2. DACH ANCA VASKULITIS FORUM 2024
22. & 23. NOVEMBER 2024 | MÜNCHEN

CSL Vifor

Therapie der AAV in speziellen Situationen (DAH)

PD Dr. Björn Christian Frye
Freiburg

Therapie der AAV in speziellen Situationen

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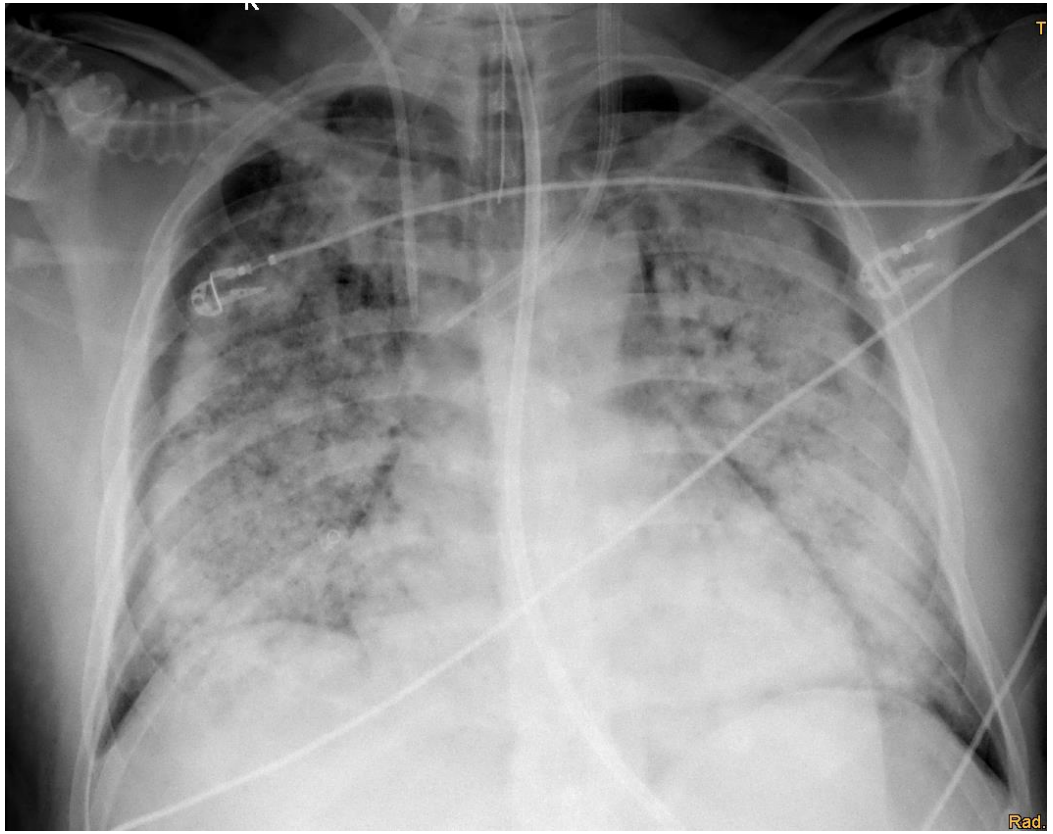
4. Dezember 2024

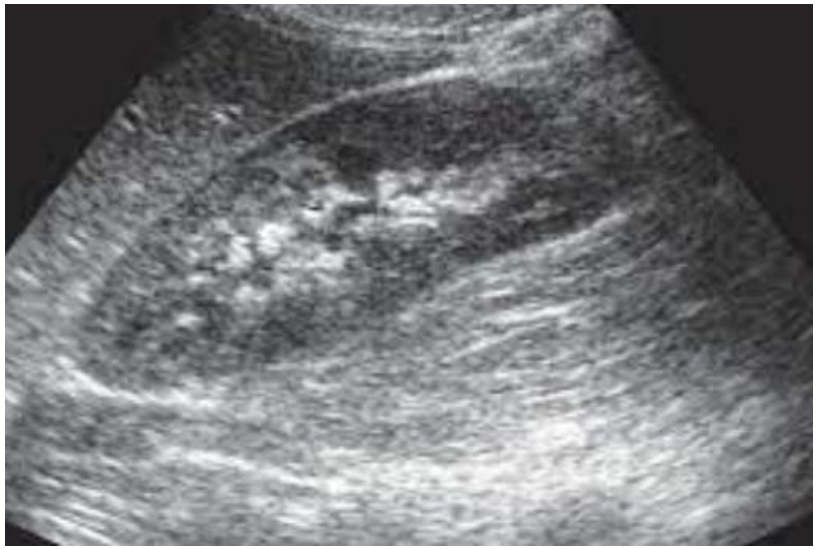
Interessenskonflikte

- **Beratungs- bzw. Gutachtertätigkeit:**
  - Advita Lifescience, Boehringer Ingelheim, Relief, Roche
- **Honorare:**
  - Advita Lifescience, Astra Zeneca, Boehringer Ingelheim, Novartis, Relief, Roche, Vifor
- **Finanzierung wissenschaftlicher Untersuchungen:**
  - Advita Lifescience, BMS

Therapie der AAV in speziellen Situationen



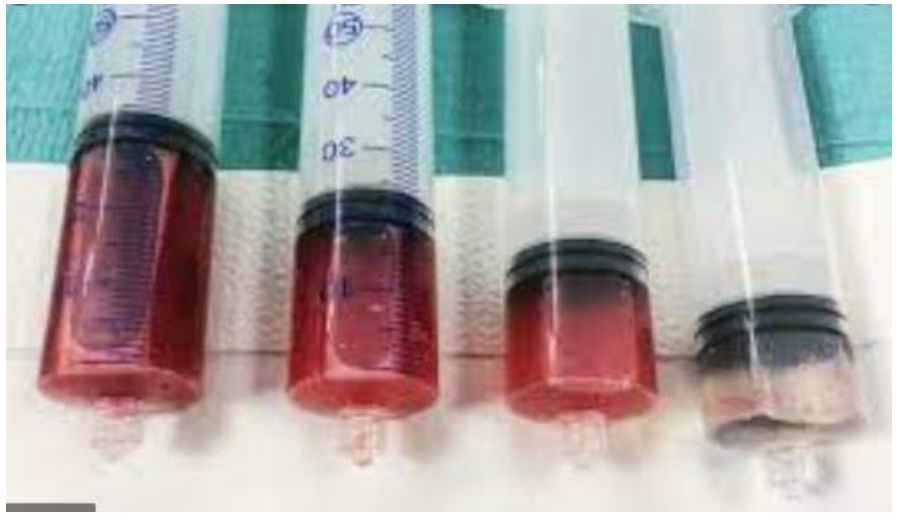
Alveoläre Hämorrhagie



Glomerulonephritis

Therapie der AAV in speziellen Situationen

Alveoläre Hämorrhagie

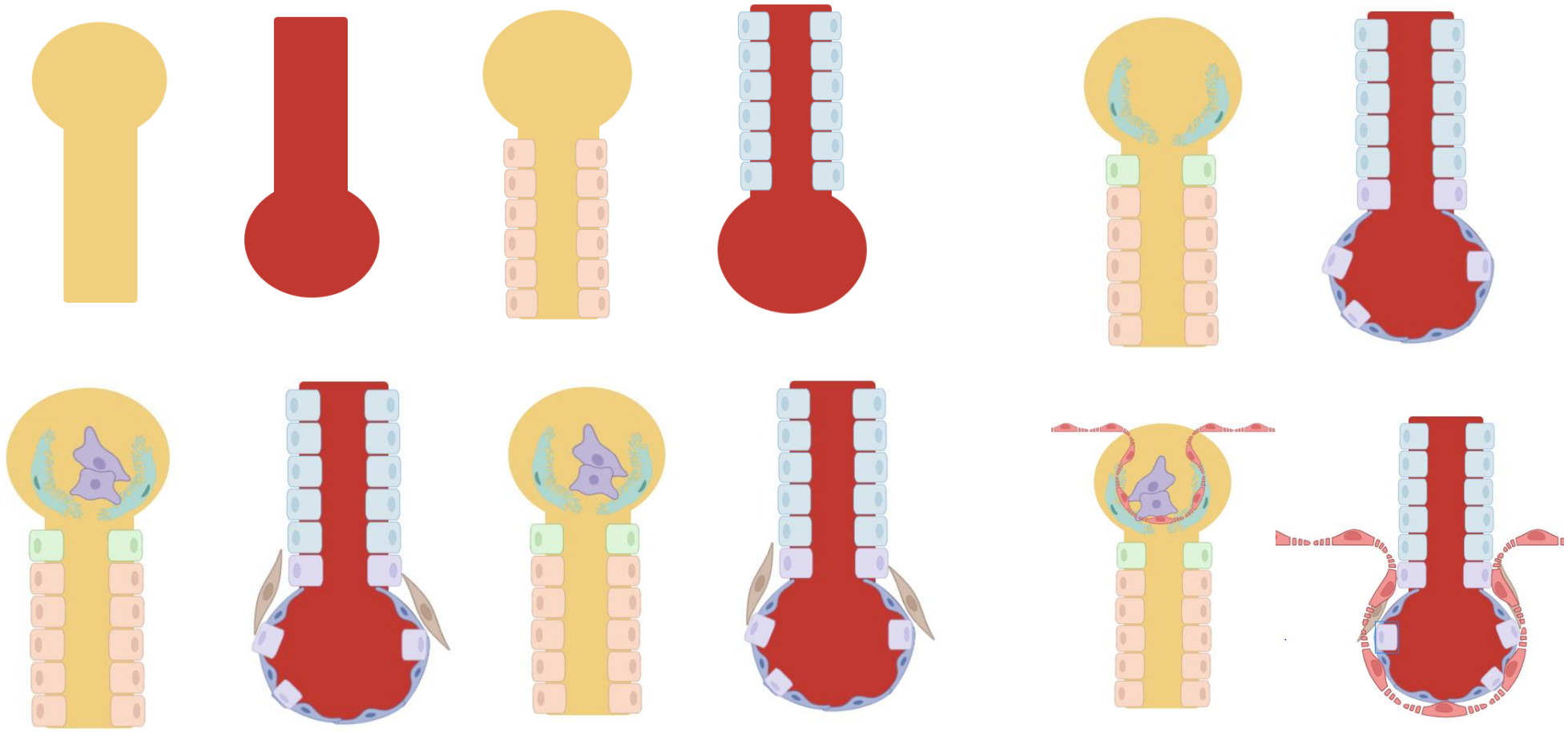


Glomerulonephritis

Befund:
Schnellbefund:
Leukos: -
Nitrit: -
Protein: +++
pH 6,5
Blut: +++

Mikroskopie (Angaben sind semiquantitativ oder pro Gesichtsfeld, 400 x):
RBC: 3-8/GF ; 24 % eumorph, 75 % dysmorph, 1 % Akanthozyten
WBC: 0-2/GF
Zylinder: vereinzelt feingranulierte Zylinder

