VASCULARITES À ANCA : MALGRÉ LES CONSENSUS, TOUJOURS DES CONTROVERSES ?

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CanVasc

Disclosures (past 5 years)

Name: Christian Pagnoux

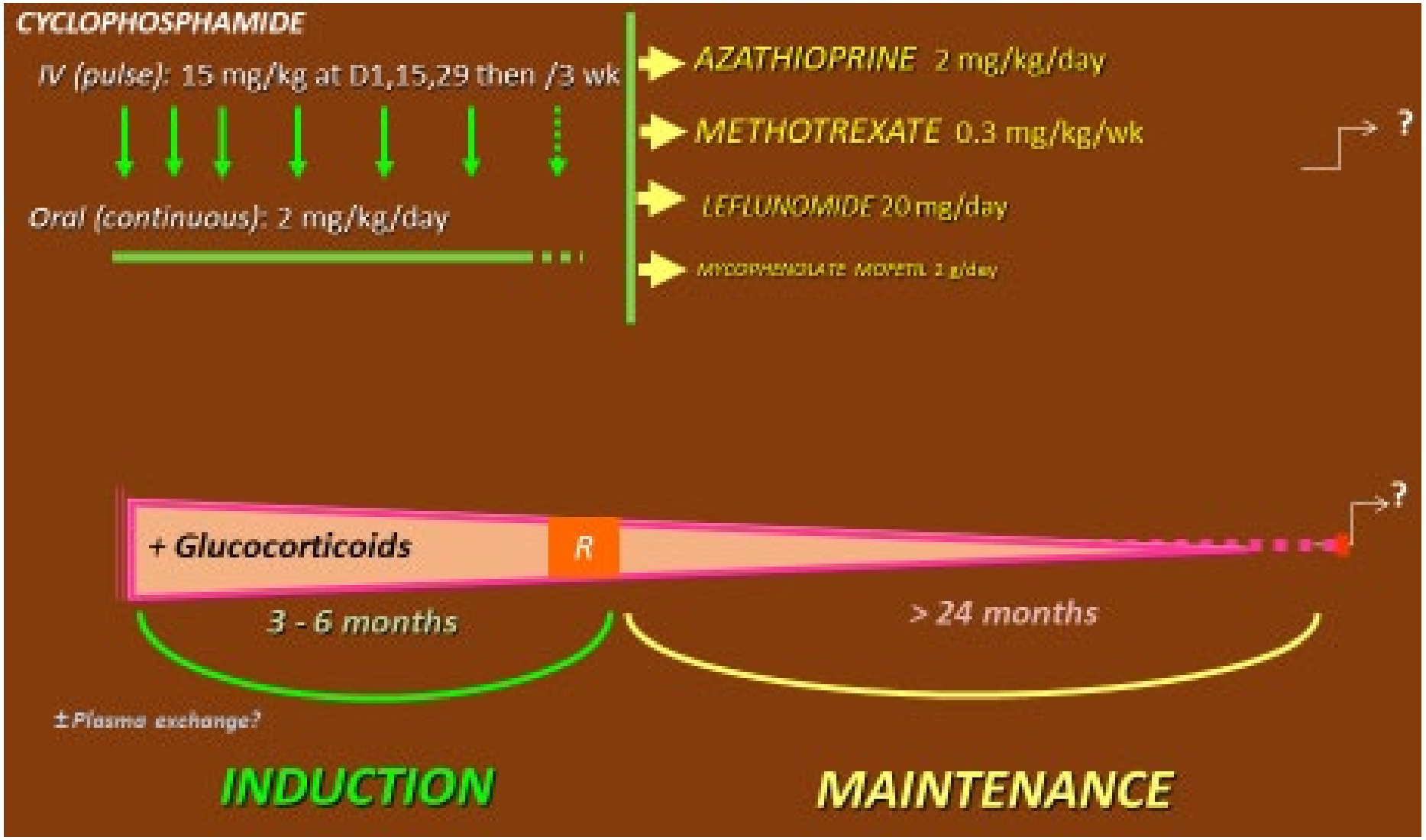
Credentials: MD, MPH, MSc, Vice-President of CanVasc, member of VCRC steering committee

- **Consulting fees:** Astra-Zeneca, Hoffmann-La Roche, BMS, GSK, Sanofi, ChemoCentryx, Otsuka
- **Grants/research support** (CanVasc): Hoffmann-La Roche, GSK, AmGen, Pfizer, TEVA, Otsuka
- **Speaker's honoraria:** Hoffmann-La Roche, GSK, Astra-Zeneca, Otsuka

Objectifs

- Réviser brièvement les messages-clés des plus récents consensus internationaux sur les choix de thérapies d'induction, de maintien et lors de rechutes, pour les vascularites à ANCA.
- Définir ce que représente la rémission clinique, et démontrer le bénéfice et les limitations d'utilisation de marqueurs d'atteinte de rémission clinique dans le suivi clinique de patients atteints de ces vasculites
  - indice BVAS
  - taux d'ANCA
  - suivi radiologique
  - fonction rénale, examen d'urine ou autre paramètre de suivi utile.
- Discuter des conduites cliniques à adopter lors de différentes situations cliniques
  - rémission clinique et persistance des taux sanguins d'ANCA
  - rémission clinique et réapparition des taux sanguins d'ANCA
  - hématurie en réapparition malgré une rémission clinique
  - régression partielle de lésions de la sphère ORL (croûtes nasales, lésion trachéales)
- Débattre de la durée de la thérapie de rémission, à la suite d'un traitement d'induction ou lors d'une rechute, et des risques inhérents à sa poursuite ou son arrêt.
- Élaborer un schéma d'investigation et de traitement en présence de formes réfractaires.

Il y a 15 ans…


CYCLOPHOSPHAMIDE
IV (pulse): 15 mg/kg at D1,15,29 then /3 wk
Oral (continuous): 2 mg/kg/day
AZATHIOPRINE 2 mg/kg/day
METHOTREXATE 0.3 mg/kg/wk
LEFLUNOMIDE 20 mg/day
MYCOPHENOLATE MOFETIL 1 g/day
?
+ Glucocorticoids
R
?
3 - 6 months
> 24 months
± Plasma exchange?
INDUCTION
MAINTENANCE

Principles of treatment of severe, systemic G/MPA

CYCLOPHOSPHAMIDE

IV (pulse): 15 mg/kg at D1,15,29 then /3 wk

Oral (continuous): 2 mg/kg/day

RITUXIMAB

1 g at D1 & 15 (or 4 x 375 mg/m²/wk)

+ Glucocorticoids

R

3 - 6 months

± Plasma exchange?

INDUCTION

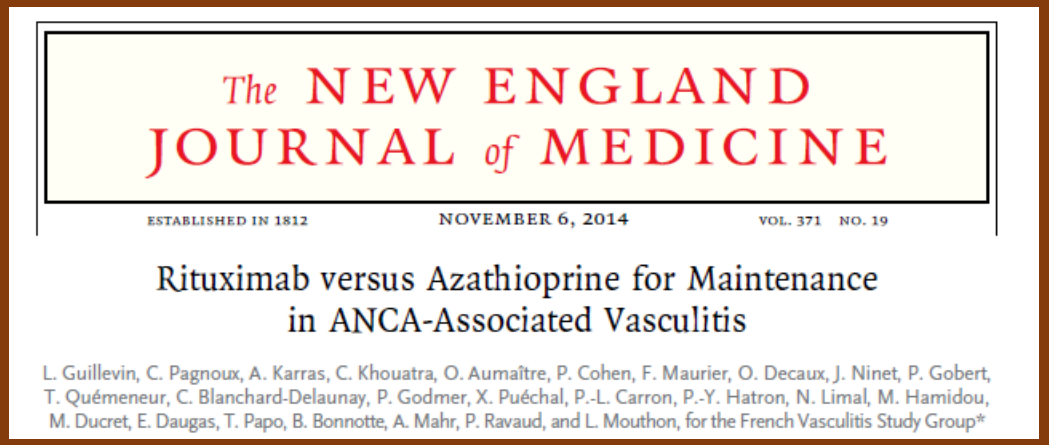
MAINTENANCE

ORIGINAL ARTICLE 2010

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. Tchao, M.D., Lisa Webber, R.N., Linna Ding, M.D., Ph.D., Lourdes P. Sejismundo, 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*

Principles of treatment of severe, systemic G/MPA



(CYCLOPHOSPHAMIDE)

RITUXIMAB
1 g at D1 & 15

RITUXIMAB 500-1000 mg q4-6 months

?

+ Glucocorticoids

R

3 - 6 months

> 18 months

± Plasma exchange?

INDUCTION

MAINTENANCE

The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE

Plasma Exchange and Glucocorticoids in Severe ANCA-Associated Vasculitis

M. Walsh, P.A. Merkel, C.-A. Peh, W.M. Szpirt, X. Puéchal, S. Fujimoto, C.M. Hawley, N. Khalidi, O. Floßmann, R. Wald, L.P. Girard, A. Levin, G. Gregorini, L. Harper, W.F. Clark, C. Pagnoux, U. Specks, L. Smyth, V. Tesar, T. Ito-Ihara, J.R. de Zoysa, W. Szczeklik, L.F. Flores-Suárez, S. Carette, L. Guillevin, C.D. Pusey, A.L. Casian, B. Brezina, A. Mazzetti, C.A. McAlear, E. Broadhurst, D. Reidlinger, S. Mehta, N. Ives, and D.R.W. Jayne, for the PEXIVAS Investigators*

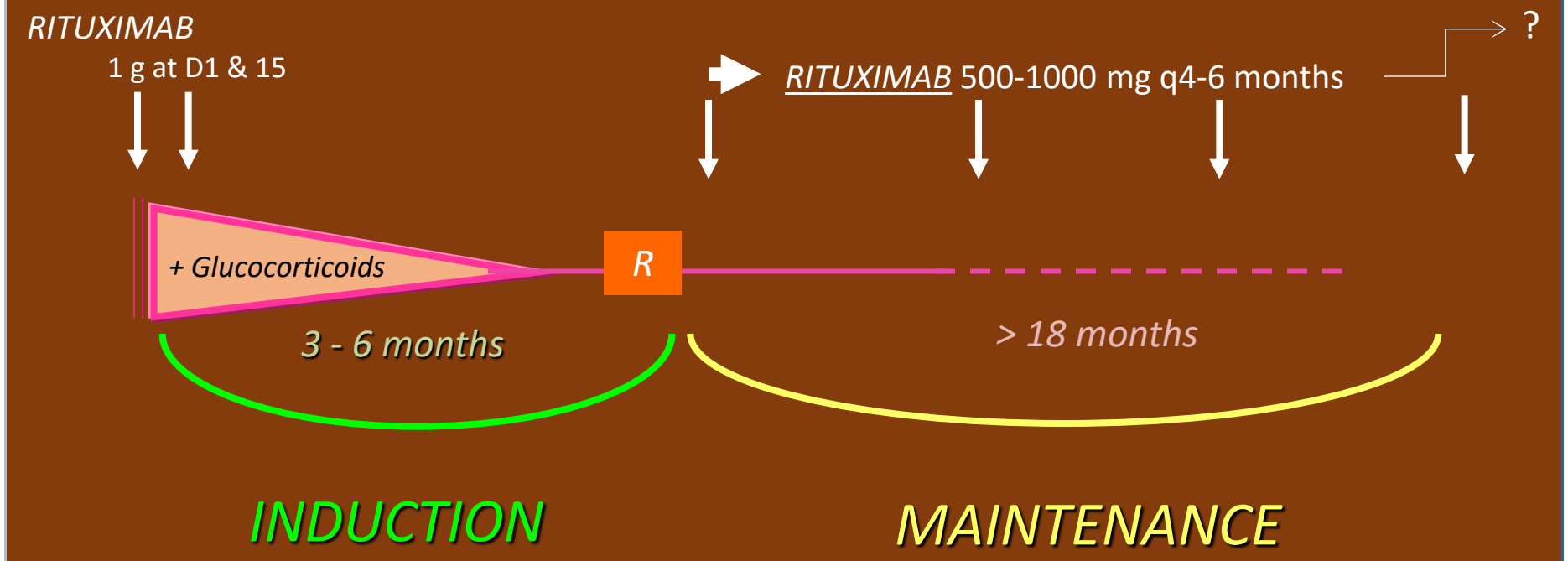
February 13, 2020

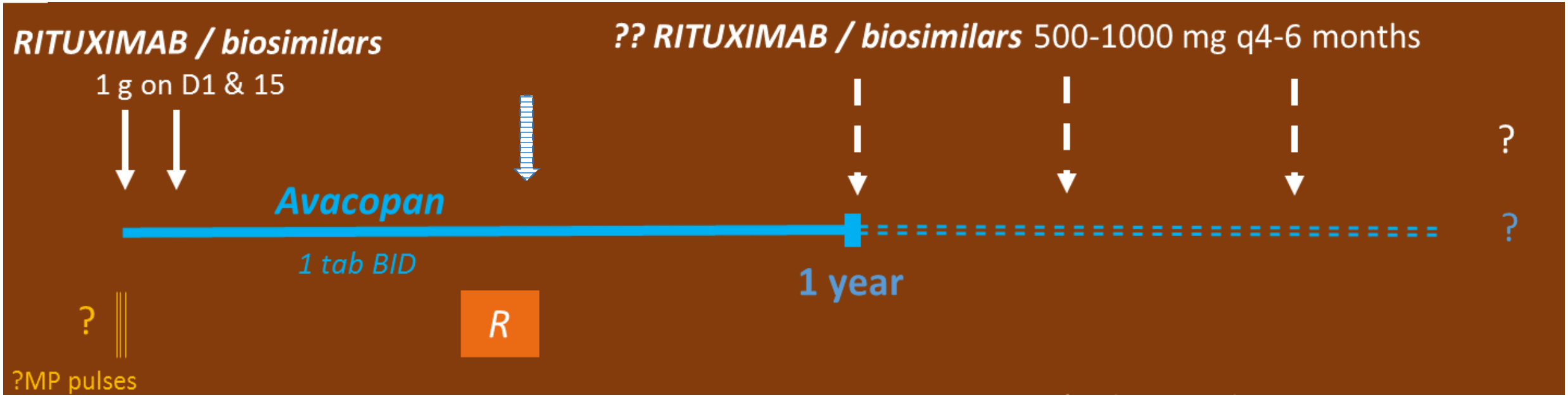
Death from any cause or ESRD

27% at 3 years

.. Où en est-on aujourd’hui?

