Objectives: To determine the frequency of adverse reactions to PCV13 in patients with BS who were candidates for TNF inhibitor therapy, together with ankylosing spondylitis (AS) and rheumatoid arthritis (RA) patients as controls.

Methods: All of our patients who are candidates for TNF inhibitor therapy have been offered vaccination with PCV13 since 2016. We surveyed all patients with BS, AS and RA who were vaccinated with PCV13 in our infectious diseases outpatient clinic since 2016. Patients’ charts were reviewed and additionally patients were telephoned to identify any adverse local or systemic reactions. Local reactions were defined as redness, swelling, pain, and limitation of arm movement. Systemic reactions were defined as fever, headache, chills, rash, vomiting, joint pain, and muscle pain.

Results: A total of 88 patients with BS, 143 patients with AS and 133 patients with RA had been vaccinated in our infectious diseases outpatient clinic. Among these, 55/88 (62%) patients with BS, 86/143 (60%) patients with AS and all 98/143 (68%) patients with RA could be reached. Twenty-one of 55 (38%) patients with BS, 18/86 (20%) patients with AS and 27/98 (27%) patients with RA reported at least one local and/or systemic reaction after vaccination. Patients with BS reported more systemic reactions than the other two groups (48%, 12%, 23% respectively). On the other hand local reactions were less common among patients with BS (52%, 88%, 77% respectively). The local reactions were confined to erythema at injection site, pain and difficulty in moving among patients with AS and RA while 2 patients with BS had severe papulopustular skin lesions at injection site, in addition to erythema, pain and difficulty in moving. Both of these patients were pathergy positive at the time of the diagnosis.

Conclusion: Severe papulopustular skin lesions at PCV13 injection site were observed only, but rarely, in patients with BS. Possibility of recall bias due to the retrospective nature of our study and the lack of other vaccines as controls are limitations of our study: Whether the skin lesions are caused by the skin pathergy reaction needs to be studied prospectively, as the pathergy status at diagnosis may be changed by the time these patients become candidates for TNF inhibitor therapy.

References:

Disclosure of Interests: Bena Yurttas: None declared, Sidki Safa Taflan: None declared, Nese Salhotu: None declared, Gulen Hatemi Grant/research support from: Roche, Bristol, Consultant of: Gedeon, Cristina Hidalgo: None declared, Ana Isabel Turrión: None declared, Javier del Pino Grant/research support from: Roche, Bristol, Consultant of: Gedeon, Cristina Hidalgo: None declared.

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Scleroderma, myositis and related syndromes

**AB0543**

**CLINICAL CHARACTERISTICS OF A GROUP OF CHRONIC GRAFT-VERSUS-HOST DISEASE PATIENTS WITH POSITIVE AUTOIMMUNITY.**

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**Background:** Graft-versus-host disease (GVHD) is a commonly severe multiorgan complication in patients undergoing allogeneic transplantation of hematopoietic progenitors. Its chronic form reflects a complex immune response with different degrees of inflammation, immune dysregulation and fibrosis. In some chronic graft-versus-host disease (cGVHD) patients, positive antibodies have been detected, which represent the presence of immune activity and suggest the possible involvement of B lymphocytes in the disease etiopathogenesis, but their clinical utility is controversial.

**Objectives:** To describe the clinical characteristics of a group of cGVHD patients with positive autoimmunity treated in a multidisciplinary consultation of Rheumatology-Dermatology- Hematology of GVHD.

**Methods:** Observational and retrospective study to describe the clinical characteristics of the patients with positive autoimmunity collected in the database of the multidisciplinary consultation of GVHD. The variables reviewed for this study, in addition to the demographic ones, were type of antibody, disease causing the transplant, presentation, severity and type of involvement. The statistical analysis was done with Epi-info 7.22.6.

