Disclosure of Interests: None declared

Table 1. Demographic, clinical and serological characteristics

<table>
<thead>
<tr>
<th>Sex</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean age</td>
<td>57.5</td>
</tr>
<tr>
<td>Smoking%</td>
<td>80.0</td>
</tr>
<tr>
<td>Classification Criteria%</td>
<td>2002, 64/ACR/RL 49</td>
</tr>
<tr>
<td>Shimmer test</td>
<td>83</td>
</tr>
<tr>
<td>Extraluminal features%</td>
<td>74</td>
</tr>
<tr>
<td>Serologies %</td>
<td>86/48</td>
</tr>
<tr>
<td>Hyperrheumatoid factor %</td>
<td>50</td>
</tr>
<tr>
<td>Hypocomplementemia%</td>
<td>42</td>
</tr>
</tbody>
</table>
| Acute phase reactants mean | 3.7
| ESR mean | 13.8  |
| ESSPRI mean | 9.6   |
| ESSDAl mean | 2     |

Disclosure of Interests: None declared


**THURSDAY, 13 JUNE 2019**

**Vascultis**

**OP0139**

**ASSESSMENT OF ULTRASOUND DEFINITIONS AND A SCORING SYSTEM FOR SALIVARY GLAND DISEASE IN PRIMARY SJÖGREN’S SYNDROME: AN OMERACT PATIENT RELIABILITY EXERCISE**

Stephanie Finzel1, Sandrine Jousse-Joulin2, Annamaria Iagnocco3, Esperanza Naredo4, Antonietta d’Agostino5, Felicia Costantino6, Lene Terslev7, Helen Keen8, George Bruyn9.

1University Clinic of Freiburg, Department of Rheumatology and Clinical Immunology, University Medical Centre, Freiburg, Germany; 2Cavaliere Blanche Hospital and Brest Occidentale University, EA 2216, ER1 29, Rheumatology Department, Brest, France; 3Universita degli Studi di Torino, 3Academic Rheumatology Centre, Turin, Italy; 4Hospital Universitario Fundación Jiménez Díaz, Universidad Autónoma, Rheumatology Department, Joint and Bone Research Unit, Madrid, Spain; 5University Medical Centre Ljubljana, Rheumatology Department, Joint and Bone Research Unit, Ljubljana, Slovenia; 6Charité-Universitätsmedizin Berlin, Department of Rheumatology and Clinical Immunology, Berlin, Germany; 7Charles University Prague, Institute of Rheumatology and Department of Rheumatology, First Faculty of Medicine, Prague, Czech Republic; 8Hospital Ambroise Paré, Boulogne-Billancourt, France; 9Rigshospitalet, Centre for Rheumatology and Spinal Diseases, Copenhagen, Denmark; 10Perth University, Department of Rheumatology, Perth, Australia; 11MC Groep Hospitals Lelystad, Department of Rheumatology, Lelystad, Netherlands

**Background:** Salivary gland ultrasound (SGUS) may have the potential of facilitating diagnosis and therapy monitoring of salivary gland disease in patients with primary Sjögren’s syndrome (pSS). The aim of the Outcome Measures in Rheumatology Clinical Trials (OMERACT) US subgroup is to validate US as an outcome measurement instrument in pSS. Following the OMERACT US stepwise validation approach, preliminary consensus definitions for elementary lesions based on a systematic literature review were developed and subsequently a scoring system has been agreed upon and tested through a web-based Delphi exercise.

**Objectives:** To assess the reliability of consensus based SGUS scoring system in patients with SS.

**Methods:** Nine sonographers conducted an US reliability exercise of the parotid gland lesions in pSS showed a good to excellent reliability in patients with SS.

Conclusions: The developed US definitions and the scoring system for salivary gland lesions in pSS showed a good to excellent reliability in patients with SS. Next step is to apply the scoring system in clinical trials.