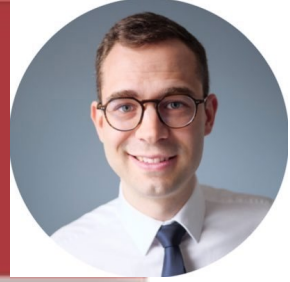
Principles of treatment of severe, systemic G/MPA





Pagnoux, Fifi-Mah. Curr Treatm Opt Rheumatol 2021(7),112-33

A predictive mortality score in ANCA-associated renal vasculitis



Developing a novel mortality prognostic score: Death in ANCA Glomerulonephritis – Estimating the Risk (DANGER)

Methods

Caucasian patients with ANCA-vasculitis and glomerulonephritis

Maine-Anjou Registry
Development cohort
n = 194

RENVAS Registry
Validation cohort
n = 185

Outcome: death

Predictive performance compared with existing scores

Results

DANGER score components

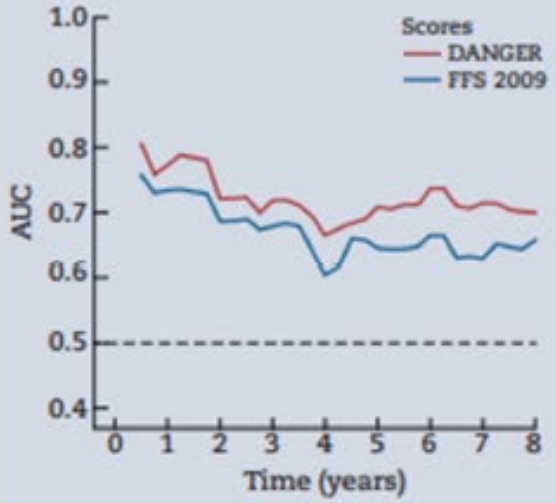
Cox proportional HR (95% CI) for death

Hypertension	Heart disease
1.86 (0.97–3.53)	1.91 (1.01–3.61)

Age (per +5 years)	Creatinine (per +50 micromol/L)	Hemoglobin (per +1 g/dL)
1.50 (1.29–1.75)	1.07 (1.03–1.10)	0.87 (0.76–0.99)

Score performance

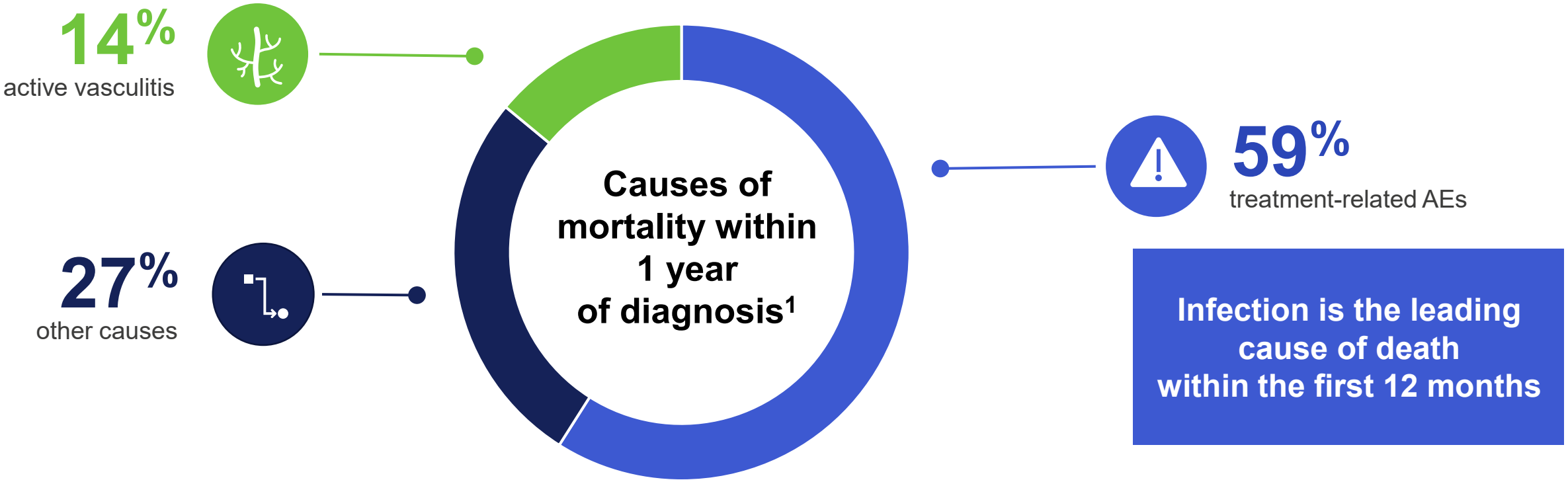
in validation cohort



Fage, N. *et al.*
NDT (2024)
@NDTSocial
@MaineAnjouReg

We developed and validated the DANGER score, with higher prognostic value than previously known scores to predict death in ANCA-associated vasculitis with glomerulonephritis.

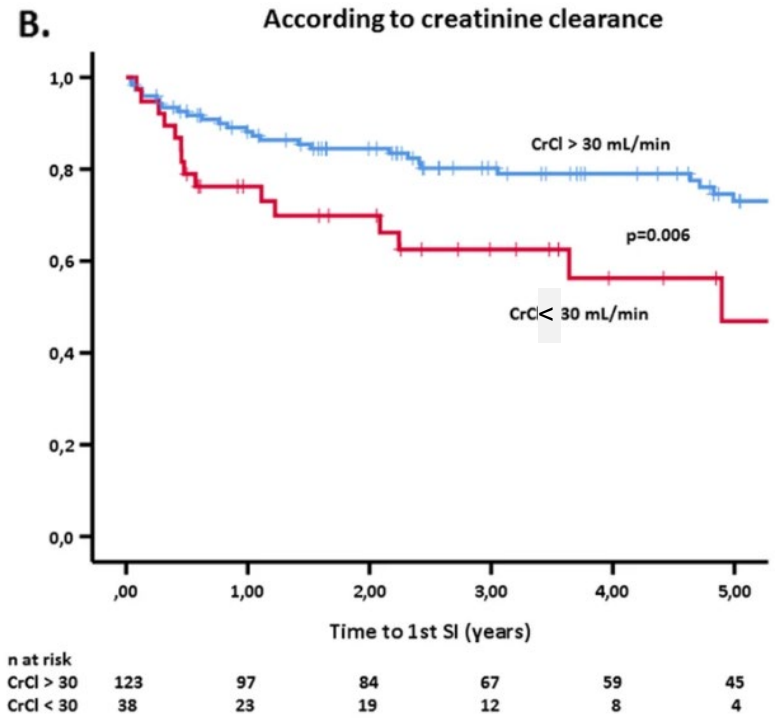
Over half of first-year mortality in ANCA-associated vasculitis is caused by treatment-related adverse events

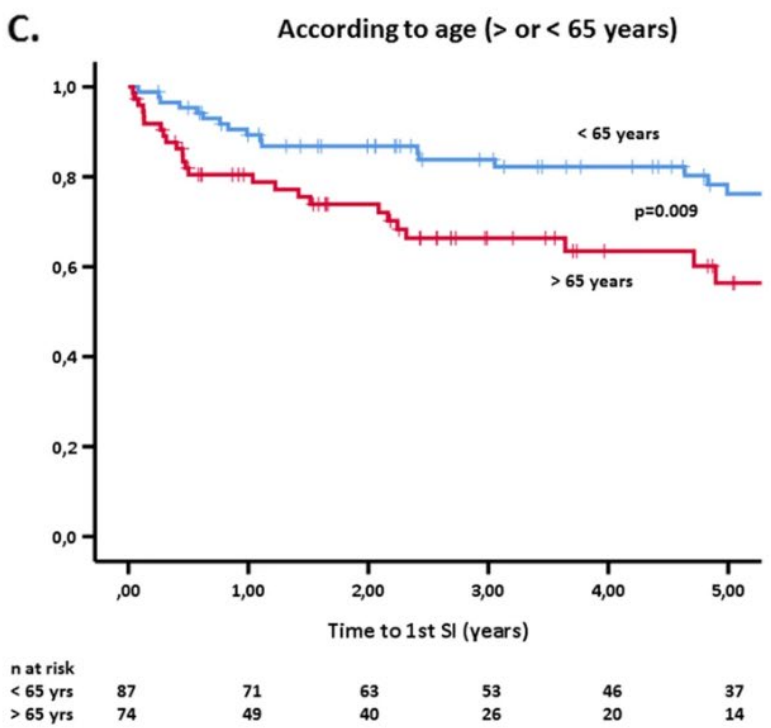


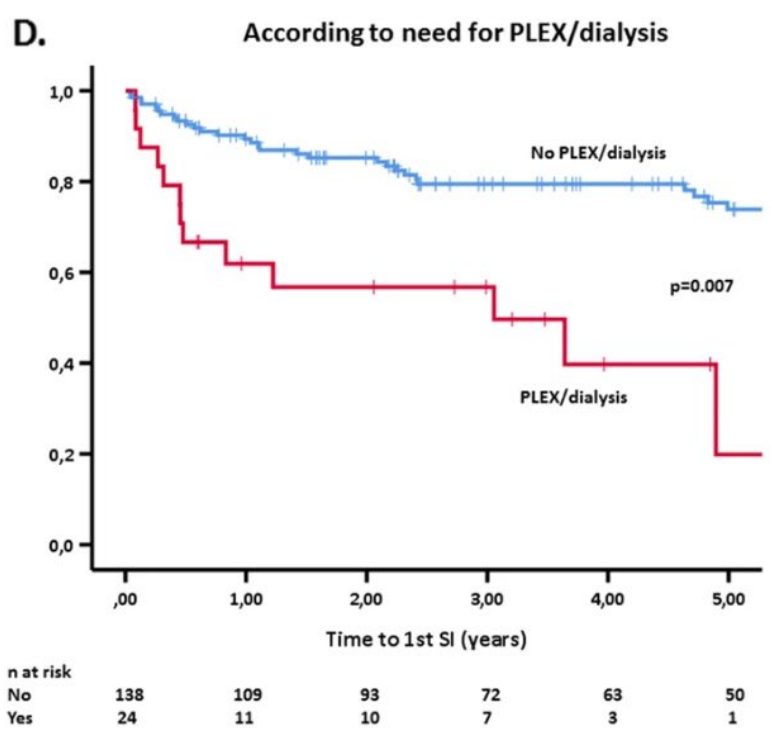
AE, adverse event; ANCA, anti-neutrophil cytoplasmic antibody; GC, glucocorticoid

1. Little MA, Nightingale P, Verburgh CA, et al. Ann Rheum Dis. 2010 Jun;69(6):1036-43.

Serious infections in 162 patients with GPA (63%) or MPA (37%)

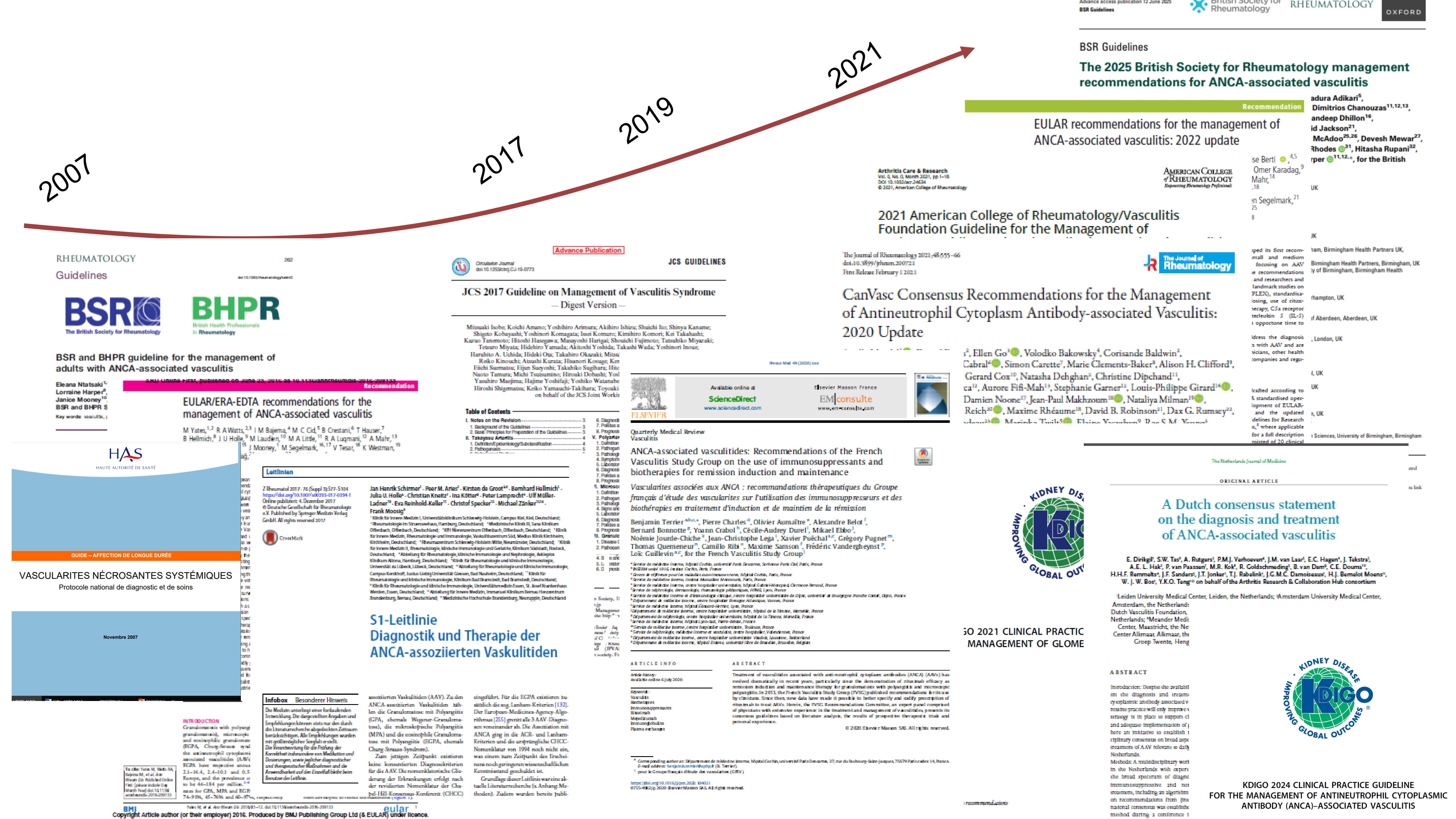






Thomas K et al. Arthritis Res Ther. 2021 Mar 20;23(1):90

RECOMMENDATIONS & GUIDELINES FOR THE MANAGEMENT OF ANCA VASCULITIS



2007
2017
2019
2021
BSR Guidelines
The 2025 British Society for Rheumatology management recommendations for ANCA-associated vasculitis
Recommendation
EULAR recommendations for the management of ANCA-associated vasculitis: 2022 update
2021 American College of Rheumatology/Vasculitis Foundation Guideline for the Management of
CanVasc Consensus Recommendations for the Management of Antineutrophil Cytoplasm Antibody-associated Vasculitis: 2020 Update
RHEUMATOLOGY
Guidelines
BSR and BHPR guideline for the management of adults with ANCA-associated vasculitis
JCS GUIDELINES
JCS 2017 Guideline on Management of Vasculitis Syndrome
— Digest Version —
EULAR/ERA-EDTA recommendations for the management of ANCA-associated vasculitis
HAS
HAUTE AUTORITÉ DE SANTÉ
GUIDE – AFFECTION DE LONGUE DURÉE
VASCULARITES NÉCROSANTES SYSTÉMIQUES
Protocole national de diagnostic et de soins
Novembre 2007
Leitlinien
S1-Leitlinie
Diagnostik und Therapie der ANCA-assoziierten Vaskulitiden
ANCA-associated vasculitides: Recommendations of the French Vasculitis Study Group on the use of immunosuppressants and biotherapies for remission induction and maintenance
Vascularites associées aux ANCA : recommandations thérapeutiques du Groupe français d'étude des vascularites sur l'utilisation des immunosuppresseurs et des biothérapies en traitement d'induction et de la rémission
KIDNEY DISEASE IMPROVING GLOBAL OUTCOMES
KDIGO
2021 CLINICAL PRACTICE GUIDELINE
MANAGEMENT OF GLOME
The Netherlands Journal of Medicine
ORIGINAL ARTICLE
A Dutch consensus statement on the diagnosis and treatment of ANCA-associated vasculitis
KDIGO 2024 CLINICAL PRACTICE GUIDELINE FOR THE MANAGEMENT OF ANTINEUTROPHIL CYTOPLASMIC ANTIBODY (ANCA)–ASSOCIATED VASCULITIS

2007

2017

2019

2021

Rheumatology, 2025, 64, 4470–4494
https://doi.org/10.1093/rheumatology/keaf240
Advance access publication 12 June 2025
BSR Guidelines

British Society for Rheumatology RHEUMATOLOGY OXFORD

BSR Guidelines

The 2025 British Society for Rheumatology management recommendations for ANCA-associated vasculitis

Morkeh[3,4,†], Madura Adikari[5], ul Brogan[9,10], Dimitrios Chanouzas[11,12,13], l Pero[15], Emmandeep Dhillon[16], ick[19,20], David Jackson[21], on[24], Stephen McAdoo[25,26], Devesh Mewar[27], [30], Benjamin Rhodes[31], Hitasha Rupani[32], [5], Lorraine Harper[11,12,*], for the British p[8]

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st, Birmingham, UK
Southampton, UK
Free Hospital, London, UK

Recommendation

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

Bernhard Hellmich [1], Beatriz Sanchez-Alamo,[2] Jan H Schirmer,[3] Alvise Berti [4,5]
Ilmer Karadag,[9]
ahr,[14]
Segelmark,[21]

Arthritis Care & Research
Vol. 0, No. 0, Month 2021, pp 1–16
DOI 10.1002/acr.24634
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AMERICAN COLLEGE of RHEUMATOLOGY
Supporting Rheumatology Professionals

2021 American College of Rheumatology/Vasculitis Foundation Guideline for the Management of Antineutrophil Cytoplasmic Antibody–Associated Vasculitis

y M. Archer,[7]
L. Merkel,[12]
I. Vitobaldi,[17]
. Kalot,[22]
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focusing on AAV
recommendations
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The Journal of Rheumatology 2021;48:555–66
doi:10.3899/jrheum.200721
First Release February 1 2021

The Journal of Rheumatology

CanVasc Consensus Recommendations for the Management of Antineutrophil Cytoplasm Antibody-associated Vasculitis: 2020 Update

Arielle Mendel[1], Daniel Ennis[2], Ellen Go[3], Volodiko Bakowsky[4], Corisande Baldwin[5], Susanne M. Benseler[6], David A. Cabral[6], Simon Carette[7], Marie Clements-Baker[8], Alison H. Clifford[9], Jan Willem Cohen Tervaert[9], Gerard Cox[10], Natasha Dehghan[2], Christine Dipchand[11], Navjot Dhindsa[2], Leilani Famorca[12], Aurore Fifi-Mah[13], Stephanie Garner[13], Louis-Phil Clode Lessard[15], Patrick Liang[16], Damien Noone[17], Jean-Paul Makhzoum[18], Nataliya Mi Christian A. Pineau[1], Heather N. Reich[20], Maxime Rhéaume[18], David B. Robinson[21], I Tanveer E. Towheed[8], Judith Trudeau[23], Marinka Twilt[5], Elaine Yacyshyn[9], Rae S. Lillian B. Barra[24], Nader Khalidi[12], and Christian Pagnoux[7]

ABSTRACT. ***Objective.*** In 2015, the Canadian Vasculitis Research Network (CanVasc) created recommendations for management of antineutrophil cytoplasm antibody (ANCA)-associated vasculitides (AAV) in Canada. current update aims to revise existing recommendations and create additional recommendations, as nece based on a review of new available evidence.