Therapie der AAV in speziellen Situationen



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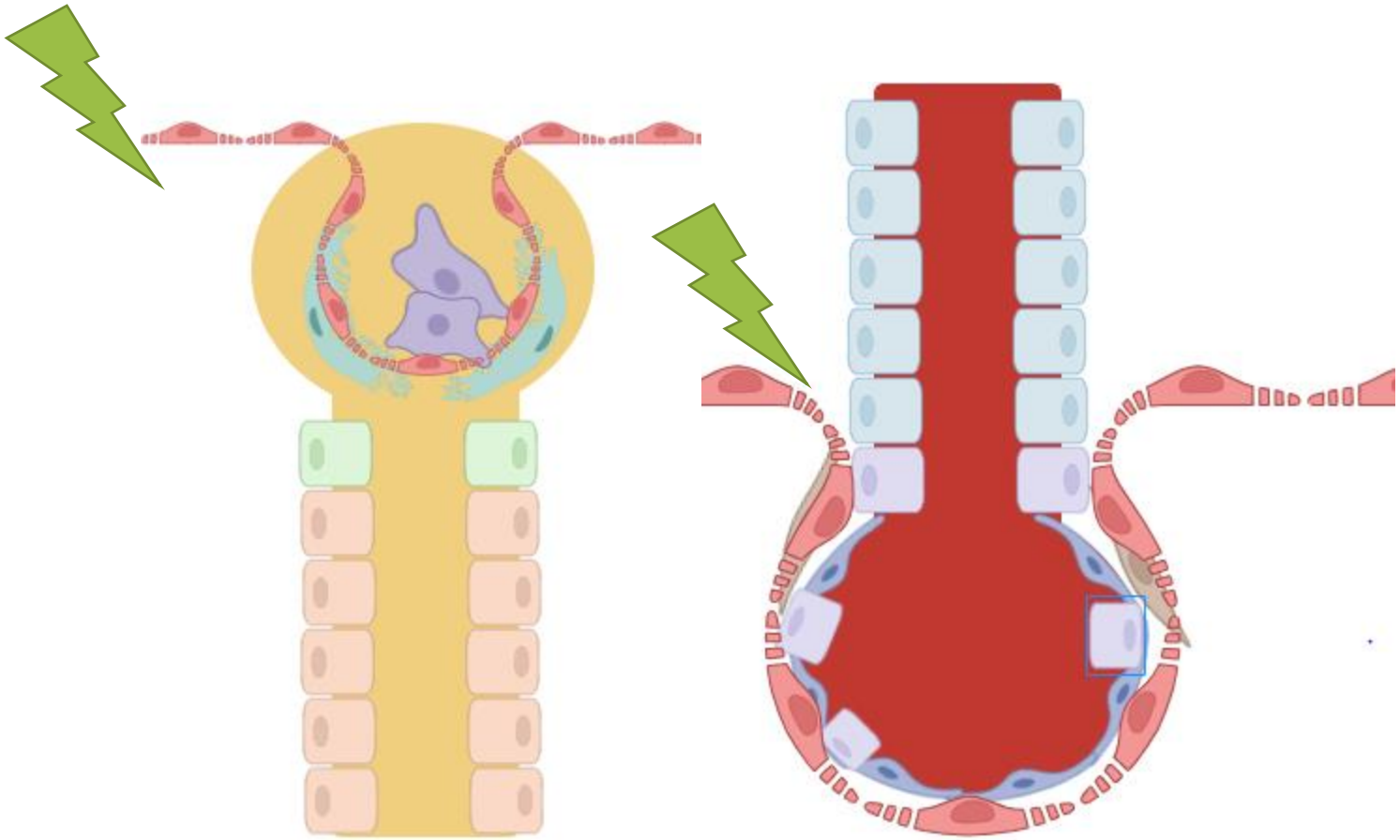

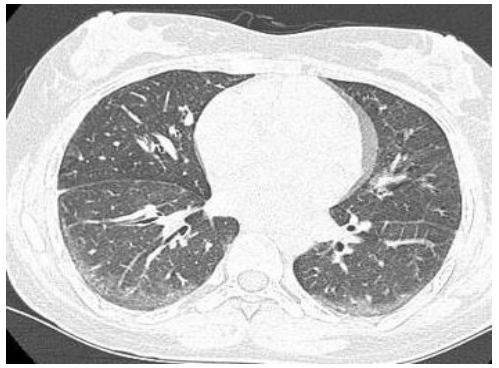

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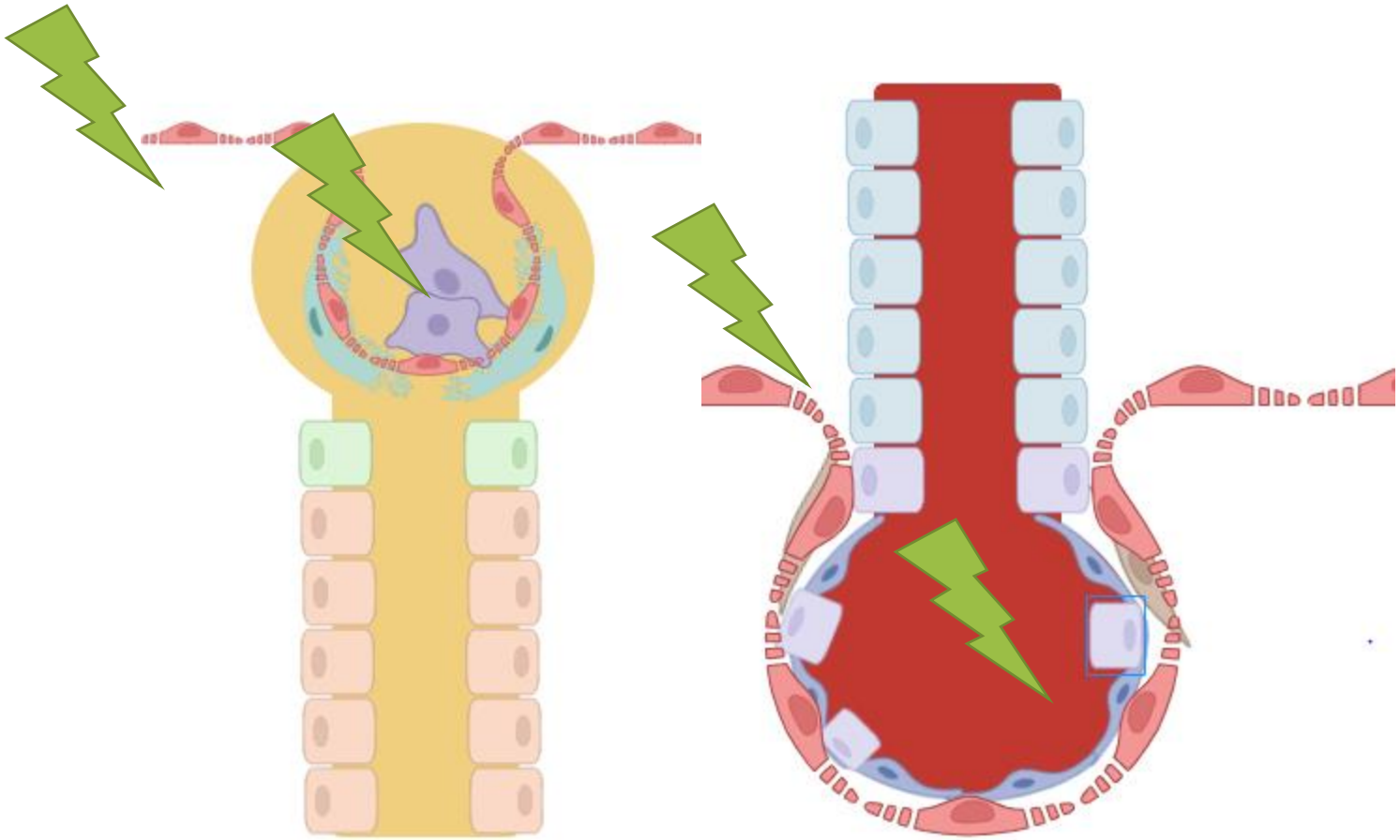