**Disclosure of Interests:** Stephanie Finzel: None declared, Sandrine Jousse-Joulin: None declared, Annamaria Iagnocco: None declared, Esperanza Naredo Consultant for: Abbvie, Speakers bureau: Speakers fee from: Roche, Bristol-Myers Squibb, Pfizer, UCBB, Lilly, Novartis, Janssen, and Cellgene GmbH, ALOIZIA HOCEVAR: None declared, Sarah Ohndorf: None declared, Petra Hanova: None declared, Maria-Antonietta d’Agostino: None declared, Felicia Costantino: None declared, Lene Terslev: Speakers bureau: Speakers fee from: Roche, Novartis, Pfizer, MSD, BMS, Cellgene, Helen Keen: None declared, George Bruyn: None declared

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**OP0140**

**LONG-TERM OUTCOMES OF TOCILIZUMAB FOR PATIENTS WITH GIANT CELL ARTERITIS: RESULTS FROM PART 2 OF THE GIACTA TRIAL**

John H. Stone1, Min Bao2, Jian Han3, Martin Aringer4, Daniel Blockmans5, Elisabeth Brouwer6, Maria C. Cid7, Bhaskar Dasgupta8, Jürgen Reich9, Carlo Salvatori10, Robert Spiera11, Sebastian Unizony12, Mass Gen Hosp Rheumatol Unit, Harvard Med Sch, Boston, United States of America; 2Genetech, South San Francisco, United States of America; 3Rheumatology, Med III, U Med Cte TU Dresden, Dresden, Germany; 4Dept Gen Internal Med, U Hosp Gaschulbsig, Leuven, Belgium; 5Dept Rheumatol Clin Immunol, U Groningen, U Groningen, Groningen, Germany; 6Dept Autoimmune Dis, Hosp Clin, U Barcelona, Inst d’Invest Biomol August Pi i Sunyer, Barcelona, Spain; 7Southend U Hosp, NHS Fndh Trust, Westcliff-on-Sea, United Kingdom; 8Friedrich-Alexander-Universität Erlangen-Nürnberg, Dept Internal Med 3–Rheumatologl Immunol U Erlangen, Erlangen, Germany; 9Div Rheumatol, Arcispalde Santa Maria Nuova-IRCCS, Reggio Emilia, Italy; 10Hosp Special Sugr, Cornell, United States of America

**Background:** Tocilizumab (TCZ) 162 mg administered subcutaneously weekly (QW) or every-other-week (Q2W) plus 26-wk prednisone tapering resulted in higher rates of sustained glucocorticoid (GC)–free remission in patients (pts) with giant cell arteritis (GCA) than placebo plus 26-wk (PBO+26) or 52-wk (PBO+52) prednisone tapering in the 52-wk, double-blind, randomized controlled GIACTA trial.

**Objectives:** Determine long-term safety and explore maintenance of efficacy in GCA pts in a 2-year long-term extension (part 2) of this trial.

**Methods:** At the end of the double-blind period, pts in clinical remission (CR) were instructed to stop double-blind TCZ treatment upon entering part 2. CR was defined as absence of flare per investigator assessment. GCA therapy, which could include prednisone initiation/termination of open-label TCZ and/or GC, was at the investigator’s discretion per disease status. Outcomes included maintenance of CR (no flare during part 2), flare, time to first flare, treatments received, cumulative GC dose, and safety. Treatment groups refer to originally assigned treatment (PBO or TCZ).

**Results:** Among 250 pts treated in the double-blind period, 215 entered part 2 and 197 (92%) completed 3 years in the trial. Among the 81 TCZ QW and 36 TCZ Q2W pts in CR at wk 52, 38 (47%) and 13 (36%) pts, respectively, maintained CR during part 2. Of these 51 original TCZ pts, 33 (65%) were treatment-free (no TCZ and no GC treatment), which was higher than the treatment-free proportion of original PBO pts who maintained CR in part 2 (17/38, 45%). Median time to first flare while not receiving TCZ was longer for pts in the original TCZ groups (TCZ QW, 575 days; TCZ Q2W, 428 days) than for pts in the original PBO groups (PBO+26, 162 days; PBO+52, 295 days); TCZ QW pts remained flare-free the longest (Figure 1). Retreatment with TCZ (with or without GC) for flare was effective for restoring CR in part 2. Cumulative GC dose over the 3-year study was lowest in the TCZ QW group (median dose [mg/day]: TCZ QW, 2372.8; TCZ Q2W, 2863.0; PBO +26, 5006.0; PBO+52, 5322.5). Rates of serious adverse events per 100 pt-years over 3 years (double-blind period + part 2) were comparable for pts who never received TCZ (23.2) and who did receive ≥1 dose of TCZ (25.4), and rates of
serious infections were 4.6 and 3.5 per 100 pt-years, respectively. Additional results will be presented for original PBO pts.