Methods. A needs assessment survey of CanVasc members informed questions for an updated systematic erature review (publications spanning May 2014 to September 2019) using Medline, Embase, and Cochr New and revised recommendations were developed and categorized according to the level of evidence strength of each recommendation. The CanVasc working group used a 2-step modified Delphi procedur reach > 80% consensus on the inclusion, wording, and grading of each new and revised recommendatio

Results. Eleven new and 16 revised recommendations were created and 12 original (2015) recommendati were retained. New and revised recommendations are discussed in detail within this document. Five orig recommendations were removed, of which 4 were incorporated into the explanatory text. The supplemen material for practical use was revised to reflect the updated recommendations.

Conclusion. The 2020 updated recommendations provide rheumatologists, nephrologists, and other spe ists caring for patients with AAV in Canada with new management guidance, based on current evidence consensus from Canadian experts.

Key Indexing Terms: antineutrophil cytoplasm antibody-associated vasculitis, eosinophilic granulomat with polyangiitis, glomerulonephritis, granulomatosis with polyangiitis, microscopic polyangiitis

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[10]G. Cox, MB, Firestone Institute for Respiratory H University, Hamilton, Ontario; [11]C. Dipchand, M Nephrology, QEII Health Sciences Centre, Dalhou Nova Scotia; [12]L. Famorca, MD, S. Garner, MD, Division of Rheumatology, McMaster University, [13]A. Fifi-Mah, MD, Division of Rheumatology, Un Calgary, Alberta; [14]L.P. Gérard, MD, MSc, Division of Nephrology, University of Calgary, Calgary, Alberta; [15]C. Lessard, MD, Centre de Recherche Musculo-Squelettique, Trois-Rivières, Québec; [16]P. Liang, MD, Division of Rheumatology, Centre Hospitalier Universitaire de Sherbrooke, Université de Sherbrooke, Sherbrooke, Québec; [17]D. Noone, MB, BCh, BAO, MSc, Division of Nephrology, Hospital for Sick Children, University of Toronto, Toronto, Ontario; [18]J.P. Makhzoum, MD, M. Rhéaume, MD, Division of Internal Medicine, Hôpital du Sacré-Cœur de Montréal, Université de Montréal, Montréal, Québec; [19]N. Melman, MD, MSc, Division of Rheumatology, The Ottawa Hospital, University of Ottawa and Ottawa

KIDNEY DISEASE IMPROVING GLOBAL OUTCOMES — KDIGO

KDIGO 2024 CLINICAL PRACTICE GUIDELINE
FOR THE MANAGEMENT OF ANTINEUTROPHIL CYTOPLASMIC
ANTIBODY (ANCA)–ASSOCIATED VASCULITIS

- Recommendation: For patients with active, severe GPA/MPA, we conditionally recommend treatment with **rituximab over cyclophosphamide for remission induction**.

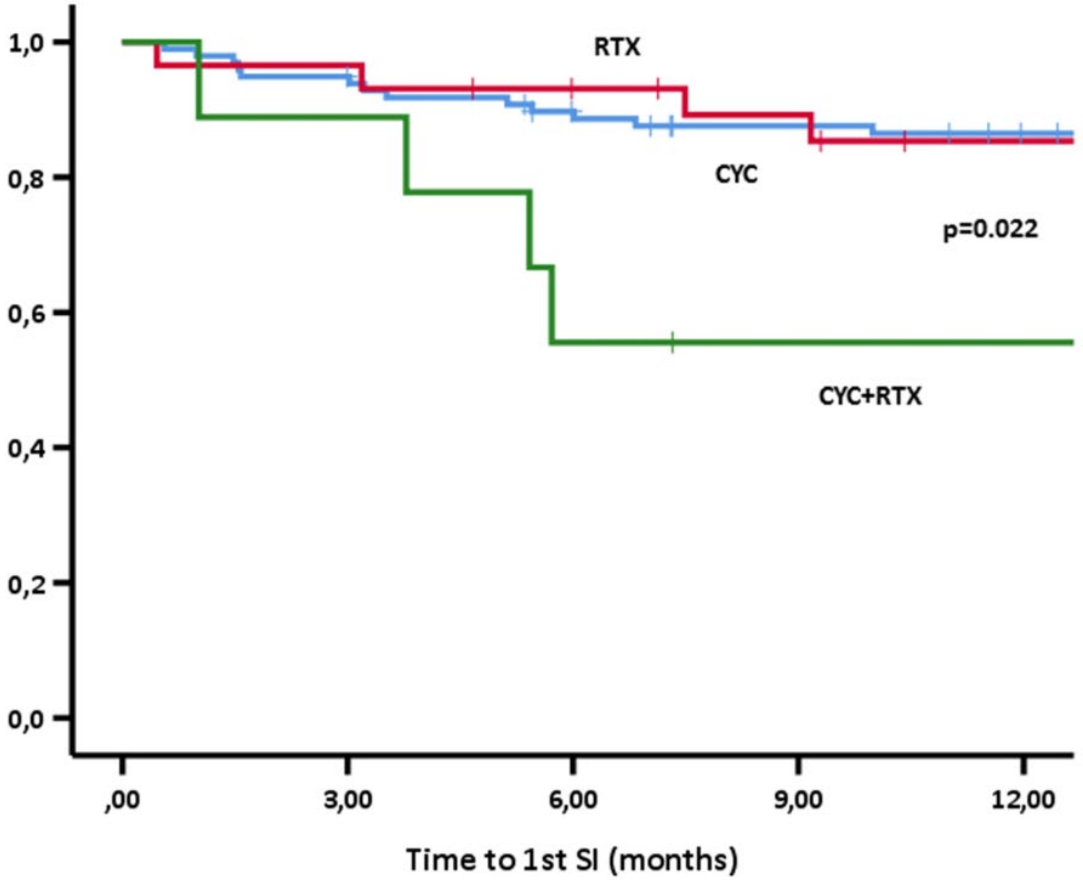
- In patients with severe, newly diagnosed GPA or MPA, we recommend GC plus either CYC or RTX for first-line remission induction therapy. **RTX is preferred for remission induction** in patients with severe GPA or MPA **in whom CYC is contraindicated**, including those with a risk of infertility. Category 1B, Strength A.

- For induction of remission in patients with new-onset or relapsing GPA or MPA with organ-threatening or life-threatening disease, we recommend treatment with a combination of glucocorticoids and **either rituximab or cyclophosphamide**.* Rituximab is **preferred in relapsing disease**.

- Recommendation 2b The recommended options for immunosuppression for remission induction of newly diagnosed GPA or MPA are intravenous pulsed cyclophosphamide (CYC) or rituximab (RTX) (GRADE 1A, SoA 98%).
- Recommendation 2c For active relapsing disease, treatment with RTX is preferred (GRADE 1B, SoA 97%).
- Recommendation 2d A combination of both CYC and RTX can be considered for organ-threatening or lifethreatening disease (GRADE 2C, SoA 98%).

Chung et al. Arthritis Rheumatol. 2021 Aug;73(8):1366-1383
Mendel et al. J Rheumatol. 2021 Apr;48(4):555-566
Hellmich et al. ARD. 2023
Biddle BSR 2025

Serious infections



n at risk

CYC	98	92	83	78	74
RTX	29	27	24	22	19
CYC+RTX	9	7	5	4	3

Thomas K et al. Arthritis Res Ther. 2021 Mar 20;23(1):90

KDIGO

Practice Point 9.3.1.2: In patients presenting with markedly reduced or rapidly declining glomerular filtration rate (GFR) ([SCr] >354 mmol/l]), there are limited data to support rituximab and glucocorticoids.

Both cyclophosphamide and glucocorticoids, and the combination of rituximab and cyclophosphamide can be considered in this setting.

Limited, non-severe GPA

Recommendation

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

For induction of remission of non-organ-threatening or non-life-threatening GPA or MPA, treatment with a combination of glucocorticoids and rituximab is recommended. Methotrexate or mycophenolate mofetil can be considered as alternatives to rituximab. (1b, B)

Recommendation 2e. Certain individuals with active GPA or MPA, with no evidence of life or organ-threatening disease, may be considered for alternative induction therapy with methotrexate (MTX) or mycophenolate mofetil (MMF) (GRADE 1A, SoA 96%).

British Society for Rheumatology

- A GC tapering protocol should be initiated within 2 weeks of induction therapy in patients with severe GPA or MPA. A **reduced-dose GC tapering protocol can be considered in adult patients** with severe GPA or MPA who are receiving CYC or RTX induction therapy, to reduce cumulative GC exposure and infection risk.
- Recommendation: For patients with active, severe GPA/MPA, we conditionally recommend a **reduced-dose glucocorticoid regimen** over a standard-dose glucocorticoid regimen for remission induction.
- As part of regimens for induction of remission in GPA or MPA, we recommend treatment with oral glucocorticoids at a starting dose of 50–75 mg prednisolone equivalent/day, depending on body weight. We recommend **stepwise reduction in glucocorticoids according to table 4** and achieving a dose of 5 mg prednisolone equivalent per day by 4–5 months.

Table 4 Glucocorticoid dosing (mg/day, prednisolone equivalent) with rituximab or cyclophosphamide-based regimens for remission induction in GPA or MPA according to the PEXIVAS Study[93]

	Body weight (kg)		
Weeks	<50	50–75	>75
1*	50	60	75
2	25	30	40
3–4	20	25	30
5–6	15	20	25
7–8	12.5	15	20
9–10	10	12.5	15
11–12	7.5	10	12.5
13–14	6	7.5	10
15–18	5	5	7.5
19–52	5	5	5
>52	Individual taper	Individual taper	Individual taper

*Consider use of intravenous methylprednisolone at a cumulative dose of 1–3 g on days 1–3 in patients with severely active disease, including but not limited to renal involvement with a documented estimated glomerular filtration rate <50 mL/min/1.73 m^2 and/or diffuse alveolar haemorrhage.

KDIGO: Practice Point 9.3.1.6: Recommendations for oral glucocorticoid tapering are given in Figure [9].

Chung et al. Arthritis Rheumatol. 2021 Aug;73(8):1366-1383
Mendel et al. J Rheumatol. 2021 Apr;48(4):555-566
Hellmich et al. ARD. 2023.

- Recommendation 4d. Despite common place use, there is a lack of supporting trial evidence for intravenous methylprednisolone (IV MP) pulses. Therefore, IV MP pulses are not routinely recommended but can be reserved as an option for the management of organ-threatening manifestations, including active renal disease and diffuse alveolar haemorrhage (GRADE 2C, SoA 97%).

- 5b. Pulse IV MP (0.5–1 g/day for 1–3 days) can be considered in severe, organ- or life-threatening GPA or MPA, but lacks proven efficacy and carries a potential risk of adverse effects. (Category 3, Strength D)

Mendel et al. J Rheumatol. 2021 Apr;48(4):555-566

Biddle BSR 2025

Glucocorticoid Dosing and Outcomes in ANCA Associated Vasculitis With Kidney Involvement

ISN INTERNATIONAL SOCIETY OF NEPHROLOGY

Cohort

Retrospective, single center study

Biopsy proven renal AAV (n=138) (2010-2023)

Standard Glucocorticoid therapy (Pre 2020 era)

Reduced Glucocorticoid therapy (Post 2020 era)

Findings

Primary outcomes

- ESKD/death within a year post biopsy.

Secondary outcomes

- Serious infections
- Remission + patient & kidney survival
- Change in eGFR

Mean age 65.2 years

Females 46.4%

Anti MPO 68.1%

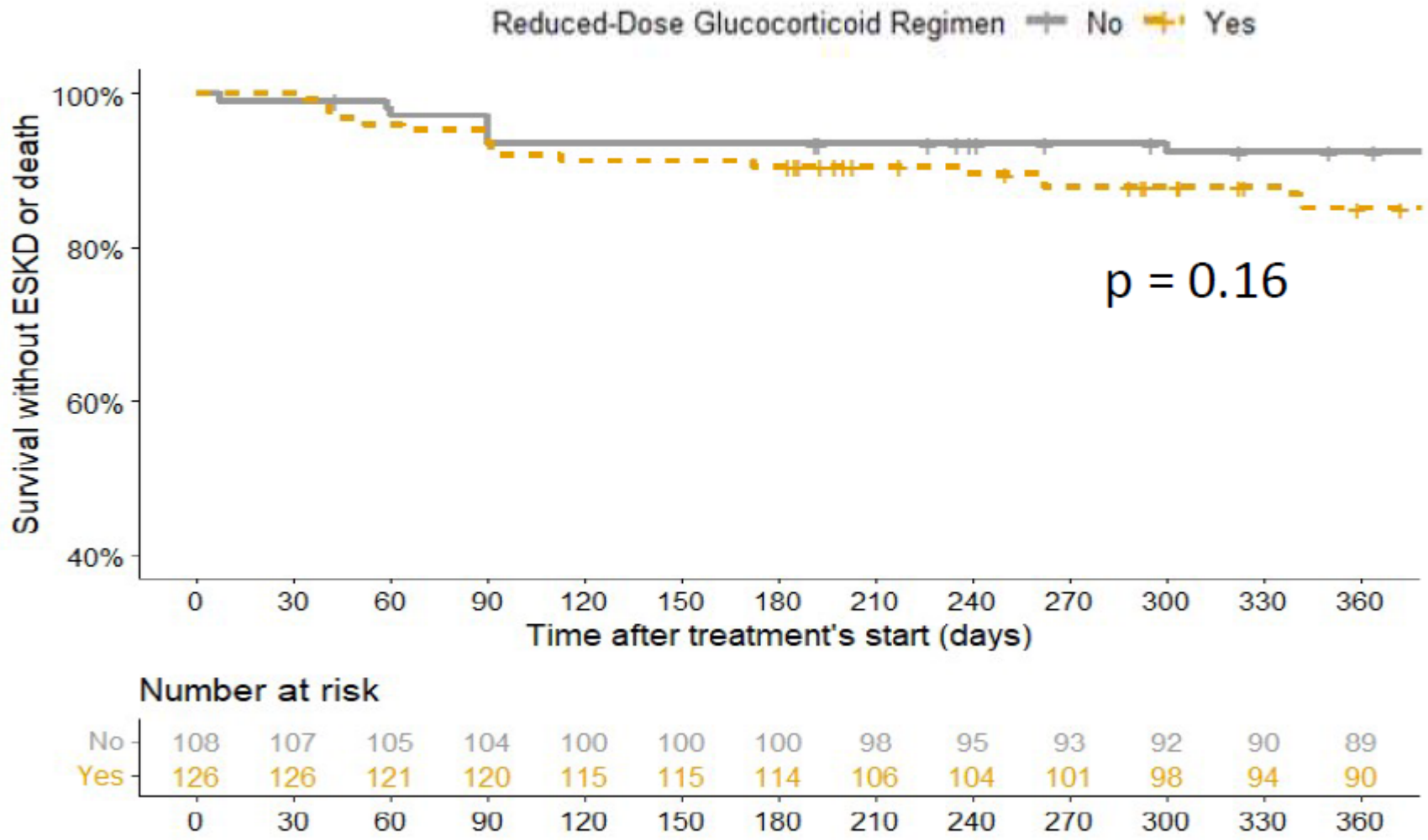
	Reduced Glucocorticoid (n=41)	Standard Glucocorticoid (n=97)
Age	70.3 yrs	63.1yrs
ESKD/death	24.4%	21.6%
Serious Infections	14.6%	17.5%
Remission	63.4%	63.9%

KI REPORTS
Kidney International Reports

Maria S et al, 2025

Visual abstract by:
Priya John MD, DM
@drpriyajohn

Conclusion: Reduced-GC during induction therapy in individuals with organ-threatening kidney involvement was not associated with a change in outcomes or kidney function recovery. This supports data from a large randomized trial.

