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Medication adherence and real-world factors of first line induction treatment selection in ANCA-associated vasculitis

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Abstract. *Background & Objective:* This study aimed to explore real-world factors that may influence the selection of first-line remission-induction therapy in ANCA-associated vasculitis (AAV), as well as to evaluate physician compliance with guideline recommendations and patient adherence to maintenance therapies. *Methods:* A retrospective analysis of 112 patients with AAV, including granulomatosis with polyangiitis (GPA, 67%), microscopic polyangiitis (MPA, 13%), and eosinophilic granulomatosis with polyangiitis (EGPA, 20%), was conducted at a single tertiary care center using electronic health records from 2018 to 2023. Treatment regimens, patient demographics, organ involvement, and adherence to EULAR guidelines were analyzed. Patients receiving rituximab (RTX) or cyclophosphamide (CYC) as first-line remission-induction therapy were compared. Compliance was defined as alignment with guideline-recommended dosing and timing of induction therapy, while adherence during maintenance was evaluated based on consistency with prescribed regimens, assessed via prescription refill data. *Results:* Of the 102 patients included in the study, 85 received CYC and 17 received RTX as first-line remission induction therapy. Compared to the RTX group, those receiving CYC were significantly older (median age 57 vs. 44 years, $p < 0.05$), had higher BVAS scores (median 12 vs. 10, $p = 0.02$), and exhibited more comorbidities (74% vs. 35%). Organ involvement rates were similar in both groups. No significant differences in major organ involvement were observed between the two groups. RTX adherence was 100% in both the induction and maintenance phases, whereas adherence to oral maintenance therapies was notably lower, at 66% for methotrexate, 36% for mycophenolate mofetil, and 28% for azathioprine. *Conclusion:* In real-world practice, older age, higher BVAS, and a greater comorbidity burden appear to influence clinicians' preference for CYC over RTX as first-line induction, despite similar organ involvement between groups. Overall compliance with induction guidelines was high, but adherence to oral maintenance regimens remained suboptimal. These findings underscore the need for personalized treatment strategies and targeted measures to enhance long-term medication adherence in AAV.

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Introduction

Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) encompasses granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA), and eosinophilic granulomatosis with polyangiitis (EGPA) (1, 2). These diseases

primarily affect small and medium-sized vessels, leading to multiorgan involvement. AAV is characterized by neutrophil-driven vascular inflammation, resulting in endothelial and tissue injury (3). Loss of tolerance to neutrophilic proteins, namely proteinase 3 (PR3) and myeloperoxidase (MPO), plays a central role in disease pathogenesis (2, 3). Immunofluorescence typically reveals minimal immunoglobulin and complement deposition, hence the term 'pauci-immune' vasculitis (2). GPA and MPA commonly present with necrotizing crescentic glomerulonephritis and pulmonary capillaritis (4), whereas EGPA is classically characterized by late-onset asthma, nasal polyposis, and eosinophilia in peripheral blood and/or tissue (5). AAV treatment consists of two distinct phases: remission-induction and remission-maintenance (6). The primary goal of induction therapy is to rapidly suppress inflammation and prevent irreversible organ damage. Current standard induction regimens for severe AAV involve glucocorticoids (GCs) combined with either rituximab (RTX) or cyclophosphamide (CYC), as recommended by the latest EULAR guidelines (1, 7, 8). In non-severe cases, methotrexate (MTX), azathioprine (AZA), and mycophenolate mofetil (MMF) are considered alternative options (1). Following successful induction, the primary goal of maintenance therapy is to sustain remission and prevent relapse. This phase typically involves low-dose corticosteroids combined with AZA, MMF, MTX, or RTX (9). While MTX and AZA demonstrate comparable efficacy in remission-maintenance, RTX has shown superior relapse prevention (9). Frequent clinical relapses often necessitate repeated induction therapy, increasing the risk of treatment-related adverse effects (10).

