

2. DACH ANCA VASKULITIS FORUM 2024

22. & 23. NOVEMBER 2024 | MÜNCHEN

CSL Vifor

Intensivierte Induktionstherapie bei ausgewählten Patienten

Prof. Dr. Gunter Aßmann



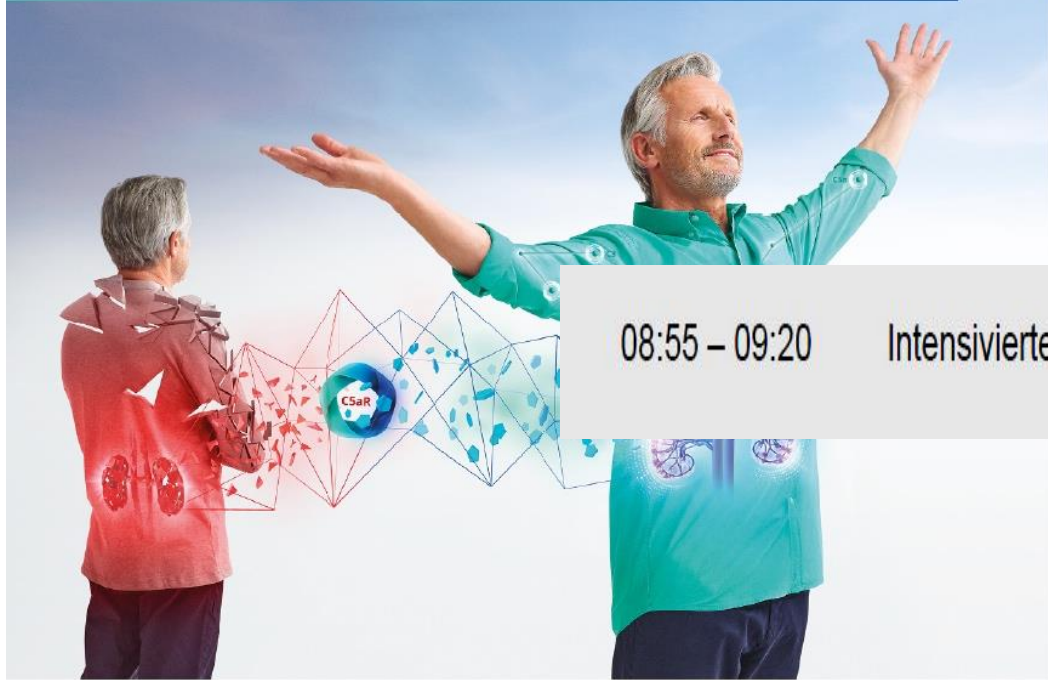
Minden





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22. & 23. NOVEMBER 2024 | MÜNCHEN



08:55 – 09:20

Intensivierte Induktionstherapie bei ausgewählten Patienten

G. Aßmann, Minden (DE)



Vifor Pharma Österreich GmbH
Vifor Pharma Switzerland AG
Vifor Pharma Deutschland GmbH

CSL Vifor



Interessenskonflikte

Grants für Grundlagen-Forschung 2019-2024 von:
Pfizer, Novartis, AbbVie, Chugai

Vortragshonorare 2019-2024:
**Pfizer, MSD, Novartis, AbbVie, GSK, Boehringer-Ingelheim,
Janssen, UCB, Astrazeneca, CSL Vifor Pharma**

Univ.-Prof. Dr. Gunter Aßmann
Klinik für Rheumatologie
und klinische Immunologie
RUB-Universitätsklinikum Minden JWK
D-32429 Minden

Intensiviert:

- (1) Kombinationstherapie: CYC + RTX + GC (+ AVAC)
Dosisescalation: CYC, GC
- (2) Off-label/exp. Therapien

Ausgewählte Patienten:

Klinische Konstellationen

- Hoher BVAS
- Terminale Organbedrohung
- Multiple „major items“

Intensivierte Induktionstherapie bei ausgewählten Patienten

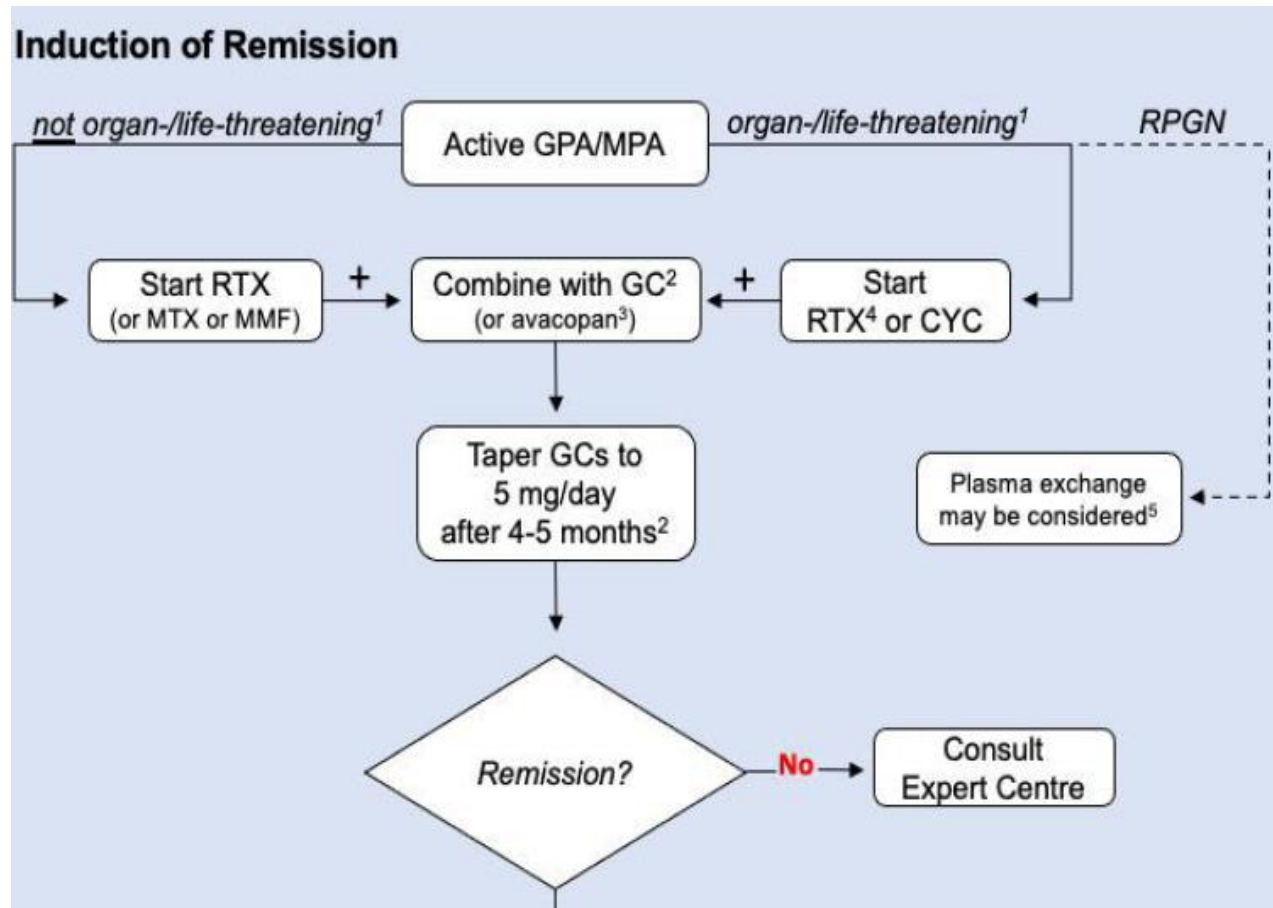
Induktionstherapie:

Remissionsinduktion

- bei ED
- im Rezidiv
- bei Refraktärität

Recommendation

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



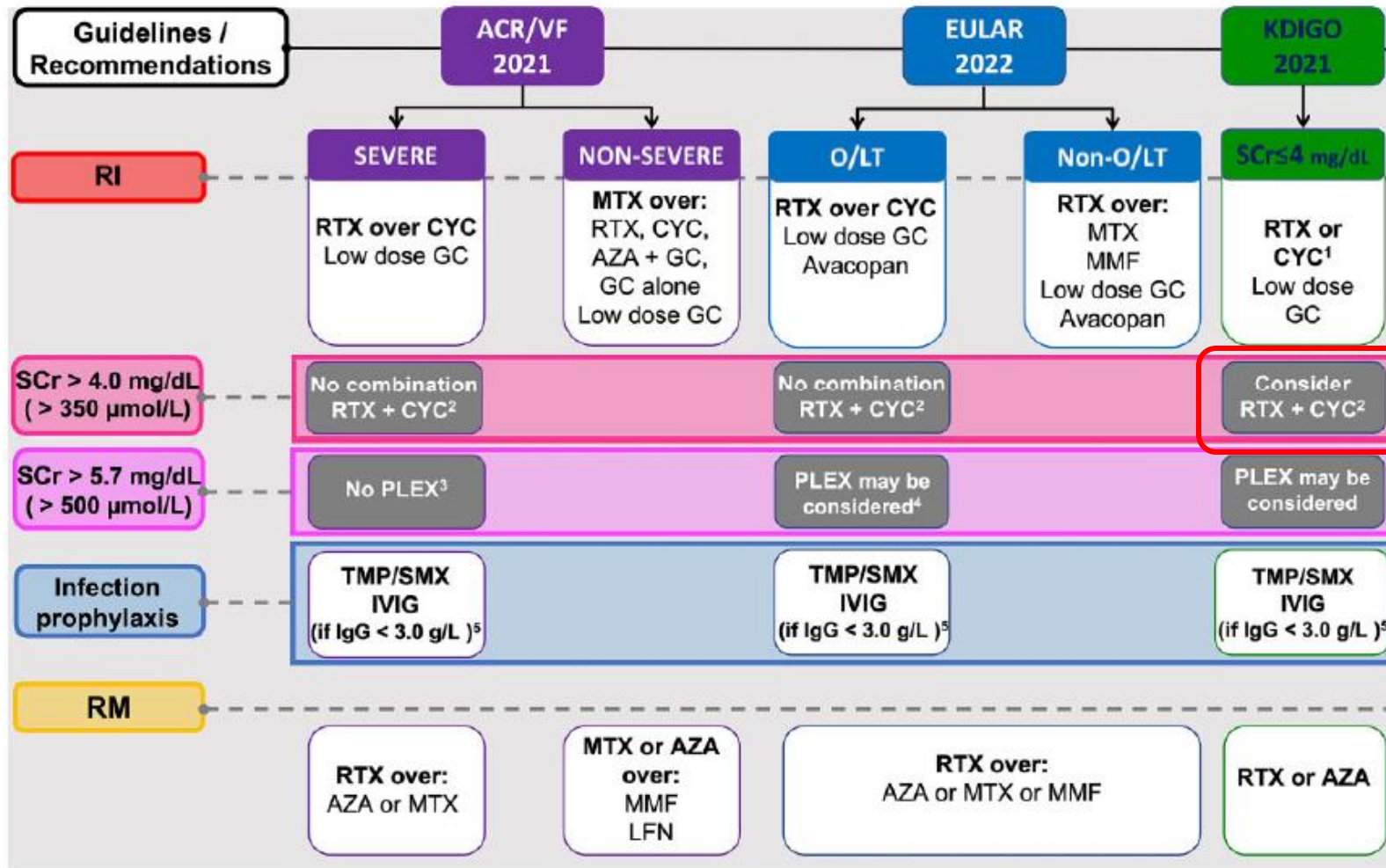
Ausgewählte Patienten:
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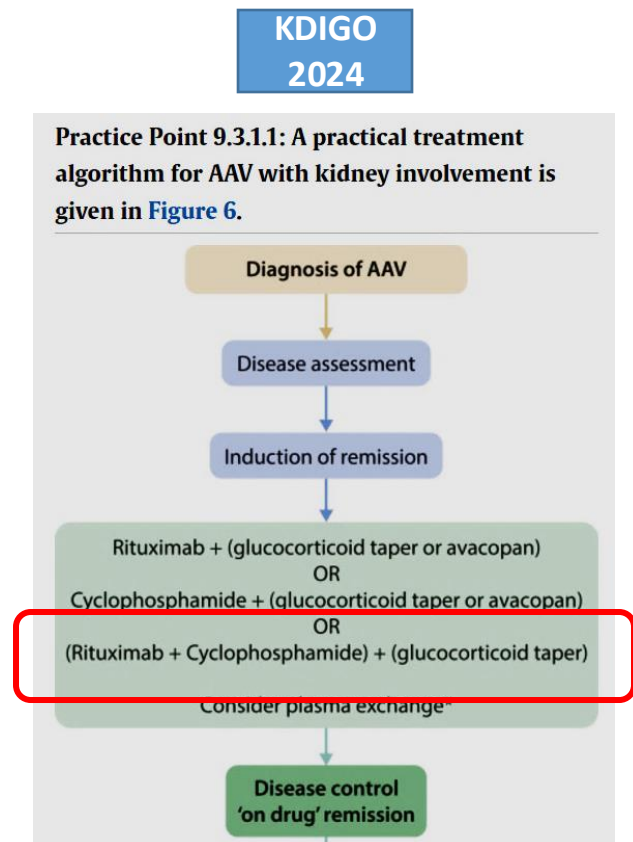
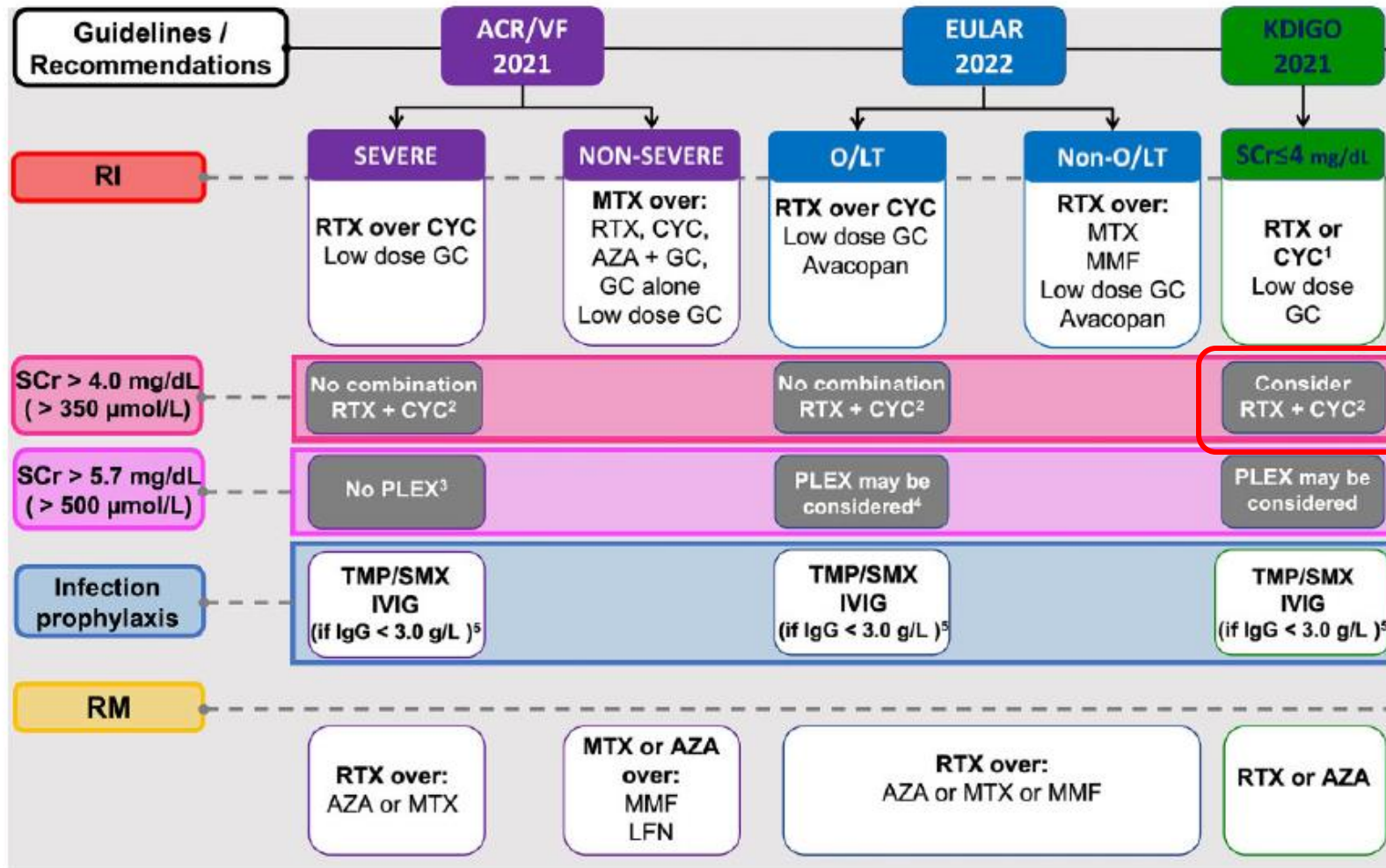
Standardschema vs. Individualschema

Induktionstherapie:
 Remissionsinduktion

Intensiviert:
 Kombinationstherapie: CYC + RTX + GC + AVAC
 Dosisescalation: CYC, GC