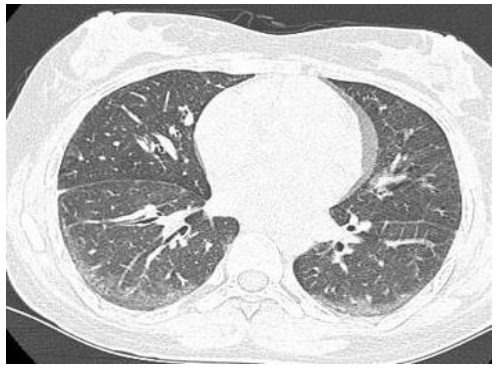

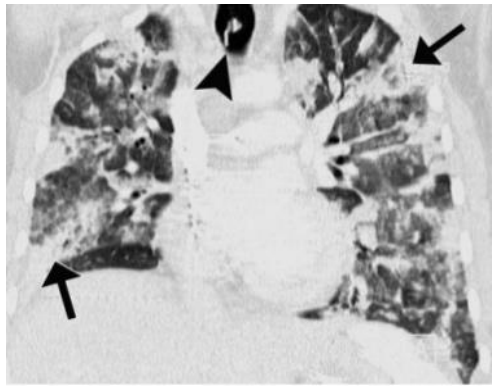

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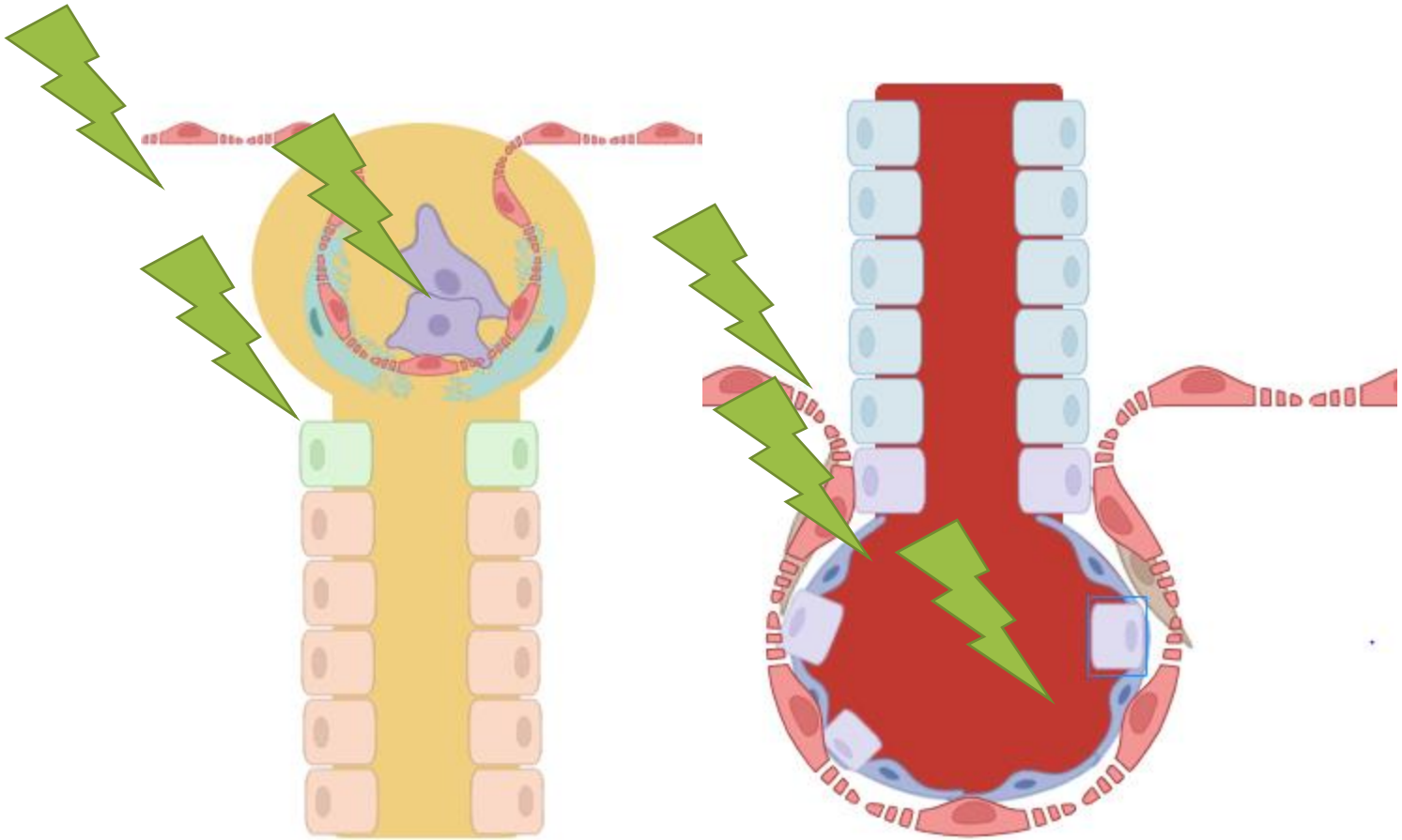
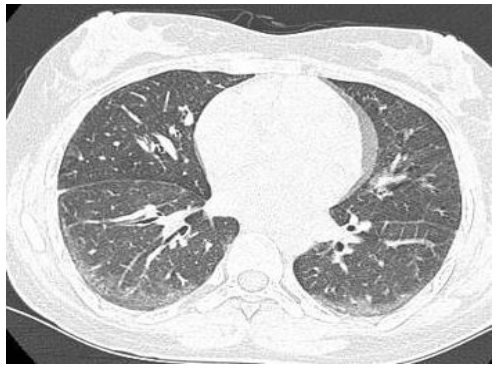
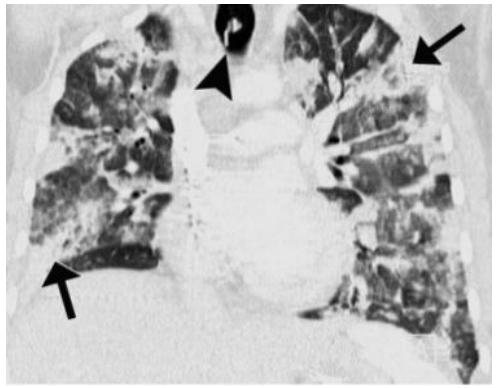
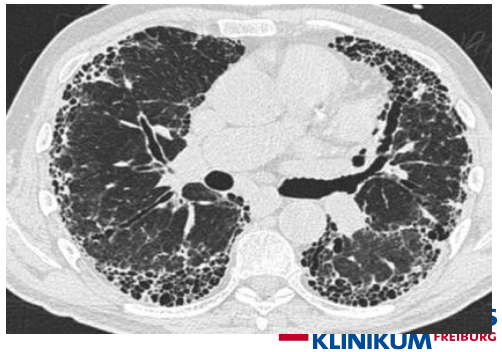








Therapie der AAV in speziellen Situationen

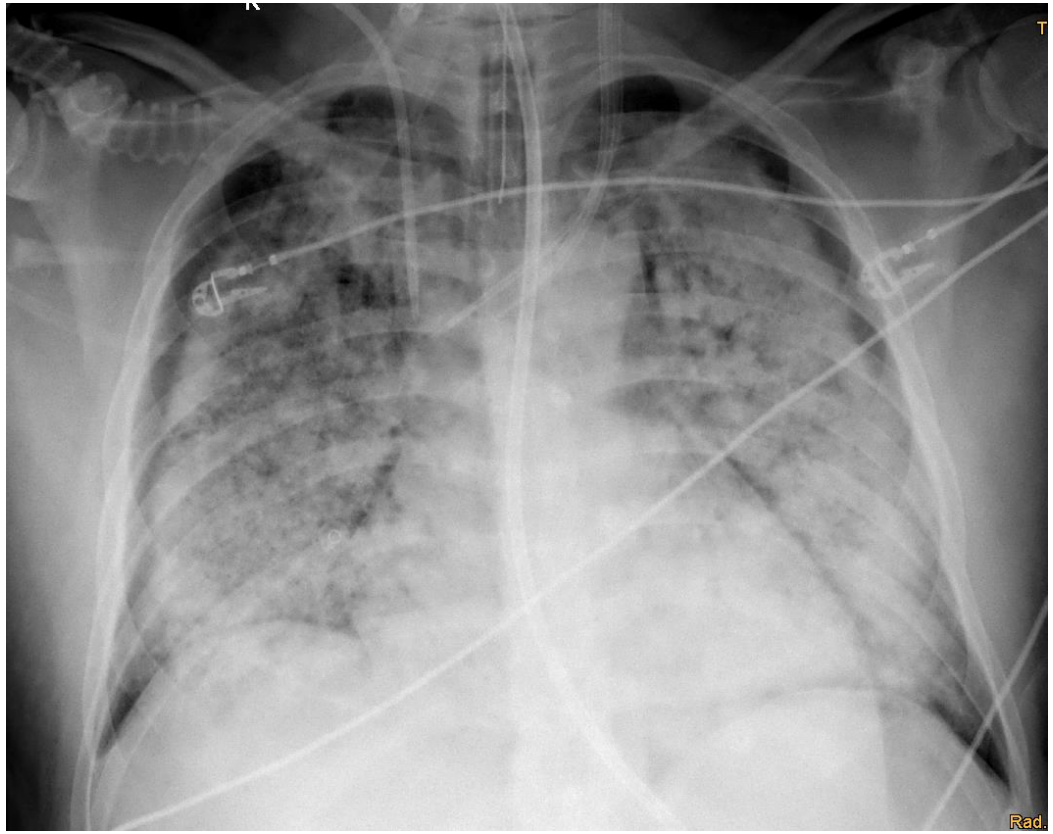
Alveoläre Hämorrhagie

- Kapillaritis
- Rheumatische Erkrankungen
- Medikamente
- Infektionen

Glomerulonephritis

- Kapillaritis
- Rheumatische Erkrankungen
- (Post-) Infektionen

Therapie der AAV in speziellen Situationen



IgG [g/l]	10,20
IgA [g/l]	1,15
IgM [g/l]	0,79
C3 [g/l]	1,09
C4 [g/l]	0,17
C3d [mg/l]	31,60
Phospholipid/ß2GP AK IgG [GPL-U/ml]	3
Phospholipid/ß2GP AK IgM [MPL-U/ml]	12
ß2-Glycoprotein-1 AK IgG [U/ml]	0
HEp-2 (Kern 1:50)	negativ
HEp-2 (Nukleolen 1:50)	negativ
HEp-2 (Chromosomen 1:50)	negativ
HEp-2 (Cytoplasma 1:50)	negativ
ANCA (EthOH-fixiert, IgG, 1:10)	+! c
ANCA (formaldehyd-fixiert, 1:10)	+!
Anti-Myeloperoxidase (Suchtest, Euroimmun)	negativ
Anti-Proteinase-3 (Suchtest, Euroimmun)	+++
Anti-glomeruläre Basalmembran (Suchtest, Euroimmun)	negativ
Anti-Myeloperoxidase (IgG) [E/ml]	< 2
Anti-MPO (Myeloperoxidase) [U/ml]	1
Anti-PR3 hs (Proteinase 3) [U/ml]	> 200

Therapie der AAV in speziellen Situationen

- Intensivmediziner*in
  → Steroid
- Pneumolog*in:
  → Cyclophosphamid
- Rheumatolog*in:
  → Avacopan
- Nephrolog*in:
  → Plasmapherese

Therapie der AAV in speziellen Situationen

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Und wie therapieren wir den Patienten?

Therapie der AAV in speziellen Situationen

- Steroide:

5.3	Empfehlung	Modifiziert Stand (2023)
Empfehlungsgrad **A für „Standarddosis vs. dosisreduziert", 0 für 0,5mg/kg KG Initialdosis**	Zur Remissionsinduktion bei organbedrohender GPA/MPA soll eine begleitende Glucocorticoidtherapie mit 50-75 mg/Tag Predniso(lo)n-äquivalent (je nach Körpergewicht) begonnen werden. Die GC-Therapie soll schrittweise reduziert und nach 15-16 Wochen auf 5-7,5 mg/Tag Predniso(lo)näquivalent reduziert werden (siehe Tabelle 13).	
	Bei schwerer organbedrohender GPA/MPA mit RPGN und/oder alveolärer Hämorrhagie kann initial eine i.v. Methylprednisolon (MP)-Pulstherapie, erwogen werden, an die sich eine orale GC-Therapie mit schrittweiser Dosisreduktion anschließt (siehe Tabelle 13).	
	Bei nicht-organbedrohender GPA/MPA kann bei Remissionsinduktion mit RTX eine niedrigere initiale GC-Dosis mit 0,5 mg Prednisolonäquivalent/kg/Tag erwogen werden.	