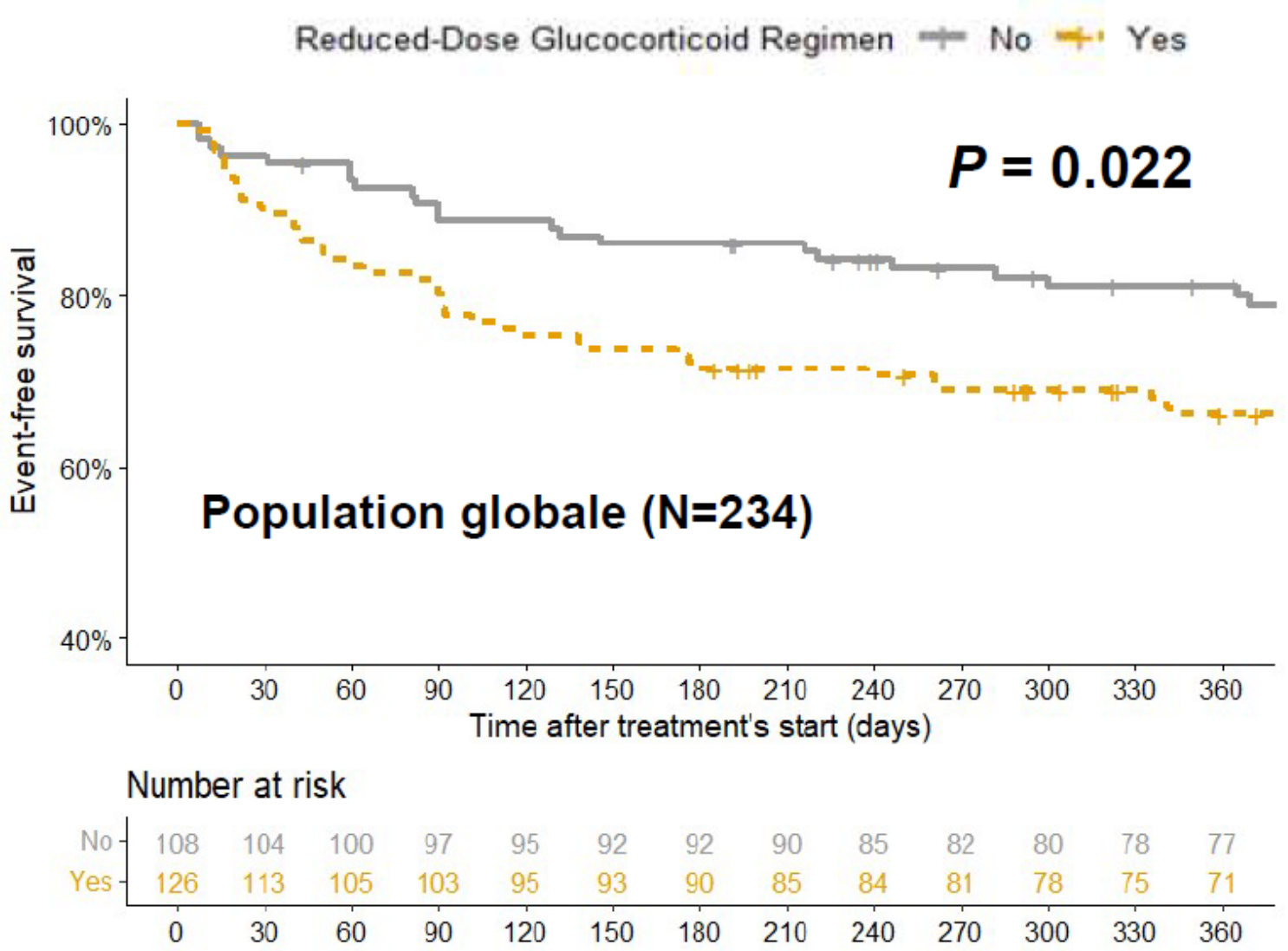


S. Nagle, Ann Rheum Dis 2025 Feb;84(2):319-328

EVENT

- Minor relapse
- Major relapse
- Progression (before remission)
- ESRD
- Death

Concerns especially if
RTX induction
and/or creatinine >300 µmol/L



S. Nagle, Ann Rheum Dis 2025 Feb;84(2):319-328

Principles of treatment of severe GPA/MPA

CYCLOPHOSPHAMIDE ?

± MP pulses?

± PLEX?

RITUXIMAB
1 g at D1 & 15 (or 375 mg/m²/wk)

3 - 6 months

INDUCTION

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*

ADVOCATE: Phase 3 study comparing an avacopan-based regimen to a GC-based regimen[1,2]

Screening & randomization
- Age ≥12 years
- Newly diagnosed or relapsing GPA/MPA
- PR3 or MPO-ANCA+
- Active disease
- eGFR ≥15

Avacopan-based regimen (n=164)
- Avacopan, 30 mg twice daily + add-on immuno-suppressants
- Placebo prednisone taper over 21 weeks

GC-based regimen (n=166)
- Placebo, twice daily + tapered prednisone + add-on immuno-suppressants
- Prednisone, 60 mg/day with standard tapering to zero over 21 weeks

Add-on immuno-suppressants consisted of:*
- RTX (4 weeks)
- - OR -
- CYC (13–14 weeks) → AZA

PRIMARY ENDPOINTS[1,2]
- **Remission** (BVAS 0 & no GC use in prior 4 weeks) at Week 26
- **Sustained remission** (BVAS 0 & no GC use in prior 4 weeks, no relapse Weeks 26–52)

KEY SECONDARY ENDPOINTS[1,2]
- **Change in GC-related toxicity†** at Week 52
- **Change in HRQoL** over 52 weeks
- Renal disease: **change in eGFR and UACR** over 52 weeks

* Non-study GCs supplied for: i) ANCA-AV worsening; ii) non-ANCA-AV related reasons. †According to the Glucocorticoid Toxicity Index.
ANCA, anti-neutrophil cytoplasmic antibody; AZA, azathioprine; BVAS, Birmingham Vasculitis Activity Score; CYC, cyclophosphamide; eGFR, estimated glomerular filtration rate; GC, glucocorticoid; GPA, granulomatosis with polyangiitis; HRQoL, health-related quality of life; MPA, microscopic polyangiitis; RTX, rituximab.

1. Merkel PA, Jayne DR, Wang C, et al. JMIR Res Protoc. 2020 Apr 7;9(4):e16664.
2. Jayne DRW, Merkel PA, Schall TJ, et al. N Engl J Med. 2021 Feb 18;384(7):599-609.
3. Jayne DRW, Merkel PA, Schall TJ, et al. N Engl J Med. 2021 Feb 18;384(7):599-609. Supplement.

Advocate
Baseline Characteristics

~~ICU-DAH, eGFR<15ml/min~~

55% GPA : **45%** MPA[2]

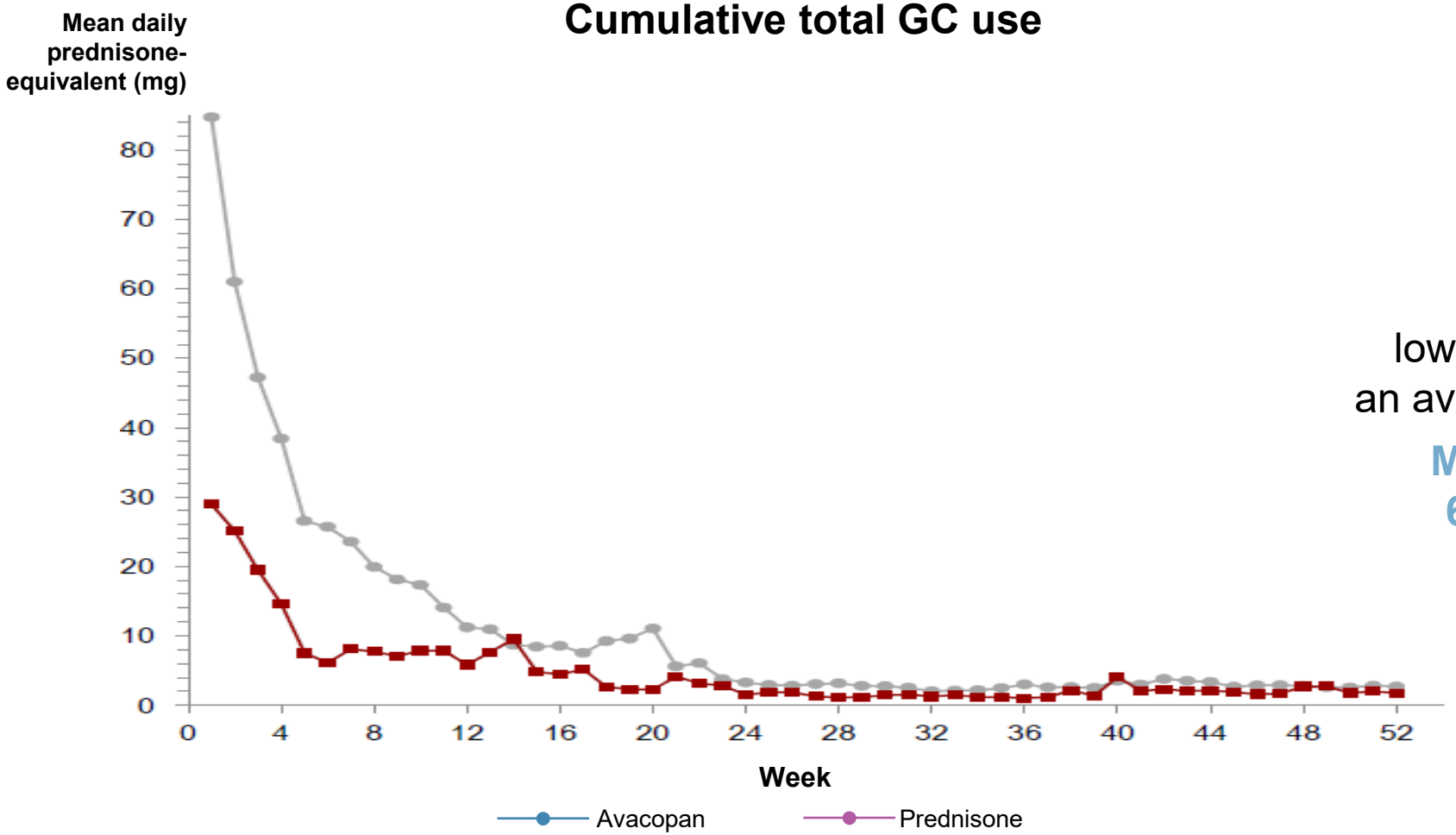
43% PR3 positive : **57%** MPO positive[2]

81% of patients had renal vasculitis[2]

Conducted in **198 study locations across 19 countries**[3]

Characteristic (n=330)	Avacopan (n=166)	Prednisone (n=164)
Type of vasculitis – no. (%)		
GPA	91 (54.8)	90 (54.9)
MPA	75 (45.2)	74 (45.1)
Immunosuppressant induction treatment – no. (%)		
Intravenous rituximab	107 (64.5)	107 (65.2)
Intravenous cyclophosphamide	51 (30.7)	51 (31.1)
Oral cyclophosphamide	8 (4.8)	6 (3.7)
Renal involvement – no. (%)	134 (80.7)	134 (81.7)
eGFR	51 (31)	53 (33)

An avacopan-based induction regimen lowered mean cumulative GC use vs. a GC-based regimen



56%

lower overall GC dose with an avacopan-based regimen[1–3]

Median total GC dose:
600 mg vs 3,098 mg

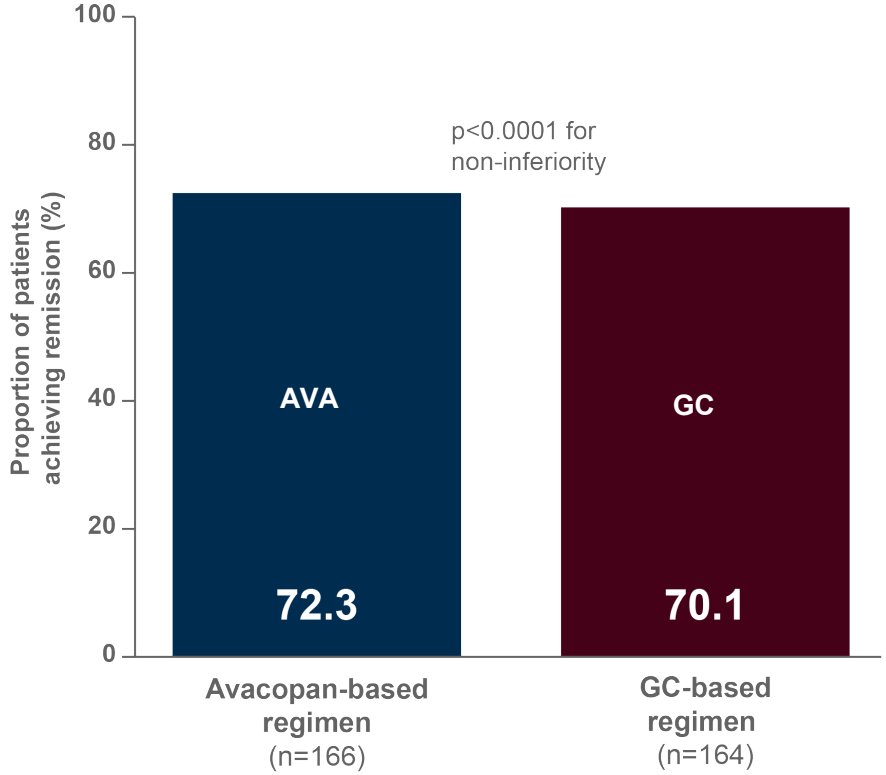
GC, glucocorticoid.

1. Jayne DRW, Merkel PA, Schall TJ, et al. N Engl J Med. 2021 Feb 18;384(7):599-609.
2. Jayne DRW, Merkel PA, Schall TJ, et al. N Engl J Med. 2021 Feb 18;384(7):599-609. Supplement.
3. Erratum for: N Engl J Med. 2021 Feb 18;384(7):599-609. N Engl J Med. 2024 Jan 25;390(4):388.