Although treatment guidelines outline remission induction and maintenance strategies for AAV, they do not establish strict criteria for choosing between CYC and RTX as first-line induction therapy. In real-world practice, treatment decisions appear to be shaped by multiple factors, yet whether physicians follow a specific determinant or rely on broader clinical intuition remains uncertain. Given the chronic nature of AAV and the necessity for long-term immunosuppression, ensuring patient adherence is crucial for achieving relapse-free survival. Recently, lower age and higher BVAS, upper respiratory tract involvement, renal involvement and non-adherence to treatment have been shown to be predictors of vasculitis-related damage (11). While adherence variability is well-documented in other rheumatic diseases, ranging from 34% to 93% depending on the condition (12, 13), specific data on adherence rates in AAV remain scarce.

To address this gap, we conducted a real-world analysis to explore factors that may influence the selection of first-line remission-induction therapy in AAV. Additionally, we evaluated physician compliance with guideline recommendations and patient adherence to maintenance therapies, aiming to provide insights into long-term treatment strategies in clinical practice.

Methods

Study design and participants

This retrospective study analyzed data from 112 patients diagnosed with ANCA-associated vasculitis (AAV) at a single tertiary care rheumatology center in between January 1, 2018, and December 31, 2023. Inclusion criteria required a clinically confirmed diagnosis of AAV in patients aged 18 years or older. Patients were excluded if they had incomplete medical records and unverified diagnoses. Patient data were extracted from electronic medical records using a standardized data collection form by researchers blinded to the study hypotheses. Collected data included demographic details, laboratory findings, comorbidities, disease activity measures (assessed using the Birmingham Vasculitis Activity Score (BVAS)), and specific organ involvements. Organ involvement was defined according to the BVAS criteria (14, 15). Treatment-related data included details on remission-induction and remission-maintenance therapies, medication types and doses, treatment intervals, and any modifications made during therapy. Treatment choices were compared against the EULAR guidelines applicable at the time of decision-making. The remission induction period was defined as the six months following diagnosis. In our center, we used a standardized steroid protocol for AAV patients. All patients received an initial dose of IV methylprednisolone (500–1,000 mg daily for 1–3 days), followed by oral prednisone (0.5–1 mg/kg/day) with a planned tapering schedule as clinically indicated. Compliance with remission-induction therapy was defined as receiving the guideline-recommended dose and timing of intravenous CYC or RTX within the six-month induction period, with

no more than a two-week delay in any individual dose, based on clinical expert consensus. Any significant deviation from these regimens, such as dose reductions or delays without clinical justification, was classified as non-compliance. Adherence to remission-maintenance therapy was assessed using both prescription refill data and physician documentation of patient-reported non-adherence or missed doses. For intravenous RTX and CYC, adherence was evaluated based on the timing of scheduled doses, with delays of more than two weeks considered non-adherent. For oral therapies, including azathioprine, methotrexate, and mycophenolate mofetil, adherence was assessed based on pharmacy refill records. First, we examined whether there were differences in organ involvement, disease severity, and demographics between patients receiving CYC or RTX as first-line remission-induction therapy. Additionally, we assessed physician compliance with guideline-recommended induction therapy and patient adherence to maintenance regimens to gain insights into long-term treatment patterns in real-world practice. The study was conducted in accordance with the principles of the Declaration of Helsinki and was approved by the Institutional Review Board of Ankara Bilkent City Hospital (E1-23-3801). Due to its retrospective design, informed consent was waived. All collected data were anonymized to ensure patient confidentiality.

Statistical analysis

Statistical analyses were performed using SPSS Statistics for Windows, Version 21 (IBM Corp., Armonk, NY). Descriptive statistics were used to summarize patient characteristics. Continuous variables were assessed for normality using the Shapiro-Wilk test and presented as means with standard deviations (SD) for normally distributed data or medians with interquartile ranges (IQR) for non-normally distributed data. Categorical variables were presented as frequencies and percentages. Comparisons of normally distributed continuous variables were conducted using independent t-tests, while non-normally distributed variables were analyzed using the Mann-Whitney U test. Categorical variables were compared using the Chi-square test or Fisher's exact test when expected cell counts were less than five. A p-value of <0.05 was considered statistically significant.

