITSAN, MAINRITSAN2, RITUXVAS, MYCYC, IMPROVE), n=571

Renaler Relapse

Nach der Induktions-Therape

- Persistierende Hämaturie 30 %
- UPCR >0.05 g/mmol 34 %

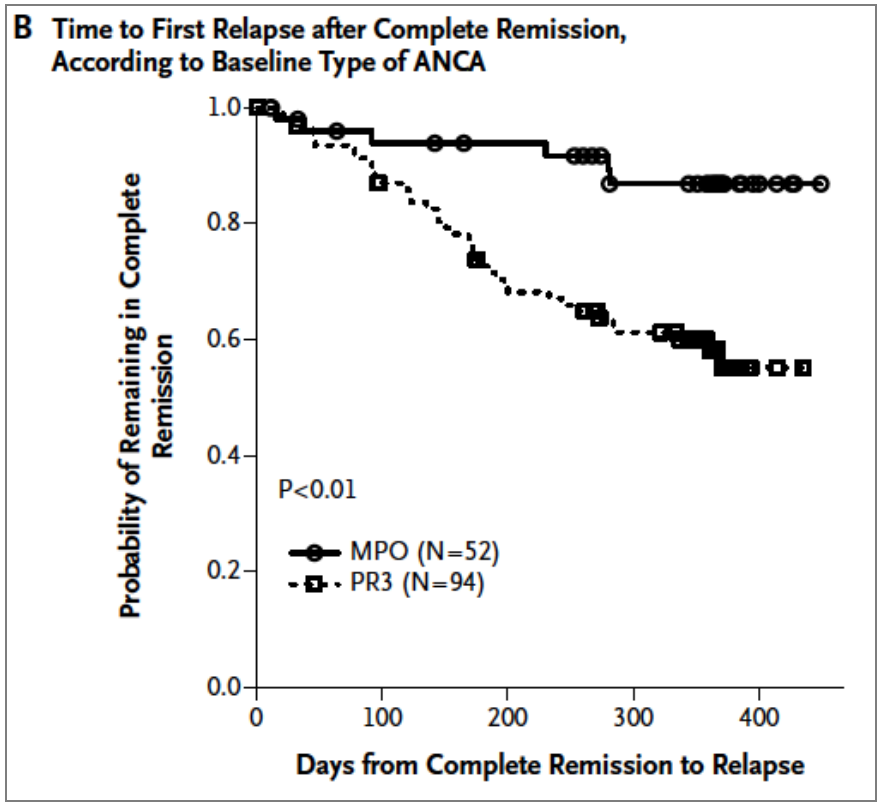
	Kidney relapse		
Variable	Adjusted sHR	95% CI	*P*
Older age (per 1-yr increase)	1.02	0.99–1.04	0.170
ANCA type (reference: MPO)			
Negative	1.27	0.32–4.95	0.730
PR3	1.40	0.68–2.88	0.360
Maintenance therapy: other (vs. RTX)	7.11	2.15–23.52	0.001
sCreat after induction therapy (per 10 µmol/l)	0.98	0.94–1.03	0.490
Hematuria after induction therapy	2.16	1.13–4.11	0.020
UPCR after induction therapy ≥0.05 g/mmol	2.22	1.16–4.24	0.016