Avacopan-based Regimen was Superior to GC-based Regimen in Sustaining Remission At 52 Weeks

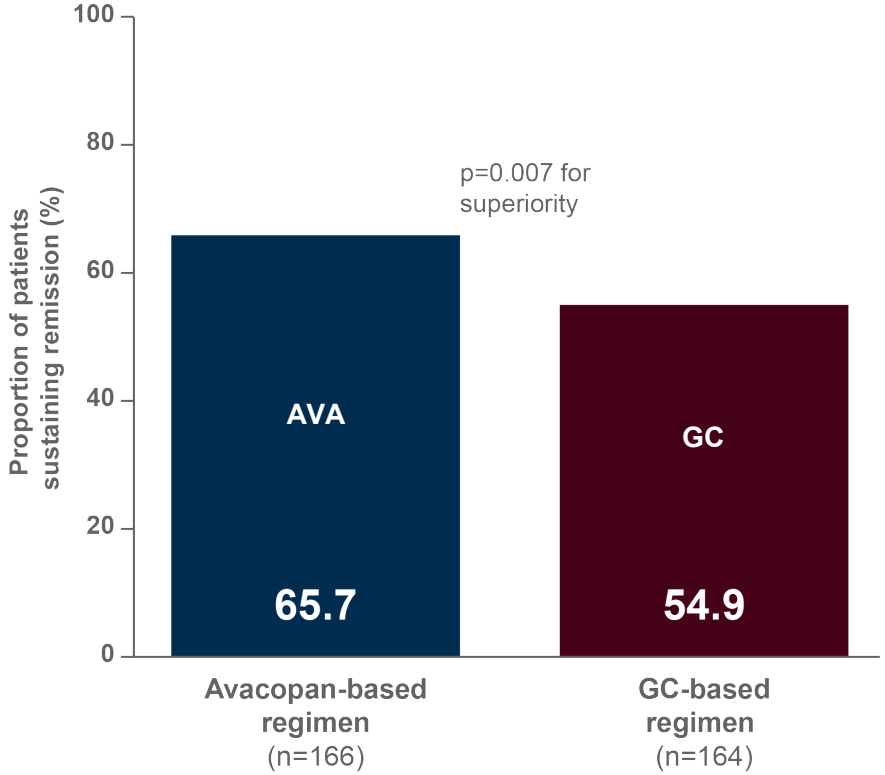
NON-INFERIOR DISEASE REMISSION (WEEK 26)

Avacopan-based regimen demonstrated numerically higher remission at 26 weeks vs GC-based regimen



SUPERIOR SUSTAINED REMISSION (WEEK 52)

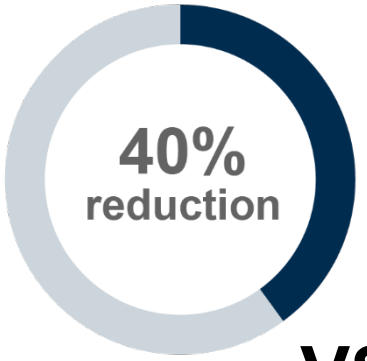
Avacopan-based regimen demonstrated superior sustained remission at 52 weeks vs GC-based regimen



Jayne D, et al. *N Engl J Med* 2021;384(7):599–609.

Avacopan-based Regimen Resulted in Statistically Significant Improvements in Renal Function Vs. GC-based Regimen

DECREASED UACR AT WEEK 4



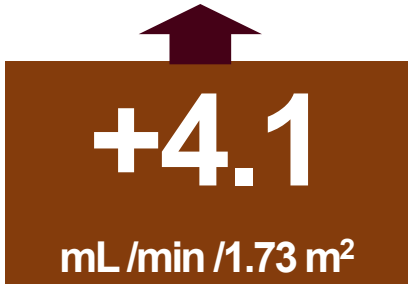
Avacopan-based regimen (n=166)

VS

GC-based regimen (n=164)

Baseline to Week 4 (P<0.0001)

STATISTICALLY eGFR IMPROVEMENT AT WEEK 52



Avacopan-based regimen (n=119)

GC-based regimen (n=125)

Mean eGFR improvement at Week 52 (p=0.029)
Average eGFR in both groups at baseline: 45.1 mL/min/1.73 m^2

SIGNIFICANTLY IMPROVED eGFR IN STAGE 4 CKD PATIENTS AT WEEK 52

Avacopan-based regimen

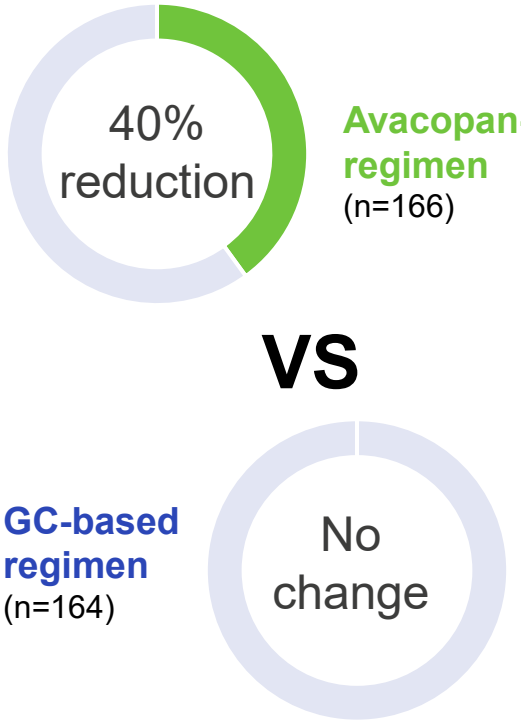
GC-based regimen

Mean eGFR improvement for patients with stage 4 kidney disease (<30 mL/min/1.73 m^2) at Week 52 (p=0.01)

Jayne D, et al. *N Engl J Med* 2021;384(7):599–609.

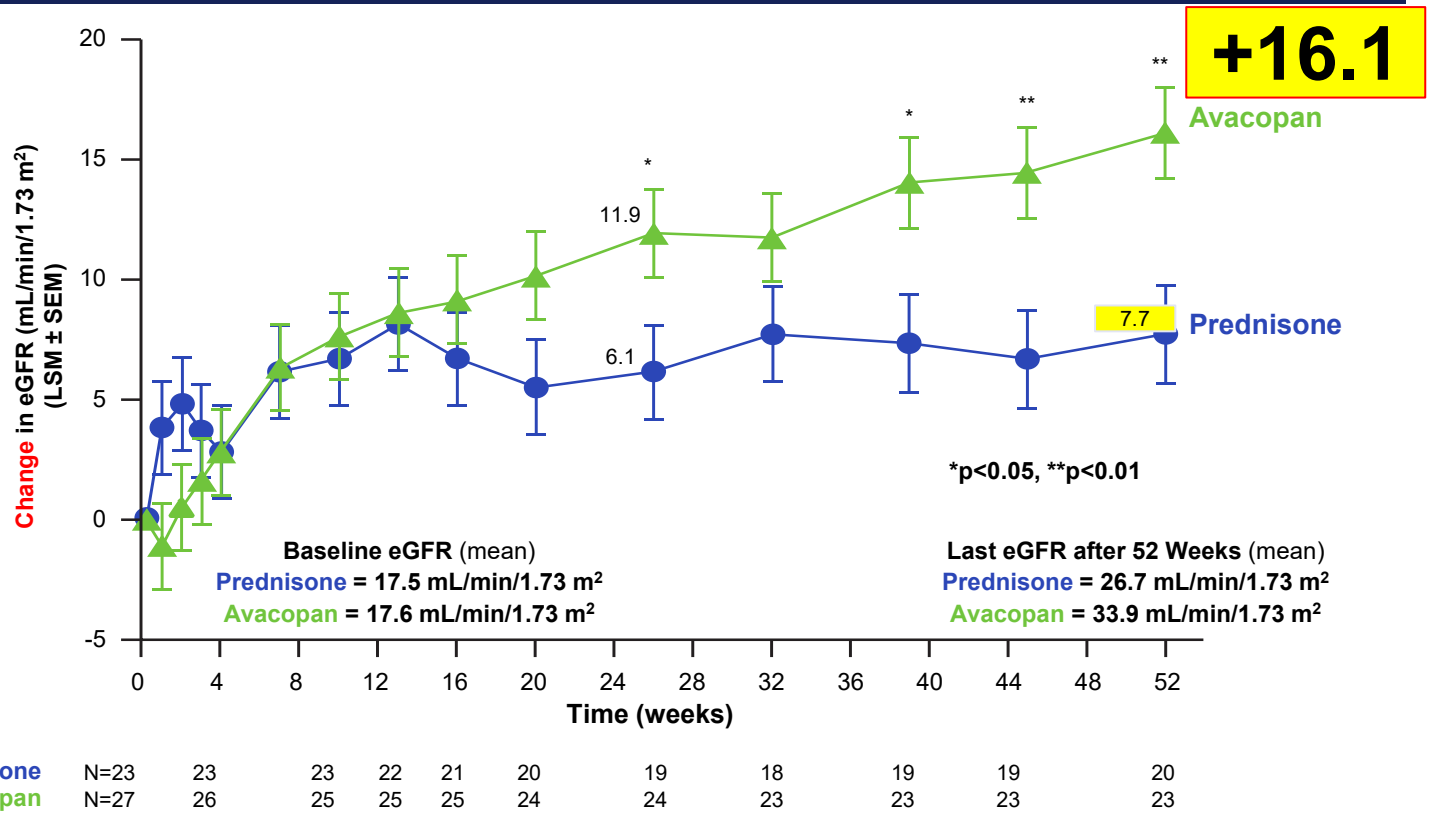
An avacopan-based regimen resulted in statistically significant improvements in renal function vs. a GC-based regimen

DECREASED UACR AT WEEK 4[1]



Baseline to Week 4 (P<0.0001)

STATISTICALLY SIGNIFICANT eGFR IMPROVEMENT AT WEEK 52[2]
(Renal recovery for patients with baseline eGFR ≤20)



eGFR, estimated glomerular filtration rate; LSM, least mean squares; GC, glucocorticoid; SEM, standard error of mean.

1. Jayne DRW, Merkel PA, Schall TJ, et al. N Engl J Med. 2021 Feb 18;384(7):599-609.
2. Adapted from Cortazar FB, Jayne DRW, Bruchfeld A, et al. Renal recovery for patients with baseline eGFR ≤20 in the avacopan ADVOCATE trial. Poster FR-PO651 presented at American Society of Nephrology Kidney Week, November 2-6, 2022.

JOURNAL ARTICLE ACCEPTED MANUSCRIPT

CanVasc consensus recommendations for the use of avacopan in antineutrophil cytoplasm antibody-associated vasculitis: 2022 addendum FREE

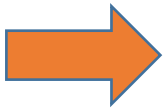
David Turgeon ✉, Volodko Bakowsky, Corisande Baldwin, David A Cabral, Marie Clements-Baker, Alison Clifford, Jan Willem Cohen Tervaert, Natasha Dehghan, Daniel Ennis, Leilani Famorca, Aurore Fifi-Mah, Louis-Philippe Girard, Frédéric Lefebvre, Patrick Liang, Jean-Paul Makhzoum, David Massicotte-Azarniouch, Arielle Mendel, Nataliya Milman, Heather N Reich, David B Robinson, Carolyn Ross, Dax G Rumsey, Medha Soowamber, Tanveer E Towheed, Judith Trudeau, Marinka Twilt, Elaine Yacyshyn, Gozde K Yardimci, Nader Khalidi, Lillian Barra, Christian Pagnoux

Rheumatology, kead087, https://doi.org/10.1093/rheumatology/kead087

Published: 21 February 2023 **Article history** ▾

1. The addition of oral avacopan (30 mg twice daily) can be considered for induction of remission in patients with newly diagnosed or relapsing GPA or MPA treated with cyclophosphamide or rituximab (Category 1B, Strength B, LoA 9.54 ± 0.69).

The cost-effectiveness of avacopan as first-line therapy for all patients is unknown and may be a limiting factor for its widespread use in practice. Patients at increased risk of GC toxicity (e.g., pre-existing diabetes, metabolic syndrome, cardiovascular disease, osteoporosis, glaucoma, cataracts, neuropsychiatric disorders, susceptibility to recurrent or severe infections) would especially benefit from strategies to limit the use of GC such as avacopan. Patients with renal involvement and those refractory to conventional treatments may also potentially benefit from avacopan.



2. After starting avacopan, a faster glucocorticoid tapering protocol aiming for discontinuation by the end of week 4 should be considered (Category 1B, Strength B, LoA 9.25 ± 0.93).

3. When initiated as part of induction therapy, avacopan can be continued for one year (Category 1B, Strength B, LoA 9.43 ± 0.79).

KDIGO 2024 CLINICAL PRACTICE GUIDELINE
FOR THE MANAGEMENT OF ANTINEUTROPHIL CYTOPLASMIC
ANTIBODY (ANCA)–ASSOCIATED VASCULITIS

Practice Point 9.3.1.7

Avacopan may be used as an alternative to glucocorticoids.

Patients with an increased risk of glucocorticoids toxicity are likely to receive the most benefit from avacopan.

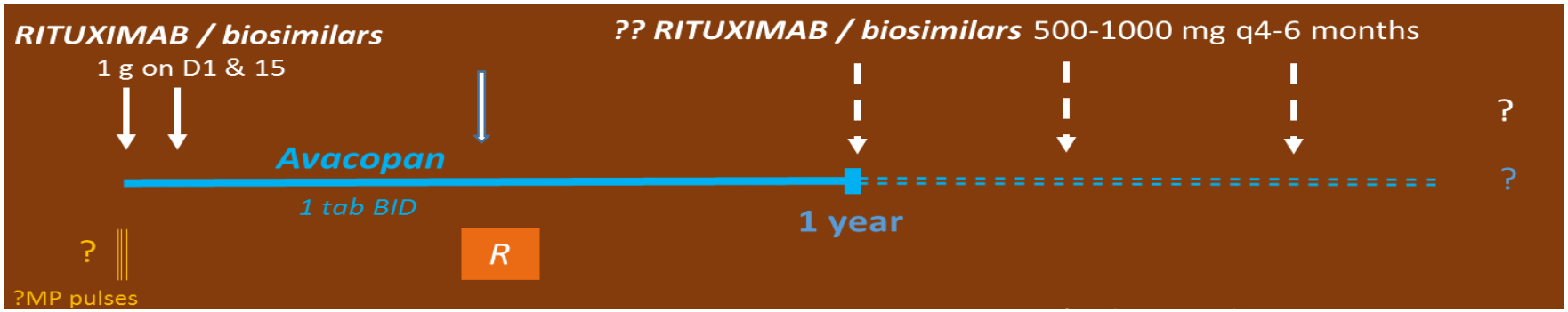
Patients with lower GFR may benefit from greater GFR recovery.

British Society for Rheumatology

Recommendation 2a. All people with lived experience of active GPA or MPA should be assessed for induction of remission treatment with immunosuppressants combined with glucocorticoids (GC) or Avacopan (GRADE 1A, SoA 99%).

Recommendation 5. Patients with active GPA or MPA should be considered for avacopan use as a steroid sparing agent, with or without a short course of glucocorticoids (tapering over four weeks) (GRADE 1A, SoA 96%).

Discussion points



Pagnoux, Fifi-Mah. Curr Treatm Opt Rheumatol 2021(7),112-33

- Are renal and non-renal benefits of avacopan maintained after stopping it?

- How to combine avacopan with rituximab?
- What is the optimal duration of avacopan? and others?

Dr. S Faguer

REVERSE

Newly diagnosed or relapsing active AAV (BVAS≥3) with eGFR 0-29 mL/min/1.7m2 at inclusion

Randomization with stratification on:

- eGFR (< 15 or ≥ 15 mL/min/1.7m²)

*RTX: 375 mg/m2 weekly for 4 weeks

**CYC: 0.5 g/m2 at D0 and weeks 2, 4, 7, 10 and 13 (reduced to 0.5 g according to kidney function)

Control group

H H H Cs H Cs H Cs Cs

D1 W4 W8 W12 W24 W36 W48 W52 W64

RTX* or CYC** ± PEx

RTX RTX

Prednisone

Placebo

Randomization

Experimental group

H H H Cs H Cs H Cs Cs

D1 W4 W8 W12 W24 W36 W48 W52 W64

RTX* or CYC** ± PEx

RTX RTX

Prednisone

Avacopan 30 mg b.i.d

Glucocorticoids or Avacopan in peopLe with high-risk renal Anti-Neutrophil cytoplasm antibody associated vasculitis Trial (GALANT)

To study the effects of avacopan compared to the PEXIVAS reduced-dose prednisone regimen in patients with AAV at high-risk of kidney failure in a pragmatic randomized open-label blinded end point trial

… and is definition of remission the same today?

- Survival
- BVAS or BVAS/WG = 0 with 5, 10 mg or 0 mg of prednisone?
- At month 3 or month 6?
- With less damage?

… and is definition of remission the same today?

- Survival
- BVAS or BVAS/WG = 0 with 5, 10 or 0 prednisone
- At month 3 or month 6
- With less damage?

- Should ANCA be negative?
- Repeat renal biopsy, hematuria, proteinuria?
- Renal biomarkers: uCD163, urinary T cells, serum C4 levels, uDKK-3 and uPRO-C6 and fibrosis

ANCA persistants: valeur prédictive ?

