

2. DACH ANCA VASKULITIS FORUM 2024

22. & 23. NOVEMBER 2024 | MÜNCHEN

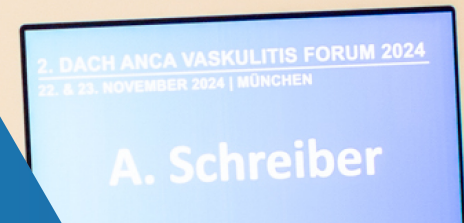
CSL Vifor

**Neue innovative
Therapieoptionen
(CAR-T, Complement)**

Prof. Dr. Adrian Schreiber

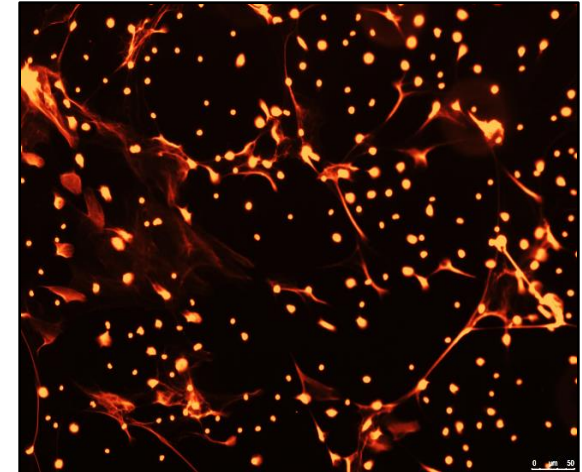
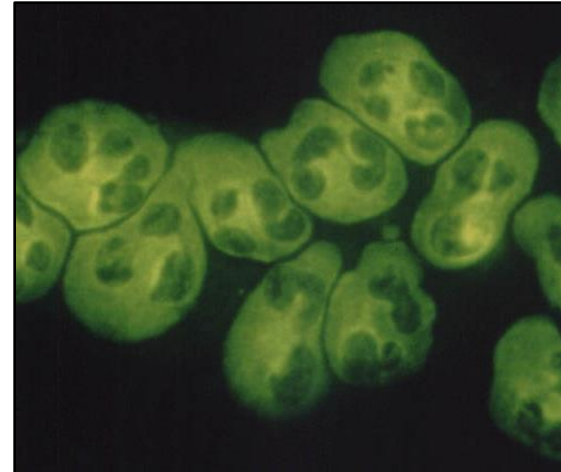
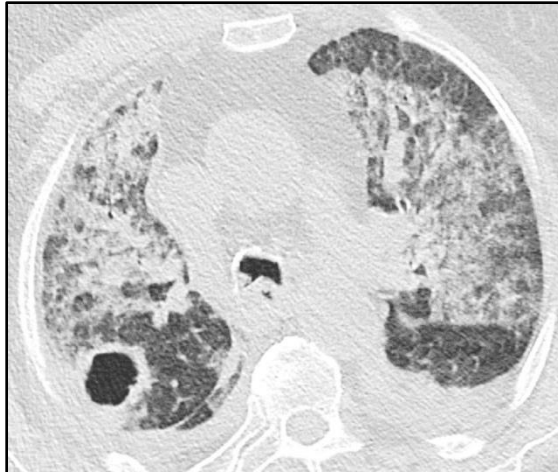
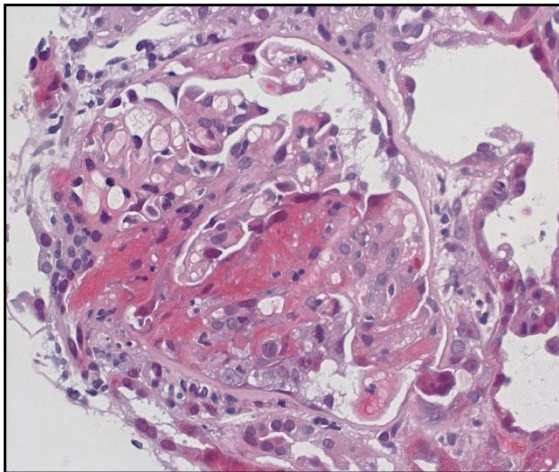


Berlin



Neue innovative Therapieoptionen (CAR-T und Anderes)

Adrian Schreiber



Im Rahmen dieses Vortrages bestehen keine Interessenskonflikte

Referenten-/Beraterhonorare/Forschungsunterstützung

Vifor CSL, Novartis, Alexion, Hansa Biopharm, Otsuka, Stadapharm, Astra Zeneca

PI in klinischen Studien bei AAV

Therapierefraktärität-Induktionstherapie

The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE

Rituximab versus Cyclophosphamide for ANCA-Associated Vasculitis

John H. Stone, M.D., M.P.H., Peter A. Merkel, M.D., M.P.H., Robert Spiera, M.D., Philip Seo, M.D., M.H.S., Carol A. Langford, M.D., M.H.S., Gary S. Hoffman, M.D., Cees G.M. Kallenberg, M.D., Ph.D., E. William St. Clair, M.D., Anthony Turkiewicz, M.D., Nadia K. Tchau, M.D., Lisa Webber, R.N., Linna Ding, M.D., Ph.D., Lourdes P. Sejsmundo, R.N., B.S.N., Kathleen Mieras, C.C.R.P., David Weitzenkamp, Ph.D., David Ikle, Ph.D., Vicki Seyfert-Margolis, Ph.D., Mark Mueller, B.S., C.C.R.P., Paul Brunetta, M.D., Nancy B. Allen, M.D., Fernando C. Fervenza, M.D., Ph.D., Duvuru Geetha, M.D., Karina A. Keogh, M.D., Eugene Y. Kissin, M.D., Paul A. Monach, M.D., Ph.D., Tobias Peikert, M.D., Coen Stegeman, M.D., Ph.D., Steven R. Ytterberg, M.D., and Ulrich Specks, M.D., for the RAVE-ITN Research Group*

Stone **NEJM** 2010

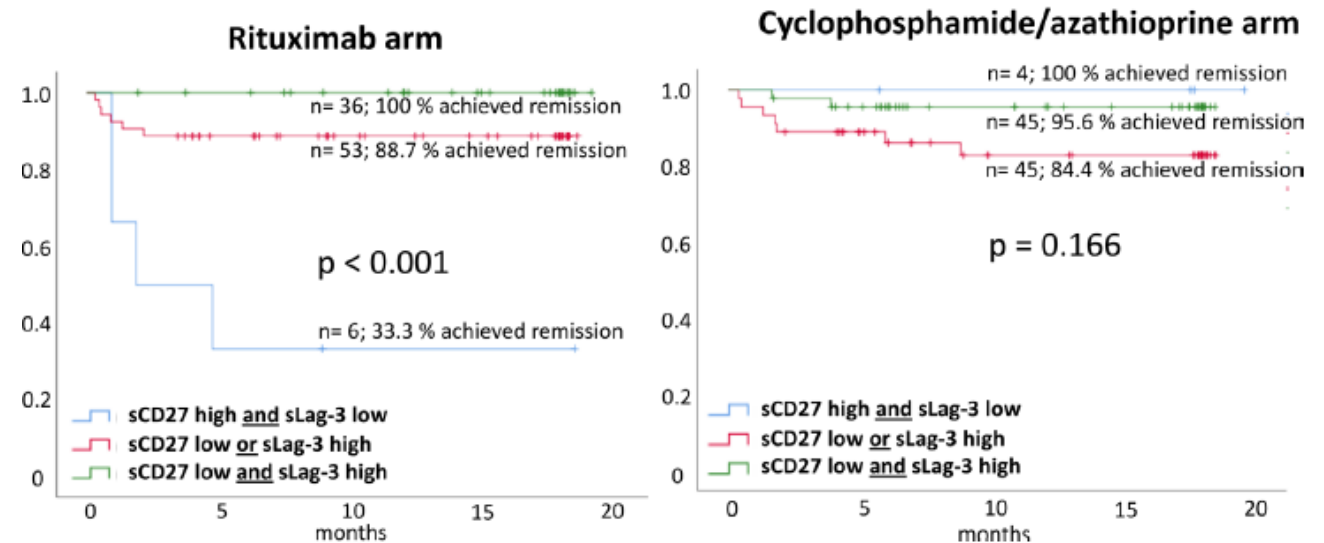
TRANSLATIONAL SCIENCE

Association of baseline soluble immune checkpoints with the risk of relapse in PR3-ANCA vasculitis following induction of remission

Gabriele Gamerith,¹ Finn Mildner,¹ Peter A Merkel,² Kristina Harris,³ Laura Cooney,³ Noha Lim,³ Robert Spiera,⁴ Philip Seo,⁵ Carol A Langford,⁶ Gary S Hoffman,⁶ E William St Clair,⁷ Fernando C Fervenza,⁸ Paul Monach,⁹ Steven R Ytterberg,¹⁰ Duvuru Geetha,¹¹ Arno Amann,¹ Dominik Wolf,¹ Ulrich Specks,¹² John H Stone,¹³ Andreas Kronbichler¹⁴

Gamerith **AnnRheumDis** 2022

Efficacy Measure	Rituximab (N = 99)	Cyclophosphamide- Azathioprine (N = 98)	Difference percentage points (95% CI)	P Value
	number (percent)			
Complete remission				
6 mo	63 (64)	52 (53)	11 (-3 to 24)	0.13
12 mo	47 (47)	38 (39)	9 (-5 to 22)	0.22
18 mo	39 (39)	32 (33)	7 (-7 to 20)	0.32
Remission and <10 mg/day of prednisone				
6 mo	70 (71)	60 (61)	10 (-4 to 23)	0.16
12 mo	59 (60)	60 (61)	-2 (-15 to 12)	0.82
18 mo	54 (55)	52 (53)	2 (-12 to 15)	0.84
Complete remission at any time†	76 (77)	70 (71)		0.15



Induktionstherapie-Avacopan

The NEW ENGLAND
JOURNAL of MEDICINE

ESTABLISHED IN 1812 FEBRUARY 18, 2021 VOL. 384 NO. 7

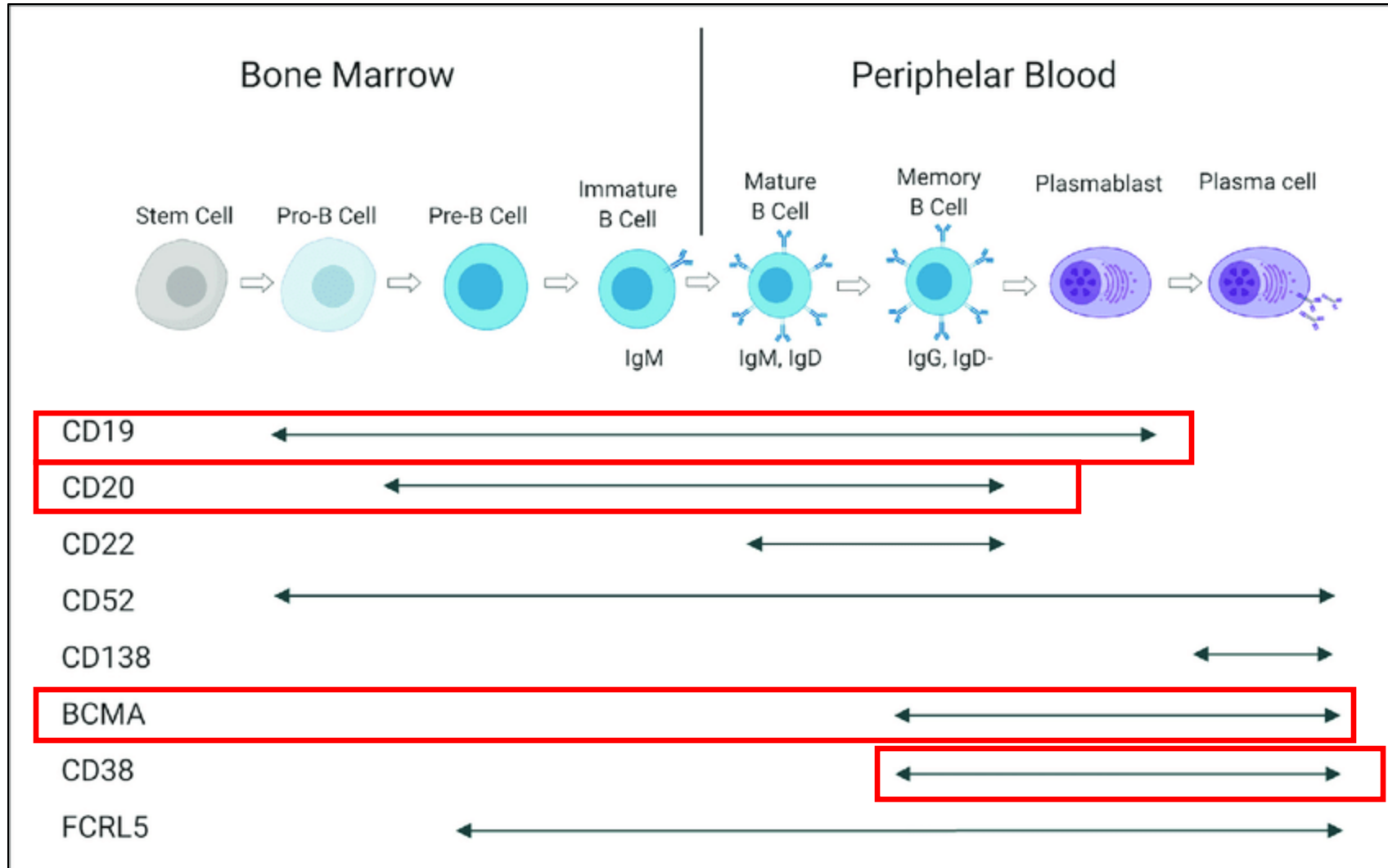
Avacopan for the Treatment of ANCA-Associated Vasculitis

David R.W. Jayne, M.D., Peter A. Merkel, M.D., M.P.H., Thomas J. Schall, Ph.D., and Pirow Bekker, M.D., Ph.D.,
for the ADVOCATE Study Group*

Remission 52w

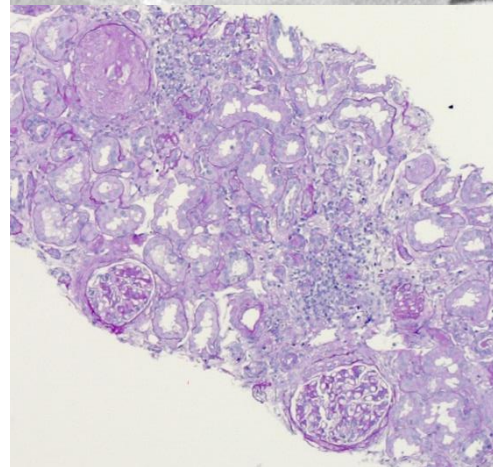
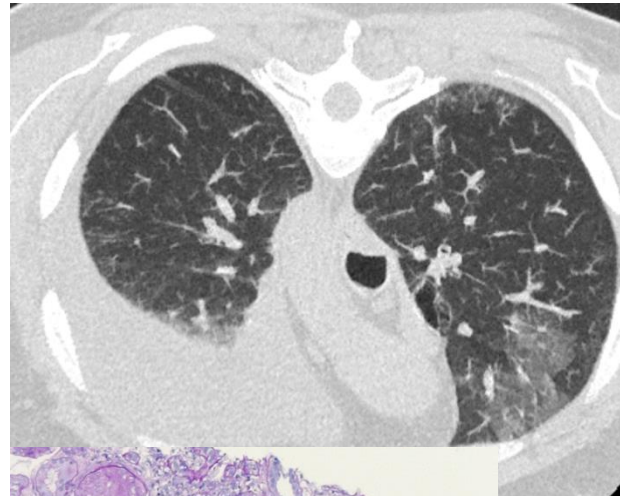
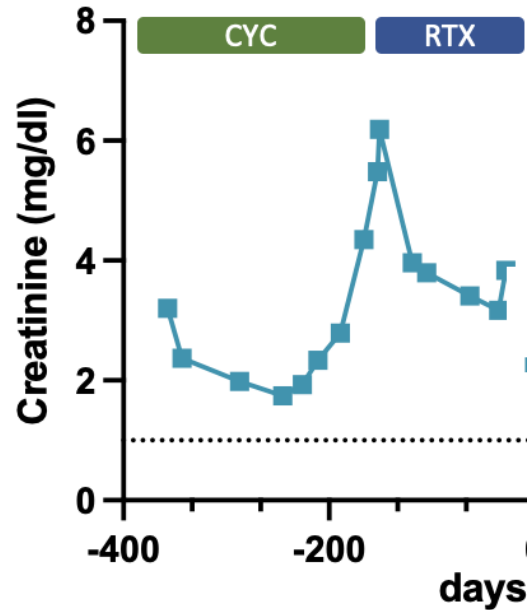
	Prednisone (N=164)	Avacopan (N=166)
Disease Status		
Newly diagnosed subjects	90 / 164 (54.9%)	109 / 166 (65.7%)
Relapsing disease	24 / 50 (48.0%)	39 / 51 (76.5%)
ANCA Status		
Anti-PR3 positive	40 / 70 (57.1%)	43 / 72 (59.7%)
Anti-MPO positive	50 / 94 (53.2%)	66 / 94 (70.2%)
Background Treatment		
Cyclophosphamide	30 / 57 (52.6%)	33 / 59 (55.9%)
Rituximab	60 / 107 (56.1%)	76 / 107 (71.0%)
Type of AAV		
GPA	52 / 90 (57.8%)	56 / 91 (61.5%)
MPA	38 / 74 (51.4%)	53 / 75 (70.7%)

Surface antigens

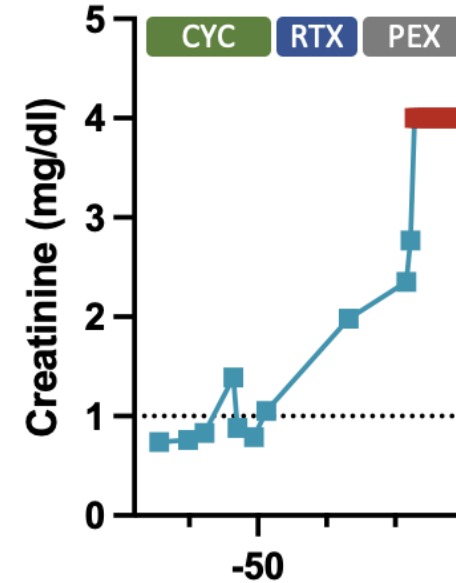


Novel treatment approaches: Daratumumab

Patient 1: 57j, male
MPO-ANCA+ MPA
active kidney disease
active pulmonary disease