Benichou N, KI 2023

Relapse Risko

RAVE study
CYC/AZA vs. Rituximab 18 Mo

Time to first relapse



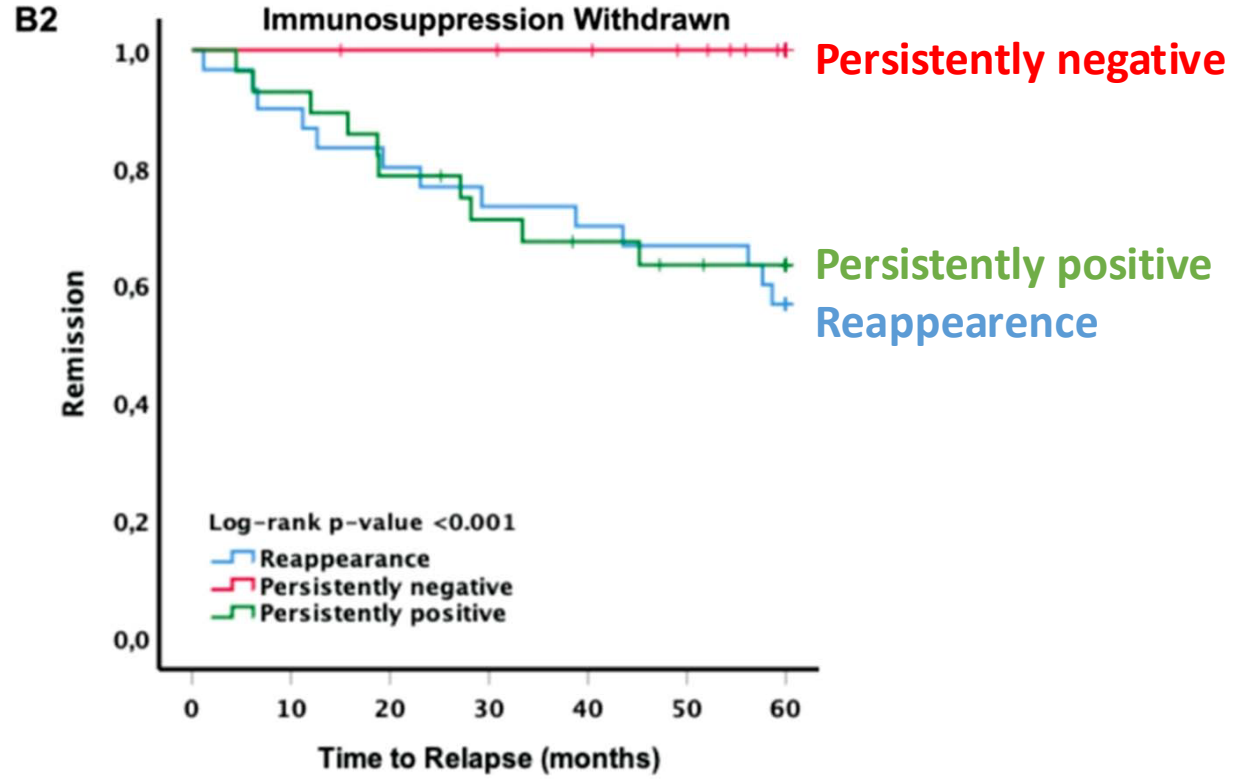
MPO

PR3

Specks NEJM 2013

Mayo Clinic cohort

Remission by MPO-ANCA titer



Persistently negative

Persistently positive

Reappearence

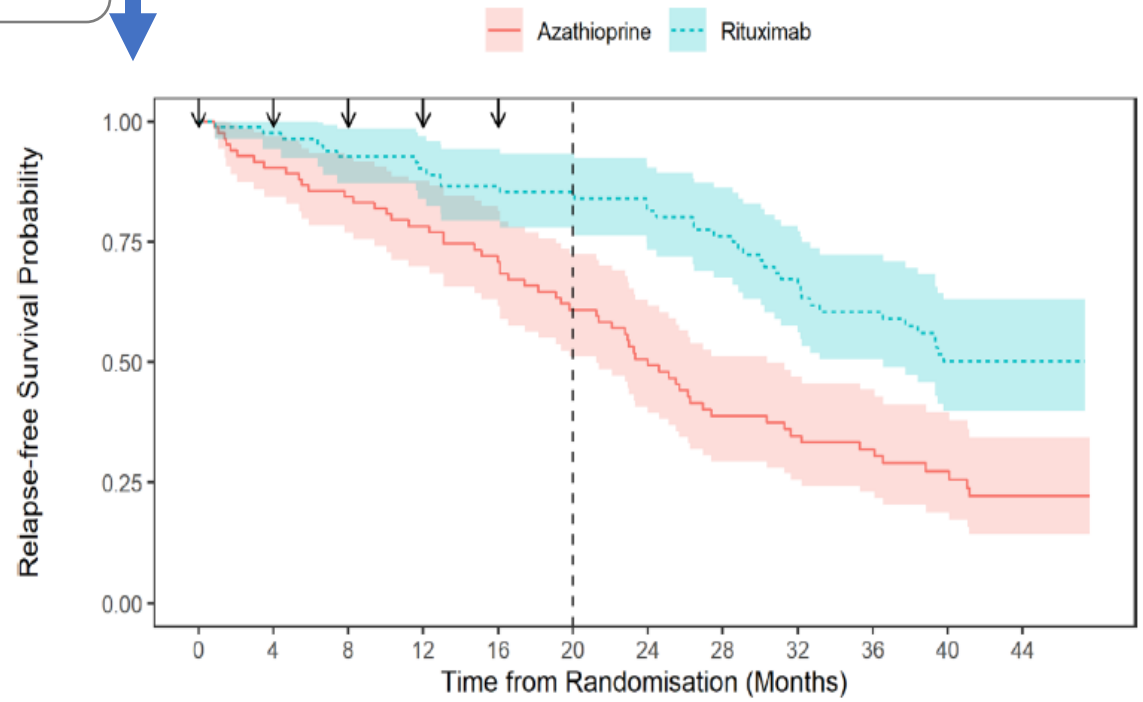
Moura MC , CJASN 2023

Rituximab – Relapse Prevention

RITAZAREM (1g RTX/4Mo vs AZA)

170 **relapsing** GPA/MPA

Induction RTX
Remission at months 4



Smith RM, Ann Rheum 2023

Long-term outcome in AAV
….Wo stehen wir?

- Höhere Mortalität im Vergleich zur Allgemeinbevölkerung (20% nach 10 Jahren)
  - alle Altersgruppen

- 22% ESKD nach 10 Jahren
  34% Recovery nach initialer Dialyse

- Gutes Transplant-und Patienten ÜL
  (86% nach10 J)

Sánchez Álamo, EUVAS, NDT 2023
Sánchez Álamo, EUVAS, NDT 2024