- Taux de rechute + élevé

  - Kyndt, AmJMed 1999 : 43p - 54% de rechutes, surtout si ANCA persistant : 86% vs 20% (p = 0.0001)

  - Slot, Arthritis Rheum 2004: 33p PR3-ANCA+, lors du switch CYC → AZA: RR rechute = 2.6 si ANCA+

  - Girard, Rheumatol 2001 : 55p dont 43 ANCA+ au diagnostic: 9/9 rechutes si ANCA+, vs 13/34 si neg

ARTHRITIS & RHEUMATISM
Vol. 58, No. 9, September 2008, pp 2908–2918
DOI 10.1002/art.23800

Predictors of Treatment Resistance and Relapse in Antineutrophil Cytoplasmic Antibody–Associated Small-Vessel Vasculitis

Comparison of Two Independent Cohorts

Christian Pagnoux,[1] Susan L. Hogan,[2] Hyunsook Chin,[2] J. Charles Jennette,[2] Ronald J. Falk,[2] Loïc Guillevin,[1] and Patrick H. Nachman[2]

1/ PR3-ANCA+

2/ lung involvement (and ENT)

3/ age ≤75 years and eGFR ≥30mL/min/1.73m²

→ *Relapses +*

RMD Open
Rheumatic & Musculoskeletal Diseases

ORIGINAL RESEARCH

Score to assess the probability of relapse in granulomatosis with polyangiitis and microscopic polyangiitis

Maxime Samson ,[1] Hervé Devilliers,[2] Sara Thietart,[3] Pierre Charles,[4] Christian Pagnoux,[5] Pascal Cohen,[6] Alexandre Karras,[7,8] Luc Mouthon,[6] Benjamin Terrier ,[6] Xavier Puéchal ,[6] Loic Guillevin[6]

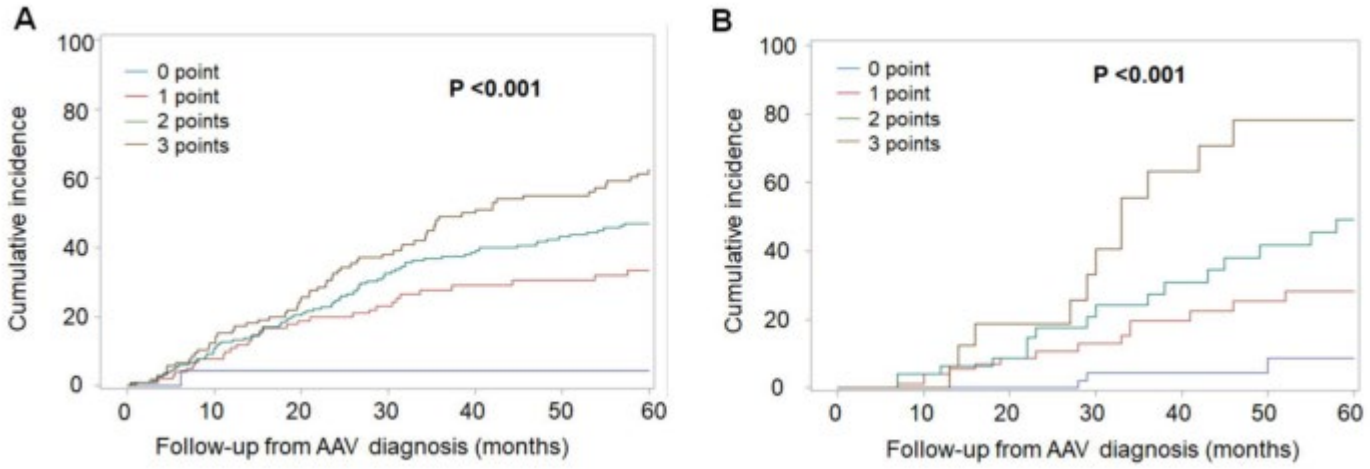


Table 4 Risk of relapse (cumulative incidence function) in each cohort depending on the FRS at AAV diagnosis

	Development cohort (n=427)			Validation cohort (n=209)		
FRS	Risk of relapse 2 years after diagnosis	Risk of relapse 3 years after diagnosis	Risk of relapse 5 years after diagnosis	Risk of relapse 2 years after diagnosis	Risk of relapse 3 years after diagnosis	Risk of relapse 5 years after diagnosis
0 point	2 (0.4–13.8)	3 (0.6–20.4)	4 (0.6–25.8)	2 (0.6–6.0)	5 (1.5–14.4)	8 (2.3–26.8)
1 point	18 (13.1–24.2)	27 (19.8–37.0)	31 (23.5–41.3)	8 (4.1–15.9)	19 (11.2–31.2)	30 (19.8–47.0)
2 points	25 (19.4–31.3)	37 (30.6–43.8)	42 (36.1–47.9)	14 (7.4–26.4)	31 (20.2–47.4)	48 (33.0–69.1)
3 points	34 (28.5–41.1)	49 (41.0–58.6)	55 (47.6–63.2)	29 (17.3–47.0)	56 ((39.8–79.0)	76 (58.2–100)

Seminars in Arthritis and Rheumatism 51 (2021) 1011–1015

Contents lists available at ScienceDirect

Seminars in Arthritis and Rheumatism

journal homepage: www.elsevier.com/locate/semarthrit

ANCA status and renal parameters at month 12 post-diagnosis can help predict subsequent relapses in patients with granulomatosis with polyangiitis

Lindsay K. Cho*, Simon Carette, Christian Pagnoux

Table 3

Univariable and multivariable survival analyses for the risk of late relapses (after 12-month follow-up) in patients with GPA based on predictors of relapse at the time of diagnosis and M12.

	Univariable HR	*P* value	Multivariable HR (95% CI)	*P* value
Characteristics at diagnosis				
ANCA status at diagnosis				
PR3-ANCA	0.45	0.02		
MPO-ANCA	0.76	0.53		
Serum creatinine level (μmol/L)	0.99	0.15		
BVAS-v3	0.99	0.55		
Characteristics at M12				
ANCA status at M12				
PR3-ANCA	1.16	0.60	1.20 (0.66–2.22)	0.55
MPO-ANCA	2.35	0.04	3.54 (1.29–9.74)	0.01
Atypical ANCA	0.99	0.99	2.30 (0.45–11.8)	0.32
PR3- MPO-ANCA	1.28	0.81	1.46 (0.19–11.1)	0.71
Microhematuria	1.48	0.18	1.91 (1.03–3.52)	0.04
Serum creatinine level (μmol/L)	0.99	0.08	0.99 (0.98–0.99)	0.04
Relapses ≤ M12	0.86	0.62	1.06 (0.54–2.1)	0.87

EPIDEMIOLOGICAL SCIENCE

The effect of achieving serological remission on subsequent risk of relapse, end-stage renal disease and mortality in ANCA-associated vasculitis: a target trial emulation study

Gregory McDermott [1], Xiaoqing Fu,[2] Claire Cook,[2] Catherine Ahola,[2] Brett Doliner,[3] Jennifer Hanberg,[3] John H Stone [4], Hyon K Choi [2], Yuqing Zhang [2], Zachary S Wallace [2]

Patients treated to a negative ANCA assay within 180 days had HR 0.55 (95% CI 0.38 to 0.81) of relapse

Outcome	Serological remission within 180 days HR (95% CI)	Persistently positive titre at 180 days
Adjusted HR* for relapse		
PR3-ANCA+	0.52 (0.20 to 1.33)	1.0 (Ref)
MPO-ANCA+	**0.62 (0.40 to 0.96)**	1.0 (Ref)
Renal involvement at baseline	0.64 (0.39 to 1.03)	1.0 (Ref)
RTX or RTX/CYC treated	**0.55 (0.33 to 0.92)**	1.0 (Ref)
CYC only treated	0.47 (0.21 to 1.03)	1.0 (Ref)
TPE treated	0.49 (0.10 to 2.49)	1.0 (Ref)

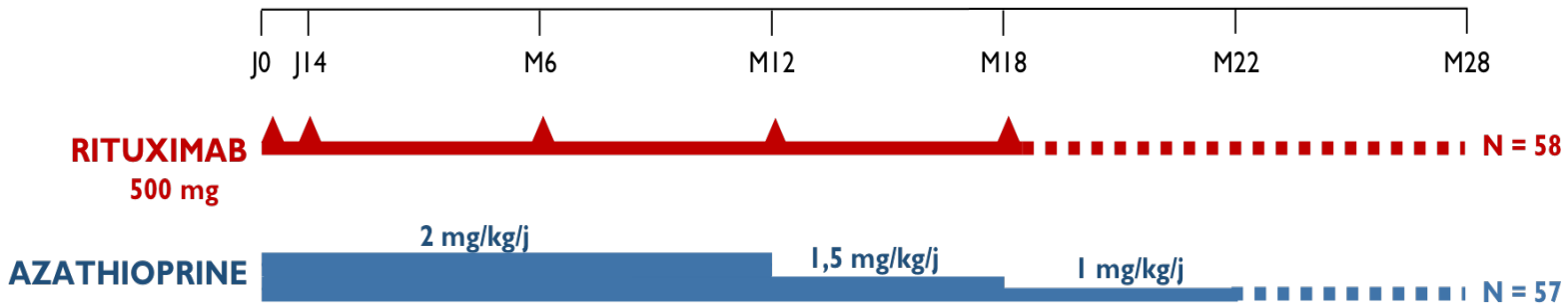
MAINRITSAN 1

The NEW ENGLAND JOURNAL of MEDICINE

ESTABLISHED IN 1812 NOVEMBER 6, 2014 VOL. 371 NO. 19

Rituximab versus Azathioprine for Maintenance in ANCA-Associated Vasculitis

L. Guillevin, C. Pagnoux, A. Karras, C. Khouatra, O. Aumaître, P. Cohen, F. Maurier, O. Decaux, J. Ninet, P. Gobert, T. Quémeneur, C. Blanchard-Delaunay, P. Godmer, X. Puéchal, P.-L. Carron, P.-Y. Hatron, N. Limal, M. Hamidou, M. Ducret, E. Daugas, T. Papo, B. Bonnotte, A. Mahr, P. Ravaud, and L. Mouthon, for the French Vasculitis Study Group*

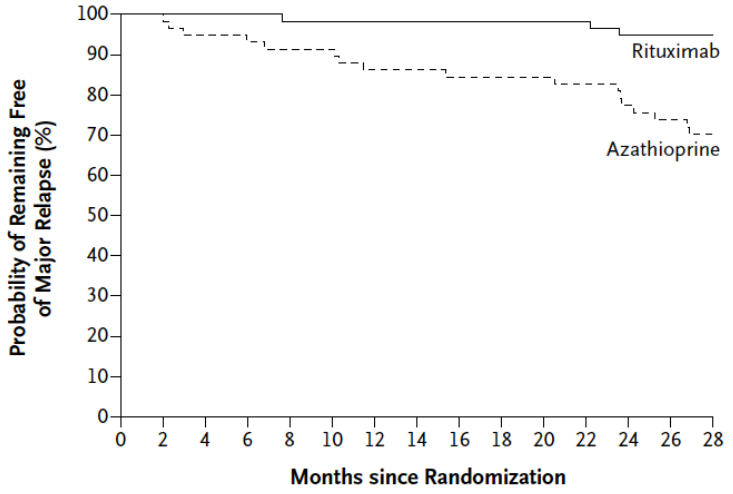


115 patients

Major relapses :

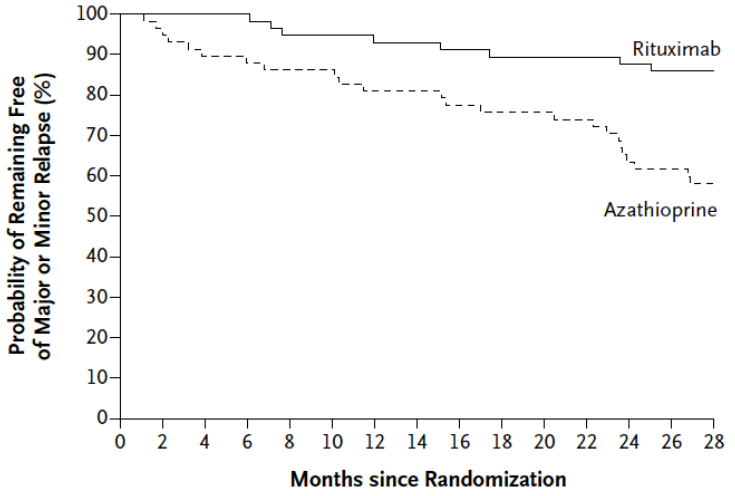
RTX 5 %
AZA 29 % } HR 6.6 (95% CI 1.6-27.9)

No difference in SAE



No. at Risk

Rituximab	57	57	57	57	56	56	56	56	56	56	56	56	54	52	39
Azathioprine	58	58	55	54	53	53	50	50	48	48	48	47	44	41	33



No. at Risk

Rituximab	57	57	57	57	54	54	53	53	52	51	51	51	50	47	36
Azathioprine	58	56	52	51	50	50	47	47	44	43	43	42	36	35	30

- In patients with GPA or MPA who received CYC or RTX induction therapy, **RTX (infusions every 4–6 months) is recommended as first-line maintenance therapy.**
- Recommendation: For patients with severe GPA/MPA whose disease has entered remission after treatment with cyclophosphamide or rituximab, we conditionally **recommend treatment with rituximab** over methotrexate or azathioprine **for remission maintenance**..
- For maintenance of remission of GPA and MPA, after induction of remission with either rituximab or cyclophosphamide, we **recommend treatment with rituximab**. Azathioprine or methotrexate may be considered as alternatives.
- Recommendation 6a. Following induction of remission with an RTX or CYC-based treatment regimen, we recommend maintenance of remission with RTX in preference to other agents (GRADE 1A, SoA 98%).

→ For how long?

→ For everyone?

Chung et al. Arthritis Rheumatol. 2021 Aug;73(8):1366-1383
Mendel et al. J Rheumatol. 2021 Apr;48(4):555-566
Hellmich et al. ARD. 2023
Biddle BSR 2025

9.3.2 Maintenance therapy

Recommendation 9.3.2.1: We recommend maintenance therapy with either rituximab, or azathioprine and low-dose glucocorticoids after induction of remission *(1C)*.

This recommendation places a higher value on prevention of relapses and a relatively lower value on adverse events related to immunosuppressive drugs.

KDIGO 2024 CLINICAL PRACTICE GUIDELINE
FOR THE MANAGEMENT OF ANTINEUTROPHIL CYTOPLASMIC
ANTIBODY (ANCA)–ASSOCIATED VASCULITIS

Rituximab preferred	Azathioprine preferred
• Relapsing disease • PR3–ANCA disease • Frail older adults • Glucocorticoid-sparing especially important • Azathioprine allergy	• Low baseline IgG (<300 mg/dl) • Limited availability of rituximab

Maintenance in renal patients with ESKD?

- Internists/Rheumatologists vs. nephrologists?

Practice Point 9.3.1.5: Discontinue immunosuppressive therapy after 3 months in patients who remain on dialysis and who do not have any extrarenal manifestations of disease.

?

Practice Point 9.3.1.5: Consider discontinuation of immunosuppressive therapy after 3 months in patients who remain on dialysis and who do not have any extrarenal manifestations of disease.

- MASTER-ANCA

Recommendation 7b. People living with severe renal involvement who remain dialysis dependent have a high risk of infection. Patients with renal limited disease who remain dialysis dependent may not require ongoing immunotherapy. Maintenance of remission therapy to prevent relapse should be balanced against the risks of immunosuppression (GRADE 2C, SoA 98%).