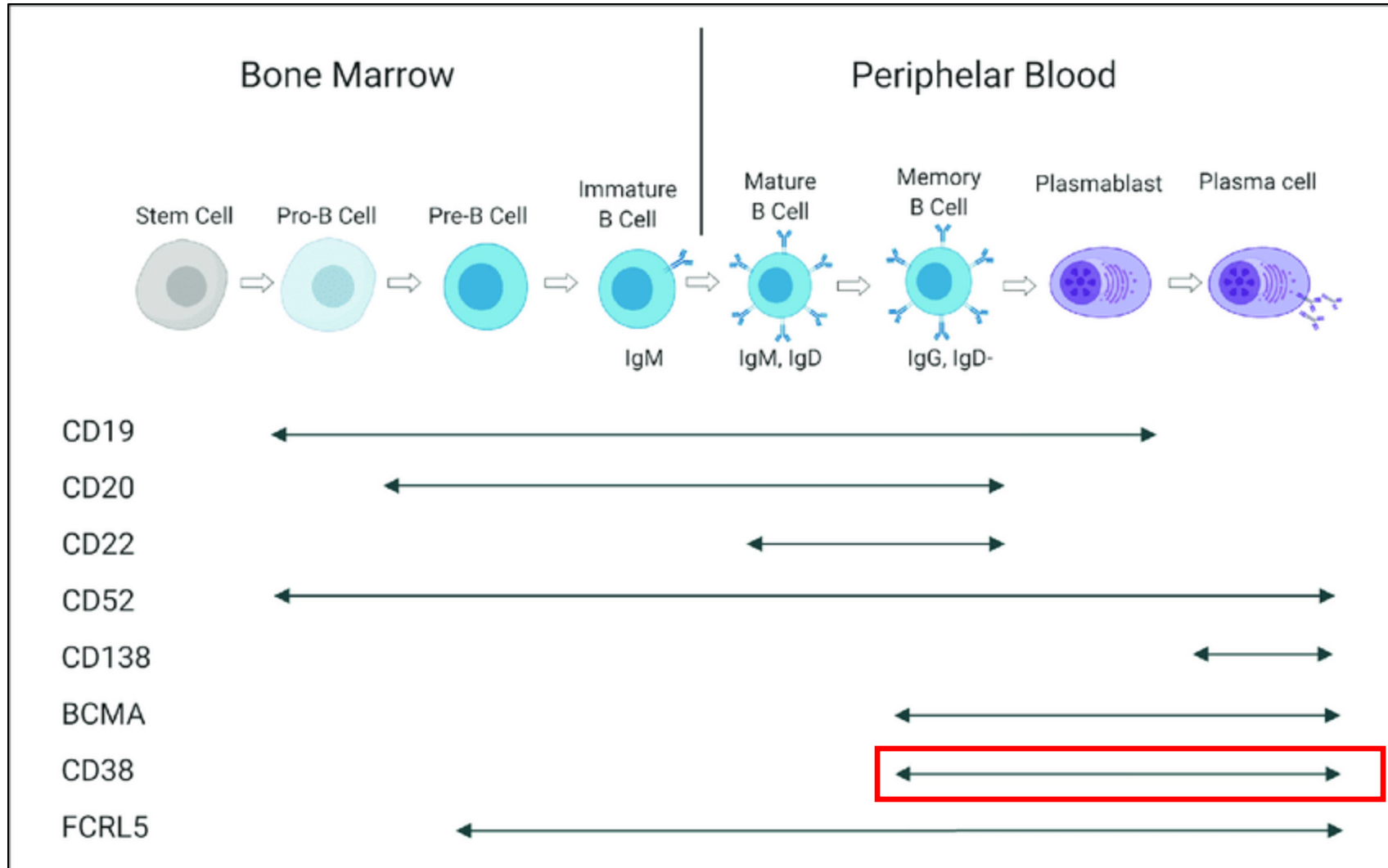
Patient 2: 41j, male
PR3-ANCA+ GPA
dialysis-dependent AKI,
ARDS with vvECMO,
Cardiac manifestation (aortic stenosis)



Lennard Ostendorf

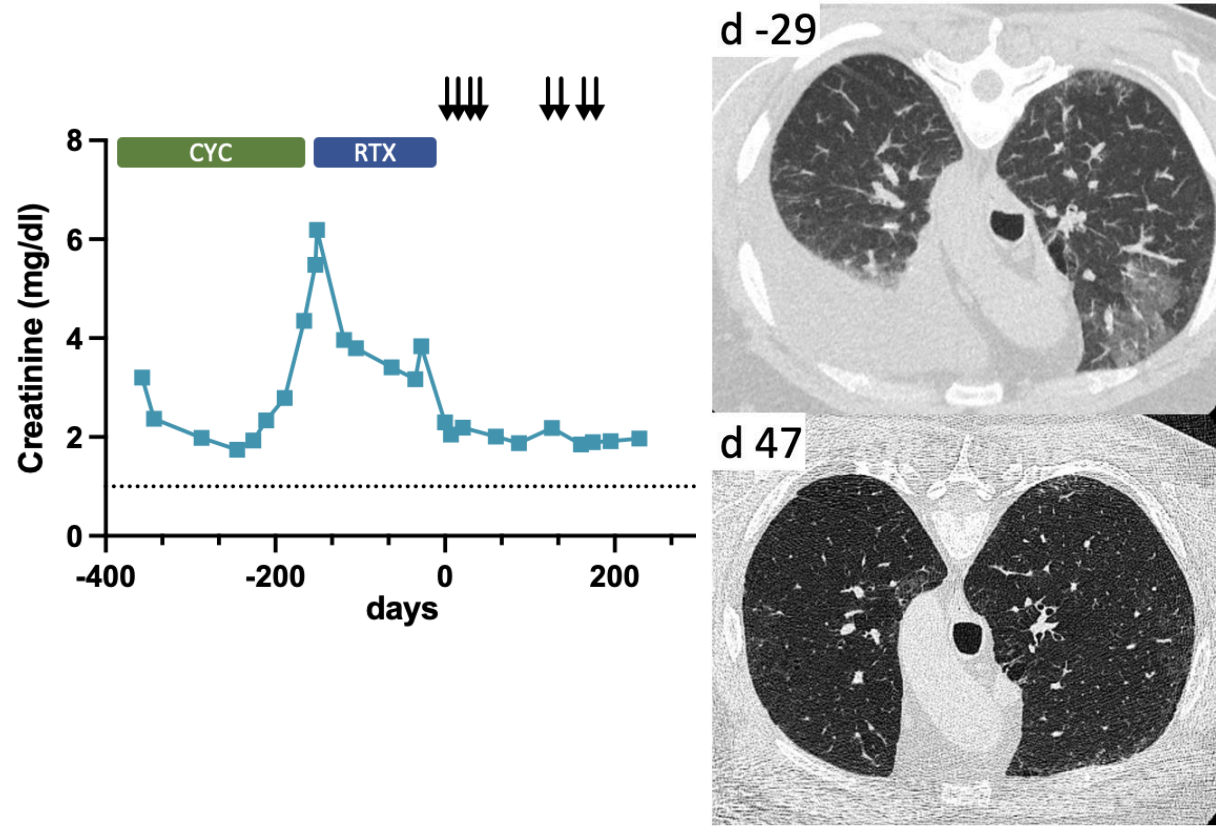


Surface antigens

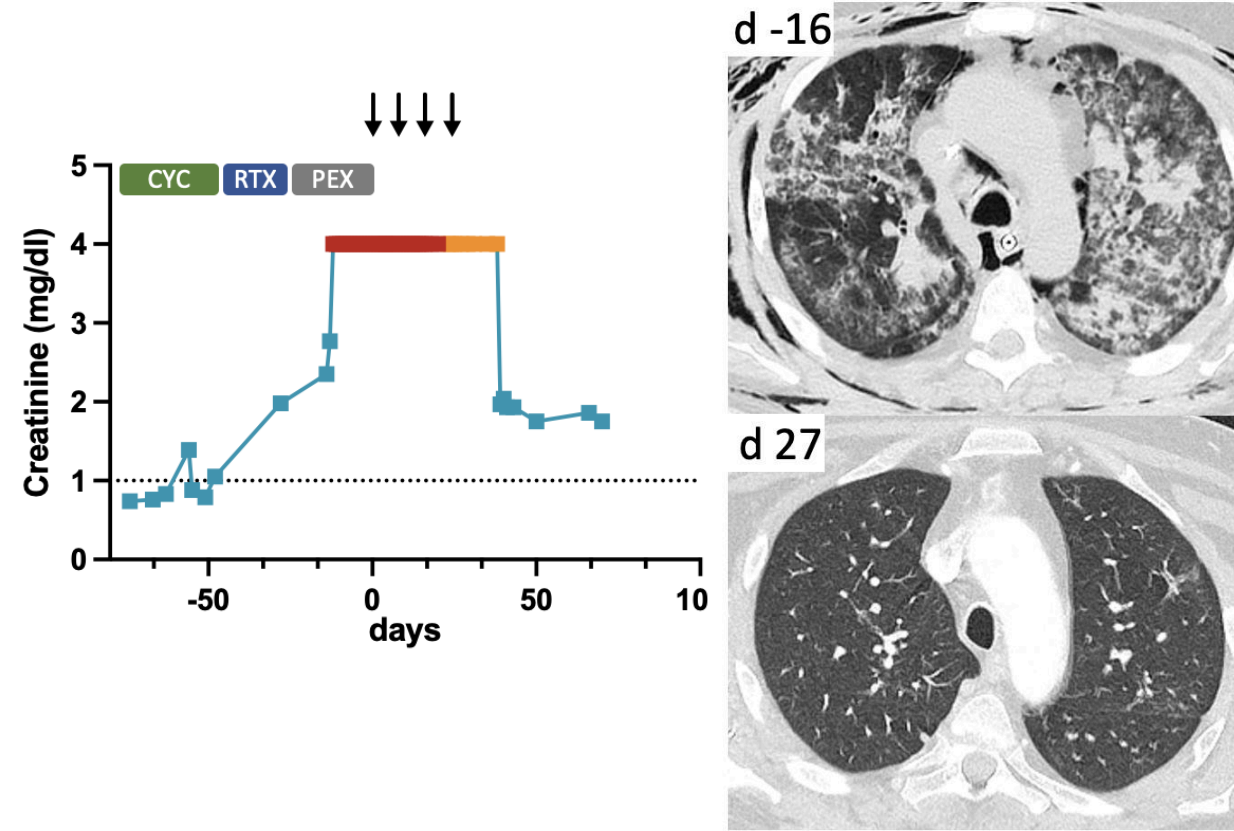


Novel treatment approaches: Daratumumab

Patient 1



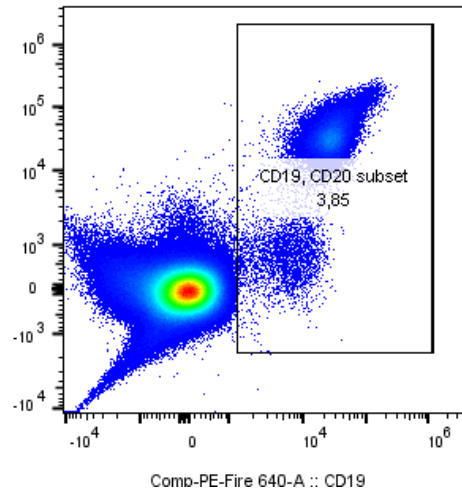
Patient 2



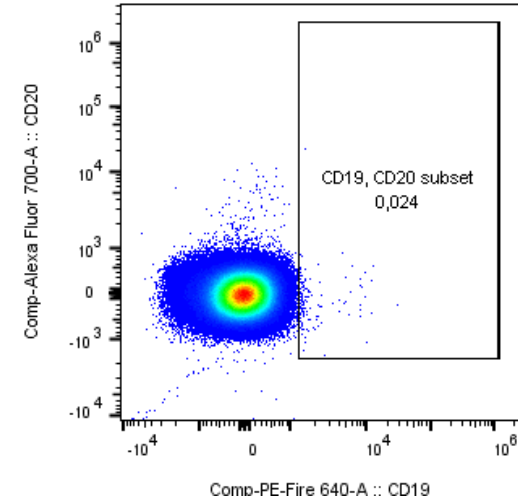
Insufficient depletion of tissue antibody-producing cells

blood

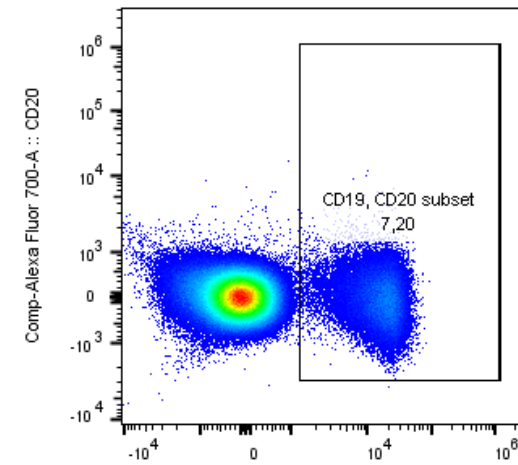
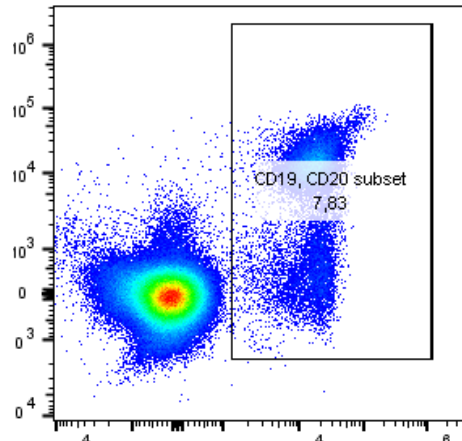
Kontrolle



RTX Behandlung

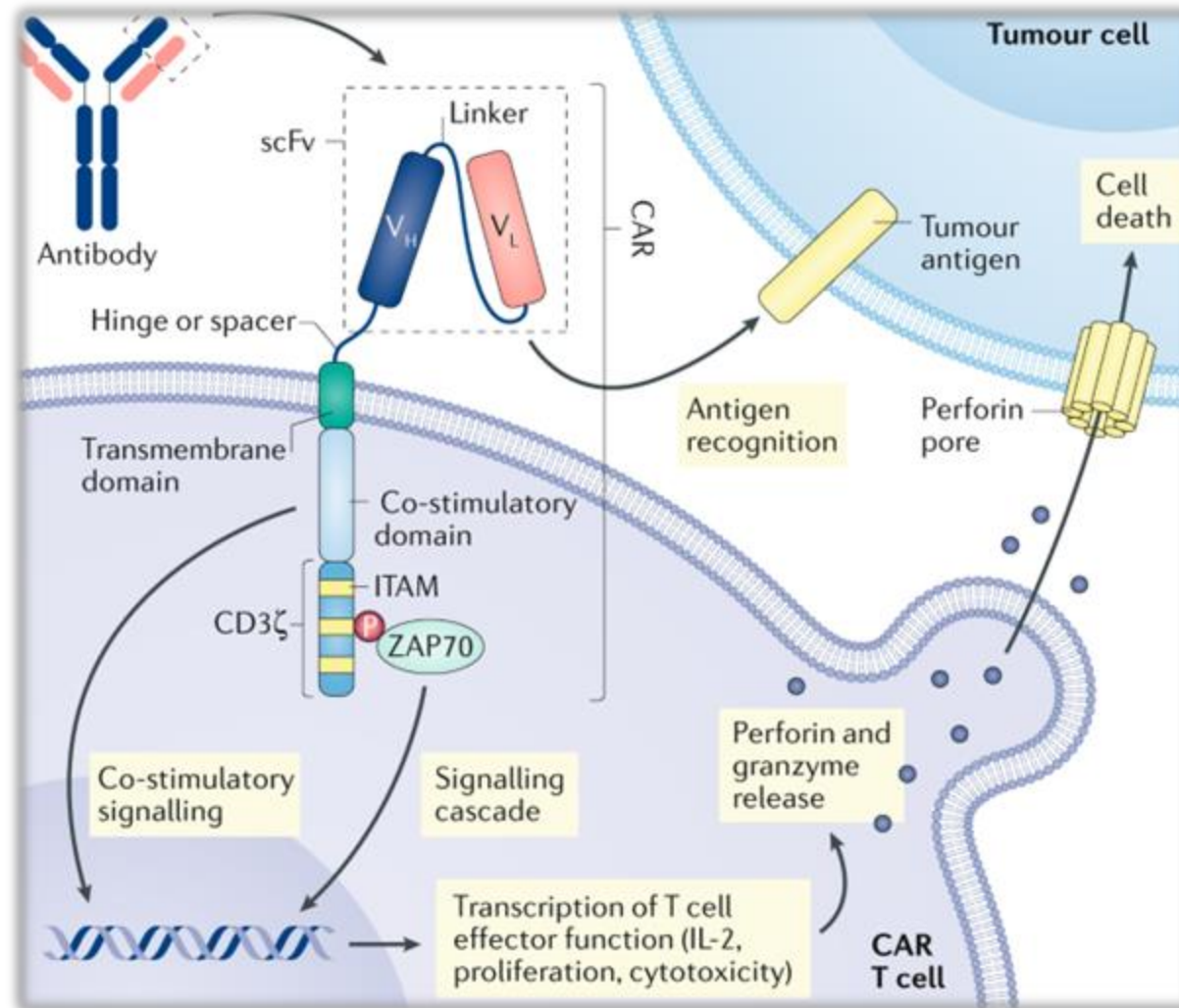


**Bone-
marrow**

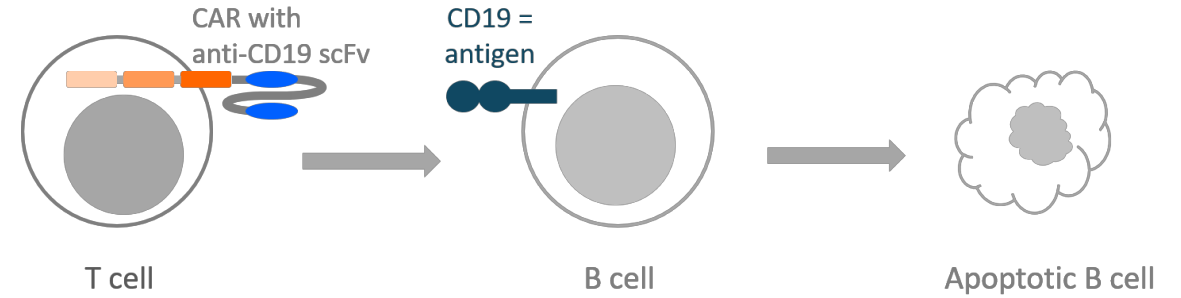
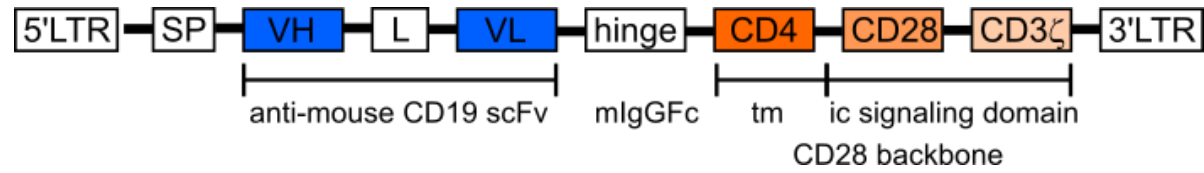


