Objectives: With sparse data on prognostic factors in IgAV, we investigated whether pre-existing conditions are risk factors for mortality in adult IgAV patients. **Methods:** Observational population-based cohort study using state-wide linked longitudinal health data for adults with IgAV (n=267) and matched controls (n=1080) between 1980-2015. Charlson comorbidity index (CCI) and serious infections (SI) were recorded over an extensive lookback period prior to diagnosis. Date and causes of death were extracted from the WA Death Registry. Mortality rate (deaths/1000 person-years) ratios (MRR) and time dependent survival analysis assessed the risk of death. Age and gender specific mortality rate data were obtained from the Australian Bureau of Statistics.

Results: During 9.9 (± 9.8) years lookback IgAV patients accrued higher CCI scores (2.60 vs 1.50 $p < 0.001$) and had higher risk of SI (OR 8.4, $p < 0.001$), not fully explained by CCI scores. During 19 years follow-up, the risk of death in IgAV patients (n=137) was higher than in controls (n=397) (MRR 2.06, CI 1.70-2.50, $p < 0.01$) and the general population (SMRR 5.64, CI 4.25, 7.53, $p < 0.001$). Survival in IgAV was reduced at five (72.7 vs. 89.7 %) and twenty years (45.2% vs. 65.6 %) (both $p < 0.05$). CCI (HR 1.88, CI: 1.25 - 2.73, $p = 0.001$), renal failure (HR 1.48, CI: 1.04 - 2.22, $p = 0.03$) and prior SI (HR 1.48, CI: 1.01 - 2.16, $p = 0.04$) were independent risk factors. Death from infections (5.8 vs 1.8%, $p = 0.02$) was significantly more frequent in IgAV patients.

Conclusion: Premorbid accrual of comorbidity is increased and predicts premature death in IgAV patients. However, comorbidity does not fully explain the increased risk of serious infections prior to diagnosis or the increased mortality due to infections in IgAV.

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AB0394 CLINICAL CHARACTERISTICS OF EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS: A SINGLE-CENTER RETROSPECTIVE ANALYSIS ON 52 CASES OF CHINESE PATIENTS

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Background: Eosinophilic granulomatosis with polyangiitis (EGPA) is a rare and heterogeneous systemic vasculitis. Different patients or the same patient in different stages show different manifestations, which may lead to misdiagnosis and delay treatment.

Objectives: To analyze the clinical characteristics in Chinese patients with EGPA.

Methods: EGPA patients who fulfilled the 1990 ACR classification criteria were included between December 2003 and April 2020. The demographic and clinical characteristics were collected and analyzed retrospectively.

Results: There were 52 EGPA patients recruited, 34 (65.4%) patients were males and the median age at onset was 47(38-55) years. The median duration from disease onset to diagnosis was 30(4-96) months. For initial symptoms, respiratory manifestations (61.5%) were the most common, including 42.3% patients beginning with asthma, followed by 21.2% with nose/paranasal sinuses manifestations. Respiratory medicine (53.8%) were the most common department at first visit, followed by rheumatology medicine (11.5%, Figure 1A). There were 44.2% EGPA patients definitely diagnosed at the department of rheumatology or after consultation by rheumatologists.

During the whole disease process, the most common clinical manifestations were asthma (88.5%), then nose/ paranasal sinuses (84.6%), pulmonary (76.9%) and nervous system (61.5%) manifestations, followed by constitutional symptom (44.2%), heart (36.5%) and skin (23.1%) involvement. Only 9.6% patients had gastrointestinal tract involvement and 3.8% had renal involvement (Figure 1B).

There were 46 (88.5%) patients showing the ratio of peripheral blood eosinophils $> 10\%$ at diagnosis. Among the rest 6 patients, 3 had higher eosinophil ratio before diagnosis, while the other 3 patients had been treated with glucocorticoid before diagnosis, of whom 2 patients showed pathological eosinophil infiltration in lung or paranasal sinuses mucosa, respectively. There were 8(15.4%) patients with positive ANCA. Compared with EGPA patients with negative ANCA, they had lower incidence of asthma (62.5% vs. 93.2%), but higher incidence of constitutional symptoms (87.5% vs. 36.4%), arthralgia (50.0% vs. 6.8%) and renal involvement (25.0% vs. 0.0%), higher peripheral eosinophil count [$2.06(0.80-4.51) \times 10^9/L$ vs. $1.81(0.93-3.32) \times 10^9/L$], ESR [$20(7-77)$ mm/h vs.