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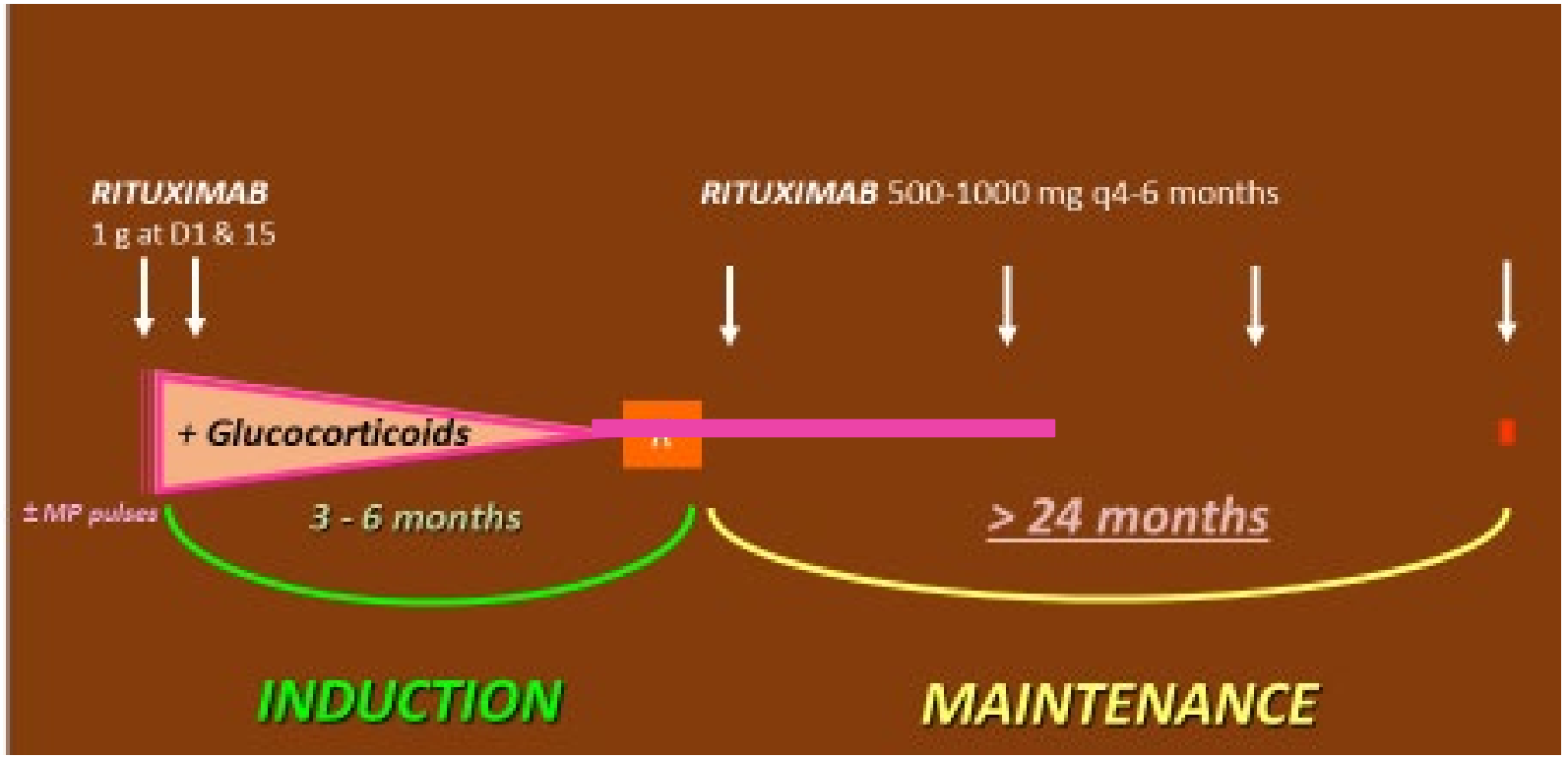
Duration of maintenance

- In patients with GPA or MPA, maintenance with RTX (or conventional immunosuppressants) should be continued for a minimum of 2 years (1b, B); extended maintenance therapy can be considered, especially in high-risk clinical subgroups
- We recommend that therapy to maintain remission for GPA and MPA be continued for 24–48 months following induction of remission of new-onset disease. (1a, B) *Longer duration of therapy should be considered in relapsing patients or those with an increased risk of relapse, but should be balanced against patient preferences and risks of continuing immunosuppression (4, D)
- Ungraded position statement: The duration of non-GC [or GC] remission maintenance therapy in GPA/MPA should be guided by the patient's clinical condition, preferences, and values.
- Recommendation 7a. Maintenance of remission should be continued for a period of 24-48 months (GRADE 1A, SoA 97%)

Table 4 Glucocorticoid dosing (mg/day, prednisolone equivalent) with rituximab or cyclophosphamide-based regimens for remission induction in GPA or MPA according to the PEXIVAS Study[93]

	Body weight (kg)		
Weeks	**<50**	**50–75**	**>75**
1*	50	60	75
2	25	30	40
3–4	20	25	30
5–6	15	20	25
7–8	12.5	15	20
9–10	10	12.5	15
11–12	7.5	10	12.5
13–14	6	7.5	10
15–18	5	5	7.5
19–52	5	5	5
>52	Individual taper	Individual taper	Individual taper

*Consider use of intravenous methylprednisolone at a cumulative dose of 1–3 g on days 1–3 in patients with severely active disease, including but not limited to renal involvement with a documented estimated glomerular filtration rate <50 mL/min/1.73 m^2 and/or diffuse alveolar haemorrhage.



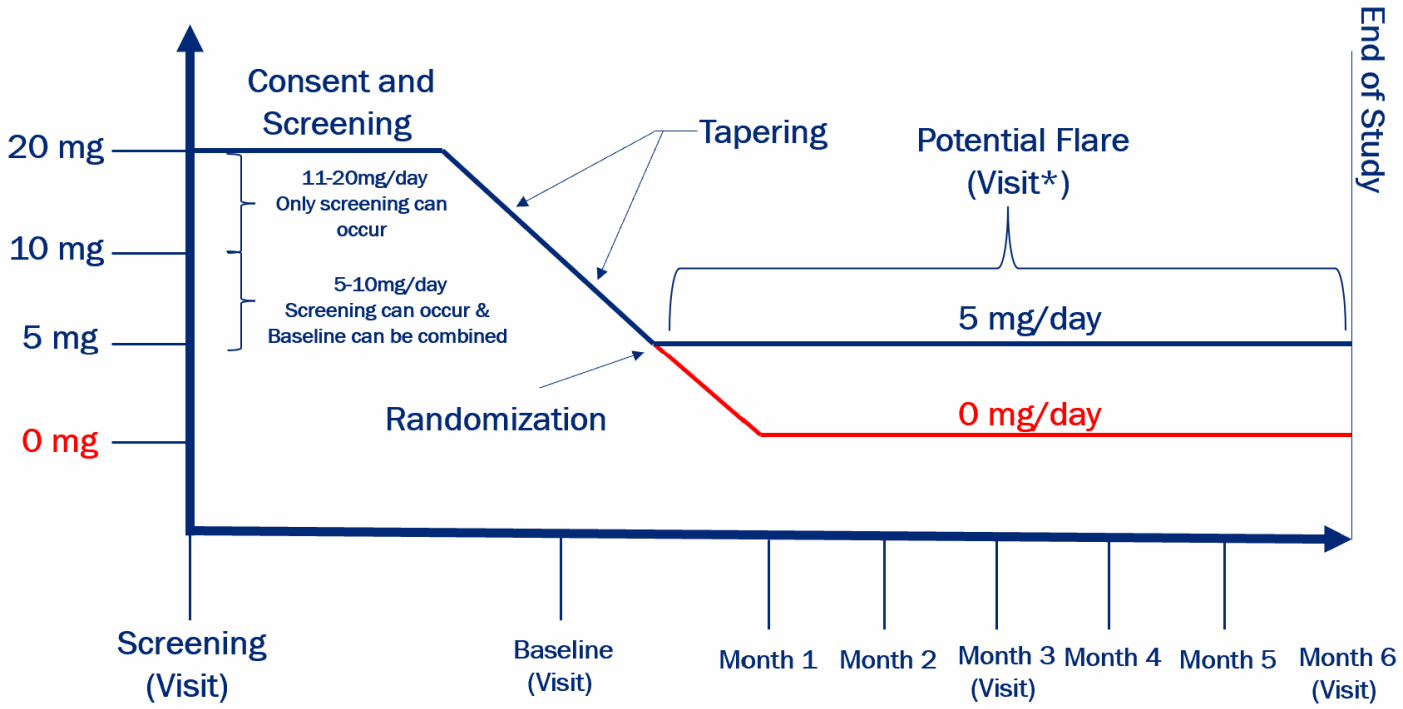
BSR 2025	The optimum length of treatment with GC during the maintenance phase is uncertain. Depending on concurrent immunosuppression, complete GC withdrawal may be possible within 6–12 months following induction of remission treatment	GRADE 2B, SoA 98%
CanVasc 2020 (Add. 2022)	Low-dose GC should be part of the initial remission maintenance therapy in GPA and MPA after remission is achieved; the optimal duration of low-dose GC for remission maintenance is not known.	Category 4, Strength D
EULAR 2022	*In text only: "there is little evidence […] and dosage need to be individualised on a shared decision basis."*	-
KDIGO 2024	*In text only: "[With AZA] Glucocorticoids should also be continued at 5–7.5 mg/d for 2 yr and then slowly reduced by 1 mg every 2 mo. [Following RTX induction] prednisolone can be withdrawn by 6 months."*	-
PANLAR 2023	For patients with GPA or MPA who achieved remission after induction therapy, we recommend against treatment with low doses of prednisone indefinitely.	Conditional, Very low, 100%

TAPIR: The assessment of prednisone in remission trial

Peter Merkel, Christian Pagnoux, Nader Khalidi, Ulrich Specks, Curry Koening, Carol Langford, Larry Moreland, Paul Monach, Jason Springer, Shubhasree Banerjee, Simon Carette, Rennie Rhee, Medha Soowamber, Kenneth Warrington, Renée Borchin, Cristina Burroughs, Carol McAlear, David Cuthbertson, Jeffrey Krischer, for the Vasculitis Clinical Research Consortium

TAPIR

Study Design and Objective



Revised from Merkel. ACR Convergence 2024,16S48: Plenary I (0774)

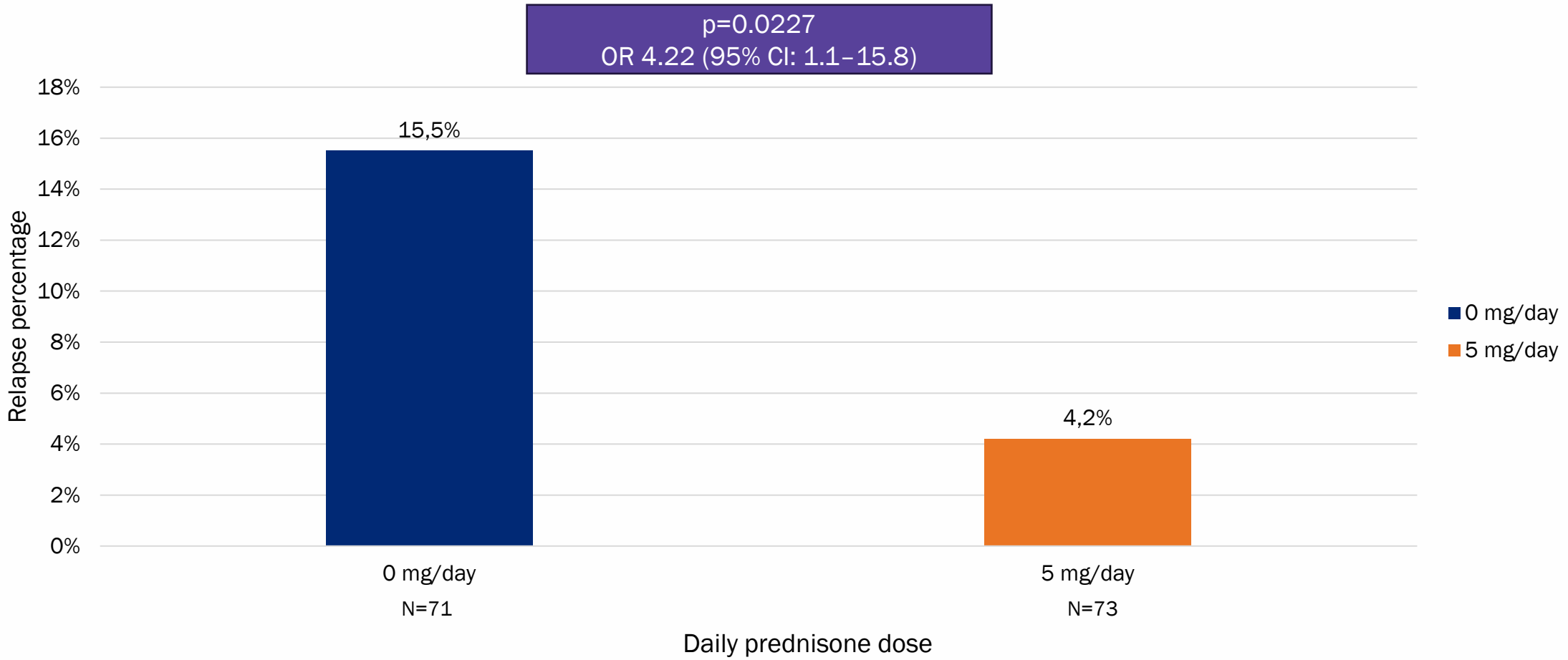
Objective

Evaluate the efficacy and safety of low-dose glucocorticoids for maintenance of remission of GPA.

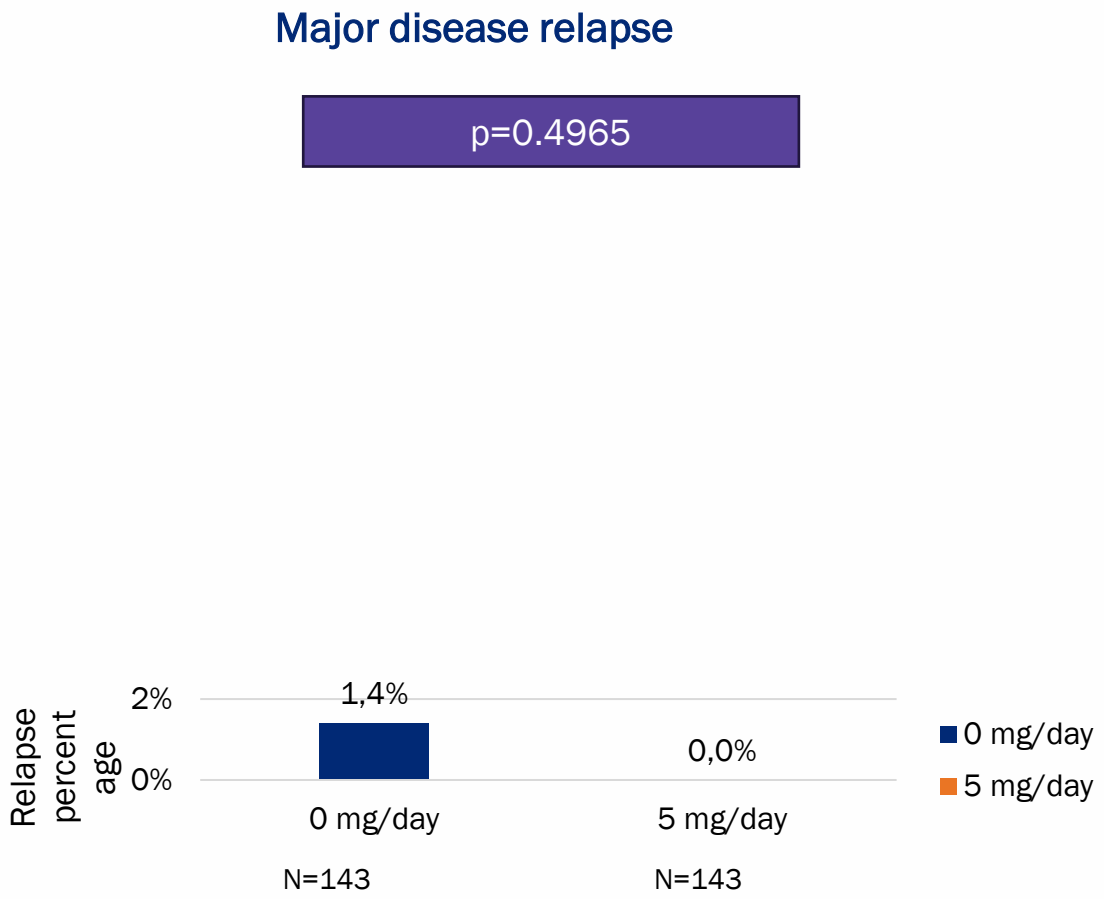
→ Primary hypothesis is a difference of ≥20% in the relapse rate *(13.2% to 32.9%)*