Holle, …., Moosig, Diagnostik und Therapie der ANCA-assoziierten Vaskulitiden S3-Leitlinie, AWMF

Therapie der AAV in speziellen Situationen

- Steroide:

5.3	Empfehlung
Empfehlungsgrad **A für „Standarddosis vs. dosisreduziert", 0 für 0,5mg/kg KG Initialdosis**	Zur Remissionsinduktion bei organbed begleitende Glucocorticoidtherapie m äquivalent (je nach Körpergewicht) be soll schrittweise reduziert und nach 1 Predniso(lo)näquivalent reduziert wer Bei schwerer organbedrohender GPA/ Hämorrhagie kann initial eine i.v. Met erwogen werden, an die sich eine oral Dosisreduktion anschließt (siehe Tabe Bei nicht-organbedrohender GPA/MP RTX eine niedrigere initiale GC-Dosis n Prednisolonäquivalent/kg/Tag erwogen werden.

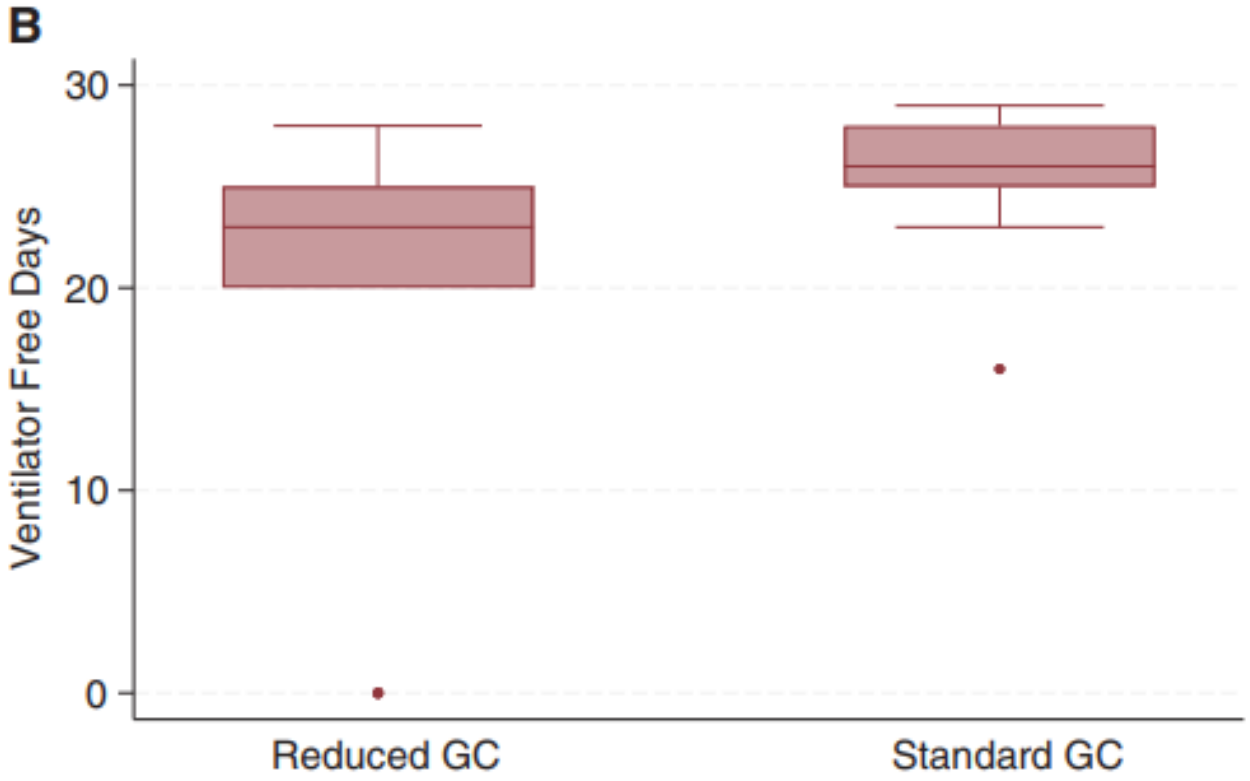
The two treatment groups received the same glucocorticoid regimen: one to three pulses of methylprednisolone (1000 mg each), followed by prednisone at a dose of 1 mg per kilogram per day. The dose was tapered so that by 5 months, all patients who had a remission without disease flares had discontinued glucocorticoids (see the Supplementary Appendix).

was made by the local investigator. With regard to the glucocorticoid regimens, all patients were treated with daily intravenous methylprednisolone for 1 to 3 days, for a maximum cumulative dose of 1 to 3 g, with the dosing decided by the local investigators. All patients then received oral

Steroid regimen. The steroid regimen was the same for both treatment groups. On days 1–3, 0.5 gm of methylprednisolone was given intravenously. From day 4 to day 14, prednisolone was administered orally at 1 mg/kg BW per day. By day 15, tapering of the steroid regimen was initiated, with a reduction of 10 mg/week. When a dosage of 30 mg/day was

Holle, …., Moosig, Diagnostik und Therapie der ANCA-assoziierten Vaskulitiden S3-Leitlinie, AWMF

Therapie der AAV in speziellen Situationen



Fussner, Flores-Suárez et al., AJRCCM 2024

Therapie der AAV in speziellen Situationen

- Cyclophosphamid:

5.1	Empfehlung	Modifiziert Stand 2023
Empfehlungsgrad **A**	Zur Remissionsinduktion bei organ- oder lebensbedrohlicher GPA oder MPA soll eine Kombinationstherapie aus Glucocorticoiden (GC) und Rituximab (RTX) oder GC und Cyclophosphamid (CYC) erfolgen. Bei Patienten mit rezidivierendem Verlauf soll vorzugsweise GC und RTX eingesetzt werden.	

Holle, …., Moosig, Diagnostik und Therapie der ANCA-assoziierten Vaskulitiden S3-Leitlinie, AWMF

Therapie der AAV in speziellen Situationen

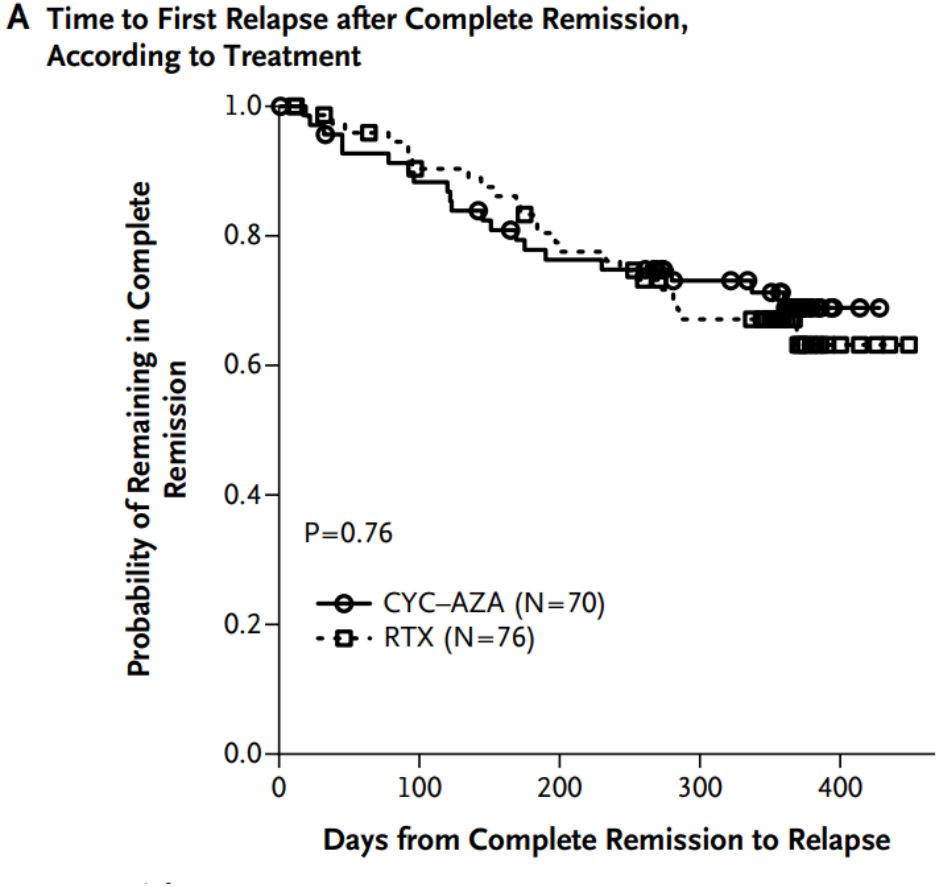
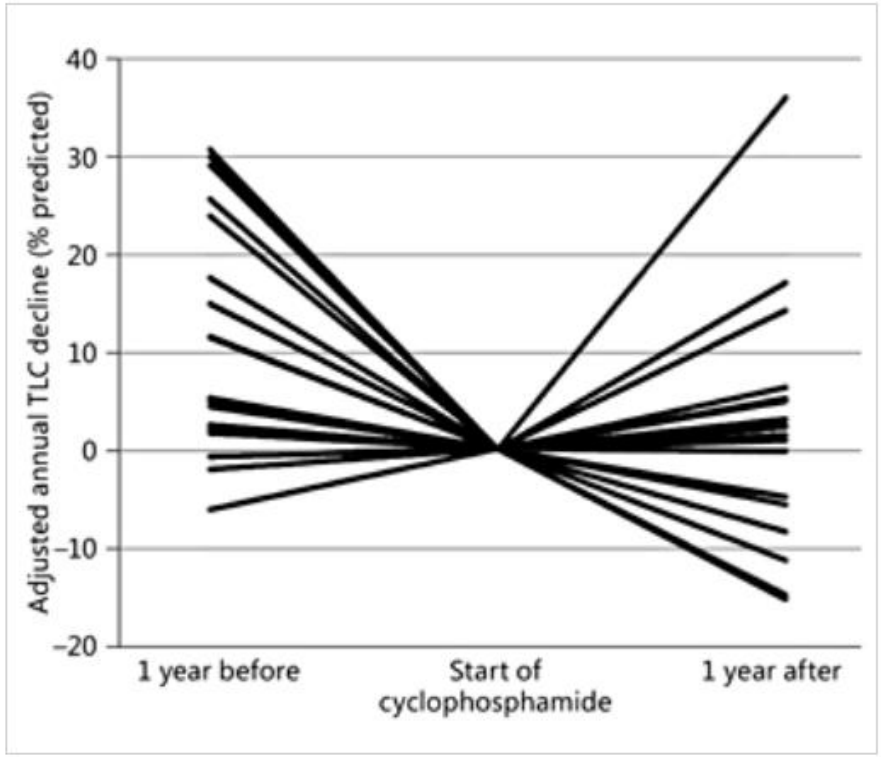


