### POS1346
**PREVALENCE, PHENOTYPICAL CLINICAL CLUSTERS AND TREATMENT OF NEUROBEHÇET’S DISEASE. STUDY IN NORTHERN SPAIN**

A. Herrero-Morant1, C. Álvarez-Reguera1, L. Sanchez-Bilbao1, D. Martinez-López1, J. L. Martin-Varillas2, G. Suárez-Amorín1, R. Fernández Ramón1, M. C. Mata Arnaiz2, M. Á. González-Gay1, R. Blanco1, J. Marqués de Valdecilla University Hospital, Rheumatology and Ophthalmology, Santander, Spain; 2Hospital Comarcal de Laredo, Rheumatology, Laredo, Spain

**Background:** Behçet’s disease (BD) may present with different clinical phenotypes. Ocular and NeuroBehçet’s Disease (NBD) are severe complications [1-4]. Data on NBD epidemiology, clinical phenotype and therapy are scarce and controversial.

**Objectives:** In a wide unselected single-center series of BD our aims were to assess a) NBD prevalence, b) associations with other clinical clusters and c) treatment.

**Methods:** Cross-sectional study of all 120 patients diagnosed with BD in Northern Spain, between January 1, 1999 to December 31, 2019. Finally, 96 patients were included in this study according to 2014 International Criteria for Behçet Disease (ICBD) [5]. NBD was diagnosed according to the International Consensus Recommendation (ICR) criteria [4].

**Results:** NBD was diagnosed in 23 of 96 (24%) patients (15 women/8 men) (mean age: 44±13.9 years). NBD was classified as parenchymatous (n=10, 43.5%), non-parenchymatous (n=10, 43.5%) and mixed (n=3, 13%). HLAB1 was positive in 5 out of 13 (38.4%) patients tested. The main cluster of clinical associations were oral aphthae (n=20, 87%), ocular (n=14, 60.9%), cutaneous (n=10, 43.5%), articular (n=8, 39.1%), vascular (n=4, 17.4%) and intestinal (n=1, 8.7%) involvement (Figure 1).

**Disclosure of Interests:** Alba Herrero-Morant: None declared, Carmen Álvarez-Reguera: None declared, Lara Sanchez-Bilbao: None declared, David Martinez-Lopez: None declared, José Luis Martin-Varillas Grant/research support from: Abbvie, Pfizer, Lilly, Janssen, and Roche, Ricardo Blanco Speakers bureau: Abbvie, Lilly, Pfizer, Roche, BMS, Janssen, and MSD, Grant/research support from: Abbvie, Lilly, Pfizer, Roche, BMS, Janssen, and MSD, Ricardo Blanco Speakers bureau: Abbvie, Lilly, Pfizer, Roche, BMS, Janssen, and MSD, Grant/research support from: Abbvie, Lilly, Pfizer, Roche, BMS, Janssen, and MSD, Janssen, and MSD, Roche

**DOI:** 10.1136/annrheumdis-2022-eular.2671

---

### POS1347
**BIOLOGICAL THERAPY IN NEUROSARCOIDOSIS. STUDY OF 30 PATIENTS FROM A SERIES OF 234 SYSTEMIC SARCOIDOSIS FROM A UNIVERSITY HOSPITAL**

A. Herrero-Morant1, D. Martinez-Lopez1, L. Sanchez-Bilbao1, I. Gonzalez-Mazon1, J. L. Martin-Varillas2, R. Fernández Ramón1, C. Alvarez-Reguera1, M. A. Gonzalez-Gay1, R. Blanco1, J. Marqués de Valdecilla University Hospital, Rheumatology and Ophthalmology, Santander, Spain; 2Hospital Comarcal de Laredo, Rheumatology, Laredo, Spain

**Background:** Neurosarcoidosis (NS) is a severe complication of sarcoidosis [1,2]. NS may be classified according to several subtypes [1]. Data on therapy, including biological therapy (BT) is scarce.

**Objectives:** To assess efficacy and safety of BT in refractory NS subtypes.

**Methods:** Study of NS from a large cohort (n=234) of all consecutive patients diagnosed with sarcoidosis in a single university hospital from January 1, 1999 to December 31, 2019. Diagnosis of sarcoidosis was established according to ATS/ERS/WASOG criteria [3].