Prognose - Room for Improvement

Ursachen für Mortalität

- Infektion
- CVD, Maligne Erkrankungen

→

- Steroide reduzieren, Avacopan
- Immunsuppression
- Prävention
- Management - Co-Morbiditäten

Risikofaktoren

- Alter
- ↓ eGFR baseline
- Histo - Chron Veränderungen
  - % normal Glomeruli
- MPA

→

- Awareness!
- Frühe Diagnose
- Rascher Therapiebeginn

Klinische Parameter

- Hämaturie, Proteinurie
- ANCA

→

- Erhaltung der Nierenfunktion
- Regelmäßige Evaluierung, follow-up

Relapse Prävention

→

- Individualisierte Therapie

PEXIVAS

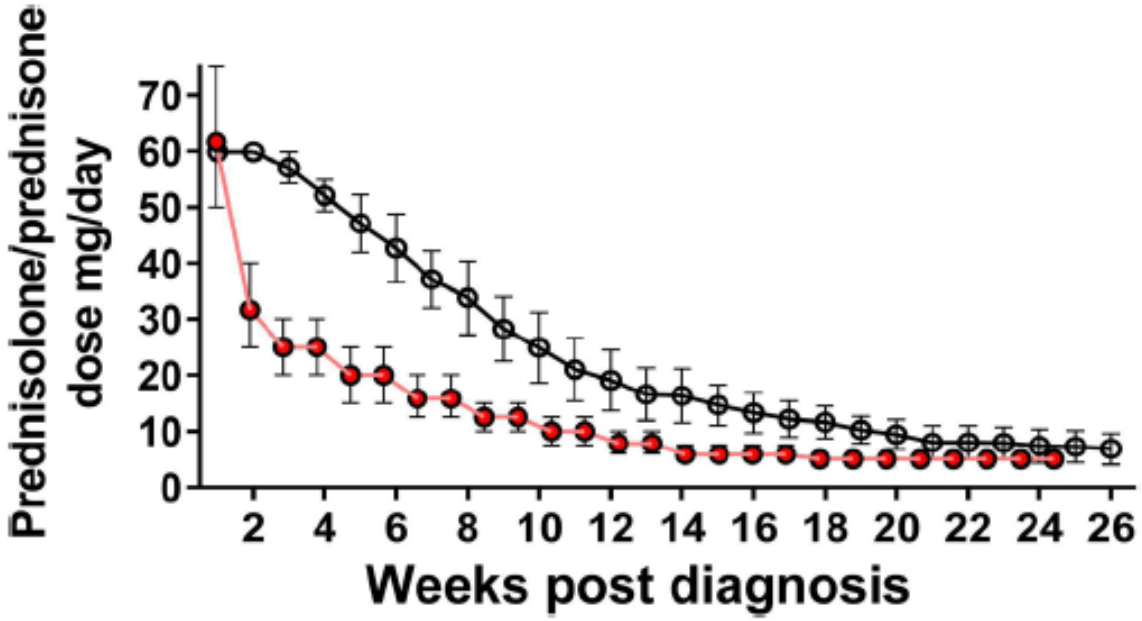


Figure 2 Protocol target glucocorticoid (GC) doses in AAV induction trials[81 106 113 221–226] (black line), illustrating how these compare with the reduced GC group from the PEXIVAS trial (red line). The line and