Conclusion: Nearly half the pts treated with TCZ QW maintained CR for the entirety of part 2, though flares still occurred in the remaining pts once they discontinued TCZ treatment. Among pts who maintained CR in part 2, higher proportions of those originally assigned to TCZ were treatment-free compared with those originally assigned to PBO. Retreatment with TCZ restored CR in pts who experienced flare. Cumulative GC doses over 3 years were lower in pts originally assigned to TCZ than in those originally assigned to PBO. No new safety signals were observed with TCZ exposure in GCA pts during the 3-year study.

REFERENCES:

Disclosure of Interests: John H. Stone Grant/research support from: F. Hoffmann-La Roche, Genentech, Xencor, Consultant for: Chugain, F. Hoffmann-La Roche, Genentech, Xencor, Min Bao Shareholder of: F. Hoffmann-La Roche Ltd., Employee of: Genentech, Jian Han Shareholder of: F. Hoffmann-La Roche Ltd., Employee of: Genentech, Martin Aringer Grant/research support from: Roche, Consultant for: AstraZeneca and Eli Lilly, Daniel Blockmans: None declared, Elisabeth Brouwer Speakers bureau: Dr. Brouwer as an employee of the UMCG received speaker fees and consulting fees from Roche which were paid to the UMCG, Maria C. Cid Grant/research support from: Kiniksa Pharmaceuticals, Consultant for: Roche, GSK, Janssen, Abbvie, Speakers bureau: Roche, Jürgen Rech Grant/research support from: Bristol-Myers Squibb and Celgene (greater than $10,000), Consultant for: Bristol-Myers Squibb, Celgene, Chuigui, GlaxoSmithKline, Janssen, Eli Lilly, Novartis, Roche, Sanofi Aventis, and UCB (in total more than $10,000), Speakers bureau: Bristol-Myers Squibb, Celgene, Chuigui, GlaxoSmithKline, Janssen, Eli Lilly, Novartis, Roche, Sanofi Aventis, and UCB (in total more than $10,000), Carlos Saivarani: None declared, Robert Spiera Grant/research support from: Roche-Genentech, GlaxoSmithKline, Bristol-Myers Squibb, Boehringer Ingelheim, Cytori, Chemo-centrix, Corbux, Consultant for: Roche-Genentech, GlaxoSmithKline, CSL Behring, Sanofi Aventis, Sebastian Unizony Grant/research support from: F. Hoffmann-La Roche, Genentech, Consultant for: Kiniksa, Sanofi, GSK.


OP0141
CD28 AS A POTENTIAL THERAPEUTIC TARGET FOR GIANT CELL ARTERITIS

Ryu Watanabe1, Hui Zhang2, Gerald Berry3, Steven Nadler3, Jörg Goronzy4, Cornelia Weyand1.

1Stanford University School of Medicine, Department of Medicine, Division of Immunology and Rheumatology, Stanford, United States of America; 2Stanford University School of Medicine, Department of Pathology, Stanford, United States of America; 3Bristol-Myers Squibb, Princeton, United States of America

Background: Giant cell arteritis (GCA) is a granulomatous vasculitis of medium and large arteries. In GCA-affected arteries, vascular wall is destroyed by tissue-infiltrating CD4 T cells and macrophages, which leads to intramural neoangiogenesis, intimal hyperplasia and luminal occlusion.

Objectives: This study aimed to examine how CD28 signaling plays a role in vasculitis induction and maintenance and which pathogenic processes are dependent on CD28-mediated T-cell activation.

Methods: We engrafted human arteries into immunodeficient NSG mice and induced vasculitis by transferring GCA immune cells. Human-NSG chimeras were treated with anti-CD28 domain antibody or control Ab. Using tissue transcriptome analysis, immunohistochemistry, flow cytometry and immunometabolic analysis, treatment effects were examined in vivo and in vitro.

Results: Treating such humanized mice with an anti-CD28 domain antibody profoundly reduced tissue-infiltrating T-cells and effectively suppressed vasculitis. Mechanistic studies revealed that CD28 regulated AKT signaling, T-cell proliferation and differentiation of IFN-γ and IL-21-producing effector T-cells. Blocking CD28 signaling disrupted T-cell metabolic fitness; particularly, glucose utilization. Expression of the glucose transporter Glut1 and of glycolytic enzymes as well as mitochondrial oxygen consumption all rely on CD28 signaling. CD28 blockade effectively suppressed vessel wall remodeling processes such as adventitial microvascular formation and intimal hyperplasia as well as induction and maintenance of CD4+CD103+ tissue-resident memory T cells.