CD19

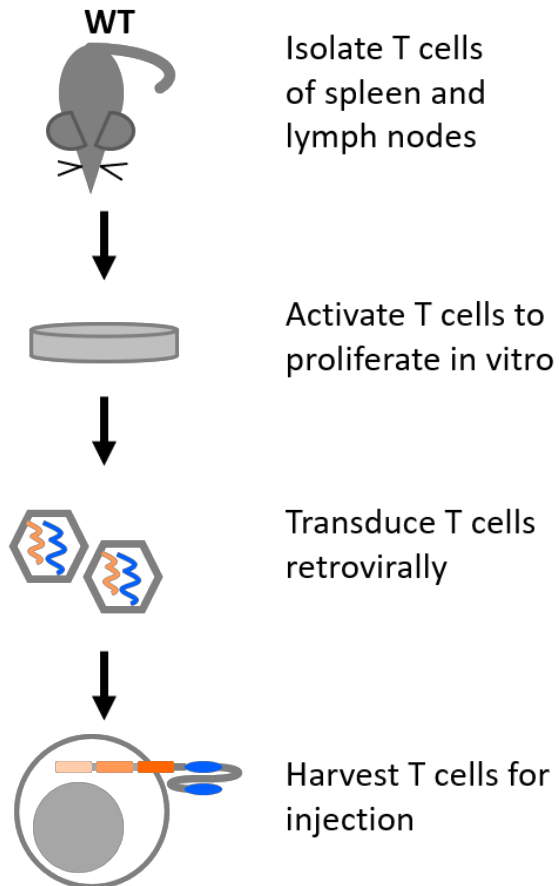
Cellular therapies as a new option for improved depletion of B cells



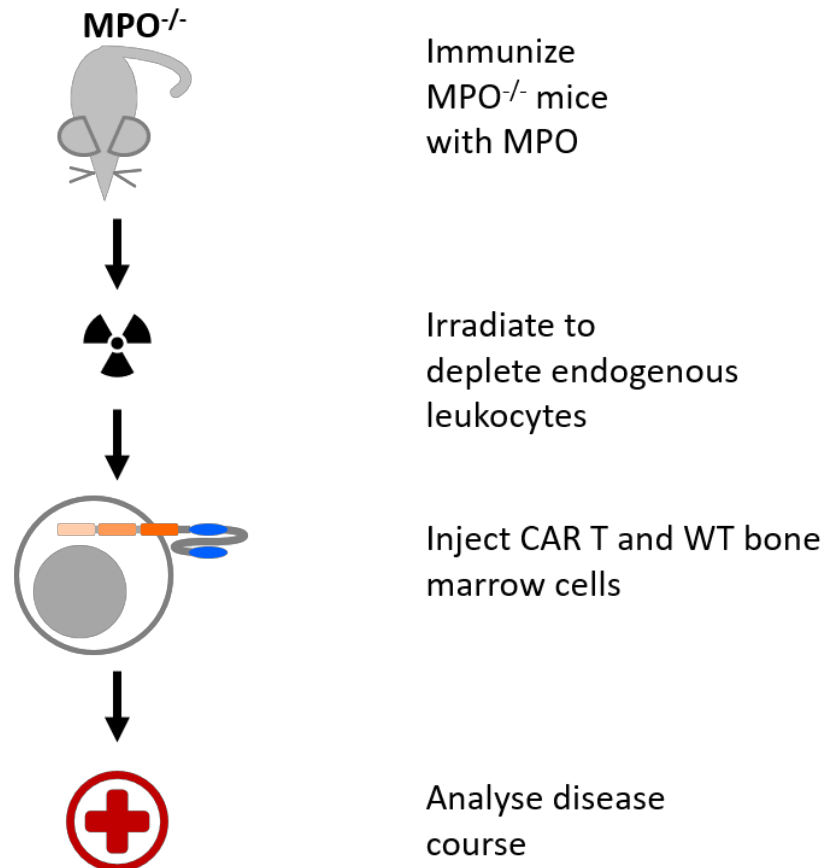
Chimeric antigen receptor (CAR) T cells



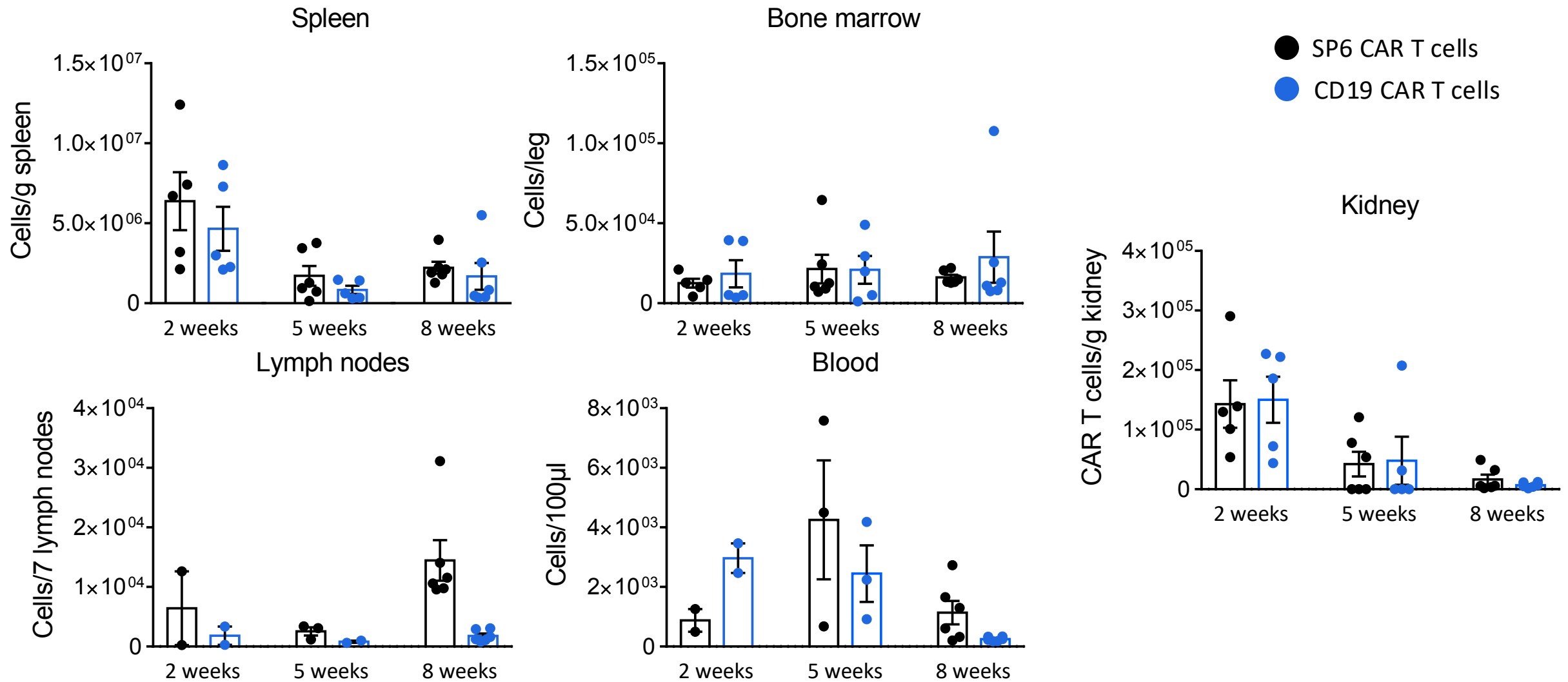
Höpken lab



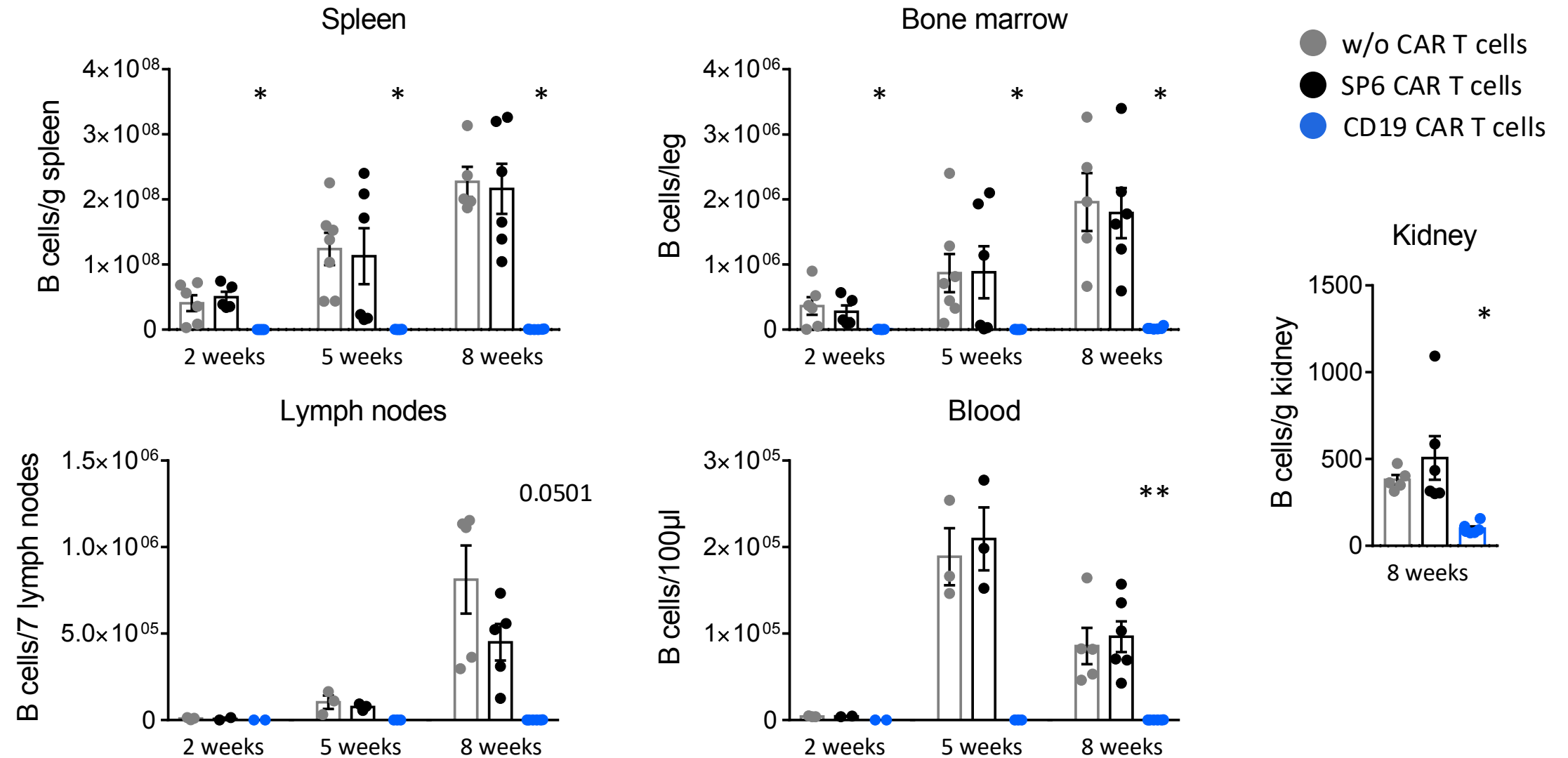
Schreiber lab



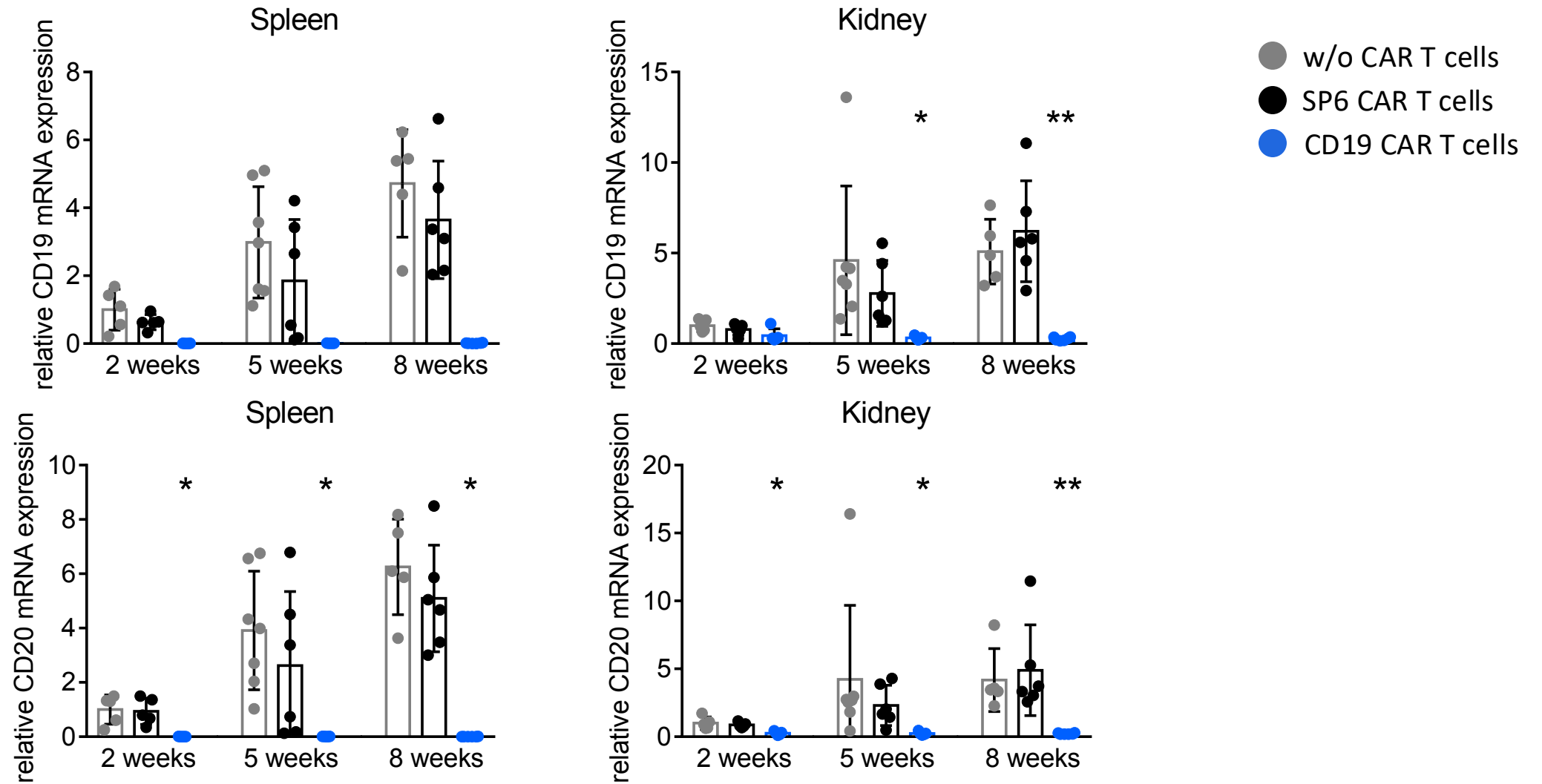
CD19 CAR T cells detectable in all analyzed organs



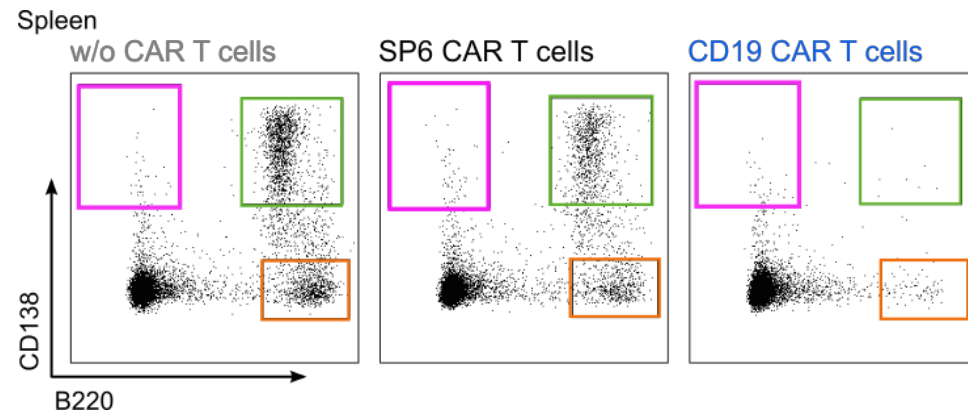
CD19-expressing B cells depleted in CD19 CAR T-treated mice



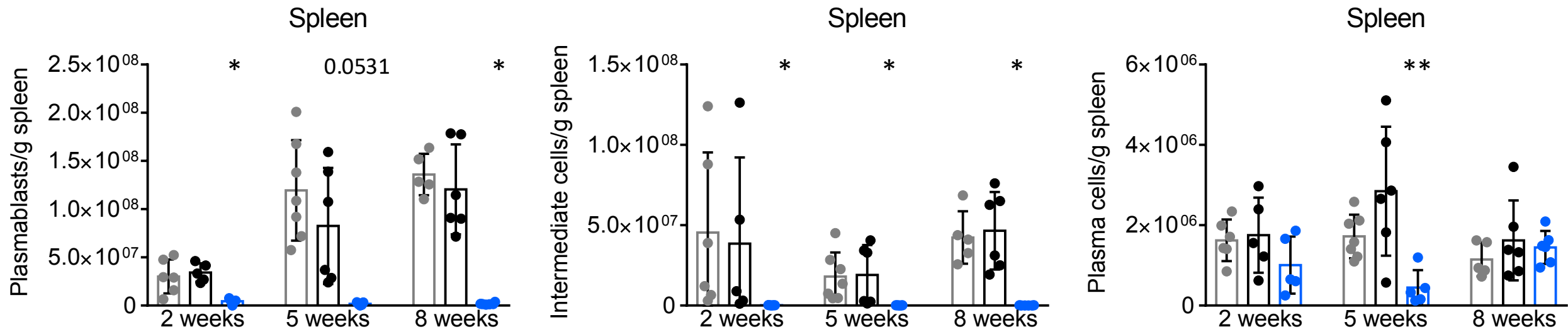
CD19-expressing B cells depleted in CD19 CAR T-treated mice