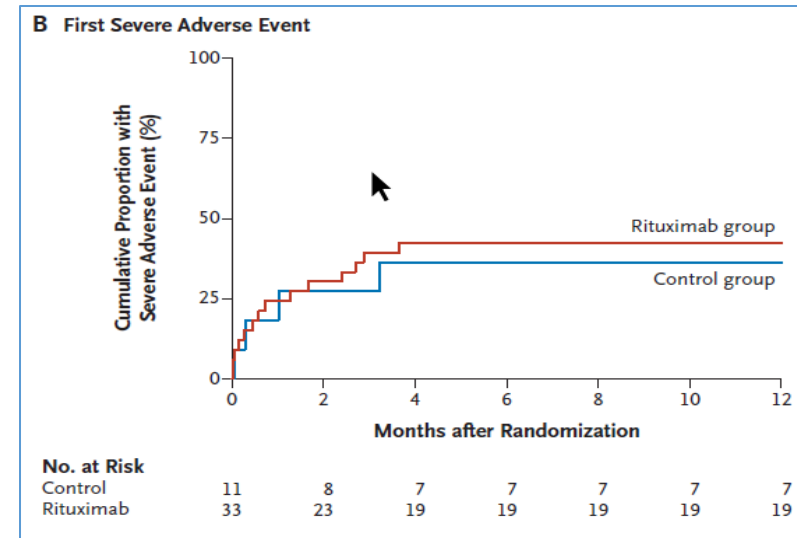
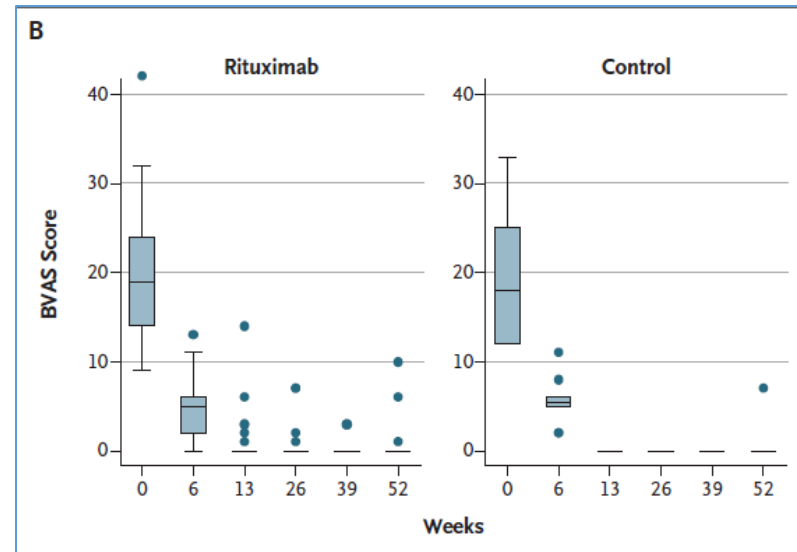
Die Nephrologie führend in Empfehlungen:
 „CYC+RTX in besonderer Situation zu erwägen“



RITUXIVAS

Schema: RTX 375/qm 4w + CYC 2x15mg/KG iv
 vs. CYC (standard, 3-6 mon). N=44

Charakteristika: (prä-)terminale NI, hoher BVAS:19



RITUXIVAS

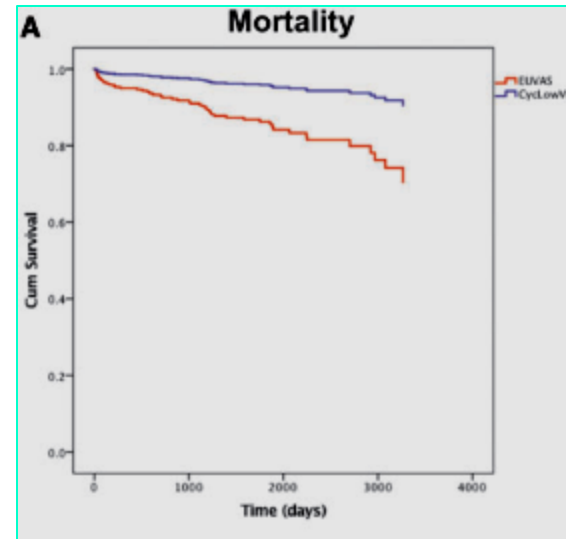
Schema: RTX 375/qm 4w + CYC 2x15mg/KG iv

CycLowVas-Studie

Schema: RTX 1g Tag1+14 + 6xCYC 500mg (n=66)

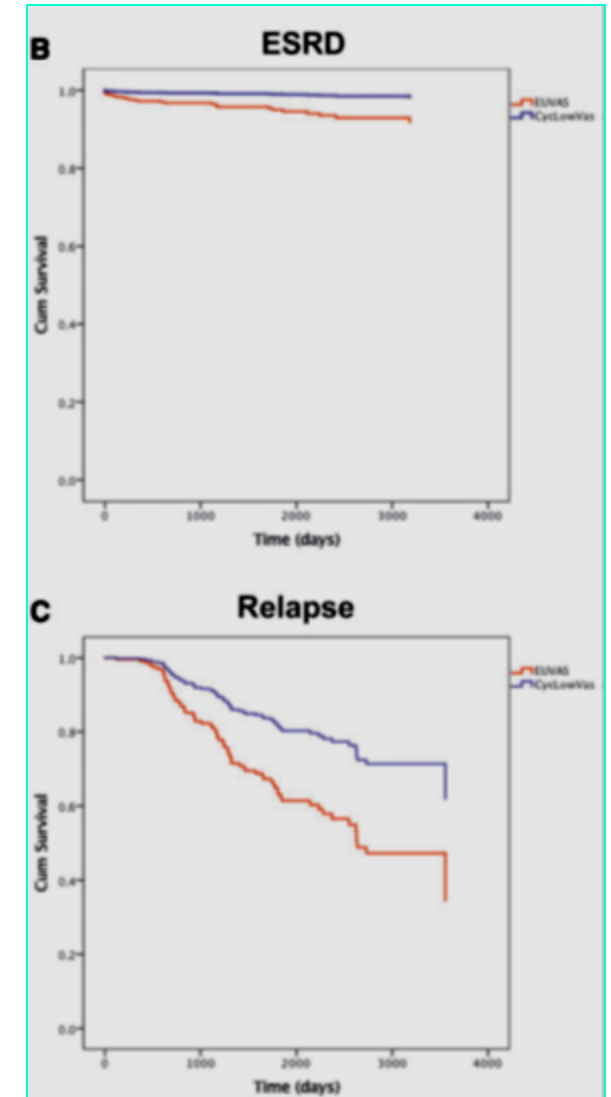
Vergleich mit historischen Daten EUVAS (n=198)

Charakteristika: follow-up 56m



Zusätzlich:

- signifikant <GC
- signifikant <CYC Dosis
- Hohe Signifikanz: bei PR3+
- Infektraten: idem



Jones RB, NEJM 2010

McAdoo et al, Nephrol Dial Transplant 2019

RITUXIVAS

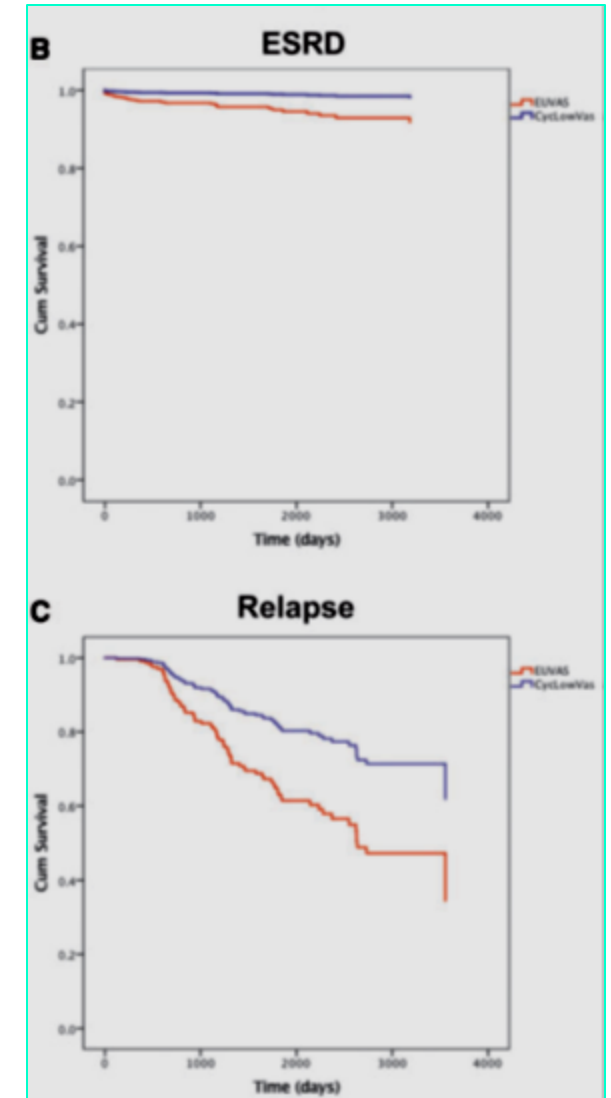
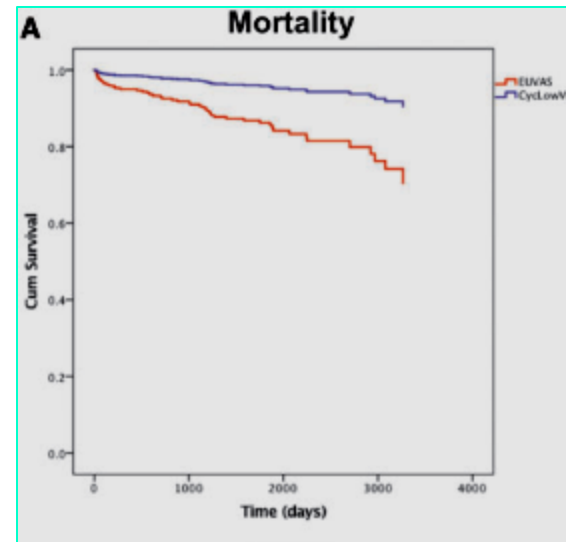
Schema: RTX 375/qm 4w + CYC 2x15mg/KG iv