Conclusion: CD28 stimulation provides a metabolic signal required for pathogenic effector functions in GCA, implicating CD28 signaling as a promising therapeutic target.

REFERENCES:
[1] MMP (Matrix Metalloprotease)-9 Producing Monocytes Enable T Cells to Invade the Vessel Wall and Cause Vasculitis.


[4] Inhibition of JAK-STAT Signaling Suppresses Pathogenic Immune Responses in Medium and Large Vessel Vasculitis.


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OP0142
THE IMPACT OF DISEASE EXTENT AND SEVERITY DETECTED BY QUANTITATIVE ULTRASOUND ANALYSIS IN THE DIAGNOSIS AND OUTCOME OF GIANT CELL ARTERITIS: RESULTS FROM THE TEMPORAL ARTERIOBYPY VERSUS ULTRASOUND (TABUL) STUDY AND VALIDATION IN AN INDEPENDENT COHORT

Sara Moni1,2, Cristina Ponte1, Claudio Pereira3, Federica Rumi4, Greta Carrara5, Catherine Klersy6, Andrew Hutchinsons, Wolfgang A. Schmidt8, Roberto Caporali1, Carlomaurizio Montecucco1, Raashid Luqmani4, Catherine Klersy6, Andrew Hutchinsons, Wolfgang A. Schmidt8, Roberto Caporali1, Carlomaurizio Montecucco1, Raashid Luqmani4.

1University of Pavia, IRCCS Policlinico S. Matteo, Department of rheumatology, Pavia, Italy; 2University of Pavia, PhD in Experimental Medicine, Pavia, Italy; 3Hospital de Santa Maria, Instituto de Histologia y Rheumatology Research Unit, Instituto de Medicina Molecular, Faculdade de Medicina da Universidade de Lisboa, Lisbon Academic Medical Centre, Department of Rheumatology, Lisbon, Portugal; 4University of Oxford, NDORMS, Rheumatology Department, Nuffield Orthopaedic Centre, Oxford, United Kingdom; 5Italian Society of Rheumatology, Epidemiology Unit, Milan, Italy; 6IRCCS Policlinico S. Matteo Foundation, University of Pavia, Biometry and Clinical Epidemiology, Pavia, Italy; 7London School of Hygiene and Tropical Medicine, Department of Health Services Research and Policy, London, United Kingdom; 8Medical Centre for Rheumatology Berlin-Buch, Immanuel Krankenhaus Berlin, Berlin, Germany; 9Southend University Hospital, NHS Foundation Trust, Westcliff-on-Sea, United Kingdom

Background: Colour duplex sonography (CDS) is used in patients with giant cell arteritis (GCA) to detect inflammatory oedema of the vascular wall, known as “halo”. A quantitative analysis of halo characteristics to grade the severity and extension of vascular involvement detected by CDS could improve GCA assessment.

Objectives: To develop a quantitative CDS score to improve the diagnosis of GCA, and to correlate the score with histologic findings and clinical outcome. To determine the additional role of clinical signs/symptoms to the CDS score.

Methods: We selected patients with a positive CDS and a diagnosis of GCA recruited into the Temporal Artery (TA) Biopsy (TAB) vs Ultrasound in Diagnosis of GCA (TABUL) study. Due to collinearity we fitted 4 different CDS models including combinations of the following items: number of sites and distribution of halos, average and maximum intima-media thickness (IMT) of the level of the TA and auxillary arteries (AX) and halos bilaterality. We fitted 4 models with clinical and laboratory findings. We combined the best CDS and clinical models (according to the Akaiki Information Criterion) to identify independent correlates of a TAB diagnostic for GCA and of clinical outcome at 6 months (visual loss + VDI ocular + glucocorticoids > 10 mg/day and/or need for immunosuppressants) and performed a 10-fold cross-validation of the model. We validated the clinical outcome model on an independent cohort referred to the fast-track ultrasound clinics of two European rheumatology centres.

Results: We included 135 patients with GCA from TABUL (female: 68%, age 73 ± 8) and 72 patients from an independent cohort (female: 46%, age 75 ± 7). The