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±8.8) 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<0.001) and the general population (SMR 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 prematurity of 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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Disclosure of Interests: None declared

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

Disclosure of Interests: None declared

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AB0397

EPIGENOMICS 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 1.08) 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.

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

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

Clinical characterisitics of ocular Behçet’s disease.

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

<table>
<thead>
<tr>
<th>n (%)</th>
<th>Unilateral, n (%)</th>
<th>Bilateral, n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Uveitis</td>
<td>44 (88.0)</td>
<td>26 (59.1)</td>
</tr>
<tr>
<td>Anterior</td>
<td>17 (38.6)</td>
<td>13 (30.5)</td>
</tr>
<tr>
<td>Intermediate</td>
<td>2 (4.5)</td>
<td>-</td>
</tr>
<tr>
<td>Posterior</td>
<td>14 (31.8)</td>
<td>9 (43.5)</td>
</tr>
<tr>
<td>Panuveitis</td>
<td>11 (25.0)</td>
<td>4 (36.4)</td>
</tr>
<tr>
<td>Retinal vasculitis</td>
<td>5 (10.0)</td>
<td>-</td>
</tr>
<tr>
<td>Dry eye</td>
<td>5 (10.0)</td>
<td>-</td>
</tr>
<tr>
<td>Cystoid macular edema</td>
<td>6 (13.0)</td>
<td>2 (69.2)</td>
</tr>
<tr>
<td>Episcleritis</td>
<td>1 (2.4)</td>
<td>1 (2.4)</td>
</tr>
<tr>
<td>Optic neuritis</td>
<td>3 (6.0)</td>
<td>3 (6.0)</td>
</tr>
</tbody>
</table>

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.