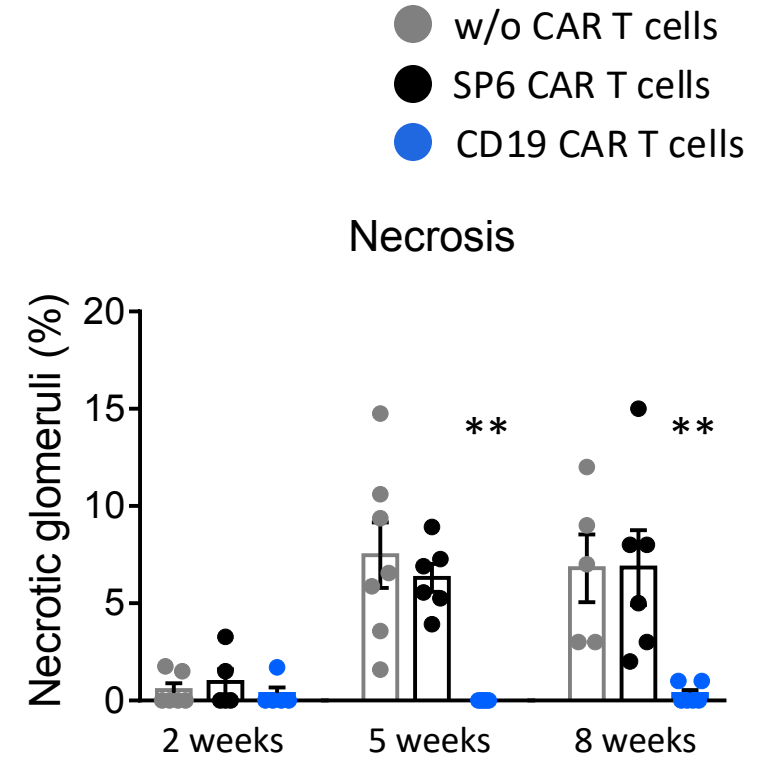
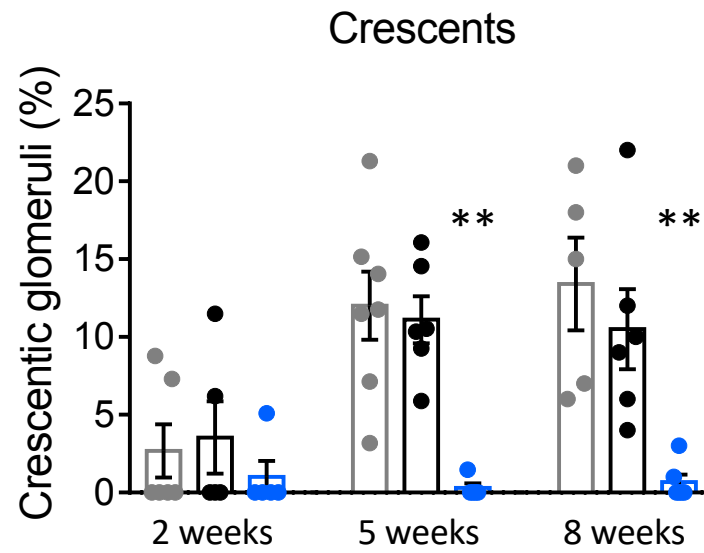
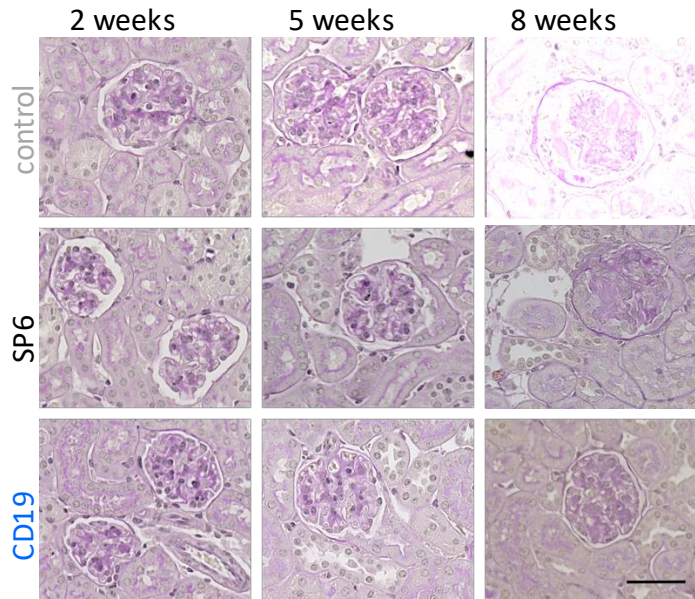
Plasmablasts & intermediate plasma cells decreased after CD19 CAR T-treatment



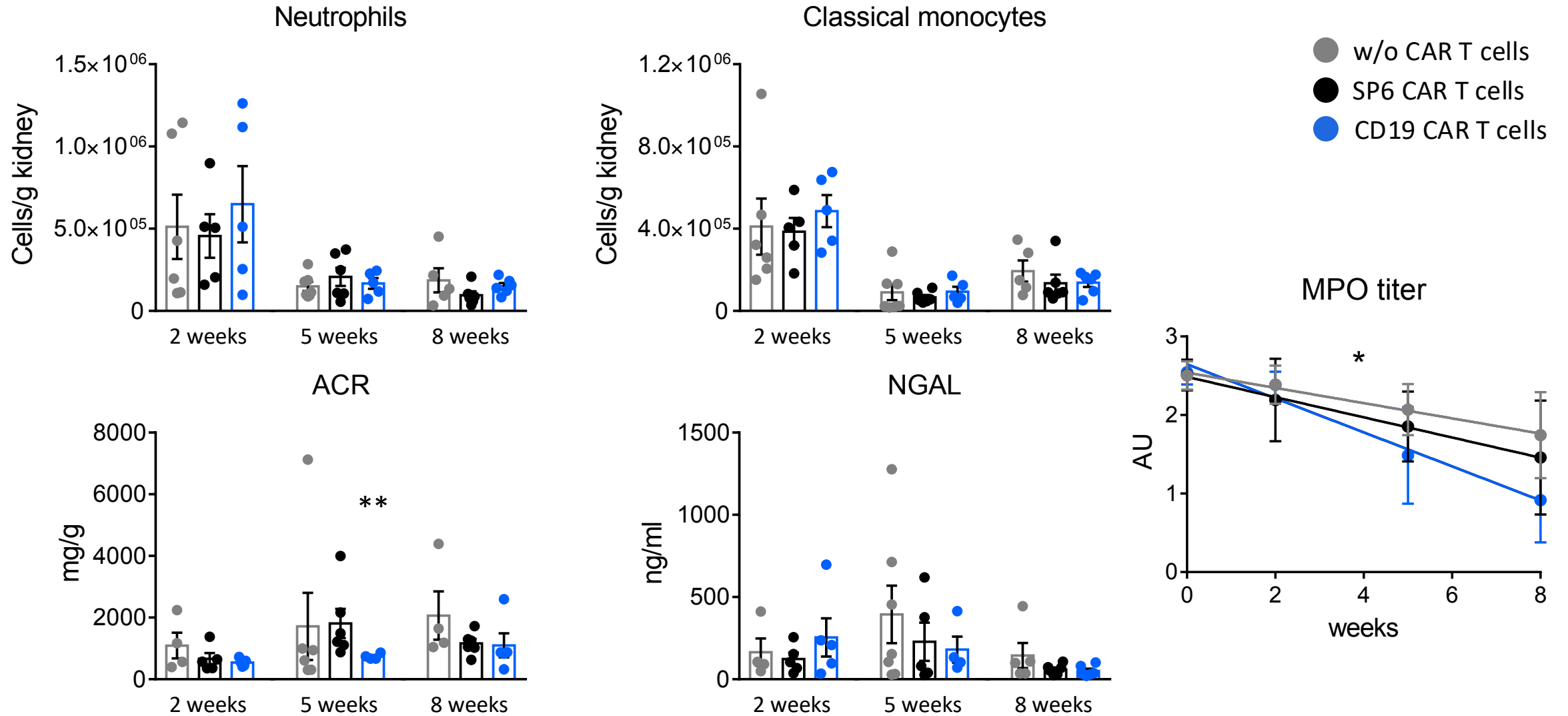
- w/o CAR T cells
- SP6 CAR T cells
- CD19 CAR T cells



Histopathological changes strongly decreased in CD19 CAR T-treated mice



Kidney infiltrating cells, urine parameter, MPO titer

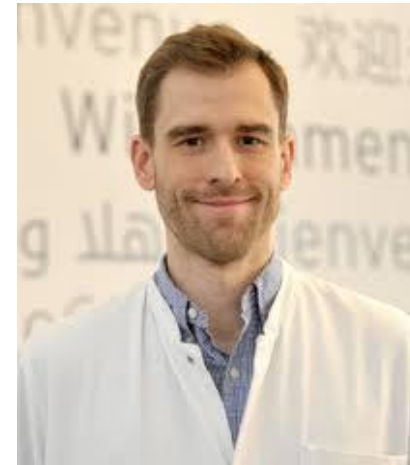


Summary

- CD19-targeting CAR T cells are effective in treating ANCA-induced glomerulonephritis
 - CAR T cells migrate into organs
 - CD19-expressing cells markedly diminished
 - Development of kidney damage strongly decreased

PR3-ANCA-positive granulomatosis with polyangiitis (GPA) with kidney, lung, joint, ENT, eye, and skin involvement, ED 12/1994 (Hamburg)

- 52 yrs, male
- Medical history:
 - 1995-2001 Endoxan (po) and Prednisolon (po)
 - 03/06 Relapse with skin, joint, and ENT involvement
 - 3x Endoxan i.v. (Berlin Buch)
 - o cumulative Cyclophosphamide: 150 g !!
 - Azathioprin 5-11/06 -> Intolerance
 - MMF 11/06-08/09 -> Arthralgias
 - MTX 25 mg/Woche s.c. 08/09-12/14
 - 12/2014 Relapse with skin vasculitis
 - RTX 4x 375mg/m² 12/14 & 1/2015
 - RTX 04/15, 05/15, 11/15



David Simon



Gerhard Krönke

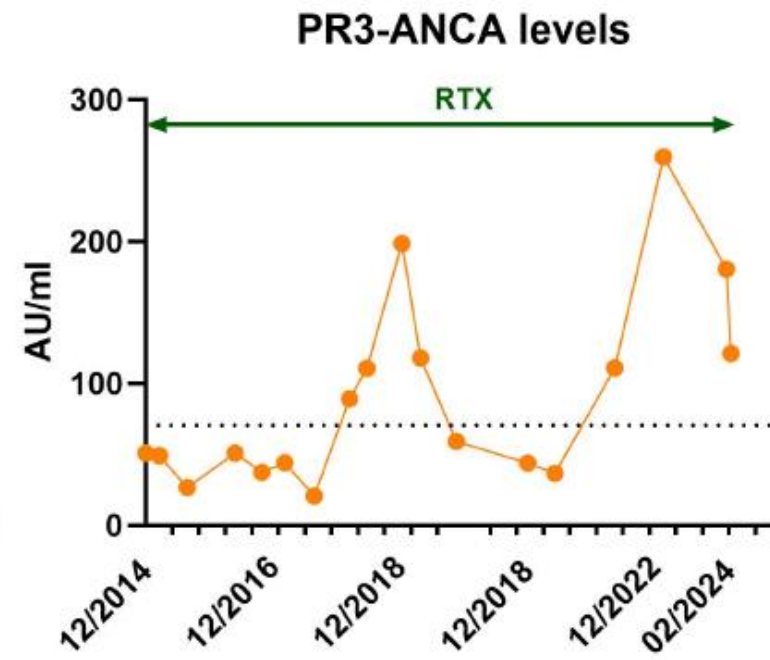
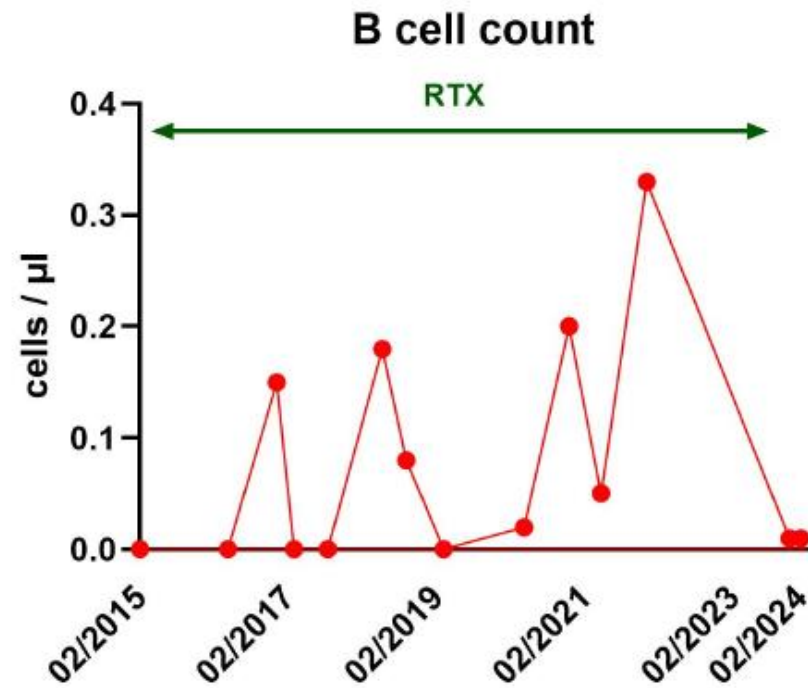
Rheumatologie und Klinische Immunologie
Charité

PR3-ANCA-positive granulomatosis with polyangiitis (GPA)

- Relapse 8/16 Skin vasculitis and arthralgias
- RTX 1g 8/16
- 12/16 additional MTX -> Intolerance (Nausea, headaches)
- 3/17 RTX, additional Azathioprin
- 6/17 persistent biopsy-proven leukocytoclastic cutaneous vasculitis with necrosis
- MMF 2g/d
- 11/17-2/19 Pause of therapy because of Therapy fatigue
- 02/2019 Majorrelapse Lung
- 3/19 Rituximab 4x 375mg/m²
- 10/19: Rituximab 500mg
- 4/20: Rituximab 500mg
- 12/20: Rituximab 500mg
- 5/22: Rituximab 500mg
- 11/23 Minor Relapse with rising ANCA, Arthralgias, erythrozyturia
- 11/23 Rituximab 1000mg

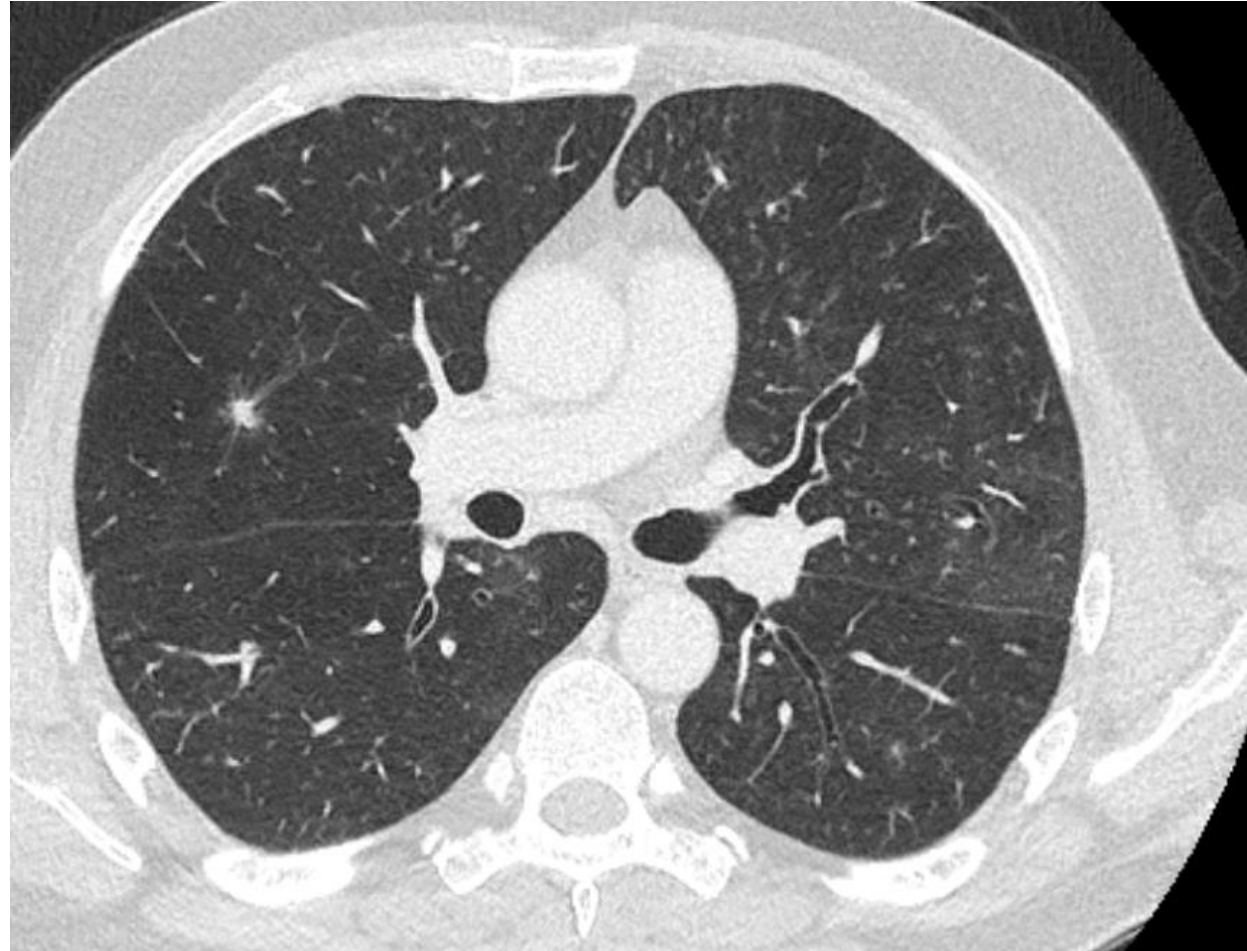
PR3-ANCA-positive granulomatosis with polyangiitis (GPA)

Positive PR3-ANCA and incomplete B cell depletion despite continuous treatment with Rituximab



PR3-ANCA-positive granulomatosis with polyangiitis (GPA)

New lung granuloma formation, dyspnea and B-symptomatology despite treatment with rituximab



