Table 1. Overview of studies on treatment for ENT-involvement.

ENT-involvement	AAV type per study	Therapy	Results	LoE
Otitis Media / Hearing loss	OMAAV (n=6)	RTX	Remission 100% Mean hearing gain AC 22 dB	2b
	OMAAV (n=104)	Steroids	Response (hearing) 56% BC 11 dB	4
	OMAAV (n=122)	Steroids + csDMARD	Relapse 47%	4
	OMAAV (n=8)		Relapse 36%	4
	GPA (n=11)		Response (hearing) 68% Relapse 0%	4
Vestibular symptoms Subglottic stenosis	OMAAV (n=3)	Steroids + CYC	Response (hearing) 81% Response (hearing) 100%	4
	GPA (n=9)	Dilatation	Remission (patient-reported) 100%	4
	GPA (n=15)		Relapse 78%	4
	GPA (n=34)	Intralesional steroids + dilatation	Mean number of procedures 4	2b
	GPA (n=8)		Median response 34 months	4
			Response 88% Median response 12 months	4
			Mean number of procedures 3	2b
	GPA (n=18)	Intralesional steroids + dilatation	Mean intervention-free interval 26 months	2b
	GPA (n=5)	+ resection	Response 80%	4
	GPA (n=13)	resection Surgery	Response (dyspnea) 100%	4
	GPA (n=11)		Relapse 39%	4
	GPA (n=6)		Higher QoL 77%	4
Sinonasal symptoms	GPA (n=6)	Surgery or dilatation + intralesional steroids or csDMARDs	Response (airway) 91% Relapse 83.3%	4
	GPA (n=8)	RTX	Remission 38%	2b
	GPA (n=3)	RTX	Response 50% Response: 67%	2b
	EGPA (n=29)	Steroids + csDMARDs	Remission 82%	2b
	EGPA (n=44)		Remission 21%	2b
ENT symptoms NOS	GPA (n=19)	Surgery	Response 32%	2b
	EGPA (n=65)	Mepolizumab	Response 76%	4
	AAV (n=7)	RTX	Remission 28%	1b
	GPA (n=11)		Relapse 22%	2b
	EGPA (n=61)		Drop in daily steroid dosage	4
			Response 93%	2b
	GPA (n=28)	Steroids + csDMARDs	Relapse 17%	4
	EGPA (n=15)		Remission 96%	4
	GPA (n=51)	RTX (n=51) MTX	Relapse 42%	4
			Remission 86%	4
			92% of observational period in remission	2b
			54%	

AC: air-conduction, BC: bone-conduction, csDMARD: conventional synthetic disease modifying antirheumatic drug, CYC: cyclophosphamide, dB: decibel, EGPA: eosinophilic GPA, GPA: granulomatosis with polyangiitis, MTX: methotrexate, NOS: not otherwise specified, OMAAV: otitis media associated ANCA-vasculitis, QoL: quality of life, RTX: rituximab.

Conclusion: On the treatment of ENT manifestations in AAV 25 studies were available, mainly with small patient numbers and low LoE (1b-4). Heterogeneity in treatments and outcome measures impaired comparison between studies. More placebo-controlled studies are needed to guide treatment.

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AB0388

USE OF MESNA PROPHYLAXIS IN CYCLOPHOSPHAMIDE-TREATED PATIENTS WITH ANCA-ASSOCIATED VASCULITIS: RESULTS OF AN ELECTRONIC SURVEY

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Background: Co-prescribing 2-mercaptoethane sodium sulfonate (mesna) with cyclophosphamide (CYC) for ANCA-associated vasculitis (AAV) aims to prevent

the potential urotoxic effects of CYC. The evidence for this practice is often considered weak, and there may be some diversity in what practitioners do in clinical practice.

Objectives: To investigate current clinical practice related to prescribing mesna prophylaxis or not and the underlying rationale for CYC-treated patients with AAV. **Methods:** We searched MEDLINE for publications with the MeSH term "ANCA-associated vasculitis" over a 10-year period up to October 2020. Email addresses of authors of these publications were extracted from the online information available in MEDLINE. These authors were invited by email to participate in an anonymous online SurveyMonkey survey of 21 to 24 questions asking about the characteristics of the respondent, their experience with AAV, and their practice in using CYC to treat AAV and using mesna in CYC-treated patients with AAV and the underlying rationale. Respondents were eligible to take the full survey if they were involved in deciding and/or monitoring therapy with CYC for patients with AAV. We compared 15 response variables to identify factors associated with the use or not of mesna. Response variables with multiple categories were first analyzed across all categories; if the omnibus test result was significant, additional analyses were used to identify the categories, which were the sources of group separation. We analyzed by-country variations for only countries with ≥ 10 respondents. Statistical analyses involved Pearson's chi-square test or Fisher's exact test, as appropriate. For multiple-response variables, the Rao-Scott correction was applied.