Week	<50kg	50-75kg	>75kg
1	50	60	75
2	25	30	40
3-4	20	25	30
5-6	15	20	25
7-8	12,5	15	20
9-10	10	12,5	15
11-12	7,5	10	12,5
13-14	6	7,5	10
15-16	5	5	7,5
17-18	5	5	7,5
19-20	5	5	5
21-22	5	5	5
23-52	5	5	5

Hellmich B, et al. Ann Rheum Dis 2023

Walsh et al, NEJM 2020

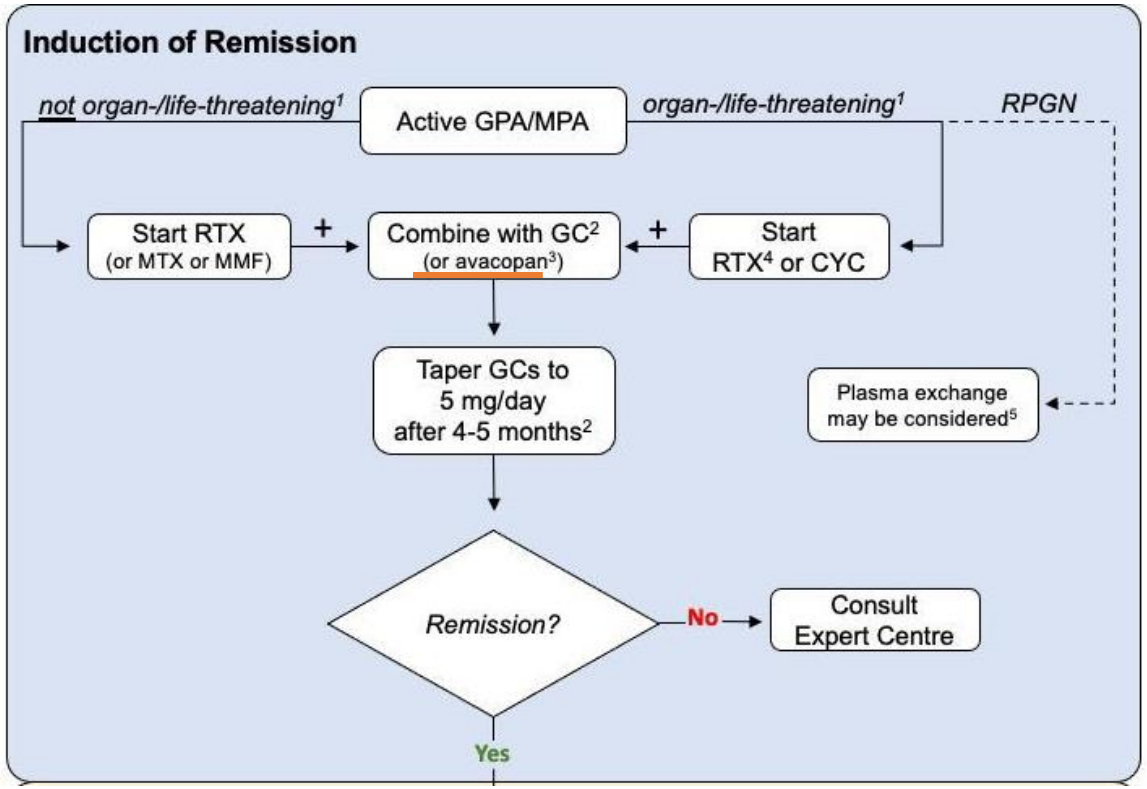
Table 3. Secondary Outcomes.*

Secondary Outcome	Plasma Exchange vs. No Plasma Exchange	Reduced-Dose vs. Standard-Dose Glucocorticoid Regimen	
	effect size (95% CI)		
Death from any cause	0.87 (0.58–1.29)	0.78 (0.53–1.17)	
End-stage kidney disease	0.81 (0.57–1.13)	0.96 (0.68–1.34)	
Sustained remission	1.01 (0.89–1.15)	1.04 (0.92–1.19)	
Serious adverse events	1.21 (0.96–1.52)	0.95 (0.75–1.20)	
Serious infections at 1 year	1.16 (0.87–1.56)	0.69 (0.52–0.93)	P 0.02

