extended oligoarticular (oJIA) and persistent polyarticular (pJIA). Furthermore, IFNγ producing CD4+ T cells were decreased in fibromyalgia secondary to Sjögren’s with respect to fibromyalgia patients and HC. Counts of TNFα producing CD4+ T cells were increased in fibromyalgia with respect fibromyalgia secondary to Sjögren. IL-10 producing CD4+ T cells were normal in fibromyalgia but decreased in fibromyalgia secondary to Sjögren.

**Conclusion:** Fibromyalgia patients show an abnormal circulating activation stages of CD4+ T cells, as well as, express unusual elevated counts of CD4+ T cells producing IL-17A, IL-4 and IFNγ. These alterations could differentiate two pathologic and inflammatory behaviors of the T cell compartment between fibromyalgia and fibromyalgia secondary to Sjögren patients. References:


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**AB0036**

**CHILDREN WITH EXTENDED OLGARTICULAR AND POLYARTICULAR JUVENILE IDIOPATHIC ARTHRITIS HAVE A CYTOKINE PATTERN FAVOURING B CELL ACTIVATION IN CIRCULATION**

R. A. Moura1, F. Oliveira-Frannes1, C. Marques1, A. Brito1, R. L. Teixeira1,2, V.C. Romão1,2, R. Campanhão-Marques1,2, V. Teixeira1, M. J. Saavedra1, C. Ponte1,2, N. Khmelinskii1,2, J. Fonseca1,2,1, Instituto de Medicina Molecular João Lobo Antunes, Faculdade de Medicina, Universidade de Lisboa, Lisbon, Portugal; 2Rheumatology Department, Hospital de Santa Maria, Lisbon Academic Medical Centre, Lisbon, Portugal

**Background:** Juvenile idiopathic arthritis (JIA) is the most common rheumatic disease in children. The majority of polyarticular JIA (pJIA) and a large fraction of extended oligoarticular JIA (oJIA) patients fulfill classification criteria for rheumatoid arthritis (RA) in adulthood. B-cells play several important roles in RA pathogenesis, but it is still unclear if the pattern of B-cell involvement in pJIA and extended oJIA follows what has been described for adults with RA.

**Objectives:** The main goal of this study was to determine the concentration of cytokines potentially relevant for B-cell activation in serum from children with pJIA and extended oJIA when compared to children with persistent oJIA, adult JIA, early and established RA.

**Methods:** Serum samples were collected from children with extended oJIA (n=8), persistent pJIA (n=6), pJIA (n=6), adult JIA (n=8), untreated early RA (<1 year of disease) (n=12), established RA patients treated with synthetic disease-modifying anti-arthritic drugs (DMARDs) (n=10) and two groups of age- and sex-matched healthy donors (children, n=6 and adults, n=10). A proliferation-inducing ligand (APRIL), B-cell activating factor (BAFF), interleukin (IL)-6 and IL-21 serum levels were measured by enzyme-linked immunosorbent assay (ELISA).

**Results:** Children with extended oJIA, early and established RA patients had significantly higher BAFF serum levels when compared to controls, but no significant differences were observed in children with persistent pJIA, pJIA and adult JIA when compared to all groups included. APRIL serum levels were significantly increased in early and established RA patients when compared to both controls and children with persistent pJIA. No significant differences were found in APRIL concentrations between children with JIA, adult JIA and controls. IL-6 serum levels were significantly increased in children with extended oJIA, pJIA, early and established RA when compared to controls, but no significant differences were found in children with persistent oJIA and adult JIA patients. IL-21 serum levels were significantly increased in early RA when compared to controls, but no significant differences were observed between any of the other groups included.

**Conclusion:** The similarity in B-cell cytokine pattern found between extended oJIA, pJIA, early and established RA patients, contrary to what was observed in persistent pJIA, suggests an early B-cell involvement in the pathogenesis of extended oJIA and pJIA as described for RA.