CycLowVas-Studie

Schema: RTX 1g Tag1+14 + 6xCYC 500mg

Endurance-Trail (Ph3)

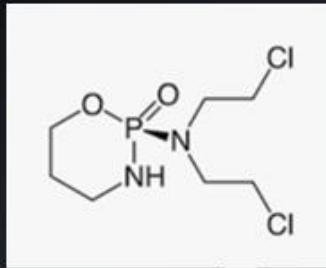
Schema: RTX 1g Tag1+14 +/- 6xCYC 500mg



Jones RB, NEJM 2010

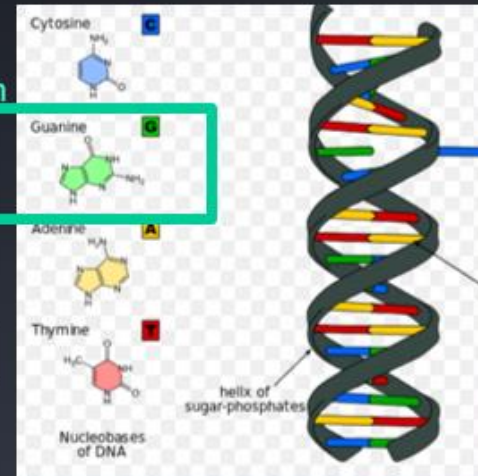
McAdoo et al, Nephrol Dial Transplant 2019; Dirikgill et al, BMC open 2022

Cyclophosphamid

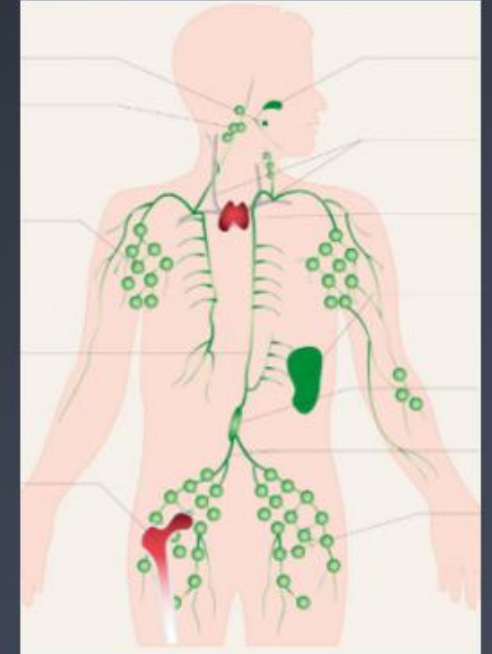


4-Hydroxy-Cyclophosphamid

Alkylierung von Guanin

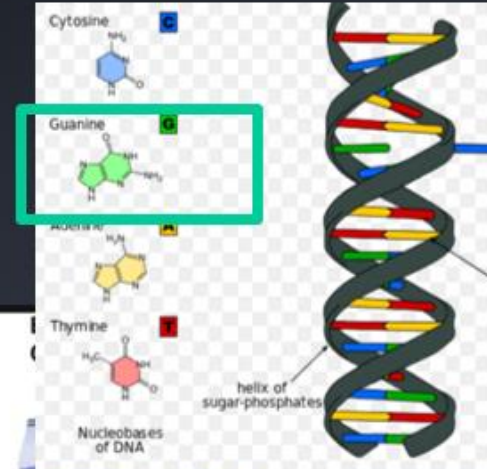


Apoptose der Zellen



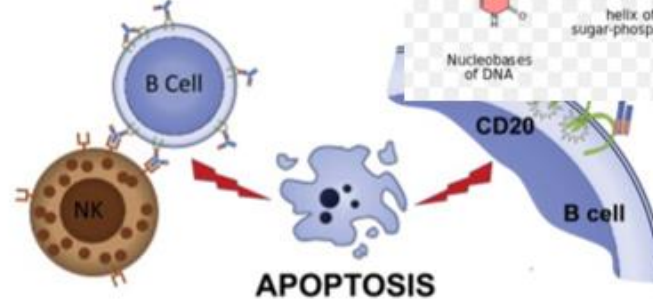
Lymphatisches System
überproportional genetisch
repräsentiert mit Guanin

Rituximab (RTX) + Cyclophosphamid

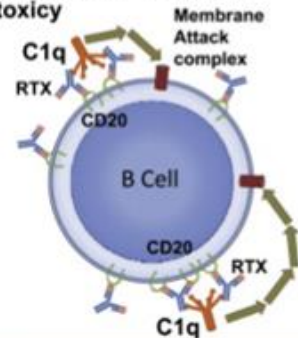


Apoptose der Zellen

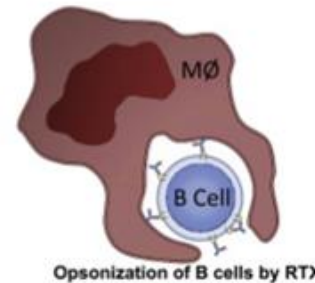
A. Antibody-dependent cellular cytotoxicity (ADCC)



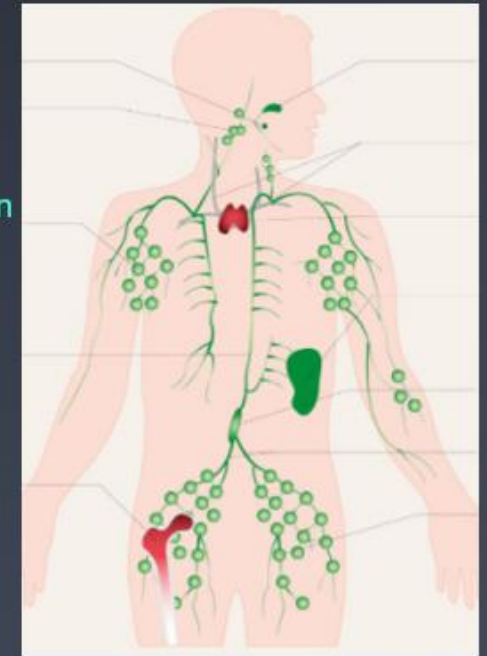
C. Complement dependent cytotoxicity



D. Phagocytosis by Macrophages

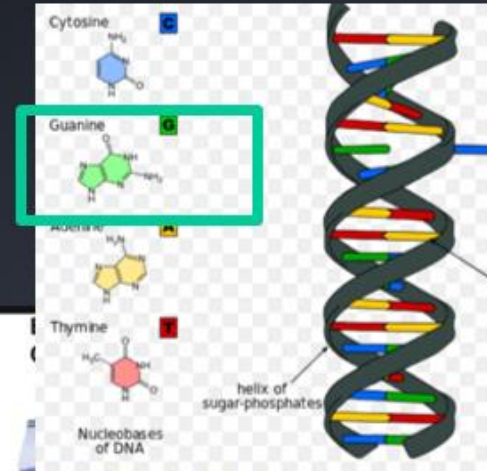


B-Zell-Depletion



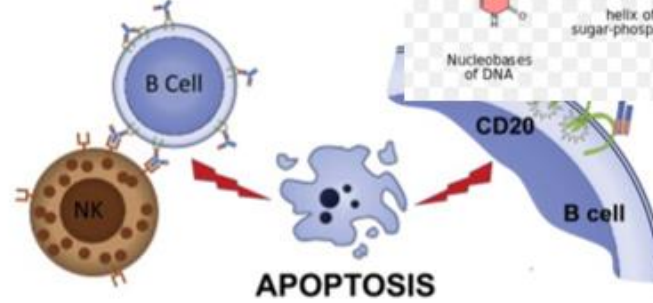
Hoch effiziente Depletion des lymphatischen Systems (B>T Lymphozyten)