Fig. 1



Specks, Merkel et al., NEJM 2013, Schupp, Köhler et al., Respiration 2016

Therapie der AAV in speziellen Situationen

	Cyclophosphamide (n = 31)	Rituximab (n = 37)	*P*
Characteristic			
Age, median (IQR) years	67 (56–74)	58 (42–67)	0.01
Sex, no. (%) male	17 (55)	20 (54)	0.94
GPA, no. (%)	16 (52)	27 (73)	0.06
MPA, no. (%)	15 (48)	10 (27)	0.08
BVAS/WG score, median (IQR)	10 (9–13)	10 (8–12)	0.38
Spo_2:Fio_2 at presentation, median (IQR)	288 (155–442)	205 (115–452)	0.49
APACHE III score, median (IQR)†	50 (33–101)	50 (31–72)	0.59
Mechanical ventilation, no. (%)	13 (42)	18 (49)	0.63
Active renal disease, no. (%)	14 (45)	16 (43)	0.87
Requiring new renal replacement therapy, no. (%)	7 (23)	3 (8)	0.16
Outcome			
Hospital mortality, no. (%)	4 (13)	2 (5.4)	0.40
Length of hospital stay, median (IQR) days	9.8 (6–15)	9.2 (4.9–17)	0.90
Length of ICU stay, median (IQR) days	8 (2–16)	5.2 (3–8.6)	0.56
Duration of mechanical ventilation, median (IQR) days	4.2 (1.2–6.4)	3.6 (0.8–5)	0.77
Complete remission at 6 months, no. (%)	21 (68)	33 (89)	0.02

Cartin-Ceba, Diaz-Caballero et al., Arthritis Rheumatol 2016

Therapie der AAV in speziellen Situationen

- Plasmapherese:

5.5	Empfehlung	Modifiziert Stand (2023)
Empfehlungsgrad **0**	Eine additive Plasmaaustauschbehandlung (PLEX) in Kombination mit GC und CYC oder RTX kann für ausgewählte GPA-/MPA-Patienten mit einer aktiven Nierenbeteiligung und einem Kreatininwert von >300 µmol/l (3,4 mg/dl) erwogen werden. Eine additive PLEX-Behandlung zur Therapie der alveolären Hämorrhagie sollte nicht standardmäßig erfolgen.	

Holle, …., Moosig, Diagnostik und Therapie der ANCA-assoziierten Vaskulitiden S3-Leitlinie, AWMF

Therapie der AAV in speziellen Situationen

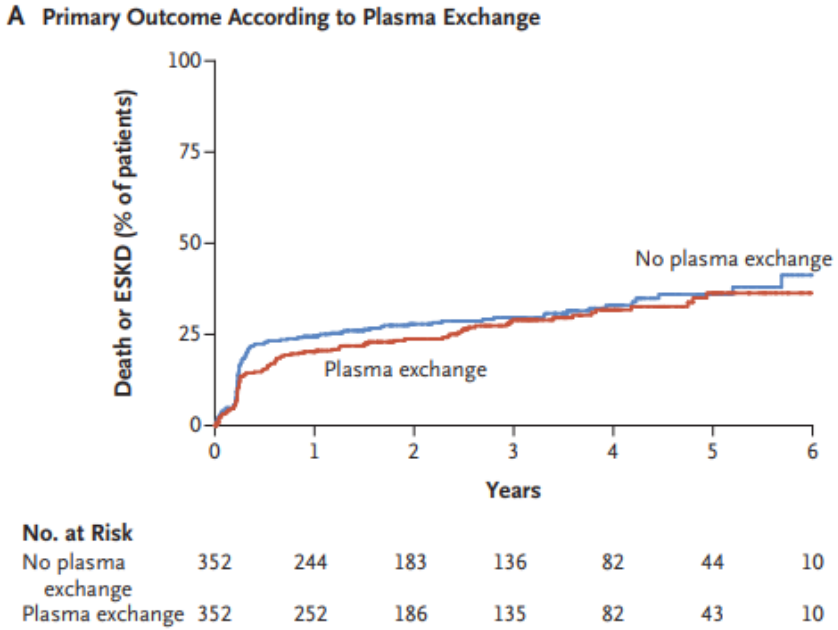
Table 4. Clinical characteristics and outcomes in patients with DAH secondary AAV who were treated with plasma exchange and those who were not treated with plasma exchange*

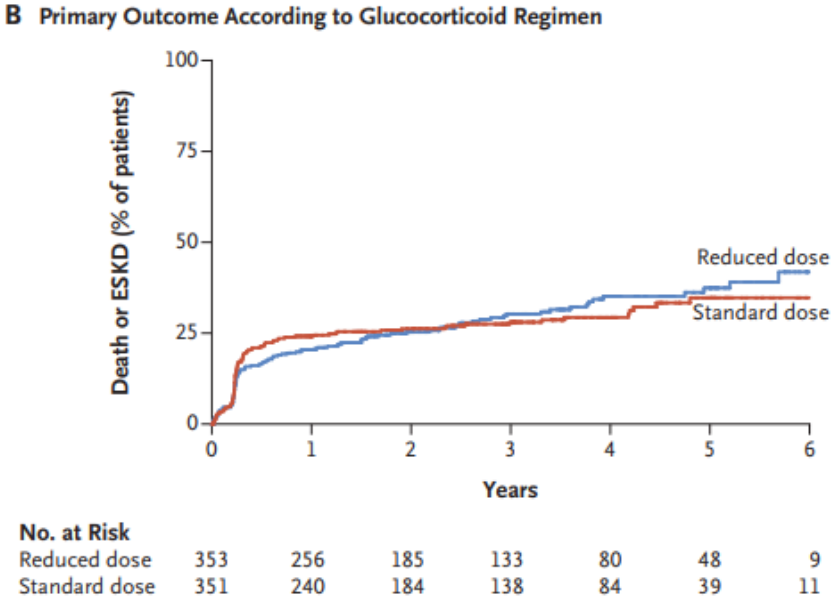
	No plasma exchange (n = 41)	Plasma exchange (n = 32)	*P*
Characteristic			
Age, median (IQR) years	63 (48–73)	62 (49–72)	0.70
Sex, no. (%) male	22 (54)	19 (59)	0.62
GPA, no. (%)	27 (66)	17 (53)	0.27
MPA, no. (%)	14 (34)	15 (47)	0.33
BVAS/WG score, median (IQR)	10 (8–12)	12 (10–18)	0.002
Spo_2:Fio_2 at presentation, median (IQR)	292 (158–457)	167 (96–395)	0.03
APACHE III score, median (IQR)†	50 (41–54)	51 (29–61)	0.76
Mechanical ventilation, no. (%)	13 (32)	21 (66)	0.003
Active renal disease, no. (%)	14 (34)	19 (59)	0.03
Requiring new renal replacement therapy, no. (%)	3 (7)	9 (28)	0.01
Hemosiderin-laden macrophages, median (IQR) %	60 (21–93)	55 (28–89)	0.62
Neutrophils in BAL fluid, median (IQR) %	21 (7–66)	32 (15–66)	0.34
Creatinine, median (IQR) mg/dl	1.1 (1–2.6)	1.9 (0.9–4.4)	0.20
GFR, median (IQR) ml/minute	52 (26–75)	26 (11–55)	0.12
Outcome			
Hospital mortality, no. (%)	3 (7)	5 (16)	0.28
Length of hospital stay, median (IQR) days	7.7 (4–15.3)	10.8 (7–18)	0.06
Length of ICU stay, median (IQR) days	6.1 (2–11)	6 (4.2–9.1)	0.83
Duration of mechanical ventilation, median (IQR) days	2.8 (0.7–5.1)	3.7 (2.4–5.4)	0.48
Complete remission at 6 months, no. (%)	32 (78)	23 (72)	0.54

* Spo_2 = oxygen saturation measured by pulse oximetry; Fio_2 = fraction of inspired oxygen (see Table 1 for other definitions).
† For 41 patients admitted to the ICU.