$18(9-42)$ mm/h] and CRP [$18.5(3.2-65.9)$ mg/L vs. $3.3(3.2-13.0)$ mg/L], higher Birmingham vasculitis activity score [$17(10-22)$ vs. $13(9-15)$] and vasculitis damage index [$3(1-3)$ vs. $2(1-2)$, all $P < 0.05$]. There were 21.2% EGPA patients showing poor prognostic factors according to five-factor score (FFS) and 34.6% patients according to the revised FFS.

Conclusion: EGPA patients may have no asthma especially those with positive ANCA. Multi-disciplinary collaboration especially based on rheumatologists and pulmonologists should be emphasized for early identification and prompt treatment.

Figure 1. Department at first visit (A) and clinical manifestations during the whole disease process of 52 Chinese EGPA patients (B).

Disclosure of Interests: None declared

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AB0395 RENAL INVOLVEMENT IN HENOCH-SCHÖNLEIN PURPURA

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Background: Henoch-Schönlein purpura (HSP), also known as rheumatic purpura, is an immune complex vasculitis affecting small vessels with dominant IgA deposits. Clinical manifestations mainly involve cutaneous purpura, arthralgias and/or arthritis, acute enteritis and glomerulonephritis. HSP is more common among children than adults. Kidney damage is the principal prognostic determinant in HSP. The aim of this study is to evaluate the renal manifestations of HSP.

Objectives: The aim of this study is to evaluate the renal manifestations of HSP.

Methods: This was a retrospective study of 35 patients with diagnosed HSA, who were admitted between the time period may 2008 and november 2018. All of these patients met the American College of Rheumatology classification criteria for a diagnosis of HSP. Renal involvement was defined as the presence of proteinuria or hematuria.

Results: The study group consisted of 35 patients, 20 women (57%) and 15 men (43%). The mean age was 48.8 ± 18.7 years. The incidence rates of skin, joint, gastrointestinal and central nervous system involvement were 91.4, 77.1, 54.2 and 2.8%, respectively. Elevated serum IgA levels occurred in 5.7 %. Six patients (17.1%) were given corticosteroids during the first 3 months of their HSP, which were indicated for severe gastrointestinal (n=5) or renal involvement (n=1). Eighteen of the thirty five patients (51.42%) had renal involvement. There were 12 women and six men. All patients had proteinuria (100%). Hematuria has been identified in 11 patients (61.1%) and renal insufficiency in 4 patients (22.2%). The renal involvement was mainly detected in the first month. Renal biopsy was affected in four patients (22.2%). An endocapillary glomerulonephritis was found in 3 patients and an extra capillary necrotizing glomerulonephritis in one patient. The immunofluorescence showed the presence of IgA deposits in 4 patients.

Conclusion: Renal involvement dominates the prognosis for the disease. The most common clinical expression is the association of microscopic hematuria with proteinuria.

Disclosure of Interests: None declared

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AB0396 RAPID IMPROVEMENT IN CYSTOID MACULAR EDEMA WITH HIGH DOSE INTRAVENOUS METHYLPREDNISOLONE IN NON-INFECTIOUS UVEITIS OF DIFFERENT IMMUNE MEDIATED INFLAMMATORY DISEASES

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Background: Cystoid Macular Edema (CME) is the most important cause of blindness in non-infectious uveitis (NIU) (1). Corticosteroids and conventional and/or biological immunosuppressant may be required (1-6). High-dose intravenous methylprednisolone (IVMP) pulse therapy may induce a rapid improvement.

Objectives: To evaluate the efficacy and safety of IVMP pulse therapy in CME of different immune mediated inflammatory diseases (IMID).

Methods: Multicentre study of 66 patients with severe ocular inflammation who received IVMP. The underlying diseases were: Vogt Koyanagi Harada disease (VKHD)(n=24), Behçet's disease (BD) (19) and idiopathic NIU (23). The main outcome variable was macular thickness and macular edema (Optical coherence tomography [OCT] >300 µm); that was assessed at 0 (basal), 2-5, 7, 15 and 30 days after IVMP.

The results are expressed as mean ±SD for normally distributed variables, or as median [IQR] when are not. Comparison of continuous variables was performed using the Wilcoxon test.