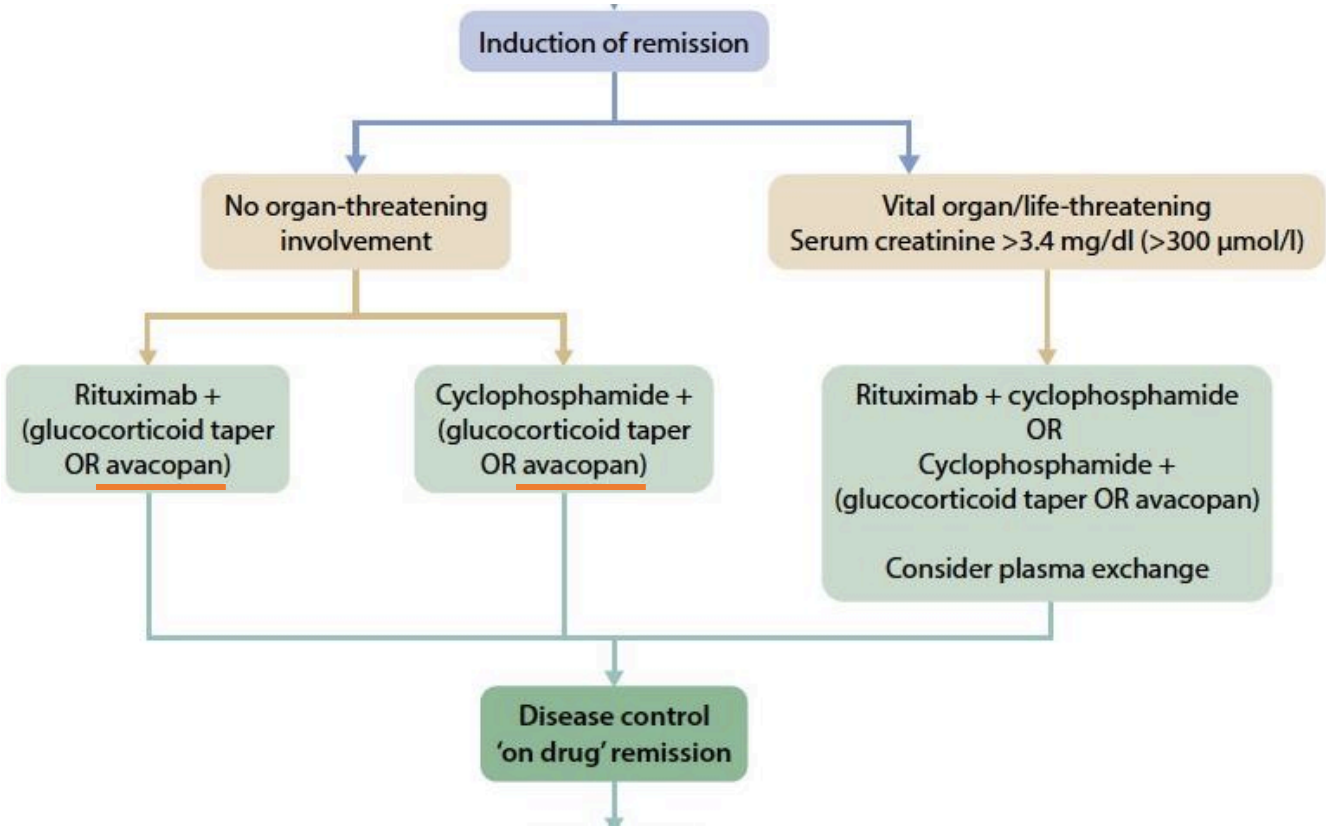
30% Reduktion von schweren Infektionen

EULAR recommendations for the management of ANCA-associated vasculitis: 2022 update

Bernhard Hellmich ,[1] Beatriz Sanchez-Alamo,[2] Jan H Schirmer,[3] Alvise Berti ,[4,5] Daniel Blockmans,[6] Maria C Cid ,[7] Julia U Holle,[8] Nicole Hollinger,[1] Omer Karadag