Results

The study included 112 patients with a confirmed diagnosis of AAV. The cohort comprised 75 (67%) patients with GPA, 22 (20%) with EGPA, and 15 (13%) with MPA. Detailed demographic and clinical characteristics, stratified by AAV subtype, are presented in Table 1. Female patients accounted for 45% of GPA, 55% of EGPA, and 67% of MPA cases. The median age (IQR) varied by subtype: 53 years

Table 1. Baseline Characteristics in Patients with AAV Subtypes

Characteristics	GPA (n=75)	EGPA (n=22)	MPA (n=15)
Gender, Female, n (%)	34 (45%)	12 (55%)	10 (67%)
Age, Median (IQR)	53(43-60)	56 (49-63)	64 (61-73)
Smoking Status, Ever Smoked (%)	22 (29.3%)	6 (27.3%)	2 (13.3%)
At Least One Comorbidity, n (%)	43 (57%)	18 (82%)	15 (100%)
Diabetus Mellitus (%)	12 (16.0)	2 (9.1)	2 (13.3)
Hypertension (%)	23 (30.7)	9 (40.9)	8 (53.3)
Hyperlipidemia (%)	2 (2.7)	2 (9.1)	2 (13.3)
Coronary Artery Disease (%)	6 (8.0)	2 (9.1)	4 (26.7)
Chronic Kidney Disease (%)	5 (6.7)	0 (0.0)	4 (26.7)
Kidney Failure (Dialysis) (%)	9 (12.0)	0 (0.0)	2 (13.3)
Congestive Heart Failure (%)	1 (1.3)	4 (18.2)	1 (6.7)
Hypothyroidism (%)	6 (8.0)	2 (9.1)	4 (26.7)
Osteoporosis (%)	5 (6.7)	2 (9.1)	3 (20.0)
Malignancy (%)	2 (2.7)	0 (0.0)	0 (0.0)
Thrombosis (%)	5 (6.7)	4 (18.2)	5 (33.3)
PR3-ANCA Positivity (%)	89%	0%	0%
MPO-ANCA Positivity (%)	5%	32%	100%
BVAS Score, Median (IQR)	12 (8–16)	10 (7–14)	17 (12–18)
Life-Threatening Organ Involvement (%)	71%	41%	87%

Abbreviations: AAV: ANCA-associated vasculitis; GPA: Granulomatosis with Polyangiitis; EGPA: Eosinophilic Granulomatosis with Polyangiitis; MPA: Microscopic Polyangiitis; BVAS: Birmingham Vasculitis Activity Score; IQR: Interquartile Range; PR3-ANCA: Proteinase 3-Antineutrophil Cytoplasmic Antibodies; MPO-ANCA: Myeloperoxidase-Antineutrophil Cytoplasmic Antibodies.

