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Background: Ocular and NeuroBehçet’s Disease (NBD) are the most severe manifestations of Behçet’s disease (1-4). NBD can be classified as a) primary neural parenchymal lesions, also known as parenchymal NBD (p-NBD) or b) secondary to vascular involvement or non-parenchymal NBD (np-NBD) (4). Response to biological therapy (BT) in these two refractory subtypes of NBD is unknown.

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

Methods: Open-label multicenter study of refractory NBD from 21 different National Hospitals. NBD diagnosis was based on the International Consensus Recommendation criteria (4). Efficacy was determined by complete or partial response and no-response. Complete, partial or no response was defined according to the resolution of the neurological syndrome (signs and/or symptoms) after the BT onset.

Results: We studied 41 patients (21 women/20 men; mean age: 40.6±10.8 years). NBD was classified as p-NBD (n=33, 80.5%) and np-NBD (n=17, 41.5%). There were no significant differences in baseline general features and in neurological clinical response in both subgroups (Table 1 and Figure 1). The first BT used in p-NBD were Infliximab (IFX) (n=15), Adalimumab (ADA) (n=11), Golimumab (GLM) (n=3), Tocilizumab (TCZ) (n=2) and Etanercept (ETN) (n=2) and in np-NBD were IFX (n=9), ADA (n=8), TCZ (n=1) and ETN (n=1).

Table 1. Main features of p-NBD and np-NBD

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Total</th>
<th>p-NBD</th>
<th>np-NBD</th>
<th>p-nPBD vs np-NBD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at biological therapy initiation, years (mean±SD)</td>
<td>44±13.9</td>
<td>41±14.96</td>
<td>39±10.6</td>
<td>0.412</td>
</tr>
<tr>
<td>Gender, n (m/f) (%)</td>
<td>21/20</td>
<td>21/15</td>
<td>5/12</td>
<td>0.091</td>
</tr>
<tr>
<td>HLAB51+ patients tested, n (%)</td>
<td>15/31 (48.8/52.2)</td>
<td>15/27 (45.5/35.5)</td>
<td>0/10 (0/0)</td>
<td>0.391</td>
</tr>
<tr>
<td>Oral aphthae, n (%)</td>
<td>40 (37.6)</td>
<td>32 (97)</td>
<td>8 (22.2)</td>
<td>0.323</td>
</tr>
<tr>
<td>Cutaneous involvement, n (%)</td>
<td>28 (63.4)</td>
<td>23 (69.7)</td>
<td>5 (15.6)</td>
<td>0.003</td>
</tr>
<tr>
<td>Articular involvement, n (%)</td>
<td>21 (48.8)</td>
<td>15 (45.5)</td>
<td>6 (30.3)</td>
<td>0.161</td>
</tr>
<tr>
<td>Vascular involvement, n (%)</td>
<td>9 (22)</td>
<td>10 (30.3)</td>
<td>7 (11.1)</td>
<td>0.442</td>
</tr>
<tr>
<td>Previous conventional Immunosuppressive drugs to BT</td>
<td>5/12</td>
<td>5/12</td>
<td>0/0</td>
<td>0.001</td>
</tr>
<tr>
<td>Azathioprine</td>
<td>24 (58.5)</td>
<td>20 (60.6)</td>
<td>4 (26.7)</td>
<td>-</td>
</tr>
<tr>
<td>Methotrexate</td>
<td>16 (39.0)</td>
<td>12 (36.4)</td>
<td>4 (26.7)</td>
<td>-</td>
</tr>
<tr>
<td>Cyclophosphamide</td>
<td>13 (31.7)</td>
<td>13 (39.4)</td>
<td>0 (0)</td>
<td>-</td>
</tr>
<tr>
<td>Cyclosporine A</td>
<td>9 (22.0)</td>
<td>8 (24.2)</td>
<td>1 (6.3)</td>
<td>-</td>
</tr>
<tr>
<td>Mycophenolate Mofetil</td>
<td>2 (4.9)</td>
<td>2 (6.1)</td>
<td>0</td>
<td>-</td>
</tr>
</tbody>
</table>

Figure 1. Response to biological therapy according to NBD subtypes.

After an overall mean follow-up of 575±50.9 months BT was switched in 22 patients due to inefficacy (n=16) or Adverse Effects (AE) (n=6) and in 4 cases was definitively discontinued because of complete prolonged remission (n=3) or AE (n=1). AE were observed in 7 (17.1%) patients. Severe AE were observed in 2 cases, one due to demyelinating disease and the other due to pulmonary tuberculosis, both in patients undergoing IFX therapy. The other 6 AE were infusion reaction to IFX (n=1), IFX-induced psoriasis (n=1), IFX-induced acneiform eruption (n=1), infusion reaction to TCZ (n=1), intolerance to IFX and recurrent mild infections (n=1) and erosive lichen planus and bullous impetigo (n=1).

Conclusion: In our series, BT seems equally effective and safe in both refractory p-NBD and np-NBD.