2 small new granulomas in the right lower lobe

CAR T cell therapy

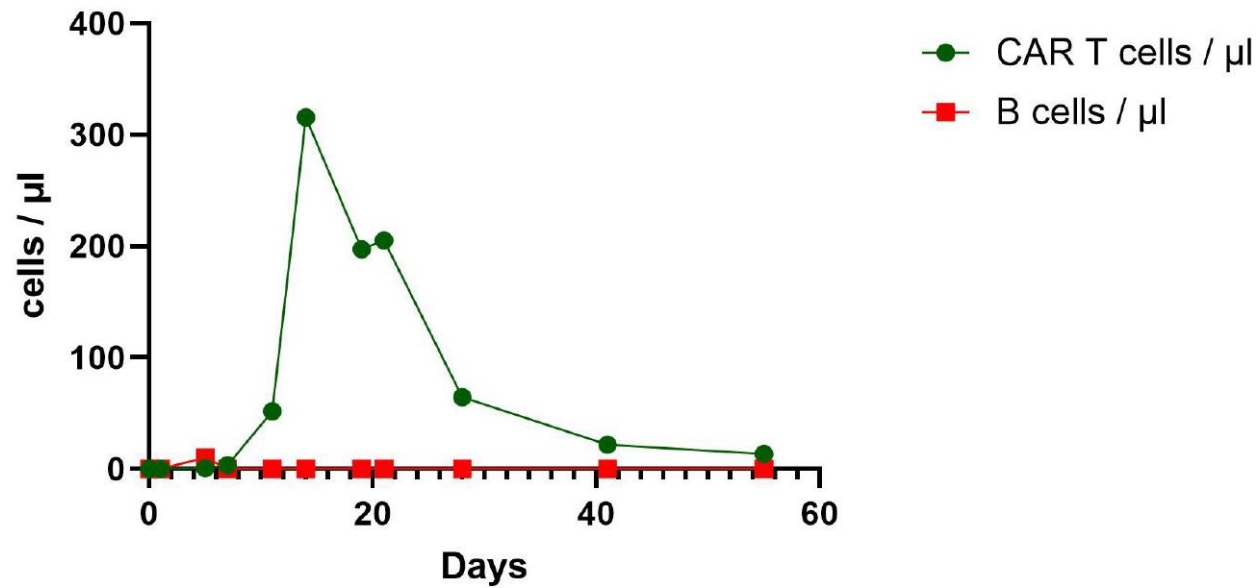
Date infusion	20.06.2024
Cell count	1.0 x 10 ⁸ / 10 ⁶ CAR-positive viable T-Zellen
Karnofsky-Index before therapy	100%
ECOG Score before therapy	0
Lymphodepletion	Cyclophosphamid Fludarabin
Cytokine Release Syndrome (CRS)	Grad I
Neurotoxicity (ICANS)	No ICAN
CAR-T-induzierte Zytopenie	NCI Grad 0
Antiinfektive Prophylaxe	Aciclovir / Cotrimoxazol

Safety

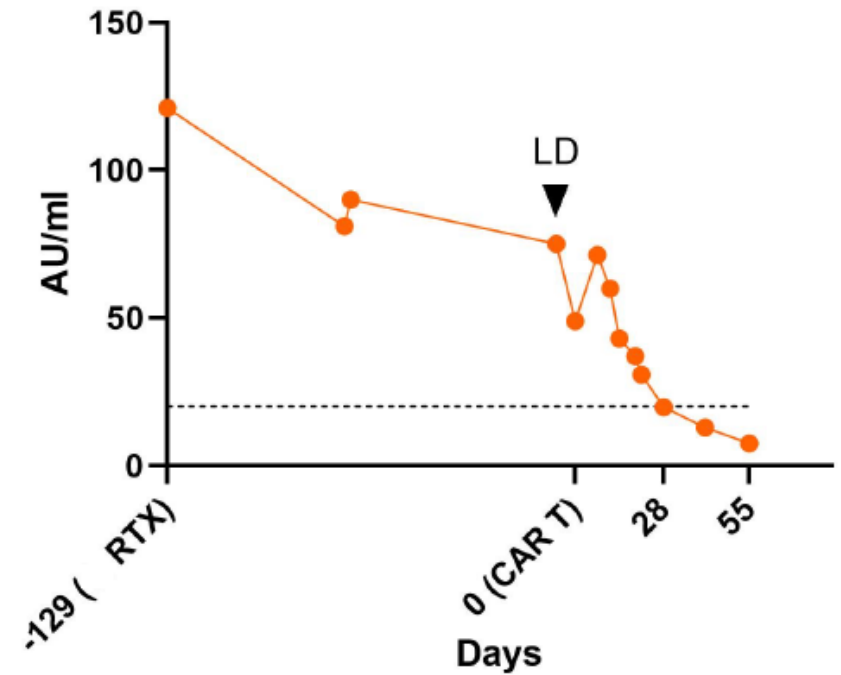
- CRS Grade 1
 - Resolved after tocilizumab (3x 800 mg)
- No ICANS
- Neutropenia
 - grade 3 (minimum neutrophil count of 0.57/nl) 13 days after therapy
 - resolved within 2 days after administration of 2 x 48.000.000 E filgrastim.

CAR T cells, B cells and PR3-ANCA in the peripheral blood after CAR T cell therapy

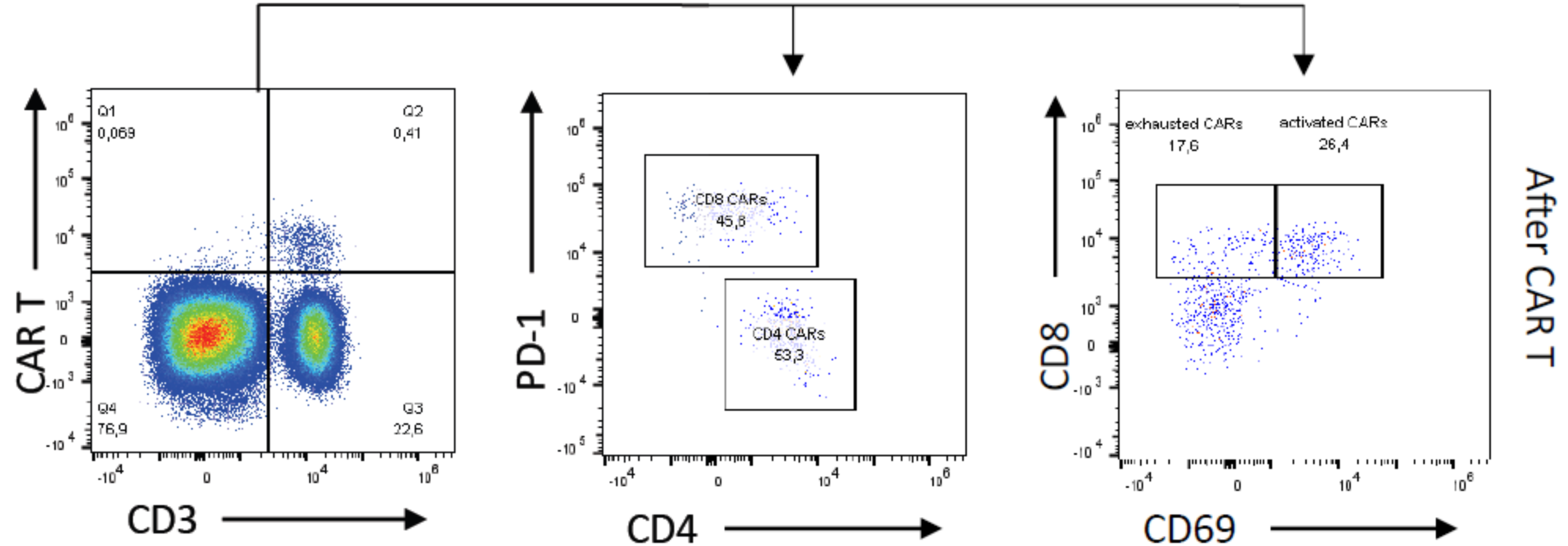
CAR T and B cell count



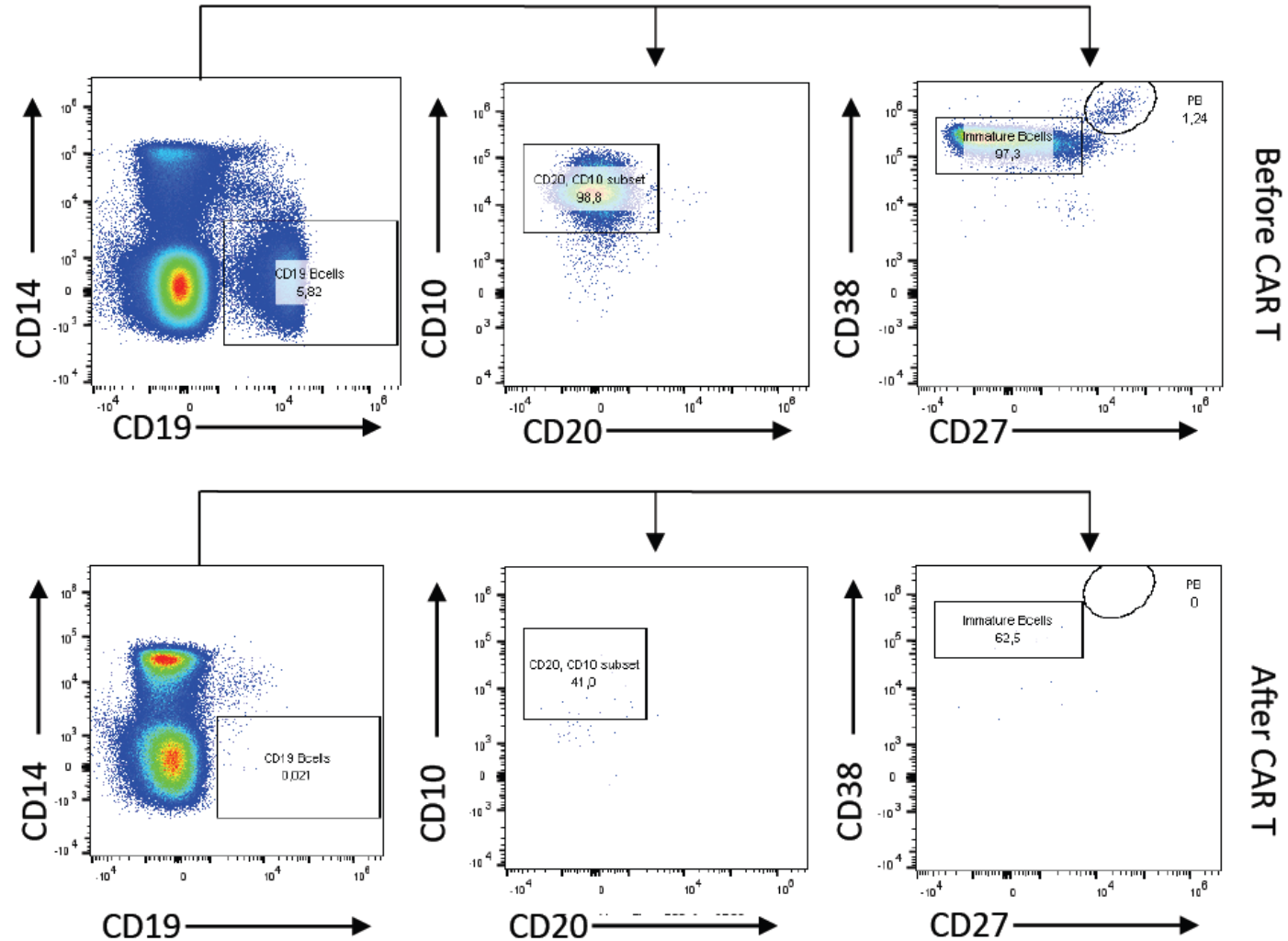
PR3-ANCA levels



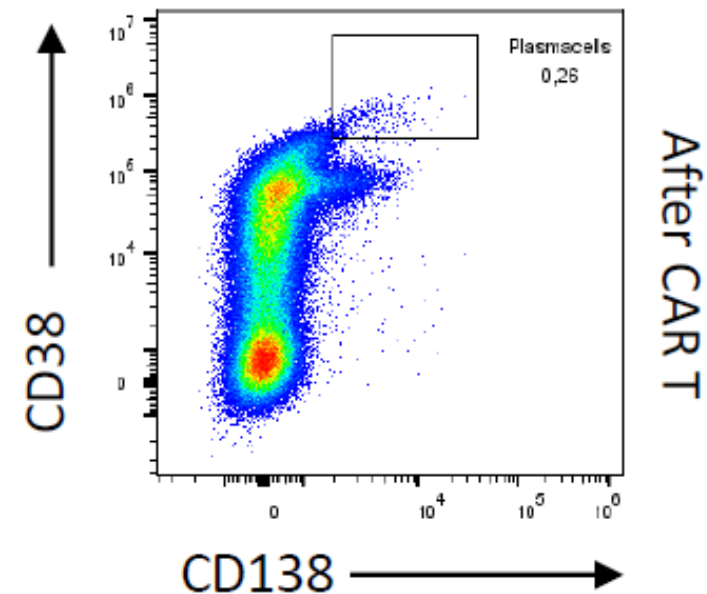
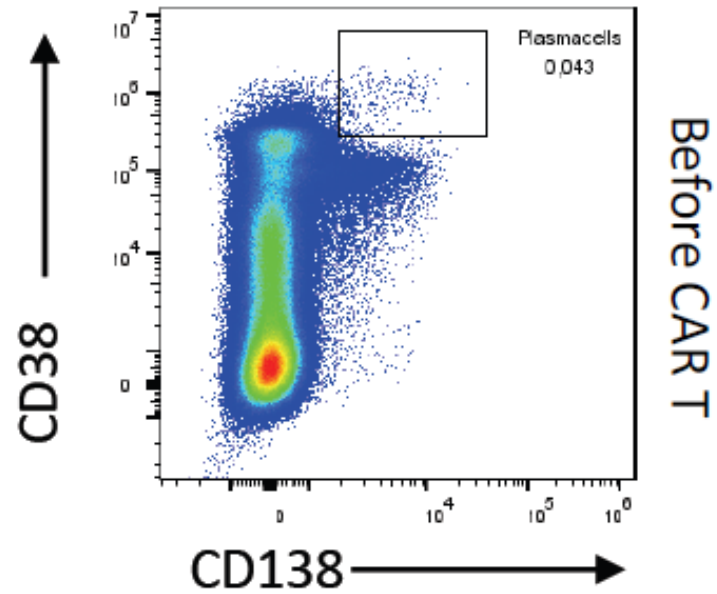
CAR T cells are detected in the bone marrow 48 days post-treatment



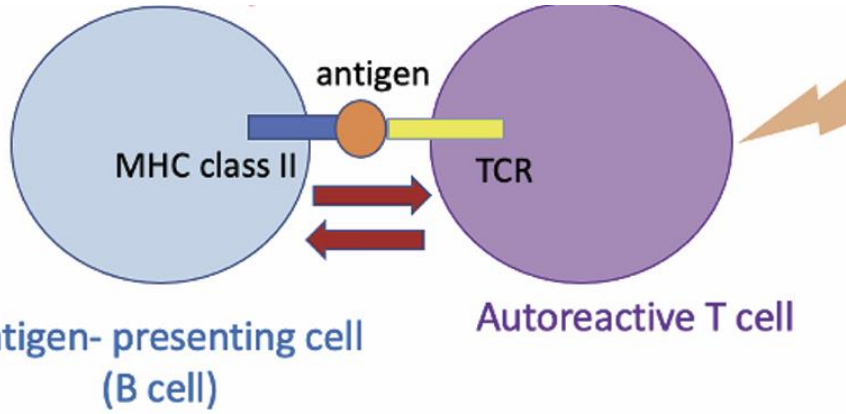
CD19+ B cells in the bone marrow are diminished post-treatment



CD138+/CD19- plasma cells persist in the bone marrow

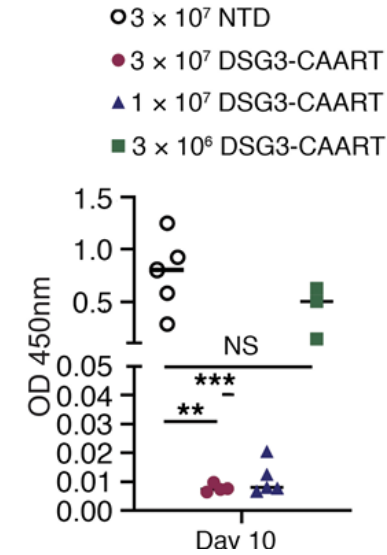


Ausblick: CAAR-T als spez Therapieoption?