KDIGO 2024



Hellmich B, et al. Ann Rheum Dis 2023

Floege J et al. KI 2024

Treatment Individualisation

Factors that increase relapse risk for AAV

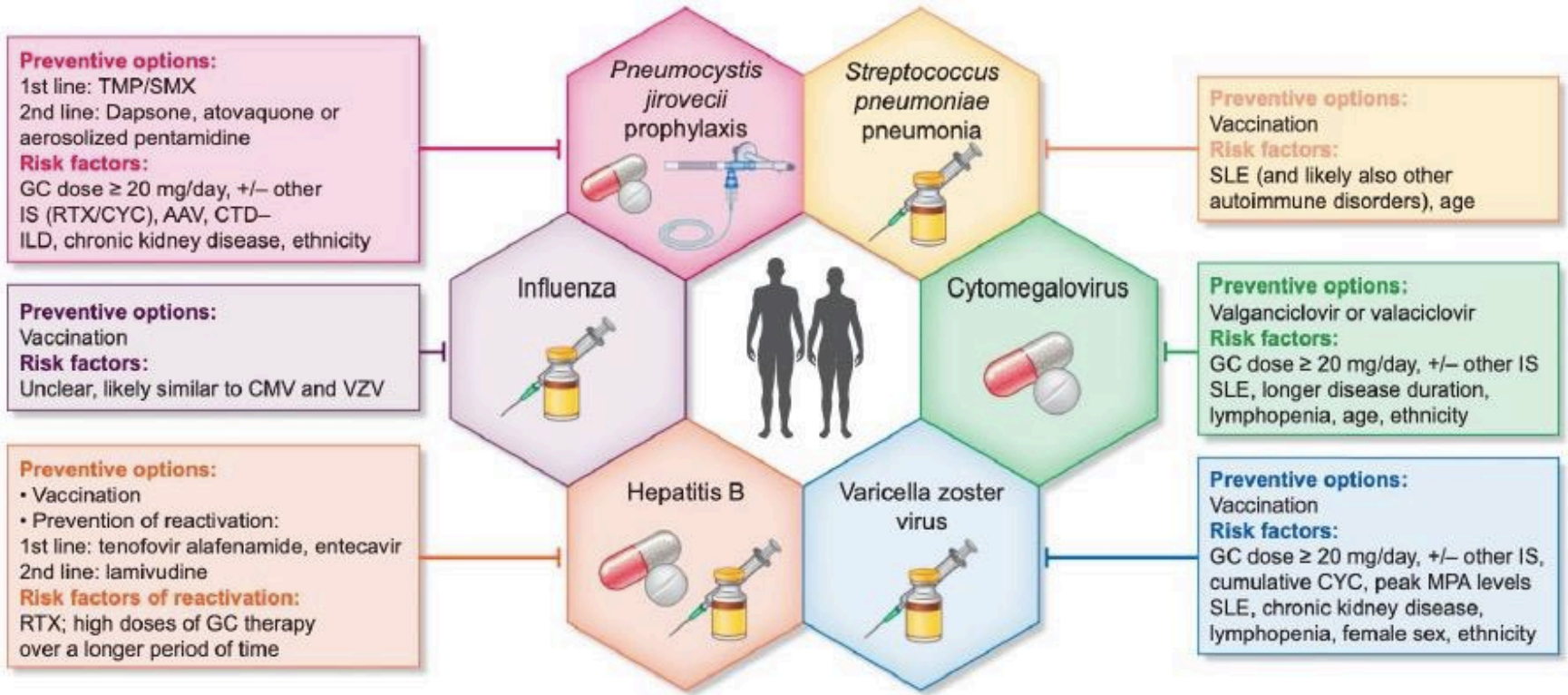
Baseline factors	Factors after diagnosis	Treatment factors
• Diagnosis of granulomatosis with polyangiitis • PR3–ANCA subgroup • Lower serum creatinine • More extensive disease • Ear, nose, and throat disease	• History of relapse • ANCA positive at the end of induction • Rise in ANCA	• Lower cyclophosphamide exposure • Immunosuppressive withdrawal • Glucocorticoid withdrawal

- Persisting haematuria after induction
- No RTX maintenance

ANCA is the strongest predictor for relapse after RTX

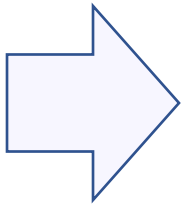
2024

Preventive strategies



Windpessl M et al NDT 2023

…. eine optimale Therapie



- Rasche, potente Wirkung
- Günstiges Nebenwirkungsprofil
- Steroidfreie Startegie

- Erhaltung der Nierenfunktion
- Reduktion von Proteinurie

- Prävention von Rezidiven

- Optimierung der Lebensqualität

Thank You !!

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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.