Rituximab (RTX) + Cyclophosphamid

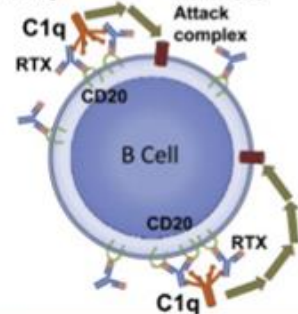


Apoptose der Zellen

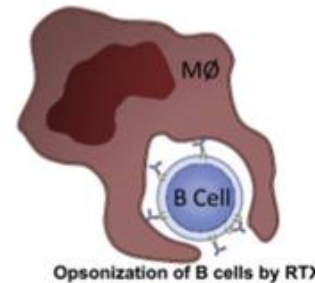
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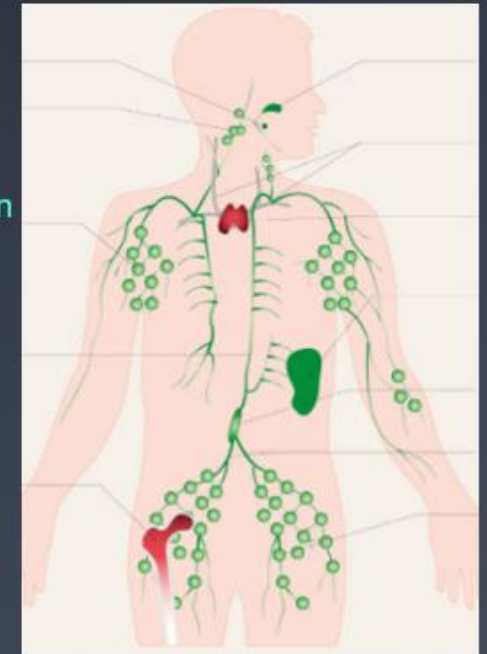
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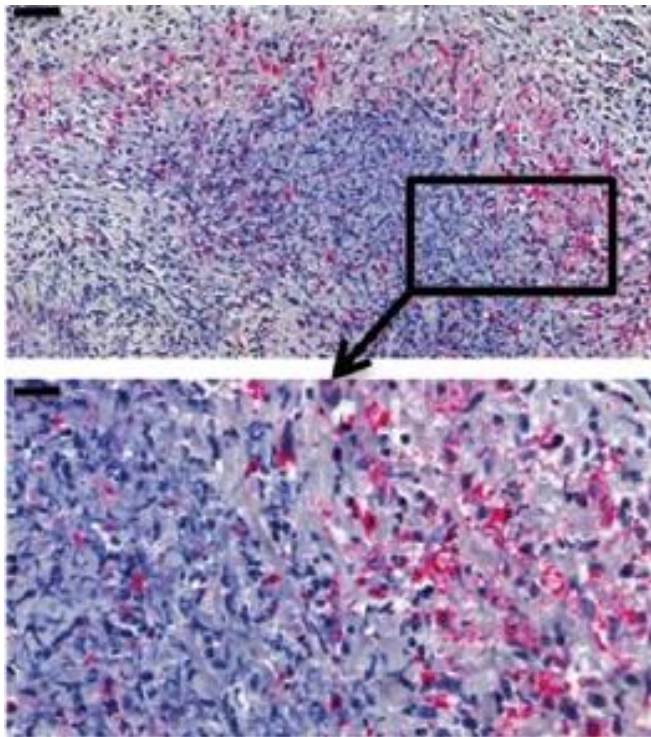
B-Zell-Depletion



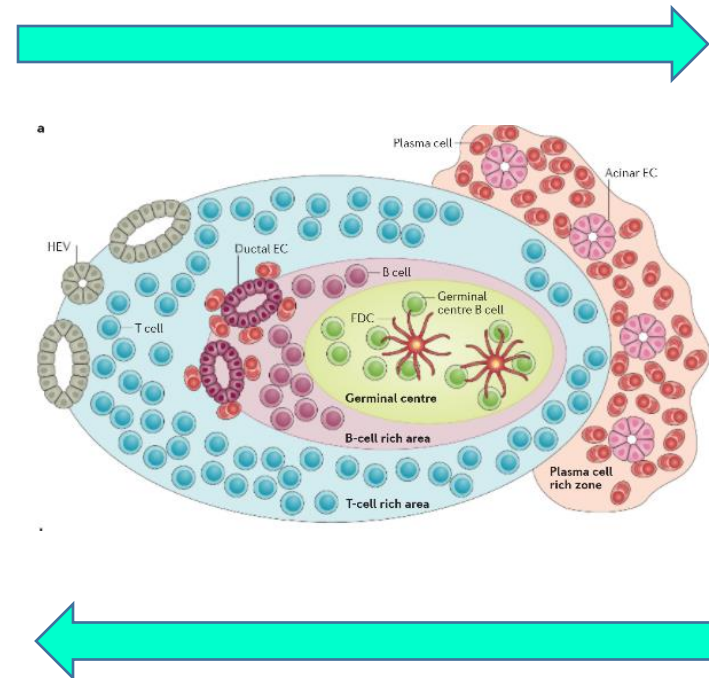
Hoch effiziente Depletion des lymphatischen Systems (B>T Lymphozyten)

- ➡ „target“: Lymphom
- ➡ „target“: ELS/cluster

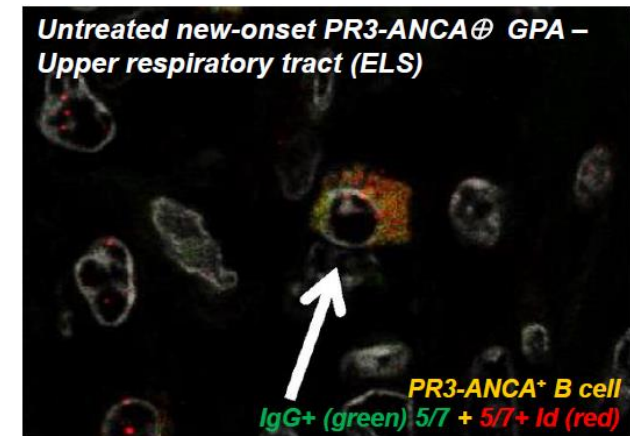
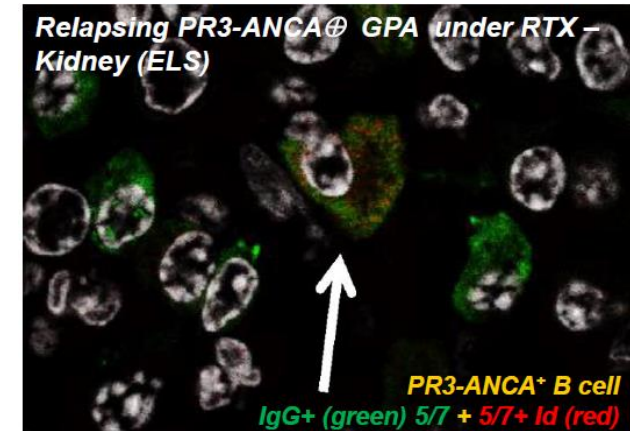
Bioptat: GPA



Architektur: ELS

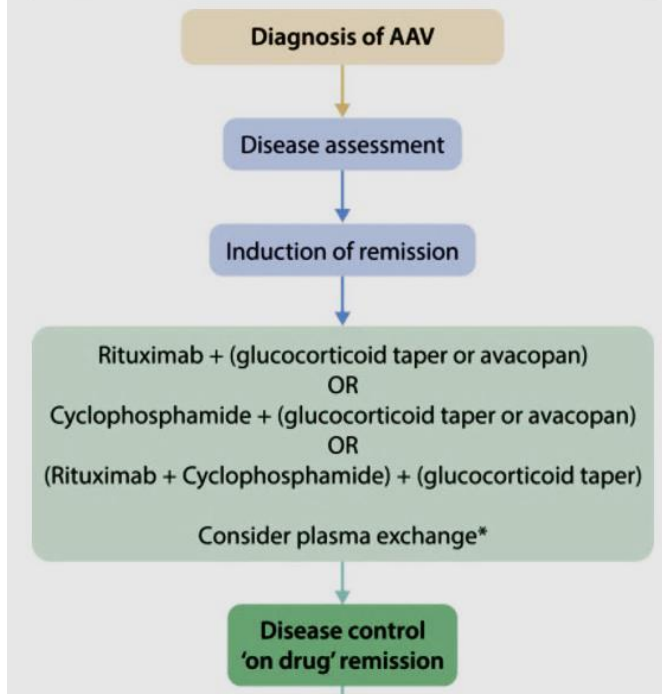


„therapierefraktäre“ autoimmune Cluster



KDIGO
2024

Practice Point 9.3.1.1: A practical treatment algorithm for AAV with kidney involvement is given in Figure 6.



KDIGO 2024

Practice Point 9.3.1.1: A practical treatment algorithm for AAV with kidney involvement is given in Figure 6.