PV patient 1

Coating	anti-		
	DSG3	hlgG	BSA
# Plated cells	100 K	1000	1000
PV B cells + NTD	156	88	0
PV B cells DSG3-CAART	12	208	0
PV B cells + CART19	2	7	1
HD B cells + NTD	4	118	2
HD B cells DSG3-CAART	1	142	1
HD B cells + CART19	4	11	4

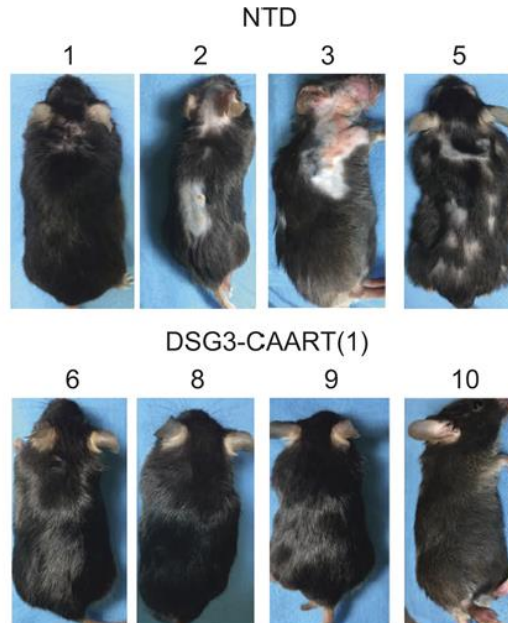


The Journal of Clinical Investigation

CONCISE COMMUNICATION

Antigen-specific B cell depletion for precision therapy of mucosal pemphigus vulgaris

Jinmin Lee,¹ Daniel K. Lundgren,¹ Xuming Mao,¹ Silvio Manfredo-Vieira,¹ Selene Nunez-Cruz,² Erik F. Williams,³ Charles-Antoine Assenmacher,⁴ Enrico Radaelli,⁴ Sangwook Oh,¹ Baomei Wang,¹ Christoph T. Ellebrecht,¹ Joseph A. Fraietta,³ Michael C. Milone,² and Aimee S. Payne¹



MPO/PR3- CAAR-T ?
Erste Versuche laufen

Imlifidase

Endopeptidase Cleavage of Anti-Glomerular Basement Membrane Antibodies *in vivo* in Severe Kidney Disease: An Open-Label Phase 2a Study

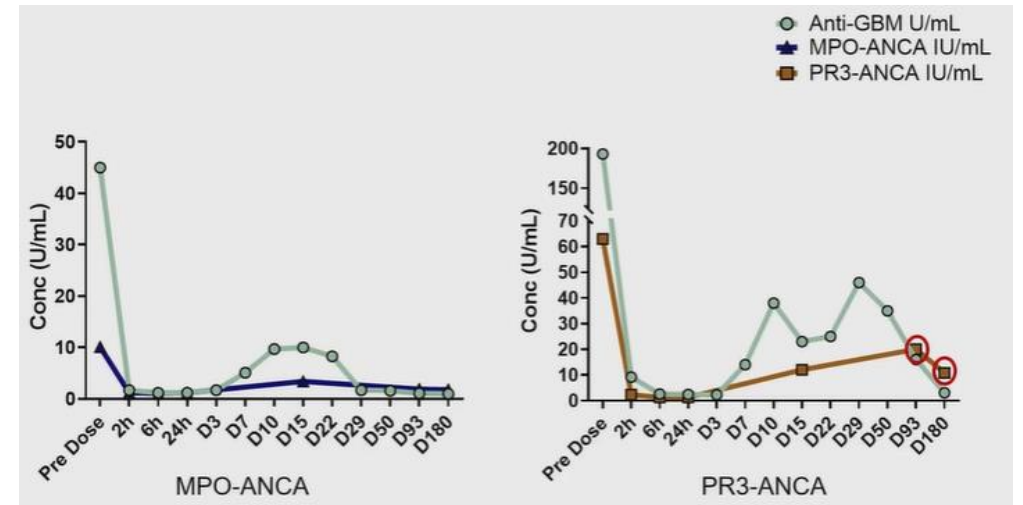
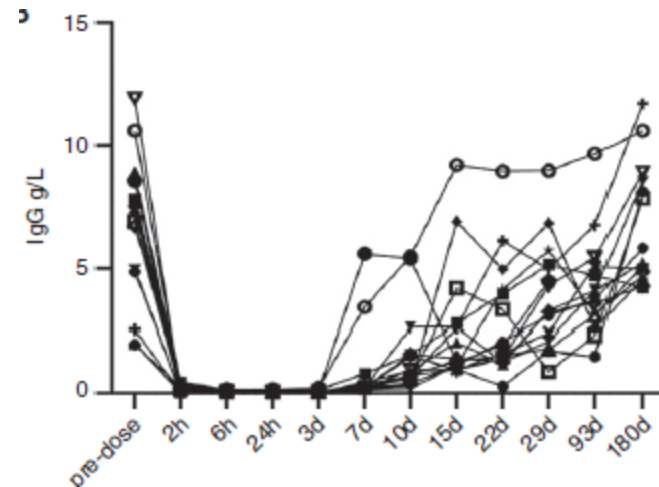
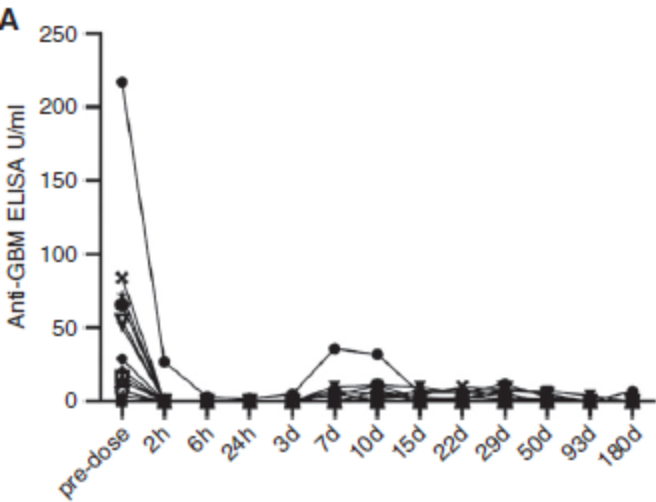
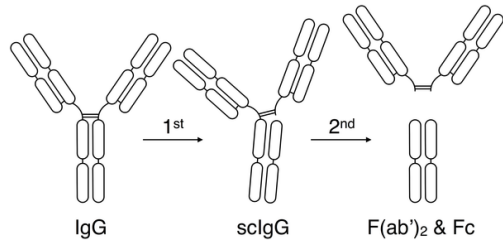
Fredrik Uhlin,^{1,2} Wladimir Szpirt,³ Andreas Kronbichler,⁴ Annette Bruchfeld,^{1,5} Inga Soveri,⁶ Lionel Rostaing,⁷ Eric Daugas,⁸ Arnaud Lionet,⁹ Nassim Kamar,¹⁰ Cédric Rafat,¹¹ Marek Mysliveček,¹² Vladimír Tesar,¹² Anders Fernström,¹ Christian Kjellman,¹³ Charlotte Elfving,¹³ Stephen McAdoo,¹⁴ Johan Mölne,¹⁵ Ingeborg Bajema,¹⁶ Elisabeth Sonesson,¹³ and Märten Segelmark^{1,17}

Table 1. Baseline clinical characteristics

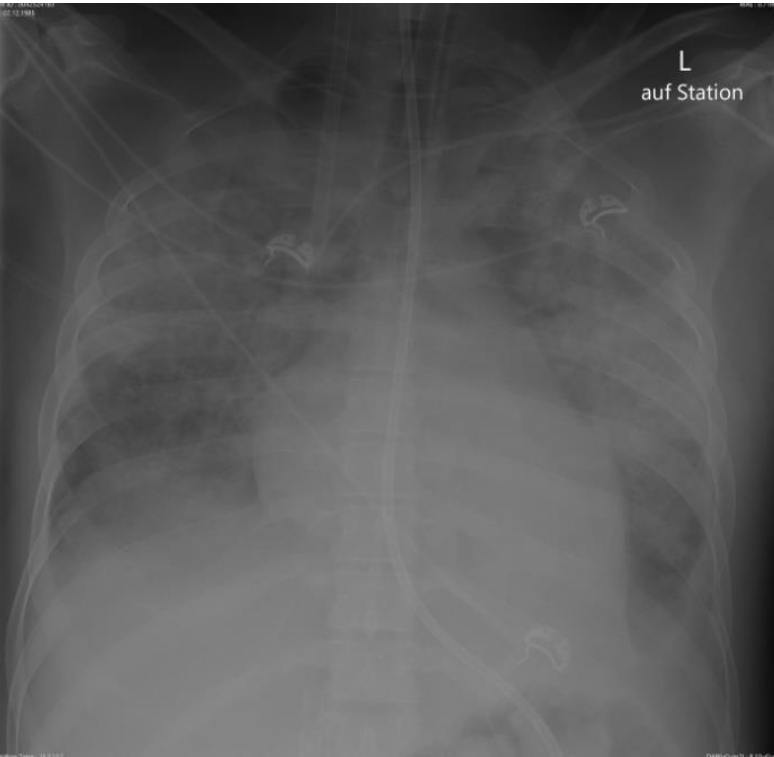
Characteristics	Men	Women	All	Historical Controls
Number, n (%)	9 (60)	6 (40)	15 (100)	50 (100)
Country: France/Sweden/Denmark Austria/Czech Republic, n	3/1/2/1/1	2/3/1/0/0	5/4/3/2/1	NA
Age years median (range)	60 (19–77)	66 (32–71)	61 (19–77)	63 (16–88)
ANCA MPO/PR3/none, n (%)	3/2/5 (33/22/56)	1/0/4 (17/0/67)	4/2/9 (27/13/60)	15 ^a /7 ^a /29 (30/14/58)
ANA positive, n (%)	3 (33%)	0	3 (20%)	NA
Renal function oliguria/dialysis/no dialysis, n (%)	4/4/1 (44/44/11)	1/1/4 (17/17/67)	5/5/5 (33/33/33)	41 ⁷ /9 (82/18)
Pulmonary disease AH/other/none, n (%) ^b	1/2/6 (11/22/67)	1/2/3 (17/33/50)	2/4/9 (13/27/60)	21/NA/NA (42/NA/NA)
Smoking class current/previous/never, n (%)	2/7/0 (22/78/0)	0/3/3 (0/50/50)	2/10/3 (13/67/20)	NA
Urinary albumin-creatinine, mg/g median (range) Reference range <27 mg/g ^c	2230 (434–30,531)	1932 (195–3540)	1982 (195–30,531)	NA
CRP mg/L median (range) Reference range <0.3 mg/dL ^c	3.4 (<0.07–5.2)	1.2 (<0.07–8.9)	3.2 (<0.07–8.9)	NA
Hb g/dL median (range) ^c	8.4 (6.9–11.1)	9.3 (7.3–11.5)	8.8 (6.9–11.5)	NA
Platelets 10 ⁹ /L median (range) ^c	318 (131–505)	320 (201–384)	320 (131–505)	NA
Berden class crescentic/mixed/sclerotic/focal, n (%)	6/2/0/0 (75/25/0/0)	3/2/1/0 (50/33/17/0)	9/4/1/0 (65/29/7/0)	25/3/1/0 (86/10/3/0)
Normal glomeruli, % median (range)	11% (0–29)	9% (0–35)	9.5% (0–35)	6.5% (0–43)
Linear staining for IgG, n/n (%)	5/8 (62)	6/6 (100)	11/14 (79)	27/29 (93)

Imlifidase (Idefirix, Hansa Biopharma)

- Cystein Protease von *Strep. pyogenes*

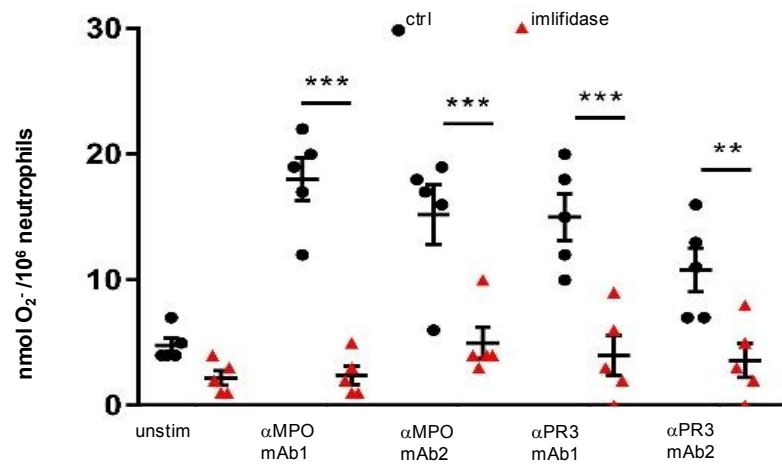
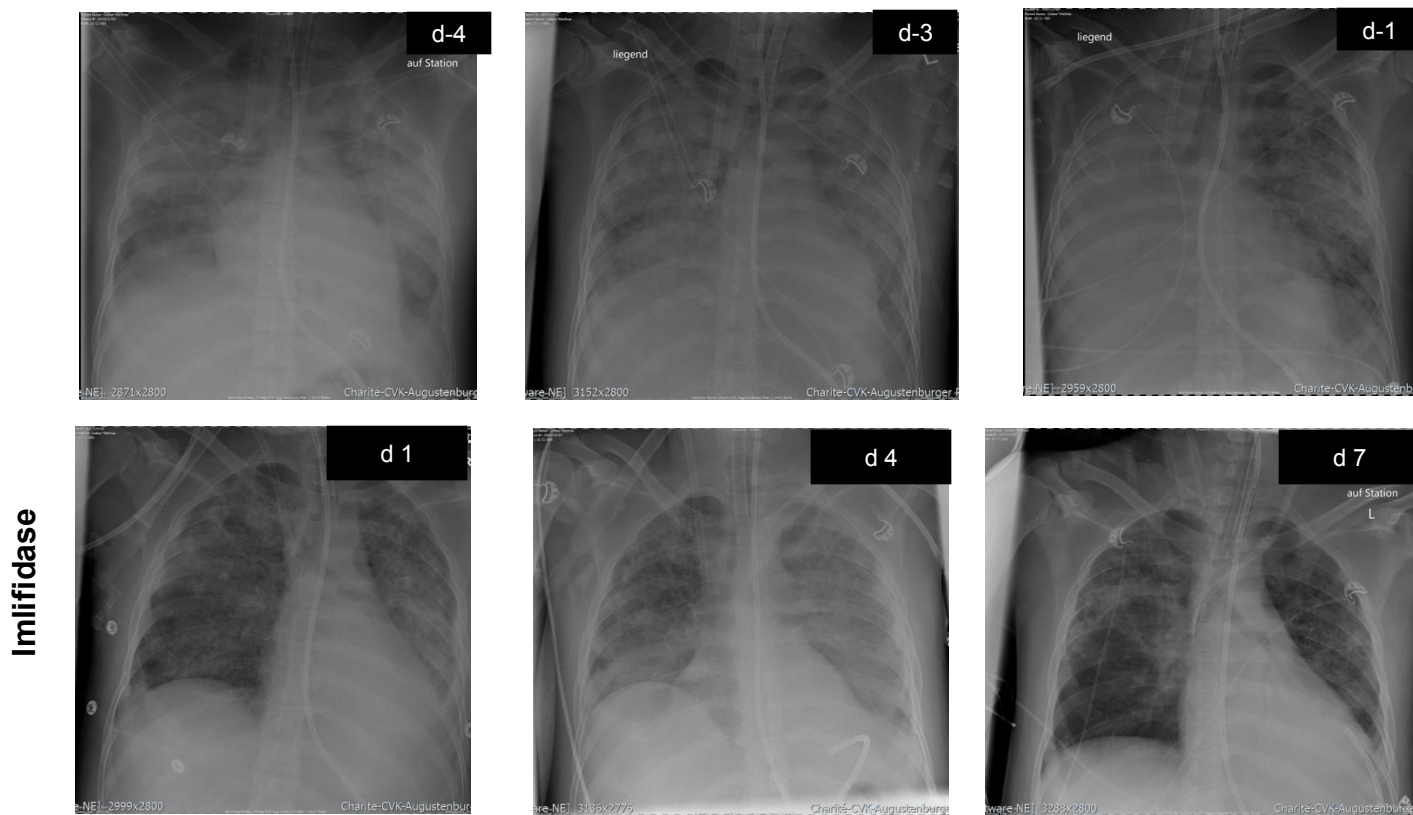
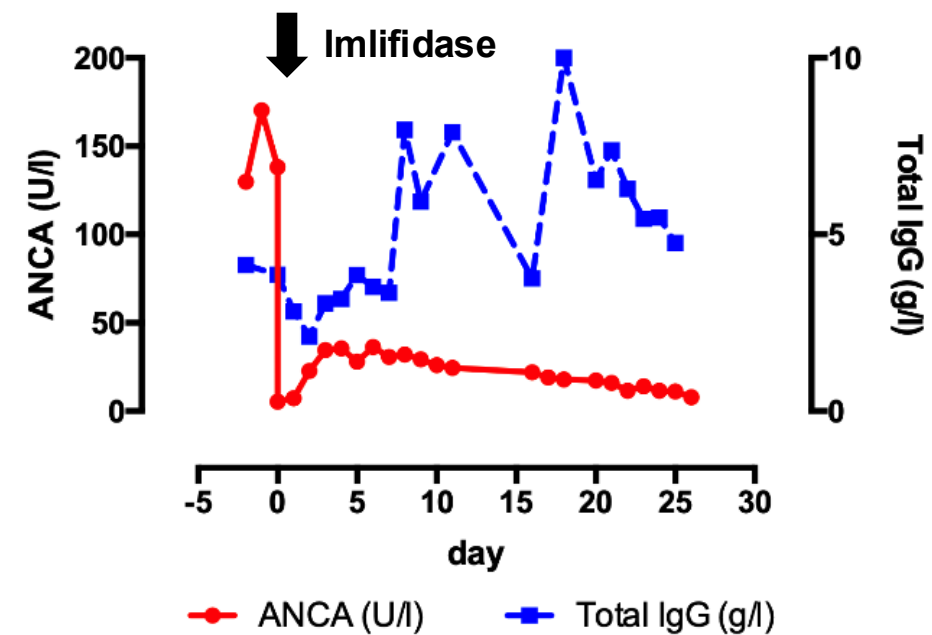


Imlifidase



Immunsuppression: Plex, Steroid, Cyclophosphamid, Rituximab

Novel treatment approaches: Imlifidase



Name of Study	Imlifidase in ANCA-associated vasculitis		
Protocol Number:	Phase: Investigator- Sponsored Trial	Country: Germany	
Principal Investigator	Adrian Schreiber / Philipp Enghard		
Indication	Severe ANCA associated vasculitis with Acute Respiratory Distress Syndrome due to pulmonary hemorrhage		
Investigational Agent	Imlifidase for injection (Hansa Biopharma AB)		
Study center(s)	Charité – Universitätsmedizin Berlin <ul style="list-style-type: none">• Charité Campus Mitte, Charitéplatz 1, 10117 Berlin• Campus Virchow Klinikum, Augustenburger Platz, 13353 Berlin• Campus Benjamin Franklin, <u>Hindenburgdamm</u> 30, 12203 Berlin		
Sample size	10 patients		