**Efficacy was considered as complete or partial response and no-response according to the resolution of the neurological syndrome (signs and/or symptoms) after the BT onset.**

**Results:** NS was observed in 30 (19 women/11 men) of 234 (12.8%) patients; mean age, 55.0±15.8 years. NS subtypes were chronic headache (n=13, 43.4%), peripheral neuropathy (n=6, 20%), cranial neuropathy (n=5, 16.7%), spinal cord abnormalities (n=3, 10%) and aseptic meningitis (n=3, 10%). A total of 26 (86.7%) patients received oral corticosteroids (CT) (mean maximum dose: 50±19.2 mg/dL) and 7 (23.3%) IV CT. In addition, conventional immunosuppressants were administered to 18 (60%) patients and BT to 12 (40%) patients. No treatment was administered to 4 (13.3%) patients. Table 1 shows treatment according to NS subtypes. A total of 12 patients received treatment with 22 BT. Most used BT were monoclonal anti-TNFα (n=18, 81.8%), infliximab (IFX) (n=10, 45.5%) and adalimumab (ADA) (n=5, 22.7%). After 12 months since the initiation of BT, complete, partial or no response was observed in 14 of 17 (82.4%), 2 (11.8%) and 1 patient (5.9%), respectively (Figure 1).

**Disclosure of Interests:** A. Herrero-Morant Speakers bureau: Abbvie, Amgen, Celgene, Janssen, Lilly, Roche, Sanofi, UCB, MSD, Pfizer, Roche, BMS, Janssen, and MSD, MSD, Pfizer, Roche, BMS, Janssen, and MSD, Roche

**DOI:** 10.1136/annrheumdis-2022-eular.2671

---

**Table 1. Main clinical features and treatment of 23 patients with NBD**

<table>
<thead>
<tr>
<th>NBD Type</th>
<th>n (%)</th>
<th>Mean maximum oral prednisone dose, (SD) mg/day</th>
<th>Conventional immunosuppressants, n (%)</th>
<th>Monoclonal anti-TNFα, n (%)</th>
<th>Ticlozilumab, n (%)</th>
<th>Anakinra, n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Parenchymal phenotype</td>
<td>10 (43.5)</td>
<td>51.7±19.3</td>
<td>6 (46.2)</td>
<td>4 (57.1)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>-Hemiparesis</td>
<td>5 (50)</td>
<td>52.5±75</td>
<td>2 (50)</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>-Optic neuropathy</td>
<td>3 (30)</td>
<td>52.3±26.3</td>
<td>2 (66.7)</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>-ENCEPHALOPATHY</td>
<td>1 (10)</td>
<td>45</td>
<td>1 (16.7)</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>-Optalmparessis</td>
<td>1 (10)</td>
<td>1 (100)</td>
<td>1 (100)</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Non-parenchymal phenotype</td>
<td>10 (43.5)</td>
<td>42±12.5</td>
<td>5 (38.5)</td>
<td>2 (28.6)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>-Aseptic meningitis</td>
<td>10 (43.5)</td>
<td>42±12.5</td>
<td>5 (38.5)</td>
<td>2 (28.6)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Mixed</td>
<td>3 (13)</td>
<td>45±15</td>
<td>2 (66.7)</td>
<td>1 (33.3)</td>
<td>1 (100)</td>
<td>0</td>
</tr>
<tr>
<td>-Aseptic meningitis and ophthalmparessis</td>
<td>1 (33.4)</td>
<td>60</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>-Aseptic meningitis and other cranial nerve involve</td>
<td>1 (33.4)</td>
<td>0</td>
<td>1 (50)</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>-ENCEPHALOPATHY and intracranial hypertenison</td>
<td>1 (33.4)</td>
<td>30</td>
<td>1 (50)</td>
<td>1 (100)</td>
<td>1 (100)</td>
<td>1 (100)</td>
</tr>
</tbody>
</table>

**REFERENCES:**

---

**Figure 1. Clusters of clinical associations of NBD**

Treatments were oral corticosteroids (n=16; 69.6%; mean maximum dose 42±12.5 mg/day, conventional immunosuppressants (n=13, 56.5%) and Biological Therapy (BT) (n=7; 30.4%). BT was used in patients who were refractory to conventional immunosuppressants. Monoclonal anti-TNFα were used as the first option in all patients who received BT. In 3 out of 7 (42.7%) patients BT was switched due to inefficacy. Table 1 shows the main NBD clinical subtypes and treatment.

Complete remission was achieved in 18 of 23 cases (78.2%), partial response in 2 out of 23 cases (8.7%). No severe adverse effects were observed.

**Conclusion:** NBD was observed in 24% of patients with BD. The most frequent clinical clusters of NBD were oral aphthae and ocular involvement. All patients treated with either conventional immunosuppressant or BT achieved clinical remission.