Background: Multisystem inflammatory syndrome in children (MIS-C) is a rare complication of SARS-CoV-2 infection in the pediatric population, caused by extensive activation of immune system. The understanding of the distorted immune response is still in the early stages.

Objectives: To analyze comprehensively immune profile in MIS-C patients including detailed serologic response to SARS-CoV-2 in comparison with control groups.

Methods: Blood samples of consecutive MIS-C patients were collected at admission. Flow cytometric analysis of all lymphocyte populations including T and B cell differentiation was performed. Immunophenotyping was performed by six-color panels for the detection of lymphocyte subpopulations. Anti-SARS-CoV-2 specific antibodies were measured in the patients serum. The IgA and IgG antibodies against S protein, the IgG S1 and S2 specific antibodies, antibodies against nucleoprotein and neutralising antibodies were measured. Patients were assessed for a wide range of auto-antibodies, namely ANA, anti-ENA (Jo-1, PL-7, PL-12, SRF, Mi-2, Ku, Prm/Sci 100, Sci-70), myositis specific antibodies (EJ, MDA-5, TH-Y, Ro52, SAE-1, SAE-2, NXP-2), anti-dsDNA, anti-phospholipid antibodies (aCl IgA, IgG, IgM, anti-2GPI IgG IgM) and ANCA. Control groups were compared to specific antibody response consisted of 14 healthy children and 19 healthy adults, who had SARS-CoV-2 infection in the last 2 months.

Results: Samples of 20 patients were included (14/20 boys, median age 12.4 years). Patients had higher percentage of double negative T cells and low numbers of of cytokinge producing T cells Th1, Th2 and Th17. Numbers of immune competent and CD21+ transitional B cells were also lowered. All patients had positive antibodies against SARS-CoV-2 including neutralising antibodies. Nine (9/19, 47 %) patients had high titer (>1:160) of neutralising antibodies. Results were compared with 2 control groups; 14 healthy children (7/14 boys; median age 8 years) and 19 healthy adults, who all experienced SARS-CoV-2 infection in the last two months. Patients with MIS-C had significantly higher levels of anti-S IgA (p=0.0001), patients with MIS-C and healthy children had significantly higher titers of anti-S1 (p=0.001) and significantly lower titers of anti-S2 (p=0.016) in comparison to adults (Figure 1). No differences were found in the titers of neutralising antibodies and anti-N antibodies. All patients were ANA negative, 19/20 patients were anti-ENA negative, whereas 1 patient had anti-Ro antibodies in low titre. Three patients had aCL IgG in medium titre and 2 patients anti-beta2GPI IgG in low titre. Patients were negative for all other autoantibodies.

Conclusion: The immune response in MIS-C patients is specific with most prominent differences in elevated percentage of double negative T cells and low numbers of Th1, Th2, Th17 and CD21+ transitional B cells. MIS-C patients have distinct serologic response with high anti-S IgA, high anti-S1 and low anti-S2 titres.

Methods:

Objectives:

Results:

Conclusions:

References:

Disclosure of Interests:

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POS0073 CAN WE PREDICT THE DEVELOPMENT OF NEPHRITIS IN PEDIATRIC IGA VASCULITIS PATIENTS?

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Background: IgA vasculitis (IgAV) is the most common systemic vasculitis of childhood, characterized by palpable purpura, arthritis, gastrointestinal and renal involvement. It is a relatively self-limited disease apart from the renal involvement which is associated with the long term morbidity.

Objectives: We aimed to define a marker at disease onset to predict the renal involvement.

Methods: In this pilot study, we analyzed a targeted panel of vascular inflammation markers (eT2, RAGE, Tie-2, sCD40L, Tie-1, sFlt-1, LIGHT, TNF-a, PIGF, IL-6, IL-1B, IL-10 and MCP-1) in the plasma samples of eight patients IgAV at the onset of the disease, before any treatment was initiated. At the time of sample collection, none of the patients had renal involvement; four of these patients subsequently developed nephritis and were defined as the IgAVN group. The levels of the markers were studied by a cytometric bead-based multiplex assay panel according to manufacturers’ instruction (LEGENDplex HU Vascular Inflammation panel 2 (13-plex); catalogue number 740966, Biolegend) and analyzed by Novocyt 3005 flow cytometer.
Results: There were no significant differences in gender, age, clinical manifestations, and laboratory findings between IgAV and IgAVN patients (Table 1). sCD40L levels were higher (median 1938.1 vs 754.9 pg/mL, p = 0.04) whereas sST2 levels were lower (median 862.8 vs 2302.8 pg/mL, p=0.02) in the patients who developed IgAV nephritis (Figure 1). sRAGE levels were higher and IL18 levels were lower in IgAVN patients but did not reach statistical significance, probably due to the low number of patients. The other parameters did not show any specific pattern.