Cartin-Ceba, Diaz-Caballero et al., Arthritis Rheumatol 2016

Therapie der AAV in speziellen Situationen

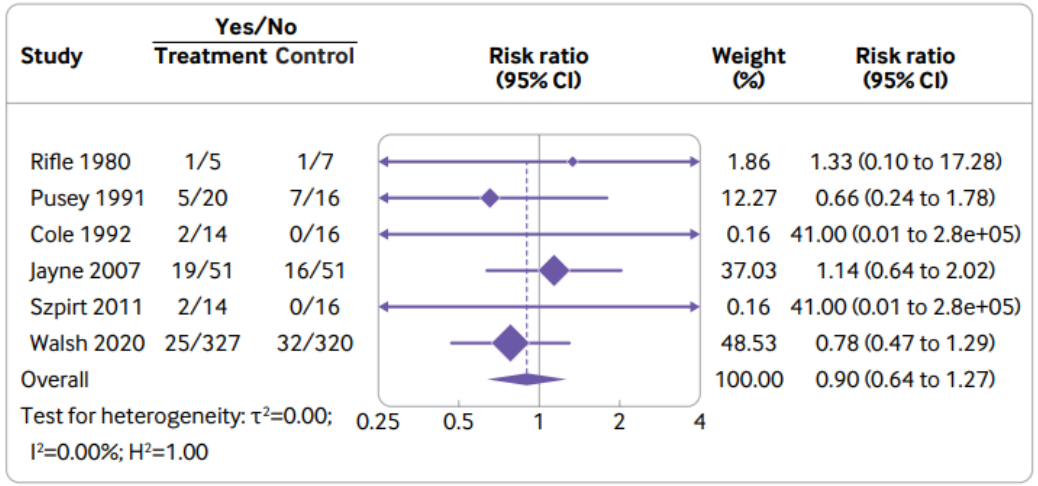




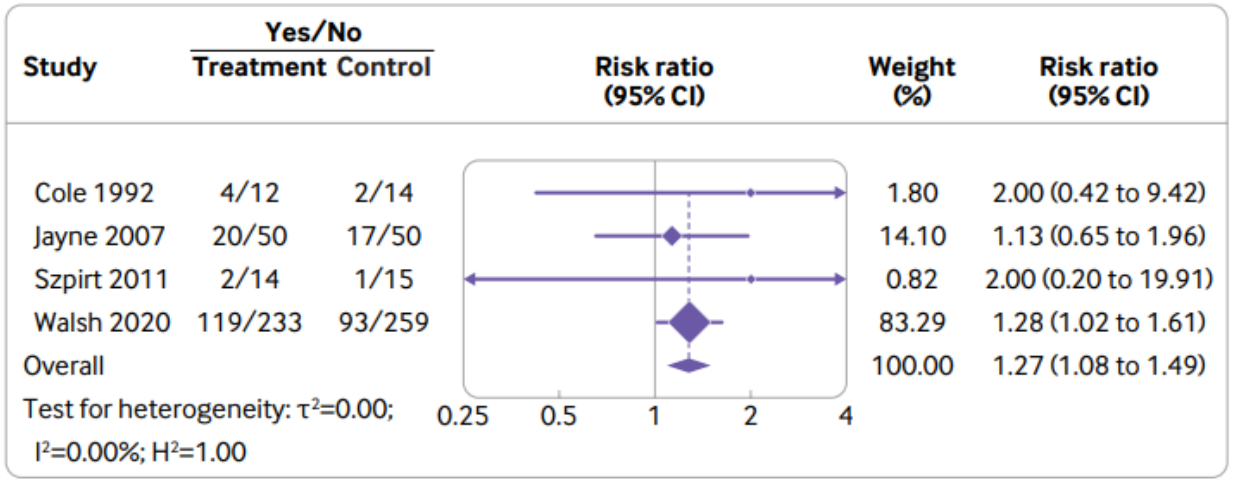
Walsh, Merkel et al., NEJM 2020

Therapie der AAV in speziellen Situationen

Mortalität

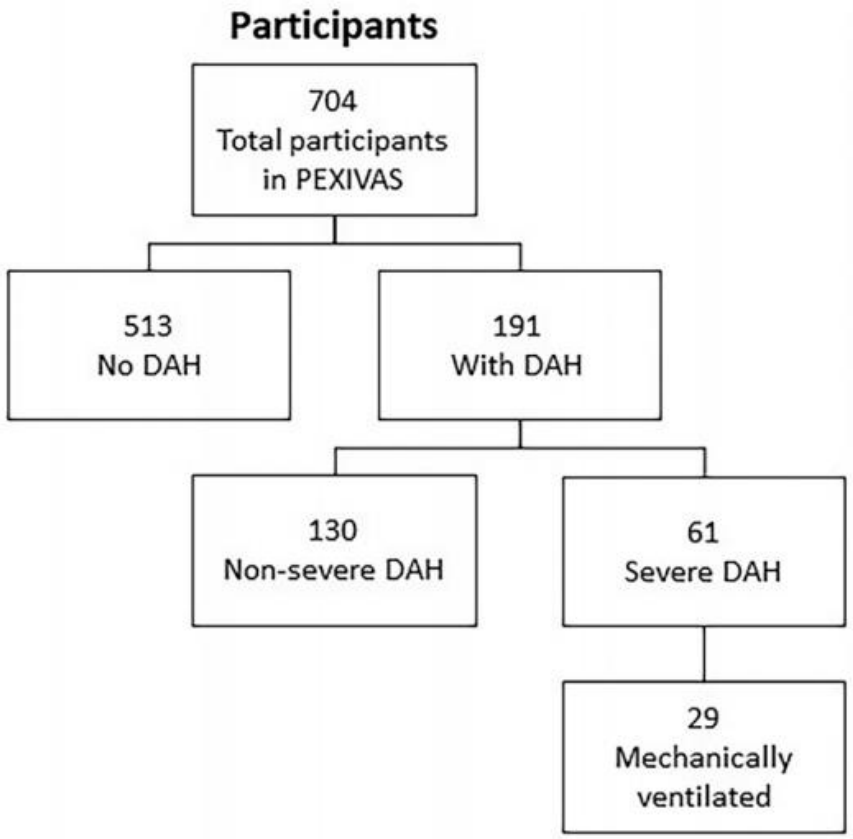


Infektionen



Walsh, Collister et al., BMJ 2020

Therapie der AAV in speziellen Situationen



Treatment Groups

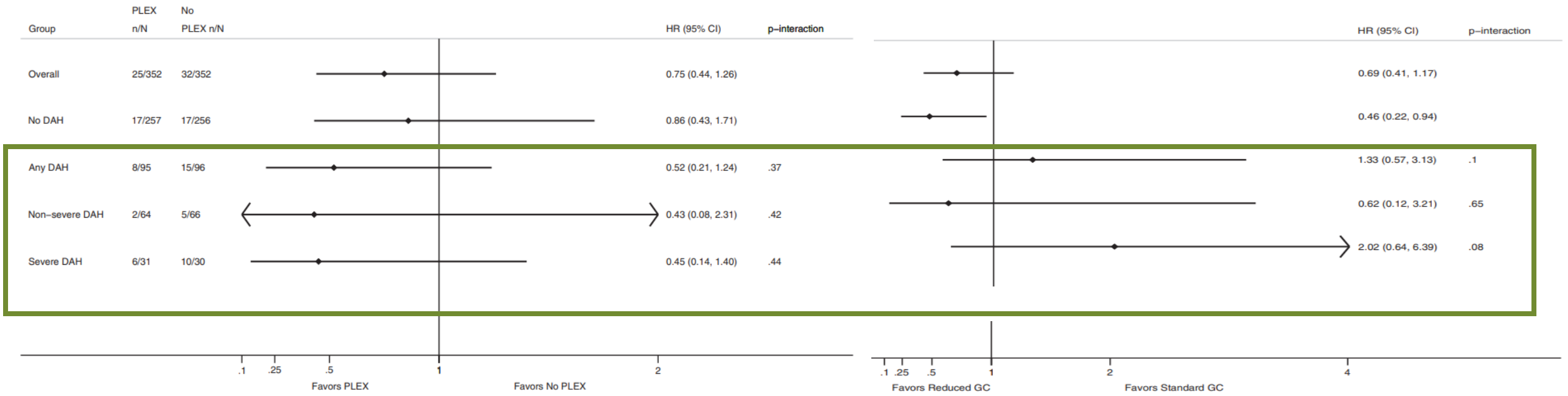
PLEX + Standard GC		PLEX + Reduced GC		No PLEX + Standard GC		No PLEX + Reduced GC	
N = 176		N = 176		N = 175		N = 177	
No DAH	129	No DAH	128	No DAH	127	No DAH	129
Non-severe DAH	32	Non-severe DAH	32	Non-severe DAH	33	Non-severe DAH	33
Severe DAH	15	Severe DAH	16	Severe DAH	15	Severe DAH	15
Ventilated	7	Ventilated	8	Ventilated	6	Ventilated	8

	Any DAH (*n* = 191)	No DAH (*n* = 513)	*P* Value
Age, yr, mean (SD)	61.1 (15.4)	63.9 (13.4)	0.018
Female	78 (40.8)	229 (44.6)	0.37
ANCA			<0.001
Anti-PR3	99 (51.8)	187 (36.5)	
Anti-MPO	92 (48.2)	326 (63.6)	
New diagnosis of AAV	164 (85.8)	477 (93.0)	0.004
BVAS/WG	11 (9–13)	7 (6–9)	<0.001
Creatinine, μmol/L	184 (115–320)	304 (212–442)	<0.001
Dialysis at baseline	48 (25.3)	92 (18.0)	0.033
Randomized to PLEX	95 (49.7)	257 (50.1)	0.93
Randomized to reduced GC	96 (50.3)	257 (50.1)	0.97
Immunosuppression			<0.001
Oral cyclophosphamide	44 (23.0)	197 (38.4)	
Intravenous cyclophosphamide	107 (56.0)	247 (48.2)	
Rituximab	40 (20.9)	69 (13.4)	

Fussner, Flores-Suárez et al., AJRCCM 2024

Therapie der AAV in speziellen Situationen

1-Jahres-Überleben



+ PLEX - PLEX Reduced GC Standard GC

Fussner, Flores-Suárez et al., AJRCCM 2024

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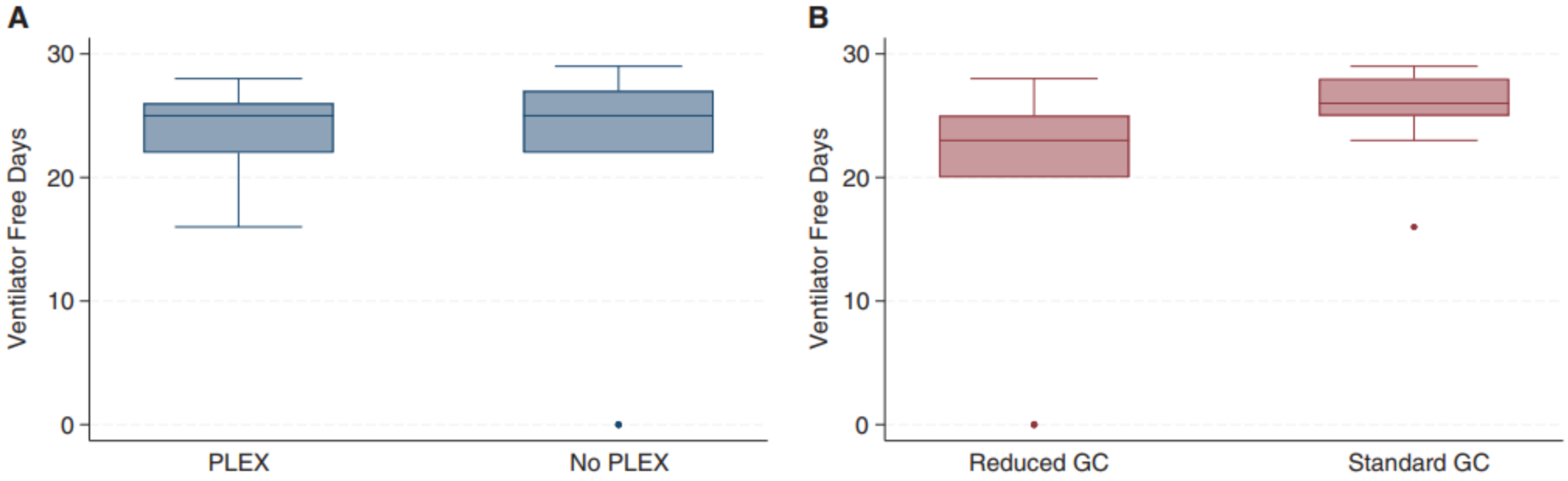
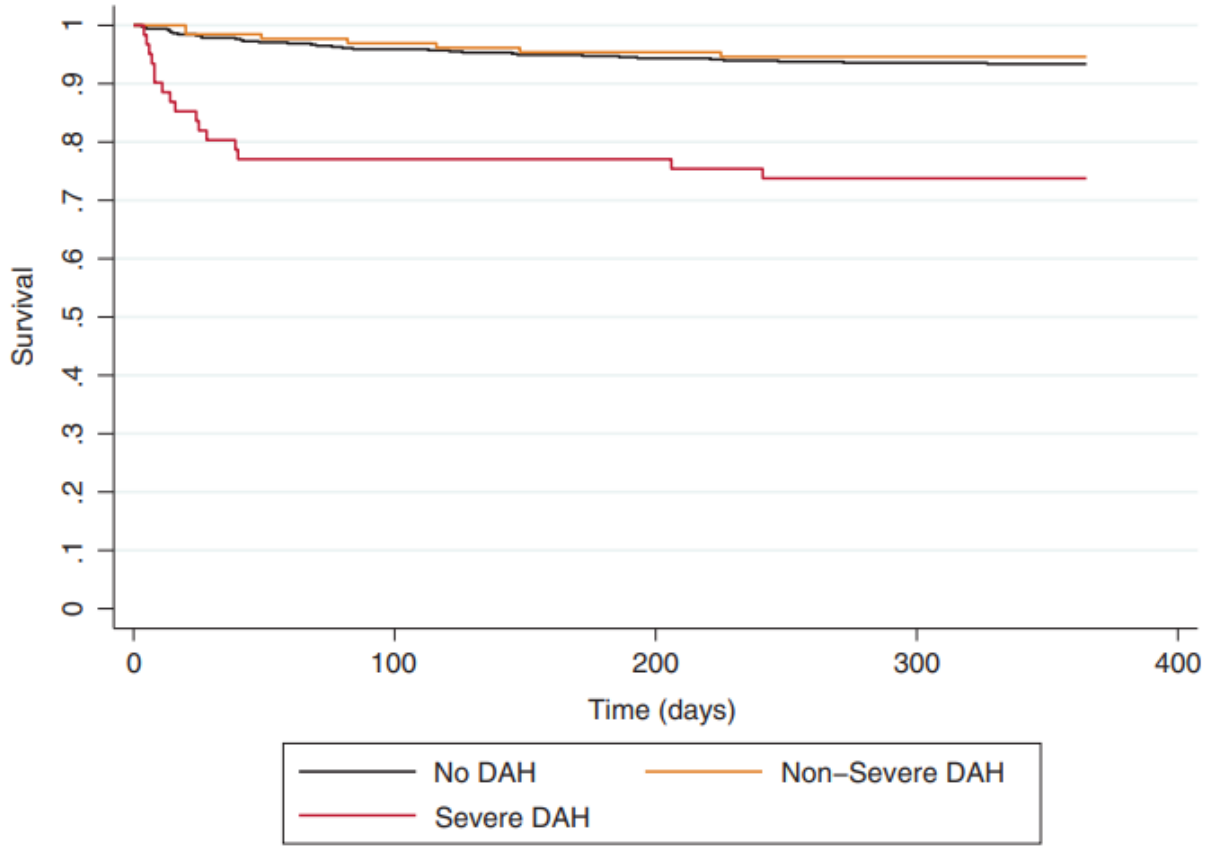