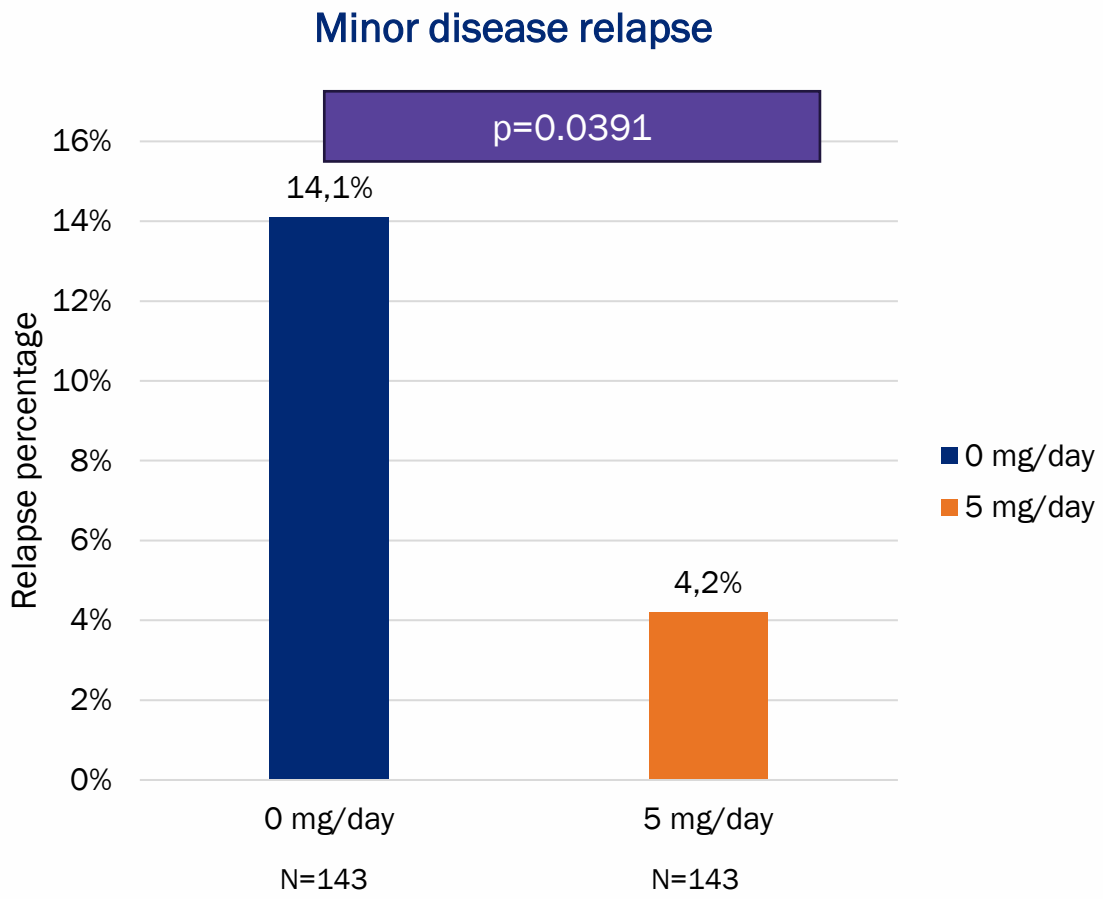
→ 142 patients 1:1
with 80% power, 2-tailed p of 0.0516

Relapse by month 6

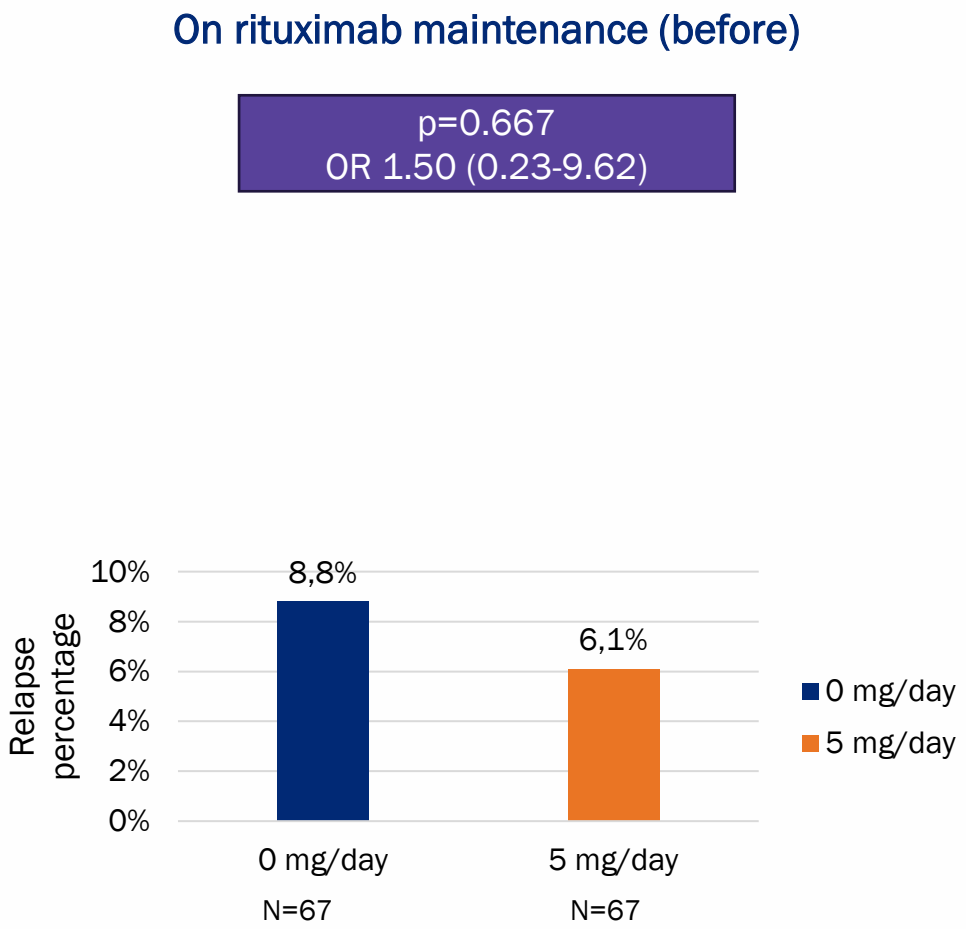


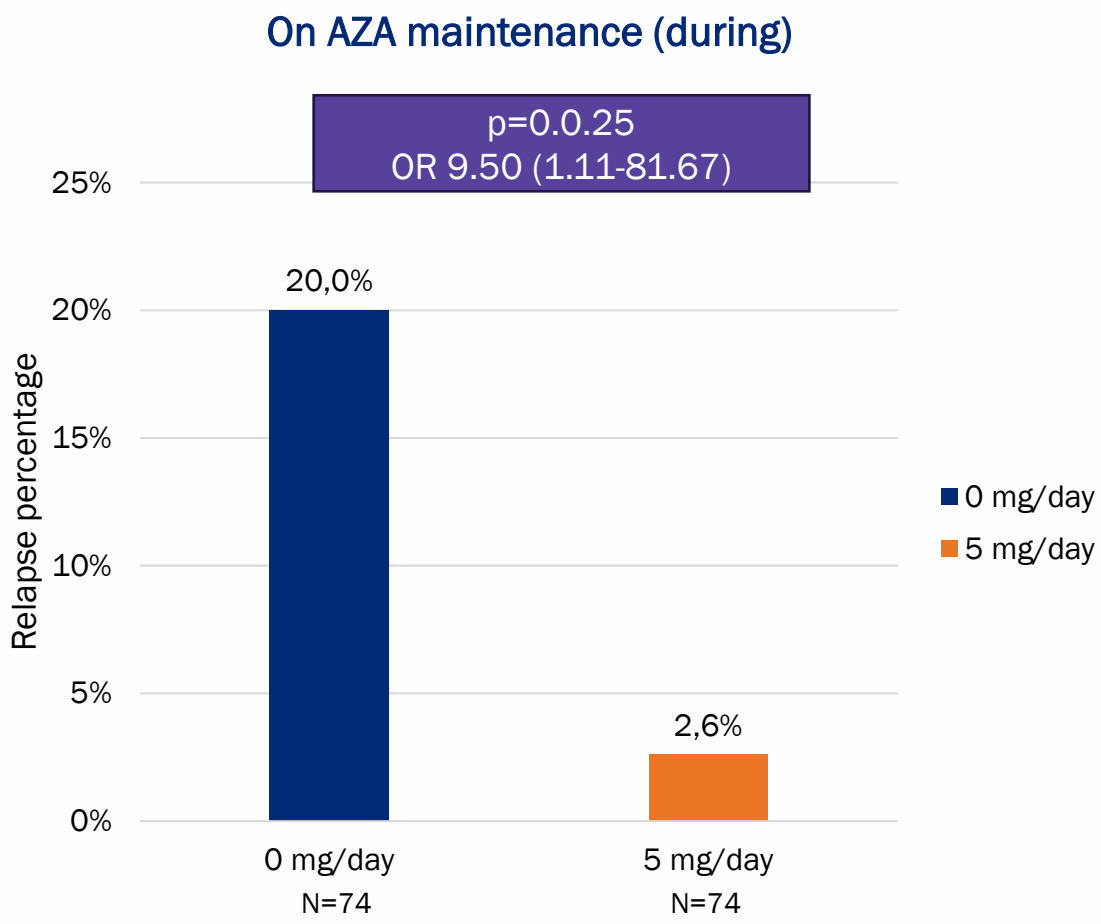
Relapse by month 6 stratified by type of relapse





Relapse by month 6 stratified by use of rituximab





OR, odds ratio.

Suivi des ANCA : valeur prédictive ?

- Réascension ou réapparition fréquentes des ANCA avant les rechutes

- Cohen Tervaert, Arch Intern Med 1989 : 35p – 49% de rechutes, toutes précédées d'une réascenscion du taux ANCA (dans les 3-6 mois pour moitié)

- Jayne, QJM 1995 : 60p - 38% de rechutes dont 57% précédées d'une réascension des ANCA (83% ANCA+ lors du diagnostic de rechute)

- Boomsma, Arthritis Rheum 2000 : 100p, 37% de rechutes; 43% avec réascension des cANCA et 29% de ceux avec ré↑ des PR3-ANCA n'ont pas rechuté

- Kemna, J Am Soc Nephrol 2015 : mesure répétée des ANCA associée aux rechutes chez les patients avec atteinte rénale (HR 11.09 [95%CI 5.01–24.55]) mais pas les autres (HR 2.79 [1.30–5.98])

- Fussner, Arthritis Rheum 2016 : réascenscion des ANCA associée aux rechutes sévères chez les patients sous RTX (HR 5.80 [95%CI 2.06–19.77]) mais pas sous CYC-AZA (HR 2.84 [0.87–11.40])

CONCLUSION

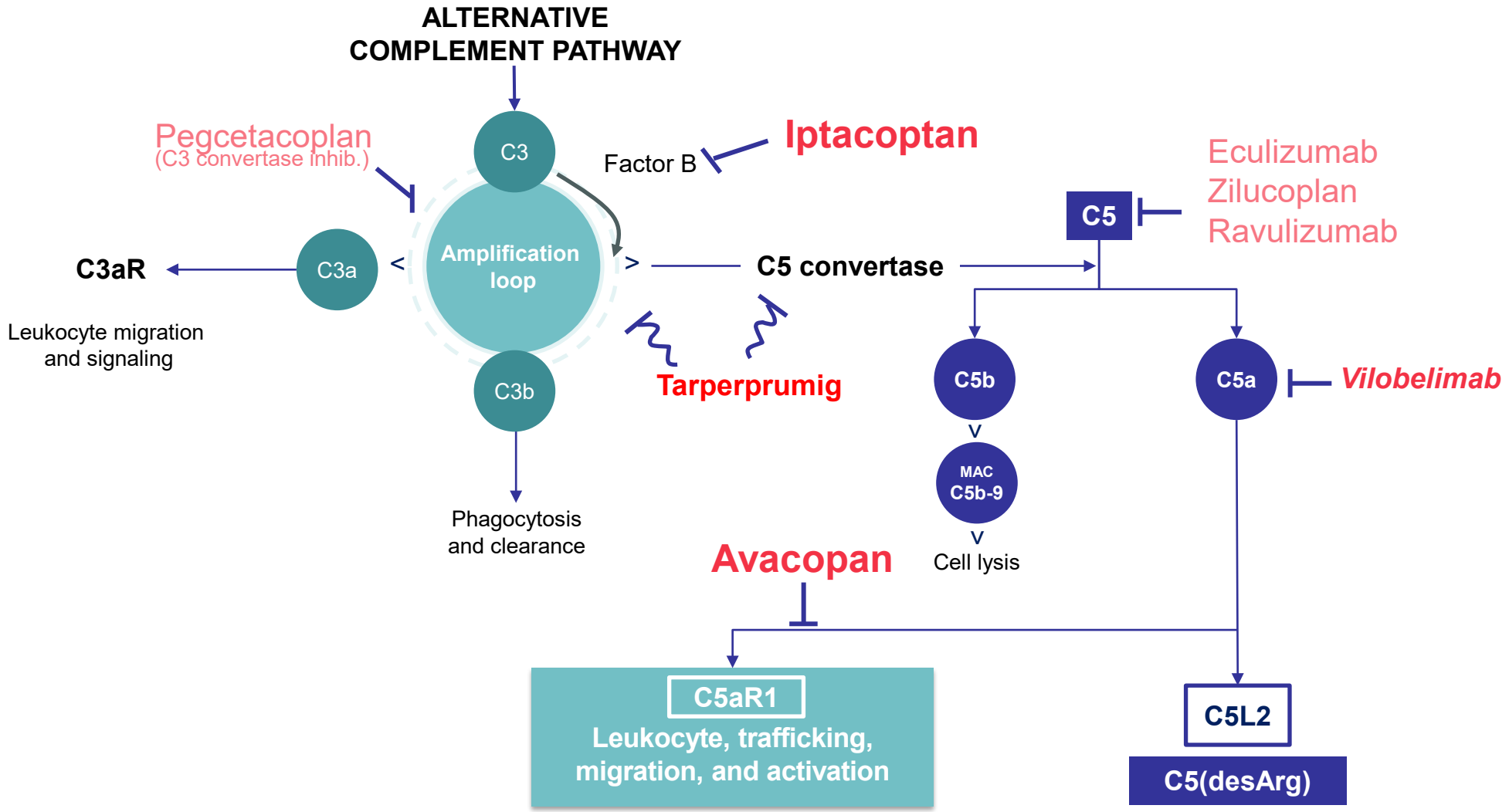
Refractory disease

- Patients with refractory disease, and those in whom the aforementioned therapies are contraindicated or poorly tolerated, should be managed in a referral center for vasculitis in collaboration with subspecialists, for consideration of alternate, additional, and/or experimental therapies. Category 4, Strength D

- Recommendation 23a. People living with AAV should be cared for in cohorted* rather than general clinics (*where people living with AAV are grouped together and seen in a dedicated clinic) (GRADE 1B, SoA 98%).

Can we do better now / in the future (new drugs / trials)?

- Obinutuzumab, daratumumab, antiBLys/APRIL
- Complement: factor B inhibitor, factor H agonist, C3 inhibitor, anti-properdin



C5a, complement component 5a; C5aR1, C5a receptor 1; MAC, membrane attack complex.

1. Bekker P, Dairaghi D, Seitz L, et al. PLoS One. 2016 Oct 21;11(10):e0164646.
2. Thurman JM, Holers VM. J Immunol. 2006 Feb 1;176(3):1305-10.
3. TAVNEOS Product Monograph. 2022/04/19. Otsuka Canada for: VIFOR FRESENIUS MEDICAL CARE RENAL PHARMA LTD.
4. SOLIRIS Product Monograph. 2021/03/25. Alexion Pharma GmbH.
5. Tesar V, Hruskova Z. Front Immunol. 2022 Jul 8;13:888816.

Can we do better now / in the future (new drugs / trials)?

- Obinutuzumab, daratumumab, antiBLys/APRIL
- Complement: factor B inhibitor, factor H agonist, C3 inhibitor, anti-properdin
- Anticlaudin-1 & antifibrotic agents
- FCγRn (and IDeS)
- Anti-GM-CSF

- Bispecific Abs / BITE
- Cathepsin C or G inhibitors
- *SAL-T*

- CAR-T cells (CD19, BCMA)

22nd International Vasculitis Workshop

21 - 25 February 2026
Melbourne, Australia

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Supported by Melbourne Convention Bureau, Melbourne Australia

Key Dates

Early bird registration opens
28 March 2025

Call for abstracts open
28 March 2025

Call for abstracts close
19 September 2025

Early bird registration closes
31 October 2025

Presenter registration deadline
31 October 2025

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