(43–60) for GPA, 56 years (49–63) for EGPA, and 64 years (61–73) for MPA. Disease activity, assessed by BVAS, was highest in MPA (median 17, IQR 12–18), followed by GPA (12, IQR 8–16) and EGPA (10, IQR 7–14). Life-threatening organ involvement was present in a majority of GPA (71%) and MPA (87%) patients, but less frequent in EGPA (41%). Of the 112 patients initially identified, 102 received either CYC or RTX as first-line remission-induction therapy. The remaining 10 patients, as detailed in Table 2, received alternative initial treatments: 6 with GPA received methotrexate, and 4 with EGPA received azathioprine. 85 patients received CYC and 17 patients received RTX as first-line remission induction therapy) (Table 2). CYC-treated patients were significantly older than those treated with RTX (median age 57 vs. 44 years, p = 0.03). Comorbidities were also more prevalent in the CYC group (74.1% vs. 35.3%, p = 0.002). BVAS scores was higher in CYC-treated patients (median 12 vs. 10, p = 0.02). Constitutional symptoms were significantly more frequent in the CYC group (48.2% vs. 17.6%, p = 0.03). Although skin involvement was more frequent in RTX-treated patients (29.4% vs. 11.8%), this difference did not reach statistical significance (p = 0.14). No significant differences were found in gender distribution (p = 0.67), AAV subtype (p = 0.45), ANCA serotype (p = 0.52), Five Factor Score (p = 0.21), or other organ involvement rates (p = 0.38) (Table 3). A case bycase analysis was performed to evaluate whether any other condition were noted that may influenced treatment selection. This analysis revealed that RTX was preferentially used in one patient with risk of CYC's gonadal toxicity, one patient with a history of malignancy, one patient with cytopenia, and one patient with chronic liver disease, suggesting that these factors played a role in induction therapy selection.

Table 2. Chosen Agents for Induction and Maintenance in ANCA-Associated Vasculitis

Agent	GPA (n=75)	EGPA (n=22)	MPA (n=15)
Remission-Induction			
Cyclophosphamide	56 (75%)	15 (68%)	14 (93%)
RTX firstline	13 (17%)	3 (14%)	1 (7%)
Methotrexate	6 (8%)	0	0
Azathioprine	0	4 (18%)	0
Plasmapheresis	20 (27%)	1 (5%)	6 (40%)
IVIg	6 (8%)	5 (23%)	1 (7%)
Remission-Maintenance			
Methotrexate	14 (19%)	3 (14%)	1 (7%)
Mycophenolate mofetil	8 (11%)	3 (14%)	4 (27%)
Azathioprine	37 (49%)	12 (54%)	7 (47%)
Rituximab	16 (21%)	4 (18%)	3 (20%)

Abbreviations: CYC: Cyclophosphamide; RTX: Rituximab, GPA: Granulomatosis with Polyangiitis; EGPA: Eosinophilic Granulomatosis with Polyangiitis; MPA: Microscopic Polyangiitis.

Table 3. Characteristics of Patients: CYC Firstline vs. RTX Firstline

Characteristic	CYC (n=85)	RTX (n=17)	P value
Gender, Female, n (%)	40 (47.1%)	9 (52.9%)	0.66
Age, Median (IQR)	57 (47-63)	44 (33-55)	0.03
At Least One Comorbidity, n (%)	63 (74.1%)	6 (35.3%)	0.002
GPA	56 (65.9%)	13 (76.5%)	0.57
EGPA	15 (17.6%)	3 (17.6%)	1
MPA	14 (16.5%)	1 (5.9%)	0.45
PR3-ANCA Positivity (%)	49 (57.6%)	13 (76.5%)	0.18
MPO-ANCA Positivity (%)	20 (23.5%)	3 (17.6%)	0.75
BVAS Score, Median (IQR)	12 (8-17)	10 (7-12)	0.02
Five Factor Score ≥1 (%)	62 (73%)	14 (70%)	0.55
Constitutional Symptoms, n (%)	41 (48.2%)	3 (17.6%)	0.03
Skin Involvement, n (%)	10 (11.8%)	5 (29.4%)	0.14
Peripheral Nervous System Involvement, n (%)	12 (14.2%)	2 (11.8%)	1
Pulmonary Involvement, n (%)	63 (74.1%)	13 (76.5%)	1
Renal Involvement, n (%)	39 (%45.9)	5 (29.4%)	0.28
Life-Threatening Organ Involvement (%)	56 (69.4%)	11 (64.7%)	0.70

Abbreviations: CYC: Cyclophosphamide; RTX: Rituximab; PR3-ANCA: Proteinase 3-Antineutrophil Cytoplasmic Antibodies; MPO-ANCA: Myeloperoxidase-Antineutrophil Cytoplasmic Antibodies; BVAS: Birmingham Vasculitis Activity Score; IQR: Interquartile Range; Chi-Square, Fisher exact.