<table>
<thead>
<tr>
<th>Parameter</th>
<th>IgAV (n=4)</th>
<th>IgAVN (n=4)</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender</td>
<td>2F, 2M</td>
<td>1F, 3M</td>
<td>0.49</td>
</tr>
<tr>
<td>Arthritis/arthralgia</td>
<td>0.0±5.4</td>
<td>10.3±3.4</td>
<td>0.68</td>
</tr>
<tr>
<td>Hb level (g/dL)</td>
<td>12.4±2.3</td>
<td>12.3±0.6</td>
<td>0.95</td>
</tr>
<tr>
<td>White blood cells, x10^9/L</td>
<td>1150±1933</td>
<td>852±1497</td>
<td>0.08</td>
</tr>
<tr>
<td>Platelets, x10^12/L</td>
<td>290±3520</td>
<td>269±3874</td>
<td>0.68</td>
</tr>
<tr>
<td>C-reactive protein (mg/dL)</td>
<td>0.63±0.03</td>
<td>1.16±0.96</td>
<td>0.39</td>
</tr>
</tbody>
</table>

Data are given as mean±SD.

Disclosure of Interests: No declared.

Background: Golimumab (GOL) is approved for treatment of polyarticular juvenile idiopathic arthritis (pJIA) in patients 2 years and older. Data on long-term safety of GOL in this indication are limited.

Objective: To assess long-term safety and efficacy of GOL in pJIA patients.

Methods: In this ongoing non-interventional observational study, clinical characteristics, disease activity and safety parameters were analysed using the German Biologics in Paediatric Rheumatology (BiKeR) registry. 81 pJIA-patients treated with GOL were body weight-matched with 162 patients receiving TNFi or MTX. No new safety signals were reported.

Results: Baseline parameters of GOL patients differed from the alternative TNFi and MTX cohorts. In patients starting with GOL treatment, disease duration was longer, corticosteroid use was less and disease activity measured by the mean number of active joints and the JADAS10, was lower (Table 1).

The long-term clinical efficacy of GOL in pJIA is highlighted by a decrease of JADAS 5.2 from baseline to 5.0 after 14 months. After 2 years, a JADAS 10 minimal disease activity was reached by 44.4% of patients, whereas 22.2% of patients were in remission and the JIA ACR 30/50/70/90 response rates were 77.8/72.6/66.7/55.6% respectively.

AE, SAE and infectious AE rates between the three cohorts were comparable (Table 1). In the GOL cohort, 4 SAE (1 uveitis, 1 arthritis flare, 1 fibromyalgia syndrome and 1 abscess) were reported, while in the all TNFi group 7 SAEs and in the MTX cohort 1 SAE were noted (Table 1). One serious infectious event (1 abscess) was documented in the GOL cohort, 2 events with GOL and MTX patients had influenza and no serious infectious events were seen in the MTX control group.

Several autoimmune processes occurred: 2 incident events in the GOL cohort (1 uveitis, 1 psoriasis), 3 cases in the all TNFi group (2 uveitis, 1 psoriasis) and 1 event in MTX-patients (celiac disease) (Table 1). Out of the 20 GOL patients with preexisting uveitis at baseline, 6 had flare events; there were no reported uveitis flares of the 17 patients in the alt. TNFi group and no patients with preexisting uveitis in the MTX-group.

No malignancies or deaths were reported.

Conclusion: Our interim results show an acceptable safety profile of GOL therapy, comparable to treatment with alt. TNFi or MTX. No new safety signals occurred. The efficacy outcome data confirm long-term benefits of GOL treatment in pJIA patients.

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