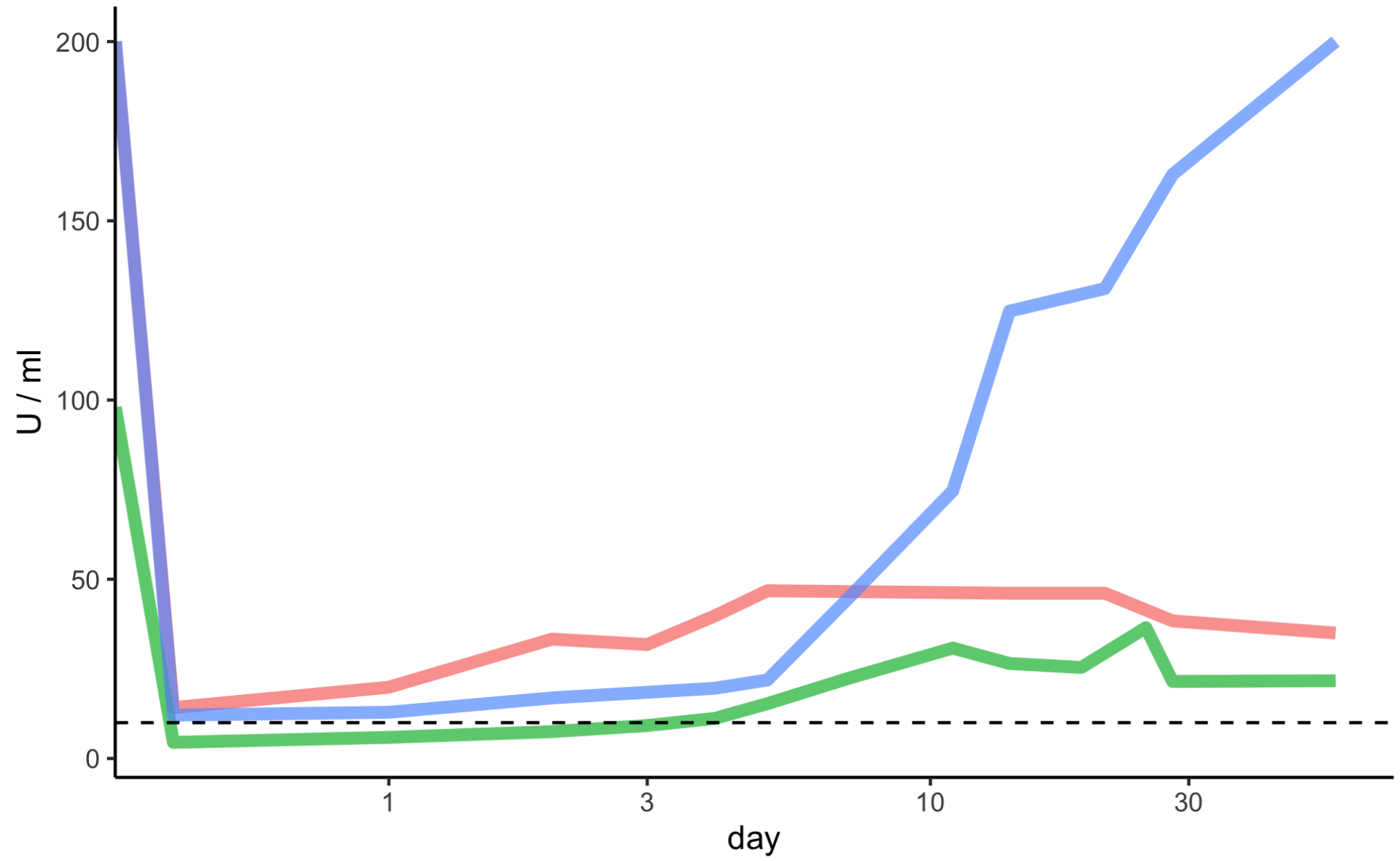
Janis Sonnemann



Philipp Enghard



ANCA titer after imlifidase



dashed line indicates reference range

Aknowledgements

AG Kettritz/Schreiber

Tanja Filipowski

Uwe Jerke

Lovis Kling

Dörte Lodka

Sylvia Lucke

Susanne Rolle

Janis Sonnemann

Jonas Zimmermann



Höpken Lab

Maria Zschummel

Mario Bunse

ECRC

FC Luft

Dominik Müller

Nicola Wilck

Volker Siffrin

Department of Nephrology

Kai-Uwe Eckardt

External collaborators

David Simon & Gerhard Krönke, Charité

Achim Leutz & Alexander Mildner, MDC Berlin

Kerstin Amann, FAU Erlangen

Markus Bieringer, Helios Klinikum Berlin

Udo Schneider, Charité Berlin

Philipp Enghard, Charité Berlin

Matthias Mack, UKR Regensburg

Andreas Linkermann, Mannheim

Ulf Panzer, UKE Hamburg

Liste der Referenzen

- Alberici, Federico et al. "Treatment goals in ANCA-associated vasculitis: defining success in a new era." *Frontiers in immunology* vol. 15 1409129. 13 Jun. 2024, doi:10.3389/fimmu.2024.1409129
- Assmann, Gerd et al. "Effects of ezetimibe, simvastatin, atorvastatin, and ezetimibe-statin therapies on non-cholesterol sterols in patients with primary hypercholesterolemia." *Current medical research and opinion* vol. 24,1 (2008): 249-59. doi:10.1185/030079908x253663
- Basu N, et al. Fatigue: a principal contributor to impaired quality of life in ANCA-associated vasculitis. *Rheumatology (Oxford)*. 2010 Jul;49(7):1383-90. doi: 10.1093/rheumatology/keq098.
- Basu, Neil et al. Markers for work disability in anti-neutrophil cytoplasmic antibody-associated vasculitis, *Rheumatology*, Volume 53, Issue 5, May 2014, Pages 953–956, <https://doi.org/10.1093/rheumatology/ket483>
- Bate, Sebastian et al. "The Improved Kidney Risk Score in ANCA-Associated Vasculitis for Clinical Practice and Trials." *Journal of the American Society of Nephrology : JASN* vol. 35,3 (2024): 335-346. doi:10.1681/ASN.0000000000000274
- Benichou, Nicolas et al. "Proteinuria and hematuria after remission induction are associated with outcome in ANCA-associated vasculitis." *Kidney international* vol. 103,6 (2023): 1144-1155. doi:10.1016/j.kint.2023.02.029
- Berden, Annelies E et al. "Histopathologic classification of ANCA-associated glomerulonephritis." *Journal of the American Society of Nephrology : JASN* vol. 21,10 (2010): 1628-36. doi:10.1681/ASN.2010050477
- Brar, Sandeep et al. "Association of Angiotensin-Converting Enzyme Inhibitor or Angiotensin Receptor Blocker Use With Outcomes After Acute Kidney Injury." *JAMA internal medicine* vol. 178,12 (2018): 1681-1690. doi:10.1001/jamainternmed.2018.4749
- rix SR. The Challenge of Assessing Remission and Relapse in ANCA Kidney Disease. *J Am Soc Nephrol*. 2024 Apr 1;35(4):395-397. doi: 10.1681/ASN.0000000000000331.
- Caravaca-Fontán, Fernando et al. Sodium-glucose cotransporter 2 inhibition in primary and secondary glomerulonephritis, *Nephrology Dialysis Transplantation*, Volume 39, Issue 2, February 2024, Pages 328–340, <https://doi.org/10.1093/ndt/gfad175>
- Cartin-Ceba, Rodrigo et al. "Diffuse Alveolar Hemorrhage Secondary to Antineutrophil Cytoplasmic Antibody-Associated Vasculitis: Predictors of Respiratory Failure and Clinical Outcomes." *Arthritis & rheumatology (Hoboken, N.J.)* vol. 68,6 (2016): 1467-76. doi:10.1002/art.39562
- Chalkia, Aglaia et al. "Avacopan for ANCA-associated vasculitis with hypoxic pulmonary haemorrhage." *Nephrology, dialysis, transplantation : official publication of the European Dialysis and Transplant Association - European Renal Association* vol. 39,9 (2024): 1473-1482. doi:10.1093/ndt/gfae020
- Chapman, Lara et al. Management of foot health in people with inflammatory arthritis: British Society for Rheumatology guideline scope, *Rheumatology*, Volume 61, Issue 10, October 2022, Pages 3907–3911, <https://doi.org/10.1093/rheumatology/keac340>
- Clifford, Alison H, and Jan Willem Cohen Tervaert. "Cardiovascular events and the role of accelerated atherosclerosis in systemic vasculitis." *Atherosclerosis* vol. 325 (2021): 8-15. doi:10.1016/j.atherosclerosis.2021.03.032

Liste der Referenzen

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

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

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

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

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

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

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

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

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

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

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

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

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

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

Liste der Referenzen

- Gopaluni, Seerapani et al. "Effect of Disease Activity at Three and Six Months After Diagnosis on Long-Term Outcomes in Antineutrophil Cytoplasmic Antibody-Associated Vasculitis." *Arthritis & rheumatology* (Hoboken, N.J.) vol. 71,5 (2019): 784-791. doi:10.1002/art.40776
- Grahammer, Florian et al. "The podocyte slit diaphragm--from a thin grey line to a complex signalling hub." *Nature reviews. Nephrology* vol. 9,10 (2013): 587-98. doi:10.1038/nrneph.2013.169
- Hakroush, Samy et al. "Bowman's capsule rupture links glomerular damage to tubulointerstitial inflammation in ANCA-associated glomerulonephritis." *Clinical and experimental rheumatology* vol. 39 Suppl 129,2 (2021): 27-31. doi:10.55563/clinexprheumatol/7eol6d
- Harper, Lorraine et al. "Treatment of fatigue with physical activity and behavioural change support in vasculitis: study protocol for an open-label randomised controlled feasibility study." *BMJ open* vol. 8,10 e023769. 30 Oct. 2018, doi:10.1136/bmjopen-2018-023769
- Harper, Lorraine et al. Management of fatigue with physical activity and behavioural change support in vasculitis: a feasibility study, *Rheumatology*, Volume 60, Issue 9, September 2021, Pages 4130–4140, <https://doi.org/10.1093/rheumatology/keaa890>
- Hayek, Salim S et al. "Soluble Urokinase Receptor and Acute Kidney Injury." *The New England journal of medicine* vol. 382,5 (2020): 416-426. doi:10.1056/NEJMoa1911481
- Heijl, Caroline et al. "Long-term patient survival in a Swedish population-based cohort of patients with ANCA-associated vasculitis." *RMD open* vol. 3,1 e000435. 13 Jul. 2017, doi:10.1136/rmdopen-2017-000435
- Hellmich, Bernhard et al. "EULAR recommendations for the management of ANCA-associated vasculitis: 2022 update." *Annals of the rheumatic diseases* vol. 83,1 30-47. 2 Jan. 2024, doi:10.1136/ard-2022-223764
- Heron, Vanessa et al. "The impact of antineutrophil cytoplasmic antibody-associated vasculitis on employment and work disability in an Australian population." *International journal of rheumatic diseases* vol. 24,7 (2021): 904-911. doi:10.1111/1756-185X.14131
- Hiepe, Falk, and Andreas Radbruch. "Plasma cells as an innovative target in autoimmune disease with renal manifestations." *Nature reviews. Nephrology* vol. 12,4 (2016): 232-40. doi:10.1038/nrneph.2016.20
- Hoffman, G S et al. "Wegener granulomatosis: an analysis of 158 patients." *Annals of internal medicine* vol. 116,6 (1992): 488-98. doi:10.7326/0003-4819-116-6-488
- Hollander, D, and R T Manning. "The use of alkylating agents in the treatment of Wegener's granulomatosis." *Annals of internal medicine* vol. 67,2 (1967): 393-8. doi:10.7326/0003-4819-67-2-393
- Holle et al. S3-Leitlinie der Deutschen Gesellschaft für Rheumatologie und Klinische Immunologie e. V. (DGRh). Diagnostik und Therapie der ANCA-assoziierten Vaskulitiden, Version: 1.1; Stand: 12.08.2024
- Hruskova, Zdenka et al. "Characteristics and Outcomes of Granulomatosis With Polyangiitis (Wegener) and Microscopic Polyangiitis Requiring Renal Replacement Therapy: Results From the European Renal Association-European Dialysis and Transplant Association Registry." *American journal of kidney diseases : the official journal of the National Kidney Foundation* vol. 66,4 (2015): 613-20. doi:10.1053/j.ajkd.2015.03.025
- Iudici, Michele et al. "Granulomatosis with polyangiitis: Study of 795 patients from the French Vasculitis Study Group registry." *Seminars in arthritis and rheumatism* vol. 51,2 (2021): 339-346. doi:10.1016/j.semarthrit.2021.02.002
- Becherucci, Francesca et al. "A Clinical Workflow for Cost-Saving High-Rate Diagnosis of Genetic Kidney Diseases." *Journal of the American Society of Nephrology : JASN* vol. 34,4 (2023): 706-720. doi:10.1681/ASN.0000000000000076

Liste der Referenzen

- Jayne, David R W et al. "Avacopan for the Treatment of ANCA-Associated Vasculitis." *The New England journal of medicine* vol. 384,7 (2021): 599-609. doi:10.1056/NEJMoa2023386
- Jayne, David R W et al. "Randomized Trial of C5a Receptor Inhibitor Avacopan in ANCA-Associated Vasculitis." *Journal of the American Society of Nephrology : JASN* vol. 28,9 (2017): 2756-2767. doi:10.1681/ASN.2016111179
- Jayne, David R W et al. "Avacopan for the Treatment of ANCA-Associated Vasculitis." *The New England journal of medicine* vol. 384,7 (2021): 599-609. doi:10.1056/NEJMoa2023386
- Jayne, David R W et al. "Randomized trial of plasma exchange or high-dosage methylprednisolone as adjunctive therapy for severe renal vasculitis." *Journal of the American Society of Nephrology : JASN* vol. 18,7 (2007): 2180-8. doi:10.1681/ASN.2007010090
- Jayne, David R W et al. "Avacopan for the Treatment of ANCA-Associated Vasculitis." *The New England journal of medicine* vol. 384,7 (2021): 599-609. doi:10.1056/NEJMoa2023386
- June, Carl H et al. "CAR T cell immunotherapy for human cancer." *Science (New York, N.Y.)* vol. 359,6382 (2018): 1361-1365. doi:10.1126/science.aar6711
- Junek, Mats L et al. "Risk of Relapse of Antineutrophil Cytoplasmic Antibody-Associated Vasculitis in a Randomized Controlled Trial of Plasma Exchange and Glucocorticoids." *Arthritis & rheumatology (Hoboken, N.J.)* vol. 76,9 (2024): 1431-1438. doi:10.1002/art.42843
- Kaufeld, Jessica et al. "Atypical Hemolytic and Uremic Syndrome Triggered by Infection With SARS-CoV2." *Kidney international reports* vol. 6,10 (2021): 2709-2712. doi:10.1016/j.ekir.2021.07.004
- Klapa, Sebastian et al. "Low Concentrations of C5a Complement Receptor Antibodies Are Linked to Disease Activity and Relapse in Antineutrophil Cytoplasmic Autoantibody-Associated Vasculitis." *Arthritis & rheumatology (Hoboken, N.J.)* vol. 75,5 (2023): 760-767. doi:10.1002/art.42410
- Kochi, Masako et al. "Chronic kidney disease, inflammation, and cardiovascular disease risk in rheumatoid arthritis." *Journal of cardiology* vol. 71,3 (2018): 277-283. doi:10.1016/j.jcc.2017.08.008
- Krasselt, Marco et al. "48-jährige Patientin mit krustenartigen Veränderungen an den Füßen" [48-year-old woman with crusty alterations on the feet]. *Deutsche medizinische Wochenschrift (1946)* vol. 148,18 (2023): 1155-1156. doi:10.1055/a-2061-5060
- Krasselt, Marco L, and Julia U Holle. "ANCA-assoziierte Vaskulitis" [ANCA-associated vasculitis]. *Innere Medizin (Heidelberg, Germany)* vol. 63,9 (2022): 947-960. doi:10.1007/s00108-022-01386-w
- Kronbichler, Andreas et al. "Diagnosis and management of ANCA-associated vasculitis." *Lancet (London, England)* vol. 403,10427 (2024): 683-698. doi:10.1016/S0140-6736(23)01736-1
- Kronbichler, Andreas et al. "Plasma exchange in ANCA-associated vasculitis: the pro position." *Nephrology, dialysis, transplantation : official publication of the European Dialysis and Transplant Association - European Renal Association* vol. 36,2 (2021): 227-231. doi:10.1093/ndt/gfaa311
- Kronbichler, Andreas et al. "Diagnosis and management of ANCA-associated vasculitis." *Lancet (London, England)* vol. 403,10427 (2024): 683-698. doi:10.1016/S0140-6736(23)01736-1

Liste der Referenzen

- L'Imperio, Vincenzo et al. "Bowman's capsule rupture on renal biopsy improves the outcome prediction of ANCA-associated glomerulonephritis classifications." *Annals of the rheumatic diseases* vol. 81,6 (2022): e95. doi:10.1136/annrheumdis-2020-217979
- Lejeune, Margaux et al. "Bispecific, T-Cell-Recruiting Antibodies in B-Cell Malignancies." *Frontiers in immunology* vol. 11 762. 7 May. 2020, doi:10.3389/fimmu.2020.00762
- Little, Mark A et al. "Early mortality in systemic vasculitis: relative contribution of adverse events and active vasculitis." *Annals of the rheumatic diseases* vol. 69,6 (2010): 1036-43. doi:10.1136/ard.2009.109389
- Mach, François et al. "2019 ESC/EAS Guidelines for the management of dyslipidaemias: lipid modification to reduce cardiovascular risk." *European heart journal* vol. 41,1 (2020): 111-188. doi:10.1093/eurheartj/ehz455
- Manolio, Teri A et al. "Finding the missing heritability of complex diseases." *Nature* vol. 461,7265 (2009): 747-53. doi:10.1038/nature08494
- Maunz, Annika et al. "Association of the AAV-PRO questionnaire with established outcome measures in AAV." *Rheumatology (Oxford, England)* vol. 63,1 (2024): 174-180. doi:10.1093/rheumatology/kead199
- McAduo, Stephen P et al. "Long-term follow-up of a combined rituximab and cyclophosphamide regimen in renal anti-neutrophil cytoplasm antibody-associated vasculitis." *Nephrology, dialysis, transplantation : official publication of the European Dialysis and Transplant Association - European Renal Association* vol. 34,1 (2019): 63-73. doi:10.1093/ndt/gfx378
- McGregor, JulieAnne G et al. "Adverse events and infectious burden, microbes and temporal outline from immunosuppressive therapy in antineutrophil cytoplasmic antibody-associated vasculitis with native renal function." *Nephrology, dialysis, transplantation : official publication of the European Dialysis and Transplant Association - European Renal Association* vol. 30 Suppl 1,Suppl 1 (2015): i171-81. doi:10.1093/ndt/gfv045
- Monti, Sara et al. "Factors influencing patient-reported outcomes in ANCA-associated vasculitis: correlates of the Patient Global Assessment." *Seminars in arthritis and rheumatism* vol. 56 (2022): 152048. doi:10.1016/j.semarthrit.2022.152048
- Moran, Sarah M et al. "The Clinical Application of Urine Soluble CD163 in ANCA-Associated Vasculitis." *Journal of the American Society of Nephrology : JASN* vol. 32,11 (2021): 2920-2932. doi:10.1681/ASN.2021030382
- Casal Moura, Marta et al. "Management of antineutrophil cytoplasmic antibody-associated vasculitis with glomerulonephritis as proposed by the ACR 2021, EULAR 2022 and KDIGO 2021 guidelines/recommendations." *Nephrology, dialysis, transplantation : official publication of the European Dialysis and Transplant Association - European Renal Association* vol. 38,11 (2023): 2637-2651. doi:10.1093/ndt/gfad090
- Casal Moura, Marta et al. "Maintenance of Remission and Risk of Relapse in Myeloperoxidase-Positive ANCA-Associated Vasculitis with Kidney Involvement." *Clinical journal of the American Society of Nephrology : CJASN* vol. 18,1 (2023): 47-59. doi:10.2215/CJN.06460622
- Casal Moura, Marta et al. "Maintenance of Remission and Risk of Relapse in Myeloperoxidase-Positive ANCA-Associated Vasculitis with Kidney Involvement." *Clinical journal of the American Society of Nephrology : CJASN* vol. 18,1 (2023): 47-59. doi:10.2215/CJN.06460622
- Nicholls, Stephen J et al. "Effect of Evolocumab on Progression of Coronary Disease in Statin-Treated Patients: The GLAGOV Randomized Clinical Trial." *JAMA* vol. 316,22 (2016): 2373-2384. doi:10.1001/jama.2016.16951
- O'Malley, Lucy et al. "The Longitudinal Course of Fatigue in Antineutrophil Cytoplasmic Antibody-associated Vasculitis." *The Journal of rheumatology* vol. 47,4 (2020): 572-579. doi:10.3899/jrheum.190113