Diagnosis of AAV

Disease assessment

Induction of remission

Rituximab + (glucocorticoid taper or avacopan)
OR
Cyclophosphamide + (glucocorticoid taper or avacopan)
OR
(Rituximab + Cyclophosphamide) + (glucocorticoid taper)

Consider plasma exchange*

Disease control
'on drug' remission

Ausgewählte Patienten:

Klinische Konstellationen

- Hoher BVAS
- **Terminale Organbedrohung**
- **Multiple „major items“**

Standardschema vs. Individualschema

Induktionstherapie

Intensiviert:

Kombinationstherapie: CYC + RTX + GC + AVAC

Dosiseskalation: CYC, GC

	Diagnosis	
	GPA (n=2434)	MPA (n=1434)
Demography		
Age, years	53.4 (16.3; n=2413)	63.6 (14.6; n=1422)
Gender		
Men	1301/2433 (53.5%)	705 (49.2%)
Women	1132/2433 (46.5%)	729 (50.8%)
Diagnosis		
GPA	2434 (100%)	0
MPA	0	1434 (100%)
Organ pattern		
Constitutional	1534/2384 (64.3%)	740/1356 (54.6%)
Musculoskeletal	1375/2384 (57.7%)	660/1356 (48.7%)
Skin	619/2429 (25.5%)	303 (21.1%)
Mucosa	95/2411 (3.9%)	12/1420 (0.8%)
Eyes	533/2412 (22.1%)	50/1422 (3.5%)
Ear-nose-throat	1697/2433 (69.7%)	172 (12.0%)
Lung	1470/2432 (60.4%)	507/1433 (35.4%)
Cardiovascular	185/2430 (7.6%)	103/1433 (7.2%)
Gastrointestinal	170/2429 (7.0%)	103/1431 (7.2%)
Kidney	1403 (57.6%)	1188 (82.8%)
Central nervous system	228/2414 (9.4%)	63/1420 (4.4%)
Peripheral nervous system	395/2410 (16.4%)	289/1420 (20.4%)

Data-driven subclassification of ANCA-associated vasculitis: model-based clustering of a federated international cohort



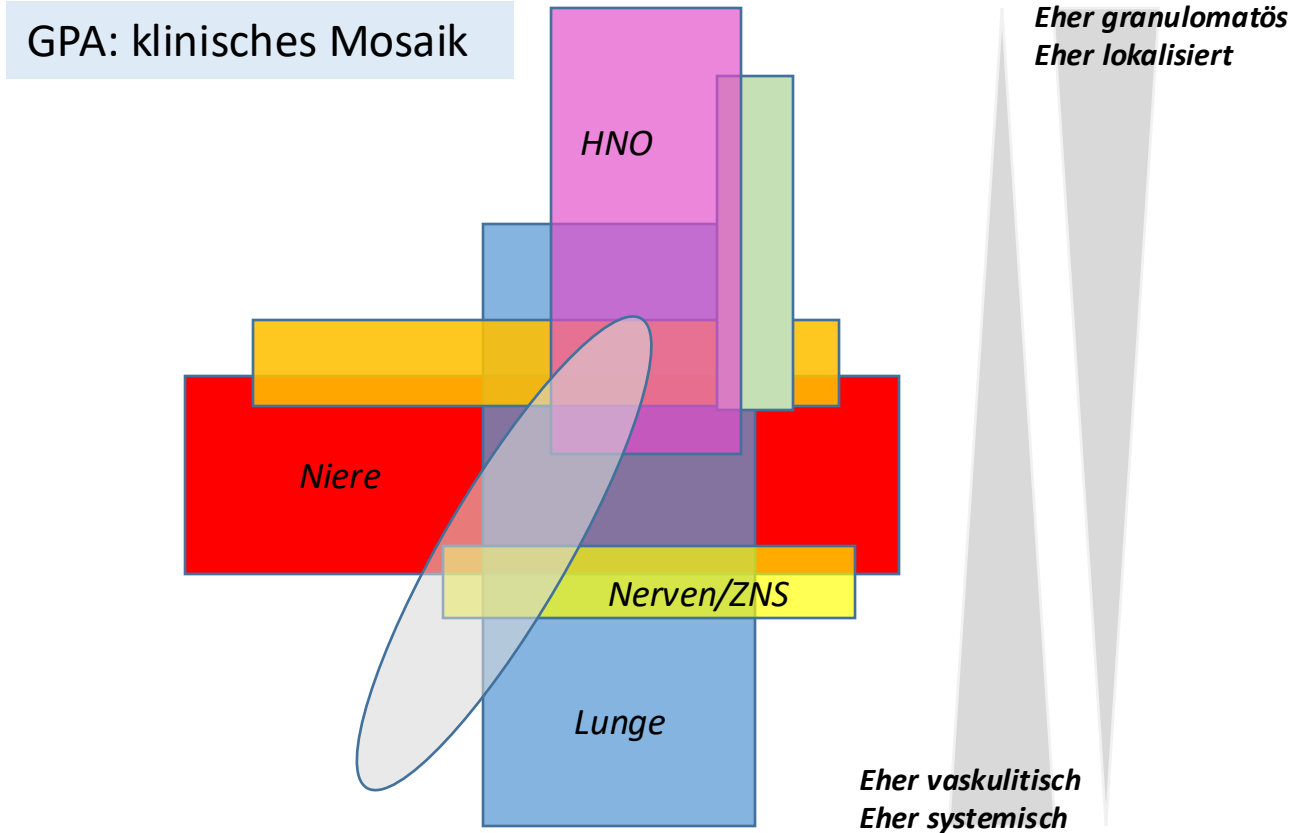
Karl Gisslander, Arthur White, Louis Aslett, Zdenka Hrušková, Peter Lamprecht, Jacek Musiał, Jamsheela Nazeer, James Ng, Dedan O'Sullivan, Xavier Puéchal, Matthew Rutherford, Márten Segelmark, Benjamin Terrier, Vladimír Tesáľ, Michelangelo Tesi, Augusto Vaglio, Krzysztof Wójcik, Mark A Little*, Aladdin J Mohammad*, on behalf of the FAIRVASC consortium†

Nieren: „major item“ und „Schlüssel“-Manifestation:

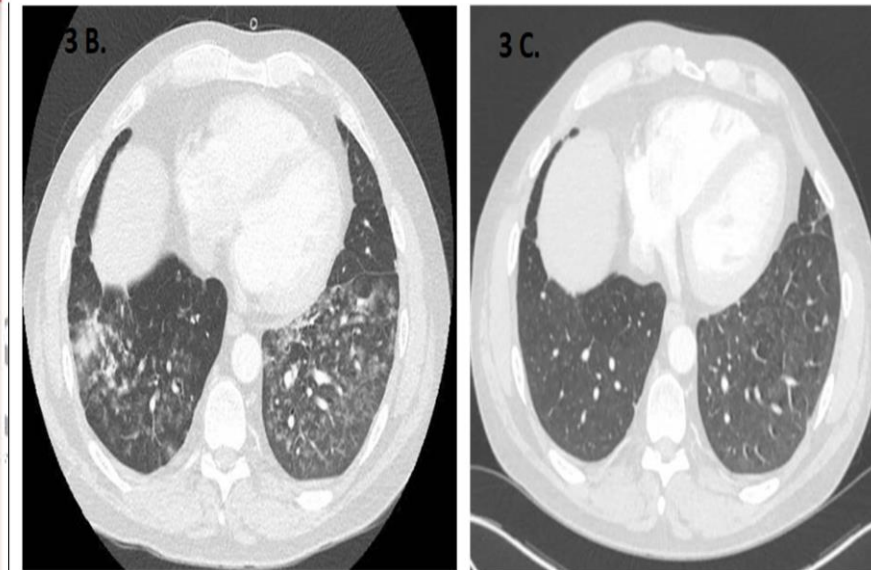
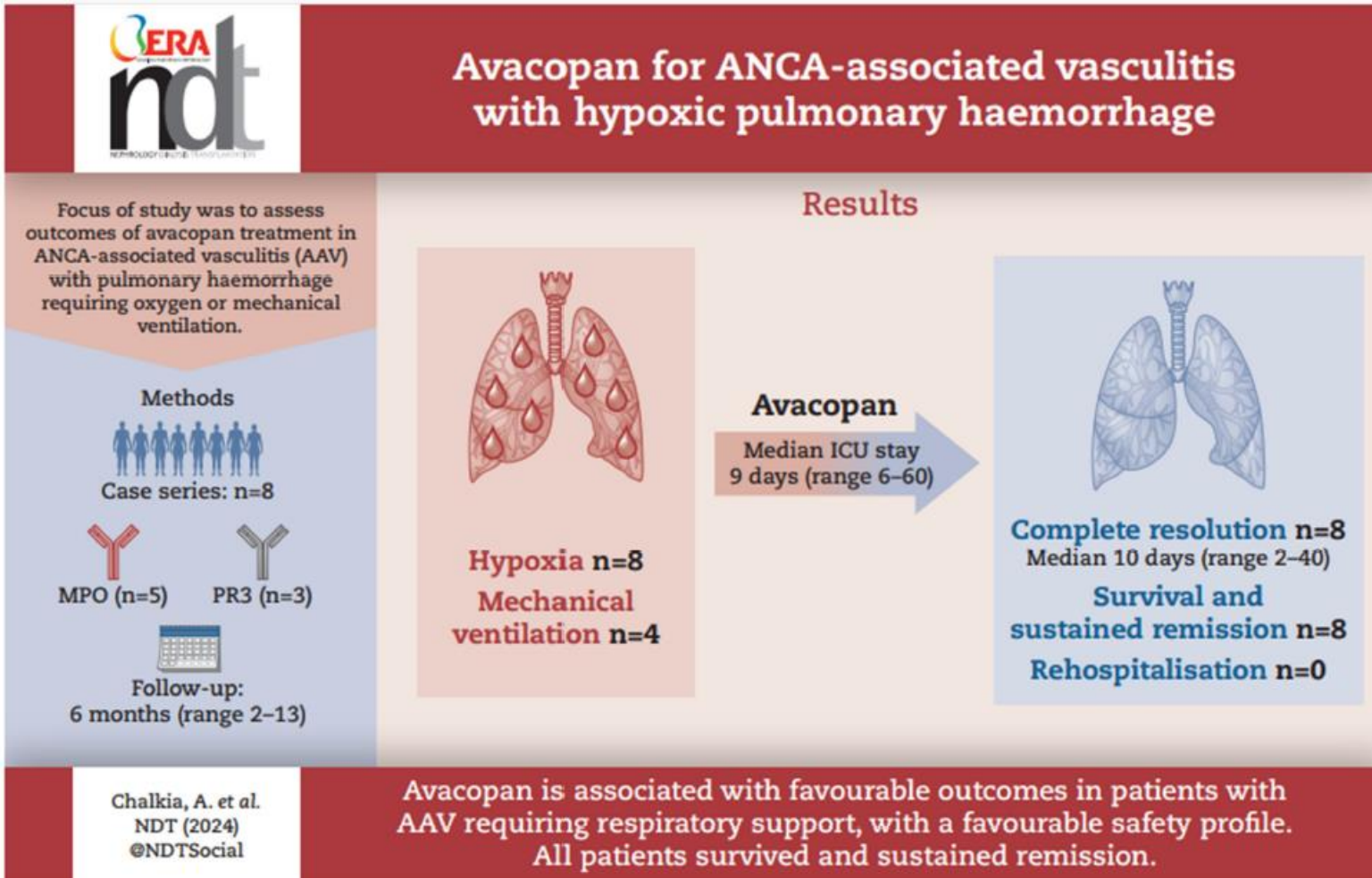
- Krankheitslast
- Outcome/Prognose
- Richtungsweisend für Therapieauswahl ?