**Results:** Only 16 (16%) of the 100 patients included in the database had positive autoimmunity. Twelve (75%) tested positive to ANA, although 5 (31.25%) in a lower titer (1/80). The most common immunofluorescence pattern was the nuclear in 88.89% (66.67% nucleolar and 22.22% nuclear + cytoplasmic). Other antibodies detected were: 6 anti-Ro/SS2, 2 anti-diSDNA, 1 anti-RP1S5, 1 anti-Fibrillarin, 1 anti-SAE1, 1 p-ANCA and 1 anti-NOR-90. The mean of age was 51.31±14.03 years. As for sex 4 (25%) were female and 12 (75%) were men. The most frequent disease that caused the transplant was acute myeloid leukemia (58.3%). Ten (62.5%) patients presented de novo cGVHD, 1 (6.25%) progressive and 5 (31.25%) quiescent. The time since receiving the transplant until the first visit was 14 to 79 months. Ten (62.5%) patients had nonspecific symptoms (arthralgia and myalgia), 2 (12.5%) edema, 8 (50%) contractures, 8 (50%) fasciitis and 6 (37.5%) eosinophilic. Eight (50%) patients had ocular involvement and 6 (37.5%) of the oral mucosa in the form of dry syndrome (Sjögren-like syndrome). Ten (62.5%) patients had limitation of joint mobility detected by the range of motion scale (ROM), of which 6 were mild and 4 moderate. Only 5 (31.25%) patients had general condition impairment. As for the skin involvement 10 (62.5%) patients had sclerodermiform involvement (8 of them being eosinophilic fasciitis-like), 2 (12.5%) lichenoid, and 3 (18.5%) mixed (sclerodermiform + lichenoid). Only 1 patient didn’t meet diagnostic criteria for GVHD. The sclerodermiform was the most common type of involvement in the positive ANA patients. Regarding the severity according to the Of the American National Institute of Health (NIH) classification: 8 (50%) had serious affection, 5 (31.25%) moderate and 2 (12.5%) mild, with 4 (25%) exitus.

**Conclusion:** In our cohort of patients with cGVHD, serum detection of autoantibodies is uncommon, being the ANA with nucleolar pattern the most frequent. Although the small sample size does not allow correlations with the clinical variables it’s worth highlighting a greater positivity of autoantibodies in the sclerodermiform skin forms.

References:

**Disclosure of Interests:** Marisa Elisa Acosta: None declared, Luis Gómez-Lechón: None declared, Olga Compañ: None declared, Sonia Pastor: None declared, Carlos A. Montilla-Morales: None declared, Olga Martínez González: None declared, Ana Isabel Turrón: None declared, Javier del Pino Grant/research support from: Roche, Bristol, Consultant of: Gedeon, Cristina Hidalgo: None declared.

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**EVALUATION OF MACULAR AND OPTIC DISC MICROVASCULAR NETWORK IN PATIENTS WITH SYSTEMIC SCLEROSIS: AN OPTICAL COHERENCE TOMOGRAPHY ANGIOGRAPHY STUDY.**

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**Background:** Systemic sclerosis (SSc) is characterized by fibrosis of the skin, internal organs and vasculopathy. Invivo, the retina provides a unique opportunity to assess the microcirculation in the eye. Previous studies have been evaluated the changes in the retinal and choroid layer and showed thinning of the choroid layer and reduced retinal microvascular density.

**Objectives:** To analysis the retinal and optic disc capillary network in patients with SSc without clinical signs of retinal involvement by using optical coherence tomography angiography.

**Methods:** In total 40 SSc patients who classified according to the ACR/EULAR criteria and 40 healthy control subjects were included in the analysis. All patients underwent a detailed ophthalmologic examination by the same ophthalmologist. After pupil dilatation, macular angiography was performed with 6x6mm area scanning using standardized system and images of the retinal capillary plexus were analyzed by Cirrus OCTA software. Mean macular thickness, retinal nerve fiber layer (RNFL) and the Ganglion cell inner plexiform...
layer (GC-IPL), vessel density (VD), perfusion density (PD), optic disc PD, retinal thickness, nasal and inferior RNFL thicknesses were significantly thinner in SSc patients (Table). Additionally GC complex thicknesses were significantly thinner in all quadrants compared to controls.

Central vessel density (CVD) and central perfusion density (CPD) values were found significantly decreased in all regions in patients with SSc. Optic disc perfusion density values were also decreased in SSc group. An inverse correlation was found between central macular thickness, FAZ area and perimeter values (rho=-0.300, p<0.007; rho=-0.276, p=0.013, respectively). There was no relationship between the disease duration and the OCTA measures.

Conclusion: Vascular and perfusion density were found decreased in patient with SSc at the results of OCTA measures. These findings may help to understand vasculopathy in the pathogenesis of the disease and OCTA may be a new method providing objective and non-invasive information about capillary network in SSc.