Fussner, Flores-Suárez et al., AJRCCM 2024

Therapie der AAV in speziellen Situationen



Fussner, Flores-Suárez et al., AJRCCM 2024

Therapie der AAV in speziellen Situationen

- Avacopan:

5.4	Empfehlung	Neu Stand (2023)
Empfehlungsgrad **0**	Der Einsatz von Avacopan kann zusätzlich zu einer Remissionsinduktion mit CYC oder RTX erwogen werden, um die kumulative GC-Dosis zu reduzieren.	

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Therapie der AAV in speziellen Situationen

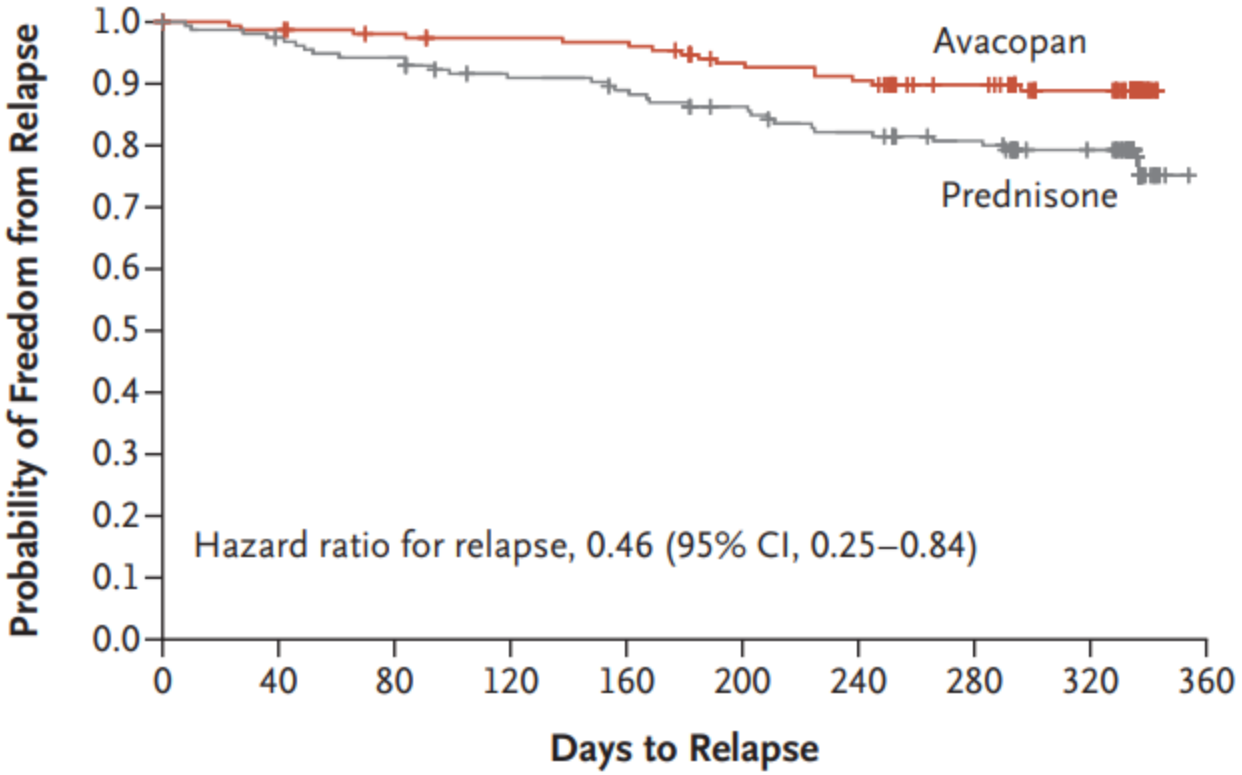
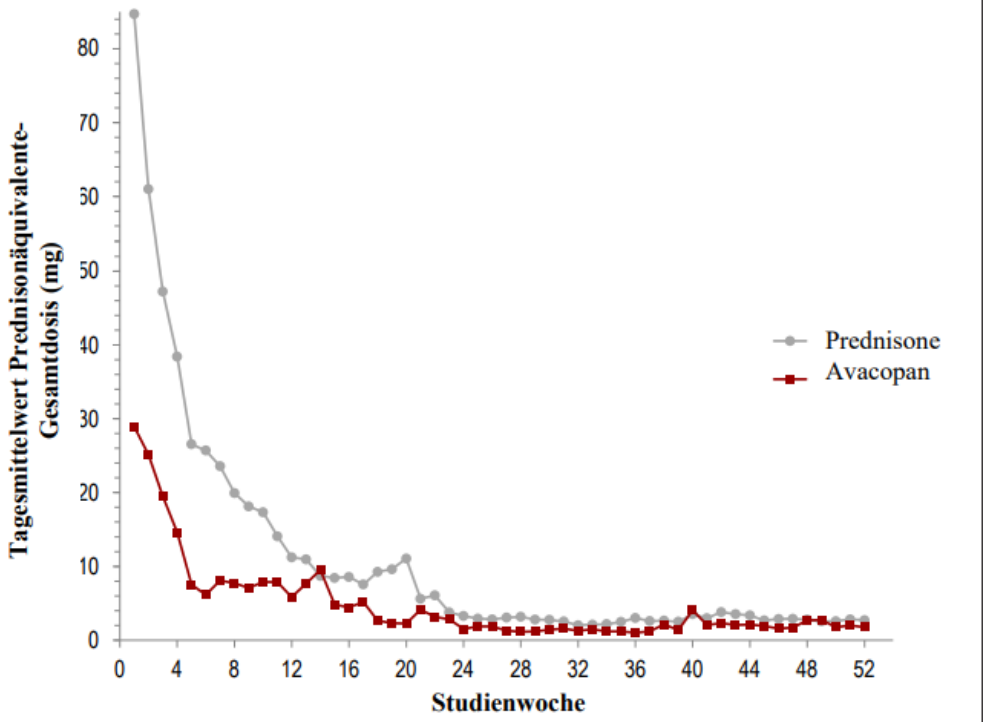


Abbildung 2: Durchschnittliche tägliche Gesamtdosis von Prednison-äquivalent Glukokortikoid pro Patient pro Studienwoche in der ADVOCATE-Studie (Intent-to-treat-Population)



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Therapie der AAV in speziellen Situationen

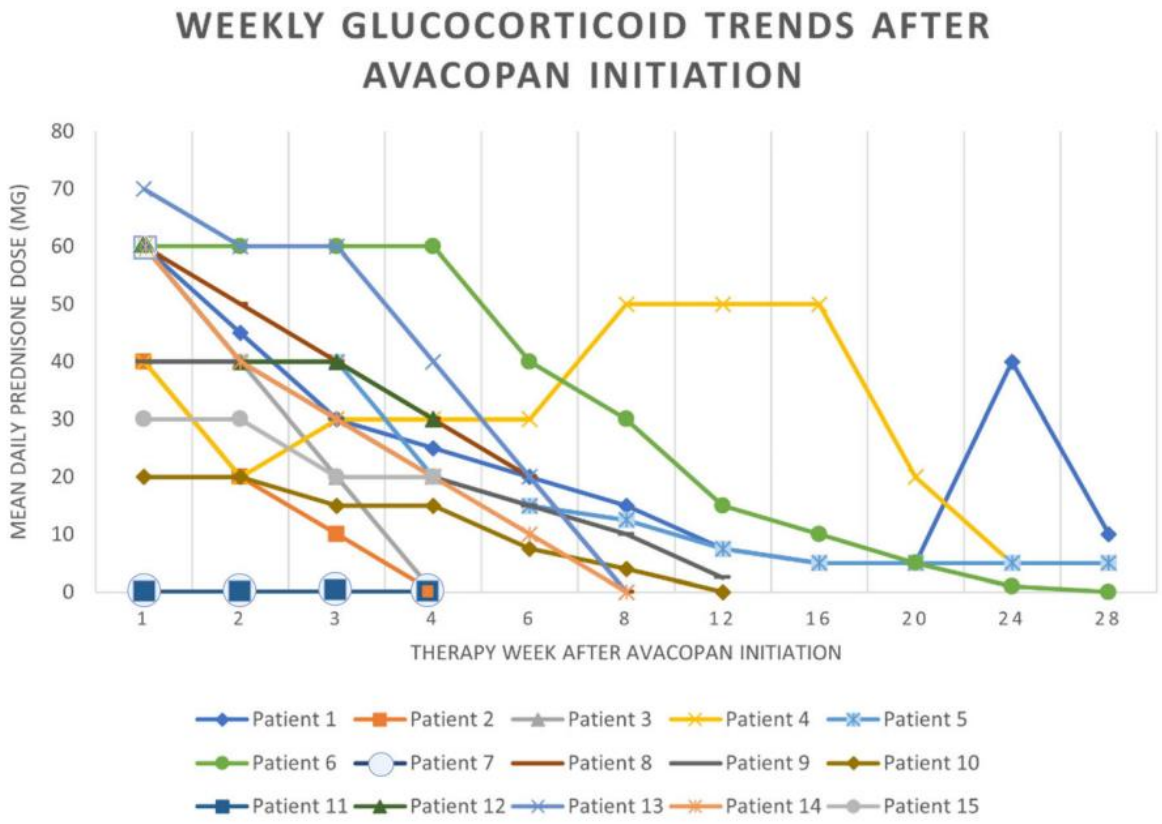

Table 2. Clinical characteristics and outcomes stratified by time of administration of avacopan during presentation with diffuse alveolar hemorrhage*