AAV oft mit extra-renalen aber inklusive renalen Manifestationen

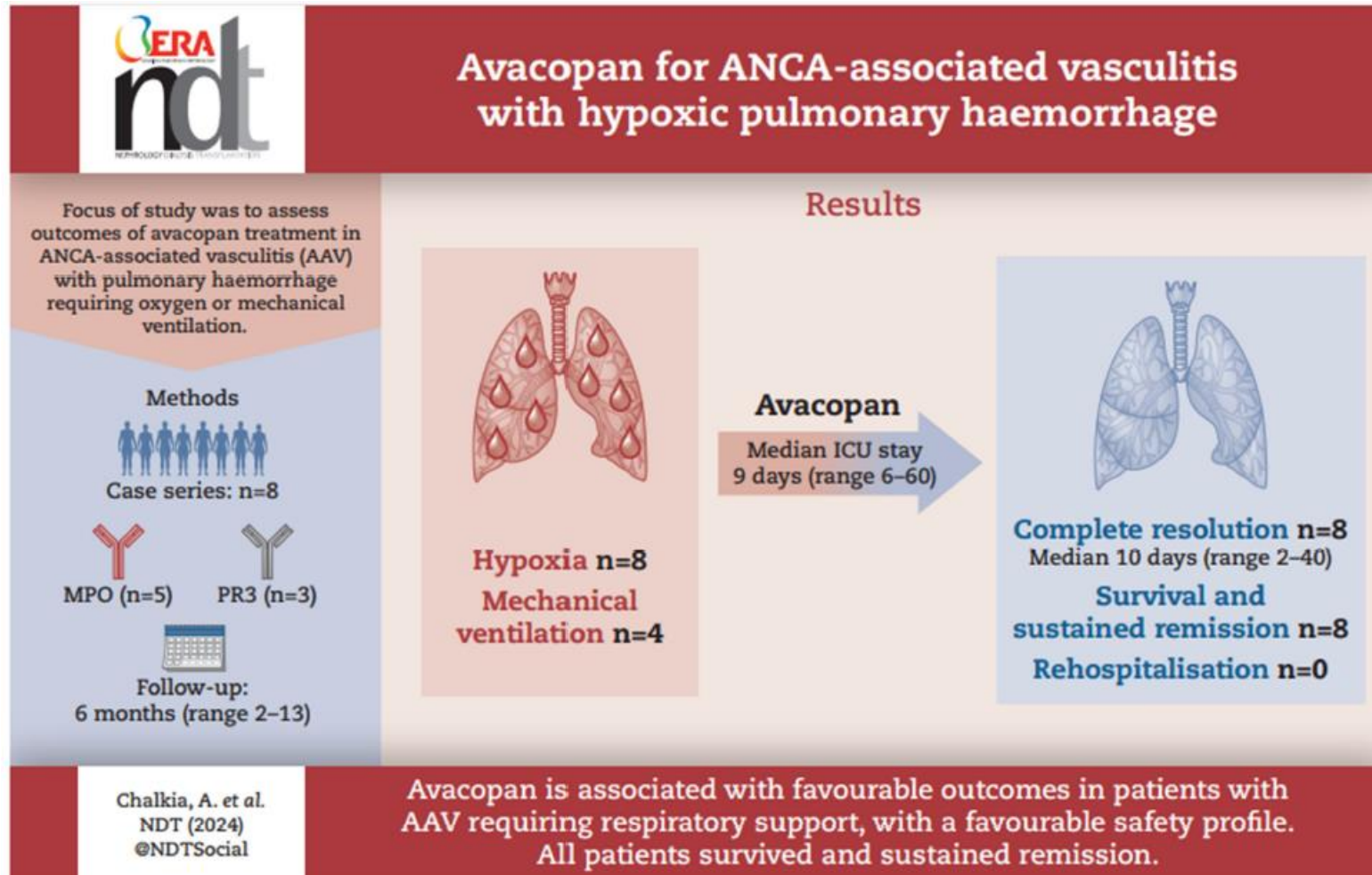
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AAV mit schwerer Lungenbeteiligung (major item): alveolärer Hämorrhagie: maximierte Therapie



AAV mit schwerer Lungenbeteiligung (major item): alveolärer Hämorrhagie: maximierte Therapie



Maximierte Therapie

CYC n=1

RTX n=4

CYC+RTX n=4

PLEX n= 4

ECMO n=2

Avacopan add on

Tag 10 (2-40)

GC fre n=3 von 8

AAV mit schwerer Lungenbeteiligung (major item): alveolärer Hämorrhagie: maximierte Therapie

Table 1. Clinical characteristics of 15 patients with AAV with DAH treated with avacopan*

Demographics and clinical presentation	Patient data
Age at presentation with DAH, median (IQR), y	66 (52–72)
Female, n (%)	9 (60)
Body mass index, median (IQR) ^a	27 (21–36)
Ethnicity, n (%) ^b	
White	14 (93)
Unknown ^c	1 (7)
Smoking status, n (%)	
Never smoked	11 (73)
Former smoker	4 (27)
Vasculitis disease status, n (%)	
Newly diagnosed	9 (60)
Relapsed	6 (40)
Immunosuppression before admission	2 (13)
ANCA ELISA, n (%)	
PR3	8 (53)
MPO	7 (47)
AAV subtype, n (%)	
GPA	8 (53)
MPA	7 (47)
BVAS/WG, median (IQR) ^d	8 (5–10)
Organ involvement, n (%)	
Renal	9 (60)
General ^e	10 (66)
ENT	7 (46)
Nervous system	1 (7)
BAL confirmation, n (%)	14 (93)
SpO ₂ /FIO ₂ on presentation, median (IQR) ^f	449 (397–461)
Respiratory failure, n (%)	3 (20)
Renal failure, n (%)	2 (13)
Remission induction therapies, n (%)	
Rituximab	14 (94)
Plasma exchange	5 (33)
Cyclophosphamide (IV)	3 (20)

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AMERICAN COLLEGE
of RHEUMATOLOGY
Empowering Rheumatology Professionals

Treatment of Antineutrophil Cytoplasmic Antibody–Associated Vasculitis With Diffuse Alveolar Hemorrhage With Avacopan

Sam D. Falde,¹ Amos Lal,¹ Rodrigo Cartin-Ceba,² Lester E. Mertz,² Fernando C. Fervenza,¹ Ladan Zand,¹ Matthew J. Koster,¹ Kenneth J. Warrington,¹ Augustine S. Lee,³ Nabeel Aslam,³ Andy Abril,³ and Ulrich Specks¹

Table 2. Clinical characteristics and outcomes stratified by time of administration of avacopan during presentation with diffuse alveolar hemorrhage*

	Early avacopan ^a (n = 7)	Late avacopan ^b (n = 8)	P
MPA, n (%)	6 (85)	1 (13)	0.005
MPO-ANCA, n (%)	6 (85)	1 (13)	0.005
BVAS/WG, median (IQR)	7 (3–10)	8 (6–10)	0.44
Organ involvement, n (%)			
General	5 (71)	5 (62)	0.78
Renal	5 (71)	4 (50)	0.65
ENT	2 (25)	7 (71)	0.07
Nervous system	0 (0)	1 (13)	0.34
SpO ₂ /FIO ₂ on presentation, median (IQR)	442 (363–458)	449 (427–458)	0.69
Respiratory failure, n (%)			
HFNC	1 (14)	0 (0)	0.33
IMV	2 (28)	0 (0)	0.15
Renal failure, n (%)	1 (14)	1 (13)	0.92
Achievement of remission, ^c n (%)			
Remission	7 (100)	7 (88)	0.33
Complete remission	5 (71)	5 (62)	0.71
Death, n (%)	0 (0)	1 (13)	0.919

AAV mit schwerer Nieren- und Lungenmanifestation (major items) unter CYC+RTX+Avacopan

Table 1.