During the remission induction phase, RTX was administered at a dose of 1,000 mg on days 1 and 15. CYC was given at 15 mg/kg every two weeks or once a month, depending on renal clearance. We observed that compliance with CYC induction therapy was high, with rates of 93% in GPA, 85% in EGPA, and 75% in MPA. RTX induction therapy compliance was 100% across all AAV subtypes.

The minimum recommended starting dose of oral glucocorticoids was 24 mg/day, with a maximum dose of 100 mg/day. For pulse steroid therapy, the minimum total dose per cycle was 500 mg, while the maximum reached 3,000 mg. Patients were recommended to continue steroid treatment for at least 3 months, following a tapering schedule based on clinical response. Despite recommended dosing guidelines, there was significant variability in the doses of maintenance therapies used. Following remission induction, we noted that some patients did not proceed with any maintenance therapy. Despite recommended dosing, there was significant variability in the doses of maintenance therapies used. Azathioprine was administered at doses ranging from 50 mg to 200 mg daily. Methotrexate doses varied between 10 mg/week and 15 mg/week, while MMF ranged from 1,000 mg/day to 2,000 mg/day. During the remission-maintenance phase, adherence was lower for oral therapies, with rates of 66% for MTX, 36% for MMF, and 28% for AZA, while RTX adherence remained at 100%.

Discussion

This retrospective study provides real-world insights into the factors influencing physician selection of induction therapies in AAV and highlights the issue of lower adherence to long-term oral medications. In remission-induction treatment, RTX was selected as the first-line therapy in 15% of patients (n=17/112). Patients receiving CYC as first-line therapy were significantly older, had more comorbidity, and higher BVAS scores than those treated with RTX. Major organ involvement, like renal or pulmonary, did not significantly differ between RTX and CYC groups. However, a review of patient records indicated that RTX was preferentially chosen when concerns existed regarding CYC's potential for gonadal toxicity(16), or in patients with pre-existing cytopenia or chronic liver disease. Compliance with RTX therapy was 100% across all AAV subtypes in both induction and maintenance phases, whereas adherence to oral maintenance therapies was considerably lower, with rates of 66% for MTX, 36% for MMF, and 28% for AZA. The primary goal of AAV treatment is to achieve remission and prevent relapse while minimizing long-term drug toxicities and associated comorbidities (3). When selecting the most appropriate remission induction therapy, factors such as disease severity, comorbidities, drug contraindications, and potential toxicities should guide decision-making. Following evidence from the RAVE and RITUXVAS studies, RTX may be preferred over CYC in specific populations, including patients aiming to preserve fertility, frail older adults, children, adolescents, particularly in relapsed or PR3-ANCA-positive disease (1, 7, 8, 17, 18). In a study, RTX was found to be more effective than CYC for inducing remission in relapsing disease. Also, RTX was found equally effective in treating severe manifestations, such as major renal disease or alveolar hemorrhage with CYC. Adverse event rates were similar between the two groups, yet the optimal choice remains debated, particularly in rapidly progressive kidney disease. Although RTX's efficacy in renal failure has not been directly tested in clinical trials, existing evidence suggests equivalence between RTX and CYC in severe kidney involvement (17, 19-21). Our data support the selective use of RTX in practice. While rates of specific organ involvement (e.g., renal or pulmonary) did not significantly differ between the CYC and RTX groups, patients receiving CYC had significantly higher BVAS scores. This finding suggests that, while the presence or absence of involvement in a single organ system may not have been the primary driver of treatment choice, the overall burden of disease and the extent of multi-system involvement, as reflected by the higher BVAS, might be associated with a preference for CYC. This observation may align with hypothesis that clinicians in a real-world setting may be more inclined to select CYC for patients with a greater overall disease burden, reflecting a more aggressive approach in cases with more extensive vasculitis. To determine whether this pattern is replicated across diverse healthcare settings with varying patient populations and established treatment paradigms, further multi-center studies are essential. The preference for RTX in younger patients avoids the well-documented risk of CYC-induced gonadal toxicity, including early menopause and primary ovarian failure(16). The overall compliance with