**Disclosure of Interests:** None declared

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**AB0037**

**EXPRESSION OF NEGATIVE CHECKPOINT MOLECULES BTLA AND HVEM IS DYSREGULATED IN AUTOIMMUNE DISEASES**

S. Nagpal1, S. Cole1, A. Floudas2, M. Wechalekar3, Q. Song1, T. Gordon4, R. Caricchio5, D. Veale6, U. Fearon7, N. Rao8, L. Y. Hao1, 1Janssen Research & Development, Spring House, United States of America; 2Trinity College Dublin, Dublin, Ireland; 3Finders University, Adelaide, Australia; 4Temple University, Philadelphia, United States of America; 5University College Dublin, Dublin, Ireland

**Background:** Immune checkpoint blockade with agents targeting CTLA4 and PD-1/PDL-1 alone or in combination has demonstrated exceptional efficacy in multiple cancer types by "unleashing" the cytotoxic action of quiescent, tumor-infiltrating T cells. However, the therapeutic action of these immunotherapies goes hand in hand with the loss of immune tolerance and appearance of immune-related adverse events such as colitis, arthralgia and inflammatory arthritis in responsive patients. Therefore, immune checkpoint molecules have been proposed as targets for the treatment of autoimmune diseases.

**Objectives:** Herein, we interrogate the potential of BTLA/HVEM axis as a target for restoring immune homeostasis in rheumatoid arthritis (RA), Systemic Lupus Erythematosus (SLE) and Sjögren’s Syndrome (SjS) by examining their expression patterns in autoimmune disease tissues. **Methods:** Message and protein expression of BTLA and HVEM were examined in RA and SLE synovial tissues, SLE cutaneous lesions, SjS salivary glands and peripheral blood samples of autoimmune disease by RNA sequencing and flow cytometry.

**Results:** Tissue dysregulation of the BTLA-HVEM axis was observed: Increased BTLA RNA level in RA synovium, SLE-affect ed skin, and SjS salivary gland samples, whereas HVEM level was affected only in the RA synovium compared to unaffected tissues. Detailed immunophenotyping of B, T, and myeloid cell populations in RA, SLE, SjS and healthy control PBMCs revealed differential modulation of the BTLA+ or HVEM+ immune cell subsets in a disease-context dependent manner. SjS patients showed an overall decrease in memory B cells and most of the BTLA+ B cell subsets while a decrease in HVEM+ B cells was observed only in SLE PBMC samples and not RA and SLE samples. Immuno-phenotyping with a T cell panel exhibited decreased BTLA and HVEM expression on T cell subsets in SjS and SLE but not in RA patients. In addition, protein levels of HVEM were differentially decreased in SLE myeloid cell subsets. Finally, we demonstrate tissue-specific surface expression patterns of BTLA in RA and SLE samples: higher surface BTLA levels on RA and SLE PBMC B cells than matched tissue-derived B cells.

**Conclusion:** Our results demonstrate a dysregulation of the BTLA/HVEM axis in either lesional tissue or peripheral blood in an autoimmune disease context-dependent manner. These results also indicate the potential of targeting BTLA- HVEM axis for the treatment of multiple autoimmune diseases.


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**AB0038**

**IMMUNE PHENOTYPING OF ERDHEIM-CHESTER DISEASE THROUGH MASS CYTOMETRY**

M. Papp1, A. Corneau1, F. Cohen1, B. Robin2, J. F. Emile1, M. Miyara2, C. Blanc1, Z. Amoura1, O. Hermine1, J. Haroche1, T. Troval Macle1, 1Sorbonne Université, Assistance Publique-Hôpitaux de Paris, Hôpital Pitéï-Salpêtrière, Service de Médecine Interne 2, Institut E3M, Centre National de Référence des Histio cytoses, Paris, France; 2Imagine Institute, Laboratory of Molecular Mechanisms of Hematologic Disorders and Therapeutic Implications, Imagine Institute, INSERM UMR1163, Paris, France; 3Sorbonne Université, INSERM U9307 PASS, Plateforme de Cytométrie (CyPS), Paris, France; 4Université de Versailles, AP-HP EA4340-BECCH, Départment de Pathologie, Boulanger-Billancourt, France; 5Sorbonne Université, Assistance Publique-Hôpitaux de Paris, Centre d’Immunologie et des Maladies Infectieuses (CIMI-Paris),