Results: We studied 43♀/ 23♂ patients. The main features are shown in TABLE 1. IVMP dose ranged from 250 to 1000 mg/day administered for 3-5 consecutive days, the dose was established according to the presence or not of other systemic manifestations apart from uveitis. All of them had active intraocular inflammation at the moment of the study.

Table 1. Main features of 66 patients with cystoid macular edema.

	VKHD (n=24)	Idiopathic (n=23)	Behçet's disease (n=19)	Overall (n=66)
Men/Women, n	5/19	9/14	9/10	66
Mean age (years)	42 ± 11	47 ± 15	33 ± 10	-
Unilateral/Bilateral, n (%)	2 (8.3) /22 (91.7)	10 (43.5) / 13 (56.5)	4 (21) / 15 (79)	16/50
Inflammatory ocular patterns, n (%)				
Posterior uveitis	6 (25)	9 (39.1)	3 (15.8)	18
Panuveitis	18 (75)	14 (60.9)	16 (84.2)	48
Laboratory data, n (%)				
ANA	2 (8.34)	2 (8.7)	0 (0)	4
HLA B27	0 (0)	4 (17.4)	0 (0)	4
HLA B29	0 (0)	1 (4.3)	0 (0)	1
HLA B51	0 (0)	5 (21.7)	8 (42)	13

A rapid and maintained statistically improvement was observed in OCT values in all underlying diseases (FIGURE 1). No major side effects were observed.

Conclusion: High-dose IVMP pulse therapy is useful and safe in the prompt control of CME, regardless of the underlying IMID.

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Figure 1. Improvement of OCT in 66 patients with cystoid macular edema.

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AB0397 EPIDEMIOLOGY AND CLINICAL FEATURES OF OPHTHALMOLOGICAL MANIFESTATIONS IN BEHÇET'S DISEASE. STUDY OF 50 PATIENTS OF A SERIES OF 120 PATIENTS IN A REGION IN NORTHERN SPAIN

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Background: Ophthalmological involvement is one of the most feared manifestations of Behçet's disease [1-6].

Objectives: Our aim was to define the main demographic and clinical features of ophthalmological involvement in a well-defined cohort of patients with Behçet's disease.

Methods: Descriptive study of a cohort of 120 patients diagnosed with Behçet's disease from January 1, 1980 to December 31, 2019. Finally, following the 2014 International Criteria for Behçet's Disease (ICBD) (J Eur Acad Dermatol Venereol 2014; 28:338-47) 94 patients were chosen for this study.

Results: 50 patients (28 men/22 women; male to female ratio of 1:0.8) had ocular involvement. Mean age at diagnosis was 37.6±11.8 years. Mean ICBD score was 5.8±1.3 points. Neurological involvement was the most frequent manifestation in this group. Genital ulcers were more frequent in the non-ophthalmological involvement group. Systemic clinical domains are shown in Figure 1.

The most frequent ocular manifestations were uveitis (n=44, 88.0%), retinal vasculitis (n=5, 10%) and dry eye (n=5, 10%). Likewise, the most frequent type of uveitis was anterior (n=17, 38.6%), followed by posterior uveitis (n=14, 31.8%). 26 (59.1%) of all uveitis were unilateral. Panuveitis was more frequent among patients under 60 years. Similarly, anterior uveitis was predominant in patients older than 70 years. There were no remarkable differences between genders. Main clinical features are shown in Table 1.

Table 1. Clinical characteristics of ocular Behçet's disease.

	n (%)	Unilateral, n (%)	Bilateral, n (%)
Uveitis	44 (88.0)	26 (59.1)	18 (40.9)
Anterior	17 (38.6)	13 (76.5)	4 (23.5)
Intermediate	2 (4.5)	-	2 (100)
Posterior	14 (31.8)	9 (64.3)	5 (35.7)
Panuveitis	11 (25.0)	4 (36.4)	7 (73.6)
Retinal vasculitis	5 (10.0)	-	-
Dry eye	5 (10.0)	-	-
Cystoid macular edema	4 (8.0)	-	-
Episcleritis	3 (6.0)	3 (100)	-
Optic neuritis	2 (4.0)	-	-

Conclusion: Ophthalmological involvement in Behçet's disease was more frequent in men. Uveitis and retinal vasculitis were the most frequent ocular manifestation. No remarkable differences in clinical features were observed between genders.