**Table. Demographic and ocular parameters of study population**

<table>
<thead>
<tr>
<th>Parameters</th>
<th>SSc (n = 40)</th>
<th>Control (n = 40)</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, years; mean (SD)</td>
<td>472 (8.6)</td>
<td>472 (8.1)</td>
<td>0.631</td>
</tr>
<tr>
<td>Male sex, n (%)</td>
<td>24 (60)</td>
<td>16 (40)</td>
<td>0.087</td>
</tr>
<tr>
<td>Disease duration, months; mean (SD)</td>
<td>813 (38.0)</td>
<td>N/A</td>
<td>0.003</td>
</tr>
<tr>
<td>Perfusion density, 6 mm total area; mean (SD)</td>
<td>246.3 ± 19.4</td>
<td>252.6 ± 15.3</td>
<td>0.032</td>
</tr>
<tr>
<td>Vessel density (mm⁻¹), 6 mm total area; mean (SD)</td>
<td>280.8 ± 12.4</td>
<td>285.6 ± 8.7</td>
<td>0.008</td>
</tr>
<tr>
<td>Vessel density (mm⁻¹), 6 mm total area; mean (SD)</td>
<td>176.0 ± 1.3</td>
<td>186.6 ± 0.64</td>
<td>0.006</td>
</tr>
<tr>
<td>Circularity index; mean (SD)</td>
<td>0.72 ± 0.09</td>
<td>0.73 ± 0.06</td>
<td>0.049</td>
</tr>
<tr>
<td>RNFL total (µ); mean (SD)</td>
<td>74.37 ± 12.34</td>
<td>74.09 ± 8.48</td>
<td>0.011</td>
</tr>
<tr>
<td>RNFL inferior (µ); mean (SD)</td>
<td>122.62 ± 17.87</td>
<td>127.40 ± 12.63</td>
<td>0.023</td>
</tr>
<tr>
<td>Inferior nasal GCC(µ); mean (SD)</td>
<td>85.72 ± 8.53</td>
<td>85.82 ± 4.84</td>
<td>0.066</td>
</tr>
<tr>
<td>Inferior temporal GCC(µ); mean (SD)</td>
<td>82.70 ± 8.62</td>
<td>84.95 ± 4.15</td>
<td>0.016</td>
</tr>
<tr>
<td>Superior nasal GCC(µ); mean (SD)</td>
<td>86.35 ± 7.73</td>
<td>86.8 ± 5.70</td>
<td>0.012</td>
</tr>
<tr>
<td>Superior temporal GCC(µ); mean (SD)</td>
<td>472 (8.6)</td>
<td>475 (8.1)</td>
<td>0.631</td>
</tr>
</tbody>
</table>

**Disclosure of Interests:** None declared

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**AB0545**

**GASTROINTESTINAL INVOLVEMENT IN SYSTEMIC SCLEROSIS**

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**Background:** Systemic sclerosis (SSc) is a chronic, connective tissue disease with an autoimmune pattern characterized by inflammation, fibrosis and microcirculation changes leading to internal organs malfunctions. The gastrointestinal tract (GIT) is affected in up to 90% of patients with SSc. Any part of the GIT from the mouth the anus can be affected. There are few descriptive studies about SSc-related GIT involvement.

**Objectives:** To assess immediate (Day 4) and intermediate (Day 42) efficacy and safety of protaglandins E1 (PGE1) inhibitors- Alprostadil in treatment of symptomatic Raynaud's in systemic sclerosis: single tertiary rheumatology centre experience from Sri Lanka

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**Background:** Systemic sclerosis (SSc) is a multi-system connective tissue disease1. Raynaud phenomenon (RP) is a frequent (~90%) manifestation in SSc2 & if untreated could lead to complications such as digital ulcers, gangrene and amputation.

Calcium antagonists are considered as first-line therapy for symptomatic SSc-RP. In refractory cases phosphodiesterase type 5 (PDE-5) inhibitors- Sildenafil and PGII analogue- Iloprost are used. A meta-analysis of trials indicate that PDE-5 inhibitors reduce the frequency and severity of SSc-RP attacks.

In addition a study done with PGE1 inhibitors- Alprostadil 60 micrograms infusion given for six consecutive days, had shown immediate efficacy in SSc-RP; as demonstrated by increased blood flow, digitally measured by teleangiography. Furthermore there was a reduction of the number, frequency and severity of attacks2.

Due to unavailability of Iloprost in Sri Lanka, based on the above evidence some rheumatology centres use Alprostadil 60 microgram infusion for 3 consecutive days in refractory symptomatic SSc-RP.

**Objectives:** To assess immediate (Day 4) and intermediate (Day 42) efficacy of Alprostadil 60 microgram infusions given for 3 consecutive days in treating SSc-RP.