REFERENCES:

Disclosure of Interests: Alba Herrero-Morant: None declared, José Luis Martín-Vallinas Grant/research support from: AbbVie, Pfizer, Lilly, Janssen, UCB and Celgene, Santos Cañadell Paid instructor for: Assistant professor of the Catedra UAM-ROCHE, EPID-Future, UAM, Madrid, Spain, Olga Maiza-Alonso: None declared, Julio Sanchez-Martín: None declared, Norberto Ortega: None declared, Enrique Raya: None declared, Águeda Príncipe-Español: None declared, Clara Moriano: None declared, Rafaél Meleo: None declared, Jenaro Graña: None declared, ANA URRUTICOECHA-ARANA: None declared, Angel Ramos Calvo: None declared, Marta Loredo Martínez: None declared, Eva Salgado-Pérez: None declared, Francisca Sivera: None declared, Ignacio Torre-Salaberri: None declared, J. Navarreza-Spezza: bureau: Bristol-Myers Squibb, Jose Luis Andreu Sanchez: None declared, Olga Martinez Gonzalez: None declared, Ricardo Gomez de la Torre: None declared, Sabela Fernandez: None declared, Susana Romero-Yuste: None declared, Iñigo Gonzalez-Mazon: None declared, Carmen Alvarez-Reguera: None declared, David Martinez-Lopez: None declared, J Luis Hernandez: None declared, Miguel A Gonzalez-Gay: bureau: Abbvie, Roche, Sanofi, Lilly, Celgene, Sobi, and MSD, Grant/research support from: Abbvie, MSD, Janssen, and Roche, Ricardo Blanco: bureau: Abbvie, Lilly, Pfizer, Roche, BMS, Janssen, and MSD, Grant/research support from: Abbvie, MSD, and Roche.

The Cochrane revised tool for assessing risk of bias for RCTs (RoB2), ROBINS-I for observational studies and AMSTAR II for meta-analyses were used for bias assessment. Evidence was categorized based on the GRADE system as per EULAR SOP.

Results: Based on the results of the Delphi, 11 topics related to treatment interventions were identified that were transformed into PICO questions (Table 1). Other items that received lower scores were added in the format of subquestions. Based on these research questions, string searches for the SLR were created.

Table 1. Key topics of interest for treatment strategies identified in the Delphi exercise grouped according to the PICO format.

<table>
<thead>
<tr>
<th>Patients</th>
<th>Intervention &amp; Comparators</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diagnosis</td>
<td>Cyclophosphamide</td>
<td>Disease-related outcomes</td>
</tr>
<tr>
<td>Granulomatosis with Polyangiitis</td>
<td>Rituximab</td>
<td>Treatment-related adverse events</td>
</tr>
<tr>
<td>Microscopic Polyangiitis</td>
<td>Mycophenolate</td>
<td></td>
</tr>
<tr>
<td>Eosinophilic Granulomatosis with Polyangiitis</td>
<td>Methotrexate</td>
<td></td>
</tr>
<tr>
<td>Disease severity</td>
<td>Azathioprine</td>
<td></td>
</tr>
<tr>
<td>New-onset disease</td>
<td>Glucocorticoids</td>
<td></td>
</tr>
<tr>
<td>Relapsing disease</td>
<td>Avacapan</td>
<td></td>
</tr>
<tr>
<td>Organ- or life-threatening disease</td>
<td>Mepolizumab</td>
<td></td>
</tr>
<tr>
<td>Not organ- or life-threatening disease</td>
<td>Plasma exchange</td>
<td></td>
</tr>
</tbody>
</table>

The SLR was still ongoing at the time this abstract has been written and results of the SLR will be presented at the meeting.

Conclusion: This SLR identified recent developments affecting key areas of AAV treatment, that provide systematic evidence to inform the 2022 update of the EULAR recommendations for the management of AAV, which will also be presented at this meeting.

REFERENCES:

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Disclosure of Interests: Jan Schirmer: None declared, Beatriz Sanchez-Alamo: None declared, Sara Monti: None declared, Bernhard Hellmich Speakers bureau: Abbvie, BMS, Chugai, GSK, MSD, Novartis, Pfizer, Roche, Vifor; Consultant of: Boehringer, BMS, Chugai, GSK, InfraRx, Novartis, Roche, Vifor, David Jayne Speakers bureau: Vifor, Consultant of: Astra-Zeneca, Boehringer, BMS, Chemo-centrix, Chugai, GSK, Novartis, Roche

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POS0831 IGA VASCULITIS IN ADULTS, PEDIATRICS AND NON-VASCULITIC IGA NEPHRUPATHY

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Background: IgA vasculitis (IgAV) has been extensively studied in children, while its natural history remains poorly studied in adults. Sparse data comparing childhood and adult-onset disease has shown significant differences in their clinical presentation, especially in the severity of renal involvement, which accounts for the major long-term morbidity. IgAV shares similar renal histologic features with IgA nephropathy (IgAN), while clinically IgAN is a chronic kidney disease which may lead to end stage renal disease and dialysis. The extent of kidney injury among adults with IgAV is still a matter of uncertainty.

Objectives: We aimed to evaluate clinical manifestations, laboratory data, treatment patterns and long-term outcomes of pediatric and adult-onset IgAV in comparison to IgAN.

Methods: This retrospective collaborative study examined medical records of adults and children with IgAV and IgAN adult patients admitted to rheumatology clinic and in hospital pediatric departments in a 13-year period (2007-2019). Diagnosis of adults with IgAV relied on the Ankara criteria and was confirmed by a consistent skin biopsy with positive IgA staining by immunofluorescence. Children with IgAV were included in our study on a clinical basis. All IgAN patients had a kidney biopsy proven disease. We analyzed and compared frequencies of clinical manifestations, laboratory findings, treatment regimens and long-term outcomes at one year follow-up. Finally, we assessed long term outcomes, such as time to dialysis and all-cause mortality, till the end of the follow-up time.

REFERENCES:

Disclosure of Interests: None declared

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