Results: The invitation for the electronic survey was emailed twice in October 2020 to 1,374 unique email addresses; 156 individuals responded; 139 were eligible and completed the survey. The 139 participants were from 34 countries and were essentially MDs (98%) who mainly worked in rheumatology (50%), nephrology (25%) or internal medicine/clinical immunology (18%). Mesna was given in conjunction with CYC systematically, never, or on a case-by-case basis by 68%, 19% and 13% of respondents, respectively. As compared with systematic mesna-prescribers, never/occasional mesna-prescribers reported a longer time since receiving their degree as a health professional (≥ 15 years: 80% vs 50%, $P<0.001$), were more frequently based in England/United States (than in France/Germany/Italy) (78% vs 21%, $P<0.001$), had longer involvement in care of patients with AAV (≥ 15 years: 62% vs 37%, $P=0.006$), had less practice in using intermittent pulse therapy as the exclusive/predominant CYC administration scheme (62% vs 89%, $P<0.001$), and, as a rationale underpinning their mesna practice, had less adherence to local operational procedures (47% vs 73%, $P=0.002$) or (inter)national management guidelines for AAV (16% vs 49%, $P<0.001$). Never/occasional versus systematic use of mesna did not differ across medical specialties (5 categories, $P=0.192$) or healthcare settings (3 categories, $P=0.437$), and was not associated with prior experience of CYC-related urotoxic events (3 categories, $P=0.495$) or severe mesna toxicity issues (3 categories, $P=0.957$). The confidence that their practice reflected the best possible patient care did not differ between never/occasional and systematic mesna-prescribers (7-point Likert scale, $P=0.794$).

Conclusion: Practice with regard to prescribing mesna in conjunction with CYC to treat AAV is heterogeneous, although systematic mesna use prevailed over never or occasional use. The decision to prescribe or not mesna may be based more on circumstantial than structural reasons.

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AB0389

A COMBINED RHEUMATOLOGY-OPHTHALMOLOGY CLINIC IMPROVED DIAGNOSIS AND TREATMENT OF IMMUNE-MEDIATED AND INFLAMMATORY DISEASES AMONG PATIENTS PRESENTING WITH INFLAMMATORY EYE DISEASES

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Background: Patients presenting with an inflammatory eye disease may have underlying immune-mediated and inflammatory diseases (IMIDs). However, IMIDs are challenging to diagnose. A combined rheumatology-ophthalmology evaluation could enhance detection of IMIDs in this group.

Objectives: To investigate whether the implementation of a combined rheumatology-ophthalmology clinic could increase the sensibility of IMIDs diagnosis.

Methods: A combined rheumatology-ophthalmology clinic started in January 2020. A rheumatologist with expertise in IMIDs assessed outpatients of a tertiary care ophthalmologic clinic specialised in inflammatory eye disease consecutively referred between 01/01/20 and 31/12/20. Two separate visits confirmed

both ophthalmology and rheumatology diagnoses. The same period of the year 2019 was used as a comparator.

Results: We noticed a three-fold increase in the number of ophthalmology patients who underwent an evaluation with a rheumatologist in 2020 compared to 2019 (41 vs 13, +315%; Table). There were no differences in sex distribution or age between the two years. However, the median time before rheumatology assessment was shorter in 2020 than 2019 (median days 0 [range 0-6] vs 12 [range 0-91]). IMIDs diagnoses increased by 27% in 2020 compared to 2019 (29/41 vs 7/13, 71% vs 54%), mainly driven by new diagnoses of systemic vasculitis and Behcet's disease (Figure 1). There was also an increase in diagnoses of non-IMIDs systemic conditions (3 cancers and one infection in 2020 vs none in 2019). After rheumatology evaluation, immune-modulating therapy was initiated in 18/41 (44%) vs 3/13 (23%).