remission-induction therapies in our study was high. RTX demonstrated the highest compliance in both induction and maintenance phases, with 100%, likely due to its less frequent dosing schedule and favorable side effect profile. For remission-maintenance treatments, adherence rates were significantly lower, with 66% for MTX, 36% for MMF, and 28% for AZA. Our findings highlight a critical gap in AAV management, emphasizing the need to improve long-term adherence to maintenance therapy. Currently, data on adherence in AAV are limited, with most insights derived from studies in other chronic autoimmune diseases, such as systemic lupus erythematosus (SLE). Non-compliance in SLE ranges from 43% to 75%, with key contributing factors including depression, low education levels, polypharmacy, and rural residence (22) . Similar barriers may impact adherence in AAV, as patients may reduce or discontinue medications when symptoms improve, or side effects become intolerable. Additionally, physician hesitation to discontinue maintenance therapy due to relapse concerns may contribute to prolonged treatment durations, although further research is needed to confirm this. Polypharmacy, often linked to multiple comorbidities, is a well-established risk factor for medication non-adherence in chronic conditions. Patients with AAV, especially those managing numerous medications for various comorbidities, may struggle with complex treatment regimens, leading to unintentional non-adherence. In our study, the higher prevalence of comorbidities, and the resulting polypharmacy, may have contributed to lower adherence rates to oral maintenance therapies. Given that relapses significantly increase morbidity and mortality in AAV, improving adherence to maintenance therapy is critical. Relapse events not only contribute to progressive organ damage and increased mortality but also necessitate high-dose corticosteroid use, further exacerbating treatment-related complications. Therefore, ensuring high compliance with maintenance medications is essential to minimize these risks and promote better long-term outcomes. RTX stands out in this regard. One of the primary limitations of this study is its observational design, which restricts the ability to establish causal relationships between medication adherence and clinical outcomes. Another significant limitation of our study is its single-center design. While this inherently restricts the generalizability of our findings to other institutions, it simultaneously reflects the real-world clinical context we sought to investigate. Furthermore, reliance on patient self-reports and medical records for adherence data may introduce bias or inaccuracies. While more direct measures of adherence, such as pill counts or electronic monitoring, could provide more accurate assessments, these were not feasible within the retrospective design of our study. Additionally, socioeconomic factors, which could significantly influence treatment choices and adherence, were not accounted for in this study. Nonetheless, future prospective studies should incorporate direct assessment of patient-reported barriers to adherence, including factors such as experienced side effects, access to medications, financial constraints, health literacy, and the patient's understanding of the need for long-term therapy.

We also acknowledge the heterogeneity within ANCA-associated vasculitis and the clinical and genetic distinctions between GPA/MPA and EGPA. Combining these subtypes in some descriptive analyses could potentially obscure subtype-specific differences. Future studies with larger cohorts should aim to analyze these subtypes independently, particularly when evaluating factors influencing treatment decisions. In this real-world cohort of AAV patients, older age, higher BVAS scores, and greater comorbidity burden—rather than specific organ involvement—were associated with the selection of CYC over RTX as first-line induction therapy. As expected, patients with specific clinical conditions, such as malignancy, cytopenia, or chronic liver disease, were preferentially treated with RTX. Despite high compliance with RTX, adherence to oral maintenance therapies remains a major challenge, underscoring the need for interventions to improve long-term adherence, particularly for oral agents. A T2T approach tailored to AAV may optimize treatment selection, minimize premature switches, and enhance maintenance therapy adherence. Further prospective studies are needed to validate these findings and develop personalized adherence strategies for improved long-term disease control.

Conflict of Interest: Each author declares that he or she has no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article. The authors declare no competing financial interests in relation to the work described.

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