Liste der Referenzen

- Hildreth, Andrew D et al. "Single-cell sequencing of human white adipose tissue identifies new cell states in health and obesity." *Nature immunology* vol. 22,5 (2021): 639-653. doi:10.1038/s41590-021-00922-4
- O'Sullivan, Kim M, and Stephen R Holdsworth. "Neutrophil Extracellular Traps: A Potential Therapeutic Target in MPO-ANCA Associated Vasculitis?." *Frontiers in immunology* vol. 12 635188. 15 Mar. 2021, doi:10.3389/fimmu.2021.635188
- Odler, Balazs et al. "Risk factors for serious infections in ANCA-associated vasculitis." *Annals of the rheumatic diseases* vol. 82,5 (2023): 681-687. doi:10.1136/ard-2022-223401
- Odler, Balazs et al. "Challenges of defining renal response in ANCA-associated vasculitis: call to action?." *Clinical kidney journal* vol. 16,6 965-975. 11 Jan. 2023, doi:10.1093/ckj/sfad009
- Oristrell, J et al. "Relapse rate and renal prognosis in ANCA-associated vasculitis according to long-term ANCA patterns." *Clinical and experimental immunology* vol. 203,2 (2021): 209-218. doi:10.1111/cei.13530
- Ostendorf, Lennard et al. "Daratumumab for the treatment of refractory ANCA-associated vasculitis." *RMD open* vol. 9,1 (2023): e002742. doi:10.1136/rmdopen-2022-002742
- Perkovic, Vlado et al. "Alternative Complement Pathway Inhibition with Iptacopan in IgA Nephropathy." *The New England journal of medicine*, 10.1056/NEJMoa2410316. 25 Oct. 2024, doi:10.1056/NEJMoa2410316
- Perna, Alessandro et al. "Kidney transplantation in patients with ANCA-associated vasculitis is associated with a high incidence of post-transplant cancer." *Journal of nephrology* vol. 37,6 (2024): 1611-1619. doi:10.1007/s40620-024-01951-6
- Pittam, Bradley et al. "The prevalence and impact of depression in primary systemic vasculitis: a systematic review and meta-analysis." *Rheumatology international* vol. 40,8 (2020): 1215-1221. doi:10.1007/s00296-020-04611-7
- Prskalo, Luka et al. "Urinary CD4 + T Cells Predict Renal Relapse in ANCA-Associated Vasculitis." *Journal of the American Society of Nephrology : JASN* vol. 35,4 (2024): 483-494. doi:10.1681/ASN.0000000000000311
- Quartuccio, Luca et al. "Unmet needs in ANCA-associated vasculitis: Physicians' and patients' perspectives." *Frontiers in immunology* vol. 14 1112899. 23 Feb. 2023, doi:10.3389/fimmu.2023.1112899
- Quartuccio, Luca et al. "Alveolar haemorrhage in ANCA-associated vasculitis: Long-term outcome and mortality predictors." *Journal of autoimmunity* vol. 108 (2020): 102397. doi:10.1016/j.jaut.2019.102397
- Quinn, Kaitlin A et al. "An international Delphi exercise to identify items of importance for measuring response to treatment in ANCA-associated vasculitis." *Seminars in arthritis and rheumatism* vol. 55 (2022): 152021. doi:10.1016/j.semarthrit.2022.152021
- Ridker, Paul M et al. "Inhibition of Interleukin-1 β by Canakinumab and Cardiovascular Outcomes in Patients With Chronic Kidney Disease." *Journal of the American College of Cardiology* vol. 71,21 (2018): 2405-2414. doi:10.1016/j.jacc.2018.03.490
- Ridker, Paul M et al. "Inhibition of Interleukin-1 β by Canakinumab and Cardiovascular Outcomes in Patients With Chronic Kidney Disease." *Journal of the American College of Cardiology* vol. 71,21 (2018): 2405-2414. doi:10.1016/j.jacc.2018.03.490
- Robson, Joanna et al. "Damage in the anca-associated vasculitides: long-term data from the European vasculitis study group (EUVAS) therapeutic trials." *Annals of the rheumatic diseases* vol. 74,1 (2015): 177-84. doi:10.1136/annrheumdis-2013-203927

Liste der Referenzen

- Sagmeister, Michael S et al. "Kidney transplantation in ANCA-associated vasculitis." *Journal of nephrology* vol. 32,6 (2019): 919-926. doi:10.1007/s40620-019-00642-x
- Salmela, Anna et al. "Prognostic Factors for Survival and Relapse in ANCA-Associated Vasculitis with Renal Involvement: A Clinical Long-Term Follow-Up Study." *International journal of nephrology* vol. 2018 6369814. 16 Oct. 2018, doi:10.1155/2018/6369814
- Sachez-Alamo, Beatriz et al. "Long-term outcome of kidney function in patients with ANCA-associated vasculitis." *Nephrology, dialysis, transplantation : official publication of the European Dialysis and Transplant Association - European Renal Association* vol. 39,9 (2024): 1483-1493. doi:10.1093/ndt/gfae018
- Sachez-Alamo, Beatriz et al. "Long-term outcome of kidney function in patients with ANCA-associated vasculitis." *Nephrology, dialysis, transplantation : official publication of the European Dialysis and Transplant Association - European Renal Association* vol. 39,9 (2024): 1483-1493. doi:10.1093/ndt/gfae018
- Sánchez Álamo, Beatriz et al. "Long-term outcomes and prognostic factors for survival of patients with ANCA-associated vasculitis." *Nephrology, dialysis, transplantation : official publication of the European Dialysis and Transplant Association - European Renal Association* vol. 38,7 (2023): 1655-1665. doi:10.1093/ndt/gfac320
- Sayer, Matthew et al. "Cardiovascular Disease in Anti-neutrophil Cytoplasm Antibody-Associated Vasculitis." *Current rheumatology reports* vol. 26,1 (2024): 12-23. doi:10.1007/s11926-023-01123-8
- Schäfer, Ann-Kathrin et al. "Case Report: High-dose immunoglobulins prior to plasma exchange in severe pulmonary renal syndrome." *Frontiers in immunology* vol. 14 1210321. 9 Jun. 2023, doi:10.3389/fimmu.2023.1210321
- Scherbacher, Paul J et al. "Prospective study of complications and sequelae of glucocorticoid therapy in ANCA-associated vasculitis." *RMD open* vol. 10,1 e003956. 29 Feb. 2024, doi:10.1136/rmdopen-2023-003956
- Schunk, Stefan J et al. "Measurement of urinary Dickkopf-3 uncovered silent progressive kidney injury in patients with chronic obstructive pulmonary disease." *Kidney international* vol. 100,5 (2021): 1081-1091. doi:10.1016/j.kint.2021.06.029
- Schunk, Stefan J et al. "Association between urinary dickkopf-3, acute kidney injury, and subsequent loss of kidney function in patients undergoing cardiac surgery: an observational cohort study." *Lancet (London, England)* vol. 394,10197 (2019): 488-496. doi:10.1016/S0140-6736(19)30769-X
- Speer, Thimoteus et al. "Urinary DKK3 as a biomarker for short-term kidney function decline in children with chronic kidney disease: an observational cohort study." *The Lancet. Child & adolescent health* vol. 7,6 (2023): 405-414. doi:10.1016/S2352-4642(23)00049-4
- Schunk, Stefan J et al. "WNT-β-catenin signalling - a versatile player in kidney injury and repair." *Nature reviews. Nephrology* vol. 17,3 (2021): 172-184. doi:10.1038/s41581-020-00343-w
- Schupp, Jonas Christian et al. "Usefulness of Cyclophosphamide Pulse Therapy in Interstitial Lung Diseases." *Respiration; international review of thoracic diseases* vol. 91,4 (2016): 296-301. doi:10.1159/000445031
- Shochet, Lani et al. "Animal Models of ANCA Associated Vasculitis." *Frontiers in immunology* vol. 11 525. 9 Apr. 2020, doi:10.3389/fimmu.2020.00525
- Silva, Rita M et al. "Renal Transplantation in Antineutrophil Cytoplasmic Antibody-Associated Vasculitis: A Single-Center 10-Year Experience." *Transplantation proceedings* vol. 55,6 (2023): 1396-1399. doi:10.1016/j.transproceed.2023.04.018

Liste der Referenzen

- Simms-Williams, Nikita et al. "Effect of combination treatment with glucagon-like peptide-1 receptor agonists and sodium-glucose cotransporter-2 inhibitors on incidence of cardiovascular and serious renal events: population based cohort study." *BMJ (Clinical research ed.)* vol. 385 e078242. 25 Apr. 2024, doi:10.1136/bmj-2023-078242
- Slot, Marjan C et al. "Renal survival and prognostic factors in patients with PR3-ANCA associated vasculitis with renal involvement." *Kidney international* vol. 63,2 (2003): 670-7. doi:10.1046/j.1523-1755.2003.00769.x
- Smith, Rona M et al. "Rituximab versus azathioprine for maintenance of remission for patients with ANCA-associated vasculitis and relapsing disease: an international randomised controlled trial." *Annals of the rheumatic diseases* vol. 82,7 (2023): 937-944. doi:10.1136/ard-2022-223559
- Sonnemann, Janis et al. "Urinary T Cells Identify Renal Antineutrophil Cytoplasmic Antibody-Associated Vasculitis and Predict Prognosis: A Proof of Concept Study." *Kidney international reports* vol. 8,4 871-883. 18 Jan. 2023, doi:10.1016/j.ekir.2023.01.013
- Specks, Ulrich et al. "Efficacy of remission-induction regimens for ANCA-associated vasculitis." *The New England journal of medicine* vol. 369,5 (2013): 417-27. doi:10.1056/NEJMoa1213277
- Stone, John H et al. "Rituximab versus cyclophosphamide for ANCA-associated vasculitis." *The New England journal of medicine* vol. 363,3 (2010): 221-32. doi:10.1056/NEJMoa0909905
- Specks, Ulrich et al. "Efficacy of remission-induction regimens for ANCA-associated vasculitis." *The New England journal of medicine* vol. 369,5 (2013): 417-27. doi:10.1056/NEJMoa1213277
- Strand, Vibeke et al. "The impact of treatment with avacopan on health-related quality of life in antineutrophil cytoplasmic antibody-associated vasculitis: a post-hoc analysis of data from the ADVOCATE trial." *The Lancet. Rheumatology* vol. 5,8 (2023): e451-e460. doi:10.1016/S2665-9913(23)00092-9
- Tampe, Désirée et al. "Different Patterns of Kidney Fibrosis Are Indicative of Injury to Distinct Renal Compartments." *Cells* vol. 10,8 2014. 6 Aug. 2021, doi:10.3390/cells10082014
- SPRINT Research Group et al. "A Randomized Trial of Intensive versus Standard Blood-Pressure Control." *The New England journal of medicine* vol. 373,22 (2015): 2103-16. doi:10.1056/NEJMoa1511939
- Trivioli, Giorgio et al. "Slowly progressive anti-neutrophil cytoplasmic antibody-associated renal vasculitis: clinico-pathological characterization and outcome." *Clinical kidney journal* vol. 14,1 332-340. 6 Sep. 2020, doi:10.1093/ckj/sfaa139
- Trivioli, Giorgio et al. "Genetics of ANCA-associated vasculitis: role in pathogenesis, classification and management." *Nature reviews. Rheumatology* vol. 18,10 (2022): 559-574. doi:10.1038/s41584-022-00819-y
- van Eeden, Charmaine et al. "Fatigue in ANCA-associated vasculitis (AAV) and systemic sclerosis (SSc): similarities with Myalgic encephalomyelitis/Chronic Fatigue Syndrome (ME/CFS). A critical review of the literature." *Expert review of clinical immunology* vol. 18,10 (2022): 1049-1070. doi:10.1080/1744666X.2022.2116002
- van Eeden, Charmaine et al. "Myalgic encephalomyelitis/chronic fatigue syndrome (ME/CFS) and fibromyalgia: PR3-versus MPO-ANCA-associated vasculitis, an exploratory cross-sectional study." *Lancet regional health. Americas* vol. 20 100460. 27 Feb. 2023, doi:10.1016/j.lana.2023.100460
- Visseren, Frank L J et al. "2021 ESC Guidelines on cardiovascular disease prevention in clinical practice." *European heart journal* vol. 42,34 (2021): 3227-3337. doi:10.1093/eurheartj/ehab484

Liste der Referenzen

- Wallace, Zachary S et al. "Disease Activity, Antineutrophil Cytoplasmic Antibody Type, and Lipid Levels in Antineutrophil Cytoplasmic Antibody-Associated Vasculitis." *Arthritis & rheumatology (Hoboken, N.J.)* vol. 71,11 (2019): 1879-1887. doi:10.1002/art.41006
- Walsh, C L et al. "Imaging intact human organs with local resolution of cellular structures using hierarchical phase-contrast tomography." *Nature methods* vol. 18,12 (2021): 1532-1541. doi:10.1038/s41592-021-01317-x
- Walsh, Michael et al. "Plasma Exchange and Glucocorticoids in Severe ANCA-Associated Vasculitis." *The New England journal of medicine* vol. 382,7 (2020): 622-631. doi:10.1056/NEJMoa1803537
- Walsh, Michael et al. "The effects of plasma exchange in patients with ANCA-associated vasculitis: an updated systematic review and meta-analysis." *BMJ (Clinical research ed.)* vol. 376 e064604. 25 Feb. 2022, doi:10.1136/bmj-2021-064604
- WALTON, E W. "Giant-cell granuloma of the respiratory tract (Wegener's granulomatosis)." *British medical journal* vol. 2,5091 (1958): 265-70. doi:10.1136/bmj.2.5091.265
- Weiner, Maria et al. "Outcome and treatment of elderly patients with ANCA-associated vasculitis." *Clinical journal of the American Society of Nephrology : CJASN* vol. 10,7 (2015): 1128-35. doi:10.2215/CJN.00480115
- Weppner, Gesche et al. "In situ detection of PR3-ANCA+ B cells and alterations in the variable region of immunoglobulin genes support a role of inflamed tissue in the emergence of auto-reactivity in granulomatosis with polyangiitis." *Journal of autoimmunity* vol. 93 (2018): 89-103. doi:10.1016/j.jaut.2018.07.004
- Windpessl, Martin et al. "Preventing infections in immunocompromised patients with kidney diseases: vaccines and antimicrobial prophylaxis." *Nephrology, dialysis, transplantation : official publication of the European Dialysis and Transplant Association - European Renal Association* vol. 38,Supplement_2 (2023): ii40-ii49. doi:10.1093/ndt/gfad080
- Windpessl, Martin et al. "Preventing infections in immunocompromised patients with kidney diseases: vaccines and antimicrobial prophylaxis." *Nephrology, dialysis, transplantation : official publication of the European Dialysis and Transplant Association - European Renal Association* vol. 38,Supplement_2 (2023): ii40-ii49. doi:10.1093/ndt/gfad080
- Xiao, Hong et al. "C5a receptor (CD88) blockade protects against MPO-ANCA GN." *Journal of the American Society of Nephrology : JASN* vol. 25,2 (2014): 225-31. doi:10.1681/ASN.2013020143
- Zimmermann, Jonas et al. "Avacopan in Anti-Neutrophil Cytoplasmic Autoantibodies-Associated Vasculitis in a Real-World Setting." *Kidney international reports* vol. 9,9 2803-2808. 6 Jul. 2024, doi:10.1016/j.ekir.2024.07.007
- Zimmermann J, Sonnemann J, Jabs WJ, et al. Avacopan in Anti-Neutrophil Cytoplasmic Autoantibodies-Associated Vasculitis in a Real-World Setting. *Kidney Int Rep.* 2024;9(9):2803-2808. Published 2024 Jul 6. doi:10.1016/j.ekir.2024.07.007
- Zonozi, Reza et al. "Real-World Experience With Avacopan in Antineutrophil Cytoplasmic Autoantibody-Associated Vasculitis." *Kidney international reports* vol. 9,6 1783-1791. 26 Mar. 2024, doi:10.1016/j.ekir.2024.03.022

Gekürzte Verschreibungsinformationen

Schweiz:

Tavneos®. Z: Avacopan. **I:** Tavneos, als ergänzende Therapie zu einer immunsuppressiven Standardbehandlung auf Basis von Rituximab oder Cyclophosphamid mit Glukokortikoiden, ist für die Behandlung erwachsener Patienten mit schwerer aktiver ANCA Vaskulitis (GPA/MPA) indiziert. **D:** Orale Einnahme morgens und abends 2x täglich 30 mg (3 Kapseln zu je 10 mg) mit Nahrung. **KI:** Überempfindlichkeit gegen den Wirkstoff oder einen der Hilfsstoffe. **VM:** Hepatotoxizität; Angioödem; Überwachung des Blutbildes (weisse Blutkörperchen); Schwere Infektionen; Reaktivierung des Hepatitis-B-Virus; Herzbeschwerden; Bösartige Tumore; Macroglycerinhydroxystearat. **S/S:** Eine Anwendung während der Schwangerschaft und bei Frauen im gebärfähigen Alter, die keine Verhütungsmethode anwenden, ist nicht empfohlen. Es ist nicht bekannt, ob Avacopan in die Muttermilch ausgeschieden wird. Der Nutzen des Stillens für das Kind sollte gegen den Nutzen der Behandlung für die Patientin abgewogen werden. **UW:** Sehr häufig: Infektion der oberen Atemwege, Nasopharyngitis; Kopfschmerzen; Erbrechen, Durchfall, Übelkeit; erhöhter Lebertest; verminderte Anzahl weisser Blutkörperchen. Häufig: Lungenentzündung, Infektion der unteren Atemwege, Influenza, Bronchitis, Zellulitis, Infektion der Harnwege, Herpes zoster, Sinusitis, orale Candidose, Herpes im Mundbereich, Otitis media, Rhinitis, Gastroenteritis; Neutropenie; Oberbauchschmerzen; Anstieg der Kreatinphosphokinase im Blut. Gelegentlich: 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.