IMIDs diagnoses after rheumatology assessment

Ophthalmology assessment and referrals to rheumatology	Referrals 2019 (n=13)	Referrals 2020 (n=41)
Inflammatory eye disease, n (%)	6 (46)	36 (88)
Uveitis, n	6	21
Vasculitis, n	2	4
AIION, n	3	5
Conjunctivitis, n	1	0
Scleritis, episcleritis, n	0	1
Other, n	1	5

Conclusion: An integrated approach with rheumatologist appeared to remarkably increase the likelihood to detect IMIDs among patients presenting with inflammatory eye diseases.

Disclosure of Interests: None declared

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AB0390

CHARACTERISTICS AND PROGNOSIS OF AGE-DEPENDENT ANCA-ASSOCIATED VASCULITIS IN JAPAN

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Background: It is known that most of ANCA-associated vasculitis (AAV) patients are elderly. A cohort study showed that the mean age of onset was 71.1 years, especially in patients with microscopic polyangiitis (MPA)¹. However, the characteristics and prognosis of age-dependent AAV patients are still unclear.

Objectives: To clarify the differences in age-related characteristics and prognosis between Japanese patients with AAV.

Methods: We enrolled 44 patients with AAV who underwent remission induction therapy at our hospital from January 2016 to December 2020. They were divided with under 70 years old group (<70 yo group, n=12) and over 70 years old group (≥70 yo group, n=32). We investigated between two groups as follows; clinical characteristics and laboratory data at diagnosis, rates of complete remission (CR) at 6 months, defined as Birmingham Vasculitis Activity

Score (BVAS)=0 and prednisone ≤7.5 mg/day, adverse events, and relapse free survival.

Results: Mean age were 61.9 ± 9.74 years old in <70 yo group and 77.5 ± 4.61 years old in ≥70 yo group (p<0.01). There were significantly fewer newly diagnosed cases in the <70 yo group, and half of the patients with relapsing disease. There was no difference in the type of ANCA, organ involvement, or BVAS. Remission induction therapy was performed with CY or RTX, and no difference was observed between these two groups. In addition, there was also no difference in maintenance therapy (Table 1). CR rate in <70 yo group and in ≥70 yo group were 55% and 46 % respectively (p=0.73). Severe infections occurred with no patient (0%) in <70 yo group and with 5 patients (16%) in ≥70 yo group (p=0.30). 5 patients of relapse were observed in the <70 yo group and 1 patient in the ≥70 yo group, and relapse free survival was significantly lower in the <70 yo group (p=0.001) (Figure 1).

Table 1. Clinical characteristics at baseline and treatment drugs between <70 yo group and ≥70 yo group.

	<70 yo 群 (n=12)	≥70 yo 群 (n=32)	p value
Age (year)	61.9 ± 9.74	77.5 ± 4.61	< 0.01*
Female, n (%)	10 (83)	28 (87)	0.66
AAV type	6 (50) / 6 (50)	26 (81) / 6 (19)	0.06
MPA, n (%) / GPA, n (%)			
Newly diagnosed, n (%)	6 (50)	27 (84)	0.045*
ANCA positivity			
MPO, n (%) / PR3, n (%)	11 (92) / 0 (0)	30 (94) / 3 (9)	1 / 0.55
negative, n (%)	1 (8)	1 (3)	0.48
CRP (mg/dl)	3.34 ± 4.01	8.15 ± 6.86	0.03*
eGFR (mL/min)	55.8 ± 25.3	57.7 ± 24.7	0.93
BVAS	12.0 ± 8.0	14.8 ± 6.8	0.23
Remission induction therapy	5 (42) / 7 (58)	16 (50) / 16 (50)	0.74
CY, n (%) / RTX, n (%)			
Maintenance therapy			
AZA, n (%)	7 (58)	14 (44)	0.50
RTX, n (%)	1 (8)	6 (19)	0.65
others, n (%)	2 (17)	3 (9)	0.60
without IS, n (%)	2 (17)	9 (28)	0.70

The p-value was estimated using Fisher's exact or Wilcoxon rank sum test. yo, years old; AAV, ANCA-associated vasculitis; MPA, microscopic polyangiitis; GPA, granulomatosis with polyangiitis; CY, cyclophosphamide; RTX, rituximab; AZA, azathioprine; IS, immunosuppressants. *p<0.05

Conclusion: There was no difference in remission rate between two groups. Severe infections were observed only in the ≥70 yo group. We suggest that younger AAV patients need attention to relapse after the remission because of lower relapse free survival in <70 yo group.

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Figure 1. Relapse free survival Relapse free survival was calculated by Kaplan-Meier method and compared by log-rank test. *p<0.05

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