AAV patients' characteristics on avacopan

age (years, median, min-max)	59, 20-81
gender female/male (n/n)	15/15
ethnics (white/non-white) (n/n)	30/0
primary diagnosis/relaps (n/n)	27/3
AAV type GPA (n)/ MPA (n)	20/10
PR3-/MPO-ANCA positive (n/n)	20/10
renal biopsie: GN (n)	30
extra-renal major organ involvement (n)	30
pulmo (n)	26
cns (n)	4
cardiac (n)	2
eye (n)	19
mesenterical(n)	3
neuritis (n)	0
BVAS (mean, min-max)	23.2, 13-43

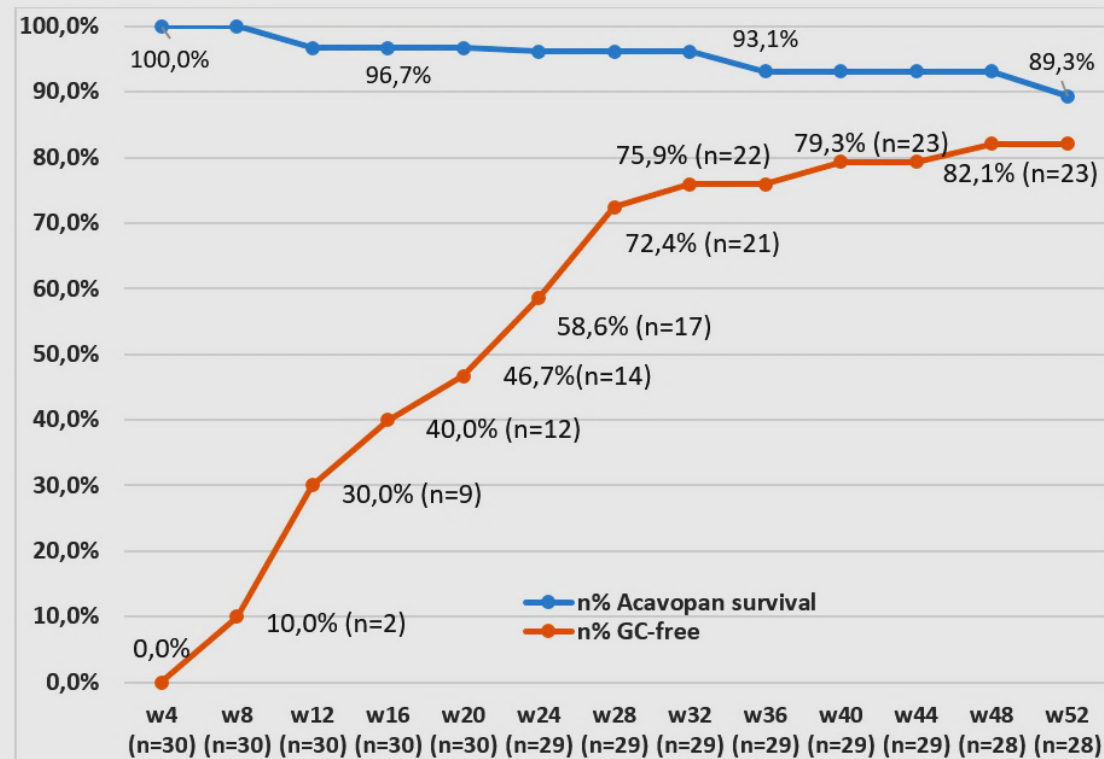
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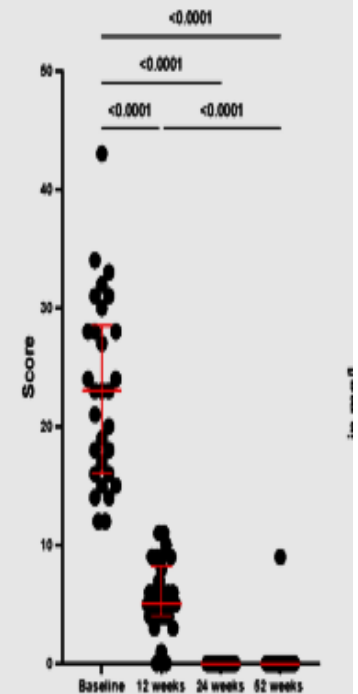
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extra-renal major organ involvement (n)	30
pulmo (n)	26
cns (n)	4
cardiac (n)	2
eye (n)	19
mesenterical(n)	3
neuritis (n)	9
BVAS (mean, min-max)	23.2, 13-43

Fig: survival of AAV patients on avacopan and glucocorticoids over 52 weeks.



BVAS V3.0



AAV mit schwerer Nieren- und Lungenmanifestation (major items) unter CYC/RTX/Avacopan

Real-World Experience With Avacopan in ANCA-Associated Vasculitis



Methods & Cohort		Baseline Characteristics		Outcomes	
	Multicenter study from United States		Renal involvement 77%	PRIMARY WEEK 26 Clinical remission at week 26 90%	SECONDARY WEEK 52 Clinical remission at week 52 84%
	Retrospective analysis N=92		Relapsing vasculitis 34%		
	Avacopan in newly diagnosed & relapsing AAV		Baseline eGFR<15ml/min/1.73m ² 23%		
	Median time to Start avacopan 3.6 wks Stop steroid 5.6 wks		Baseline KRT 10%		
	Study period: Oct 2021 to May 2023		Patients who received PLEX 14%		
			Induction regimen 1. Rituximab 48% 2. Cyclophosphamide(CP) 2% 3. Rituximab & low dose CP 47%	Change in eGFR (baseline to week 26) +12.2	Change in eGFR (baseline to week 52) +19.8
				Stopped Avacopan due to adverse events 20%	

AAV-ANCA associated vasculitis, PLEX-Plasma exchange, KRT-Kidney replacement therapy

KIREPORTS
Kidney International Reports

Reza Z et al, 2024

Visual abstract by:
Priya John MD,DM
@drpriyajohn

Conclusion: A high rate of remission and an acceptable safety profile were observed with the use of Avacopan in the treatment of AAV in this post-marketing analysis, including the populations excluded from the ADVOCATE trial

AAV unter CYC/RTX/Avacopan: Daten zu Komplikationen: Infekte

RITUXIVAS

Schema: RTX 375/qm 4w + CYC 2x15mg/KG iv
vs. CYC (standard, 3-6 months). N=44

Charakteristika: (prä-)terminale NI, hoher BVAS:19



Keine Unterschiede: Infektionen (12 Mon)

CycLowVas-Studie

Schema: RTX 1g Tag1+14 + 6xCYC 500mg (n=66)

Vergleich mit historischen Daten EUVAS (n=198)

Charakteristika: follow-up 56m



Keine Unterschiede: Infektionen trotz signifikant
Länger andauernder B-Zell-Depletion (56 Mon)

AAV unter CYC/RTX/Avacopan: Daten zu Komplikationen: Infekte

RITUXIVAS

Schema: RTX 375/qm 4w + CYC 2x15mg/KG iv
vs. CYC (standard, 3-6 months). N=44

Charakteristika: (prä-)terminale NI, hoher BVAS:19



Keine Unterschiede: Infektionen (12 Mon)

CycLowVas-Studie

Schema: RTX 1g Tag1+14 + 6xCYC 500mg (n=66)

Vergleich mit historischen Daten EUVAS (n=198)

Charakteristika: follow-up 56m



Keine Unterschiede: Infektionen trotz signifikant
Länger andauernder B-Zell-Depletion (56 Mon)

Mögliche Effekte der Kombinationstherapie



GC-Einsparung

- Intensivierte Induktion
- GC-Reduktion (PEXIVAS)
- AVACOPAN

Tiefe Remission ohne Re-Therapie/Induktion

Organerhalt mit pos. Sekundäreffekt

Intensiviert:
Kombinationstherapie: CYC + RTX + GC + AVAC

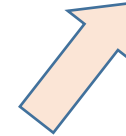
...scheint erfolversprechend als ...



Intensivierte Induktionstherapie bei ausgewählten Patienten

Ausgewählte Patienten:

- Terminale Organbedrohung: Niere
- Ggf. auch mit multiple „major items“



...als ...



Remissionsinduktion

- bei ED
- Ggf. auch im Rezidiv

CAVE: retrospektive, aber umfassende Daten für AAV der Niere, retrospektive kleine Fallzahlen für nicht-renale AAV

Intensiviert:
Kombinationstherapie: CYC + RTX + GC + AVAC

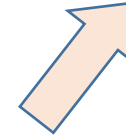
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AVACOPAN eher mit positiven Effekt auch in der Kombination CYC+RTX

Intensiviert:

Kombinationstherapie: CYC + RTX + GC + AVACOPAN

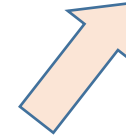
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AVACOPAN eher mit positiven Effekt auch in der Kombination CYC+RTX

Nach Cluster- und ELS-Hypothese der AAV intensiviert Kombinationstherapie inkl. AVACOPAN auch zu diskutieren:

- bei hohem BVAS /multiplen „major items“
- gravierender Lungenmanifestation



Vielen Dank für Ihre Aufmerksamkeit !

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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: Angioödem. **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.