	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

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Therapie der AAV in speziellen Situationen

- Intensivmediziner*in
  → Steroid
- Pneumolog*in:
  → Cyclophosphamid
- Rheumatolog*in:
  → Avacopan
- Nephrolog*in:
  → Plasmapherese

Methylprednisolon

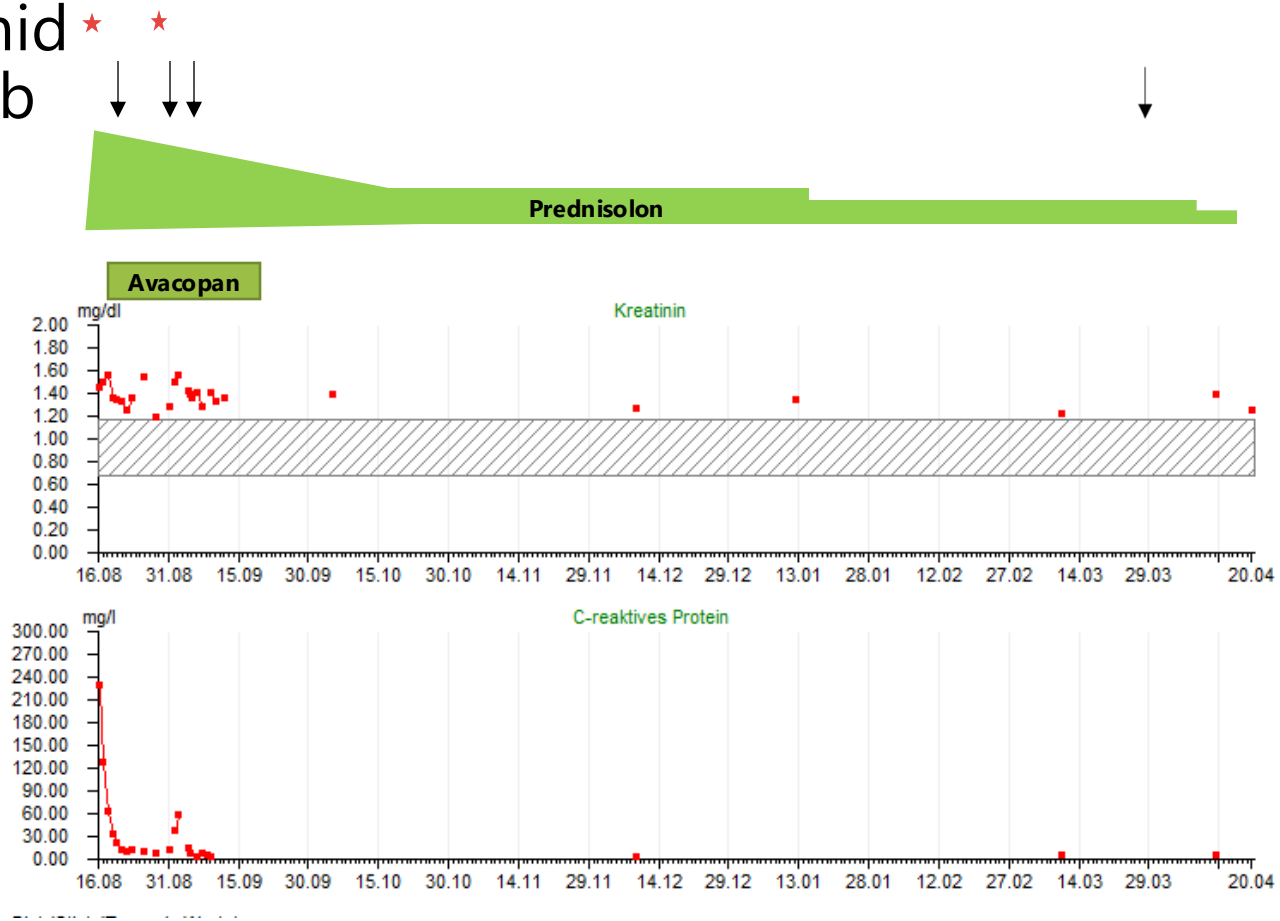
Cyclophosphamid

Avacopan nach Extubation

Rituximab

Therapie der AAV in speziellen Situationen

Cyclophosphamid
Rituximab



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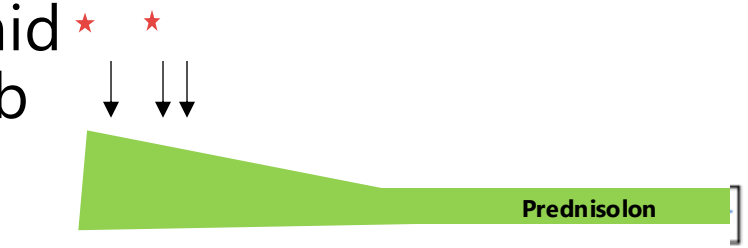
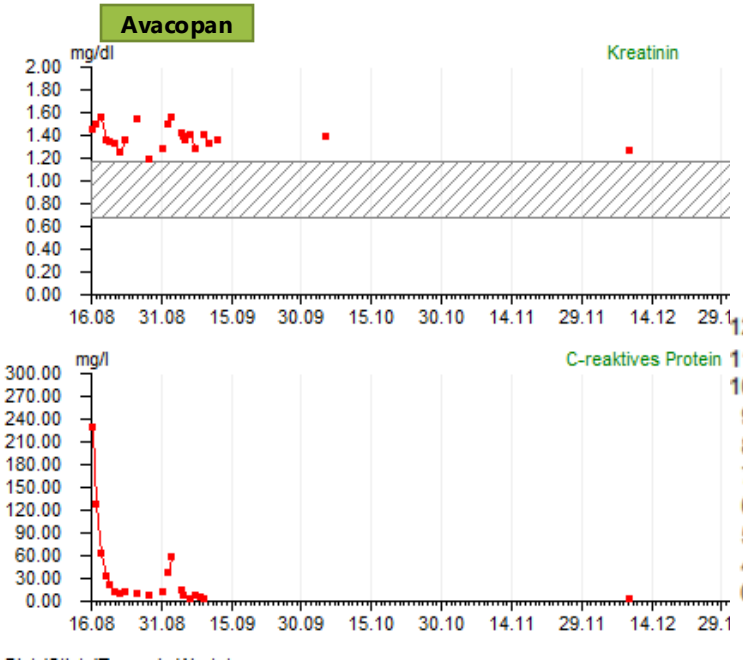
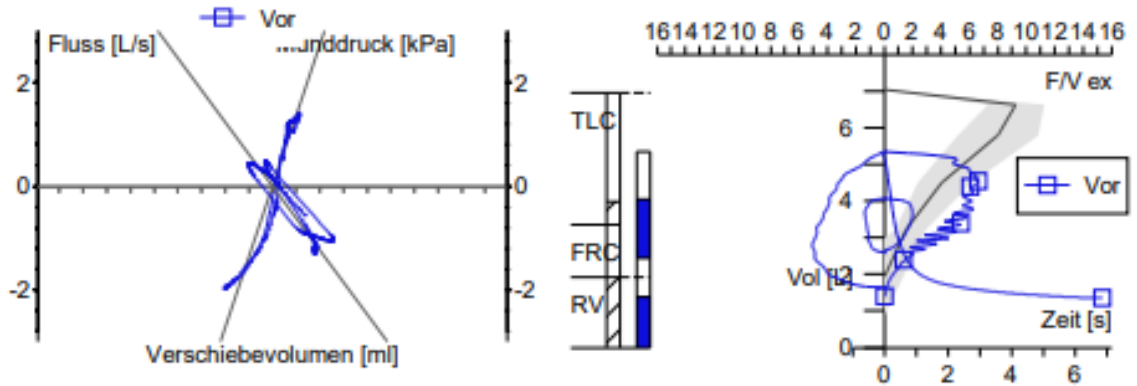
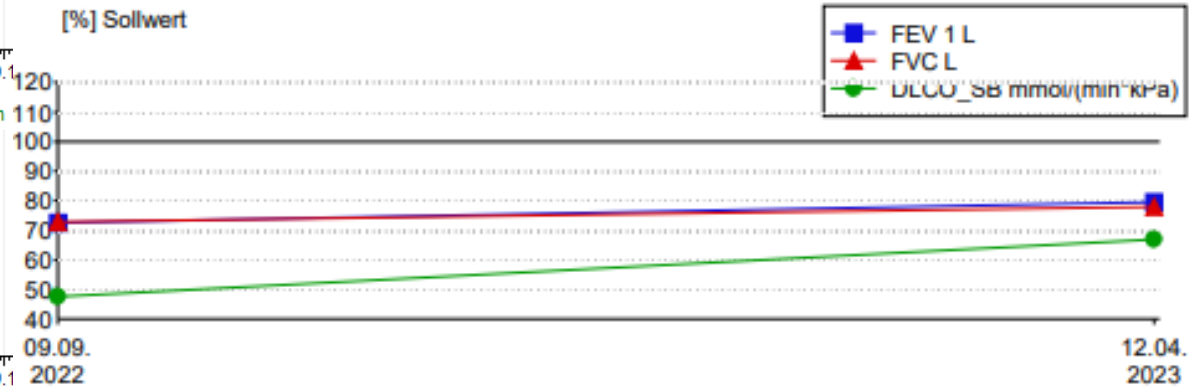
Therapie der AAV in speziellen Situationen

Cyclophosphamid
Rituximab









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Therapie der AAV in speziellen Situationen

Diffuse alveoläre Hämorrhagie

- Bronchoskopische Diagnose
- Nicht nur Vaskulitis
- Meist schlecht

- Hit hard and early
- Immunsuppression (Glukokortikoide) schnell reduzieren
- Daten zur besten Therapie weiter unklar

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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.